Scott-Brown's Otolaryngology

Fifth edition

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Historical introduction

About 36 years ago Bill Scott-Brown suffered a major coronary infarct and being strictly ordered to 'rest' for six months set himself to create, as Editor (not author, because that would have been too strenuous, he thought) this work of his own inspiration. In 1952 I was among the first generation of FRCS candidates for whom it was the Bible. We all revered 'Negus' for the nose and throat (some of us still do) but Scott-Brown, in two volumes as it then was, provided the first post-war text for otolaryngology across the board. SB (as he was known) was probably the only person to be at all surprised by the success of his achievement, and to find himself in due course under notice from Butterworths to prepare a second edition. It was at this stage that he recruited John Ballantyne and myself and the second, third and fourth editions were produced by the two of us under his friendly eye. For the third edition we succumbed to the inevitable by expanding two fat volumes into four (slightly) thinner ones, only to find that the fourth edition in its turn required four fat ones.

Throughout this 20 year period John Ballantyne and I derived constant satisfaction and pleasure from the ongoing association with so many willing friends and contributors past and present. We than them warmly.

We know that the ENT fraternity world-wide has pleasure in the knowledge that SB continued in his retirement still to take satisfaction from the perpetuation of his work. The sad news of his death came just as this new edition went to press. Those who knew him will perhaps see in this Fifth Edition, and the 35th year of his book, a memorial to his achievement.

John Groves
Advisory Editor
Introduction

When I was first invited to edit the Fifth edition of Scott-Brown's Otolaryngology, I thought I was aware of the enormity of the task and my own limitations. As time progressed, I realized that I had misjudged both.

This work has represented the mainstream of British otolaryngological thinking for over thirty years. However, the increase in the breadth and depth of our specialty is such that only a gifted few can be conversant with all aspects of it. Hence, I realized that I could not undertake the task without help. I have been most fortunate in having such a distinguished group of volume editors, all of whom are already well-known in British otolaryngology, and all of whom have been delightful and stimulating colleagues in this work. It has been a joy to work with them.

Modern otolaryngology has widened in recent decades, and procedures are now being performed that are no longer covered by the term 'ear, nose and throat surgery'. This work attempts to embrace all the areas that so-called ear, nose and throat surgeons are covering at the present time, and hence the change of the title to Scott-Brown's Otolaryngology.

For the new edition Scott-Brown has grown from four to six volumes. An entirely new volume has been introduced in recognition of the subspecialty of paediatric otolaryngology and the amount of material in audiological medicine is now great enough to justify its separation from the Ear volume. Although these are now specialties in their own rights, they are also, and will continue to be beyond the lifetime of this edition, part of the routine practice of most British otolaryngologist. To enable these new volumes to stand alone, a certain amount of overlap with other volumes has been necessary.

In any multi-author and multi-volume production, overlap is always necessary if each chapter is to be developed freely, and if there is to be easy reference to subjects dealt with in more than one volume. Consequently, I ask for the reader's indulgence in those sections where overlap has been planned and deliberate. Where it has occurred as a result of my ineptitude, I apologize.

The editorial team have been very pleased at the response of those invited to contribute, although, unfortunately, a few leading members of our specialty were unable to accept the invitation. However, by and large, those asked were both cooperative and energetic in their responses, and have given freely of themselves in their contributions. I have been most impressed by the spirit of goodwill among the otolaryngologists in this country, and I am grateful to them.

In the production of this edition, I have seen myself as custodian of a great British institution. I have always been aware of the privilege and responsibility of my position, and am grateful for the advice I have received from many senior and not so senior members of our specialty. I am particularly indebted to the Advisory Editor, John Groves, and to his formed editorial colleague, John Ballantyne. My respect and admiration for these colleagues has risen, not simply because of the invaluable help they have given so freely in this edition, but because I now realise the enormity of their accomplishment and their contribution to British otolaryngology in editing the last three editions.
I also wish to express my thanks to those in Belfast who have helped with, or suffered because of, the Fifth Edition. Some have done both, and without their backing and encouragement this work would not have been possible. It would be invidious to try to name everyone. Various secretaries have been of enormous help, and without this I could not have produced this edition. My consultant colleagues have advised and encouraged me, and my junior colleagues have given very practical advice in their down-to-earth comments and invaluable help with proof-reading. My family have been both encouraging and remarkably tolerant of the long hours required to edit such a work as this.

The staff at Butterworths were helpful and encouraging throughout. Initially, Peter Richardson set the wheels in motion. He was followed as publisher by Charles Fry, who was assisted by Anne Smith and Jane Bryant. The sub-editors have been Anne Powell and Jane Sugarman. The general spirit of pleasant cooperation and tolerance has been delightful.

I am sufficiently optimistic to believe that there will be a Sixth Edition. I do not know who will be editing it. However, if the reader has any constructive comments or criticisms, I should be pleased to have them ... in writing! I can not guarantee to acknowledge these, but I promise that, if I am the editor, I shall give them due consideration, and, if not, I shall make them available to my successor.

Alan G. Kerr
Chapter 1: Methods of examination of the pharynx and larynx

M. S. McCormick

History

Most diseases of the upper aerodigestive tract have an uncomplicated history. However, the duration of symptoms may give some indication as to the origin, or aggressiveness of the disease process. Leading questions relating to dysfunction are asked.

Mouth disorders may cause:
- pain, swelling or ulceration
- loose or ill-fitting dentures
- bleeding, numbness or weakness of the tongue and lips
- difficulty with speech or swallowing if tongue movement is faulty or tethered.

Pharyngeal lesions may present with:
- dysphagia (difficulty in swallowing) which may be
  - acute, progressive or recurrent
  - for solid only
  - for solids and liquids
- odynophagia, that is pain on swallowing
- weight loss
- voice alteration or hoarseness when the larynx is invaded
- referred otalgia
- a dry swallow is often noted in globus pharyngeus
- hypernasal speech and regurgitation of food into the nose results from velopharyngeal incompetence.

Laryngeal disorders may give rise to:
- altered voice production, that is abnormalities of strength, pitch, tone and quality
- hoarseness (a rough voice) - the commonest disorder
- stridor or noisy breathing
- dysphagia if the extrinsic larynx is invaded.

A lump in the neck is a common symptom with many causes.

The patient's response to questions will also give the otolaryngologist an indication of the disability caused by the disease. Knowledge of the patient's general condition and attitude to his symptoms must also be obtained at this point. Most important is the patient's cardiorespiratory reserve. Direct questions about exercise tolerance, angina, cough and sputum production will usually reveal whether the patient's management will be determined by his general health or whether the disease can be treated on its own merits. Note should also be taken of diseases which might complicate management, for example diabetes, specific allergies, or any regular medication. It should be remembered that the patient will also be assessing his attending surgeon so that these early exchanges may dictate the future conduct of the examination and management of the problem.
Armed with this information regarding general and specific ill health the otolaryngologist may now proceed to examine the patient's upper respiratory tract.

**Examination**

**Lighting**

The fundamental prerequisite for this examination is adequate lighting. The standard concave head mirror is 9 cm in diameter with a central aperture of 2 cm and a focal length of 18 cm. The light source for this mirror is normally positioned about 30 cm behind and lateral to the patient's ear, the side being determined by the surgeon's preference. The normal arrangement is shown. This system provides brilliant illumination of the area under study and also leaves both hands free for use in the examination.

Alternatives to the reflective head mirror include various types of head lights which obtain their light from either low voltage DC bulbs or even fibreoptic systems. Preference for each particular lighting system is personal, but it is customary to use one of the latter type of head lights in the operating theatre, as much greater independence of movement of the surgeon's head is necessary.

**General**

It is customary to examine the patient in the sitting position unless this is impossible for some reason. The patient should be comfortable in the examination seat, bending slightly forward with the hands resting on his knees. With the use of a good light, a relaxed cooperative patient and, if necessary, a darkened room, the examiner is now able to proceed to gain maximum information from the examination.

The various aspects of examination taught by surgical tutors such as the site, size, shape, texture as well as other physical characteristics appropriate to each specific pathology should be remembered. Cysts should be transilluminated. The scientific method also encourages accurate recording of the observations so that the effect of treatment or any change in physical characteristics can be assessed, for example size of surface ulceration, 'mobility' of neck masses, etc.

It is important to obtain adequate exposure of the area to be examined. It is probably best to have the patient remove enough clothing to expose the neck and shoulder tips. This allows the examiner not only to observe but also to palpate any area under suspicion. Induration associated with ulceration may be assessed, as can fixity of tumours to bone. The obscure origins of some neck swellings can be confirmed or refuted using bimanual palpation with a gloved finger in the mouth. Biopsy of suspicious mouth ulcers can be performed under direct vision using local anaesthesia if necessary. When taking a biopsy it is important not to crush or distort the specimen. The biopsy, whether obtained by scalpel and dissecting forceps or punch biopsy forceps should be placed directly, without over manipulation, into the specimen pot which usually contains formalin or other fixative according to the wishes of the histopathologist, some preferring an unfixed fresh specimen.
Examination of the mouth and oropharynx

Lips

The lips should be observed for pallor or angular stomatitis as these may indicate anaemia. The nature of ulceration of the lip is easily diagnosed by the history, site and its physical characteristics, for example an acutely painful aphthous ulcer, the recurring nature of herpes labialis, the persistent squamous carcinoma with raised rolled edges, etc.

Buccal cavity, teeth and tongue

A relaxed cooperative patient will usually permit examination of all parts of the oral cavity including the buccal mucosa, teeth and tongue. The examiner may wish to restrain the patient's head with his left hand while retracting the lip or cheek with a spatula in the right hand. Alternatively the nurse may support the patient's head from behind, thus freeing the examiner's other hand. Both sides of the mouth may be inspected by using wooden or metal spatulas, or special lip retractors. An orderly and thorough examination of the mouth is essential as so many disease processes affecting the mucosa may be systemic, for example Addison's disease; or multifocal, for example carcinoma in situ. The various fossae within the mouth should be inspected in a regular and systematic fashion. The author's preference is to examine the buccal surface of the lower lip followed by the lower buccogingival sulcus as far back as the last lower molar tooth on that side. The cheek is then retracted superolaterally to allow inspection of the upper buccogingival sulcus from posterior to anterior along the gingival surface of the upper lip and then the examination proceeds to the other side of the mouth. The opening of the parotid duct can be seen opposite the second upper molar tooth. Any thickening of the duct or abnormal secretions should be noted. The various forms of disease affecting the mucosa inside the mouth including stomatitis are dealt with in Chapter 4.

The examiner now turns his attention to the teeth and the surrounding structures of the upper and lower jaw. Loose teeth, unhealed sockets and ill-fitting dentures are common symptoms of an expansive lesion within the jaw. The patient can usually point accurately to any specific point of complaint in this regard.

Malocclusion of teeth of the upper and lower jaw should be noted. This is important as strain may be placed on the temporomandibular joints and cause referred otalgia. Poor dental hygiene may result in carious teeth in the younger patient or more commonly gingivitis in adults.

Next the patient is asked to open the mouth widely so that the dorsum of the tongue is seen. The shape and symmetry of the tongue should be noted as well as fasciculation, seen in motor neuron disease. The patient is asked to move the tongue voluntarily in all directions. Paralysis of a hypoglossal nerve may be easily overlooked in the early stages but becomes more obvious after disuse atrophy of the affected tongue muscles. The tongue may also be tethered by malignant infiltration.

The patient is then asked to raise the tip of the tongue to expose its ventral surface. This step leads naturally to examination of the anterior part of the floor of the mouth. On
either side of the frenulum may be seen the opening of the submandibular salivary gland duct which may be affected in stone formation within the salivary gland or in malignant disease of the floor of the mouth. The examiner now retracts the tongue gently to one side and applies countertraction to the cheek preferably using two spatulas. An adequate view of the glossogingival sulcus can not be obtained as far back as the area between the last molar and the lateral border at the base of the tongue. This area is known by some as 'coffin corner' as an early cancer in this site is easily missed unless a thorough examination is undertaken. The same might be mentioned for the area between the last upper and lower molar teeth, the retro- or interdental trigone. Palpation of this area is particularly easy and should not be omitted. This examination is repeated on the other side of the mouth. Finally the hard palate is inspected. Mouth breathing in children may be the result of nasal or nasopharyngeal obstruction and is often associated with a high arched palate.

**Oropharynx**

A tongue spatula is placed in the midline of the dorsum of the tongue and gentle pressure is applied so that the tonsillar pillars, tonsils, soft palate and uvula can be seen. Care is necessary at this point not to stimulate the posterior one-third of the tongue as this will usually induce a gag response even in those tolerant to examination. After observing the mucosa the otolaryngologist's main concern in this area is usually the oropharyngeal tonsil. There is tremendous variation not only in the appearance of tonsils, but also in the interpretation of this appearance by otolaryngologists. Most accept that the diagnosis of recurring acute tonsillitis is made on history alone, and therefore the appearance of the tonsil on the day of examination is probably of very limited value; for example size can vary with the degree of inflammation present and may be apparently altered by protrusion of the tongue.

**Soft palate**

The soft palate is a muscular curtain covered on both sides with mucosa. It can be functionally assessed by asking the patient to phonate or to sniff. Asymmetry of movement is seen in glossopharyngeal paralysis. Its posterior surface is examined by a nasopharyngeal mirror.

The posterior pharyngeal wall is also easily visible. Prominent lymphoid follicles are seen in pharyngitis. Mucopurulent discharge descending from the nasal cavities and nasopharynx may be seen in rhinosinusitis. It is now necessary to use reflective light mirrors to assess the nasopharynx and the remains of the oropharynx, hypopharynx and larynx.

**Nasopharynx, hypopharynx and larynx**

To view the nasopharynx it is necessary to see behind the soft palate. The mirror must be small enough to pass below and behind the soft palate, but large enough to reflect sufficient light to examine the area and to be able to orientate the image seen. To facilitate this examination the handle is offset and the mirror is angled on the handle. It is important to warm the glass surface of the mirror or condensation will take place and obscure vision. This is commonly undertaken with the flame from a spirit lamp, but other thermostatically controlled devices are available. It is reassuring for the patient to know that the mirror will
not burn him so it is usual for the examiner to place the back of the mirror on his own cheek or hand prior to proceeding with the examination.

With the patient sitting forward and relaxed, the tongue is depressed with a Lack's tongue depressor and the mirror passed behind the soft palate. By reflecting light from the head mirror on to the angled nasopharyngeal mirror a view can be obtained of the roof of the nasopharynx, the posterior choanae, the posterior aspect of the nasal septum and the posterior ends of the inferior turbinates. A view of the eustachian cushions can be seen by rotating the handle of the mirror. The posterior surface of the maxillary tuberosity can also be examined using this method. While introducing the mirror into the mouth the movement of the soft palate should be observed for asymmetry or whether it is unduly hyposensitive. These may indicate a glossopharyngeal dysfunction. Some patients automatically contract their soft palate against the posterior pharyngeal wall occluding any view. This can be overcome by asking the patient to breathe through the nose while keeping his mouth open. Topical local anaesthesia may be necessary for those with a hypersensitive gag reflex. This can be achieved by using either lignocaine spray or asking the patient to suck a benzocaine lozenge. This examination is one of the most difficult skills to acquire but, with practice, adequate information can be obtained for diagnosis. When in doubt, flexible nasendoscopy under local anaesthetic or examination under general anaesthesia may be necessary.

The posterior pharyngeal wall can be seen on depressing the tongue. However, it is necessary to use a mirror to examine fully the remainder of the oropharynx, hypopharynx and larynx including the base of the tongue, the lateral pharyngeal walls, the pyriform fossae and the aditus to the larynx. The size of the patient's mouth and soft palate should be examined so that the largest mirror that can be comfortably introduced in the mouth is used. This facilitates the use of the most light as well as giving a better overall view. In patients with a very sensitive gag reflex it may be necessary to use local anaesthetic as described previously.

The patient should be asked to protrude his tongue, and this is then held with a gauze swab either by the thumb and index finger or thumb and middle finger depending on the ability of the patient to protrude the tongue and also on the examiner's preference. It is advisable to be firm but not rough, quick without rushing, and thorough in the order of the clinical inspection permitted with the introduction of the mirror. Care should be taken not to catch the frenulum of the tongue over the cutting edge of the lower teeth. The patient's upper lip can be gently elevated with the free index or middle finger, and if necessary the patient may be asked to smile. This has the effect of pulling the oral commissure upwards and backwards and increases the area of vision.

A previously warmed laryngeal mirror is introduced into the mouth after testing its temperature as described previously. It is best held like a pen, and care is taken not to soil the surface of the mirror with saliva, etc. The mirror is then gently placed against the soft palate and uvula and using gentle steady pressure these structures are elevated bringing into view in the mirror the base of the tongue and epiglottis, and behind this the larynx and hypopharynx. It is now usually possible to view the anterior surface of the epiglottis, the vallecula divided by the glossoepiglottic fold and also the irregular surface of the base of the tongue. By tilting and raising the mirror the laryngeal aditus is seen comprising the epiglottis, the aryepiglottic folds and the arytenoids posteriorly.
It is common in children, but less so in adults, that the epiglottis may overhang the larynx obscuring the view. The white vocal cords stand out in contrast to the surrounding pink mucosa. They are seen to connect the arytenoids with the base of the epiglottis at the anterior commissure. The bulging false cords or ventricular bands appear to meet the true cords and obscure the ventricles. The cords are observed at rest, during quiet respiration and phonation. In the fully relaxed patient it is quite possible to see between the cords and down the trachea where two or three rings of the tracheal cartilages may be seen anteriorly.

Further rotation of the mirror shows the interarytenoid area and the posterior pharyngeal wall. Inferiorly lies the postcricoid region which can only be seen when the patient swallows and hence is not visible on indirect laryngoscopy. On each side of this lies the pyriform fossa. It is possible to see both medial and lateral walls and occasionally the apex of the fossa. ‘Pooling’ of secretions may be a sign of a tumour in the postcricoid space or cervical oesophagus. It is important therefore to ask the patient to swallow any retained secretions and then to re-examine.

Despite reassurance, local anaesthesia and deftness of movement of the examiner's hands it is sometimes impossible to obtain adequate information from indirect laryngoscopy and further information must be obtained either by flexible nasendoscopy or by direct examination of the larynx under general anaesthesia. Methods of grasping the epiglottis with Mackenzie's laryngeal forceps have been superseded to some extent by the fibreoptic instrument, but may well still find use when this equipment is not available.

It is sometimes necessary to perform a direct examination of the larynx under local anaesthesia, for example, the injection of Teflon into a paralysed vocal cord (see below), or to obtain a biopsy from a patient with a bulky friable tumour of the supraglottis. It is well known that a tracheostomy prior to definitive treatment is a bad prognostic factor in laryngeal cancer patients. Endoscopy under general anaesthesia could possibly precipitate complete obstruction of the airway and necessitate an emergency tracheostomy. The technique of local anaesthesia is relatively simple but with the more general use and increased safety of general anaesthesia, this technique described below is not used by many today.

After spraying the oropharynx with lignocaine (10%), pledgets of cotton wool soaked in cocaine (4%) are introduced into the pyriform fossae using Mackenzie's forceps, and held in place for 3-4 minutes on each side. The superior laryngeal nerve is anaesthetized here as it passes through the thyrohyoid membrane. A spray of cocaine is then applied directly to the vocal cords using an angled spray while observed with a laryngeal mirror. With a cooperative patient it is possible to perform direct examination of the larynx, hypopharynx, the postcricoid space and cervical oesophagus. It may also be necessary to apply some more anaesthesia directly to the vocal cords once the laryngoscope is in position.

The neck and salivary glands

While examination of the interior of the mouth, oropharynx and hypopharynx can give information as to the presence of disease, it is also of the utmost importance to examine thoroughly the external larynx and the areas of lymphatic drainage. The examiner should stand behind the patient who should remain comfortably seated with the head slightly flexed. Enough clothing should be removed so that the supraclavicular fossae and tips of the
shoulders can be seen. It is best to examine the neck in triangles and while each surgeon has his own preferences it is important to do this methodically. The examination should commence with the posterior triangle superiorly by defining the mastoid tip and then feeling for lymph nodes along the anterior border of the trapezius muscle. It is also possible to palpate under the muscle by gently pressing the fingers under the muscles so that the flesh between the thumb and fingers can be palpated. The examining fingers will eventually reach the clavicle. At this point the floor of the posterior triangle can be examined by rolling the tissues between the fingertips and the muscular floor of the triangle, gradually moving medially until the sternomastoid muscle is reached. It is now possible to feel the lymph nodes associated with the internal jugular vein by firmly pressing the fingers underneath the muscle. It is also possible to palpate the muscle mass itself using the thumb and fingers as described for the trapezius. The fingers eventually arrive once again at the mastoid tip. The medial side of the sternomastoid muscle as far as the suprasternal notch should then be examined again palpating for any pathological lymph nodes. The clavicle and the suprasternal notch are palpated, and at this point the trachea can be felt in the midline.

The external features of the larynx should also be assessed. The most prominent of the cartilages is the cricoid, and it may be just possible to palpate a normal thyroid isthmus overlying the second and third tracheal rings. The cricothyroid membrane, the alae of the thyroid cartilage, the thyrohyoid membrane and the hyoid itself should be palpated. Deep in the groove between the sternomastoid and the larynx lie the great vessels of the neck and associated with the internal jugular vein lies the deep cervical plexus of lymph nodes. Mobility of the larynx on the prevertebral fascia can be tested by grasping the thyroid alae between the thumb and fingers and then moving the whole structure from side to side. The gritty sensation so produced by this manoeuvre is termed ‘laryngeal crepitus’ and is absent in lesions which push the larynx forward, away from the prevertebral fascia, for example postcricoid carcinoma or retropharyngeal abscess. The examination is continued with the submental triangle bounded by the ramus of the mandible and the line that would represent the line of the omohyoid muscle to the point of the chin. In this triangle lies the submandibular salivary gland and as the fingers come gently forward, the facial artery crossing the mandible and the associated pre- and postfacial lymph nodes can be felt. The examiner stands behind the patient, cupping the fingers under the mandibular ramus and palpating the floor of the mouth for other lymph nodes or direct extension of intraoral tumours. Palpation is carried forward to the point of the chin and then finally the tissues of the anterior triangle are rolled against the muscles of the floor of the mouth. If swelling of the submandibular gland is felt, it is mandatory to examine the gland bimanually with a gloved finger in the mouth while externally supporting the gland with the fingers.

To examine the parotid gland adequately assessment should be made not only of the gland itself, but also of facial nerve function, of the neck for metastases and of the parotid duct. A swelling in the parapharyngeal space or soft palate may be of parotid origin. Both parotid glands should be palpated including the finger-like processes that extend up in front of the ear and posteriorly over the mastoid tip. Any swelling deep to the skin at this point must be considered to be parotid until proven otherwise. To examine facial nerve function completely it must be recalled that the facial nerve not only innervates the muscles of facial expression but also secretomotor fibres to the lacrimal and the submandibular glands, special sensory fibres to subserve taste to the anterior two-thirds of the tongue and also the motor fibres to the stapedius muscle. To test facial movements it is imperative to immobilize the
contralateral side of the face which is under assessment. Then each section of the face must be tested in turn: forehead, eye closure and corneal protection, the patient is asked to move the cheek with a smile or blowing, the nose with a sniff, the upper and lower lip with a whistle and the chin by imitating shaving movements.

**Special investigations**

The reader is referred to the appropriate chapters on radiology with regard to discussion of plain radiographs, tomograms, laryngograms, computerized axial tomography, magnetic resonance imaging and radio isotope scanning. Fibreoptic endoscopy and direct examination of the larynx and microlaryngoscopy are dealt with in Chapter 3.
Chapter 2: Radiology of the pharynx and larynx

P. D. Phelps

Radiological evaluation of the mucosal surfaces of the upper aerodigestive tract has always been secondary to clinical examination, but can be useful for showing foreign bodies or the lower limits of a tumour when this cannot be assessed endoscopically. Traditionally the radiographic techniques used have been (a) plain films to show the air/soft tissue interface and surrounding bony structures, (b) intraluminal contrast examinations. High kilovolt techniques, tomography and xeroradiography, singly or in combination, are used to give a better demonstration of the outlines of these air-filled cavities; the first two techniques by partially eliminating the overlying bony structures from the image, and xeroradiography by the edge enhancement effect. However, interpretation is fraught with difficulty because of variations in airway contour, poor radiographic contrast and superimposition of areas of interest. The barium swallow remains the most important radiological examination for the oro- and hypopharynx, especially when combined with cineradiography. It is a simple and easy means of examination, although obviously or more value for lesions below the cricopharyngeus which cannot be assessed with a laryngeal mirror.

High resolution computerized tomography (CT) has greatly improved and extended the imaging capabilities in this region. Not only is the clearest demonstration of the air/soft tissue interface given in axial sections, but also the surrounding fascial planes, muscles and vessels in the parapharyngeal region, are shown. Thus anatomy which formerly could not be assessed, except by invasive contrast examinations such as angiography or sialography, can be seen and CT has proved capable of showing deep pathology that is undetectable by physical examination and sometimes even by biopsy.

The radiographic anatomy and a brief account of some features of disease of the mucosal surface of the nasopharynx are considered in Volume 4, Chapter 2. It is stressed there that demonstration of infiltration by carcinomata into the deep tissue planes of the parapharyngeal region was the most significant advance made by the latest sectional soft tissue imaging techniques. The parapharyngeal space bridges the nasopharynx and oropharynx and so a discussion of the CT anatomy and the fascial spaces and compartments below the skull will be considered, together with a brief account of the relations of the salivary glands to these spaces.

Magnetic resonance (MR) promises to give even better demonstration of these soft tissue structures. Differentiation between mucosal and lymphoid tissue is possible, and there is clear delineation of tumours from surrounding soft tissues. The relationship of a mass to major blood vessels is also shown better with MR than by enhanced CT (Lloyd and Phelps, 1986). Above all, direct three-plane imaging is a major advantage especially in the demonstration of the limits of the tumour.
Imaging techniques

Plain radiography

The best plain film view of nasopharynx, larynx and pharynx is given by the lateral projection with the pharynx and larynx clear of the cervical spine. The film is placed against the shoulder and the incident beam is centred on the angle of the jaw if the nasopharynx is the region of interest, or the thyroid cartilage if the larynx is being examined. Xerograms give a clearer demonstration. Alternatively, a high kilovolt technique may be used utilizing 'low dose' film with screens, or a wedge filter to allow visualization of the range of densities from the neck to the thoracic inlet.

To show the structures low in the neck and upper mediastinum, the central ray is directed to a point below the middle of the clavicle at the level of the thoracic inlet and to the centre of the film, placed in the Bucky tray.

Even with xerography or high kilovolt technique the anteroposterior projection is usually less informative as the air-filled structures of the nasopharynx and larynx are largely obscured by the cervical spine, but it may show tracheal displacement or compression, or a fluid level in a pouch or abscess. To demonstrate a laryngocoele the exposure is made during the Valsalva manoeuvre, but tomography may be necessary. Long ossified styloid processes are best shown on an anteroposterior film taken through the open mouth. All examinations should be performed erect if possible to show fluid levels if present.

Structures demonstrated

On the lateral projection, air in the upper respiratory passages outlines the valleculae and cavities of the larynx and trachea. Soft tissue structures such as the soft palate, base of tongue, epiglottis and aryepiglottic folds are silhouetted against this air background. Occasionally enlarged tonsils may be seen as oval densities, and the cartilaginous eustachian tube may present as a narrow dark slit when filled with air having a rim around it due to the eustachian cushion. The hyoid bone and, if ossified, the thyroid and cricoid cartilages, can usually be clearly seen. Careful note should be made of the thickness of the soft tissues in the nasopharynx and of the prevertebral soft tissues. These are important, as a bulge or increase in thickness may indicate oedema, abscess, haematoma, cyst or tumour. Loss of the normal spinal curvature should also be noted. Surgical emphysema shows as linear streaks of air in the prevertebral plane. In adults the soft tissues in the roof of the nasopharynx should measure not more than 1 cm; they should be regular in outline and thickness. In children, hypertrophy of adenoids may be so pronounced as to obliterate the air space between the posterosuperior wall and the soft palate, and enlarged adenoids may be visible into early adult life. Imaging of the adenoids is considered in Volume 6, Chapter 24.

In infants, particularly in the first weeks of life, the prevertebral soft tissues normally look thick, especially on full expiration, and this should not be mistaken for a retropharyngeal infection. Films should always be taken on inspiration and, if there is any doubt, the film should be repeated on expiration, or a cine run taken with the infant held in an erect lateral position.
The depth of the cervical prevertebral soft tissues normally increases slightly from the level of the anterior arch of the atlas down to the lower border of the fifth and sixth cervical vertebrae, where it blends with the thicker soft tissue shadow of the cricopharyngeus and upper oesophagus. After the age of 2 or 3 years it should not measure more than 4 mm in depth. Below the cricoid the soft tissue thickness between the air-filled trachea and the spine should not normally exceed three-quarters of the diameter of the corresponding cervical vertebra, and should it do so a tumour or inflammation in the postcricoid region or upper oesophagus should be suspected.

Foreign bodies and growths in the larynx or trachea may be silhouetted against intraluminal air. At the thoracic inlet the trachea usually lies centrally in the anteroposterior projection. On the lateral view, it normally lies equidistant between the anterior vertebral border and the posterior profile of the upper manubrium. These relationships may be disturbed by scoliosis or kyphosis, the trachea being displaced towards the concavity of the scoliosis, and forwards with kyphosis. With a straight cervical spine, displacement of the air-filled trachea (or barium-filled oesophagus) indicates extrinsic pressure.

Ossification commonly occurs in one or more of the laryngeal cartilages, although there is considerable individual variation both in age of onset and extent. It is uncommon before the third decade. Ossification occurs most commonly in the thyroid cartilage in which it starts posteriorly and slowly extends forwards and upwards. The cricoid tends to ossify from behind forwards. The stylohyoid ligament not infrequently ossifies in its upper part, and occasionally throughout its whole extent. The styloid processes are best shown on an anteroposterior film taken through the open mouth. Rarely, ossification occurs in the epiglottis and occasionally the cricothyroid ligament is ossified. Compression of the trachea is usually obvious.

The arytenoids may ossify in the absence of ossification in other laryngeal cartilages and present as dense triangular opacities. If superimposed they should not be mistaken for a swallowed foreign body when the examination has been performed to try to demonstrate one. The same applies to ossified triticeous cartilages. The corniculate and cuneiform cartilages are unlikely to be so mistaken because they lie more anteriorly.

The region of the anterior commissure is often difficult to assess endoscopically and lesions of this part of the larynx may be shown best by lateral xerography or by CT which can also demonstrate extension of a tumour into the pre-epiglottic space.

**Tomography**

Conventional tomography still has an important place in the examination of the larynx especially in the frontal projection, although a similar demonstration may be obtained by high-kilovolt techniques. Anteroposterior tomographic studies of the larynx demonstrate the true and false vocal cords and the laryngeal ventricle between them. The cords should be demonstrated in full adduction by obtaining a picture with the patient phonating 'ee' and as near as possible in full abduction in the phase of quiet respiration. Linear tomography is preferred because of the short exposure time and good radiographic contrast. Further views may be obtained in inspiratory phonation to distend the laryngeal ventricles, or with the
patient performing a Valsalva manoeuvre which produces distension of the supraglottic larynx and hypopharynx.

Immobility of one vocal cord due to paralysis or fixation shows loss of the normal acute angle between the undersurface of the cord and the subglottis when the patient phonates 'ee', but such immobility is of course normally assessed clinically. Tomography is of more value for lesions below the cords, especially when there is narrowing as by a tumour or previous intubation.

**Computerized tomography**

Computerized tomography of the larynx is now a valuable addition to available imaging techniques although it has not replaced the traditional ones, especially linear tomography. The axial CT sections produce an image similar to the one seen by the surgeon. Respiratory movement is less of a problem with the new fast scanners, but acceptable images can be obtained in quiet respiration with long scanning times. Scanning is begun at the level of the hyoid bone and sequential scans are viewed. Above the rounded hypopharynx it is bisected by the crescentic epiglottis. Further down the median and lateral glosso-epiglottic folds delineate the valleculae. Below this the airway assumes a triangular shape and the pyriform fossae are seen as two lateral appendages separated by the aryepiglottic folds. At the level of the cords the shape changes to the characteristic glottic chink or boat shape with the sharp anterior commissure extending right up to the thyroid cartilage in the midline. In the subglottic area there is an even symmetrical oval shape which gives way at the level of the first tracheal ring to an oval flattened posteriorly, which may be likened to the shape of a horseshoe. Further sections may be taken with the patient holding his breath or performing a modified Valsalva manoeuvre. A good demonstration of distended pyriform fossae and the supraglottic structures may be obtained.

Computerized tomography provides a non-invasive, quick and effective radiological investigation of the larynx, and is not uncomfortable for the patient. It can be carried out without risk in the face of respiratory obstruction or after suspected laryngeal injury. It gives an accurate assessment of laryngeal anatomy and involvement by tumour, particularly at the glottic level. The value of such an assessment is greatly increased if partial laryngectomy is contemplated, but this is an unusual operation in the UK where carcinoma of the larynx is treated by radiotherapy and/or total laryngectomy. Because CT and conventional tomography present images in different planes, the two are complementary. Whereas CT is better for showing the laryngeal cartilages and structures at the glottic level, conventional tomography is superior for showing subglottic extension of growths, and gives a more satisfactory demonstration than reformatted CT images in the coronal plane.

Fractures of the larynx, as from direct contact with the steering wheel of a motor vehicle, are difficult to demonstrate radiologically and even harder to assess clinically. Xerography may be useful but CT is now the optimum method for showing fractures of the thyroid cartilage, displacement of the arytenoids and the size and state of the airway.

Computerized tomography can assist the endoscopic assessment of tumours both benign and malignant, although it is not required routinely. Some rare benign tumours may show characteristic features. Chondromata arise from one of the laryngeal cartilages and show
a typical appearance on CT with speckled calcification. Plasmacytoma appears radiologically as a smooth rounded tumour, homogeneous in consistency and often indistinguishable from carcinoma of the larynx. Lipoma is a very rare tumour which may be suspected preoperatively because of low attenuation of the fat.

Malignant tumours are nearly always squamous cell carcinomata, and CT may help to evaluate deep laryngeal and paralaryngeal soft tissue invasion, thereby altering the staging of the disease. The paraglottic space, that is the space between the mucosa and the cartilage, is well demonstrated and is seen as a translucent line just deep to the thyroid lamina. When this line is absent on one side, it contrasts with the normal side and indicates tumour infiltration of the thyroarytenoid muscle, up to the thyroid cartilage. The most important role for CT is for showing invasion of the cartilaginous skeleton of the larynx by the tumour. Such involvement is a strong indication for total laryngectomy. Gross involvement of the thyroid laminae is usually obvious on CT, but minor degrees of destruction by tumour invasion may be difficult to evaluate, since the thyroid laminae may show considerable unevenness of density in the normal. This may give rise to both false positives and false negatives in interpretation (Lloyd, Michaels and Phelps, 1981). Another feature which has been associated with invasion of the cartilages, confirmed after surgical excision of the larynx, has been the presence of increased density and ossification. This may involve the arytenoid, cricoid and thyroid lamina. In the author's series of 26 cases, increase in the density of the cartilage was shown to be due to increased ossification and was most often seen in the arytenoid. Previous radiotherapy appeared to be an important factor in most but not all cases.

**Magnetic resonance**

Magnetic resonance images can be obtained in three planes, but this modality has not been fully evaluated at present.

**Soft tissue imaging in the parapharyngeal region**

An axial CT scan with 5-mm contiguous sections forms the basis of the examination. Contrast enhancement, by bolus injection or infusion, is usual to show the position of the major vessels or the presence of a vascular tumour. There is normally clear delineation of the parotid gland from the muscles and from fat in the parapharyngeal space, but sialography may help to define the limits of the deep lobe of the parotid. Magnetic resonance shows the position of the vessels more clearly.

**Infratemporal and pterygopalatine fossae**

The infratemporal fossa is an irregularly shaped space behind the maxilla and medial to the ramus of the mandible. It is situated below the zygoma, the greater wing of the sphenoid and part of the squamous temporal bone. The medial wall of the fossa is the lateral pterygoid plate. The anterior and medial walls meet below, but are separated above by the pterygomaxillary fissure through which the infratemporal fossa communicates with the pterygopalatine fossa. The upper end of the pterygomaxillary fissure is continuous with the inferior orbital fissure. The pterygopalatine fossa is well shown by axial CT and the sphenopalatine foramen, which is an opening in its medial wall, that is the lateral wall of the nasal cavity, can sometimes be seen. The normal fossa contains fat.
Spread of tumours along the axis of the pterygomaxillary fissure with expansion of the walls is an important concept, particularly in the natural history of juvenile angiofibroma. This tumour arises in the region of the sphenopalatine foramen and spreads through the fissure into the infratemporal fossa (Lloyd and Phelps, 1986). This is a much more important sign than the traditional 'bowing' of the posterior wall of the antrum, but erosion of bone in the region of the sphenopalatine foramen appears to be pathognomonic.

The infratemporal fossa contains all muscles of mastication except the laterally placed masseter and the small depressors of the mandible. Most laterally and superiorly is the head of the temporalis muscle which inserts on the coronoid process of the mandible. The pterygoid muscles are protractors of the mandible and fill the bulk of the infratemporal fossa. A significant portion of the maxillary artery also lies within the infratemporal fossa.

The 'spaces' that have been considered so far have some bony boundaries, although inferiorly they are continuous with other described potential spaces whose boundaries are not bony but the deep fascial planes of the neck. Unfortunately these fascial boundaries are not demonstrated by imaging techniques; neither do they interfere with the spread of malignant neoplasms. Nevertheless they must be considered, as expansion of benign tumours and the resultant CT appearances depend very much on the site of origin of the tumour. The two most important of these spaces will be referred to as the parapharyngeal space and the carotid space.

*The parapharyngeal space*

This is the most anterior space and extends from the base of the skull to the hyoid bone. The fat in the space is a prominent feature of axial CT scans at this level. The lateral wall of the parapharyngeal space is formed by the pterygoid muscles. The medial wall is made up of the swallowing muscles. At the level of the nasopharynx, these are principally the tensor and levator palati, but below this the medial wall is formed by the constrictor muscles of the pharynx. The muscles arising from the styloid process form the posterior wall.

As described in the account of the nasopharynx in Volume 4, Chapter 2, carcinomata of the nasopharynx have a marked tendency to spread into and obliterate the parapharyngeal fat space, but this space is also encroached upon by benign tumours arising laterally in the deep lobe of the parotid gland, or medially in the wall of the nasopharynx. When the tumour becomes large the fat planes are obliterated, but before this stage is reached the position of the fat in the parapharyngeal space in relation to the mass is an important indication of whether this mass arose in the pharynx or in the deep lobe of the parotid gland.

*The carotid space*

The carotid space or sheath is a potential fascial space containing the internal jugular vein, the carotid arteries, and the lower four cranial nerves, as well as the sympathetic chain and various lymph nodes. Benign tumours arising in this space are usually vagal neuromata or rarely glomus tumours. These tumours tend to respect the fascial margins of the carotid space and to separate the major blood vessels. The relationship of the blood vessels to the mass is better shown by MR. Lower down at the hyoid level, a densely enhancing but smoothly outlined mass shown by CT is almost certain to be a carotid body tumour.
Rarely a mass may arise more posteriorly in the so-called 'paraspinal space' and displace all the vessels as well as the anterior scalene muscle anteriorly.

Regimen for investigating parapharyngeal masses

At the present time CT in the axial plane with 5-mm contiguous sections and contrast enhancement, preferably by infusion, appears to be the optimum means of investigating these lesions. In selected cases, further sections in the coronal plane may be an advantage. At least one series (Schaefer et al, 1985) has demonstrated better definition of these tumours by MR than by CT, and another series (Lloyd and Phelps, 1986) has shown a better demonstration of the relationship of the major vessels. Angiography is clearly indicated if the lesion appears to be vascular, but seems to have little to offer if there is no contrast enhancement on the CT scan.

Dynamic CT scanning is discussed in Volume 1, Chapter 17 but there has been little experience or success in differentiating neck masses by use of this technique. Som et al (1984) described a protocol based on differentiating a deep lobe parotid mass and an extraparotid lesion. The best way of making this distinction is by demonstrating a fat plane between the deep lobe and the posterolateral aspect of the mass. This fat plane represents the compressed fibrofatty supporting matrix of the parapharyngeal space and, when it is seen, the mass is extraparotid in origin. When the fat plane is not seen between the mass and the parotid gland, the lesion is probably a parotid tumour.

Further discrimination can be made by the degree and timing of the contrast enhancement by plotting attenuation against time curves. Immediate enhancement followed by rapid washout is a feature of vascular anomalies and vascular tumours with little stroma such as glomus tumours, whereas meningiomata show a less rapid but more persistent degree of enhancement. Neuromata almost always enhance less but the degree of enhancement is very variable and difficulty may be encountered in distinguishing between extraparotid benign mixed tumours and the neuromata that do not show enhancement. Their similar appearances on CT scans and the dynamic scan findings do not, in most cases, allow a confident distinction to be made. The internal carotid artery is usually, but not always, displaced anteromedially by neuromata and posteriorly by minor salivary gland tumours. Som et al also found that most extraparotid benign pleomorphic salivary adenomata arise within salivary cell rests in the parapharyngeal space and, therefore, are separate from both parotid and the pharyngeal muscular wall. This has also been the experience of the author.

Few authorities in the UK use CT for assessment of cervical lymph nodes, but at least one series from the USA (Stevens et al, 1985) has shown that CT has greater accuracy than the clinical examination of the neck for the staging of patients with nodal metastases. Their criteria on the CT scan were:

(1) if there was a lymph node greater than 1.5 cm in size;

(2) if the node had central necrosis regardless of size, in the absence of clinical infection, or
(3) if a group of three or more nodes, each smaller than 1.5 cm, was seen in the primary drainage station of the primary tumour, as sometimes enlarged nodes may be demonstrated replacing the parotid gland or within the carotid sheath obscured by the overlying sternomastoid muscle.

It appears that magnetic resonance is inferior to CT for this demonstration, and is unable to show any nodes that cannot be palpated.

**Barium swallow examination**

Assessment of the mucosal surfaces of the oro- and hypopharynx is, as with the nasopharynx, almost entirely by inspection. Barium swallow is of more value for showing lesions of the oesophagus, pharyngeal pouches, fistulae, and for neurological swallowing problems especially when cine is used.

Normally barium flows rapidly through the pharynx and down the oesophagus. It may coat the sides of the pharynx for a short time and, after the first swallow, a little may remain in the valleculae and pyriform fossae only to be quickly cleared by a subsequent swallow. Any degree of stasis beyond this should be suspect. The normal larynx will appear as a 'filling defect' in the frontal projection with contrast in the pyriform fossa on either side. This is well shown on the oblique projection, obtained with the patient swallowing while his head is turned to one side. Tumours of the pharynx will be well outlined by a coating of barium and masses demonstrated in the pyriform fossae, which are sometimes difficult to see with a mirror.

When the larynx fails in its primary function as a protective sphincter for the lungs, 'spillover' will occur to give a 'barium laryngogram'. This problem is seen more and more in an ageing population when dysphagia is often the result of a mild stroke. Cineradiography at four frames per second gives a good demonstration of deglutition. Passage of the bolus across the back of the tongue, with elevation of the larynx and tilting of the epiglottis down over the closer larynx, is shown. Contrast then passes through the open cricopharyngeus into the oesophagus. Minor functional disorders of swallowing can only be shown by this technique, but it is wasteful of film and should probably only be used to try to solve a particular swallowing problem. A good account of pharyngeal deglutition in patients with functional disorders of the act of swallowing, compared with a group of volunteers with no complaint of dysphagia, is contained in two papers from Malmo, Sweden (Ekberg and Nylander, 1982a, b). These authors, using cineradiography at 50-100 frames per second, found a high incidence of epiglottic dysmotility and cricopharyngeal incoordination in the patients with dysphagia. Although only a small percentage had severe disturbance, such as complete paralysis of the pharyngeal constrictors or aspiration into the trachea, nevertheless contrast was frequently seen to enter the vestibule of the larynx.

**Oesophagus**

This is a tubular organ the diameter of which varies from a potential space to the size needed to accommodate whatever can be swallowed. It normally contains no air, although occasionally a small triangular air shadow can be seen on a lateral film just below the level
of the cricopharyngeus. Air is almost always visible in this region when there is a foreign body present in the pharynx or upper oesophagus.

The conventional radiological examination is the barium swallow. It is often stated that barium should never be used if it is thought that spillover into the trachea may occur, but provided small amounts only are used, there seems to be little danger, especially if postural drainage is used afterwards.

Barium is insert and less irritating than most water-soluble contrast media, and usually provides better radiographic detail. In infants, if atresia or a fistula is suspected, it is better to pass a soft rubber tube and to inject a water-soluble opaque fluid, of the kind used for bronchography, only if necessary. Communication between the oesophagus and trachea involves the anterior wall of the oesophagus so that injection of opaque fluid should be made in prone and prone oblique positions under X-ray screen control and films taken as required.

When undertaking a screen examination of the oesophagus, the patient is usually given barium fluid to swallow, although occasionally barium paste is used. The radiologist follows the passage of a mouthful of barium from the mouth to the stomach. The situation, extent, and form of physiological and pathological constrictions or filling defects, or any hesitation, hold-up or diversion of the normal flow are noted. Attention is paid to the form and amplitude of peristaltic waves. Some authorities test the lower oesophageal sphincter for competence by watching the patient swallowing prone while abdominal compression is applied. However, such an unphysiological manoeuvre has little relevance as a test for reflux. Reflux of barium into the oesophagus when the patient swallows water in the supine position is probably a better test.

Zaino and colleagues (1970) have demonstrated that there is a sphincter 1-2 cm long at the upper end of the oesophagus, below the cricopharyngeus, in which increased pressure can be measured manometrically, and stated that this provides the true sphincter mechanism for the upper oesophagus and not the circular fibres of the cricopharyngeus. At times the cricopharyngeus muscle may produce a pronounced indentation of the posterior outline of the filled oesophagus as barium passes through, and this is often especially marked when there is neuromuscular incoordination present, as with bulbar lesions.

Foreign bodies in the upper aerodigestive tract are usually fish or meat bones lodged in the upper oesophagus. It may be difficult to differentiate the foreign body from ossification in the laryngeal cartilages. Air in the soft tissues or in the oesophagus, held open by a non-opaque foreign body, is an important sign. Less often objects such as pins, coins, dentures, buttons, etc may lodge in the pyriform fossa, the nasopharynx or between the cords. Stridor is the presenting feature in such cases. Perforation, either by the foreign body or by instrumentation, can lead to inflammatory changes in the para- and retropharyngeal tissue planes. Widening of the retropharyngeal soft tissue space is seen in such cases, sometimes with surgical emphysema or an abscess cavity.

When one attempts to demonstrate or localize a non-opaque swallowed foreign body such as a fish bone, success can sometimes be achieved by persuading the patient to swallow a sandwich of teased-out dry cotton wool with a centre of cotton wool soaked in barium. The
The upper oesophagus normally deviates a little to the left at the level of the thoracic inlet. It may be indented, compressed or displaced by an enlarged thyroid, or parathyroid gland, or enlarged lymph nodes, as well as by mediastinal tumours or aneurysms of the aortic arch. It is indented anteriorly, and on its left side, by the normal aortic arch and left main bronchus and, when present, the oblique indentation of an anomalous right subclavian artery above the level of the aortic impression is diagnostic. Malformations of the aortic arch, such as right-sided aorta or double aorta, may be suspected or diagnosed by the different impressions they produce on the barium-filled oesophagus.

The oesophagus is usually loosely attached to the descending aorta and tends to maintain this relationship throughout life, so that when the aorta becomes elongated and unfolded with atheroma, the oesophagus tends to be displaced with it. An atheromatous aorta may compress the oesophagus at its lower end where they cross, and this is particularly liable to happen when the thoracic aorta is tortuous and heavily calcified.

Barium swallow is complementary to oesophagoscopy in the diagnosis of obstructive lesions at the lower end of the oesophagus and is the principal means of identifying achalasia of the cardia.

Subsidiary imaging techniques

Other imaging techniques which have a minor or obsolete role in the examination of pharynx, larynx and oesophagus have been briefly considered in Volume 1, Chapter 17. Laryngography is time consuming, uncomfortable for the patient and contraindicated in airway obstruction and stridor. It has not been almost entirely replaced by newer imaging methods for showing the mucosal surfaces of the larynx.

**Sinography**

Cysts of congenital origin in the neck arise laterally from branchial cleft remnants or in the midline along the course of the thyroglossal duct. On CT scans they will appear as well circumscribed low density lesions with normal fascial planes around them unless the cyst has become infected. When a cyst breaks through to the skin it produces a fistula. Valuable information about the situation and extent of fistulae, sinuses and tracts in the neck can be obtained by injecting an oily contrast medium (Lipiodol Ultra Fluid) through a small catheter.

**Sialography**

Pathological changes in the salivary glands are traditionally investigated by plain radiography, or after injection of a contrast medium into the parotid or submandibular ducts. The technique of sialography and its importance for showing abnormalities of the duct systems are discussed in Volume 1, Chapter 17.

Mass lesions within or around the salivary glands are now best demonstrated by CT. The precise location of a mass within the parotid gland can be shown, and the position of the
facial nerve inferred. CT demonstrates whether a mass is circumscribed or invasive and suggests the histological nature of a cyst or lipoma. It can differentiate masseter muscle hypertrophy from diffuse non-inflammatory enlargement of the parotid gland. It is not, however, reliable for differentiating benign from malignant neoplasms. Most intraparotid masses, even those differing little in density from the surrounding gland, are detectable by CT, but intravenous or intraductal contrast injection may be helpful to delineate the tumour more clearly and confirm that the deep lobe is not involved. Adenolymphoma (Warthin's tumour) tends to have multiple lobules and is often wholly or partially outside the gland. Isotope studies with technetium-99m ($^{99m}$Tc) may be diagnostic as Warthin's tumour accumulates the radionuclide intensely. This differs from other tumours which show as areas of decreased activity in the concentration of isotope which occurs in normal salivary glands. Malignant tumours may be well defined, or they may have indistinct margins at their interface with the parotid tissue and may infiltrate outside the gland into the surrounding fat if they are invasive.

**Angiography**

Angiography, by means of transfemoral catheterization, is now largely replaced for the study of neck masses by enhanced CT, MR and digital vascular imaging. There seems to be little point in performing angiography unless the lesion shows significant contrast enhancement on CT scanning. A sparsely vascularized mass, such as a neurofibroma, will show some 'puddling' of contrast on the angiogram, and characteristically there is anteromedial displacement of the internal carotid artery. The paragangliomata or glomus tumours make up the next most common group of enhancing extraparotid parapharyngeal masses (Som, 1984). They may also displace the internal carotid anteriorly, but the angiogram will demonstrate the intense and typical vascularity of the lesion. Carotid body tumours occur lower in the neck. They arise medial to the carotid bifurcation and displace the carotid artery laterally, or at a higher level splay the carotid bifurcation. Although the internal jugular vein can usually be recognized displaced posteriorly on the CT scan, the carotid arteries are usually incorporated in the hyperdense mass.

Vascular masses such as glomus tumours and haemangiomatica, which also occur in the neck, in the salivary glands or the base of the tongue, are extremely difficult to excise and embolization techniques may be necessary to reduce the blood supply prior to surgery. These techniques are described briefly in Volume 1, Chapter 17.

**Summary**

Plain radiography will continue to have a limited role for assessing the outlines of the upper aerodigestive tract, for showing foreign bodies and occasionally for cysts and tumours, especially those presenting problems of assessment for endoscopy. Barium swallow examination still has a most important role for the study of the oesophagus and for some swallowing problems in the hypopharynx as well as diagnosis of pharyngeal pouches and achalasia of the cardia. Computerized tomography has a limited role in the investigation of laryngeal pathology. It is rarely required to assist the endoscopic assessment of tumours but may help to confirm persistence of disease and erosion of the laryngeal cartilage after radiotherapy. Assessment of laryngeal trauma by CT may be of value. Severe injuries to the framework of the larynx require open exploration, but CT may show that this is not required
in less severe case (Schaefer and Brown, 1983). Computerized tomographic scanning is especially useful for demonstrating the anatomical relations and extent of deep neck masses, but has frequently proved unreliable for distinguishing between benign and malignant tumours. Nevertheless, the combination of physical examination and CT gives the opportunity to stage these lesions more accurately than before.

Magnetic resonance promises to give even better discrimination of such masses and normal structures in the soft tissues below the skull. Malignant tumours of the oropharynx seem particularly well suited to assessment by magnetic resonance imaging, especially those in the base of the tongue. The extent of local deep infiltration should be demonstrated and any spread to adjacent areas such as the hypopharynx or pre-epiglottic space. The lateral projection possible with MR, but not directly with CT, is particularly useful. Carcinoma of the floor of the mouth is assessed clinically but its extent, including involvement of the mandible, can be well shown by MR. It is to be hoped that in the future better discrimination of pathology will be possible by magnetic resonance imaging and especially better differentiation of benign and malignant neoplasms.
Chapter 3: Endoscopy

R. P. E. Barton

Since the afternoon in Paris in September 1854 when Manuel Garcia, a professor of singing, saw his own vocal cords with the aid of two mirrors, physicians have been fascinated by the art of endoscopy. At about the same time, Desormeaux, a Frenchman, pioneered the first practical endoscope for examining the urethra, bladder, vagina and rectum, but it was not until 1868 in Freiburg, Germany, that Kussmaul, using a modified Desormeaux instrument, carried out the first oesophagoscopy. His patient was a professional sword swallower! The remainder of the nineteenth century was a time of great interest and advance in oesophagoscopy and laryngoscopy and it is pleasant to record that Garcia lived to the age of 102 and was recognized as the 'Father of laryngology'.

Killian, also working in Freiburg, became known as the 'Father of bronchoscopy', while in the USA in 1907 Chevalier Jackson published the first major text book on endoscopy entitled, *Tracheobronchoscopy, Esophagoscopy and Gastroscopy*. Endoscopes have been modified and greatly improved by modern lighting and telescopic systems, but the two major contributions to endoscopic techniques, in recent years, have been the use of the operating microscope and the introduction and development of flexible fibreoptic endoscopes.

**Outpatient endoscopy**

*Flexible nasopharyngolaryngoscopy*

Early fibreoptic laryngoscopes suffered from poor optical systems, and otolaryngologists were often reluctant to use them. However, as experience was gained both by the manufacturers and users, early problems were overcome and fibreoptic endoscopes became increasingly sophisticated. Present day instruments give excellent images and, with practice, are easy to handle.

The major advantage of the flexible laryngoscope is that it may be used immediately, even in a busy outpatient session, without disruption to the clinic. In the great majority of patients in whom the larynx and pharynx must be seen, and in whom indirect (mirror) laryngoscopy is unsatisfactory, admission for direct examination under general anaesthesia and its attendant expense is obviated. The economic advantages can be readily demonstrated and there should be no difficulty in persuading those who hold the purse strings that these instruments will rapidly pay for themselves (Welch, 1982).

**Indications**

Outpatient fibreoptic laryngoscopy is reserved for inspection only and therefore the indications do not differ significantly from those for indirect laryngoscopy (Chapter 1). Any patient, including cooperative children, with symptoms referable to the upper air and food passages, may be examined. These symptoms include hoarseness or other voice disorder, the feeling of a lump or discomfort in the throat, and problems associated with swallowing solids or liquids. An advantage over indirect laryngoscopy is that the nose and nasopharynx may be inspected during the passage of the endoscope. This is obviously relevant where symptoms
in the larynx or pharynx may be due to nasal pathology. An important advantage over direct laryngoscopy under general anaesthesia is that the dynamic function of the larynx can be more readily assessed.

Failed mirror examination is not the sole indication for flexible endoscopy. As the procedure is non-invasive, it is often useful for teaching or documentation purposes. Excellent still or video pictures can be taken with the wide range of cameras now available. While it is not proposed that the technique is a 'psychological' weapon it is often a valuable reassurance to patients with non-organic disease. They clearly see it as a 'better' investigation than indirect laryngoscopy. An additional bonus is that patients may view their own larynx and pharynx and thus be reassured that nothing has been concealed.

The flexible nasolaryngoscope is also a valuable instrument in assessing patients with neck lumps referred by general surgeons before excision or biopsy of the lump.

**Equipment**

The design of fibreoptic endoscopes and the various accessories, of which there is a wide range, are constantly being updated and refined. At the time of writing the smallest diameter nasolaryngoscope is the Olympus ENF-P2, outside diameter 3.4 mm. A heavier duty model, with biopsy/suction channel, is the same manufacturer's ENF-IT10, outside diameter 5.0 mm. The former costs approximately £3,500 ($5,250) and the latter £7,900 ($11,850) (prices early 1986), but the economic advantages have been mentioned. With regular but careful use and maintenance these instruments should last at least 5 years. When considering purchase of flexible endoscopes it is sensible to take into account other instruments, light sources and camera units already available within the department or hospital. This is probably more important than minor variations in price and specifications between the instruments of different manufacturers. If still or video records are to be made it is strongly recommended that advice is sought from the medial illustration department. Without such advice many inappropriate and expensive purchases have been made.

**Anaesthesia**

Many people can tolerate a fibreoptic laryngoscope passed through the nose and into the pharynx without topical anaesthesia provided that the instrument is carefully manipulated. However, many patients undergoing fibreoptic examination will have failed indirect laryngoscopy and are therefore likely to gag easily. Various methods of obtaining adequate topical anaesthesia have been described: the author's technique is as follows.

The nose is first inspected and the wider side chosen if there is a significant difference. This is sprayed with up to 1 mL of 10% cocaine solution and the oropharynx and back of the tongue are sprayed with 10% lignocaine solution. Five to six sprays (50-60 mg) of Xylocaine spray are a suitable dose. This side of the nose is then packed with a pledget of cotton wool soaked in a further 1 mL of 10% cocaine to anaesthetize further and constrict the nasal mucosa. It is helpful, but not essential, if the patient then sucks a benzocaine lozenge (10 mg) for 10 minutes. Adequate time should be allowed to obtain good anaesthesia as this results in a very low failure rate. This is not time wasted as other patients may be seen in the meanwhile.
Technique

The patient sits upright in a chair with a tilt facility in case of vasovagal attacks or vomiting due to excessive manipulation in the pharynx or cocaine reaction. Similarly, suction and resuscitation equipment must be available even though complications are extremely rare. The nasal pack and any residual lozenge are removed. Normally the examiner stands to the right of the patient and, after lubricating the endoscope suitably, according to the manufacturer's instruction, and applying a demister to the objective lens, passes the endoscope along the floor of the nose, under vision, through the posterior choana and into the nasopharynx. Using the thumb control the tip is gently flexed downwards at the posterior border of the soft palate. Early attempts from this stage onward may not be entirely successful and, as in all things, expertise comes only with practice. The basic movements are advancement and retraction of the cable of the endoscope with the leading (left) hand, combined with rotation of the lens housing by the right hand, the thumb of which controls the lever flexing the tip of the cable (this assumes a right-handed operator). After general inspection of the region the patient should say 'Eee', or just speak. A close view of the cords can be obtained, but in simple outpatient fibreoptic laryngoscopy, the instrument should not be inserted through the vocal cords.

If the objective lens mists over it need not be withdrawn as it may be cleared by asking the patient to swallow. On completion, the endoscope is gently withdrawn and, as with all endoscopy, inspection is maintained on the way out. If topical anaesthesia has been used the patient must not swallow until full sensation has returned, usually in 1.5-2 hours.

Problems and difficulties

Failed examination

In a large personal series (unpublished data) of several hundred patients, examination has failed on three occasions only. In one patient, extreme anxiety prevented introduction of the laryngoscope and a second patient gagged severely whenever the tip of the laryngoscope was advanced beyond the soft palate, despite apparent adequate topical anaesthesia. On the third occasion, the objective lens misted so persistently that no view was obtained. a large overhanging epiglottis may prevent full exposure of the anterior commissure and this may necessitate direct laryngoscopy.

Occasionally, a posterior deviation of the septum, not apparent on anterior rhinoscopy, may impede passage of the endoscope. If so, the other nostril should be anaesthetized, but it is too rare an occurrence to merit bilateral preparation routinely.

Misinterpretation of findings

It should almost always be possible to say whether the larynx and hypopharynx are normal or abnormal. Correct interpretation of abnormality is gained with experience, when correlation between findings at fibreoptic laryngoscopy and direct laryngoscopy should be high. Misdiagnoses at fibreoptic laryngoscopy should not result in harm to the patient, but only deflated pride and education for the examiner when direct laryngoscopy reveals the true diagnosis (see below).
**Lack of biopsy channel**

This is in fact less of a problem than it appears. Anaesthesia and biopsy of the larynx in a busy outpatient clinic are time consuming, potentially hazardous because of bleeding or airway obstruction and, as biopsy forceps in fibreoptic laryngoscopes with such a channel are very small, the histological interpretation of tiny fragments of tissue may be difficult or misleading. If abnormalities are noted at fibreoptic laryngoscopy the patient should be examined later by direct laryngoscopy under general anaesthesia.

**Telescope pharyngolaryngoscopy**

Some laryngologists favour the use of a rigid telescope to examine the pharynx and larynx as a primary means or to attempt to obtain a view when mirror examination has failed. Advantages of rigid telescopes include an excellent view, especially with camera and video, speed of examination and relative economy compared with flexible endoscopes. But their main disadvantage is that they must be passed through the mouth and they may not be tolerated by patients who have failed indirect laryngoscopy.

**Stroboscopy**

Even greater appreciation of vocal cord action may be obtained by examination with a stroboscopic light source. By synchronizing an intermittent flash of light with the vibrations of the cords on phonation their movement can be effectively 'frozen' or reduced to slow motion. Both the fibreoptic endoscope and telescope may be used but, while this is an outpatient procedure, it is too time consuming for a routine clinic. The investigation of voice disorders is discussed fully in Chapter 7.

**Day patient endoscopy**

**Flexible bronchoscopy and oesophagogastroscopy**

The term 'day patient' is used for these procedures as they take considerably longer than simple fibreoptic nasolaryngoscopy and therefore cannot be fitted into a busy outpatient clinic on an *ad hoc* basis. Specific outpatient sessions can be designated for fibreoptic procedures in this area, whereas rigid system endoscopies necessarily entail general anaesthesia with access to an operating theatre and recovery area. An ideal solution is a day ward on a half-session basis, with the morning patients being discharged by 1300 h to allow afternoon admissions. This maximizes the use of beds and caters both for patients requiring topical anaesthesia, with or without sedation, and for those who need general anaesthesia. It is important, particularly for the latter, that the general state of health of the patient is assessed beforehand and that the patients are warned that they will not be able to drive themselves home.

A further important difference compared to fibreoptic laryngoscopy is that a trained assistant is essential. Flexible bronchoscopes and oesophagogastroscopes require the operator to use both hands on the control housing simultaneously during long periods of the procedure as both these instruments incorporate suction and biopsy channels.
In addition to the tip control, as found on the flexible laryngoscope, channels for the passage of instruments, suction and insufflation are also present. The assistant is therefore needed to advance and retract the endoscopes as instructed and also helps with the introduction and removal of instruments from the biopsy channel.

Flexible system bronchoscopy and gastroscopy are normally undertaken by cardiothoracic surgeons or chest physicians and gastroenterologists respectively, rather than by otolaryngologists, and therefore it is not intended to give any great detail as to technique in this chapter. Batch (1985) has suggested that otolaryngologists could usefully employ the gastro-oesophagoscope to examine the gastrointestinal tract, even as far as duodenum, but it seems unlikely that a practising otolaryngologist, even if he correctly diagnosed pathology that far from his own area, would want to trespass on the territory of the gastroenterological physicians or general surgeons so far as treatment was concerned. However, there are patients with symptoms referable to our own area in whom it is impossible to obtain an adequate view by the traditional rigid system pharyngo-oesophagoscopy in whom fibreoptic examination provides the diagnosis. Even so it is the author’s practice to refer such patients to a colleague who is undertaking many such procedures each week. Correct interpretation of visual findings lies in the experience of the observer (Batch, 1985).

If oesophagogastroscopy is of little practical application for otolaryngologists it is well worth our while learning to use a fibreoptic bronchoscope. Possibly the commonest reason for otolaryngologists to use a bronchoscope is for patients with a hoarse voice due to a paralysed vocal cord. If this is caused by a carcinoma of the bronchus, the thoracic surgeon usually cannot offer a surgical cure and, thus if confirmation of the diagnosis can be obtained at the time of laryngoscopy the patient is spared a second procedure. Significant advantages of the fibreoptic bronchoscope over the rigid instrument are its smaller diameter and its innate flexibility thus giving much greater access within the tracheobronchial tree for inspection and biopsy.

The other ways in which the otolaryngologist will appreciate its value are in the removal of foreign bodies or secretions from the chest. Few departments are equipped with the full range of grasping forceps which have been designed over the years to remove the wide variety of foreign bodies which may be inhaled, and it is a reassurance to have a flexible bronchoscope available to remove small foreign bodies, particularly from the periphery of the bronchial tree. Inspissated and impacted secretions may act in the same way as foreign bodies and cause segmental collapse. The fibreoptic bronchoscope may prove vital if they cannot be moved by conventional suction and physiotherapy.

**Instruments and techniques**

There is now a wide range of instruments available from several different manufacturers most of whom are prepared to put together a 'package' tailored to the user's requirements. No advice will be given here on particular instruments except to recommend very strongly that individuals or departments hoping to buy flexible endoscopes should, if appropriate, talk to others within their hospital already using these. Interdepartmental compatibility is not only sensible but may also have considerable economic advantages, as with nasolaryngoscopes it is also essential to discuss the purchase of any photographic or video equipment with the department of medical illustration.
The actual techniques are not difficult to learn, but should be acquired from someone already experienced in the procedures. As mentioned, the skill often lies in the interpretation of the visual findings and this only comes from practice. Briefly and for completeness, gastro-oesophagoscopy is normally carried out with sedation - oral or intravenous diazepam is commonly used - and topical anaesthesia of the pharynx by lignocaine spray or benzocaine lozenges. It can, of course, also be performed under general anaesthesia. The endoscope is passed through the mouth, but no anaesthesia of the oesophagus and beyond is needed.

Fibreoptic bronchoscopy is best carried out on the conscious patient. Individual techniques obviously vary, but an acceptable method is to begin as for fibreoptic laryngoscopy and supplement the topical anaesthesia by spraying 10% lignocaine, via the bronchoscope, into the larynx when seen and similarly, after the cords have been passed, into the tracheobronchial tree as the endoscopy progresses.

**Rigid system endoscopy**

Provided there is no risk of airway obstruction or perforation of the pharynx or oesophagus there is no reason why rigid system endoscopies cannot be carried out in a day case operating theatre. However, as patients undergoing these procedures have traditionally been admitted, partly for administrative reasons and also as many need thorough medical and anaesthetic assessment, this subject will be covered in the next section.

**Inpatient endoscopy**

**Indications**

Rigid system endoscopy has been a standard method of diagnosing disease of the pharynx, oesophagus and laryngotracheobronchial complex throughout this century. Despite the recent increase in the use of flexible endoscopy, the place of rigid system endoscopy has not decreased, perhaps with the exception of exclusion endoscopy where flexible endoscopy may provide the answer in a less invasive manner. There is no likelihood, in the foreseeable future, that rigid endoscopic techniques will become obsolete or of less use and it is therefore most important that all trainees should have a thorough grounding in them. As with the fibreoptic endoscope this can only be learned with practice, initially under close supervision, and is even more important with rigid endoscopes as it is much easier to do damage with them. Serious complications with flexible endoscopes are rare, but many patients have had their oesophagus perforated with a rigid endoscope.

The following three indications are suitable for rigid system endoscopy.

**Diagnosis**

Rigid system endoscopy is suitable for obtaining tissue for histological examination from patients with symptoms referable to the upper air and food passages. (It is worth mentioning here that if lymphoma is a possible diagnosis many pathologists now prefer tissue dry, rather than in formalin or other fixative, to facilitate immunological typing.)
Treatment

The human race will certainly never cease to insert different foreign objects into its various orifices. For a century now, rigid system endoscopy has been the method of choice for removing foreign bodies from the pharynx, oesophagus, larynx or tracheobronchial tree. Endoscopic dilatation of benign or malignant stricture of the oesophagus is another traditional way in which rigid system endoscopy has been used for treatment and benign lesions such as polyps or nodules are readily removed endoscopically from the larynx. More recently, the laser has been employed in this manner and now, particularly in some conditions such as juvenile papillomatosis and premalignant lesions of the larynx, it is probably more efficient than the older methods of ultrasound, suction diathermy or removal with forceps. The endoscopic diathermy of pharyngeal pouches is described in Chapter 14, and a further example of the use of rigid system endoscopy for the purpose of treatment is submucosal injection of Teflon into paralysed vocal cords to improve the voice.

As part of other procedures

An essential step in the treatment of a pharyngeal pouch is to pack it with ribbon gauze endoscopically to give bulk to the pouch and thus facilitate its location and dissection.

'Pan'-endoscopy

Pan-endoscopy here refers to a full examination of the upper air and food passages when, for example, a search is being made for a primary cancer in a patient presenting with a neck lump which is possibly malignant. It should include careful examination of the mouth, oro- and hypopharynx, oesophagus, larynx, tracheobronchial tree and nasopharynx. The latter is covered in Volume 4, Chapter 3 and will not be referred to further in this chapter despite its importance. Furthermore, it may not be necessary to carry out a full examination on every patient, but it is more convenient to present pan-endoscopy here as one entity and the author's preferred technique will be described.

Preoperative assessment

Except in an emergency the following routine is carried out:

Clinical examination

The pure anatomy of the region will not be repeated as this is covered in Volume 1 and standard anatomy textbooks. However, clinical examination of the mouth, indirect laryngoscopy and palpation of the neck allow the surgeon to appreciate the shape of each patient. Factors which may influence the endoscopy such as capped or loose teeth, unusual configuration or stiffness of the jaw and neck should be noticed. Severe trismus may affect surgeon or anaesthetist and the latter will wish to be warned of any suspected or significant narrowing of the airway. A general medical history is also taken and the patient's chest, heart and abdomen are examined in the routine manner. If there is any doubt about the general health then electrocardiography is arranged, blood is taken for haemoglobin, urea and electrolytes and other investigations as appropriate.
**Radiology**

X-rays of the chest and a lateral view of the neck are carried out. Aortic aneurysms were once a significant worry to endoscopists. It is certainly helpful to be forewarned of any pathology within the mediastinum.

**General measures**

The patient is starved for a minimum of 4 hours and suitable premedication is ordered by the anaesthetist.

**Instruments**

Many hospitals still use the standard Negus instruments and these are normally quite adequate, with modern fibreoptic lighting, for pan-endoscopy. However, as systems for cleaning, sterilizing and storing instruments and then presenting them to the surgeon vary widely it is essential that these are checked carefully on each occasion before the induction of anaesthesia. It is annoying for the surgeon and dangerous for the patient to suspend the endoscopy in mid-procedure while someone is sent to hunt for a suction tube or biopsy forceps of the correct length. Similarly light sources and suction apparatus must be checked before starting.

**Anaesthesia**

This is clearly the province of the anaesthetist and each technique will vary to some extent. However, it is essential to understand clearly that the anaesthetist has overall control and the right to the airway. Where a surgeon and anaesthetist are used to working together there will normally be no problems of communication, but where the pair do not know each other's habits well, the surgeon must be prepared to demur immediately to his colleague. In a full pan-endoscopy the present author carries out the laryngoscopy and bronchoscopy using an 'apnoeic' technique with no tube, thus maximizing the view of the larynx. Intravenous thiopentone and suxamethonium are used to keep the patient asleep and paralysed. Oxygenation is maintained by intermittent manual ventilation via face mask during the laryngoscopy and by Venturi injection down the bronchoscope during this part of the procedure. When these are completed a standard endotracheal tube is passed and anaesthesia then maintained by appropriate concentrations of halothane, oxygen, and nitrous oxide while the pharyngoscopy and oesophagoscopy are performed.

A small tube does allow a more leisurely examination of the larynx, but this is normally reserved for patients with potential airway obstruction as the anaesthetic technique described above still allows ample time for diagnosis, assessment of lesions, biopsy and teaching.

**Surgical techniques**

It is pointless to pretend that pan-endoscopy is a sterile technique and therefore to scrub up, put on gown and gloves and towel the patient is a waste of time and money. Indeed, covering up all but the patient's mouth with green towels can be positively dangerous as
important danger signs such as cyanosis or poor chest movements may be missed. Except in small children in whom a long procedure is expected, when cooling may become an important factor, it is better to leave the head and neck completely exposed and nothing more than a thin gown covering the chest. The lower half may be blanketed as appropriate. It is sensible to wear a pair of clean, but not necessarily sterile, gloves to prevent acquiring any infection from the patient.

When anaesthetized the patient's head and neck are extended. This can be achieved most simply by pushing the patient's ordinary pillow under the shoulders and allowing gentle extension. Great care should be taken to prevent the head from 'hanging' and particular gentleness must be exercised in manipulating the head and neck of patients with cervical osteoarthritis. Even if no long-term damage is caused it is unkind to inflict a stiff neck, even temporarily, on a patient.

Specially designed pieces are available to fit the ends of tables for endoscopy but seem less fashionable nowadays. It is certainly worthwhile learning to endoscope patients without these to be prepared for endoscopy in another operating theatre.

When the patient is adequately oxygenated the face mask is removed, a swab or silastic gum shield used to protect the upper gum or teeth and a lubricated Negus laryngoscope inserted. The epiglottis is identified and the laryngoscope passed posterior to this and on to view the larynx. If the patient's configuration prevents a good view, an assistant, with direction if necessary, can apply pressure externally on the laryngeal complex. The surgeon must never rotate the endoscope on the fulcrum of the upper teeth and the instrument must only be lifted (upwards as the patient lies on the operating table). The Negus laryngoscope gives a good general view of the larynx and, being broader at its distal end, is often the best endoscope through which to take biopsies. However, to examine the larynx fully it is also necessary to use what is conventionally known as the 'anterior commissure' laryngoscope as, with its narrower distal end, a more precise view is obtained of the less accessible parts of the larynx, and in particular, the anterior commissure, subglottic region, ventricles and posterior surface of the epiglottis which must be carefully inspected. For the removal of polyps or nodules and the taking of biopsies, an upward cutting pair of punch forceps is usually ideal. These small biopsies must be handled very carefully to prevent the histology report returning as 'crush artefact' and should therefore be transferred directly from the jaws of the punch forceps to a histology pot using only a hypodermic needle.

After completion of laryngoscopy a suitable size of bronchoscope is passed through the larynx turning the instrument through 90° to facilitate passage of the tip of the instrument past the vocal cords and then returning it to its normal orientation once it is in the trachea. The anaesthetist is informed, and the Venturi injector attached and used as necessary to maintain oxygenation. It is wise to keep a thumb on the connector if one's eye is anywhere near the proximal end of the bronchoscope. It can fly off at dramatic velocity despite the normal screw lock attachment. The bronchial tree is then inspected systematically beginning with the normal side if disease is thought to be unilateral. As each main bronchus is entered the patient's head must be turned gently to the opposite side. Telescopes to alter the angle of vision may be used as necessary, and biopsies are taken of any suspicious lesions. Straight forceps are usually more effective in the bronchial tree unlike in the larynx. If appropriate, bronchial washings using 20 mL of normal saline and collecting the aspirate in a suction trap
may be carried out. Up to this point in the endoscopy the patient has remained paralysed and therefore if vocal cord movement is to be assessed the anaesthetist should be informed so that the effect of the muscle relaxant can wear off. A MacIntosh anaesthetic-type laryngoscope is then inserted with the tip in the vallecula and the vocal cords are observed. This is more accurate than using the heavier Negus laryngoscope behind the epiglottis as this may in itself cause distortion of the larynx and impairment of the vocal cord movements.

The patient is now intubated and examination of the pharynx and oesophagus carried out. The Negus oesophageal speculum, approximately 22 cm in length, will give a view of the oropharynx, hypopharynx and the upper oesophagus. The upper teeth or gum are protected and a full inspection carried out. It does not really matter in what order the inspection is carried out, but it is important to develop a system early in training so that no area is overlooked. The exception to this is when a lesion is noted or when it is suspected bleeding may occur from one part. Then the remainder of the area should be examined first and the biopsy, or close inspection of a particular site, left until last. Again, no rotational force must be used on the upper gum or teeth. Occasionally these are so prominent that examination with the endoscope in the midline is impossible. Often only the incisor teeth remain and it is perfectly in order to examine the hypopharynx from either side.

After examination of the hypopharynx the tip of the speculum is passed behind the larynx and advanced as far as the cricopharyngeal sphincter, approximately 15 cm from the incisor teeth in adults. If this is open, or opens easily, the endoscope is introduced into the upper oesophagus. An aid to easy passage through the sphincter is to slip the left thumb behind the lower teeth or gums and lift, that is protrude, the mandible. If the cricopharyngeus is in spasm, further intravenous suxamethonium may be helpful but, as in all endoscopic work it is essential that no undue force is used. It is normally possible to see approximately 5 cm of the upper oesophagus with this instrument and, particularly when looking for small foreign bodies, it is essential to keep looking as the instrument is withdrawn.

To examine the mid or lower oesophagus longer instruments are needed depending on the size of the patient and it is important to remember that though these instruments are rigid and straight the oesophagus is not! In order to examine the whole length safely, the help of an assistant to hold and move the patient's head is normally necessary. Initially, to pass the longer instruments through the cricopharyngeus the head is gently flexed. Once the instrument is in the cervical oesophagus the head can be extended and, as the oesophagus curves to the left in the upper chest the head is turned to the right.

As the cardia is reached and the oesophagus swings back to the midline the head can be straightened. These movements should obviously be reversed as the endoscope is removed, again carefully observing the oesophageal lumen during removal of the endoscope. The treatment of pathology within the oesophagus, such as dilatation of strictures, is discussed in Chapter 22. As stated earlier these procedures need not all necessarily be carried out each time on each patient and can all be carried out independently.

There seems general reluctance to palpate the mouth of patients either awake or asleep. It is surprising how much information may be obtained by careful palpation within the mouth and neck at the time of endoscopy. For example, if one patient in whom jaw shape and very prominent teeth prevented any view of the pharynx with a rigid instrument, but in whom there
was a high level of suspicion of a carcinoma of the pyriform fossa because of an overlying neck lump, the present author made the diagnosis by sliding a finger down the corner of the mouth, feeling the tumour and obtaining a truly 'blind' biopsy. The neck should also be repalpated as it is not unusual to feel enlarged cervical nodes under anaesthesia which have not been felt previously, particularly in people with short necks who may be somewhat nervous before surgery.

Postoperative care

Many patients undergoing endoscopic procedures will do so once only, for example to remove a foreign body, but many will also return for further procedures. It is therefore most important to write an accurate account of the findings immediately. Many surgeons prefer to draw their own diagram of the larynx and pharynx, but rubber stamps and printed diagrams are available and are particularly helpful for accuracy in the bronchial tree. Any lesion, particularly if it is possibly malignant, should be assessed meticulously as to its extent and this then drawn into the notes with an accurate description. 'Fair-sized ?SCC right larynx', is far from helpful without a good diagram. The notes should also contain postoperative instructions to the nursing staff and likely dispersal of the patient on discharge.

Postoperative instructions should always state when the patient may eat or drink. Something should be written in the notes, even if it is only 'See standard instruction for "X" procedure on ward'. Such a list of standard instructions for each endoscopic procedure on the ward is a useful device provided that people read it. In uncomplicated endoscopies it is now the author's practice to allow clear fluids when the patient is fully recovered from the anaesthetic - usually 2-3 hours - and, if tolerated well, a light meal a further 2-3 hours later. However, where there has been any difficulty, particularly if there is a possibility of a perforation or tear in the pharyngeal or oesophageal mucosa nothing is given by mouth for 6-12 hours, while the temperature, pulse and blood pressure are carefully monitored. Again, unless there has been any cause for concern during surgery or in the postoperative period, a routine postoperative chest X-ray is not obtained, although some surgeons still prefer to do so.

Complications of surgery

Laryngospasm

This is fairly common immediately after operation and is due to blood or other secretions in the larynx or irritation of the larynx by passage of endoscopes or tubes. Most anaesthetists prefer not to use local anaesthetic on the vocal cords as it is important that the patient's cough reflex should recover immediately in view of the possibility of blood in the larynx or pharynx. Patients should therefore be kept in the recovery area with full facilities for resuscitation and reintubation until the danger of laryngospasm is past.

Missing teeth

Even if great care has been taken with loose teeth during the endoscopy, the use of a sucker in the postoperative period or the patient biting on an airway may break or dislodge teeth. If there is a possibility of a tooth, or fragments of one, having been inhaled, then a
chest X-ray must be obtained. If any such fragment has entered the bronchial tree the patient must be returned to theatre to have it removed by bronchoscopy.

**Perforation of the pharynx or oesophagus**

If care is taken with the endoscopy this should be rare. If there is a possibility that it has occurred the patient should be starved and observed carefully until its presence or absence is proved. Suggestive symptoms and signs of perforation include pain in the neck or in the chest radiating to the back, a rising temperature and pulse, air emphysema in the neck and widening of the mediastinum or pneumothorax on chest X-ray. A nasogastric tube should not be used initially as this may be passed through the perforation. A thoracic surgeon should be consulted. The perforation can be treated surgically or medically by intravenous fluids with broad-spectrum antibiotics via the same route; nothing should be allowed by mouth. Most perforations heal rapidly on this regimen, but it is wise to confirm this with a contrast swallow before letting the patient resume a normal diet. However, if the perforation has occurred through a carcinoma then emergency arrangements must be made to carry out the surgical procedure appropriate to the particular tumour in order to minimize the risk of seeding tumour into the neck or mediastinum. Instances are related, some doubtless apocryphal, of perforation into an aortic aneurysm, the great vessels or even the heart. The operator is hardly likely to fail to notice such an event, but the patient is unlikely to remember it.

**Microlaryngoscopy**

Kleinsasser introduced and helped to popularize the use of a new design of laryngoscope to be used in conjunction with the operating microscope in the early 1960s. These instruments are now familiar and have three particular features, namely, the matt black finish to prevent glare and reflection from the microscope, the broader lumen, and their ability to be stabilized allowing the operator both hands free for manipulation with the larynx. Kleinsasser's design has since been modified by several surgeons, but the basic features persist. Other advantages stem from the magnification and excellent light provided by the microscope which, in turn, also allows more lucid teaching of students and trainees, either via the side arm or through closed-circuit colour television with a camera in place of the side arm. Permanent records in the form of video tapes, which have now largely replaced colour cine film, and still photographs add to the teaching value. Furthermore, accurate laser therapy is now available by linking an appropriate machine to the microscope.

Compared to conventional laryngoscopy there are two major disadvantages. The first is the cost: although most otolaryngology departments possess a microscope it may not be possible to modify it for laser work. The second is that procedures inevitably take very much longer, particularly if photography is undertaken. Many otolaryngologists still feel that they have adequate vision, lighting and instrumentation with the conventional instruments to remove small laryngeal polyps and nodules without resorting to the microscope for procedures which take a matter of 5 minutes or less, including induction of an recovery from the anaesthetic. Certainly, assessment of tumours should still be carried out with the traditional instruments as the laryngoscope is dynamic in this role whereas in microlaryngoscopy it is static.
Procedures particularly suited to microlaryngoscopy include laser excisions of laryngeal lesions and microdissection, as in stripping of polypoid or premalignant vocal cords where it is necessary to grip mucosa with one instrument and dissect simultaneously with a second. In neonates or small infants magnification is often helpful.

**Anaesthesia and surgical technique**

Preoperative preparation is as for conventional laryngoscopy. A small endotracheal tube (5-6 mm) rarely obstructs the surgeon's view within the larynx, thus most anaesthetists prefer to intubate for microlaryngoscopy as the procedures last considerably longer than conventional laryngoscopy. However, a recent innovation of high pressure, rapid pulsed oxygen via a small needle for oxygenation while maintaining anaesthesia intravenously gives a completely unobstructed view (Smith, 1982).

When the patient is anaesthetized he is positioned as described previously. Again there is no advantage in scrubbing and towelling up just because the procedure may appear more 'sophisticated'; bacteriologically it is no different. Unless the patient is well known to the surgeon a diagnostic endoscopy with Negus instruments should be carried out first to alert the surgeon to any likely difficulties resulting from unusual anatomy or pathology. A suitably sized microlaryngoscope is then inserted using, initially, a conventional lighting system. The patient's chest is then padded or, alternatively, a rigid bar fixed to the side of the operating table. When the larynx is clearly seen the laryngoscope is fixed with a Lewy, or other suitable holding device. The microscope with a 400 mm objective lens is swung into place and it is often helpful at this stage to have some head-up tilt on the operating table to make the operating position as comfortable as possible. Another advantage is to have an experienced assistant who puts the right instrument in the endoscopist's hand at the right time, thus minimizing the length of the procedure and strain on the eyes from constant looking up and down with its attendant necessity to refocus.
Chapter 4: The mouth

I. J. Mackenzie

The normal mouth

The normal adult mouth contains 32 teeth, supporting structures, tongue and oral mucosa. All are easily examined and any abnormalities may be a good indicator of local or systemic disease.

The ideal mouth has both perfect appearance of the soft tissues and occlusion, the teeth having erupted into their correct positions. Structural defects of the teeth are not common: anomalies of the development of teeth include an abnormal number (less or more than normal), disorders of eruption, and defects of structure.

The third molars, second premolars or the upper lateral incisors are the most common teeth to be missing in the adult. If young girls have missing upper lateral incisors the upper canines are very prominent. Anodontia or the complete failure of the growth of any permanent teeth is extremely rare; such patients keep their deciduous teeth for a long time, but need dentures at an early age. Anhidrotic hereditary ectodermal dysplasia and Down's syndrome are both associated with anodontia or hypodontia, as in cleft palate.

The deciduous teeth usually begin erupting at about 6 months; the process is complete by about 2.5 years. The permanent dentition starts erupting at the age of 6 years and usually by the age of 21 all the teeth have erupted.

Delayed eruption of the teeth is associated with cretinism, rickets, cleidocranial dysplasia and cherubism. In hereditary gingival fibromatosis the teeth appear not to have erupted because of the nature of the proliferating tissue of the gingiva. More commonly, delayed eruption is the result of local obstruction.

Structural defects

Hypoplasia and hypocalcification of the teeth are the most common diseases. The deciduous teeth are not often affected except in severe illness such as neonatal jaundice when the teeth erupt with a yellow colour; in congenital porphyria the teeth may be red, and after large doses of tetracycline the teeth may be discoloured.

The permanent teeth may be affected by local or systemic influences. The systemic disease amelogenesis imperfecta is often divided into two groups: hereditary enamel hypoplasia and hereditary enamel hypocalcification. Both have a variable pattern of inheritance. In the former, the teeth although of an unusual appearance are reasonably strong, whereas in the latter the enamel is very weak and the teeth crumble.

Dentinogenesis imperfecta, an uncommon autosomal dominant hereditary disease, is associated with osteogenesis imperfecta. The enamel of the teeth appears normal but because of the weak underlying dentine the enamel has no support and the teeth collapse.
In congenital syphilis the appearance of the teeth is characteristic: the incisor teeth of the permanent dentition have a notch in the incisal edge (Hutchinson's incisors) and the first molars may be dome-shaped (Moon's molars).

Although rickets, hypothyroidism and scurvy are uncommon, during the time of the illness the structure of the developing teeth is altered and areas of defective enamel with pits and grooves can be seen when the tooth erupts. The antibiotic tetracycline, although known to cause discoloration of teeth, is still often given to children, causing subsequent abnormality of the teeth. In these patients the teeth are of normal strength but the bright yellow banding turns to brown, and the teeth may need to be crowned to cover the unsightly appearance.

Dental fluorosis is a result of too much fluoride in the water supply, appearing at levels greater than 2 parts per million, which is achieved in certain geographical areas. The teeth have a mottled appearance and these are usually permanent teeth. The mottling may be minimal but in severe cases the enamel is grossly affected.

Dental caries is probably the commonest disease of western society and results in the premature loss of millions of teeth each year. Caries insidiously causes the breakdown of the enamel, then rapidly spreads in the softer dentine and ultimately affects the dental pulp causing the infection and death of the neurons and blood supply to the teeth. The infection so caused will affect the supporting tissues of the teeth resulting in abscess formation.

Although many theories are adduced it is generally agreed that the carbohydrates in the diet are the underlying cause of the disease and that three factors are important: a bacterial plaque containing cariogenic bacteria, a bacterial substrate (commonly sugar), and susceptible tooth surfaces.

The importance of cariogenic bacteria was confirmed by experiments on germ-free animals on a sugar rich diet who subsequently had cariogenic bacteria introduced and then developed caries. It is probable that the most likely mediators of dental caries are strains of Streptococcus mutans producing insoluble glucans.

Much research work has been directed to find a suitable vaccine that might control these bacteria but the work is still only showing success in monkey models.

Dental plaque is the adherent deposit which forms on teeth and consists of bacteria in a matrix of organic material, and is instrumental in starting both dental decay and periodontal disease. If teeth are cleaned correctly the plaque is removed from most of the surface of the teeth, although it is very difficult to remove from the pits and fissures. Plaque forms very quickly and if left alone for 48 hours it contains active growth of streptococci, lactobacilli, actinomyces, diphtheroids and various Gram-negative anaerobes.

Sugars (sucrose) diffuse rapidly into plaque and it has been shown that the pH of plaque after an oral rinse of 10% glucose will drop within 5 minutes from a pH of 6-8 to that of about 5 and the return to normal pH takes over an hour. When the pH is so low the enamel may become decalcified by the acid production of the plaque. Of course if the mouth is
exposed longer to sugar the pH of the plaque remains low for longer, which is why toffees or a high carbohydrate diet are associated with a high caries rate. A famous study was carried out in Vipeholm in 1947 in which children were given different diets and those fed toffees between meals had a very marked increase in their caries rate. In addition, a study in Turku indicated that caries activity fell by 90% when xylitol was substituted for sucrose.

Dental caries affecting the enamel usually causes no symptoms and although examination might show some slight discoloration, there is initially no bacterial invasion. When the caries reaches the dentine there may be some symptoms and if there is a break in the enamel, the dentine, having a good nerve supply, responds to hot and cold stimulation. This stage is treated conservatively by removal of the decay and replacement with either an amalgam restoration or with a composite repair of an anterior tooth. It is important at this stage to remove all the decay and to line the cavity with a material so that heat or cold are not passed to the dentine through the metal restoration.

If the caries is allowed to progress it leads ultimately to a pulpitis or inflammation of the pulp, resulting in a severe toothache often only relieved by analgesics. Even at this stage the vital nature of the tooth may occasionally be saved by the removal of the irritant caries and placing a calming dressing over the pulp. The dentine layer can grow actively to protect the pulp, and a layer of secondary dentine may be laid down.

If the pulp is left alone, the pain disappears as the inflammation turns to necrosis and the pulp dies. The tooth is now non vital. When the caries has reached this extent it has destroyed a large amount of the dentine of the tooth, the support for the enamel disappears, the enamel starts to fall away and the patient is left with a big cavity.

As the infection reaches the apex of the tooth an abscess may form causing pain, made worse by touching the tooth. There may be also a generalized reaction and inflammation affecting the relevant jaw. In the upper jaw, the swelling may reach the orbit and may mimic a frontal sinus infection. Cervical lymphadenopathy may also be found.

Although at this stage it may be prudent to remove the tooth, the pain will be eased by cleaning out the canal of the root and releasing the pus at the apex. If the tooth settles down it may later be saved by root filling the canal down to the apex.

**Oral hygiene (periodontal disease)**

Periodontal disease is nearly as destructive of teeth as caries and is characterized by gingivitis, inflammation of the gums, destruction of the periodontal membrane and loss of alveolar bone. It is classified into gingivitis (acute and chronic) and periodontitis (acute, chronic and juvenile).

Gingivitis may start in childhood. Although it may not be obvious at first, it progresses insidiously and leads eventually to periodontitis with the subsequent loss of the teeth.

It is very difficult to draw a dividing line between gingivitis and periodontitis. Local factors cause most periodontal disease but systemic disease such as diabetes, leukaemia and scurvy have a very profound effect on the gingiva and supporting tissues.
Gingivitis is usually chronic but acute ulcerative gingivitis or Vincent's gingivitis is occasionally seen. This infection of the mouth is caused by *Borrelia vincentii* and *Fusobacterium nucleatum*, both part of the normal flora of the mouth. The infection is more common in young adults and in neglected mouths. The mouth is painful, the dental papillae bleed and are ulcerated, and the smell of the breath is usually quite startling. Fortunately, the infection is quickly controlled by good oral hygiene and metronidazole.

Chronic gingivitis, which is extremely common, is the result of accumulation of plaque and may be controlled by good oral hygiene: good tooth brushing, interdental flossing and ensuring that all infectants are removed. In man, the severity of gingivitis is related to the amount of plaque present on the surface of the teeth; even if plaque has been present for some time, the inflammation around the teeth will subside if it is removed. In chronic gingivitis the bacterial flora around the teeth changes as the inflammation becomes more severe: it is thought that endotoxins from these bacteria cause the damage to the tissue which leads to periodontitis.

Periodontitis, like gingivitis, may be acute or chronic. In acute periodontitis the tooth is traumatized by biting heavily on something and the supporting periodontal ligament is stretched and damaged. A periodontal abscess forms by infection in a deep periodontal packet and causes acute pain.

Chronic periodontitis is characterized by destruction of the periodontal fibres, resorption of the alveolar bone, migration of the epithelial attachment along the root towards the apex and the formation of pockets around the tooth. The most common feature is the formation of plaque which extends into the pocket: some of this plaque is calcified, forming calculus. The attached epithelium migrates away from the enamel and subsequently exposes the cementum, and the periodontal membrane and alveolar supporting bone are destroyed. The subgingival calculus is an important feature of the disease: it is a source of infection, helps to perpetuate inflammation and retards healing.

Often the first symptom is loosening of the teeth because the destruction below the gingival surface is not painful and to the patient's eye the gingiva looks no different than it has done for years. To save the teeth, treatment at this stage must be vigorous: the subgingival calculus is removed and good oral hygiene instituted. Diet, irregularities of the teeth, prosthetic appliances, cavities and poor restorations are all aggravating factors which should be corrected.

Surgery may be needed including removal of the pockets around the teeth, and exposure of the teeth to remove the subgingival calculus. There are several procedures but the reattachment of the gingival and periodontal tissues to the tooth is the aim; after a successful operation there is evidence of new bone formation and repair of the periodontal membrane.

Nowadays, dental surgeons are much more aware of the ravages of periodontal disease, the care of the gingival tissue has a high priority, and instruction in good oral hygiene is now very important. (The method of payment of dental surgeons in the National Health Service is changing to take account of this new emphasis on mouth care.)
Prevention of dental caries and periodontal disease depends on the common denominator of dental plaque, and patients must understand the purpose of plaque control. Although fluoride and measures to remove stagnant areas from the teeth help to combat these diseases, good oral hygiene with removal of all plaque keeps caries and periodontal disease away.

**Stomatitis**

Stomatitis is a collective name for inflammatory disease of the oral mucous membrane. The skin and oral mucous membrane are both epithelial surfaces so that some diseases affect both.

**Primary herpetic stomatitis**

Herpetic infection is the commonest acute stomatitis in children and is caused by the virus herpes simplex type 1. It begins with vesicles distributed singly or in clumps over the oral mucous membrane. The vesicles are normally 3-4 mm in diameter, dome-shaped and circular; as they rupture they ulcerate to form lesions with a red edge. At this stage they are painful. The patient is often pyrexial and does not feel well, but usually within a week the lesions are fading. Direct smears from early lesions confirm the diagnosis, but this is usually made on the clinical appearance. Antiviral agents (idoxuridine and acyclovir) may be used locally on the lesions but this is difficult with small children.

**Herpes labialis**

Recurrent herpetic infections after primary herpes is thought to affect 30% of those initially infected. The recurrence usually affects the lip. These so-called 'cold sores' are common after a cold, local infection, emotional upsets, heat and various unknown factors. The virus is thought to be dormant in the ganglion of the trigeminal nerve. The lesions are usually preceded by a prickly sensation, then a cluster of vesicles appear which may discharge exudate. The vesicle ruptures and then crusts over. Treatment should be started as soon as the prickly sensation begins and idoxuridine 1% or acyclovir should be applied. Although they do not always abort the lesions, the cycle of events does not appear to be so severe.

**Hand, foot and mouth disease**

This viral infection is common among school children. It presents with ulceration of the mucosa and a vesicular rash on the extremities. It has no connection with the disease of similar name in cattle. It is caused by A strains of the Coxsackie virus, has an incubation period of between 3 and 10 days and is highly contagious. The small scattered ulcers in the mouth are not very painful and the rash often affects the hands and feet. No treatment is required and there is often a mini-epidemic in a school when every child has the disease.

**Herpes zoster**

The infection caused by the varicella-zoster virus usually affects the skin of the face in the distribution of the trigeminal nerve in adults. When it affects the mucous membrane the patient experiences pain before the rash appears and thinks that he has a toothache. The
vesicles are present in the mouth but there is a sharp demarcation line in the midline; they may coalesce within the mouth and form a large ulcerative area. A cervical lymphadenopathy is frequent. The treatment of the intraoral lesions is difficult but analgesics help until the infection resolves.

**Candida**

Acute and chronic candidiasis is common in the mouth and nowadays a systemic Candida infection is often seen in those patients undergoing treatment for leukaemia or malignancy with powerful drugs. This is a 'disease of the diseased'.

The Gram-positive hyphae of Candida are a normal commensal in about 40% of mouths, the most common strain being *Candida albicans*.

The most usual infection is acute oral candidiasis, commonly referred to as thrush. It presents in the mouth as soft, creamy-yellow patches which form on the surface of the oral mucous membrane. It is common in the elderly and the newborn and those on antibiotic therapy, particularly the tetracyclines. If the plaques are rubbed off a red area of mucosa remains, with the antibiotic form the whole mucosa is red, oedematous and sore, the *Candida albicans* not being immediately obvious. Nystatin pastilles or amphotericin B lozenges are both used to combat this infection.

In chronic candidiasis, persistent white plaques, which on initial observation are indistinguishable from leukoplakia, form on the tongue. The plaques are thick layers of parakeratotic epithelium invaded by hyphae of Candida, with chronic inflammation beneath. These chronic lesions cannot be wiped off and their most common site is at the angle of the mouth; the infection is often associated with angular stomatitis. Scrapings of the lesions submitted for histology confirm the diagnosis.

The management of chronic candidiasis is difficult. In those patients with diseases of cell-mediated immunity or Addison's disease, the underlying condition should be corrected first; in those with chronic mucocutaneous candidiasis the most recent drug to show any improvement in the condition is miconazole.

Angular stomatitis is a cracking of the lips at the commissure. Although it is often associated with iron deficiency anaemia, it is a common manifestation of oral candidiasis. It presents in older people and is the result of the general lack of tone in the facial muscles, allowing the skin of the face to sag causing folds at the corners of the mouth. There is a great temptation to keep these cracks moist by touching with the tongue. Both bacterial and Candida infections thrive in these cracks. The condition may be aggravated by grossly overclosed dentures; some patients wear the same dentures for 30 years, they wear down a great deal and this may add to the problem. The lesions are treated locally with antifungal cream and any disease within the mouth is also treated. The bacterial element must be treated too, with antibiotic creams.
**Denture stomatitis**

This very common condition affects those patients who wear their dentures for long periods. It is characterized by an area of bright uniform erythema exactly corresponding to the upper denture-bearing area of the mucous membrane. It is not seen under lower dentures as these are not so tightly fitting and saliva may wash under them. Occasionally the condition is associated with iron deficiency anaemia and is rarely a reaction to the material of which the dentures are made. The treatment is to encourage the patient to remove the dentures as much as possible, to keep the dentures clean, to take antifungal pastilles or lozenges when the dentures are out of the mouth, and to practice good oral hygiene after the infection has settled.

**The acute specific fevers**

Chicken-pox, and previously smallpox, cause a skin rash as well as intraoral vesicles. In the prodromal stage of measles tiny white spots on a bright red base, called Koplik's spots, are seen. Ulceration of the oral mucosa may also be seen in glandular fever.

**Benign oral ulceration**

There are three main types of this common condition: minor aphthae, which probably affect about 10% of the population, herpetiform aphthae, and the major aphthae which are uncommon.

**Minor aphthae (Mikulicz's aphthae)**

Small painful shallow ulcers appear at more or less regular intervals in the mouth. Their underlying cause is not known but several have been suggested.

No viral or bacterial infection has been isolated, but the ulcers in the mouth may be secondarily infected by bacteria. In women it has been noticed that the aphthous ulcers disappear during pregnancy. Oestrogens have been tried but no real progress in treatment has been made.

Several immunological abnormalities have been reported in patients with aphthous ulcers but the findings are not consistent. It has been shown that patients with these ulcers have higher titres of circulating antibodies, mainly IgM, to crude extracts of fetal oral mucosa. In other reports circulating lymphocytes sensitized to fetal oral mucosa have been found and it is postulated that the antigen responsible is in or on the epithelial surface. It may also represent a cross reaction between an antigen of an oral microorganism and an antigen of oral epithelium. The sequence of cellular changes described in biopsies of aphthous ulcers is regarded as characteristic of a delayed hypersensitivity reaction (type IV). The ulcers to not respond well to immunosuppressive drugs.

About 10% of patients with these ulcers have iron, folic acid or vitamin B12 deficiency. Of the three, folic acid deficiency seems the only one that clears the ulcers up for good when it is corrected.
Most patients who suffer from aphthous ulcers come from the clerical, semiprofessional and professional groups of people and it is well observed that periods of stress are associated with exacerbations of the ulcers. The disease also appears to be more common in non-smokers, and is particularly apparent in people when they first give up smoking. Aphthous ulcers are uncommon in those who wear full dentures or in manual workers. The ulcers are more common in women. Because there is often a considerable period of time between each attack it is often difficult to assess the benefits of treatment. It is unusual for the lesions to affect the tongue and they are usually located on the non-keratinized areas of the oral mucosa. The ulcers are usually about 2-3 mm in diameter with a yellow floor and a sharply defined red margin; they also have a crater appearance.

**Herpetiform ulcers**

These ulcers are characterized by a very large number of small ulcers which are less sharply defined than the minor aphthae. The ulcers may be confluent and there is a widespread erythema of the mucous membrane. Compared with the similarly named herpes simplex ulcers there are no preceding vesicles and the ulcers are much less regular in size and shape than the simplex ulcers. These ulcers are not caused by the herpes virus and the term herpetiform describes the appearance of the lesion. The condition is uncommon.

**Major aphthae**

These ulcers are usually several centimeters in diameter, persist for many months, and scarring may follow their resolution. They involve the masticatory mucosa. Because of their appearance they may be confused with carcinoma: a biopsy may be the only way to confirm the diagnosis. These aphthae are not indurated like malignant ulcers.

**Management of benign oral ulceration**

The management of these ulcerative conditions is difficult: nothing has been found to cure them although many drugs have been tried. Those patients with blood deficiencies must be treated effectively, but for most the treatment is empirical.

Corticosteroids in different forms give relief: they can be taken topically as hydrocortisone hemisuccinate 2.5 mg pellets or as a topical paste in the form of triamcinolone in orabase, which sticks to the lesion. Although not encouraged, the continual use of intraoral steroids does not give rise to the side-effects of systemic steroids.

Mouthwashes containing tetracycline, chlorhexidine 0.2%, or choline salicylate have all been tried. They relieve symptoms in some people probably because of their anti-inflammatory effect.

Diazepam has been tried, to reduce the anxiety of the patients with recurring ulcers, but the results are poor.

Sodium cromoglycate as a topical agent theoretically cuts down the release of histamine from the ulcers, reducing the amount of pain. It has helped some patients but the incidence of the ulcers is not reduced.
Levamisole enhances both cellular and humoral immune responses. For reasons not understood it reduces the number, frequency and severity of attacks of aphthous ulcers, although it has little effect on major aphthae.

Despite the large number and different types of treatment available the ideal medicine has yet to be found.

**Miscellaneous oral lesions**

**Tuberculous ulceration**

When advanced pulmonary tuberculosis was more common, ulcers were seen on the tongue, but they are now rare. The ulcer has a stellate shape with overhanging edges and watery granulations in its floor. It may be mistaken for a malignancy, because it may be indurated. Biopsy confirms the diagnosis. The oral lesions heal with systemic treatment.

**Syphilitic ulceration**

Oral manifestations of syphilis may be seen in the three phases of the disease. Although not common, the initial infection may arise from oral sexual contact and a primary chancre may then be found on the lips or within the mouth. The chancre begins as a firm nodules 3-4 weeks after infection and when the surface breaks down a few days later a rounded ulcer with raised indurated edges remains. The ulcer is characterized by being painless, although often there are enlarged lymph nodes in the neck. The chancre eventually heals and the diagnosis of syphilis may be made by demonstrating *Treponema pallidum* in material from the chancre because serological tests are not positive at this stage.

Secondary syphilis develops a few months after the primary infection, and in this phase the patient may experience constitutional upset with a mild febrile illness, headache and sore throat. There may be a skin rash of pinkish macules and the oral lesions, which particularly affect the tongue and palate, take the form of flat ulceration covered by a greyish membrane.

These ulcers may lie all over the palate and are described as snail track ulcers. They may coalesce to form well-defined rounded areas described as mucous patches. The diagnosis may be confirmed at this stage by direct examination for spirochaetes, and by serology.

Tertiary syphilis usually appears after 3 years or considerably longer. The characteristic lesion is the gumma, affecting any part of the skin or oral mucous membrane and starting as a swelling which undergoes necrosis leaving a deep ulcer. The ulcer is rounded with soft punched out edges, its floor is pale and resembles wash leather. Although the lesion may heal, it may cause severe scarring of the oral tissues.

In congenital syphilis, painful fissures may also develop at the mucocutaneous junctions and these result in characteristic radiating scars at the angles of the mouth. Saddle nose, frontal bossing and interstitial keratitis may also be seen in this condition. Abnormal teeth have already been described.
**Behçet's syndrome**

Behçet's syndrome consists of anterior uveitis and genital and oral ulcers. The oral ulcerations are usually very similar in appearance to the major aphthae and their treatment is the same.

Associated with the condition are erythema nodosum, erythema multiforme, non-suppurative arthritis, and neurological involvement in 20% of patients.

**Lichen planus**

This condition is very common, particularly in women beyond middle age; its cause remains a mystery. Lymphocytic infiltration and the so-called liquefaction degeneration of the basal layer suggest an immunological cause of a cell-mediated type but experimental immunological tests have been inconclusive. Some studies have shown a correlation with diabetes and rheumatoid arthritis and, like aphthous ulceration, there is a known link with anxiety.

The lesions are characteristic both in their appearance and distribution. Three types are commonly seen: the strial, atrophic and erosive types. In the strial type the striae show hyperkeratosis, parakeratosis, starry and annular patterns.

The atrophic lesions are often combined with the strial type. The epithelium is severely thinned and red, but in the erosive type there are shallow irregular areas of total destruction of the mucous membrane. This type heals by fibrosis. In all types the underlying rete pegs tend to be pointed and saw-toothed and there may be degeneration and liquefaction with beads of fluid accumulating along the basement membrane at the junction between the epithelium and connective tissue. There is also infiltration by chronic inflammation.

In the mouth the lesions are usually symmetrical, commonest on the buccal mucosa and tongue, but uncommon on the floor of the mouth. The symptoms vary: in some patients it is an incidental finding, others complain of roughness of the mouth. The atrophic and erosive types are more painful and sometimes eating is difficult.

Patients with lichen planus of the skin rarely have oral disease as well, although the disease is the same. Lichen planus, particularly the atrophic type, commonly affects the gingiva. Because of the discomfort on cleaning the teeth the gingiva becomes more inflamed, gingivitis ensues setting up a vicious circle. If there is any doubt about the diagnosis particularly with the erosive type, then a biopsy should be taken. This disease is often difficult to differentiate from leukoplakia. The erosive variety has a poorly defined relationship with the development of oral cancer.

The treatment is by pellets of beclomethasone valerate three times a day. The results are good. In severe cases potent steroid preparations may be necessary.
**Pemphigus vulgaris**

Pemphigus vulgaris is rare and is fatal if untreated. It is characterized by vesicles or bullae on the skin and mucous membranes. Of all the oral diseases pemphigus vulgaris has the most compelling evidence that it is caused by immunological failure. The two main findings to support this are, first, a raised titre of antibodies (predominantly of the IgG class) to the intercellular substance of the epithelium, and second, antibodies that can be demonstrated by fluorescence in the intercellular area of the epithelium.

Histology shows that the epithelial cells lose their attachment to each other, a phenomenon called acantholysis. These intraepithelial changes begin just above the basal layer appearing as clefts within the epithelium and as this split widens vesicles and bullae form. The epithelial cells which lose their attachments become rounded in shape and the cytoplasm contracts around the nucleus. These cells are characteristic of pemphigus vulgaris.

The disease may affect either sex, and the mouth lesions often precede the skin lesions. Nikolsky's sign is diagnostic: stroking the mucous membrane induces a vesicle or bulla to appear.

The disease may be mild or severe and in severe cases there is widespread ulceration in the mouth and of the skin. After rupture of the vesicles the underlying erosions are painful. In severe cases protein, fluid and electrolytes are lost causing a systemic upset.

Immunosuppressive therapy has made this disease less life threatening: corticosteroids and azathioprine have both been used successfully, but treatment must be continued for life. When the disease is at its worst the patient cannot eat or drink, and food and fluid must be given by nasogastric tube.

**Mucous membrane pemphigoid**

Although it has a very similar name this disease must be separated from pemphigus vulgaris. It may be serious, but it is not fulminating like pemphigus vulgaris. Bullae and erosions of the mucous membrane are found but the skin is not usually affected.

As a vesicle forms there is a loss of attachment of the epithelium to the connective tissue. The epithelium after separation remains intact, and unlike pemphigus vulgaris there is no acantholysis. Whereas in pemphigus vulgaris the separation is intraepithelial, in mucous membrane pemphigoid the individual cells do not separate from each other. Although an immunological cause is suspected the evidence is not as strong as in pemphigus vulgaris.

The disease affects mainly women, the mouth being the commonest site; it also affects the eyes, larynx, pharynx and oesophagus. After the vesicles have become erosions the membrane heals with a certain amount of scarring. It may be difficult to distinguish this disease from pemphigus vulgaris as in certain cases Nikolsky's sign is positive. A biopsy may be needed to confirm the diagnosis. The most serious sequela of this disease is affection of the eye for the scarring may cause blindness.
Fibrosis and then stenosis of the larynx and pharynx have been reported. In most cases treatment is of limited value but if the eyes are affected systemic corticosteroids may have to be used.

**Acute erythema multiforme (Stevens-Johnson syndrome)**

This disease generally affects young people. It usually has a prodromal period with fever and constitutional disturbances, and the patient may be acutely ill. The oral lesions include split, crusted and bleeding lips and widespread erythema and erosions within the mouth. The eyes and the skin are often affected, the skin lesions being red macules with a bluish cyanotic centre, and these lesions may become bullous in nature. The disease tends to recur every few months for 2 or 3 years.

The cause of the disease is still being studied: reaction to drugs, particularly the sulphonamides and barbiturates, has been implicated. It is also thought that the disease is a sequel to herpes and mycoplasmal pneumonia. Histologically the oral lesions are characterized by widespread necrosis with eosinophilic colloid change in the superficial epithelium, and an inflammatory response in the cells of the dermis.

**Fordyce's spots**

Small creamy spots representing small sebaceous glands are very common and seem to increase with age. They have no significance.

**The effects of drugs on the oral mucosa**

**Local effects**

Patients commonly treat toothache with aspirin, but instead of swallowing it they place it beside the affected tooth, causing local damage with superficial necrosis of the mucosa. The subsequent damage resembles leukoplakia. Other tablets have a similar effect.

Antibiotic mouthwashes, especially tetracycline, alter the local oral flora and a Candida infection may supervene 2 or 3 days after antibiotic therapy begins.

**Systemic effects**

**Bone marrow depression**

Several drugs suppress white cell production, the most severe form being agranulocytosis. In the latter form the gingiva and pharyngeal mucosa undergo necrotizing ulceration. The drugs which have this potential are the antibacterials, analgesics (amidopyrine), phenothiazines and the anti-thyroid agents. The resultant leucopenia has the same oral presentation as in leukaemia described later in the chapter.

Red cell production may be reduced, this being a known side-effect of phenytoin leading to anaemia and, in some cases, to aphthous ulceration and Behçet's syndrome. If folic
acid deficiency is found to be the underlying cause it should be corrected and the drugs stopped. The ulcers then heal.

**Drugs affecting the immune system**

Patients taking steroids in large doses are prone to infections, herpes and Candida infections in the mouth are common in such patients. These oral manifestations are common in patients having transplant operations.

**Specific effects**

Some drugs give specific reactions. Gold, sometimes used in the treatment of rheumatoid arthritis, may cause oral lesions resembling lichen planus. Sulphonamides and barbiturates are held responsible for erythema multiforme. Phenolphthalein drug derivatives commonly used in purgatives may cause sharply circumscribed skin lesions, the lesions appearing every time the drug is used. Arsenical, mercurial and gold-containing drugs may give rise to a serious systemic reaction with exfoliative stomatitis and dermatitis. The epithelium of the skin and the oral mucous membrane are lost and, if not corrected, the disease may be lethal. Similar reactions have been reported to the barbiturates and phenylbutazone.

Phenytoin is used commonly in the treatment of epilepsy. A well-known side-effect is progressive fibrous hyperplasia of the gingiva. Metals such as mercury, bismuth and lead may cause pigmentation of the gingiva, usually affecting the gingival sulcus, a result of the formation of the metal sulphides and bacterial products. The most commonly seen nowadays, though rarely, is the blue line along the gingiva caused by lead. Topical antibiotics may give the tongue a dark appearance due to an overgrowth of pigment-forming bacteria.

**Oral manifestation of systemic disease**

**Haematological disorders**

**Anaemia**

In a patient presenting with a history of a sore tongue, anaemia must be excluded as an underlying cause. Deficiency states characterized by low serum levels of iron, folic acid or vitamin B₁₂ but without actual anaemia may be found in patients complaining of sore tongue.

The mechanism by which these deficiencies affect the oral mucosa and lingual tissues is not understood, but these factors have an important trophic effect in the oral cavity, as shown by the gross atrophic changes of the tongue seen in anaemia. Iron, folic acid, and vitamin B₁₂ are essential for the nutrition of the tongue mucosa, and even though the haemoglobin levels remain normal, deficiency of some of these other factors shows in the mucosa of the tongue. Thus glossitis may be characterized by virtually complete atrophy of the filiform papillae of the dorsum of the tongue. A deficiency must be investigated further.
Acute leukaemia

The oral symptoms in many cases of this disease may be quite dramatic. The gingiva becomes very swollen and ulcerated, and painful. The gingiva may also bleed as a result of both the infection and the associated purpura. The gingiva are infiltrated with leukaemic cells. Because of the associated anaemia the oral mucosa may appear pale.

Chronic leukaemia

The oral mucosa is not often affected.

Agranulocytosis

This is a very uncommon condition with symptoms similar to acute leukaemia.

Thrombocytopenia purpura

Sudden bleeding from the gingiva may be the first sign of this disease which is characterized by a low platelet count, normal clotting and an extended bleeding time. Evidence of bleeding may be seen at the posterior border of an upper denture where it presses on the palate.

Haemophilia and Christmas disease

Although bleeding may be experienced in the mouth after the shedding of deciduous teeth, the diagnosis has usually been made previously. But this disease must be suspected with persistent haemorrhage after tooth extraction.

Endocrine disorders

Pituitary hyperfunction

Acromegaly and gigantism usually have obvious presenting features but a change in the occlusion of the teeth may indicate the onset of acromegaly. The condylar growth centre of the mandible is affected so that eventually the lower jaw protrudes. A comfortable eating position is not easy to find as the teeth change position. The patient may also complain that his dentures no longer fit.

Pituitary hypofunction

The only effect in the mouth is that the teeth erupt slowly and late.

Hypothyroidism

Eruption of the teeth is delayed and the tongue is large in cretinism.
Hyperparathyroidism

This is usually caused by a tumour of the parathyroid glands. Areas of decalcification cause osteitis fibrosa cystica, that is cystic areas of bone resorption.

Hypoparathyroidism

In this rare condition of hypoparathyroidism in children, the disease is present during the development of the teeth and there may be aplasia or hypoplasia of the enamel.

Addison's disease (adrenal hypofunction)

The main clinical features of this disease are lassitude, anorexia, loss of weight and low blood pressure. Pigmentation, either brown or black, is distributed over the gingiva, buccal mucosa and the lips, and it may be an early sign of the disease.

Diabetes

Although these patients are more prone to infections than normal, periodontal disease is common only in severe uncontrolled diabetes.

Acquired immune deficiency syndrome (AIDS)

Although this disease was only recognized in 1981, infection with human immunodeficiency virus (HIV), formerly known as human T-cell lymphotrophic virus type III (HTLV-III), is increasing at an alarming rate and over the next few years patients will present outside specialist centres. The oral cavity may be an initial indicator of the disease and when examining the mouth this should be considered (Croser and Farthing, 1986).

Suspicion should be raised if the patient, particularly a young male, has evidence of oral thrush, angular cheilitis, gingivitis, a scaly red dermatitis or persistent aphthous ulceration. These symptoms, evidence of possible immunosuppression, are called an AIDS-related complex (ARC). To meet the definition of AIDS the patient either must have a Kaposi's sarcoma or have had a life-threatening opportunistic infection such as pneumocystis. Hairy leukoplakia, particularly on the side of the tongue, is very suggestive of AIDS infection. The only totally safe way to treat such patients is to assume that all blood and saliva are infected. The Government guidelines define appropriate precautions.

Vitamin deficiencies

Riboflavin (vitamin B₂ deficiency)

Angular stomatitis consisting of red, painful fissures at the angles of the mouth and shiny redness of the mucous membranes are characteristic. Although not common, it is seen in patients with the malabsorption syndrome.
Nicotinic acid deficiency

This disease, commonly known as pellagra, is seen in the UK in alcoholics. Stomatitis and glossitis are presenting features as well as defects of the skin and gastrointestinal tract. The lip, lateral and gingival margins of the tongue become red, swollen and ulcerated.

Vitamin B\textsubscript{12} deficiency

Pernicious anaemia, which is a result of lack of absorption of vitamin B\textsubscript{12}, is characterized by a tongue that is red, painful and smooth caused by atrophy of the papillae. The oral symptoms may be the first sign of the disease.

Folic acid deficiency

The oral symptoms resemble those of pernicious anaemia but the disease is also associated with severe aphthous ulceration as seen in Behçet's syndrome.

Vitamin C deficiency

Commonly known as scurvy this disease is rarely seen nowadays. The main features of the disease are dermatitis and purpura but the gingiva may also swell and bleed.

Vitamin D deficiency

Rickets, although not common in the UK, is still seen in other parts of the world. Although the development of the teeth is not hindered, the structure of both the dentine and enamel is altered causing grooving or pitting of the enamel. The zone of uncalcified pre-dentine is altered making the teeth weaker.

Miscellaneous conditions

Rheumatoid arthritis

Rheumatoid arthritis may be associated with Sjögren's syndrome and a persistently dry mouth.

Rare conditions

Wegener's granulomatosis may produce gingival changes; polyarteritis nodosa may produce oral lesions; and sarcoidosis and Crohn's disease have been found in the mouth.

Effects of radiotherapy on the mouth

Irradiation is one of the main treatments of malignancy of the mouth and neck and it has well recognized effects on the oral tissues. Both cobalt and neutron therapy affect the tissues of the mouth directly and indirectly. The direct effects are reduction of mitosis of the normal cells: some cells undergo total degeneration and then die. The indirect effects are the
result of local damage to the blood vessels, causing thrombosis of the vessels and ischaemia of the oral tissues which therefore take a long time to heal.

The oral mucosa initially shows an inflammatory reaction with a developing area of erythema. The cells which die produce a yellowish-white membrane over the surface of the mouth. If the damage is more severe the full layers of the epithelium is lost producing raw painful areas. The oral discomfort is further increased because saliva is quantitatively and qualitatively reduced making the mouth dry and uncomfortable. Direct effects of radiation make the teeth brittle, and combined with relative xerostomia, hygiene is compromised and rampant caries may develop. It is therefore of prime importance that the mouth and teeth be kept as clean as possible during radiotherapy.

**Ranula**

A ranula is a uni- or multilocular, mucus-filled cyst in the floor of the mouth. It is classified into two varieties according to its lining and extent:

1. a 'simple' ranula has an epithelial lining and is confined to the floor of the mouth, in contrast to
2. a 'plunging' ranula or 'burrowing' ranula which is lined by connective tissue and not epithelium. Although it may be confined to the floor of the mouth, it can extend through the mylohyoid muscle into the neck.

These cysts are considered to arise from the sublingual salivary gland, although several alternative theories have been proposed, such as development from embryonic epithelial rests, a submucosal bursa, or a form of dermoid cyst. The plunging variety results from extravasation of cyst contents into the tissues.

There is no age limit, but they are generally seen in children and young adults. Typically, the appearance is of a bluish, translucent cyst on one side of the frenulum. This appearance has led to the name 'ranula' because of the likeness to the belly of a small frog (Latin rana, meaning a frog). They can burst and recur, and may reach a large size before attention is sought. Examination should include bimanual palpation, in particular, looking for extension into the neck.

The treatment of a ranula is surgery. Marsupialization is often used, and several operations are needed in many cases. Although marsupialization may be effective for a simple ranula, it will invariably fail if used alone for the plunging variety, because the remaining sublingual gland continues to secrete. However, differentiation of a simple from a plunging ranula can be difficult clinically. The following treatment plan has therefore been proposed (Black and Croft, 1982):

1. a ranula confined to the mouth should be excised (the sublingual gland is often included in the excised tissue)
(2) a ranula extending into the neck should be marsupialized, and the sublingual gland excised. More extensive surgery is not only unnecessary, but carries the risk of damaging the submandibular duct and lingual nerve.

**Disorders of the temporomandibular joint**

The temporomandibular joint lies between the condyle of the mandible and the articular surface of the temporal bone. The capsule of the joint is attached below to the neck of the condyle and above to the margins of the articular area of the temporal bone, extending to the anterior edge of the articular eminence in front and to the squamotympanic fissure behind. The articular disc or meniscus is a dense sheet of fibrous tissue dividing the joint cavity into a large upper and smaller lower compartment. The lateral pterygoid muscle is inserted partly into the front of the disc and partly into the fossa on the front surface of the neck of the condyle below the articular surface. Therefore, when the muscle contracts the disc and condyle are pulled forwards. The commonest disorders of the temporomandibular joint are limitation of movement, pain and clicking sounds.

Trismus means limitation of movement: this feature may be temporary or long term. Temporary trismus may be the result of a bone infection caused by either a buried or an erupting wisdom tooth giving rise to pericoronitis. Following visits to the dentist an injection of local anaesthetic directed at the inferior dental nerve may be the cause of local inflammation near the joint and a fracture of the condyle near the joint also causes trismus. Although acute trismus is not common, tetanus should always be excluded as a cause.

Permanent limitation of movement of the temporomandibular joints may be caused by extra- or intra-articular causes. Fibrosis may occur around the temporomandibular joints during radiotherapy and in the more severe forms may result in complete ankylosis. Although not common in the UK but common in India, oral submucous fibrosis results in the formation of a dense collagenous connective tissue of the buccal and palatal mucous membranes causing thick, hard mucosa and later difficulty in opening the mouth.

Intra-articular fracture followed by bleeding and acute pyogenic arthritis may permanently limit movement of the joints. Rheumatoid arthritis may affect the temporomandibular joints, with crepitus and some limitation of movement, and the extent of the disease is related to the severity of the general illness. Pain does not seem to be a major feature, but X-rays may show erosions of the bone and flattening of the joint surfaces. Treatment is as for the generalized disease.

Pain around the joint may be the result of infection or temporal arteritis but the most common cause is given the name of temporomandibular pain dysfunction syndrome.

This condition, which is commoner in women than in men, consists of pain, clicking of the joints and limitation of opening of the mouth. The onset of the disease is gradual, although some patients report that the symptoms start after yawning. The pain is usually related to one side, is not severe but is worse on eating. The patients may complain of otalgia but point to the pain being first in front or just behind the temporomandibular joint. The joint is heard to click when the mouth is opened and closed and the patient finds it difficult to open the mouth fully.
Abnormalities of occlusion are said to be a common cause of this condition but have not been proven. Missing posterior teeth cause overclosure and a forward posture of the lower jaw. This causes joint strain during mastication. Bruxism of the teeth may also strain the temporomandibular joint and its surrounding musculature. These factors, although not proven, may give rise to a neuromuscular incoordination causing spasms or areas of fatigue in the muscles and subsequent pain.

X-rays are of limited help but at least they can exclude organic disease. It is also important to check that the teeth themselves are sound and that it is not toothache, and the ear should also be examined to exclude otalgia. On palpation of the masseter muscle between the fingers the patient will complain of pain and one can feel the tenseness of the muscle.

The management of these cases is often difficult, but the majority of the symptoms will settle with conservative measures in this self-limiting condition. If an occlusal cause is suspected, detailed occlusal reconstruction may be carried out, but a more practical approach is the fitting of an acrylic overlay appliance which allows free occlusion without cuspal interference. These appliances although simple, will replace missing posterior teeth, correct a severe overbite and often stop grinding of the teeth. Although these appliances may be regarded initially as cumbersome, difficulties of speech and eating are soon overcome and they usually provide an effective treatment. The spasm associated with the masseter muscle may be eased by heat treatment to the area.

The temporomandibular joint pain dysfunction syndrome is a disease of the young. If an elderly patient presents with similar symptoms then the diagnosis of temporal arteritis, osteoarthritis or rheumatoid arthritis should be considered. Irradiation for malignancies of the head and neck may result in fibrosis of the structures surrounding the joints and may result in difficulty in opening or even complete ankylosis. It may require surgery to release the joint or even create a false joint. Trauma to the jaw may result in meniscal tears which can be demonstrated by arthrography.

Although a variety of operations have been described for this disease, including injections of steroid, condylotomy, condylectomy and arthroplasty, they are now regarded as a last resort and conservative treatment is tried for as long as possible.

Cysts of the jaw

Cysts are common in the jaws, arising from proliferation of epithelial rests left from the developing teeth. Cysts may be described as odontogenic or non-odontogenic depending on whether they are in tooth-bearing regions of the jaws. Most cysts have an epithelial lining with the exception of the solitary bone cysts.

The commonest cysts of the jaws, accounting for about 68% of these lesions, are periodontal cysts. They are rarely seen before the age of 10 years and are commoner in men than women. They affect the maxilla three times more often than the mandible and develop as a result of irritation originating at the apex of an infected root canal associated with chronic inflammatory periapical changes. The cyst wall consists of collagenous fibrous connective tissue. The fluid present is usually watery and opalescent and it may contain cholesterol crystals which give it a shimmering appearance. As they grow they give rise to
slowly progressive painless swellings, and there are no symptoms until the cyst becomes large enough to be conspicuous. When the overlying bone thickness has become thinned a crackling sensation may be felt on pressure with the fingers over the cyst.

If a dead tooth, which has been a cause of the cyst, has been extracted for some reason, the lesion that is left is called a residual cyst. X-rays will confirm a rounded, clearly radiolucent area with a sharply defined outline, the only difficulty in outlining the cyst is in the region of the maxillary antrum and the demarcation between the cyst and the antral wall may be difficult.

Dentigerous cysts surround the crown of a tooth and are attached to the neck of the tooth. They account for about 15% of the cysts of the jaws. These cysts are common in the first 10 years of life and usually cause no symptoms. X-rays confirm that the crown of a tooth is usually not in its normal erupting position. It lies in a rounded and unilocular cavity and the cyst is surrounded by sclerotic bone.

Eruption cysts, as the name implies, may involve the deciduous or permanent molar teeth and the cysts lie superficially in the gingivae and appear as soft, rounded, bluish swellings.

Primordial cysts or odontogenic keratocysts which are uncommon, account for about 5% of jaw cysts and usually present in the second and third decade in men. These cysts arise from the remains of the dental lamina or the enamel organ and the cyst is characterized by the growth of finger-like processes along the lines of least resistance and, therefore, the cysts may be of considerable size before they become clinically apparent. X-rays show a radiolucent, multiloculated, area with a scalloped margin; this may cause confusion with ameloblastoma. The bony wall is sclerotic.

Nasopalatine cysts, although uncommon, form in the midline of the anterior part of the maxilla and are thought to arise from the epithelium of the nasopalatine ducts in the incisive canal. They are slow growing and give rise to a salty taste in the mouth when they discharge. Nasolabial cysts are rarely seen.

The treatment for all these cysts is enucleation, but with very large cysts it may be necessary to marsupialize the cyst so that the jaw is not weakened enough to fracture. Marsupialization allows the formation of new bone and when this has formed the cyst may be enucleated. Although ideally all involved teeth should be extracted, this may not be acceptable, particularly when anterior teeth are involved, and therefore conservative management of the teeth may be carried out with root filling and apicectomy. It is uncommon for cysts to recur, except primordial cysts, of which up to 60% recur; this is common daughter cyst formation, thus it is important that these particular cases are followed-up long term.

Solitary bone cysts are characterized by not having an epithelial lining. The cause of these cysts is unknown, although trauma is often thought to be the origin. The cyst cavity has a rough bony wall and the lining may be of thin connective tissue. On X-ray, these lesions are rounded, unilocular radiolucent areas which may arch upwards between the roots of the teeth. They usually present in teenagers, are frequently symptomless and are more common
in females. Attempted removal will find a cavity containing no fluid and as a result of the bleeding so incurred, the cyst cavity will heal up.

Cysts of the soft tissues of the mouth are commonly seen. The most common are called mucous retention cysts which originate in the minor salivary glands of the lip, and the ranula which arises from the sublingual salivary gland in the floor of the mouth. A sublingual dermoid cyst is seen in a similar position to a ranula but the cyst is filled with desquamated keratin and it has a semi-solid feel to it. They are also seen between the hyoid and the mandible and are symptomless until difficulty is experienced with eating and drinking. All these cysts are treated by enucleation, but the mucous retention cysts of the lips have a habit of recurring, probably because of damage to adjacent mucous glands.

Tumours of the jaw

Apart from the tumours normally found in bone in other parts of the body, both benign and malignant, the maxilla and mandible contain odontogenic tissues from which tumours may develop.

Fibromata of the jaws are uncommon and may be either endosteal or subperiosteal and they grow slowly often with no symptoms until they are conspicuous by their size or are picked up by routine X-ray. Treatment is by removal and the histology is confirmed by a mass of fibroblasts and collagen fibres. An ossifying fibroma is sometimes seen in children and may present as facial deformity and histologically the calcified masses closely resemble cementum of the tooth.

Chondromata and osteomata of the jaws are uncommon but exostoses of the hard palate, called torus palatinus, and of the mandible, called torus mandibularis, are seen frequently. These exostoses consist of lamellae or compact bone. The torus palatinus is found at the posterior end of the hard palate and is usually apparent when dentures are first fitted, and the torus mandibularis is found on the lingula side of the mandible in the region of the mental foramen. Both these exostoses are removed easily with either a drill or chisel.

Giant cell granuloma of the jaw is uncommon. It arises in young people and mainly affects the mandible. Histologically the lesion forms a lobulated mass which consists of proliferating connective tissue in which there are rounded foci of giant cells. Although the presenting complaint is usually a swelling in the mandible, because of its rapid growth it may present as paraesthesia of the mental nerve. Biopsy is the only way to confirm the diagnosis, for its X-ray appearance of loculation and erosion makes it difficult to differentiate from malignancy. It also has a similar appearance to the effects of hyperparathyroidism; a blood test will confirm normal calcium levels.

Odontogenic tumours arise from either epithelial or connective tissue of the mandible. The most common epithelial tumour is the ameloblastoma, this is a benign tumour which is locally invasive. The tumour originates in odontogenic epithelium, is slow growing and invades cancellous bone. Histologically, the tumour consists of islands of epithelial cells in a connective tissue stroma. The outer layer of epithelial cells has an ameloblast appearance with basally placed nuclei. Cyst formation is common within the ameloblastoma. The X-ray appearance is very similar to that of a multilocular cyst although there may be a honeycomb
pattern of radiolucency. These tumours are more common in middle-aged individuals and, in 80% of cases, are found in the region of the mandibular ramus as expanding swellings, which will destroy much bone and may even present as pathological fractures of the mandible. Biopsy will confirm diagnosis. Complete removal of the tumour achieves a cure but if any of the tumour is left then it will recur. Very large tumours may need extensive surgery with bone grafting if large amounts of the mandible are removed.

Adenomatoid odontogenic tumour is a benign tumour which affects young people and has a well-defined capsule and contains whorls and strands of epithelial cells. Some of the cells look very similar to ameloblasts and give rise to its previous name of adenoameloblastoma, but it has no real relationship to ameloblastoma and simple enucleation is the treatment of choice.

Calcifying epithelial odontogenic tumours are very rare but histologically very interesting in that they consist of epithelial cells in a connective tissue stroma, but they have homogeneous hyaline areas within the tumours which look like amyloid. There are also areas of calcification in which tooth-like structures may be seen. Biopsy is required to differentiate from malignancy and treatment is by excision.

Cementomata or cemental dysplasia are lesions characterized by continued proliferation of cementum. Usually four named types are known - benign cementoblastoma, cementifying fibroma, peripheral cemental dysplasia, and gigantiform cementoma. These lesions are characterized by their radiopaque nature on X-ray and rarely result in expansion of the bone. All can be readily removed and recurrence is not a problem.

Myxomata of the jaws are seen in young people and have a characteristic soap bubble appearance on X-ray. They are slow-growing tumours and consist of spindle-shaped cells scantily distributed in a loose mucoid intercellular material and clinically there is a fusiform swelling of the jaw. Like ameloblastoma wide excision is necessary as they tend to recur.

Other benign masses presenting in the jaw may be odontomas and these are hamartomata. They appear as a radiopaque structure on X-ray and, usually, like teeth, when fully developed grow no further and have no symptoms.

A variety of odontomas are recognized based on their development. The commonest are enameloma, dentinoma and cementinoma, which develop as their names imply from the various tissues of the teeth.

More complicated odontomas called germinated, compound composite and complex composite are reported, which are varieties of distorted teeth. The germinated odontoma is only slightly abnormal and the complex composite odontoma is made up of multiple small simplified teeth called denticles.

**Malignant tumours of the jaws**

Primary malignant tumours of the jaws are uncommon. The jaws are involved in 4% of cases of osteogenic sarcoma. It tends to occur in slightly older patients than those with long bone disease and in the mandible or maxilla of the elderly as a complication of Paget's
disease (Garrington et al, 1967). Histologically the tumours contain osteoblasts, fibroblasts and cartilage cells, but osteoid is also present. The cellular activity is very irregular. Characteristically a fast growing swelling in the mouth is accompanied by severe pain in some instances. X-ray show patchy irregular bone formation and resorption, and biopsy confirms the diagnosis. These tumours have a high metastatic potential and so the prognosis from any treatment is poor, but is better than long bone disease.

Non-Hodgkin's lymphoma, sarcomata and chondrosarcomata of the mandible have been reported very rarely. Multiple myeloma is rarely diagnosed from jaw involvement, but commonly the characteristic lytic areas are found on a total body scan for evaluation of the extent of the disease. Burkitt's lymphoma most frequently affects the jaws of African children. It is characterized by a proliferation of undifferentiated stem cells which cause destruction and swelling of bone and loosening and movement of teeth. The Epstein-Barr virus is well known as a possible aetiological factor. Treatment with methotrexate has had some success.

The mandible and maxilla are uncommon sites for secondary carcinoma but should be considered particularly in patients with carcinoma of the bronchus, thyroid and kidney. The patients usually present with pain and, if the inferior alveolar nerve is involved, paraesthesia may be present. The previous medical history of the patient will give the clue as to whether the disease is primary or secondary although a primary tumour is rarely found. X-rays show an area of bone resorption with a hazy outline. Treatment may be hormonal or by radiotherapy: surgery is not often recommended.

**Osteodystrophies of the jaws**

Rickets, hyperparathyroidism and scurvy may cause metabolic bone disease but in all three, mandibular or maxillary involvement is uncommon. Rickets, although not seen in the UK very often, does not cause soft teeth as is sometimes thought, and those children affected do not have a higher incidence of caries. However, the teeth may be slow to erupt. The gingival inflammation of scurvy is well known but the deformation of collagen and osteoid matrix does not cause any bony lesion.

The jaws are a common site for the cyst-like lesions found in hyperparathyroidism, although the renal symptoms usually bring the patient to the doctor. X-rays of the jaws show generalized bone resorption and a loss of density giving a ground glass appearance. A blood test will often confirm the diagnosis, and histology distinguishes a giant cell granuloma from an ameloblastoma. Removal of the parathyroid adenoma is the primary treatment.

Osteogenesis imperfecta is an hereditary disorder transmitted as an autosomal dominant trait in which bones are poorly formed and fragile. Blue sclerae, deafness and dentiginous imperfections are associated with the disease. Dental extractions must be carried out with great care.

Achondroplasia is a failure of normal proliferation of cartilage in the epiphyses and the base of the skull. It causes the characteristic face of the dwarf: the middle third of the face is retrusive and the profile concave. Disparity in the growth of the jaws causes a severe dental malocclusion.
Cleidocranial dysplasia is characterized by a deformed clavicle and retrusion of the mandible. It is also a well-known cause of the delayed eruption of teeth.

Paget’s disease of bone, more commonly seen in the elderly, may affect the mandible and maxilla. The alveolus of the jaws are symmetrically and grossly enlarged as a result of the continual resorption and deposition of the bony tissue in a rapid, irregular and exaggerated fashion. Gross hypercementosis of the tooth roots is also seen and the first indication may be difficulty in extracting a tooth. On X-ray the bones have a cotton wool appearance. Osteogenic sarcoma is a well-known but rare complication of this disease.

Monostotic fibrous dysplasia may present as a painless swelling in the mandible or maxilla. The disease is usually self-limiting and is thus probably better left alone, apart from confirming the diagnosis, unless the swelling becomes very large and unsightly. The lesions of polyostotic fibrous dysplasia may be multiple and are accompanied by pale pigmentation of the skin; like the monostotic type, the disease is self-limiting when associated with endocrinopathies.

Cherubism (Albright's syndrome) is a disease inherited as an autosomal dominant trait and is not now regarded as a fibrous dysplasia. The lesions are highly cellular with many giant cells. As the lesions resolve new bone is laid down at the angle of the lower jaws and also in the region of the tuberosities of the upper jaws. This new bone causes the fullness of the cheeks. The maxillary swelling causes the eyes to turn upwards producing the characteristic cherubic appearance. Like the other two types there is a tendency to natural regression, although surgery may be necessary for severe abnormalities. In fibrous dysplasia and cherubism blood chemistry is normal.

**Tumours of the mouth**

**Benign tumours of the mouth**

Although not true tumours, polyps, epulides and granulomata are common swellings within the mouth, usually the result of chronic inflammatory hyperplasia caused by injury or infection. The lesion begins as a nodule covered by epithelium; the original granulation tissue then becomes replaced by collagenous fibrous tissue. A fibrous epulis or denture granuloma are the different names for a similar histological reaction. A fibrous epulis is usually seen protruding between the teeth on the buccal or labial aspect of the gingival margin whereas a denture granuloma lies near the edge of a denture which is irritating the mucosa. These lesions may be easily excised and any known cause should be removed.

A giant cell epulis is seen during the period when the deciduous teeth are being shed. The swelling is rounded, soft and purplish in colour, may grow rapidly and bleed easily. Histologically the epulis consists of multinucleated cells in a vascular stroma of plump spindle-shaped cells, and the lesions may be easily excised. It may be difficult to differentiated these lesions from an endosteal giant cell granuloma which has eroded through bone and appears as a broad-based flattened nodule. Radiographs show evidence of the underlying bony destruction in the case of the granuloma.
Benign epithelial lesions

A squamous cell papilloma presents as a warty white or pink swelling with finger-like processes of stratified squamous epithelium supported by a core of vascular connective tissue. It is treated by removal.

Pleomorphic adenoma

These tumours are usually well circumscribed or encapsulated and over 60% of the intraoral adenomata arise from the hard palate. They can usually be excised by a large defect in the palate may require flap repair or obturation.

Benign connective tissue tumours

Fibromata are commonly seen on the buccal mucosa, are slow growing, sessile or pedunculated and may be soft or hard. Neurofibromata are rarely found in the mouth but when present appear as a soft lobular swelling. Lipomata are uncommon in the mouth. Various types of haemangiomata may be seen in the mouth: the capillary haemangioma consists of a mass of fine capillary vessels in a connective tissue stroma, and the cavernous haemangioma consists of large, thin-walled blood sinuses separated by septa of connective tissues and the mass resembles a blood-filled sponge. These lesions are often associated with similar skin lesions. Small or moderate-sized haemangiomata may be removed simply by surgery or cryosurgery. The treatment of large lesions is very difficult: injection of sclerosing agents may be helpful. Lymphangioma may affect the tongue and result in a very large tongue; there may also be small lesions of the buccal mucosa. Small lesions may be easily removed, but large lesions of the tongue are difficult to treat.

Premalignant lesions of the mouth

Under normal circumstances healthy oral epithelial cells progress towards the surface and desquamate without appreciable keratin accumulation. Thickening of the superficial mucosal layer (stratum corneum) is called hyperkeratosis; increase of the proportion of nucleated cells near the surface is called parakeratosis, and accentuation of the basal layer (stratum granulosum) with elongation of the rete pegs into the submucosa is called acanthosis. All of these changes are common but harmless.

A white patch in the mouth is commonly called leukoplakia. Such a patch can persist and a few become malignant. The term 'premalignant' is not really appropriate for it is only in retrospect when a carcinoma is present that the leukoplakia has been premalignant. There is no doubt about the potential for malignant change in leukoplakia, but the difficulty lies in estimating the risk. In one large series of white patches in the mouth only 2.4% became malignant in the first 10 years and 5% in 20 years (Banoczy and Sugar, 1972). This rate is 50 times the risk of malignant change in the normal mouth. In another large series of biopsies taken from areas of leukoplakia, the diagnosis was carcinoma \textit{in situ} in 2% and invasive squamous cell carcinoma in 8%. Leukoplakia of the anterior floor of the mouth should be regarded with suspicion as the incidence of malignant change is in the region of 20-25%. Red lesions are called erythroplasia, a disease with a greater risk of malignant change than leukoplakia.
The basis of the potential of these lesions to change their nature is related to the degree of dyskeratosis of the tissue. Although the interpretation of biopsies varies widely between pathologists, dyskeratosis causes disordered proliferation, maturation and organization of the epithelium and may be present in small or large lesions. The features which may be present are nuclear hyperchromatism, nuclear pleomorphism, mitosis, loss of polarity, deep cell keratinization and loss of intercellular adherence. In cases where there is little disorganization of the epithelium it is called mild atypia, but in true dyskeratosis or severe atypia the epithelium is conspicuously disorganized with irregular-shaped nuclei scattered throughout the thickness of the epithelium.

The lesions of erythroplasia are red and sharply defined and the texture of the lesion is very different from the surrounding mucosa. Most of these lesions when biopsied are carcinoma in situ or carcinoma.

**Candidal leukoplakia**

Speckled leukoplakia are white lesions on an atrophic erythematous base and are thought to be a combination of leukoplakia, erythroplasia and candidal infection. The potential for malignant change lies somewhere between that for leukoplakia and erythroplasia.

If the dyskeratosis is described as moderate or severe, the lesions must be observed closely over a long period of time and any change in nature should be confirmed by biopsy. It is not practical to remove large areas of leukoplakia, particularly in the elderly in whom these patches are common and which are unlikely to undergo malignant change in the patient's lifetime.

Carcinoma in situ is the worst type of dyskeratosis: here the abnormalities extend through the thickness of the epithelium. All the cellular abnormalities characteristic of malignancy may be present but the underlying connective tissue is not invaded. The disease is not often seen in the mouth but should be treated as a true carcinoma.

**Carcinoma of the lip**

The lips form the upper and lower anterior walls of the oral cavity and are composed of the mucosa stretching from the mucocutaneous junction to the line of contact of the opposing lips, including the commissures. They may be divided into upper and lower lips and right and left commissures.

The 600 cases a year reported in the UK are predominantly in men, with a male:female ratio of 8:1. Keller (1970) confirmed an association with farming and outdoor occupations as well as confirming a higher incidence in tobacco users. The lower lip was more vulnerable to the effects of the sun either because of the angle at which the ultraviolet rays strikes the vermilion surface or because the lower lip was larger. Carcinoma of the lip is twice as common in Israel as in the UK. Farmers in Texas and Australia have a particularly high incidence and the risk of acquiring the carcinoma doubles for every 250 miles nearer the equator the patient lives (Szipak, Sonte and Frenkel, 1977).
Squamous cell carcinoma is often preceded or accompanied by a white or reddish plaque, called leukoplakia and erythroplasia respectively. Generally carcinomata remain localized for extended periods, and tend to grow slowly. The tumour can invade the deeper layers of the lip, adjacent skin and oral sites, and bone, with extension down the mental nerve.

The UICC classification is shown in Table 4.1.

**Table 4.1 UICC classification of lip cancer**

*TNM pre-treatment clinical classification*

**T - primary tumour**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tis</td>
<td>Pre-invasive carcinoma (carcinoma <em>in situ</em>)</td>
</tr>
<tr>
<td>T0</td>
<td>No evidence of primary tumour</td>
</tr>
<tr>
<td>T1</td>
<td>Tumour limited to the lip: 2 cm or less in its greatest dimension</td>
</tr>
<tr>
<td>T2</td>
<td>Tumour limited to the lip: more than 2 cm but not more than 4 cm in its greatest dimension</td>
</tr>
<tr>
<td>T3</td>
<td>Tumour limited to the lip: more than 4 cm in its greatest dimension</td>
</tr>
<tr>
<td>T4</td>
<td>Tumour extending beyond lip to neighbouring structures, eg, bone, tongue, skin of neck, etc</td>
</tr>
<tr>
<td>Tx</td>
<td>The minimum requirements to assess the primary tumour can not be met.</td>
</tr>
</tbody>
</table>

The lymphatic drainage from the lower lip anastomoses across the midline and therefore a lesion of the lower lip may spread to nodes on both sides or the opposite side of the neck.

Removal of the tumour is the accepted treatment, particularly if the tumour has spread to the mandible or lymph nodes. Leukoplakia or erythroplakia should be watched carefully.

A lesion smaller than 1 cm can usually be removed with adequate clearance by a modified V-shaped incision followed by primary closure. There is usually enough elasticity in the tissues of the lip to prevent compromise of function or aesthetics. Because the mucosa of the lip is usually widely damaged by actinic cheilitis a lip shave is usually performed in addition. The Abbe-Estlander flap is useful for reconstructing the lip after removal of the carcinoma, allowing up to two-thirds of one lip to be resected. Modifications of this flap operation are described for upper and commissure lesions of the lips. Removal of larger lesions may require reconstruction by a cheek or tongue flap. Radiotherapy may be used for the infirm and for very extensive lesions, but because of early detection most may be removed surgically. For early lesions the prognosis is excellent with either surgery or radiotherapy.

**Cancer of the oral cavity**

Over 90% of all malignancies of the oral cavity are squamous cell carcinomata, the remainder include tumours of the minor salivary glands, sarcomata, lymphomata and melanomata.
In the UK and the USA malignant tumours of the oral cavity account for less than 2% of all malignancies, whereas in India they account for more than 40% (Mahboubi, 1977). On average in the UK almost 1500 cases of intraoral cancers and about 600 cases of cancer of the lip are seen in one year.

**Squamous cell carcinoma of the oral cavity**

The mouth is divided anatomically into the lip, the buccal or cheek mucosa, the gingivae, the hard palate, the oral tongue and the floor of the mouth. The floor of the mouth is the crescent-shaped area between the lower gingiva and the under surface of the tongue. There is also an area called the retromolar trigone which is the small area of mucosa attached to the anterior surface of the ascending ramus of the mandible posteriorly, running forward to the upper and lower molar teeth including the edge of the maxillary tuberosity.

**Aetiology**

Over 95% of patients with squamous cell carcinoma are over the age of 40 years and, in this age group, one in 20,000 of the general population will be affected, whereas the chances of acquiring the disease over the age of 75 are one in 1100.

Smokers are six times more likely to develop intraoral cancer than non-smokers (Silverman and Griffiths, 1972; Wynder and Stellman, 1977). In the USA, it has been shown that 40% of those who develop a squamous cell carcinoma of the mouth and who continue to smoke after treatment will develop a second head and neck malignancy, while those who give up smoking have only a 6% chance of a second primary tumour (Moore, 1971). These results have not been confirmed in the UK where there has been no apparent increase in the incidence of oral carcinoma over the last few years. Fewer men and more women are now affected, and the large increase in smoking over the last 20 years does not correlate with the falling or static incidence of the disease.

The decline in pipe smoking has mirrored the decline in mouth cancer. Reverse smoking in India, Sardinia, Venezuela and Panama is associated with a high incidence of hard palate malignancy (Reddy, 1974), and the habit of holding raw tobacco against the mucous membrane in one area of the mouth is also associated with a high incidence of the verrucous variant (Stecker, Devine and Harrison, 1964).

In India and Southeast Asia the high incidence of oral cancer is related to the habit of chewing betel nut, which is a mixture of the nut, tobacco and slaked lime wrapped in betel leaves. This mixture is extremely cheap and available at every street corner. Its interesting and refreshing taste particularly after eating, makes it practically addictive. It is taken in India by whole cross sections of society, being packaged very much like peanuts in the UK.

In the USA the relation between squamous cell carcinoma of the mouth and high alcohol consumption has been known for some years. Whether this is because of a direct effect or whether it reflects an underlying nutritional deficiency is not known (Wynder, 1971; Lowenfels, 1974). In the UK the figures are not so convincing and, as with tobacco consumption, there has been no corresponding increase in oral cancer with the increasing alcohol consumption in recent years.
The aetiology of oral cancer is still little understood, but dietary and metabolic factors have been studied. There is an increased incidence in patients with cirrhosis of the liver, particularly in France. This may be because of the lack of the vitamin riboflavin which may result in a degenerative mucosa. There is a higher incidence of disease in Scandinavia in women and related to the Plummer Vinson syndrome (Wynder et al, 1957; Wynder and Stellman, 1977), and a higher incidence is also reported in textile workers exposed to wool fibres.

Although theoretically poor oral hygiene, mechanical irritation by sharp teeth or dentures would be thought to be implicated, there is no evidence to prove this. The decline of oral cancer over the years may be attributed to less pipe, smoking, improvement in oral care and the effective treatment of syphilis.

Pathology

The typical squamous cell carcinoma is an obvious ulcerated lesion with a greyish rough base. The tumour may protrude above the surface (exophytic) or infiltrate deeply with minimal projection above the surface (endophytic).

Histologically there is invasion of the deeper tissues of epithelial cells with cellular abnormalities, including pleomorphism and intense nuclear staining. Tumour cells invade deeper tissues including muscle, glands, nerves and eventually bone. Low grade tumours (well-differentiated) are those with minimal pleomorphism and few mitoses, and high grade (poorly differentiated) tumours have much cellular and nuclear pleomorphism and negligible keratinization. This differentiation is important in prognosis (Arthur and Farr, 1972). Verrucous carcinoma refers to certain exophytic tumours that have a papillary and micronodular appearance and tend to spread laterally. Some regard these tumours as the most well-differentiated squamous cell carcinomata.

The lateral border of the tongue is the commonest site inside the mouth. Over 70% of intraoral tumours lie in the lower half of the mouth. Because symptoms may be minimal or absent when an oral cancer is small, diagnosis and treatment are often delayed until the cancer is large or cervical lymph node metastasis is apparent.

Clinical features

Bruun (1976) at the Royal Dental Hospital in Aarhus, Denmark, found that the average delay from the onset of symptoms in a patient with a malignant lesion in the oral cavity to actual consultation with a general practitioner was 4.9 months. He also showed that a further 5.6 months elapsed after this consultation before actual therapy began.

General dental and medical practitioners may only see one or two intraoral malignancies in their careers; 70% of carcinomata had not been suspected of being malignant at the patient's first visit to their practitioner. Not all the blame is to be laid on the practitioners as the patients are often late in presenting with their complaints. Education of patients, medical and dental practitioners might produce early diagnosis of these tumours.
Lesions of the lip, anterior cheek, floor of the mouth and gingivae tend to spread to the submandibular lymph nodes; carcinomata of the tongue and retromolar trigone spread to the upper digastric nodes. The presence of lymph node metastases at diagnosis halves the survival changes of the patient. Distant metastases are uncommon in cancer of the oral cavity, but some have put the distant metastases rate at post-mortem as high as 47% (Merino, Lindberg and Fletcher, 1977).

Reporting of survival is based on accurate staging of the disease. The problem with staging is that there is a large subjective element, particularly in the assessment of oral cancer and it is often forgotten that these tumours are three dimensional. The TNM classification of the UICC is shown in Table 4.2 (Manual for Staging of Cancer, 1977). The N status is the same for other head and neck tumours and is shown in Table 4.3.

Table 4.2 TNM classification for oral carcinoma

TNM pre-treatment clinical classification

<table>
<thead>
<tr>
<th>Tis</th>
<th>Pre-invasive carcinoma (carcinoma in situ)</th>
</tr>
</thead>
<tbody>
<tr>
<td>T0</td>
<td>No evidence of primary tumour</td>
</tr>
<tr>
<td>T1</td>
<td>Tumour 2 cm or less in its greatest diameter</td>
</tr>
<tr>
<td>T2</td>
<td>Tumour more than 2 cm but not more than 4 cm in its greatest dimension</td>
</tr>
<tr>
<td>T3</td>
<td>Tumour more than 4 cm in its greatest dimension</td>
</tr>
<tr>
<td>T4</td>
<td>Tumour with extension to bone, muscle, skin, antrum, neck, etc</td>
</tr>
<tr>
<td>Tx</td>
<td>The minimum requirements to assess the primary tumour can not be met.</td>
</tr>
</tbody>
</table>

Survival curves show that the highest mortality is in the first 2 years after diagnosis. The actuarial survival for carcinoma of the lip for 5 years is 77% and that for the tongue 26%. Over 10% of patients with oral cancer develop a second primary tumour.

Table 4.3 N status - TNM classification

N - regional lymph nodes

<table>
<thead>
<tr>
<th>N0</th>
<th>No evidence of regional lymph node involvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>N1</td>
<td>Evidence of involvement of movable homolateral regional lymph nodes</td>
</tr>
<tr>
<td>N2</td>
<td>Evidence of involvement of movable contralateral or bilateral regional lymph nodes</td>
</tr>
<tr>
<td>N3</td>
<td>Evidence of involvement of fixed regional lymph nodes</td>
</tr>
<tr>
<td>Nx</td>
<td>The minimum requirements to assess the regional lymph nodes can not be met.</td>
</tr>
</tbody>
</table>

Features of specific sites

Carcinoma of the tongue

Most squamous cell carcinomata arise from the lateral borders or central aspect of the tongue, origin from the dorsum is uncommon and midline lesions are rare. Fewer than 1% of patients are less than the age of 30 years, although cases in adolescence have been reported (Byers, 1975). Usually the lesion starts as a small white or red patch and the only symptom
the patient experiences is some discomfort with spicy foods. As the ulcer forms it is firm with raised edges, has an inflamed granular floor which bleeds easily and the base is indurated. The lesion is usually larger on palpation than thought on observation. In larger lesions the ulcer spreads into the adjacent floor of the mouth and mandible and may be so extensive that the exact site of origin cannot be ascertained. Unfortunately most lesions are already T2 in size at diagnosis. It is often only when eating and swallowing become difficult that the patient seeks help.

The lymphatic network of the tongue is well developed and, classically, lymph drainage from the tip of the tongue is to the submental nodes and then to the jugular omohyoid group, and from the side of the tongue to the submandibular nodes and then the jugulodigastric nodes.

It is important to palpate both sides of the neck as tumour may spread by an abnormal route to the other side. Nearly 30% of patients at presentation have evidence of neck involvement.

Carcinoma of the floor of the mouth

These are tumours of the sixth and seventh decades, and over the last 10 years the male:female ratio has dropped (Fu, Lichter and Galante, 1976). As the carcinoma grows, ulceration becomes more prominent with lateral spread, and the tongue and mandible are invaded. If the tumour is not freely mobile the periosteum of the mandible is invaded until proven otherwise and an orthopantomogram may give an indication of such invasion. Pain indicates deeper infiltration. At presentation more than half the lesions are classified T3 and T4 and fewer than 10% are T1.

Carcinoma of the alveolar ridge

Over 80% of these lesions affect the lower alveolus and the first symptoms are a poorly fitting denture. As for the floor of the mouth fewer than 20% of the lesions are T1 in size at diagnosis (Cady and Catlin, 1969). The important point in pathology of tumours arising from or extending to the lower alveolus is invasion of the mandible. The tumour can erode directly into bone, frequently through the healed extraction sockets, or may enter the inferior dental canal through the mental foramen. The foramen lies close to the mucosal surface in edentulous patients whose alveolar ridge has atrophied. Once in the canal the tumour spreads along the inferior alveolar nerve as far back as the base of the skull. Invasion of the lower cortical strut of bone is uncommon.

The 5-year survival rate for T1 lesions is over 78% but for T4 lesions is less than 15% (MacComb and Fletcher, 1967).

Carcinoma of the buccal mucosa

This is the lesion common in betel nut chewers in India. Small lesions are completely asymptomatic but if the lesion is posterior, trismus may be the presenting feature caused by invasion of the pterygoid muscles (Paymaster, 1956). Over 40% of these patients present with
lymph node enlargement (Conley and Sacoyanna, 1973). There is a 90% cure rate for T1 lesions and only 15% for T4 lesions (MacComb and Fletcher, 1967).

**Carcinoma of the hard palate**

Over 80% of these tumours present in patients over the age of 70 years, affecting men and women equally. They are common in those parts of the world where reverse smoking is practised (Reddy, 1974) and particularly in India. One in five reverse smokers has either a premalignant or malignant condition.

**Carcinoma of the retromolar trigone**

The trigone is a very small area so that at presentation most of these carcinomata have already spread to the surrounding areas of the soft palate, tonsil, buccal mucosa and tongue. These tumours are aggressive and over half have neck nodes at diagnosis.

**Clinical diagnosis of oral malignant conditions**

The initial evaluation of a patient with a suspected malignant oral condition is a comprehensive history, directed to the duration of the symptoms and their nature. The history must include questions about the social habits, particularly smoking and drinking, and also any relevant occupation or hobbies. It is important that any previous exposure to chemical or radioactive irritants be noted.

The patient's medical history must be carefully recorded, especially any previous syphilitic, severe cardiac, nervous or respiratory problems. Surgery or medication in the past for an illness may limit the use of one of the treatment modalities available. Prior treatment for cancer should be reviewed in great detail and if radiotherapy has been used, the exact dosage and treatment fields must be available for treatment planning. The history should indicate the sort of support that will be available to the patient after treatment for this could equally influence the treatment.

The physical examination of the patient should include not only the oral lesion and the neck, but also the cardiopulmonary, renal, gastrointestinal, hepatic and metabolic systems. It is important at the beginning of the clinical evaluation to examine the dental and gingival status of the mouth, an examination which is often totally neglected.

The superficial structures of the oral cavity, lip and tongue are amenable to inspection and palpation: a biopsy of the mouth or lip may be easily taken without general anaesthesia, and the histological diagnosis may be made very early in the assessment of the patient. The biopsy usually confirms the high suspicion of malignancy on examination.

The patient's general health must be assessed and any evidence of malignant spread to other sites in the body or the possibility of a second primary tumour must be sought.

The haemoglobin, white cell count, erythrocyte sedimentation rate, electrolytes, urea, total protein, albumin and glucose levels in the blood are recorded. Urine analyses should also be carried out. Anteroposterior and lateral X-rays of the chest are important to exclude
secondary disease or a primary malignant condition. Orthopantomograms of the mandible will show any evidence of bony invasion. Roughly half of the patients with clinical involvement of the lower alveolus by an intraoral carcinoma show radiological evidence of bone involvement. This is usually an irregular invasive bony defect, so-called 'moth-eaten appearance', but a smooth pressure erosion is also common. 'Floating teeth', displaced bony fragments, pathological fractures, and rarely an expanded inferior canal are less common radiological signs. The floor of the mouth is often difficult to evaluate directly and computerized tomography will show the extent of submucosal infiltration, although teeth, if present, distort the image. Computerized tomography is also useful in determining response of the tumour to chemotherapy and radiotherapy during treatment.

The place of magnetic resonance in diagnostic imaging has not yet been determined and its full potential has yet to be realized. Xeroradiography and ultrasound have little place in intraoral lesions. Radionucleotide scanning of bone and liver may be used to exclude metastasis.

An examination under anaesthetic to demonstrate the extent of disease is useful and a full panendoscopy of the upper respiratory and alimentary tracts including bronchoscopy can be carried out to search for a second tumour. It is also useful while the patient is anaesthetized to examine the neck again.

Most lesions are either squamous cell carcinomata or of minor salivary gland origin. If the biopsy confirms a rare malignant melanoma, the prognosis is very poor and surgery is the only treatment that might control the disease. Lymphomata of the oral cavity are very uncommon: they must be typed accurately, and further investigations of the lymphatic system and bone marrow carried out. Treatment depends on the exact histology and extent of the disease: chemotherapy or radiotherapy or a combination of both are the mainstays of management.

**Treatment of mouth cancer**

The selection for treatment for squamous cell carcinoma of the mouth must be based on a number of factors in addition to the extent of the disease. General health, nutritional status, smoking, drinking habits and the patient's mental and psychological make up must also be considered in deciding individual treatment modalities. The prognosis of advanced oral carcinoma is universally very poor: furthermore mutilation and loss of function from either surgery or radiotherapy can be conspicuous.

**Irradiation**

The radiotherapeutic methods for treating squamous cell carcinomata are telecobalt units or low megavoltage linear accelerators. Most squamous cell carcinomata are radiosensitive and exophytic lesions are more radiosensitive than erosive lesions. Bone and muscle invasion adversely affects the radio responsiveness of the tumours and therefore subsequently decreases the radiocurability (Chu and Fletcher, 1973).

T1 and T2 lesions in all sites within the mouth respond equally to surgery and radiotherapy, the latter usually accomplished with a dose of about 6,500 cGy given in
fractions over 4-5 weeks. Lesions of the lateral border of the tongue and anterior floor of the mouth can be successfully treated with interstitial implants to deliver a dose of up to 10,000 cGy. A combination of external beam and implants may be used. If nodes in one side of the neck are enlarged, surgery should be considered as the first line of treatment.

Radical radiotherapy can be used for T3 and T4 lesions: some are radiosensitive but the overall survival rates are poor. Preoperative or postoperative radiotherapy has been advocated in an effort to increase survival. Preoperatively radiotherapy is advocated to reduce tumour size and to limit seeding and embolic spread of tumour cells, but this has not been demonstrated in practice. Roscuit et al (1972) using a preoperative radiation dose of 4,000 cGy showed no increased survival in the large T4 tumours. In 1986 Robertson et al presented a series of patients given postoperative radiotherapy 6 weeks after removal of T3 and T4 tumours and showed a reduction of recurrence at 18 months from 50% to 18% compared to a previous series. Snow et al (1980) found no difference in recurrence rates at 18 months between preoperative, postoperative or radical radiotherapy.

The use of neutron therapy with the cyclotron is still controversial. Initial results from both the USA and the UK were encouraging; recurrence of disease at the primary site was reduced, but the survival of the patients was not increased. Further trials have shown that the survival rates for fast neutrons are no better than those for photons.

**Surgery**

The standard procedures for mouth cancer are a glossectomy (partial or total) and a resection of the mandible with the adjacent floor of the mouth, called a commando procedure by the Americans or, more elegantly, a pelvimandibulectomy by the French.

**Hemiglossectomy**

Before any operation on the tongue, the use that the patient makes of his voice must be considered. If he depends on his voice for his livelihood then attempts must be made to leave a good tip to the tongue and to supply a sulcus so that he can wear teeth, either immediately or later.

The proposed lines of clearance of the tumour inside the mouth are tattooed, since it is easy to become disoriented while removing the primary tumour from the tongue if there are not marks to follow.

If the patient has enlarged cervical nodes a standard radical neck dissection is performed and left attached at the submandibular region. When the radical neck dissection is completed it may be thought best to split the lip to improve access. It is quite possible to do this operation without splitting the lip, which is preferable since it prevents oedema of the lip and a scar. The skin is elevated off the mandible and bleeding from the mental foramen prevented by first isolating and dividing the vessels which pass through the foramen. The mucosa in the gingivobuccal sulcus is cut thereby opening the mouth.

Excision of the primary tumour, if it is large, may require a mandibulotomy.
The primary tumour is removed by holding the tip of the tongue in a towel clip and pulling it forward. Cutting diathermy is used to remove the tumour. It will be necessary to stop and tie the lingual artery. A margin of at least 3 cm is needed and the margin must be in depth as well as width.

**Operations for tumours of the floor of the mouth and alveolus**

Tumours of this area are dealt with in a very similar manner by the commando operation or pelvimandibulectomy.

Under general anaesthesia, a temporary tracheostomy is inserted. If the patient has an enlarged node in the neck a radical neck dissection is carried out and left with its pedicle on the submandibular area.

The superior cervical flap is raised well up over the mandible, with or without splitting the lower lip. The oral mucosa is divided in the gutter lateral to the mandible well away from the tumour.

It is often possible to preserve the lower border of the mandible thus preserving bony continuity (marginal resection) which is superior to division of the mandible and grafting.

If the tumour affects the alveolar ridge the alveolus is removed in the appropriate area using a Stryker saw or a fissure burr. The segment of mandible is left in continuity with the primary tumour which is removed with a wide margin, including part of the tongue if necessary.

After removal of a small tumour (less than 2 cm in diameter) it is often possible either to close the defect primarily or to repair it with a split skin graft.

The quilting technique is particularly useful. Its principles are as follows. A split skin graft must be immobilized and fluid, mainly serum and blood, must be prevented from collecting between it and its bed. This can best be achieved in the mouth by stitching the graft with numerous black silk sutures about 1 cm apart, and making small nicks in the graft between these stitches.

Resection of the vast majority of tumours produces a large defect. While it is technically possible to close the defect in the mouth by suturing the remnant of the tongue to the cheek, this results in a very crippled tongue, particularly in two circumstances: in resection of a carcinoma of the lateral border of the tongue; or in resection of tumours involving the anterior part of the floor of the mouth. In the first case loss of the greater part of the bulk of the tongue leads to severe crippling if the small remnant is sewn down, and in the second instance, if the tongue is sewn to the internal surface of the lower lip to close the defect of the anterior part of the floor of the mouth, the tip of the tongue is immobilized leading to unsightly swellings and subsequent difficulty in articulation of dentolabial consonants. This result can be improved by a later epithelial inlay, but for the last 20 years or so it has been generally agreed that the defects remaining in the mouth after a partial glossectomy or resection of a tumour involving the anterior part of the floor of the mouth (a
pelvimandibulectomy) require reconstruction of the soft tissue defect, with local or distant flaps. The principles of reconstruction are as follows:

1. the reconstructive techniques should neither interfere with nor limit the excisional surgery
2. form and function should be quickly restored
3. the morbidity and mortality should not be increased by the reconstructive phase
4. a secondary cosmetic deformity should not be produced
5. the reconstructive phase should be completed as quickly and as simply as possible, especially if a cure is doubtful
6. prolonged reconstructive procedures should not ordinarily be carried out in patients for whom a prosthesis would provide satisfactory rehabilitation. Numerous different types of flap are available for achieving those ends.

**Local mucosal flaps**

The main local flap for reconstruction within the mouth is the lingual flap. It requires that all or virtually all of the tongue has been preserved. The tongue flap is outlined by marking a line lengthwise on the tongue, of about 20-40% of the width of the tongue to include the lingual artery in its base. The lingual flap is then rotated into the defect of the floor of the mouth or of the oropharynx. The edges of the anterior one-third or free portion of the tongue are then closed, primarily avoiding producing a pointed tip of the tongue. The raw edges of the remaining part of the donor site are usually covered by a split skin graft to prevent the mobile portion of the tongue adhering to the flap. This flap has a high success rate as a result of the very rich vascular supply to the tongue from the lingual artery; there is unfortunately little cross-over across the midline except for one small artery which crosses the midline at the tip of the tongue and on which thin bipedicled flaps can occasionally be raised.

**Regional random flaps**

The following regional random flaps have been described for the repair of defects within the mouth: nasolabial flaps, lateral cervical flaps and median cervical flaps based superiorly.

The nasolabial flap is particularly useful for reconstruction of the anterior part of the floor of the mouth because simple closure of the remaining part of the tongue to the lip binds the tongue, makes speech and the fitting of a denture difficult and causes salivary incompetence. At the end of the resection, inferiorly based flaps are raised from the nasolabial fold. A tunnel is then developed beneath the mucosa of the cheek and the lingual alveolar sulcus so that the flap can be turned downwards to be passed through the tunnel into the floor of the mouth. The point at which the flap appears in the mouth is marked by methylene blue, the flap is taken out again and the epidermis is removed with an electric dermatome. The
flaps are then re-introduced into the mouth and sewn in place one behind the other, the one placed anteriorly being sewn to the edge of the incision in the lip and the posterior one being sewn to the edge of the defect in the tongue. If the pedicle is denuded in this way the flaps can be regarded as 'subcutaneous pedicle flaps' and the defect can be closed primarily so that it is not necessary to divide the pedicle and return it. Alternatively a small orocutaneous fistula can be left as originally described, 3 weeks later the flap is divided, the pedicle returned and the fistula closed. The defect in the nasolabial area is easily closed by primary suture and the scar rapidly becomes inconspicuous since it is placed in a skin crease. Indeed, the enthusiasts for this operation claim that the patient derives a bonus since he gets a face-lift at the same time!

Lateral cervical flaps and the median cervical (apron) flap are now rarely used.

Distant axial flaps

Two axial flaps have proved extremely useful for reconstruction of large defects in the mouth and the oropharynx: the temporal flap and the deltopectoral flap. The temporal flap is a true axial flap based on the superficial temporal vessels. The flap is raised at the end of the excisional phase, turned into the mouth through the zygomatic foramen, and is sewn into the intraoral defect. Three weeks later the temporary fistula is closed by division or return of the pedicle. Although only just over half of the forehead is required to resurface most deficiencies within the mouth, the cosmetic defect on the forehead is less if the entire forehead skin is removed and replaced by a split skin graft laid directly on the pericranium which must be carefully preserved. This flap can be used for reconstructing any defect in the mouth or oropharynx. It is very successful, rarely undergoing necrosis, and has enjoyed great popularity. Facial palsy is a potential hazard but is rare. The external salivary fistula is not a real complication since it is placed high in the mouth. A persisting external fistula after the second stage is uncommon. The skin of the flaps is difficult to keep clean at first but it settles rapidly, softens and then behaves like a free skin graft within the mouth. The only real disadvantage is the very obvious cosmetic defect on the forehead (McGregor, 1963).

The deltopectoral flap may be used in several ways, but in the original description it was passed upwards beneath the cervical skin, the distal end was sewn into the defect in the mouth with its skin surface facing into the pharynx, and the rest of the flap was formed into a tube, skin surface inwards, running beneath the cervical skin, although it is now more usual to create an external skin tube. At a second stage, 3 weeks later, the pedicle was divided and the orocutaneous fistula closed (Bakamjian, Long and Rigg, 1971). The deltopectoral flap and the temporal flap were until recently the most successful soft tissue replacements available. The temporal flap, however, leaves an unsightly appearance to the forehead of the patient. The process of insertion and dividing of these flaps takes time and patients are in hospital for some time.

Over the last few years axial flaps have been largely superseded by myocutaneous flaps particularly the pectoralis major flap.

The myocutaneous flap, as with any other, may be lost through technical error or vascular compromise. If technical errors are avoided, the security, speed of development, and success rate of undelayed myocutaneous flaps surpasses any direct cutaneous or microvascular
flap. The donor site of a myocutaneous flap is closed primarily if possible, or covered by a split skin graft from the thigh 5 days later.

**Pectoralis major myocutaneous flap**

The pectoralis major myocutaneous flap (PM flap) is now the 'workhorse' of most oral cavity and oropharyngeal reconstruction. The primary blood supply is from the pectoral branch of the acromiothoracic artery.

When developing the flap, the incisions must be so placed that they will not compromise a deltopectoral flap if it should be needed later. The technique of elevation of the flap is described in Volume 1.

The defect is closed primarily with suction drainage in most circumstances. When the skin of the chest wall overlying most of the pectoralis muscle is used for reconstruction, the defect is usually closed by a skin graft.

The skin paddle can be orientated to the defect not only in contour but also in its relation to the muscle. For the oropharynx, the skin is usually orientated in the long axis of the muscle, whereas in the floor of the mouth it is orientated in the horizontal axis. Between the horizontal and vertical axis, the surgeon can orientate the paddle of skin obliquely. Various designs for conformation to tongue, floor or mouth, and retromolar trigone defects can easily be accomplished without jeopardizing vascularity.

Intraoral and extraoral defects can be lined by splitting the flap and folding in upon itself. The skin paddle is made equal to the area of the two defects.

Sternomastoid and trapezius myocutaneous flaps are now little used.

**The free vascular forearm flaps**

The free vascular forearm flap is an axial pattern free flap based on the radial artery or one or two of the forearm veins (basilic or cephalic vein or one of the interconnecting branches). The blood vessels are large. The radial artery measures 2 or 3 mm in diameter and the veins measure 3-5 mm. Each has a long pedicle and consistent anatomy.

The hand and fingers derive their blood supply mainly from the ulnar artery by means of the deep palmar arch. Allen's test of compressing the radial artery at the wrist may prove whether the hand's arterial supply is sufficient through the ulnar and interosseous arteries.

**Technique of raising the flap**

The required size and shape of the flap are mapped out on the flexor or radiodorsal surface of the forearm. A pneumatic tourniquet is placed around the upper arm to facilitate dissection. The radial artery is first identified at the wrist, and the flap is then raised by following the course of the radial artery proximally. Care must be taken to include the fascia of the forearm in the flap. The deeper branches to muscles and tendon sheaths must be ligated. The venae commitantes should also be ligated, for they do not drain much of the
venous blood of the forearm. The flap may be raised up to the level of the elbow joint, identifying the bifurcation of the brachial artery. The artery is normally raised throughout the entire length of the flap, but it can be extended proximally or distally as desired. The veins of the flap are ligated distally, and one of the major veins of the forearm (cephalic or basilic vein) is dissected beyond the upper edge of the skin flap as far as is necessary. Anastomosis of one vein is sufficient to provide drainage for the flap.

At this stage the tourniquet is released and the viability of the flap is assessed. The remaining blood supply of the hand may also be tested by clamping the radial artery at its origin and at the wrist before dividing it finally. Simultaneous preparation of donor and recipient sites by two teams saves much operating time. After the flap has been set in place, either end of the radial artery may be anastomosed to the recipient vessel. As stated earlier, one venous anastomosis is enough to drain the entire flap.

The donor site in the forearm is either closed directly or covered with split skin graft.

Immediate reconstruction of the radial artery with a vein graft has not been abandoned.

**The mandible**

When excising a large tumour in the mouth it may be possible to preserve a rim of mandible maintaining the shape of the face. It is often necessary to excise the horizontal ramus posterior to the mental foramen, but this defect does not require reconstruction. However, removal of the anterior arch causes a severe deformity (Andy Gump) and must be reconstructed.

Two main groups of indications for mandibular reconstruction may be described:

1. restoration of the function of the resected part of the mandible
2. rehabilitation of cosmesis, that is prevention of an unacceptable change in the shape of the face after partial or total resection of the jaw.

In conjunction with mandibular resection a considerable part of the soft tissue of the mouth, the tongue and the oral vestibule are also resected. It is therefore not sufficient merely to bridge the bony defect but, in addition, the soft tissue must be replaced. The spread of cancer of the mandible is prevented by the cortical bone, but the mandibular nerve serves as a pathway for spread of the tumour. This nerve should therefore be followed up to the base of the skull and divided at that point. It can be concluded from this that carcinoma of the floor of the mouth lying close to the mandible does not always require resection of the whole width of the mandible and a strut of cortical bone can often be preserved.

This technique has been in use for some time, but osteomyelitis can arise from the poor blood supply of the strut of cortical bone and as a result of the effects of irradiation. The remaining strut of bone which is responsible for maintaining the shape of the face must then be resected later because of infection (Flynn, 1977).
Temporary replacement of the mandible

Metal plates can be used for this purpose and they can even be used to replace the head of the condyle. The main principles are absolute stable anchoring of the plate to the mandibular remnant and tension-free closure of the soft tissues of sufficient thickness over the plate. If these principles are observed such plates can provide a sound mandibular prosthesis which is stable for many years.

The temporary mandibular reconstruction can provide stable scar tissue which maintains the mandibular fragments in an acceptable position after the plate has been removed. However, the plate can also be used for a planned replacement of the bone. This technique is now largely obsolete.

Definitive mandibular reconstruction

Free autologous, homologous and pedicled autologous transplants may all be used. A large portion of the transplanted bony tissue of a free autologous transplant is not primarily revitalized, but is revived by insidious replacement and it provides a framework for new bone formation. The transplanted bone requires a good recipient area from which new bone formation can arise. The distance to be bridged by regenerating bone should not exceed 8 cm. If longer defects are bridged, the free graft is usually resorbed over several months without being replaced by bone.

Free bone grafts may be inserted intraorally in certain favourable situations such as benign bony tumours and osteomyelitis. In the hands of an experienced surgeon the danger of infection is no greater by the intraoral route than by the extraoral route. Young women in particular are grateful for the absence of visible scars achieved by a procedure of this type. Donor sites for free grafts include the iliac crest, the ribs with or without pieces of cartilage, and the vault of the skull.

Absolute stability of the opposing bony surfaces of the bone and the graft are a prerequisite for success. This stability is best achieved with plates and screws. However, these plates must be removed 6 months later, otherwise the bone graft undergoes disuse atrophy because of an absence of functional load.

Attempts have been made to broaden the indication for free grafts using cancellous bone which has an enormous osteogenic potential. Metal carrier systems or dacron mesh trays (perforated) have been developed in the shape of the mandible which are sufficiently stable, despite the presence of perforations, and which hold cancellous bone chips. The mesh system consists of titanium. The metal gutter is removed after 4-6 months. This method of reconstruction is particularly suitable for the anatomically difficult region of the chin. The success rate is variable.

Homologous lyophilized mandibular bone may also be used for reconstruction. Sensational results have been claimed in the Soviet literature. Unfortunately this method appears only to be applicable in the USSR.
Pedicled bone grafts

Rib from the anterior wall of the thorax combined with a pectoralis major flap can be considered, but a combined osteomyocutaneous latissimus dorsi flap has proved superior. The donor defect is less noticeable for the patient and the rib is well vascularized. The rib is fastened into the defect with a plate. The latissimus dorsi island flap guarantees a good blood supply to the rib and also offers sufficient soft tissue for reconstruction of the floor of the mouth or other soft tissue defects. The muscle pedicle of this flap is led under the pectoralis major muscle to the lateral part of the neck to increase the length of the pedicle.

The vault of the skull is an important donor site of bone to the mandible. The blood supply of this bone is provided by the temporalis muscle. A temporary osteotomy of the zygoma is carried out and the bone together with the muscle is then transplanted into the mouth.

Free bone grafts

The microvascular free bone transfer shows a rapid rate of healing and subsequent strength and tolerance to stress is good. These bone flaps have the ability to retain their normal bony architecture with the survival of their original osteogenic cells. The operative time is increased because of the microvascular anastomosis but the use of two teams, one carrying out the excision, the other raising the necessary flap, speeds up the procedure. The forearm flap is the most popular. The segment of bone available lies distal to the insertion of pronator teres and the maximum length available is about 10-12 cm. It can be difficult to modify the bone to the shape of the mandible but good immobilization after insertion increases the chances of success. The bone is also often too thin. Iliac crest free bone grafts, and recently the scapular free bone graft, have become popular.

Other treatments

Cryosurgery

The use of cryosurgery for intraoral carcinoma has waned as its results are usually poor. This method has been found useful in palliation because it decreases the size of the tumour, reduces the tendency to bleed, and it relieves pain.

Laser

During the last 10 years surgical laser systems have become available for clinical application; the continuous wave CO₂ laser is the most commonly used now. The CO₂ laser fulfills the requirements of the surgeon for removal of soft tissue and in some units is the preferred method for removal of intraoral tumours (Strong et al, 1979).

The laser has many advantages; there is little bleeding, no postoperative oedema, the wound heals quickly, tissue can be sent for histology, there is little postoperative pain, and the patients are often ready for discharge earlier than those treated surgically. Leukoplakia, erythroplakia, carcinoma in situ, and early invasive carcinomata are suitable lesions for
treatment; precise and orderly dissection may be carried out. Removal of larger lesions has been reported but there are few results available for comparison.

Chemotherapy

Chemotherapy has not been used as a curative method for carcinoma of the oral cavity, but many trials have been carried out using cytotoxic drugs in combination with radiotherapy (adjuvant or induction therapy); many tumours respond but increased survival has not been demonstrated (Wolf, Jacob and Makuch, 1984).

Methotrexate and cisplatin are recognized as the most active drugs in controlling oral cancer, and these drugs have been used alone or in combination with bleomycin and 5-fluorouracil, and initial response rates as high as 90% have been reported.

Management of neck metastases

The spread of cancer from the oral cavity to the lymph nodes of the neck is a poor prognostic sign (Kalnins et al, 1977). Most patients have no neck metastases at diagnosis and fewer than 10% of these patients die later because of neck disease. Thus prophylactic radical neck dissection has not been proved to increase survival, but may be useful as a means of acquiring adequate access to the oral cavity for surgical resection. If the primary site is being irradiated then elective irradiation of the neck may be carried out and this has been shown to reduce the incidence of later metastases, but the effect on survival is less certain (Stell, 1979).

In the presence of unilateral neck glands, surgical removal of the primary lesion and radical neck dissection is the treatment of choice provided the patient is fit and the primary tumour is resectable. A further advantage of radical neck dissection in these cases, apart from improving access, is that it provides space for the pedicle of myocutaneous flaps and vessels for anastomoses in free flaps.

Bilateral neck nodes carry a dismal prognosis whichever treatment method is used. Irradiation to both sides of the neck is possible and bilateral radical neck dissection may also be carried out usually with preservation of one jugular vein. This operation has many dangers including raised intracranial pressure if a jugular vein is not preserved.

The prognosis of a fixed mass in the neck is very poor, with survival to 5 years at less than 5%. Resection of the mass may be extremely difficult, but not impossible. Internal jugular vein invasion means the disease is not curable, but the common carotid artery may be resected and replaced with a graft, or even tied off if a good retrograde flow has already been shown on angiogram. Skin invasion may require the use of a flap to repair the defect. On occasion fixed cervical nodes can be radically irradiated and any residual disease may then be excised.

The problems of the dentition

The dental status and awareness of the patient must be assessed at an early stage of treatment planning.
When surgery is to be undertaken the suitability of the teeth to support a future prosthesis must be considered. The use of an obturator, whether immediate or definitive, has to be planned in advance and cooperation between the prosthodontist and surgeon is required. The use of precision dental attachments on healthy teeth has become more popular and very acceptable dental reconstruction can be carried out of both upper and lower jaws. If part of the mandible is removed the jaw deviates to the resected side and it may be necessary to fit an appliance to the maxilla to guide the mandible into its previous intercuspal position.

Of those patients who have received radiotherapy approximately 20% have a significant morbidity from osteoradionecrosis and this must be considered in treatment planning (Coffin, 1983). Pain, trismus, loss of taste and dry mouth are common after-effects of radiotherapy and these should be treated symptomatically. Anaesthetic mouthwashes may help and artificial saliva makes the mouth more comfortable. It is unlikely that anyone who has neglected his teeth in the past is suddenly going to mend his ways, so that all broken and loose teeth should be removed. Extraction of teeth should be carried out as gently as possible with the minimum damage to the supporting bones under cover of prophylactic antibiotics. Ten days should be left between the extraction and the start of treatment to ensure good healing (Starcke and Shannon, 1977). During treatment of patients with good dentition scrupulous oral hygiene must be carried out including regular fluoride treatment to protect the enamel of the teeth. A mouthwash of 0.2% chlorhexidine is both comforting and bacteriostatic and may control infection.

The ravaging form of dental caries which can follow irradiation can be eliminated in cooperative patients. In young patients with a full good dentition, construction of acrylic overlay splints has been shown to protect the teeth (Coffin, 1973). If teeth must be removed after treatment, once again atraumatic removal with good wound closure and prophylactic antibiotics should be carried out. It is often forgotten that osteoradionecrosis of the jaws may be stimulated by ill-fitting dentures. Thus it is imperative that dentures are kept clean, any signs of mucosal irritation should be investigated, and the dentures trimmed or relined accordingly. New dentures should be reviewed at 24 and 48 hours to ensure that no area is being traumatized.

In those patients receiving chemotherapy the mucous membranes are particularly susceptible to breakdown under the toxic influence of the chemotherapeutic agents and severe stomatitis often occurs. Dental extractions during and after treatment should be carried out with the same care as for patients receiving radiotherapy. It is important to maintain meticulous oral hygiene during treatment. The effect of chemotherapy is short-lived, whereas the effects of radiotherapy persist.

**Management of recurrent disease**

Recurrence of disease at the primary site is the result of inadequate surgery or failure of radiotherapy in the initial management of the patient. After failed surgery, radical radiotherapy is rarely effective. Surgical salvage after failed radiation is possible but the complication rate is high and survival poor, with a median survival time of 8-10 months (Hong and Bromer, 1983; Krespi, Wurster and Sisson, 1985)
Chemotherapy is being used with some success in the treatment of recurrent disease. Cisplatin increases survival times but no dramatic breakthrough in disease control has been identified. It is important that the symptoms of depression and pain in patients with recurrent disease are treated effectively and the patient made as comfortable as possible.

The second primary tumour

Cancer of the mouth tends to be multiple in both time and space: second tumours within the mouth at the time of presentation or later are common. The incidence of a second primary tumour is about 10%; the interval between diagnosis of the first cancer and the second may be many years but most will show themselves within 3 years. The most common site for a second primary is another site in the head and neck particularly in the mouth itself, but the oesophagus and lung are also other common sites (Tepperman and Fitzpatrick, 1981).

Management of carcinoma of the minor salivary glands

Within the oral cavity there are 400 to 500 secreting minor salivary glands, about half of which lie on the palate. These glands all have the potential for malignant change forming 10% of malignant tumours of the oral cavity that are not squamous cell in origin.

These lesions usually present as an asymptomatic swelling, but if there is perineural involvement, pain or palsy may be the presenting symptom.

The most common tumour of these glands accounting for over half of these malignancies is adenoid cystic carcinoma and this usually arises from the hard and soft palate. The tumour tends to invade adjacent structures so that total removal is difficult, but they are sensitive to irradiation. Spread to the neck and the rest of the body is a late sequel.

Mucoepidermoid carcinomata are found predominantly in the palate and are classified as high and low grade: the low grade metastasise late and are treated effectively by excision. The high grade tumour is locally aggressive and will metastasise early to the lymph nodes. Treatment is by a combination of surgery and irradiation.

Adenocarcinomata occur most commonly on the hard palate and in about the sixth decade of life. Local recurrence after either surgery or radiotherapy is common and the prognosis is very poor.

Malignant mixed cell tumours are more common in females and must be suspected on the sudden growth of a long-standing slowly growing mass. They have a high recurrence rate after treatment and distant metastases are common.
Chapter 5: Acute infection of the pharynx and tonsils

J. Hibbert

Although acute infection of the pharynx and tonsils must be one of the most common conditions encountered in medicine, it is one of the most poorly understood. Certain well-defined clinical entities do exist but many others are described for which there is little or no scientific basis. Many difficulties arise and several questions remain unanswered.

(1) Do virus infections in the pharynx and tonsils predispose to bacterial infection?

(2) Is it possible to have an infective condition involving the pharyngeal lymphoid tissue without affecting the tonsils?

(3) Is there such a condition as chronic tonsillitis?

(4) Is there an infective condition called chronic pharyngitis?

(5) Why are some patients susceptible to acute pharyngitis and acute tonsillitis and others not?

(6) Does the tonsil become irreversibly diseased after many episodes of acute tonsillitis.

One of the most misleading aspects of acute infection of the tonsils and pharyngeal lymphoid tissue is the interpretation and importance attached to throat swabs and virus cultures. The presence of an organism in a patient's throat and its culture from a swab does not mean that it is pathogenic. Studies comparing bacteriological isolates from the throat swabs of patients with acute tonsillitis and acute pharyngitis, and from asymptomatic controls, show very little difference (Box, Cleveland and Willard, 1961; Reilly et al, 1981). This particularly applies to aerobic organisms such as streptococci and Haemophilus. Similarly, virus cultures when positive do not imply that the organism is actually causing an infection; conversely patients with a clinically obvious infection may not have a positive culture from a throat swab. It has therefore been suggested that many infections may be caused by anaerobic organisms (Reilly et al, 1981; Toner et al, 1986) or by viruses (Everett, 1979). It has also been suggested that most sore throats are initiated by a virus infection which gives little in the way of signs in the throat and that secondary bacterial infection prolongs the illness and produces more in the way of signs, for example, pus in the tonsillar crypts. Most authorities distinguish between acute pharyngitis and acute tonsillitis, others talk of streptococcal pharyngitis to include infection of both.

In general, acute pharyngitis is most often felt to be an acute viral infection involving the pharyngeal lymphoid tissue and including the tonsil. Acute tonsillitis is reserved for infection with most of the signs seen in the tonsil, but it is likely that the pharyngeal lymphoid tissue is also involved. It seems unlikely that infection of some lymphoid tissue of the pharynx can occur without infection of the remainder. There is no doubt that patients who have had their tonsils removed are still susceptible to infections, both viral and bacterial.
One other area of confusion in this field is the appearance of the throat on clinical examination. It must be accepted that the normal tissues of the posterior pharyngeal wall, the pharyngeal lymphoid tissue and the tonsils are normally pink. In some asymptomatic patients the lymphoid tissue of the pharynx appears abnormally hyperaemic. The appearances of acute follicular tonsillitis are unmistakeable and easily diagnosed as abnormal, but many patients with a severe sore throat have little in the way of physical signs in the throat. Thus, the interpretation of the appearances of the pharynx and tonsils is very difficult and inaccurate. Some surgeons claim to be able to diagnose 'diseased or chronically infected' tonsils on inspection. This has never been put to test of a trial and it is the opinion of the author that the appearance of the tonsils is not related to the amount of infection which has occurred. Certainly it has been shown (Weir, 1972) that the size of the tonsils is not related to the frequency of previous infection.

In the present chapter the discussion is restricted to well-defined clinical entities for which there is at least a sound clinical, if not scientific, basis and conditions such as parenchymatous tonsillitis, chronic tonsillitis, streptococcal pharyngitis and chronic hypertrophic pharyngitis will be assigned to a non-proven category.

**Acute tonsillitis**

Acute infection of the tonsils is most frequent in childhood, presumably because immunity to common organisms has not been established. Acute tonsillitis, however, does occur in adults, but one should always be aware of the differential diagnosis and any predisposing factors.

**Causative organisms**

It is on this topic that there is a good deal of controversy because the culture of a particular organism from the throat swab of a patient with acute tonsillitis does not necessarily imply that it is pathogenic. The most common bacterium implicated in acute tonsillitis is the beta-haemolytic streptococcus. Other streptococci may be responsible on occasions, but the role of other organisms such as staphylococci, *Haemophilus* and anaerobic organisms (Toner et al, 1986) is yet to be determined. The part played by viruses in acute tonsillitis is unknown. It has been felt that an initial viral tonsillitis may predispose to a superinfection by bacteria (Everett, 1979) or that viruses alone may be responsible for tonsillitis on many occasions. Certainly a large variety of viruses has been cultured from tonsils (for example, influenza and parainfluenza viruses, adenoviruses, enteroviruses and the rhinoviruses).

**Clinical features**

Prior to onset of sore throat there may be a day or so of a prodromal illness with pyrexia, malaise and headache. The predominant symptom, however, is sore throat which is made worse by swallowing. The voice may change, partly due to the accumulation of saliva but also due to the patient's efforts to restrict movement of the soft palate and tongue. Pain may radiate to the ears or may occur in the neck due to enlargement of the jugulodigastric lymph nodes. In classical acute follicular tonsillitis, the tonsils are hyperaemic and pus accumulates in the tonsillar crypts (it should be noted that an identical appearance may occur in infectious mononucleosis). Rarely, the debris in the crypts coalesces to form a purulent
membrane. Tonsillitis does occur without pus in the crypts and in this case the tonsil is very hyperaemiac. Untreated acute tonsillitis will subside over the course of about one week, but with appropriate treatment the illness is shorter.

**Differential diagnosis**

Scarlet fever is a streptococcal tonsillitis in which the streptococcus produces the erythrogenic toxin which results in an erythematous rash. Infectious mononucleosis or acute diphtheria are the conditions most likely to be mistaken for acute tonsillitis and, of course, they do produce an acute tonsillitis. The tonsillitis and pharyngitis associated with leukaemia or agranulocytosis should always be considered.

**Treatment**

Benzylpenicillin 600 mg 6-hourly intramuscularly or ideally intravenously is the most effective treatment for acute tonsillitis. After an initial response it is usual to discontinue the parenteral antibiotic and continue with penicillin V by mouth. More commonly the antibiotic is given only by mouth.

Analgesics should also be given and soluble aspirin gargles are suitable. Many consider paracetamol to be a more suitable analgesic as this avoids gastric irritation in adults and Reye's syndrome in children. In a patient allergic to penicillin, erythromycin (500 mg 6-hourly) should be given. Ampicillin should never be used to treat acute tonsillitis in case the patient has infectious mononucleosis (see below).

**Complications**

These are best considered as either local or general factors.

**Local**

Severe swelling with spread of infection and inflammation to the hypopharynx and larynx may occasionally produce increasing respiratory obstruction, although this is very rare in uncomplicated acute tonsillitis. Peritonsillar abscess is one of the complications of acute tonsillitis and its development means that the infection has spread outside the tonsillar capsule.

Spread of infection from the tonsil or more usually from a peritonsillar abscess through the superior constrictor muscle of the pharynx results first in a cellulitis of the tissues of the neck and, later, in a parapharyngeal abscess. Alternatively, such an abscess in the parapharyngeal space can arise following suppuration in a cervical lymph node. Once infection has spread to involve the tissue spaces in the neck it can spread rapidly through these tissue spaces and into the mediastinum. These infections are often due to a number of organisms together (see below) and surgical drainage is as important in the management of the patient as are antibiotics. These neck space infections following acute tonsillitis occasionally occur in fit young patients, but are much more common in debilitated patients with conditions which predispose to infection, for example diabetes and immunosuppressed states such as lymphoma or cytotoxic therapy.
General

Systemic complications of acute tonsillitis such as septicaemia are excessively rare in adults as are the complications of acute glomerulonephritis and rheumatic fever. These are discussed under the section Tonsillitis in Volume 6.

Peritonsillar abscess (quinsy)

A peritonsillar abscess (quinsy) is a collection of pus between the fibrous capsule of the tonsil usually at its upper pole and the superior constrictor muscle of the pharynx. It usually occurs as a complication of acute tonsillitis or it may apparently arise de novo with no preceding tonsillitis. There are a number of interesting and unanswered questions regarding peritonsillar abscess, one being why does it mainly occur in young adults and rarely in children when acute tonsillitis is a disease of childhood. Although a patient with a peritonsillar abscess may have a previous history of recurrent episodes of acute tonsillitis, on occasions, there is no such history and it remains to be explained why the patient should suddenly develop a quinsy.

Bacteriology

The bacteriology of acute tonsillitis and peritonsillar abscess is different and, although one is a complication of the other, it may be that the complication only occurs in the presence of certain organisms. Although a beta-haemolytic streptococcus is the most frequent organism isolated in both acute tonsillitis and peritonsillar abscess, in the latter it is hardly ever isolated on its own. The bacteriology of peritonsillar abscess is characterized by a mixed bacterial flora with multiple organisms, both aerobic and anaerobic, being isolated. A large variety of different organisms is involved (Jokinen et al, 1985) and it may be that the reason a peritonsillar abscess develops as a complication of acute tonsillitis is because of the involvement of anaerobic organisms with the infection spreading through the tonsillar capsule.

Clinical features

The usual patient with a quinsy is a fit young adult who may have a previous history of repeated attacks of acute tonsillitis, however, the patient may never have had tonsillitis previously. Usually a quinsy is preceded by a sore throat for 2 or 3 days which gradually becomes more severe and unilateral. This heralds the development of a quinsy which is almost always unilateral but occasionally can be bilateral. At this stage the patient is ill with a fever, often a headache and severe pain, made worse by swallowing. There may be referred earache and pain and swelling in the neck due to infective lymphadenopathy. The patient's voice develops a characteristic 'plummy' quality as a consequence of the oropharyngeal swelling and an accumulation of saliva in the mouth. Examination reveals an ill-looking patient with pyrexia and often with severe trismus. The classical appearance of the oropharynx is the striking asymmetry with oedema and hyperaemia of the soft palate, and enlargement, hyperaemia and displacement of the tonsil on that side. There are usually tender enlarged lymph nodes in the jugulodigastric region on the same side.
**Differential diagnosis**

Any condition which produces swelling or oedema of the soft palate may be mistaken for a quinsy. An abscess related to an upper molar tooth is probably the most likely condition to be confused with a quinsy because it will also produce trismus and earache. Any of the causes of a parapharyngeal swelling (see Chapter 21) may also mimic a quinsy, although a carefully taken history and examination will usually make these conditions obvious.

**Treatment**

The patient should be admitted to hospital and treated with analgesics and antibiotics. In general, the antibiotic should be administered by the intravenous route and benzylpenicillin (600 mg 6-hourly) is the first choice; in those patients allergic to penicillin, erythromycin (500 mg 6-hourly) should be used. Although infection is usually with a mixture of aerobic and anaerobic organisms most are sensitive to penicillin. In a patient with an early peritonsillar abscess which is really a peritonsillar cellulitis, incision and drainage are not to be recommended. It is difficult to be certain when a discrete abscess has formed, but marked bulging of the soft palate, rather than a diffuse oedema, usually indicates this and at this stage incision and drainage should be performed. Failure of an assumed peritonsillar cellulitis to respond to adequate antibiotics within 24 hours is also an indication for incision. This is undertaken at the point of maximum swelling of the soft palate above the upper pole of the tonsil. The mucosa should be anaesthetized with a lignocaine spray and incised with a no. 15 blade with all but the terminal 0.5-1.0 cm guarded using zinc oxide tape. Usually pus will gush out of the incision. A pair of sinus forceps should be introduced through the incision and opened to break down any loculi.

The majority of otolaryngologists advise a patient who has had a quinsy to undergo tonsillectomy at a suitable interval (usually 6 weeks) to avoid a recurrence. The evidence from follow-up studies on patients who have had a quinsy shows that only about 20%, at the most, have a second peritonsillar abscess, so perhaps the policy of tonsillectomy after this condition should be reviewed (Beeded and Evans, 1970; Brandon, 1973; Hold and Tinsley, 1981; Herbild and Bonding, 1981; Tucker 1982b).

The alternative method of managing a quinsy is to perform emergency abscess tonsillectomy. The advantage of this is that incision and drainage are avoided and only one hospital admission is necessary. This assumes that all patients who have a quinsy will go on to need tonsillectomy to avoid recurrence and, as stated above, this may not be necessary. The risks of abscess tonsillectomy are mainly theoretical, namely increased haemorrhage and spread of infection. None of the studies of abscess tonsillectomy shows an increased incidence of these complications (Moesgaard Nielson and Griesson, 1981), whereas cold tonsillectomy at an interval following a quinsy has been shown to have an increased incidence of primary haemorrhage (Kristenson and Tveteras, 1984). However, if the recurrence rate after quinsy is only 20%, then abscess tonsillectomy means that many patients are having unnecessary surgery.
Complications

There is no doubt that a peritonsillar abscess is a potentially lethal condition. Rapidly increasing oedema and spread of infection can lead to pharyngeal and laryngeal oedema with respiratory obstruction, and on occasions tracheostomy is necessary. Spread of infection through the constrictor muscles of the pharynx can lead to a parapharyngeal abscess which may involve the carotid sheath leading to jugular vein thrombosis or even fatal carotid artery haemorrhage. If a parapharyngeal abscess is suspected because of tender swelling in the neck it must be incised and drained through a neck incision. In this situation there is a real risk of a salivary fistula which again puts the carotid artery at risk. However, neglect of a parapharyngeal abscess results in spread of infection and possible mediastinitis.

Parapharyngeal abscess

The parapharyngeal space lies on either side of the superior part of the pharynx - the oropharynx and nasopharynx. It is bounded laterally by the parotid gland and parotid fascia and by the medial pterygoid muscle. Medially, this space is bounded by the pharynx and separated from it by the superior constrictor muscle. Posterior to the pharynx the parapharyngeal space communicates with the retropharyngeal space. Superiorly, the parapharyngeal space is limited by the base of the skull and inferiorly, by the fascia surrounding the submandibular gland. The parapharyngeal space contains the carotid sheath with the internal carotid artery, internal jugular vein, vagus nerve, the styloid group of muscles and last four cranial nerves. It also contains some lymph nodes. Infection can spread to the parapharyngeal space from the retropharyngeal space, the peritonsillar space and from the submaxillary space. The commonest causes of a parapharyngeal abscess are tonsillitis, peritonsillar abscess or a dental infection. Rarely mastoiditis or a pharyngeal foreign body can give rise to a parapharyngeal space abscess.

Clinical features

The symptoms and signs of a parapharyngeal abscess are very similar to those of a peritonsillar abscess except that the maximum swelling is more inferiorly placed and the soft palate is less oedematous. The other striking feature of a parapharyngeal abscess is the tender firm swelling in the upper part of the neck. This allows it to be distinguished from a peritonsillar abscess in which swelling in the neck is due to enlarged lymph nodes rather than an abscess. Of course, if a parapharyngeal abscess complicates a peritonsillar abscess the clinical features are very similar.

Treatment

The patient is treated with intravenous penicillin 600 mg 6-hourly; erythromycin is the drug of choice if the patient is allergic to penicillin. If pus is present in the neck then this must be drained. If this does not seem likely, observation for 24 hours to assess the effects of antibiotic therapy is reasonable. Drainage of a parapharyngeal abscess is not without risk as trismus and pharyngeal oedema make general anaesthesia difficult. If there is any doubt about the capability of the anaesthetist to pass an endotracheal tube then an initial tracheostomy should be performed under local anaesthesia. The abscess is drained through a collar incision in the neck at the level of the hyoid bone. The abscess is widely opened and,
if it has arisen from an adjacent space, this should be opened also. Tracheostomy is performed if there is doubt about the adequacy of the patient's airway. If the infection has arisen from a dental abscess, expert advice should be sought and this should be dealt with at the same time.

**Complications**

A parapharyngeal abscess can lead to involvement of the carotid sheath with thrombosis of the internal jugular vein or rupture of the carotid artery. Spread of the abscess into the mediastinum can occur if treatment is not instituted rapidly. The possibility of airway obstruction has already been mentioned.

**Retropharyngeal abscess**

A collection of pus in the retropharyngeal space occurs in three situations. Acute suppuration in a retropharyngeal lymph node following an upper respiratory tract infection in childhood gives rise to a suppurrative retropharyngeal abscess. This condition is very rare in adults because these lymph nodes atrophy in adult life; retropharyngeal abscess of this type is considered in Volume 6. Occasionally a foreign body which has perforated the posterior pharyngeal mucosa will give rise to an abscess in this situation. In adults an abscess in the retropharyngeal space is uncommon and nearly always due to tuberculous disease of the cervical spine which has spread through the anterior longitudinal ligament of the spine to reach the retropharyngeal space. This condition is virtually confined to adult patients and is nearly always due to a reactivation of a dormant focus which has arisen from a previous infection of tuberculosis. The previous focus of infection which was almost certainly blood-borne has been controlled by the immune response and the reactivation of the focus must be due to a change in the immune system.

Usually the infection begins as a destructive process involving the intervertebral disc and then the anterior portion of the vertebral body. Histologically, the lesion is a granuloma with caseation and with epithelioid cells, giant cells and a surrounding zone of lymphocytes and fibrous tissue.

**Clinical features**

In the early stages of the disease there may be few symptoms and little to be seen on examination. Pain often occurs and later the patient may have fever. As the process progresses there may be neurological signs and, occasionally the patient may present with the signs and symptoms of spinal cord compression. The pharynx may appear normal or there may be a marked bulge of the posterior pharyngeal wall. Usually there will be nothing to feel in the neck unless the swelling is huge. Radiology usually shows evidence of bone destruction and loss of the normal curvature of the cervical spine. It must be remembered that the spine may be quite unstable and undue manipulation may precipitate a neurological event. The diagnosis is made by the radiological appearances supplemented by needle biopsy. Surgical drainage of the abscess is not normally necessary but should be carried out through a cervical incision and approach in front of and medial to the carotid sheath. Occasionally exploration of the neck is necessary to obtain biopsy material in order to make the diagnosis. Treatment should be with chemotherapy and at all times expert advice should be sought regarding the stability
of the spine. Spinal fusion is rarely necessary to stabilize the cervical spine. Occasionally surgery is required to decompress the spinal cord if there is a progressive neurological deficit.

**Acute lingual tonsillitis**

This is a rare condition, although it would be surprising if some degree of infection did not occur as part of most episodes of acute pharyngitis and acute tonsillitis. It may well be that this is the case, but the infection gives little in the way of signs and is overshadowed by the more obvious infection of the tonsils or pharyngeal lymphoid tissue. Thus lingual tonsillitis is only usually recognized in patients who have had their palatine tonsils removed. The bacteriology and clinical features of the disease are similar to those of acute tonsillitis except that the sore throat tends to be made worse by speech and protrusion of the tongue and the voice has a more striking 'plummy' quality. The lingual tonsil, best visualized on indirect laryngoscopy, is hyperaemic and pus can be seen in the follicles. Treatment is as for acute tonsillitis as are the complications, except that respiratory obstruction due to swelling and oedema of the base of the tongue and supraglottic larynx is more likely to occur.

**Diphtheria**

Diphtheria is a specific infection caused by the Gram-positive bacillus *Corynebacterium diphtheriae*. In countries with a well-developed immunization programme it is a rare disease (200-300 cases/year in the USA), although it does still occur in epidemics in underdeveloped societies where it carries a mortality of 10%. The disease spreads rapidly in conditions of overcrowding where, in addition, medical care tends to be poor. The disease itself varies in severity depending upon the immunity of the host and also the virulence of the infecting organism. Clinically, it can vary from an asymptomatic carrier state to a rapidly fatal toxic disease.

**Clinical features**

The infection remains localized to the primary site of infection, usually pharynx, larynx and nasal cavities, although in tropical and subtropical countries it gives rise to a cutaneous infection. The systemic effects of the infection are all related to the production of an exotoxin. Spread of infection is by infected droplets of nasal, nasopharyngeal or pharyngeal secretions and the incubation period is 2-6 days. In a host with well-developed immunity to the exotoxin there may be minimal symptoms or none at all, although the organism is still capable of being transmitted. The onset of the illness is heralded by malaise, pyrexia and headache. Anterior nasal diphtheria gives rise to a mucopurulent haemorrhagic discharge with nasal obstruction due to a membrane in the nasal cavity or nasopharynx. Oropharyngeal diphtheria produces a severe sore throat with a greyish-green membrane on both tonsils, posterior pharyngeal wall and soft palate. Tender bilateral cervical lymphadenopathy occurs in the jugulodigastric region. The membrane may spread from the pharynx to the larynx causing rapidly increasing airway obstruction necessitating endotracheal intubation or tracheostomy. Most of the deaths from diphtheria are related to the toxaemia which causes a myocarditis, cardiac conduction defects and arrhythmias producing acute circulatory failure. The exotoxin may also produce a fatal thrombocytopenia.
Neurological complications may appear 3-6 weeks after the onset of diphtheria and give rise to paralysis of the soft palate, diaphragm, external ocular muscles and occasionally a Guillain-Barré syndrome. Patients who recover from diphtheria may show severe scarring of the nasopharynx and oropharynx including the soft palate and the larynx, with fibrous bands and adhesions.

**Differential diagnosis**

The diagnosis of diphtheria is unlikely to be missed if the physician considers it, but it may be confused with streptococcal tonsillitis, infectious mononucleosis, the acute manifestations of leukaemia or agranulocytosis.

**Treatment**

The diagnosis of diphtheria must be made on clinical grounds and cannot await evidence of culture, although microscopic examination of the membrane may be helpful. Treatment involves neutralization of toxin with equine antitoxin (20,000-120,000) units depending upon the severity of the illness) together with benzylpenicillin (600-1200 mg 6-hourly). If the membrane is confined to the tonsils 20,000 units of antitoxin are usually adequate. Higher doses are required if it extends beyond the tonsil. With highly purified antitoxin anaphylactic reaction is rare, but small intramuscular doses can be given, followed in 1-2 hours by full intravenous dose if there is no reaction.

**Immunization**

Active immunization against diphtheria is by injection of toxoid (produced by formalin denaturation of toxin) in three doses beginning at 3 months of age. It is this immunization which has so reduced the incidence of diphtheria.

**Infectious mononucleosis**

This is an acute infection caused by the Epstein-Barr virus which has been isolated from the blood, lymph nodes and saliva of patients with the disease and the latter is probably the mode of transmission. Transmission of the disease to volunteers using the virus has not been accomplished, although there is little doubt that this virus is the causative agent and individuals with antibody to the capsular antigen of the virus will not develop the disease. In general, infectious mononucleosis is a disease of young adults, being very rare in childhood.

**Clinical features**

The clinical manifestations of infectious mononucleosis are variable ranging from an asymptomatic state to a severe systemic illness with hepatosplenomegaly.

The incubation period is of the order of 5-7 weeks and usually there is a prodromal phase of 4-5 days with malaise, fatigue and headache. The most common manifestation of infectious mononucleosis is tender enlargement of cervical lymph nodes (hence the synonym glandular fever) which, in most patients, is accompanied by a sore throat.
The pharyngeal signs are variable. Often there is an acute follicular tonsillitis, indistinguishable from a streptococcal tonsillitis. On other occasions a limited membrane may form in the oropharynx and there may be petechiae on the soft palate. Occasionally enlargement of pharyngeal and base of tongue lymphoid tissue together with a membranous slough may produce progressive respiratory obstruction necessitating tracheostomy.

Pyrexia usually accompanies the severe form of the disease and lymph nodes in other regions may be enlarged. Splenomegaly occurs in 50% of patients and hepatomegaly in 10%. Liver function tests are frequently abnormal in infectious mononucleosis and clinical jaundice occurs in about 10% of patients.

A rubelliform skin rash sometimes occurs and this is almost invariable if ampicillin is mistakenly prescribed for the condition. Ampicillin should therefore never be prescribed for a patient with a sore throat unless it is certain that this is due to acute epiglottitis.

A small proportion of patients may show a periorbital oedema particularly involving the lower eyelid and occasionally this may be mistaken for sinusitis, especially in young patients. Other more rare manifestations of infectious mononucleosis include lesions such as facial palsy, Guillain-Barré syndrome, meningoencephalitis, myelitis, myocarditis, pericarditis, nephritis and pneumonitis. The blood picture in infectious mononucleosis usually shows a leucocytosis of which 50% are mononuclear cells and 10% are atypical with pleomorphic nuclei. Occasionally haemolytic anaemia, an aplastic anaemia or thrombocytopenia will occur.

The diagnosis is made from the clinical picture together with the finding of a mononucleosis in the peripheral blood. The white blood count may be normal in the first week but rises in the second. The common serological tests depend upon the development of heterophile antibodies, the most useful being agglutinins to sheep and horse red cells and these antibodies are the basis of the Paul Bunnell and monospot tests. These tests are usually positive in the first week of the disease although around 10% of patients never develop a positive test. This proportion of negative results may be even higher in children.

**Tonsillar debris and cysts**

Caseous debris may accumulate in the tonsillar crypts and particularly in the supratonsillar cleft. The patient may notice the accumulation and may express it. This debris is of no significance and should be ignored.

Accumulation of debris in a crypt may form a tonsillar cyst. These appear as yellow-coloured inclusion cysts and again are of no significance and can be ignored.

**Unilateral tonsillar enlargement**

It is not unusual for the tonsils to be somewhat different in size, but a gross difference should always be viewed with suspicion, particularly if the patient feels the difference is of recent onset. If the larger tonsil appears abnormal a biopsy should be performed; if it looks normal then observation is indicated. Gross asymmetry with enlargement of the tonsil implies neoplasia, usually squamous carcinoma or lymphoma. Occasionally a peritonsillar abscess will give rise to confusion. When examining a patient with an apparent unilateral enlargement of
the tonsil great care must be taken to exclude a parapharyngeal mass which is displacing the tonsil medially (see Chapter 21).

**Ulceration of the tonsil**

The differential diagnosis of an ulcerative lesion of the tonsil is interesting. In theory many different disorders can give rise to an ulcerative lesion of the tonsil but, in practice, most of these can be excluded on history and clinical examination. Other investigations which may be useful are blood picture, chest radiograph, specific blood serological tests and, ultimately, biopsy. Possible causes are listed below.


2. Infection:
   a. acute - acute streptococcal tonsillitis, peritonsillar abscess, acute diphtheria, infectious mononucleosis, Vincent's angina.
   b. chronic - syphilis, tuberculosis.


**Acute pharyngitis**

The implication when this diagnosis is made is that the infection appears to be restricted to the pharyngeal lymphoid tissue and that the tonsils if present are not grossly infected. This is almost certainly an over-simplification and it seems more logical to assume that all the lymphoid tissue of the nasopharynx and oropharynx is infected at the same time. The organisms involved are similar to those in a predominantly tonsillar infection except that viruses probably represent a much higher proportion of the infecting organisms, although there is no doubt that streptococcal pharyngitis, for example, can occur in the absence of tonsils.

**Viral pharyngitis**

Many different viruses give rise to a predominantly upper respiratory tract infection which involves a rhinitis, a pharyngitis and, in some cases, a laryngitis. The possible causative organisms are shown in Table 5.1.

**Table 5.1 Causative organisms in viral pharyngitis**

- Rhinoviruses
- Coronavirus
- Influenza A and B viruses
- Parainfluenza viruses
- Adenoviruses
- Enteroviruses
- Respiratory syncytial virus.
When nasal infection and symptoms predominate the illness is designated the common cold or coryza. When the symptoms are fever and malaise with pharyngitis, influenza is usually diagnosed. In an individual patient the signs and symptoms do not allow one to predict which virus is responsible. This can only be ascertained by serological tests showing rising antibody titres and this, of course, is rarely of practical importance. Most children and adults will suffer three or four virus infections per year and these often occur in epidemics. Spread of viruses is by droplet infection and once in the upper respiratory tract the virus will enter the epithelial lining cells. It may be prevented from doing this by the presence of specific secretory antibodies in the mucociliary blanket lining the upper respiratory tract, non-specific antiviral mucoproteins or by the mechanical effect of ciliary action. Once within the epithelial cells lining the upper respiratory tract the virus divides and the cells die. The systemic effects of an acute viral illness such as fever, headache, myalgia, arthralgia, anorexia are either a result of haematogenous and systemic spread of the virus or the release of intracellular factors caused by the necrosis of epithelial cells.

**Clinical features**

The sore throat may be the initial symptom of an acute pharyngitis or it may be preceded by a rhinitis and/or conjunctivitis or by a day or so of malaise, fever and headache. The sore throat is made worse by swallowing and the pain may radiate to the ears. Cervical lymphadenopathy may occur and the clinical manifestations depend upon the nature of the virus and the resistance of the host. Thus the larynx may be involved, producing hoarseness, and the lower respiratory tract may also be colonized producing cough. It is not unusual for acute otitis media of the suppurative or non-suppurative type to be part of the illness. Secondary bacterial infection probably does occur on occasions and this will also affect the clinical picture. Examination of the patient may reveal a rhinitis, with secretion in the nasal cavities and hyperaemic congested turbinates. The nasopharynx may be hyperaemic and the mucosa covered by mucopus. The posterior pharyngeal wall also shows streams of mucopus with hyperaemic islands of lymphoid tissue which may occasionally show pustular follicles. The tonsils may be inflamed as may the larynx. The course of the illness again depends upon the host resistance and virulence of the organism, but usually the disease is self-limiting, lasting for 3 to 4 days.

**Treatment**

None of these illnesses is sufficiently severe to warrant antiviral agents and so treatment must be symptomatic with bed-rest, analgesics (for example aspirin) and fluids by mouth. If a significant bacterial complication has occurred antibiotics are indicated.

**Complications**

These are mainly local complications such as sinusitis, otitis media, laryngitis, tracheobronchitis and pneumonia. The laryngitis may occasionally result in sufficient oedema to produce a degree of respiratory obstruction, but this is much more common in children than adults. General complications are rare but include meningitis, encephalitis and myocarditis. The usual cause of death in patients with an upper respiratory virus infection is a viral pneumonia with secondary infection and this is much more likely in elderly or debilitated patients.
**Herpes simplex**

The herpes simplex virus occurs in two forms; type I usually affects the oral cavity and oropharynx and type II generally gives rise to genital infection. Primary infection with the type I herpes simplex virus usually affects children and causes a severe vesicular and ulcerative stomatitis affecting the lips, gums, tongue, buccal mucosa, soft palate and occasionally spreading to the oropharynx. Children with this condition are ill with pyrexia, tachycardia and cervical lymphadenopathy. Diagnosis is normally obvious, although occasionally it can be confused with Stevens-Johnson syndrome (see below), and can be confirmed by isolation of virus from an unruptured vesicle. The virus can be identified using fluorescent antibody or can be seen as an intranuclear inclusion in the scrapings.

Usually, the treatment of primary herpes is non-specific, namely analgesics and fluids, which may need to be given intravenously. Secondary herpetic infection occurs when the herpes virus resides within the posterior root ganglion following a primary stomatitis. Intercurrent illness then results in the appearance of herpetic vesicles usually on the lips or at the angles of the mouth as a typical cold sore.

**Herpes zoster**

Zoster virus is the same virus as that which causes chicken-pox (varicella). Herpes zoster probably arises by the reactivation of virus particles which have remained in the cranial nerve nuclei, ganglia or spinal root ganglion following a previous attack of chicken-pox. Thus it resembles the reactivation of herpes simplex virus in cold sores. Also, however, herpes zoster can occur during an epidemic of chicken-pox. In the pharynx, eruption of zoster can occur in the distribution of the fifth, ninth and tenth nerves. Thus the palate can be affected or the tonsil and posterior pharyngeal wall. This is often associated with herpes zoster oticus (see Volumes 2 and 3) and the pharyngeal manifestations are very transient and easily overlooked. They may give rise to pain on swallowing and vesicles and shallow ulcers, which heal rapidly, may be seen on the soft palate, hard palate, tonsil or posterior pharyngeal wall.

**Vesicular and bullous eruptions of the pharynx**

Herpangina, herpes simplex (less commonly) and herpes zoster may give rise to vesicles in the pharynx which break down and form small ulcers which heal rapidly. Other conditions which may occur are Stevens-Johnson syndrome, pemphigus, pemphigoid and benign mucous membrane pemphigus. These conditions affect the skin and mucous membrane of the mouth and on occasions the oropharynx. Only very rarely is the pharynx involved without involvement of the oral cavity and these conditions are discussed in Chapter 4.

**Other specific viral illnesses**

*Hand, foot and mouth syndrome*

This is an illness probably caused by a coxsackie virus (an enterovirus). The disease is characterized by a vesicular eruption in both the oral cavity and the oropharynx accompanied by vesicles on the hands and feet. There is also usually pyrexia with malaise. The illness is short-lived and self-limiting and mainly affects children.
**Herpangina**

This is a self-limiting vesicular eruption which occurs in the oropharynx and a number of enteroviruses have been implicated. It is distinguished from herpes simplex which is almost always restricted to the oral cavity and very rarely spreads to the pharynx.

**Cytomegalovirus infections**

This virus is usually transmitted by blood transfusion, although it can occur without, especially in immunosuppressed states. It gives rise to an illness which is very similar to infectious mononucleosis. The diagnosis is made by serial antibody levels.

**Other causes of acute pharyngitis**

**Acute gonococcal pharyngitis**

This disease is acquired following orogenital sexual intercourse and the majority of patients who acquire it are asymptomatic. Some may have a transient sore throat and occasionally an exudative gonococcal tonsillitis will occur with an appearance similar to streptococcal tonsillitis or infectious mononucleosis. There will also be tender enlarged cervical lymph nodes. If the diagnosis is suspected the patient should be treated with 4.8 mega units of procaine penicillin given intramuscularly usually with probenecid. A swab should be taken for microscopy and for culture and sensitivity tests. Some gonococci produce a beta-lactamase (penicillinase) and are therefore relative resistant to penicillin treatment. This should be suspected in patients who give a history of sexual contact in the Far East. Thus oral tetracycline should be used for those patients who do not improve with penicillin, and the treatment failures seem to be more frequent in patients with gonococcal pharyngitis than in those with genital infections. The dangers of gonococcal infection are those of septicaemia with septic foci in many organs but particularly in joints and tendons. As well as a septic arthritis, there occurs with gonococcal infections an arthritis which seems to be a hypersensitivity reaction and may be associated with an iritis.

**Oedema of the uvula**

Acute oedema of the uvula (Quincke's disease) is unusual without an obvious precipitating cause. The patient complains of the onset of a tickle or irritation in the throat together with a sensation of gagging. Examination shows oedema of the uvula which on occasions may be very severe indeed. The aetiology is unknown but may be related to an inhaled or ingested allergen. The oedema usually settles down of its own accord (perhaps assisted by an intravenous injection of hydrocortisone) unless it is part of a more serious allergic reaction such as angioneurotic oedema.
Other causes which produce oedema of the uvula are numerous and include:

(1) trauma: foreign bodies, surgery, endotracheal intubation;

(2) infection:
   - acute - peritonsillar abscess, viral pharyngitis, candidiasis;
   - chronic - syphilis, tuberculosis;

(3) tumours: squamous carcinoma;

(4) radiotherapy;

(5) allergic: angioneurotic oedema;

(6) blood diseases: agranulocytosis, acute leukaemia;

(7) miscellaneous: aphthous ulceration, Behçet's syndrome.

**Chronic pharyngitis**

This title implies a long-standing infection or inflammation of the pharynx; classically the disease is divided into specific or non-specific types. Specific chronic infections are due to a well-defined pathological entity, although sometimes the differentiation between acute and chronic is a little blurred. Non-specific pharyngitis is a much more difficult entity to define and diagnose and therefore to treat. It is discussed first.

**Chronic non-specific pharyngitis**

The clinical picture of this condition is the patient who complains of a long-standing discomfort in the throat, pain on swallowing and occasionally earache. This must be distinguished in the history from patients who have recurrent acute episodes. The usual description of this condition is that the patient with the above symptoms has islands of lymphoid tissue on the posterior pharyngeal wall which are hyperaemic and enlarged. Occasionally the term chronic hypertrophic pharyngitis is used. Many normal patients with nothing in the way of throat symptoms have islands of pink lymphoid tissue on their posterior pharyngeal wall, however, and in this situation it may be very difficult to decide what is normal and what is abnormal - laboratory tests such as blood picture or throat swabs are of no value. There are a number of sources of infection which are liable to produce chronic infection of the lymphoid tissue of the posterior pharyngeal wall. One is chronic sinusitis in which puss passes from the nose into the nasopharynx, oropharynx and hypopharynx. Clinically this can usually be seen particularly in the nasopharynx and the diagnosis refuted or confirmed by sinus radiographs and proof puncture. Patients with bronchiectasis or chronic bronchitis are producing infected sputum which can infect the pharynx. Gingivitis or dental caries when very severe may give rise to infected lymphoid tissue in the pharynx. Irritants which cause an inflammatory condition of the pharynx are tobacco smoking and industrial fumes.
When confronted with a patient who complains of chronic sore throat, a history, examination and investigation are designed to exclude patients with a primary carcinoma, chronic specific pharyngitis (see below) and those with chronic non-specific pharyngitis with a predisposing cause as outlined above. With these exclusions a proportion (probably the majority) of patients will still remain undiagnosed. Some of these will be classified as globus pharyngitis (see Chapter 10). The remainder of the patients are often classified, erroneously in the author's opinion, as having chronic pharyngitis. These patients should be followed-up (it is not unusual for a carcinoma to be overlooked) and examined at regular intervals. Some otolaryngologists will prescribe gargles, mouthwashes or other remedies such as Mandl's paint (iodine and potassium iodide in glycerine). There is no evidence that any of these measures is of therapeutic benefit.

Chronic specific pharyngitis

These entities are discussed below but not in order of incidence, simply in an order of convenience.

Syphilis

This is an infection by the spirochaete Treponema pallidum and apart from the congenital form is acquired by sexual intercourse. The disease progresses through primary, secondary and tertiary stages with the secondary most likely to give rise to pharyngeal symptoms. The disease manifests itself in a great many ways and clinical presentations, particularly in the tertiary stage where the presenting symptom may involve virtually any organ system. The lesion of primary syphilis is at the site of the initial inoculation and the organism can penetrate both normal mucosa and mucosal abrasions. The primary pathology in syphilis is an endarteritis with an increase in adventitial cells, a proliferation of endothelial cells and an inflammatory focus of lymphocytes, plasma cells and monocytes. Usually in parts of the lesion healing with fibrous tissue is taking place. In the secondary and particularly in the tertiary phase of the disease a granulomatous reaction takes place with necrosis of tissue and occasional giant cells.

Primary syphilis

The lesion is the chancre which develops after an incubation period, about 21 days on average. The most frequent extragenital sites for a chancre are the lips, tongue, buccal mucosa and tonsil. The lesion begins as a papule which breaks down to form a painless ulcer with indurated margins. At the same time there may be a unilateral or bilateral cervical lymphadenopathy and the glands are non-tender. Although the chancre is characteristically painless secondary infection can render it painful. The ulcer persists for a variable period, 2-6 weeks as a rule, and then heals. While the primary lesion is present the individual is capable of transmitting the disease.

Secondary syphilis

The secondary stage occurs several weeks (usually 4-6 weeks) after the primary lesion and about 30% of patients in the secondary stage will have evidence of a healing chancre. The features of the secondary stage are fever, headache and malaise with generalized
lymphadenopathy and a mucocutaneous rash and sore throat. The pharynx and soft palate show hyperaemia and inflammation, and may show lesions which have been described as mucous patches or snail-track ulcers. These lesions are most commonly seen in the oral cavity and are ulcerated lesions covered with a greyish-white membrane which when scraped off has a pink base with no bleeding. The secondary stage of the disease lasts a few weeks and here again the lesions in the mouth and pharynx are infectious. About 30% of patients will go on to develop the tertiary stage of the disease.

**Tertiary stage**

This develops some years (5-25) after the initial infection and is characterized by lesions which may be widespread throughout the body or restricted to one or two organ systems. In the upper respiratory tract the manifestations are those of the gumma. This is the granulomatous necrotic lesion which begins as a nodule and then breaks down to form an ulcer. It can occur in the hard palate, the nasal septum, the tonsil, posterior pharyngeal wall or in the larynx. The gumma, whether ulcerated or not, is typically painless. There is usually no lymphadenopathy associated with these lesions unless they are secondarily infected. When treated with penicillin the gumma will rapidly heal.

**Diagnosis**

In the primary or secondary stage of the disease spirochaetes can be identified by dark field illumination microscopy in smears taken directly from the lesion. The spirochaetes can also be identified in biopsy specimens using silver stains or fluorescein-labelled antibody. Biopsy of a tertiary lesion gives a typical histopathological picture. Serological tests for syphilis fall into two main groups: those used to identify non-specific antibodies to cardiolipin (VDRL tests) and those to detect specific treponemal antibodies (TPI and FTA).

The VDRL (Venereal Disease Research Laboratory) tests use an antigen extracted from beef heart in a slide flocculation test. The VDRL reaction begins to become positive during the first or second week after the development of the chancre and 99% of patients with secondary syphilis give a positive reaction as do a similar proportion with tertiary syphilis. Unfortunately a proportion of patients with other diseases also give a positive reaction. These include other infections involving non-syphilitic treponemes (yaws, bejel or pinta which are cutaneous infections) and also infections such as atypical pneumonia, malaria, smallpox and leprosy. Some patients with disordered immune systems such as those with systemic lupus erythematosus or rheumatoid arthritis will give a positive reaction and occasionally elderly patients who have never been exposed to syphilis will show a positive reaction.

Of the tests for specific antibody, the TPI (*Treponema pallidum* immobilization) is the most specific but also the most expensive. It depends upon the ability of antibody in the patient's serum to immobilize spirochaetes which are observed by dark ground illumination. The TPI test is 100% positive in patients with established secondary and tertiary syphilis and, if carried out correctly, is entirely specific. The FTA (fluorescent treponemal antibody) test involves the absorption from the patient's serum of cross-reacting antibodies using non-pathogenic treponemas and then absorption of specific antibody by dried *Treponema pallidum* preparations. The absorbed antibody is identified by a fluorescein-labelled antihuman gamma globulin. The FTA test is positive in 100% of patients with secondary and tertiary syphilis.
and the false positive rate is not as high as in the VDRL reaction but occasional patients with systemic lupus erythematosus or rheumatoid arthritis will be positive.

**Treatment**

In primary and secondary syphilis a dose of 2.4 mega units of benzathine penicillin in a single or two intramuscular injections is satisfactory. In tertiary syphilis a total of 7.2 mega units of benzathine penicillin is given usually as 2.4 mega unit injections at around 7 and 14-day intervals.

**Tuberculosis**

The pharynx is not a common site for clinically manifest tuberculosis. However, it is the site of primary infection which nearly always occurs in children and results in an asymptomatic primary focus in the pharynx (usually the tonsil or adenoid) with cervical lymphadenopathy.

Secondary tuberculosis affects the pharynx but only in patients with massive sputum positive and usually cavitating pulmonary tuberculosis. This is in contrast with laryngeal tuberculosis when lesions do occur with low grade or inactive pulmonary pathology. The pharyngeal lesions are secondary to coughing up heavily infected sputum and consist of very painful multiple shallow ulcers in the pharynx or oral cavity. Occasionally the pharynx is involved in patients with widespread miliary tuberculosis and here the lesions may be from blood-borne as well as sputum-borne dissemination of the disease.

Lupus vulgaris is a low grade cutaneous infection of tuberculosis and has been described in the nasal cavities and in the pharynx. Tuberculous otitis media is probably a blood-borne dissemination of the disease but occasionally it can arise from pharyngeal disease by spread from the eustachian tube.

**Diagnosis**

There is little difficulty in making the diagnosis of pharyngeal tuberculosis because of the association with pulmonary disease which is obvious clinically and radiologically.

**Treatment**

Pharyngeal tuberculosis needs no special treatment; it will be treated at the same time as the pulmonary disease, namely with triple therapy usually using isoniazid, rifampicin and pyrazinamide as first line drugs.

**Toxoplasmosis**

Toxoplasmosis is a common disease of birds and mammals caused by the protozoan *Toxoplasma gondii*. The infection can be transmitted to humans by the ingestion of cysts in uncooked meat or food contaminated with animal faeces.
In immunocompetent humans acquired toxoplasmosis usually gives rise to no symptoms. Some will have a sore throat with malaise and fever and cervical lymphadenopathy and, on occasions, the patient will simply present with an enlarged cervical lymph node. The fever and malaise may last for several weeks and many organ systems may be involved, for example, lungs, myocardium, pericardium, liver, brain and skeletal muscle. The disease is usually self-limiting and death is most unusual.

In immunodeficient individuals the disease may be much more serious with multisystem failure and death.

The diagnosis of the disease can be made by a serological test which is an indirect dye or fluorescent antibody test. On occasions lymph nodes will be removed for purposes of biopsy and the histology is typical with follicular hyperplasia and typical epithelioid cells.

Treatment

Usually no treatment is necessary but in those individuals with a severe systemic upset or in immunodeficient individuals a combination of pyrimethamine and sulphadiazine is indicated.

Leprosy

Isolated leprosy of the pharynx does not occur; it spreads to the nasopharynx and occasionally to the oropharynx from the nasal cavities. Leprosy is an infection caused by *Mycobacterium leprae* which produces a chronic disease with a spectrum of clinical manifestations. Tuberculoid leprosy is a low grade lesion affecting an area of skin and its nerve supply. Lepromatous leprosy is a more florid form of the disease with massive infection of the dermis of the skin; it can affect the nasal cavities, nasopharynx and also the testis and lymphoreticular system. Disease in the pharynx spreads from the nasal cavities and gives rise to a combination of granulomatous lesions, ulcerating and healing with fibrosis. The diagnosis is usually made by biopsy and the treatment is by chemotherapy with sulphones.

Scleroma

This is a chronic infective condition caused by *Klebsiella rhinoscleromatis*. The disease begins in the nose and only secondarily spreads to involve the pharynx where it produces granulomatous lesions and scarring.

Fungal infections

*Candidiasis (moniliasis)*

*Candida albicans* is a fungus which is part of the flora of the oral cavity or oropharynx in 30-40% of normal individuals. For the organism to become pathogenic and give rise to symptoms there must be a local or systemic change in the host. It has been called a 'disease of the diseased'.

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Local changes which predispose to infection by candida may complicate local diseases such as lichen planus and leukoplakia. Systemic antibiotic administration may change the oral flora sufficiently to disturb the local balance and allow overgrowth of candida. Radiotherapy to the oral cavity and pharynx is often complicated by candida.

In patients with chronic ill health, and particularly immunocompromised individuals, candidiasis of the oral cavity or pharynx may occur. Thus diabetes, lymphomata and treatment with immunosuppressive agents predispose to candidiasis. It is a common feature in patients with acquired immune deficiency syndrome (AIDS).

Infection with candida in the oral cavity or pharynx may be asymptomatic or may give rise to severe pain with dysphagia. Clinically it gives rise to small white patches which when removed leave an erythematous ulcer.

Candidiasis can be treated by local or systemic therapy or a combination of the two. Attention must be paid to the predisposing condition if this is possible. Local antifungal agents are usually effective: nystatin 100,000 units 6-hourly, amphotericin 100,000 units 6-hourly, miconazole 125 mg 6-hourly; occasionally systemic therapy (ketoconazole 100 mg twice daily) will be more effective.

Pharyngeal symptoms of blood disease

Certain blood diseases, because of their effect on the immune system, often present with lesions in the mouth or pharynx.

Agranulocytosis

Diseases which result in a severe decrease in the number of polymorphonuclear leucocytes are unusual but often present with oral and pharyngeal symptoms. Occasionally agranulocytosis will be idiopathic but most of these disorders are hypersensitivity reactions to drugs which only affect a small proportion of patients. Some are dose related, others are not. A large variety of different drugs is likely to produce these reactions, for example, chloramphenicol, sulphonamides, phenylbutazone, thiouracil, carbimazole and chlorpromazine. Chemotherapeutic cytotoxic agents have a direct effect on bone marrow and result in marrow aplasia and this is dose related. The first symptoms associated with agranulocytosis are fever and headache with severe pain on swallowing. The lesions in the pharynx are necrotic ulcers with a slough and may be single but are usually multiple and coalescent. There is usually no cervical lymphadenopathy. There is often severe halitosis and the patients become very ill. Culture of the ulcers rarely gives any information usually only yielding normal commensal organisms. The diagnosis of agranulocytic pharyngitis is made on the blood count and film which should always be performed on any patient with a sore throat. Treatment involves withdrawal of the precipitating agent, high dose steroids and systemic antibiotics. Blood transfusions are often necessary.

Acute leukaemia

Acute leukaemia may be of three types (lymphoblastic, myeloblastic and monoblastic) although the clinical presentation is indistinguishable. The lymphatic type is most common
in children, myeloid most common in young adults and the monocytic can occur at any age. These diseases present with fever, anaemia and bleeding disorders. Often the oral or pharyngeal manifestations are the first symptoms. Ulceration with slough and membrane formation occurs on the gums, in the oral cavity and in the pharynx. There is nothing specific about these lesions except for their severe extent and the fact that they are often associated with haemorrhage. Often there is an associated bilateral cervical lymphadenopathy. The diagnosis is made by a blood picture and bone marrow examination.

**Vincent's angina**

Vincent's angina (trench mouth) is an infection with a spirochaete, *Borellia vincenti* and an anaerobic organism *Bacillus fusiformis*. In general, Vincent's angina occurs in patients with very poor dental hygiene and a generally debilitated condition. It can occur, however, without these predisposing factors. It is essentially a necrotizing gingivitis with ulceration and bleeding of the gums which are covered with a necrotic membrane. The lesions are painful and associated with marked fetor and the patient may be pyrexial and often has tender enlarged cervical glands. The lesions may spread to involve the tonsil but, on occasions, the tonsil may be involved without a gingivitis or stomatitis. The lesion of the tonsil is a deep ulcer with a grey slough in its base. The diagnosis is made by taking a scraping from the ulcer or gingiva on a slide, staining with gentian violet and identifying both the spirochaete and the fusiform bacillus. Treatment of Vincent's angina is usually local with peroxide mouth washes, care from a dental hygienist, together with benzyl-penicillin. In addition metronidazole (500 mg 8-hourly intravenously) should be given.

**Pharyngeal stenosis**

Stenosis of the pharynx is not common. When it occurs there is fibrous tissue formation with adhesions and fibrous strands covered by mucous membrane. It can occur in the nasopharynx involving the posterior choanae or the eustachian openings. It may also arise in the oropharynx and the soft palate may be firmly fixed to the posterior pharyngeal wall. Stenosis in the nasopharynx and oropharynx may affect nasal respiration, speech and eustachian tube function. Swallowing is usually affected by hypopharyngeal stenosis. Involvement of the larynx by the same pathological process which caused stenosis of the pharynx is much more serious and results in dysphonia and airway difficulty. The cause of stenosis may be varied (Table 5.2) and usually stenosis is the end result of an inflammatory process. Occasionally the stenosis may occur at the same time as the active pathological process (for example, scleroma, leprosy, Wegener's granuloma).

Treatment of established stenosis of the nasopharynx or oropharynx is rarely necessary or successful. Hypopharyngeal stenosis can be treated by repeated dilatation.

**Acquired immune deficiency syndrome (AIDS)**

Otolaryngological symptoms occur in about 40% of patients presenting with AIDS (Marcussa and Sooy, 1985). The importance of this is first in correctly diagnosing and therefore treating the patient, and second, in recognizing the condition and avoiding surgery without precautions to safeguard members of staff. The agent known to be responsible for AIDS is the human T-cell lymphotrophic virus type III (HTLV III) which is transmitted by
sexual contact or through blood products. In the case of medical staff needle-stick injuries or contamination of cuts, abrasions or skin lesions or possibly conjunctival contamination with blood or secretions are the modes of infection. (At the time of writing the virus is still generally referred to as HTLV III. However, the internationally agreed name is now the human immunodeficiency virus and it is likely that HIV will become the routine expression.)

The most likely presentation of AIDS to the otolaryngologist is the patient who has persistent cervical lymphadenopathy. This may occur alone or may be part of a generalized lymphadenopathy associated with pyrexia, diarrhoea, weight loss, oral or pharyngeal candidiasis and lymphopenia. Some patients with AIDS may present with recurrent episodes of acute tonsillitis, acute pharyngitis, sinusitis or rhinitis. None of these show any typical features. Candidiasis is much more frequent in patients with AIDS and its occurrence in young males must be viewed with great suspicion unless there are other predisposing factors. Candidiasis affects the oral cavity and pharynx in patients with AIDS and two other lesions may be seen in the oral cavity. Hairy leukoplakia was first described by Greenspan et al in 1984 and consists of white patches on the tongue which seem to regress and then recur. Histologically, the leukoplakia shows marked keratinization giving the hairy appearance. The other lesion which occurs in the oral cavity or oropharynx in patients with AIDS is Kaposi's sarcoma. This was a rare lesion until AIDS was described, previously being confined to elderly male patients particularly of eastern European descent, children and young adults in Africa and patients who were immunosuppressed. About 30% of patients with AIDS have Kaposi's sarcoma and 25% present with it. The lesions affect the skin and oral cavity as flat or raised red patches or nodules which may ulcerate. The lesions should be excised; on occasions lymph nodes are involved and there may be systemic involvement. The disease is rarely lethal in the patient with AIDS; death occurring as a result of AIDS rather than the sarcoma.

If the otolaryngologist suspects a patient may have AIDS the patient should be referred to a venereologist or physician. The diagnosis is made by the multiple manifestations of the disease together with a lymphopenia and antibodies to the HTLV III virus. The latter may be positive in homosexual males who can transmit the virus and it is felt that approximately 30% will go on to develop AIDS. When the otolaryngologist is required to perform surgery on a patient who has high HTLV III antibody levels the patient should be managed in the ward and in the operating theatre in the same way as a patient with hepatitis B infection (Youngs, Stafford and Weber, 1986).

**Obstructive sleep apnoea**

This is a subject which is incompletely understood at the present time and there are many questions which remain unanswered about apnoea during sleep. There is no doubt that normal individuals have apnoeic episodes when asleep and that these episodes are completely harmless. On the other hand, there are examples of patients who have develop right-sided heart failure and pulmonary hypertension secondary to upper airways obstruction with apnoea. The difficulty lies in the area between these two extremes and how one should define it. Apnoea during sleep has been divided into obstructive, central, and mixed; obstructive apnoea being preceded by upper respiratory obstruction with increasing respiratory effort. Central apnoea occurs without this increasing respiratory effort but the mechanism could still be obstructive; it is simply that the drive to respiration is lacking. Purely central apnoea with no
obstructive element does occur but is probably very rare. Mixed apnoea is a combination of failure of central control and upper airway obstruction.

The mechanism of the upper airway obstruction is unknown but it has certainly been demonstrated that in sleep there is reduced activity in the muscles of the tongue, soft palate and pharynx and this reduced activity may have a central component (Remmers et al, 1978). The loss of tone in the muscles of the tongue, soft palate and pharynx results in a collapse of the airway due to a suction effect and, as respiratory effort increases, the suction and therefore the obstruction increases. As the patient becomes hypoxic there is an arousal response which increases muscle tone and improves the airway. This loss of tone in upper airway musculature during sleep has been demonstrated in normal asymptomatic individuals and the factors which lead to the development of the full-blown obstructive sleep apnoea syndrome are probably multiple. It has arbitrarily been accepted that obstructive sleep apnoea occurs when a patient has 30 apnoeic episodes each lasting 10 seconds or more during 7 hours of sleep. This is reinforced if it can be demonstrated that during these apnoeic episodes the oxygen saturation levels in arterial blood fall. This standard for sleep apnoea is by no means universally accepted and much more investigation remains to be done.

A number of studies cast doubt upon the validity of this standard. For example, an investigation by Black et al (1979) of normal individuals has shown that up to 50% of these would fit the above criteria for sleep apnoea, with oxygen desaturation levels falling as low as 60%. The relationship between snoring and sleep apnoea is also ill-defined. Snoring is more common in the elderly male and occurs in a surprisingly high proportion of the population (Lugaresi et al, 1980). Snoring is associated with a high incidence of systemic hypertension and simple snorers can develop obstructive sleep apnoea after alcohol excess (Remmers et al, 1978). Thus, the supposition has been raised that a proportion of patients who snore may go on to develop obstructive sleep apnoea.

Clinical features

The classical features of the Pickwickian syndrome, which is the first described example of sleep apnoea syndrome, occur only rarely (Gastaut, Tassinari and Duron, 1966). The classical description is of a squat obese individual with a short fat neck who snores heavily with increasing respiratory effort until prolonged apnoeic episodes occur with cyanosis. The patient then begins to breathe again with semi-arousal only to drift back into sleep and a further apnoeic episode. This disturbed sleep with oxygen desaturation leads to daytime somnolence such that the individual is falling off to sleep during the daytime and it may be difficult to distinguish the condition from narcolepsy.

Obstructive sleep apnoea can and does occur in non-obese individuals, although it seems to be more prevalent in elderly obese males. The reported incidence in the USA is considerably higher than that in the UK and Europe. This may be a real difference between the populations or may be an apparent one generated by the fact that many more sleep laboratories exist in the USA for the investigation of such disorders. Examination of the oropharynx of patients with obstructive sleep apnoea may reveal no abnormality although a proportion has a rather elongated soft palate and uvula. Examination may also show a cause for nasal obstruction such as a deviated nasal septum or nasal polyps. It should be remembered that patients with asthma and those with obstructive lung disease may show sleep
disturbance and nocturnal oxygen desaturation. After full clinical examination, investigation of patients with obstructive sleep apnoea requires sinus and chest radiography, full blood picture and pulmonary function tests. Polysomnography is essential but the facilities for this are sparse in the UK at present. Thus monitoring during sleep with ECG, EEG, EOG (electro-oculogram), ear oximetry, oral and nasal thermistors and thoracoabdominal strain gauges is necessary. Patients with obstructive sleep apnoea will show apnoeic episodes preceded by increasing respiratory effort but reduced air flow. Oxygen desaturation is often accompanied by bradycardia and cardiac arrhythmias eventually leading to right ventricular failure and pulmonary hypertension.

**Treatment**

This is a controversial subject and even more so if one considers the treatment of snoring as well. Patients with severe obstructive sleep apnoea with oxygen desaturation certainly should be treated and the options are as follows.

In patients who are obese, weight loss may help and also may make other treatment more successful. Medical treatment of obstructive sleep apnoea includes the continuous administration of oxygen at increased pressure (CPAP), although this is not comfortable and not well tolerated by patients. Protriptyline has been shown to decrease the number of apnoeic episodes.

The most successful surgery for obstructive sleep apnoea is tracheostomy, although this has major disadvantages. Correction of obvious upper airway defects such as a severely deviated nasal septum or removal of nasal polyps may help some patients and should be carried out before more drastic measures are undertaken. Recently, excision of part of the soft palate and uvula (palatopharyngoplasty) has been described (Blair-Simmons, Guilleminault and Silvestri, 1983) and the results documented. The surgery involves resection of the tonsils if they are still present together with a portion of the free edge of the soft palate and uvula. It is yet to be determined how much tissue should be resected and depends upon the amount which it is thought is redundant (in some patients as has already been said the soft palate looks normal and yet resection of some soft palate will improve the symptoms of obstructive sleep apnoea). In general 10-15 mm of soft palate should be resected and this is the full thickness of the free border of the soft palate. More should be resected medially than laterally. If the tonsils are removed at the same time, suturing the anterior to the posterior pillar has been advised. If the tonsil is not present but the pillars seem excessively flaccid a portion should be resected. The bare surface of the soft palate remaining after resection is sutured so that the mucosa of the posterior surface is brought into contact with the anterior surface.

The results of palatopharyngoplasty are variable. Some patients are cured of sleep apnoea and oxygen saturation levels of 60% before surgery may return to normal after surgery. Some patients are helped only to a marginal extent and some not at all. The implication in these patients is that the obstruction is at another level and mandibular osteotomies, hyoid bone expansion or wedge resection of parts of the base of tongue have on occasion been helpful (Guilleminault, 1984).

In patients with very severe apnoea and desaturation levels down to 50% tracheostomy is probably the treatment of choice. The complications of palatopharyngoplasty are similar to
those of tonsillectomy with the addition of palatal incompetence in some. Postoperatively approximately 30% of patients will have regurgitation of fluid after palatopharyngoplasty but this proportion improves until only a small percentage remains. However, in these patients nasal regurgitation of fluids and speech with nasal escape is permanent.

**Tonsillectomy**

Tonsillectomy, like all other surgical procedures, should never be advised unless it is virtually certain that the patient will benefit. The indications for the procedure should be stringent and loose indications such as halitosis, globus hystericus and others are to be condemned (Tucker, 1982a). Such indications simply expose the patient to totally needless trauma and risk, and serve to damage the reputation of otolaryngology as a surgical speciality. Nevertheless, when performed for the correct indications adult tonsillectomy is a very worthwhile procedure.

**Indications**

**Recurrent tonsillitis**

Tonsillitis does occur in adults but it is not common. Repeated attacks (five per year for 2-3 years) are a definite indication for tonsillectomy. Acute tonsillitis in an adult is a significant illness and can be diagnosed from the history of pyrexia, severe sore throat lasting several days and constitutional upset. Repeated episodes of minor sore throats, or a more or less constant discomfort in the throat rarely, if ever, arise from the tonsils and do not constitute indications for tonsillectomy. If there is doubt about the diagnosis in a patient with repeated sore throats and the surgeon is not certain that these represent acute tonsillitis then arrangements must be made for the patient to be examined by the surgeon during an acute attack. Only in this way can the situation be elucidated and the patient advised correctly. There is absolutely no place for a policy of performing the surgery because the patient desires it or because the surgeon cannot think what to advise for a particular symptom.

**Peritonsillar abscess**

There is no doubt that peritonsillar abscess is a serious illness with a definite mortality. There is also no doubt that a patient who has had one quinsy is liable to have a second or third. However, this liability to recurrence is closer to 20% than 100% and a quinsy is not an absolute indication for tonsillectomy. Each patient must be judged individually; one who has had repeated episodes of acute tonsillitis who then develops a quinsy is perhaps in need of tonsillectomy more than a patient who has a quinsy with no previous tonsillitis. A second quinsy is probably the point at which to decide absolutely on tonsillectomy. Abscess tonsillectomy is discussed earlier in this chapter but in the author's opinion has little to recommend it.

**Tonsillectomy for biopsy purposes**

When a tonsil is thought to be the site of a neoplasm biopsy is essential. Thus the patient with a unilateral enlarged and abnormal looking or ulcerated tonsil should be advised to have a biopsy. This can be undertaken as an incisional biopsy and is the method of choice
when a squamous carcinoma is suspected. When the swelling is thought to be a lymphoma (a large hyperaemic, fleshy-looking tonsil) the approach may either be incisional biopsy or tonsillectomy. Certainly tonsillectomy provides more tissue for the histopathologist to study but is more hazardous than incisional biopsy. If the patient has obvious lymph nodes in the neck, biopsy of one of these may be preferable to the patient and will enable the pathologist to determine lymph node architecture.

Other indications

It is usual to perform a tonsillectomy for sleep apnoea syndrome when the patient is undergoing palatopharyngoplasty.

When the glossopharyngeal nerve in treatment of glossopharyngeal neuralgia or the styloid process for Eagle's syndrome are being approached through the pharynx the tonsil must be removed to give access.

Contraindications to tonsillectomy

Bleeding disorders

Tonsillectomy should not be performed when the patient has a bleeding tendency. This bleeding disorder must be investigated, diagnosed and treated prior to tonsillectomy. Patients with a vague history of bleeding easily or with a family history of bleeding disorder must be thoroughly screened and usually their clotting mechanism will be found to be normal. In patients with a known bleeding disorder the advisability of surgery must be discussed with the patient and haematologist and the risks discussed and estimated. If it is still felt that surgery is indicated then the patient's deficiency must be corrected by the haematologist prior to surgery.

Recent infection

A recent (within 4 weeks) episode of tonsillitis makes the surgery more difficult and increases the risk of haemorrhage. A recent upper respiratory tract infection probably increases the risks of pulmonary complications of general anaesthesia and the operation should be postponed for 2 or 3 weeks.

Oral contraceptives

The risks of deep vein thrombosis after tonsillectomy are very low indeed and it is unknown whether this risk is increased in patients who are taking oestrogen-containing oral contraceptives. The three options open to the surgeon in this instance are to ignore the risk and carry on; advise the patient to stop the contraceptive 6 weeks prior to surgery and to use some other form of contraception; cover the period of surgery with low-dose heparin (5000 units 12 hours before, 5000 units with the premedication and at 12 and 24 hours postoperatively). The author favours the second option and in patients who refuse this uses the third choice.
Preoperative considerations

A patient undergoing tonsillectomy must have a carefully taken history and full clinical examination prior to surgery. In particular recent upper respiratory tract infection, bleeding tendency, coincidental anaemia or heart murmur must all be ruled out. The state of the patient's dentition must be assessed and in particular carious or loose front teeth or capped teeth must be looked for and the patient suitably informed of the risks. Urinalysis is the minimum in the way of preoperative investigations, some surgeons require also a haemoglobin estimation, blood grouping and chest radiographs.

The operation of tonsillectomy

Premedication should be determined by the anaesthetist, but Omnopon (papaveretum) 20 mg and scopolamine (hyoscine) 0.4 mg are suitable for the average adult. General anaesthesia is the routine for tonsillectomy in the UK and a nasotracheal tube is the usual method of maintaining the airway.

A detailed description of a particular surgical technique for tonsillectomy is not desirable as this is a procedure which must be taught practically and because many different variations of technique are used. It is up to each surgeon to develop his own method under close instruction and supervision. It is worthwhile, however, making some general points which are common to most techniques.

1) Most surgeons use a technique of dissection because it allows a more careful and thorough removal of all lymphoid tissue. Guillotine tonsillectomy is a dying art and perhaps this is a good thing.

2) A Boyle-Davis gag (which is not hot) should be positioned with care not to damage the lips, teeth or posterior pharyngeal wall and so that the tongue blade lies centrally along the dorsum of the tongue.

3) The mucosa of the anterior and posterior tonsillar pillars should be incised in such a way as to preserve as much mucosa as possible.

4) The plane of dissection of the tonsil is in the loose alveolar tissue between the capsule of the tonsil and the superior constrictor muscle of the pharynx. It is essential that the dissection is performed in this plane, if too deep a plane is dissected the muscle will be damaged and bleeding will be increased, and if the plane is not deep enough then lymphoid tissue will remain. It is usual to begin this dissection at the upper pole of the tonsil, but if difficulty is encountered the plane should be found more inferiorly and then followed superiorly. Most of the difficulty in tonsillectomy is because this plane is not identified and, in some situations, particularly after a quinsy this identification is quite difficult. Some muscle fibres of the superior constrictor insert into the fibrous tissue of the tonsillar capsule and they must be gently dissected away. The dissection is carried inferiorly to the junction of the tonsil and the base of the tongue and the final attachment of the tonsil is severed often using a snare.
(5) The most essential part of tonsillectomy is the control of haemorrhage and the operation is not complete until all bleeding has ceased. The fossae should be absolutely dry at the end of the procedure. It is usual to pack the fossa with gauze immediately following removal of the tonsil while the other tonsil is removed or, in the case of the second tonsil, while bleeding from the first fossa is dealt with. Much of the bleeding will have subsided by the time the gauze pack is removed. Bleeding points are identified and either ligated or diathermized depending upon the surgeon's preference. Diathermy is quicker (Haase and Noguera, 1962), but great care must be taken not to inflict an unwanted diathermy burn on the patient. The incidence of secondary haemorrhage may be greater after the use of diathermy (Carmody, Vamadenan and Cooper, 1982; Siodlak, Gleeson and Wengraf, 1985).

(6) When all haemorrhage has been controlled the blood clot must be aspirated from the nasopharynx where it will have accumulated and all swabs must be removed and counted.

The recovery of the patient from anaesthesia must be as gentle as possible, coughing and retching are liable to cause haemorrhage. The patient is recovered in the tonsil position, that is on his side with his head down so that if haemorrhage does begin the blood will flow out of the nose and mouth and not downwards to the larynx. The airway must be maintained at all times and noisy respiration may mean that there is blood in the airway. This should be inspected and the blood and secretions aspirated. At the same time the tonsillar fossae should be inspected for signs of haemorrhage.

**Postoperative care**

The postoperative care of a patient following tonsillectomy is directed towards early detection of haemorrhage. Thus regular observation of pulse (every 15 minutes for 4 hours, every 30 minutes for a further 4 hours, then hourly is a suitable regimen), and blood pressure (hourly for 12 hours) is essential. Haemorrhage is less likely to be concealed in an adult than in a child, but in the semi-conscious patient careful observation is essential and noisy respiration may indicate blood in the pharynx and the fossae should be examined. Vomiting of blood or a rise of pulse rate means the tonsillar fossae must be inspected for signs of haemorrhage. Postoperative analgesia must be given and two or three doses of Omnopon (20 mg) are suitable. After this paracetamol is usually sufficient, aspirin increases the risk of haemorrhage (Carrick, 1984).

**Complications of tonsillectomy**

These may be conveniently divided into perioperative and postoperative and the latter subdivided into immediate, intermediate and late, but this is a somewhat artificial classification. The most significant complication is haemorrhage and most of the deaths associated with tonsillectomy are directly or indirectly associated with this complication.

**Perioperative**

**Haemorrhage**

The amount of bleeding during tonsillectomy varies with individual patients and surgeons. Recent infection, previous peritonsillar abscess, and severe scarring are the factors
which increase the haemorrhage in any one patient. The surgeon can minimize the haemorrhage by a careful, gentle dissection technique. Excessive haemorrhage from both fossae raises the question of a coagulation defect which was previously unsuspected and this must be excluded. When haemorrhage is excessive and cannot be controlled by the usual means (ligation, diathermy) other methods are available. A single difficult bleeding point may need undersewing and ligating. If excessive haemorrhage occurs throughout the fossa it can be controlled by leaving a pack in the fossa and oversewing the pillars. This is hazardous because of the risk of dislodgement of the pack in the immediate recovery period. A pack of absorbable material, for example Calgitex or Gelfoam, can be used or, alternatively, a gauze pack can be left in the fossa and removed the following day by dividing the sutures. If the latter is chosen a tie can be brought out of the mouth and taped to the cheek. Occasionally an aberrant internal carotid artery will be encountered in the tonsillar fossa and this leads to massive haemorrhage which can only be controlled by ligation of the internal carotid artery in the neck. Occasionally uncontrollable haemorrhage during tonsillectomy, possibly due to a large tonsillar branch of the facial artery, requires external carotid artery ligation and this should be performed inferior to the origin of the facial artery.

**Trauma**

As has been said already, capped or carious teeth are at risk during tonsillectomy and the patient should be warned of this. Dental advice may be necessary before and after surgery. Gentle insertion of the gag minimizes the risk of damage to the teeth and also of damage to the posterior pharyngeal wall by a hurried or careless insertion of the tongue blade. Badly placed mucosal incisions can lead to excessive loss of mucosa of the soft palate, and occasional damage to the arterial supply of the uvula bilaterally may lead to loss of the uvula. Insertion of the gag can sometimes lead to dislocation of the temporomandibular joint or pain due to joint dysfunction in the postoperative period.

**Postoperative**

**Immediate complications**

**Haemorrhage**

The most significant immediate complication of tonsillectomy is so-called reactionary haemorrhage. By definition this occurs up to 24 hours postoperatively, but the vast majority of reactionary or primary haemorrhages occur within the first 8 hours.

Reactionary haemorrhage is dangerous in two ways; first, in the phase during which the patient is recovering from anaesthesia before the cough reflex is fully established, blood in the airway can result in laryngeal spasm or can asphyxiate the patient by mechanically occluding the airway. Second, haemorrhage results in hypovolaemia which, if not corrected, results in peripheral circulatory failure (shock) and eventually death.

Reactionary haemorrhage after tonsillectomy is unusual, occurring in about 0.5-1% of operations (Nesbitt, 1934; Williams, 1967). The cause is unknown but it must represent bleeding from an artery or vein which had stopped bleeding at the time of surgery. The possible causes of renewed bleeding may be dislodgement of blood clot from the lumen of
the vessel or vasodilatation of a vessel which was in spasm at the time of surgery. This is probably a local problem but it is possible that changes in blood pressure or state of vessels by anaesthetic agents may play a part. Bleeding from a vein postoperatively may be due to excessive venous pressure induced by coughing or retching. However much one speculates on the causes of reactionary haemorrhage, it must be admitted that the precise cause in the vast majority of patients is unknown.

In order to avoid disaster any bleeding following tonsillectomy must be taken very seriously. Blood must be cross-matched at the first sign of haemorrhage. The tonsillar fossae must be inspected with great care, the clot removed if possible and a bleeding point may be seen. If the bleeding is minor it may cease once the clot has been removed or it may stop with a little pressure with a swab, possibly soaked in 1:1000 adrenaline. Even if all haemorrhage then ceases the fossae should be inspected from time to time over the next few hours as bleeding can start again. If there is any doubt, the patient must be prepared for a second anaesthetic and the bleeding point ligated under general anaesthesia. This second anaesthetic is hazardous and carries a significant mortality (Davies, 1964) because the patient has already had one anaesthetic, has blood in his airway, is hypovolaemic from blood loss and also may have a stomach full of blood.

**Anaesthesia**

The anaesthetic complications in the immediate postoperative period relate to the maintenance of an adequate airway, free of secretions and particularly of blood. A swab left in the airway may obviously lead to asphyxiation and it is the surgeon's responsibility to be certain that no such oversight occurs.

**Intermediate complications**

**Haemorrhage**

The most significant intermediate complication is secondary haemorrhage which by definition, is any haemorrhage which occurs more than 24 hours after surgery and classically occurs at 10 days. This time course implies that the most likely cause is infection. Secondary haemorrhage after tonsillectomy is not common, occurring in about 1% of patients. It is not usually as severe as primary haemorrhage but, nevertheless, must be taken very seriously as it does have a definite mortality.

The patient should be treated with systemic antibiotics (intravenous penicillin or erythromycin), a haemoglobin estimation should be performed and blood should be cross-matched.

It is usual to remove blood clot from the tonsillar fossa and perhaps hold a swab soaked in 1:1000 adrenaline in the fossa. How much these measures influence the course of events is not known.

It is most unusual for secondary haemorrhage to be severe enough or sufficiently prolonged to require formal ligation under anaesthesia. If this is contemplated a coagulation screen should be performed because a minor coagulation defect which did not cause problems
at initial surgery can still cause problems at the secondary stage. In a tonsillar fossa which is infected and friable it is difficult to find and ligate a bleeding point. Occasionally this will be possible but more usually diathermy or suturing of a bleeding point will be necessary.

**Haematoma and oedema of the uvula**

Excessive surgical trauma can produce marked bruising and oedema of the uvula and, although alarming to look at, this settles down without a problem. Damage to the arterial supply to the uvula bilaterally is very rare but when it occurs the uvula may necrose.

**Infection**

Postoperatively the tonsillar fossae contain whitish slough which one would expect to be an ideal culture medium for bacteria. In fact, the fossae rarely become seriously infected. When this does occur it can be recognized by increasing, rather than decreasing pain around the end of the first postoperative week, and earache seems to be particularly common. The fossae look surprisingly normal but the combination of pyrexia with increasing pain leaves little doubt that there is infection. Untreated this may lead to secondary haemorrhage but usually haemorrhage is the first sign that infection has occurred.

**Pulmonary complications**

Pulmonary atelectasis postoperatively leading to pneumonia is very rare. It is more likely to occur if the patient has an upper respiratory tract infection at the time of surgery and inhalation of blood or fragments of tonsil tissue may precipitate it. Very rarely a lung abscess will supervene.

**Subacute bacterial endocarditis**

Tonsillectomy leads to a transient bacteraemia at the time of surgery. If the patient has an abnormal heart valve or rarely, a septal defect, subacute bacterial endocarditis may complicate the operation. For this reason all patients with a heart murmur found at the preoperative investigation should have been examined by a cardiologist and, if necessary, the surgery and immediate postoperative period should be covered by systemic penicillin.

**Earache**

Postoperative earache is fairly common after tonsillectomy and is usually referred pain from the tonsillar fossa, although occasionally this will herald the onset of a secondary infection. Acute otitis media has been described as a complication of tonsillectomy and the ears should always be examined in any patient who complains of otalgia.

**Late complications**

**Postoperative scarring**

Careless, traumatic surgery with loss of mucosa, particularly on the soft palate can result in scar tissue in the palate which limits mobility. In an extreme case it is conceivable
that this could affect the voice of a patient, producing nasal escape. The latter is of course much more common after a combined tonsillectomy and adenoidectomy.

**Tonsillar remnants**

Incomplete dissection can leave behind small islands of lymphoid tissue. Failure to remove the lower pole of the tonsil right down to the base of the tongue can result in an appearance which looks as though the tonsils have not been removed. Usually small tonsillar remnants are asymptomatic and discomfort in the throat or minor sore throats should not readily be ascribed to tonsillar remnants. Large masses of tonsil tissue remaining after tonsillectomy can result in acute tonsillitis and peritonsillar abscess has been described in a patient whose tonsils had been removed. If it is felt that tonsillar remnants are the site of recurrent acute infection then they should be removed.

**Malignancy following tonsillectomy**

A report by Vianna, Greenwald and Davies in 1971 suggested that patients whose tonsils had been removed were almost three times more likely to develop Hodgkin's disease when compared with patients who still had their tonsils. In fact the statistics of the study were felt to be subject to bias and further studies since have shown no difference in the occurrence of Hodgkin's disease after tonsillectomy (Ruuskanen, Vanha-Pertula and Kavalainen, 1971; Johnson and Johnson, 1972).
Chapter 6: Acute and chronic laryngitis; leucoplakia

Paul van den Broek

Acute laryngitis

Acute laryngitis is usually of infectious origin, either viral or bacterial, but can also be due to exogenous noxious agents. In some instances autoimmune processes can manifest themselves in the larynx, simulating an acute inflammatory reaction. Swelling of the laryngeal mucosa and the underlying tissue is the common factor in all these conditions. They can be divided into several well-defined clinical entities which are discussed in this chapter.

Acute (simple) laryngitis

Aetiology

This is the most usual form of laryngitis and occurs as a symptom of a common cold. The disease is often associated with and secondary to an acute inflammation of the nose, throat or paranasal sinuses. It is an airborne infection usually caused by adenoviruses and influenza viruses. These can damage the respiratory mucosa to such an extent that secondary bacterial infection supervenes. The bacteria involved most commonly are *Streptococcus pneumonia*, *Haemophilus influenzae* and the haemolytic streptococcus, which are common inhabitants of the aerodigestive tract in some patients. Unfavourable climate and diminished resistance through undue physical and psychological strain may be predisposing factors.

Pathology

The laryngeal mucosa shows all the signs of acute inflammation. There is extravasation of fluid. In the early phase, polymorphonuclear leucocytes, and later lymphocytes and plasma cells, predominate the picture. The underlying muscles and even the perichondrium and the cricoarytenoid joints may be affected by the process.

Areas of epithelium may be destroyed and exfoliated. Full recovery is usual but, in some instances, fibrosis will result and there can be permanent damage to the laryngeal mucosa with loss of its original structure. This can be the beginning of a chronic laryngitis.

Symptoms

The main symptoms are hoarseness, discomfort and pain in the larynx; usually there is also an irritant paroxysmal cough. The voice is hardly ever completely lost, but speaking causes discomfort and phonation often results in a high-pitched husky voice. The voice varies in strength and pitch. The irritating cough may persist after the voice has returned. The degree of temperature elevation and general symptoms depends very much on concomitant infections in the other parts of the respiratory tract. The infection is often limited to the larynx with very few general symptoms.
Clinical diagnosis

The diagnosis is made by a careful history and examination of the upper and lower respiratory tracts. The presence of a generalized infection is usually apparent. It may be necessary to substantiate this further by radiographs of the sinuses and chest. The larynx is investigated by indirect laryngoscopy, which can be difficult in the presence of acute infection as a result of hypersensitivity of the surrounding structures. A local anaesthetic consisting of 4-10% xylocaine spray may be helpful.

When the larynx can be seen, a red and swollen mucosa is found which may prevent a deeper view into the larynx. The true vocal cords lose their whitish and contrasting colour and are also swollen, sometimes partly obstructing the laryngeal lumen. In adults this hardly ever results in impaired breathing with stridor; however, in children, the clinical course can be rapidly alarming. The presence of inspissated mucus, or sometimes a true purulent discharge, is pathognomonic of a bacterial infection which needs more aggressive treatment.

Treatment

The treatment of acute laryngitis depends on the presence of concomitant infections in the upper respiratory tract and the degree of local changes in the larynx. The treatment in all cases consists of simple supportive measures such as voice rest, medicated steam inhalations and the avoidance of cold, draught, tobacco and alcohol. Expectoration of mucus can be assisted by administration of mucolytic agents.

With these measures most cases of viral laryngitis subside within a few days. After a viral laryngitis, it may sometimes be necessary to suppress persisting coughs with codeine.

The presence of bacterial infections, apparent by the presence of pus and general symptoms, is usually an indication for antibiotics. These should be broad spectrum whenever they are given without prior culture and sensitivity testing. The most appropriate antibiotics are broad-spectrum penicillins 500 mg four times daily or doxycycline 200 mg/day.

Local application by sprays with astringent agents should be avoided. When professional activities occasionally prevent taking full vocal rest, the discomfort of speaking can be overcome by a local anaesthetic spray. This should, however, always be restricted to one or two applications to prevent further irritation.

Acute laryngitis is mostly a disease with a short and benign course. Adequate treatment along the lines described above is mandatory to prevent permanent damage of the laryngeal mucosa which can be the beginning of a chronic laryngitis.

In children, symptoms and signs of acute laryngitis may be much more alarming because the laryngeal lumen is much smaller and the laryngeal tissues are more prone to oedematous swelling. This applies especially to the separate entities known as acute laryngotraacheobronchitis and acute epiglottitis.
Acute (fibrinous) laryngotracheobronchitis

Aetiology

In children, an acute respiratory infection may run a fulminant course spreading to the entire respiratory system. Small children up to the age of 7 years are most often affected. The infection can be caused by any of the microorganisms commonly involved in respiratory infections, but the haemolytic streptococcus is predominant. It usually superinfects on an infection by the influenza virus.

Pathology

Acute laryngotracheobronchitis affects the entire respiratory tract. The production of tenacious mucus which can hardly be expectorated, thus adding to the respiratory distress, is characteristic. The formation of pseudomembranes is also common, which, unlike diphtherial membranes, can be wiped off without causing bleeding. The inspissated secretions may cause total obstruction of the small bronchi leading to atelectasis.

Clinical features

Any mild common respiratory infection can lead to the complete picture of an acute laryngotracheobronchitis with its sometimes fulminant course. This generalization of a limited infection complicated by a bacterial superinfection must be recognized early to allow adequate treatment. The patient’s temperature sometimes rises up to 41°C and toxaemia may develop rapidly.

Most commonly, during or after a common cold, the child's temperature rises further and this is combined with a dry and harsh cough, hoarseness and an evident stridor. The production of tenacious secretions which can be hardly expectorated, and the mucosal swelling are the main causes of obstruction in the airway, which is most prominent at the narrow laryngeal inlet. It is at this stage that painstaking observation is necessary to prevent the child developing respiratory failure which can be rapidly fatal. The increased muscular energy consumption required for breathing and coughing, together with the retention of carbon dioxide, leads to a combination of metabolic and respiratory acidosis which paralyses the central regulation of respiration. During the initial phase the child is restless and sometimes cyanotic; in the later stages there may be an apparent improvement when the child becomes tired and calm. The retention of carbon dioxide causes a change of colour from cyanotic to pale and these are the first and often only signs of imminent disaster.

A small child with a temperature higher than 38.5°C and stridor should be admitted to hospital for observation. The clinical picture is usually dominated by the laryngeal stridor, the degree of which can scarcely be investigated objectively. The sequelae of rapidly developing or continuing stridor can only be properly judged by objective blood analysis which gives information on the degree of oxygenation, carbon dioxide retention and acidosis. It must be realized that any value can change within a very short time. Mirror examination in these children is impossible. Auscultation of the lungs is often difficult to interpret because of the stridor and massive secretions. A chest X-ray is required to investigate the degree of
involvement of the lower respiratory system. First and most important remains sound clinical judgement based on clinical examination and laboratory investigations.

**Treatment**

Acute laryngotracheobronchitis should be treated vigorously. Treatment should start immediately with antibiotics, preferably a broad-spectrum penicillin; this can be given orally or by injection depending on the general condition of the child. The value of corticosteroids in reducing the inflammatory reaction is debatable. However, when children are in distress the use of intravenous steroids will certainly do no harm and is probably beneficial. They should not be used for longer than is required to relieve the most acute symptoms; this is usually a few days.

The child should be isolated in a room or tent with moist air. Mucolytic agents can be added by mouth or in aerosols to facilitate expectoration of the tenacious mucus. If feeding by mouth is difficult a nasogastric tube should be introduced. The child must be adequately hydrated and should be carefully monitored for cardiac or respiratory failure; regular blood samples should be taken for analysis. Any sign of deterioration should lead to consultation about the necessity of airway assistance either by endotracheal intubation or tracheostomy and, if necessary, by assisted respiration. It is outside the scope of this chapter to discuss the merits of intubation versus tracheostomy. Neither method is safer than the other. They both require good instrumentation and technique, but above all well-trained personnel to observe and nurse a child with a tracheostomy or endotracheal tube.

**Subglottic laryngitis (pseudocroup, spasmodic cough)**

**Aetiology**

Acute laryngotracheobronchitis should not be confused with the condition generally known as subglottic laryngitis (pseudocroup). Subglottic laryngitis is common in young children below the age of 3 years. The symptoms are usually alarming. The exact aetiology is unknown, but the disease is often associated with an infection by one of the influenza viruses. However, certainly in view of the clinical picture, the association with a microorganism or virus seems at least doubtful. The main intralaryngeal changes, consisting of a substantial swelling of the mucosa, are found on or near the undersurface of the true vocal cords and in the subglottic region.

**Clinical features**

An attack of pseudocroup starts abruptly in a child, who might have a mild respiratory infection with some cough. Usually after the child has gone to bed and has fallen asleep, he/she wakes up again with a dry cough and a rapidly increasing stridor. The complete clinical picture develops in a very short time and seems alarming. There is usually no or only mild fever, the voice is raw and the sound resembles the barking of seals. The cough is dry. Secretions may be present but are not marked. The child becomes restless, nervous and tends to cry. The anxiety of the parents is usually projected onto the child and the clinical signs then worsen. The child may have a red appearance from exertion and perspiration. There are
no further diagnostic aids to ensure the diagnosis of this generally benign and self-limiting condition.

**Treatment**

The child and sometimes the parents need treatment. The child should be comforted as much as possible because further exertion during crying and coughing stimulates intralaryngeal swelling. Sedatives should never be given to the child because they suppress respiratory reflexes essential for maintaining oxygen and carbon dioxide levels within normal limits. Sedatives are probably best given to the parents! The value of corticosteroids for the treatment of the child is still debated. There is no objection to parenteral administration of corticosteroids but the effect is doubtful and injection can distress the child.

If possible the child should be taken to a room with moist air which helps to ease coughing and irritation. A bathroom with running hot water to produce steam is probably the best place. The child should be observed carefully until the worst symptoms settle. If any doubt is present about the child's breathing capabilities he should be admitted to hospital. Only rarely will there be a real emergency. Occasionally there may be progression to complete acute laryngotracheobronchitis, requiring more aggressive treatment.

In an emergency the treatment of choice is endotracheal intubation which can usually be limited to 1 or 2 days. Whenever this is necessary it is of paramount importance that the right diameter of tube should be used to prevent local damage to the mucosa which may otherwise lead to permanent stenosis: it is better to use too small than too large a diameter. Siliconized tubes have the advantage of a very low friction coefficient and therefore have a less traumatic effect on the mucosa.

Although the progress of the stridor should be carefully monitored, it is hardly ever necessary to proceed to aggressive treatment. The stridor usually subsides within a few hours and the next day the child may be entirely normal. There is a tendency towards recurrence and it seems that some children have a predisposition for this condition. Possibly an allergic reaction in the subglottic region is a contributing factor.

**Membranous laryngitis**

**Aetiology**

Another rare form of laryngitis, probably closely linked with acute laryngotracheobronchitis, is known as membranous laryngitis, sometimes also called croup or pseudomembranous croup. It is not caused by *Corynebacterium diphtheriae* (Klebs-Loeffler bacillus) but by various microorganisms including streptococci, *Pseudomonas aeruginosa* or Vincent's microorganisms.

**Pathology**

The presence of a confluent membrane covering the laryngeal surface is the most characteristic sign. No bleeding occurs when this is removed and no ulceration remains. The
main site is the supraglottis or the laryngeal vestibule. Only rarely does it spread to the vocal cords. It can descend from above as part of Vincent's infection of the pharynx.

**Clinical features**

The clinical picture is similar to that of other forms of laryngitis. The constitutional disturbance is often accompanied by anorexia and thirst; there is moderate fever; swallowing is painful and coughing is usually present. Later there may be stridor due to laryngeal spasm and obstruction by oedema or obstructing membranes. The disease should be differentiated from classical diphtheria which it resembles. A bacteriological investigation will establish the diagnosis and differentiate it from other forms of laryngitis.

**Treatment**

Antibiotics or sulphonamides are given depending on the sensitivity of the microorganism. Modern chemotherapy has altered the outlook of most forms of acute laryngitis.

**Acute epiglottitis**

Acute epiglottitis is a distinct form of acute inflammation of the larynx. As the name implies the epiglottis is the main site of involvement. The inflammation of the epiglottis leads to extensive swelling in the laryngeal inlet.

**Aetiology**

Acute epiglottitis has been shown to be caused by infection with *Haemophilus influenzae* type B. In general, this bacterial disease is secondary to a virus infection which has rendered the larynx more sensitive to bacteria. The disease mainly affects children but is also seen in adults.

**Clinical features**

The history is usually short and starts with an upper respiratory tract infection. There is a rapid rise in the patient's temperature sometimes exceeding 40°C, with signs of severe illness. The patient is quiet, and has pain the throat which inhibits swallowing and appetite. There is often a rapid and potentially fatal increase of stridor which is most marked in children. Unlike pseudocroup the child prefers the sitting position, the tripod sign, and usually drools.

The epiglottis is often directly visible in the throat as a rounded swollen red mass. Care should be taken when depressing the tongue as this can cause fatal glottic spasm.

**Treatment**

Acute epiglottitis should be considered a surgical emergency and the patient should be admitted to hospital. The possibility of rapid deterioration requires careful and skilled observation in order to be able to take adequate measures when necessary. When the airway
is sufficient the main treatment consists of inhalation of moist air, and antibiotics, preferably amoxycillin, should be given. Airway obstruction may develop very rapidly and some experienced surgeons advocate direct establishment of an airway. There is still much debate whether this should be by intubation or tracheostomy (Fearon and Cinnamond, 1977; Oh and Motoyama, 1977; Kinnefors and Oloffson, 1983). Both methods seem to give equivalent results.

In general, the monitoring of patients, especially children, with airway obstruction due to laryngeal infection or other causes has become a separate speciality. The otolaryngologist should be a regular observer in intensive care wards where his endoscopic and surgical skills may be needed when the airway becomes obstructed. The observation of a stridulous patient should include constant monitoring of heart and respiratory function, temperature, and regular analyses of the gaseous content of the blood.

A child has a relatively small respiratory reserve compared with an adult; the oxygen consumption per unit of bodyweight is twice as high. Furthermore, the smaller diameter of the airways results in a higher peripheral airway resistance and a greater risk of obstruction.

The relief of life-threatening obstruction of the airways can usually be effected by passing an endotracheal tube. Except in cases of a foreign body in the airway, the laryngeal opening can be found and a tube inserted. This procedure require a laryngoscope with good light and tubes of different sizes. After the airway has been re-established a decision should be taken as to whether an indwelling tube should be left or a tracheostomy performed. Siliconized tubes of various makes cause very little local irritation if they are of the correct size. Many paediatricians and otolaryngologists have accepted prolonged intubation as the method of choice for the first 10-14 days, provided that adequate monitoring facilities are available.

Successful results with prolonged intubation have led to less frequent use of tracheostomy for short-term airway relief in many centres. Whenever long periods of assisted ventilation are foreseen, tracheostomy should still be considered as a good and probably preferable alternative. Modern synthetic semirigid materials reduce the chance of complications which were common when metal tracheostomy tubes were used.

Nowadays, tracheostomy is rarely performed as an emergency procedure, because endotracheal intubation has usually been carried out first. Yet, in most centres, a regular tracheostomy in the trachea is preferred.

The differential diagnosis of different types of acute laryngitis in children is very important for the institution of adequate treatment in cases where this is necessary. Some conditions need rapid and aggressive treatment, others can be observed without danger.

The main features of the different forms of laryngitis are summarized in Table 6.1.
Table 6.1 Acute laryngitis in children

<table>
<thead>
<tr>
<th></th>
<th>Subglottic laryngitis</th>
<th>Laryngotracheobronchitis</th>
<th>Epiglottitis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong></td>
<td>Any</td>
<td>1-4 years</td>
<td>1-8 years</td>
</tr>
<tr>
<td><strong>Onset</strong></td>
<td>Gradual</td>
<td>Rapid</td>
<td>Gradual (after common cold)</td>
</tr>
<tr>
<td><strong>Aetiology</strong></td>
<td>Viral</td>
<td>?</td>
<td>Bacterial (secondary)</td>
</tr>
<tr>
<td><strong>Temperature</strong></td>
<td>&lt; 39°C</td>
<td>&lt; 38°C</td>
<td>&lt; 38°C</td>
</tr>
<tr>
<td><strong>Voice</strong></td>
<td>Hoarse</td>
<td>Harsh</td>
<td>Hoarse</td>
</tr>
<tr>
<td><strong>Posture</strong></td>
<td>Indifferent</td>
<td>Restless</td>
<td>Lying</td>
</tr>
<tr>
<td><strong>Treatment</strong></td>
<td>Supportive</td>
<td>Moist air, supportive</td>
<td>Antibiotics, rarely intubation</td>
</tr>
<tr>
<td><strong>Monitoring</strong></td>
<td>No</td>
<td>No</td>
<td>Yes</td>
</tr>
</tbody>
</table>

**Oedema of the larynx**

Oedema of the mucosa can accompany any inflammatory reaction of the larynx and is, therefore, not a specific disease but rather a sign. It may be a solitary reaction to different types of exogenous stimuli or to unknown factors. Trauma, infections and tobacco are the most important contributors. Another important cause of oedema is the physical trauma of radiation treatment. Most forms of laryngeal oedema persist over a prolonged period of time with only a limited tendency to spontaneous resolution. Several clinical entities require special mention.

**Reinke's oedema**

The accumulation of fluid under the epithelium of the true vocal cords is generally known as Reinke's oedema, named after the German anatomist, Reinke, who first described the loose areolar tissue in this region. The attachment of the vocal ligament along the medial edge and underneath the vocal cord by the lamellar fibres extending into the conus elasticus restricts the oedema to the superior surface of the cords (Mayet, 1961).

**Aetiology**

The precise cause of Reinke's oedema is not known. Allergy, infections and especially local irritants probably play a major role. Tobacco is one of the major culprits and it has been shown that an important percentage of patients are heavy smokers (Myerson, 1950). Chronic sinusitis has also been implicated, but most studies fail to mention this.
Clinical features

The condition is fairly common and comprises about 10% of benign laryngeal pathology. The disease is commoner in men but the percentage may vary greatly. The patients are mostly aged between 30 and 60 years old. The oedematous swelling of the vocal cords can easily be recognized with indirect laryngoscopy. The vocal cords are red and swollen and have a slightly translucent appearance. Sometimes the mucosa becomes redundant and polypoid projections are visible. Rarely these may be so voluminous as to cause stridor. The oedema prevents normal vocal cord vibrations causing hoarseness often with deepening of the voice. The vocal range diminishes and the voice become monotonous. There is frequently a dry cough or clearing of the throat.

Treatment

The treatment of Reinke's oedema should consist of a combination of surgery and vocal rehabilitative measures. Naturally all known causative factors, especially smoking, should first be eliminated. If smoking is not stopped the results of any treatment will be very disappointing.

Surgery consists of microsurgical removal of strips of vocal cord mucosa by microlaryngoscopy (Kleinsasser, 1976). First the mucosa is incised in a sagittal direction and the fluid, which may be either thin or mucoid, is sucked out. A strip of vocal cord mucosa is then removed with a microforceps and scissors.

There is some controversy whether both vocal cords should be stripped at the same session or whether an interval should be allowed. The present author agrees with Kleinsasser that it is perfectly safe to treat both cords during the same operation, but that care should be taken not to extend the incisions into the anterior commissure. After the procedure absolute vocal rest is advocated for one week. Healing is usually rapid and new epithelium develops with a firmer attachment to the vocal cord muscle which prevents recurrence. Speech therapy is instituted after 2-3 weeks and is continued for as long as it is felt to be beneficial. Recurrences are generally uncommon.

Some patients only come for treatment after a long history, often of many months. Delay of treatment for too long can result in the development of chronic laryngitis.

Angioneurotic oedema (angio-oedema)

This condition is characterized by recurring attacks of local swelling in various parts of the body particularly the face, larynx, extremities and buttocks. Death may result from life-threatening oedema of the larynx. Gastrointestinal disturbances presenting as colic, nausea and vomiting, are almost invariably associated with the oedema.

Generally, angio-oedema can be divided into an allergic and a non-allergic form which can be either hereditary and non-hereditary.
Angioneurotic oedema of allergic origin

This form is usually accompanied by urticaria. It presents as an acute allergic reaction to food, medicines or inhaled allergens. The diagnosis is based on the history and typical concomitant symptoms. The oedema rarely leads to laryngeal obstruction. The allergic reaction can be alleviated with antihistamines and corticosteroids. In severe cases, a subcutaneous injection of adrenaline (1:1000) 1 mg can be life saving. It is very important that the allergens are identified in order to prevent future attacks.

Hereditary angio-oedema

Less frequently, angioneurotic oedema is of non-allergic origin: it can be both hereditary and non-hereditary. The hereditary form with all its clinical manifestations was described by Sir William Osler in 1888. He recognized the life-threatening character of this condition. The underlying mechanism of the disease has been recognized to be a serum deficiency of the C1-esterase inhibitor protein (C1-iNH). This enzyme is one of a series of naturally occurring inhibitors of complement activation, kinin formation and fibrinolysis.

The complement system is composed of nine serum components which are activated by each other in a strict sequence leading to the release of polypeptides which enhance vascular permeability. When C1-iNH is not present as in hereditary angio-oedema, minor events such as trauma or emotional strain, can elicit a chain of reactions resulting in the release of complement. The mortality from concomitant laryngeal oedema is high if treatment is not rapidly instituted.

Blok and Baarsma (1984) described a family in which 35 members over three generations were traced, 14 (40%) of whom appeared to be affected. This indicates an autosomal dominant inheritance with a high penetrance. The typical triad (abdominal pain, peripheral non-pitting oedema and laryngeal oedema) was present in four patients. The laryngeal oedema being the least common symptom.

The treatment can be divided into that of the acute attack and short- or long-term prophylaxis. The acute attack is treated with an intravenous injection of C1-iNH 36,000 units. Patients should keep this at home for use in an emergency. This preparation can also be used as short-term prophylaxis in these patients before operations such as dental extractions.

Patients suffering from frequent attacks should have long-term prophylaxis which is best effected by the fibrinolytic inhibitor epsilon aminocaproic acid (EACA) and its derivative tranexamic acid, or by the androgen methyltestosterone and its derivative danazol. These stimulate the production of C1-iNH.

Laryngeal perichondritis

Perichondritis is an inflammatory reaction in the tissues covering the laryngeal cartilages. A primary form, usually developing as a blood-borne infection, used to be quite common when typhus, typhoid and smallpox were still prevalent. Immunization programmes have eliminated these diseases from major areas in the world. Perichondritis can also be
secondary to a superficial infection in the larynx spreading to the deeper tissues. In the presence of abscesses or cellulitis the cartilage may also be involved.

At present, one of the major causes of perichondritis is radiotherapy. Although the improved dosage schedules and the introduction of megavoltage radiation sources have greatly diminished the chances of serious complications, perichondritis remains a potentially dangerous hazard of this form of treatment. Perichondrium and cartilage can be affected by the radiation beam resulting in a sterile inflammatory reaction with very little tendency to spontaneous resolution. When the cartilage is uncovered after the tumour resolves, the cartilage with its inherent avascularity is liable to serious infection with little healing tendency.

**Clinical features**

Perichondritis usually develops slowly. The characteristic sign is dull pain over the entire laryngeal skeleton. The thyroid and cricoid cartilage are thickened and tender on palpation. The swollen red laryngeal mucosa can be seen on indirect laryngoscopy. The swelling may be so pronounced as to impair vocal cord function or obstruct the airway. Sometimes the cartilage is exposed. A foul smell indicates tissue necrosis. Occasionally pieces of necrotic cartilage are expectorated.

The clinical course is usually slow and protracted but, occasionally, the clinical signs of perichondritis develop very quickly, especially during radiotherapy, necessitating emergency treatment.

**Treatment**

Medical treatment, consisting of high doses of broad-spectrum antibiotics effective against anaerobes, should be instituted directly. Furthermore, corticosteroids should be given in high doses for one week with gradual withdrawal (prednisone 30 mg/day). Whenever airway problems predominate, tracheal intubation or a tracheostomy should be performed.

Although acute perichondritis can resolve quickly, especially with regard to airway obstruction, the laryngeal oedema may last for weeks to months. Laryngeal abscesses should be drained.

Separate mention should be made of the treatment of laryngeal perichondritis after radiotherapy (Stell and Morrison, 1973). This condition is generally serious and may be the cause of very resistant trouble. It may be necessary to maintain a tracheostomy for weeks, months, or even years. Resolution is usually slow and often leaves a gradually progressive narrowing of the larynx from scar formation.

**Relapsing polychondritis**

Relapsing polychondritis is a rare condition which was first described in 1923 by Jacksh-Wartenhorst. There is a recurrent inflammation of cartilage especially of the auricle, nose and trachea. The aetiology is unknown, but it is thought to be an autoimmune disease linked with the collagen-vascular group of diseases. Occasionally, patients with relapsing
polychondritis suffer from rheumatoid arthritis, systemic lupus erythematosus or ankylosing spondylitis.

The recurrent inflammation in the cartilages of the head and neck region such as the pinna, nasal cartilages, larynx or trachea, is the most prominent manifestation, but inflammatory lesions such as scleritis, conjunctivitis, keratitis, arthropathy and vasculitis can be found in other supportive tissues. The erythrocyte sedimentation rate is often raised except in the very early stages.

Laryngeal and tracheal lesions manifest themselves with signs of laryngitis and tracheitis. The mucosa is swollen especially around the epiglottis and aryepiglottic folds and descending down into the trachea. In the later stages the cartilage disappears and the epiglottis may be shrunken. The other laryngeal cartilages may also be soft and tender. Loss of cartilage of the larynx and the trachea can lead to segmental narrowing through collapse and fibrosis.

Histologically there is degeneration of the cartilage tissue due to invasion by inflammatory tissue. Several authors have described the microscopic aspects of these lesions. It seems that the process is preceded by a degeneration of the marginal chondrocytes (Valenzuela et al, 1980). The ground substance of the cartilage become acidophilic. Erosion by inflammatory tissue takes place around the cartilage and compression of lacunae can be found. Initially, the exudate is mainly composed of neutrophils but later lymphocytes, plasma cells and sometimes histiocytes can be found. In the end stages progressive fibrosis is found. Histochemical staining has shown a deficiency of matrix acid polysaccharide (Verity, 1963).

Although anticartilage antibodies have been found in the serum of patients with relapsing polychondritis, the exact meaning of these immunological findings in relation to the cause of the disease is not known (Michaels, 1984). This also applies to the finding of autoantibodies to type II collagen, a constituent of both eye and cartilage tissue, which have been found in cases of relapsing polychondritis. Although there is growing evidence that relapsing polychondritis is an autoimmune disease, definite proof is still lacking.

The presenting symptoms of laryngeal involvement are hoarseness and dyspnoea due to oedema of the mucosa. Usually there are signs of acute inflammation with fever and pain. Without treatment there is progression to serious stenosis of the larynx. The course can be very slow and relatively benign but can also be rapid and fatal. The mean survival of 27 patients found in the literature who died before 1971 was 5.25 years, but varied from one month to 23 years (Hughes et al, 1972).

The treatment of choice is still considered to be the administration of corticosteroids; initially high dosages are required, prednisone 30-60 mg/day. After the acute symptoms have subsided a maintenance dose is necessary to prevent exacerbation, prednisone 5-10 mg/day. The therapy can rarely be withdrawn entirely. When exacerbations occur the dose must be increased. The use of other drugs such as antimetabolites and immunosuppressive drugs seems logical if the autoimmune cause is accepted but, at present, not enough evidence for their usefulness is available.
Any chronic non-specific inflammatory reaction of the laryngeal mucosa may be called a chronic laryngitis. The patient suffering from chronic laryngitis complains of hoarseness over a long period of time and, on inspection of the laryngeal surface, changes in the laryngeal mucosa are always visible. The clinical picture may show variations so that over the years many descriptions have been given to clinical entities which were thought to be different. These were given separate names mainly referring to the macroscopic or microscopic appearance, and date back to the time of Virchow, the German pathologist of the second half of the nineteenth century. He introduced the term 'pachydermia' to designate local changes in the vocal cord which on microscopic appearance showed thickening of the epithelial layers. Depending on the history and the site of the lesions he further divided this entity into 'pachydermia verrucosa' and 'contact pachydermia'. Although these names are not much used today the condition is still well known to every laryngologist.

Since Virchow's time many other names have been used such as hyperplastic laryngitis, leucoplakia, keratosis and hyperkeratosis and others meant to describe chronic laryngeal disease with certain clinical and histomorphological features. However, most are so ill-defined that they are of limited value for a clinician, although they may be informative with regard to aetiology, natural history and treatment. Furthermore, the microscopic appearance of the laryngeal mucosa and the surrounding tissues may show some differences, although these are not pathognomonic for any clinical entity. On the contrary, in most of these non-specific laryngeal conditions the microscopical picture is rather uniform, being characterized by hyperplasia of the squamous epithelium with differences only at the cellular level. The mucosa of the true vocal cords is normally covered by squamous epithelium whereas the remainder of the larynx is normally covered by respiratory epithelium. During life this respiratory epithelium is subject to progressive metaplasia towards squamous epithelium. It has been known for a long time that epithelial changes in the laryngeal mucosa are enhanced by tobacco and environmental pollution and can result in the development of an infiltrating carcinoma. In 1923 the American laryngologist Chevalier Jackson stated that chronic laryngitis and what he called 'keratosis' could be precancerous. These lesions should be detected early and eradicated. Several authors (Putney and O'Keefe, 1953; McGavran, Bauer and Ogura, 1960; Gabriel and Jones, 1962; Norris and Peale, 1963; Crissman, 1979) have carried out studies to investigate the possibility of malignant degeneration, and it has been recognized that certain microscopic changes, especially at cellular level, can be regarded to be predictive for later malignant transformation.

Kleinsasser (1963) first stressed the importance of a classification system of histological grades which would help to alert the clinician to those lesions which have a higher chance of malignant degeneration and thus would need more aggressive treatment or a closer follow-up. The introduction of microlaryngoscopy by the same author around 1961 allowed well delineated and representative biopsies to be taken from the suspicious lesions in the laryngeal mucosa and was an important step towards accurate clinical assessment of these lesions. Over the past 20 years the value of this method has been repeatedly confirmed.

However, a uniform grading system has not yet been adopted and subjective interpretations of the degrees of change found in the laryngeal mucosa under pathological circumstances are an obstacle to a reliable and reproducible grading system. Perhaps more
objective means of quantification of the changes at the cellular level, for instance, morphometric or photometric evaluation (Hellquist and Oloffson, 1984; Hellquist et al, 1984), will enhance the reliability of such a system and make it more acceptable.

**Aetiology**

Chronic laryngitis primarily affects middle-aged men but the variation in age is wide. The median age is approximately 57 years, that is about 5 years less than the average age of patients with laryngeal carcinoma.

Many factors, both endogenous and exogenous, have been incriminated as causative. The exogenous stimuli may be physical, chemical or infective, the most important being inhaled irritants and, notably, cigarette smoke. In many studies it has been shown that metaplastic changes in the surface epithelium of the airways in heavy smokers is more marked than in non-smokers. This also explains the sex differences observed.

The changes in the mucosa are most marked on the ventricular bands and the true vocal cords. Auerbach, Hammond and Garfinkel (1970), in a study of larynges at post-mortem found epithelial changes in 6% of non-smokers, 22% of smokers of 20 cigarettes and 44% of smokers of 40 cigarettes a day. The degree of cellular atypia is also strongly related to smoking habits. The presence of cellular atypica is observed in 85% of heavy smokers. After smoking has been stopped the hyperplasia remains, but the cellular atypia gradually diminishes.

In series of patients with chronic laryngitis the percentage of smokers is usually high (Putney and O'Keefe, 1953, 89.8%; Norris and Peale, 1963, 94%). Alcohol is also often mentioned as a causative factor (Hinds, Thomas and O'Reilly, 1979), but much less solid evidence is available. In the author's own data on smoking and drinking habits in a large number of patients with head and neck cancer, the alcohol consumption among the laryngeal cancer patients is much lower than among patients with carcinoma of the oral cavity and pharynx. This could point towards a direct surface effect. In a study by Stevens (1979), hamsters exposed to benzpyrene and alcohol were affected more frequently by laryngeal carcinoma than those not receiving alcohol.

Chronic laryngitis is more frequently found in patients suffering from a chronic infection of the upper or lower respiratory tract. Stell and McLoughlin (1976), studying a group of 58 patients with chronic laryngitis, found a history of infection in 53%. It is not clear whether this association is due to the increased incidence of coughing in these patients resulting in mechanical trauma, or whether a more generalized involvement of the respiratory mucosa including that of the larynx is the main reason. Probably both factors play a role. The stubborn nature of chronic respiratory tract infections means their elimination plays an important role in the treatment of chronic laryngitis.

Besides coughing, vocal abuse is an important physical factor which contributes to the development of inflammatory lesions of the larynx due to mechanical irritation. Muscular strain, venous congestion and forced vocal attack are probably involved. Virchow recognized the factor of vocal abuse when describing the picture of pachydermia verrucosa in a Prussian army officer. It is certain that abnormal vocal strain especially in those who use their voice
professionally can be a source of tissue changes which have a disastrous effect on the voice. These are discussed in Chapter 7.

Finally endogenous factors must also be taken into account. These may be constitutional or metabolic. Short, heavily-built people are more prone to chronic laryngitis. Diabetes, hypothyroidism and vitamin A deficiency can also be contributory.

**History and clinical signs**

Chronic laryngitis is usually of insidious onset and rarely develops after an acute laryngitis, although this may be the trigger in a larynx which has already been affected by asymptomatic epithelial changes. When no acute infection has been present it is difficult for the patient to pinpoint the exact time of onset.

There are no general symptoms and no fever. Hoarseness is the most frequent and often the only symptom. This complaint tends to vary with the time of the day and with the intensity with which the voice is used. Typically the patient complains that the voice is worse in the morning. Drying of the laryngeal mucosa during the night through mouth breathing and a decreased frequency of swallowing is probably the reason for this. Inspissated mucus which has to be cleared causes dryness and the feeling of a foreign body in the throat. When the throat has been cleared and the mucosa is moistened again the voice gradually improves. However, it remains harsh with varying pitch and volume. There can be periods of complete aphonia, although these are rare. The vocal range is reduced, especially in the higher frequencies. There may be a cough caused by local irritation as a result of mucus, dryness or intraepithelial changes, which can worsen the other symptoms. Pain is rarely present unless undue strain by coughing has damaged the mucosa.

The complaints of a patient with chronic laryngitis tend to develop slowly and then become stationary. There may be variations over short periods but, in general, chronic laryngitis remains constant over a long period.

**Clinical picture**

Chronic laryngitis is diagnosed from the history and by indirect laryngoscopy. Direct inspection of the larynx is the corner-stone of any diagnosis. Furthermore, a histopathological examination of tissue removed from the laryngeal lesions is indispensable. Without these investigations a diagnosis cannot be made.

Chronic laryngitis can be divided into several clinical conditions.

**Simple diffuse chronic laryngitis**

The patient complains of hoarseness and sometimes a slight cough over a long period of time. These complaints start insidiously, occasionally during an upper respiratory tract infection, and persist although they are not always present.

Examination shows a reddened hyperaemic laryngeal mucosa. The true vocal cords lose their white colour and become pink or red, sometimes with a glossy appearance or with
submucosal oedema. Diagnosis is made on the findings at indirect laryngoscopy. If the laryngeal mucosa is smooth and regular, a biopsy should be avoided in the early stages to prevent damage to the laryngeal mucosa.

Simple chronic laryngitis can best be treated by vocal rest, inhalations with mentholated air and, if the slightest signs of infection are present, an appropriate antibiotic should be given. Furthermore, all possible noxious agents should be avoided especially tobacco and alcohol.

This form of laryngitis is reversible within a few weeks with adequate measures.

**Chronic diffuse hyperplastic laryngitis**

The most important contributing factors are chronic infection of the sinuses and lower airway; tobacco and alcohol; occupational, chemical or physical irritants; and mouth breathing. The onset is insidious and these patients often have a history of coughing.

The laryngeal picture is determined by more conspicuous changes of the laryngeal mucosa, especially the true vocal cords, which lose their normal appearance. The mucosa is clearly swollen and the white colour replaced by red, deep red or sometimes grey. The surface of the mucosa is hardly ever completely smooth. Patches of epithelial thickening and broad-based polypod lesions can be found. The picture is much more alarming than that of simple laryngitis and it may be difficult to differentiate its appearance from carcinoma or specific laryngitis.

This form of laryngitis is usually associated with chronic respiratory infections such as sinusitis and bronchitis.

**Keratosis, leucoplakia, pachydermia, squamous cell hyperplasia**

These terms are based partly on clinical appearance and partly on histological features. They are still often used for local or more diffuse lesions of the larynx, primarily the vocal cords. Many clinicians would like to see these terms abandoned because they are ill-defined and confusing. The lesions are often well circumscribed and well demarcated from the surrounding tissue. One or both cords can be affected as well as the anterior commissure.

Very often the surface of the lesion is white in colour as a consequence of thickening of the squamous epithelium covered by excess keratin. The elevation of the lesion from the surrounding tissue can be seen clearly under the operating microscope. The surrounding mucosa may be normal or may resemble simple chronic laryngitis. The keratinization may be so abundant that the picture may simulate a benign tumour, a squamous papilloma or a verrucous carcinoma. When the lesion lies in the posterior part of the glottis where the mucosa is redundant to allow movement of the cords, it has also been named 'posterior laryngitis'. Its possible relation with nocturnal regurgitation of gastric acid has led to the name 'acid laryngitis'. At this site the epithelium is already normally slightly thickened and the transition towards abnormal is very gradual, so that care should be taken not to overdiagnose this condition. The presence of symptoms including gastro-oesophageal reflux usually helps to make a diagnosis.
Contact ulcers - contact pachydermia

These terms were coined by Jackson (1928) and Virchow (1887) respectively and are still in use today. Although Jackson later included vocal cord granuloma, this should be considered as a separate entity.

Contact ulcers are saucer-like lesions on the medial edge of the vocal cord exactly at the vocal process. They can be bilateral and symmetrical, often with a small projection on one cord which fits the saucer of the other side. There is no epithelial defect, thus the word 'ulcer' is not correct. The lesions are made up of thickened epithelium with a central indentation exactly at the site of the mucoperichondrial covering of the vocal process. Patients with a contact ulcer may complain of pain locally. This disease usually occurs in tense personalities and it is agreed that vocal overuse and abuse are important aetiological factors.

Histology of chronic laryngitis

The importance of an accurate histological diagnosis in cases of chronic laryngitis cannot be overestimated. The relation between chronic epithelial changes and carcinoma has been repeatedly demonstrated, although the percentage is generally only between 3 and 5% (Crissman, 1979).

The chances of malignant degeneration are related to certain histomorphological characteristics which can help to divide the lesions into high and low risk groups (Oldekalter, 1986). A reliable diagnosis demands two criteria to be fulfilled. The first is that the biopsy is taken from a representative site of the lesion. If the lesion is small a total removal will allow examination of the entire specimen. When the lesion is extensive, biopsies are taken from the most aggressive looking part of the lesion. Epithelial changes such as carcinoma in situ are often found in the vicinity of a squamous cell carcinoma and care should be taken not to overlook the site where a carcinoma may be present, for instance, subglottic or in the ventricle (McGavran, Bauer and Ogura, 1960). The second requirement regards the way the specimen is sent to the pathologist. If possible the removed undamaged piece of mucosa should be orientated to allow the pathologist to make sections vertical to the mucosa to avoid tangential sectioning which may simulate infiltration of tumour tissue into the underlying tissue.

Although it is recognized that squamous epithelium can undergo changes which may eventually lead to squamous cell carcinoma and that certain histological and cellular features indicate a higher chance of malignant transformation, a uniform and internationally accepted classification of these lesions is still lacking, mainly because each classification depends on a subjective interpretation. The results of objective methods such as morphometry (Oldekalter et al, 1985), photometry (Hellquist and Olofsson, 1984) and others have not yet found widespread clinical application.

In Europe the most accepted classification is that originally proposed by Kleinsasser (1963), who introduced a histological grading system. The normal squamous epithelium of the larynx is non-keratinizing in several layers. Adjacent to the subcutaneous tissue lies the basal layer (stratum germinativum), consisting of cylindrical cells with ovoid nuclei. Mitoses are frequent in this germinal layer of the epithelium. From this layer the cells move to the
surface, gradually changing from round to flat cells which are shed from the surface. Although the more superficial cells contain intracellular keratohyalin granules, there is no full development towards keratin. Under normal circumstances this maturation process follows a regular pattern with normal cells grouped in layers. A disturbance of this normal maturation underlies the pathological changes found in chronic laryngitis. The degree of disturbance of this maturation process, also called dysplasia, forms the basis of Kleinsasser's and other classification systems.

*Grade I: simple squamous cell hyperplasia or keratosis*

There is thickening of the entire epithelium. The basal cell layer becomes undulated, sometimes with deep projections in the stroma (acanthosis). There is further differentiation of cells towards intracellular keratin formation. The nuclei extend into the keratin layer (parakeratosis) and keratic covers the lesion to a varying degree (hyperkeratosis), but the regular maturation pattern is retained.

*Grade II: squamous cell hyperplasia or keratosis with atypia*

In the second stage there is early disorganization of the maturation process, but this is not very extensive; the loss of the normal organization is limited and never affects all layers at the same time. There is atypia at the cellular level, including altered nucleus/cytoplasm ratio, abnormal DNA content, abnormal mitoses and other aberrations. Acanthosis, dyskeratosis and parakeratosis are also present.

*Grade III: carcinoma in situ*

This third stage shows the most serious disorganization of the squamous epithelium identical to that of severe dysplasia. Frequent mitoses and cellular anomalies are found. The entire epithelium shows all cellular changes compatible with squamous cell carcinoma, but without infiltration through the basal membrane.

**Treatment**

It is important to make an early diagnosis and classify the lesion in order to institute adequate treatment. Squamous cell hyperplasia should be removed locally. Microlaryngoscopy as introduced by Kleinsasser is the most appropriate method. The lesion can usually be peeled off the underlying muscular tissue and vocal ligaments adequately and accurately. Both cords can be treated in the same session if the anterior commissure is left untouched. Occasionally the lesion may be so diffuse that total removal is not possible. In these cases removal is performed as far as possible (Kleinsasser, 1976).

Histological examination is always performed. Class I and II lesions normally need no further treatment. For class III lesions opinions differ. The increased chance that an infiltrating carcinoma will develop from such a lesion means that less risk can be taken. If possible, total removal by microlaryngoscopy is the method of choice and should be performed as soon as possible. A laryngofissure is rarely indicated. Furthermore, a good inspection of the surrounding mucosa is mandatory, especially in the subglottic region, in order not to overlook an infiltrating carcinoma. Radiotherapy is only indicated when removal
must be repeated for recurring or diffusely spreading lesions. Although some authors prefer radiotherapy as the primary treatment of choice, others feel that the recurrence rate after radiotherapy is too high and therefore advocate surgical removal or laser treatment.

Local removal is the method of choice in previously untreated carcinoma in situ. Radiotherapy should be given if the disease is so diffuse that local removal cannot be performed, or for recurrence of lesions previously removed.

Any patient suffering from squamous cell hyperplasia of the larynx needs careful follow-up. Class II and III lesions carry an increased risk of developing squamous cell carcinoma and need regular, life-long follow-up (Hellquist, Lundgren and Oloffson, 1982).

**Atrophic laryngitis**

This rare entity is also called laryngitis sicca. It is characterized by atrophic changes in the respiratory mucosa with loss of the mucus-producing glands. It is usually part of an atrophic rhinitis caused by *Klebsiella ozaenae*, but is much rarer.

**Pathology**

Fibrosis of the corium of the mucosa leads to anaemia and glandular atrophy. The respiratory epithelium shows squamous metaplasia with loss of cilia. Inspissated mucus adheres to the epithelium, dries and forms thick crusts. The most common sites are the false cords, the posterior region and the subglottic region.

**Clinical features**

An irritable cough and hoarseness are the most important signs. Crusts which are sometimes blood stained are expectorated. The crusts can readily be seen in the larynx and are the most important diagnostic feature. If the nose and sinuses show similar pathology the diagnosis is made easily.

In far advanced stages, when repeated crusting has led to total atrophy, there may be reactive inflammation of the cartilage structures with progressive fibrosis and eventually serious stenosis of the larynx.

**Treatment**

Treatment is directed at underlying causes such as generalized infections, poor nutrition or, rarely, syphilis. Local treatment consists of the stimulation of secretions and the removal of crusts. Secretions can be encouraged by small doses of ammonium chloride or iodide. The mucus so produced is less viscid, it softens the crusts and facilitates expectoration. The larynx can be sprayed with solutions of mucolytic agents. Local irritation especially by smoking should be strictly forbidden.
Contact granuloma (intubation granuloma)

A separate entity is formed by localized granulomata, nearly always unilateral, situated medially or superiorly on the vocal process of the arytenoid cartilage.

These lesions are often confused with contact ulcers. Jackson (1923) made no mention of granuloma when he first presented the clinical and pathological features of contact ulcers. Later he added the description of granulomata and considered them as part of the healing process. Benjamin and Croxson (1985) consider granulomata as a separate clinical entity. The granuloma has a typical polypoid appearance which is a local reaction to trauma. Granulation tissues can develop if the perichondrium is damaged either by vocal trauma or through trauma from an endotracheal tube. The granuloma may develop a long time after intubation.

Clinical features and diagnosis

Slight hoarseness is the most important symptom; the diagnosis is readily made by indirect laryngoscopy. The lesion is usually small but can become quite large and sometimes partially obstructs the laryngeal lumen. It is most restricted to one side and is usually attached on the superior edge. The granuloma can be pedunculated and move up and down between the cord. The colour is red, sometimes stained with dark areas from haemorrhage.

Treatment

These granulomata are not easy to treat. Simple removal by microlaryngoscopy seems the method of choice but local recurrences are common. The carbon dioxide laser advocated in recent years for treatment of this condition has not really improved matters; repeated treatment is often necessary. A conservative approach of 'wait and see' is sometimes as effective and should be considered in every case in view of the very resistant nature of this condition in spite of surgical removal.

Amyloidosis

Amyloid is an eosinophilic hyalin material with a strong affinity to certain dyes such as Congo red. Amyloidosis is a disease which has been known for over 140 years in which infiltration of different organs may occur.

Laryngeal amyloidosis is rare. It may be part of a generalized amyloidosis with involvement of many organs, in particular the heart, kidneys, gastrointestinal tract, blood vessels, liver. There are two main forms. Type A is the secondary type which is found in patients with long-standing inflammatory diseases; type B is the primary which is sporadically found in the larynx. This latter form may also be found in patients with multiple myeloma or macroglobulinaemia. With modern immunohistochemical methods it is possible to differentiate further the different types of amyloid, which morphologically, are all the same.

The clinical presentation of amyloid in the larynx is not characteristic. It may present as a solitary polyp on the vocal cord or as a more diffuse swelling in any region of the larynx or even trachea. Ulceration is not present. A biopsy of the lesion will lead to a diagnosis. A biopsy of the wall of the rectum is necessary to exclude a generalized form.
In rare instances there can be extensive infiltration from the larynx into the trachea, with progressive obstruction of the lumen of the airway.

Treatment is directed towards any underlying disease. The local lesion can be removed by microlaryngoscopy with sharp instruments or with the laser. The disease usually only progresses very slowly and repeated removal may be necessary.

**Granulomatous infections**

Both specific and non-specific granulomatous diseases can be found in the larynx. Sometimes the exact nature of the disease may be evident especially when a diagnosis has already been made elsewhere. However, it may be very difficult to make a diagnosis if only the larynx is involved. History, histopathological investigations and blood chemistry are essential for a correct diagnosis.

**Tuberculosis**

Laryngeal tuberculosis used to be commonly associated with pulmonary tuberculosis. In the western world, improved socio-economic circumstances and the advent of chemotherapy have resulted in a marked decline in tuberculosis, which is now rare in these areas. However, in developing countries the situation is quite different (Manni, 1982).

Laryngeal tuberculosis is almost exclusively found in patients suffering from open pulmonary tuberculosis and, in most cases, is a result of contamination by sputum containing acid-fast bacilli. Laryngeal tuberculosis only rarely develops by a blood-borne infection which causes diffuse involvement of the laryngeal mucosa with extensive ulceration. The frequency of the involvement of the larynx is difficult to estimate and varies in the different series published. Auerbach (1946) in his historic publication found laryngeal involvement in 37.5% of patients with pulmonary tuberculosis at autopsy, but today the percentage of involvement is probably much lower. There is no sex predominance. The age of patients with laryngeal tuberculosis used to be between 20 and 40, but is now generally higher.

**Pathology**

The pathway of infection is not known exactly, it is believed that contact with sputum containing tubercle bacilli plays an important role. The possibility of haematogenous or lymphogenous infection has also been suggested (Ormerod, 1939). The infection starts in the subepithelial space with exudation and hyperaemia followed by round cell infiltration. There is an inflammatory reaction of the mucosa and tubercles are found consisting of a granulomatous reaction with Langhans' giant cells, caseation and necrosis. The covering mucosa has an irregular appearance. Eventually confluence of these tubercles leads to necrosis of the overlying epithelium which sloughs and ulcerates. The ulcers are shallow with undermined edges, but there may be infiltration of cartilages, especially that of the epiglottis and the arytenoids. Acid-fast bacilli may be found with special stains, but are not always present.

Tuberculosis is to some extent self-limiting and heals with fibrosis which may result in serious stenosis of the larynx. Sometimes tumour-like swellings are found with reparative
processes called tuberculomata. Occasionally, there may be a diffuse oedematous reaction consistent with an allergic response to the tubercle bacillus.

**Clinical features**

Laryngeal tuberculosis should be suspected in any patient with pulmonary tuberculosis, especially in countries where tuberculosis is still endemic. Pain in the throat and referred earache are common. Cough, often productive, and hoarseness are nearly always present.

In advanced cases with extensive ulceration, the symptoms are very severe. The voice may be reduced to a harsh whisper. The pain and dysphagia can become unbearable. Only rarely is oedema so severe as to cause dyspnoea.

Laryngeal tuberculosis presents in many different forms. Mucosal hyperaemia and oedema are common first signs, often with irregularities of the mucosal surface. When tubercles are formed, granulomatous masses can be seen, ulceration appears later although it is relatively rare (15%, Manni, 1982). All regions of the larynx can be affected but there is a certain predilection for the posterior commissure, the arytenoids and the vocal cords.

**Diagnosis**

Patients with pulmonary tuberculosis should undergo laryngoscopic examination. A chest radiograph is performed to assess pulmonary lesions and a sputum smear for acid-fast bacilli will usually be sufficient to confirm the diagnosis.

Other forms of non-specific laryngitis and scleroma may resemble laryngeal tuberculosis as well as the ulcerative lesions found in lupus vulgaris, syphilis and carcinoma. If there is any doubt a biopsy should be performed. Tuberculosis and a malignant tumour may present simultaneously.

**Treatment**

As a result of improved socio-economic standards and the discovery of several very effective drugs, laryngeal tuberculosis is now rare. The drugs include streptomycin, para-aminosalicylic acid and rifampicin. Usually a combination of these drugs is used for maximum effect. Toxicity is still a problem, and all otologists should be aware of the serious side-effects of streptomycin on the auditory and vestibular organ. As well as these drugs, vocal rest should be advocated. Previously, application of local preparations containing local anaesthetic and astringents such as formaldehyde were advised, but these seem to be of limited benefit compared to chemotherapy.

Historically, galvanocautery was applied and even nerve blocks were performed in the superior laryngeal nerves for intractable pain. The recurrent nerve used to be injected with alcohol to immobilize the cord to promote better healing. These measures have now been abandoned.
The prognosis has altered entirely since the introduction of antituberculous drugs and nowadays a laryngeal infection with adequate treatment will subside within a few weeks, but treatment must be continued over a long period.

**Sarcoidosis**

Sarcoidosis is a chronic idiopathic granulomatous disease, also known as Besnier-Boeck disease. It may affect several organs and the mediastinal lymph nodes are usually involved. Head and neck manifestations are found in 10% of patients of whom only a minor proportion have laryngeal disease (Neel and McDonald, 1982). Symptoms are generally mild notwithstanding extensive tissue involvement. The disease is usually self-limiting. Laryngeal sarcoidosis may be the sole site in 50% of cases affecting the larynx.

**Pathology**

The pathology of laryngeal lesions resembles a non-specific granuloma similar to the lesions in other organs. The granulomata are composed of epithelioid cells with a varying number of lymphocytes and plasma cells. Giant cells with inclusion bodies are few and necrosis or caseation are not found. Later fibrosis and hyalinization and possibly encapsulation by fibrous tissue are more apparent.

**Clinical features and diagnosis**

Usually the patient has a history of hoarseness, dysphagia and dyspnoea. In most cases the main site is the supraglottis. The epiglottis and the false cords are swollen, oedematous and pale, and the rim of the epiglottis is full and rounded. The true cords and the subglottis are only rarely affected. The lesion can progress rapidly and lead to life-threatening airway obstruction. The diagnosis is made by biopsy which reveals the granulomatous nature. Further suspicion is raised by systemic manifestations. When the diagnosis is suspected, confirmation should be obtained by a full physical and laboratory investigation. This may be very difficult, especially if no other organs are involved. In rare instances sarcoidosis can present with a neuritis of the recurrent laryngeal nerve through involvement of cervical or mediastinal nodes. A positive Kveim skin test is highly suggestive but a negative reaction does not exclude the diagnosis. An elevated serum angiotensin converting enzyme (SACE) is found in about 60% of the patients. A gallium-67 scan can be very helpful in localizing enlarged lymph nodes.

**Treatment**

Opinions still differ as to whether sarcoidosis should always be treated. In general, sarcoid is very sensitive to high doses of corticosteroids, however, the recurrence rate is high and many lesions will regress spontaneously. The main indication for treatment for laryngeal sarcoidosis is airway obstruction and, to a lesser degree, severe dysphagia or hoarseness. Steroids may be given systemically or by local application, but their effect remains difficult to estimate. If the airway is seriously compromised a tracheostomy may be necessary and may have to remain in place for many months.
Syphilis

With the improvement in the treatment of syphilis, laryngeal syphilis is now rare. Involvement of the larynx is present in about 5% of cases. A syphilitic infection of the larynx should always be considered whenever an unexplained infection is present.

All stages of this disease can manifest in the larynx. A primary lesion has been described rarely. A small mucosal erosion develops into a typical primary chancre. The secondary stage is pluriform: vesicles and papular lesions often extend into the larynx from the pharyngeal mucosa. The third stage appears after a symptom-free period, sometimes of many years, and is the most important. Granulomata are found in the mucosa and form a gumma. These are characterized by a centre of necrotic amorphous tissue surrounded by an infiltrate of plasma cells and lymphocytes, sometimes with eosinophils and giant cells. There is periarterial infiltration and obliterative endarteritis.

The lesions have a predilection for the anterior parts of the larynx - the epiglottis and the aryepiglottic folds - compared to tuberculosis, which more often lies in the posterior part of the larynx. The mucosa is swollen and infiltrated and later undergoes deep ulceration with central sloughing. Abundant tenacious necrotic tissue reaches and penetrates the cartilage. The vallecula and the lateral pharyngeal wall are also involved. Considerable destruction can be found which, after healing, leaves a bizarre deformation of the larynx.

Clinical features

The presentation of syphilis in the larynx is very similar to other granulomatous laryngeal diseases. Hoarseness and sometimes dysphagia are the primary symptoms. Pain is rare and indicates very rapid destruction of deeper structures. Swelling of the mucosa causes some degree of stridor.

The laryngeal appearances vary widely. Laryngeal syphilis can easily be confused with a malignant tumour or with other chronic granulomatous infections, such as tuberculosis. At one time, the simultaneous occurrence of laryngeal syphilis and a malignant tumour was not rare. Nowadays laryngeal syphilis has become so rare that the diagnosis is usually only suspected after a biopsy has excluded carcinoma, which is so much more frequent.

Very rarely congenital syphilis can affect the larynx in the infant.

Treatment

The treatment of laryngeal syphilis should conform to the normal treatment of syphilis. This usually means prolonged treatment with high doses of penicillin. Local treatment by inhalations may be beneficial by removing necrotic tissue, which must sometimes be carried out to ensure an adequate airway. Local irritants such as tobacco and alcohol should be avoided.
Scleroma of the larynx

Scleroma, better known as rhinoscleroma, is a chronic granulomatous infection caused by *Klebsiella rhinoscleromatis*. The disease was recognized as an inflammatory process by Mikulicz in 1882 who described the characteristic foamy cells which carry his name. Initially it was considered as a lesion of the nose alone, but later, other sites of this infection were described which led to the change of name. The disease occurs worldwide with a low incidence but it is endemic in certain parts of central Europe, North East Africa and Central America. In a fully developed infection the pathology is characterized by Mikulicz cells, Russell bodies and Gram-negative bacteria but, in the initial stages especially, the inflammatory reaction is non-specific and can be difficult to diagnose. Repeated biopsies may be necessary before the diagnosis is made. Other granulomatous infections including tuberculosis, leprosy and granuloma inguinale can give a similar picture with the presence of macrophages resembling Mikulicz cells.

The symptoms and signs are non-specific and, as in many other chronic laryngeal infections, the diagnosis is usually first suspected after the discovery of the characteristic findings in a biopsy specimen. The presence of nasal lesions, which are found in 95% of cases will help to make a diagnosis. Laryngeal involvement is found in 14-80% of the cases. Laryngeal scleroma is rarely isolated (Jay, Green and Lucente, 1985).

Treatment consists of prolonged administration of bactericidal drugs. The spore forming properties of the organism necessitate the combination of an aminoglycoside, such as gentamicin, with an antimetabolite, such as tetracycline. Occasionally, endoscopic removal of granulomatous tissue is necessary to prevent obstruction. Relapse is common and makes close observation for longer periods necessary.

Wegener's granulomatosis

Wegener's granulomatosis is a diffuse systemic disease of unknown cause. It includes a triad of necrotizing granulomatous lesions in the upper and lower respiratory tracts manifesting themselves as a sinusitis or rhinitis, generalized vasculitis involving arteries and veins, and a necrotizing glomerulonephritis (MacKinnon, 1970). Probably about 25% of the patients also develop laryngeal manifestations during the course of the disease. The larynx is rarely the source of primary manifestation (Terent et al, 1980).

The lesion usually lies in the subglottis and may cause laryngeal obstruction. The mucosa is swollen, has a granular appearance, bleeds easily and is sometimes ulcerated.

If untreated Wegener's granulomatosis can be rapidly fatal. Corticosteroids can change the course of the disease if given early. Nowadays there is strong evidence that immunosuppressive drugs, especially cyclophosphamide, are very active against this disease and they are the treatment of first choice. In view of the toxicity of these drugs they should be used with proper precautions and only under medical supervision. Cures lasting up to 10 years have been reported.
**Leprosy**

Lepromatous lesions can be found in the larynx and these resemble the tuberculoid and syphilitic granulomata. The disease, caused by *Mycobacterium leprae* (Hansen's acid-fast bacillus), still holds many secrets with regards to the mode of infection. There are two forms - lepromatous and tuberculoid - both of which can arise in the larynx.

The epiglottis and aryepiglottic folds are most often affected. There is granulomatous swelling, and often ulceration and destruction, primarily in the supraglottic region. The epiglottis may be curled into a hollow rod. The mucosa may be studded with tiny nodules which may also occur in the trachea. Microscopically, the mucosa is thickened and foamy histiocytes are found (Virchow cells).

Modern chemotherapy can alter the otherwise fatal outcome to a certain extent. Dapsone, clofazimine and rifampicine are commonly used. The treatment should be prolonged over many years.

**Mycosis of the larynx**

Fungal infections have become much more common, partly through the widespread use of antibiotics and cytotoxic agents, which may influence the bioequilibrium allowing fungi, some of which are normal saprophytes, to spread. Also, generalized diseases such as diabetes, hypovitaminosis, malnourishment, hepatic disease and disseminated malignant disease predispose to fungal infection. It is likely that these infections will become increasingly important with the increasing incidence of acquired immune deficiency syndrome (AIDS).

Other mycoses have become more common as a result of the increase in worldwide travel, which has spread fungal infections from regions where they are endemic (Lyons, 1966).

An isolated involvement of the larynx is very rare. Contamination of the larynx is usually part of a fungal infection of the aerodigestive tract or of a systemic infection. These infections may be either superficial and limited to the mucosa, or infections spreading deep into all tissues.

The following mycoses can also affect the larynx.

**Candidiasis (moniliasis)**

Laryngeal involvement by *Candida albicans* is usually secondary to candidal infection of the oropharynx or of the lower airways. *Candida albicans* is essentially a saprophyte commonly found in the mouth and pharynx which can assume pathogenic properties under altered circumstances.

Manifestations of candidiasis in the larynx include oedema and erythema of the mucosa, a whitish-grey adherent fibrinous pseudomembrane and superficial ulceration
surrounded by squamous cell hyperplasia. Microscopically the yeast, with its hyphae and pseudohyphae, is easily recognized.

Treatment is primarily directed towards correction of the underlying causes. Drugs containing nystatin or miconazole are given by topical application, as lozenges or as an aerosol.

**Coccidioidomycosis**

This infection, caused by *Coccidioides immitis*, is endemic in certain parts of California, especially the San Joaguin Valley. Primary infections are common and present as mixed respiratory infections. The disseminated form is very rare.

There have been incidental reports of involvement of the larynx presenting as a granulomatous lesion clinically identical with other granulomata such as tuberculosis. The fungus can usually be seen in biopsy specimens. Administration of amphotericin B is the treatment of choice.

**Paracoccidioidomycosis (South American blastomycosis)**

*Paracoccidioides brasiliensis* is the causative organism of this fungal infection, also called South American blastomycosis. It is endemic in Central and South America.

The disease manifests itself with oropharyngeal or skin lesions and bronchopulmonary infections. Laryngeal involvement is quite common.

Ulcerations of the larynx can sometimes lead to strictures. The organism can be identified in a smear or in sputum. Amphotericin B is the treatment of choice.

**Histoplasmosis**

Histoplasmosis is caused by *Histoplasma capsulatum*: it is worldwide and is endemic in certain regions of the USA such as in the valleys of the Ohio and Mississippi rivers. Many people have been infected with *Histoplasma* during a previous respiratory infection leaving pulmonary calcification which can be found on a chest radiograph. There is also a disseminated form attacking organs such as the liver, spleen and bone marrow. Mucosal lesions are described with increasing frequency.

In the larynx the characteristic lesions of a chronic granulomatous infection are indistinguishable from tuberculosis. Oedema, erythema and granulomata are present, but the lesions lie more anteriorly.

It is not always easy to identify *Histoplasma*, especially in the chronic forms. Special staining tests with methenamine-silver or culture in Sabouraud's medium can be helpful. Further confirmation can be obtained by skin tests or complement fixation tests. The clinical course is variable and characterized by exacerbations and remissions.
The most important drug is amphotericin B 30-50 mg, four to six times a day. Careful monitoring is necessary as serious nephrotoxicity may occur. A newer drug, ketoconazole, is less toxic but needs further investigation into its efficacy.

Other mycotic infections

Other fungal infections which have been described in the larynx include: North American blastomycosis caused by Blastomyces dermatitidis, cryptococcosis caused by Cryptococcus neoformans, rhinosporidiosis caused by Rhinosporidium seeberi, and aspergillosis caused by Aspergillus niger. The diagnosis can be made by using fungal staining techniques.

Actinomycosis

Actinomyces israelii is the causative agent of actinomycosis. It is not a true fungus and, according to most pathologists, should be classified in the group of the higher bacteria (Michaels, 1984). Involvement of the larynx is rare (Brandenburg, Frisch and Kirkham, 1978) and is usually secondary to a suppurative cervical lymph node. The characteristic yellow 'sulphur granules' are seen and under the microscope Actinomyces are identified as long slender Gram-positive branching filaments.

Parasitic infections

Parasitic diseases in the larynx are very rare. Leishmaniasis, trichinosis, schistosomiasis and ascariasis are all parasitic infections which can be found in the larynx. The diagnosis is suspected by the general manifestations of the disease, the lesions in the larynx being of the granulomatous type. Detailed descriptions can be found in textbooks of tropical diseases.
Chapter 7: Disorders of the voice

P. H. Damsté

The relation of voice, speech and language

The human voice serves a number of communicative functions, some in connection with speech and some that are not directly related to speech and language: voice is important in several non-verbally communicated messages.

Non-language use of the voice

Many examples of non-verbal use of the voice can be observed in daily life:

(1) A baby attracting attention and inviting care from its mother. This also happens in the adult world: babies of all ages continue to use sounds for this purpose.

(2) Confirming one's identity: a boy on the beach shows himself off to family and playmates, dancing and shouting at the top of his voice. In much the same way a radio or television appearance of a popular celebrity may have more impact because people 'hear his voice' rather than because of what he has to say.

(3) The tone of the voice is more important in singing than the words.

(4) The tone of the voice expresses in attitude such as intimacy, authority, submission, dominance towards the person to whom the message is directed. The quality of the voice gives the background against which the contents of the message must be interpreted. A call of 'be careful' can convey an attitude of genuine solicitude, a reproach or a threat.

(5) The voice, even without words, can express emotions such as grief (weeping), frustration or anger (crying). When a person cries out in anger, pain, indignation or astonishment, the tone of the voice is more important than the words.

(6) The quality of the voice is related to other psychomotor means of communication, such as posture, gait, gestures and facial expressions. A person is characterized by all these psychomotor manifestations which are permanent personality traits. Moreover, they can express transient emotional states.

(7) Voice, like the other non-verbal means of expression, is partly under voluntary control. It is also part of involuntary 'body language'. The messages conveyed in an unintended way by body posture, movements, facial expression and voice play an enormous role in human interactions. The amount of non-verbal communication is grossly underestimated compared to the importance of verbal language, which is usually overrated. C'est le ton qui fait la musique (the tone makes the music) is a French expression meaning that the contents of a message depend on the way in which it is delivered.

(8) Throat and voice complaints are often non-verbal messages of emotional (psychological) disturbance. The consultant must be able to identify them as such.
If loss of voice, a sore throat or the feeling of a lump are not detected as a signal of worry or distress, the patient and the doctor both start off on the wrong foot. When both are reluctant to face the true origin of the discomfort, a psychological game develops. The patient avoids the issue by describing only his 'cold', 'feeling a lump', or 'having no voice'. The doctor avoids the danger zone by statements about 'red mucosa', 'laryngitis' and by prescribing medicine. This is more likely to happen if a more adequate assessment will cost too much time or is simply out of the range of the consultant's abilities.

Thus, even when it is absent, the voice is eloquent in its non-verbal language. Some people unfortunately are deaf to its meaning or pretend not to hear it. This is probably the most frequent cause of error in diagnosis and of failure in the treatment of voice disorders.

**Use of the voice in language**

The linguistic significance of the voice is obvious. Pairs of speech sounds such as p-b, s-z are characterized by the discriminative feature voiceless-voiced. The meaningful coding of language makes use of 'voicing' as one of the distinctive features of speech sounds (phonemes). 'Tie' differs from 'die' only with respect to voicing of the initial apico-alveolar plosive. In whispered speech the distinction is therefore harder to make.

Besides being a contributing feature to articulated speech the voice adds an element of intonation to spoken language. This is the pattern of voice-pitch in the flow of a sentence or phrase. The code for producing meaningful intonation differs in various languages and dialects. The codes apply to spoken phrases as well as to understanding a phrase, for example, the rising tone at the end of a phrase when a question is intended. Obviously the listener must attach the same meaning to a code as the speaker has intended to convey. The intonation or prosody aspect suffers when a voice is very weak or is out of control in any other way. Monotonous speech or unusual intonation of accents is prevalent in neurological voice and speech disorders.

The emotional expression of the voice can sometimes be understood across language barriers, just like the meaning of certain gestures is understood by all mankind. Speech, on the contrary, is only understood by those who are familiar with the particular language environment in which the speaker has grown up. Summing up, it can be said that voice is a natural medium well adapted to communicate emotional content, whereas speech is a cultural medium that is suitable to convey intellectual content. Speech may be used to express feelings but also to hide, disguise or deny them.

**Anatomy and physiology of voice control**

For clarity, the phonatory system can be divided into three levels and a control function:

1. the voice activating air-stream (the respiratory system);
2. the voice generator (the larynx with its vocal folds) which causes the air to vibrate and thus produces the tone;
(3) the voice resonator (the pharyngeal and oral cavity) which selectively transmits some frequency bands (called formants) and weakens others (antiresonances);

(4) the coordinating and controlling function (the central and peripheral nervous system).

The voice activating air stream

During phonation there is a difference in air pressure below and above the glottis. This pressure difference provides the energy that overcomes the resistance of the adducted cords, and causes them to vibrate. In efficient use of the speaking voice the pressure drop is small (the equivalent of 5-10 cm of water pressure) and the air flow is also low (less than 200 mL of air/s). A speaker or singer can learn to exert control over the subglottic pressure, the degree of glottal closure, and the flow of air. These three parameters are, of course, mutually dependent. Too weak closure of the glottis causes a high flow, sometimes audible as a breathy voice with a rush of air. Too strong glottis closure accompanies a high pressure or a low flow, or both. This is audible as a hyperkinetic or croaking voice.

Patterns of breathing

At the end of an expiration when the muscles of the thorax and the abdomen relax, there is a short pause before the onset of inspiration. This is called the expiratory pause. It occurs only when the body and the mind are completely at rest. Under this condition the organism has a low rate of oxygen and carbon dioxide exchange and is not expecting an approaching effort or excitement. The respiratory frequency is low and the displaced volume of air is small. Slight contractions of the diaphragm suffice to meet the demand for air. The only respiratory movements that can be seen are movements of the abdominal wall as it is displaced outward by the descent of the diaphragm.

The respiration at rest as just described can change into a more active form as a consequence of:

(1) physiological adjustment to increase CO₂ production in the tissues when metabolism increases;

(2) emotional anticipation preparing the organism for action. The latter can also occur as a poorly adapted conditioned anxiety response. When it is not followed by increased activity it can lead to neurotic hyperventilation.

Normally the first visible sign of deeper breathing is the outward movement of the flanks that is added to the forward movement of the abdomen. When the large pillars of the diaphragm contract, the dome of the diaphragm is flattened and the lower ribs are pushed outwards. The flanks can also be expanded by active contraction of the external intercostal muscles. This happens when the need for air increases and breathing becomes deeper. Elevation of the ribs widens the lower thoracic aperture increasing the diameter of the thorax; it also assists the movement of the diaphragm to displace more air.
Another very effective inspiratory movement is stretching of the curved vertebral column: when a cervical and lumbar lordosis and a thoracic kyphosis are straightened, the volume of both the abdomen and the thorax is increased. Part of the voice therapy repertoire is correction of body posture: establishing contact with the ground by planting the feet firmly and cancelling a lumbar lordosis by tilting the pelvis backwards. This increases the distance between the insertions of the diaphragm and increases the range of contraction of that powerful muscle.

Generally speaking, the respiratory pattern mounts from abdominal to thoracic and accessory respiratory musculature with increasing alertness or arousal. Strong emotions can give rise to overbreathing. They prepare the body for action by autonomic and endocrine changes, and when the anticipated action is put off (for example, by fear of its consequences) and the hyperventilation continues, too much CO₂ will be washed out of the system. A low CO₂ level in the blood and the tissues thwarts the availability of calcium ions and this can cause various problems. In its acute form it can lead to a regulatory deficit of the circulation and to fainting (collapsing is in itself an extreme withdrawal and flight response; see voice reactions to stress). In a less acute form nervous overbreathing can cause symptoms of the so-called hyperventilation syndrome: lightness in the head, dizziness, headache, irritability, paraesthesia (a tingling feeling in the extremities), muscular spasm of the hands and the face. Hyperventilation is a regular occurrence in voice pathology, for example, in functional dysphonia, when the patient speaks all day long with a great waste of air and in organic paralytic dysphonia, when the patient is unable to close the glottis as a result of vocal cord paralysis on both sides.

**Effect of insufficient control of the airflow on the voice**

A well-controlled voice is produced by relaxed and supple cords caused to vibrate by a moderate stream of air under low pressure. In vocal dysfunction the air pressure and the airstream are not well controlled during phonation: the vocal cords will not be closed fully during phonation and will allow passage to a large airflow. This is observed in indirect laryngoscopy as an oval or triangular glottic opening. It is incorrect to interpret such an image when seen with a laryngoscopic mirror as a paresis of the internal arytenoid muscles: after some instruction for improved voice production the cords are seen to close perfectly, which indicates that the paresis has no organic origin. Incomplete closure of the glottis is often seen when the intrinsic laryngeal musculature is in a state of excessive tension that is inappropriate for effective function. Under laryngostroboscopic observation the vocal folds do not vibrate over their full width; the resulting sound lacks resonance as a consequence of a lack of overtones.

When focal dysfunction is caused by vocal fold closure not being in tune with breath control it is customary to speak of hyperfunction in the case of excessive closure of the glottis and of hypofunction in the case of insufficient closure. In both conditions there is a lack of breath support. This notion is important for voice therapy. It explains how excessive airflow is kept in check during phonation.
Breath support

In respiration at complete rest the expiratory phase is caused entirely by the elastic force of lung tissue that has been stretched during the inspiration and that resumes its neutral position. No extra muscular force is needed to drive the air out, so long as the neutral starting point is not reached.

After a deep inspiration (when preparing to speak or to sign a long phrase) the elastic expiratory force is rather large as a result of the strongly distended condition of the lungs. If this force were allowed to drive the air out through the adducted vocal folds the air pressure would be greatly in excess of that required for good phonation. Therefore the excess pressure and flow are checked by a counter/inspiratory force:

(1) the weight of the abdominal contents, when the individual is standing upright;

(2) a certain tone of the inspiratory musculature - the diaphragm and the external intercostal muscles. The checking activity is strongest at the beginning of phonation and can diminish gradually as the expiration progresses and the stretched tissues approach their natural starting position. When all complementary air has been spent the expiratory muscles may enter into play to drive out the reserve volume of air (Mead, Bouhuys and Proctor, 1968).

The inspiratory 'rein' during phonatory exhalation is called breath support. Most professional speakers and singers are well aware of some form of indirect control of the resonant properties of their voice. Some feel it in the abdomen, others in the sides or the back. Some report that the back of the neck feels like a powerful control centre for the quality of their voice. The following paragraphs explain how the curvature of the neck affects the length and tension of the vocal cords.

The vibrating glottis: the voice generator

The expiratory airstream brings the vocal folds into vibration. The impressive range of intensities, tonal qualities and pitch of the human voice is the result of:

(1) the movements in the cricothyroid articulations which stretch or shorten the vocal ligaments;

(2) the movements of the arytenoid cartilages, each of which is in the centre of intrinsic laryngeal muscles which cause them to rotate and glide over the articulatory surface of the cricoid cartilage;

(3) the tendinous membrane that covers the inner surface of the intrinsic musculature of the vocal folds. It inserts on the inner side of the cricoid and ends on the free margin of the vocal ligament. This membrane has also been called the conus elasticus or the cricovocal membrane.

The coordinated activity of the intrinsic laryngeal muscles causes the vocal folds to take on a firmness, a certain length and a degree of closure (firmly or loosely adducted folds during the production of voice sound). When close together the vocal folds narrow the airway.
- the site is called the glottis. The folds act like a fluttering valve: they are alternately pushed apart by air pressure and sucked together by airstream. The vibratory cycle (consisting of an open and a closed phase) repeats itself in a rapid succession of 80-800 cycles (or more) per second. The closing phase is caused by the Bernoulli effect. When the air speed in the narrowing between the folds is at its highest, the pressure exerted on the walls of the glottis is minimal, giving rise to an abrupt closure. It is this shock wave which, in the frequency of the glottal tone, excites the resonating cavity in rapid succession. The more abrupt or steep the wave, the more harmonic overtones are generated.

In contrast with the chest register, which is the normal mode of vibration for the male speaking voice, the production of a falsetto voice is entirely different. Here the vocal ligaments are stretched to their full length. The thyroarytenoid muscles in the vocal folds are fully relaxed and do not resist the stretching of the vocal cords. The vibrating mass is reduced to the medial rim of the vocal folds because of the tension in the ligaments. Also the folds are not completely closed as a rule; consequently the flow of air is interrupted in a less abrupt way than was the case in chest voice. The resulting pressure wave is smoother and gives rise to only one or two harmonic overtones, which gives the voice a flute-like character.

**The suspension of the larynx**

The way in which people use their voice is largely governed by habit. Although phonatory behaviour is in principle a voluntary activity, it is in part automatic and therefore hard to change. Most of the voice disorders that are seen in the clinic stem from a faulty use of the voice. Changing voice habits is therefore the most important therapy in this chapter on voice disorders. In order to understand how a patient can, with the guidance of a therapist, attain a better control of his voice technique and how this is achieved, the inside of the larynx, and its suspension system must be described. This will enable us to see how the voice generator is linked to the respiratory system. Also the properties of the generator in relation to the volume and shape of the resonating cavities will become clearer.

Because of the firm connection of the larynx to the hyoid bone these structures can be considered as one complex suspended between the base of the skull and the upper aperture of the thorax. It can be moved up and down in front of the cervical vertebrae by long muscles such as the stylohyoid, omohyoid and geniohyoid muscles. From the highest position, as in swallowing, to the lowest, as in yawning, the larynx remains securely fastened to the vertebral column by an ingenious suspension system on which the larynx glides like a sledge over the vertebrae. The carriage consists of a tendinous centre that is held by the constrictor muscles. It extends from the hyoid to the cricoid, and slides over the peristomeum in front of the vertebral column. Short muscles - the middle and lower pharyngeal constrictor muscles, the most caudal of which is the cricopharyngeus muscle - connect the larynx and hyoid to the tendinous sheet. The lower of these muscles has a particular significance in voice control. When the larynx is in a low position it tilts the cricoid forward, thus shortening the vocal folds (detente of the ligament). Together with its antagonist, the cricothyroideus, it controls the delicate balance of vocal cord tension. If the larynx-hyoid complex is held in an elevated position, the direction in which the contracting cricopharyngeus pulls is changed - it will pull in a horizontal or even caudal (downward) direction, and tilt the cricoid backwards. Its effect is then parallel to that of the cricothyroideus and results in stretching of the vocal ligament. This effect of elevating the larynx is observed in people without vocal training who try to
reach high notes when singing or crying. The result is a shrill and thin high tone, because the manoeuvre, apart from stretching the vocal cords, reduces the length of the vocal tract and diminishes the volume of the resonator.

**Influence of respiratory tract on voice generator**

How the muscles immediately related to the larynx and the hyoid affect the length and tension of the vocal cords has been described previously. More distant forces also influence the shape of the glottis and the consistency of the vocal folds. The effect of the contracting diaphragm on the length of the vocal cords is discussed next. When the diaphragm contracts during inspiration, it moves downwards and pulls the bronchial tree with it. The caudally directed force of the trachea (which according to Zenker and Zenker (1960) can be in the order of 1000 g) pulls the anterior part of the cricoid downwards. The anterior part is moved because the main force is applied anterior to the cricothyroid joint, situated on the posterior half of the cricoid. The cricoid is thus tilted forward by the inspiratory force of the diaphragm, and the vocal folds are shortened.

The tracheal pull is maximal when the diaphragm is in the full inspiratory position and when the thorax is wide with expanded and elevated ribs. In this ready-for-phonation position, the external frame in which the larynx is suspended provides complete freedom for the finest adjustment by the intrinsic laryngeal musculature. Messchaert, a great Dutch baritone, described the singer beginning a tone with his chest full of inspired air as feeling that he is, like the tone, light and floating on air.

Understanding the control of the glottis by muscle forces near and far is important for the diagnosis of voice dysfunctions. It has yet another application. Patients who have a bilateral recurrent nerve paralysis with the cords in a paramedian position suffer dyspnoea as a result of high air resistance of the glottis during inspiration. When they stick out the tongue they elevate the hyoid-larynx complex. If, at the same time, they apply abdominal breathing with a well descended diaphragm the vocal folds will be passively abducted (Zenker and Zenker, 1960). Even though the widening of the glottis may be slight, it has been sufficient to help some patients through a difficult period without having to resort to tracheostomy.

**The oropharyngeal cavity: the voice resonator**

When tracheal pull is applied during the inspiratory activity of the diaphragm, it can be combined with contraction of the pretracheal muscles. The effect is one of widening and lengthening of the lower pharynx and this has a considerable effect on the voice sound. When the volume of the pharyngeal cavity increases, lower harmonics are selected from the sound spectrum generated by the vocal fold vibrations. The transmission of lower harmonics is perceived as a full and dark sound. This is called covering of the voice, as contrasted with the open voice.

In producing an open voice the larynx is held in an elevated position (short resonator) and the vowels have a clear ai-like quality as opposed to the dark o-like quality in covered voice.
Untrained singers have a habit of moving the larynx upwards when they attempt to reach high notes. Trained singers, on the contrary, maintain the larynx at practically the same level throughout the entire range of their voice. The covering mechanism is as it were contained within the mechanism for higher pitch. By gradually mixing the colours of the voice registers they avoid any gap or sudden transition between registers. This is attained by keeping the volume of the resonator fairly constant, and by allowing the vibrating mass of the folds to decrease gradually, not suddenly as in transition to the falsetto mode.

Three parts of the oropharyngeal resonator are of special interest: the laryngeal entrance immediately above the glottis; the middle part with the velopharyngeal valve; and the outermost part between the lips. The steep waves of air pressure that emanate from the glottis do not flow out into a wide pharynx immediately, but have to pass the narrowing between the ventricular folds. A rather strong closing of the ventricular folds has been observed especially during the production of clear ringing tones. The situation is not unlike that in the bell-shaped cup of a trombone or other brass instrument. The shock waves are funnelled into a narrow enclosure and thereby reach high pressures than if they had not met this resistance. The ventricles and the interventricular space act as a transformer or a filter of the primary glottic sound which can be modified, for example, by raising or lowering the larynx.

The consistency of the walls of the resonator is important. Firm walls transmit the sound without loss of high frequency components, whereas soft walls absorb parts of the sound energy spectrum. The soft palate or velar valve is a 'soft spot' that can be varied at will. When the velum is firmly closed and the velar musculature firmly contracted, transmission is complete and the sound issuing from the mouth is clear. When the musculature is relaxed or thin (in the case of a congenital insufficiency) a part of the spectrum is filtered out and absorbed by the soft spot. It is not even necessary for the velar porch to be open; when an area as large as that of the soft palate is soft, it works as a filter: low frequencies are comparatively unaffected, frequencies above 1200 Hz disappear from the spectrum. The result is perceived as a hypernasal sound that lacks lustre.

The voice sound generated at the glottal source and transmitted through the resonating tube is finally imparted to the ambient air through the mouth. The vocal intensity (as measured) or the loudness (as perceived) are roughly proportional to the area of the mouth opening. This explains the importance of practising mandibular opening in singing and speaking.

Integration and control by the central nervous system

When a person is actively engaged in oral communication, be in song or speech, the entire body participates in the activity. Not only the intrinsic laryngeal musculature, but also the soft palate, the tongue, the floor of the mouth, the muscles of the neck, the diaphragm, the trunk and the pelvis (for breath support) all take part in expressive phonation. All this is controlled by the central nervous system. Signals arrive at the central nervous system carrying information about the condition of stretch of muscles and ligaments, the position of joints (static signals) and the changes that take place during movement (dynamic signals). Other afferent signals arrive from internal organs such as the mucosal surfaces of the respiratory
tract (Wyke, 1973). The central nervous system integrates the information and sends out signals for the necessary adjustment of muscular tone.

The key to voice therapy is to teach a patient voluntary control over involuntary behaviour. It is therefore important to realize that at the top of the hierarchy, governing the entire process of phonation, is not the central nervous system, but the individual himself. This point of view is sometimes neglected by those whose attention is more narrowly focused on the mechanism of the voice. A patient can normally be held responsible for his use of the voice. If he has temporarily lost control of his voice it is usually as a consequence of some form of stress in facing his environment. Stress is usually not a result of a vocal disorder, but the cause of it. Stress is translated into psychomotor disturbances that affect posture, respiration and voice control. This in turn gives rise to organic changes of the folds caused by faulty use or overloading of the vocal folds.

The examination of patients with voice disorders

History

When taking the history in the case of a voice disorder, the principal complaint is elicited first, in the patient's own words. It is supplemented by questions on the following points: the date of onset (gradual or abrupt), the course, previous treatments; what was the voice like before the trouble began; have there been earlier similar troubles; which activities in the patient's job or free time put demands on the voice? In a complete history all the remaining relevant data required for the diagnosis are assembled. At the very least the following are enquired for the diagnosis are assembled. At the very least the following are enquired about: general health and life-appreciation, respiratory and digestive tracts, cranial nerves (swallowing, hypernasality) and, most important, relationships in the family, at school or in the patient's job. Sataloff (1981, 1984) has summed up points of relevance in the history and examination of professional singers.

Examination

The examination begins during the history taking. An impression is gained of the patient as a communicator by his conduct, facial expressions and eye contact. The opportunity is taken to listen to the sound of the voice and to note its peculiarities. For comparison at a later visit the qualities of the voice should be recorded. A language has to be developed for this purpose, the following metaphors are more or less descriptive. Is the voice:

- low or high
- loud or soft
- powerful or weak
- clear or hoarse, husky
- sharp or dull
- sonorous or thin
- resonant or falsetto
- periodic or raw, harsh
- relaxed or tense, pinched.
It is usual to include in the description an interpretation of the manner of voice production, for example:

hyperkinetic, that is a tense voice with forceful closure of the glottis and high subglottic air pressure;

hypokinetic, that is a voice with little energy, and with air waste.

The posture of the body is noted - tense, relaxed or slouched - and the breathing habit - quiet movements of the abdominal wall or high thoracic breathing, using the accessory muscles of respiration.

Functional assessment

Assessing the voice function by eliciting various kinds of voice production is important. Often the crucial point in phoniatric diagnosis is deciding how far the functional capabilities of the voice go, and to what degree they are limited by an organic factor. Taking a phonetogram is a thorough way of eliciting a large variation of voice sounds. As a preliminary examination, the way in which the patient’s voice alter in response to the following instructions is observed: coughing, phonating while yawning (check whether the larynx is held in a really low position), voice production successively with a relaxed sigh, with a falsetto voice, and in the case of hypofunction during an attempt to produce a sharp, piercing voice. In this way the functional potential of the organ, the extent to which the patient can control his voice, and his readiness or resistance to change his voice can be quickly assessed.

The function of the voice is also evaluated during the examination of the ear, nose and throat. The patient's attention is then distracted from the performance of his own voice. The oral cavity and the pharynx are inspected, and the length of the velum is noted in relation to the depth of the pharyngeal isthmus. The air conductance of both sides of the nose is tested by alternately closing off one of the nasal orifices, the neck is inspected and palpated and indirect laryngoscopy performed.

Indirect laryngoscopy

During indirect laryngoscopy the vocal cords are observed in quiet and deep breathing and while performing the following manoeuvres: producing a low voice, coughing with a short dry cough, producing a high falsetto and a sharp loud voice. Thus, an impression is gained of the motor capabilities of the larynx, and the value of an isolated abnormal phenomenon is reduced to its proper proportion. For example, if the vocal cords do not close completely during phonation, but do close completely on coughing and phonating in a harsh voice, one can conclude that closure of the glottis is possible, that the incomplete closure is of functional (habitual) origin. The result of the inspection is described and illustrated with a simple drawing. An estimate of the length is made and noted (long, short, or cords of average length).
The observed findings are then summed up in a conclusion which relates them to each other and ascribes them to their probable cause(s) - the diagnosis. A phoniatic diagnosis has to account for:

(1) the condition of the vocal folds;

(2) capabilities and limitations of voice function;

(3) contributing factors of constitution and temperament;

(4) factors maintaining the dysfunction: psychological, habitual and environmental factors.

Finally, the diagnosis is discussed with the patient and advice formulated. Whether the choice of therapy be medical, surgical or behavioural, the patient should be helped to make a decision for himself based on the available information. The motivation to begin the treatment determines in part a successful outcome, particularly when the patient is referred for psychotherapy or voice therapy. The referring specialist can influence that motivation.

In most patients it will be possible to arrive at a decision with the method of examination just described. Further instrumental techniques can only supply superfluous information in most cases. They should be omitted from the routine examination, lest they should give the patient a wrong impression that he has a more serious disorder than is actually the case. If the examiner wishes to collect data for scientific reasons, he should tell the patient, and ask his permission. It is morally and ethically wrong to subject the unknowing patient to all sorts of recordings that benefit science and industry more than the patient. With this restriction, some further methods of examination can be used in particular cases.

**Special methods of examination**

**Magnetic tape recording of the voice**

Recording the voice has several advantages (Yanagisawa, Casuccio and Suzuki, 1981; Gould, 1983):

(1) it provides a document for later comparison;

(2) when replayed the examiner can focus all his attention on features of the voice and articulation, and on characteristics in the use of language, without having his attention distracted by conducting conversation;

(3) when replaying the recording in the presence of the patient some features can be discussed which make the patient aware of the nature of the dysfunction and motivate him to accept treatment; at the same time the examiner obtains an idea about the patient's ability to discriminate between the abnormal and desirable qualities of the voice, which is important in prognosis;
(4) the recording allows various forms of acoustic analyses without further burden to the patient.

The phonetogram

The examiner's subjective assessment of the loudness, pitch and quality of the voice can be supplemented by objective measurements. There are user-friendly but costly apparatus on the market that combine a number of acoustic measurements and calculations. A simple sound-intensity meter and a musical instrument which can produce a series of tones in the range between 60 and 1400 Hz will suffice for the examiner with a musical ear. The sound intensity and the pitch can be plotted against each other in a graph, which then shows clearly the range of the voice and its intensity span for all frequencies that have been tested.

The voice quality or timbre is harder to quantify. Attempts to derive a representative index for voice quality from the acoustic signal will yield results in the near future.

In clinical judgement of voice dysfunction data from many sources are combined. One of these sources is laryngostroboscopy.

Stroboscopy

Mirror examination of the vocal folds under intermittent light is an excellent clinical tool for observing details of the epithelium and the deeper structures during phonation. The vibratory motion itself and the different phases of opening and closure of the vocal folds can be seen.

The action of the laryngostroboscope rests on the fact that the vibrations of the vocal cords are periodic, so that the vocal cords return at regular intervals to the same position. If the vocal cords are illuminated in exactly the same phase of vibration by a short burst of light they appear to be standing still. Because of the speed with which the flashes of light follow each other, and because of the afterimage effect of the eyes the vocal cords appear to be stationary. A microfilm signal ensures synchronization of the flashes of light with the vibrations of the vocal cord. A delayed image of the vibrations is produced by slightly reducing the frequency of the flashes of light compared with the vibrations of the vocal cord. The vocal cords are then illuminated in a successively later and later phase of movement by the pulses of light. For a description of the technique the reader is referred to Kitzing (1985).

Stroboscopic examination of the vocal folds is carried out in the same manner as conventional laryngoscopy, using a forehead mirror with an observation hole and a laryngeal mirror. However, the normal light source is replaced by the flashing xenon-tube. An optical system is used for video display and recording of the image and the light is transmitted fro the source to the pharynx by a quartz fibre bundle. Mainly the upper surface of the vocal cords is seen, whereas the lower surface remains out of sight. However, the medial surfaces which are directed towards each other are easily seen, at least during the open phase of the vibration. This is an advantage over conventional laryngoscopy in which the fast vibrations during phonation only allow a blurred view of the medial edges. Small irregularities can then escape the examiner. In stroboscopy it is possible to see accurately where an epithelial abnormality is situated and how it participates in, or impedes, the phonation. In stroboscopic
Laryngoscopy the vibrating part of the vocal folds is sharply defined, and everything which protrudes from their medial surfaces is distinctly observed.

With the help of a well adjusted stroboscope the extremely fast vibratory movement appears as a gentle waving motion.

*The pattern of the wave motion*

In chest or modal register the vocal folds have a soft or elastic consistency. A wave edge appears from the underside, moving the medial surfaces apart. It ebbs away when it reaches the upper surface of the folds. At the same moment the glottic chink has already begun to close from the underside. In the lower tones of the vocal range the folds are soft, and the amplitude of vibration is large. As the pitch is increased the substance of the folds becomes gradually firmer, and the amplitude of vibration smaller. In untrained voices there is a sudden transition towards the falsetto register.

**Diagnosis and treatment**

**Synoptic diagram**

*Table 7.1* displays the various voice disorders. It is divided into four columns: the right columns shows the disorders or primarily organic origin, such as laryngeal paralysis and papilloma. The left column shows the psychogenic and habitual voice disorders; there is no laryngeal pathology, and from an organic point of view the phonatory system is capable of function. The cause of dysfunction is either emotional or habitual. In the former case the voice is inhibited by psychological stress, in the latter case faulty use or overloading of the voice causes the dysfunction. Functional dysphonia of long standing may give rise to organic adaptations to the misuse.

These secondary organic affections of the vocal cords are displayed in the middle columns. These are the consequences of temporary or chronic abuse of the vocal cords and, so long as they have not progressed too far, are still reversible. For this reason they belong to the large class of functional dysphonias, and not to the primary organic dysphonias.

**Psychogenic voice disorders**

Emotionally-conditioned voice disorders are by no means rare and they often demand skilful and time-consuming professional assistance before the patient can resume control over his voice. The voice is the mirror of the soul and in these cases the loss of control of the voice shows the soul to be in disarray. A questionable form of therapy still often used by otolaryngologists consists of suggestive laryngological actions executed with magic and/or authority. If the loss of voice has allowed the patient sufficient time to re-establish mental composure, a suggestive treatment may be appropriate. However, the chances for recurrence are high if the causes of the stress and the resulting disarray are left untreated. In such cases the needs of the patient are best answered by an appropriate form of counselling, for which the patient can be referred to the family physician, a knowledgeable speech therapist or, if applicable a priest. Every specialist should be aware of his limitations; if a laryngologist feels
that he is inexperienced in counselling patients in emotional turmoil, he should help the
patient to find a suitable source for moral support.

In a number of cases the pseudoparalysis is the consequence of a lack of assertiveness.
A person who knows how to hold his own in difficult circumstances is less vulnerable to
psychogenic dysfunctions than a person who lacks self-reliance. A course in assertiveness
training can provide immunization to episodes of aphonia and prevent recurrences.

**Table 7.1 Overview of voice disorders**

<table>
<thead>
<tr>
<th>Functional</th>
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<tr>
<td>Psychogenic (phononeurosis)</td>
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<td>Habitual</td>
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<th>Organic</th>
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<tr>
<td>Secondary organic (phonoponosis)</td>
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<tr>
<td>Primary organic</td>
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</table>

Emotional or psychotraumatic interference with voice control
Improper use of the voice (habitual dysphonia) leads to -->
Overloading, leading to abnormal adaptations of the cords such as:
Congenital web or asymmetry

Anxiety neurosis
Neuromotor disorders (peripheral or central)

Compensation neurosis (reinforced by the effect)
Habitual dysphonia -->
Irritation of the mucosa, recurrent laryngitis
Trauma (also surgical and intubation granuloma)

Psychogenic aphonia or dysphonia
Hypofunction -->
Oedema, nodules (chronic nodular laryngitis)
Cysts, polyps, infections:
common cold, tracheobronchitis, syphilis, systemic
diseases

Spastic dysphonia
Hyperfunction -->
Chronic laryngitis with hyperplasia of epithelium: leucoplakia,
pachydermia (contact ulcer)

*Disorders related to the mutation*
Prolonged mutation
Mutation falsetto
Incomplete mutation -->
Hypoplasia of muscular and connective tissue
Tumour benign/malignant
Endocrine disorders
Congenital weakness (sulcus glottidis).
Voice reactions to stress

A common reaction to frustration and grief is crying. This behaviour can be described as 'agonistic', that is a response to a situation that the individual is incapable of coping with in a more adult way. He or she then resorts to a primitive behavioural pattern of withdrawal or flight: crying solicits pity and help from bystanders, appeases a threatening dominant person and is one way to solve an unbearable situation.

Even more common than crying is responding to frustration and anger by shouting. Both weeping and becoming angry belong to the same family of agonistic behaviours. In contrast to crying which is linked to flight and submission, shouting in anger is linked to fight and aggression.

Shouting and crying are generally felt as socially undesirable or even unacceptable. Many people have been so conditioned by their upbringing that they reject consistently any show of emotions. Such an attitude can easily lead to repression of emotions and inhibition of vocal expression of feelings. The trouble that led to these feelings thus has a minimal chance to be solved, and the source of stress continues to demand attention.

In this vicious circle of bottled-up feelings and unsolved displeasures, conversion symptoms may develop. The whispering voice of psychogenic aphonia is one example. It is of the submissive 'flight' type; hypokinesia prevails and it is prevalent in women. Another form more prevalent in men is vocal stuttering, also called spastic dysphonia. It is a sometimes querulous, sometimes aggressively protesting form of speech with hyperkinetic characteristics. It can have a gradual onset in which the patient feels slight irritation or involuntary contractions in his throat. These eventually give rise to scarcely perceptible interruptions of the voice in the middle of a word, or to an irregular tremor during voiced sounds. As the stress does not improve and the voice disorder only makes it worse, the patient starts a new vicious circle of his own design and possibly with the help of his therapist or consultant.

In his forceful attempts to cope with the vocal inhibitions and interruptions, the patient will strain to push the sound out. New symptoms are added, and the disorder may worsen, as can happen in developmental stuttering: the inadequate attempts to avoid or to overcome the difficulties cause even more secondary symptoms.

The diagnosis is difficult (Stoicheff, 1983). The early stage of spastic dysphonia can be similar to dysarthric voice complaints as seen in degenerative disease of the motor system (amyotrophic lateral sclerosis, multiple sclerosis). Their course is different. The voice dysfunction will persist only if the communicative stress is not solved and the patient's coping abilities are not improved. However, a neurological degeneration will show deterioration over months or years. In case of doubt an audiometric brainstem evoked response will discover an organic process. In the author's cases of spastic dysphonia of psychogenic origin, the audiometric brainstem evoked response has always been normal. A better term would be pseudospastic dysfunction from the spastic dysarthric voice dysfunctions caused by medullary and brain disease.
Guidelines for the treatment of psychogenic dysphonia

(1) Help the patient find out what may have caused him to lose control of his voice.

(2) Assist him/her to clear up misinterpretations of the complaint in the home and family environment. It is better to admit emotional stress as the cause of the voice suffering than to continue the game of 'laryngitis' or 'neurological disorder'.

(3) Prevent unnecessary diagnostic examination, which draws attention to the conversion symptom, promotes somatic fixation, and leads the attention away from the emotional inadequacies which have caused it.

(4) If the events and circumstances that caused the emotional stress have been discovered, the patient should receive support and instruction on how to cope with similar stressful events.

(5) If patients cannot summon up the courage to look into the emotional causes of the failure of their voice function, they need a strengthening of their self-reliance and training of their personal resiliency.

When the person's coping ability is improved, rather than treating the symptom, the symptom will soon disappear by itself.

Habitual dysphonia

When a person in normal circumstances nearly always uses a poor voice, this is termed habitual dysphonia. The quality of the voice has no relation with stressful events and seems to be a habit. Just like an emotionally-conditioned dysphonia it is a learned behaviour which can be changed. However, there are differences. For the correct diagnosis the consultant should obtain answers to several questions. If the following series is answered in the affirmative, the complaint is probably a habitual dysphonia:

(1) has the quality of the voice always been poor?
(2) has the voice problem had a very gradual onset?
(3) is the quality of the voice nearly constant?
(4) has the voice failed repeated after prolonged speaking?

If the following questions are answered with 'yes' the diagnosis is more likely to be psychogenic dysphonia:

(1) before the voice problems began was the voice quality good?
(2) has the change in voice quality arisen abruptly?
(3) is the quality of the voice inconstant, changing with the circumstances?
(4) has the voice failed repeatedly in situations of emotional stress?

There is no sharp division between habitual and psychogenic voice disorders. Anyone can suffer a temporary loss of control of the voice as a result of stressful circumstances, and people with poor voice habits are probably more vulnerable than others.
Perello (1962) has expressed this idea by assigning patients with functional voice disorders a place on the following scale:

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phononeurosis and phonoponosis
<--------------------------------------------------------------->
emotional problem                       misuse and overloading
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Diagnostic analysis can determine the extent to which emotional factors, voice use and voice load are responsible in an individual case. A combination of these factors was indicated by an old term 'phonasthenia'. A typical example was the teacher with a poor voice technique, who loses his voice in the course of the day owing to emotional stress in his occupation. Instead of abiding by this ill-defined term, the therapist should see clearly whether he is working on improving the patient's voice technique and, when he is, bracing weaknesses of personality and improving the patient's skill in communicating with the environment. The two approaches can be applied in combination by the same therapist.

Some patients need exercises and very little counselling. Others are cured by a few counselling sessions and no voice practice at all.

**Disorders of the voice in relation to the mutation**

The voice changes at the start of adolescence, when a boy is between 12 and 15 years of age. A low voice, divided into a chest and a falsetto register is one of the sex characteristics of the human male. It is a result of the growth of the larynx and the vocal folds under the influence of testosterone. Increased production of testosterone by the testicles is in turn initiated by the decrease in gonadotrophin, secreted by the hypophysis. The voice change begins a short time before the growth of the larynx becomes noticeable. After about a year the speaking voice has fallen on the average eight semitones (from around 268 Hz to 173 Hz). These are average values but there are great individual variations. The mutation is not only a change of the tonal range but is especially a change of the sound quality: the light boy's voice gives way to the heavy man's voice.

The popular association of functional mutation disorders with feminity or sexual immaturity is incorrect: cases of intersex and eunuchoidism are extremely rare. They will be mentioned in the section on primarily organic voice disorders. In order to ascertain that a patient is developing normally, the first question can be whether he has started to shave. Some hair under the nose and on the chin and the presence of acne are reliable indications of androgenic hormonal activity. A prominent junction of the thyroid alae (the Adam's apple) indicates sexual maturation of the larynx. If laryngoscopy shows vocal cords of normal length this is another sign that the voice disorder under investigation is not of hypogonadal origin.

The above applies to all three of the common mutational disorders: prolonged mutation, mutation falsetto voice and incomplete mutation. All three are abnormal functional adjustments to the change in size of the vocal folds. The audible characteristics in each are different. In prolonged mutation the heavy and the light register alternate. For every part of a sentence that is spoken in falsetto voice there are a few syllables or words spoken in the chest voice. Sometimes the young man is not aware of the inconsistency in timbre and pitch of his voice which may be one reason why has not achieved control over his new voice.
Inconstancy of the voice also occurs in normal cases of mutation; it is only when the duration extends over 6 months or so and does not show a tendency to decrease, that it is termed a prolonged mutation.

A person with a mutation falsetto voice speaks continuously in a falsetto voice. There may be brief periods, for example during laughing or coughing when the voice drops into a heavy register. This event greatly facilitates the diagnosis because only a mature male larynx can generate separate registers. Hearing the heavy register, even for one short moment, confirms that a normal chest voice can be produced by the larynx and that hypogonadism is out of the question.

The diagnosis is more difficult in cases of incomplete mutation. An important fact drawn from the voice history is that the mutation has proceeded imperceptibly. The sound of the voice is less characteristic than in the previous two conditions. There is no question of split registers, the pitch of the voice is too high, the tone is dull, it lacks the low resounding quality of the modal or heavy register and the origin of the poor voice quality goes back to the time of the mutation.

An abnormal use of the voice during puberty is found in the history of some cases of incomplete mutation. Patients who have continued to sing in a boys' choir long after the pubertal change of the larynx has set in may have impeded a normal transition of the vocal fold tissues to the postadolescent state. Also patients who have suffered from chronic non-specific respiratory diseases such as asthma often have a dysphonia similar to incomplete mutation. Tense breathing habits may have prevented these persons from establishing a pattern of relaxed and well controlled phonation.

The treatment of incomplete mutation is very different from that of prolonged mutation and mutation falsetto voice. In the latter is always possible to elicit a voice sound in normal register, by lower the larynx and pressure on the Adam's apple. The change to a normal voice should be abrupt, not gradual. The duration of treatment can usually be short. In incompletely mutated voices the treatment is aimed at achieving more supple cords: the structure of connective tissue and muscles in the folds must change. This requires intense work on the part of the patient over a period of months or years.

Hypo- and hyperkinetic forms of habitual dysphonia and the resulting organic laryngeal pathology

In many cases of poor vocal habits the delicate balance between subglottic pressure, breath flow and glottal resistance (firmness of closure) is neglected. When the vocal folds are not sufficiently closed and too much air is used, the voice sounds breathy. This is called hypokinetic dysphonia. When the glottis is firmly constricted, the resulting voice sound is harsh or croaking and is called hyperkinetic dysphonia. Sometimes it is hard to distinguish hypokinesis at the level of the glottis, hyperkinesis at the ventricular and pharynx level (constriction) and high thoracic breathing. In that case the issue is left undecided and termed 'dyskinesis'.

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Prolonged use of improper vocal habits may cause the vocal folds to adapt to the strain by forming nodules, oedema and various forms of hyperplasia. The former are seen more in children and women, the latter in men.

Vocal nodules

Vocal cord nodules are a frequent disorder in children and adults. In children and adolescents they consist of spindle-shaped thickenings of the edges of the vocal cords, whereas in adults they constitute more localized thickenings, varying from small points to nodules, typically at the junction of the anterior and middle thirds of the vocal cords and always symmetrically on both cords.

Vocal nodules originate from a combination of overtaxing and incorrect use of the voice (habitual dysphonia). They can be prevented or cured by voice rest or by learning to use the voice properly. So strong is the influence of function on the form of the vocal cords, that the nodules can come and go in a matter of weeks, as has been observed many times in salesmen during a busy season or in women in a period of emotional stress. If the factors which have led to the nodules persist for a long time the nodules become permanent. A preliminary phase of submucous transudation is followed by an ingrowth of vessels, then by fibrous organization. At this stage resumption of normal functions leads to a slow return to normal.

The consistency of the nodules can be observed by laryngoscopy under stroboscopic light. A local oedematous swelling of recent onset vibrates in phase with the whole vocal fold whereas an older and more fibrous swelling can so impede the vibrations that only a part of the cord vibrates. The improvement in the vibratory pattern during restoration of the voice can be followed well by stroboscopy.

The treatment of choice is re-education of the voice by a suitable training programme that motivates the patient to practise in his home and work environment (Boone, 1981). Voice rest can be used in varying degrees. A complete voice rest, intended to be carried out literally, is too difficult for most patients to follow, because they would need to take sick leave and seclude themselves. The nodules will improve or disappear if this treatment is followed, but the question is what will happen as soon as the talking is resumed. The patient is then exposed again to the full burden of his daily life and work, still with his incorrected vocal habits. If they have not been changed in the interim, there is a high chance that the disorder will recur. There are two indications for voice rest: when the cause of the trouble has been a short-lived overtaxing of the voice; and as a preliminary to voice therapy. The patient who must learn new vocal habits is more likely to succeed if he is encouraged to give up his old habits. This is the case if he is freed from any vocal use apart from his exercises at the start of the retraining period. It has the best chance of success in a residential setting where all aspects of personal growth, resistance to stress and voice training are taken into account.

Vocal fold oedema and laryngeal polyps

A thin layer of loose connective tissue separates the epithelium of the folds from the underlying ligament and muscles. The potential space under the epithelium is called the subepithelial space of Reinke. Accumulation of fluid in this space is called vocal fold oedema;
if the accumulation is concentrated at one point and balloons the epithelium out in front of it, this is known as a vocal fold polyp.

Polyps can occur along the whole membranous parts of the vocal folds, but are most common near the anterior commissure. A polyp at that point can be easily overlooked because it is out of sight behind the epiglottis. It can be brought into view by having the patient lower his larynx during the mirror examination by yawning and falsetto phonation. In most cases the cause is unknown; some polyps may have originated from phonating with excessive subglottic air pressure and incompletely closed cords, so that the mucous membrane at the anterior commissure is sucked on and ballooned out. The accumulation of fluid in the subepithelial layer is later followed by the ingrowth of young connective tissue, so that the polyp eventually becomes firm in consistency.

Oedema of the vocal cords usually affects both sides symmetrically. It can be an after effect of acute laryngitis, particularly when the voice has not been spared during the inflammatory phase. In other patients it is the consequence of chronic overtaxing of the voice. Oedema has been seen to develop in professional speakers who were in the habit of speaking with an incompletely closed glottis. The oedema can be explained as an organic compensation, an attempt to close the chin. If this hypothesis is correct, it can be concluded that there is a risk in advising people to whisper in order to save their cords from damage during an inflammation. They can easily acquire a habit of speaking loudly with a breathy voice, while being convinced that in so doing they are sparing their voice. It is better to advise them to speak with a clear voice of low intensity and to avoid noisy places and large gatherings.

The pale appearance of swollen, oedematous vocal folds is unmistakable. It is more difficult to recognize a slight degree of vocal cord oedema. Listening to the voice may give a hint when the range is a semitone deeper than usual and high tones and falsetto tones become almost impossible. Stroboscopic examination shows a floppy wave edge and an enlarged vertical component of the vibratory pattern. A slight oedema will resolve in time; when there is a large colloidal mass beneath the epithelium resorption may take a long time and the patient may prefer to have the swollen mucosa removed, after which healthy epithelium will grow over the bare area. Voice therapy to control the breath flow and ensure sufficient glottal closure is indicated to prevent recurrence.

A vocal fold polyp never resolves and should be removed. Spontaneous cure may occur in a very early stage of a polyp, by rupture of the epithelium and escape of the contents which were not yet organized by connective tissue.

Hyperplasia of the epithelium

Improper use of the voice can play an important role in the inception of a series of epithelial changes, particularly when it coincides with smoking and alcohol intake. These three causes of chronic irritation combine to elicit adaptation reactions that are rather damaging to the voice: inflammation, acanthosis, hyperkeratosis. These conditions are discussed at length in a separate chapter and only the voice aspect will be mentioned here.
When examining the voice field and listening to the voice, it is observed that the patient cannot phonate at a low level of intensity. Because of the thickening of the epithelium, a higher than normal airflow is needed to start vocal fold vibration. The threshold of quiet phonation may lie as high as 60 dB in the phonetogram. The intensity range and the tonal range are limited. The hoarseness improves but does not disappear when posture and breath control are corrected.

Laryngostroboscopic examination is useful to determine the degree and the extent of the thickening and above all to check for malignancy. In the latter case there is absence of vibration of the infiltrated cord. In this way, follow-up with stroboscopic laryngoscopy can prevent unnecessary biopsies, a fact which is naturally important for preserving the voice function.

In principle, all secondary organic affections of the vocal cords, even those with epithelial hyperplasia are reversible if all causative factors are removed. Examples of complete remittance of severe forms of hyperplastic laryngitis have been observed when patients were admitted to the author's clinic, when they stopped smoking and drinking and learned new voice habits. Such a regimen is rarely practised and most patients prefer surgery combined with voice correction.

Voice disorders of primarily organic origin

In the voice problems discussed hitherto, the prime source of the trouble was always an incorrect use of the voice. In the diagnoses discussed here misuse of the voice has not necessarily been a causal factor. A summary of these diagnoses is found in Table 7.1 under the title Primary organic voice disorders.

Vocal cord paralysis

An experienced examiner can suspect a vocal cord paralysis from the sound of the voice. Even if there is no clearly audible air waste, the voice sounds weak and thin, the lower register is lost and chest resonance is absent. This is explained by the difference in levels between the two cords: when the arytenoid of the normal cord adducts it also tilts forward, bringing the cord in a more caudal position than its paralysed counterpart. Thus the vibrating folds do not touch over their full medial surfaces.

During laryngostroboscopic examination a hypotonia of the paralysed cord can be seen: the amplitude of vibration of the affected cord is larger during phonation than that of the healthy side. When the glottis is not fully closed, the paralysed cord flutters like a flag in the wind.

Breathing can be disturbed in a bilateral paralysis with the cords in the paramedian position. The increased airway resistance is most noticeable on inspiration. Particularly rapid inspiration results in inspiratory stridor. It is for this, and other reasons a distressing syndrome (Holinger, 1981). In the opposite situation, with the cords fixed in an intermediate position, the glottis cannot be closed and offers too little resistance to the respiratory air, also during
phonation. If the patient speaks a good deal, the excessive displacement of respiratory air can result in hyperventilation. A decreased tension of carbon dioxide in the blood and tissues leads to peculiar complaints, which the patient seldom connects with his voice problem. These complaints include tingling in the fingers and feet (paraesthesia), feeling light in the head or dizzy, headache, irritability and emotional liability. It is wise to enquire about symptoms of the hyperventilation syndrome before beginning voice exercises.

**Treatment**

In a unilateral paralysis in the intermediate position, the aim of treatment is better glottic closure which can be achieved with exercises that make use of the remaining innervation and that stimulate compensation by adjoining muscles. The exercises consist of short well-controlled expiratory thrusts that bring the vocal folds into vibration. The Bernoulli (suction) effect narrows the glottis. External pressure by two fingers to the side of the larynx can improve the sound. Progress is measured by the duration of phonation. When proper care is taken development of a false vocal cord voice can be prevented.

Surgical methods for improving the voice when exercises have not had the desired effect are discussed in Chapter 10.

A bilateral paralysis in the paramedian position can develop into a situation in which the vocal cords lie closer together, in the median position. The voice improves, but the patient now has shortness of breath on inspiration and audible stridor. It is probable that this is caused by gradual reinnervation of the cricothyroid muscles allowing their adduction effect to come into action. When stridor and shortness of breath become serious there comes a point at which relief must be provided, either by a tracheostomy or by an operation to widen the glottis - lateral fixation of an arytenoid. In borderline cases, exercises for the improvement of breath control can forestall the necessity of an operation. When the glottic resistance is abnormally high, an improvement of the posture can provide just that little extra widening of the glottis which makes satisfactory breathing possible.

After an operation to widen the glottis one should not expect too much of the vocal function, because of the air escape during speaking. Voice exercises now serve to provide clear articulation and to reduce air waste to a minimum to prevent the symptoms of hyperventilation.

**Voice disturbances caused by endocrine dysfunctions**

A child's voice, the voice of an adult woman and that of a man sound different mainly because the sizes of the vocal folds and of the resonant tubes are small, medium and large respectively. The differences between the sexes (sexual dimorphism) are induced by genetic influence in the early embryonic development and are later elaborated by hormonal action during the period of rapid growth of puberty and adolescence. At a more mature age the effect of the sex hormones is more limited. Oestrogens administered to a grown man change the texture of the skin and hair and increase subcutaneous fat deposits on the breast and hips but do not change the timber and pitch to a perceptible degree. Androgens administered to a grown woman have a more marked effect. They cause extra nitrogen uptake and protein synthesis of the organs that harbour the male secondary sex characteristics: the size of the
vocal folds increases changing the voice in timbre and range, hair starts to grow in unusual places and there may be an enlargement of the clitoris. The effects of oestrogens on men and of androgens on women are different in another respect. In the former, feminization effects rapidly disappear after the intake of a hormone-like substances is discontinued. In the latter, the virilization effect on the voice persists.

**Intersexuality**

Aberrant sexual development may be caused by chromosomal abnormalities. XO and XXY are the most frequent abnormal chromosome sets. In the presence of only one X chromosome the individual develops into a girl with recognizable physical characteristics and decreased fertility (Turner' syndrome). The XXY constitution leads to individuals of male appearance who are infertile (Klinefelter's syndrome). During puberty they acquire fat deposits on the breasts and hips, the mutation of the voice is absent or incomplete, and there is little growth of facial hair. In this respect they resemble the eunuchoid syndrome. Testosterone stimulates development in a more virile direction. It has been suggested that the XYY constitution (an extra Y chromosome) contrasts with Klinefelter's syndrome by creating male individuals who are mostly mentally handicapped, over-aggressive and infertile.

Chromosomal abnormalities are comparatively rare and they are not the only cause of aberrant sexual development. The embryo's endocrine glands start to function during intrauterine life. The hormones thus produced play a role in the further development of the reproductive organs. The delicate balance of growth and differentiation can be rudely disturbed by a the relatively large amounts of estrogens produced by the placenta. It is assumed that a male embryo in a very early stage may undergo an influence from placental oestrogens which prevent full masculinization from taking place. The tissues do not respond to androgenic stimulation; this leads to pseudohermaphroditism (the non-virilizing testes syndrome). The baby is born with tests hidden in the labia or in the pelvis, the genitalia resembling those of the female type. The child is often reared as a girl until adolescence, when testosterone production by the hidden tests becomes strong enough to induce male sex characteristics. The voice changes, facial hair starts to grow and interest in girls appears. At that point it has to be admitted that at birth he had been registered with the wrong sex. In these cases a request to change the birth certificate is usually granted.

**Eunuchoidism**

In 1863 the Italian composer Rossini, not long before his death, wrote a 'Petite Messe Solennelle'. More than a century later it is impossible to comply with his advice for the performance: 'Twelve singers of three sexes will suffice: men, women and castratos'. The third sex is no longer trained in our schools of music. In order to enter a career as a castrato singer, boys under the age of puberty underwent an operation which deprived them of their testes. Thus, when at puberty the hypophyseal gonadotrophic and growth hormones were disinhibited and released into the circulation the gonadotrophic hormones did not find target organs that could start the production of testosterone. The growth hormone, however, induced a growth spurt. As a consequence no secondary male sex characteristic developed, but the limbs increased in length by growth uninhibited by the action of testosterone. Hence the tall stature of castratos or eunuchs; others of the third sex amassed large amounts of body fat and grew to extremely heavy proportions. The sexless, unearthly and heavenly quality of their voices
appealed to audiences, as at present many are enchanted by the extraordinary musical possibilities of male alto voices.

The few castratos that still occur are usually caused by accidents. Other causes of testicular hypogonadism are infectious diseases that lead to atrophy of the testes. When this is recognized in time the hormonal defect can be substituted for by the administration of testosterone.

In addition to the testicular form of hypogonadism, there is the (rare) condition of hypophyseal hypogonadism. Klinefelter's syndrome has already been mentioned as a genetic aberration with a deficient output of gonadotrophins, and subsequent hypogonadism and eunuchoidism.

**Sexual orientation and gender identity**

In some conditions of abnormal sexual behaviour there is no evidence of genetic or hormonal abnormality. It is very likely at the least that environmental factors have conditioned the abnormality, possibly in interaction with internal predisposing factors.

Homosexuality does not give rise to any problem about sex characteristics of the voice, because homosexuals, men and women alike, accept their gender and they feel no need to change their appearance. The popular conception that male homosexuals would like to have high-pitched voices was disproved by Lerman and Damsté (1969) who compared the mean fundamental frequency of the speaking voice of 13 homosexuals and 13 heterosexuals.

In trans-sexuals there is a discrepancy between the somatic gender and the gender as subjectively experienced. Some trans-sexuals feel a strong urge to live as a person of the opposite sex, others have the firm conviction that they belong to the sex opposite to that indicated by their bodies. Trans-sexuals may wish to change their physical sex characteristics by surgical and hormonal treatment. In women, the transformation proceeds by surgical removal of the breasts and the uterus and adnexa. Supplementary treatment by testosterone causes facial hair to grow and the voice to change. In men, the surgical treatment consists of removal of the penis and the testes and the construction of an artificial vagina. Treatment by oestrogens develops breasts but will have little influence on the voice. Two courses of action are available to change the male timbre of the voice: surgery, with the hazard of a limited voice range and a poor quality of the voice, and training to attain habitual use of a light mid-register instead of a low chest voice. This can lead to a satisfactory result if the client has the required auditory discrimination ability and the motivation to practise intensively.

**Virilization of the voice in women**

Masculinization can occur in women of all ages. In girls before or at puberty it is called perverse mutation. It is extremely rare and the cause is endogenous - a tumour of the ovaries or the adrenal glands that produces testosterone.

A critical period for a woman's voice begins at the menopause, when periodic ovulation and menstruation end. The hormonal balance shifts and with it come alterations in the body chemistry inducing changes in elastic and collagenous fibres. These physiological changes
ageing processes can be compensated by good use of the voice. If the woman is a singer, she can take precautions that the chest register does not take control of her voice. If she practises her middle register daily in a mezza-voice, she can continue to use her feminine voice into old age.

At the present time women have to be even more on their guard if they want to follow a natural development of the voice after the menopause. Some women are prescribed testosterone at this time of their lives, because it relieves certain disagreeable symptoms related to the climacteric. This treatment always affects the voice sooner or later. The individual sensitivity to androgens varies and so does the sensitivity for noticing the first change in the voice. Some notice a difference after one or two injections or a few weeks of oral administration; others do not complain even though they suffer gross alterations of the voice.

The anabolic steroids are related to the androgenic hormones. They are prescribed in chronic debilitating diseases and in the presence of metastases of ovarian cancer. Virilization by drugs such as these is marked by voice change before other symptoms of virilization appear. These other symptoms, especially hirsutism (hair on the face and legs) are usually prominent and early signs when the virilization is the result of an endogenous cause (the Stein-Leventhal ovarian dysfunction, or ovarian tumours).

It is important that the initial symptoms of voice virilization are recognized, because only then can the drug be stopped in time to prevent more severe damage to the voice. The early signs are so inconspicuous that the patient is often more impressed by a slight change of her voice than the doctor, to whom the voice still seems within the normal range of pitch and quality. Only by careful questioning and listening to the patient's subjective report, can the doctor discriminate between this incipient organic disorder and other more passing organic or functional disorders. One listens to unsteadiness of the timbre: repeated changes between a full resonant and a thin falsetto-like voice sound within one spoken sentence. The difference between the two qualities of the timbre are very slight in the beginning. When virilization progresses a split in normal and falsetto register will become evident. In case of doubt the reaction of the patient to the almost imperceptible change of her voice is the decisive cue: the voice sounds strange to her, it is not under control as it used to be, especially in the high tones. People who sing, professionally or as amateurs, will of course perceive this at an earlier stage than others.

In an advanced stage of voice virilization the lower part of the voice range has a distinct heavy quality like the chest register that is normal for men. Some victims of virilization feel extremely self-conscious about this chest voice and try to avoid the low tones, even during the examination of the voice range.

Most authors agree that the structural change in the vocal folds caused by the virilizing agent cannot be reversed, even over a long period. The organic structure is irreversibly changed and demands a functional compensation. Blending the registers is an important goal of practice. The prognosis depends on the stage to which masculinization has progressed, the age of the patient, and her ability to compensate the damaged function, based on a good musical ear and phonatory control.
Other hormonally-induced voice disorders

Laryngopathia gravidarum is a disorder of pregnancy, a time during which oestrogens are produced in great quantities. These may cause mild oedema of the vocal folds, as in premenstrual hoarseness. The voice is somewhat lower and gruffer than normal. In predisposed women the disorder can assume a more serious form, with redness and oedematosous swelling of the epithelium of the vocal folds, the ventricular bands and the aryepiglottic folds, sometimes with haemorrhage and loss of the epithelium. In another form, crust formation is the most prominent symptom.

The voice disorder can proceed to complete aphonia with stridor and shortness of breath. The inflammatory appearance has a hormonal cause: once the pregnancy is over the disorder usually resolves completely.

_Hypothyroidism_ (decreased function of the thyroid gland) in early childhood leads to dwarfism and mental retardation - cretinism. In later life it causes myxoedema, that is a thickening of the subcutaneous tissue; the patient is sensitive to cold, sluggish and shows little mental initiative; the voice is monotonous, low and dull; speech is slow with laborious articulation. All this is often incorrectly interpreted as normal symptoms of old age. If correctly interpreted, substitution therapy brings a marked improvement. Anginal chest pains, resembling cardiac complaints, may accompany the symptoms and deflect attention away from the true cause.

_Hyperthyroidism_ produces the opposite symptoms: the voice is clear, high and animated, but tends to instability and is quick to tire. Poor coordination between respiratory and laryngeal control may produce hoarseness. Other symptoms of thyrotoxicosis are irritability, anxiety and tenseness.

Thyroid enlargement without toxic symptoms can also cause voice complaints as a result of displacement of the pretracheal muscles, and in serious cases by compression of the trachea. The dysphonia after thyroidectomy, described in the section on vocal cord paralysis, is notorious.

_Congenital abnormalities of the larynx_

When the voice has been abnormal since early childhood a congenital cause should be suspected, particularly when a functional examination shows that the intensity and the range of the voice are limited. An indirect or direct laryngoscopy may be necessary to confirm a suspected congenital may be necessary to confirm a suspected congenital disorder, but this can be too radial, especially in a very young child, if nothing more is gained than confirmed diagnosis of a condition that cannot be corrected. The author recommends this investigation only if there is a reasonable expectation that laryngoscopy will lead to corrective surgery, as in the case of laryngeal web.

Webbing of the anterior commissure considerably reduces the freedom of the vocal folds to vibrate. The voice sounds unusually high and breathy and cannot produce a powerful tone. The treatment is surgery, followed by re-education of the voice.
Another congenital condition is asymmetry of the vocal folds as a result of:

1. unequal length of the cords;
2. unequal mobility of the cords, which can be explained as a congenital paresis;
3. a difference in the level of the cords which can result from either of the above.

Finally two rare conditions which may lead to a mild form of voice handicap should be mentioned. One is the 'sulcus glottidis', a groove along the length of one or both vocal folds, perhaps a hypoplasia of the connective tissue (Itoh et al, 1983). The other is over-elasticity of the vocal cord ligament that is sometimes observed in boys and girls with hyperextensible joints in Ehler-Danlos syndrome. It results in a peculiar low and monotonous voice. These disorders do not usually require surgical treatment. Injection of Teflon suspension may be considered, but should only be carried out by a specialist in phonosurgery.

**Other primary organic voice disorders**

**Senile atrophy**

Women of advanced age may speak in a voice with a lower than normal pitch because of vocal fold oedema or hypothyroidism (Honjo and Isshiki, 1980). In some old men the voice may assume a light timbre and a high pitch. At the same time they lose the ability to phonate in the chest register. The cause of this is probably shrinking of the muscle mass of the cords or stiffening of the vocal membrane and the vocal fold ligaments, so that they lose the suppleness required for the production of the chest register.

It is certainly not a general symptom of old age.

**Cysts of the vocal folds**

Two cysts of importance in phoniatrics are the mucous retention cyst and the epithelial inclusion cyst. Both can result from laryngitis and they can interfere with the voice function to a greater or lesser degree, depending on their site. As this is usually halfway along the membranaceous part of the folds the symptoms can be considerable. They may be visually inconspicuous and may appear as a unilateral nodule or hide under 'monocorditis' (Monday et al, 1983). Small cysts near the edge of a vocal fold can cause a diplophonia; that is a double or interrupted tone, caused by the inequality of the vibrating masses of the two folds. Cysts are removed assuming that the underlying connective tissue is not damaged. After the operation a period of voice therapy is usually necessary to correct habits formed while the cyst was still present.

**Inflammation**

Specific inflammations of the larynx, such as tuberculosis, syphilis and granulomata are dealt with in Chapter 6.
Papillomatosis of the larynx

When dysphonia has been present for some time and shows a gradual progression, laryngoscopy may reveal a warty epithelial mass on one of the cords. The diagnosis of a papilloma is confirmed by histological examination of the specimen, which shows a characteristic arrangement of the cells in this benign epithelioma. In adults, surgical removal of the growth is unlikely to be followed by a recurrence. However, in children these tumours are apt to recur, and multiple papillomata may occur on both vocal cords and the false cords, sometimes extending to the epiglottis and the trachea.

The prognosis improves with age: the chance of extension and recurrence becomes much less with the attainment of adulthood. The quality of the voice should then be reasonable provided that multiple operations have not caused permanent scarring and stenosis. Treatment must therefore ensure that damage to the growing larynx is avoided and that subepithelial layers are not damaged. Training in non-traumatic use of the voice makes sense because continuous trauma caused by incorrect voice use might stimulate the growth of papillomatous tissues.

Trauma to the larynx

Direct trauma to the larynx can be caused by traffic accidents for example the neck can be struck by a projecting part of a car or motor bicycle. A cartilaginous fracture, a ligamentous tear or a rupture of the trachea may occur. After the life-threatening condition has healed the effects on the function of the vocal folds must be considered. Mucosal adhesions can impede the movement and vibration of the vocal folds. In addition, the slightest interference with the cricothyroid or cricoarytenoid joints can influence the regulation of tension of the cords unfavourably, resulting in huskiness of the voice.

The examination presents certain difficulties. Since there are numerous other causes of dysphonia, it may not be easy in a particular case to decide if the symptoms are connected with an accident, especially if this occurred a long time ago. This problem is particularly difficult if there is question of financial compensation.

In testing the vocal function it is important to determine the margin between the available functions on the one hand and to what extent they are being used on the other. The margin can be reduced by retraining of the vocal function.

The prognosis for the voice depends on the findings at indirect laryngoscopy with respect to the position and mobility of the arytenoids, the aperture and closure of the glottis and difference in the levels of both vocal cords. Adhesions can be removed or a cord can be moved to a more favourable position by injection of a suspension of Teflon to improve the voice. On the other hand, unexpected results have been obtained by continued voice exercises. Experience has shown that once one has succeeded in making a vocal fold vibrate in the airstream, regular voice use will bring about a further improvement. The form of the vocal fold is undoubtedly modified by the use of the voice. Some feel that this can be ascribed to the Bernoulli effect, the medially directed sucking force of the airstream by its modelling action, progressively broadening a narrow fold. To produce a tolerable voice with a somewhat rudimentary instrument is a greater art than when the normal cords are present. Re-education
of the voice requires considerable skill on the part of the therapist and tenacity and optimism on the part of the patient.

Stenosis of the larynx in children before the onset of speech gives rise to compensatory voice mechanisms. In the absence of laryngeal voice, these children develop a form of buccal voice, a 'frog's speech'. With the help of movements of the tongue and the bottom of the mouth, air is pressed along the pharyngeal arch which is thus made to vibrate. The sound so produced is only capable of abnormal articulation, but it can, nevertheless, be understood by persons closely associated with the child.

In a number of cases of laryngeal stenosis a satisfactory lumen can be obtained by plastic reconstruction. Where this has not been possible the remaining lumen has been fitted with an acrylic tube which allowed sufficient air to be forced through the larynx to produce voice. In that case, the patient remains dependent on a tracheostomy for respiration.

Cancer of the vocal cords

Hoarseness is so common that despite this early symptom, vocal cord cancer is often ignored for a long time. Also, this disease does not usually strike like a bolt from the blue, but affects mainly smokers for whom a morning cough and break in the voice from collected mucus is nothing unusual. The hoarseness must then have reached a certain degree of seriousness and persistence before it becomes a reason for the patient to see his doctor. It is therefore a good rule that if a patient has been hoarse for 6 weeks he should be examined by a throat specialist. Typically this form of hoarseness is constant (not intermittent as in functional dysphonia), and does not improve but rather progressively worsens. Stroboscopic laryngoscopy may aid in the differential diagnosis from hyperplastic laryngitis: malignant infiltration must be suspected when the affected vocal cord is seen not to vibrate in stroboscopic light. Stroboscopic examination can also be used for follow-up after radiotherapy: if the hoarseness has not resolved and the vibrations of the vocal fold have not returned to normal, it is justifiable to suspect that the tumour has not completely resolved. Radical surgery will have to follow without too much delay.

Rehabilitation of the voice after laryngectomy

Since the prognosis for cancer of the larynx is better than for some other forms of carcinoma, a successful effort at rehabilitation is especially rewarding. Through all the remaining years the patient enjoys an improved quality of life if he has learnt to use a good substitute voice.

The appropriate time to begin speech training after surgery depends on the local condition of the wound and on the general condition of the patient. The wound should be healed without fistula and the mirror should show no trace of fibrin in the pharynx. For his general condition the patient is encouraged to take walks and other forms of exercise. The condition of his (false) teeth is checked. An audiogram is made as a preliminary to voice and speech training.

Informing the patient and his/her spouse is an essential part of the rehabilitation programme. Usually the nature and the consequences of the operation have been discussed
before the operation. The spouse should always be present in such an informative session: the partners can discuss matters between them and what one has missed will have been heard and understood by the other. In the stress of the preoperative period much of what has been said is not remembered. It is a good idea to repeat the information in the more quiet period after the patient has recovered from the surgery. Among the points to be discussed are:

(1) the altered anatomy;

(2) moisturizing and cleaning the stoma;

(3) types of substitute voice;

(4) useful aids and appliances;

(5) the address of the Association of Laryngectomized Patients;

(6) other questions brought up by the patient and his spouse.

Depending on the organization of the clinic, most of the informative sessions will be held by a doctor, a nurse or a speech therapist. The person responsible should be aware that these sessions do more than furnish matter-of-fact information. Counselling after radical surgery includes guiding a person through phases of recovery from a heavy loss. The counsellor will have to cope with periods of depression, anxiety or protest. With his or her help and the cooperation of an understanding partner or relative, the patient will ultimately accept his handicapped condition. Only then will he be ready to concentrate on assignments and exercises for his rehabilitation.

The artificial larynx

After laryngectomy the patient is offered the choice between using an external voice prosthesis or learning oesophageal speech. The third and perhaps best choice is practising oesophageal speech while using an artificial larynx for speech situations in which oesophageal speech cannot yet meet the demands. The artificial voice generators are essentially of two types: the pneumatic, activated by the respiratory air from the tracheostoma, and the electric, driven by batteries. The first has several advantages: it can be used the day after the operation, provided it can be connected to the tracheal cannula; by carefully choosing the material for the vibrating membrane a suitable pitch and quality of the voice can be selected; because the pitch varies with the breath pressure, the fundamental frequency approaches normal speech intonation; it has no battery that can run out; it is cheap. Some patients dislike the polyethylene or metal tube that transmits the sound from the vibrator at the stoma to the mouth, and see this as a disadvantage. Typical examples of pneumatic voice prostheses are the Tokyo artificial larynx and the Memacon DSP 8.

Of the many electric external vibrators that are available most have an adjustable pitch and intensity level. Servox has the option of a manually operated tonal accent device, the use of which requires linguistic as well as other skills. All voice prostheses have the disadvantage that one hand is occupied when speaking. Every year new developments are reported so that
hope for further improvements is entirely justified. Details about training in the use of artificial larynges are discussed by Keith and Darley (1986).

**Oesophageal speech**

Surgeons who performed laryngectomies early in the 20th century reported that, to their surprise, several of their patients sooner or later, developed intelligible speech, but only those patients with no external pharyngeal fistula. In the early days of the operation a pharyngostoma was a frequent complication which interfered with food intake and with development of substitute speech. The mysteries of speech without a larynx have been solved in subsequent years. At the present time every laryngectomized patient should have access to voice rehabilitation immediately after the surgical wound has healed and the feeding tube has been removed.

When a patient is left to his own devices after laryngectomy and he attempts to speak, the result is initially poor. His lips move, but no sounds come out of his mouth - no vowels because there is no voice, and no consonants because there is no airstream with which they can be articulated. After some practice the patient can learn to move the floor of the mouth and the root of the tongue in such a way that some air is displaced, enough to produce short sibilant and plosive sounds. This can lead to pseudo-whispered speech which is not a real whisper because it lacks the sighing sound of the glottis. It is produced with effort and lacks some indispensable voice sounds, so that it is difficult to understand. As has been mentioned under stenosis of the larynx there are people who learned to vibrate the pharyngeal arch with such a mechanism, producing a buccal speech or 'frog's' speech. It is a type of speech which can only be understood by the initiated and there is no reason to encourage its use by laryngectomees. On the contrary, as a sequel to pseudo-whispered speech the patient may develop so-called pharyngeal voice, which is equally unsatisfactory and is a blind alley on the road to oesophageal voice. It is produced by contractions of the pharynx, which is just the thing to be avoided when learning to produce oesophageal speech. The correct manner in which the patient can generate a properly understandable substitute voice, is by taking air into the oesophagus. When, immediately after the intake, the air is returned, it makes the mouth of the oesophagus vibrate resulting in a vowel-like sound, resounding in a wide pharyngeal and oral cavity.

**The initial stage**

When attempting to make his first oesophageal sounds, any manner or manoeuvre by which the patient succeeds is correct for that particular patient. There is no single method that will be suitable for all. There are several possibilities:

1. drawing air into the oesophagus by inhalation; the entrance of the oesophagus should be opened during an inspiration;

2. pumping air into the oesophagus by injection; the tongue and the floor of the mouth compress the air, while the entrance to the oesophagus is relaxed;

3. carbonated water is used to help the patient form his first recognizable words with 'second voice'.
A patient who is well instructed in his rehabilitation will soon find out which method of air intake suits him best. A few experienced laryngectomized speakers take air in by inhalation. Most experienced patients can speak two to five syllables on one intake. Eventually, replenishing the air in the oesophagus is achieved so unobtrusively that it is hardly noticed. The basic sound is usually learned within 1 or 2 days. It takes 2-10 weeks before the patient can use his newly acquired voice outside his own home and the quality further improves over the next year.

### Difficulties encountered in acquiring oesophageal voice

The most frequent problem for beginners is that the oesophageal mouth does not relax at the moment that the air must pass through it on the way in or out. When the air does not get farther than the pharynx, the sound produced is tense, high pitched and of short duration that is unsuitable as a substitute voice. Sometimes a patient succeeds in injecting air into the oesophagus but is unable to return it at will. When more than a fraction of a second elapses between injection of air and phonation, the air is passed on to the stomach and is lost for phonation. The problem is sometimes solved by active relaxation training of the muscles of the neck and jaw: an indirect approach to relax the oesophageal sphincter. In stubborn cases dilatation by an oesophageal bougie and insufflation of air by a catheter can help to overcome the sphincteric contraction reflex.

A diverticulum of the pharyngeal wall can have an adverse influence on the sound of the voice which may be moist and bubbling. Such a diverticulum is best prevented by stitching the pharyngeal wall with the greatest care and ensuring that the patient does not swallow in the first days after operation.

A distracting symptom in some oesophageal speakers is an audible sound made by air passing through the stoma or the tracheostomy tube. It can sometimes be improved by teaching the patient a more controlled method of breathing during speech.

Some patients complain that their voice is too weak to be of any use in other than the quietest environments. This can be caused by flaccidity of the pharyngeal wall at the level of the vibrating structures. If light finger pressure on the skin above the tracheostoma improves the quality of the voice a special neckband that applies permanent pressure can be fitted.

### Measures for restoring other functions

The sense of smell, that is often absent in laryngectomees, can be restored. The patient learns to ventilate the nose by making the same pumping movements as he uses to inject air in the oesophagus, this time with the soft palate lowered. It is attempted as follows: the root of the tongue is moved up and down and one nostril is closed off by a finger. A hissing noise through the other nasal opening betrays that air is moving. Mastery of this skill ensures that the patient can detect noxious gases in his home, can enjoy agreeable smells in the countryside and can enjoy the taste of his food better.

Swimming is not necessarily impossible for laryngectomees who engaged in this sport before the operation. A cannula with an inflatable cuff is inserted in the trachea. An airhose connected to the cannula at one end leads to a snorkel at the other fixed to the head by an
elastic band. After the cuff has been inflated, a thorough check is made so that no air escapes past the cannula when the swimmer breathes out with the neck under water. The snorkeler will have to become used to the larger dead space, which requires him to breathe more deeply, by practice sessions on dry land before actually entering the water. Being able to swim again means much for many laryngectomees: it reduces the sense of being handicapped and enables those for whom many other sports have become impossible to stay in good health.

*A surgical prosthetic method to restore the voice after laryngectomy*

A disadvantage of the oesophageal voice is that the small supply of air that is available for phonation necessitates repeated interruptions of the speech flow for the intake of air. This drawback has been met by a procedure described by Singer, Blom and Hamaker (1981). The wall between the trachea and the upper oesophagus is punctured during endoscopy. A tube of about 4 mm diameter ending in a valve is inserted in the opening. It has dual functions: it keeps the fistula open and it allows air from the trachea to pass into the oesophagus when the patient closes off his stoma and breathes out. The voice that is produced is generated at the pharyngo-oesophageal junction, as is oesophageal voice. The difference is that more air is available for speech. The advantages are that speech is immediately available after operation, and that sustained speech is produced with a fluent quality. Disadvantages are that one hand is occupied during speech and the patient remains dependents on the hospital for replacements of the tube every 3 or 4 months.
Chapter 8: Trauma and stenosis of the larynx

A. G. D. Maran

Acute laryngeal trauma

Epidemiology

There are basically two types of laryngeal trauma - penetrating wounds and blunt injuries. The blunt injuries can be high velocity or low velocity injuries. Penetrating wounds are caused by knives, bullets, wires and agricultural implements. High velocity blunt injuries are usually caused by road traffic accidents or injuries at work. The velocity, however, may be so high that the wound becomes compound. Low velocity blunt injuries rarely become compound and are due to blows with fists and as a result of sports injuries. The sports that are particularly associated with laryngeal injury are snow mobile racing, motor cycle racing, basketball, karate and injuries have even been reported due to contact with golf balls and cricket balls. Reports have also come from the sport of ice hockey where garrotting with the hockey stick is evidently practised in the professional game.

The type of individual who suffers from laryngeal trauma is usually a young male who indulges in sport, is involved in fights, or who drives cars fast and dangerously.

In North America and western Europe, the condition of laryngeal trauma was first associated with road traffic accidents. This was when no seat belts were used or lap-type seat belts were in vogue. Nowadays, the incidence in these countries of laryngeal damage from road traffic accidents is only a fraction of what it was. This is due to the crossover seat belt and the institution of speed limits and other safety features in cars, such as collapsible steering wheels, mirrors etc. In developing countries, however, when driving by a large number of people is a relatively new feature, laryngeal injuries as a result of road traffic accidents present a significant problem to the practising otolaryngologists. Furthermore, in these countries there is an improving delivery of medical care and more patients are rescued from road traffic accidents and removed from the site of the accident to the hospital, where previously they may have died at the roadside.

Biomechanics

Any classification of types of laryngeal injury is an unhelpful exercise unless it is confined to injuries to the supraglottis, the glottis, the subglottis or mixed injuries. Basically, one must consider injuries to the surrounding skeleton of the larynx, that is the hyoid, thyroid, cricoid and tracheal rings, and injuries to the internal soft tissues. Damage to both the skeleton and the soft tissues creates different problems and requires different modes of management.

Penetrating wounds tend to bounce off the more solid pieces of the larynx, that is the supporting skeleton. It would be usual for a penetrating instrument to slide off the thyroid cartilage and penetrate the thyrohyoid membrane superiorly or go between the cricoid and the thyroid inferiorly to penetrate the cricothyroid membrane. Each of these presents different functional problem.
Penetration of the thyrohyoid membrane causes bleeding in the paraglottic space and thus airway obstruction. It does not affect the voice in any way. A little bleeding or oedema will resolve with the normal scavenging macrophage process of the body but, if there is any significant amount of bleeding, then it will not all resorb and will be organized to cause some degree of stenosis of the supraglottis.

This does not happen to such an extent if the cricothyroid membrane is penetrated. The most immediate effect of this will be that air will leave the respiratory tract and will cause some surgical emphysema in the neck. The penetrating wound, however, may be covered with thyroid tissue which may act as a valve. The bleeding may fill the subglottic space causing respiratory obstruction, but it is more likely to run down the trachea through a clean cut and cause coughing.

Low velocity blunt injuries are unlikely to fracture the thyroid or the cricoid, but fractured hyoids are not uncommon in karate and basketball. Again there will be bleeding into the soft tissues of the paraglottic space and, if the ends of the fractured hyoid are in close apposition, then movement during swallowing will cause pain which may require treatment. The patient may also have swelling of the base of the tongue and some dysphagia.

Even though the thyroid and cricoid are not fractured, there may well be bleeding within the paraglottic space and bleeding within Reinke's space on the vocal cords. The interarytenoid space, which must be present to allow gliding and separation of the arytenoids, can fill with blood ultimately causing stenosis, but the problem is usually one of oedema and minimal bleeding rather than oblitative bleeding causing airway damage or later stenosis.

High velocity blunt injuries will fracture the skeleton of the larynx. The fate of the thyroid cartilage depends on its degree of calcification and, thus, on the age of the patient. If the thyroid cartilage is pushed backwards over the cervical spine, then it splays apart. A minimal injury like this with an elastic thyroid cartilage will result in no fracture, but if there is any rigidity in the thyroid cartilage or if the force is great enough, then the cartilage will usually split down the front or down the thyroid prominence. The inherent elasticity of the uncalkified cartilage will allow it to spring back into place, and there may be little damage or there may be disruption of the anterior commissure. The classically described situation of detachment of the tendon of the anterior commissure and the petiole of the epiglottis is hardly ever seen, but is so dramatic that it demands inclusion in any text on laryngeal injury. In this case, the epiglottis falls backwards and the vocal cords literally roll up on themselves towards the arytenoid. Usually in an elastic thyroid with a linear fracture down the prominence, however, there is little in the way of disruption of the anterior commissure, but there will be bleeding into the pre-epiglottic space and posterior displacement of the epiglottis. More important is the fate of the arytenoids. As the thyroid becomes compressed against the cervical spine, the arytenoids are sandwiched. This can result in them being displaced at worst, but at best there will be bleeding into the interarytenoid space and subsequent swelling.

If the thyroid is calcified and is then compressed against the cervical spine, it is unlikely to have enough inherent elasticity to return to its original position. It will, therefore, shatter rather like an egg and there will be loss of the thyroid prominence. There will be similar arytenoid injury as described above.
The cricoid is the most important part of the laryngeal skeleton. It is the only complete ring in the upper or lower respiratory tract. The thyroid, the hyoid and tracheal rings are all U-shaped with soft tissue attachments posteriorly. If the cricoid is disrupted then it will constrict. Even a linear fracture in the cricoid will cause some resorption of cartilage and reduction of the calibre of the airway at the level of the cricoid. This has severe effects on airflow as a consequence of Poiseuille's law relating the airflow to the fourth power of the radius of the airway. This is probably why high tracheostomies have such a deleterious effect on the airway. There is nothing magic about the first ring, but a tracheostomy tube put through an opening made by excision of the first ring will be contiguous with the cricoid cartilage and may result in enough resorption of that cartilage for the cricoid to stenose. It is rare that acute injuries damage the soft tissue within the cricoid but, if a high velocity acute injury damages the integrity of the cricoid cartilage, then there will be a very difficult defect to repair.

The final soft tissue injury from high velocity blunt injuries takes the form of separation of the trachea from the cricoid. This usually results in death at the roadside, but it is quite possible for enough lumen to remain for the patient to breathe long enough to come into hospital. Several tracheal rings can be damaged with this sort of injury and the cricotracheal membrane sheared off.

Pathological consequences of injury

Soft tissue

Any injury to the larynx will result in some oedema of soft tissue. This usually has no permanent effects other than in Reinke's space, where permanent oedema of the vocal cord can result or resolve into a laryngeal polyp.

Far more important is the effect of organized haematoma. This is most marked in the supraglottic space where there is the most scope for expansion of soft tissue and obliteration of the airway.

The interarytenoid area is also a very large potential space with debilitating consequences if organization occurs within the area.

The anterior parts of the vocal cords at the anterior commissure may be detached, but more commonly abrasions of the mucosa here can result in anterior web formation.

The subglottic space in children is very much more important than it is in the adult, in whom subglottic space obliteration narrowing their airway is rare. It is usually the result of disorganization of the surrounding skeleton, especially the cricoid.

Glottic competence can be lost for several reasons. The most common cause is fixation of an arytenoid in an unsatisfactory position, but it can also be made incompetent by resorption of the thyroarytenoid muscle and atrophy of the cord, and also by vocal cord paralysis due to damage to the recurrent laryngeal nerve in subglottic injuries.
Skeletal injuries

The hyoid, the only bone in the respiratory tract, may be fractured and may well heal without the patient knowing anything has happened apart from a few days of discomfort. On rare occasions, the fractured ends form a bursa which results in continual movement of the fractured edges together and this requires excision.

The thyroid cartilage, if fractured, will heal using fibrous tissue and, provided it is in a good position, this is just as satisfactory as wiring or stitching it together. If, however, it is compressed, as in a calcified thyroid cartilage, then it has to be reconstituted and held outwards with a stent.

The effects of disruption of the cricoid cartilage have already been described, and any rehabilitation of this area must involve widening the cricoid cartilage and keeping the edges apart with some material which does not resorb.

At this point, it is pertinent to point out the effect of blood on cartilage. If cartilage is allowed to stay in contact with blood for any length of time, then the blood is absorbed by the cartilage. This is especially important in the trachea where loss of the U-shaped rings perhaps causes no observable abnormality in the airway until the patient takes exercise or a deep breath. The increased velocity of airflow pulls in the weakened tracheal walls and the patient will have dyspnoea on exercise due to tracheomalacia.

If cartilage is left denuded of mucosa and is in contact with secretions, then the surface of the cartilage will become inflamed. This will result in the formation of granulations and is most frequently seen in intubation injuries where the vocal process of the arytenoid is sometimes damaged and an intubation granuloma results. Similarly, if too large an intubation tube is used, then the anterior commissure is split and cartilage becomes bared in that area resulting in an anterior intubation granuloma.

Treatment principles

Protection of the airway

This is obviously the most important feature and is probably the reason that most victims of road traffic accidents are now saved. If there is merely oedema present, with no suggestion of intraluminal bleeding or tracheal damage, then the patient can be kept at bed rest with or without steroids or steam inhalations.

More likely, however, the airway will be at risk and, rather than performing an immediate tracheostomy, the patient should be intubated. In any review of chronic laryngeal stenoses, there is always a hint of criticism in publications that anaesthetists at this point missed an acute laryngeal injury. If anyone has had a neck injury, there will be contusion and perhaps bleeding in the throat and it is quite impossible with the equipment available to him, to recognize intraluminal or skeletal damage to the larynx. Even though an endotracheal tube is not much smaller than many of the stents that are used in the later reconstruction of a larynx, neither they nor stents do anything to stop intraluminal bleeding, especially in the supraglottic area, nor to prevent webs in either the posterior or anterior glottis.
In the first aid situation, a tracheostomy may be needed but this is very much less favoured than immediate intubation.

**Protection of laryngeal function**

Although the larynx has functions related to swallowing closure and effort closure, by far its most important functions are in relation to breathing and speaking. It is these functions that should be protected as far as possible in the management of laryngeal injury. In the assessment of results of treatment, a success with regard to breathing is a patient who does not have to wear a permanent tracheostomy tube and who is able to lead a normal life with no or minimal dyspnoea. On the other hand, if the vocal cord has been damaged, a normal speaking voice is unlikely. Success in this function, therefore, can range from normal voice to audible phonation as opposed to a whisper.

Emphasis has been laid on the importance of preventing bleeding in the laryngeal spaces in the treatment of acute injury to the larynx. In this regard it is appropriate to mention the use of stents. Many stents have been described for use in laryngeal injury, but their role should be isolated to the scaffolding of a reconstituted skeletal structure. They have no part to play in stopping bleeding and the subsequent organization of laryngeal spaces. A much better technique for this is to open the spaces and obliterate them with quilting sutures. If there is any significant degree of bleeding within the larynx, then it should be opened by way of a midline approach (laryngofissure) and the spaces evacuated and quilted with 3-0 Vicryl sutures. Inserting drains into the spaces is quite useless.

If there has been damage to the skeletal structure then a principle of minimal debridement should be practised. There is not very much cartilage in the larynx and excision of any tracheal rings, and certainly of the cricoid cartilage, carries with it grave consequences. Although much of this cartilage may resorb, it is better to cover it with mucosa and see if it forms a scaffold for firm fibrous tissue. The worst that can happen is what one would achieve with debridement.

In general terms, the arytenoid will be swollen in nearly every moderately severe laryngeal trauma and so the patient should be fed with a nasogastric tube to stop inhalation from glottic incompetence certainly for a few days.

**Assessment**

**History**

The most important step in diagnosing an acute laryngeal injury is to be aware of the possibility in every patient who has had trauma to the upper half of the body. Dyspnoea and dysphonia are the main features leading to suspicion, with dysphagia and pain as lesser indicators.

**Examination**

Marks on the neck may or may not be present and their absence does not rule out a fractured larynx, but it makes such a diagnosis unlikely.
Surgical emphysema confined to the neck is almost pathognomonic of a breach in the airway. Loss of landmarks such as a thyroid prominence is also diagnostic. It should be borne in mind that any neck wound carries with it the associated possibilities of damage to the great vessels and to the cervical spine.

**Radiology**

Plain X-rays of the neck are helpful in confirming the presence or absence of air in the soft tissues. Tomography and laryngography are usually impractical in acute injuries.

**Laryngoscopy**

This should be performed in all patients. If ordinary mirror examination is impossible, flexible laryngoscopy may yield valuable information.

**Treatment**

**Penetrating injuries**

Injuries such as those due to knife wounds, wire wounds and wounds from agricultural or industrial implements will only require treatment if there is bleeding into the supraglottic area. Nearly every such case will require to have the larynx opened and the supraglottic area drained and quilted.

Bullet wounds most certainly require exploration with debridement of cartilage, which will probably also be fractured, and exploration of the neck vessels and nerves. Reconstruction will follow the same principles as outlined previously. On occasion the injuries from bullet wounds are so severe that total laryngectomy is necessary.

It is usual for patients with supraglottic injury to end up with a reasonably good voice and no permanent tracheostomy.

**Low velocity blunt injuries**

The majority of these patients do not require open exploration of the larynx, but most will require observation in hospital at least overnight in case of laryngeal oedema and airway obstruction. As well as sports injuries, similar pathological consequences can follow attempted strangulation and the inhalation of fumes during a conflagration. Provided both the airway and the voice are reasonable then these patients can be observed. If either of these functions is disturbed, however, then the larynx should be intubated and perhaps later explored and reconstructed.

Many of these patients will end up with a poor voice if the glottis has been damaged, because there may well be later minor web formation or arthrodesis of an arytenoid, but it is unusual for these patients to require a permanent tracheostomy.
High velocity blunt injuries

About half the patients who have laryngeal injuries as a result of road traffic accidents will require laryngeal exploration and reconstruction. Skeletal damage is repaired by reconstruction usually using stents, and soft tissue injuries are dealt with by reducing bleeding, evacuating spaces and using quilting sutures.

If the cricoid is injured, then primary repair should be attempted. Only when primary repair has failed should one of the many techniques applied to chronic cricoid stenosis be applied.

Separation of the cricotracheal membrane is an unusual injury and one which is dealt with fairly reasonably by dropping the larynx in the neck and freeing the trachea down to the carina, and pulling it upwards for an end-to-end anastomosis, excising and damaged tracheal rings.

Most high velocity blunt injuries will result in combined injuries to the glottis and subglottis. If only the glottis is involved then the results with regard to breathing should be good, but if the subglottis is involved, then the patient faces future surgery for chronic subglottic stenosis.

Chronic laryngeal stenosis

Epidemiology

This section will be confined to chronic laryngeal stenosis in the adult. The condition, if it manifests itself in childhood, is quite different and is considered in Volume 6.

Common causes of chronic laryngeal stenosis in western Europe and the USA are failed treatment or non-recognition of acute trauma, but stenosis is also seen as a complication of tracheostomy, intubation and partial laryngectomy. In Egypt, and other parts of the Middle East, scleroma is probably the most common cause of laryngeal stenosis. Tracheostomy is an operation performed well by nearly every medical practitioner involved in the care of trauma patients in western Europe and North America, but there are still places in the world where tracheostomies can be performed badly, leading to laryngotracheal stenosis. Other systemic diseases, such as Wegener's granuloma, polychondritis, and various types of autoimmune thyroiditis, can also damage the subglottic area resulting in stenosis, but they are rare.

While supraglottic and glottic stenosis do occur, the most common site is the subglottic area. The main cause of this is, therefore, disruption of the supporting skeleton of the cricoid and the tracheal rings. The associated soft tissue narrowing usually reflects the lack of integrity of the supporting structures.

Pathological considerations

Much the same pathological considerations apply to chronic laryngeal stenosis as to the acute injury. The soft tissue damage is due to mucosal loss and adhesions but, most
importantly, to organization of haematoma within the paraglottic, the preepiglottic and the interarytenoid space.

Glottic competence is affected by web formation anteriorly, and posteriorly an arthrodesis of the arytenoid can result in an unsatisfactory position. Furthermore, the recurrent laryngeal nerves, if they are not injured in the initial trauma, stand a very high chance of injury in the ensuing treatment of chronic laryngeal stenosis, and arytenoidectomy or cordopexy almost always forms part of the treatment of chronic laryngeal stenosis.

A factor in chronic laryngeal stenosis that does not, however, apply in the acute injury is that of tissue memory. If a cartilaginous framework has been disrupted, it heals with fibrous tissue, the fibrocytes of which have a directional memory. Thus, merely incising and separating scar tissue will lead to the tissue attempting to replace itself in its original scarred state. Reconstruction must be more sophisticated than incision and replacement. As much scarred tissue as possible should be excised, but the danger of repositioning will be ever present. This is most important in the cricoid where the interruption of the ring causes narrowing. The forces within the cricoid are altered, probably permanently, from this narrowing, and excision of the scarred area and separation of the cricoid ends with support from intervening tissue is probably the single most difficult problem in the management of chronic stenosis.

Excision of scarred soft tissue is not nearly so difficult. Wide excision of scarred tissue is, of course, necessary but grafting with split skin or mucosa usually gives good results. It must be re-emphasized, however, that no amount of satisfactory soft tissue healing will take place if the skeletal framework is disrupted or resumes its scarred altered position.

Stents are useful in supporting a reconstituted laryngeal framework and, to an extent, in separating mucosal surfaces that have been adherent. It bears repetition that stents are of little value in preventing haematoma formation within soft tissue.

Treatment principles

Most patients presenting for treatment for chronic laryngeal stenosis will already have a tracheostomy. They should be warned that the results of treatment of chronic laryngeal stenosis are at best unrewarding and their tracheostomy may be permanent. In the postoperative period with resultant swelling, the patient will almost certainly have to be fed with a nasogastric tube at least for some days. They should also be warned that it is unlikely that they will regain a normal voice, especially if the glottis has been damaged.

There is almost universal dissatisfaction with the surgical treatment of the systemic conditions that cause laryngotracheal stenosis, such as a scleroma, Wegener's granuloma and polychondritis. It is debatable whether these patients should be treated with any surgery other than occasional dilatations.
Assessment

History

The cause of the chronic stenosis is obviously important. If it is a result of an excessively zealous partial laryngectomy, then it is unlikely that enough tissue will be found to augment the lumen of the larynx. Again, if it is a systemic disease that has caused the laryngeal stenosis, it is unlikely that surgery has any place to play in the management. Exceptions to this might be confined segments of scleroma in the advanced fibrotic stage, but this would be a very rare occurrence.

Perhaps the most important communication to establish between the surgeon and the patient is a mutual sense of realism. Both should realize what is and what is not possible with surgery. Both should realize that dynamics of tissue healing can alter any result and this should be taken into account in timing the operation. No attempts should be made to increase the laryngeal lumen until 18 months have passed from the time of the initial injury. Finally, the patient must be quite clear as to what his objectives from surgery will be. He must evaluate how much a good voice means to him and similarly whether he wants to be rid of the tracheostomy tube so much that he is willing to undergo surgery. More minor cases should also realize that the additional scarring of surgery could, in rare instances, result in the patient having a tracheostomy for the rest of his life.

Examination

The surgeon should establish with mirror or flexible endoscopy the extent of the laryngeal, glottic or subglottic stenosis, but this is not always possible and is probably the least important part of the examination. Perhaps the most important part of the physical examination is in the assessment of the length of the neck and, therefore, how much trachea is available for mobilization in the neck without having to go into the mediastinum.

Radiography

The first investigation should be tomography and, in cases where the subglottis cannot be demonstrated, then laryngography should be used. This is perhaps the only place that laryngography now has to play in the investigation of laryngeal disease. Both laryngography and tomography will give a good idea of soft tissue scarring and distortion, but this is better shown by xeroradiography. Computerized tomographic (CT) scanning will give a very much better idea of the state of the laryngeal cartilages and should, if possible, be carried out in all instances.

Endoscopy

This is necessary to establish, as accurately as possible, the extent of laryngeal damage, but it is also useful to ascertain the lower extent of subglottic stenosis and to test for the state of the tracheal cartilages. These have to be examined from as high as possible without creating any splinting and with the anaesthetist blowing high airflows into the lungs using a Venturi system. In this way, tracheomalacia can be assessed.
The state of the arytenoids must be ascertained to see if they are fixed or not and oesophagoscopy should be carried out in every case.

**Treatment**

**Supraglottic stenosis**

There are three choices in the treatment of this condition: first, there is supraglottic laryngectomy; second, a laryngeal widening procedure; and third, laser excision. The author does not think there is any place now for supraglottic laryngectomy in the treatment of this condition. It defies all the basic tenets of the surgery of laryngeal trauma, namely minimal excision. There is usually nothing wrong with the supraglottic skeletal framework and the lesion is nearly always of soft tissue. The choice lies between serial excisions of the soft tissue with the laser and the laryngeal widening operation. Laser excision allows the patient to keep the tracheostomy tube and to evaluate the effect of serial excisions. An alternative is the laryngeal widening procedure where the larynx is opened in the midline and as much as possible of the submucosal scarred tissue removed. The remaining mucosa is stitched back against the laryngeal framework with quilting sutures or areas of scarred tissue are grafted either with skin or buccal mucosa.

**Glottic stenosis**

The anterior glottic web can be dealt with either by laser excision, by repeated endoscopic excision or by external excision and separation of the anterior glottis with a silastic or tantalum keel (McNaught keel). If an external approach is used, then the keel is kept in place for at least 5 weeks. It can then be removed with minimal reopening of the neck wound. The external approach is probably the preferred one when there is also a stenosis of the anterior parts of the false cord but, if the webbing is limited to the glottis, then laser excision or endoscopic removal is probably best in the first instance.

Posterior stenosis of the glottis is more difficult to treat. The glottis consists of roughly 50% cartilage from the medial face and vocal processes of the arytenoids and 50% of membranous vocal cord from the vocal ligament and the attached mucosa and thyroarytenoid muscle. Posterior glottic stenosis lies between the arytenoids. This is usually accompanied by fixation of at least one arytenoid. The stenosis may be excised and the arytenoid separated with a modified keel with silastic stenting on the end of it to keep the posterior glottis open. For this to succeed, both arytenoids must be mobile and capable of achieving glottic competence when the keel is removed. If the arytenoids are not mobile then one should be removed by a laryngofissure and the cord stitched laterally with stenting applied to stop further adhesions.

**Subglottic stenosis**

**Cricoid stenosis**

Enough has already been written about the biomechanics of cricoid stenosis to make it clear how a free graft in this area must work. It must keep the cricoid ring open and, to do this on a permanent basis, it must adhere to the cartilaginous ends. It is unlikely that free
bone or cartilage grafts, taken from ribs, can ever achieve this objective in a satisfactory and regulated manner. Furthermore, it is certain that allografts have no place.

Perhaps the best method is to swing down the body of the hyoid bone on a muscle pedicle of sternohyoid and hope that this, wired into the arch of cricoid, can keep it open. When this is done, the soft tissue scarring must also be removed and replaced with a skin graft and a stent applied either in the form of rolled up silastic above a tracheostomy tube, or in a modified tracheostomy tube. If a Montgomery T-tube is used for this, then the greatest care must be taken to see that it does not crust.

For greater degrees of cricoid stenosis, where the ring cannot realistically be reconstituted, then it is best to remove the cricoid leaving part of the posterior lamina on which sit the arytenoids. The larynx is then dropped and the trachea pulled up and joined to the lower end of the thyroid lamina anteriorly and to the arch of the cricoid posteriorly. This tends to give something of a lump in the back of the immediate subglottic space, but it is a fairly reliable procedure and can usually allow the patient to be extubated.

**Tracheal stenosis**

The more minor degrees of tracheal stenosis are best treated with dilatations. Very often the problem is one of tracheomalacia, rather than true stenosis, and the true stenosis cannot be seen on endoscopy or X-ray. Very often these patients are frustrated by the lack of a medical diagnosis when they know full well that they are dyspnoeic on exertion. If they are only dyspnoeic on exertion, however, they must consider very carefully whether or not to have surgery just because an operation exists to excise the weak area of trachea. This operation will almost certainly damage one or both of the recurrent laryngeal nerves and result in further surgery for vocal cord paralysis. Attempts to strengthen the tracheal wall with marlex mesh or other external devices, although intuitively attractive, are not often successful.

If a tracheal stenosis is severe enough to warrant the wearing of a tracheostomy tube, then it is a relatively easy matter to excise up to 4-5 cm of trachea and to join the trachea on to the cricoid or first tracheal ring.

Freeing the trachea into the mediastinum presents little problem, provided the operator keeps close to the wall of the trachea and does not stray outside the plane of the peritracheal fascia. Pulling the trachea up is easy because it acts as a concertina. The surgeon must remember, however, that the same pull is then applied downwards after the anastomosis. The most important part of this operation is dropping the larynx in the neck. This is done by cutting off the superior cornu of the thyroid cartilage on both sides. This releases the pull of the stylopharyngeus, salpingopharyngeus and palatopharyngeus muscles. The preepiglottic space should be entered by dividing the thyrohyoid membrane and the thyroid cartilage distracted from the hyoid. The middle constrictor should also be removed from the posterior lamina of the thyroid cartilage. There is enough slack in the false cords on the interior part of the larynx to allow several centimetres of displacement.

During this procedure, attempts should be made to find the recurrent laryngeal nerve on either side. If a damaged nerve is found, then it is best to perform a Woodman's operation.
at the time of the initial anastomosis but, if both laryngeal nerves are intact, then it can be expected that any vocal cord paralysis is due to neuropraxia and will recover.

If this manoeuvre is not enough to close the gap of an extensive stenosis, then a procedure, described over 20 years ago by Dr Grillo of Boston, can be utilized. In the UK, it is often called Barclay's procedure and consists of carrying out a right thoracotomy and removing the right mainstem bronchus from the carina, closing the hole at the carina and joining the right mainstem bronchus on to the left mainstem bronchus at a lower level. This gives several more centimeters of length to the trachea and does not result in stenosis further down.

If localized stenosis occurs further down the trachea, then laser excision can be used.

Results

The results from supraglottic stenoses are usually good. It is usually possible to remove the tracheostomy tube and leave the patient with a reasonable voice. Similarly the results from the treatment of glottic stenosis should also be good and it would be a rare event for the patient to have a permanent tracheostomy.

The results of the treatment of subglottic stenosis, however, are universally poor. Although isolated claims of remarkably good results in the treatment of this lesion are made by the occasional surgeon, they cannot be reproduced consistently by experienced laryngologists of long-standing merit. The key to the subglottis is the cricoid, and it does appear that we have not yet found a satisfactory solution to restoring the dynamic elastic forces necessary to preserve the integrity of the only complete ring in the respiratory tract.
Chapter 9: The obstructed airway

P. J. Bradley

Attempts to save man's life from suffocation have been made from ancient times. These early attempts occasionally succeeded but more often failed. The ancient Egyptians, who established the first civilization, were pioneers in solving problems. Two engravings in the Abydos and Sakkarah regions of Egypt (3600 BC) record the performance of a tracheostomy (Shehata, 1981). Homer around 1000 BC reported that Alexander the Great saved the life of one of his soldiers from suffocation, by making an opening in the trachea using the tip of his sword. Hippocrates (460-377 BC) suggested a type of pharyngeal intubation using a straight cannula passed orally to maintain an airway. In the tenth century, Avicenna (980-1037) advocated intubation of the larynx using bent tubes of silver and gold. Tracheostomy during this period was reserved for the hopeless cases.

Towards the end of the 19th century Schrotter (1876) and MacEwen (1878) revived the idea of peroral intubation and with the invention of the laryngoscope by Bozzini (1807) perfected by Kirstein (1895), techniques improved, many lives were saved, and inhalation anaesthesia became a possibility. In 1885 successful intubation of the larynx was performed in the USA by O'Dwyer (1841-1898) in preference to a tracheostomy for laryngeal diphtheria. The manufacture of the endotracheal tubes was modified and improved in the succeeding years, and the original hard metal tubes were replaced by a flexible metal one, introduced on a curved guide wire through the mouth. This was replaced in 1907, by Barthelemy and Dufour of France, by a rubber catheter which could be guided into the trachea by touch. In 1928, Magill, the father of endotracheal anaesthesia, published his experience of blind nasal intubations gained during World War I. After the Second World War, the attitude to intubation was radically altered by the use of muscle relaxants which made intubation relatively easy, quick and atraumatic. The Macintosh laryngoscope, introduced in 1943, has now come into daily and universal use. The added help of electric light and fiberoptic illumination and the Magill's intubation forceps have made laryngeal intubation a simple, safe and routine procedure.

Tracheostomy, derived from two Greek words meaning 'I cut the trachea', has probably been known for about 3,500 years (Frost, 1976). In 1833 Trousseau described 200 tracheostomies in patients with diphtheria. Fifty of these patients survived, and the operation of tracheostomy became a legitimate procedure with wide acceptance. However, it was not until the early 1900s when Chevalier Jackson attempted to standardize indications, techniques, and the instrumentation for tracheostomy, that the procedure became practical and relatively safe (Jackson, 1909). At present the mortality for tracheostomy as the primary operation varies from 0.5 to 3.0%. These variations can be explained by the differing ages and conditions of the patients, as well as by the urgency of the procedure and the skill of the personnel involved.

Clinicians who have to share the airway in the operating room, whether under routine or emergency circumstances, have come to appreciate the need to achieve and maintain a patent airway if the patient is to survive. It therefore behoves us all to have alternative methods available to achieve a patent airway and be willing to use these alternative methods with speed should the method being used fail. Otolaryngologist have, over the years,
accumulated much experience of the grave complications of emergency and long-term laryngeal intubation. The laryngologist functions as the primary source of expertise in the long-term management of the intubated airway and in the treatment and rehabilitation of the complications caused by artificial airways.

**Intubation of the larynx**

**Instruments**

Certain basic instrumentation is required for successful laryngeal intubation. The equipment should always be checked in advance if time and the needs of the patient allow. A source of 100% oxygen and a bag-valve assembly should also be available to ventilate the patient. Tightly-fitting anaesthetic face masks in a variety of sizes should be to hand to help gain initial control of the airway. Suction is essential and a large bore Yankauer tip type suction head is ideal to clear the airway of mucus, blood or vomit. The Macintosh laryngoscope must be available - the handle should be checked for battery integrity - with a variety of fitting blades. A selection of different sized endotracheal tubes should also be available, with a metal malleable stylet which may occasionally be needed to aid with the insertion of the tube. The Magill forceps is often invaluable in directing the endotracheal tube if any difficulties are encountered in finding or exposing the laryngeal inlet. The selection of the appropriate bore of endotracheal tube is based roughly on the patient's age, especially in children. An adult woman will usually accept a tube of internal diameter of 8.5 mm and a man, one of 9.5 mm. It is a mistake to select a small bore tube in the belief that it will be easier to insert. Not only is this usually incorrect, but there will be a large air leak past the tube, together with an increase in the resistance to spontaneous respiration should the latter be desired. On the other hand, an over-large tube inserted with force will damage the vocal cords with possible serious long-term results. The correct length of the endotracheal tube which will be required can be estimated by measuring the distance from the lobe of the ear to the angle of the mouth and doubling it.

**Indications**

**(1) Protection of the trachea from contamination in:**

(a) non-anaesthetized patients with a depressed cough reflex, for example head injury, cerebrovascular accidents, overdoses and some central nervous system diseases;

(b) anaesthetized patients who are likely to vomit or soil the lower airway, for example obstetric patients or patients with intestinal obstruction;

(c) patients at risk of soiling the trachea or lower airway from an operative site, as in head and neck or dental procedures.

**(2) Severe upper airway obstruction.**
Relative

(1) To facilitate controlled ventilation.

(2) To facilitate tracheobronchial toilet.

(3) To maintain a clear airway under difficult circumstances, for example the prone position for neurosurgery.

(4) For diagnostic procedures - angiography or bronchography.

Technique

Careful but rapid preparation of the patient will aid intubation unless the situation dictates immediate action. The occiput is elevated about 4-6 cm off the table with a pad, providing that cervical spine injury is not suspected. The neck should be flexed on the trunk and the head extended on the neck in the 'sniffing position'. Pre-oxygenation of the patient should be performed with a tightly fitting anaesthetic mask and ventilation assisted as necessary by the bag-valve-mask assembly. Sedation or topical anaesthetic both can be used at this stage to decrease patient discomfort.

The head is placed so that the angle between the mouth and the trachea is reduced, until it forms, as near as possible, a straight line. It is a mistake to hyperextend both the head and neck, as this produces misalignment of the mouth and tracheal axes, so making intubation more difficult.

Endotracheal intubation in the hands of experts takes less than 10 seconds, however, the inexperienced may take much longer. Oxygen uptake continues during the period of intubation and the arterial oxygen tension declines. The lungs should therefore be inflated if possible with oxygen up to the moment when the laryngoscope is inserted into the mouth. Similarly, prolonged attempts at intubation must always be interrupted to prevent hypoxaemia.

To achieve the correct degree of extension at the atlanto-occipital joint, the forefinger of the right hand is applied to the patient's hard palate and the upper jaw is pulled towards the operator. The lips can then be pushed away from the teeth by the middle finger and thumb.

The laryngoscope is held in the left hand, and is introduced into the right side of the patient's mouth so that the tip of the blade approaches the midline from the right. The 'Z'-shaped cross-section causes the tongue to be pushed out of the way to the left.

The handle of the laryngoscope is lifted in the direction to which it points. It must not be rotated, as this movement will damage the teeth, gums and the mucous membrane of the pharynx. The uvula is seen at the tip of the blade which is advanced in the midline while elevation of the soft tissues is maintained. The tip will eventually come to lie in the valleculae between the tongue and the epiglottis. Elevation of the root of the tongue will indirectly elevate the posteriorly placed epiglottis, and the laryngeal opening will come into view ready for intubation.
The endotracheal tube is passed between the vocal cords. If the patient is making respiratory movements, the tube should be passed during inspiration when the cords are separated maximally. If the larynx is difficult to see anteriorly, it may be brought into view by an assistant gently pressing the thyroid cartilage posteriorly.

At this stage the cuff of the endotracheal tube should be inflated with air via a syringe until the tracheal leak is just eliminated.

The tube should be fixed to the head with a bandage or tape to prevent displacement.

Finally, the endotracheal tube is attached, as the situation demands, to an Ambu bag, ventilator or anaesthetic circuit.

Oral intubation is the most direct route, requires less time and is indicated in the moribund or apnoeic patient. An awake patient must be cooperative or adequately sedated to allow oral intubation because of the profound stimulation of the throat by direct laryngoscopy. Nasotracheal intubation is useful when the patient is uncooperative or if manipulation of the neck is considered unsafe (Danzl and Thomas, 1980). This technique can be used if the patient has trismus, severe mandibular injuries, cervical spine rigidity, distortion or masses in the oral cavity. The nasotracheal approach has disadvantages in that it demands greater technical expertise, and the patient must have good spontaneous respirations because the tube is guided by breath sounds. The complication rate of this procedure is less than 3%, but includes epistaxis, sinusitis, nasal necrosis, retropharyngeal lacerations and otitis media (Tintinalli and Claffey, 1981). Fibreoptic techniques can be used to aid oral and nasal intubation but are usually reserved for elective intubations because they require more expertise and time as the larynx can be easily obscured. Successful translaryngeal intubation is achieved if the clinician is flexible, comfortable with multiple techniques and is aware of the possible complications and difficulties that may be encountered with each technique used. Emphasis is placed on the expeditious control of the airway in a rapidly deteriorating patient. If the initial approach to intubation is unsuccessful, prompt use of alternative approaches must be instituted (Salem, Mathrubhutham and Bennett, 1976).

**Intubation difficulties**

Difficulties in intubation arise from three sources:

1. errors of techniques (*Table 9.1*);
2. anatomical variations;
3. transient physiological and structural abnormalities.

The incidence of difficult intubation has been estimated to be approximately 1:750 cases in experienced hands (Edems and Sia, 1981). Of these airways with difficulties, 90% can be anticipated and in the remaining 10% the problem is discovered unexpectedly.

Preintubation assessment can often identify high risk patients. Patients with head and neck problems sometimes have a number of factors which would suggest possible intubation
problems. A mass can alter the normal anatomy of the pharynx, larynx or trachea so that the glottis is impossible or extremely difficult to expose by direct laryngoscopy. Oedema and scarring of the airway secondary to radiotherapy or prior to surgery may interfere with the ability to expose the glottis directly. The presence of temporomandibular joint ankylosis, tumour, masseteric spasm or a small mouth with a large tongue will compromise adequate exposure. Lesions that limit neck mobility such as cervical spondylisis, Klippel-Feil syndrome, as well as congenital abnormalities which produce a very anteriorly placed larynx (micrognathia, Treacher Collins and Goldenhaar's syndromes) will make alignment of the laryngeal and oropharyngeal axes difficult (White and Kander, 1975).

Table 9.1 Common errors of orotracheal intubation

<table>
<thead>
<tr>
<th>Step</th>
<th>Error</th>
</tr>
</thead>
<tbody>
<tr>
<td>Positioning</td>
<td>Axes not aligned</td>
</tr>
<tr>
<td>Opening of mouth</td>
<td>Mouth not wide enough</td>
</tr>
<tr>
<td>Insertion of blade</td>
<td>Wrong size or type of blade</td>
</tr>
<tr>
<td></td>
<td>Blade badly positioned</td>
</tr>
<tr>
<td>Exposure of cords</td>
<td>Leverage rather than traction</td>
</tr>
<tr>
<td>Introduction of tube</td>
<td>Obscured line of vision</td>
</tr>
<tr>
<td></td>
<td>Failure to maintain natural curve</td>
</tr>
<tr>
<td></td>
<td>Trachea angulated by traction.</td>
</tr>
</tbody>
</table>

Temporary abnormalities often present a problem in an emergency. These abnormalities include blood, vomit and foreign bodies that interfere with the view of the airway; trismus secondary to head injuries, seizures or drug ingestion; trauma of the mouth or face; a neck injury; and hypoxia caused by shock or drugs in a conscious but uncooperative patient.

Physicians who are likely to encounter emergency situations which require rapid control of the patient's airway should be able to secure an airway with techniques other than the standard oro- or nasotracheal intubation. Alternative techniques include the fibreoptic laryngoscope, guided retrograde transcricoid intubation, oesophageal devices, cricothyroidotomy, tracheostomy and transtracheal jet ventilation (Iserson, Saunders and Kaback, 1985). Many techniques and devices have been used to aid blind intubation including forceps, hooks, catheter, guides and drugs to increase the respiratory flow, and agents to sedate or paralyse the patients. The most frequently used device to facilitate the positioning of the tube during orotracheal intubation is the malleable guide wire. In its most common form the blunt ended wire is used to mould and hold the tube in a pronounced curve. However, this technique has the potential of lacerating the vocal cord or trachea.

More recently, the above techniques have been added to the flex-end tube or trigger tube. The tube is of standard design; the wall contains a wire that allows anterior flexion of the tip by means of a trigger at the proximal end. While internal guides are generally used in orotracheal intubation, external guides are most often employed in nasotracheal intubation. The most common external guide is the Magill forceps, an instrument that allows the distal end to be grasped and be positioned in the vocal cords under direct vision. The clear
polyvinyl chloride (PVC) tracheal tube is commonly used nowadays, and it allows the clinician to see the condensation of expired air on the inside of the tube.

However, at times oral and nasal intubation are difficult to achieve even with the techniques described. The fibreoptic laryngoscope has been used for difficult intubations since it was first described by Davis (1973), who reported it as an alternative to blind nasal intubation. The flexible fibreoptic laryngoscope is small enough to allow the adult endotracheal tube to be passed around the laryngoscope. The laryngoscope acts as an introducer and allows direct exposure of the larynx. The distal 5 cm of the scope can be manoeuvred in an anterior and posterior direction using the controller. Neither neck movement nor spontaneous respiration is required. With nasopharyngoscopes it is now possible to remove secretions which in the past interfered with the identification of structures (Davidson, Bone and Nahum, 1975).

Correct placement of endotracheal tube

In an emergency, and sometimes in difficult elective intubation, the endotracheal tube is occasionally misplaced in the oesophagus. If this situation is not recognized quickly the patient will die. There are very few absolute signs that the tube is in the trachea; in most cases reliance is placed on a good view of the larynx during intubation, the 'feel' of the lungs during bag compression, the presence of reasonable breath sounds, and the appropriate thoracic movements. The restarting of spontaneous respiration with the appearance of bag movements is reassuring. Bronchospasm or pneumothorax can confuse the picture, but the presence of these diagnoses should be accepted only on good evidence. Difficulty in ventilating is more likely to be a consequence of a tube in the oesophagus than because of bronchospasm. The detection of breath sounds is notoriously misleading, as these may be mistaken for the sound of air passing along the oesophagus. If a blue patient goes pink the tube is unlikely to be anywhere but in the trachea. Patients do not die from 'failure to intubate', they die either from failure to stop trying to intubate or from undiagnosed oesophageal intubation. If difficulty is encountered during intubation the airway can nearly always be maintained by an oral airway and a ventilating mask and bag.

Preventing complications

The design of the current plastic disposable endotracheal tubes has greatly reduced the risks of complications compared with the old cuffed, reusable, red-rubber types. The types of injury following endotracheal intubation have been well described (Blanc and Tremblay, 1974). The incidence and severity of mucosal damage correlates with the duration of the intubation. Minimal damage is reported in patients intubated for less than 48 hours. With longer intubation, the incidence of mucosal damage increases especially in the glottic and subglottic areas. Severe late complications of endotracheal intubation include glottic granulomata, laryngotracheal synechiae, vocal cord paralysis and, the most severe, tracheal stenosis. Tracheal stenosis, although uncommon has been reported in at least one in 342 long-term intubated patients (Hawkins and Luxford, 1980).

The risk of complications greatly increases after intubation for more than 48 hours (Johnsen, 1973; Kane et al, 1982). Major recent improvements that have reduced the complications of endotracheal intubation are an improved understanding of the pathogenesis
of intubation injuries and advances in mechanical ventilation, respiratory therapy and endotracheal tube design and care. Prevention of airway injury from prolonged intubation depends on preventing excessive pressure on the airway structures by the cuff, and developing preshaped endotracheal tubes which conform to the airway anatomy. Laryngeal injury from long-term nasotracheal intubation is lower than that of oral intubation, probably as a result of the smaller sized tubes and the greater stability which reduces the frictional forces of the tube in the larynx during positive pressure ventilation.

The incidence of tracheal stenosis produced by cuff-induced lesions has diminished significantly with the development of the high residual volume (high volume, low pressure, high compliance or 'floppy') cuff (Arola, Inberg and Puhakka, 1981). Problems reported with the high residual volume cuff include aspiration around the endotracheal tube, a higher incidence of sore throats as a result of the larger mucosal contact, increased difficulty of intubation because the cuff obstructs the view of the larynx during insertion, obstruction of the endotracheal tube lumen on account of overinflation of the cuff and high peak pressure during coughing, and increased incidence of postintubation stridor after prolonged intubation. Animal studies have shown that ventilation by endotracheal tube with low pressure, high volume cuffs causes different but significant tracheal damage when compared to the old tubes with high pressure, low volume cuffs (Sanada, Kojima and Fonkalsrud, 1982).

The advent of the CO₂ laser microlaryngeal surgery has made it imperative to protect the plastic intubation tubes from laser-induced ignition. Wrapping the tube in metal tape will protect the plastic sufficiently, as long as it is applied correctly. Even with these precautions lasers have caused fires by ignition of dry protective material, loose metal wrapping or inadequate wrapping of the endotracheal tube (Hirshmann and Smith, 1980).

Hoarseness after an operation is relatively common and has been reported in as many as 70% of patients after operation (Stauffer, Olson and Petty, 1981; Gleeson and Fourcin, 1983). Typically the hoarseness disappears after a few days and neither the patient nor the surgeon remains concerned about the quality of the voice. However, in some cases hoarseness or even aphonia persists associated with pain on swallowing. The following factors contribute to the development of postoperative hoarseness (Jaffe, 1972):

(1) the act of intubation;

(2) the endotracheal tube during the operation;

(3) the indwelling endotracheal tube after operation;

(4) intubation and concomitant bronchitis or bronchopneumonia;

(5) an allergic reaction involving the larynx;

(6) any operation in the neck or upper thorax.

Other symptoms may include sore throat, cough, sputum production and haemoptysis in descending frequencies ranging from 40% to 10% of patients studied. The incidence of
granuloma varies from one in 800 to one in 30 as reported in the literature, but most complications resolve spontaneously (Stauffer, Olson and Petty, 1981).

One-quarter of patients ventilated and intubated in intensive care units require ventilatory support for more than one week, and 10% require aid for more than 2 weeks. Any inflatable cuff, no matter how soft, is potentially hazardous when confined within the tracheal lumen without a safety mechanism. Pressure control is essential, preferably by some means that does not require much attention. Nasotracheal intubation is generally better tolerated than the orotracheal route and can usually be maintained for 3-4 weeks.

Laryngotomy/cricothyroidotomy

In an emergency, failure to clear or secure the airway with an endotracheal tube may result in the death of the patient. Entry into the airway can be achieved rapidly through the cricothyroid membrane because it is superficial and an easily identifiable landmark (Roven and Clapham, 1983).

Indications

Emergency

An emergency arises when an obstructed airway cannot be secured through the laryngeal route for whatever reason.

Elective

Cricothyroidotomy has been condemned because of the high incidence of subglottic stenosis. In the presence of laryngeal pathology and/or prolonged intubation the incidence of subglottic stenosis is high and cricothyroidotomy as an elective procedure should not be performed (Brantigan and Grow, 1982).

Techniques

There are three techniques for laryngotomy/cricothyroidotomy:

(1) using an intravenous catheter;

(2) using a cricothyrotome;

(3) a formal surgical procedure.

The patient's head is placed in the extreme extended position by placing a pillow beneath the shoulders to provide better exposure. In an emergency, a 14-gauge intravenous needle can be inserted, through the cricothyroid membrane, into the trachea and is aspirated until air is returned to signify correct position. A catheter is then directed 45° caudally and advanced over the needle into the trachea. The cricothyrotome is used in a similar manner but it is usually not readily available. Sometimes a skin incision may need to be performed to make insertion easier. In the formal surgical procedure, the instruments needed are a scalpel,
an artery forceps and an endotracheal or tracheostomy tube. The operator steadies the thyroid cartilage with the thumb and middle finger using the non-dominant hand and the cricothyroid space is identified with the index finger. The scalpel should be inserted perpendicularly through the cricothyroid membrane. A stab and twist movement without reaching the posterior cricothyroid ring gains access to the airway. With the non-dominant hand the artery forceps is slightly opened and passed around the scalpel blade into the airway to widen the hole. The scalpel is exchanged for an endotracheal tube or a tracheostomy tube. If the cricothyroid membrane is cut transversely and parallel to the tracheal rings more bleeding can be expected because of the veins crossing this area.

After a surgical cricothyroidotomy when an endotracheal or tracheostomy tube has been placed, ventilation can be managed in the normal way using an Ambu bag and an adaptor. With the intravenous catheter or cricothyrotome with Luer connection two immediate problems arise - connection to an inflation device, and producing adequate gas flow through a small bore tube with high resistance. In the casualty or resuscitation environment several methods are available: a 3 mm endotracheal tube using a 3 mL Luer lock; a syringe with a 7 mm endotracheal tube adaptor inserted into the barrel; a 10 mL syringe with the endotracheal tube inserted into the barrel with the cuff inflated. Enough oxygen is then supplied for tissue oxygenation but CO₂ removal is inefficient. If the minute ventilation is inadequate, the surgeon can at least 'buy time' for the performance of a surgical cricothyroidotomy or tracheostomy. The resistance to flow through the small bore cannula can be overcome by a high pressure oxygen source (at least 400 kPa). Flow must be intermittent and allow adequate time for exhalation. The Jet-vent or a high pressure flow system is needed to overcome these problems. An obvious hazard is hyperinflation with high intrathoracic pressures, possibly leading to cardiac decompensation or pneumothorax. High-frequency jet ventilation is the safest way to solve the problems of hyperinflation and cardiac decompensation.

Once the emergency has been controlled, the clinician can convert to a translaryngeal tube or to a tracheostomy. The long-term management plan should be specific to the patient's needs (Weymuller and Cummings, 1982). Severe laryngeal injury should mandate a tracheostomy, whereas in an obstructed laryngeal lesion the blockage may be removable endoscopically.

Complications

Complications may occur during catheter placement or during ventilation. Haemorrhage may occur from an artery or vein during placement (McGill, Clinton and Ruiz, 1982). The main problems with ventilation are subcutaneous emphysema - if the catheter slips out of the trachea - and hyperinflation. Hyperinflation occurs when laryngeal obstruction limits expiration so that the chest becomes more expansive with each inflation. Subglottic stenosis may follow cricothyroidotomy, although its true incidence is difficult to assess. It is probably uncommon unless prolonged intubation in assisted ventilation is needed (Sise et al, 1984).

A 'minitracheostomy' or cricothyroidotomy has been suggested for patients who are at risk of sputum retention (Matthews and Hopkinson, 1984), which is a major cause of morbidity and mortality following thoracic surgery and in the chronic bronchitis patient. It
has been suggested that the use of the minitracheostomy allows permanent access to the trachea for suction, while avoiding the disadvantages of tracheostomy or endotracheal intubation. The minitracheostomy preserves the function of the glottis. Patients therefore retain an explosive cough with minimal loss of expiratory air volume. However, in a recent long-term follow-up of such patients (Gleeson et al, 1984), up to 75% had subjective and objective voice changes following minitracheostomy.

**Tracheostomy**

**Indications**

Tracheostomy may be performed for any of three basic reasons:

1. ventilatory insufficiency as a result of secretions;
2. mechanical respiratory insufficiency;
3. upper airway obstruction - real or anticipated.

Ventilatory insufficiency may complicate many medical conditions including infections, congestive heart failure, pulmonary oedema, chronic lung disease, or bulbar disease secondary to cerebrovascular insufficiency complicated by pneumonia. Secretory ventilatory insufficiency is now the most common indication for tracheostomy. Acute respiratory failure requiring tracheostomy may occur with a variety of conditions, including drug intoxication, head and chest trauma, elective surgery, neuroparalytic disorders and chronic obstructive pulmonary disease. In these cases requiring long-term treatment, tracheostomy may provide the easiest means of providing ventilatory assistance; it eliminates upper respiratory 'dead-space' and allows frequent and accurate pulmonary aspiration and toilet. Upper airway obstruction is now the least common indication for tracheostomy. Infectious processes and cancer of the larynx and hypopharynx, foreign bodies of the trachea or larynx, subglottic oedema from any cause, and occasionally infections of the oropharynx may require a tracheostomy. Laryngeal trauma is also an indication for immediate tracheostomy.

In general, once the tracheostomy has been performed the underlying disease is no longer an immediate threat to the airway. Since Moser's dictum that 'the time to do a tracheostomy is when you first think about it', it has been well known that in upper airway obstruction, the operation should be performed if the patient has stridor at rest, is restless, cannot lie flat, is using accessory muscles of respiration or has a rising pulse rate. A tracheostomy is indicated in patients with respiratory failure if serial measurements of the vital capacity fall to 25% of normal value. The length of time to wait after oroendotracheal intubation before performing tracheostomy is controversial. In general with the new low pressure cuffed endotracheal tubes most clinicians would currently recommend a wait of 3 weeks. In contrast if prolonged intubation is expected, an early tracheostomy may be preferable.
Surgical technique

All tracheostomies should be performed in the operating room if possible. The procedure should be carried out under sterile conditions. If the tracheostomy is carried out with adequate preparation, meticulous surgical technique and excellent postoperative care, it is safe and reliable. The procedure can be performed under local anaesthetic, but if the patient's condition does not preclude an endotracheal intubation the procedure should be performed in an orderly controlled environment.

Exposing the trachea requires a skin incision between the lower border of the cricoid cartilage and the suprasternal notch. Excellent rapid exposure is obtained through a vertical midline incision. In an elective situation, a horizontal incision gives a better cosmetic result. Under emergency circumstances, cosmesis becomes a lesser consideration when rapid control of the airway is needed. One exception is the patient admitted with stridor secondary to laryngeal tumour who should have a high tracheostomy even if this involves going through the tumour (Stell, 1973). The tumour should then be removed as quickly as possible as the tracheostomy may become the site of tumour recurrence. After the skin has been opened the strap muscles can be separated by blunt dissection in a vertical plane through the linea alba. Dissection through this area should be relatively bloodless, although communicating venous channels may be encountered. All bleeding should be controlled at entry as identification of bleeding areas can be impossible once the tracheostomy is placed in position. The isthmus of the thyroid gland should be clamped with artery forceps, transected in the midline, and transfixed, exposing the trachea. The thyroid gland must be divided in the midline, as deviation from this can result in profuse haemorrhage and even recurrent laryngeal nerve damage. The technique of retracting the thyroid isthmus superiorly or inferiorly may be quicker, but the risk of airway obstruction during early tube change is real and this procedure should be avoided unless the surgeon carries out the first tube change.

The opening into the trachea should lie at the level of the second to the fourth tracheal ring to avoid damage to the cricoid cartilage which can result in subglottic stenosis. The tracheal incision should be of the type that least disturbs the tracheal anatomy (Bryant et al, 1978). The vertical incision is more than adequate for this purpose. Some surgeons recommend a superiorly or inferiorly based flap (Bjork) or even the removal of a segment of cartilage; these procedures are advised to facilitate retention and changing of the tube. However, the disadvantages of these procedures include tracheal stenosis, delayed healing after removal of the tube with the formation of a tracheocutaneous fistula which frequently requires formal closure. In the patient with a short fat neck the use of the cricoid hook to elevate and stabilize the trachea can be invaluable in making the tracheal incision. If the trachea is soft and malleable as in the young the insertion of laterally based stay sutures at the tracheal opening can be helpful and aid with changing the tube later. In the elderly, it may occasionally be necessary to use the Mayo scissors or even bone cutting forceps to open into the tracheal lumen. If the tracheal cartilages are found to be calcified it is better to remove an adequate portion of anterior tracheal wall to allow easy entry of the tracheostomy tube rather than outfracture the tracheal rings.

The tracheostomy tube should have been previously selected and checked for leaks in the cuff before insertion. In general, tubes of size 33 or 36F are suitable for women and tubes of 36-39F for men. Once the tube has been inserted into the trachea the obturator is removed
immediately, and the blood and mucus aspirated from the lumen. The wound must not be closed too tightly at the end of the operation, as this can lead to rapid development of alarming surgical emphysema of the neck. The flanges of the tracheostomy tube should be sutured to the skin using strong silk to prevent dislodgement. The wings or flanges of the plastic tracheostomy tube can be shortened so the tapes cannot be used during the initial recovery period. The practice of tying in the tracheostomy tube with a surrounding gauze pad should be discouraged for the first 24 hours, at least, as the pad may obscure any sign of bleeding and may even encourage surgical emphysema as the tissue become swollen.

**Emergency tracheostomy**

A tracheostomy performed in an emergency under local anaesthetic can be a harrowing experience for all concerned and should be avoided wherever possible by prompt decisions to perform elective procedures. However, the patient and the situation sometimes dictate otherwise, for example a cautious anaesthetist who is reluctant to attempt intubation in a stridulous patient with a known mass in the upper airway.

The patient should be placed in the usual position if possible with the head extended and the shoulder supported to give maximum exposure. Local anaesthetic (lignocaine 1%) should be injected into the incision area and into the strap muscles. Care should be exercised that the local anaesthetic is not placed into the trachea otherwise paroxysmal coughing may turn a semi-emergency into an absolute emergency. The local anaesthetic should not be placed in the paratracheal gutter as the recurrent laryngeal nerves may be paralysed, increasing respiratory distress. During the procedure the patient should be reassured that all is proceeding according to plan. When the trachea has been exposed the thyroid isthmus becomes a problem. There is a tendency to mobilize the thyroid off the trachea and move it upwards or downwards rather than dividing it. It is recommended that the thyroid isthmus be divided rather than mobilized as the thyroid can occlude the airway during early postoperative changing of the tube. It is important that the thyroid is divided in the midline as deviation from the path will cause torrential haemorrhage. The thyroid isthmus should be clamped in the midline using two artery forceps, it is divided and the cut ends transfixed by silk sutures. Once this stage has been carried out local anaesthetic should be instilled into the trachea to suppress the cough. The patient should be warned at this stage that he can no longer speak. This warning can help to minimize the panic that often accompanies this type of operation. If the patient becomes distressed or loses his airway during an emergency tracheostomy an airway must be secured without delay, otherwise the patient will die.

Once the skin has been incised, the trachea needs to be identified and stabilized using the thumb and the index finger of the non-dominant hand. Holding the scalpel in the dominant hand the strap muscles should be divided in the midline until the tracheal cartilages are identified. By retracting the skin and the strap muscles the trachea should be opened longitudinally. Using an artery forceps the trachea can be opened to allow the correct placement of the tracheal tube in the tracheal lumen. The cuff is then inflated to protect and control the lower airway. The emergency situation should now be controlled.

During an urgent tracheostomy the bleeding can be profuse and sometimes terrifying to the inexperienced. The surgeon must not waste time in attempting to stop the bleeding, but first secure the airway. To the surgeon's surprise the bleeding dramatically subsides once the
trachea is opened. It cannot be emphasized often enough that it is the surgeon's responsibility to make sure that the assistants do not retract the trachea from the midline during the surgery. The surgeon must dissect down on to the trachea because if exploration deviates from the midline major complications may result.

**The tracheostomy tube**

The selection of the tracheostomy tube is usually governed by the requirements of the operation and the postoperative care. Usually a plastic tube with an inner cannula and a built-in cuff is preferred if the patient requires protection of the lungs from aspiration or haemorrhage. If the patient requires controlled ventilation, a cuffed Shiley or Portex tube with a high volume, low pressure cuff system is satisfactory. A fenestrate tube permits the passage of air upwards through the glottis thereby allowing the patient to speak.

There are two types of tracheostomy tube.

**Metal tubes**

Metal tubes, whatever their design, have several basic common principles: an obturator, an outer tube, and an inner tube. The inner tube is always slightly longer than the outer tube so that crusts can collect on the protruding end. Cleaning of the inner tube can be performed unhurriedly because the outer tube maintains the patency of the airway. Newer tracheostomy tubes have a flange which is not rigidly attached, allowing free movement of the neck. In unusual situations special tubes are available - Koenig's tube for extensive and low narrowing of the trachea, and Durham's tube in which the position of the flange is adjustable, so that it can be used for patients with either thin or very fat necks. The main disadvantages of the metal tubes are that they do not have a cuff, and cannot therefore produce an airtight seal. If the metal tubes do not fit properly, the end of the tube can erode the anterior tracheal wall. It must be remembered that the metal tracheostomy tubes are manufactured and supplied as a 'set'. Therefore only complete sets should be used otherwise complications may result.

**Non-metal tubes**

Non-metal tubes are made of rubber or silastic. Their advantage is that they almost all have an inflatable cuff and can be connected to an anaesthetic machine or a respirator. They do not produce mechanical damage to the trachea. Paradoxically the main disadvantage of these tubes is the inflatable cuff: it should be blown up to the point where there is a slight air leak past it and it should be deflated for 5 minutes in every hour. It is absolutely essential to maintain a permanent airtight seal, this can be achieved by the use of a Salpekar tube, which has two cuffs, one above the other, allowing alternate deflation and inflation of each cuff.

**Elective tracheostomy**

Any major operation on the mouth, pharynx and larynx always constitutes a danger to the airway, both as a result of direct surgical trauma and by physiological disturbance of the swallowing mechanism. In many of these patients with uncertain general condition,
particularly cardiovascular or pulmonary deficiency and advanced age, elective tracheostomy should be considered. Generally there is nothing to lose by its use - better too often than too late (Shaw, Stylis and Rosen, 1974).

**Postoperative management**

Much of the morbidity and some of the mortality attributed to tracheostomy can be prevented by meticulous postoperative care by the surgeon / physician in charge, the nursing staff, the patient and his family. Initially, frequent suction is necessary because the tracheostomy robs the patient of his ability to cough and he cannot clear the secretions from his tracheobronchial tree. The secretions are profuse for the first few days and may require a full time special nurse to perform very frequent suction if lower respiratory infections are to be avoided. The secretions result from the trachea being exposed to the cooler and drier air that it has been used to. The trachea therefore needs to be sucked out at frequent intervals. No specific time interval can be set and suction is required when indicated. The attending staff must obey the usual aseptic ritual: wash, wear gloves, and the suction tube should be sterile and preferably disposable. Changing the tracheostomy tube should not be necessary for at least 36-48 hours. At this time the tract will have epithelialized and the hole in the trachea will usually be readily found. However, even in experienced hands the trachea may occasionally be difficult to find, and therefore it is important that the surgeon who has performed the tracheostomy should do the first tube change. When the nursing staff are expected to do the tracheostomy changes the whereabouts of a medical staff member should be known in case the airway is lost.

Patients who are having their tube changed are placed in the tracheostomy position, that is lying flat and with the head extended. When the tube is withdrawn many patients experience paroxysmal coughing and the patient is therefore instructed first to inhale deeply, so that on removal of the tracheostomy tube all the secretions and debris are blown out rather than aspirated.

During the early postoperative phase the inspired air needs to be adequately humidified to prevent crusting. The use of saline or sodium bicarbonate instillations into the trachea, 1-2 mL/h, helps to reduce the likelihood of such complications and aids with suction clearance of the mucus secretions (Schild, 1970).

**Complications**

As with any other operation the complications of tracheostomy may be immediate, that is during or immediately after the operation; intermediate, happening during the rest of the patient's stay in hospital; or late, commencing after the patient has gone home (Conley, 1979). Table 9.2 lists some of the more common of these complications.

**Immediate**

**Haemorrhage**

Haemorrhage during the operation is frequent, arising from the anterior jugular veins or the thyroid gland. Bleeding should be controlled at once by diathermy or ligation. If the
bleeding is profuse and difficult to control digital pressure should be applied and the wound extended to allow a direct view of the bleeding area. Blind groping and grasping in a small hole is to be condemned as further bleeding and other tissue damage can result (Stemmer et al, 1976).

**Table 9.2 Complications of tracheostomy**

*Immediate*

Haemorrhage
- thyroid veins
- jugular veins
- arteries

Air embolism

Apnoea

Cardiac arrest

Local damage
- cricoid / tracheal cartilage
- recurrent laryngeal nerves

*Intermediate*

Dislodgement / displacement of the tube

Surgical emphysema of the neck

Pneumothorax / pneumomediastinum

Scabs and crusts

Infection

Tracheal necrosis

Tracheoarterial fistula

Tracheo-oesophageal fistula

Dysphagia

*Late*

Stenosis of the trachea

Difficulty with decannulation

Tracheocutaneous fistula / scars.

**Air embolism**

Air embolism is a serious complication but fortunately is very rare. During surgery large neck veins can be inadvertently opened with large volumes of air sucked in and passing rapidly into the right atrium. This situation can produce a critical situation leading to tamponade and death if not recognized. The complication can be minimized by meticulous surgery with good access and visibility.

**Apnoea**

Apnoea is though to be the result of the sudden discharge of the pent-up carbon dioxide from within the lungs once the obstruction has been suddenly bypassed. A quick way
of resolving this difficulty is to make the patient breathe a mixture of 95% oxygen and 5% carbon dioxide.

Cardiac arrest

Cardiac arrest may occur during a tracheostomy. The three most important factors appear to be excessive adrenaline production in the anxious patient; a rapid rise of the pH, consequent upon washing out of retained CO₂; and hyperkalaemia consequent upon respiratory alkalosis.

Local damage

In a short chubby neck great difficulty can be experienced with placing the tracheal incision in the correct position. It is better that the incision be placed lower than through the cricoid or the first ring as there is an increased risk of subsequent tracheal stenosis. Unilateral or bilateral vocal cord paralysis may arise from inadvertent injury to the recurrent laryngeal nerve during an emergency tracheostomy, particularly if the dissection deviates from the midline.

Intermediate

Dislodgement / displacement of the tube

The length of the tracheostomy tube and the thickness of the soft tissues of the neck are clearly the most important factors, however, the modern tubes are of sufficient length to obviate accidental withdrawal of the tube. Nevertheless the silver tubes of Negus and Chevalier Jackson are shorter and should be used only in patients with thin necks. Postoperative oedema, haematoma and emphysema will cause a broadening of the distance between the skin surface and the anterior tracheal wall, the process of expansion dragging the tube out of the trachea. The technique of suturing the flanges to the skin will help to minimize the possibility of dislodgement during the early period. In the later period the tracheostomy tapes should be tied with the neck in flexion, in which the girth of the neck is the smallest; if tied in extension the tapes will be loose once the head comes forward.

Subcutaneous emphysema

Subcutaneous emphysema can be alarming but is seldom fatal. Many factors may contribute to this complication - an over-large incision in the trachea, depression of the superior flap of the trachea above the incision, obstruction to the egress of air by glottic or pharyngeal obstruction, a tube that is partially obstructed or diverts air into the soft tissues of the neck, too tight closure of the subcutaneous tissues and skin about the tracheostomy tube causing a ball-valve effect, and excessive coughing. The emphysema is most often confined to the neck but can extend to the face and the chest wall. It usually presents within the first day and is self-limiting by the around the seventh day, unless the precipitating factors persist. The patient may develop a low grade pyrexia with localized cellulitis and a feeling of discomfort caused by stretching of the skin. The most frequent causes are tight skin closure and an improperly fitting tracheostomy tube; they should be rectified at once. In this situation
the risk of the tracheostomy tube being dislodged is increased because of the local increase in neck swelling.

**Pneumothorax / Pneumomediastinum**

These conditions may arise after any operation in the root of the neck. Usually this complication occurs in patients who are having surgery under local anaesthetic and are struggling, gasping and coughing. Occasionally the apex of the lung can be high in the neck and may be punctured accidentally. The diagnosis should be considered in all patients after the creation of the tracheostomy if the dyspnoea has not improved. A chest X-ray will confirm the diagnosis. Under severe circumstances, immediate needle aspiration with a 14-16 gauge needle into the upper anterior thorax will confirm the diagnosis and improve the patient's condition. The patient almost always needs aspiration and drainage.

**Scabs and crusts**

A tracheostomy alters the basic physiology of the inspired air from filtered, warm and humidified to dry, cold air coming into direct contact with the trachea. This alteration dries the tracheal and pulmonary secretions which interfere with the ciliary capacity to move the mucus blanket, and thus causes a production of thick, tenacious, mucus scabs and crusts. This basic interference with the movement of the ciliary blanket and the perpetuation of the drying process, is one of the most serious aspects in the postoperative course of tracheostomy. If not corrected, this sequence of events leads to infection, obstruction, atelectasis and pneumonia. If the situation is not controlled the scabs will increase in size, with the result that they are difficult or impossible to cough out or even remove by suction. Therefore the air supplied to the tracheostomy needs to be humidified artificially.

Air saturated with water vapour may be supplied by an old fashioned but cheap and reliable steam tent. Commercially available humidifiers are available in most hospitals nowadays. A simple method of droplet infusion is to introduce, in an adult, 15 drops/minute of saline via a fine bore plastic catheter or to instil 5 mL/h via a syringe directly into the trachea. Suction is applied as necessary with a sterile, disposable, smooth-tipped catheter; the nurse should wear sterile gloves. Many of these patients aspirate their oropharyngeal secretions and thus have an excessively wet trachea. There is a constant slow adaptation process over weeks, and ultimately the trachea adapts to its new dry environment. The patient should be encouraged to regulate and adjust his own humidification before discharge from hospital. In some instances it may be necessary to insert a bronchoscope to clear out the trachea and bronchi.

**Infection**

All tracheostomy wounds become locally contaminated within hours. However, all tracheostomy wounds should be attended with the strictest of local hygiene. Local dressings applied around the tracheostomy wound help to reduce the pressure on the neck skin and avoid necrosis. However, during the early period these dressings need to be changed frequently as secretions and blood accumulate. Barrier creams applied to the skin help to reduce the risk of local skin infections. Some patients develop Pseudomonas infections locally which may progress to fatal septicaemia. Prophylaxis is the best counteraction to infection.
Fortunately infection in the neck wound is usually local, indolent and produces local cellulitis with some granulation tissue. As the wound is open drainage is adequate and seldom are antibiotics necessary.

**Tracheal necrosis**

This complication most frequently follows local pressure secondary to infection. The pressure is derived from over-sized tracheostomy tubes, an improper curve of the tube, impingement of the tip of the tube or the pressure of the balloon on the trachea. The effect of this pressure begins as an ulcer on the wall of the mid or low cervical trachea. Previous irradiation, low grade infection and poor physiological status exaggerate the condition. This may lead to necrosis of the trachea with subsequent stenosis, tracheo-oesophageal or tracheoarterial fistula. It is therefore essential that the regulation of the pressure in the cuffed tubes in the intensive care units or on the ward receives the most careful attention. Any sign of bleeding, pain or obstruction should attract immediate investigation and remedial action by the elimination of the pressure factors, careful inspection of the ulcer, with a decision to allow the ulcer to heal by secondary intent or to excise the ulcer and attempt primary closure. Occasionally in large ulcers, the great vessels are at risk and need to be protected with a muscle flap with withdrawal or change in position of the ventilatory apparatus.

**Tracheoarterial fistula**

This tragic complication occurs in about 0.4% of tracheostomies. It is associated with an improper position of the tracheostomy tube against the vessel, improper curve and length of the tube, or is secondary to pressure from the cuff. It is almost always fatal. One of the essential prophylactic measures in tracheostomy is to evaluate the position of the innominate artery by digital pressure during the procedure. A significant warning sign before exsanguinating haemorrhage is slight bleeding from the trachea any time from 3 days to 3 weeks before the catastrophe.

**Tracheo-oesophageal fistula**

There are two significant factors contributing to the production of a tracheo-oesophageal fistula. A combination of these factors causes necrosis of the posterior wall of the trachea and the anterior wall of the oesophagus, thus creating a fistula. These factors are an overinflated or improperly fitting cuffed tube, causing pressure on the posterior tracheal wall and, usually, an indwelling nasogastric tube in the oesophagus. Positive pressure ventilation is a significant contributing factor. The diagnosis is suspected clinically by violent coughing during eating, chronic coughing associated with the swallowing of saliva and occasionally air escaping into the hypopharynx. Endoscopic examination is the best method for confirming the site and the presence of the fistula. The use of contrast media for diagnosis is often confusing because it is difficult to differentiate between aspiration of the contrast through the fistula and aspiration of the contrast material into the larynx as a result of loss of the swallowing reflex. Once the diagnosis is confirmed the best approach is surgical closure.
**Dysphagia**

Difficulty in swallowing is often encountered for the first few days after tracheostomy. This situation can be managed by feeding the patient through a Ryles tube or by inflating the cuff of the tracheostomy tube during feeds. This difficulty with swallowing may be related to the original indication for tracheostomy but other factors may contribute: tethering of the larynx so that it cannot move upwards during swallowing; the pressure of an inflated cuff on the oesophagus, or very rarely, as a result of ulceration of the tracheo-oesophageal wall producing a fistula (Bonanno, 1971).

**Late**

**Stenosis of the trachea**

Most tracheal stenoses result from the inflatable cuff on the orotracheal or tracheostomy tubes, others from scar contracture caused by improperly placed incisions, repetitive incisions, tracheal resections and trauma, tracheal infections or organic disease of the trachea.

**Difficulty with decannulation**

Most tracheostomies are temporary; the patient is ultimately decannulated and the tracheocutaneous fistula is closed. In the early phase of the tracheostomy the tube can be withdrawn without much difficulty because the wound is clean, no granulations have formed and the skin edges are still raw which leads to rapid closure. When the tube has been in place for weeks, months or even years removal may be difficult - granulations may have formed developing into fibromata resulting in tracheal strictures. Patients with tracheostomy of long duration should be carefully examined before the tube is removed. Lateral neck X-ray or xerograms with or without tomography can show granulations or fibromata gathering at the entrance of the tracheostome. Sometimes the trachea needs to be inspected directly to assess the lumen. Some tracheal narrowing can be demonstrated in more than 90% of patients after tracheostomy, and is most often seen near the site of the stoma (Lulenski, 1981). Functional impairment is rare unless the trachea has been narrowed more than 50% as measured in biplane radiographs. If the airway appears adequate, tube sizes can be decreased until the patient is breathing through the glottis. The smaller tube may be closed over with tape, or a plug can be inserted for several days before final removal of the tube. If the wound is recent it will close in 1-2 days, but if it is of longer duration it may be necessary to close it surgically.

**Tracheocutaneous fistula and scar**

The tracheal wound and the skin incision usually close by secondary intention, but occasionally fistulae persist particularly if the tracheostomy remained for a long period. A persistent fistula causes continual tracheal secretions with skin irritation, disturbed phonation, frequent infections and poor cosmesis. The scar after secondary closure is usually cosmetically unacceptable to the patient and family. The skin is usually thin and frequently becomes attached to the trachea by fibrous tissue so that the scar moves up and down when the person swallows and causes a tug on the trachea. Most fistulae heal spontaneously if they have been
present for less than 16 weeks, but if they have been present for longer most will need to be
closed. Surgery is best performed with endotracheal anaesthesia; the old scar is excised and
the strap muscles mobilized. The wound should be closed in layers with a small corrugated
drain to avoid the risk of haematoma formation or surgical emphysema (Kulber and Passey,
1972).
Chapter 10: Neurological affections of the pharynx and larynx

David Howard

Neuroanatomy

Neuronal control of the pharynx and larynx begins in the cerebral cortex at the lower part of the precentral gyrus. Additional fibres also arise from some frontal and parietal areas. The majority of fibres descend in the pyramidal tracts, with subsequent partial decussation at the upper border of the medulla to reach the nucleus ambiguus. Most palsies are produced by lesions of the nucleus ambiguus or the vagus nerve trunks and their branches. Lesions can cause unilateral or bilateral paralysis of the pharynx, often associated with palatal paralysis, and/or paralysis of the larynx and cricopharyngeus.

The cranial portion of the accessory nerve contains motor fibres destined for the muscles of the soft palate (except tensor veli palatini), pharynx, and intrinsic larynx.

**Glossopharyngeal nerve**

The glossopharyngeal nerve is essentially sensory. The nerve is not motor to the palate (it only supplies the stylopharyngeus which cannot be tested clinically) so that when the gag reflex is tested the stimulus is carried by the ninth nerve but the resulting palatal movement is mediated by the tenth nerve.

**Vagus nerve**

The vagus nerve leaves the cranium in the same sheath of dura as the accessory nerve. The glossopharyngeal nerve lies in front of these. The vagus is swollen during its exit by the superior ganglion cells. Below the jugular foramen the nerve is swollen again for approximately 2.5 cm by the inferior ganglion. This latter ganglion is crossed by the cranial root of the accessory nerve which blends with the vagus below the ganglion. The cells of the ganglion are sensory only.

The vagus nerve descends in the neck within the carotid sheath, between the internal jugular vein and the internal carotid artery, as far as the superior border of the thyroid cartilage and then between the vein and the common carotid artery to the root of the neck. Its subsequent course differs on each side.

**Pharyngeal branch**

This arises from the upper part of the inferior ganglion and runs between the external and internal carotid arteries to the superior border of the midline constrictor. It forms the pharyngeal plexus along with branches from the sympathetic trunk, glossopharyngeal and external laryngeal nerve.
**Superior laryngeal nerve**

This arises from the lower portion of the inferior ganglion, and descends on the side wall of the pharynx posterior and then medial to the internal carotid artery. It divides into the internal and external laryngeal nerves.

**Internal laryngeal nerve**

This descends to the thyrohyoid membrane piercing it above the superior laryngeal artery. It divides and supplies sensory innervation to the supraglottis, valleculae, and posterior surfaces of the arytenoid cartilages. Inferiorly it communicates with ascending branches of the recurrent laryngeal nerve. It also carries afferent fibres from neuromuscular spindles and cricoarytenoid joint receptors (Wyke and Kirschner, 1976).

There are abundant laryngeal receptors, both tactile and pain, providing a high degree of sensitivity particularly of the supraglottic mucosa. The afferent loop of the reflex initiates a bilateral laryngeal reflex. The chemoreceptors in the larynx have unknown functions. The subglottis has a similar receptor, served by the recurrent laryngeal nerve. These subglottic chemoreceptors apparently modify breathing, since some chemical stimuli applied to them initiate slow deep breathing, as does carbon dioxide applied to this part of the tract when it is isolated. The articular joint afferent fibres are particularly well developed, producing a highly sensitive and rapid monitoring response. Some reflexes which control respiration and phonation may arise from stretch receptors in the musculature of the larynx, demonstrable both histologically and by electrophysiology. Debate continues with regard to the exact role of all the sensory receptors and the reader is referred to an excellent contribution by Wyke and Kirschner (1976). Debate also remains as to whether the internal laryngeal nerve is entirely sensory: Williams (1951) described motor fibres innervating the interarytenoids. Certainly no obvious interarytenoid contracture takes place on stimulation of the superior laryngeal nerve.

**External laryngeal nerve**

This branch descends deep to the superior thyroid artery on the inferior constrictor. It then pierces the muscle, passes around the inferior thyroid tubercle and supplies the cricothyroid muscle. It also gives branches to the pharyngeal plexus and inferior constrictor muscle.

**Recurred laryngeal nerves**

The nerve on the right side arises from the vagus in front of the first part of the subclavian artery, winds backwards around the vessel and ascends obliquely behind the common carotid to the side of the trachea. On the right the nerve is commonly in front of the tracheo-oesophageal groove and may even be considerably lateral to the trachea as it reaches the lower pole of the thyroid gland. Near the lower pole both recurrent nerves are always intimately related with the terminal branches of the inferior thyroid artery. Authors differ in reporting the percentage of each variation, but on the right side the nerve occurs with approximately equal incidence behind, in front of, or intermingling with, the terminal branches of the inferior thyroid artery.
On the left side the nerve curves around the arch of the aorta and ascends commonly in a more protected position within the tracheo-oesophageal groove. On this side it is most likely to run behind the branches of the inferior thyroid artery, and least likely to pass in front of them.

Both nerves are intimately related to the medial surface of the thyroid lobes and their fascial coverings before passing under the lower border of the inferior constrictor to enter the larynx behind the cricothyroid joint.

As they ascend, each nerve gives branches to the mucous membrane and muscular coat of the oesophagus and trachea. The nerves may divide before entering the larynx. They supply all the muscles of the larynx, except the cricothyroid, and sensation to the mucous membrane below the vocal cords. They also give branches to the inferior constrictor and communicate with the internal laryngeal nerve.

**Aetiology**

It is difficult to produce an exhaustive list of causes of neurological deficits in the pharynx and larynx. The cause of many neural lesions, particularly of the larynx, are unknown.

**Supranuclear lesions**

Cortical lesions producing laryngeal and pharyngeal palsies are rare, and little is known about the effects on the larynx. It requires a bilateral symmetrical lesion of the cortex to produce a pharyngeal or laryngeal palsy, and in such cases respiratory and reflex laryngeal movements may be unaffected. Hemiplegia does not impair vocal cord movements.

**Nuclear lesions**

The nucleus ambiguus may be involved in posteroinferior cerebellar artery thrombosis, tumours of the medulla, bulbar palsy, syringobulbia, motor neuron disease, encephalitis, poliomyelitis, cranial polyneuritis, tabes and rabies. The most common cause of bilateral laryngeal paralysis arising in the nucleus is progressive bulbar palsy or cranial polyeuritis. Before the advent of lead-free pain it was a rare manifestation of lead poisoning. Nuclear lesions usually cause a combined paralysis of the soft palate, pharynx and larynx, but the larynx may be spared if only the superior part of the nucleus is affected. This may produce a palatopharyngeal paralysis - the syndrome of Avellis.

**Posterior fossa and jugular foramen lesions**

These involve the vagus nerves as they emerge from the brainstem and leave the skull. Nerves IX, XI and XII may also be affected and a large number of laryngopharyngopalatal palsies combined with lesions of these nerves have been named. They come under the headings of posterior fossa syndrome and jugular foramen syndrome. Although the many eponyms are of historical interest their use can lead to confusion.
The commonest combinations of associated cranial nerve lesions in the region are:

1. IX, X, XI, in the jugular foramen, Vernet's syndrome;
2. X, XI, Schimdt's syndrome;
3. X, XI, XII, Hughlings Jackson syndrome;
4. IX, X, XI, XII, Collet-Sicard syndrome;
5. IX, X, XI, XII and Horner's syndrome, Villaret's syndrome.

Having established the combination of nerve palsies the presence of an additional Horner's syndrome indicates a lesion outside the skull as the cervical sympathetic ascends to the base of the skull but does not pass through the jugular foramen. Evidence of additional brainstem compression obviously indicates an intracranial lesion.

Lesions in this region may arise from a wide range of diseases. These include skull fractures, primary tumours (particularly glomus tumors, meningioma) of the temporal bone and postnasal space, metastases, cholesteatoma, extension of infection from the middle ear, jugular bulb thrombophlebitis, tuberculosis, and syphilitic meningitis.

**Extracranial lesions**

In the cervical region the list of causes includes penetrating injuries; tumours of the hypopharynx, upper oesophagus, thyroid and parapharyngeal space; enlargement of/or surgery on the regional lymph nodes; and surgery of the thyroid gland.

Arising in the thorax the left recurrent laryngeal nerve has a longer course and is more exposed to compression by an aortic aneurysm, an enlarged left atrium in mitral stenosis, carcinoma of the bronchus, other mediastinal tumours and enlarged mediastinal glands.

The right recurrent laryngeal nerve is more vulnerable to injury during surgery on the thyroid gland because of its more anterior and lateral position at the inferior pole of the gland, rather than being protected in the tracheo-oesophageal groove.

**Paralysis of the palate and pharynx**

**Symptoms**

Unilateral palatal paralysis may not give rise to any symptoms because of compensation by the unparalysed muscles of the opposite side. Direct questioning may reveal slight changes in phonation (especially in professional voice users), snoring, postnasal drip and occasionally a unilateral hearing loss because of eustachian tube dysfunction on the affected side.

Bilateral palatal paralysis prevents the palate from closing off the oropharynx from the nasopharynx and prevents control of airflow through the nose - fluids, and sometimes solids,
thus regurgitating through the nose during deglutition. The voice has a nasal quality (rhinolalia aperta), as is heard in an untreated cleft palate. There is also a tendency to mouth breathing, snoring at night, and mucoid rhinorrhoea.

The patient with a bilateral palatal paralysis can usually swallow sufficient for his needs unless the pharynx is also involved. A combined paralysis is much more serious, particularly if the pharyngeal paralysis is bilateral; the patient cannot swallow and attempts to do so result in spasms of coughing as a result of overspill into the larynx. Soft bulky foods are usually more easily swallowed than liquids and normal solids. Even the patient's ordinary secretions may enter the lower respiratory tract and cause inhalation pneumonia.

In unilateral pharyngeal paralysis the compensation by the opposite constrictors is often efficient, but the patient needs to swallow in a deliberate manner, has bouts of coughing to 'clear' the throat, and may find it easier to sleep on the affected side to prevent laryngeal irritation from pharyngeal secretions.

**Signs**

Unilateral palatal paralysis is detected by examination of the oropharynx. When the patient says 'ah' the palate does not rise on the affected side and the uvula is drawn to the normal side. With a bilateral palatal paralysis the palate remains immobile during phonation. Occasionally it may be difficult to decide whether there is any movement in a bilateral paralysis. The patient cannot whistle or blow up a balloon.

Sensation is tested by touching each side of the palate gently with an orange stick and asking the patient to compare the two. Pain sensation may also be tested, with care, using a long pin. The posterior pharyngeal wall is tested in the same manner. With a unilateral pharyngeal paralysis the pharyngeal reflex is lost on the affected side and the pharyngeal wall droops. In both unilateral and bilateral pharyngeal paralysis pharyngeal secretions collect in the hypopharynx and around the laryngeal inlet and may be clearly seen on indirect laryngoscopy. If the symptoms are strongly suggestive of bilateral pharyngeal paralysis care should be taken on indirect laryngoscopic examination as it may produce sudden aspiration of these pooled secretions.

**Diagnosis**

Rarely, palatal paralysis in children may be referred to the otolaryngologist because the symptoms are believed to be caused by 'adenoids'. The history of enlarged adenoids is very different: the child has nasal obstruction but no nasal regurgitation. The thick nasal intonation caused by enlarged adenoids produces the contrasting speech defect of rhinolalia clause.

Reduced motility, and occasionally fixation, of the palate may be produced locally by scarring following unsatisfactory cleft palate repair, adenoidectomy or tonsillectomy, syphilis, scarlet fever, and tumours.
**Investigation**

Paralysis of the palate is unlikely to require investigation other than for the underlying cause. Accurate recordings of the degree and type of nasopharyngeal closure with palatal movement have been studied using video recordings obtained via flexible nasoendoscopy. This evaluation is only necessary when palatal or nasopharyngeal surgery is proposed for a long-term and stable palatal paralysis.

Pharyngeal paralysis with varying degrees of dysphagia has been the subject of physical, radiological, endoscopic and electrophysiological investigations. By far the most common in clinical use has been the radiological evaluation of pharyngeal and oesophageal swallowing using radiopaque contrast media such as barium, or the water soluble material gastrografin. Modern cineradiology and cine-fluorography techniques have been further enhanced by the addition of videotape facilities allowing detailed and repeated analysis of each phase of swallowing. The degree of overspill can be seen and cricopharyngeal spasm and notable hold-up clearly shown. This latter feature may help to identify those patients who would benefit from a cricopharyngeal myotomy, but sadly the radiographic appearances and subsequent postmyotomy results do not always correlate.

Pharyngeal pressure measurements, pH monitoring, electromyographic recording and endoscopic examination, although used in research, have yet to prove their worth in most clinical situations.

**Treatment**

Treatment differs according to the cause and extent of the lesion. Isolated palatal paralysis rarely requires treatment, although troublesome nasal regurgitation may be helped by holding the nose during swallowing or by an upper dental palate with a soft palate extension.

Pharyngeal paralysis requires treatment to provide adequate nutrition and prevention of inhalation pneumonia. Temporary help may be obtained by a fine bore nasogastric tube for feeding and repeated pharyngeal suction to protect the airway. This is unsatisfactory in the long term, and tracheostomy and gastrostomy may be necessary. A cuffed tube is used in the tracheostomy but even this requires regular deflation and may not provide adequate protection against pharyngeal secretions. Under these circumstances total laryngectomy has been used to separate the food and air passages. The neurological deficit can be unpredictable in the long term and spontaneous improvement may occur depending on the aetiology. If this is at all likely, a satisfactory compromise is provided by epiglottopexy (Brookes and McKelvie, 1983) in which the laryngeal inlet is occluded by stitching the epiglottis to the supraglottis via a lateral pharyngotomy leaving a small posterior defect. A valved tracheostomy tube allows speech, while the epiglottopexy prevents any significant overspill. The operation can be reversed if subsequent neurological improvement takes place.
Pharyngeal neurological lesions

_Globus pharyngeus (globus 'hystericus', idiopathic globus_

This functional disorder is most common in middle-aged women and is associated with a variety of sensations in the pharynx and larynx. Emotion, particularly fear, can cause a marked sensation of a lump, dryness, or 'blockage' of the throat, so it is not surprising that people who are emotionally unstable should refer symptoms to this region. However, the term globus hystericus is misleading as true hysteria is rare and hysterical dysphagia is uncommon even in these patients.

The most common feature in globus pharyngeus is the sensation of a lump in the throat. Often the patient's attention has been drawn to the throat by a previous minor throat infection, a transient incident with food 'sticking' in, or 'catching' the throat, or a relative or friend dying of 'cancer of the throat'. The symptom is most obvious when the patient attempts to swallow his own saliva to 'see if the lump is still there', but there is no true dysphagia and the symptoms often disappear while eating a meal. Repeated attempts at swallowing saliva may lead to aerophagy, with gastric distension and discomfort. The symptoms may have been present for many months and are usually worse if the patient is under any form of stress.

Examination often reveals a notably anxious patient with a pronounced gag reflex, but the pharynx, oral cavity, larynx and neck do not show any evidence of disease.

It is advisable to investigate these patients by a blood count to exclude anaemia, and by a barium swallow which may show obvious cricopharyngeal spasm without any other abnormality, but the role of this finding in the pathogenesis of the problem remains unclear. The spasm may be associated with lesions at the lower end of the oesophagus such as hiatus hernia, carcinoma or achalasia. There is no doubt that many globus patients can be demonstrated to have a hiatus hernia and it is postulated that the lower oesophageal pathology causes reflex vagal stimulation and subsequent alteration in the tone of the hypopharyngeal musculature, particularly the cricopharyngeus. However, many patients have hiatus hernias without globus type symptoms and research has not yet explained the cause of globus pharyngeus.

If symptoms persist direct rigid, or flexible endoscopy may be indicated as these methods are more likely to detect early organic disease, particularly of the postcricoid area, than radiology.

If no organic disease is shown by adequate investigation the patients are often rendered symptom free by reassurance and explanation. Occasionally, an anxiolytic, such as diazepam, may help troublesome symptoms but this type of medication is only required for a small proportion of patients. Speech therapists may have a useful part to play, particularly if there is an associated functional dysphonia. Rarely, the more emotionally crippled patient requires psychiatric referral.
Cricopharyngeal spasm and pharyngeal pouch

These pharyngeal disorders almost certainly have a neurogenic basis but are more fully discussed in Chapter 14.

Glossopharyngeal neuralgia

Apart from the distribution of the pain, glossopharyngeal neuralgia resembles the much commoner condition of trigeminal neuralgia. The pain occurs in brief agonizing stabs which may be of great severity. They usually start in relation to the tonsil and radiate down the side of the neck, in front of the ear and to the back of the mandible. Very rarely the pain may begin deep in the ear.

The attacks are usually precipitated by swallowing or by protruding the tongue, but when the ear is the main site external stimulation of the ear or skin may provoke an attack. Similar, but more continuous pain may be caused by tonsil or pharyngeal neoplasm and this must be carefully excluded.

When the throat is the main site of pain, some relief may be obtained by direct application of cocaine to the lateral pharyngeal wall and posterior third of the tongue. Carbamazepine (Tegretol), given in increasing dosage up to 200 mg four times daily, may control the attacks and additional relief may be obtained with sedatives. If these measures prove unsuccessful operation on the nerve may be necessary. It is always wise to regard the neuralgia as evidence of an underlying lesion until proved otherwise by events or by surgical exploration. The list of findings at surgical exploration includes a long styloid process, aberrant vessels coursing over the nerve, unsuspected neurofibromata and cholesteatomata. Skull base and lateral X-rays may be useful and selected cases may require computerized tomographic (CT) scanning.

The nerve may be approached via the tonsillar fossa, skull base, or intracranially. Wilson approached the nerve through the tonsillar fossa where it lies on the stylopharyngeus (Wilson and McAlpine, 1946). It may be avulsed or divided, but adequate safe dissection to the base of the skull is not possible by this route and only symptoms referable to the throat are relieved. A long styloid process may be fractured or partially removed during the same procedure (after tonsillectomy) but the vogue for this procedure seems to be declining.

Approach to the nerve at the skull base is difficult owing to its relatively small size, depth and relation to other important structures. It must be avulsed to remove the jugular and petrosal ganglia otherwise the connections with the tympanic plexus remain intact and any ear symptoms continue.

Injection of the nerve with alcohol at the point of emergence from the skull base can be undertaken but this is a difficult and hazardous procedure which has not gained wide acceptance.

The most reliable results are obtained with modern neurosurgical techniques involving craniotomy and division of the nerve fibres soon after emergence from the medulla.
Additional division of the upper two rootlets of the vagus nerve containing auricular and pharyngeal branches may be necessary.

**Herpes zoster**

Rarely, the neurotrophic varicella zoster virus may affect the distribution of the ninth and tenth cranial nerves. The vesicular eruption usually affects the palate, the pharynx and the laryngeal inlet on one side. The pharyngeal lesions may be isolated or be accompanied by other eruptions on the auricle or the anterior pillar of the fauces such as occur in geniculate herpes (Ramsey Hunt's syndrome).

The eruption is usually preceded by general malaise, fever and anorexia, particularly in the elderly. Herpes zoster may occur at any age, but most patients are over 50. Initially the throat is sore for a few hours before typical vesicles appear. These break down to form shallow ulcers, and they may be accompanied by severe pain. They heal without scarring but intractable pain may persist for many years in elderly patients. Local treatment with an antiseptic mouthwash keeps the pharynx clean, and local analgesics are useful before meals. Systemic steroids have been advocated but are of no proven value. Strong analgesics such as pethidine may be required in the acute stages but are contraindicated for persistent postherpetic pain. Carbamazepine (Tegretol) is rarely useful in postherpetic pain and nerve division or injection often provides only temporary relief. A combination of chlorpromazine and dihydrocodeine can be given for long periods and antidepressants may be required to treat the depression which may accompany the persistent pain.

**Diphtheria**

This disease is caused by the Klebs-Loeffler bacillus, *Corynebacterium diphtheriae*. It produces a membranous exudate at the initial site of infection which is later followed by distant toxic effects, of which polyneuropathy and circulatory failure are the most important.

It has a worldwide distribution but only isolated outbreaks occur in the UK following the introduction of active immunization. Because of the rarity at the present time the first cases of recent outbreaks have been missed and the subject deserves careful consideration.

Diphtheria is most common in the 2-10-year-old age groups and is spread by droplet infection from carriers and patients. Active immunization has been followed by the disappearance of carriers.

**Pathology**

The organisms remain at the site of infection and do not become invasive. The powerful exotoxin initially causes epithelial necrosis, followed by an inflammatory reaction with the necrosis forming the characteristic membrane. The membrane is 'false' as it consists of invaded and necrotic layers of mucosa and is not an exudate. The membrane is adherent and is difficult to remove. The corynebacteria are present at the margin of the membrane and swabs should be taken from this area.
The neurological symptoms are associated with fatty degeneration of the myelin sheaths of involved nerves with a consequent slowing of nerve conduction velocity. The fauces are the commonest site of diphtheric infection and as absorption of toxins is more rapid than from the other sites (nose, nasopharynx, larynx), faucial diphtheria is usually associated with most toxaemia. Palatal paralysis is attributed to the ascent of toxin from the common faucial site of infection to the medulla. (In cutaneous infection local ascent of the nerves by the toxin is responsible for the local development of paralysis.) Paralysis of accomodation, generalizedpolyneuropathy and myocardial damage are the consequences of dissemination of the toxin via the blood stream.

**Symptoms and signs**

Although infection can also occur in the nose, nasopharynx and larynx, the faucial site is much the commonest and paralysis of the palate is usually the earliest neurological symptom. The onset of faucial diphtheria is usually insidious: the child becomes quiet and anorexic but rarely complains of a sore throat. Lassitude and general malaise are associated with a normal temperature or mild pyrexia seldom above 38.4°C (101°F). Initially the membrane is absent, but may become extensive within 24 hours. It usually begins on one or both tonsils and spreads on to the fauces, uvula and palate. It may vary from cream to yellow to grey, glistens, and bleeds with removal. Paradoxically it is usually thinner and less well defined in the more severe cases but covers a wider area of the pharynx. Other sites of membrane formation, notably the nasopharynx, must always be checked. Fetor is striking and characteristic and there is usually a cervical adenitis. Early signs of toxicity are marked pallor, drowsiness, vomiting and tachycardia.

Palatal paralysis, usually bilateral, may occur within a few days but is commonest during the second or third week. It leads to regurgitation of fluids through the nose and a nasal voice. The palatal reflex is absent. Paralysis of accomodation usually develops in the third or fourth week and generalized neuropathy between 5 and 7 weeks after infection. This generalized neuropathy (which is not necessarily preceded by palatal and eye palsies) involves paralysis of the pharynx, intrinsic muscles of the larynx and the diaphragm. It is a serious complication because dysphagia may be complete and there is risk of aspiration pneumonia and respiratory failure.

**Diagnosis**

Any membranous throat condition should be regarded with suspicion particularly if it is difficult to remove and associated with a low grade fever but a relative tachycardia. Infectious mononucleosis can produce a grey membrane and oedema in the throat identical to diphtheria and is the commonest infection to cause real difficulty in diagnosis. However, other signs such as generalized lymphadenopathy, splenomegaly and the finding of abnormal mononuclear cells in the peripheral blood help to distinguish the condition. Agranulocytosis and acute leukaemia may produce lesions of the throat resembling diphtheria.

Quinsy, Vincent's angina, thrush and herpes zoster should be readily distinguishable.

Nose and throat swabs should be taken in every suspected case, but negative cultures do not exclude diphtheria and bacteriological confirmation of virulent diphtheria may take 3
or 4 days. Paralytic complications increase in frequency and extent with increasing delay of treatment by antitoxin, and this therapy must never be withheld while awaiting bacteriological confirmation of the suspected clinical diagnosis. Once toxin is fixed in heart muscle or peripheral nerves it is not affected by antitoxin which can only neutralize circulating toxin.

**Treatment**

The importance of rest in this condition cannot be overemphasized. All patients should be nursed flat in isolation, often for many weeks. They should be mobilized gradually and only when clinical and electrocardiographic evidence of myocarditis is absent. The old fashioned adage of 'a pillow a week' until the patient is sitting up remains a good regimen. As complete recovery can ensure in many cases, it is tragic when death results from too early and too great an exertion because of impatience.

The dose of antitoxin required depends on the site of the disease, the extent and duration of the membrane. It varies from 20,000 units intramuscularly to 200,000 units intravenously. A repeat dose of antitoxin is unnecessary if the correct initial assessment is made but can be given after a 2-3 day delay if the membrane continues to spread. Penicillin should be given to all cases to limit the spread of infection. It does not affect any preformed toxin and is in no way a substitute for antitoxin. Intravenous injections of hydrocortisone and 1:1,000 adrenaline drawn up into syringes must be available when administering antitoxin as it is a horse-serum preparation. A history must be taken of previous injections and allergy.

Palatal paralysis usually calls for no treatment other than that dictated by the general condition of the patient, but when the pharynx is affected a nasogastric tube must be passed and left *in situ* to provide feeding and to lessen the risk of inhalation pneumonia. A sucker may be used to remove pharyngeal secretions, but in severe cases a tracheostomy with a cuffed tube may be necessary. If both respiratory and pharyngeal muscles are affected intermittent positive pressure ventilation can be carried out via the tracheostomy.

Although paralysis recovers spontaneously, the restoration of movements is said to be hastened by exercises, such as whistling and blowing up balloons.

**Prognosis**

This depends on the virulence of the infecting organism, the position and extent of the membrane, and the delay in administering antitoxin. Death from diphtheria during the first week is caused by circulatory failure or laryngeal obstruction. Myocarditis may prove fatal in the second or third weeks and respiratory failure is responsible for most later deaths. Complete recovery without sequelae is usual but may take up to 6 months in severe cases. Most patients are out of danger within 10 weeks. Active immunization with triple vaccine, commencing at 4 months of age, usually prevents or modifies diphtheria but fatal attacks may still occur.

Most convalescent and long-term carriers can be treated with a 7-day course of erythromycin. Tonsillectomy is rarely required.
Acute anterior poliomyelitis

This acute viral disease is characterized by local or widespread muscular paralysis resulting from destruction of anterior horn cells in the spinal cord and corresponding cells in the nuclei of the cranial nerves. It is of particular interest to the otolaryngologist because paralysis of the pharynx and larynx occur in the bulbar type of disease, with involvement of the brainstem nuclei.

Aetiology

This disease is also known as infantile paralysis and in areas of the world where the virus is uncontrolled and sanitation poor, young children are still the main victims. However, in developed countries there has been a shift of the age of onset towards young adults. In the UK and many other countries which have had active immunization campaigns, poliomyelitis occurs only sporadically and the classical midsummer and autumn epidemics are of historical interest only.

The neurotropic virus exists in three known types which have different antigenic properties. It is transmitted by pharyngeal secretions and food contaminated by virus excreted in the faeces. The virus enters the body via the nasopharynx or gastrointestinal tract and has an incubation period of 7-14 days. Axonal spread occurs, but the virus is also bloodborne and this is the main route of transmission to the central nervous system. Aycock and Luther (1929) indicated that the virus easily gained entry via the raw tonsillar bed after tonsillectomy. There is also good evidence that physical exertion during the stage of incubation predisposes to increased paralysis, particularly in those muscle groups used in the exercise. Paralysis may also develop in limbs into which injections are given. All these factors are obviously best avoided during an epidemic. Detailed reviews of the virology and epidemiology are available (Cohen, 1969; Drouhet, Debre and Celers, 1970).

Pathology

The virus is often widespread throughout the brain and spinal cord but has a particular affinity for motor nuclei of cranial nerves and the anterior horn motor cells. Depending on the severity of the attack these cells necrose, usually within the first few days of the disease, following which no further destruction occurs. Other cells show evidence of damage by loss of Nissl granules but these are presumably capable of recovery. Necrosed cells are removed by neurophages, and examination of the spinal cord many years after the acute episode may show almost complete loss of anterior horn cells in the affected areas. Approximately one-third of cells have to be destroyed to induce clinical signs of paralysis. Paralysed muscle undergoes severe neurogenic atrophy.

Symptoms and signs

There are three possible distinct phases of this illness. Entirely subclinical infections and trivial illnesses lasting a few days with headache, diarrhoea and sore throat occur in most cases. The illness aborts before the paralytic stage and these apparently healthy individuals greatly outnumber paralytic cases and probably provide the greatest source of infection. Depending on factors such as the type and virulence of the virus, a variable proportion of
patients who recover from the initial mild illness enter a second stage of meningitic involvement, the so-called pre-paralytic stage, 2-3 days after recovery from the first. Headache and general malaise return, are far more severe and are accompanied by lumbar and limb pain, and cervical rigidity. In children delirium and convulsions may occur. This group of symptoms resembles that of other forms of viral meningitis.

Finally, a small group of patients continues into the third, paralytic phase. Unfortunately, some adults who develop paralysis do not go through two clearly defined preceding phases. Prediction of the extent of paralysis is difficult but severe limb pain and loss of tendon reflexes suggest a bad prognosis. Rest in bed at the first sign of an attack is essential.

In most patients maximum paralysis is reached within 3 days and occurs in a random manner. Spinal paralysis may affect the respiratory muscles and early recognition is important. A useful simple test is to ask the patient to count as far as possible with a single breath, a number below 15 suggests serious respiratory insufficiency.

Brainstem damage is serious, swallowing becomes impossible and pharyngeal secretions are aspirated. Combined bulbar and respiratory paralysis is still often fatal despite the best treatment.

Fortunately, except in severe cases, only some of the muscles originally affected remain paralysed. Recovery begins after about a week, and may continue for over 3 months.

**Treatment**

Barrier nursing is essential. Both bulbar paralysis and respiratory paralysis may require the attention of the otolaryngologist. Respiratory embarrassment caused by accumulation and aspiration of pharyngeal secretions must be distinguished from true paralysis of the muscles of respiration. Respiratory weakness requires artificial respiration. If there is no bulbar palsy a tracheostomy is not necessary if a negative pressure cuirass or cabinet respiratory is used. These respirators once widely used with good results, require critical day-to-day management with a high level of skill on an appropriate unit. Nowadays they have been largely replaced by intermittent positive pressure ventilation via a naso- or oral endotracheal tube, or through a cuffed tracheostomy tube. The advantage of intermittent positive pressure ventilation is that, if bulbar paralysis is also present, the cuffed intratracheal tube prevents the aspiration of pharyngeal secretions, food and vomit. Nasogastric feeding is obviously important in patients with bulbar paralysis. These patients require detailed management of blood gases, bronchial secretions, secondary infection, nasogastric feeding, care of the skin, bladder, bowels, and satisfactory communication. These many factors are best undertaken by an experienced team on an intensive care unit.

**Prevention**

Salk introduced the first vaccine by killing the three virus strains with formalin and injecting the preparation systemically. The value of this preparation became well established in the 1950s but it did not prevent colonization of the gut by wild virus and an immune patient could still be a carrier. It has therefore been superseded by the oral administration of
an attenuated live vaccine of the Sabin type, given as one or two drops on a sugar lump. Immunity appears to last for at least 3 years and in children and young adults booster doses are needed at intervals of a few years. There can be no doubt that in countries where a sustained vaccination programme has been pursued paralytic poliomyelitis has been virtually abolished but it is important that standards are not relaxed allowing the problem to grow again (Paul, 1971).

**Rabies**

This viral disease of mammals is usually transmitted to man by the bite of a dog with infected saliva. Rabies was eradicated from the UK 80 years ago but remains enzootic in foxes in mainland Europe. It has been estimated to cause 15,000 deaths a year in other parts of the world. 'Furious' rabies is characterized by intense arousal and hydrophobia associated with inspiratory muscle and pharyngeal spasm. 'Dumb' rabies presents as an ascending paralysis.

**Pathology**

The virus enters via a bite and is transmitted along nerve trunks in both directions to infect many organs including salivary and lacrimal glands. Brain and spinal cord show ganglion cell degeneration and marked perineural and perivascular round cell infiltration.

**Symptoms and signs**

The incubation period varies widely from 10 days to years (usually 1–2 months depending on the distance of the infected bite from the central nervous system. The first general symptoms are depression, apprehension and insomnia. Then follows hydrophobia, the classical sign of rabies: attempts to drink water produce gross spasm of the pharynx, larynx and respiratory muscles. This spreads to all the muscles of the body producing opisthotonus. At its height even the sound or thought of water will elicit the spasm. Other signs are hypersalivation, dysphagia, cranial nerve palsies, cardiac arrhythmias and severe psychiatric disturbance. Patients sink into a coma after a few days and even with intensive care only a few patients survive proven rabies.

The condition must be distinguished from tetanus which has a much shorter incubation period of 2–3 weeks. Trismus is the early symptom and pharyngeal spasm is absent. In hysteria true pharyngeal spasm is absent and the problem is amenable to drugs and suggestions.

**Prophylaxis and treatment**

Animal bites must be thoroughly cleaned, and the wound left open. The indications for vaccine treatment are clear (WHO Committee, 1966) and it should begin as early as possible after the bite. The newer inactivated vaccine given in human diploid strain COI38 has been shown to be safe and effective (Wiktor, Plotkin and Grella, 1973; Aoki et al, 1975). Hyperimmune gamma globulin is now given as a rule with vaccine.
Patients with established rabies require heavy sedation to control spasm and suffering. Total paralysis with curare and artificial respiration offer the only hope.

**Myasthenia gravis**

This disease occurs in all ages and all races but is twice as common in women as men. It is usually seen in young adults and may present to the otolaryngologist when the bulbar muscles are involved. Current evidence suggests it is an autoimmune disease with damage to the acetylcholine receptors in the motor end-plate. Any group of muscles may be affected and the disease tends to remit and relapse. There is abnormal muscle fatiguability, sometimes confined to an isolated group of muscles. The extraocular muscles are those most commonly involved. If it is restricted to the bulbar muscles there is dysarthria, dysphagia, regurgitation of fluid through the nose, and movements of the tongue, palate and pharynx are decreased. The symptoms typically appear in the evening when the patient is tired, particularly towards the end of a meal. They improve after a night’s rest. If the patient is asked to count aloud the voice becomes less distinct and more nasal.

Diagnosis depends on the typical clinical picture, the evaluation of the effects of intravenous edrophonium (Tensilon test), modern electromyographic measurements, or the detection of antiacetylcholine receptor antibodies in the blood.

The standard symptomatic treatment for myasthenia gravis is still neostigmine or pyridostigmine, the dosage of which is steadily increased until the maximal effect is obtained. This does not affect the underlying natural history. Immunosuppressive treatment with prednisone, with or without azathioprine (Imuran) may be required. Plasmapheresis has been life saving.

Most remissions occur in the first few years of the disease but unfortunately relapses are common. Most deaths also occur in the early years of the disease. Neonatal myasthenia is occasionally seen in infants of affected mothers but usually recovers in a few weeks.

Thymic enlargement is often found in young patients and may be caused by a thymoma, some of which are malignant. The place of thymectomy remains debatable. The results are best in young women with a short and severe history, but both sexes may benefit and operation is indicated if there is rapid deterioration with optimum medication. If a thymoma is present radiotherapy may be used before surgery. Only one-third of patients with thymoma survive beyond 5 years.

Myasthenia is closely related to thyrotoxicosis and less commonly with sarcoidosis, diabetes mellitus, rheumatoid arthritis and systemic lupus erythematosus. Accumulating evidence suggests that the disease has several different clinical, immunological, and genetic forms (Feltkamp et al, 1974; Fritze et al, 1974).

**Motor neuron disease (progressive bulbar palsy)**

This is a disease of middle age or later in which degeneration affects motor neurons in the anterior horns of the spinal cord, certain somatic motor nuclei of cranial nerves, and in the cerebral cortex. Both upper and lower motor neurons are affected and symptomatology
is diverse. The cause is, as yet, unknown. Pathological fasciculation can occur in any situation where some motor neurons degenerate and others persist, but it is most common in motor neuron disease.

The most common presentation is wasting of the muscles of the hand and upper girdle, but another common presentation, of interest to the otolaryngologist, is progressive bulbar palsy. This is more common in women causing dysphagia and dysarthria. It is often combined with upper motor neuron signs and evidence of pseudobulbar palsy. If weakness begins with bulbar palsy it usually spreads to the shoulders and arms.

The classical sign of the bulbar presentation is fasciculation and wasting of the tongue as a result of degeneration of the hypoglossal nuclei. This sign is unreliable in a protruded tongue, and it must be elicited with the tongue on the floor of the mouth. Differential diagnosis from myasthenia gravis is rarely difficult.

Dysphagia with aspiration is the most dangerous symptom and the course of the disease is more rapid if this is the initial symptom.

No treatment has any effect on the course of this relentless and depressing disease. Cricopharyngeal myotomy has its proponents, but does not always help the dysphagia and any improvement is usually short lived. The knowledge of a fatal creeping paralysis can lead to a complete collapse of morale, and requires careful handling.

**Palatal myoclonus**

In this condition there are rhythmical movements of the soft palate, occurring 60-180 times per minute (palate nystagmus). They develop insidiously, on one or both sides, and interfere with sleep, swallowing and respiration. They persist during sleep and may be inhibited initially by voluntary control. It appears to be a disorder of the olivocerebellar modulatory projection on the rostral brainstem and is seen in multiple sclerosis and brainstem infarction.

It may occur in association with myoclonus of the pharynx, larynx, eyes and diaphragm. The palatal myoclonus may be audible as a clicking sound and is sometimes abolished by anticonvulsants.

**Laryngeal neurological lesions**

**Functional disorders**

Many patients of a type similar to those with globus pharyngeus present with disorders of laryngeal sensation or voice production. These are related to social upsets, anxiety, smoking and vocal misuse. Following exclusion of organic disease local measures, reassurance, and advice from a speech therapist are usually successful.
'Spastic' larynx

Paralysis of the larynx may be caused by peripheral or central damage. As mentioned previously the latter is rare at a cortical level and the lesions are 'spastic'. This must not be confused with the so-called 'spastic' dysphonia which has been discussed at length in recent years. Spastic dysphonia is a rare bizarre dysphonia, possibly hysterical in origin, with overaction of the interarytenoid muscle against which the patient produces a markedly interrupted flow of speech. Improvement in some of these patients may be obtained by cutting the recurrent laryngeal nerve.

Brainstem paralysis of the larynx is, by contrast with cortical paralysis, flaccid.

Laryngeal spasm

This rather ill-defined condition is rarely neural and in young children is usually infective in nature. In the past, poor living conditions, rickets, whooping cough, upper respiratory tract sepsis, poliomyelitis, tetanus, and rabies have all been involved. Nowadays these are all rare in the UK.

Adults may suffer from 'choking' attacks with considerable panic and functional overlay. If these episodes are severe and there is concomitant vagal activity, so-called 'laryngeal vertigo' and a frank fainting episode may occur. These episodes may arise at night in individuals with marked oesophageal reflux and cricopharyngeal incompetence from any cause. It is presumed that a small amount of stomach content is aspirated into the larynx and the patient awakes with a 'choking' attack. This is an unusual disorder and other commoner causes of nocturnal dyspnoea should be excluded.

Superior laryngeal nerve palsy

Paralysis of this nerve whether on one or both sides is often clinically unrecognized and it has been somewhat neglected in laryngeal studies.

In unilateral superior laryngeal nerve paralysis, the voice is not severely affected and compensation occurs quickly. The disability to a professional voice user, in particular a singer, may, however, be significant. This form of paralysis is more readily recognized on examination because of the asymmetric tilt and shift of the larynx and the ipsilateral, slowly bowed, and flabby cord. Deprived of one of its tensors the affected cord also appears longer. Arytenoid movements are unimpaired. The voice sometimes fails to regain its former strength even though its quality returns.

Bilateral superior laryngeal nerve palsy is even more rarely recognized. The symmetry of the larynx at rest and during phonation makes the presence of this combined paralysis difficult to detect. The absence of anterior tilt allows the epiglottis to hang more over the endolarynx and the slightly flaccid, bowed and hyperaemic cords are more difficult to see. The bowed cords allow excess leakage of air during phonation and the voice is lower, weaker, breathy and lacks inflection. With good compensation the speaking voice returns to normal but the singing voice is severely compromised.
The detection of both these paralyses has been aided by modern fibreoptic laryngeal endoscopes, stroboscopy, and video recording (Howard and Lund, 1986).

Superior laryngeal nerve palsy occurs in a significant number of patients undergoing thyroid surgery and a detailed knowledge of the variable anatomy of this nerve is essential. Dissection and division of the superior pole vessels immediately adjacent to the capsule of the upper pole of the thyroid gland will lessen the chances of damage. Trauma to the neck, particularly from road traffic accidents, can cause superior laryngeal nerve palsy.

**Recurrent laryngeal nerve palsy**

The eventual static position and appearance of a paralysed vocal cord will depend on the degree and permanency of denervation and the degree of associated muscle atrophy and fibrosis. Semon (1881) and his contemporaries, and otolaryngologists up to this day have failed to recognize these variables, and produced an unnecessary amount of disagreement and controversy. Indeed accurate recording of the position of a paralysed vocal cord is difficult because only 3-6 mm separate the median, paramedian and so-called 'cadaveric' positions. Therefore, many cases are reported inaccurately and this may help to explain the apparent inconsistencies between the various theories.

The 'cadaveric' position does not always occur after death and is best replaced by the term 'lateral'. By far the most satisfactory method of describing the position of the paralysed cord is to state clearly, in millimetres, the distance it lies from the median. If detailed follow-up and evaluation is to be undertaken for research and publication, then endoscopic photographic documentation should be used to ensure accuracy of long-term measurements.

It is also best to describe the paralysis, whether unilateral or bilateral, in terms of abductor or adductor paralysis, that is the direction of movement which the cord cannot make.

**Semon's law**

Semon's law is of historical interest only. The weight of neurohistological and neurophysiological evidence against it is now overwhelming, and it plays no part in modern laryngology. Briefly, Semon proposed that the motor fibres innervating the adductor and abductor muscles lay in separate bundles in the recurrent laryngeal nerve and that they had difference susceptibilities to an advancing lesion of the nerve, giving rise first to an abductor paralysis with the cord in the median position, and then adductor paralysis so that the cord came to rest in the lateral (cadaveric) position. There are many clinical inconsistencies to this, in addition to neurohistological and neurophysiological evidence that this is not the case. The reader is referred to the excellent account by Wyke and Kirschner (1976) for further study.

**Wagner and Grossman theory**

Wagner (1890) and Grossman (1897) proposed a theory which has had more recent experimental confirmation by Arnold (1962) and Dedo (1970). In the absence of cricoarytenoid joint fixation, an immobile cord in the paramedian position has a total pure unilateral recurrent nerve paralysis, and an immobile vocal cord in the lateral (cadaveric)
position has a combined paralysis of superior and recurrent nerves (the adductive action of cricothyroid is lost).

However, clinically it is not uncommon to see patients with intrathoracic lesions (which should produce a pure recurrent palsy) with the paralysed cord in the lateral (cadaveric) position. Purported explanations for this are stretching of the nerve by the intrathoracic lesion thus pulling the vagus down from the skull base and injuring the superior laryngeal nerve; and possible retrograde atrophy of the vagus to the nucleus ambiguus. Damage to the laryngeal nerves produces loss of sensation as well as motility. This may produce an incompetent laryngeal sphincter and possible life-threatening aspiration.

No doubt controversy with regard to the above considerations will continue but, from a practical point of view, the management of a patient with a vocal cord palsy depends on the aetiology of the lesion and on the defect it causes. In the case of a unilateral cord palsy the patient may obtain excellent vocal compensation without treatment, only the timbre of the voice being altered. 'Idiopathic' defects may recover up to 3 years after onset and it is necessary to wait a minimum of 6 months to see if recovery occurs in a paralysed cord before undertaking any definitive treatment. It is also wise to regard the diagnosis as 'unknown' rather than idiopathic for at least 18 months after onset.

**Aetiology**

Stell and Maran (1978) reviewed five large series of paralysis of the recurrent laryngeal nerve in the literature. The following groups were discernible.

**Malignant disease**

This accounted for 25% of cases, one-half being caused by carcinoma of the lung.

**Surgical trauma**

This was responsible for 20% of cases with surgical procedures on the lung, heart, oesophagus and mediastinum now outnumbering those produced by thyroidectomy.

**Idiopathic**

In 13% no cause could be found although virus infections such as influenza and infectious mononucleosis have been suggested as aetiological agents. In this idiopathic group all patients who are smokers should be considered to have carcinoma of the lung until proved otherwise.

**Inflammatory**

Another group accounting for 13% of cases with pulmonary tuberculosis still being the major cause.
Non-surgical trauma

In 11% of cases the cause was stretching of the nerve by enlargement of the left atrium, aortic aneurysm, or neck trauma.

Neurological

A proven neurological cause was found in 7%, that is cerebrovascular disease, Parkinson's disease, multiple sclerosis, head injury, alcoholic and diabetic neuropathies.

Miscellaneous

The remaining 11% of cases did not fit neatly into any of the above categories and they included a wide range of illnesses such as rheumatoid arthritis, haemolytic anaemia, syphilis, and collagen diseases.

Owing to its long course the left recurrent laryngeal nerve is affected in approximately three-quarters of the cases and the right in about 15%, the remainder being bilateral.

Symptoms and signs

Evaluation begins with listening to the patient's voice as he gives the history. A faint whisper suggests a functional adductor paralysis, a forced whisper an organic adductor paralysis. A voice which tires with use suggests a unilateral abductor paralysis and stridor and aspiration occur with a bilateral abductor paralysis. The patient's age and occupation are of paramount importance.

Additional symptoms in the upper respiratory and gastrointestinal tracts, and the rest of the head and neck are obviously of relevance and the patient's past medical history and smoking habits are of particular importance.

Investigations

Haematological tests

A full blood count, erythrocyte sedimentation rate and serology are useful screening tests, but other indices such as viral studies and blood glucose tests are rarely useful.

Radiology

Plain X-rays and tomograms of the chest, and a good submentovertical skull base view yield the best results. Further plain X-rays of the nasopharynx, neck and petrous bones are occasionally useful. A barium swallow and thyroid scan may be required if the cause remains undetected. Computerized tomographic scanning is becoming increasingly available but is rarely indicated in these investigations.
**Endoscopy**

Panendoscopy with rigid and flexible instruments may be used to elucidate or confirm the diagnosis and the causative lesions. Particular points of note are the need for biopsy of the fossa of Rosenmüller and the bronchial carina even if these appear normal. This is particularly necessary in those patients whose history and radiology have not suggested a cause. The affected arytenoids must be palpated to distinguish vocal cord paralysis from cricoarytenoid fixation.

**Treatment**

Evaluation of these patients will show them to belong to one of the four following groups. The treatment may vary even within the same group depending on the general state of the patient and the aetiology of the lesion. Many patients will be helped by speech therapy, with or without associated surgery.

**Unilateral abductor paralysis**

This is an important group to consider first because speech therapy may be the only treatment necessary. The single palsied cord lies in the paramedian position and any initial hoarseness may well disappear as the unaffected cord compensates. When the left cord is involved, the aetiology is commonly that of carcinoma of the lung and presentation with the vocal cord palsy means that the carcinoma is already inoperable.

If the dysphonia persists and is distressing to the patient then treatment is by Teflon injection. The voice, although reasonable, tends to tire with repeated use, and professional voice users are particularly likely to request treatment. Teflon injection is most likely to produce a good result when the cord is in the paramedian position but the amount used is critical and the technique is more fully described later. The post-injection voice is rarely entirely 'normal', although the volume and quality are improved. It is important that the patient with a paralysed cord is advised before surgery that, although there is a good chance of improving the voice, it will not necessarily return to its previous 'normal' level.

**Unilateral adductor paralysis**

The flaccid palsied cord lies in the lateral position and gives rise to a weak husky voice sometimes no more than a whisper. In addition, as this lesion is most common the result of damage to the vagus or both superior and recurrent laryngeal nerves, the laryngeal sphincter is incompetent, part of the larynx insensitive, and consequently aspiration may occur.

Both the type and timing of treatment depend on the aetiology of the unilateral adductor paralysis. If the cause is a carcinoma, particularly of the bronchus, then the patients appreciate the improvement of voice and ability to cough that can be obtained with a prompt Teflon injection. Many of these patients with carcinoma will have only a few months to live and delay to await any possible laryngeal compensation is unwarranted. There is also increasing support for immediate injection of palsied cords resulting from major thoracic operations, thus enabling the patient to cough satisfactorily during the postoperative period.
Teflon injection for an adductor paralysis gives overall poorer results than for abductor lesions. This is a result of the difficulty of closing the posterior part of the glottis between the arytenoids where there will always be some air wastage. Overspill may be helped by injection but the results are more difficult to predict and require experience and care on the part of the surgeon.

Unilateral adductor paralysis not caused by carcinoma requires a waiting period of at least 6 months to allow for compensation or evidence of recovery. Speech therapy should be given during this time. If the unilateral adductor paralysis is a result of laryngeal trauma there is usually considerable scarring, particularly of the thyroarytenoid muscle and it may not be possible to displace the cord medially by means of a Teflon injection. In order to alter the position of the scarred cord from lateral to medial a number of alternative procedures have been advocated. The best results are obtained using a laryngofissure approach and moving the paralysed cord medially by inserting either muscle or cartilage between it and the adjacent thyroid lamina. To achieve a similar result, needles of nasal septal cartilage may also be inserted endoscopically using microlaryngoscopic techniques. This latter procedure is technically more difficult but avoids the complications occasionally associated with laryngofissure in an already scarred larynx. A reversed type cordopexy has been used fixing the cord in the midline via an external approach.

Bilateral abductor paralysis

This lesion is usually the result of damage to both recurrent laryngeal nerves at thyroidectomy. Other treatable causes are rare. The cords lie in the paramedian position and the voice is good but the degree of stridor is very variable. In the acute situation stridor may be life threatening and a tracheostomy required. However, if the lesion develops more slowly and the patient is relatively inactive there may be little or no stridor. Nevertheless, at some point, usually associated with an upper respiratory tract infection, all these patients will develop stridor. If the lesion is diagnosed soon after thyroidectomy immediate re-exploration of the neck is indicated. This is not so much to re-anastomose sectioned nerves (which gives overall poor results), but more in the hope of finding the nerves caught in a ligature. Removal of this often allows good long-term recovery. The author advocates a positive approach to post-thyroidectomy paralysis, but re-exploration of the nerves is pointless after delay of more than 6 months because of motor end-plate degeneration and fibrosis of laryngeal muscles.

The choice of treatment for patients with established bilateral abductor paralysis is wide and numerous operations have been described. No operation should be attempted until at least 6 months after the onset of paralysis, thus allowing for any possibility of spontaneous recovery. If there is recovery of any movement in one cord then possible operations should be undertaken only on the other cord.

When considering treatment it is important to remember the basic point that the patient has a good voice but poor airway. Any operative procedure on the cord to improve the airway will decrease the quality of voice and on occasions fail to improve the airway. Many of these patients will require or present to the laryngologist with a permanent tracheostomy. If this is fitted with a speaking valve they have the excellent situation of a good airway and a good voice. The only disadvantage to this being the actual wearing of the tube which some patients are unable to accept. A permanent speaking valve tracheostomy is usually the best choice in
the professional voice user. It is the author's experience that patients who have become used
to wearing such a tube will decline further surgical intervention and they should not be
persuaded into undergoing any of the following numerous procedures.

**Arytenoidectomy**

This procedure was first used by veterinary surgeons in the 19th century on race
horses with unilateral cord palsies. Ivanoff (1913) undertook the first human arytenoidectomy. King (1939) described an extralaryngeal approach to mobilize the arytenoid and affected cord laterally and attach it to a severed omohyoid muscle and the thyroid ala. Woodman (1946) modified this procedure by excising the arytenoid and suturing the residual vocal process to the inferior cornua of the thyroid cartilage. This latter operation became popular during the following 30 years but is associated with variable results of voice and airway and complications of infection and stenosis.

Arytenoidectomy can also be accomplished via a laryngofissure or lateral thyrotomy approach, combined with lateral cord mobilization and fixation. The thyrotomy approach allows good access to the arytenoid and a submucosal dissection.

Endolaryngeal arytenoidectomy was first described by Thornell in 1949 and, like the other procedures, gives variable results. It could be a difficult operation to perform with microsurgical instruments but can now be carried out bloodlessly and precisely using the CO₂ laser. It may be combined with partial or complete cordectomy using the laser and is now the operation of choice in the author's unit.

**Cordectomy**

Since the early part of this century various open and endoscopic methods of cordectomy have been advocated but variable results and, in particular, granulation formation have made surgeons wary of the procedure. With the advent of the CO₂ laser the endoscopic operation is precise and quick to perform and may be combined with arytenoidectomy. Removal of the posterior half of one vocal cord and the arytenoid gives a good compromise of voice and airway. The operation is completed in a haemostatic field and is associated with minimal postoperative oedema and granulation. The in-patient stay is very short and more tissue can easily be removed at a second endoscopy if necessary. The operation can be undertaken in those patients who have a compromised airway but who have so far managed without a tracheostomy.

**Reinnervation procedures**

In 1927 Colledge and Ballance demonstrated in monkeys, baboons and a single human case that bilateral recurrent nerve damage could be repaired by anastomosis of the phrenic nerve to the recurrent laryngeal nerve. This resulted in abduction of the cords in quiet respiration with an adequate airway. Crumley (1982) has demonstrated split phrenic nerve to recurrent nerve anastomosis giving respiratory abduction of the vocal cord while preserving diaphragmatic motion. This procedure deserves further evaluation but the nerve-muscle pedicle technique first reported in humans by Tucker in 1976 has not produced good results in other surgeon's hands. This latter technique involves transferring a portion of sternohyoid
muscle with its nerve supply from the ansa cervicalis into the posterior cricoarytenoid muscle to try to produce reinnervation and movement.

**Bilateral adductor paralysis**

Apparent bilateral adductor paralysis may occur in patients with psychiatric disturbances. There may be a severe dysphonia but the commonly used term 'hysterical aphony' is often inappropriate. The milder cases may be considerably helped by speech therapy particularly if the underlying problems (usually social) can be resolved. A few patients may need psychiatric referral and recurrence of symptoms is not uncommon.

Organic disease producing this bilateral lesion is fortunately rare but is usually a serious central nervous system disease or neoplastic process involving the medulla, skull base or upper neck. With both cords in the lateral position these patients are not only aphonic but are unable to cough well and have incoordinated swallowing. This laryngeal incompetence results in life-threatening aspiration in a very short time. Initial management involves a tracheostomy with a cuffed tube and a nasogastric tube for feeding. Further management is usually required depending on the aetiology. Total laryngectomy is the only sure way to protect the lungs but may not be undertaken if there is a possibility of neurological improvement. Epiglottopexy (Brookes and McKelvie, 1983) is the procedure of choice in neurological problems where subsequent neurological improvement may occur.

Other methods of management have been advocated such as Teflon injection of the cords and supraglottis, closure of the glottis by suturing the cords, and cricopharyngeal myotomy. The results of these other methods are variable and often poor and the author does not recommend them in a complete bilateral adductor palsy.

**Teflon injection in vocal cord paralysis**

The injection of Teflon paste into the paralysed vocal cord is now firmly established as an effective treatment with minimal complications. Indications for its use and the correct timing of surgery have already been considered in the previous section on vocal cord palsy.

In 1911, Brünings successfully treated patients by injecting paraffin into the laterally placed vocal cord. A modification of the injection syringe he described is still the instrument of choice today. Subsequent to his description, injected paraffin in other parts of the body was found to cause embolization and paraffinoma and its use was abandoned. Over 40 years later Arnold (1955) revived the technique using cartilage particles as the injection material. Following this a variety of substances such as bone paste, silicone, tantalum oxide powder, tantalum and Teflon were evaluated. Teflon paste emerged as the material of choice. Its permanence, low tissue reactivity and lack of carcinogenicity in over 20 years of use are well documented. The reader is referred to the excellent review of the characteristics of Teflon paste by Montgomery (1979). After injection the Teflon particles are walled off by foreign body reaction and the suspending glycerol is slowly absorbed. Initial accompanying oedema abates and an immediate good postoperative voice may deteriorate a little. A second injection may be necessary and even with experienced surgeons approximately 20% of patients will require this. A firm rubbery mass persists at the injection site as the paste migrates minimally, but it does not appear in lymph nodes and appears to be almost totally inert.
Several techniques have been used to evaluate the functional results of the injection such as aerodynamic studies, high speed photography of the vocal cords, video recordings of the vocal cords, voice spectrographs and phonation times. However, the simplest and most useful measure is simply to record the talking and singing voice of the patient pre- and postoperatively. The injection should produce a clearer, stronger voice (and decrease aspiration if present) without significant impairment of the patient’s airway.

**Method**

The injection can be performed under local or general anaesthesia and each method has its advantages and disadvantages together with surgeons who strongly advocate each method. The final choice will depend on the patient, his or her overall condition, and the local methods of practice and facilities. It is preferable for the surgeons to be familiar with both techniques.

Dedo, Urrea and Lawson (1973) gave an excellent account of the method used under local anaesthesia with results in 135 patients. This technique is used with the patient in a sitting position with the larynx and pharynx anaesthetized by sprays and droplets of cocaine. The surgeon views the larynx by indirect laryngoscopy and injects Teflon from a Brünings syringe into the paralysed vocal cord. Following withdrawal of the needle the patient can be asked to phonate and the quality of the voice checked. Further injection can be undertaken if necessary. This method undoubtedly gives good results but is not tolerated well by all patients and, indeed, is refused by some. The ability to assess the right amount of Teflon paste should not be too highly regarded since oedema of the surrounding cord occurs rapidly within 2-3 minutes and unless the injections can be completed in this time the true depot size is unpredictable. Subsequent resolution and glycerol absorption also occur in postoperative weeks.

The method is of course most useful in patients whose overall condition contraindicates general anaesthesia. Recently Ward, Hanson and Abemayor (1985) described a transcutaneous technique, injecting the cord via a needle placed through the anterior neck into the larynx, in a patient with severe trismus. The result was checked using a flexible fibreoptic telescope passed into the pharynx and attached to a videocamera.

The major criticism of the technique under general anaesthesia has been the likelihood of over-injection. However, this rarely occurs when the basic requirements are observed. These are that the other cord should be fully functional and the injected cord should not be pushed right up to the midline. The author's preference is for the following technique. Under general anaesthesia with fine catheter intubation or Venturi ventilation a Dedo laryngoscope is placed with accompanying Loewy suspension. A Brünings syringe is loaded with Polytef Teflon paste and care taken to ensure that the paste has filled the attached needle and passes easily. The first injection is made in the posterolateral corner of the middle third of the vocal cord just anterior to the vocal process. With each click of the syringe rachet 0.1 mL of paste is deposited and usually 0.3 to 0.4 mL is required to push the cord medially to lie close to the midline. It is most important to realize that paste is compressible and is not all delivered immediately the handle is squeezed. Therefore several seconds should be allowed to elapse following each click so that all the paste is extruded and the cord accurately assessed. The injection should cause a gentle fusiform enlargement and care should be taken to avoid
placing the paste in Reinke's space, too far laterally into the ventricular floor, or too deeply into the subglottis. A second injection is usually required in the anterolateral corner of the middle third of the vocal cord, that is the midpoint of the membranous cord. Because of the firm adherence of the mucous membrane to the vocal process it is impossible to inject posteriorly to close any gap between the arytenoids.

An intramuscular injection of dexamethasone 8 mg is given at the time of the procedure to minimize oedema but routine antibiotics are unnecessary. The majority of these operations can be carried out on a day-case basis, but a postoperative overnight stay may be indicated by the general condition of the patient.

**Complications**

The morbidity from this procedure is small but in all large series there is an incidence of about one in 100 cases requiring tracheostomy. The patient should be warned of this possibility preoperatively. Tracheostomy may be required because of acute progressive oedema which usually responds promptly to steroids, antibiotics and humidification allowing subsequent decannulation after only a few days. Less often a hemilaryngitis will occur 7-10 days postoperatively and requires antibiotic treatment.

Over-injection or injection into the subglottis both produce airway impairment but should be rare if care is taken. The author has successfully treated this complication by removal of the injected Teflon and any necessary additional tissue with a CO₂ laser.

The minimal soreness and foreign body sensation experienced by many patients subsides in a few days and an improvement in the voice should be obtained in over 90% of patients. As stated previously, the best results are in those with a pure unilateral recurrent nerve lesions.
Chapter 11: Tumours of the larynx

P. E. Robin and Jan Olofsson

In their broadest sense the terms tumour, swelling or space-occupying lesion have a significance in the larynx beyond that in most other sites, not only because of the early prejudice of the airway, but also because of interference with function in some cases, even when the lesion is minuscule. It is for this latter reason that many lesions are identified which are tumours but not true neoplasms.

Pseudotumours

Cysts

Cysts of the larynx may be congenital or acquired. They may arise in the vocal cords (55%), ventricular bands (25%) or in the epiglottis (20%) (Kleinsasser, 1978). They may be lined by a squamous or columnar epithelium.

Congenital cysts

Congenital cysts are rare and are most often in the ventricular bands or aryepiglottic folds. They may be diagnosed in the neonatal period as a consequence of breathing difficulties directly after birth. They may originate from a sequestration of embryonic cells in the saccule or laryngeal ventricle or arise from the seromucinous glands.

Incision of the cysts may be sufficient or excision can be performed if possible. If the airway is secured the intubation tube may be removed, otherwise it has to be left in place to allow repeat laryngoscopies. However, if the clinical course is prolonged, a tracheostomy is necessary.

Retention cysts

Retention cysts of the larynx are squamous or columnar; both forms may originate from obstructed seromucinous salivary glands. The squamous variant is common on the lingual surface of the epiglottis, the valleculae and on the aryepiglottic folds. These cysts may reach a considerable size before being diagnosed by minor cysts are often incidental findings at a routine otolaryngological examination. If possible these cysts should be excised entirely.

Squamous cysts also present on the squamous-lined portion of the vocal cords. They are most common on the undersurface of the anterior part of the cords. Minor cysts on the vocal cords are filled with clear mucus. Larger cysts contain a yellowish, thick fluid, which sometimes includes cholesterol crystals.

The laryngoscopic appearance of vocal cord cysts and vocal cord polyps may be very similar and it is microscopic examination that reveals the true nature of the lesion. Larger cysts are easier to recognize with their yellow colour and location under a thin translucent epithelium.
The treatment consists of excision of minor vocal cord cysts and marsupialization of larger ones.

Cysts of the ventricular band or cavity may be misinterpreted as neoplasms - differential diagnosis which must be ruled out. Cysts are most common above the age of 60 years and are lined by columnar or sometimes oncocytes cells. Under light microscopy oncocytes are large cells with an abundant pale, dark, or 'colloid' cytoplasm which is more or less acidophilic, and a small dense darkly staining nucleus. Electron microscopic studies show that the cytoplasm of the oncocytes contains large numbers of tightly packed mitochondria accounting for the granular or homogeneous appearance of the cells (Hamperl, 1962). Histochemically the oncocytes are characterized by the abundance of oxidative enzymes (Balogh and Roth, 1965; Johns, Regezi and Batsakis, 1977). The oncocytes tend to appear with increasing frequency in ageing individuals and occasionally form the predominant component of cysts and tumours. Such oncocytic lesions have most commonly been reported in the parotid glands. In the larynx a variety of names such as oncocytic cysts, oncocytic papillary cystadenomata, oncocytic adenomatous hyperplasia, oxyphilic granular cell adenoma, oncocytoma, and oxyphilic adenoma, has been given to these lesions. Nohteri (1946) found oncocytes in eight out of 37 (22%) laryngeal autopsy specimens. De Santo, Devine and Weiland (1970) in an analysis of material at the Mayo Clinic over 20 years found that 11% (33% of the saccular and 4% of the ductal cysts) were lined with oncocytes or contained such cells.

Granulomata

Non-specific granulomata are nearly always caused by trauma. Postoperative granulomata may occur after laryngeal endoscopic procedures or partial laryngectomies. Sometimes a stitch is found in the granuloma. A microlaryngoscopy should be performed to rule out recurrence in patients treated for malignant disease. The more commonly used laser surgery sometimes shows a tendency to excessive granulomatous tissue formation during the healing period.

Intubation granulomata are often caused by long-term intubation with artificial respiration. The granulomata are caused by an ulceration of the mucosa overlying the vocal process. The duration of intubation, as well as the size and type of the tube, and the degree of relaxation of the patients, are all causative factors. Prolonged intubation in adults is frequently discussed in the literature. Most intubation granulomata are diagnosed within a few weeks after extubation. Hoarseness, irritation and sometimes pain may occur. A microlaryngoscopy and excision should be performed, however, recurrences may occur. Intubation ulcerations and granulomata are certainly more common than is realized, but most of these granulomata may be coughed up and the base heals spontaneously.

Contact ulcers and granulomata located over the vocal processes, and often on both sides, probably have a multifactorial aetiology and the patients should be considered from several different aspects. They are nearly exclusively seen in males over the age of 30 years (Kleinsasser, 1978; Öhman et al, 1983). Vocal abuse has been considered to be the most important aetiological factor as suggested by Jackson (1928), who first described this lesion. He recommended a treatment consisting of vocal rest for a long period of time and, in some cases, vocal rest combined with surgical excision (Jackson and Jackson, 19350. Peacher and
Holinger (1947) reported good results with voice therapy, which have been confirmed in many reports. Patients with contact ulcers or granulomata have a low-pitch pressed quality of voice as their most pronounced feature. They often have an irritation and pain localized to the larynx, cough frequently and need to clear the throat. Emotional stress is considered another aetiological factor (Peacher, 1961). Other factors, such as hiatus hernia and gastro-oesophageal reflux have been discussed (Cherry and Margulies, 1968; Goldberg, Noyek and Pritzler, 1978; Ward et al, 1980). In an oesophageal manometric study, 74% of the patients examined with contact ulcers or granulomata, were found to have oesophageal dysfunction as hiatus hernia, gastro-oesophageal reflux, dysmotility, etc. About 30% of the general population of the corresponding age have oesophageal dysfunction (Öhman et al, 1983). It is difficult to know whether oesophageal dysfunction is an aetiological or concomitant factor of contact granulomata. Ward et al (1980) reported good results with antireflux therapy. Biopsy should be performed to rule out cancer which rarely occurs at this site. No difference in healing could be seen when voice therapy combined with surgery was compared to voice therapy alone (Öhman et al, 1983). Excessive granulomata should be excised to facilitate voice therapy.

Amyloidosis

The pathogenesis of amyloidosis is unknown but it is characterized by extracellular deposits of a proteinaceous substance. The disease was first described by Rokitansky in 1842, and the term 'amyloidosis' was introduced by Virchow in 1851. Burow and Neumann reported the first patient with laryngeal amyloidosis in 1875. Many hundreds of cases of laryngeal amyloidosis have been reported since that time.

Amyloidosis can be either generalized - primary or secondary - or localized. The larynx is rarely involved in generalized primary amyloidosis. It is, however, the usual site for amyloidosis of the respiratory tract. However, the real nature of amyloidosis is still an enigma. Amyloidosis makes up 1% of all benign laryngeal 'tumours'. It is slightly more common in males than in females and usually occurs between the ages of 40 and 60 years (Stark and New, 1949; McAlpine and Fuller, 1964). The sites of occurrence are, in descending order of frequency, the false vocal cords, aryepiglottic folds and the subglottis (Leroux-Robert, 1962; d'Arcy, 1972), but Ryan, Pearson and Weiland (1977) cited the vocal cords as the prime site.

Amyloidosis within the larynx occurs in two forms: one tumour-like and the other displaying diffuse infiltration. The symptomatology will of course, depend on the site of involvement. Hoarseness will occur if the vocal cords are involved, and increasing inspiratory problems are typical of subglottic deposits; patients with supraglottic amyloidosis have uncharacteristic and more diffuse symptoms.

Histopathology

Congo red is the most commonly used staining reaction for amyloid and gives a bright red colour. In polarizing light an apple-green birefringence is obtained. Low-angle X-ray diffraction is the third principal method, after light and electron microscopy, for identifying the amyloid substance (Kyle and Bayrd, 1975). The congo red reaction is sensitive, and false
positives and negatives are rare. However, Phorwhite BBU is more sensitive with even fewer false staining reactions (Waldrop, Puchtler and Valentine, 1973).

The following differential diagnoses have to be ruled out: hyalinized myxomatous polyps, benign and malignant tumours beneath an intact overlying mucosa, retention cysts and laryngocoele. Plasmacytoma with amyloid deposits is another differential diagnosis.

**Treatment**

The treatment for laryngeal amyloidosis is surgery, which can be performed microlaryngoscopically. Localized lesions may be removed entirely. In diffuse submucosal deposits repeated excisions may be necessary to restore the airway and to preserve the voice. Extra care should be taken when removing amyloid tissue at the level of the cricoid ring to avoid subglottic stenosis. A laryngofissure approach may be indicated for extensive lesions. The use of the carbon dioxide laser should not be overlooked. In amyloidosis of immunoglobulin origin the use of immunosuppressive or cytostatic agents has been suggested (Jones et al, 1972).

**Benign mesodermal tumours**

**Vascular neoplasms**

Vascular neoplasms arise from blood or lymphatic vessels. The tumours arising solely from lymph vessels are extremely rare within the larynx. Combined lymphangiomata and haemangiomata may be present. The blood vessel neoplasms may be benign (haemangioma) or malignant (haemangiosarcoma). In addition haemangiopericytomata and Kaposi’s sarcoma also occur.

**Haemangiomata**

Haemangiomata are rare in adults. Vascular but non-neoplastic lesions occur such as the 'telangiectatic' vocal cord polyp, which is filled with thin-walled blood vessels. Some of these vessels may be filled with old or recently formed thrombi. Around such polyps older submucous haemorrhage may be seen at microlaryngoscopy.

Another differential diagnosis is the pyogenic granuloma, often located on the posterior part of the vocal cord and related to a previous intubation.

**Infantile haemangiomata**

These are discussed in Volume 6, Chapter 26.

**Chondromata**

Since 1816, when a cartilaginous tumour of the larynx was first described by Travers (van de Catsijne, 1965), more than 200 such tumours have been reported in the literature and approximately 20% of these have been chondrosarcomata (Fombeur et al, 1974; Zismor, Noyek and Lewis, 1975). These tumours tend to occur between the ages of 40 and 70 years
and are more frequent in males than in females, with a ratio of four to one. Most of the
tumours originate in the cricoid cartilage (70%) and most often from the posterior cricoid
plate (van de Catsijne, 1965; Barsocchini and McCoy, 1968).

**Symptomatology**

The symptomatology of laryngeal cartilaginous tumours is generally non-specific, with
hoarseness and dyspnoea as prominent features, their degree depending on the site and size
of the tumour. Tumours arising from the cricoid cartilage often extend into the subglottic
space and thereby cause progressive inspiratory stridor. Hoarseness may occur if the vocal
cord mobility is impaired. Extension of the tumour posteriorly into the hypopharynx may
result in dysphagia. A swelling may be noted externally if the tumour is located in the cricoid
ring or in the thyroid cartilage.

**Clinical findings**

Indirect laryngoscopy usually reveals a smooth mass covered by an intact overlying
mucosa. While radiological examination may disclose peripheral or central calcific stippling,
coarse irregular calcification is the rule. This feature is considered to be pathognomonic of
cartilaginous tumours and is found in about 75% (Zismor, Noyek and Lewis, 1975). Because
the tumour may be so hard and difficult to penetrate, biopsy specimens may be
unrepresentative often consisting of the overlying mucosa only.

**Histopathology**

The histopathological evaluation of cartilaginous tumours often presents considerable
difficulties as regards both classification and grading of malignancy. A difficulty in
distinguishing between chondroma and highly differentiated chondrosarcoma lies in the fact
that pronounced cellularity and polymorphism often occur only in small foci (Lichtenstein,
1965). DNA measurements may assist in a correct diagnosis (Kreicbergs, 1981) but are not
available in all laboratories.

**Treatment**

Surgery is the treatment of choice, radiotherapy being of little value (van de Catsijne,
1965; Ackerman and del Regato, 1970). The operation of choice has generated much
discussion (Goethals, Dahlin and Devine, 1963; Al-Saleem et al, 1970; Hyams and Rabuzzi,
1970; Lawson, Bryce and Briant, 1972). Conservative surgery, whenever possible, has been
recommended for both chondromata and chondrosarcomata on account of the slow growth rate
of these tumours and the low incidence of metastases of the latter.

**Myogenic tumours**

**Leiomyomata**

Leiomyomata comprise three different types - common, vascular and 'bizzare'. The
latter had not been described in the larynx (Kleinsasser and Glanz, 1979). The leiomyoma is
one of the most common benign tumours in the human being.
Leiomyomata have been reported in children but they occur more often in adults of all ages. They seem to be most common in the supraglottic region, have been of pea to pigeon-egg size and have been removed endoscopically or by an external approach.

**Rhabdomyomata**

Rhabdomyomata of the true adult type are extremely rare tumours of the human body. Kleinsasser and Glanz (1979) found only eight descriptions of confirmed cases of rhabdomyomata in the larynx and added one of their own. Most rhabdomyomata in the larynx originate in the vocal cord region and appear as a polypoid mass but may extend above and below the cords.

Microscopic examination shows a tumour composed of round to oval cells with a pale, faintly granular cytoplasm. There are many large vacuoles in the cytoplasm and mainly placed in the periphery. The nuclei are round to oval and vesicular with prominent nucleoli, also located in the periphery. Cross-striations are usually visible in some cells with ordinary haematoxylin-eosin staining, but are accentuated by phosphotungstic acid-haematoxylin (PTAH) staining. Electron microscopic examination may be useful especially in uncertain cases. The differential diagnosis is primarily granular cell tumour.

Fetal rhabdomyoma is extremely rare in the larynx. The precise nature of this tumour is unknown. The lesion may be a hamartoma and not a true neoplasm. It usually presents shortly after birth but may occur in adults and present as a vocal cord polyp (Michaels, 1984).

The treatment for these lesions is surgery and endoscopic measures may be sufficient.

**Granular cell tumours**

In the past, benign granular cell tumours have been considered to be of mesenchymal origin. Abrikossoff (1926) suspected a myogenic origin and suggested the term 'myoblastic myoma'. The exact histogenesis of this tumour, however, still remains uncertain. A Schwann cell origin has been suggested (Azzopardi, 1956). Ackerman and Rosai (1974) concluded that the multiplicity of current aetiological data suggests that a granular cell tumour is the consequence of degradation and that it is not a specific neoplastic entity.

The most common location for granular cell tumours of the larynx is the true vocal cords and they may be managed endoscopically.

**Fibromata**

Fibromata are composed of fibrillar connective tissue. In the large series of benign laryngeal tumours presented by New and Erich (1938) only six of their 722 tumours were fibromata. Eight of the miscellaneous tumours listed in *Table 11.1* are fibromata (Shaw, 1979). The appearance may vary. New and Erich (1938) described them as soft and pedunculated and Shaw (1979) as round, firm, smooth and sessile.

The treatment is endoscopic removal in most cases.
Table 11.1 Benign tumours of the larynx seen at the Institute of Laryngology and Otology, London, 1948-1969 (Shaw, 1979)

<table>
<thead>
<tr>
<th>Non-neoplastic</th>
<th>Neoplastic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vocal cord polyps</td>
<td>Papilloma*</td>
</tr>
<tr>
<td>Retention cysts</td>
<td>Adenoma</td>
</tr>
<tr>
<td>Tuberculous granuloma</td>
<td>Chondroma</td>
</tr>
<tr>
<td>Intubation granuloma</td>
<td>Miscellaneous</td>
</tr>
<tr>
<td>Contact ulcer granuloma</td>
<td></td>
</tr>
<tr>
<td>Amyloid deposit</td>
<td></td>
</tr>
<tr>
<td>Wegener's granuloma</td>
<td></td>
</tr>
<tr>
<td>Granular cell myoblastoma</td>
<td></td>
</tr>
<tr>
<td>Miscellaneous</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>1300 (86%)</td>
</tr>
<tr>
<td></td>
<td>205 (14%)</td>
</tr>
</tbody>
</table>

* Approximately 25% were multiple juvenile papillomata.

Lipomata

Neoplasms may arise from the adipose tissue present, especially in the false cords. Many of the reported lipomata arose in the hypopharynx and extended into the larynx (Michaels, 1984).

Macroscopically, lipomata are light-coloured, encapsulated and lobulated tumours. Microscopically they are composed of fat cells of varying size and a fibroreticular stroma.

The treatment is endoscopic removal or by an external approach depending on the size and location.

Benign ectodermal tumours

Adenomata

Benign tumours arising from the seromucinous glands of the larynx are rare. The statistics given by Friedman (1975) from the Institute of Laryngology and Otology, London reported 16 cases seen during a 21-year period (see Table 11.1). Sabri and Hajjar (1967) reported on 37 neoplasms of the larynx of which 19 were mixed tumours. Most of these occurred in the subglottic larynx (Som et al, 1979).

Symptoms may be few until the tumour obstructs the breathing. The differential diagnoses should be limited to those lesions that are expansile masses with smooth overlying mucosa - a retention cyst, internal laryngocoele, angioma or adenoid cystic carcinoma.

The treatment is by surgery and the approach depends on the size and location of the adenoma within the larynx.
Neurogenic tumour

Along with the other benign tumours, neurilemmoma of the larynx is not common. New and Erich (1938), in their major review of 722 benign laryngeal tumours, reported one neurilemmoma. Holinger and Johnston (1951) reported one in a series of 1197 benign laryngeal tumours. Nanson (1978) found 87 reported in the literature.

Neurilemmoma is a benign tumour arising from the Schwann cells of the axon sheath. The term was coined by Stout (1935). It is usually a well-encapsulated, slowly growing tumour, the size of which can be fairly large. A neurilemmoma with a diameter of a few centimeters is obviously more serious in the larynx than growing subcutaneously. Symptoms develop insidiously but can be prominent if degeneration and haemorrhage occur into the tumour causing a life-threatening situation.

Treatment

Treatment is by surgery. Small tumours may be removed endoscopically, others via a laryngo-fissure procedure or a lateral laryngotomy, depending on the size and location of the neurilemmoma.

Paragangliomata

More than 30 paragangliomata of the larynx have been reported in the literature with an equal sex incidence and with a peak in the fifth decade of life (Olofsson et al, 1984). Most of these tumours arise from the supraglottic paraganglia and less frequently from the inferior ones. The location of the tumours means that they often do not give symptoms until they have reached an advanced stage. Haemoptysis may occur. Angiography can provide information about the vascularity of these tumours. Computerized tomography or magnetic resonance imaging, if available, are the best radiological methods to determine the extent of these tumours. Paragangliomata arising from the inferior paraganglia may present as thyroid tumours depending on the close relationship to the thyroid capsule. The diagnosis is made entirely on the microscopic examination. Strikingly few of the laryngeal paragangliomata have been diagnosed preoperatively, which to some extent may be the result of too superficial biopsies but also of the rarity of this entity.

Histopathology

Important criteria for making the diagnosis of paraganglioma include the typical ‘Zellballen’ pattern in light microscopy, which is best demonstrated by reticulin stain. The presence of argyrophilic granules is revealed by Grimelius’ stain. Ultrastructural examination shows neurosecretory granules. The main differential diagnoses are haemangiopericytoma, carcinoid tumour, granular cell tumour, salivary gland tumours, haemangioma and thyroid carcinomata, which may invade the larynx and trachea. A relatively high percentage of laryngeal paragangliomata show malignant behaviour with regional and distant metastases (Wetmore et al, 1981).
Treatment

The treatment for paragangliomata varies considerably because of the rarity of the tumour and the often incorrect diagnosis. Conservative surgery should be performed whenever possible.

Malignant tumours

Cancer of the larynx is a particularly important malignancy. In the UK it represents approximately 1% of all malignancies (Powell and Robin, 1983) in men, although somewhat less in women (Table 11.2). It has, in common, with many other head and neck cancers, a predominantly squamous pathology as well as early interference with both function and emotion. It shares with only a few other types of cancer (such as of the cervix, skin, lymphoma, perhaps colon) a high rate of cure which, in certain subsites, may reach over 85% (Table 11.3) and overall exceeds 50% (Powell and Robin, 1983). Carcinoma of the larynx, therefore, places upon the clinician a much greater responsibility than usual, for careful evaluation and treatment bring a probability of cure while, in common with a number of other head and neck neoplasms, failure may be followed by a relatively uncomfortable and unsavoury death. Even further demands are made at the present time, for not only the survival of the patient but the rehabilitation of speech is becoming of even greater importance than formerly. Thus the selection of treatment and types of surgery must be made with more insight than previously into the implication of the disease, its behaviour and response. Because of the disparity of the prognosis, not only between laryngeal tumours and those of neighbouring sites, but also of the various sites within the larynx, a greater than usual attention must be paid to the accurate assessment of each tumour, so that the appropriate management may be instituted.

Table 11.2 Incidence of malignancy of head and neck (from Powell and Robin, 1983, courtesy of Castle House Publications)

<table>
<thead>
<tr>
<th></th>
<th>Males (%)</th>
<th>Females (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Skin</td>
<td>8.2</td>
<td>7.0</td>
</tr>
<tr>
<td>Oral cavity</td>
<td>0.6</td>
<td>0.3</td>
</tr>
<tr>
<td>Oropharynx</td>
<td>0.4</td>
<td>0.1</td>
</tr>
<tr>
<td>Nasopharynx</td>
<td>0.1</td>
<td>0.1</td>
</tr>
<tr>
<td>Hypopharynx</td>
<td>0.3</td>
<td>0.2</td>
</tr>
<tr>
<td>Nasal cavity and sinuses</td>
<td>0.2</td>
<td>0.2</td>
</tr>
<tr>
<td>Larynx</td>
<td>1.3</td>
<td>0.2</td>
</tr>
<tr>
<td>Thyroid</td>
<td>0.2</td>
<td>0.6</td>
</tr>
<tr>
<td>Lymphoma</td>
<td>0.3</td>
<td>0.3</td>
</tr>
<tr>
<td>Sarcoma</td>
<td>0.1</td>
<td>0.1</td>
</tr>
<tr>
<td>All head and neck</td>
<td>11.7</td>
<td>9.1</td>
</tr>
<tr>
<td>All sites: total number</td>
<td>9177</td>
<td>8585</td>
</tr>
</tbody>
</table>
Incidence

Carcinoma of the larynx is not common, nor is it rare. Incidence world-wide varies (Waterhouse et al, 1982), and a number of areas of relatively high (>10/100.000) incidence can be identified, for example Brazil (Sao Paulo), the black populations in parts of the USA, Honk Kong, India (Bombay, Poona), France (Bas Rhins, Doubs), Italy (Varesa), Poland (Katowice), Spain, and Switzerland (Geneva), while low incidence areas (<3/100.000) include Japan, Norway, Sweden, New Guinea, and Senegal (Dakar). The UK suffers from an intermediate to low incidence (4/100.000). Incidence, it must be remembered, is as reliable as the statistical infrastructure, and a number of less developed countries may suffer from underreporting. It is clear, nevertheless, that significant differences do occur between and within various countries. In Hawaii the Caucasians suffer an incidence (8.1/100.000), more than five times that of the Chinese (1.4), four times the Filipino (2.0) and nearly three times that of the Japanese (3.3), who live in the same place (these figures are for men, those for women are proportionately less). Where the urban population can be identified separately from the rural population the incidence is almost always higher in the former. Where racial groups can be identified, the respective incidence appears to follow that of the country of origin, for example in Hawaii the Japanese incidence is similar to that of the Japanese in Japan, the Caucasians to that in the USA. The racial characteristic extends between countries, for example the incidence in the Chinese in Shanghai and the USA generally is similar. Black populations in the USA have a higher incidence of carcinoma of the larynx than their Caucasian counterparts in the same area. One invariable characteristic of carcinoma of the larynx is its greater predominance in men, compared with women, 6:1 at its lowest in Canada (Mannitoba) and 32:1 at its maximum in Italy (Varesa). Indeed, it is in the higher incidence areas that the male/female disparity is greatest.

Table 11.3 Histological differentiation: 5-year survival, age-adjusted, males only 1957-1976

<table>
<thead>
<tr>
<th>Squamous carcinoma</th>
<th>Supraglottis Total 5-year survival (%)</th>
<th>Glottis Total 5-year survival (%)</th>
<th>Subglottis Total 5-year survival (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>In situ</td>
<td>3 0.0</td>
<td>89 88.5</td>
<td>2 59.2</td>
</tr>
<tr>
<td>Well differentiated</td>
<td>128 38.8</td>
<td>531 71.1</td>
<td>15 29.8</td>
</tr>
<tr>
<td>Moderately differentiated</td>
<td>120 33.8</td>
<td>215 67.2</td>
<td>17 13.6</td>
</tr>
<tr>
<td>Poorly differentiated</td>
<td>197 29.3</td>
<td>195 54.7</td>
<td>26 44.8</td>
</tr>
<tr>
<td>Not specified</td>
<td>99 37.4</td>
<td>276 71.4</td>
<td>11 72.3</td>
</tr>
<tr>
<td>Total number</td>
<td>547 33.8</td>
<td>1306 69.3</td>
<td>71 38.8</td>
</tr>
</tbody>
</table>

Source: Birmingham and West Midlands Regional Cancer Registry, 1986.

It is difficult to draw firm conclusions from observations of incidence beyond remarking that the social and racial differences probably reflect different habits, and in the case of cancer of the larynx along with those of the mouth and pharynx, tend to reflect the already recognized effects of tobacco and alcohol (Newhouse, Gregory and Shannon, 1980). Of interest is the relatively low rate of laryngeal cancer in the UK compared with the definitively high rates of (smoking-related) lung cancer. A small rise in incidence of laryngeal
cancer has been observed in England and Wales 1960-1968 and 1972-1978, a rise proportionately greater in females (McMichael, 1978; Muir, Nectoux and Stukonis, 1983) attributed to the rise in smoking during the 1939-1945 war. This rise is not apparent when only glottic tumours are considered. The already high and increasing levels of mortality from laryngeal cancer in France, Italy and Spain contrast with the rest of Europe, and examination of birth cohorts in France have led to the suggestion (Tuyns and Audigier, 1976) that alcohol plays a promoting role, particularly in the supraglottis.

**Presentation**

The *incidence* (as opposed to mortality) (Ramadn et al, 1982) of cancer of the larynx, in common with most head and neck cancers, increases with increasing age, but because of the reducing numbers of persons surviving as age progresses, the actual number of cases *presenting* for treatment falls with age. The peak age of presentation is therefore younger than that of maximum incidence (that of glottis in the UK being approximately 62 years). Supraglottic lesions present at an earlier age than those of the glottis. The trend of increasing incidence with age is not universal and does not apply, for example, in Spain (Zaragosa) (Waterhouse et al, 1982) and Finland (Taskinen, 1969) where the cancer is more often supraglottic. These observations suggest that there may be a significant difference in the aetiology and behaviour of these cancers compared with the glottic tumours which predominate in the UK.

**Classification**

Laryngeal lesions are diverse in their behaviour and prognosis and thus classification is particularly important. Attempts at classification were begun as long ago as 1876 (Isambert). Over a period of years (since 1954) the International Union against Cancer (UICC) has undertaken the task of establishing a classification of a number of cancers, the larynx being one of the first, and now agreement has been reached generally with the American Joint Committee (AJC) and other similar bodies (UICC, 1978) about what may become a definite classification, at least for a decade. The basis of the UICC classification is anatomical.

It is important to understand the purpose of classification. The original intention of classification was to enable different sources to standardize their material in order that numbers and extent of tumours could be compared. The various sites were selected because of both their anatomical ease of identification and also their behavioural homogeneity. The definitions of the original UICC proposals have changed slightly to achieve the original intent. There was never any primary intention to use the classification to promote any treatment, although clinicians have found it useful to use the UICC terminology for this purpose. Classification is fundamental in studies of epidemiology.

Tumours were originally designated 'intrinsic' and 'extrinsic' laryngeal tumours. The latter, and also the term 'laryngopharynx' have been discarded for they represent the present day hypopharynx (sometimes oropharynx). Even now there is some difficulty in definition, and the term 'marginal zone' has been selected to designate the area of difficulty. The TNM (UICC, 1978) classification of larynx (International Classification of Diseases for Oncology, 1977) is given in Appendix 11.1.
There is a lack of precise definition of the 'vocal cord'. Several interpretations are permissible: the free edge; the point where the vestibule meets the upper surface to a point or line 1 cm below; the free edge to 1 cm below; the free edge and the subglottis down to the upper border of the cricoid; that area of the cord beneath which lies Reinke's space, that is the upper surface and free edge of the membranous cord. The lack of definition of the cord may undermine the credibility of the TNM system, yet in practice the use of the last interpretation seems to create little real difficulty. This was the agreed definition at the Centennial Conference on Laryngeal Cancer in Toronto (1974).

Of all the sites of cancer the larynx not only has one of the more detailed and precise TNM classifications, but it is also one of the most well tried and useful. It is for this reason it justifies some degree of study.

**Staging**

Staging is the grouping together of (TNM) features which may share a level of prognosis or a certain treatment. Staging has not been utilized in this and subsequent discussion.

**Aetiology**

The cause of cancer of the larynx is not known. A number of possibly related factors (male predominance, some radial predilection, a greater incidence among urban dwellers) have been designated, and there is an indisputable relationship between tobacco and alcohol (US Surgeon General, 1979; Hinds, Thomas and O'Reilly, 1979). Radiation (Sakamoto, Sakamoto and Sugano, 1979), asbestos (Hinds, Thomas and O'Reilly, 1979), and a number of occupational factors have been cited (Elwood et al, 1984), but none can be regarded as conclusive in the manner of those causing, for instance, lung cancer. Laryngeal keratosis and leucoplakia (Hellquist, Olofsson and Gröntoft, 1981; Crissman, 1982) are related to carcinoma of the larynx, but metaplasia (although smoking-related) has not satisfied the criteria for designation as a clear aetiological factor (Auerbach, Hammond and Garfinkel, 1970). One of the highest alcohol-consuming populations in Europe - the French - also show the highest laryngeal carcinoma incidence in that continent. Contrary to common belief, the perceived consumption of spirits (Scotland, UK) does not have a similarly matching laryngeal cancer incidence (Waterhouse et al, 1982). There is almost invariably an associated social relationship between alcohol and tobacco, and thus a distinction between the two factors is difficult to make, and cohort studies in France (Tuyns and Audigier, 1976) seem to postulate a reduction of laryngeal cancer as a result of deprivation (during World War II) of both alcohol and tobacco (the opposite of the features of the apparent rise in incidence, particularly in women (UK), as a result of their increase in smoking and alcohol consumption during the same war period).

It is reasonable to accept that although no close and irrefutable aetiological factor can be designated, there are several often related environmental factors which are clearly associated with an increased incidence of cancer, that is tobacco, alcohol, environmental (urban) pollution, asbestos, therapeutic radiation (thyroid), as yet unidentified social and possibly genetic factors which affect racial groups, and certain uncommon occupational influences.
Symptoms of carcinoma

The symptoms of carcinoma of the larynx are not greatly different from those of any space-occupying lesion of the larynx, but certain features make a carcinoma more distinguishable.

Progressive and unremitting dysphonia. The feature of a malignant tumour is its relentless advance, although in the early stages, in particular, the dysphonia may be intermittent. A further consideration is the cancer which develops in one who suffers from chronic laryngitis, and these individuals are particularly at risk from delay in diagnosis.

Dyspnoea and stridor are more frequent, and as a sequel to neglected dysphonia, almost invariably indicate an advanced tumour. Subglottic carcinoma may present with these as the only symptoms.

Pain is a relatively uncommon and late symptom and more typical of supraglottic lesions. Pain referred to the ear is particularly sinister and should always promote a high suspicion of cancer.

Dysphagia is relatively rare but carries a worse prognosis because it almost invariably indicates invasion of the pharynx.

Swelling of the neck or larynx may reflect the direct penetration of a tumour outside the larynx and as such its origin, without other symptoms, may initially be difficult to distinguish conclusively. Secondary malignant deposits in the lymph nodes of the neck have few distinctive features other than their predominantly ipsilateral situation, usually in the upper/middle deep cervical chain. A prelaryngeal or tracheal lymph node must be distinguished from thyroid disease.

Cough and irritation of the throat may be early non-descript symptoms; haemoptysis is rare and is most often seen in a lesion of the (margin of the) epiglottis; while anorexia, cachexia or fetor are usually late symptoms.

Examination and diagnosis

Diagnosis will be made after consideration of

(1) history
(2) examination of the larynx
(3) examination of the neck
(4) general examination of the patient
(5) radiology
(6) clinical investigations
(7) histological examination.
History

The history follows from the symptoms already discussed. Of no small importance is the temporal factor, unfortunately not considered in the UICC classification. The rate of advance of a cancer is important in its progress and prognosis - thus a small lesion with a long history of symptoms suggests a slowly growing lesion, whereas a massive cancer with a short history inevitably has a correspondingly poor outlook. Cancer can coexist with or supervene in leucoplakia, chronic laryngitis, tuberculosis, etc, and the symptoms of cancer or of any of the other disorders are not necessarily distinguishable from each other.

Examination of the larynx

This is generally, initially, performed with a mirror (see Chapter 1). Any focal abnormality of the larynx can be a tumour, but typically a vocal cord lesion may appear as a warty enlargement on one cord, but variation from a nodule or thickening of a vocal cord through extensive hyperkeratotic sheets of epithelium to gross ulceration may be seen. In the supraglottis there is focal swelling, redness or ulceration, while a mass may be visible in the subglottis. The subglottis is often difficult to see, but a tumour may appear, most usually an asymmetrical swelling often masked by mucoid debris. A second difficult area may be the posterior surface of the epiglottis hidden by the backward curve of the tip. The laryngeal ventricle is a third area that it is difficult to assess, where the initial or only clue to the presence of cancer may a slight fullness. A feature associated with ulceration of these hidden areas can be pain, referred to the (ipsilateral) ear. It cannot be too strongly emphasized that any focal abnormality may prove to be malignant.

A most important feature to be assessed is the mobility of the larynx. Mobility of any moving part of the larynx can be impaired by invasion of the tumour into muscle layers and it carries a much more sinister prognosis. Mobility may occasionally be impaired by the sheer bulk of a large tumour, but as distinct from deeper invasion, this is uncommon and often uncertain. Subglottic lesions often limit vocal cord movement by invasion of either the muscles or the cricoarytenoid joint, and both indicate an advanced tumour.

Indirect laryngoscopy is satisfactory for most patients, and provides added information about mobility. However, flexible endoscope allow examination of almost every patient without anaesthesia which may prejudice the assessment of mobility. Furthermore, the subglottis can sometimes be examined.

Direct laryngoscopy is necessary for most patients for biopsy purposes; microlaryngoscopy is desirable in many.

Examination of the neck

This must be carried out carefully to identify the possible spread of tumour beyond the larynx either directly or by metastasis to the regional lymph nodes. The most frequent site of secondary deposits is the ipsilateral deep cervical chain, usually in the upper/middle region but confined to this area. Glottic tumours rarely metastasize (Table 11.4), while deposits in the lymph nodes are more frequent from subglottic and particularly supraglottic lesions. Nodes
invaded by subglottic lesions are often found in the upper mediastinum, an added reason for the relatively poor prognosis of these lesions. The frequency of ipsi-, and indeed, bilateral deposits derived from supraglottic cancer is also reflected in the prognosis. Occasionally deposits can be identified in the prelaryngeal nodes and, even more infrequently, beyond the cervical region. Examination must include an assessment of the number, mobility and level of the lymph nodes.

Table 11.4 Incidence of nodal metastases of carcinoma of larynx (males) 1957-1976

<table>
<thead>
<tr>
<th>Node status</th>
<th>Number with known node status</th>
<th>Node positive (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supraglottis</td>
<td>598</td>
<td>38.8</td>
</tr>
<tr>
<td>Glottis</td>
<td>1394</td>
<td>4.8</td>
</tr>
<tr>
<td>Subglottis</td>
<td>77</td>
<td>13.0</td>
</tr>
</tbody>
</table>

Source: Birmingham and West Midlands Regional Cancer Registry, 1986.

Some swelling of the larynx, whether widening or as a result of penetration of tumour through the cricoarytenoid membrane, may be felt. An enlarged thyroid lobe should suggest invasion by tumour.

General examination

A general physical examination is required to identify metastases, for example to the liver, and to assess the overall physical status of the individual who is likely to be subjected to an anaesthetic and biopsy, possibly surgery, radiotherapy or chemotherapy. This examination applies to any case of malignancy and need not be detailed here.

Radiological investigations

Chest

In many respects this is the most important investigation at this stage, for assessment of the chest not only indicates the presence or likely absence of distant metastases (and the chest is undoubtedly the most common site of such metastases in cancer of the larynx); but may also indicate the presence of other disorders. It is indeed, nowadays, an integral part, together with a clinical examination, of the assessment of the general physical status of the older adult.

Larynx

Radiological examination of the larynx is undertaken in order to attempt to delineate the extent of the tumour. It is described in detail in Chapter 2.
Clinical investigation

It is appropriate at an early stage to undertake laboratory investigations, that is a full haematological screen and biochemical profile including liver functions and serum proteins; in the past, serological tests for syphilis were regarded as essential, although in the UK the yield is unrewarding. A (urine) screen for diabetes, and electrocardiography are also indicated.

Histological examination

Currently it is normal to acquire a specimen (biopsy) of the tumour by direct laryngoscopy, and this is usually carried out under general anaesthesia allowing a careful and thorough direct examination of the tumour. Direct examination should include the use of a microscope in most circumstances. Biopsy material should include an adequate amount of tissue both from ulcerated areas and elsewhere if practicable. If the tumour is very small, care must be taken, first to obtain sufficient material, and second, not to damage normal tissue.

Table 11.5 Histological confirmation of cancer of larynx 1957-1976

<table>
<thead>
<tr>
<th></th>
<th>Histologically confirmed (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supraglottis</td>
<td>90.9</td>
</tr>
<tr>
<td>Glottis</td>
<td>94.1</td>
</tr>
<tr>
<td>Subglottis</td>
<td>89.0</td>
</tr>
</tbody>
</table>

Source: Birmingham and West Midlands Regional Cancer Registry, 1986.

The biopsy material is important on three grounds:

(1) Definitive diagnosis of malignancy is required. It is not possible in every case either to diagnose or to exclude the presence of malignancy purely by inspection, and all experienced surgeons have found this. Even the most benign-looking polyp or nodule has occasionally been found to be malignant. Conversely, some quite active looking keratoses may not be malignant. Tuberculosis in former times was a diagnostic complication (Hautant, 1937).

(2) Identification of the type of tumour. While squamous carcinoma is undoubtedly the most common, other rare forms of malignancy are found and need individual consideration.

(3) Differentiation, Often neglected, the degree of differentiation may be significant. Biopsy material may, unless representative, be misleading, and thus where possible several
biopsies should be taken. The degree of differentiation may scarcely be worthy of consideration when a prognosis is to be made (see Table 11.3).

**Difficulties in diagnosis**

**Negative biopsy**

If a biopsy of a malignant-looking lesion is found to be negative, the biopsy should be repeated, but on some occasions reliance is placed on clinical diagnosis alone (usually advanced cases).

**Keratosis**

Sometimes a keratosis yields a non-malignant histopathological diagnosis. It is often difficult to decide just when such a lesion becomes malignant.

**Previous radiation**

Previous radiation is a common source of dilemma. A low-grade perichondritis may prevent a larynx from returning to normal. Careful observation is required. If a larynx does return to normal after radiation treatment and changes later, recurrent cancer is the most likely cause.

**Miscellaneous conditions**

Various conditions such as chronic laryngitis, tuberculosis, syphilis and benign tumours may give rise to diagnostic confusion or difficulty (Hautant, 1937).

**Pathology**

The vast majority of malignant tumours arising in the larynx are squamous cell carcinomata. All other types of malignancy arising in the larynx are rare. The histopathological classification can be performed according to Broders (1920, 1932). To determine whether the initial biopsy specimen yielded prognostic information, Jakobssen et al (1973) introduced an eight-factor malignancy grading system and applied it to glottic carcinomata. Nuclear polymorphism, mode of invasion and the total malignancy score were the factors that were most important in predicting the outcome for the patients (Jakobssen, 1973).

A distinct variant of well-differentiated squamous cell carcinoma is the verrucous carcinoma (Ackerman's tumour), which is most frequently reported arising in the oral cavity, but makes up a small proportion of all laryngeal carcinomata (van Nostrand and Olofsson, 1972; Ferlito and Recher, 1980).

**Spread of laryngeal carcinoma**

The growth and spread of laryngeal carcinoma is determined to a great extent by the site of origin of the primary tumour. Important factors in determining the directions and extent
of tumour growth are the anatomic barriers produced by the laryngeal compartments described by Pressman (1956), Tucker and Smith (1962) and Pressman, Simon and Monell (1969). The growth and spread of laryngeal carcinoma is well described by Ogura (1955), Kirchner (1969) and Olofsson and van Nostrand (1973).

**Glottic carcinoma**

Most of the tumours arising in the glottic region originate on the free margins of the vocal cords which are covered by a squamous epithelium. According to the agreements reached at the Centennial Conference on Laryngeal Cancer in Toronto (1974), the glottic region comprises the free margins and the horizontal surfaces of the vocal cords and the commissures. The anterior commissure is defined as a line between the vocal cords measuring a few millimetres in height. The subsurfaces of the vocal cords belong to the subglottic region.

Glottic carcinomata may arise in, or extend to, the anterior commissure area, where there is only a thin layer of submucosa and a fibrous cord, 'the anterior commissure tendon', that separates the mucosa from the underlying cartilage. This explains the increased risk of cartilage invasion in the anterior commissure area compared with that for tumours involving other parts of the vocal cords where muscle and perichondrium intervene.

The anterior midline is the most frequent location for invasion of the laryngeal framework (Ogura, 1955; Olofsson and van Nostrand, 1973). When the framework is invaded this occurs most frequently in the ossified parts of the cartilage. Local bone destruction by osteoclasts active at the margins of the tumour precedes the tumour invasion (Carter and Tanner, 1979). The vascularization of the cartilage is of great importance too. Unvascularized cartilage has a great resistance to tumour invasion (Olszewski, 1976; Guerrier and Andrea, 1977).

Tumours involving or crossing the anterior commissure often extend below the cords and may then escape outside the larynx through the cricothyroid membrane anteriorly, sometimes using the preformed vascular channels. Tumours may also extend laterally to the conus elasticus and can then escape through the cricothyroid triangle - bounded by the cricothyroid membrane, the thyroid cartilage and the medial edge of the cricothyroid muscle.

When the vocal cord muscles are invaded the tumour may extend along the muscle bundles anteriorly or posteriorly and may then reach lateral to the arytenoid cartilage where the tumour comes close to the mucosa of the pyriform sinus. Invasion of the posterior cricoarytenoid muscle may occur. Tumour extension lateral to the arytenoid cartilage is difficult to assess by conventional laryngoscopical and radiological means, but may be assessed by computerized tomography. A widening of the thyroarytenoid space indicates such tumour spread. The mucosa of the pyriform sinus on the affected side should be included in the laryngectomy specimen in these cases to avoid a recurrence above the stoma.

Vertical extension of glottic carcinomata to the subglottis and/or supraglottis seems to occur more frequently than extension to the opposite side (Pressman, Simon and Monell, 1969; Olofsson and van Nostrand, 1973).
Fixation of the vocal cord indicates deep invasion with involvement at least of the thyroarytenoid muscle. When the posterior part of the vocal cord is involved, fixation of the cord may be the result of invasion of the arytenoid or cricoid cartilages or the crico-arytenoid joint. Perineural invasion may be another aetiological factor but is seen mainly in major carcinomata. Fixation of the vocal cord indicates that the laryngeal framework is invaded and/or that the tumours have spread outside the larynx through cartilage and/or through the cricothyroid membrane or spaces as in about 50% of the cases (Olofsson and van Nostrand, 1973; Olofsson, Lord and van Nostrand, 1973). This means that fixation in most cases is a contraindication to partial surgery. Impaired mobility indicates a more superficial invasion of the thyroarytenoid muscle and is often not a contraindication to conservative surgery.

With previously used diagnostic methods, more than 50% of patients with vocal cord fixation in glottic carcinoma were under-assessed, as methods were lacking to determine cartilage invasion and spread outside the laryngeal framework. Computerized tomography has become a valuable complement in the radiological diagnosis of laryngeal carcinoma and adds important information about deep tumour invasion, cartilage destruction and extension of tumour outside the larynx (Archer et al, 1978; Archer and Yeager, 1979; Gregor, Lloyd and Michaels, 1981; Mancusa, Calcaterra and Hanafee, 1978; Sökjer and Olofsson, 1981).

Several reports have stressed the risk of cartilage invasion for tumours which traverse the laryngeal ventricle to occupy at least the glottic and supraglottic regions and often also the subglottis causing a fixed hemilarynx. These tumours extend within the paraglottic space and spread outside the larynx through the cartilage and the cricothyroid space.

Supraglottic carcinoma

The growth and spread of supraglottic carcinoma is well studied by the use of whole organ (specimen) serial sections (Kirchner and Som, 1971; Olofsson and van Nostrand, 1973; McDonald, de Santo and Weiland, 19760.

McGavran, Bauer and Ogura (1961) noted that supraglottic carcinomata often had 'pushing margins'. They often involve both sides of the supraglottic larynx. Bocca, Pignataro and Masciario (1968) stressed that supraglottic carcinomata seldom extend to the glottic region because 'the larynx consists of two distinct parts, an upper and a lower part, whose line of demarcation runs at the level of the vocal cords'. The different embryological derivations and the various lymphatic supplies were also stressed. Supraglottic carcinomata do not always respect the glottic region, but the various opinions in reported series depend to a certain extent on the selection of the material examined (Szlezak, 1966; Olofsson and van Nostrand, 1973). Exophytic supraglottic lesions do not often extend to the glottic region and seldom invade the thyroid cartilage. Ulcerative lesions may extend down below the anterior commissure, and when doing so they have a great tendency to invade the thyroid cartilage (Kirchner and Som, 1971). Invasion of the cartilage does not seem to occur unless the tumours can be seen macroscopically extending below the anterior commissure. This observation is certainly important in selecting the patients for horizontal supraglottic laryngectomies.

Invasion of the pre-epiglottic space is a prominent feature of supraglottic carcinomata and especially for those that involve the posterior (laryngeal) surface of the epiglottis. The tumours may extend into the pre-epiglottic space through the fenestration in the epiglottic
cartilage or by destruction of the cartilage. The lateral parts of this space are in direct continuity with the paraglottic space (Tucker and Smith, 1962) and this is another, but not so common, pathway for the tumours to reach the pre-epiglottic space. Nearly all tumours that invade the pre-epiglottic space involve the laryngeal surface of the epiglottis, which in most cases can be assessed at laryngoscopy (Olofsson and van Nostrand, 1973). Invasion of the pre-epiglottic space occurs in 40% of all supraglottic carcinomata and in 70% of all epiglottic tumours (Ogura, 1955; Szlezak, 1966; Kirchner, 1969; Olofsson and van Nostrand, 1973; McDonald, de Santo and Weiland, 1976).

Supraglottic carcinomata may extend cranially to the valleculae and to the base of the tongue. Posteriorly, the tumours may extend to the arytenoid cartilage, invasion of which seems to occur only when the arytenoids are grossly involved by tumour (Kirchner and Som, 1971). The pyriform sinus can be involved by tumours riding over the aryepiglottic folds. The pyriform sinus may also be reached by deep invasion.

**Subglottic carcinoma**

Primary subglottic carcinomata are rare and are characterized by a tendency to grow circumferentially and to be extensive before symptoms, such as inspiratory stridor, occur. Invasion of the vocal cords may cause impairment of their mobility and thereby hoarseness for which the patient may seek medical advice.

Subglottic carcinomata can spread through the cricothyroid membrane anteriorly or through the cricotracheal space, for example posteriorly, or invade the trachea caudally.

**Lymph node involvement**

The lymphatics within the larynx can be divided into a supraglottic and a subglottic network, separated by the free margin of the vocal cords, which has a minimal lymphatic drainage (Rouvière, 1931). This explains the low incidence of lymph node metastases for tumours confined to the vocal cords.

The supraglottis is rich in lymphatics, which accounts for the high incidence of lymph node metastases for supraglottic carcinomata, 32% as reported by Som (1970) and 73% as reported by Baclesse (1949). Sand Hansen (1975) found that 44% of the patients with supraglottic carcinomata, but only 5% of those with primary glottic carcinomata and 6% of those with subglottic carcinomata, had palpable cervical lymph nodes at the time of initial diagnosis. In total 18% of patients with laryngeal cancer had lymph node metastases at the time of referral.

The incidence of palpable lymph nodes increases with the extent of the primary tumour. Of patients with T1 supraglottic carcinomata 17% had lymph node metastases compared with 47% for the T2-T4 tumours. For glottic carcinomata the highest incidence of lymph node metastases was found when the sub- and supraglottic regions were involved (17%) (Sand Hansen, 1975).
Distant metastases

Few patients present with distant metastases at the time of diagnosis of their laryngeal carcinoma. Secondary distant metastases are more common. Sand Hansen (19750 reported 11% with distant metastases, most of which occurred in the lung (6.8%). The occurrence of pulmonary metastases seemed to some extent to be influenced by the presence of lymph node metastases, the macroscopic appearance and histology of the tumour. Poorly differentiated, necrotic tumours and tumours with lymph node metastases had the highest incidence of pulmonary metastases.

Multiple primary tumours

A number of papers stress the occurrence of synchronous and metachronous second and third primary tumours in patients with head and neck cancer. Wagenfeld et al (1980, 1981) found 6.5% of second primary carcinomata within the respiratory tract in patients with glottic carcinomata and 12.3% in those with supraglottic carcinomata; more than half of these tumours were located in the lungs and have to be separated from metastases.

Prospective panendoscopic examinations in patients with neoplasms arising in the upper respiratory tract have yielded a high percentage of synchronous multiple primary carcinomata (McGuirt, Matthews and Kourman, 1982) and introduce interesting aspects in the clinical management of patients with upper aerodigestive tract malignancies.

Treatment

Almost all carcinomata of the larynx are treated. Like most head and neck tumours, the therapeutic problem is local, not merely because in the majority of cases there is no evidence of spread beyond the local tissue or regional nodes, but also because even in those cases where dissemination is evident, the local lesion is the one causing symptoms that require some form of management. Even temporary control of the local and regional disease may require energetic treatment.

Most cancers of the head and neck show a reduced or uncertain response to second treatments (the separate treatment of regional nodes which appear after the treatment of the primary tumour is excepted from this statement), but the larynx is in this respect rather different. The so-called 'salvage' surgery of the primary lesion is thus a legitimate approach and must be considered in the treatment planning (Lederman and Dalley, 1965; Bryce, 1972; Stell et al, 1982).

Treatment may fall into the following categories:

(1) no treatment
(2) palliation
   (a) pain relief
   (b) tracheostomy
   (c) other surgery
   (d) radiotherapy
   (e) chemotherapy
(3) curative (radical)
   (a) radiotherapy
       radioactive implants
       megavoltage
       neutron
   (b) surgery
   (c) (chemotherapy)
(4) rehabilitation.

Since squamous carcinoma makes up the overwhelming proportion of malignancy of the larynx, the discussion will refer almost exclusively to this disorder. Rare forms of malignancy of the larynx will be dealt with separately.

No treatment

A few patients require no treatment, including those presenting in extremis, who are no longer conscious of pain or distress, or in whom disseminated tumours cause their death without the primary tumour or regional disease causing symptoms. In a retrospective sense this category also includes those who do have cancer of the larynx but who die of other disorders, the laryngeal cancer unrecognized. The fact that cancer of the glottis, especially the small lesion of the vocal cord, has a relatively long natural history may occasionally raise the question of the necessity of any treatment in those who have another debilitating or lethal disorder, and a careful judgement is required before instituting treatment at all. Approximately 7-8% of patients presenting clinically with carcinoma of larynx receive no treatment (Stell, Morton and Singh, 1983).

Palliation

It is appropriate to consider palliation of cancer of the larynx at an early stage, not because of its importance numerically, but because it is necessary to consider carefully the purpose of the treatment and the means used. Palliation indicates the attempt to suppress the carcinoma and its symptoms but without expectation or intent to cure. It may be of little value if the result is short-term, and allows the disease to recur in a more distressing or painful form. Palliation which, for instance, suppressed the disseminated disease but not the troublesome primary lesion could be unwelcome. Palliation is commonly used in the later stages of disease.

Pain relief

Pain is not particularly common in laryngeal cancer. Paradoxically it is the slowly growing, sometimes underestimated and thus undertreated, lesion which can be the most intractable. A carcinoma with a long previous history, often of recurrences, is likely to take a long time to destroy the sufferer, and once pain develops a long period of control may be required. such patients need the benefit of pain control specialists and the various adjuvant psychological, electrical and pharmaceutical methods. Such patients, too, may be subjects for treatment by a combination of methods including radiation, surgery and chemicals.
**Tracheostomy**

The relief of airway obstruction in a patient with incurable cancer often provides a dilemma. Dyspnoea is indeed distressing and its relief in such patients may merely delay for a very short period the inevitable death and on some occasions preserve a life temporarily only for other suffering and pain. These patients often provide problems, not only because a decision may be required rapidly, but also because the individual may be unable to indicate his/her wishes. Discussion with relatives is important, and often the time gained by tracheostomy is required for the relatives and the patient to come to terms with the unpleasant reality. It is this situation which requires the most informed and sympathetic management by the surgeon.

Tracheostomy may well be required as a preliminary before the possibility or practicability of treatment has been assessed. Tracheostomy in these cases also requires considerable thought and judgement, but is more appropriately discussed in relation to curative management.

The third instance where tracheostomy may be considered as a palliative procedure is in conjunction with other palliative procedures. Certain lesions are incurable - examples are all those with disseminated metastases, or those already treated where the airway becomes prejudiced and recurrence(s) are identified which may be temporarily controlled by chemotherapeutic agents, allowing a period of relative well-being. Each case must be considered upon its own merits.

**Other surgery**

Too often surgery, otherwise regarded as 'radical', is neglected as a palliative procedure. On occasions a total laryngectomy is the most reliable method of pain control, and a radical neck dissection, even in those with disseminated metastases, may remove a fungating or painful local lesion which is otherwise difficult to control and which is the source of the current symptoms. Such treatment, permitting sometimes months of relative well-being, may be a simple substitute for debilitating and time-consuming radiotherapy or chemotherapy, the modes of treatment usually invoked for palliation but which often impose more discomfort than the cancer itself.

**Radiotherapy (see Volume 1, Chapter 20)**

Radiation therapy is commonly used for palliation. Radiation is of particular value under these circumstances because it can be applied locally and selectively, focusing on the area or cause of symptoms; thus cases in which the primary lesion is obtrusive, or those where local (or even distant) metastatic lesions are the major problem, may be treated sufficiently to suppress those symptoms.

Usually the approach to palliation is simpler than for curative therapy. A complicating factor may be larger volumes requiring treatment (the larger the volume the less radiation per unit volume can be given) and less likelihood of complete control - often the original reason for the palliative nature of treatment. Sometimes the detailed planning and preparation is waived since these exercises are designed to promote maximum dosage to an accurately
limited volume, the need for accuracy being heightened by the desire to treat to tolerance, while tolerance is not sought when palliation is required. Radioactive implants of gold are useful for local treatment, especially of secondary deposits. Palliative courses of radiation can be delivered in fractions, but over a shorter period, since extension of the period is desirable only where treatment to tolerance is sought. Even so, there are limitations to the value of palliative radiotherapy and, as a general rule, one course to a particular area is offered, the patient, like any other, being at risk from the destructive effects of radiation if the appropriate dosage is exceeded.

**Chemotherapy (see Volume 1, Chapters 21 and 22)**

As yet (with a few anecdotal exceptions) no carcinoma of the larynx has been cured by drugs. Nevertheless, chemotherapy has become an integral component of the armamentarium used to manage cancer, although in realistic terms it is predominantly a palliative therapy. Its use as an adjuvant will be discussed in the section on radical treatment. For some 40 years single drug regimens, for example, nitrogen mustards, have been used in a variety of cancers particularly in the field of lymphomata. Intra-arterial perfusion was intended to maximize the effect in the tumour-bearing area, minimizing the toxic effects with a systemic antidote. The larynx was not suitable for this type of regimen. More recently, again following the lead from the lymphoreticular tumour, where protocols were based on biologically-observed cell kinetics, the trend has been towards (usually) the intravenous use of a multiple drug course of therapy, timed strictly over a prescribed period and repeated after an interval (Price and Hill, 1977; O’Connor et al., 1977). A variety of drugs has now been used including methotrexate, cyclophosphamide, 5-fluorouracil, bleomycin, hydroxyurea, vincristine, and cisplatin. In those cases where some response can be recorded, the inconvenience and, indeed, the risk of complications and side-effects, may be worthwhile and there are numerous reports describing the results of treatment with one or a variety of these agents (for example Perry et al., 1982).

Response to chemotherapy may be measured by partial or complete regression of tumours (as in phase II trials) or by the survival of the patient (phase III). Complete regression is rare while partial response, even then in only one patient in five, can demonstrate the effect of the chemical treatment, but is, in effect, a complete failure. Chemotherapy can in no way be compared with radiation or surgery, for survival has not yet been shown to be improved; rather it is an alternative to analgesics.

Chemotherapy has two other major disadvantages. First it is a 'blunt instrument' influencing cancer wherever it may be, that is both local and distant (where the local lesion is the one causing symptoms); indeed it may be negatively selective, being more effective against the metastases (perhaps better perfused) than the local lesion. The second objection is the tendency towards significant side-effects and the associated malaise, rather than a promotion of well-being, in a significant proportion of patients. It is as well to consider that suffering induced by non-curative treatment is hard to justify.

**Curative treatment**

Curative or radical treatment may involve radiotherapy, surgery, or chemotherapy, of which the last is not used alone. The first two, may be used either separately or together.
Radiotherapy

At the present time a radiotherapist is a specialist in his own right skilled in the application of all forms of radiation treatment, trained in medical physics and internal medicine at least to the extent of appreciating the interaction of disease with the treatment proposed and the variety of complications both early and late. The radiotherapist is also, essentially, an oncologist with experience of a very wide range of cancers, and until relatively recently the main author of chemical treatment. It is thus that the radiotherapist has a major role in the assessment and selection of treatment for most cancers, as well as cancer of the larynx. Joint clinics are now regarded as almost indispensable in the management of cancer of the larynx, particularly because most patients are treated by radiation. Radiotherapy is described in detail in Volume 1, Chapter 20.

Radiation is biologically most effective where the tissues are well oxygenated. This would seem to imply that it is most valuable in small lesions and where the vascular supply is undamaged, for example, where it has not been preceded by surgery. (There is no real evidence to suggest that surgery is actually prejudicial.) Attempts to use hyperbaric oxygen have not borne fruit (Hurley, Richter and Torrens, 19720. Conversely, the less attractive cases for radiotherapy are those where the tumour is large or widespread, not only because the volume is large and a tumoricidal dose more difficult to achieve without unacceptable side-effects, but also because the centre of larger tumours is often avascular and tends to be necrotic. In addition, the characteristics, even of megavoltage radiation, are such that tumours in bone are less responsive, and the cure of lymph nodes is uncertain. Radiation is thus theoretically more applicable on the oxygenated periphery of the tumour, while surgery could deal with the mass.

Selection of cases

Radiation is chosen in those cases where cure is likely with preservation of function. It may be used in a few circumstances where surgery is contraindicated or refused. Preliminary radiation of unresectable tumour is only rarely helpful. Radiation may be chosen, even if cure is uncertain, but with surgery in reserve. Radiation may be used for a majority of cases of cancer of the larynx.

The combination of radiation with chemotherapy may be considered, although the results of such treatments are not yet fully validated (Coker et al, 1981). Chemotherapy before radiation may increase the response but not survival.

Few circumstances contraindicate radiotherapy: active perichondritis, where cartilage is invaded; and where radiation has been used previously.

Interstitial radiation

Historically, radium-226 needles were implanted (Finzi and Harmer, 1928) by surgically removing part of the thyroid ala, with good results. This has now been superseded by external radiation.
Radioactive gold-198 grains can be inserted using a special gun in a pattern which can give a very high dose (100 Gy) localized to nodes or nodules in the neck, with little damage to normal structures.

Radiation reactions

Radiation reactions are frequent but usually not severe. They may be minimized by the avoidance of smoking, alcohol and careful attention to the skin and to nutrition. It is usual for the patients to develop mucositis, or painful erythematous reactions in the larynx and pharynx. In many cases, it is sufficient to require hospitalization for a week or so towards and after the end of treatment. Local and systemic analgesic mixtures are generally sufficient to control the symptoms. Rarely antibiotics and steroids are required. Similarly, erythema or moist desquamation of the skin may develop, and patients are discouraged from washing or abrading the skin until a little time after treatment is complete. Severe reactions may progress to necrosis of the skin, although this is very rare except where the tumour originally involved the skin.

Perichondritis is a deeper inflammatory reaction of the laryngeal skeleton. Whether true necrosis of cartilage occurs is uncertain. The symptoms are persistent pain, earache, more severe dysphagia and, where sepsis supervenes, severe illness. Perichondritis may require the suspension of radiation treatment. Mild symptoms during or after the treatment usually respond to steroids and antibiotics. If perichondritis is severe, urgent removal of the larynx is required. Mild symptoms may occur some years after radiation, provoked usually by a respiratory tract infection. When laryngeal oedema follows radiation, it can be hard to establish whether perichondritis or local residual disease is the cause, and it can be immensely difficult to reach a decision. It is essential, however, to monitor such patients carefully, for if residual tumour is present but allowed to extend beyond the larynx, the prognosis is poor.

Generally, a patient who undergoes radiotherapy for carcinoma of the larynx must undergo a relatively strenuous treatment, because of hospitalization or travelling and the almost invariable interference with nutrition, together with general side-effects of malaise, weakness, anorexia, insomnia, followed by dryness of the mouth and throat, loss of taste, and sometimes pigmentation of the skin and telangiectasia with some subcutaneous fibrosis.

Radiation is not an easy course or 'soft option', and many patients who have been subjected to both surgery and radiation will readily volunteer that radiation was by far the most miserable form of treatment. On the other hand, treatment for many small glottic tumours is relatively easy.

Most patients who have undergone successful radiation therapy retain a good or useful voice. A number are subject to dysphonic episodes with respiratory tract infections. Treatment of larger tumours, particularly if the reaction was severe, may be followed by some change in the voice, but this is preferable to its loss by surgery.
**Neutrons**

Fast neutron radiotherapy (Catterall, 1977) has shown a capacity for local control of malignancy apparently in excess of the success of photons in some sites. Its value has yet to be the subject of closely supervised trials and it is not generally available.

**Surgery**

*Microendolaryngeal and laser surgery*

The advent of the carbon dioxide laser used with the surgical microscope has added further impetus to the treatment of the smaller cancers of the larynx. Carcinoma *in situ* can be treated by microsurgical excision (Kleinsasser, 1978; Hellquist, Olofsson and Gröntoft, 1981) and laser surgery makes this even easier. Ideal cases are uncommon and require long and careful follow-up. There is an attraction to avoid radiation in younger persons where a sufficient length of life remains during which the risk of radiation-induced or other cancers is significant. Certain localized supraglottic lesions may be excised using a laser (Schlechter and El Mahdi, 1984). The real value of this method is still not fully evaluated.

Lasers can be used for biopsy, for tumour reduction (Vaughan, Strong and Jako, 1978) and staging (Fried, 1984), cordectomy (Davis et al, 1982), treatment of local tumours (Strong, 1975) and, of course, many benign tumours, especially vascular ones (Strong et al, 1976, 1979).

*Excision surgery*

Surgical treatment of carcinoma of the larynx historically precedes radiation treatment (Gussenbauer, 1874). Almost invariably there is some risk, if not some prejudice to, or loss of, the voice - and in supraglottic laryngectomy risk to the protection of the airway. In contrast with this, radiation generally preserves function. Today surgery is not associated with the major risks of former years, and physical fitness or age are not often the limiting factors. Most patients are fit for treatment and all may generally be offered surgery or radiation, the choice being made according to the likely effective control of the cancer and the relative consequences of the treatment.

Surgery is effective in almost all cases where the lesion can be encompassed. As a primary or sole treatment it is more effective than radiation in larger tumours and where there are secondary deposits of carcinoma in the lymph nodes of the neck. Rehabilitative procedures do much to minimize the disability after surgery. Surgery, however, may be used in combination with, or as a sequel to radiation therapy, especially in its role as 'salvage' or secondary surgery for recurrence, in many cases without detriment to the expectation of cure (Bryce, 1972; Stell et al, 1982). Partial resection of the larynx may also maintain near normal function with high rates of cure (Ogura, Sessions and Spector, 1975). After radiation failure, surgery is the appropriate course for most potentially curable cases.
Selection of treatment

With the principles outlined above, the selection of treatment of cancer of the larynx can be outlined.

Those cases of doubtful malignancy, keratosis and those with carcinoma in situ in the glottis and supraglottis may be treated by microendoscopic removal (Kleinsasser, 1978) and with the help of lasers (Strong, 1974, 1975). Small tumours of the marginal zones (suprahypoid epiglottis, aryepiglottic fold, sometimes false cord) may also be candidates for such surgery. Once the need arises to remove tissue which may interfere with function (for example, the vocal cord), alternatives should be considered.

Smaller tumours of the supraglottis and glottis (especially T1a and T1b lesions) are usually treated by radiation with good results, although some regard many such lesions of the supraglottis, especially those arising from the base of the epiglottis and the false cords, as most appropriately treated by conservative (horizontal partial) laryngectomy (Schechter and El Mahdi, 1984) with good results (Ogura and Mallen, 1967; Bocca, Pignataro and Masciaro, 1968). The relative values of radiation versus surgery are not clear. T2 lesions are treated by radiation - the alternative being total laryngectomy. Larger T3,4 lesions may best be excised if surgery is the only mode of treatment to be used. T3 glottic lesions may be treated by radiation (Hunter and Palmer, 1980) with salvage surgery without a reduction in cure (Bryce, 1972; Stell et all, 1982). Subglottic lesions too may be irradiated (Lederman, 1970) with possible secondary surgery if required, but follow-up with endoscopy, is a necessary part of management. Where there are secondary nodal deposits the use of primary surgery is much more appropriate, particularly if conservative surgery is possible for, although small or subclinical nodes are effectively controlled by radiation (Fletcher, 1972), the expansion of the field may make cure more difficult, and conservative surgery after radiation is less attractive (Radcliffe and Shaw, 1978).

Other less common forms of malignancy of the larynx are almost invariably treated by laryngectomy; these include adenocarcinoma (Whicker et al, 1974), verrucous carcinoma (van Nostrand and Olofsson, 1972), salivary adenocarcinoma (Spiro et al, 1976), fibrosarcoma (Flanagan, Cross and Libcke, 1965), melanoma (Moore and Martin, 1955), chondrosarcoma (Swerdlow, Som and Biller, 1974) and chemodectoma (Adlington and Woodhouse, 1972).

Emergency laryngectomy

This controversial issue was proposed by Kiem, Shapiro and Rosin (1965) as a result of their studies of 116 laryngectomy patients of whom 17 developed peristomal recurrence. Peristomal recurrence occurred without particular reference to the original site (supra/subglottic) but more frequently in those who had tracheostomy as a preliminary procedure at the time of laryngectomy (13.9%) and even more frequently (40.9%) in those whose tracheostomy preceded laryngectomy by 2 days or more. Peristomal recurrence was uniformly fatal. Baluyot, Shumrick and everts (1971) described the policy, and also the role of radiotherapy.

The difficulties in achieving the proposed ideal management are, naturally, the possible lack of availability of frozen section histology, and the doubt whether a patient with a
compromised airway can give rational consent to a laryngectomy. Nor has the effectiveness of the policy been demonstrated either in terms of reduced peristomal recurrence or mortality.

**Surgical techniques**

Many different techniques for treatment of laryngeal cancer have been devised since Buck in 1853 first performed a successful partial laryngectomy by laryngofissure in the USA, and Billroth of Vienna carried out the first total laryngectomy for cancer in 1873 (Gussenbauer, 1874). All the subsequent techniques have derived from the work of these early pioneers, and later modifications have concerned mainly such details as anaesthesia, skin incisions, succession of stages and methods of closure.

In the early years of the present century, although these operations were increasingly used, serious complications were the rule and operative mortality caused by haemorrhage, wound infection and bronchopneumonia was at times as high as 25% for the smaller procedures and 50% or more after complete removal of the larynx.

From about 1910 onwards this toll was gradually reduced through the work of pioneers such as Gluck (1922), and Thomson and Colledge (1930). Their achievements were obtained not merely by technical skill, but by a realization of the importance of adequate preparation of the patient before surgery and careful postoperative nursing to combat the dangers of infection.

After 1940, with the advent of surgical aids such as antibiotics, safer anaesthetics and blood transfusion, the whole scene changed. There are now few definite contraindications to laryngeal surgery and serious complications are rare. Operative mortality for major laryngeal operations is no more than about 1%.

The following types of procedure are now used:

(1) vertical partial resection  
(a) cordectomy  
(b) frontal partial laryngectomy  
(c) lateral partial laryngectomy  
(d) frontolateral partial laryngectomy  
(e) extended frontolateral partial laryngectomy  

(2) horizontal partial resection  
(a) epiglottectomy  
(b) supraglottic partial laryngectomy  
(c) extended supraglottic partial laryngectomy  

(3) total resection  
(a) total laryngectomy alone  
(b) total laryngectomy with partial pharyngectomy or partial glossectomy.

The operation of radical block dissection of cervical lymphatics on one or both sides may need to be combined with any of these procedures. It is seldom required in the first group owing to the paucity of lymphatics in the true cords and therefore the improbability of
metastatic spread. In addition, partial or total thyroidectomy may be obligatory in the major resections.

Lateral or vertical partial laryngectomy, often termed 'laryngofissure', is today performed less frequently owing to the equally effective results achieved by radiation in suitable cases. However, it still has a very definite place in situations where good radiation is not available, in some cases of radiation failure, possibly for limited cordal tumours in young adults, where radiation may provoke future neoplastic changes, and perhaps also in a few older patients unsuitable for prolonged radiation. Primarily it is indicated for T1a lesions of one vocal cord which should not extend into the anterior commissure or on to the arytenoid cartilage. It is also a suitable operation for the removal of many large benign laryngeal tumours.

Frontolateral partial laryngectomy may be useful where a glottic tumour crosses the anterior commissure to involve the anterior third of the opposite cord and without any reduction of mobility ('horseshoe tumour') (Som and Silver, 1968).

The older more anatomical hemilaryngectomy is no longer employed, but provided that there is no evidence of deep infiltration, an extension of the lateral partial laryngectomy technique to include the whole ventricular band and arytenoid cartilage may on occasion be used in highly selected cases (Ogura and Mallen, 1967), and is often termed 'hemilaryngectomy'. This term is also rather loosely applied to other types of partial resection.

Pharyngotomy, either by the anterior transverse approach or by the lateral route (Trotter, 1926), provides a satisfactory access to limited T1 or T2 supraglottic tumours. The anterior pharyngotomy approach is also useful in excising small tumours of the tip of the epiglottis and marginal aryepiglottic folds (Martin, 1957). The larger operation of supraglottic horizontal partial laryngectomy for T1a and T1b lesions of the epiglottis and laryngeal vestibule is more modern in concept. Although popular in Europe and centres in the USA (Bocca, Pignataro and Masciaro, 1968; Ogura et al, 1969; Some, 1970; Cachin, 1974), it is still not widely practised in the UK partly because of the relative rarity of suitable cases. However, it deserves greater acceptance in view of the consistently good results published compared with those of radiation. Primarily it is indicated for T1a or T1b lesions confined to the supraglottis. Transglottic extension or involvement of the tongue base usually contraindicates the procedure. It is also inadvisable in poorly differentiated lesions, in patient much over 65 years and those with reduced pulmonary function (Som, 1970), for these accommodate relatively poorly to the changed swallowing and compromised laryngeal sphincters.

Consent for total laryngectomy must always be obtained before attempting any type of partial resection and prudence indicates that the operation should start with a direct endoscopy. Previous full-dosage radiation is certainly no bar to lateral partial laryngectomy but greatly increases the hazards of the horizontal supraglottic operations. Nor should surgery be performed without a precise knowledge of the extent of the lesion before radiation (Radcliffe and Shaw, 1978; Stell and Ranger, 1974).

Preparation for surgery. It is a most important preoperative measure in this type of surgery to ensure the health of the mouth as far as possible. Unhealthy teeth should be treated
and particular attention must be paid to the periodontal tissues. Operation ought not to be undertaken until the mouth has healed. Nasal sepsis should be eliminated as far as is practicable. Attention must be given to general health, haemoglobin level, chest, and the control of other medical conditions. A systemic antibiotic should be given commencing at the beginning of operation (for example, ampicillin).

From a psychological point of view the surgeon will naturally assess each case individually. It is highly desirable that the patient is informed in some detail as to what the procedure is and what to expect, for thus is obtained not only the necessary informed consent but the better cooperation of the patient during the postoperative period.

Anaesthesia. The operations can be performed under local anaesthesia, but general anaesthesia is preferable, and essential for the more exacting supraglottic (horizontal) partial laryngectomy.

Where the resting airway is seriously reduced by the tumour, there should be no hesitation in carrying out a preliminary tracheostomy under local anaesthesia with insertion of an angled cuffed anaesthetic tube. As soon as the trachea is exposed, the cuffed tube is inserted and general anaesthesia is induced.

Position of the patient. This should be similar to that used for tracheostomy; the head is extended by placing a roll or a sandbag beneath the shoulders. It is important that the pillow or sandbag be evenly placed in order to ensure that the larynx and trachea are strictly in the midline.

Lateral (vertical) partial laryngectomy

Using this technique it is possible to remove completely many tumours confined to the vocal cord, with an adequate margin of healthy tissue and without removal of the arytenoid cartilage. The principles of operative surgery for malignant disease elsewhere apply, with the exception that it is unnecessary to remove the associated cervical lymph nodes for reasons already given.

The tumour must be removed in one piece with as wide a margin of apparently healthy tissue as is practicable. It has been suggested that this margin should be at least 0.5 cm and, although this suggestion is of some practical value, the tissue removed must be as much as is possible and prudent. Even in the earliest case it should consist of the whole of the side of the larynx anterior to the arytenoid cartilage, including its tip, and from the upper border of the cricoid cartilage below to the upper border of the thyroid ala above. It may be more than 1 cm above, but may be less anteriorly if the tumour approaches the commissure, or less posteriorly if it approaches the tip of the arytenoid cartilage.

Naturally, the smaller margin of healthy tissue removed, the less satisfactory are the results likely to be.

If, on histological examination of the specimen removed at operation, any doubt exists as to the complete removal of the lesion with an adequate margin of healthy tissue, a full course of radiation should at once be given if the larynx has not previously been irradiated.
However, in the latter event, vigilant follow-up alone is permissible with recourse to total laryngectomy at the first sign of recurrent disease.

**Incision.** A transverse incision over the upper border of the cricoid cartilage gives adequate exposure with a separate lower one for the tracheostomy.

The strap muscles are separated and retained for later use. The thyroid isthmus is divided. The tracheostomy is prepared by excising a disc of the anterior tracheal wall or by a midline incision only.

Excision of the thyroid ala is achieved by first separating the outer perichondrium as far as the oblique line, and sectioning the cartilage (with a saw) in the midline and laterally.

**Excision of tumour.** The midline (or just to the contralateral side if the tumour reaches the commissure) is incised, and the growth excised, above through the ventricular band, below just above the cricoid, and posteriorly including the tip of the vocal process of the arytenoid.

**Closure.** After haemostasis, and using the sternohyoid muscle placed inside the preserved outer perichondrium to reduce the dead space, the perichondrium is sutured in the midline and the wound closed in layers. A nasogastric tube is inserted.

**Postoperative care.** The main complications of haemorrhage and chest infections are avoidable, the former by meticulous haemostasis, the latter by care of the tracheostomy (see Chapter 9). Early activity should be encouraged. Swallowing is painful at first, and nutrition is by nasogastric tube for a few days. The tracheostomy may be closed when the airway is demonstrably adequate. Antibiotics should be continued for some weeks, especially if there has been previous radiation. Granulations may form over unepithelialized cartilage and will need to be removed.

**Late results of operation.** The larynx heals by slow fibrosis and epithelialization, and this rarely results in stenosis, especially if the thyroid ala has been removed. Usually, however, a fibrous replica of the cord is produced which, after a year, may become a very passable substitute for the true cord. Conley (1961) claims better healing and an improved voice by fashioning a new cord from an inturmed flap of cervical skin used to line the raw side of the larynx. Other reconstructive techniques have been tried.

There is no satisfactory movement of this band of scar tissues and, although the voice will be useful, it is husky, variable in strength and usually cannot be said to approach the normal. Singing is impossible. There is often some stenosis of the anterior part of if the opposite cord has been removed as in the frontolateral operation. Marked stenosis may limit the airway, and in these cases the tracheostomy is retained using a valve to permit phonation.

Local recurrence either in the scar or in adjacent tissue is uncommon. If a growth later occurs on the opposite cord it is difficult to decide whether it is a recurrence or a second primary growth. Metastasis is rare but may become evident in the cervical or mediastinal lymph nodes months or years after operation and with no evidence of local recurrence in the larynx.
Supraglottic partial laryngectomy

Incision. The T-shaped incision described by Som (1970) or a simple transverse incision is adequate. A tracheostomy is performed.

Approach to supraglottis. After elevation of skin flaps, the strap muscles are divided from the hyoid bone and the superior laryngeal vascular bundles ligated, defining the hyoid, superior cornua, and posterior borders of the thyroid cartilage. If a neck dissection is required, or is indicated, it is performed at this stage. The external perichondrium is dissected and the cartilage sectioned horizontally.

Exposure of the tumour. The pharynx is entered laterally just below the greater cornu of the hyoid. The hyoid bone is removed. The incision is carried across the base of the tongue, maintaining an adequate margin until the tumour can be viewed from above.

Excision of the tumour. Working from above, the tumour is excised with scissors with a blade each side of the aryepiglottic fold, preserving the arytenoid. Extended operations (Ogura, Sessions and Spector, 1974) are described.

Cricopharyngeal myotomy. A myotomy is performed, and a nasogastric tube inserted.

Closure. After haemostasis, closure is of the raw surfaces first by suturing the cut edges of mucosa and then of the pharynx in layers by approximating the base of the tongue to the cut edge of the thyroid cartilage, and the infrahyoid muscles to the cut suprahyoid muscles, and finally of the skin.

Postoperative care. A routine similar to that for partial laryngectomy and tracheostomy is followed, but even with particular care, spillover and aspiration are almost invariable.

Rehabilitation. Gradual use of the voice is begun after about 7-10 days. Once spillover is reduced, oral food, initially semisolids, is begun. This aspect of care is paramount for patients after horizontal partial laryngectomy, and it is because of this difficulty that the operation may not be appropriate in those with reduced pulmonary function and who are aged much over 65 years; previous radiation may also be a contraindication (Radcliffe and Shaw, 1978).

Total laryngectomy

Billroth of Vienna first removed the larynx as a treatment for cancer in 1873 (Gussenbauer, 1874).

At first, the results of operation were bad and not a single patient survived for one year in the first 25 cases recorded. Postoperative complications were frequent and severe, the most common causes of death being general septicaemia, spreading cellulitis or mediastinitis, septic pulmonary complications, haemorrhage and shock.
An attempt to improve on these poor results was made by performing the operation in two stages, the first consisting of the establishment of a tracheostomy, the larynx being removed a few weeks later.

Single-stage laryngectomy was suggested in 1921 by Moure and Portmann and, since that time, most surgeons have practised a one-stage operation with increasing safety.

**Incisions.** A commonly used incision was first recommended by Gluck (1922) and modified by Soerenson (1930). The incision commences on the anterior border of the sternomastoid muscle about the level of the hyoid bone, passes down along the anterior border of the muscle for about 6–7 cm and then curves across the midline at the level of the second or third ring of the trachea.

Other incisions are used, especially for combined total laryngectomy and radical neck dissection, and all have advantages. Jackson's single vertical incision is not now used. A single horizontal incision midway between the hyoid bone and the sternum with a separate one for the tracheostomy, is adequate and can be adapted for neck dissection.

**Exposure of larynx.** The larynx is dissected from the sternomastoid and carotid sheaths on each side, tying the vascular bundles. The strap muscles are section low in the neck, and usually the contralateral lobe of the thyroid freed from the specimen. The infrahyoid muscles are section from the bone.

**Tracheostomy.** The trachea is divided below the tumour and a tracheostome fashioned now or later.

**Removal of the larynx.** This is easier from above downwards, sectioning the constrictor muscles from the thyroid cartilage, and preserving mucosa as far as appropriate. Care should be taken to ensure adequate margins, especially over the cricoarytenoid joints.

**Closure.** Closure of the pharynx is in layers in the shape of either I or Y, ideally with a Connell suture. A long posterior myotomy (Singer and Blom, 1981) may avoid segments of spasm. The skin is closed in layers over suction drains.

**Postoperative care.** This is as before, and much as for tracheostomy (see Chapter 9). Feeding is by nasogastric tube. Antibiotic cover (for example ampicillin) may be continued for a few days.

**Complications.** The most frequent complication (apart from those associated with any surgery) is pharyngocutaneous fistula. This may develop at 4–10 days with a collection under the skin which requires drainage. Most fistulae close spontaneously after some days or weeks, assisted by frequent dressing, adequate nutrition and maintenance of the haemoglobin level. They are prevented by good technique, haemostasis, antisepsis, and a well-prepared patient. Rarely, extensive sloughing and large fistulae need repair by full thickness distant flaps.
Other complications include haemorrhage, wound infection, pulmonary and cerebral embolism, cardiac infarction, tracheal crusting and stomal recurrence, and the chest complications associated with any laryngeal operation and tracheostomy.

**Thyroid insufficiency.** In some cases a total thyroidectomy is necessary as part of the surgical procedure. In such cases, thyroid replacement in the form of L-thyroxine is required, 0.1-0.3 mg daily. Replacement therapy is not necessary immediately but can await the resumption of swallowing in 1-2 weeks.

Rather more insidious is the hypothyroidism which may follow surgery and radiotherapy. While the patient may have retained one lobe of the thyroid, previous or later radiotherapy may on occasions impair the function, and hypothyroidism may supervene even after several years. Treatment is straightforward once the condition is recognized.

**Parathyroid insufficiency.** This usually, but not invariably, follows total thyroidectomy. The problem may be acute in the immediate postoperative period. It may be delayed, or it may be temporary and may occur even after partial thyroidectomy.

In cases where it is anticipated, pretreatment with a vitamin D preparation may reduce the postoperative problem; otherwise intravenous calcium maintenance is necessary until either control is achieved by vitamin D medication or the condition resolves.

**Respiration.** Most surgical procedures on the larynx require a tracheostomy to maintain the airway during and often after the surgical procedures. Total laryngectomy requires a permanent end-tracheostomy, and the maintenance of respiration depends on a good formation of the tracheostome and good healing without crusting. Most patients require no special care afterwards but, not infrequently, a respiratory tract infection is followed by tracheitis and crusting, and a short period of treatment may be required to control it.

A partial laryngectomy ideally does not require a permanent tracheostomy, and the tube may be withdrawn once the natural airway can be shown, by occluding the tube, to be adequate. However, the benefits of retaining part of the natural larynx in respect of phonation are so great that the retention of the temporary tracheostomy should not be regarded as of overriding concern and should be infinitely preferable to a total laryngectomy if the control of the cancer permits it. However, the retention of a tracheostomy in a horizontal partial laryngectomy may in itself be prejudicial to the recovery of the effective protective function of the glottic sphincter. Fine judgement may be required.

**Swallowing after total laryngectomy.** As a rule this presents no difficulty. Even while a nasal feeding tube is in position a patient can swallow his saliva and drink fluids quite satisfactorily, although the act of swallowing is painful for the first few days. If healing has proceeded well and a fistula is not anticipated, it should be possible to remove the feeding tube after about 6 days, but if there has been previous radiation it would be much wiser to leave the tube in position for about 10 days since it is often not until the tenth day or later that the pharyngeal wound tends to break down. If the wound does break down, the tube should be left in position until the full extent and size of the fistula is revealed.
**Functional rehabilitation of voice.** Because the voice is impaired by operations on the larynx, the cooperation of a speech therapist is essential in maximizing its recovery.

Partial vertical and horizontal laryngectomy are operations designed to retain at least part of the glottis. The benefit of this is quite clear in that it utilizes the remains of the glottic vibrator in order to phonate, and the pulmonary reservoir to permit the production of a flow which can be modulated, and sufficient to produce sentences rather than a few words. The cords are often intact after horizontal partial laryngectomy and the voice is relatively little impaired, while vertical partial laryngectomy rarely allows a normal voice. In both cases a speech therapist can improve the production and efficiency in voice production.

**Oesophageal voice.** Many patients who have undergone a total laryngectomy have been able to develop abnormal but functionally satisfactory oesophageal speech. This mechanism relies upon the subject charging the oesophagus with air and utilizing the vibrations, at either the cricopharyngeal sphincter level or another level, to phonate (Dworkin and Banton, 1982). The preoperative and postoperative assistance of a speech therapist is essential in most cases, not only to teach the mechanism of voice production, but also to improve and teach other normally subsidiary communication methods. While it is optimistically said that 60% of laryngectomees develop satisfactory phonation, it is evident that a significant minority does not. Often the elderly do not have the capacity or even the will to phonate adequately, and even those who do so are restricted, because of the limitation of the oesophageal air reservoir, in the length of word sequences they can achieve. It is not surprising therefore that much ingenuity has been applied to assist laryngectomees to improve upon the relatively poor outlook (for speech) after total laryngectomy. Conservative surgery has developed because of its capacity to preserve speech (Bocca, Pignataro and Masciario, 1968; Ogura et al, 1960; Som, 1970) and has been applied in vertical partial laryngectomy since inception.

An early method of improving speech or voice production has been to develop a small tracheo-oesophageal fistula (Conley, de Amnesti and Pierce, 1958) which allows the oesophagus to be charged with air more continuously from the trachea (that is the pulmonary 'bellows' is brought back into play). Such a procedure relies upon the development of the oesophageal or pharyngeal vibrator, which is not developed by all patients.

**Neoglottis.** More definite attempts at laryngeal voice replacements are represented by the method of Staffieri (1974). This procedure also develops a tracheo-oesophageal fistula. One method uses the fistularized redundant mucosa derived from the postcricoid region positioned over the cut end of the trachea, and the second prepares a fistula through the posterior wall of the trachea. In each case the tracheostomy is made through the anterior wall of the trachea lower down and no end-tracheostomy is fashioned. Both these methods are intended to provide a form of neoglottis which itself generates a voice, but the second method, particularly, may act as a recharging mechanism for the oesophagus, utilizing a phonatory vibration in the pharynx.

The construction of such a neoglottis is as follows. A total laryngectomy is carried out without prejudicing the lower margin of the tumour, that is an adequate margin of the subglottis must be removed with the tumour. A tracheostome is constructed at a lower level in the anterior wall of the trachea. The mucosa of the postcricoid/anterior oesophageal wall
is preserved and positioned using the index finger over the cut end of the trachea. A vertical incision is made through the fasciomuscular layer and the mucosa drawn through, incised, and sutured to line the fistula. A filament is passed through the nose, fistula and tracheostomy to mark the fistula, the pharyngeal wall is sutured to the end of the tracheal and then closed as is usual after laryngectomy and the skin closed. Drains should not be placed too close to the reconstruction. Phonation is achieved by temporarily closing the tracheostome and directing air into the pharynx.

The Asai technique (Asai, 1965) is an ingenious method using a tube of skin from the upper end of the trachea to recurved under the based of the tongue, using, for example, a medially-based (deltopectoral) chest flap.

All these techniques are attractive but have the same problems. Their attraction is that, in the majority of cases, speech at least as good as the oesophageal speech is obtained, and often better because the reservoir (the lungs) of air is large. In addition, it is easy to ensure that phonation is achieved rapidly just as a patient with a tracheostomy but retaining the larynx is quickly returned to speaking normally. However, the disadvantages are those associated with a tracheo-oesophageal fistula, and no method has so far been described where aspiration has not been experienced - if the neoglottis is too wide, aspiration may occur; if too narrow, phonation is impaired.

Valves. In order to improve upon the achievements associated with the construction of a neoglottis, Singer and Blom (1980) and Panje (1981) devised replaceable valves which, when inserted, behave in a manner which is an advance on the quality of phonation usually produced by a neoglottis. Since they are one-way valves, aspiration may be reduced. Their disadvantage is the need to replace the valve regularly and the associated cost; nor are these methods entirely free of the risk of aspiration. However, both the Blom-Singer and the Panje valves can be inserted as a secondary procedure (Singer and Blom, 1980) after a standard laryngectomy and end-tracheostomy have been performed. An essential part of the procedure is the prevention of muscular spasm of segments of the pharynx, and Singer and Blom (1981) emphasized the need for a (posterior) myotomy of the pharyngeal wall at laryngectomy or as a secondary procedure. Hamaker et al (1985) have summarized the results.

A reasonable approach to voice rehabilitation may be as follows. A total laryngectomy with an end-tracheostomy is carried out and healing allowed to be completed. A myotomy of the pharyngeal musculature from the level of the tongue to the upper oesophagus is performed. This minimizes the operative risk and allows any adjuvant radiotherapy and chemotherapy to take place. Voice rehabilitation should be pursued and will be achieved by a proportion of patients. A later reassessment will permit the identification of those whose rehabilitation is inadequate, and a neoglottis or valve preparation can be provided as a secondary procedure. A practical compromise may be to construct a tracheostomy in the anterior wall of the trachea, leaving the end prepared with a pharyngeal wall cover, but not fistularized, in order to be able to construct the neoglottis more satisfactorily at a second stage if a Staffieri type is preferred to a replaceable valve.

Disability after total laryngectomy. The main disability from which the patient will suffer after operation is naturally the loss of the normal voice. The sense of smell is also
impaired because there is no regular air current through the nose, and taste and the appreciation of flavours are reduced.

The patient must take care that water does not enter the tracheostome when bathing or washing, and swimming must be prohibited.

Heavy lifting or strenuous digging are not possible as these actions entail fixation of the chest wall by closure of the larynx, but light physical work is possible and, occasionally, the patient can partially close the tracheostome by contracting any muscle remnants surrounding it. Some young women who have undergone this operation have subsequently married and borne children without difficulty (Shaw, 1965).

If radical neck dissection has been necessary, some reduction in the usefulness of the arm at the shoulder level may be expected, especially in the older age groups, together with a variable amount of persistent discomfort more evident in those whose range of movement is most impaired (Ewing and Martin, 1952).

Apart from these disadvantages, patients generally come to terms with their disabilities, adaptation being usually good in age groups up to 65 years. Unfortunately, many patients are older and less adaptable but with instruction and encouragement from surgeons, speech therapists, laryngectomee clubs and associations, the majority lead happy and useful lives although they are often socially limited.

**Other malignant ectodermal tumours**

**Verrucous carcinoma**

This rare carcinoma (approximately 1% of laryngeal malignancies) (Ryan et al, 1977) has a male predominance, causes the usual laryngeal symptoms, frequently has an extensive warty papillomatous appearance, and its site is glottic more often than supraglottic. Lymphatic spread is rare (Biller, Ogura and Bauer, 1971). Traditionally treatment is surgical (Burns, van Nostrand and Bryce, 1976) by partial or total laryngectomy. Radiotherapy has been employed, but anaplastic change and death have been reported after this (Burns, van Nostrand and Bryce, 1976), while uniformly good results follow surgery (and this may include local or biopsy excision) (Maw, Cullen and Bradfield, 1982).

**Carcinosarcoma**

An equally rare lesion (less than 1% of laryngeal malignancies) (Goellner, Devine and Weiland, 1973), carcinosarcoma presents with typical symptoms and usually a polypoid or pedunculated appearance (Brodsky, 1984). Treatment is by excision, with either partial or total laryngectomy. Radiation has rarely been used and is difficult to evaluate (Hyams and Rabuzzi, 1970).

**Adenoid cystic carcinoma**

Adenoid cystic carcinoma of the larynx is also very rare (Olofsson and van Nostrand, 1977) with the typical rather long history of laryngeal symptoms. The subglottis, in contrast
with other laryngeal malignancies, is the commoner site. Local or wide-field surgery is the generally favoured treatment, although radiotherapy is used on some occasions. The slow and relentless course of this tumour makes 5-year ‘cures’ meaningless and the vast majority eventually succumb.

Other rarities include anaplastic small cell carcinoma (Olofsson and van Nostrand, 1972). These tumours are treated with primary radiotherapy and adjuvant chemotherapy (Baugh et al, 1986).

**Malignant mesodermal tumours**

*Chondrosarcoma* is exceedingly rare, and usually occurs in the posterior segment of the cricoid. Surgery, inevitably in the form of total laryngectomy, is the only useful treatment in most cases, although tumours in certain locations could be excised conservatively (Hellquist, Olofsson and Grontöft, 1979).

*Non-Hodgkin's lymphoma* is rare, but once diagnosed is treated, as other extranodal lymphomata, by radiotherapy and/or chemotherapy. *Melanoma* (Conley, 1967) and *fibrosarcoma* are equally rare and are treated by excision.

**Summary and selection of treatment**

Assessment of the relative value of differing treatment methods for laryngeal cancer is not easy. Despite the gradual international adoption of the TNM classification system, confirmed in 1978 for the larynx, and greater uniformity in end-result reporting, most accounts are still based on retrospective studies. Adequate conclusions by randomized controlled prospective studies are still lacking, in part as a consequence of difficulty of application in cancer therapy. Despite this, much knowledge has been gained in recent years by extensive clinical experience using increasingly refined methods of surgery and radiation; also by the histological study of serially-sectioned laryngeal specimens (Kirchner, 1969; McDonald, de Santo and Weiland, 1976; Olofsson and van Nostrand, 1973). Important knowledge of lymphatic pathways within the larynx (Pressman and Simon, 1961) and patterns of local spread and cervical lymph node involvement has also been gained (Kirchner, 1974; Olofsson and van Nostrand, 1973).

Forty years ago radiotherapy was condemned for limited cordal cancer (Colledge, 1940). Today it is established as the treatment of choice in such lesions. In early glottic cancer (T1 and T2), modern radiation techniques still allow the surgeon to perform partial or total laryngectomy for failures with good salvage rates (Lederman, 1970; Bryce, 1972). In support of radiation for early glottic lesions, Lederman (1970) reported 5-year survival rates of at least 77% in his large series. Recurrence of glottic cancer after radiation and still treatable by lateral partial laryngectomy, can give a salvage for 3-year survivals of about 60% with preservation of voice and natural airway (Radcliffe and Shaw, 1978). In more advanced recurrent glottic or transglottic cancer after radiation, a 5-year survival rate of 47% by total laryngectomy can still be obtained (Stell et al, 1982).

On the other hand, 5-year survival results for initial treatment by radiation of smaller supraglottic and epilaryngeal cancer without nodal spread are very satisfactory (Lederman,
1970; Fletcher et al, 1970; Fletcher and Goepfert, 1977), but salvage for failures will generally require total laryngectomy. Such cases treated by supraglottic partial laryngectomy may give a 5-year survival of 70% with voice preservation and equally good chances of salvage by total laryngectomy for the failures (Som, 1970; Shumrick, 1971; Leroux-Robert, 1975; Ogura, Marks and Freeman, 1980). Primary conservation surgery therefore compares favourably with primary radiation treatment (de Santo, Willie and Devine, 1976). Supraglottic laryngectomy may be preferable in those cancers which include the false cord and base of epiglottis and those with metastatic nodes (Ogura, Sessions and Spector, 1975).

T3 glottic lesions, once automatically recommended for primary surgery or planned combined radiotherapy and surgery (Lederman, 1970; Sissons, 1974), should now be considered for primary radiation treatment with 'salvage' surgery if there is recurrence (Bryce, 1972; Stell et al, 1982). Larger T4 lesions are still much more appropriately treated by surgery, although some clinicians would challenge this.

The rare cases of true subglottic cancer are generally advanced when diagnosed and planned combined treatment by radical surgery with postoperative radiation to the lower neck and mediastinum probably gives the best chance of cure (Harrison, 1971; Bryce, 1972). However, Lederman (1970) has shown that where the disease is limited to the anterior half of the subglottic space with no palpable neck nodes and no vocal cord fixation, a 5-year survival rate of 60% can be obtained by radiation, particularly in women.

A more recently emerging treatment, apparently of value when used as an adjuvant method in advanced lesions (T3 and T4), is cytotoxic chemotherapy. Since 1974, the use of a kinetically based multidrug protocol has been on trial either before or after conventional therapy (Price and Hill, 1977) or integrated with radiation (O'Connor et al, 1977). Results are beginning to emerge from both techniques with minimal side-effects (O'Connor et al, 1979).

These results illustrate certain definite trends in the management of laryngeal cancer. First, that radiation gives excellent results in the early glottic lesions with absolute preservation of function. Second, that the balance of judgement between conservative and radical surgery of the larynx is becoming more refined, with the results of the former approximating to the latter for supraglottic tumours and at least equalling the results of radiation. Third, that the attack on cancer at the molecular level through cytotoxic drugs is now being applied to the larynx as an aid to conventional methods or combined treatment, again with cure and functional conservation as the objectives.

Perhaps in contrast with the foregoing has been the development of endolaryngeal procedures with the laser, permitting more accurate excision and, combined with careful follow-up, an even more conservative approach to non-malignant lesions, to carcinoma in situ and to a few very small invasive malignancies. The development of a more positive attitude to rehabilitation, including the neoglottis and valve techniques, should encourage a more definitive and constructive approach to laryngeal cancer in general. Whether still further advances can be made by prevention (that is alcohol reduction and a non-smoking generation) and by further therapeutic developments, remains to be seen.
Appendix 11.1 Larynx (ICD-0 161) (classified 1972, confirmed 1978)

Rules for classification

The classification applies only to carcinoma. There should be histological verification of the disease. Any unconfirmed cases must be reported separately. The minimum requirements for assessment are:

T (site): clinical examination, laryngoscopy and radiography
N (node): clinical examination
M (metastases): clinical examination and radiography
X indicates that minimum requirements for assessment cannot be met.

Anatomical regions and sites

(1) Supraglottis (161.1)

Epilarynx including marginal zone

(i) Posterior surface of suprahypoid epiglottis (including the tip)
(ii) Aryepiglottic fold
(iii) Arytenoid

Supraglottic excluding epilarynx

(iv) Infrahypoid epiglottis
(v) Ventricular bands (false cords)
(vi) Ventricular cavities

(2) Glottis (161.0)

(i) Vocal cords
(ii) Anterior commissure
(iii) Posterior commissure

(3) Subglottis (161.2)

Regional lymph nodes

The regional lymph nodes are the cervical nodes.
TNM pretreatment classification

* T - primary tumours

### Supraglottis

- **Tis** Preinvasive carcinoma (carcinoma *in situ*)
- **T0** No evidence of primary tumour
- **T1** Tumour confined to the region with normal mobility
  - **T1a** Tumour confined to the laryngeal surface of the epiglottis *or* to an aryepiglottic fold *or* to a ventricular cavity *or* to a ventricular band
  - **T1b** Tumour involving the epiglottis and extending to the ventricular cavities or bands
- **T2** Tumour confined to the larynx with extension to adjacent site or sites *or* to the glottis without fixation
- **T3** Tumour confined to the larynx with fixation *and/or* other evidence of deep infiltration
- **T4** Tumour with direct extension beyond the larynx
- **Tx** The minimum requirements to assess the primary tumour cannot be met.

### Glottis

- **Tis** Preinvasive carcinoma (carcinoma *in situ*)
- **T0** No evidence of primary tumour
- **T1** Tumour confined to the region with normal mobility
  - **T1a** Tumour confined to one cord
  - **T1b** Tumour involving both cords
- **T2** Tumour confined to the larynx with extension to either the supraglottis *or* the subglottis regions with normal *or* impaired mobility
- **T3** Tumour confined to the larynx with fixation of one *or* both cords
- **T4** Tumour with direct extension beyond the larynx
- **Tx** The minimum requirements to assess the primary tumour cannot be met.

### Subglottis

- **Tis** Preinvasive carcinoma (carcinoma *in situ*)
- **T0** No evidence of primary tumour
- **T1** Tumour confined to the region
  - **T1a** Tumour confined to one side of the region
  - **T1b** Tumour with extension to both sides of the region
- **T2** Tumour confined to the larynx with extension to one *or* both cords with normal or impaired mobility
- **T3** Tumour confined to the larynx with fixation of one *or* both cords
- **T4** Tumour with destruction of cartilage *and/or* with direct extension beyond the larynx
- **Tx** The minimum requirements to assess the primary tumour cannot be met.
**N - regional lymph nodes**

N0  No evidence of regional lymph node involvement  
N1  Evidence of involvement of movable homolateral regional lymph nodes  
N2  Evidence of involvement of movable contralateral or bilateral regional lymph nodes  
N3  Evidence of involvement of fixed regional lymph nodes  
Nx  The minimum requirements to assess the regional lymph nodes cannot be met.

**M - distant metastases**

M0  No evidence of distant metastases  
M1  Evidence of distant metastases  
Mx  The minimum requirements to assess the presence of distant metastases cannot be met.

(Note: The TNM system, with particular reference to N (nodes) is due for revision in 1987.)
Chapter 12: Tumours of the oropharynx

P. M. Stell and J. R. G. Nash

The oropharynx is that part of the pharynx extending from the level of the hard palate above to the hyoid bone below. Its boundaries for the purposes of classification of tumours are shown. The subdivisions of the oropharynx are shown in Table 12.1.

Table 12.1 Oropharyngeal sites (UICC and AJC)

(1) Anterior wall (glosso-epiglottic area)
   (i) tongue posterior to the vallate papillae (base of tongue or posterior third)
   (ii) vallecula (UICC only)
   (iii) anterior (lingual surface of epiglottis (UICC only))
(2) Lateral wall
   (i) tonsil
   (ii) tonsillar fossa and faucial pillars
   (iii) glossotonsillar sulci
(3) Posterior wall
(4) Superior wall
   (i) inferior surface of soft palate
   (ii) uvula.

A further subdivision, into the palatine arch and the oropharynx proper is of practical importance because squamous carcinoma of the palatine arch is less aggressive and metastasizes later than that elsewhere in the oropharynx.

Two further points of surgical anatomy require emphasis. First, some authors include the retromolar trigone as part of the oropharynx while others do not. The retromolar trigone is included in the oral cavity in the UICC/AJC (Union Internationale Contre Le Cancer/American Joint Committee) definition, and tumours of the trigone should not be considered as oropharyngeal tumours. Second, tumours of the anterior (lingual) surface of the epiglottis and the vallecula are regarded as laryngeal tumours and are classified as such by the AJC. They behave and are treated in a similar way to such tumours. However, these structures are part of the oropharynx in the UICC classification.

The oropharynx contains three structures of importance for the development of tumours:

   (1) a lining of squamous epithelium;
   (2) the paired tonsils, and the collection of minor lymphoid tissue in the base of the tongue;
   (3) collections of minor salivary tissue within the epithelium concentrated in the soft palate, the uvula and the capsule of the tonsil.

The surgical anatomy of the oropharynx including its lymphatic drainage is described in Volume 1, Chapter 10.
Pathology

Approximately one-third (35%) of pharyngeal carcinomata arise in the oropharynx. The oropharynx contains squamous, lymphoid and salivary tissues. Tumours may arise from each of these, and their relative incidence is shown in Table 12.2.

Table 12.2 Oropharyngeal tumours

<table>
<thead>
<tr>
<th>Tumour Type</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Squamous carcinoma</td>
<td>70%</td>
</tr>
<tr>
<td>Non-Hodgkin's lymphoma</td>
<td>25%</td>
</tr>
<tr>
<td>Salivary tumours</td>
<td>5%</td>
</tr>
</tbody>
</table>

Epithelial tumours

benign tumours such as papillomata arise from squamous epithelium but are of little pathological interest. The most important epithelial tumour is the squamous carcinoma; the lymphoepithelioma, which is a variant of this, will also be discussed.

Squamous carcinoma

The age and sex incidence of squamous carcinoma of the oropharynx are shown. As might be expected in an area with a rich lymphatic drainage, lymph node metastases are common and may be the presenting feature.

Bilateral nodes are less common than fixed nodes in this disease (fixation is, of course, a subjective assessment varying from one examiner to another). Enlargement of lymph nodes in squamous carcinoma is not influenced by the histological type of the tumour, and is as common in anaplastic tumours as in well-differentiated lesions.

The detection of lymph node metastases is very inaccurate because of differences between examiners, and because some palpable nodes do not contain tumour, whereas some impalpable nodes do. The level of the lymph nodes in the neck is shown.

The sites of origin of squamous carcinoma are listed in Table 12.3. Tumours of the oropharynx are staged under the UICC/AJC scheme, with the following conditions:

1. the classification applies only to carcinoma. It is not clear whether the classification includes all carcinomata or only squamous carcinoma

2. there must be histological verification of the disease. The histological features are variable: keratinization may be present or absent, and the degree of differentiation ranges from poorly differentiated or anaplastic tumours to well-differentiated examples reminiscent of normal structures. Spindle and basal cell variants are occasionally seen

3. the extent of disease must be assessed clinically, radiographically and endoscopically. The T classification for both UICC and AJC is shown in Table 12.4, and that for classification of nodes in Table 12.5.
Table 12.3 Site incidence of squamous cell carcinoma of the oropharynx

<table>
<thead>
<tr>
<th>Site</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tonsil/facial pillars</td>
<td>50%</td>
</tr>
<tr>
<td>Base of tongue</td>
<td>20%</td>
</tr>
<tr>
<td>Soft palate</td>
<td>10%</td>
</tr>
<tr>
<td>Vallecula and lingual epiglottis</td>
<td>10%</td>
</tr>
<tr>
<td>Posterior wall</td>
<td>5%</td>
</tr>
<tr>
<td>Lateral wall</td>
<td>5%</td>
</tr>
</tbody>
</table>

Another primary tumours in the upper alimentary tract or lung is found in about 10% of cases of squamous cell carcinoma of the mouth and oropharynx.

Table 12.4 Classification of the primary tumour (UICC and AJC)

Tis  Pre-invasive carcinoma (carcinoma in situ)
T0   No evidence of primary tumour
T1   Tumour 2 cm or less in its greatest dimension
T2   Tumour more than 2 cm but not more than 4 cm in its greatest dimension
T3   Tumour more than 4 cm in its greatest dimension
T4   Tumour with extension to bone, muscle, skin, antrum, neck etc
Tx   The minimum requirements to assess the primary tumour cannot be met.

Table 12.5 Nodal classification

(A) UICC Classification
N   Regional lymph nodes
N0  No evidence of regional lymph node involvement
N1  Evidence of involvement of movable homolateral regional lymph nodes
N2  Evidence of involvement of movable contralateral or bilateral regional lymph nodes
N3  Evidence of involvement of fixed regional lymph nodes
Nx  The minimum requirements to assess the regional lymph nodes cannot be met

(B) AJC
Nx  Minimum requirements to assess the regional lymph nodes cannot be met
N0  No clinically positive node
N1  Single clinically positive homolateral node 3 cm or less in diameter
N2  Single clinically positive homolateral node more than 3 cm but not more than 6 cm in diameter or multiple clinically positive homolateral nodes, none more than 6 cm in diameter
N2a Single clinically positive homolateral node more than 3 cm but not more than 6 cm in diameter
N2b Multiple clinically positive homolateral nodes, none more than 6 cm in diameter
N3  Massive homolateral node(s), bilateral nodes, or contralateral node(s)
N3a Clinically positive homolateral node(s), one more than 6 cm in diameter
N3b Bilateral clinically positive nodes (in this situation, each side of the neck should be staged separately - N3b: right, N2a: left, N1)
N3c Contralateral clinically positive node(s) only.
**Lymphoepithelioma**

The term lymphoepithelioma was first used by Regaud and Schmincke independently but simultaneously in 1921. The tumour is widely permeated by lymphocytes: the Regaud type contains nests of non-keratinizing squamous cells and the Schmincke type exhibits isolated transitional cells (hence the alternative term of transitional cell carcinoma).

The important clinical characteristics of this tumour are its extreme radiosensitivity, its tendency to metastasize and its sites - the nasopharynx, the tonsil and the base of the tongue. Quick and Cutler recognized it in 1927 as a subdivision of squamous carcinoma, and evidence for and acceptance of this view have grown steadily since. Sometimes these tumours may be confused with lymphoma, as the squamous cell component may be extremely undifferentiated. Immunohistology using the leucocyte common and cytokeratin antibodies may be helpful in doubtful cases. Surface marker studies of the lymphocyte population in lymphoepithelioma have shown these cells to be reactive, because they are composed of B cells, T-helper and T-suppressor cells and show no evidence of monoclonality or phenotypic restriction. Post-mortem studies on patients who died of this disease also show that the tumour is a variant of squamous cell carcinoma, with a coincidental lymphocytic content, because metastases to non-lymphatic sites, particularly the liver, contain tumour cells resembling the primary tumour only and do not contain lymphocytes.

**Aetiology**

Squamous cell cancer is uncommon before the age of 50 but increases in frequency thereafter. Men are affected about five times more often than women. Keratosis with dysplasia is well recognized as a precancerous condition in the mouth, but it seldom occurs in the oropharynx except on the palatine arch. As with oral cancer, tobacco and alcohol are thought to be important causal factors: however, those who believe this fail to answer the embarrassing question as to why the incidence of oral cancer has fallen to 10% of its former incidence in the last 50 years, at a time when the use of tobacco and alcohol have increased. The mucosal atrophy associated with iron-deficiency anaemia might be the precursor of oropharyngeal cancer in women. Syphilis and dental sepsis are no longer important factors in the UK.

**Tumours of lymphatic origin**

Hodgkin's disease is very rare in the oropharynx. Non-Hodgkin's lymphoma accounts for about 15% of tumours in this site, and most cases are of B-cell type. These have features in common with other tumours of MALT (mucosa associated lymphoid tissue) sites, remaining localized to the MALT sites longer than nodal lymphomata. The commonest type of B-cell lymphoma in the oropharynx is the large cell lymphoma of high grade malignancy.

**Aetiology**

As for most lymphomata, the aetiology is largely unknown. Certain associations are recognized, such as the development of lymphoma, often of lymphocytic or immunocytic type (*see below*), in long-standing cases of Sjögren's disease. African Burkitt's lymphoma, a B-cell high grade lymphoma of lymphoblastic type, shows a well-known association with
immunosuppression due to chronic malaria infection. Epstein-Barr virus is present in nearly all cases, and the lymphoma appears to favour sites of current epithelial proliferations: the odontogenic tissues in the young child, and the breast in pregnant women. T-cell lymphomata, rare in the oropharynx, show the well-documented association with human T-lymphotropic virus type I (HTLV-I), especially in Japan and the Caribbean countries.

Many of the lymphomata of the oropharynx are primary in the sense that there is no demonstrable deposit elsewhere in the body at the time of diagnosis. Sometimes, however, they appear to be secondary to, or coincident with deposits at other sites. These may include the gastrointestinal tract, lung and testis.

Classification

Until the early 1970s, classification of lymphomata was based on morphological features only. A bewildering variety of schemes was used, the best known being that of Rappaport (Rappaport, 1966). When immunological data on the cellular immunoglobulins, and later surface markers, of the lymphoma cells became available, it was apparent that the morphological criteria did not always correspond with the cells' immunological functional characteristics. In particular, Rappaport's 'histiocytic' group of large cell lymphomata was found to be principally of B-cell type, not of the macrophage origin implied by the term. The Kiel classification (Lennert et al, 1975, 1978) is widely used in Europe, and incorporates immunological and morphological data (Table 12.6). In over 10 years of use, only minor modifications have been required. The conceptually similar classification of Lukes and Collins (1974) is more widely used in the USA.

Table 12.6 Kiel classification of non-Hodgkin's lymphomata (simplified from Lennert et al, 1978)

(1) Low grade malignant lymphomata
   (a) lymphocytic lymphoma (B cell, T cell, hairy cell leukaemia, mycosis fungoides and Sezary's syndrome, T-zone lymphoma)
   (b) immunocytoma
   (c) plasmacytoma
   (d) centrocytic lymphoma
   (e) centroblastic/centrocytic lymphoma (follicular or diffuse)

(2) High grade malignant lymphomata
   (a) centroblastic lymphoma
   (b) lymphoblastic lymphoma (B or T)
   (c) immunoblastic lymphoma (with or without plasmablastic differentiation, B and T).

NB: Centroblastic and immunoblastic lymphomata may be difficult to distinguish and are sometimes grouped together as 'large cell lymphomata'.

5
Table 12.7 Histological types of non-Hodgkin's lymphoma of the oropharynx (based on a personal series of 100 patients)

Low grade
- Centroblastic/centrocytic B-cell lymphoma: 16%
- Immunocytoma: 13%
- T-cell low grade: 7%
- Lymphocytic lymphoma: 2%
- Centrocytic lymphoma: 2%

High grade
- Other B-cell lymphomata (lymphoblastic immunoblastic): 3%
- Centroblastic lymphoma: 53%
- T-cell high grade: 4%

The commonest types of lymphoma in the oropharynx (Table 12.7) are centroblastic (high grade) and centroblastic/centrocytic (low grade) (CB/CC). The centroblastic lymphoma is composed predominantly of large follicle centre transformed lymphocytes with prominent nucleoli, together with variable numbers of infiltrating reactive T cells. This tumour behaves in a highly aggressive manner, but if at an early stage (I and II) may be curable by local treatment. The low grade lymphomata are less aggressive, and contain mixtures of B cells of various types: follicle centre cells in CB/CC lymphoma, lymphocytes in lymphocytic lymphoma and immunocytes (lymphoplasmacytoid cells) in immunocytoma. All types also contain T cells, presumably of reactive nature. Paradoxically, these low grade tumours may be less curable than high grade lymphoma because, even though they progress more slowly, they are frequently disseminated by the time of diagnosis (stage III and IV).

The relative site incidence of non-Hodgkin's lymphoma of the head and neck and the staging system (AJC) are shown in Tables 12.8 and 12.9.

Table 12.8 Site incidence of non-Hodgkin's lymphoma of the head and neck

- Lymph nodes: 25%
- Oropharynx: 45%
- Nasopharynx: 15%
- Nose and sinuses: 10%
- Miscellaneous: 5%

Salivary gland tumours

Salivary gland tumours can occur in the oropharynx, but it is obvious from all reported series that few oropharyngeal tumours are of salivary origin, and that only a small proportion of all salivary tumours arise in the oropharynx.

In a large series of 530 minor salivary gland tumours, 3% affected the oropharynx, almost all of them arising in the tonsillar area. It is difficult to draw up a histological classification because of the small numbers involved, the different histological terms, and different referral patterns, but at least one-half of the salivary gland tumours of the
oropharynx are malignant, adenoid cystic carcinoma forming the vast majority. At this site the tumour behaves in the same way as elsewhere: it spreads by permeation along nerve sheaths, metastasizes to lymph nodes, bone and lung and has a very poor long-term prognosis.

**Staging classification of non-Hodgkin's lymphoma**

Stage I  
Involvement of a single lymph node region (I) or of a single extralymphatic organ site or site (IE)

Stage II  
Involvement of two or more lymph node regions (number to be stated) on the same side of the diaphragm (II); or, localized involvement of an extralymphatic organ or site of one or more lymph node regions on the same side of the diaphragm (IIE)

Stage III  
Involvement of lymph node regions on both sides of the diaphragm which may also be accompanied by local involvement of extralymphatic organs or site (IIIE) by involvement of the spleen (IIIS), or both (IIIE + S)

Stage IV  
Diffuse or disseminated involvement of one or more extralymphatic organ tissues with or without associated lymph node enlargement. The reason for classifying the patient as stage IV is identified further by specifying the site.

**Clinical features**

**Symptoms**

In 10% of patients the presenting symptom is a lymph node metastasis in the neck. Other symptoms include sore throat, pain on swallowing and referred earache. Large tumours of the base of the tongue give the voice a curious muffled quality. The routine clinical examination of the upper respiratory tract and the neck, not forgetting the axilla, groins, liver and spleen in patients with a lymphoma will not be discussed further.

**Clinical examination**

Two main clinical types of squamous cell carcinoma may be recognized: the exophytic and the ulcerative. The exophytic type spreads superficially and the ulcerative type infiltrates deeply, but exceptions do occur. An adenocarcinoma presents as a smooth non-ulcerated swelling and the malignant lymphomata as enlargements in the tonsillar fossae or base of the tongue. Ulceration eventually supervenes.

The growths can be easily seen provided that good lighting is used, though for those originating in the base of the tongue a laryngeal mirror is needed. Fixation of the palate or tongue should be noted and the area should be palpated with the forefinger to estimate the extent of infiltration. A postnasal mirror should be used to detect extension into the nasopharynx or onto the upper surface of the soft palate. Sometimes a carcinoma may occur deep in the base of the tongue which may not be associated with any abnormality of the surface mucosa. The presenting symptoms may be pain, possibly of the glossopharyngeal neuralgic type, or a node in the neck. It is important to remember that a carcinoma of the
base of the tongue cannot be excluded merely by inspection with a spatula or a laryngeal mirror - it is essential to palpate the tongue as well if growths of this type are not to be missed.

The neck must be examined carefully for lymph nodes both from in front and, most particularly, from behind the patient using the tips of the fingers. Another primary tumour should be looked for in the upper respiratory tract.

Investigation

Radiological examination includes the chest, but radiographs of the pharynx are mandatory for staging carcinoma under the UICC/AJCC scheme. What is to be achieved by this is not clear: tumours of the tonsil have no specific radiographic features and rarely invade the mandible. Computerized tomographic (CT) scans are more useful for showing the extent of the tumour, extension into the pterygoid fossa, and the presence of lymph nodes. The radiological assessment is described more fully elsewhere (Chapter 2). The assessment of the non-Hodgkin's lymphomata is unclear at the moment and is changing rapidly. Broadly speaking investigations should follow routes given below.

Biopsy

A biopsy is necessary to confirm the diagnosis and to establish the histology of the growth, since this is of importance in planning treatment. In most cases the biopsy can be performed under topical analgesia, but a general anaesthetic may be needed for lesions of the base of the tongue and to allow the extent of the growth to be determined by palpation. The biopsy specimen needs to be of adequate size to give a reasonable area of tissue for assessment of architecture, as well as cytological detail; needle biopsies are therefore not generally suitable. The lymph node must not be traumatized during removal or subsequent handling: the node consists of a delicate framework containing a fluid population of lymphocytes, and requires more care than perhaps any other pathological specimen. This also applies to biopsies from extranodal lymphoma sites. Fixation must be adequate: the authors' preference is to slice the tissue about 2 mm thick with a sharp, new blade, and then fix it for 24 hours in formalin with added 2% acetic acid (Curran and Gregory, 1980). Embedding is preferably done by a vacuum method, and cutting carried out with a sharp knife to give sections 2-3 microm thick. Plastic sections are not essential, but may add detail in certain cases, notably T-cell lymphomata.

Part of the biopsy should be snap frozen, unfixed in liquid nitrogen or carbon dioxide/isopentane, then stored at below -70°C for immunocytochemical studies (Nash, 1986). Few antibiotics work satisfactorily on fixed tissue; the availability of frozen tissue enables diagnosis in many otherwise difficult cases. Appropriate precautions must be taken to safeguard staff from infection when handling such material. Immunostaining can then be carried out when convenient, usually after the paraffin sections are available. The preferred current antibody panel is listed in Table 12.10. In the future, increasing use of gene probes will no doubt allow characterization of lymphomata according to gene re-arrangement and oncogene expression: stored frozen material is also suitable for these studies.
Table 12.10 Monoclonal antibodies used in the study of lymphomata

*B-cell related*
- Pan-B
- Immunoglobulins (IgA, IgG, IgD, IgM, kappa and lambda light chains)
- Dendritic reticulum cell
- Common leukaemia antigen
- HLA-Dr(Ia)

*T-cell related*
- Pan-T
- E-receptor
- Mature thymocyte
- Prethymic cells
- T-helper
- T-suppressor

*Other*
- Leucocyte common antigen
- Epithelial antigens (various)
- Macrophage.

**Blood investigation**

This should include complete blood count, erythrocyte sedimentation rate (ESR) urinalysis, liver function tests, plasma proteins and immunoglobulins.

**Bone marrow aspiration and biopsy**

The bone marrow biopsy is a vital part of lymphoma staging, and it is important that a trephine be taken as well as an aspirate: the proportion of positives in bone marrow aspirates from patients with CB/CC lymphoma may be as low as 10%, while trephines give a figure of at least 50%. The bone marrow is handled in a similar manner to the tumour biopsy specimen, but decalcification is required. EDTA has been used for this purpose, which although slower than strong acids, gives much better morphology. Bone marrow may also be examined by immunocytochemical methods on frozen sections where appropriate.

Bone marrow involvement by lymphoma may take several forms (Bartl et al, 1984). In involvement by lymphocytic and immunocytic lymphoma, the pattern is frequently diffuse, which makes minimal involvement difficult to detect. Some small nodules may be seen: these must not be confused with the lymphoid nodules frequently present in normal marrow, especially in elderly people. CB/CC lymphoma frequently gives a nodular picture in the marrow, but these are large pale nodules containing follicular centre cells, and are clearly abnormal. A diffuse pattern may also occur, and this does not seem closely related to the presence of nodularity in the primary lymphoma. Centroblastic lymphoma is less frequently seen in the marrow, but forms clumps or sheets of the large transformed follicle centre cells. Individually scattered centroblasts are less frequently seen and, if present in small numbers, might be confused with myeloid precursors.
**Percutaneous liver biopsy**

While a wedge biopsy at the time of laparotomy gives the best results, an early percutaneous needle biopsy gives such a high yield of positive results that it usually precludes the need for laparotomy.

**Radiology**

This should include chest radiograph, skeletal survey, gastrointestinal series.

**Pedal lymphography**

Laparotomy was a standard procedure in the management of Hodgkin's disease but it certainly has no place in the management of the non-Hodgkin's lymphomata. Exploratory laparotomy with splenectomy may reveal disseminated disease, but this can usually be performed by simpler means, notably by lymphography the accuracy of which is about 95%.

**Treatment of squamous carcinoma of the oropharynx**

At least six forms of management are available for oropharyngeal tumours: no specific treatment; radiotherapy; surgery; combined surgery and radiotherapy; chemotherapy; and cryosurgery.

**No specific treatment**

It is noticeable, but perhaps not surprising, that very few accounts of the treatment of any head and neck cancer contain a clear definition of those patients not treated, the reasons for withholding treatment, and the fate of those who do not receive specific therapy. Yet a fairly large proportion of patients with any head and neck cancer are not treatable with any prospect of success - about 20% of patients with oropharyngeal carcinoma in the authors' experience. The question of untreatability of carcinoma is inevitably coloured by personal philosophy; the authors' contraindications to surgery are shown in Table 12.11, but it is realized that there may be controversy over these indications. Patients with one of the first four conditions shown have a very poor chance of survival: 10% or less at 1 year, and 5% or less at 5 years, and it is doubtful whether they should be offered an operation. Radical surgery inevitably mutilates and deprives the patient of important functions. Is it, therefore, justifiable to operate on the many who do not survive, for the sake of those few who do? Each surgeon must decide this for himself.

**Table 12.11 Contraindications to surgery of oropharyngeal tumours**

<table>
<thead>
<tr>
<th>Anaplastic tumours</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fixed bilateral neck glands</td>
</tr>
<tr>
<td>Trismus</td>
</tr>
<tr>
<td>Horner's syndrome</td>
</tr>
<tr>
<td>Distant metastases</td>
</tr>
<tr>
<td>Patient's refusal</td>
</tr>
<tr>
<td>Advanced age (over 75 years)</td>
</tr>
<tr>
<td>Poor general condition</td>
</tr>
</tbody>
</table>
The temptation to give 'palliative treatment' for the patient who has an incurable tumour must also be mentioned. Usually all that is palliated is the surgeon's inability to accept reality! Of course, there are occasional unexpected successes, and young patients should seldom be denied the chance of treatment, but it should be remembered that radiotherapy causes a very sore mouth and a general upset lasting for months. The authors' figures show that the average period of survival of a patient with a head and neck cancer which is not suitable for radical treatment is 11 weeks. This is rather less than the period taken for the radiotherapy reaction to settle, so that the net result of palliative radiotherapy may be in fact an increase in the patient's discomfort. It should also be remembered that the vast majority of the general population is well aware of the site and significance of the regional radiotherapy unit. If such a unit pursues a frequent policy of giving palliative therapy which is followed rapidly by death, the local population can be forgiven for regarding the regional radiotherapy centre with dread.

**Radiotherapy**

Radiotherapy is undoubtedly the treatment of choice for non-Hodgkin's lymphoma without disseminated disease, and for many squamous carcinomata. Reviews of treatment of large series of squamous carcinomata are available.

Ledermann's series extends over a longer period from 1933 and shows an overall survival of 20% (Ledermann, 1967), whereas Fletcher and Lindberg's (1966) series from the USA, starting in 1954, shows 5-year survival of 35% for tonsillar carcinoma, and of 50% for palatal tumours. The chance of control of the primary tumour increased by about 20% with the introduction of supervoltage therapy, although this increase may not be reflected in an increased survival rate. Probably the greatest improvement over the last few decades has been the extension of first-class facilities to cover the whole population in the western countries - a political and not a medical achievement.

**Surgery**

Surgery was first described for tonsillar carcinoma at the end of the 19th century but, at that time, must have been a fearful undertaking for both patient and surgeon. There is little wonder that, when radiotherapy became available, surgery was abandoned by all but a few.

Until antibiotics became available it was impossible to open the neck and the pharynx at the same time because of the danger of infection. It was thus impossible to treat most of these patients surgically because of the presence of enlarged lymph nodes.

In the 1940s sulphonamides and then penicillin became available, and Hayes Martin was thus able to extend the operation of hemimandibulectomy and resection of the floor of the mouth to the treatment of tonsillar carcinoma, and to put the operation on a safe permanent footing. In his textbook published in 1957 he described how the term commando operation came to be used for this operation: the technical definition of the operation was too long-winded for everyday use and was therefore shortened to commando (combined neck dissection, mandibulectomy and resection of the oropharynx). At that time the commandos were recruited for aggressive warfare in difficult circumstances. His house staff borrowed the term and applied it to the operation (perhaps the analogy was close!).
Now that surgery can provide a cosmetically and functionally satisfactory end-product, what should be its place in treatment of carcinoma of the oropharynx, particularly of the tonsil? The patients who should not be offered surgery, for example those with trismus or fixed bilateral neck glands, have been mentioned above (see table 12.11). Furthermore, most patients with an oropharyngeal tumour without a palpable lymph node of the neck do tolerably well when treated by radiotherapy and should be offered this form of treatment initially. There thus remain two types of patients who should have surgery: those who have failed to benefit from radiotherapy, and those with a tonsillar carcinoma and a palpable gland in one side of the neck. Although the latter patient is treated in many centres by radiotherapy followed by a radical neck dissection, in the authors’ experience the gland in the neck may become inoperable by the time the radiotherapy reaction has settled, and such a policy gives poor results; a 5-year survival of 12.5% for instance in Ledermann's series. It has been shown fairly clearly that surgery achieves better results than radiotherapy for larger tumours with nodes in the neck: 60% compared with 0%.

For tumours of the lingual surface of the epiglottis, supraglottic laryngectomy gives very good results. Tumours at this site are uncommon but treatment with radiotherapy is not very successful; a recent series did not contain a single survivor at 5 years. Better results are obtained with supraglottic laryngectomy, and this form of treatment deserves wider use and recognition for this tumour; tumours at this site must certainly be treated surgically.

**Treatment of neck nodes**

Few surgeons would disagree that palpable unilateral nodes should be treated by radical neck dissection. Many would treat bilateral nodes similarly but the results are not good. Fixed nodes are occasionally worth resecting, but the natural history of the disease can rarely be affected in the presence of bilateral fixed nodes. The treatment of the neck in patients with no palpable nodes must also be discussed. It is well known from pathological studies that a proportion of such patients have lymph nodes which contain tumour but which are not palpable. On this fact rests the policy of prophylactic neck dissection or prophylactic neck irradiation. Sadly, it has been shown by controlled trials that neither policy improves the survival rate compared to a 'wait and see’ policy, although the local control rate might be improved.

**Combined surgery and radiotherapy**

An analysis of the failures of radiotherapy shows that there is a high incidence of local recurrence, and this has led some surgeons and radiotherapists to wonder if combined treatment would offer better results. As surgery fails at the periphery and radiotherapy at the centre of the tumour, a combination of the two might do better than either. Much of the early scientific work on preoperative radiotherapy was done by Powers and Tolmach (1964) who showed that a very high proportion of viable cells in a tumour (as many as 99% or more) were killed by 1000 cGy, higher doses being necessary to achieve eradication because of the exponential relationship between cure and dose. The argument advanced was that low dose preoperative radiotherapy, because it killed nearly all of the viable cells, should drastically reduce the chances of local implantation and dissemination, and at the same time not increase the morbidity. Powers and Tolmach investigated this in mice and showed that a low dose of
preoperative radiotherapy led to a significant increase in survival after surgery for a variety of induced tumours.

The causes of failures due to surgery and radiotherapy are as follows:

**Surgical failures**

1. The primary tumour may be cut across because some carcinomata proliferate along tissue planes
2. Regional spread in the lymphatics may not be encompassed by the operated field
3. Distant undetected vascular spread may have occurred before operation
4. Surgical manipulation may spread neoplastic cells into the lymphatics, blood vessels or the wound.

**Radiation failures**

1. The central portion of the primary tumour may be relatively anoxic, and hence most radioresistant. While peripheral, better oxygenated tumour might be killed, the central portion could regain its malignant potential after a period of quiescence
2. Metastases to lymph nodes are relatively radioresistant
3. Local or distant spread outside the treated field may have taken place before therapy, and radiation then does not encompass the lesion
4. The tumour may be radioresistant. That is, may be better able to resist ionizing radiation than surrounding vital tissue.

If these factors are considered together it seems possible that radiation followed by surgery planned to encompass the original estimated extent of the tumour may offer higher survival rates. For example, the peripheral extensions of malignant cells at the primary site inadvertently left behind by the surgeon in the unirradiated patient may be eliminated by the combined approach. The core of poorly oxygenated neoplasms destined to recrudescence in irradiated patients (after lurking undetected beneath an intact mucosa) may be eliminated by surgery. Furthermore, lymphatic metastases leading to failure of radiotherapy are removed by combined therapy. Cells spilled at operation after radiation therapy may be less plentiful because the greater part of the tumour has been killed, and less able to resume production. Finally, radiation may 'seal off' lymphatic channels and thus reduce the chance of manipulative dissemination of tumour cells.

It must also be recognized that there are patients who cannot be helped by preoperative radiotherapy, including patients with metastases (evident or unrecognized) and patients with a non-recurring carcinoma, for example, small skin tumours.
One of the main criticisms of preoperative irradiation is that the optimal dose and the benefit, if any, have seldom been investigated by rigidly controlled, randomized prospective trials. Most investigators have contented themselves with comparing survival rates at different times in their own institutions or retrospectively with other institutions. Since survival rates can vary widely depending on referral patterns, philosophy, selection, skill and many other factors, such a 'trial' produces no evidence of any value.

Only one controlled trial has been carried out and sadly this showed that preoperative radiotherapy does not increase survival (Strong et al, 1978).

**Reconstruction after surgery**

There have been four phases of repair of the surgical defect: using local flaps, axial flaps, musculocutaneous flaps, and free flaps.

*Local cervical flaps* tend to be of poor viability; one flap in three is lost partially or completely because of necrosis; furthermore, the use of these flaps leads to a temporary fistula on the neck, and possibly exposure of the carotid vessels. This method has now been abandoned.

*Axial flaps* include the *temporal* and *deltoidal* flaps. The *temporal* flap has few of the disadvantages of cervical flaps. The flap is virtually always viable; the temporary fistula does not leak saliva because it is placed high in the mouth, and the carotid sheath is not exposed. The main disadvantage of this method is the very obvious defect on the forehead. Visible defects of this sort make many patients self-conscious and may even make some reclusive. 'Surgery scars not only the face but also the mind'.

The *deltoidal* flap can be used for repair after excision of tumours of the oropharynx. The flap is led in through a temporary fistula and sewn to the edges of the defect. Three weeks later the flap is divided; the distal end is rearranged in the mouth and pharynx; the proximal end of the flap is returned to the chest, and the fistula is closed. It is important at the primary operation to protect the carotid sheath with a levator scapulae muscle graft and to close the lower compartment of the neck. This technique gives quite good results. The necrosis rate of the flap is less than 10% if proper precautions are taken while lifting it; the functional result for speaking and swallowing is good and the appearance of the patients is satisfactory. However, axial flaps have now been largely abandoned.

*Musculocutaneous* flaps are now one of the two standard methods of repair. Much the commonest for repair of tonsillar defects is the *pectoralis major* flap. At the end of the excisional phase a *pectoralis major* flap is raised in the standard manner (Volume 1, Chapter 24) and led through the neck into the oropharynx, where it is sutured in place.

*Free* vascularized flaps have also been described, the commonest being the forearm flap. The flap is raised during or at the end of the excisional phase (Volume 1, Chapter 24) and is then transferred to the oropharynx. Its artery and at least two veins are reanastomosed in the neck, the facial artery and vein often being convenient sources. This flap is not as
reliable in most surgeons' hands as the pectoralis major flap. Furthermore, the latter flap provides bulk, and it remains the workhorse of reconstruction for tumours at this site.

Chemotherapy

In contrast with the rosy picture in the lymphomata, the results of chemotherapy for carcinomata are still disappointing. Chemotherapy may be used for the sole treatment of advanced/recurrent (end stage) disease or as an adjunct to radiotherapy or surgery. Both types of treatment have been used in numerous phase II trials, that is where the response of the tumour is measured, but the survival of the patient is not. Such trials show that 25-50% of head and neck cancers respond to a wide variety of agents. Sadly these trials do not contain untreated controls. Only rarely have phase III trials been carried out, that is with untreated controls and with measurement of the patients' survival. These have shown that only cisplatin is effective, and that some agents actually reduce survival besides being toxic and expensive. Furthermore, the prolongation of median survival by cisplatin in patients with end stage disease is only 3 months. The results of phase III trials of adjuvant chemotherapy with radiotherapy for treatable tumours have been even more depressing: a high proportion of tumours respond but there is no effect on survival.

Cryosurgery

The destructive properties of freezing have been known for a long time, but cryosurgery has been used clinically only in the last decade, and cryosurgical units for clinical use are now available. Cryosurgery appears to achieve its effective by rupture of nuclear and cellular membranes, alterations in the lipoprotein components of cell membranes, pH changes, toxic concentration of electrolytes, polymerization and denaturation of proteins, and vascular stasis and microthrombi leading to ischaemia. It appears that there must be rapid repeated freezing and thawing with sufficient duration of freezing to a temperature of at least -20°C for a good iceball to form. The advantages claimed for cryosurgery are relative avascularity with little or no postoperative bleeding, minimal tissue response, pain relief mediated by destruction of sensory nerve endings, resistance of some tissues such as bone to the effect of freezing and possibly an immunological effect due to the altered antigenicity of the tumour tissue.

Cryosurgery does have a place in the treatment of tumour of the oral cavity and oropharynx, but this is mainly for benign lesions, hyperkeratosis, haemangioma and tonsillectomy in haemophiliacs, for instance. Small salivary tumours may respond well in unfit patients and small recurrences of squamous carcinoma will also sometimes resolve. But, for the average large recurrence of squamous carcinoma cryosurgery can at best achieve reduction in size of the tumour and pain relief. For the vast majority of patients with squamous carcinoma this form of treatment is at best only palliative.

Treatment of non-Hodgkin's lymphoma

Localized lymphoma is potentially curable by radiotherapy. The use of radiotherapy followed by chemotherapy is another approach to treatment, and the available data show an advantage for patients treated by the combined approach compared with radiotherapy alone.
For disseminated non-Hodgkin's lymphomata, the treatment of choice is systemic chemotherapy.

**Single agent chemotherapy**

A wide variety of agents is active in non-Hodgkin's lymphomata. They include the alkylating agents (nitrogen mustard, cyclophosphamide and chlorambucil), the vinca alkaloids (vincristine and vinblastine), procarbazine and prednisone, bleomycin, doxorubicin and the nitrosoureas (BCNU and CCNU). The response to single agent chemotherapy, the duration of response and the overall survival, vary with histological type, results consistently being superior in patients with nodular lymphomata in contrast to those with diffuse disease. Single agent chemotherapy with an alkylating agent may be the treatment of choice in patients with advanced 'favourable' histology.

**Combination chemotherapy**

Combination chemotherapy is the treatment of choice in patients with unfavourable histological types which, in contrast to the favourable histology lymphomata, show a rapid progressive and fatal course unless complete remission is achieved. Once complete remission has been maintained for 2 years, the probability of relapse is low, suggesting that a cure is possible. A partial response in these histological types conveys little survival benefit.

The first successful combination regimen for non-Hodgkin's lymphomata was cyclophosphamide, vincristine and prednisone, forming the CVP and COP regimens. CVP consists of short pulses of chemotherapy at 21-day intervals with cyclophosphamide on days 1-5 at a dose of 400 mg/m² per day. The Southwest Oncology Group called their regimen COP and administered cyclophosphamide at a dose of 800 mg/m² every 2 weeks for six courses. Other modifications have included cyclophosphamide on day 1 only, a day 1-8 schedule or on a day 1-4 schedule. CVT and COP produce complete responses in 60-90% of patients with favourable histology lymphomata and may be used as an alternative to single agent therapy in this group, although the superiority of combination therapy has not been established.

In an attempt to improve results in the unfavourable histological types more aggressive regimens including MOPP and C-MOPP (cyclophosphamide replacing mustine), and regimens developed by adding bleomycin and doxorubicin to the CVP regimen to form BACOP, have been tested. Each of these regimens have complete response rates of more than 40%.

The Southwest Oncology Group (SWOG) added doxorubicin to a modified COP to produce the CHOP regimen. In a study comparing CHOP to COP, the complete remission rate of 67% achieved with CHOP was superior to the 60% remission rate on COP for patients with diffuse lymphomata. The addition of bleomycin does not appear to improve the results of treatment.

Involvement of the central nervous system is a common problem in lymphomata, being the site of first relapse in 26% of the patients in one series. It is suggested that, in parallel with acute lymphoblastic lymphoma, prophylactic intrathecal therapy should be a component

**Treatment of salivary tumours**

Benign salivary tumours are almost exclusively pleomorphic adenomata, and are treated by local excision with a generous margin. Malignant tumours, generally adenoid cystic carcinomata, are treated by radical surgery as for a squamous carcinoma, followed by appropriate reconstruction. Radiotherapy has no place in the primary treatment of these disease, but is very useful if tumour remains at the surgical margins, and may be useful for palliation.
Chapter 13: Tumours of the hypopharynx

P. M. Stell and A. C. Swift

Surgical anatomy

The hypopharynx extends from the lower limit of the oropharynx at the level of the hyoid bone down to the opening of the oesophagus at the lower border of the cricoid cartilage. It is divided into three anatomical sites:

- the pyriform fossa (sinus);
- the postcricoid area;
- the posterior pharyngeal wall.

The definition of these sites is shown in Table 13.1. The larynx projects into the hypopharynx from the front so that grooves, known as the *pyriform fossae*, are formed on either side. These are shallow above and are separated from the valleculae by the pharyngoepiglottic folds. Lower down, the pyriform fossae become deeper. The upper shallow part of the pyriform fossa is bounded laterally by the thyrohyoid membrane and medially by the aryepiglottic fold. The lower deeper part of the fossa is related laterally to the thyroid cartilage. An important inferior relation is the paraglottic space which is the potential space bounded by the thyroid ala laterally and the conus elasticus and the quadrangular membrane medially: tumours passing through the aryepiglottic fold easily gain entrance to this space and then pass inferiorly lateral to the vocal cord. The pyriform fossae are lined by squamous epithelium, with a rich underlying network of lymphatics. These drain upwards with the superior laryngeal pedicle to the upper deep cervical nodes.

<table>
<thead>
<tr>
<th>Table 13.1 Definition of the hypopharynx (UICC)</th>
</tr>
</thead>
<tbody>
<tr>
<td>(1) Pharyngo-oesophageal junction (postcricoid area) extends from the level of the arytenoid cartilages and connecting folds to the inferior border of the cricoid cartilage.</td>
</tr>
<tr>
<td>(2) Pyriform sinus (fossa) extends from the pharyngo-epiglottic fold to the upper end of the oesophagus. It is bounded laterally by the thyroid cartilage and medially by the surface of the aryepiglottic fold and the arytenoid and cricoid cartilages.</td>
</tr>
<tr>
<td>(3) Posterior pharyngeal wall extends from the level of the floor of the vallecula to the level of the cricoarytenoid joints.</td>
</tr>
</tbody>
</table>

The *postcricoid* area is that segment which extends from the level of the arytenoid cartilages and connecting folds to the inferior border of the cricoid cartilage.

Although the hypopharynx, by definition, ends at the oesophageal opening at the lower border of the cricoid cartilage there is no change in the mucosa at that level and tumours spread readily across the junction. Many carcinomata in this part of the alimentary canal are pharyngo-oesophageal and there would be merit in tumour classifications incorporating the cervical oesophagus into the 'pharyngo-oesophageal region' in the same way that the International Union Against cancer (IUCC) incorporates the posterior third of the tongue into
the 'oropharynx' for oncological classification. The lymphatic drainage of the postcricoid area is less rich than that of the pyriform fossa: it passes to the paratracheal nodes.

The posterior pharyngeal wall is less well defined. It can best be categorized as that part lying between a posterior projection of the vocal cords in the cadaveric position. It ends superiorly at the level of the hyoid bone and inferiorly at the level of the arytenoids. It is separated from the prevertebral muscles by a fascial space.

Pathology

As elsewhere tumours may be mesodermal or ectodermal in origin and benign or malignant in behaviour. A résumé of the possibilities and their frequency is shown in Table 13.2.

Table 13.2 Tumours of the hypopharynx

**Mesodermal**

<table>
<thead>
<tr>
<th>Classification</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign</td>
<td>0.25%</td>
</tr>
<tr>
<td>fibrolipoma</td>
<td></td>
</tr>
<tr>
<td>leiomyoma</td>
<td></td>
</tr>
<tr>
<td>haemangiomia</td>
<td></td>
</tr>
<tr>
<td>Malignant</td>
<td>0.25%</td>
</tr>
<tr>
<td>leiomyosarcoma</td>
<td></td>
</tr>
<tr>
<td>non-Hodgkin's lymphoma</td>
<td></td>
</tr>
<tr>
<td>malignant synovioma</td>
<td></td>
</tr>
</tbody>
</table>

**Ectodermal**

<table>
<thead>
<tr>
<th>Classification</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malignant</td>
<td>0.25%</td>
</tr>
<tr>
<td>adenocarcinoma (including salivary carcinomata)</td>
<td></td>
</tr>
<tr>
<td>transitional cell carcinoma</td>
<td>0.25%</td>
</tr>
<tr>
<td>undifferentiated carcinoma</td>
<td>0.25%</td>
</tr>
<tr>
<td>oat cell carcinoma</td>
<td>0.25%</td>
</tr>
<tr>
<td>carcinosarcoma</td>
<td>1%</td>
</tr>
<tr>
<td>squamous carcinoma</td>
<td>97.5%</td>
</tr>
</tbody>
</table>

Benign tumours are very rare. The commonest of these are the fibrolipoma and the leiomyoma. Both are polypoid tumours which usually present with dysphagia. On barium swallow they present a typical appearance of a smooth constant mass lying in the lumen of the oesophagus. They are treated by surgery: the tumour is removed at its pedicle which is exposed by a lateral pharyngotomy. Malignant tumours, as would be expected in an area lined by squamous epithelium, are virtually all squamous carcinomata. Unlike tumours of the oropharynx and nasopharynx, squamous carcinomata of the hypopharynx are usually well differentiated - for example, 85% in Ledermann's (1967) extensive series.
**Surgical pathology of squamous carcinoma**

Squamous cell carcinomata may be exophytic or ulcerated. An exophytic carcinoma most commonly lies in the upper pyriform fossa and aryepiglottic fold regions, whereas the ulcerated form is typical of the other parts of the hypopharynx. Carcinomata in this region are usually classified under the anatomical site (see Table 13.1) from which they are thought to have arisen.

Often the tumour is so advanced when first seen that it is difficult to determine its site of origin, and placing it in one of the above categories is rather a matter of guesswork. This may explain the reported variations in frequency of tumours at each site, but there is undoubtedly a wide geographic variation. In the UK, most series of patients show that approximately one-half is considered to arise in the pyriform fossa and the other half from the remaining sites, tumours of the posterior wall being very uncommon. On the other hand, in Canada and the USA the incidence of pyriform fossa tumours is much higher. The differences are well illustrated in the figures of four series of patients from London, Toronto, Houston, USA and Liverpool (Table 13.3).

Table 13.3 Relative site incidence of hypopharyngeal carcinoma

<table>
<thead>
<tr>
<th>Number of patients</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pyriform fossa (%)</td>
<td>39</td>
<td>61</td>
<td>75</td>
<td>38</td>
</tr>
<tr>
<td>Postcricoid area (%)</td>
<td>24</td>
<td>24</td>
<td>2</td>
<td>45</td>
</tr>
<tr>
<td>Posterior pharyngeal wall (%)</td>
<td>10</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cervical oesophagus (%)</td>
<td>7</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Others (%)</td>
<td>37</td>
<td>15</td>
<td>23</td>
<td>0</td>
</tr>
</tbody>
</table>

1. Royal Marsden Hospital, London (Dalley, 1968).
2. Toronto General Hospital (Bryce, 1967).
3. M. D. Anderson Hospital, Houston, Texas (MacComb and Fletcher).
4. Authors’ personal series.

In most series the incidence of carcinoma is much higher in men than in women, but in the postcricoid region the reverse is true.

Tumours of the pyriform fossa can be subdivided into those affecting the lateral and those affecting the medial wall. Tumours arising on the lateral wall extend through the thyrohyoid membrane to invade the carotid sheath and thyroid gland, so that a palpable neck mass in this disease may be direct extension of the tumour and not an enlarged lymph node. This should be confirmed by asking the patient to swallow. Tumours of the medial wall rapidly invade the aryepiglottic fold, a structure rich in lymphatics, and pass through it to enter the paraglottic space. They then pass inferiorly in this space lateral to the vocal cord, but beneath the mucosa. Spread in this space fixes the vocal cord causing hoarseness.

The submucosal spread of the disease is obviously of surgical importance: its average extent is 10 mm in pyriform fossa tumours, and 5 mm in postcricoid tumours (Harrison,
This spread, affecting the oesophageal stump and the thin membranous tracheo-oesophageal wall, is one of the most potent causes of recurrent. In the same report, Harrison showed that deposits in the paratracheal nodes were common. Some surgeons have thus advocated mediastinal dissection but, as enlarged nodes in the tracheo-oesophageal groove can be removed by finger dissection from above, it is probably unnecessary.

Paralysis of a vocal cord may be due to invasion of the recurrent laryngeal nerve or immobility of the cricoarytenoid joint.

Carcinoma of the posterior pharyngeal wall has not been studied in detail. In the authors' series 60% remained localized to the posterior wall, and 40% invaded the pyriform fossa. Fixation to the prevertebral fascia, surprisingly, was rare. One patient in three with this disease has swollen glands in the neck.

Cancer of the hypopharynx has a notorious propensity to metastasize to the lymph nodes of the neck, and indeed an enlarged node may be the presenting symptom. The incidence of lymph node metastases in hypopharyngeal carcinoma (using the authors' series as a guide) is shown in Table 13.4. Although the upper deep cervical nodes are usually invaded first, the lymph may spread up and down so that almost any node in the deep cervical chain may be the first to be enlarged and once one node has been invaded the distribution may be even more eccentric.

### Table 13.4 Incidence of lymph node metastases (%)

<table>
<thead>
<tr>
<th></th>
<th>N0</th>
<th>N1</th>
<th>N2</th>
<th>N3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pyriform fossa</td>
<td>35</td>
<td>35</td>
<td>5</td>
<td>25</td>
</tr>
<tr>
<td>Postcricoid area</td>
<td>70</td>
<td>20</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Posterior pharyngeal wall</td>
<td>60</td>
<td>30</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Cervical oesophagus</td>
<td>95</td>
<td>0</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Overall</td>
<td>55</td>
<td>25</td>
<td>5</td>
<td>15</td>
</tr>
</tbody>
</table>

Since the lymphatic network freely crosses the midline, bilateral lymph node metastasis can occur, particularly if the growth itself crosses the midline as in postcricoid carcinoma.

Lymph node invasion occurs early in pyriform fossa tumours and there may be a large lymph node in the neck while the primary growth is still small. About two-thirds of patients already have palpable cervical lymph node involvement when they are first seen. Of more importance to the surgeon, however, would be the ability to estimate the likelihood of occult metastases when no nodes are palpable. Ogura and Mallen (1965) showed that 38% had microscopic invasion although clinically no nodes were palpable. They concluded that this figure is high enough to justify an elective neck dissection. Occult lymph node metastases in postcricoid carcinoma are too uncommon to justify elective neck dissection.

Distant metastases to bones and viscera are becoming increasingly recognized, possibly because patients are surviving for longer after their primary lesion.
Other malignant tumours

Malignant tumours of the hypopharynx other than squamous cell carcinomata are shown in Table 13.2. The most important is the carcinosarcoma or pseudosarcoma. This is a polypoid tumour usually arising from the aryepiglottic fold. Its stroma resembles a sarcoma but it is covered by a layer of squamous carcinoma. Despite its bizarre and aggressive appearance the sarcomatous element never metastasizes and the carcinomatous part does so late, to the lymph nodes of the neck. Some pathologists think the 'sarcomatous' element is merely an unusual stromal reaction to squamous carcinoma. Whatever the case, these tumours are not particularly aggressive and localized removal without sacrificing the pharynx and larynx is usually sufficient to control the disease for long periods.

Another interesting rare tumour of very low-grade malignancy, paraganglioma, which arises in the hypopharynx is characterized by pain of neuralgic type which often extends to the ear, mediated via the vagus nerve. In small tumours this can be relieved dramatically by biopsy excision. The pain may be triggered by eating or drinking sharp or bitter substances. Most of these very uncommon tumours have arisen in the arytenoid region, although some have developed on the epiglottis. Macroscopically they usually appear as small reddish or purplish swellings which may be almost pedunculated. Histologically they appear to be paraganglioma. These tumours also tend to metastasize, possibly after several years, to regional lymph nodes and also in a very unusual manner to subcutaneous tissues and subserous layers such as under the peritoneum and pleura. These metastases are also painful and sometimes exquisitely so. They tend to be multiple, and one of Ranger's (1979) patients had 120 metastases removed at one operating session from subcutaneous sites scattered throughout the body. The patient obtained great relief from the procedure. The tumours are not radiosensitive and local excision appears to be the treatment of choice.

Aetiology

Anaemia

In 1919 both Paterson and Brown-Kelly described a syndrome which is now taken to consist of anaemia, glossitis, pharyngeal web, koilonychia and splenomegaly. The association of this disease with postcricoid carcinoma was shown by Ahlbom in 1937. More recently MacNab-Jones (1961) showed that between one- and two-thirds of patients with postcricoid carcinoma have a history of this syndrome. The risk of an individual patient with the Paterson-Brown-Kelly syndrome of developing postcricoid carcinoma is small, of the order of 2%. The anaemia is not necessarily microcytic and may be macrocytic (Jacobs and Kilpatrick, 1964). Of 266 patients with postcricoid carcinoma, 35% had had dysphagia for more than 5 years at the time of diagnosis and a benign stricture of the upper oesophagus had been demonstrated in 9% before the carcinoma developed (Richards, 1970).

The condition is better called sideropenic dysphagia but it is also known as the Paterson-Brown-Kelly syndrome after the two workers who described the condition separately in 1919. In some part of the world it is also called the Plummer-Vinson syndrome.

Sideropenic dysphagia is characterized by thinning of the mucosa of the upper alimentary tract with loss of the rete pegs and a reduction or absence of glycogen in the cells.
This condition is most obvious on the tongue and lips, presenting as superficial glossitis and angular stomatitis with cracking of the mucosa at the corners of the mouth. In the pharyngo-oesophageal region the same histological features are evident and in some patients a mucosal fold develops from the anterior wall of the upper oesophagus referred to as an oesophageal web. In a few patients with long-standing sideropenic dysphagia a dense fibrous stricture may form in the upper oesophagus (Figure 13.4) and histologically there may be necrosis of the muscle as well as fibrosis.

Most patients have a low haemoglobin concentration, mean corpuscular haemoglobin, and serum iron. The iron-binding capacity is raised in about one-half of the patients. In some the serum iron may be low even though the haemoglobin may be within normal limits. In about 20% of patients, vitamin B₁₂ absorption is reduced to within the pernicious anaemia range and in about 5% the level of serum B₁₂ is pathologically low, indicating that the patient has pernicious anaemia in addition to sideropenic anaemia. The word sideropenia reflects the commonest, most striking and most readily demonstrable deficiency (iron), but many patients have other metabolic abnormalities. Approximately one-half of the patients have pyridoxine deficiency as shown by estimations of pyridoxine-dependent transaminases, and have evidence of abnormal tryptophan metabolism as shown by the excretion of excess abnormal metabolites in the urine after oral administration of tryptophan. Riboflavine metabolism remains normal in these patients.

These biochemical abnormalities found in patients with sideropenic dysphagia do not occur in patients with other strictures of the oesophagus, such as those resulting from ingestion of caustics, and for this reason they appear to be causal and not secondary to the dysphagia as was originally thought by some workers such as Vinson (1922). In addition, patients undergoing gastrectomy develop multiple deficiencies and women in this category develop webs in the upper oesophagus eight times more commonly than a matched group of the population who have not undergone gastrectomy (Jacobs and Kilpatrick, 1964).

The dysphagia in those patients who do not have a fibrous stricture is unexplained. Most of them do not have a demonstrable web on barium swallow and in them it is considered that the dysphagia results from incoordination of muscular contraction due to interference with the sensory side of the reflex arc. The same explanation is probably true of those patients who have a definite web. The web in itself does not explain the dysphagia because it may be just as prominent for a time after the dysphagia has been completely relieved by the administration of iron with vitamin B complex.

**Smoking and drinking**

Inevitably heavy smoking and drinking have been said to be aetiological factors of pharyngeal cancer. However, this unproven opinion must be interpreted with great caution since the incidence of pharyngeal cancer has not increased during this century, despite the enormous increase in tobacco consumption, and despite the fact that a higher proportion of the population now survive into the cancer age group.
Radiation

In the 1920s and 1930s, there was a vogue, particularly in the North West of England, for treating thyrotoxicosis with small doses of radiotherapy at weekly intervals for several months. This treatment was effective, particularly when one remembers that surgical treatment of thyrotoxicosis at that time was an adventure with a fair mortality. A small proportion of these patients are now developing a pharyngeal carcinoma, with an average latent interval of 25 years.

Clinical features

Symptoms of carcinoma

A carcinoma in the hypopharynx first affects swallowing. Since the pyriform fossae are fairly capacious and since carcinoma is at first painless, symptoms may not arise until the growth is well advanced. The first symptoms may be indefinite but discomfort or pain on swallowing develop, initially for solid food only. The discomfort begins simply as a soreness or slight pricking sensation as solid food is going down, but it gradually progresses to pain, which may be referred to the ear, and to obstruction to all acts of swallowing.

As has already been pointed out, an enlarged lymph node may appear in the neck while the primary tumour is still small and before it has given rise to any throat symptoms.

Carcinomata of the postcricoid area and below cause obstructive symptoms earlier than those at a higher level but discomfort and pain are not so evident. In early cases the patient may say that she feels as though the food is passing over a ledge or ridge while fluids go down without difficulty. In most cases, however, obstructive symptoms are quite severe by the time the patient first seeks advice and loss of weight is usually more marked than in carcinomata arising in the other sites.

In men the tumour is commonest in the sixth to seventh decades but in women postcricoid lesions often appear earlier, and patients in their late twenties and thirties are fairly common.

Clinical examination

The pharynx and the larynx are examined with a laryngeal mirror or a fibreoptic laryngoscope in the usual way.

Tumours of the pyriform fossa are generally easily observed, and some idea of the extent of the tumour can be seen. Fixation of the hemilarynx is an important sign.

In tumours of the postcricoid area the tumour is not often seen unless it extends into the pyriform fossae. Retained secretions may be seen in the pyriform fossa. Immobility of the vocal cord is also important to elicit.

The neck must be examined for lymph node metastases in the usual way. Laryngeal crepitus is lost in postcricoid tumours, and tumours extending out of the thyrohyoid membrane.
can also be palpated occasionally. Widening of the laryngeal framework may be felt in advanced postcricoid tumours.

**Investigation**

**Radiology**

Radiological methods of investigation of tumours of the hypopharynx are considered in Chapter 2.

**Endoscopy**

Endoscopy is extremely valuable in the assessment of a hypopharyngeal tumour. It also serves of course to provide a specimen of tumour for biopsy, but this should be the last and, in some ways, the least important part of the endoscopy. The hypopharynx, the larynx, the trachea and the oesophagus are examined using rigid endoscopes.

The purpose of the examination of the pharynx is to determine the presence of a tumour and its extent. In a pyriform fossa tumour the main areas of interest are whether both walls of the pyriform fossa are involved, but more particularly the lower limit of the tumour in relation to the postcricoid area and the lateral limit of the tumour in relation to the posterior pharyngeal wall. A tumour which enters the postcricoid area or approaches the midline of the posterior wall of the pharynx cannot be dealt with by partial pharyngectomy and requires total pharyngectomy. The other area of interest in pyriform fossa tumours is to assess whether the tumour spreads superiorly above the pharyngoepiglottic fold to invade the base of the tongue. This spread can often be better felt, using a gloved finger, than seen. A postcricoid carcinoma is assessed for its upper and lower limits measured in relation to the upper incisor teeth. The upper limit is easy to assess, but it is usually very difficult to pass a rigid oesophagoscope through a postcricoid carcinoma to assess the lower limits. This difficulty can almost always be overcome by passing a filiform bougie through the tumour and then passing a small bronchoscope blind over this. This allows the lower limit of the tumour to be assessed and also permits inspection of the remainder of the oesophagus which, on occasion, harbours a second primary tumour. It is also necessary to note whether the tumour spreads into the pyriform fossa: this information has no relation to prognosis and treatment but it is vital in staging the tumour using the UICC and American Joint Committee (AJC)(1973), classification, despite the fact that the information is completely irrelevant. Lastly, the trachea should be examined for a tracheo-oesophageal fistula, which is rare.

The larynx is examined in patients with tumours of the pyriform fossa to look for extension into the vestibule of the larynx and rigidity of the false cord area. Mobility of the cords cannot of course be reliably assessed under general anaesthesia.

Once the extent of the tumour has been assessed a large specimen of the tumour is taken for histology, preferably using a punch biopsy forceps. Finally, the examiner should put on a rubber glove and examine the inside of the pharynx, assessing mobility of the tumour over the prevertebral fascia, and extension of the tumour over the pharyngoepiglottic ligament into the base of the tongue.
As has been mentioned already in the section on pathology, carcinoma of the hypopharynx tends to spread in submucous tissues well beyond the surface limits of the growth and the extent of this can be difficult to judge. It is never easy to assess possible involvement of deep structures in the neck with any accuracy, although this will be clear if there is a paralysis of the recurrent laryngeal nerve. While involvement of the thyroid gland may be revealed on a thyroid scan there may still be spread into the gland even when the scan is normal.

**Laboratory studies**

These patients require very careful general assessment including routine clinical examination, usually by a physician, to assess their general health. This group of patients also has a very high incidence of blood and electrolyte abnormalities and requires a full routine assessment from this point of view.

**Treatment**

**Untreatable patients**

Probably because of the magnitude of the surgical attack required, there is a higher proportion of untreatable tumours among hypopharyngeal than among other head and neck cancers. Of the authors' series of 500 patients, 31% were untreatable, 10% because of poor general condition, 2% because of metastases, 2% because the patient refused treatment and 17% because of an advanced tumour. Palliative treatment of patients with advanced disease must be governed by the predominant symptom for which relief is necessary and in many of these patients there is little that can be done. A gastrostomy which enables the patient to obtain nourishment but which does nothing to allow the swallowing of saliva or to avoid inhalation pneumonia cannot be regarded as palliation. The treatment of these patients by cytotoxic agents is discussed in Chapter 24.

**Radiotherapy**

The largest series of patients to receive radical radiotherapy alone is that of Ledermann (1967), who showed that patients without enlarged nodes (the minority) stood a 20% chance of surviving 5 years when treated with radiotherapy. However, in the presence of enlarged nodes the cure rate was negligible - less than 5% - which was also the case with anaplastic tumours. Radiotherapy therefore should be reserved for the patient without enlarged nodes, and with a small tumour, that is with a vertical length less than 5 cm and not producing a vocal cord paralysis. Conversely, surgery will be needed as the primary form of treatment for tumours longer than 5 cm, in those patients with enlarged cervical nodes, and for tumours which recur after radiotherapy.

Much is heard at the moment about the possibility of combining radiotherapy or surgery with cytotoxic chemotherapy. Although considerable advances have been, and are still being, made in the field of cytotoxic agents the position at present can be summarized by saying that the stage has not been reached when chemotherapy could be advocated as an adjuvant method of treatment for carcinoma of the hypopharynx: although the administration of chemotherapy before radiotherapy induces response in about 25% of patients, controlled
trials have shown that the ultimate survival is unaltered. Furthermore, the 75% of patients who do not respond to chemotherapy suffer the toxic effects of the drugs (and the State the expense) and their treatment is delayed.

**Combined surgery and radiotherapy**

About 15 years ago there was a fashion in the treatment of head and neck cancer to administer preoperative irradiation in order to reduce local recurrence by implantation and dissemination by veins and lymphatics. The concept is attractive but there is general disagreement on the dose, different authorities using doses ranging from 1000-5000 cGy. Furthermore, despite the claims of some authors, there is no evidence that combined treatment improves survival rates, and it has become much less popular since Strong et al (1978) showed by a controlled trial that it does not prolong survival.

**Surgery**

Treatment of the lymph nodes and of the primary site must both be considered.

For the patient with secondarily involved lymph nodes the operation of radical neck dissection has stood the test of time. More recently many surgeons have advocated prophylactic neck dissection, that is radical neck dissection in the absence of palpable lymph nodes, on the grounds that, although impalpable, some may be histologically involved.

Pathological studies have shown a high incidence of impalpable but invaded neck nodes in pyriform fossa tumours and it is probably advisable therefore to carry out prophylactic neck dissection for these patients.

There is no evidence, from carefully controlled trials, that prophylactic neck dissection improves the chances of survival in postcricoid carcinoma. Since it can cause a stiff shoulder and at worst may kill the patient, the operation is not justifiable. In the authors' series only 5% of patients undergoing pharyngolaryngectomy later developed an enlarged lymph node and they are the only patients who therefore stand to benefit from the operation. As a postcricoid carcinoma lies in the midline a neck dissection of both sides is logically needed, but this would increase the mortality by roughly the same order as the theoretical gain and the overall benefit would be negligible.

**Antibiotic chemoprophylaxis**

Resection of a hypopharyngeal tumour exposes the wound to the existing oral/pharyngeal microflora and any potential pathogens. The wound is therefore at risk of becoming infected and fistulae or massive breakdown may complicate healing. The incidence of wound infection after major head and neck operations in which the mouth/pharynx is opened is often substantial, ranging from 14 to 87% (Swift, Bartzokas and Corkill, 1984). In spite of this, prophylactic antibiotics are used to cover such surgery routinely by only about one-half of the otorhinolaryngologists in the UK (Raine and Swift, 1984). Before the introduction of antibiotic prophylaxis on the authors' unit, 22 of 32 patients developed a wound infection after major oncological surgery (Bartzokas et al, 1984). Cultures of the saliva and throat before surgery were of limited value in predicting sepsis. After surgery, all of the
wound infections were polymicrobial, and consisted of Gram-negative aerobic bacilli (34.4%), *Staphylococcus aureus* (21.3%), anaerobes (13.6%), pneumococci and *Haemophilus influenzae* (10%). A subsequent controlled trial with an antibiotic suitable for covering these organisms (Augmentin: amoxycillin and clavulanic acid), substantially reduced the incidence of wound infection and antibiotics have since been used routinely (Raine et al, 1984).

Of the microorganisms grown from infected wound, intestinal aerobic Gram-negative bacteria are commonly found. The source of these bacilli was originally thought to be exogenous contamination after surgery but they have since been shown to originate from the stomach, and their subsequent ascent is facilitated by the nasogastric feeding tube (Swift, Bartzokas and Corkill, 1984). An attempt to interrupt this gastro-oral pathway by regular instillation of 40% ethyl alcohol into the stomach after surgery has since been undertaken, but was unsuccessful. These intestinal bacteria are isolated from the mouth and pharynx in most patients after surgery in spite of broad-spectrum antibiotic prophylaxis, and yet the number of wound infections remains low. This is probably because they are not present during surgery, but colonize the pharynx after surgery, when the wound has been closed. Their importance in the aetiology of wound infections is therefore not as great as once thought, although they are still likely to contribute to existing infections. Anaerobes are generally the commonest group of commensal bacteria in the oropharyngeal flora, and although they are not prevalent pathogens on the authors’ unit, they are considered to be important nevertheless.

The choice of the prophylactic antibiotic regimen is determined from the potential pathogens present at the time of surgery when the wound is exposed. Providing an adequate regimen is given perioperatively, the intricacies of the choice are probably not crucial.

The chosen regimen should therefore cover at least the oral anaerobes, streptococci and staphylococci, and combinations of metronidazole and cephalosporin or one of the penicillin group of agents are eminently suitable. On the authors’ unit, four doses of erythromycin 1 g intravenously 6-hourly, starting during the early stages of surgery are given. Erythromycin is active against the likely pathogens, especially the oral anaerobes, although it does not cover intestinal aerobic Gram-negative bacteria. However, as stated previously, these microorganisms are rarely present at the time of surgery, unless a tracheostomy or nasogastric tube has been previously inserted, in which case broader spectrum cover is necessary. The advantage of this regimen are that effective prophylaxis is obtained using a single agent for a short period; the induction of bacterial resistance will be unlikely; the oral flora will not be grossly disturbed and thus resistant pathogens will be prevented from proliferating freely.

The regimen is also employed when free small bowel loops are used to replace the pharynx after total pharyngolaryngectomy. Cultures from the bowel have all been sterile, so there has been no need to expand the spectrum of chemoprophylaxis.

**Resection of the primary tumour**

The operation advised obviously depends on the site of the primary tumour. Patients with a tumour on the posterior pharyngeal wall are often advised to have a total pharyngolaryngectomy; but Ogura, Watson and Jurema (1960) showed that it is usually unnecessary to sacrifice the larynx. Such tumours can be removed by a lateral pharyngotomy; the resultant deficit is covered by a split skin graft stitched over the prevertebral fascia.
Preliminary results indicate that this operation is satisfactory, and has the enormous advantage of preserving the voice.

Operations for tumours of the pyriform fossa vary from partial laryngectomy and partial pharyngectomy to total pharyngolaryngectomy. Ogura, Saltzsrein and Spjut (1961) attempted to apply supraglottic laryngectomy, removing the affected part of the pharynx and the suprastructure of the larynx, but preserving the patient's vocal cords and hence his voice. However, pathological studies have repeatedly shown that most of these tumours invade the larynx, notably the paraglottic space, making the operation highly dangerous. A total laryngectomy is therefore always needed but it is not necessary always to perform a total pharyngectomy. Indeed, in the authors' series, it was possible to preserve enough pharyngeal mucosa in 67% of patients for closure of the defect, in the same manner as after total laryngectomy for laryngeal carcinoma. The operation is thus very similar to that of total laryngectomy (see Chapter 11) including a hemithyroidectomy. The patient is thus spared the problem of a pharyngeal reconstruction. When this is not possible (usually because the tumour has invaded the postcricoid area) a total pharyngolaryngectomy with a pharyngeal reconstruction is necessary.

There is no dubiety about the treatment of postcricoid carcinoma - a total pharyngolaryngectomy is needed, with resection of part of the oesophagus, or all of it, if the tumour extends into the cervical oesophagus, which it often does.

**Restoration of continuity of the alimentary tract after laryngopharyngectomy**

The oesophagus has a segmental blood supply and, despite a few reported successes, it is not usually possible to mobilize the oesophagus sufficiently to allow it to be brought up for anastomosis to the pharynx. The methods which are currently in use to restore continuity can be classified as follows:

1. plastic strips;
2. skin flaps;
3. visceral repair:
   a) pedicled;
   b) free.

These will now be considered in more detail.

**Repair with a plastic tube**

Silicone rubber tubes are advocated by Stuart (1979) and they have the advantage of simplicity and the fact that the lower end can be inserted into the oesophageal lumen without sutures being required and so a considerable length of oesophagus can be resected without opening the chest. Difficulties arise in obtaining a water-tight seal between the pharynx and the upper end of the tube.
**Repair using skin**

While split skin grafts wrapped around a moulded tube have been used, they are liable to give rise to fistulae and to stenosis and cannot be used after radiotherapy.

The most suitable skin flap is the pectoralis major musculocutaneous flap. At the end of excision of the pharynx and larynx, the flap is raised and passed upwards between the skin of the upper chest and lower neck. The skin island is made into a tube, with the skin surface inwards. The upper and lower ends of the tube are then anastomosed to the pharynx and oesophagus respectively. This method has the advantage that an abdominal operation is not required but the disadvantage that only a limited length of oesophagus can be resected and reconstituted without opening the chest. Also, fistulae and stenosis are common. The deltopectoral flap has already passed into history, and repair with this flap probably will be abandoned soon.

**Visceral repair**

Visceral transposition has the great advantage that it effects an immediate repair of the pharynx and oesophagus and the whole length of the gullet can be removed so that there is no lower limit to the level of resection. Swallowing is restored within a few days and most patients are eating almost a normal diet within a week or two. Also, the repair can be regarded as durable in the sense that swallowing tends to be maintained even when recurrent growth develops in the neck. Fistulae are uncommon and stenosis is unknown when stomach is used and is seldom seen when colon is transposed. However, the procedure does impose a major operation on the patient and it requires the cooperation of an experienced surgeon who is well versed in the techniques of handling abdominal viscera.

Both stomach and colon have their own advantages and disadvantages which may determine the choice in a particular patient but, in many, the decision may merely reflect the particular experience and preference of the surgeon concerned.

Two particular advantages of transposing the stomach are that it has an excellent blood supply and when it is used only one anastomosis, in the neck, is required. On the other hand, mobilization of the stomach requires extensive and meticulous dissection and in a few patients there is some tendency to reflux and to dumping. On balance, stomach is the viscus of choice and the colon has now been abandoned by all but a few.

Repair procedures of this type should not be undertaken by the 'occasional' surgeon in the field of laryngopharyngectomy and it is necessary to discuss only the general principles of the operations rather than the minute details which are, nevertheless, critical to the success of the procedure.

At operation the two surgical teams should start their procedures synchronously, but if a bilateral radical neck dissection is included the abdominal surgeon will complete the mobilization before the neck dissection is finished. This has advantages in that the viscus can remain in its natural environment for a while after mobilization before being moved to the neck. The main steps of the procedure are as follows. The pharynx and larynx are mobilized from the carotid sheath on each side as described for total laryngectomy (see Chapter 11), the
superior and inferior thyroid pedicles being divided. Both lobes of the thyroid gland are also mobilized. The trachea can be divided at this stage, and the tracheostome is created. The upper thoracic oesophagus is mobilized by blunt finger dissection, and the paratracheal lymph nodes are also mobilized *en bloc* with the oesophagus. Simultaneously, the abdominal team opens the abdomen through an upper paramedian incision. The abdominal oesophagus is freed, followed by mobilization of both curvatures of the stomach, dividing the left gastric artery but preserving the right gastro-epiploic artery. Finally the first two parts of the duodenum are freed, and a pyloromyotomy is performed. The thoracic oesophagus is now mobilized by blunt finger dissection from above and below. Once it is free the stomach is pulled into the neck. The pharynx is divided above the level of the hyoid, the oesophagus is divided from the stomach and this hole is oversewn. A fresh opening is made in the fundus of the stomach which is anastomosed to the base of the tongue.

At the end of the operation and before the patient leaves the theatre a radiograph of the chest must be taken, and if pneumothorax is present, this must be treated appropriately by aspiration or insertion of an intercostal drain attached to an underwater seal unless the pneumothorax is very small. In any event the radiograph must be repeated after a few hours.

In recent years great strides have been made in microvascular surgery and it is now possible to restore continuity between the pharynx and the oesophagus by the use of revascularized segments of intestine, with success in about 95% of patients. At the end of the excisional phase a loop of ileum is taken by a second team taking care to preserve an artery and vein in its entirety. A suitable adjacent artery and vein of the same calibre as the ileal vessels are freed in the neck - the facial artery and vein are often suitable. The ileal loop is then freed, and the artery and vein reanastomosed in the neck. Finally the ileum itself is anastomosed, the upper anastomosis to the oropharynx often being end-to-side. Small fistulae can occur but the method is remarkably free of complications and is the treatment of choice where the tumour is confined to the neck. Larger tumours require visceral repairs which are in effect limited to transposition into the neck of either the stomach or the colon. Both procedures have had considerable success and many surgeons regard them as providing the most satisfactory method of restoring swallowing and speech after laryngopharyngectomy.

**Postoperative care**

**Speech therapy**

Only a tiny minority of patients develop 'oesophageal' speech after pharyngolaryngectomy. The best hope is one of the vibrating machines, but the results are generally poor.

**Thyroid and parathyroid replacement**

As the patient must undergo total thyroidectomy (and therefore parathyroidectomy) he must take thyroxine for life (usually 0.2 mg/day), and the serum calcium must be maintained within the normal limits by supplements of calcium and calciferol.
Diagnosis of recurrence

As has been discussed, both radiotherapy and surgery are used in the treatment of carcinoma of the hypopharynx and upper oesophagus and they are not mutually exclusive. Accordingly, after either form of treatment it is necessary to try to detect the presence of any residual or recurrent disease at the earliest possible moment while alternative treatment is still possible. Unfortunately there are difficulties in detecting further growth before it has reached an advanced stage and this is well illustrated by the very large number of patients reported in numerous series who succumb to their disease after being treated by only one method. Although there may be differences of opinion about the relative merits of each form of treatment used as a primary measure or as a salvage procedure, there is ample evidence to support the use of a second form of treatment if the first fails and, if this is not employed, it is usually because recurrent disease has advanced to the stage of being untreatable before being positively identified.

Treatment of any type alters the tissues and subsequent examination is a matter of determining whether the abnormalities are those which would be expected or whether they indicate that active tumour is still present. While positive biopsies confirm the presence of growth (unless taken within a few weeks of the completion of a course of radiotherapy) negative biopsy findings in no way exclude the possibility. After both radiotherapy and surgical excision there may well be healing of the surface while there is still active growth in the deeper tissue.

Mortality after pharyngolaryngectomy

Pharyngolaryngectomy is the most major head and neck procedure, and it has the highest mortality. Unlike survival, which is quoted at 5 years to ensure that the figure is representative, mortality rates are quoted on a very vague basis. The mortality ranges from 0.5% to 25% depending on the criteria chosen, from a death during operation at one extreme, to counting any patient who does not go home alive and swallowing at the other. There is an urgent need to standardize reporting so that different methods of treatment can be compared. Death within one week is probably a reasonable period to encompass those causes immediately connected with the operation. The mortality by this definition should be 0.5-1.0% for skin flaps and revascularized loops, and 10-15% for gastric transposition.
Chapter 14: Pharyngeal pouches

D. A. Bowdler

The leading symptom of diseases of the pharynx and oesophagus is dysphagia, one of the less common causes of which is a diverticulum. Pharyngeal diverticula may be posterior, posterolateral or lateral, but the most commonly encountered type is the posterior pharyngeal pulsion diverticulum.

Diverticula can present at any age, are benign lesions, and are therefore eminently curable. The diagnosis and treatment is well established, but the aetiology of the different types of diverticulum is still uncertain and controversial.

Embryology and anatomy

Knowledge of the embryological development of the branchial apparatus is essential when considering a classification of pharyngeal diverticula.

Each of the six branchial arches consists of a cartilaginous bar, from which the skeletons of the mandible, pharynx and larynx are derived, and is surrounded by mesoderm from which the muscles differentiate; an artery and cranial nerve supply each arch. Between the arches are depressions which, on the pharyngeal aspect, are lined by endoderm and called pouches, and on the external surface are lined by ectoderm and called clefts.

The fifth arch and pouch regress during development. The remaining four pouches, except the first, each grow laterally into a dorsal and ventral component, which contribute to the structures of the head, neck and mediastinum, although the development of the lower two pouches is not fully understood.

Splanchnic mesoderm migrates around the pharynx, investing it with a cover of three constrictor muscles; these are deficient anterolaterally where the neurovascular bundle to each branchial arch enters, representing potentially weak areas.

The constrictor muscles overlap each other; the superior lying innermost and the inferior outermost, inserting into a posterior midline raphe. A gap lies above the superior constrictor through which the eustachian tube passes.

A potential gap, overlain by the tonsil in life, remains between the superior and middle constrictors, bounded anteriorly by the hyoglossus muscle. The neurovascular bundle of the third arch, of which the nerve is the glossopharyngeal, passes through this gap.

The inferior constrictor is described in two parts, the thyropharyngeus and the cricopharyngeus. Thyropharyngeus arises from an oblique line on the thyroid ala and a fibrous arch between the thyroid and cricoid cartilages, its upper fibres overlapping the superior and middle constrictors increasingly as they pass posterosuperiorly, its lowest fibres lying horizontally edge to edge with the cricopharyngeus. A potential weakness exists between the middle and inferior constrictors, bounded anteriorly by the thyrohyoid muscle, and is pierced by the fourth arch neurovascular bundle, of which the nerve is the superior laryngeal. Below
the level of the vocal cords the thyropharyngeus is unsupported by the other constrictors, resulting in an area of weakness posteriorly, which is known as Killian's dehiscence.

The cricopharyngeus muscle is thicker and bulkier than thyropharyngeus, passing uninterrupted from one side of the cricoid to the other around the back of the pharynx. The sixth neurovascular bundle, of which the nerve is the recurrent laryngeal, enters the pharynx under the lower border of cricopharyngeus. The circular fibres of the oesophageal musculature lie below and parallel to the cricopharyngeus, but the longitudinal muscles at the upper end sweep forward to insert into the cricoid cartilage, leaving a relatively weak area, first described by Laimer and Hackermann, whose names are given to this area. The cervical oesophagus is not exactly midline tending to veer to the left.

The motor supply to the constrictors arises from the pharyngeal branch of the vagus, nerve, which forms a plexus on the middle constrictor. The cricopharyngeus muscle is an exception being supplied by the recurrent laryngeal and external laryngeal nerves. The sensory supply of the pharynx is via the glossopharyngeal nerve, the internal laryngeal nerve and the recurrent laryngeal nerve.

To summarize there are several relatively weak areas through which mucosal bulges or diverticula may develop.

Lateral
(1) Above the superior constrictor
(2) between the superior and middle constrictors
(3) between the middle and inferior constrictors

Posterolateral
(4) Laimer-Hackermann point

Posterior
(5) Killian's dehiscence.

Classification

Classifications are based on developmental, anatomical and aetiological grounds (Korkis, 1958; Wilson, 1962). In earlier classifications an anterior pharyngeal diverticulum was described but is now recognized to be an overdeveloped or deep vallecula, and not a diverticulum. The classification below is based primarily on anatomical site, and then subdivided by aetiology (Table 14.1).

Lateral pharyngeal diverticulum

Lateral pharyngeal diverticula are uncommon and arise from the posterior faucial pillar, and the upper or lower pyriform fossa. They are best observed with frontal cineradiographic views using a contrast material, while being emphasized by raised intrapharyngeal pressure.
**Congenital**

Congenital lateral pharyngeal diverticula are extremely rare. In most reported cases the aetiology has been attributed to a branchial cleft remnant which opens into the pharynx, usually at the lower pole of the tonsillar fossa by the posterior faucial pillar. Most arise from the second branchial cleft, the track passing between the internal and external carotid arteries before opening into the pharynx, but some have been reported in association with the third and fourth clefts.

**Table 14.1 Classification of pharyngeal diverticula**

**Lateral**

1. Congenital
2. Acquired
   
   a. normal bulges
   b. traumatic
   c. raised intrapharyngeal pressure 'pharyngocoeles'

**Posterolateral**

**Posterior**

1. Congenital
2. Acquired
   
   a. traumatic
   b. posterior pharyngeal pulsion diverticulum (Zenker's diverticulum).

They are usually unilateral and diagnosed in the first two decades of life, although occasionally in much older patients. The patients present with a history of recurrent infected swellings in the neck, previously treated with antibiotics or by incision and drainage, before the correct diagnosis is established. The signs are a tender fluctuant swelling in the anterior triangle of the neck, pyrexia, mild dysphagia and occasionally mild stridor. Investigation with radiological contrast studies after the infection has settled with antibiotics may demonstrate the narrow tract leading to a cyst in the neck.

Treatment is by excision of the cyst and tract to the pharyngeal wall, where the neck is closed and oversewn. Histological examination of the sac reveals an epithelial lining of stratified squamous or columnar epithelium surrounded by chronic inflammation with lymphocyte infiltration.

**Acquired**

Argument about the aetiology of acquired lateral pharyngeal diverticula still continues. Some authorities consider the basic defect to be a congenital weakness and so classify these diverticula as congenital, but the potential weak areas are present in all individuals, the variable or precipitating factor being raised intrapharyngeal pressure causing protrusion of pharyngeal mucosa through those areas which are developmentally deficient in muscle. These
diverticula do not seem to occur in the absence of such a predisposing factor and are usually found in patients well into adulthood, supporting the argument that they are acquired lesions.

Normal bulges

Frequent and incidental findings on routine barium swallows are small lateral pharyngeal bulges which can be seen either arising in the pyriform fossa or, more rarely, in the tonsillar fossa. They are more common in the elderly, probably due to reduced muscular tone and loss of elasticity of the tissues, and they are usually asymptomatic and bilateral, which is the reason they are thought by many to be normal variants.

Radiological contrast studies demonstrate the bulges as smooth, hemispherical prominences arising from the pyriform fossa or tonsillar fossa, the appearance lending itself to the name pharyngeal 'ears'. These are largest when a modified Valsalva manoeuvre is performed (blowing hard against pursed lips), collapsing down to a normal pharyngeal contour under normal pressure. They are most easily seen in frontal views. At this stage they require no treatment, but it is probable that they represent an early stage in the evolution of larger symptomatic diverticula.

Traumatic

Colonel M. Morris (late RAMC), while in India, recorded certain nomadic groups of habitual criminals from the Central and United Provinces, with self-inflicted diverticula. These were produced by pushing a piece of lead, the size of a pigeon's egg, into the tonsillar fossa. Repeated use eventually created a diverticulum that could be kept patent with a finger. Coins or jewellery could be hidden in this diverticulum and be delivered when required, by tilting the head forwards and effecting a vomiting motion. The diverticulum probably lies between the middle and superior constrictors, and if not used or maintained constantly would rapidly disappear (Atkinson, 1952).

Raised intrapharyngeal pressure (pharyngocoeles)

As stated above these large and occasionally symptomatic diverticula may arise from precursor pharyngeal 'ears'.

Two factors are implicated in their development, either separately or together: the first is frequent repetitive increases in intrapharyngeal pressure and the second is loss of muscular resilience associated particularly with advancing years. The diverticula evolve, from large outpouchings of the lateral pharyngeal wall, into sacs with obvious necks that are sometimes called 'pharyngocoeles'.

A pharyngocoele was first described by Wheeler (1886), in a patient who had been in the army and took considerable pride in his ability to command a full brigade on the parade ground with his own voice. Pharyngocoeles were also noted in Egyptian muezzins who sang verses of the Koran from the minarets of mosques. Many developed pharyngocoeles which often became so marked, that special collars were required to restrain them, must as a man wears a truss for an inguinal hernia. These uncommon pulsion diverticula are usually unilateral and asymptomatic affecting men more frequently than women, in a ratio of 8:1.
Occupations in which the intrapharyngeal pressure is raised, such as glassblowers or trumpeters, have been implicated in the aetiology of this condition.

Symptoms may develop due to food entrapment in the diverticulum, and are insidious in onset, so that the patient presents with long-standing problems. The main symptom is dysphagia which is usually intermittent and mild, the patient complaining of a sensation of food sticking in the throat, although it may become severe. Occasionally there is regurgitation of undigested food, with associated foul taste and fetor, which may lead to nocturnal coughing and choking. Dysphonia results from the effects of spillage into the larynx, although it is also suggested that this is due to a compression of the recurrent laryngeal nerve by the sac. Chronic pulmonary problems may be the first hint of the condition.

Signs are few. There may be a palpable lump in the neck, just anterior to the sternomastoid muscle, which is soft, compressible and which may gurgle due to a mixture of air and fluid within the diverticulum sac. Indirect laryngoscopy shows little although a slit-like ostium may be observed in the region of the posterior faucial pillar or the upper part of the pyriform fossa. The sac can usually be inflated voluntarily by the patient by blowing against closed lips in a modified Valsalva manoeuvre.

**Investigation and diagnosis**

Plain radiograph may show the sac as a translucency, lateral to the pyriform fossa which can be increased in size by a modified Valsalva manoeuvre. However, the mainstays of diagnosis are fluoroscopic and cineradiographic techniques which are most efficient in delineating the diverticulum, using high density barium which coats the mucosa more effectively and is retained longer. The patient is asked to take a small amount of contrast material and then to perform a modified Valsalva manoeuvre to emphasize the diverticulum. It will be seen as a rounded, contrast-lined opacity communicating with the pyriform fossa by an isthmus or neck. Ultrasound has also been used to make the diagnosis. Occasionally barium studies fail to reveal the sac and direct inspection is necessary. Direct pharyngoscopy shows a slit-like opening to a diverticulum, the search being concentrated in the areas where they are known to occur, although they can still be easily missed.

**Treatment**

If the diverticulum causes no symptoms, no treatment is required except for diligent follow-up. In those who develop symptoms, the diverticulum should be surgically excised through a neck incision. The sac is followed through the thyrohyoid membrane to its opening in the pharynx, amputated at its neck and the pharyngeal mucosa oversewn. Postoperatively, it is advisable to feed the patient by nasogastric tube for 3-5 days to ensure healing and to diminish the risk of fistulae.

**Differential diagnosis**

The main lesions that can be confused with a pharyngocoele are: branchial cyst; posterior pharyngeal pulsion diverticulum; laryngocoele; external or internal jugular phlebectasia; and pseudodiverticulum.
While the history and examination may allow exclusion of most other cervical lesions, laryngocoeles and venous phlebectasia closely mimic pharyngocoeles altering in size with a Valsalva manoeuvre.

**Posterolateral pharyngeal diverticulum**

This diverticulum, which protrudes through the Laimer-Hackermann point, may be either true or false being differentiated by its collapsibility with the passage of peristalsis. It presents in older people, again probably due to weakness of musculature. It is almost always asymptomatic, requiring no treatment. These are probably more common than reported. Being asymptomatic they are rarely noted endoscopically or radiologically and vary in size according to the position of the peristaltic wave (Ekberg and Nylander, 1983).

**Posterior pharyngeal diverticulum**

*Congenital*

Congenital posterior pharyngeal diverticula are very rare and were first described by Brintnall and Kridelbaugh (1950) in two infants who, soon after birth, developed symptoms similar to oesophageal atresia. Radiological evidence of air in the stomach, in the absence of a tracheo-oesophageal fistula, confirms oesophageal patency. The diverticular sac arises from above the cricopharyngeus and is lined by normal pharyngeal mucosa, distinguishing it from a traumatic pseudodiverticulum. The whole sac is covered with muscle, differentiating it from a pulsion diverticulum. The treatment is excision of the diverticulum to restore pharyngeal continuity.

*Acquired*

**Traumatic pharyngeal pseudodiverticulum**

This rare condition usually presents in newborn infants, but has been reported in adults. The aetiological factor is hypopharyngeal trauma, either from damage caused by the obstetrician's finger during breech delivery, or vigorous instrumentation with suction tubes or endotracheal tubes, although in one of the adult cases the cause was spontaneous rupture of a retropharyngeal abscess in an immunosuppressed patient (Morton, 1983). Patients develop symptoms some hours after the traumatic episode, which causes a hypopharyngeal mucosal tear, either transmural or submucosal, the former causing more severe symptoms as a result of a false passage tracking down into the posterior mediastinum in the prevertebral space. This leads to cricopharyngeal spasm which precipitates the early symptoms of increasing dysphagia with excessive oropharyngeal secretions and inability to ingest feeds. Eventually, attempts to feed cause coughing and choking due to spillage into the larynx, sometimes resulting in cyanotic episodes and, at worst, aspiration pneumonia. The patient becomes pyrexial and increasingly ill, developing symptoms and signs of mediastinitis. There may also be cervical subcutaneous emphysema. The symptoms and signs mimic other conditions including oesophageal atresia or duplication, and congenital posterior pharyngeal diverticula, although a clear antecedent history helps distinguish it from the above conditions.
The management is not clearly established as few cases have been reported. To confirm the diagnosis an attempt is made to pass a nasogastric tube into the stomach; if gastric fluids can be aspirated, oesophageal atresia is excluded. Contrast material is then passed down the tube which is slowly withdrawn to give a retrograde picture of the upper alimentary tract, to demonstrate the pseudodiverticulum. If a nasogastric tube cannot be passed into the stomach, contrast material is injected down the tube at pharyngeal level which again should demonstrate the diverticulum. Alternatively, the patient sucks a contrast material feed. However, with the two latter methods, there is a considerable risk of inhalation and worsening of the pulmonary condition.

The radiological appearance of a traumatic pharyngeal pseudodiverticulum is an irregular elongated tract originating in the pharynx and passing behind the oesophagus into the posterior mediastinum.

The management is either conservative or surgical. Conservative treatment consists of intravenous antibiotic therapy and nutritional support either by nasogastric tube or intravenously. The surgical treatment consists of drainage of the pseudodiverticulum either via a cervical incision or a thoracotomy, at which time it is usual to perform a gastrostomy for nutritional support. Antibiotic cover is given. The excised track shows fibrous or inflammatory tissue rather than epithelium.

**Posterior pharyngeal pulsion (Zenker's) diverticulum**

**History**

The posterior pharyngeal pulsion diverticulum is known by many names including pharyngo-oesophageal pouch or diverticulum, retropharyngeal pouch or diverticulum, posterior pharyngeal pouch or diverticulum, and Zenker's diverticulum.

The first case of posterior pharyngeal pulsion diverticulum was described in 1764 by Mr A. Ludlow, a surgeon from Bristol, in a letter to Dr William Hunter of the Society of Physicians in London. A paper entitled 'A case of obstructed deglutition, from a preternatural dilatation of, and bag formed in the pharynx' was published in 1769 in *Medical observations and Inquiries*. The description of the symptoms and morbid anatomy is remarkably accurate when compared to present-day knowledge. From the history and examination Ludlow though that the obstruction was in the cervical region of the oesophagus due to a stricture. Initially he attempted to treat the patient by asking him to swallow a weighted silk thread, but this caught in the diverticulum. Blind dilatation was then attempted using whalebone bougies, but this also failed, next, half a pound of quicksilver or mercury was given but, alas, this again lodged in the diverticulum without passing into the stomach; in spite of the treatment the patient died after 13 days. A post-mortem was carried out revealing that the 'bag' or diverticular sac lay between the oesophagus and the vertebral column, originating from the posterior wall of the pharynx. The diverticulum was thought to have been caused by a cherry stone lodging in the posterior wall some time earlier.

A further 22 cases were described between 1764 and 1874. Zenker and Van Zeimssen collected these reports and with five patients of their own described the site of origin of the diverticular sac as between the pharynx and oesophagus, its possibly aetiology, and
differentiated between pulsion and traction diverticula. Zenker's name is still associated with the posterior pharyngeal pulsion diverticulum. Early surgical attempts at curing the condition were unsuccessful but in 1886, Wheeler reported the first successful excision of a posterior pharyngeal pulsion diverticulum, performed the year before in 1885. It was an incidental finding while operating on a patient with a large acquired lateral pharyngeal diverticulum or pharyngocoele. The patient recovered completely and deglutition and voice returned to normal. However, the mortality rate due to sepsis, in particular mediastinitis, was extremely high, making the procedure hazardous. Other methods were therefore advocated including inversion of the sac, diverticulopexy, dilatation and endoscopic division of the party wall, and a two-stage diverticulectomy. The two-stage procedure was popular in the USA and the UK but since the advent of antibiotics it has become normal to perform a one-stage excision, usually combined with cricopharyngeal myotomy, which today carries a low mortality rate. However, despite all the surgical advances in the relief of symptoms the aetiology of the condition remains uncertain.

Aetiology

Swallowing is a complex mechanism by which a bolus is passed from the mouth to the pharynx, and into the oesophagus and stomach, in a quick, coordinated fashion. The shortest phase is the pharyngeal, or second stage of swallowing, lasting less than one second. The mechanism of the second stage comes into action as the first stage is completed with the bolus being ejected into the oropharynx. The larynx is elevated with the hyoid to lie under the mandible and base of the tongue. The aryepiglottic folds contract to complete closure of the larynx, already closed at glottic level due to adductor contraction. The epiglottis is inverted by the bolus passing down the pharynx, and the pharynx is elevated over the bolus simultaneously. A pharyngeal peristaltic or stripping wave then pushes the bolus down through the pharynx and cricopharyngeal sphincter, which is passively or reflexly relaxed as the second stage of swallowing begins. Most of the bolus has preceded this wave, which clears only the more solid particles.

The aetiology of posterior pharyngeal pulsion diverticula is still unknown, although many theories have been advanced. There is much conflicting evidence from investigations using radiographic and manometric techniques, even when using the same method of study. The early theories fall into four main categories:

1. tonic spasm of the cricopharyngeus (Negus, 1950)
2. lack of inhibitory stimuli to the cricopharyngeus (Dohlman and Mattson, 1959)
3. the second swallow (due to pharyngeal laxity) (Wilson, 1962)
4. neuromuscular incoordination and congenital weakness (Korkis, 1958).

1. Negus was the proposer of this theory. As man evolved to an erect position, so the larynx and pharynx moved lower in the neck and the circular muscles or constrictors became more oblique with the exception of the lower fibres of the inferior constrictor or cricopharyngeus. A weakened area, Killian's dehiscence, was left between the cricopharyngeus and the thyropharyngeus. Negus believed the weakness was compounded by the lack of
longitudinal muscle support posteriorly. However, the anatomical factor is common to all humans so could not alone account for sac formation. One factor is variable, that of incoordination of the second stage of swallowing, in particular relaxation of the cricopharyngeus in front of the pharyngeal stripping wave. Negus believed that a predisposing factor to diverticular formation was cricopharyngeal sphincter spasm due to chronic inflammation, stenosis from healed inflammation, or an unknown neurological deficit. When the bolus reached the sphincter, which was in spasm, it would be forced posteriorly and cause a mucosal bulge. As this enlarged between the cervical oesophagus and vertebral column the sphincter would move anteriorly and so the bolus would impinge directly on the posterior pharyngeal wall with resultant expansion of the pharyngeal diverticulum.

(2) Dohlman and Mattson felt that the sphincter failed to relax, rather than being in active spasm. They believed that the cricopharyngeus was normally tethered to the prevertebral fascia, but that this attachment weakened with ageing so that, the larynx was elevated on deglutition pulling the cricopharyngeus with it rather than stretching the muscle, which would normally trigger off a reflex relaxation of the sphincter in readiness for the bolus. Failure of relaxation would increase intrapharyngeal pressure causing mucosal bulging posteriorly, which was further compounded by a negative pressure in the prevertebral space, sucking the mucosa outwards.

(3) The second swallow theory was proposed by Wilson, who noted that in his patients there was a high incidence of enlarged pharynges or megapharynx which, he believed, was due to a lax pharyngeal musculature. Furthermore, he observed that there was always a residue of barium in the pharynx after the swallow was completed. He speculated that part of the bolus failed to pass into the oesophagus before the sphincter closed and so the patient made a second voluntary swallow in order to clear the residue, but that this occurred against a closed sphincter, resulting in an area of high cricopharyngeal pressure between the sphincter and pharyngeal stripping wave, causing the mucosa to bulge posteriorly, leading to the development of a posterior pharyngeal pulsion diverticulum.

(4) Korkis believed that there was some neuromuscular incoordination which was compounded by congenital weakness and he supported the argument by recalling the congenital posterior pulsion diverticulum described by Brintnall and Kridelbaugh.

However, all agree that the weakness lies at Killian's dehiscence and that, in order for it to occur, there should be raised intrapharyngeal pressure.

Both Negus and Dohlman's proposals have been shown to be inaccurate. Kodicek and Creamer (1961) first described manometric studies using open-tipped recording tubes in the pharynx and oesophagus attached to a capacitance manometer. They were able to show that the cricopharyngeus relaxed normally and that there was no incoordination, although others have subsequently questioned this premise. However, their results are supported by Hunt, Connell and Smiley (1970) who, while demonstrating a high resting cricopharyngeal pressure, showed normal relaxation of the cricopharyngeus in relation to swallowing in patients with diverticula. More recently Knuff, Benjamin and Castell (1982), using modern manometric techniques including a low compliance infusion system and oval-shaped catheters, studied nine patients with known posterior pharyngeal pulsion diverticulum initially diagnosed by barium swallow studies. The result of this research showed normal relaxation of the
cricopharyngeus which did not contract until the end of the pharyngeal stripping wave, although a low resting pressure was found in the sphincter or pharyngo-oesophageal segment.

Ardran, Kemp and Lund (1964) argued that since most studies were performed in patients with established diverticula, which must upset the normal physiology of swallowing, the results might be due to the disease and not reflect the cause. Using contrast cineradiography with a speed of 25 frames/second on 35 mm film, they examined 16 patients with diverticula of differing sizes and 17 normal subjects. After the initial part of the swallow, during which barium passed easily into the oesophagus, the main bolus descended into the pharynx to be moved on by the pharyngeal stripping wave. In patients with diverticula they found this to be defective in two ways; first, the oropharyngeal contraction was weak or absent, and second, pharyngeal constrictor function below this level was disturbed, which together with premature cricopharyngeal closure, caused a residue to remain in the pharynx with a bulge or diverticulum forming as a result. Consequent upon their studies they proposed a mechanism of diverticulum formation. The cricopharyngeus contracts prematurely and the posterior wall bulges backwards. As the stripping wave descends to the closed sphincter, it pushes the posterior wall down and forwards to meet the back of the cricopharyngeal sphincter, which is facing upwards and forwards, and so a dimple is produced which might well go on to enlarge and become a diverticulum.

In some patients they noted failure of complete cricopharyngeal relaxation which led to a nipping between the sphincter and the stripping wave with a consequent bulge, which they suggested could also exacerbate diverticulum formation. In their study they noted no megapharynges in normal subjects and only one in 16 patients with known diverticula, which contradicts the findings of Wilson. Also, the cricovertebral distance was the same when comparing normal subjects with patients with diverticula, which does not support the Dohlman and Mattson theory.

Ellis et al (1969), and later Lichter (1978), confirmed by manometric techniques the early cricopharyngeal closure proposed by Ardran, Kemp and Lund. Lichter found that there was also premature relaxation of the sphincter, which was followed by early contracture leading to raised intrapharyngeal pressure. They did not demonstrate any weakness of pharyngeal contraction, but in some patients noted a repetitive swallowing pattern, probably as a result of the obstruction. They examined patients, all of whom had had cricopharyngeal myotomy, after surgery and demonstrated reduced resting pressures in the region of the sphincter. They suggested that cricopharyngeal myotomy should be performed whenever it was noted to be radiologically prominent.

Smiley, Caves and Porter (1970) noted that in 32 out of 34 patients with Zenker's diverticulum, there was associated oesophageal reflux or hiatus hernia. On studying patients with oesophageal reflux alone, they noted a high resting sphincter pressure but no incoordination of swallowing. They proposed that oesophageal reflux or hiatus hernia might well play a causal role in the formation of Zenker's diverticulum.

A familial tendency has also been noted by McNab-Jones (1959) in a report on a patient whose mother, sister and two brothers all had posterior pharyngeal pulsion diverticulum. Groves (1968) found three sisters who had diverticula, and the possibility, although unproven, of other members of the family being likewise affected.
Incidence

It is difficult to quantify the incidence of pharyngeal diverticula in the general population, as not all cases are reported, and centres with a special interest in these diverticula attract cases from other areas, thus falsifying their figures in relation to population. The only report that gives figures in this manner is from Ipswich (Juby, 1969), where 17 cases were reported in a population of 300,000 over a 12-year period; an incidence of 0.47 cases/100,000 persons per year. Most figures refer to numbers seen over a fixed period or relate them to hospital admissions, operations or radiographic studies. Shallow and Clerf (1948), stated an incidence of one in 1400 admissions, or 800 operations, MacMillan (1932) recorded finding 18 diverticula in 1000 contrast radiographs for patients with dysphagia, and Baron (1982) reported one in 800 routine barium studies.

Age, sex and race

Figures for age and sex are more certain, although there is variation between different centres. A collection of some of the largest series gives a ratio of about two men to one woman. Patients are generally aged over 50 years at diagnosis, but occasionally present as young as 29 years old. From a number of American series it is apparent that negroes are rarely affected by this condition.

Symptoms

Patients present with variable severity of symptoms, not necessarily related to the size of their diverticulum. Although some patients present after only a few months of symptoms, most complain of long-standing problems, the patient having adapted to the slowly progressive symptoms. Indeed, it is the insidious nature of the onset that causes most patients to present with a well-developed diverticulum. There have been attempts to stage symptoms in relation to diverticulum size, in particular by Lahey (193) who described three stages:

- **stage I:** small mucosal protrusions (the initial stage)
- **stage II:** a definite sac but with the oesophagus and hypopharynx still in line (the intermediate stage)
- **stage III:** a large sac with the hypopharynx in line with the neck of the diverticulum, and the oesophageal inlet pushed anteriorly.

Each stage was said to be associated with a symptom pattern: stage I, being associated with the sensation of food sticking in the throat; stage II, having regurgitation and gurgling from the pouch; and stage III, with development of severe dysphagia. The main use of this classification is in relation to the mortality and morbidity of operative procedures. The symptoms are listed in their frequency of occurrence.

Dysphagia

This symptom may be misinterpreted by the clinician, leading to prolonged periods when no treatment is administered. Initially the patient may complain of a sensation of a lump in the throat, which can frequently be misdiagnosed as globus hystericus. Other early symptoms are a feeling of food sticking in the throat, requiring repeated swallowing attempts.
Generally, however, the story is of increasing difficulty in swallowing solids, requiring the patient to chew every mouthful finely. As the condition progresses it becomes impossible to enjoy a meal with friends, due to the length of time taken to eat. Eating becomes acutely embarrassing. Eventually difficulty with semi-solid foods and then liquids develops. Occasionally a patient cannot swallow his own saliva, having to expectorate the excessive oropharyngeal secretions, which rapidly leads to dehydration. It has been suggested that pressure of the diverticulum on the upper oesophagus causes obstruction but manometric studies detected no change in oesophageal pressures, even with large diverticula.

**Regurgitation**

Patients suffer from regurgitation, undigested food welling into their mouths, sometimes during a meal, although more often afterwards. It is exacerbated by positional change, especially lying down in bed at night. This symptom can wake the patient in the middle of the night, when spillage from the diverticulum causes choking. A few adapt to this by evacuating the sac before going to bed, by pressing on the side of their neck over the diverticulum.

Associated with this symptom, the second most common after dysphagia, is a foul taste in the mouth due to the prolonged retention of undigested food. A gurgling sound on swallowing is sometimes noticed by the patient due to a mixture of air and fluid in the sac.

**Weight loss**

Due to dysphagia which may be evident for a considerable time before examination, some patients present with severe weight loss and malnutrition complicating the treatment of this benign condition.

**Hoarseness**

Overflow of sac contents into the larynx causes chemical irritation and laryngitis. It is suggested that this can be due to pressure of the sac on the recurrent laryngeal nerve, although vocal cord paralysis is more likely to be a result of the presence of a carcinoma in the diverticulum.

**Pulmonary complications**

A serious sequel to the spillage of sac contents into the larynx is aspiration pneumonia. Pulmonary complications, including pneumonitis or lung abscesses, are well recorded with large sacs, and treatment of these is necessary before surgical resection.

**Miscellaneous**

There is usually no pain except in the presence of carcinoma. Occasionally other strange presentations occur. Bleeding has been reported from a diverticulum, due to ulcers and carcinoma. Also rare, is a diverticulotracheal fistula which causes the patient to cough when eating, and leads to pulmonary complications. Resection of the tract and diverticulum cures
this problem. One odd symptom was a patient's failure to absorb medication due to the tablets lodging in the sac. This was also cured by resection.

**Signs**

The signs include emaciation, which can be severe, although it is uncommon nowadays as diverticula are usually picked up earlier. A swelling may be found in the neck, usually on the left side, in the lower part of the anterior triangle, which is soft, and may gurgle on palpation. This is known as Boyce's sign. A spasm of coughing may be caused by palpation due to spillage of contents into the larynx. Indirect laryngoscopy demonstrates red larynges and pooling of saliva in the pyriform fossa in which undigested food particles may be seen.

**Investigations**

While the history and the examination may be virtually pathognomonic, it is necessary to confirm the diagnosis with radiological evidence, generally contrast radiography, employing either fluoroscopy or cineradiography.

**Plain radiography**

Plain lateral radiology of the soft tissues of the neck may give a clear impression of a diverticulum, the features of which are a triangular translucency seen in the prevertebral soft tissues, with its apex at the cricoid level. The base of the translucent triangle has a meniscus, due to the fluid in the fundus of the sac, with the air above it, and the sides curve to join each other superiorly.

**Contrast radiology**

Single shot barium swallow radiographs may not demonstrate a diverticulum if it is small or if they are taken from the wrong angle or at the incorrect stage of the swallow. Therefore, it is normal to use continuous monitoring methods, such as fluoroscopy or cineradiography, which allow good observation from different angles of all stages of the swallowing mechanism.

While a diverticulum usually projects posteriorly, as it increases in size it may be displaced to overlie the oesophageal shadow on lateral views. Therefore, with larger sacs an oblique lateral view is frequently needed. Fluoroscopy is especially useful in this respect. Using fluoroscopy or cineradiography the sac is seen to change shape in different stages of the deglutition cycle, its classic pear-shape appearing during the contraction of cricopharyngeus at the end phase of the pharyngeal stripping wave. An anteroposterior view is required by the surgeon in order to assess to which side of the neck the sac is deviating, so that the best approach can be planned. It usually deviates to the left. Further features of interest are the formation of an upper lip to the sac, caused by the contracting thyropharyngeus, and a lower lip caused by the cricopharyngeal impression at the end stage of deglutition. The cricopharyngeus may be seen to be a pronounced bulge in the diverticulo-oesophageal septum either at this stage, or even earlier if there is incomplete sphincter relaxation, and it is in these patients that Lichter recommends a cricopharyngeal myotomy.
The radiographic study is incomplete if it does not include the lower oesophagus, stomach and duodenum, in order to look for other abnormalities such as hiatus hernia or peptic ulceration with which there is a strong association (Smiley, Caves and Porter, 1970).

Although many long-term radiographic follow-up studies have been performed no one as yet has seen a diverticulum develop in a patient. Previously untreated radiologically diagnosed diverticula have been noted to increase in size over the years, but there are only sporadic reports of a transitory diverticulum developing into a posterior pharyngeal pulsion diverticulum. Radiological staging has been attempted: stage I, small, that is, less than one vertebral body; stage II, medium; stage III, large, that is, greater than three vertebral bodies. As with the Lahey classification, the main value of staging is in relation to the mortality and morbidity of surgical procedures. As well as noting the size and position of the diverticulum, the internal contours should be examined. An irregularity or filling defect within the diverticulum itself may be caused by solid food remnants or by a carcinoma; if due to a food remnant then the defect should not be constant on repeated examinations. The presence of a constant filling defect is highly suggestive of carcinoma.

A tracheogram is often produced. Ekberg (1983), in a radiological investigation of 250 patients with dysphagia, noted epiglottic dysfunction in one-third of the subjects. This included many with cricopharyngeal spasm and one with a posterior pharyngeal pulsion diverticulum. Twenty-seven subjects had barium spillage into the larynx and trachea. It is possible that pharyngeal dysfunction is the cause of pulmonary problems rather than the diverticulum itself, although it is not possible to confirm this. Diagnosis has also been confirmed by ultrasound, but radiological investigations remain the sharp end of the diagnostic armoury.

Occasionally the diagnosis is not made by barium studies, which have a failure rate in examinations of the gastrointestinal tract, and oesophagoscopy may be required. Oesophagoscopy and biopsy are also necessary to confirm the diagnosis of suspected carcinoma of a diverticulum, which may influence the treatment.

Pathogenesis

The diverticulum starts as a small bulge at Killian's dehiscence. As it enlarges it lies between the oesophagus and the vertebral column and may remain static for many years or slowly increase in size until eventually it passes into the posterior mediastinum. The plane of the diverticular neck alters as the size increases until it, rather than the oesophagus, lies in line with the hypopharynx, such that the food will pass into the sac preferentially. This feature also makes identification of the oesophageal opening quite difficult at oesophagoscopy, and often presents blind attempts to pass a nasogastric tube into the oesophagus.

It has been suggested that a diverticulum exerts pressure on the oesophagus from behind, causing the dysphagia, but this has not been proven in manometric studies on patients with known diverticula. The diverticulum is usually in the midline but, if it deviates it usually does so to the left.

The histology shows a sac consisting of an epithelial lining which is stratified squamous epithelium and submucosa, often with fibrous tissue surrounding it. Nearer the neck
of the sac scanty muscle fibres are found in the wall. Occasionally there are variations, in particular carcinoma in situ and frank invasive squamous cell carcinoma. Other histological oddities have been reported, including ulceration of the pouch with underlying submucosal infiltration by plasma cells, lymphocytes and eosinophils. Harrison and Tighe (1970) reported a sac which appeared to be covered completely with a fibromuscular layer, as one might expect in a true diverticulum. The sac was lined with hyperplastic stratified squamous epithelium with some acute inflammation and ulceration, but underlying this were cysts lined with stratified columnar mucous-secreting epithelium. The only explanation that could be offered for this rather odd findings, was of a developmental abnormality of the diverticulum, much as the congenital posterior pharyngeal diverticula described by Brintnall and Kridelbaugh (1950).

**Treatment**

No treatment is indicated for a diverticulum with few symptoms, if the patient's general condition is poor, or for transitory diverticula. Clearly each case must be judged on its individual merits, with the patient being fully aware of the possible complications and potential benefits of operation. Some patients, especially when old, are happier, and perhaps wiser, to live with minor inconveniences rather than embark on potentially major surgery. The basis of treatment is to correct the cause and remove the effect. The method for achieving these aims may be endoscopic or external surgery. It is agreed by most surgeons that the cricopharyngeal sphincter is probably the main factor in the aetiology of posterior pharyngeal pulsion diverticulum although the interrelationship between it and the pharyngeal constrictors in the aetiology is not yet understood. Operations in which the sphincter is not divided have a higher recurrence rate, supporting the argument for myotomy in all cases.

**Endoscopic treatment**

Ever since endoscopic techniques were developed, surgeons have been evolving new procedures to avoid the necessity for external surgery. The main endoscopic techniques for treatment of posterior pharyngeal pulsion diverticula are dilatation of the sphincter and endoscopic diathermy of the diverticulo-oesophageal septum.

**Dilatation**

Early treatment of diverticula was aimed at dilating the cricopharyngeal sphincter to alleviate the dysphagia. Dilatation with bougies is effective, but only temporary, in relieving symptoms and does not remove the diverticulum, resulting in an eventual recurrence of symptoms. There is an additional risk of perforation of the sac. This was especially true in the early days when blind bouginage was practised. An alternative method of dilatation is extensive stretching of the sphincter using a hydrostatic bag. However, dilatation is not frequently used nowadays except to dilate a postoperative stenosis.

**Endoscopic diathermy (Dohlman's operation)**

This operation has failed to gain wide acceptance as the treatment of choice in the UK or the USA, but is more popular in some European countries. It was first described by Mosher (1917) in the USA, in a series of six patients in whom the septum between the diverticulum
and the oesophagus was divided with scissors. The technique was modified and made popular by Dohlman and Mattsson (1960) who used it extensively and recorded over 100 cases in which there were no deaths or serious complications, although the recurrence rate was 7%. The rationale behind this operation, in preference to an external approach, is based on the general consideration of these patients. The patients are often old and unfit due to emaciation and pulmonary complications from aspiration of the sac contents. Being elderly they also have a higher incidence of cardiac and pulmonary disease, and therefore represent a poor anaesthetic and surgical risk. Endoscopic diathermy of the diverticulo-oesophageal septum is a short operation, lasting only 5-10 minutes, and can be carried out under local anaesthetic, if a general anaesthetic is contraindicated. Recovery is rapid and the patient is normally ready for discharge 4 or 5 days after operation. Furthermore, the size of the sac does not affect the division of the septum. While the procedure does not remove the pouch, it relieves the symptoms and restores swallowing by dividing the cricopharyngeus and widening the mouth of the diverticulum. Also, re-operation is far easier than after external operations where scar tissue makes identification of recurrent diverticula hazardous.

Specialized instruments have been developed for this technique. The oesophagoscope is split distally, the upper beak being longer than the lower with a slit between them. The instruments include a diathermy forceps, which has teeth on the jaws to prevent slipping, a knife and a suction tube, all of which are insulated except at their working ends, and a pair of insulated paddles.

The patients are admitted one day before the operation and the sac is cleared of food debris by washing out with a stomach tube. The patients are restricted to clear fluids, and on the evening before the operation a weighted thread is swallowed, the proximal end being taped to the cheek, so that the oesophageal opening can be more easily found.

At operation, under either general or local anaesthesia, the long beak of the oesophagoscope is inserted into the oesophagus being guided by the thread, and the short beak into the diverticulum. The wall between the diverticulum and the oesophagus lies in the slit as a horizontal spur, containing the cricopharyngeal muscle. The diverticulum is cleared of any excess debris by suction and the spur is then grasped in the midline between the jaws of the diathermy forceps and coagulation diathermy applied until blanching occurs. The forceps are removed and a diathermy knife is then used to divide the coagulated strip of tissue longitudinally. In order to avoid coagulating or cutting the wrong wall, insulated paddles are passed down, one into the pouch, the other into the oesophagus so that the knife cuts down on to the paddle. Small bleeding points are coagulated with the suction diathermy. This process may be continued caudally until the fundus of the diverticulum is seen. With large sacs the operation is often staged, being completed after one or two repeat operations, each separated by 5-6 days. To minimize the risk of mediastinitis the procedure is stopped before the floor of the diverticulum is reached. The success of the operation is gauged by postoperative barium studies performed at 5 days, at which time a decision can be made to stop or to proceed further. Although some residue may be noted temporarily in the fundus of the sac, the operation is deemed successful if there is only a short party wall remaining with minimal delay in the emptying of the pouch into the oesophagus, and of course an absence of symptoms. The patient's fluid balance is maintained by intravenous infusion until the following morning when fluids are given orally, if there are no contraindications. If there are
no problems with fluids, soft diet is instituted on the second or third day and the patient is discharged after 5 days. Normal diet is allowed after 2 weeks.

Endoscopic division of the diverticulo-oesophageal septum provides a quick, safe technique for relieving symptoms, the major risks being haemorrhage, mediastinitis, emphysema and stenosis, the latter being treated by further division or dilatation. However, there is a risk of carcinoma in pouch being missed. The method has been criticized because of the recurrence rate and the possibility of carcinoma developing after treatment, a fact which has caused many to abandon its use.

**External surgical approach**

In the late 1800s and early 1900s the high mortality of diverticulum surgery promoted a search for differing methods of treatment to minimize the complications, so common in the pre-antibiotic era; all were variations on treatment of the sac, once dissected. Diverticulopexy, securing the fundus of the sac high in the neck, is rarely performed now. Two-stage diverticulotomy, once the mainstay of treatment, has been superseded by one-stage excision. Inversion has been reported more often in recent years and has considerable advantages, the only drawback being the possibility of missing a carcinoma. Cricopharyngeal myotomy remains a point of controversy.

**Two-stage diverticulectomy**

In 1909 Goldman ligated the neck of the sac and brought the fundus to the surface of the wound. When the sac sloughed a controlled fistula resulted which closed within a few weeks. This technique was subsequently modified by suturing the sac to the skin and opening it 2 weeks later, dissecting the mucosa down to the neck of the sac and packing the tract to leave a fistula which closed by secondary intention. This was a safe method minimizing mediastinitis and the staunchest advocates of this method were Lahey and Warren (1954), from the Mayo Clinic, where 365 operations were performed with only two deaths.

**Inversion**

Inversion of the diverticulum was first described in 1895 by Girard. Bevan, in the Sippy-Bevan operation (1917), modified the inversion by placing a series of purse string sutures along the length of the sac in order to obliterate it. This technique was developed to avoid the risk of opening the sac, associated as it was with serious problems of sepsis. Inversion is being used more often nowadays as it has considerable advantages. There is a low complication rate, in particular of fistula, and the patient has a short hospital stay of 4-6 days. The patients occasionally complain of a sensation of a lump in the throat for 2 or 3 days, but this quickly subsides. Patients are able to drink the day after surgery.

The operation is carried out routinely to the point of full mobilization of the sac and cricopharyngeal myotomy, the pouch is then invaginated into the oesophagus and its neck oversewn with interrupted catgut sutures. A small drain is placed in the wound which is closed in the normal manner.
**Diverticulo-oesophagostomy**

Described by Jackson, Slack and Williams (1960), this operation was used for a patient who had a very large diverticulum entering the mediastinum. The barium studies suggested that it was adherent to structures in the right thorax, implying potential risk when mobilizing the pouch up into the neck. The diverticulum was drained into the thoracic oesophagus at its fundus; the operation was successful and the patient rapidly gained weight.

This is clearly a specialized idea for a particular problem and will rarely be deemed necessary, but does offer an alternative for larger sacs.

**Cricopharyngeal myotomy**

This has been used to treat neurological disorders of swallowing including achalasia of the sphincter, muscular dystrophy, and myasthenia gravis. It is strongly argued that it should be a standard part of the operative treatment of diverticulum, although the evidence to support this is controversial.

The procedure is simple and relatively risk free, being advocated as the only treatment needed for small diverticula. As this procedure takes only 5 minutes to perform it should be part of the standard operation.

**One-stage diverticulectomy**

Although this was the first successful method it did not become popular until antibiotics were readily available. The preoperative examination is important, the patient being admitted 2 days preoperatively and the sac emptied by periods of resting in the head-down position. The patient is restricted to clear fluids for the last 24 hours. It is suggested that a black silk thread be swallowed, the proximal end being secured to the cheek, in order to facilitate identification of the oesophagus, down which it should pass.

The patient's general health is assessed with particular emphasis on the chest. Treatment of any chest pathology is essential prior to surgery. The operation is usually performed under general anaesthesia with the patient intubated and paralysed. The radiographic films are examined again in theatre and it is confirmed to which side the pouch has deviated. The operation is performed in two stages, the first of which is oesophagoscopy and the second, the external approach. Once the patient is anaesthetized, an oesophagoscope is passed and the openings to the sac and oesophagus identified. A nasogastric tube is passed through the cricopharyngeal sphincter to the stomach after which the diverticulum is inspected to exclude carcinoma and to suck any debris from it before packing with ribbon gauze soaked in proflavin, the proximal end of this strip being brought out through the mouth to the head of the table so that the anaesthetist can remove it at the appropriate time without disturbing the drapes. Attempts at passing the nasogastric tube after packing the pouch can be hampered by the bulk of the pack impinging on the oesophagus, it is therefore recommended that the nasogastric tube be passed first.

The patient is then placed in the reverse Trendelenburg position with a sandbag under the shoulders and the head extended and rotated away from the side of the incision. The
operation area is sterilized and draped, and a collar incision marked out on the skin with methylene blue at the level of the upper border of the cricoid from the midline to halfway across the sternomastoid muscle, preferably in a skin fold. Some surgeons advocate an incision along the anterior border of the sternomastoid muscle, but this is generally unnecessary and uncosmetic. The incision line is infiltrated with adrenaline to minimize bleeding and the incision made through skin, subcutaneous tissues, and platysma to the strap muscles and sternomastoid. The deep cervical fascia is then incised anterior to the sternomastoid muscle which is retracted laterally. The omohyoid is identified, mobilized and divided between clamps, at which point the internal jugular vein comes into view. The middle thyroid veins are clamped, divided and ligated so that the dissection may proceed medial to the carotid sheath, which is gently retracted laterally avoiding undue pressure on the carotid artery. The inferior thyroid artery is identified and divided as necessary and, if possible, the recurrent laryngeal nerve is identified at this point of the operation. The diverticulum, which is packed with proflavin gauze, may be easily identified by colour or palpation. The fundus is grasped with Babcock forceps and carefully dissected free of the oesophagus inferiorly, while, in order to improve the exposure, the thyroid gland and thyroid cartilage are retracted medially, care being taken not to put pressure on the recurrent laryngeal nerve. The sac is then held by the surgeon while sweeping muscle fibres from the proximal part of the body of the diverticulum, with special care being taken to clear the neck so that its junction with the pharynx, which is identified by palpating the nasogastric tube, can be clearly seen. Great care must be taken to avoid tearing the sac at this juncture as it can easily extend into an oesophageal tear leading to considerable difficulties. The proflavin pack is then removed and stay sutures are inserted into the neck of the sac inferiorly and superiorly, care being taken not to place them too medially, thus causing a stricture.

The cricopharyngeal sphincter and upper circular fibres of the oesophagus are then divided posteriorly thereby avoiding the recurrent laryngeal nerve. There are many ways to facilitate myotomy but the easiest method is to place an artery forceps between the submucosa and the cricopharyngeus muscle from above, the blades are opened and a knife used to divide the fibres, which are stretched between the two jaws of the forceps. Stretching the muscle by the intrapharyngeal introduction of stomach tubes or inflated Foley catheters and endotracheal tubes, has been described; in addition the operating microscope has been used to ensure division of every single muscle fibre. After the myotomy, the wound inferior to the neck of the sac is packed with gauze to catch any debris which may discharge when the sac is amputated. The mouth of the neck is then closed with a continuous inverting suture and the stay sutures removed. A second layer of interrupted catgut or silk is then used to bury the first. Haemostasis is secured before wound closure, and a drain is inserted inferiorly. Suction drainage is avoided due to the risk of salivary fistula or damage to the recurrent laryngeal nerve. The wound is closed in two layers with continuous chromic catgut subcutaneously and prolene to the skin. Antibiotics should be reserved for complications and are not routinely used. The drain is removed when there is minimal drainage, usually after 2-3 days. Nasogastric feeding is continued for 5-7 days, after which fluids are given. If there are no complications or leakage, the nasogastric tube is removed and soft diet started the next day. Normal diet is given after 10 days.
Recent advances

Debate continues about whether endoscopic or external surgery is the best form of treatment, both having their staunch advocates and, equally, both having some advantages over the other. Endoscopic surgery is rapid and minimizes the effect of an operation in an elderly person, whereas external surgery, while longer, ensures (except with inversion) excision of the sac and relieves the risk of carcinoma developing later. It also has a lower symptomatic recurrence rate. Some surgeons have therefore tried to improve on the existing techniques.

Endoscopic modifications

Knegt, De Jong and Van der Schans (1984) reported on 28 patients on whom they had used carbon dioxide laser to divide the diverticulo-oesophageal septum. Twenty-two patients had complete relief and the remainder were considerably improved. There was one of mediastinitis, which responded to antibiotics, and 21 of the 28 patients had a sharp rise in temperature for the first 24 hours, but were otherwise symptom free. This phenomenon remains unexplained. Van Overbeek, Hoeksema and Edens (1984) used laser surgery in 12 patients, but felt it conferred no advantage to the patient over traditional diathermy techniques.

External approaches

The stapling gun has been in use in bowel surgery for many years but, until recently, otolaryngologists have not taken advantage of this apparatus. It is now becoming increasingly popular for resection of the diverticulum. In the 1980s it has been reported more frequently and offers a real contribution to the safety and ease of surgery, the closure of the sac being most important in preventing infection, emphysema and mediastinitis.

Complications

Complications may disrupt the smooth recovery of the patient. They can be divided into immediate, early and late.

Immediate

(1) Haemorrhage. This usually results from poor haemostasis at surgery, or a ligature slipping from one of the vessels ligated during surgery.

(2) Pneumothorax. While this is uncommon it may occur during the mobilization of a large sac particularly if adhesions are present.

(3) Surgical emphysema. This may result if an unseen mucosal tear is left or the suture line is not complete.
Early

(1) Secondary haemorrhage is usually due to secondary infection.

(2) Hoarseness. Endoscopic surgery avoids the recurrent laryngeal nerve, but in external approaches, there is always the danger of damage to the nerve, either temporary or permanent.

(3) Wound infection or wound abscess. These are more likely if there has been spillage from the sac or oesophagus during surgery, although they may result from a leak through the suture line. Infection will predispose to fistula formation.

(4) Fistula is usually a result of infection. Saliva or food leaking from the external excision are diagnostic of this condition, which may be confirmed by a gastrograffin swallow. Fistulae usually close spontaneously if the pharynx is bypassed with a nasogastric feeding tube.

(5) Mediastinitis may result from a leak tracking downwards. If it is not noticed at surgery the patient develops symptoms several hours or days later, complaining of pain in the neck and back, and becoming severely distressed and dyspnoeic. Plain radiographs of the chest will show air in the mediastinum or neck. If doubt exists, a small quantity of gastrograffin is swallowed and radiographs repeated. Treatment should be rapidly instituted with intravenous antibiotics, and the patient should be monitored for evidence of deterioration in the vital signs. If this occurs the neck should be reopened and any leak closed with sutures, a large drain being inserted into the posterior mediastinum. It may be necessary to perform a thoracotomy for drainage.

(6) Aerocoele. This is rare but has been described by McArthur (1980), as a complication following diverticulectomy. A large aerocoele formed in the superior mediastinum communicating with the pharynx; it resolved spontaneously several weeks after surgery, having discharged briefly via the neck.

Late

(1) Persistent hoarseness occurs when the recurrent laryngeal nerve has been divided. No recovery can be expected.

(2) Stricture results from taking too much mucosa when dividing the neck of the sac, and closing the pharynx. If too much traction is applied to the sac during excision it pulls the pharynx laterally so that inadvertently zealous excision can result. It is treated by dilatation which is generally successful, although repeated applications are often necessary. Occasionally radical surgical correction is necessary.

(3) Recurrence. All methods have a recurrence rate although it is higher for endoscopic diathermy and two-stage diverticulectomy than one-stage diverticulotomy. However, it is easy to reoperate endoscopically, whereas reopening a neck to identify the recurrent sac, with fibrosis and scarring, is difficult. The recurrence rate for endoscopic diathermy is about 6-7% against 2-3% for one-stage diverticulectomy, if the latter is combined with cricopharyngeal
myotomy. If a myotomy is not performed the recurrence rate of diverticulectomy is much higher. Bertelsen and Aasted (1976) using excision of the sac, believed that there is a considerable difference between symptomatic recurrence rate, which they reported at 2% and radiological recurrence which they reported as high as 11%. However, they did not perform cricopharyngeal myotomy. Gammelgaard (1955) reported on 20 patients who had diverticulotomy without myotomy. When reviewed at up to 8 years after surgery 14 had developed radiological recurrence. Complications using external approaches are higher when the sac is larger, being greatest in those patients falling into the stage III group.

**Carcinoma of the diverticulum**

This is a rare problem with only about 30 reported cases in the English literature. The Mayo Clinic, in a series of 1249 patients, found four cases of malignancy arising from the diverticulum, which is an incidence of 0.32%. The true incidence is probably between 0.5-1.0%. It affects men predominantly in a ratio of about 5:1 and usually occurs in a long-standing diverticulum, the average duration of symptoms being greater than 7 years. The age of diagnosis is usually over 50 years. The main predisposing factor is thought to be chronic irritation and inflammation of the diverticular lining from food retention. Symptoms indicating carcinomatous change are an acceleration of dysphagia and weight loss and occasionally blood in the regurgitated food. Nodes or a mass may be found in the neck whereas there are normally few palpable signs.

The usual lesion is an invasive squamous cell carcinoma, but a few cases of carcinoma in situ have been reported in the literature.

Barium studies show a constant filling defect, unlike food debris which moves between films or repeat swallows. It is usually seen in the distal two thirds of the pouch but can easily be missed, the diagnosis frequently being made at surgery when careful examination with the oesophagoscope should be performed.

The treatment best suited to this lesion is uncertain. Radiotherapy alone has always failed to effect a cure. Most commonly a simple diverticulectomy is performed but there are only a few cases reported to be free from disease at 5 years, the longest survival being 8 years (Huang, Unni and Payne, 1984). Following the operation radiotherapy is often given. Some authors advocate a more radical treatment as for a carcinoma of the cervical oesophagus.
Chapter 15: Benign diseases of the neck

A. G. D. Maran

Thyroglossal cysts

Terminology

Since it has not been established whether or not the tract connecting the thyroid gland to the foramen caecum persists as a solid tract, a hollow tube or a duct that becomes obliterated, terms such as 'thyroglossal duct cyst' or 'thyroglossal tract cyst' are best avoided and the general term 'thyroglossal cyst' used.

A sinus is an opening between an internal structure and an epithelial surface whereas a fistula is a connection between two epithelial surfaces. The term 'thyroglossal fistula' is often used but it is erroneous. Only one case of a congenital thyroglossal fistula has been described and its existence is rather tenuous. What most authors mean is a thyroglossal sinus and this is the preferred term. A fistula would suggest an opening between the base of the tongue and the skin surface of the neck and this clearly never occurs.

The discharge from a thyroglossal sinus is secreted mucus not saliva.

Embryology

The branchial arches and pharyngeal pouches develop at the beginning of the fourth week of embryonic life. Among other things the first arch forms the lingual swellings that make up the bulk of the anterior two-thirds of the tongue; it is completed by the tuberculum impar which forms a median eminence behind the lingual swellings and eventually joins them. The posterior third of the tongue is formed by the merging of the ventral portions of the second and third branchial arch.

During the fourth week the thyroid anlage forms an outpouching from the floor of the pharynx between the tuberculum impar and the posterior third of the tongue. It enlarges caudally as a bilobed diverticulum following the descent of the heart and great vessels and grows into the loose parapharyngeal connective tissue. As it moves down it leaves a tract behind.

The hyoid bone develops later and joins from lateral to medial. It is possible, therefore, for the tract to be caught in this, resulting in the tract running through the bone. More commonly, however, the hyoid rotates to achieve its adult position and draws the thyroglossal tract posteriorly and cranially at the inferior edge of the body. Apart from this 'notch' behind the body or partly included in the body, the thyroglossal tract lies ventral to the body and the thyrohyoid membrane.

There is no natural opening of the tract. The tongue and foramen caecum form later than the descent of the thyroid and the blind tract, sometimes found in association with the foramen caecum, is the lingual duct and represents the point of union between the paired anterior and posterior segments that form the tongue base.
The thyroglossal tract normally atrophies and disappears between the fifth and tenth weeks, but the caudal attachment may remain as the pyramidal lobe of the thyroid gland.

**Pathogenesis**

The prevalence of thyroglossal cysts depends on the author and the institution. While many authors claim it is the most common non-neoplastic neck mass and others that it represents 40% of all primary tumours in the neck, it is salutary to note that Sistrunk, whose name is still attached to the operation for removal of the cyst, reported only 31 thyroglossal cysts in a series of 86,000 general paediatric patients.

The sex distribution is equal and the age range is from birth to 70 years with a mean age of 5.5 years. Of about half the published cases, 31.5% were under the age of 10, 20.4% were in the second decade, 13.5% in the third decade and 34.6% were older than 30 years. Ninety per cent lie in the midline and 10% are to one side of the midline; of those, 95% are on the left and 5% are on the right.

If this is a developmental abnormality and thus a congenital lesion, why should almost one in three not present until the patient is over the age of 30 years? The reason is not known but two possibilities are cited. The first suggests that recurrent throat inflammation may stimulate the epithelial remnants of the tract to undergo cystic degeneration. The second possibility is a retention phenomenon. A blocked thyroglossal duct may expand to form a cyst because of an accumulation of secretion. Most proponents of this theory, however, implicate the foramen caecum in the obstruction. The lingual duct has mucous and serous glands and the cyst may be a lingual cyst and not a true thyroglossal cyst. This might account for those cysts found in the tongue above the hyoid.

In the published series, where the position is maintained, 2.1% are intralingual, 24.1% suprahoid, 60.9% thyrohyoid and 12.9% suprasternal. This means that about one in four is above the hyoid and three out of four are below.

A cyst may form on both sides of the hyoid resembling a dumb-bell lesion. Missing part of the dumb-bell may account for some recurrences after a Sistrunk operation. Sometimes multiple cysts are found. These are probably pseudocysts because they have no real lining and are only granulation tissue with extravasated mucus. These probably arise from continual mucus production in a blocked duct that gradually balloons out at its distal end to form a cyst that ruptures into the surrounding tissue.

Sinus openings are always secondary due to spontaneous or surgical drainage after infection. Sinus formation may also occur after a 'lumpectomy' operation leaving the hyoid and part of the tract if it is mucus secreting.

Thyroid tissue is present in the cyst wall in more than 60% of cases. Its presence is most probably due to the origin of the duct in relation to thyroid tissue, but benign thyroid metastases (compare endometriosis) have been postulated. It is from this tissue that thyroid carcinoma can arise.
The epithelial lining is variable; most commonly it is pseudostratified ciliated columnar but it may be squamous. Squamous carcinoma has also been reported in these cysts.

**Clinical features**

The patient will present with either a painless or an infected lump. One death has been reported due to respiratory obstruction from an intralingual cyst. If uninfected, the cyst may be soft, fluctuant and mobile, but very often it is so tense that it seems solid and, in this state, it can be wedged between the hyoid and the thyroid cartilage and thus appear fixed. It is, however, in the midline or just to the left and very few malignant lesions are in this site. Most cysts are prehyoid or infrahyoid and, since all are attached even by a tract to the hyoid, they move both on swallowing and protruding the tongue. Mobility depends on size.

If infected, the lump will be painful, the patient will have odynophagia and the overlying skin will be red. Misguided attempts at abscess drainage here can cause problems when excision is attempted and the recurrence rate is measurably higher.

Sometimes a tract is seen or palpated from the cyst to the hyoid. Such a tract may also be found clinically if there is a sinus.

The differential diagnosis is from a dermoid cyst, an infected lymph node, lipoma, minor salivary gland tumour, sebaceous cyst, cartilaginous tumour of the thyroid, hypertrophic pyramidal lobe and choristomata.

If the cyst is intralingual, it might be a lingual thyroid and, if it is below the thyroid cartilage, it may be a thyroid adenoma. Finally, the possibility of a carcinoma in the thyroglossal remnant must be borne in mind.

It is almost impossible to differentiate a midline dermoid from an uninfected thyroglossal cyst before operation. The content of a dermoid is different. Instead of thick, viscous mucus, dermoids contain cheesy semisolid material. The epithelial lining is keratinizing squamous epithelium with skin appendages on the wall. There is no evidence of a duct nor of inflammation.

Lipomata are more ill defined at the edges, but are tantalizingly fluctuant and so may cause diagnostic difficulty. They also move on swallowing.

Some of these differences may be noted on needle biopsy.

Consideration should be given to ordering a $^{131}$I scan in all suprahyoid and infrathyroid lumps. In 65-75% of patients with a lingual thyroid, there is no other thyroid tissue and it represents a complete failure of the gland to descend. It is three times more common in females because of the increased demands of the thyroid at puberty, pregnancy and menopause. There is not the same necessity to scan patients with lumps in the usual position between the thyroid and front of the hyoid.
**Treatment**

Wenglowski, who performed much of the embryological study on neck cysts, first suggested that, not only the body of the hyoid, but a core of tissue between this and the foramen caecum be removed. Schlange was the first to remove the body of the hyoid, but Sistrunk still has his name applied to the present day operation. He adopted Wenglowski’s suggestion and removed a core of tissue between the hyoid and the foramen caecum.

A horizontal incision is made at the inferior border of the mass and the skin carefully dissected off the mass. Sometimes there is a close connection between the mass and the skin and cyst rupture is a possibility. This makes further dissection difficult and there is then a risk of leaving part of the wall or pseudocysts behind.

The cyst is mobilized and the tract is often found in these cysts lying low in the neck, but in the higher ones there is little point in looking for a tract in and around the hyoid. Sometimes the tract is multiple and in sections its very thin wall is sometimes striking. Furthermore, although it has been described as being in the hyoid bone, van Nostrand’s series showed the tract never to be in the bone.

The body of the hyoid between the lesser horns is divided with shears or Mayo scissors. Medial to the lesser horns there is no danger of damaging the hypoglossal nerves. The genioglossus, mylohyoid and geniohyoid are attached to the body of the hyoid bone and a segment of these muscles is removed in continuity with the bony segment. There is no possibility of identifying a tract in this core of muscle, but failure to remove the core involves the risk of recurrence. The core is removed in a line drawn at 45° to the body of the hyoid aiming at the foramen caecum. There is no need, however, to open into the oral cavity.

**The infected cyst**

It is unwise to operate on an infected cyst. On the other hand, these may be very tense and painful and antibiotic preparation may be poor. Aspiration of pus may improve antibiotic penetration and allow resolution with a view to later removal.

**Treatment of recurrence**

If the hyoid body is not removed, the recurrence rate is 85%. Even if it is removed the reported recurrence rate varies from 2% to 8%. Recurrence is easily understood if the body of the hyoid is not removed, but it is not so easily explained if Sistrunk’s procedure is performed. Possible causes of recurrence are:

1. missing a dumb-bell cyst deep in the back of the hyoid pushing the thyrohyoid membrane back
2. dealing with a cyst that has ruptured to form thin-walled pseudocysts
3. rupturing a cyst and leaving part of the wall behind
4. failing to realize that the tract may be multiple and taking too thin a core.
If Sistrunk's procedure has not been performed then revision surgery is simple and the body of the hyoid removed along with a core. If, however, Sistrunk's procedure has been carried out, further surgery is difficult and the removal of the recurrent cyst plus tract must be improvised.

**Treatment of sinus**

A sinus is due to spontaneous or surgical drainage and is always secondary. Sistrunk's operation is performed, but an ellipse of skin is removed around the sinus.

If the sinus is high in the neck then the scar is a problem. If a healing scar is mobile then hypertrophy will possibly occur. The hyoid area moves constantly during swallowing and the patient should be warned of the scar problem. Dermabrasion, or breaking the line of the scar may be necessary.

*Thyroglossal carcinoma*

Two types of carcinoma have been described - one from the thyroid elements and one from the squamous elements.

The thyroid carcinomata are the more common and over 90 have been reported. Some have been diagnosed before operation for removal of thyroglossal cyst and some have been associated with coexisting thyroid gland cancer and only seven have had metastatic neck nodes. The age range is between 6 and 81 with an average age of 39. The sex incidence is equal. The age and sex distribution is the same as for thyroid carcinoma. Eighty-five per cent are papillary adenocarcinoma and the remaining 15% are follicular adenocarcinoma, adenocarcinoma and squamous carcinoma.

With so few cases reported, no treatment plan has evolved and prognosis cannot be delineated. It does appear, however, that Sistrunk's operation plus suppressant doses of thyroxine offers a reasonable chance of a cure.

**Branchial cysts, sinuses and fistulae**

*Terminology*

It is now thought that branchial cysts, sinuses and fistulae are not all variants of the same thing originating from the branchial apparatus. The original papers linking them with the branchial apparatus did so more by acclamation than experimentation.

The cysts, also known as lateral cervical cysts, usually present in the lateral part of the neck deep to the sternomastoid at the junction of the upper third and lower two-thirds. A few cysts have a definite tract into the area of the posterior pillar of the tonsil but most have no evidence of a tract. There is doubt if these are of branchial origin.

Sinuses, or branchial pits, open along a line between the tragus and the sternoclavicular joint at the anterior border of the sternomastoid. These are almost certainly failure of completion of development of the branchial apparatus. As such, they are present at
birth and reflect failure of development of the first, second, third and fourth arches. Sinuses may also occur with an internal opening only. These, however, may not always come to light but some may produce mucus, block off and become cysts with an identifiable tract.

On rare occasions, the estimated pit is demonstrated clinically and/or radiologically to have an internal opening at the posterior tonsillar pillar. Thus with two openings between epithelial surfaces there is a fistula. No case has been described with an external opening, an inclusion cyst in the tract and an internal opening.

**Embryology**

A 2-week embryo has on each side six branchial arches, five branchial clefts and five pharyngeal pouches. These arrangements are not parallel but tend to come together at the sixth arch. The first and second arches are important, the third and fourth less so, and the fifth and sixth vestigial.

The second pharyngeal pouch forms the palatine tonsils; the second arch grows downwards on its lateral side to meet the fifth arch thus enclosing the second, third and fourth clefts forming the cervical sinus of His. By the sixth week, the branchial apparatus has disappeared having formed the ear, tongue, hyoid, larynx, tonsils and parathyroids.

**Theories of origin**

The debate as to the origin of a branchial cyst reached a climax in the 1920s and 1930s. There are four theories of origin of branchial cysts, but because of the complicated development of the neck none has been proven by embryological investigation. Most of the theories have been an attempt to correlate clinical findings with known embryological facts and none can stand close scrutiny.

**Branchial apparatus theory**

These cysts may represent remains of the pharyngeal pouches or branchial clefts or a fusion of these two elements. When branchial cysts have an internal opening, it is in the region of the tonsillar fossa indicating an origin from the second branchial pouch. Fistulae and sinuses from the second pouch would necessarily pass between the external and internal carotid arteries.

Origin from the third or fourth pouches is unlikely, as they would have to pass over the hypoglossal nerve to reach the skin and would be severed by the upward movement of that nerve during development.

A fourth arch tract would also have to pass below the subclavian artery on the right and the aortic arch on the left.

A third arch tract should have its internal opening in the pyriform fossa and a fourth arch tract below this. These have never been described, so that origin from these pouches can be discounted.
Origin from the first pouch is possible because high branchial cysts have been described lying under the parotid gland with an internal opening between the bony and cartilaginous meatus. If the branchial apparatus theory were to be upheld, a lot more cysts would be expected to have internal openings; it is a popular misconception that many branchial cysts have an internal opening. More cysts would also be expected to be present at birth, but this event has been described only once. The peak age incidence is in the third and fourth decades which is late for a congenital lesion (compare thyroglossal duct cysts).

Cervical sinus theory

This is an extension of the previous theory and considers that branchial cysts represent remains of the cervical sinus of His which is formed by the second arch growing down to meet the fifth. It is unlikely that this is true for those with an internal tract, since this is closed by fusion of its ectodermal lining from within towards the surface. This makes an internal opening difficult to achieve.

Thymopharyngeal duct theory

Cysts may be a remnant of the original connection between the thymus and third branchial pouch from which it originates. The originator of this theory presumed that the hyoid bone constituted the lower level of branchial derivatives. Not only is this false but a persistent thymic duct has never been described. Furthermore, no branchial cyst has even been described deep to the thyroid gland nor have there been any examples of tracts between the pharynx and thymus.

Inclusion theories

King (1949) stated that there was insufficient evidence to show that cysts arose from the branchial apparatus and suggested that the cyst epithelium arose from lymph node epithelium. The following facts support this theory:

(1) most branchial cysts have lymphoid tissue in the wall and are found in the parotid and pharynx as well as in the lateral neck

(2) the peak age incidence is later than expected for a congenital lesion

(3) a branchial cyst in a neonate is almost unknown

(4) most branchial cysts have no internal opening, or at best a tract with an ill-defined termination.

Pathogenesis

Cysts and sinuses are lined by stratified squamous epithelium but, on occasion, by non-ciliated columnar epithelium. The appearance of this latter epithelium probably represents a glandular metaplasia as a result of infection. This could account for the mucus production from sinuses but, if it were the sole cause of cyst formation the cysts would be expected to be filled with thick, viscous mucus like thyroglossal cysts. This is not the case, however,
because the cysts contain straw-coloured fluid containing cholesterol crystals. It is the type of fluid that could only be derived from blood rather than from a mucous gland secretion. If mucus production from metaplastic epithelium was postulated as a cause of the cysts then it is unlikely that the wall would also contain lymphoid tissue, as more than 80% of cysts do. The lymphoid tissue often shows evidence of germinal centres which could only happen if the cyst formed inside a node.

Squamous cysts within lymph nodes are also found in sites far removed from the branchial apparatus, such as the posterior triangle of the neck, the pharynx, the parapharyngeal space and even within the substance of the parotid gland.

Heterotopic salivary, thyroid and squamous epithelium within lymph nodes is well documented. Salivary tissue within lymph nodes can undergo neoplastic change to form a monomorphic adenoma. Thyroid tissue may undergo carcinomatous change in the lateral neck and ectopic thyroid cancer is well recognized. Similarly, therefore, the squamous epithelium can undergo cystic change (as in branchial cysts) or neoplastic change as in branchiogenic carcinoma.

Branchial sinuses and fistulae are present at birth as one would expect for a developmental defect of the branchial apparatus. The peak age incidence for branchial cysts is in the third decade, the range being 1-70.

Clinical features

Sixty per cent are in males and 40% in females. The peak age incidence is in the third decade for cysts; sinuses are noted at birth and no cases are reported of sinuses appearing later in life unless by virtue of spontaneous or surgical drainage, always after infection. Sixty per cent are on the left and 40% on the right. A few are bilateral. Three-quarters are in the classical upper lateral neck position, the remainder being in the lower neck, the parotid, the pharynx and the posterior triangle.

The presenting features are:

- continuous swelling 80%
- intermittent swelling 20%
- pain 30%
- infection 15%
- pressure symptoms 5%

Seventy per cent are cystic on palpation and 30% are firm, but this is probably just a measure of fluid content. Before the widespread use of fine needle aspiration biopsy, some patients had the long work-up for a metastatic node from no identifiable primary. Indeed, the author has removed a branchial cyst after such a work-up in man of 70 who presented with a neck mass and a 6-week history.

It is unknown why some cysts present suddenly as infected masses with overlying skin erythema. The infection might be blood borne, might reflect an internal opening and infection
from the pharynx or it might be a chemical reaction within a squamous-lined cyst in a lymph node.

The differential diagnosis depends on the age of the patient.

In the patient under the age of 10 years, and especially the very young, the neck is relatively smaller than in the adult and division of the lateral neck into thirds is easier on paper than on the patient. In the newborn, a lymphangioma or dermoid must be suspected; a lymphangioma is much softer than a branchial cyst and a dermoid is very firm and tense. A lymphangioma does not have well-demarcated edges while a dermoid and a branchial cyst do. If the child is a little older, rhabdomyosarcoma is a possibility and, if it is tender, it might be lymphadenitis from the tonsil or even the teeth and pharyngeal spaces.

In the patient between the age of 15 and 40 years, the most likely alternative diagnosis is adenitis from viral or bacterial causes, tuberculosis, lymphoma or a nerve sheath tumour.

In the patient above the age of 40 years, a metastatic node from a head and neck primary neoplasm is the prime diagnosis. Alternatives are lymphoma, tuberculosis, lipoma or nerve sheath tumour.

Diagnosis is by clinical examination and from needle aspiration biopsy if necessary. Radiology is not usually helpful but if it pulsates then carotid angiography might be considered.

There is no differential diagnosis for branchial sinuses or fistula. To differentiate between a sinus and a fistula is important because all the tract must be removed to avoid recurrence. A sinogram will give this information.

**Treatment of branchial cysts**

These should be removed if they present as a mass both for diagnosis and cosmetic reasons. If left there is also the danger of infection.

A transverse incision is made in a neck crease and the sternomastoid retracted. When the cyst is mobilized, attempts are made to find a tract. This is not usually possible and very often surgeons who feel they 'ought' to find a tract, manufacture compressed areolar tissue and fascia into one going through the external and internal carotid artery. If a true tract exists it traverses this path to the posterior pillar of the tonsil. Before the operation, careful examination of this area of the fauces under anaesthesia may reveal whether or not there is an internal sinus.

If the cyst presents as an infected mass it should be aspirated and treated with antibiotics. When the infection has settled excision is planned, but sometimes there is no mass to find when the time comes for incision. If this is the case, exploration of the neck should not be performed because it will be impossible to tell which of the many lymph node contains the cyst and which are reactive.
**Treatment of branchial sinus and fistula**

The mouth of the sinus is encompassed in an elliptical incision and the tract, which is often as thick as a medium-sized artery, is found just underneath the skin. It is dissected as high as possible and then another incision is made higher in the neck. Dissection is continued to the tonsillar area where the tract usually disappears.

If a periauricular pit is noted, the surgeon should be well versed in parotid surgery and also ear anatomy, because while the tract often goes up towards the temporal region, it may also go to the junction of the bony and cartilaginous external auditory meatus. The troublesome route, however, is when the tract goes towards the facial nerve. In this instance, the surgeon should stop following the tract into the parotid and should formally identify and dissect the facial nerve.

**Branchiogenic carcinoma**

In the 1940s, many cases of carcinoma in nodes in the neck were reported and the fashion was to ascribe the development of the squamous carcinoma to neoplastic growth of heterotopic squamous epithelium within lymph nodes: hence the name branchiogenic carcinoma.

It became obvious that many of these cases were metastatic deposits in lymph nodes from primary tumours in the head and neck. This was dawn of modern head and neck surgery and in order to discourage the haphazard treatment of metastatic neck nodes, it was decreed that before a branchial carcinoma could be claimed, four postulates had to be attained:

1. the carcinoma should be demonstrated as arising in the wall of a branchial cyst
2. the tumour should occur in a line running from a point just anterior to the tragus along the anterior border of the sternomastoid to the clavicle
3. the histology should be compatible with an origin from the tissue found in the branchial vestigia
4. no other primary should become evident in a 5-year follow-up.

It is patently obvious why virtually no branchiogenic carcinomata were reported over the next 20 years. The first postulate would be a matter of exquisite timing. The second is virtually meaningless. The third merely means squamous carcinoma, but was sufficiently vague as to be menacing, and the fourth is essential.

In spite of the above, several cases of branchiogenic carcinoma have been reported in the last 20 years that undoubtedly are real. There is no doubt that heterotopic squamous epithelium can exist within lymph nodes. It would also be within the scope of normal biological developments for this squamous epithelium to undergo malignant change just as heterotopic tissue anywhere can.
While emphasizing that branchiogenic carcinoma cannot be claimed until the possibility of an undiagnosed primary has been completely excluded, it is a real entity that is underdiagnosed. Perhaps many of the long-term survivors who originally present with a metastatic neck node with a primary tumour that never comes to light, have this tumour.

Treatment is that of a node with no discoverable primary, that is either radical neck dissection and postoperative radiotherapy or excision biopsy and postoperative radiotherapy.

**Neurogenous tumours**

**Terminology**

These tumours arise from the neural crest which differentiates into the Schwann cell and the sympathicoblast; this latter cell gives rise to paraganglionic cells from which arise carotid body tumours, glomus jugulare tumours, glomus vagale tumours and ganglionic cells from which arise benign and malignant gangliomata. The Schwann cell gives rise to the neurilemmoma (schwannoma) and the neurofibroma.

Neural crest:
- Schwann cell
- Neurofibroma
- Neurilemmoma
- Sympathicoblast - Paraganglionic cells
  - Carotid body tumours
  - Glomus vagale
  - Glomus jugulare
  - Glomus tympanicum.

The thin outer sheet of nerve is called the neurilemmoma and the inner sheath of Schwann is the neurolemma. Tumours arising from the inner nerve sheath are often called neurilemmomata which is incorrect, as is neuronoma. The preferred term is 'schwannoma' for tumours arising from nerve sheath. A schwannoma shows well-developed cylindrical bands of Schwann's cells and delicate connective tissue fibres with a tendency towards pallisading of the nuclei about a central mass of cytoplasm (Verocay bodies). This form is known as Antoni type A tissue, whereas Antoni type B tissue is a loosely arranged stroma in which the fibres and cells form no distinctive pattern. The two types may also be mixed. It has no clinical significance, no surgical significance and no prognostic significance and is only mentioned because it forms part of the pathological catechism of these tumours.

Neurofibromata are often seen in association with von Recklinghausen's disease if they are multiple, but they need not be multiple and can exist as discrete entities. It is suggested that they arise from a disseminated neuroblastoma or aberrantly migrating neural crest cells. It is said that they are not as encapsulated as schwannomata and have an 8-10% chance of becoming malignant. Histologically they are characterized by loose structure, abundant matrix, stout bundles of collagen fibres and spindle-shaped cells sometimes with waveform nuclei.

The difference between neurofibroma and schwannoma is histological, but it is not as clear cut as it might seem to the non-pathologist. A recent study by Horak et al (1983) has
shown that these histological pictures can be mixed in the one tumour. From a prognostic viewpoint, these authors also show that the important difference is in the degree of cellularity rather than the classification.

Both of these tumours can show a plexiform pattern of growth. This applies especially to the cellular ones rather than to those showing the typical histological picture. The typical growth pattern is a lump that either arises from the sheath and grows outwards leaving the trunk of the nerve intact, but more often the growth involves the nerve trunk and fibres can be splayed around the tumour with apparently normal clinical function. In plexiform growth, the abnormal nerve tissue grows into adjacent tissue planes and is rather like a neural lymphangioma. It is difficult to remove and is liable to recur. These are known as plexiform neuromata.

Gangliomata or ganglioneuromata are very rare. They usually arise from a cervical sympathetic ganglion and are firm, smooth and well encapsulated. Microscopically they contain ganglion cells and neurites.

Postoperative neuromata are the result of uncontrolled growth of axons from the proximal stump of a nerve that has been cut. These are not true tumours, but represent attempts by the damaged nerve to repair itself; the axon cylinders become enmeshed in Schwann's cells and scar tissue. If the process becomes hyperactive, the neuroma becomes clinically obvious and the patient experiences localized pain and tenderness.

Carotid body tumours were first termed 'chemodectomata'. This term has now lost favour because carotid body tumours appear to arise from paraganglionic cells rather than chemoreceptor cells. Paraganglionic cells are epithelioid in appearance, are derived from the neural crest and migrate in close association with autonomic ganglionic cells. They are located chiefly along the aorta and great vessels with the largest accumulation in the adrenal medulla where they are chromaffin positive producing catecholamines, adrenaline and noradrenaline and may give rise to phaeochromocytoma.

Formerly, tumours of the extra-adrenal chemoreceptor system were described as non-chromaffin paragangliomata, but in recent years catecholamines and secretory granules like the adrenal medulla have been found in the carotid body. Functioning tumours producing hypertension have been reported in the carotid body and the jugular body. Phaeochromocytoma have been described in association with carotid body tumours.

Paragangliomata have been reported in the following sites - aortic bodies, superior vagal ganglion (glomus jugulare), auricular branch of the vagus (glomus tympanicum), inferior vagal nodose ganglion (glomus vagale), superior laryngeal nerve (glomus laryngicum), mandible (alveolar body), ciliary ganglion (ciliary body), bifurcation of the pulmonary artery, pleura, femoral artery, retroperitoneal tissue, mesentery, coccyx and pineal body.

**Pathogenesis**

The growth pattern of nerve sheath tumours is either fusiform or plexiform as outlined in the previous section. Nerve paralysis on presentation is very rare, even though the nerve is found to be grossly distorted at surgery with the fibres widely stretched over the tumour.
The histology has also been described and the fact highlighted, that features of neurofibromata and schwannoma can exist in the one tumour. These tumours both arise from the Schwann cell. On occasion, peripheral nerve tumours may be difficult to distinguish from other spindle cell mesenchymal lesions. The neural crest marker antigen, S100, is common to the supporting cells of the peripheral and central nervous system. Immunocytochemical staining using antibody to S100 is positive in the majority of tumours of Schwann cell derivation, although expression of the antigen is reduced in Antoni A or malignant areas. The use of antibody to neuron specific enolase may help to distinguish neurofibromata although the method is less tissue specific.

They arise from any cranial or spinal nerve that has a sheath and this means any motor or sensory nerve other than the optic or olfactory. In the head and neck (apart from the acoustic nerve), the vagus is affected more commonly than any other nerve, but nerve sheath tumours have been described on the hypoglossal nerve, the facial nerve, the spinal accessory, the sympathetic chain, the glossopharyngeal and branches of the cervical plexus.

While multiple neurofibromata can occur in association with von Recklinghausen's disease, little is known of the aetiology of other nerve tumours. In a long-term follow-up of over 2000 patients who had been irradiated for tonsil and adenoid enlargement, Shore-Freeman et al (1983) found 29 schwannomata, two neurofibromata and one ganglion neuroma. These tumours can, therefore, be radiation induced and they can continue to occur for at least 30 years after the radiation exposure. They are more common in women than in men. Most cases present in the 30-50 year age group. Malignant change in nerve sheath tumours in the head and neck is very rare. Das Gupta et al (1969) described only one sarcomatous change in a series of 303 solitary nerve tumours. It is said that malignant change occurs in 10-15% of patients with multiple neurofibromatosis. The only thing that differentiates a neurogenous sarcoma from a fibrosarcoma is its origin from a nerve trunk. Malignant change is suggested by rapid growth, pain and paraesthesia. They do not metastasize to regional lymph nodes but do to the lungs.

The microscopic appearance of carotid body tumours does not relate to the future behaviour of the tumour. They are poorly encapsulated and extremely vascular, like a haemangiomia, with an enveloping network of capillaries and areolar tissue arranged in concentric circles similar to an onion. They encroach upon and gradually surround the carotid bulb, invade it and extend along the carotid artery for long distances drawing blood from the vasa vasorum. The carotid system becomes progressively distorted and extended with the internal carotid artery being attenuated but never occluded. Adjacent cranial nerves are encased as may be muscles and even the base of the skull.

Glomus tumours of the vagus grow in the same manner and totally distort anatomy at the skull base with bleeding tissue making dissection extremely difficult.

Less than 10% are frankly malignant with regional and distant metastases to lung and bone.

Due to the fact that chronic hypoxia at high altitudes leads to carotid body hyperplasia, there is a high incidence of carotid body tumour in Peru where the majority of population live
at altitudes of 2000-5000 metres. The average age of presentation ranges from 35 to 50 years, the youngest reported being 12 years old. The sex incidence is equal.

There is a striking family history of up to 10% and also a tendency to bilateral tumours, tumours of other similar cells and phaeochromocytoma. Twenty-five per cent are bilateral in those with a positive family history, compared with 3% in those with no family history.

**Clinical features**

Nerve sheath tumours of the vagus present as parapharyngeal space masses. They are nearly all fusiform and so do not expand out of the parapharyngeal space to cause a discrete lump at the angle of the jaw, but usually push the sternomastoid laterally. This gives an ill-defined neck mass deep to the muscle which may, however, have an anterior border but certainly no palpable superior border. Since the vagus lies deep in the parapharyngeal space it pushes the pharynx medially. Tumours of the deep lobe of the parotid push the tonsil and soft palate medially but vagal and carotid tumours push the posterior pillar and posterolateral oropharyngeal wall forward. They are painless masses that rarely cause nerve paralysis.

Tumours of the sympathetic trunk present in the same way, but tumours of the hypoglossal, accessory and cutaneous cervical nerves merely present as neck lumps in the appropriate area. Facial nerve tumours are invariably diagnosed as parotid tumours.

A postoperative neuroma is very tender and, on palpation, the patient experiences an electric shock type of sensation. There must be a previous history of neck surgery, such as a parotidectomy, or a radical neck dissection. The nerves most usually affected are the cutaneous branches of the cervical plexus, that is the great auricular, the lesser occipital and the anterior cutaneous nerve of the neck. They rarely form on a stump of the hypoglossal nerve after a XII-VII anastomosis.

Carotid body tumours present in the same space, but it is as usual for them to form a discrete neck mass as it is for them to present as a parapharyngeal mass pushing the pharyngeal wall medially. Noradrenaline-secreting tumours have been described which had properties similar to phaeochromocytoma. There is also an association with the neural crest lesions and the syndrome of multiple endocrine adenomatosis should be suspected. Cranial nerve paralysis is more common in carotid body tumours than in nerve sheath tumours.

Glomus vagale tumours may or may not present with vagal nerve paralysis but all present with a neck mass high under the sternomastoid.

On clinical examination, it is impossible to differentiate these tumours if they present, as they usually do, in the parapharyngeal space. Old clinical aids, such as moving masses side to side but not up and down, are fairly useless because any lesion attached to a structure that run vertically in the neck like a vagus nerve, a carotid artery or the sternomastoid muscle will behave in such a fashion. Pharyngeal presentation will be the same for carotid or vagus masses, namely behind the posterior tonsillar pillar. Carotid body tumours are not pulsatile.
A computerized tomography (CT) scan will show if there is a mass in the parapharyngeal space and this will be very useful for smaller tumours that only give a suspicion of a lump clinically. A CT scan will not, however, always differentiate between a nerve sheath tumour and a carotid body tumour. Angiography is nearly always advisable if there is real suspicion of a carotid body tumour. This is not only diagnostic but gives an indication of the resectability of the tumour.

Fine needle aspiration biopsy is always worth carrying out but open biopsy should be avoided. Although much less frequent now, there is ample scope for surgical disaster by exploring these tumours unprepared for major vascular surgery. The disaster potential can be maximized by an approach through the mouth.

**Treatment of nerve sheath tumours**

There are three approaches to the parapharyngeal space:

1. transparotid
2. transmandibular
3. transcervical.

The transcervical route is best employed for these tumours. The sternomastoid muscle is retracted backwards, the tail of the submandibular gland swung forwards and the tail of the parotid gland lifted upwards. Adequate access to the skull base can be achieved by this route. The author's experience is that it has not been possible to remove nerve sheath tumours from this space, keeping the nerve intact. The first step is to make sure it is not a carotid body tumour or a glomus vagale. Once this is done, the nerve is dissected off the artery and excised with the tumour. The vocal cord paralysis is subsequently rehabilitated by a Teflon injection or a laryngoplasty.

**Treatment of carotid body tumours**

Carotid body tumours should be removed if technically possible. If the tumour is not removed, then the patient is left with a progressively enlarging neck tumour of uncertain biological behaviour. The factor that limits resection is extension to the skull base. Unless there is an inch (2.5 cm) or so of carotid artery free of tumour at the skull base, it is impossible to attach a graft and resection should be abandoned. Usually it is possible to dissect these tumours from the carotid after freeing its whole length and rotating it. This is to allow access to the posterior aspect where a plane can often be entered. At all of these operations, however, there should exist the facility for immediate vascular grafting if required.

Radiation should not be used as a primary treatment. It should be reserved for poor risk patients, inoperable patients, malignant tumours and those who refuse surgery.
Lymphangiomata

Terminology

There are three types of lymphangioma:

(1) lymphangioma simplex
(2) cavernous lymphangioma
(3) cystic hygroma.

Embryology

The lymph system arises from five primitive sacs (two jugular sacs, two posterior sciatic sacs and a single retroperitoneal sac) developed from the venous system. Endothelial buds from these extend centrifugally to form the peripheral lymphatic system.

There are two theories of the origin of lymphangiomata: either they are sequestrations of lymphatic tissue derived from portions of the primitive sacs, which retain their rapid and proliferative growth potential and have no connection to the normal lymph system, or they arise from endothelial fibrillar membranes which sprout from the walls of the cyst, penetrate surrounding tissue, canalize and produce more cysts.

Pathology

Lymphogenous conditions have been classified into three groups:

(1) lymphangioma simplex - composed of thin-walled capillary-sized lymphatic channels

(2) cavernous lymphangioma - composed of dilated lymphatic spaces often with fibrous adventitia

(3) cystic hygroma - composed of cysts varying in size from a few millimetres to several centimetres in diameter.

All these can be regarded as one entity, but site may play some part in the final version - the smaller lymphangiomata occur in the lips, tongue, cheek and where the tissue planes are tighter, whereas the cystic hygroma has more space to expand into the tissue planes of the neck. Simple lymphangiomata can occur anywhere in the mouth as pale soft fluctuant lesions and form one-third of all lymphangiomatous tumours. More common are cavernous lymphangiomata which form 40% of these lesions, mainly in the tongue. At the base of the tongue they must be differentiated from a lingual thyroid, a lingual carcinoma or an internal laryngocoele. They also occur on the lateral border. Some cheek lesions reach an enormous size and are very difficult to eradicate since total excision produces an unacceptable cosmetic defect.

A cavernous lymphangioma of the floor of the mouth can be part of a cystic hygroma or a ranula. Macrocheilia usually affects only the upper lip.
Cystic hygroma consists of large multinodular cystic masses which may communicate or be isolated. The walls are thin and the contained fluid can be clearly seen. The walls are thin and the contained fluid can be clearly seen. A hygroma occurs in the cervicofacial region spreading into the cheek, mouth, tongue, parotid and even the ear canal.

Histologically the cyst is lined by a single layer of flattened endothelium with fetal fat and cholesterol crystals. They are rare tumours forming 0.5% of large series of neck lumps. There is no sex or side predominance. Two out of three are noted at birth and nine out of 10 before the end of the second year.

Thirty-five per cent of lymphangiomata of all types occur in the cheek, tongue and floor of the mouth, 25% in the neck and 15% in the axilla.

**Clinical features**

Most of these tumours manifest themselves at birth or shortly afterwards. Lymphangiomata in the mouth can first appear in adult life, as can recurrences of cystic hygromata after surgery in infancy. Recurrences usually occur on the periphery of the facial area where the main mass originally presented, such as the ear, parotid or posterior triangle.

While size alone is the prominent first symptom and sign, if the cyst is big enough it can cause stridor. In very large cysts, a lateral displacement of the trachea and even mediastinal widening may be seen on the radiograph.

Sudden increase in size by spontaneous haemorrhage may be fatal. Brachial plexus compression with pain and hypaesthesia may also occur.

The most common site is in the posterior triangle of the neck. Large masses can extend into the anterior triangle and across the midline. These anterior tumours may involve the floor of the mouth and the base of the tongue. Cystic hygromata often extend up into the cheek and parotid gland or down into the mediastinum or axilla.

If it is not the gross congenital swelling characteristically seen at birth, it can be discovered as a painless soft or semifirm swelling in the neck. The tumour usually progressively enlarges, although some fluctuation in size is common. Depending on its mass and direction of growth, the lesion can encroach on the trachea, pharynx and oesophagus causing dyspnoea and dysphagia. A sudden increase in size of the tumour may be secondary to infection or haemorrhage and has caused death. Tumour swelling is usually related to upper respiratory tract infection and tumour pain, which is an unusual complaint, occurs only in the presence of infection. One case of facial nerve paralysis due to enlargement of the hygroma has been recorded.

The diagnosis is usually made on clinical grounds. Prior to the CT era, the radiologist could contribute little. All he could identify would be displacement of the trachea or oesophagus or mediastinal extension but the size could not be imaged. Injection of radiopaque material into the cyst is unrewarding and angiography only showed an avascular mass lesion. Computed tomography can, however, show the extent and size of the lesion and also it relation to the important structures at the skull base.
Treatment

The treatment of lymphangiomata is surgical. Intraoral lymphangiomata should be removed from an external approach since they are almost certainly much more extensive than expected. Remnants will also be left in the tissue planes of the tongue. Lymphangiomata of the base of the tongue can often be dealt with by coagulation diathermy, repeated if necessary. Cryosurgery may also be helpful. Lymphangioma of the upper lip should be dealt with by a lip shave and vermilion advancement and excision of muscle and cyst to reach an acceptable size.

Recurrences usually appear within the first 9 months in about 10-15% of patients. The recurrence rate is higher with cavernous lymphangioma than with cystic hygroma.

No patient with cystic hygroma should have surgery unless he has had a chest X-ray to check for mediastinal extension. Since the child at birth looks like a monster to the mother, surgery should not be delayed. Total removal is impossible, but surgery is easiest at the first attempt, before there is infection and scarring, so as much as possible is removed. It is essential not to damage the child and so the carotid arteries, jugular vein, vagus nerve and facial nerve, if necessary, are dissected with great care. The first excision should be limited to the cervical area. After removal it will become apparent that there is a large amount of the tumour left in the parotid and cheek. Excision of this should be delayed as long as possible, taking into account the effect of an asymmetrical face on a child's psyche. The longer one waits, the bigger the facial nerve branches become and the safety of dissection increases. In the initial excision the nerves most likely to be damaged are the lower branch of the facial nerve, the spinal accessory and the vagus.

Radiotherapy should not be used because of the possible damaging effects on the growth of local structures and the potential induction of malignancy.

Injection of sclerosants has been suggested and tried, but the scarring is unpredictable because of the multiplicity of cysts and further surgery may be made very difficult.

Repeated aspirations should only be performed in the event of rapid increase in size causing pressure effects. The danger is infection and possibly haemorrhage into the cysts.

Broomhead (1964) claimed that 15% of cases undergo spontaneous regression. This view has not met with universal agreement.

After surgery, about one-third of patients will have nerve paralysis and more than half will have to undergo further surgery.
Dermoid cysts

Pathology

In the head and neck there are three varieties of dermoid cyst.

Epidermoid cyst

The epidermoid cyst has no adnexal structures, is lined by squamous epithelium and may contain cheesy keratinous material. This is the most common variety.

True dermoid cyst

The true dermoid cyst is lined by squamous epithelium and contains skin appendages such as hair, hair follicles, sebaceous glands and sweat glands. These are either congenital or acquired.

The congenital type derives from ectodermal differentiations of multipotential cells pinched off at the time of closure of the anterior neuropore. It occurs, therefore, along the lines of fusion.

The acquired type is due to implantation of epidermis at the time of a puncture type of injury and is often solid with areas of cystic spaces containing sebaceous material.

Teratoid cyst

The teratoid cyst can be lined with squamous or respiratory epithelium and contains elements formed from ectoderm, endoderm and mesoderm - nails, teeth, brain, glands, etc. This is the rarest variety in the neck and is nearly always diagnosed in the first year of life. Less than 10 cases of teratoid tumours of the neck have been described in adults.

Twenty per cent of all dermoid cysts are found in the neck, and 30% in the neck and face. Dermoids form 28% of all midline cysts, and there is no sex predominance.

Clinical features

These cysts present as solid or cystic masses in the midline of the neck between the suprasternal notch and the submental region. They can also occur lateral to the submandibular gland. Painless swelling is the only symptom, but if the cyst is large, minor obstructive symptoms can occur.

About 20% of dermoids occur in the mouth, either deep to the mylohyoid (sublingual) or superficial to it (submental). They present in the second and third decades but have probably been present since birth.
Treatment

Complete excision is usually easy and should be carried out in all cases.

Infective neck masses

Tuberculosis

Pathology

The condition is not common in USA or Europe, but it is still common in Asia or Africa. There are 32,000 new cases of tuberculosis in the USA each year and 5% of these (1,600) develop cervical lymphadenitis.

Where the incidence of tuberculosis is low, primary infections are acquired later and so it is young adults who acquire tuberculosis nodes. In the UK, the maximum age incidence is 5-9 years, but in 30% it occurs in patients over the age of 25 years.

The bacillus, which is usually the human variety, reaches the lymph nodes by direct drainage or by haematogenous spread. The incidence of coexisting pulmonary tuberculosis is less than 5%. In one series, almost half of the tonsils removed showed evidence of tuberculosis and it thus appeared that the tonsil was the source and that the cervical adenitis was precipitated by an attack of acute tonsillitis. Once the bacillus has entered the host, further exposure is not necessary to trigger off the adenopathy.

Clinical features

Most patients give a fairly long history and usually seek medical advice because the lumps have become painful. In Asia, the presentation is different: 20% have discharging sinuses, 10% a cold abscess and 10% are adherent to the skin; in these patients all have a negative chest X-ray.

Ninety per cent are unilateral and 90% involve only one node group, the most common being the deep jugular chain followed by the nodes in the submandibular region and then those of the posterior triangle.

Diagnosis is by a positive tuberculin skin test and demonstration of acid and alkali fast bacteria in the biopsy. In the USA, patients should also have histoplasmin and coccidioidin skin tests. The differential diagnosis is between lymphoma and metastatic cancer. The absence of a primary tumour in a young adult and the length of history usually makes the latter diagnosis improbable.

Treatment

The treatment is an excisional biopsy followed by 9-12 months of antitubercular chemotherapy. If the glands are very large and matted, local removal is dangerous since the nodes are often attached to the internal jugular vein. A functional neck dissection should then be carried out preserving the sternomastoid, accessory nerve, and jugular vein if possible. In
a child, it is usually wise to remove and histologically examine the tonsils before removing the lymph nodes.

If removal is not followed immediately by chemotherapy, a sinus forms with persistent drainage and, later, ugly scars.

**Sarcoidosis**

Sarcoidosis presenting as cervical adenitis with no other manifestation of the disease is extremely rare. The neck nodes are not often involved in this condition, even when it is generalized. It is almost impossible, therefore, to make a preoperative diagnosis and a biopsy is always needed. The histological characteristic is the absence of caseation. Diagnosis may be confirmed by the Kveim skin test.

**Neck space infections**

Neck space infections are very rare and there is confusion about how many neck spaces there are: estimates vary from 13 to 20. A fascial space is an area of loose connective tissue bounded by dense connective tissue called fascia. It is a matter of opinion how thick connective tissue must be before it is called fascia, and this is where the disagreement as to the number of spaces arises. Knowledge of the anatomy of the areas in which infection tended to collect was important in the pre-antibiotic days from the point of view of routes of spread, complications, and surgical drainage, but nowadays knowledge of three spaces (retropharyngeal, lateral pharyngeal and submandibular) will allow management of 90% of patients.

**Anatomy**

The spaces listed are described by Hollinshead and are shown in Table 15.1.

Nowadays, abscesses usually only occur in the retropharyngeal, lateral pharyngeal (parapharyngeal) and submandibular spaces.

**Retropharyngeal space**

This space lies between the pharynx and the posterior layer of the deep fascia which bounds the prevertebral space. It separates the pharynx from the vertebral column and extends from the base of the skull to the posterior mediastinum as far as the bifurcation of the trachea. Anteriorly, it connects with the pretracheal space so that infections can spread by way of this latter space to the anterior mediastinum. But mediastinitis due to a retropharyngeal abscess is rare. In the infant this space contains one or two lymph nodes.

**Lateral pharyngeal space**

This is more commonly known as the parapharyngeal space; it lies lateral to the pharynx connecting with the retropharyngeal space posteriorly. Laterally, it is bounded by the lateral pterygoid muscles and the sheath of the parotid gland. It extends from the base of the skull to the level of the hyoid bone where it is limited by the sheath of the submandibular
gland. This sheath is also connected to the sheaths of the stylohyoid muscle and the posterior belly of the digastric muscle.

The carotid sheath is bounded anterosuperiorly by the pterygomandibular raphe and the spaces around the floor of the mouth anteroinferiorly.

This space is prone to infection because of its close connection to the tongue, teeth, parotid, submandibular gland and tonsils.

**Submandibular space**

This is bounded above by the mucous membrane of the floor of the mouth and tongue and below by the deep fascia that extends from the hyoid to the mandible.

It is divided into two by the mylohyoid muscle and so the submandibular gland, which is wrapped around the mylohyoid muscle, extends into both parts of the space. The space superior to the mylohyoid muscle contains most of the sublingual gland. The space inferior to the muscle contains the submandibular gland. Anteriorly lies the submental space between the two anterior bellies of digastric.

Infections of this space are known as Ludwig’s angina.

**Table 15.1 Anatomy of neck spaces**

Below the hyoid
- carotid sheath
- pretracheal space
- retrovisceral space
- visceral space
- prevertebral space

Above the hyoid
- mandibular space
- submaxillary space
- masticator space
- parotid space

Peripharyngeal area
- retropharyngeal space
- lateral pharyngeal (parapharyngeal) space
- submandibular space

Intrapharyngeal area
- paratonsillar space.
Clinical features and management

Retropharyngeal abscess

This abscess in infants is due to a lymphadenitis secondary to an upper respiratory tract infection. The child has a sore throat; examination shows a swelling behind an otherwise normal tonsil. The temperature is elevated to 38-39°C (101-102°F) and the child is ill. The swelling may obstruct the posterior nares and push the soft palate down. Respiratory obstruction is an ever-present danger because the child's spine is short and his larynx is high. (In a 9-month-old infant, the epiglottis is at the level of the atlas.)

Radiographs of the neck show a large retropharyngeal swelling. Treatment is by incision and drainage in the tonsil position.

In an adult, a retropharyngeal abscess usually signifies a tuberculous infection of the cervical spine. It is of insidious onset with a low grade fever. Pus must be obtained to confirm the diagnosis which is also suggested on a radiograph of the cervical spine. Treatment is by antituberculous chemotherapy.

Parapharyngeal abscess

This is more common in adults than children. It is a complication of tonsillectomy or tonsillitis in about 60% of patients and a complication of infection or extraction of the lower third molar in a further 30%.

Infection of the petrous apex can rarely rupture directly into the space. Infection of the mastoid tip can also enter the space by way of the digastric sheath.

There is fever and marked trismus because of involvement of the medial pterygoid muscle. The tonsil is pushed medially but looks normal. The most marked swelling is in the neck at the posterior part of the middle third of the sternomastoid. Each patient should be given at least 48 hours' treatment with an antibiotic, but by this time most patients have a swollen neck and incision and drainage will be required.

Ludwig's angina

This is a rapidly swelling cellulitis of the floor of the mouth and submandibular space secondary to soft tissue infection, tonsillar infection and infection of the lower premolar and molar teeth. Over 80% of patients have dental disease and, in these patients, the lower molars are set eccentrically with the roots closer to the inner than the outer side of the jaw, or the roots of the second and third molars may lie inferior to the mylohyoid line. Root abscesses of these teeth, therefore, drain into the submandibular space. This space may be affected with minimal discomfort from the tooth; pain comes from tension within the bone, but if this gives way and drains there is no dental pain. In cases of dental origin the most usual organisms are Streptococcus viridans and Escherichia coli.

When infection spreads to the sublingual space, the floor of the mouth becomes very swollen and appears as a roll of oedematous tissue rising to the level of the biting edge of the
teeth. The tongue is elevated posterosuperiorly and respiratory obstruction is a danger. The patient is very ill with a temperature of over 38.3°C (101°F) with pain, trismus and salivation.

Treatment is by antibiotics; incision and drainage should be postponed as long as possible.

**Laryngocoele**

**Pathology**

In the UK, the incidence of laryngocoele is approximately one per 2.5 million population per year. The sex incidence is 5:1 in favour of men and the peak age incidence is at 50-60 years. Only one case has been reported in a neonate, but it is possible that this was a laryngeal cyst; 825 were in Caucasians; 85% were unilateral and 15% bilateral. They can be external (30%), where the sac arises from the laryngeal ventricle and expands into the neck through the thyrohyoid membrane, internal (20%) where it arises from the laryngeal ventricle and stays within the larynx presenting in the vallecula, or combined (50%). Laryngocoeles are lined by columnar ciliated epithelium, whereas simple laryngeal cysts are lined by squamous epithelium.

It has long been held that laryngocoeles are due to 'blowing' hobbies or jobs such as trumpet playing or glass blowing. A careful review of the published cases reveals at most four patients subject to these habits so that this theory appears to be untrue. Of more importance is the coexistence of a carcinoma or papilloma of the larynx which acts as a valve allowing air under pressure into the ventricle. External laryngocoeles can be found in 16% of laryngectomy specimens in laryngeal carcinoma, as opposed to 2% in laryngectomy specimens for pyriform sinus cancer. The ventricle in these cases of laryngeal cancer was also significantly higher than that in patients with pharyngeal cancer due to increased air pressure consequent upon obstruction by the laryngeal carcinoma.

Lower animals have air sacs, for example the cheek pouch of monkeys, the fish pouch of pelicans, the tracheal sacs of emus, the syrinx of male quacking birds, etc. Lateral laryngeal sacs are well developed in certain anthropoid apes and are a means of enabling the animal to rebreathe while holding its breath for long periods.

Laryngocoeles in man, therefore, are almost certainly atavistic remnants corresponding to these lateral air sacs. On occasion they become manifest due to an increase in intralaryngeal air pressure due to blowing or coughing.

**Clinical features**

The most common presenting features are hoarseness and a swelling in the neck. The third most common symptom is stridor, which can come on very suddenly over a period of a few days or even hours in a patient who had previously only mild symptoms for months or years. Other presenting symptoms are dysphagia, sore throat, snoring, pain or cough. Ten per cent present with infected sacs (pyocelees) and, because of the mixture of infection and air on the radiograph, a diagnosis of gas gangrene is sometimes made. On palpation, the swelling which is usually large and over the thyrohyoid membrane, can be emptied easily. A
plain radiograph of the neck is diagnostic showing an air-filled sac. Nothing else except gas gangrene can produce this picture.

The most common presenting symptom of laryngocoeles is hoarseness with apparently normal vocal cords. To diagnose smaller laryngocoeles, therefore, every patient with hoarseness and normal vocal cords should have a plain anteroposterior neck X-ray with and without a Valsalva manoeuvre. Especially if a unilateral laryngocoele is present, the patient should have a laryngoscopy to make sure there is no underlying carcinoma of the laryngeal ventricle. These tumours can act as valves allowing air into, but not out of, the saccule thus dilating it.

If an enlarged saccule does not penetrate the thyrohyoid membrane, it travels up behind the thyrohyoid membrane and the hyoid and bulges into the vallecula. This is an internal laryngocoele and, if the mouth of the sac is blocked, then it may be full of mucus and not radiolucent. These are often diagnosed during routine otolaryngological examination and may be quite symptomless. On occasion they can reach a reasonably large size and the patient complains of a feeling of a lump in the throat.

**Treatment**

Laryngocoeles discovered radiologically and that are contained within the larynx, require no treatment. The patients have the cause of the hoarseness explained to them and are kept under annual review. If surgery is attempted on these patients, the uninflated saccule is almost impossible to find and oversew.

Internal laryngocoeles may be uncapped to see if marsupialization and scarring will stop recurrence. If they recur then they should be excised with the approach used for the external laryngocoele.

An infected laryngocoele should be aspirated and treated with antibiotics. When the infection has subsided, formal excision should be carried out.

The best operation for a laryngocoele aims at excising the saccule at its neck. This is accessed by removing the upper half of one thyroid ala, or fracturing it downwards and replacing it. The method used depends on the state of ossification of the thyroid cartilage. Once access to the supraglottis is obtained, it is an easy matter to follow the neck of the laryngocoele down as far as possible. The neck is transected and closed in layers and oversewn. If the thyroid ala is not replaced then the thyroid perichondrium is sewn into the area.

Recurrence after this operation is extremely rare.
Chapter 16: Thyroid neoplasms

O. Shaheen

Incidence

Malignant tumours of the thyroid gland represent less than 0.5% of all cancers in England and Wales (Young and Addison, 1983). In southern Sweden there are roughly two cases per 100,000 population per annum (Tennval, 1984), and in the USA the equivalent figure is slightly less than four (Third National Cancer Survey, 1971).

It is predominantly a disease of females, the female: male ratio being about 2.5:1.

Aetiology

Radiation

After reviewing all the available evidence, there is little doubt that radiation stands out as the most definite of all possible aetiological factors.

The practice of irradiating the thymus was fashionable in the earlier part of the century, particularly in America, and for a time irradiation of the tubotonsillar lymphoid tissue also enjoyed a passing vogue in the years leading up to and including the Second World War.

After reviewing the histories of such patients, it was concluded that radiation to the head, neck, and thorax in small doses during childhood was liable to induce cancer of the thyroid gland in later life (Winship and Rosvoll, 1961).

Further evidence of the carcinogenic effect of radiation during childhood came from the survivors of the atomic bombs in Japan, 18% of whom developed cancers of the thyroid gland. Most patients were under the age of 10 at the time of exposure, implying that susceptibility to the disease is most pronounced before puberty; furthermore most tumours were of the papillary type, the most common of the differentiated neoplasms (Sampson et al, 1969).

Although the adolescent and adult gland is evidently less at risk, reports of cancers in adults who had previously been irradiated for Hodgkin's disease of the cervical lymph nodes 10-15 years before clearly indicate a continued susceptibility, albeit on a somewhat reduced scale (McDougall et al, 1980). There is also some evidence to suggest that benign nodular disease may in some instances be linked to a history of previous radiation, possibly as much as 35 years previously (Favus et al, 1976).

On the experimental level, Doniach (1953) demonstrated that rats fed on thiouracil and subsequently treated by radiotherapy developed thyroid cancer. This led him to postulate that two factors were necessary for carcinogenesis, namely a high level of thyroid stimulating hormone (TSH), which one would normally expect in the prepubertal subject, and secondly ionizing radiation. The finding that most differentiated thyroid cancers regress if TSH is suppressed by the administration of thyroxine, supports this contention (Crile, 1966).
Hormonal causes

Experimental work on rats has shown that excessive production of TSH over prolonged periods, whether induced by the administration of thiouracil, by partial thyroidectomy, or by iodine-deficient diets, may induce benign and malignant tumours (Doniach and Williams, 1962). The addition of a specific carcinogen such as 2-acetylaminofluorene increases the incidence of cancer (Bielschowsky, 1944).

One should therefore expect a higher incidence of cancer in glands hypertrophied by high levels of TSH over prolonged periods of time, such as in areas where goitre is endemic (Wegelin, 1928). The evidence in support of this is, however, contradictory and the issue unlikely to be resolved by comparing the statistics of one country with those of another. In favour of the view that endemic goitre predisposes to carcinoma is the finding that the predominant pathological type in such areas is the follicular neoplasm, whereas for non-endemic goitre it is more often papillary (Wohner et al, 1966).

Genetic factors

Familial medullary carcinoma, as opposed to the sporadic disease, is passed on as an autosomal dominant form of inheritance, with an increased predisposition to other tumours, mainly of neuroectodermal origin, such as phaeochromocytoma, epithelial neuroma, and parathyroid adenoma.

In the north-western coastal area of Norway, which is not particularly noted for endemic goitre and which until quite recently was isolated from the rest of the country, the incidence of differentiated thyroid cancer is high, a fact which has led to the suggestion that this may have resulted from inbreeding over a considerable period of time (Wade, 1975).

Autoimmune thyroiditis

Malignant lymphoma is a well-known sequel of Hashimoto's disease but the exact frequency of this change is difficult to assess.

Classification, pathology and natural history

Solitary non-functioning nodules of the thyroid gland are either cystic or solid, and the latter are either benign adenomata or cancers. Anything up to 20% of non-functioning solid nodules will prove to be cancerous, although a more common figure is about 10% (Katz and Warren, 1976; Burrow et al, 1978).

For a comprehensive review of the pathology of malignant tumours of the thyroid, the reader can do no better than refer to the paper by Woolner et al (1961), which reviews a very large number of cases collected in a major centre. The two most common cancers, specifically of thyroid tissue origin, are papillary and follicular carcinoma. Because of their distinctive histological features and behaviour, they are sometimes grouped together and referred to collectively as differentiated tumours.
Medullary carcinoma arises from the parafollicular or C cells and is, therefore, not a true thyroid neoplasm, but because of its anatomical location and similarities in behaviour, it is included in all discussion of thyroid neoplasia.

**Papillary carcinoma**

This tumour comprises about 60% of all malignant neoplasms in the larger American series, but figures a little less prominently in European reports. It is the type which most often follows previous irradiation. The histological appearance is one of papillary excrescences together with neoplastic follicles; in most cases encapsulation is absent.

Small papillary tumours not exceeding 1.5 cm in diameter and exhibiting marked desmoplasia and psammoma bodies are referred to as occult carcinomata. By contrast to their bulkier counterparts, they are usually detected by accident or their presence may be suspected when a metastatic node is discovered in the absence of palpable disease at the primary site.

Most papillary cancers present as discrete hard intrathyroid masses, but additional discrete deposits are found in other parts of the gland in as many as 40% of cases, most of these being microscopical. Whether these smaller deposits represent spread within the gland along lymphatic channels or entirely independent new tumours is impossible to say, but their presence has led some people to believe that papillary cancer is truly multifocal in origin.

The tumour generally grows slowly and is late to break through the capsule of the gland, although lymph node metastasis is not uncommon in many cases. The disease may occur at any age, but it is classically commoner in younger subjects, 42% being under the age of 40 (Woolner et al, 1961).

The ratio of females to males for all papillary cancers in the same study was 2.4:1, but that in cases which had broken through the capsule of the gland was 1.5:1, and in inoperable cases 0.7:1. This suggests that the prognosis may be influenced by the sex of the individual, although other factors also need to be taken into account.

Once the tumour has become extrathyroidal, it attaches itself to neighbouring structures and ultimately becomes invasive. Typically the tumour invades the strap muscles first and although lacking a capsule, it may well provoke the formation of fibrous tissue at its leading edge, thus creating the impression of encapsulation.

Gross invasion of veins is virtually absent, but invasion of the trachea, larynx, oesophagus, and recurrent nerves takes place when the disease has extended far beyond the confines of the gland.

Almost all papillary cancers exhibit a mixed papillary and follicular pattern, in varying proportions. At one end of the spectrum are cases which are wholly papillary, while at the other end the proportion of papillary elements is overshadowed by a predominantly follicular pattern. Even those exhibiting a mainly follicular appearance lack a capsule and behave in a similar way to their papillary counterparts.
Nodal metastasis occurs in as many as 40% of cases (Woolner et al, 1961). Deposits appear classically in the paratracheal nodes, but may present anywhere in the neck, so that lymphadenopathy in the carotid or supraclavicular triangles is common. Bilateral spread to the nodes is found in about 8% of patients.

Pulmonary metastasis is uncommon at presentation, and only affects 4% of all patients, chiefly those in the older group and those with extrathyroidal spread. It often presents as a diffuse coarse miliary infiltrate rather than a series of discrete rounded shadows.

**Follicular carcinoma**

Whereas the mean age for papillary cancer was 42 years in the series of Woolner et al (1961), that for follicular carcinoma was 50. The sex ratio however, was similar at 2.6:1 in favour of females, and the total group comprised 18% of all malignant thyroid neoplasms.

Follicular carcinoma is typically encapsulated and, in those lesions with minimal invasive characteristics it may be mistaken for a follicular adenoma. The extent to which the capsule is invaded, and in particular the veins located within the capsule, provides an indication of the likelihood of metastasis. The term 'micro-angio-invasive' has been coined to describe those tumours with little evidence of venous invasion, and in which the potential for local invasiveness and distant metastasis is consequently low. Gross infiltration of capsular veins is a bad prognostic sign and generally portends distant dissemination of the disease.

Besides this particular feature of follicular cancers, the pathologist may recognize a graduation from a well-differentiated to a poorly differentiated pattern, and this feature may also be used in forecasting the outcome. In some, de-differentiation assumes an even more sinister quality so that the tumour ultimately becomes anaplastic.

Once follicular carcinoma becomes extrathyroidal, the capacity for local invasion becomes more dramatic than in papillary cancer, although the target areas are essentially the same, namely the larynx, trachea, oesophagus and recurrent nerves. Curiously the only structure which defies invasion in both types of neoplasm is the carotid artery. In the case of the larynx and trachea, cartilage is destroyed and intraluminal extension of disease causes airway obstruction and bleeding. Oesophageal invasion is uncommon and late, and usually manifests as an insinuation of the disease between trachea and oesophagus, followed by migration some distance up and down the muscle coat, and between muscle and mucosa. The tumour rarely penetrates the mucosa to enter the lumen of the oesophagus and is unlikely therefore to be recognized by preoperative endoscopy.

Nodal metastasis is uncommon in purely follicular cancers being found in about 4% of the total (Woolner et al, 1961).

Metastases of angio-invasive tumours to bone and viscera are common, and the better differentiated of these may resemble normal thyroid tissue. In the lungs the appearance on X-ray is generally of multiple large rounded shadows and, in the bones, that of osteolytic lesions. The latter in fact may be extremely vascular, to the extent of resembling arteriovenous malformations.
In rare instances, the primary tumour may present as a hyperfunctioning nodule causing some degree of hyperthyroidism.

**Medullary carcinoma**

Medullary carcinoma, comprising between 5 and 10% of thyroid neoplasms, is a tumour of the parafollicular or C cells and, unlike differentiated thyroid neoplasms, is of neuroectodermal origin (Hazard, Hawk and Crile, 1959). It falls into one of two categories, namely the sporadic or the familial hereditary type of disease.

The former is generally seen between the third and seventh decades of life, with women again predominating over men. The latter, which is inherited as an autosomal dominant trait, may appear as early as the second decade, and affects both sexes equally.

Both types present as a single hard nodule, but in the familial group there is nearly always histological evidence of multicentricity. The tumour is solid and unencapsulated, and grows in sheets or nests surrounded by a hyaline fibrous stroma which stains strongly for amyloid.

The propensity for lymph node metastasis is exhibited by both familial and sporadic types, and as many as 75% of the total eventually develop lymphadenopathy. Like papillary cancer, medullary cancer may spread to any group of cervical lymph nodes, although the emphasis is frequently on the lower deep cervical, supraclavicular, and mediastinal nodes.

The natural tendency of the primary tumour is to enlarge and then to break through the capsule of the thyroid gland, first to attach to adjacent structures, and finally to invade them.

Medullary cancer is characterized by an elevation of the serum calcitonin, which tends to be relatively higher in the familial type of disease. Selective venous catheterization has been used in the past to determine the site of maximum calcitonin concentration in the gland, and is most useful for the detection of the site of any recurrent disease.

Infusion of calcium into the bloodstream, or the ingestion of whisky, are potent stimuli for the release of calcitonin, a fact which is made use of when cases of occult tumours are suspected, or when screening the siblings of a person known to suffer from the disease. The greater the bulk of the disease, both at the primary site and at sites of metastasis, the higher the level of the serum calcitonin.

When the tumour is confined exclusively to the thyroid gland, eradication of the disease should in theory reduce the serum calcitonin level to normal (less than 0.1 ng/mL), but this is seldom achieved even when distant spread is absent.

There is good evidence to suggest that serotonin is secreted by some medullary cancers since flushing, headache, breathlessness, sweating and fainting are experienced by some patients, and both the serum 5-hydroxytryptamine (serotonin) and the urinary 5-hydroxyindoleacetic levels are raised. Prostaglandins and histaminase are also secreted, the concentration of the latter serving as a useful guide to the extent of metastatic disease.
The intractable and severe diarrhoea which is so typical of many cases of medullary carcinoma is almost invariably linked to the presence of gross metastatic disease, mainly in the liver, but its precise cause is not understood. Of the patients 50% or more eventually suffer from this distressing symptom for which there is no specific antidote.

Blood-borne metastases are seen typically in the liver, but may also affect the lungs and the bones, at which site they may cause severe pain.

Other tumours are seen in about 10% of patients with familial medullary carcinoma, namely phaeochromocytoma, mucosal neuroma and parathyroid adenoma. Investigations should therefore include an estimate of the 24-hour urinary catecholamines, the level of vanillylmandelic acid in the blood and the serum calcium.

**Anaplastic carcinoma**

The average age of patients in this group is about 60 years and once again women outnumber men. The tumour only accounts for about 5% of all thyroid neoplasms, and even this may be an overestimate, since many lymphomata were previously mistaken for anaplastic cancer.

Microscopically, the tumour is characterized by large cells with markedly irregular nuclei some of which may take on a bizarre giant appearance, while others assume a spindly look, hence the subdivision into giant-cell or spindle-cell anaplastic carcinoma.

Typically there is a history of a long-standing, but asymptomatic, goitre preceding the sudden and explosive onset of the disease. The clinical course is characterized by rapid growth with pain, early invasion of surrounding structures, and a swift decline. Patients rarely survive for longer than one year after the diagnosis is made.

**Lymphoma**

Whereas anaplastic carcinoma is almost invariably fatal, the outlook in lymphoma is comparatively favourable.

This histology, which may bear some resemblance to anaplastic tumours, is usually distinctive, and there should rarely be any confusion between the two conditions.

The clinical course, however, may be almost as dramatic as anaplastic cancer, in that there is a sudden onset of swelling spreading to adjacent structures and causing acute respiratory embarrassment. Early treatment is essential for the prevention of complications, and this may then be followed by the appropriate investigations for staging the disease.

**Follicular adenoma**

This is a solitary encapsulated ovoid or rounded mass varying in consistency from firm to soft, depending on whether the growth has been the seat of haemorrhage, infarction, cyst formation, fibrosis, or calcification. The architecture of this neoplasm is one of multiple follicles of relatively uniform appearance with compression of the adjacent fibrous
parenchyma. Care must be taken not to overlook a carcinoma, the key being the absence of venous or capsular invasion.

Adenomata may be non-functioning, or may exhibit some degree of activity, varying from slight to hyperactive. True 'toxic' adenomata are uncommon: usually a toxic nodule suppresses the remaining parenchyma of a multinodular gland.

The diagnosis is generally made retrospectively when a solitary nodule is removed by a lobectomy.

Factors influencing prognosis

The two most important factors influencing prognosis in differentiated thyroid cancer are the age of the patient and the pathology of the tumour.

Most authors agree that survival is inversely related to age and that this effect is most evident from the age of 40 onwards (Staunton and Skeet, 1979).

The relation of the sex of the patient to prognosis is probably less obvious, although Doll (1969) was of the opinion that males with a differentiated thyroid cancer generally fare worse than females, and Crile (1971) concluded that under the age of 40, females unquestionably had a better prognosis.

The histopathological criteria which govern the behaviour of a thyroid neoplasm are also considered to influence prognosis. Papillary carcinoma tends to be slow growing and, although lacking a capsule, provokes a fibrous reaction as the disease becomes extrathyroidal. This may serve to lessen the tendency towards invasion, at least for a while. It would seem also that both occult tumours and papillary cancer induced by exposure to radiation in childhood carry a particularly favourable prognosis.

Follicular carcinoma exhibits a greater capacity for invasion, exemplified typically by its tendency to infiltrate the veins which lie in the tumour capsule. The degree of differentiation also has an important bearing on the final outcome, so that the expectations for a poorly differentiated follicular neoplasm must be lower than for a well-differentiated lesion.

In general, follicular tumours have a worse prognosis than purely papillary neoplasms, and papillary tumours with follicular elements are intermediate. Extrathyroidal spread carries a worse prognosis than disease confined to the gland.

Nodal spread of papillary carcinoma apparently does not affect the prognosis adversely.

Symptoms and signs

Thyroid neoplasms whether benign or malignant present in the early or intrathyroidal phase as solitary nodules, often of indeterminate consistency. They usually lie in one or other lobe, but occasionally arise in the isthmus of the gland. Even tumours which are eventually
reported by the pathologist to be multicentric generally appear to be uninodular on clinical examination. For the vast majority of patients, the principal concern about which they seek advice is the presence of a swelling.

Attempts to draw conclusions about the nature of the swelling from its consistency are generally unrewarding, for by no means every cancer possesses the hardness of squamous cancers, and in any case the interposition of numerous soft tissue layers between the palpating fingers and the neoplasms makes interpretation of the consistency difficult.

The distinction between benign and malignant in the later stages of the natural history of a thyroid swelling is usually much easier to make because adenomata do not become so overtly extrathyroidal in the way that cancers do. Conversely, large thyroid masses which blend imperceptibly into the adjacent anatomy so that their limits are indeterminate are rarely benign.

Fixation due to escape of the disease from the gland is therefore an important criterion of malignancy, and is evident by the failure of the mass to move up and down with swallowing.

Such is the diffuse extent of extrathyroidal spread in some cases, that the carotid artery may be displaced quite far laterally, or even obscured by the overlying neoplasm so that distal pulsations may be reduced. The close relationship of the cervical sympathetic nerve to the posteromedial aspect of the carotid sheath explains the presence of a Horner's syndrome when spread is as extensive as this.

Displacement of the trachea to the contralateral side does not in itself signify the presence of malignancy, since any large goitre may do this, but compression and narrowing of the trachea leading to wheezing and shortness of breath, especially when the patient lies down, should arouse suspicion of neoplasia.

Haemoptysis on the other hand is a clear indication of an invasive process which has finally broken through into the tracheal lumen, and should leave the clinician in no doubt about the nature of the disease.

Dysphagia is a late phenomenon in the natural history of thyroid cancers, and usually signifies infiltration of the gullet, rather than simple displacement such as occurs when a very bulky goitre comes into contact with the oesophagus.

The laryngeal alae or the cricoid may be infiltrated, and the hypopharynx may be invaded when disease from the posterior aspect of the upper pole of the thyroid attaches itself to the pyriform sinus.

Hoarseness is common in such circumstances, and is later followed by dyspnoea and laryngeal stridor. Indirect laryngoscopy shows an oedematous mucous membrane encroaching on the laryngeal lumen.

Hoarseness is, however, more likely due to paralysis of the vocal cord resulting from infiltration of the recurrent laryngeal nerves by cancerous deposits in its immediate vicinity.
It should be said that vocal cord mobility may be preserved for a long time in the face of obvious invasion of the recurrent nerve.

Lymphadenopathy is most apparent at those sites which are amenable to palpation, but the nodes which are most consistently involved, namely the paratracheal, are deep seated and are therefore rarely palpable.

As nodal disease enlarges, so the likelihood of fixation to the jugular vein and prevertebral fascia increases.

Retrosternal extension, a common feature of large follicular cancers, causes tracheal displacement and narrowing as well as engorgement of the neck and upper chest wall veins. At operation the retrosternal extension, unlike its benign counterpart, is often attached to adjacent structures in the mediastinum such as the pleura, trachea, recurrent nerves, oesophagus and prevertebral fascia.

The pathology of each thyroid nodule largely determines the pattern of symptoms in any given case. Follicular adenomata rarely give rise to any symptoms other than awareness of the presence of a swelling.

In papillary carcinoma, the primary tumour often fails to grow to any significant degree, and may be overshadowed by quite substantial lymphadenopathy.

Follicular and medullary cancers are similar in their behaviour since both diseases tend to be bulkier and to spread more readily. Hence the symptoms are related to the displacement and invasion of structures lying close to the thyroid gland.

Both lymphoma and anaplastic carcinoma are characterized by rapid growth, pain and respiratory obstruction.

**Diagnosis**

*Thyroid scans*

Patients undergoing scanning should not have received iodine compounds or thyroid supplements for 4-6 weeks before the procedure, as uptake of tracer is suppressed. The scan is carried out 20-30 minutes after an intravenous dose of technetium-99 or 24 hours after an oral dose of iodine-131 or iodine-123. With technetium either a rectilinear scanner or a gamma camera fitted with a pinhole collimator may be used, but with $^{131}$I the rectilinear scanner is supposedly better.

Just before the scan, the patient is given a glass of water to wash away any radionuclide which has found its way into the pharynx in the saliva. Anatomical landmarks, such as the suprasternal notch, are defined on the scan by means of a radioactive marker which is placed on the patient. This is important for the accurate localization of nodules and for confirmation of retrosternal extensions.
Non-functioning solitary nodules, often referred to as 'cold' nodules, appear on the scan as localized areas of diminished or absent tracer uptake, but this does not indicate whether they are cystic and therefore unlikely to be malignant, or solid.

The converse of the 'cold' or non-functioning lesion is the 'hot' nodule, indicative of a biologically active circumscribed area of gland parenchyma. It is rare for such a nodule to be malignant, but exceptionally malignant lesions may be 'hot' on technetium scanning and 'cold' on iodine scan.

There has been no specific radionuclide imaging technique for the demonstration of primary or metastatic medullary cell carcinoma until the recent development of \((^{131}I)\text{meta-iodobenzyl-guanidine (MIBG)}\). Being a guanethidine analogue, it is taken up by chromaffin-containing tissue of neuroectodermal origin, such as phaeochromocytoma, for the imaging of which it was in fact designed. But since medullary carcinoma arises from the parafollicular cells which also originate from the neural crest \((^{131}I)\text{MIBG}\) has been used with some success for the imaging of both primary and metastatic tumours (Clarke, Fogelman and Lazarus, 1986).

**Ultrasound**

This is a valuable complementary investigation to radionuclide imaging since it may distinguish between cystic and solid nodules. Although cancers could conceivably arise in cysts, or alternatively undergo cystic degeneration, such an eventuality is extremely rare and almost all cancers are solid.

The results of ultrasound scanning and the findings at operation are usually in agreement but disparities do occur, such as when a solid nodule has undergone cystic degeneration.

**Radiology**

X-ray of the neck and the thoracic inlet will establish whether there is any displacement or compression of the trachea and will demonstrate the calcification which is sometimes present in papillary cancers.

Barium swallow complements the posteroanterior and lateral plain radiographs and will reveal the presence of oesophageal shift or narrowing.

Chest X-ray is carried out routinely to determine if there are pulmonary metastases, which appear as rounded shadows in follicular carcinoma and as a miliary infiltrate in papillary carcinoma.

**Fine needle aspirate**

Aspiration biopsy cytology is a safe and easy preoperative investigation, but it requires the services of an experienced cytologist for the interpretation of the findings. Unlike larger needles such as the Vim-Silverman which extracts a core of tissue, aspiration with a fine
needle yields a liquid specimen which is smeared on to a slide and may be reported on in minutes.

With such a technique, it is possible to identify papillary and follicular cells as distinct entities, but the distinction between the follicular cells of an adenoma and a carcinoma is very difficult to make.

The simplicity of the technique and the fact that the percentage of false negatives may be as low as 2.2% recommends it as part of the routine work-up of patients with thyroid swellings (Lowhagen et al, 1979).

**Thyroglobulin estimation**

This antibody is detectable in the blood of all patients with thyroid cancer and its level serves as a very sensitive marker of the extent of the disease. Patients who have been treated successfully lose all trace of thyroglobulin in their serum, and those who after a period of quiescence eventually develop a recurrence, once again demonstrate its presence in their blood.

The test is therefore useful for gauging the success or otherwise of treatment.

**Calcitonin estimation**

Calcitonin levels in the blood of patients with medullary carcinoma are abnormally high and, like thyroglobulin, the greater the bulk of the disease, the higher the level.

Unlike thyroglobulin, however, it does not often drop to the normal level of below 0.1 ng/mL in patients who have apparently been treated successfully.

**Other investigations**

Patients suffering from familiar medullary carcinoma may also harbour other neoplasms, such as phaeochromocytoma and parathyroid adenoma.

To rule out the presence of such associated tumours, it is appropriate to estimate the level of vanillylmandelic acid in the blood together with that of the urinary catecholamines over 24 hours and, in addition, the serum calcium.

Patients suspected of metastases from primary follicular or mixed papillary - follicular carcinoma should undergo whole body radioiodine scans after ablation of the thyroid gland.

In the case of medullary carcinoma, scanning is usually carried out with technetium, although (131I)MIBG may also be used. Because of the expense of this agent, it is generally reserved for the identification of residual or metastatic disease rather than as a primary diagnostic measure.
In its absence, general surveillance of metastatic medullary carcinoma may be carried out with technetium scans to show soft tissue deposits and gallium scans to delineate osseous lesions.

**Principles of treatment**

With the exception of anaplastic carcinoma and lymphoma, the mainstay of treatment for all thyroid neoplasms is surgery. The contentious issue is whether the operation for differentiated lesions should be a partial or a complete removal of the gland, bearing in mind that total thyroidectomy carries a higher morbidity.

The debate hinges on two points, namely whether the more radical of the two procedures improves the prognosis and whether its complication rate is as high as is claimed.

The morbidity of the operation should theoretically be easy enough to assess by reviewing the incidence of parathyroid insufficiency and that of recurrent nerve paralysis in the literature. But as so often happens in large series, there is considerable variability in the clinical material and in the experience of those performing the surgery. If one group of patients has a significantly higher proportion of advanced cases, the morbidity in respect of the parathyroids and the recurrent nerves is bound to be higher.

Tollefsen, Shah and Huvos (1972), for example, quoted an incidence of 21% for hypocalcaemia after total thyroidectomy, whereas that in Mustard's (1970) hands was 12% and that reported by Bartolo, Kay and Talbot (1983) was nil.

Similarly the incidence of permanent recurrent nerve paralysis may be as high as 17% when secondary thyroidectomy is practised (Beahrs and Vandertoll, 1963) or as low as 4.8% after primary total thyroidectomy (Thompson and Harkness, 1970).

The point which is much more difficult to decide is whether total thyroidectomy achieves a higher cure rate than lobectomy in differentiated thyroid cancer. Arguments are marshalled in favour of each, but the truth of the matter is that comparisons between reported series are invalid because of the abundance of variables.

In order to make matters simple, the reasons for and against are discussed, and guidelines for the choice of one of the two options drawn up.

**Papillary carcinoma**

The protagonists of lobectomy argue that the incidence of recurrent disease in the contralateral remaining thyroid lobe is about 4%, in spite of the fact that as many as 40% of cases exhibit signs of multicentricity throughout the gland (Tollefsen and Decosse, 1963; Tollefsen, Shah and Huvos, 1972). It is further suggested that the recurrence rate might have been even lower if the patients had been maintained on thyroxine after operation by virtue of its suppressive effect on TSH (Crile, 1980).

On the other hand, the incidence of recurrence in the contralateral lobe after hemithyroidectomy may be considerably higher: 24% in one series (Rose et al, 1963) and
17% in another (Hirabayashi and Lindsay, 1961). The large difference in the recurrence rate can be explained on the basis of the age factor, since the prognosis of patients over the age of 40 is much worse than that in younger subjects.

There are also some who speculate that some of the recurrences may have been sparked off by the administration of postoperative radioiodine, given with the intention of ablating remaining thyroid tissue after hemithyroidectomy. It has been postulated that this, rather than the inadequacy of the original operation, was responsible for recurrence of disease and that the tumour may also have been transformed from a well-differentiated to an anaplastic lesion in the process (Crile, 1971).

As regards the management of lymph node metastasis, most authors agree that removal of overtly diseased nodes by simple rather than radical dissection suffices in all but the most advanced cases. There is also general agreement that the presence of lymph node metastasis does not make the prognosis any worse.

With all these points in mind, how should one proceed when faced with a case of papillary carcinoma of the thyroid? Patients under the age of 40 with intrathyroidal disease, especially those with a previous history of exposure to radiation, may be managed by hemithyroidectomy with the local removal of any enlarged nodes. The risk of postoperative tetany is nil and that to the recurrent nerve is halved. However, it is vital that the patient is maintained permanently on an adequate dose of thyroxine. If Crile's (1971) comments on the inadvisability of postoperative radioiodine are accepted, a matter which not all are agreed about, this treatment must clearly be withheld.

It would seem that the place for total thyroidectomy is in the management of the older patient, and in all patients who exhibit signs of extrathyroidal escape.

The case for ablating microscopical residues with radioiodine is much stronger in this group in view of the increased likelihood of recurrence if thyroid tissue is allowed to remain. The therapeutic value of such treatment lies both in the eradication of the soil in which microscopical tumour deposits reside and in the effect of the isotope on the tumour cells directly, many of which are derived from mixed papillary and follicular lesions and therefore likely to take up radioiodine.

For those cases with unequivocal papillary tumours which have escaped into the surrounding tissues, the alternative of postoperative external beam radiotherapy may be equally or more effective.

The management of neck nodes does not differ in the more advanced cases, although the need for radical neck dissection as opposed to 'berry picking' may be that much greater. It is mandatory to keep all the patients on thyroxine and to monitor subsequent recurrence by regular scanning and serum thyroglobulin estimations.

**Follicular carcinoma**

Those who support the view that follicular carcinoma with minimal invasion of capsular veins is a relatively benign disease would go on to argue that a hemithyroidectomy
is an adequate procedure for this neoplasm. Such an argument is based on the fact that the potential for spread is small and, consequently, the need for whole body scans to detect metastases after surgery is unnecessary.

However, one can never be certain that a follicular tumour, no matter how benign in appearance, will not metastasize, and since whole body scans for the detection of distant spread are unsuccessful when a significant bulk of normal thyroid tissue remains, the argument in favour of total thyroidectomy would appear to be stronger.

This, together with the fact that follicular carcinoma is seen in an older age group in whom the prognosis is considered to be generally worse, should steer the clinician in the direction of total removal rather than a hemithyroidectomy.

Any residual thyroid tissue or distant deposits which are picked up by a postoperative whole body scan are managed by radioiodine therapy, and hormone therapy is withheld until such treatment is completed.

The propensity for lymph node metastasis in follicular cancer is low, but the principles of their management are the same as for papillary carcinoma.

**Medullary carcinoma**

The prognosis for this tumour is worse than for differentiated tumours. Whereas the presence of lymphadenopathy in papillary neoplasms does not appear to affect the outcome adversely, the same cannot be said of medullary carcinoma. Quite clearly the factors which influence survival are local escape of disease from the gland and malignant lymphadenopathy, both of which herald the appearance of distant spread.

Total thyroidectomy is the treatment of choice in all cases, be they sporadic or familial, and this includes members of a family who have raised calcitonin levels but who do not show signs of overt disease on clinical examination or on scanning.

Lymph node deposits are generally more diffusely spread than in differentiated thyroid cancer and the surgical clearance must, therefore, be more ambitious, to include the upper mediastinum as well as the cervical nodes.

The question as to whether or not external beam radiotherapy is useful in the control of local disease has not been addressed adequately so far, but there is no reason to suppose that it may not be helpful in management of postoperative microscopical residues.

**Anaplastic carcinoma**

This disease advances with such rapidity that very few cases are suitable for surgical excision. Those early cases which prove to be suitable for total thyroidectomy should receive postoperative radiotherapy to control local disease, even if distant metastases have already appeared.
Inoperable disease which threatens to cause respiratory obstruction should be managed by uncapping the trachea to avert imminent asphyxiation, followed by irradiation. The operation also yields a sample of tissue for histology.

**Lymphoma**

Resection of the isthmus for the purpose of averting or relieving respiratory embarrassment should be followed by immediate radiotherapy with the expectation of rapid recovery. During treatment the patient is fully investigated for the purpose of staging the disease and deciding whether chemotherapy will be required or not.

**Flow chart for thyroid cancer**

- Thyroid swelling (Technetum scan) --> Cold solitary nodule (Ultrasound scan) --> Solid nodule (Aspiration cytology; Thyroglobulin estimation; Open biopsy and frozen section; CT scan; Calcitonin estimation) --> Carcinoma:
  - Pure papillary
    - Intrathyroidal under age of 40 --> Hemithyroidectomy + thyroxine
    - Extrathyroidal --> Total thyroidectomy + DXT + thyroxine
    - Intrathyroidal over 40 --> Total thyroidectomy + thyroxine
  - Mixed papillary/follicular --> Total thyroidectomy + radioiodine + thyroxine
  - Follicular --> Total thyroidectomy + radioiodine + thyroxine
  - Medullary --> Total thyroidectomy +/- DXT + thyroxine
  - Lymphoma + anaplastic --> DXT +/- Chemotherapy.

**Surgery**

Most solitary nodules of the thyroid gland are removed by partial thyroidectomy and the diagnosis in the majority of cases proves to be benign, the commonest diagnosis being follicular adenoma.

The diagnosis of differentiated thyroid cancer is therefore frequently arrived at by accident following lobectomy for a supposedly benign solid nodule. After consideration of all the relevant factors, a decision must then be made as to whether the treatment thus far has been adequate or whether the contralateral thyroid lobe should also be removed.

Some surgeons, however, favour the operation of subtotal thyroidectomy as an alternative to total removal, the aim being to leave a sliver of normal thyroid tissue to protect the recurrent nerve and two parathyroids on the side of least disease.

It is not popular with others, however, because it leaves very little scope for the safe removal of the remaining thyroid tissue in the event of subsequent local recurrence at that site.

The operation generally referred to as lobectomy entails the removal of the dissected lobe, usually with some part of the thyroid isthmus, and is therefore in essence a hemithyroidectomy.
A description of this procedure follows and, as total thyroidectomy is simply a double hemithyroidectomy, it will not be discussed.

**Lobectomy**

With the anaesthetized patient in the supine position, and the head and neck fully extended, a collar incision is made from one side of the neck to the other above the hollow of the suprasternal notch.

To facilitate access to the upper pole of the gland, the incision should not be too low and by setting it above the hollow between the two sternal heads of the sternomastoid muscles, a bow-string effect in the scar can be avoided.

It is deepened down to the deep cervical fascia but, thereafter, access to the gland may be obtained in one of two ways.

For the beginner and in cases of a large goitre, division of the deep cervical fascia transversely with ligation of the anterior jugular veins followed by division of the strap muscles in the same axis is the easier option.

The alternative is to divide the deep cervical fascia vertically in the midline and then to free the deep surface of the strap muscles from the underlying gland, retracting the straps in the process. Access by this method is usually a little more restricted and is more suitable for smaller goitres.

Once the thyroid gland has been sufficiently exposed by lateral retraction of the strap muscles, the lobe is displaced medially and forwards by the assistant, and the common carotid artery identified and then retracted laterally.

The object is to create enough space between the carotid and the gland for the identification and isolation of the inferior thyroid artery, which should be divided as far away from the gland as possible to avoid the recurrent laryngeal nerve. The artery is in fact an important surgical landmark because of its intimate relationship to the recurrent laryngeal nerve, and provides an accurate indication of the depth at which the nerve should be sought. Where the artery enters the gland is one of two points at which the thyroid lobe is tethered posteriorly, the other being the ligament of Berry. Hence division of the artery facilitates the forward and medial mobilization of the thyroid lobe.

An attempt is now made to find the recurrent nerve below the inferior thyroid artery and at a corresponding depth. It lies in loose fibroareolar tissue and can be felt as a cord even before it comes into view.

On the right side it passes obliquely from lateral to medial as it approaches the inferior thyroid artery, but thereafter it lies in the tracheo-oesophageal groove. The nerve either passes deep to, or superficial to, or between the terminal branches of the inferior thyroid artery, this last arrangement being the most common finding on the right.
On the left side, the nerve lies in the tracheo-oesophageal groove throughout its entire passage, and crosses behind the artery more often than in front of it or between its branches.

After identifying the nerve, attention is turned to the inferior thyroid veins which are isolated and ligated in turn, and then to the superior thyroid vascular pedicle which is divided as close to the gland as possible to avoid the external laryngeal nerve.

Once all the principal vascular channels have been secured, the surgeon can mobilize the thyroid lobe off the recurrent laryngeal nerve. This is done by releasing the extension of the surgical capsule which passes back from the posteromedial surface of the thyroid lobe to the prevertebral fascia, working from below upwards and dividing this fascial attachment in a plane just superficial to the nerve. The point of greatest adherence between the nerve and the thyroid gland is in the condensation of the surgical capsule which tethers the thyroid lobe to the trachea and to the prevertebral fascia, namely the ligament of Berry. This is the area where the nerve is at most risk since forward traction on the gland pulls the nerve anteriorly with it.

During the process of freeing the thyroid gland from its posterior attachments and from the nerve, every effort should be made to find the two parathyroid glands. The upper is generally found on the posteromedial border of the thyroid lobe, at the level of the pharyngo-oesophageal junction. It is nearly always placed alongside a pad of fat which is separate from the gland, and its feeding vessel is the upper branch of the inferior thyroid artery.

The lower is usually found a short distance from the lower pole of the thyroid and is more anteriorly and medially placed than the upper parathyroid, and therefore closer to the trachea. It too is attached to a pad of fat and is nourished by the inferior thyroid artery by way of its lower branch. Both parathyroids appear as flat, brown, pear-shaped structures, with one sharp edge, and are freely mobile when displaced. They are extremely vulnerable and must be handled very gently.

Once the thyroid lobe is freed from the side of the trachea, it is lifted forwards and medially to mobilize the isthmus of the gland, which is then transected just beyond the midline. After suturing the cut edge of the remaining isthmus with catgut the wound is drained and closed in layers.

**Postoperative management**

After a total thyroidectomy the serum calcium is estimated daily to detect hypocalcaemia, but if signs of tetany appear, calcium gluconate is given by mouth or, if a rapid effect is required, by intravenous injection. At a later stage the degree of hypocalcaemia is reviewed and a decision taken as to the need for vitamin D₃ supplements.

Patients who undergo lobectomy are prescribed thyroxine in a dosage between 0.1 and 0.3 mg daily, and the effectiveness of suppression of TSH output assessed later, by injecting TRH and estimating the level of TSH in the blood; those who are maximally suppressed by an effective dose of thyroxine will not show a rise of TSH.
By contrast, patients who have undergone total thyroidectomy and who are likely to receive radioiodine are denied replacement therapy for 3-6 weeks to allow their TSH levels to rise before ablating any residual thyroid tissue with radioiodine. It is usual to scan the patient immediately before the administration of the isotope to show the site and extent of the remaining thyroid tissue.

Once ablation with the isotope has been carried out, thyroxine is prescribed long term and is only discontinued for short periods before future diagnostic scans, at which time the serum thyroglobulin is also estimated. The need for repeat treatment with radioiodine is judged on the findings of each scan.

Complications

Recurrent nerve paralysis is an unlikely complication after lobectomy or total thyroidectomy when disease is purely intrathyroidal. It is generally due to excessive traction, rough handling, coagulating vessels to close together, or catching the nerve in a ligature.

Paralysis is likely when disease has escaped from the gland and abuts against the nerve or when a cancerous paratracheal node is stuck to it. Attempts to peel neoplastic tissue off the nerve sheath are unrewarding; they contribute little to clearance of the disease, and they are likely to paralyse a nerve which hitherto may have been functioning.

Under such circumstances a decision has to be made as to whether or not to dissect out the contralateral nerve, in view of the probability that the nerve on the diseased side is likely to be non-functional.

Unilateral nerve paralysis may require a vocal cord augmentation procedure, either by the injection of Teflon or by the insertion of cartilage strips in the paraglottic space (Shaheen, 1984). Bilateral nerve paralysis is managed by immediate tracheostomy, and later if desired by a partial corpectomy and arytenoidectomy using the carbon dioxide laser (Shaheen, 1984).

External laryngeal nerve palsy is also more likely to occur when disease is extrathyroidal and, if present on its own, is expressed by the patient as a limitation in the register of the voice or an inability to sing. In conjunction with a unilateral recurrent nerve palsy, it manifests as a weak, breathy voice, and with a bilateral recurrent nerve palsy, as a severely compromised airway, although slightly less so than when a bilateral recurrent paralysis exists on its own.

Preservation of at least two parathyroids is necessary for the prevention of tetany, and care should be taken during a total thyroidectomy to identify as many of the glandules as possible. Finding them is never easy, but attention to the finer points of surgical anatomy and the use of the operating microscope generally yield dividends. If no glandule is found during the operation, the excised specimen should be inspected carefully and any parathyroid tissue which is considered to be free of disease is re-implanted into the patient. The value of such a manoeuvre is still the subject of debate, but nothing is lost by attempting it.
Chapter 17: Cancer of the neck

David Wright and Guy Kenyon

The management of neck cancer has undergone a succession of changes, but no treatment yet devised guarantees the patient the chance of a certain cure. Various combinations of treatment have been, and are still being, evaluated to improve both the cure rate and the rehabilitation of the patient. The control of regional metastatic disease constitutes a significant part of the process of treating head and neck cancer. When surgery is employed, either as the selected method of treatment or when other treatment has failed, a decision usually needs to be made on whether a neck dissection will have to be performed in addition to removing the primary cancer. The former may be necessary if lymph nodes are clinically positive; however, the type of neck dissection and the indications for bilateral neck dissection remain far from clear. In contemplating a surgical approach, ablation of the tumour by radical neck dissection is not acceptable if, as a consequence, the social and emotional well-being of the patient is destroyed. As Conley (1983) stated:

'Every surgeon engaged in this type of work recognises that a large percentage of his attempts to cure or palliate are associated with some degree of mutilation that will interfere with physiological function and aesthetics. The true evaluation of this mutilation can only be measured by the patient and his family. The physician's estimate, at best, is technical, remote and at times compassionate. The new physician often separates the disease from the patient, and the patient from his doctor. He must, however, be sensitive from the earliest moment of his involvement to all of the facilities that can be applied to ameliorate the inherent psychology and physical stresses of ablation'.

Ever since Crile (1906) described the radical neck dissection, it has been known that this operation does not always control cervical metastasis. Massive and fixed nodes that extend into surrounding tissues, or involve the skin or skull base, as well as the relatively inaccessible nodes of the superior mediastinum or lower neck, are not resectable. Even in the clinically negative neck, the rate of recurrence is between 7.5 and 24% (De Santo et al, 1982), depending on the site and control rate of the primary lesion.

A primary carcinoma arising in most sites in the head will ultimately drain into the lymph nodes of the neck. These lymph nodes usually form an efficient barrier to the further spread of cancer with the effect that distant metastasis may not occur or, if it does, only as a late event.

Organization of lymphatic drainage

Normal anatomy

The lymph nodes of the head and neck form one interlinked continuum. Although minor and confusing differences are found in the nomenclature given in different descriptions of lymph nodes in the neck, there is common agreement that, anatomically and functionally, the nodes form two main and distinct groupings. These are best thought of as one terminal or deep cervical group which is related to the internal jugular vein, and two groups of intermediary or outlying nodes which drain into the deep cervical chain. Ultimately, therefore,
the lymph form the head and neck drains into the deep cervical group either by direct passage from the adjacent tissues or indirectly from one of the outlying groups. In turn, the afferent lymphatics from the deep cervical chain on each side coalesce to form a jugular trunk which on the right side drains to the junction of the internal jugular and subclavian veins, and on the left usually terminates in the thoracic duct.

By using this arrangement, named groups of nodes can be described. The most widely quoted system in current anatomic text considers lymph drainage in the following groups:

1. deep cervical nodes including 'jugular chain' (named nodes - retropharyngeal, jugulodigastric and jugulo-omohyoid)
2. anterior cervical nodes (named nodes - infrahyoid, prelaryngeal and tracheal)
3. superficial nodes of scalp and face
   a. occipital
   b. retroauricular (mastoid)
   c. parotid and superficial cervical
   d. facial
   e. submandibular
   f. submental.

It is important to appreciate that the peripheral nodes associated with the scalp and face, together with the retropharyngeal and submental groups, represent a continuous ring surrounding the cervical structures like a collar. The anterior and lateral cervical nodes form separate chains lying inside this shield.

**Deep cervical nodes**

Most pathological processes are encountered in the deep or lateral cervical cain of nodes, as this group is the common route of drainage from all major regional structures - from the nasopharynx superiorly to the thyroid gland inferiorly. These nodes extend from the base of the skull to the root of the neck along the length of the great vessels. Most of them are concealed by the overlying sternomastoid muscle, but the lower end is uncovered and is, therefore, palpable in the lower part of the posterior triangle of the neck. The omohyoid muscle divides this group of nodes into superior and inferior parts. The majority of the superior group lie beneath the upper end of the sternomastoid muscle, but some of the nodes are evident in the anterior triangle of the neck with the most frequently palpable of these, the jugulodigastric, lying below the angle of the jaw. This node is associated particularly with drainage of the tongue and tonsil. Efferent lymphatics from the superior group pass to the lower cervical group and hence to the jugular lymph trunks. The inferior deep cervical nodes are closely related to the brachial plexus and subclavian vessels. The jugulo-omohyoid node in this group lies on, or above, the intermediate tendon of the omohyoid muscle, and receives lymph from both the submental group and the tongue.
Group of nodes draining deeper structures

The deeper structures of the head and neck all drain to the deep cervical nodes through one or other of the local regional groups; these include the retropharyngeal as well as the paratracheal and retrotracheal groups, and the prelaryngeal and infrahyoid nodes. The retropharyngeal nodes are further split into two subgroups, a median and a lateral, which lie on the front of the atlas along the lateral border of longus capitis, and which receive afferent lymphatics from the nasopharynx, the auditory tube and the adjacent cervical vertebrae. The paratracheal glands lie alongside the trachea and oesophagus and the pretracheal nodes are a group lying anterior to the trachea in intimate relationship with the inferior thyroid veins. The prelaryngeal nodes are also included in this group, and lie on the conus elasticus of the larynx. The main infrahyoid group, which completes the nodes draining the deeper structures, is found on the thyrohyoid membrane.

The superficial ring of nodes

The superficial ring of nodes drain the skin of the scalp and face. The occipital nodes lie at the apex of the posterior triangle and are contiguous with the retroauricular and mastoid groups which lie over the mastoid process in the superficial fascia. Parotid nodes lie on and in the parotid gland, and some extend down into the neck along the external jugular vein as part of the superficial group. The facial and buccal nodes are also grouped together; a small node often lies on the buccinator muscle and another on the lower border of the mandible at the anterior border of the masseter near the mandibular branch of the facial nerve. The submental nodes comprise three or four nodes lying on the surface of the mylohyoid between the two bellies of the digastric muscle; these nodes receive afferent lymphatics from both sides of the median plane, and some of the lymph channels in this region decussate over the symphysis of the mandible to drain into the submandibular and jugulo-omohyoid nodes. The final group, the submandibular nodes, lies on the surface of the submandibular gland and receives lymph from a wide area of the centre of the forehead, the nose and adjacent cheek, the upper lip, the floor of the mouth, the gums and from the major part of the tongue.

Lymphatic drainage of the neck

Following the definition and classification of specific lymph node groups within the neck, the topographical distribution of lymph node metastases can be described. Such patterns have been investigated in block dissection specimens by first staining and then, after clearing in cedar wood oil, by transilluminating the specimen (McKelvie, 1976). Subsequent superimposition on a Rouvier's diagram allows the accurate plotting of metastatic disease.

A study of 1155 cancer patients (Lindberg, 1972) documented tumour spread from the principal primary sites. This study found that tumours of the tongue and the floor of the mouth metastasize most frequently to nodes in the upper deep cervical chain and to nodes in the submandibular group. Tumours of the oropharynx also commonly drain to the upper deep cervical group; lesions of the retromolar trigone, anterior faecial pillar and soft palate are particularly likely to involve the jugulodigastric node. However, the incidence of metastases to the midpart of the jugular chain and to the submandibular group is also high, and with tumours of the soft palate, the incidence of bilateral metastases to the upper deep cervical groups is appreciable. In the case of tumours of the tonsil, the jugulodigastric node is almost
always the first to be involved, but in this study the incidence of involvement of the mid- and lower cervical chain was also significant, as was the involvement of nodes in the posterior triangle on both the ipsilateral and contralateral sides. A survey of the location of cervical metastatic lymph nodes in relation to the possible primary sites may be of diagnostic value.

Bilateral metastases were also found with tumours of the base of the tongue and lateral and posterior oropharyngeal walls, and the most commonly involved nodes at presentation were the upper and midcervical groups.

Tumours of the hypopharynx were found to drain most commonly to the upper, the mid- and then finally the lower deep cervical chain in a decreasing order of frequency. Ipsilateral posterior triangle nodes were only occasionally seen. In contrast, carcinomata of the nasopharynx were found not only to have a very high incidence of spread to the posterior triangle, but also the highest incidence of bilateral spread. There was also a high predilection for the supraclavicular lymph nodes.

The lymphatic drainage from the larynx is divided, with the watershed lying at the level of the vocal cord. Most of the drainage is in a lateral direction above and below this level, but there is also some anastomosis on the posterior laryngeal wall. Above the vocal cords, the lymphatics pierce the thyrohyoid membrane and run with the superior laryngeal vessels to the upper deep cervical nodes. The drainage inferiorly is to the lower deep cervical chain, and the lymph passes either between the cricoid and the first tracheal ring or between the pretracheal and paratracheal nodes.

Hence, nodal involvement with metastasis within the neck originating from these primary sites is principally to the jugular chain, with the superior nodes being most commonly involved followed by the mid-jugular nodes. There is some variation between the different studies but it would seem that the submandibular nodes are rarely involved and then only in cases where there is extensive spread to deep nodes.

The role of cervical nodes

The function of the lymphatic drainage is to carry macromolecules, such as protein and particulate matter including effete cells, away from the tissues. A small amount of interstitial fluid also traverses the lymph vessels, but the removal of the bulk of tissue fluid remains a function of the blood capillaries. It is by way of the lymphatic flow that tumour cells drain to the regional lymph nodes.

The function of regional lymphatics is far from clear and is made more complicated by the evidence in favor of their playing a significant role in tumour immunity in the cancer-bearing patient. The work of some authors has suggested that the removal of nodes which are not involved with the tumour could promote the development of residual micrometastases (Fisher and Fisher, 1972). Other work has shown sinus histiocytosis in nodes involved with the draining of a primary malignancy (Black, Speer and Opler, 1958), and has considered this change and the degree of lymphocytic infiltration to be an indication of the strength of an antitumour immune reaction which will influence the prognosis. Furthermore, it has been shown that as the antigen load from a tumour increases, the effector reaction of the node is

The existence of an enlarged node does not, therefore, necessarily indicate the presence of metastatic spread and, especially if the node is soft, may represent no more than a response to coincidental infection or the mounting of an immunological reaction. However, it has been estimated that a node smaller than 1.0 cm is impalpable and yet may contain between $10^6$ and $10^7$ tumour cells. The principal problem for the clinician is how to decide whether a node which is palpably enlarged is involved with micrometastases. At present, such a decision remains purely clinical.

**TNM classification of cervical nodes**

A carcinoma arising in the head and neck is staged according to the extent of the tumour and the presence or absence of spread to the regional nodes and to distant sites. In practice, the latter event is a rare occurrence, and staging is feasible by clinical assessment of the primary site and examination of the neck. The staging system in most common usage in the UK at the present time is that proposed by the International Union against cancer (UICC, 1978), which is based on data developed by the American Joint Committee (AJC) for Cancer Staging. This system is shown in Table 17.1.

**Table 17.1 Regional lymph nodes**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>N0</td>
<td>Regional lymph nodes not palpable</td>
</tr>
<tr>
<td>N1</td>
<td>Movable homolateral nodes</td>
</tr>
<tr>
<td>N1a</td>
<td>Nodes not considered to contain growth</td>
</tr>
<tr>
<td>N2a</td>
<td>Nodes considered to contain growth</td>
</tr>
<tr>
<td>N2</td>
<td>Movable contralateral or bilateral nodes</td>
</tr>
<tr>
<td>N2a</td>
<td>Nodes not considered to contain growth</td>
</tr>
<tr>
<td>N2b</td>
<td>Nodes considered to contain growth</td>
</tr>
<tr>
<td>N3</td>
<td>Fixed nodes.</td>
</tr>
</tbody>
</table>

Although such a system is useful, many remain critical of its application. It has been shown that different observers will fail to agree on the presence of a palpable lymph node in as many as 30% of cases (Sako et al, 1964). This in turn makes even more questionable the validity of a clinical opinion as to whether a palpable node contains tumour and also the feasibility of clinically subdividing N1 and N2 regional nodes into subsets (a) and (b). The subjective nature of such an assessment is emphasized by one study in which it was demonstrated that only 60% of palpable nodes contained tumour at the time of their removal (Nichols and Greenfeld, 1968).

In addition, the present classification implies that there is a worsening of the prognosis with different stages from N1 to N3. However, this is certainly not the case, as the prognosis for bilateral nodes, N2, is usually much worse than for unilateral fixed nodes, N3. In view of these failings, a move towards the more cumbersome but possibly more realistic system, as used at the M. D. Anderson Hospital, Texas, would probably be appropriate (Table 17.2), and it seems likely that the UICC will adopt such a system in the future.
Table 17.2 Nodal staging system

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>N0</td>
<td>No clinically positive node</td>
</tr>
<tr>
<td>N1</td>
<td>Single clinically positive node &lt; 3 cm in diameter</td>
</tr>
<tr>
<td>N2a</td>
<td>Single clinically positive node &gt; 3 cm in diameter</td>
</tr>
<tr>
<td>N2b</td>
<td>Multiple clinically positive ipsilateral nodes</td>
</tr>
<tr>
<td>N3a</td>
<td>Unilateral fixed node(s), clinically positive</td>
</tr>
<tr>
<td>N3b</td>
<td>Clinically positive bilateral nodes, fixed or not fixed</td>
</tr>
</tbody>
</table>

Recent evidence has suggested that it is not so much the presence of a node which is of importance with regard to the assessment of prognosis, but rather the anatomical level of the tumour within the neck. Barrie, Knapper and Strong (1970) have suggested five levels of possible involvement (submandibular, upper, middle and lower cervical, and posterior triangle), and have shown that the 5-year survival for patients with a variety of different primary tumours dropped from 45% for those presenting with a node at level 12 (submandibular) to only 18% for those who had lymph node involvement at level 4 (lower cervical). Furthermore, no patient with involvement of a supraclavicular lymph node survived for 5 years (Spiro et al. 1974). Similar findings have been reported more recently by workers in the UK (Stell, Morton and Singh, 1983). In fairness, the failure of the present system to take node level into account has been acknowledged by the UICC which has recommended that such information should be recorded. It seems clear that the present system of staging is likely to be revised to take account of such information in the future.

Finally, the prognostic significance of extracapsular spread has recently been recognized. If patients are reassessed on the basis of a pathological examination of their block dissection specimens, it becomes apparent that spread outside the confines of the lymph nodes heralds statistically significant reduction in survival when compared with patients with either no nodes or with disease confined within nodes (Johnson et al. 1985). Furthermore, the disease-free interval between treatment and recurrence is shorter for patients with extracapsular spread.

It is true to say that no one clinical system is infallible. Nevertheless, with the advent of superior techniques - such as the possibility of delineating tumour metastases in the neck by using computerized tomography or, in the future, by means of monoclonal antibodies - it does seem likely that clinical staging preceding treatment will become more sophisticated. In this event, the drawbacks mentioned above will clearly recede; and when a partially subjective clinical impression can be objectively confirmed or refuted without recourse to the examination of a surgical specimen, a new clinical staging system will emerge. In the meantime, the present system with its inherent drawbacks will remain the one most commonly used for treatment planning and for communication between individuals and institutions.

Differential diagnosis of a suspected malignant cervical node

On account of the diversity of lesions, the neck is of special interest to several disciplines, including those of general surgery, otolaryngology, plastic surgery, maxillofacial surgery, and neurology. It is important, therefore, that a basic concept of the clinical significance of the cervical mass is fully appreciated, and that a general classification,
acceptable to all specialities, is made available. Most cervical masses fall into the following three groups:

(1) congenital and developmental

(2) inflammatory

(3) neoplastic of either primary or metastatic origin.

**Incidence**

Primary thyroid disease represents 50% of cervical masses. Within the other half, neoplastic disease accounts for roughly 70%, inflammatory disease for 6% and congenital abnormalities for 24%. Eighty-five per cent of the malignant group is composed of metastatic spread to cervical lymph nodes arising from primary epithelial cancers above the clavicle, from the skin, the salivary glands, the thyroid gland, the lower lip and from the mucous membrane areas of the upper air and food passages.

**Differential diagnosis**

A complete history and a full examination must be undertaken, with the appropriate blood films, tuberculin test and serology to help differentiate between the three major classifications. Common congenital lesions are thyroglossal vestiges and branchial and dermoid cysts, including cystic hygromata; while inflammatory lesions present as cervical lymphadenitis, tuberculous cervical adenitis, actinomycosis, sarcoidosis, non-specific inflammatory nodes of the acquired immune deficiency syndrome (AIDS) and salivary gland inflammation.

**Neoplastic lesions**

**Primary thyroid tumours**

Primary thyroid tumours are usually midline lesions which are identifiable within the thyroid gland. Parathyroid tumours are evident only from symptoms produced by metabolic disturbances.

**Salivary gland tumours**

Salivary gland tumours appear in the neck as an extension of either the tail of the parotid or of the submandibular glands.

**Tumours of neurogenous origin**

Neuroblastoma, neurofibromata, neurilemmomata and other tumours of neurogenous origin occur in the neck. These types of tumour are difficult to diagnose preoperatively, the exception being the multiple neurofibroma of von Recklinghausen's disease and its associated café-au-lait spots. Other primary mesenchymal tumours, such as lipomata, haemangiomata or
sarcomata may also be present in the neck and may be found in the anterior or posterior triangles.

**Carotid body tumour**

The paraganglioma or carotid body tumour is uncommon, but may appear at any age from 12 to 50 years, and is always associated with the carotid vessels. These tumours possess mobility in the lateral but not the vertical plane.

**Branchiogenic carcinoma**

A branchiogenic carcinoma is exceedingly rare. It is believed to arise from remnants of the branchiogenic apparatus, but its diagnosis is always in doubt and many cases represents a metastatic epithelial cancer arising from an occult primary.

**Metastatic spread**

A suspected metastatic node in the neck warrants a systemic search to locate a primary source, preceding the biopsy of the cervical mass. A small number of metastases involving supraclavicular or scalene nodes may emanate from a primary arising in the abdomen or lungs. Fine needle aspiration biopsy and incisional biopsy are discussed elsewhere in this chapter.

**Lymphomata**

The presence of a lymphoma in the neck is often only one clinical manifestation of a more wide-spread systemic disorder. Such a finding may represent either primary or secondary disease, and as most reports of lymphomata in the head and neck have failed to use modern methods for detection of occult disease below the diaphragm, the number of patients who fall into each of these categories remains obscure. In practical terms, the distinction is important because adequate management of the patient presenting with a lymphoma in the neck depends on establishing the degree of prior systemic spread so that the disease is accurately staged and the appropriate treatment given.

**Classification of lymphomata**

All lymphomata are malignant. Unfortunately, much disarray has existed in respect of the nomenclature to be adopted in the classification of these diseases; in particular, the classification of the heterogeneous group, which comprises the non-Hodgkin's lymphomata, remains unsatisfactory. Opinion is unanimous that Hodgkin's disease, in view of its distinctive histological features, should be separated from the other diseases. At present, the remaining lymphomata are classified as 'non-Hodgkin's' on the basis of the predominant cell type involved, and subdivided according to whether the neoplastic cells are arranged in nodules or in a diffuse manner, as nodular disposition is associated with a better prognosis.
Hodgkin's disease

The diagnosis of Hodgkin's disease can be made with certainty only when Reed-Sternberg cells are seen in lymph node biopsy material. Histologically, the disease is classified into four subtypes:

1. lymphocyte predominant
2. nodular sclerosing
3. mixed cellularity
4. lymphocyte depleted.

These subtypes also give an indication of prognosis, with lymphocyte predominant and nodular sclerosing types having a better prognosis than the other two categories. However, the prognostic significance of the histological subtype is of less relevance than the clinical stage of the disease which must, therefore, be accurately assessed.

Non-Hodgkin's lymphomata

The diagnosis for non-Hodgkin's lymphomata is usually made from biopsy material taken from the lymph nodes, but occasionally an extranodal site or a bone marrow examination will reveal the nature of the disease. Although histological type is currently the basis for classification of these lymphomata, immunological characterization is increasing in importance. Unlike Hodgkin's disease, the histological characteristics of a non-Hodgkin's lymphoma are of paramount importance to prognosis, and this aspect outweighs the significance of anatomical staging. The original Rappaport classification of these diseases described the basic cell type as lymphocytic or histiocytic, but it is now realized that the latter term is a misnomer and that these cells do in fact represent proliferating lymphocytes.

Since the 1960s, several new classifications have been developed on the basis of the functional and immunological characteristics of the basic cell type. One of the most recent is as follows.

Low grade

1. Malignant lymphoma, small lymphocytic
2. Malignant lymphoma, follicular, small cleaved cell
3. Malignant lymphoma, follicular, mixed small cleaved and large cell.

Intermediate grade

1. Malignant lymphoma, follicular, large cell
2. Malignant lymphoma, diffuse, small cleaved cell
3. Malignant lymphoma, diffuse, mixed small and large cell
4. Malignant lymphoma, diffuse, large cell.
High grade

(1) Malignant lymphoma, large cell, immunoblastic
(2) Malignant lymphoma, lymphoblastic
(3) Malignant lymphoma, small non-cleaved cell.

Miscellaneous

(1) Composite malignant lymphoma
(2) Mycosis fungoides
(3) Extramedullary plasmacytoma
(4) Unclassifiable.

Whichever classification of non-Hodgkin's lymphoma is used, the important prognostic factors are the subdivisions into low and high grade tumours. Features which favour a low grade rather than a high grade tumour are:

(1) a nodular rather than a diffuse histological pattern
(2) a lymphocytic rather than a histiocytic morphology
(3) well-differentiated rather than poorly differentiated cells.

Clinical features

Hodgkin's disease

Hodgkin's disease usually presents with a painless lymph node enlargement in either the neck, axillae or inguinal regions. The question of whether such a presentation is accompanied by systemic symptoms, such as sweating, weight loss and fever, is of importance. A chest X-ray commonly shows either hilar or mediastinal lymphadenopathy. Bone marrow invasion may lead to signs of pancytopenia, although in most instances a trephine biopsy to establish involvement will be necessary. Very occasionally, the enlarged nodes will give rise to complications resulting from pressure, such as obstructive jaundice or spinal cord compression. Central nervous system involvement is rare.

Non-Hodgkin's lymphomata

The presentation of non-Hodgkin's lymphomata is similar to Hodgkin's disease, but the pattern of the disease is dissimilar, with the extranodal sites being involved much more frequently. Such sites include the central nervous system, the gastrointestinal tract, the nasopharynx, bone, soft tissues and the thyroid gland. True localized disease is therefore much less common than in Hodgkin's disease, and it has been estimated that over 50% of these patients have extranodal involvement at presentation.

Staging

Staging follows the Ann Arbor classification (Table 17.3) and this is now universally applied for both non-Hodgkin's and Hodgkin's lymphomata; clinical as well as pathological criteria are used. The staging is then further subdivided into cases without systemic symptoms.
(substage A), and those cases with a weight loss greater than 10% during the 6 months preceding presentation, or with unexplained fever or sweating (substage B). Pruritus is no longer regarded as a B symptom.

Table 17.3 Ann Arbor classification of lymphoma

**Stage I**

Disease limited to one lymph node region (I) or to a single extralymphatic organ or site (IE)

**Stage II**

Disease in more than two anatomical regions on one side of the diaphragm (II) or localized involvement of extralymphatic organs or sites, and in one or more lymph node regions on the same side of the diaphragm (IIE)

**Stage III**

Disease on both sides of the diaphragm (III) with or without involvement of the spleen (IIIS) or localized extralymphatic organ or site (IIIE)

**Stage IV**

Diffuse or disseminated disease involving one or more extralymphatic organs or tissue (for example, liver, marrow, pleura, lung, bone and skin). Involvement of extranodal sites (for example, bone marrow, liver or skin).

**Investigation**

The investigation of patients with lymphoma requires a full history and clinical examination, together with a lymph node biopsy to establish the nature of the disease. Additional investigations should include a blood differential count, sedimentation rate, and liver function tests. The serum uric acid should also be estimated as hyperuricaemia may result from the early stages of treatment. Chest X-rays to demonstrate hilar involvement, mediastinal disease and parenchymal lung disease are important because such features carry a poor prognosis. Bone marrow trephine and an examination of a bone marrow smear are necessary for staging. Additional tests may include excretion urography, bone X-rays and ultrasonography. In addition, lymphangiography or computerized tomographic scanning have allowed a more comprehensive and non-invasive assessment of patients. However, in some cases with Hodgkin's disease, in spite of an acknowledged morbidity and finite but small mortality, staging laparotomies are still required in order to establish whether the spleen is involved. This procedure also allows an accurate sampling of lymph node sites along the iliac vessels and aorta, as well as wedge biopsy of the liver. In female patients, positioning of the ovaries behind the uterus is also necessary to protect them from subsequent irradiation.
In the investigation of a patient with non-Hodgkin's disease (where prognosis is less dependent on stage), the investigations are less comprehensive and staging laparotomy is not required.

**Treatment**

**Hodgkin's disease**

The treatment of Hodgkin's disease comprises radiotherapy for the treatment of localized disease and chemotherapy for a more extensive spread. Pathological stages I and II are usually treated by means of radiotherapy.

Because adjacent nodes are often involved, the treatment is usually extended to include related lymph node regions. For disease above the diaphragm, a 'mantle area' covering the cervical, clavicular, axillary, mediastinal and hilar lymph node areas is used. In contrast, stages I and II disease below the diaphragm is treated using an 'inverted-Y' to cover the para-aortic nodes from the level of the diaphragm to the bifurcation of the aorta, together with the pelvic and inguinal regions. After treatment with radiotherapy for stages I and II disease, over 80% of patients are alive and free of disease after 10 years. The only patients in this group who do badly on this regimen are those with large mediastinal masses at presentation; because the relapse rate in these patients is high, some centres now use chemotherapy for this subgroup.

The treatment of stage III disease has not yet been satisfactorily defined. A number of centres have reported good results using 'mantle' and 'inverted-Y' radiation (total nodal irradiation), but others have reported relapse rates as high as 50%; therefore, some workers combine radiotherapy with chemotherapy during the initial phase of treatment. As an alternative, chemotherapy may be given at the time of relapse.

The treatment of stage IV disease is now by means of combination chemotherapy. Various combinations have been used, including the original schedule of MOPP (nitrogen mustard, vincristine, procarbazine and prednisolone) and MVPP (nitrogen mustard, vinblastine, procarbazine and prednisolone). Other combinations have also been tested in an attempt to reduce toxicity and increase efficiency, but none has proved to be inherently better than the original treatment protocol. It is an unfortunate fact that even with the use of such agents, the death rate among patients with advanced Hodgkin's disease remains as high as 50%; and although some who suffer a relapse after one year will achieve a second remission with further treatment, those who have a relapse early on, or who fail to achieve a remission, have a poor prognosis.

**Non-Hodgkin's lymphomata**

There is little agreement about the treatment of non-Hodgkin's lymphomata. Most patients have widespread disease at presentation and are therefore unsuitable for radiotherapy. However, as these tumours are highly radiosensitive, those patients who have low-grade malignancies of clinical stages I and II are usually treated with radiotherapy alone. The results of such treatment are not as good as in comparable patients with Hodgkin's disease but, nevertheless, 50% of appropriately selected patients will be alive and disease-free at 5 years.
Chemotherapy in combination with radiotherapy has also been used in the treatment of low-grade tumours but its role remains unclear. In the more widespread diseases (stages III and IV), the treatment depends on histological classification. Patients with low-grade tumours often follow a protracted course with few symptoms. There is little evidence to suggest that cytotoxic agents improve the chance of survival and their use therefore seems unjustified. If, however, symptoms should intervene, low-dose alkylating agents will induce a response in most cases. In contrast, the prognosis pertaining to high-grade malignancies has been dramatically improved using aggressive combination chemotherapy, and the most commonly used regimens include cyclophosphamide, doxorubicin, vincristine and prednisolone. Treatment with these agents leads to remission in almost 100% of patients with stage III disease, and in just under 50% of those with stage IV disease.

The occult primary

A patient presenting with a metastatic cervical malignancy from an unknown primary source is said to have an occult primary. It is not uncommon for a metastatic node to produce minimal symptoms and to have an indefinite pattern of development which is not accompanied by any significant history. One-third of these patients have additional symptoms including pain, weight loss, malaise and fatigue, but because of the lack of significance attached to these symptoms, less than 20% of patients with a cervical mass present in the early stage of the disease. In a series of patients reported by Winegar and Griffen (1973), the average time from the first sign of a mass presenting in the neck to the time of the patient's initial evaluation was 5.1 months.

This interval is comparable with those reported by other authors. The incidence of tumour types is listed in Table 17.4.

Table 17.4 Incidence of tumour types

<table>
<thead>
<tr>
<th>Tissue type</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Squamous cell carcinoma</td>
<td>41</td>
</tr>
<tr>
<td>Undifferentiated</td>
<td>36</td>
</tr>
<tr>
<td>Adenocarcinoma</td>
<td>17</td>
</tr>
<tr>
<td>Lymphoepithelioma</td>
<td>6</td>
</tr>
<tr>
<td>Melanoma</td>
<td>3</td>
</tr>
<tr>
<td>Leiomyosarcoma</td>
<td>1</td>
</tr>
<tr>
<td>Lymphosarcoma</td>
<td>1</td>
</tr>
<tr>
<td>Chondrosarcoma</td>
<td>1</td>
</tr>
</tbody>
</table>

Current methods of investigation are unable to detect tumours in the body until about 1 g of tumour, consisting of approximately $10^9$ cells, is present. Generally speaking, as a patient is likely to die when the total tumour burden reaches between $10^{12}$ and $10^{13}$ cells (that is 1-10 kg tumour weight), it follows that the tumour will already be at least two-thirds of the way through its life span before it is detected. Using this criterion, most tumours in man must be late or at an advanced stage at the time of initial presentation.
Three points should always be kept in mind when examining a neck mass. First, most persistent lumps in the neck in the adult are the result of malignant disease. Second, most malignant tumours in the neck are metastatic and if the presence of a goitre is excluded, the frequency of occult carcinoma in patients presenting with a metastatic node is about 5%. Third, most metastases in the neck originate from primary sites in the head and neck.

Martin and Romieu (1952) reported the following primary sites which should always be under suspicion as a source for a metastatic node in the cervical region:

- nasopharynx
- tonsil
- base of tongue
- thyroid
- hypopharynx.

It is unfortunate that the three most common sites which give rise to occult cervical metastases - the nasopharynx, the tonsil and the base of the tongue - cannot be examined easily by the general physician using a simple tongue depressor. It is important that attention to the cervical mass does not distract from the search for the primary tumour, for any delay in its discovery may allow it to advance to an incurable stage.

**Premature biopsy**

Premature biopsy of a suspicious cervical mass is a common and usually serious error of management, especially if the metastatic node exhibits extracapsular rupture. It is an excellent principle that any surgeon who violates the skin of the neck to obtain a histological diagnosis by either an incisional or an excisional biopsy should be prepared to proceed with a radical neck dissection if this is indicated. Injudicious removal of a metastatic node causes surgical scarring which may prevent or preclude subsequent surgical dissection of the lymph node bearing area. Above all, this procedure may delay the proper treatment of the primary tumour.

**Search for the primary tumour**

When a patient presents with a cervical mass which is considered to be a metastatic node, the search for the primary tumor should follow an established sequence of investigation, although the order of investigations may have to be tailored to the individual. Following the taking of a careful history and a general physical examination, the buccal cavity, the base of tongue, the hypopharynx and the nasopharynx should be carefully inspected. Examination is further facilitated by palpation, particularly in areas where this can be carried out bimanually. Palpation is an important part of the examination and Conley (1971) has emphasized that palpation should be performed during examination of the head and neck wherever a finer may reach, likening its value to that of having the added facility of a third eye.
**Investigations**

Investigations should include the following:

1. Sinus and chest X-rays, contrast studies of the upper and lower gastrointestinal tract if the metastatic tumor is an adenocarcinoma.
2. Triple endoscopy: this includes laryngoscopy, bronchoscopy and oesophagoscopy.
3. In some instances blind biopsies of the lymphoid tissue from Waldeyer's ring - in particular, the postnasal space, the tonsils and also the lymphoid tissue of the posterior third of the tongue.
4. The use of specialized techniques to aid identification of an undiagnosed mass in the neck (see below):
   - CT scanning
   - Radiolabelled antibodies
   - Fine needle aspiration cytology.

Only if, following of a full diagnostic evaluation, still no evidence of a primary tumour has been found does the patient fall into the occult primary tumour category.

**Management**

The survival time for a patient presenting with an occult tumour, in whom no primary is ever found in spite of full investigation, varies considerably with different reports. Jesse and Neff (1966) reported a 34% survival (absolute) in 127 patients, but Winegar and Griffen (1973) found only 15% (16 out of 106 patients) who survived a 5-year period. It must be postulated in such cases that the primary cancer is either so small that it cannot be discovered or that it has regressed spontaneously. Alternatively, if radiation therapy has been used for the treatment of the neck metastases, it is possible that the primary tumour has been sterilized by the irradiation. Although it is an established principle of radical neck dissection that the operation should not be performed unless the primary tumour has been controlled, it is impractical to follow this edict where the primary site is unknown. However, a radical neck dissection will still be necessary to control the metastatic disease, and the patient is more likely to die from the secondary disease than from the occult primary tumour even if it subsequently becomes clinically evident.

The options open for treatment of a patient with an occult tumour will have to be carefully considered. Surgery alone should give about a 40% 5-year cure rate. Combination therapy of surgery and radiotherapy may be more effective than irradiation alone, but it is probably wiser to employ surgery initially and to save irradiation therapy until later for proven residual or recurrent disease.

Jesse and Fletcher (1977) showed in their series that surgery alone gave the patient with a single node in the upper part of the neck a 50% chance of a cure. It is preferable, therefore, that surgery should be employed as the treatment of choice in the case of an occult
primary if the lesion appears resectable. However, for patients with multiple enlarged unilateral nodes, radical neck dissection combined with irradiation therapy is probably warranted, for without adjunctive radiotherapy this group is more liable to incur a recurrence.

**Follow-up**

Careful and regular follow-up after a patient has been treated for an occult primary is mandatory. The histology of a metastatic node may or may not be helpful in deciding the areas to be closely examined. In the case of metastasis from squamous cell carcinoma, examination of the ears, nasal fossae, postnasal space and pharynx should be carried out at regular intervals of 3-4 months. Periodic endoscopy with selected biopsies of the nasopharynx, base of tongue and tonsil area should also be performed. If the histology of the node shows a glandular origin, investigations should be carried out at 6-monthly intervals to screen the upper and lower gastrointestinal tract, and should include mammography in women, CT scanning of the thyroid and salivary glands and occasionally examination of the prostatic fluid in men.

**Computerized tomography of the soft tissues of the neck**

Computerized tomographic (CT) scanning has been used in the preoperative examination of patients with head and neck cancer. Mancuso, Maceri and Rice (1981) were the first to report on the use of CT scanning in the evaluation of cervical node metastasis. They found CT scanning to be accurate in 21 out of 23 patients when correlated with pathological specimens. Such early reports on the use of CT scanning for cervical lymphadenopathy were encouraging, and the accuracy of CT scanning in the detection of nodal metastasis has been further established by Friedman et al (1984) who compared 50 consecutive patients who had undergone radical neck dissection and preoperative CT scanning. The clinical staging of the neck, the CT scanning diagnosis and the pathological findings were correlated and the findings showed that the overall accuracy of CT scanning diagnosis was 90%. Through this comparison, the CT scan was shown to be superior to clinical examination and particularly effective in detecting occult metastasis. As the technique has proved to be diagnostically accurate, CT scans can provide important clinical information for treatment and prognosis.

At present, nodal disease is routinely staged by clinical examination and reports of the accuracy of such examination vary widely. Beahrs and Barber (1962) found a 30% false positive rate and a 39.9% false negative rate following clinical examination, which underlines the potential benefits of developing highly accurate CT scanning for cervical lymphadenopathy.

Tomographic studies of patients with clinically palpable nodes can also reveal the integrity of the vasculature. Thrombosis of the jugular vein has been diagnosed but involvement of the carotid artery is more difficult to assess accurately, although the findings by CT scanning in respect of a normal carotid artery rule out arterial involvement by nodal disease.
The use of radiolabelled antibodies in the detection of squamous cell tumours

The radioimmune detection of tumours by external emission scanning has attracted interest recently with the introduction of subtraction scanning techniques. Head and neck squamous cell carcinomata are not associated with tumour-specific antigens, but the serum carcinoembryonic antigen (CEA) levels are frequently elevated and CEA has been demonstrated on the tumour cell surfaces (Toto, 1979). Tranter et al (1984) successfully used a radiolabelled scanning technique to locate accurately several areas of tumour tissue arising from head and neck squamous cell carcinoma. The resolution is probably comparable with other antibody scans, but as the neck is a good area for radionucleotide imaging, considerable improvement was anticipated with new detection systems. In this study, masses greater than 2.0 cm were detected in several parts of the body. The technique was thought to be suitable for the detection of occult primary lesions and in the assessment of lymph node spread in metastatic sites, and at the same time in determining whether an enlarged lymph node containing tumour or was merely exhibiting reactive changes. Two advantages of this type of scan over other scanning techniques are that tumour is positively identified and, provided that the facilities are available, the procedure is simple to undertake.

Fine needle aspiration cytology of cervical lymph nodes

The examination of cells for diagnostic purposes originated in the nineteenth century and was initially applied in haematological diseases. Since then it has become accepted practice to use this technique in the assessment of haematological diseases, including the diagnosis of primary and secondary malignancies of the bone marrow and in the analysis of effusions.

The use of aspiration cytology in the diagnosis of solid tumours has remained controversial, although evidence in support of this method was published as long ago as the 1930s. A real stimulus to its further development was provided by Papanicolau, who demonstrated that malignant changes were present on smears obtained from secretions and who is remembered for the development of the Pap smear test used for the detection of uterine malignancy (Papanicolau and Trant, 1943). More recently, needle aspiration cytology has developed a wider application and the cytological features associated with a variety of different malignancies have now been catalogued.

The technique of aspiration cytology is simple, quick and virtually painless. Infiltration of the skin with a simple short-acting anaesthetic (such as lignocaine 1%) may be used before insertion of the needle for tumour aspiration, but it is not mandatory and may 'blur' the outline of a small lesion. The target is then precisely defined and the area punctured with a 24-gauge needle attached to a 10 or 20 mL syringe; if a local anaesthetic has been used, it is preferable to ensure a separate track for the aspiration in order to avoid any artefact which may have been induced. Negative pressure is then applied to the plunger of the syringe and the needle tip passed to and fro in the lesion to be sampled. The cells and tissue fluid thus obtained are then expelled and smeared on the surface of a glass slide; the smear is then air dried before staining and subsequent examination under the microscope. Variations in the technique described are common, but for a more comprehensive discussion the reader is referred to a recent review (Friedman et al, 1983).
Lymph nodes and neck masses were among the first targets selected for demonstrating
the use of the technique of aspiration cytology. Multiple nodes can be sampled during the
same session and an accuracy rate of around 95% has been claimed for epithelial tumours.

**Cervical lymph node biopsy**

It is a widely accepted principle that to take a biopsy of a metastatic cervical node
mass instead of searching for a primary neoplastic source is totally incorrect. The information
obtained is, at best, only the cell of origin and its degree of differentiation and, at worst,
reveals the existence of an anaplastic lesion. In neither case does the result indicate the site
and/or extent of the primary tumour. In addition, the incidence of wound sepsis and fungation
of the tumour through the skin of the neck is higher in patients who have undergone such a
biopsy, and McGuirt and McCabe (1980) reported an increase in the incidence of distant
metastases in patients who had undergone a cervical node biopsy before definitive treatment.

**The significance of a neck node to radical neck dissection**

**The effect of metastatic node on prognosis**

In 50% of cases where one lymph node is positive on palpation there will be several
other nodes which will be microscopically positive.

Whenever evidence of cervical metastasis is lacking, the general prognosis for a
specific primary tumour is obviously more favourable. However, cancers of the nasopharynx
and tonsil, as well as malignant melanoma, show a particularly poor prognosis when a node
is involved. Regardless of the site of the primary tumour, only 33-40% of patients will
survive longer than the 5-year period if cervical metastases are present at the time of
dissection.

**Chronological appearance of significant lymph nodes**

If positive lymph nodes in the neck become apparent soon after the presentation of the
primary tumour, an attempt to control the metastatic disease through a radical neck dissection
will be less effective than in those cases where there is a delay in the spread of the metastatic
tumour after the appearance of the primary. In the former cases, the 5-year survival rate falls
below 40% (Pack and Ariel, 1964).

**The operative techniques of neck dissection**

**The classical radical neck dissection**

In many cases, a primary head and neck tumour will respond to treatment with
radiotherapy, but lymph nodes invaded by squamous carcinoma are less likely to do so and
must be treated surgically. Operation may be required to remove the nodes alone or can be
performed in continuity with removal of the primary tumour; without doubt, this is best
achieved by a classical radical neck dissection. The only indication for the local removal of
palpable carcinomatous cervical nodes is in the treatment of papillary carcinoma of the
thyroid which, during its long natural history, is slow to break out of the capsule of the
thyroid gland and does not follow the anticipated lymphatic channels taken by other head and neck carcinomata.

The surgical removal of metastatic lymph nodes by block dissection of the neck does not appear to increase the risk of general metastasis. An extended dissection into the mediastinum does not greatly increase the cure rate. It is therefore fair to say that the classical radical neck dissection of involved lymph nodes continues to offer the best possible means of control. The failure to remove all involved lymph nodes together with the primary tumour must decrease the survival rate; and, as such, a partial neck dissection can have only a limited role.

When a radical neck dissection is carried out as part of a combined procedure, it is most important that as much tissue continuity as possible is retained between the primary site and the lymphatic field. Therefore in a total laryngectomy, the neck dissection can be left attached along the whole length of the larynx to include the superior and inferior pedicles, whereas when a laryngopharyngectomy is performed, the pedicle consists of the whole length of the pharynx. In oral cancers, the specimen should be left attached along the lower border of the mandible and should include the inner layer of the periosteum. Tumours of the oropharynx drain through a pedicle to the upper deep cervical nodes, so the specimen should remain attached near the tail of the parotid.

**Incisions**

The factors which influence the planning of incisions in the neck can be divided into two main groups:

1. the site of metastatic cancer, the nature and position of the primary, and the size, mobility and anatomical level of the metastases

2. whether or not the patient has had preoperative irradiation, with consideration being given to the size of the port and the dosage given. It must be remembered that any patient who has received over 6,000 cGy of radiation will have suffered marked damage to the skin and subcutaneous structures which means that any flap will not be as viable or heal as quickly as in a non-irradiated neck.

The first approach was described by Crile in 1906. The Y-type of incision which he used remains an excellent incision, having proved its value over an 80-year period. MacFee's double horizontal incision (MacFee, 1960) which is perhaps the most aesthetic, and which is especially suitable following irradiation, does not give such good exposure. The Martin approach is also useful and was designed to produce smaller flaps in order to prevent necrosis associated with radiotherapy (Marin, 1941). The flap described by Schobinger was placed below the horizontal ramus of the mandible and extended to the top of the mastoid and over the border of the trapezius muscle to the clavicle, giving a large single lateral cervical neck flap with excellent exposure; however, it had the disadvantage that its upper part was subject to necrosis. Conley added an additional incision at the posterior curve of the Schobinger flap, to include the parotid gland and the nape of the neck. Other flaps based either anteriorly or posteriorly have not proved as reliable, and consequently have been discarded. Single flaps are not acceptable in patients who have had severe preoperative irradiation resulting in
extensive fibrosis, atrophy and telangiectasia of the skin, as all of these factors delay healing. This has been known to occur with the Martin flap and, as such, represents a serious disadvantage of the approach. The horizontal T-incision, which produces large superior and inferior flaps, is satisfactory when a combined neck dissection with the contralateral side is contemplated, but though should always be given to the question of whether an anteriorly developed flap will survive if a preoperative tracheostomy has been performed, for the blood supply will have been diminished by the tracheostomy incision. If there is any doubt about the viability of irradiated skin, or if the superficial layers have been invaded by cancer, the area of skin affected should be sacrificed and the deficit compensated for by a transposition flap.

Rogers and Freeland (1976) when investigating the arterial vasculature of cervical skin flaps by perfusion experiments in cadaveric flaps concluded that: (1) the platysma muscle should be included in the flap to conserve the vascular network; (2) the horizontal limb of an incision should lie in the watershed between the carotid and subclavian arterial systems; and (3) the upper flap should be larger than the lower flap.

**Superficial dissection**

In a routine approach using a modified Y-shaped Crile incision, the platysma muscle should be included in the flap. Following the raising of the superior skin flap, the great auricular nerve and the superior part of the sternomastoid muscle will be identified. In the anterior part of the neck there is an absence of platysma muscle separating the skin from the deeper structures in the submandibular triangle, and care must therefore be taken to preserve the mandibular branch of the facial nerve when raising the superior flap.

**Lower end of the jugular vein**

In the lower part of the neck, the supraclavicular nerves and vessels will be seen and a tunnel superficial to the anterior belly of the omohyoid will lead directly under the sternomastoid muscle to the carotid sheath. The manubrial and clavicular heads of the sternomastoid will have to be divided to expose the inferior segment of the internal jugular vein. When handling any of the large veins of the neck, care has to be taken to prevent an air embolus. In addition, the lower end of the jugular vein must be ligated at the earliest opportunity to reduce the incidence of systemic metastases caused by tumour embolus.

**The jugular lymph trunks**

On the left side, the thoracic duct ascends medially to the internal jugular vein and passes posteriorly and laterally to the vein before descending to enter the junction of the internal jugular and subclavian veins. Its position as well as the number of its tributaries may vary. In 50% of cases, the thoracic duct will be identified on the right side.

**Supraclavicular dissection**

By following the superior border of the clavicle laterally to the trapezius muscle, and after ligating the external jugular vein above, access will be gained to the posterior belly of the omohyoid muscle which can be cut and allowed to retract into the specimen. The brachial
plexus can now be identified with the phrenic nerve arising from anterior primary rami between the levels of C3 and C5. It runs under a fibrofatty pad on the scalenus anterior muscle. The three branches of the thyrocervical trunk will also be identified.

**Spinal accessory nerve and posterior dissection**

The splenius capitis muscle is identified by following the anterior border of the trapezius. The anterior and posterior branches of the transverse cervical artery, including the veins from the nape of the neck, will need to be ligated in the supraclavicular fossa.

The branches of the third cervical nerve may join the accessory nerve as it crosses the posterior triangle before its insertion into the trapezius muscle; these branches from the cervical plexus are now considered to contribute as motor nerves to the trapezius muscle and, if they can be preserved, they will help to prevent shoulder drop when the main spinal accessory nerve is divided. A shoulder drop nearly always occurs following a classical radical neck dissection, but whether this is an impediment or a serious handicap varies in different patients. By the earlier identification of the brachial plexus, the correct layer for preservation of the phrenic nerve is identified and then, by following the nerve pedicles of C2, C3 and C4 in turn, access to the posterior surface of the carotid sheath is ensured. This posterior approach gives access to the whole length of the carotid and deep jugular system.

The vagus nerve is identified lying between the internal jugular vein and the common carotid artery, and the descending branch of the ansa hypoglossi nerve, supplying the strap muscles, will be seen lying on the superior surface of the carotid sheath. The hypoglossal nerve is identified crossing the carotid artery above the bifurcation and the dissection is continued to expose the region of the axis and mastoid process as far as the digastric groove. The fibrous origin of the sternomastoid muscle is separated from the mastoid process, and the posterior belly of the diaphragm arising from the digastric groove is identified; a tunnel is developed along the muscle to the hyoid bone. The hypoglossal nerve will once again come into view between the internal carotid artery and vagus nerve; behind this lies the superior cervical ganglion.

**Anterior dissection and submandibular triangle**

The anterior part of the dissection is continued, to the hyoid bone, by dividing or ligating the ansa hypoglossi and the plexus of veins coming from the pharynx. Dissection exposes the anterior belly of the digastric and the lateral border of the mylohyoid muscle. Between 1.0 and 2.0 cm below the lower border of the mandible, the mandibular branch of the facial nerve is again identified, together with the facial vessels and related facial lymph nodes. The nerve should be preserved provided that there is no cancer in the anterior and posterior nodes associated with the facial artery. By retracting the posterior border of the mylohyoid, the lingual nerve is demonstrated. On elevation of the nerve, the lingual extension of the submandibular gland and duct is found so that the duct can be ligated as far forward as possible to prevent infection developing in any duct remnant. The deep part of the facial artery should be identified and ligated where it emerges above the posterior belly of the digastric muscle.
Parotid gland

If there is any tumour involvement of the parotid gland, the entire gland should be removed in conjunction with the facial nerve; usually only the most inferior part of the gland needs to be removed. The posterior facial or retromandibular vein will need to be ligated where it lies in relation to the posterior belly of the digastric muscle. At this stage, it is important that the subdigastric area is examined for metastatic spread. The stylohyoid muscle can be identified immediately above the digastric tendon as it passes to be inserted into the lesser cornu of the hyoid bone. Both muscles should be included in the specimen if there is metastatic disease in this area. The occipital artery will require ligation once the posterior belly of the digastric has been divided. It must be remembered that the facial nerve lies adjacent to the anterosuperior border of the posterior belly of the digastric muscle.

Carotid artery protection

The levator scapulae muscle, or part of the trapezius muscle, can be used if there is concern about cover for the carotid artery or bulb in heavily irradiated neck. These muscles can also be used to provide cover for a pharyngeal fistula.

The upper end of the internal jugular vein

The upper end of the internal jugular vein is identified and divided as it lies deep to the posterior belly of the digastric muscle. The specimen can then be removed along with the metastatic nodes which remain attached to the deep surface of the sternomastoid muscle and the internal jugular vein. The wound should be drained through two continuous suction drains. Healing is normally complete in 7-10 days.

Bilateral neck dissection

The presence of stage N3 bilateral neck glands is a serious and poor prognostic sign and, in such instances, the survival rate falls to around 5%. For example, the 5-year survival rate in cases of carcinoma of the mouth with bilateral neck glands is only 9% (Spiro et al, 1974), and in other sites, such as the oropharynx and hypopharynx, it may be as low as 4% (Lederman, 1967).

More than 90% of patients who have bilateral lymph nodes have primary cancers situated in midline structures such as the mouth, the oropharynx and the hypopharynx, and all these sites have an extremely poor prognosis. In most cases, surgery probably does not influence the natural history of the disease, but Stell (1978) indicated that patients with a supraglottic carcinoma and bilateral glands are an exception and can still have a reasonable prognosis.

In spite of this low survival rate there are many reports in the literature of synchronous bilateral neck dissection, the first being by Le Clerc and Roy in France in 1932. In the main, patients presenting for treatment who have advanced cancer of the head and neck with evidence of bilateral node involvement are felt to be beyond hope. Palliative irradiation may be offered to the patient, but there is little hope of success in respect of prolongation of life.
The same is true of those patients with advanced disease who have been treated by a neck dissection on one side, combined with an attempt to control the other side using radiation.

The procedure of bilateral synchronous radical neck dissection carries a significant morbidity, and a mortality rate of 3.4% (Ballantyne and Jackson, 1982). Many of the complications of bilateral radical neck dissection can be reduced by staging the two sides with an interval of 6 weeks or longer, but a tracheostomy is occasionally required at the time of performing the second stage of the procedure.

When a bilateral synchronous neck dissection is carried out, it should be appreciated that the operations on each side of the neck are not usually identical, and that a modified neck dissection is nearly always required on the opposite side to preserve the internal jugular vein.

If both internal jugular veins are to be ligated, it is important to understand the effect on the venous drainage of the head and neck. The anatomy of the venous circulation of the cranial cavity must be considered. In addition to the internal and external jugular veins, the alternative drainage systems in order of importance are:

1. the external and internal vertebral plexus with their communicating veins
2. the occipital vein
3. the posterior jugular vein
4. the deep cervical vein
5. the collecting veins of the posterior cervical region and pharyngo-oesophageal and pterygoid plexus.

The total cross-section of the vertebral venous system alone probably exceeds that of the jugular veins. A rich communication also exists between intracranial and extracranial structures of the head via emissary veins. In these, the flow is mainly towards the extracranial areas, thus reducing venous congestion in the brain when the jugular circulation is blocked.

Nevertheless, the most serious complication after bilateral radical neck dissection is that of increased intracranial pressure. Although cerebrospinal fluid pressure is to some extent influenced by the position of the head, the tying of one internal jugular vein in a radical neck dissection produces a threefold increase in intracranial pressure, and when the second side is tied a fivefold increase in pressure results.

De Vries, Balm and Tiwari (1986) published details of a case where a patient developed prolonged papilloedema as a result of increased cerebrospinal fluid pressure following a staged bilateral radical neck dissection. The prognosis for vision is usually good in such patients, but permanent visual impairment remains a serious complication.

The critical period for a patient with regard to the development of a raised cerebrospinal fluid pressure following a synchronous bilateral neck dissection is during the first 12 hours. During the 8-10 days after the operation, the intracranial pressure tends to fall, although it does not revert to a normal level.

The performance of a bilateral neck dissection undoubtedly increases the incidence of oropharyngeal fistula, particularly in those patients who have had previous irradiation; in
addition, there is the increased risk of postoperative wound complications resulting from sepsis and slough of skin flaps. As the survival rate is around 5% for patients presenting with bilateral neck metastasis, the indications for bilateral staged or synchronous neck dissections are obviously limited, but this should not be confused with the treatment of and the prognosis for a patient presenting with a metastatic node on the second side at a later stage. The prognosis here is totally different from that of a patient who presents with bilateral synchronous neck glands, and surgical treatment may be warranted in the case of the former group.

**Radical neck dissection combined with superior mediastinum dissection**

The majority of head and neck tumours metastasize to the jugular chain, but there are some tumours, notably those of the cervical oesophagus, the subglottic space and the thyroid gland, which are known to metastasize preferentially to lymph nodes in the mediastinum. Sissons, Edison and Bytell (1975) described an operation for dissecting the superior mediastinum based on a method given by Grillo (1966). This operation was developed to deal with recurrences affecting the stoma following a laryngectomy, and requires the development of two bipedicled upper chest flaps. These flaps are designed to protect the structures in the upper mediastinum which are exposed when the manubrium and the medial end of both clavicles are removed. According to Sissons' description of the operation, the first stage is to delay a myocutaneous flap of the pectoral muscle which, in the second stage of the operation, is then laid over the mediastinum to protect the great vessels. Although the operation has not been performed a sufficient number of times for it to be evaluated, there have been reports of success using this technique.

**Functional neck dissection**

In a 1979 report of a large series of 788 consecutive cases of radical neck dissection, nearly one-fifth of the patients experienced a major complication, with an overall morbidity rate of 50.8% (McGuirt and McCabe, 1980). As 20.4% of these patients had regional recurrences, it is not surprising that consideration has been given to the technique of 'functional neck dissection'.

The elegant technique of functional neck dissection was described by Bocca and Pignataro (1967) who removed a 'package' of lymph nodes and their vessels, but retained the internal jugular vein, accessory nerve and sternomastoid muscle. On the basis of the anatomical and surgical contributions of Suarez (1963), Bocca (1966) modified the traditional neck dissection by radically revising the historical concepts identified with the surgical treatment of regional metastasis. As a staunch opponent of conservative nodal stripping, Bocca set out to show the effectiveness of the surgical technique which he called 'the functional neck dissection'. The arguments he put forward to support his technique were as follows:

1. functional neck dissection avoids the unjustified consequences of the traditional neck dissection, including dropped shoulder, skeletal pain, the limitations of neck and limb motion and widespread cutaneous anaesthesia

2. bilateral dissection may be performed simultaneously without danger of intracranial venous congestion
(3) when the preferred treatment of the primary tumour requires combined surgery and radiation, the functional neck dissection may alter the decision for neck irradiation in patients with disease staged as N0, and may provide a reasonable alternative to radical radiotherapy of the neck. It was accepted that the presence of stage N3 fixed nodes represents an absolute contraindication to functional neck dissection.

A weakness of this operation has been exposed by McKelvie (1974) who, in his cedar wood cleared neck specimens, demonstrated that microscopically involved nodes could be identified invading the adventitia of the jugular vein. It is clear that these nodes would not have been removed in a functional neck operation.

Therefore, in spite of the elegant nature of a functional neck dissection in experienced hands, the operation has not been universally adopted and, where it might most reasonably be used, most surgeons still prefer to use radiotherapy as an alternative.

**Prophylactic/elective neck dissection**

A prophylactic radical neck dissection may be defined as a radical neck dissection performed on a neck evaluated by clinical examination as negative for metastatic cancer. The term elective radical neck dissection extends this concept to embrace neck dissection in a clinically negative neck combined with excision of the primary tumour and, as such, places emphasis on the value of the removal of clinically undetected metastatic nodes. Such an approach has the advantage of avoiding the possibility of future surgery under less favourable conditions. It should be emphasized that a prophylactic operation will not benefit patients with advanced primary disease or those with a primary that has metastasized across the midline to give contralateral cervical nodes.

**Conservation neck dissection**

Numerous attempts have been made in the past to modify the classical radical neck dissection by conserving structures that are thought not to be involved in the malignant process or the removal of which would significantly increase the morbidity of the patient after radical operation. The structures which have consistently come under review have been the internal jugular vein, the spinal accessory nerve and the sternomastoid muscle. This adaptation of the classical operation has been defined as a conservation neck dissection, but it is fraught with risk.

Skolnik and Deutsch (1983) found no statistically significant difference in the rate of neck recurrence between a group of 422 patients who underwent radical neck dissection and the 269 patients who underwent conservation surgery. These findings have been confirmed by other authors, notably Lingeman et al (1977), Molinara et al (1980), Deutsch et al (1985), all of whom have suggested that tumour recurrence in the neck in patients with N0 and N1 disease is the same regardless of whether radical or conservative neck dissection has been employed.

Some surgeons also think that it is logical to conserve the spinal accessory nerve, since the vagus, hypoglossal and lingual nerves are routinely spared in neck dissection without any increased risk of tumour recurrence. Skolnik et al (1976) showed that the inferior spinal
accessory nodes were free of metastatic tumour in 51 dissected necks in which the nerve had been preserved. However, they had studied only those nodes along the portion of the spinal accessory nerve distal to the sternomastoid muscle, whereas Schuller et al (1983), in carrying out a prospective study, found that 90% of metastases in the spinal accessory chain were located along the proximal portion of the nerve, that is, that part superior to the point of entrance of the spinal accessory nerve to the sternomastoid muscle.

If a conservation neck dissection is to be performed, a careful selection of patients is paramount, for the partial operations should be undertaken only where there is limited disease and where the natural history of the tumour is well understood. For example, the spinal accessory nerve may be preserved in the case of low-grade tumours of the thyroid, but it must be remembered that when less of the potential specimen is removed, there is more likelihood of residual disease remaining in the neck.

Limitations and failure of radical neck dissection

Complications

The incidence of major complications after radical neck dissection can be as high as 20% (Yarrington, Yonkers and Beddoe, 1973), and between 1 and 20% of patients die as a result of the various complications arising from this operation. The major and potentially lethal complications of radical neck dissection are wound infection, necrosis of the skin flaps and rupture of the carotid arteries. It is well known that the effects of these major complications are increased by prior radiotherapy.

Infection

Infection of the neck following surgery is almost always secondary to an infected haematoma, and is more likely to occur in heavily irradiated patients in whom large flaps with compromised blood supply are overwhelmed by Pseudomonas or other Gram-negative organisms. These infections rarely spread to the deep neck spaces as the spaces have usually been obliterated during the procedure. In addition to haematomata, other contributing causes may be excessive blood loss without replacement, diabetes or other debilitating diseases.

Infections with abscess formation usually take several days to develop and more frequently occur in patients in whom the oral and pharyngeal cavities have been opened. The presence of fullness under the skin flaps requires immediate action. Failure to incise or re-open the wound and to drain the serum or infected haematoma may result in the further undermining and elevation of the flaps, with flap necrosis, spread of infection, exposure of major vessels and possibly carotid artery rupture. The spread of infection into prevertebral, retropharyngeal or pretracheal spaces can lead to fulminating mediastinitis or sepsicaemia, and this can produce life-threatening complications, such as bacterial endocarditis, meningitis, brain abscess, or microabscesses in various other organs of the body. A local wound breakdown may also produce an oral, pharyngeal or oesophagocutaneous fistula.

In patients who have had a heavy dose of irradiation, the use of pre-, intra- and postoperative antibiotics, whenever the oral cavity, pharynx, larynx or oesophagus has been opened, is important in preventing the spread of infection.
**Carotid artery rupture**

Carotid artery rupture is a disastrous emergency which results in fatality unless someone close to the patient responds immediately. The gradual destruction of the vessel wall usually takes 6-10 days, with erosion most commonly occurring at the site of the carotid bulb in the region of the bifurcation. Such an event is more likely to occur if radiotherapy has been used preoperatively, as this affected the vasa vasorum of the adventitia. Immediate pressure must be applied locally over the artery to control the haemorrhage during urgent transfer of the patient to the operating theatre, where the wound can be explored and the bleeding vessel ligated. Hemiparesis remains a strong possibility after carotid artery rupture, but prevention of shock with the maintenance of adequate perfusion of the brain through the opposite internal carotid artery can diminish the risk. There are occasions in cases of advanced cancer of the neck when rupture of the carotid artery can be anticipated as the cause of death.

The incidence of carotid artery rupture can be reduced with careful planning of the neck incisions and this is an advantage of the double horizontal incision described by MacFee (1960). The carotid sheath of a patient should be protected if previous heavy irradiation has been given, particularly in cases where, because the primary carcinoma originated in the mouth, pharynx or larynx, the formation of a fistula is likely. Muscle flaps used to cover the artery were described by Schweitzer (1962) and free grafts of dermis by Corso (1963). More recently, Gardiner, Ariyan and Pillsbury (1983) have described the successful cover of an exposed infected carotid artery using an ipsilateral trapezius myocutaneous flap. In the absence of the transverse cervical artery, the flap is nourished by the occipital artery alone.

It is an ominous prognostic sign when the carotid artery is found to have been invaded by tumour. Kennedy and Krause (1977) showed that invasion occurred in 5.4% of 508 radical neck dissections. In most cases where invasion of the carotid artery by tumour is suspected before the operation, carotid angiography should be carried out with compression of the affected side in order to evaluate the collateral circulation and the likely tolerance to surgical interruption. Although numerous methods have been described, both preoperative and intraoperative, none is absolutely reliable. Serious neurological complications have been described in at least 5% of patients following ligation of the carotid artery despite a normal test (Suarez et al, 1981). The most effective tests are the use of EEG in conjunction with intraoperative clamping of the carotid artery and measurement of the resultant luminal pressure. Because of the unforeseeable sequelae which can occur as a result of occlusion of the carotid artery, it is preferable to excise and replace the vessel whenever possible. This is particularly true if the tests show tolerance to ligation and there is little risk of fistula formation. However, in adverse circumstances, for example after heavy irradiation, it is preferable to carry out a palliative excision rather than a graft because of the risk of infection with associated rupture of the graft. If the tests show tolerance in an irradiated patient, the decision to insert a graft needs to be taken carefully, although successful cases of arterial substitution in patients who have previously been irradiated have been described. It has to be said that, overall, the prognosis for these patients is discouraging whichever technique is adopted. For example, in the small series of Suarez et al (1981), only two of the 16 patients (13%) survived.
**Fistula**

Radical neck dissection combined with laryngectomy or partial laryngopharyngectomy dramatically increases the incidence of fistula formation. This in itself depends to some degree on the nature of the tumour, the dosage of preoperative irradiation, the presence of systemic disease and the operative technique used. T3 or T4 tumours of the pyriform fossa are associated with a high incidence of postoperative fistula when combined with radical neck dissection. Fistula formation is also high in patients who have received preoperative radiotherapy greater than 3,000 cGy. In general, patients with systemic disease, such as diabetes mellitus, are more likely to run a complicated clinical course and produce a fistula. In most instances, a fistula will develop within 3 weeks of surgery and it should then be exteriorized in order to prevent secretions transversing the carotid artery and elevating the skin flap. By exteriorizing the fistula medial to the carotid artery, rupture of the vessel will be prevented provided that the wound has not broken down. However, satisfactory closure of a fistula may ultimately require a myocutaneous flap for repair.

**Chylous fistula**

Chylous fistula is a rare complication of radical neck dissection (1-2%). It occurs predominantly on the left side of the neck; only 25% of fistulae appear on the right side. If the mediastinal pleura is intact, two or more days may elapse during which chyle will fill the mediastinum before rupturing into the pleural cavity. Once established, a chylothorax becomes a serious matter, because as much as 3 litres of fluid may quickly be lost, with an attendant electrolyte, protein and fat depletion.

The commonest presentation of a chylous fistula after an operation is the presence of milky fluid in the suction drainage. Chyle can be differentiated from standard wound effusions by its appearance, by an analysis of the fat content, and by the presence of fat droplets with the separation of a fat layer on standing. In addition, the specific gravity is higher and the protein content lower than effusion fluid. As chyle accumulates in the wound, it causes redness and bulging of the flap with marked induration of the tissues. Chylothorax, if it occurs, presents as either an acute or a gradual onset of dyspnoea, usually several days after the operation.

Conservative treatment is based on continuous suction drainage and rigorous substitution of lost nutritional elements.

The duration of conservative treatment depends on the general condition of the patient, as most chylous fistulae will eventually dry up over a 2-3 week period. The surgical management of a chylous fistula requires re-exploration of the neck and the identification and ligation of the leaking lymph vessel after the patient has been allowed to drink a small quantity of cream containing a colourant.

**Palliation in neck cancer**

About 60% of patients who present with cancer of the head and neck will not be cured, but will, nevertheless, require alleviation of their symptoms. Once such a policy is accepted, the choice of options for further therapy is wide, and much interdisciplinary
expertise must be available. The expansion of ablative and reparative treatment for the cure of head and neck cancer during the last 30 years has been accompanied by a substantial improvement in the scope of palliative techniques, perhaps over a broader field than for curative therapy. For example, if radiotherapy is used as a primary treatment, the problem arises of retreatment of tissues that have been previously irradiated. Sometimes it is possible for irradiation to be given through non-irradiated skin flaps which have been used to replace previously treated skin. Unfortunately, in cases of squamous cell cancer, palliative doses of radiotherapy need to be high, that is in the range of 6.000-6.500 cGy over 6-7 weeks. Inoperable neck metastases in previously irradiated areas are probably best palliated by means of either a permanent implant of radioactive gold grains or a temporary wire implant of radioactive iridium-192 and, wherever possible, by direct surgical exposure. It is prudent in such cases to remove all damaged irradiated skin at the time of placing the implant and to cover the defect with a non-irradiated well-vascularized myocutaneous flap. In the previously untreated neck, implants may be supplemented by teleradiation therapy (Shaw, 1985).

**Advanced cancer of the neck**

A gland in the neck is unlikely to be fixed until it becomes larger than 6.0 cm or more in diameter (Spiro et al, 1974). The presence of fixed glands is in itself an uncommon event, for it occurs in only 5% of all patients with head and neck cancer. Whether a neck is operable in the presence of a fixed gland or glands is a decision that must be made by the surgeon, but the presence of fixed glands does not totally preclude the possibility of surgery. If the tumour ruptures through its capsule and becomes fixed to other structures in the neck, the patient is almost certainly incurable and is likely to die from either a local recurrence or distant metastasis. Capsular rupture must, therefore, be seen as a contraindication to attempting further, be seen as a contraindication to attempting further curative treatment of the patient and, instead, thoughts must be turned towards palliation. Surgery is also contraindicated if the tumour involves either the base of the skull or the brachial plexus, but it is possible to resect a tumour involving a large area of overlying skin and replacing it with a myocutaneous flap. In some selected cases, this procedure has been shown to offer the chance of long-term survival and is helpful in palliation. Most patients are unlikely to be cured following the treatment of fixed nodes with a combination of preoperative radiotherapy and surgery, and Santos et al (1975), who analysed a small series of patients with fixed nodes, showed that the survivors were those in whom the tumour had been sterilized by radiotherapy.

If the tumour invades the carotid system, a decision has to be made as to whether resection of the common carotid artery and its replacement with a vein graft is warranted. A high operative mortality must be expected. Conley (1952) described the results in the case of 31 patients who underwent resection of the common carotid artery: in spite of an operative mortality of five out of the 15 patients, a few did survive to live a useful life for periods of up to 2 years. This is a technique that has not been generally accepted and represents an area where there is a need for further advancement and technical development.

**Terminal cases**

Almost all patients with advanced cancer in the neck will experience distressing symptoms during the weeks preceding death. These symptoms usually include a combination of physical, mental, social and spiritual ills which contribute to the concept of 'total pain' in
the dying, and were described by Cecily Saunders (1978) as a 'continuing situation, rather than a sharply demarcated event: it often has no foreseeable end and often no remembered beginning'. The principles underlying the methods adopted for the relief of the physical symptoms of advanced cancer should be in the repertoire of every otolaryngologist treating head and neck cancer, and they have been well-documented, notably by Saunders (1978).
Chapter 18: Non-neoplastic salivary gland disease

A. G. D. Maran

As with ear disease, people do not normally die of non-neoplastic salivary gland disease and thus some basic principles of pathology are still ill understood.

Pseudoparotomegaly

Hypertrophy of the masseter

This is a condition which occurs almost exclusively in females. It is very distressing because it lends a square shape to what should otherwise be an oval-shaped face. It usually affects younger females and so they are more sensitive about their facial appearance. It looks like bilateral parotid enlargement, but the diagnosis becomes obvious when the patient is asked to clench the teeth and a bulging rippling masseter becomes obvious. It is an important diagnosis to make if only to stop unnecessary parotidectomies being performed. In the group of patients in whom it occurs, it must be distinguished from true parotomegaly that results from bulimia.

The reason for the muscular hypertrophy is not truly understood. It is too simplistic to blame all cases on bruxism which is grinding of the molar teeth. Although some patients do this, it is unlikely that this can stand as a cause unless the cusps of the molars show signs of distinct wear. It is sometimes seen in patients who have undergone long-term orthodontic treatment, perhaps as the result of the masseter setting the jaw in the new position. The most recent explanation is that it is due to kissing and that this is why it is predominantly a disease of females because the male has the stronger jaw.

Treatment is difficult but often very necessary because of the distress of these young females. There is little point in approaching the problem from an external route. This causes a facial scar, requires an unnecessary parotidectomy and puts the facial nerve at risk. It is unlikely that sufficient masseter could be removed without causing at least a temporary paralysis of the facial nerve and the disease certainly does not warrant this approach.

An intraoral approach is the recommended one. The jaw is splinted open and a cut is made along the ascending process of the mandible. The three heads of the masseter are identified and the inner two heads are removed. This leaves the outer head protecting the facial nerve. Care must be taken in this operation, however, not to penetrate the posterior limit of the masseter or else the main trunk of the facial nerve will be at risk.

Ageing

With the absorption of adipose tissue in the ageing process, the salivary glands become more obvious. It is usual that one can palpate the glands in the submandibular area in elderly patients. Patients are sometimes referred to otolaryngology/head and neck units with suspected metastatic disease of the submandibular nodes on the basis of absorption of fat from the submandibular area, leaving the glands obvious and palpable.
**Dental causes**

Dental infection can spread to the lymph nodes either within or surrounding the parotid gland and also to the lymph nodes in the submandibular area. Drainage of the lower incisor or canine teeth below the mylohyoid line can, on occasion, cause Ludwig's angina which involves the submandibular glands although the swelling is much more brawny. Tissue oedema within the infratemporal fossa and between the heads of the masseter muscle in turn causes facial swelling. This is probably the basis of the old-fashioned 'gum-boil'.

**Tumours in the parapharyngeal space**

These are dealt with elsewhere in this volume. Chemodectoma, glomus vagale tumours, schwannomata of the vagus or sympathetic trunks and enlargement of lymph nodes with cyst, tuberculosis or metastatic disease can fill this space and push the parotid gland outwards. This gives fullness in the parotid area that is not a true parotid swelling, but only a displacement. These tumours can also present between the tail of the parotid and the tail of the submandibular gland at the angle of the jaw and displace both structures.

**Tumours of the infratemporal fossa**

The infratemporal fossa is anterior to the parapharyngeal space and tumours in this area can mimic parotid swellings by exiting from the space through the mandibular notch or under the zygomatic arch. The author has had personal experience of this occurring with a haemangioma, a haemangiosarcoma, a leiomyosarcoma and a hydatid cyst.

**Mandibular tumours**

Although tumours of the mandible are relatively rare, both osteosarcoma and chondrosarcoma of the ascending process of the mandible can mimic parotid enlargement as can tumours of the horizontal portion of the mandible in relation to the submandibular gland.

**Mastoiditis**

Mastoiditis in any form is now extremely rare in western and north Europe, but there are parts of the world where it still occurs. A well pneumatized mastoid, if infected, can cause a subperiosteal abscess which, in turn, can drain into the sternomastoid muscle or the digastric muscle lifting the tail of the parotid and mimicking parotid enlargement.

**Intraparotid lesions**

(1) Neuroma of the facial nerve
(2) aneurysms of the temporal artery
(3) lymph node enlargement in or around the parotid gland
(4) parotid cysts.
Parotitis

Pathogenesis

Parotitis is probably the most common infectious disease in childhood and is due to the mumps virus in this age group. What is less well known is that the incidence of viral parotitis in young adults is rising and is due to infection with one of the many strains of the echo- or coxsackieviruses. Bacterial parotitis used to be a common premortem event prior to the advent of antibiotics. The cause of this was an ascending staphylococcal infection along the parotid duct in the dehydrated patient who lacked resistance to infection. As a common oral infection, fungal parotitis is not as common as one would expect given the vicinity of thrush to the duct.

Parotitis can occur secondary to obstruction of the duct either by a stone, which is common in the submandibular gland, epithelial debris, which is common in the parotid gland, and stenosis of the parotid duct due to interdental problems. The patient usually recovers from the pain within a period of minutes or hours but, on occasion, infection can supervene and may even progress to a parotid abscess formation.

Infection of the parotid gland can be due to lymph nodes. There are 6-10 lymph nodes around the parotid gland and 4-6 within the parotid gland. These drain the skin of the side of the face, the scalp, the ear, the eye and the posterior part of the oral cavity. Lymphadenitis can thus be secondary to skin lesions and, in this regard, infected pierced ears are probably the commonest cause. Otitis externa frequently presents with preauricular pain and swelling due to lymph node enlargement. The relationship with dental infection of the molar teeth has already been mentioned and preauricular pits as a result of malformation of the branchial apparatus can result in recurrent infections which drain to the parotid lymph nodes. These nodes may resolve or they may proceed to abscess formation.

Chronic infection of the salivary glands can occur as the result of tuberculous infection. It is rare for tuberculosis to affect the stroma of the gland and, if the salivary glands do become affected by tuberculosis, then it is the surrounding lymph nodes that are infected. Sarcoid can similarly affect the area and rarely the parotid and submandibular glands can be affected by actinomycosis, leprosy or tularemia.

The symptoms from parotitis prior to abscess formation come when saliva is produced. This happens when the patient attempts to eat and creates secretomotor stimulation of the salivary glands. If the ducts are oedematous or blocked with stone or debris, then the flow of saliva is obstructed and the tense glands swell even more, causing severe pain. The parotid and the submandibular glands are covered with the investing fascia of the neck. Although this is not very obvious at surgery, it causes constriction of the salivary glands in the case of infection. It then becomes very difficult for the glands to expand causing severe pain.

Clinical features

Parotitis causes severe pain and elevation of temperature. It is made worse by eating and the patients are very hungry but do not eat because of the fear of pain. The pain and surrounding swelling cause spasm of the masseter, the temporalis and the pterygoid muscles;
this causes trismus. The effect of trismus in a viral parotitis can be to create the environment in which a superadded opportunistic bacterial infection can occur due to bad oral hygiene. The area over and around the affected salivary gland is extremely tender. The diagnosis can be substantiated by asking the patient to sip a little lemon juice when there will be an acute worsening of the pain. This will not be the case, however, if the swelling is due to a lymph node.

Apart from the examination of the salivary glands, the oral cavity should be examined for the presence of dental infection and the molar teeth especially should be palpated, moved and tested with hot and cold stimuli. Some attempts should be made to see what material comes out of the salivary ducts with moderate pressure over the glands. Pierced ears and otitis externa should be looked for.

It should also be borne in mind that the painful parotid gland may be a manifestation of Sjögren's disease, but this is not at all common.

**Laboratory investigations**

There will be an elevated white cell count with lymphocytes predominating if it is a viral infection, and neutrophils if it is a bacterial infection. The erythrocyte sedimentation rate will also be raised in keeping with the general condition of the patient but, if it is very elevated, then the possibility of Sjögren's disease arises.

Viral titres should be measured in every case and, although the mumps titre is reliable, there are so many strains of the echo- and coxsackieviruses that it is felt to test for each strain would be too expensive in terms of reagents and manpower.

**Bacteriology**

Secretions from the ducts can be used to try to identify viral infection and also can be plated for bacterial culture and sensitivity. Mycology should also be examined in every case.

If tuberculosis or sarcoidosis is suspected then the opinion of an ophthalmologist should be secured to see if the patient has uveitis. This often accompanies parotitis in these two conditions and if present completes Heerfordt's syndrome.

**Radiology**

**Plain X-rays**

Plain X-rays are useful in identifying the presence of stones. Occlusal films in the submandibular region show stones very well and are much better in this regard than lateral films. It is important to be able to assess whether or not the stone is in the oral cavity or in the gland.

Parotid stones are radiolucent and will not be seen on plain X-ray.
Sialography

The timing of a sialogram in parotitis is debatable. It would be universally agreed that a sialogram should not be performed during the acute phase. A week or two later, however, it might be therapeutic in washing out the duct system.

This examination is very useful because it will give some idea of duct blockage and will certainly give a diagnosis of sialectasis, if this is present. It may show duct distortion although this is such a variable feature that it lacks any diagnostic specificity.

Scanning

Parotid scanning with technetium-99 has largely been abandoned because of so many false positive and negative results.

Fine needle aspiration biopsy

This is only possible in departments with an experienced cytologist. The diagnosis in parotitis is usually so obvious that fine needle aspiration is not required, but it may be carried out for confirmation.

Conservative treatment

The patient will feel so ill that he probably should be confined to bed. In spite of trismus and pain, a good standard of oral hygiene should be maintained and, although eating and drinking are very painful, the least painful nutrition is with high calorie milk drinks which do not carry any flavour. All patients will require the appropriate degree of analgesia and local heat applied to the affected gland is often comforting. Adrenaline should be applied to the appropriate salivary duct in the hope of reducing the oedema and causing some lessening of tension with drainage of saliva. It is doubtful if any antibiotic is truly effective, but the only antibiotic that is secreted in saliva is clindamycin and this can be used according to the circumstances. If a diagnosis of tuberculosis is established then the appropriate chemotherapy can be begun with a high resolution rate.

Similarly, with leprosy, actinomycosis and tularaemia, a reasonable response to chemotherapy can be expected.

If the infection is due to lymphadenitis, then the primary source of infection should also be dealt with either surgically or with antibiotics.

Surgical treatment

The most successful surgical intervention is where a stone blocks the submandibular duct with secondary sialadenitis. The stone can be removed perorally to eradicate infection, but since abscess formation is often loculated any serious attempt at drainage would involve lifting a facial flap and carrying out multiple incisions over the gland. This is seldom warranted.
Metabolic parotomegaly

The following conditions have traditionally been recorded as causes of parotomegaly - gout, Cushing's disease, myxoedema and diabetes mellitus. Investigators who have examined large series of patients with these endocrine disorders with special reference to their salivary glands, have not substantiated the original relationships and endocrine parotomegaly may be anecdotal.

The parotid gland is much more closely related to nutritional abnormalities. Since the Second World War, parotomegaly and enlargement of the submandibular glands have been noted in prisoners of war and other groups subject to starvation. This is probably a similar process to the one mentioned earlier in relation to ageing, namely the persistence of the substance of the salivary glands with the disappearance of surrounding adipose tissue.

The metabolism of fat within the parotid glands is ill understood. As well as the salivary glands maintaining their shape and form and the surrounding adipose tissue disappearing, fatty infiltration is very common in obese individuals. Furthermore, if these individuals lose a lot of weight then they are left with the original fat deposition in the salivary glands causing residual parotomegaly.

Although salivary gland enlargement is not noticed in anorexia nervosa, parotomegaly is a feature of the recently recognized condition of bulimia where binge eating is followed by self-induced vomiting. Several of these patients have presented with parotomegaly and have not disclosed their binge eating. As a result some normal parotids have been removed. Histological examination of these glands, furthermore, has failed to reveal any abnormality or abnormal deposition of fat.

Drug-induced parotomegaly

In the laboratory, it has been shown that both isoprenaline and thiouracil make a rat's parotid swell. Nowadays, with the expert control of thyroid disease, it is very seldom that a patient with parotomegaly due to thyroid-related medication or isoprenaline is seen. The list made up by the Committee for the Safety of Medicines, however, shows over 40 drugs as affecting the salivary glands. Many of these reports are anecdotal and coincidental and there is little in the way of scientific evidence to show that drugs do cause parotomegaly. Although drug 'allergy' is mentioned in many reviews of parotomegaly, again there is no evidence for such an entity.

In clinical practice, the only drugs associated with parotomegaly or painful parotid with any frequency are dextropropoxyphene (Distalgesic) and high oestrogen oral contraceptive pills. The method of action of these latter drugs is to create epithelial shedding within the duct system, the creation of epithelial mud and the blockage of salivary ducts with the possible creation of stones in the submandibular area.
Sialectasis

Pathogenesis

The cause of sialectasis is unknown. It is probably best regarded as a salivary gland analogue of bronchiectasis. There is a progressive rotting and disintegration of the alveoli which ultimately coalesce forming cysts. The debris from these cysts passes along the duct and intermittently blocks areas of the duct causing hypertrophy, stenosis and duct dilatation. This is exactly what happens in bronchiectasis and, as with this disease, some have it from birth, and others develop it for no very good reason. In a few this debris is secondary to known obstructions, but occurrence in both the salivary glands and the lung is the exception rather than the rule. Congenital sialectasis has no adequate embryological explanation.

The parotid is a serous gland and is low in calcium. The epithelial debris, which could become calcified to form a stone, does not have the stimulus of the correct environment for calcification and remains as ‘mud’. This is effective in blocking the duct system but not as effective as a stone would be. It is softer and does not impact so easily and is more easily removed with a build-up of saliva. Stones can form, however, in the parotid gland but they are of low density and are radiolucent.

The submandibular gland is a mixed seromucinous gland and is high in calcium. Epithelial debris here, therefore, calcifies easily and this is why stones are more common in the submandibular than in the parotid gland. The stones are of high density and are, therefore, radiopaque.

Although some texts list calculus disease as a separate entity, it is unlikely that stones can form de novo. They are probably all formed in a radiologically negative sialectatic gland.

The symptoms are produced when the ducts are blocked. If the main duct is blocked then the whole gland will swell up in response to the secretomotor stimulation of eating, or drinking. This is especially marked with citrus drinks or fruits which cause maximal salivary stimulation. The blockage may be in a more distal duct, in which case only a portion of the gland will swell up.

In most instances, the gland will clear itself. Sometimes the swelling stays for days but usually only for minutes or hours. If the swelling stays for some days, then it may become secondarily infected and abscess formation occur. The abscess may rupture or be drained but more likely it will heal by fibrosis.

Clinical features

History

The patient typically complains of pain and swelling of the gland during a meal. The swelling is visible and can remain up for minutes, hours or days. While the gland is swollen it is painful but, when the swelling goes down the gland is not painful. The patient does not feel unwell and does not have an elevation of temperature.
The condition can be particularly troublesome in children. This is the group of children that are diagnosed as having 'mumps' more than once. Fifty per cent of these cases resolve in time and only a few adult cases require surgical treatment.

**Examination**

On examination, a stone may be seen in the submandibular duct or palpated by bimanual palpation within the gland. The mouth of the duct may be oedematous and pouting.

The parotid duct may have the same appearance and it is useful to massage each parotid to see if there is any drainage of saliva from the duct.

**Investigations**

**Laboratory investigations**

There is little information to be gained from blood tests.

**Radiology**

The sialogram is diagnostic of this condition. A plain X-ray should be performed in all cases to see if a radiopaque stone is visible and then one should proceed to a sialogram.

A sialogram may show six pictures:

1. it may be normal
2. it may be overfilled, an overfilled sialogram is often reported as sialectasis and one must be aware of this picture
3. the radiologist may fail to cannulate the duct; most radiologists know that they should not persist with difficult cannulations for any period of time because the duct becomes oedematous and it then becomes virtually impossible to cannulate; if cannulation in these cases is eventually successful then the radiologist's report will be of duct stenosis but it will be iatrogenic rather than real
4. the fourth possible picture is that of an obstructed duct; this will be obvious in the submandibular gland if a stone is seen on the plain film but in the parotid gland, the dye may enter a little way along the duct and come to a halt, in which case it can be presumed that epithelial mud is blocking the main duct
5, 6) there are two classical pictures of sialectasis: the first is cystic and the second is globular or saccular. Only one of these truly represents the pathology of the condition. Cystic sialectasis where the alveoli coalesce and form large spaces together with duct, stenosis and dilatation, is the true picture of sialectasis. Thackray (1955) has shown that globular sialectasis represents no abnormality of the duct other than abnormal leakage where the lipiodol or radiopaque medium comes out of the alveoli and lies in the stroma of the gland. Although popularly called saccular sialectasis, it is not related to the pathology that was described akin to bronchiectasis.

As stated previously scans are unreliable and a sialogram together with a computerized tomography (CT) scan is of little value in this condition.
Treatment

No treatment

In many cases the sialogram is therapeutic. It washes out the duct and alveolar system and, thereafter, the patient may be advised to finish each meal with a citrus drink which will encourage the production of saliva, and then to massage the affected gland in order to wash out any epithelial debris and stop collections. This is successful in the vast majority of cases.

Peroral removal of a calculus

This can be carried out if a calculus is seen, usually in the submandibular gland. The duct is blocked proximally to stop the stone disappearing back into the gland during manipulation and the duct is marsupialized after the stone is removed.

Marsupialization of the duct

This must accompany any peroral removal of the stone and it can be undertaken in the parotid duct stenosis of dental origin. A cannula is placed into the duct and a 2.5-5 cm (1-2 inch) segment is opened and stitched carefully to the adjacent mucosa with 6-0 absorbable suture material.

Ligation of the duct

This is mentioned only to be dismissed as an illogical and inadequate present-day treatment.

Duct dilatation

The same applies to this method of treatment.

Tympanic neurectomy

This procedure was popular during the 1950s and 1960s. The aim of treatment is to divide Jacobson's nerve which crosses the promontory to form the tympanic plexus. It joins the jugular plexus and glosopharyngeal nerve to the greater superficial petrosal nerve and forms part of the reflex arc. Some surgeons combine this with division of the chorda tympani nerve.

The procedure certainly works for up to 6 months but, like all autonomic surgery, alternative pathways develop and symptoms recur.

Removal of the submandibular gland

This is a straightforward procedure and carries little risk with it. If the gland shows evidence of sialectasis, or if there is a stone in the body of the gland, then the surgeon should have no hesitation in removing the submandibular gland as a whole. If it has undergone numerous attacks of sialadenitis, then the removal may be difficult, but care should be taken
to avoid paralysing the mandibular branch of the facial nerve. Furthermore, one should make sure that the remnant of the submandibular duct in the oral cavity is clear of stones when the gland is removed, because it is quite possible to push stones from the gland into the duct remnant during manipulation. This is of little clinical significance, but the patient may be surprised to spit out a stone some days after the operation.

**Total parotidectomy**

Superficial parotidectomy is illogical for a disease which affects the whole parotid gland. By removing half of a sialectatic parotid gland, there is a high risk of fistula. In a superficial parotidectomy for tumour, a denervated normal deep lobe is left behind. This usually ceases to function and thus fistula is rare. In sialectasis a cystic diseased deep lobe is left behind for which denervation does little. It continues to produce mucus and saliva and causes a salivary fistula.

Total parotidectomy is, therefore, the only logical operation for sialectasis and, in the gland which may be heavily fibrosed due to recurrent sialadenitis, the facial nerve is at more risk than it is in surgery for benign tumours.

**Sjögren's syndrome**

In 1888, Dr Mikulicz described the case of a 42-year-old East Prussian farmer with swelling of the submandibular, parotid and lacrimal glands. He removed two-thirds of the submandibular gland and found it to be infiltrated with lymphocytes, and then removed the whole gland some months later with recovery of the patient. This was known as Mikulicz's disease and to it were added all the symptoms of other non-neoplastic salivary gland disease over the next 50 years. Mikulicz's syndrome included tuberculosis, sarcoid, actinomycosis, gout, etc. In 1925, Gougerot, a French dermatologist, introduced the concept of dryness when he described a series of patients with dry mouth, vulval dryness, skin dryness, etc. In 1933, Henrik Sjögren, a Stockholm ophthalmologist, described 33 women with the syndrome of xerostomia and keratoconjunctivitis sicca. Twenty-three of these patients had rheumatoid arthritis. No mention was made of parotid gland disease. In 1952, Godwin at the Armed Forces Institute of Pathology in Washington, described the concept of enlargement of the parotid glands due to lymphocytic infiltration and related this to the future development of lymphoma. It was named benign lymphoepithelial lesion. In 1974, Anderson and Talal described a further variety of this called aggressive lymphocytic behaviour.

The classification now is as follows:

1. **primary Sjögren's syndrome (sicca complex)** - this consists only of xerostomia and xerophthalmia with no connective tissue component

2. **secondary Sjögren's syndrome** - this consists of xerostomia, xerophthalmia and a connective tissue disease which in nearly 50% of cases is rheumatoid arthritis but may also be systemic lupus erythematosus, scleroderma and polymyositis

3. **benign lymphoepithelial lesion**, otherwise known as myoepithelial sialoadenitis, which is localized to the parotid glands and some regard as a prelymphomatous condition
aggressive lymphocytic behaviour which again is confined to the parotid glands and is almost a pseudolymphoma.

**Epidemiology**

Sjögren's syndrome is more common in the northern than the southern hemisphere and is more common in northern than in southern Europe. A proportion of older people have many of the symptoms of Sjögren's disease, but do not have the immunological profile. In a study of octogenarians, Whaley showed that one in six males and females had keratoconjunctivitis sicca. Three per cent of men and 20% of women had xerostomia, but only 2% had the immunological profile of the sicca syndrome. Eleven per cent of patients with rheumatoid arthritis had keratoconjunctivitis sicca, 1% had xerostomia but 100% had lymphocytic infiltration of the submaxillary glands when examined at autopsy. Thirty per cent of patients with rheumatoid arthritis will develop Sjögren's disease. It has a very wide range of autoantibodies and is the second most common autoimmune disease after rheumatoid arthritis.

**Clinical features**

Sjögren's syndrome is a multisystem disease affecting every system in the body but particularly the oral cavity, the eyes and the salivary apparatus.

The oral symptoms are those of dry mouth with secondary candidiasis, stomatitis, glossitis and subsequent dental caries.

The eye symptoms are keratoconjunctivitis sicca; the patient has a foreign body sensation in the eye, burning, redness, itching, photosensitivity and an inability to tolerate contact lenses.

Only 40% feel salivary gland enlargement and only 20% show it clinically. It is nearly always in the parotid and those patients with parotomegaly from Sjögren's disease have a much higher chance of developing lymphoma. Two-thirds of the patients never have salivary gland enlargement.

Other associated systemic problems are primary biliary cirrhosis, chronic hepatitis, vasculitis, chronic graft versus host disease, cryoglobulinaemia, hypergammaglobulinaemic purpura and polyarteritis. Fifteen per cent will have thyroiditis and many will develop pancreatitis.

Achlorhydria, disorders of oesophageal motility and web formation may present to the otolaryngologist as may nasal crusting, epistaxis, serous otitis media, laryngitis sicca and a persistent cough with tenacious sputum, glazing of the oral mucosa and sticky secretions in the nasopharynx, etc.

**General examination**

The presence or absence of a connective tissue disorder should be established as should the presence of any of the above-mentioned abnormalities of other organs.
**Investigations**

**Blood examination**

The erythrocyte sedimentation rate is usually raised. A protein profile will show elevation of all the immunoglobulins especially IgG. Rheumatoid factor and antinuclear factor will probably be positive and there may well be a wide range of autoantibodies.

**Specific immunological tests**

These can only be carried out in a few places in the UK. When class 2 antigens such as HLA A1 and B8 and DR3 are examined, then almost three times as many patients with sicca syndrome have these antigens when compared with patients with the secondary syndrome. Specific antigens for Sjögren's syndrome are called SSA and SSB. Again these are more common in patients with the sicca syndrome than in those with secondary Sjögren's disease with rheumatoid arthritis. The immediate clinical relevance of these immunological abnormalities is not known and it may be that they are *in vitro* epiphenomena.

**Schirmer's test**

This is carried out by putting special strips into the lower fornix. Wetting of less than 5 mm in 5 minutes represent a diagnosis of xerophthalmia. A diagnosis of keratoconjunctivitis sicca, however, cannot be made until the ophthalmologist examines the eye with Rose Bengal dye to see the filamentary keratitis.

**Salivary flow rate**

This is measured using Carlsson-Crittenden cups; these are suction cups placed over the parotid duct. Maximum stimulation is created by getting the patient to suck a lemon. A flow of less than 0.5 mL in a minute represents xerostomia.

**Labial biopsy**

This is performed by obtaining four globules of fat from the back of the lower lip. It can be performed under local anaesthetic and is the diagnostic test for Sjögren's disease. The pathologist must grade it according to the rules laid down.

- Grade 1: slight lymphocytic infiltration
- Grade 2: less than 50 lymphocytes per 4 mm$^2$
- Grade 3: 50 lymphocytes per 4 mm$^2$
- Grade 4: more than 50 lymphocytes per 4 mm$^2$.

The distribution of lymphocytes is important also because they cannot be diffuse, but must be periductal. In this test, false positives can be obtained in rheumatoid arthritis, scleroderma, subacute lupus erythematosus, sarcoid, amyloid and graft versus host disease.
**Radiology**

Sialography either shows a normal sialographic pattern or that of 'globular sialectasis'. This does not imply that the patients with Sjögren's disease have sialectasis. What it does imply is that there is an abnormality in the duct allowing leakage of lipiodol into the stroma of the gland.

**Natural history**

One in six patients with Sjögren's disease will go on to develop lymphoma. This will be a B-cell type non-Hodgkin's lymphoma. The immunological abnormality in Sjögren's syndrome is a loss of suppressor T-cell activity and an alteration in the T-suppressor-helper cell relationship. As well as a non-Hodgkin's lymphoma they can develop Waldenström's macroglobulinaemia and immunoblastic sarcoma.

The present suggestion as to aetiology is that the cytomegalovirus infects salivary ducts and ducts elsewhere in the body. The ducts act as the antigen and B-lymphocyte proliferation occurs. As well as lymphoma, a peculiar type of anaplastic carcinoma can also develop in these patients. It has been reported predominantly but not solely in Eskimos.

**Treatment**

There is little of a specific nature that can be done to help these patients. Bouts of parotid swelling may be treated with steroids but the bouts are seldom so severe that they require other immunosuppressive drugs. Artificial tears and synthetic saliva provide limited comfort and bromhexine 40 mg/day sometimes helps a tenacious cough.

The most important feature of treatment, however, is to put these patients on a lymphoma follow-up. Those who have parotid enlargement are at a higher risk of developing lymphoma and diagnostic parotidectomy should be considered.

**Salivary gland cysts**

Similar to vascular and lymphatic malformations, salivary gland cysts are benign swellings, and merit inclusion in this section.

Most cysts within the parotid or submandibular glands are secondary to sialectasis, or salivary tumours (pleomorphic adenoma, Warthin's tumour, cystic duct adenoma, mucoepidermoid tumour, adenocarcinoma). Cysts not related to other disease more commonly arise from minor salivary glands. In a series of 483 salivary gland cysts, 77% were minor salivary gland mucoceles, and the remainder included parotid duct cysts (10.5%), lymphoepithelial cysts (6%), and ranulae (5%) (Seifert et al, 1986).

Mucoceles are spherical, painless swellings that contain mucus. There are two histological types - extravasation mucoceles (80%), which probably follow repeated minor mucosal trauma, and retention mucoceles (20%) due to duct obstruction by microliths, or inspissated secretions or to bends in the duct.
Extravasation mucocoeles (sometimes called a mucous granuloma) are commoner in younger adults, and show a predilection for certain sites (lower lip 80%, cheek and floor of mouth 15%, and palate, tongue and upper lips 5%). In contrast, retention mucocoeles present in older patients and do not have the marked predilection for certain sites. Treatment of either variety is simple excision.

A ranula is a specific type of salivary gland cyst which arises from the sublingual gland, and is discussed in Chapter 4.

Salivary fistulae

A salivary fistula usually originates from the parotid gland, although it sometimes arises from the submandibular gland. It can be internal or external, congenital or acquired. Internal fistulae drain into the mouth and are therefore often not noticed. Congenital fistulae are rare, and may arise from aberrant or accessory salivary tissue, or be associated with branchial cleft anomalies.

A parotid fistula may be due to surgery, facial trauma, or sepsis within the gland parenchyma. A fistula which follows a partial parotidectomy (especially a lumpectomy) usually arises from the gland parenchyma, and generally drains through the suture line. This type of fistula will close spontaneously in most cases, and until this time, minor leakage will occur with meals. To help prevent such a fistula from developing, the parotid duct is often divided well forward, and ligated in some cases. Saliva may collect underneath the skin flap after surgery, and should be aspirated and a pressure dressing applied. Saliva has a high amylase content compared with fluid from a seroma.

In contrast to the above, a fistula which arises from the main duct system of the parotid leaks profusely, even at the thought of food, and invariably needs an operation to close it. This type of fistula is usually due to a deep facial wound, and the facial nerve may also be damaged. Such fistulae can be difficult to control, and three categories of treatment are described:

1. reduction of saliva production; by drugs, irradiation, gland denervation, and duct ligation

2. operations on the fistula; excision, diversion into the mouth, reconstruction of the damaged duct

3. removal of the gland; partial or total conservative parotidectomy.

Reducing the output of saliva may promote closure of the fistula, and tympanic neurectomy provides a simple effective way of achieving this. Irradiation has been used, but the outcome is uncertain, and there is a risk of later carcinoma. A damaged main duct may be suitable for repair, and this may be demonstrated by a sialogram. However, the outcome of such surgery is uncertain, especially if the fistula has been present for some time, an in such cases parotidectomy may be necessary.
A salivary fistula from the submandibular gland is a much easier problem to deal with. If it does not close spontaneously the gland should be excised.
Chapter 19: Benign salivary tumours

O. H. Shaheen

Historical perspective

Salivary gland surgery in the years leading up to the Second World War was, by contrast with the present day approach, a timid and halting affair for reasons which are now all too clear.

A poor understanding of the natural history of salivary diseases in general, and tumour pathology in particular, together with a fear of damaging the facial nerve stand out in retrospect as the main obstacles to progress. Despite being well versed in the anatomy of the facial nerve, there was little enthusiasm on the part of surgeons for confronting the nerve in vivo, a diffidence born in part from the difficult operating conditions of the time, but also from the fear of dissecting around the nerve. Safeguarding the physiological integrity of nerves by delicate handling was a relatively recent concept, and seemingly the hallmark of neurosurgeons, whereas the saying that ‘a nerve exposed is a nerve damaged’ very much dominated the thinking of the general surgical circles.

Operations on the submandibular gland would have been viewed with much less reluctance, and fears there would have focused more on the danger of spread of infections in fascial planes, rather than on the bogey of damaging nerves such as the mandibular branch of the facial nerve.

The lack of a proper classification of salivary tumours and general ignorance of their natural history probably constituted greater impediments to progress. For many years, the belief existed that mixed tumours were a low-grade form of malignancy largely because of their propensity to recur after local enucleation. It was not appreciated that the degree of encapsulation of pleomorphic adenomata was variable and often incomplete, or that their surface was frequently bosselated, features which favoured recurrence if simple enucleation were practised.

Others ascribed the high recurrence rate of pleomorphic adenomata to a multifocal origin, not realizing that the appearance of separate outlying foci was an artefact created by the plane of section passing, not only through the main bulk of tumour, but also through a number of surface excrescences. Serial sections ultimately established the continuity of the small extensions to the parent tumour and redirected attention to the real cause of recurrence, namely inadequate surgical margins and spillage of tumour.

During the was years, the foundations of our present knowledge and expertise were laid down through the efforts of a number of individuals working mainly in North America, France, and the UK. Later contributions emanated from these countries and from Scandinavia.

McFarland (1942) attempted to correlate the histology of mixed tumours to their prognosis after surgery, while Foote and Frazell (1953) were largely responsible for placing the classification of salivary tumours on a sound basis and for providing a comprehensive account of their natural history.
The implantation of mixed tumours during the operation of enucleation, long suspected as a cause of recurrence, was eventually demonstrated in a most convincing manner by Patey and Thackray (1957-1958) and the belief that they were multifocal in origin finally put to rest.

Bailey (1941) was the first in the UK to practise formal dissection of the facial nerve in operations for benign parotid tumours, while Redon (1945) in France was advocating total parotidectomy with conservation of the facial nerve at much the same time. The former considered the parotid to be essentially a bilobed structure separated by an anatomical plane containing the facial nerve, a concept which is now known to be fallacious, but which in no way detracts from Bailey's valuable contribution to parotid surgery. Redon, on the other hand, believed that all mixed tumours were multifocal in origin and consequently advocated removal of both superficial and deep parts of the parotid after freeing the facial nerve of its attachments.

Although this belief was finally and irrevocably refuted, Redon's contribution should not be minimized since it broadened the technical repertoire of parotid surgeons and provided insight into the functional capabilities of the facial nerve in the face of operative trauma.

**Surgical anatomy**

**The parotid gland**

A horizontal section of the parotid gland is much more informative about the normal disposition of salivary gland tissue than might be presumed from lateral inspection. Such a section would clearly demonstrate that most of the gland actually lies in the retromandibular sulcus, rather than on the external of the masseter muscle and would thus explain why the majority of tumours are to be found in that segment. Since the facial nerve splits this area unequally into a major portion lateral to the facial nerve and a much smaller part medial to it, it is not surprising that most tumours are to be found superficial to the nerve.

The gland is incompletely invested by a continuation of the deep cervical fascia, which surrounds the sternomastoid posteriorly and overlies the masseter muscle anteriorly. The surface component of the parotid fascia is exceedingly tough, but as it branches medially over the anteromedial and posteromedial aspects of the gland it thins out progressively. Where the gland lies close to the styloid process, the fascia blends with a tough band of fibrous tissue which joins the styloid to the posterior aspect of the angle of the mandible, thus forming the stylomandibular ligament.

The toughness of the parotid fascia on the external surface of the gland inevitably means that benign tumours are slow to project outwards to any great extent, and hence it takes years for them to present as large unsightly bulges. The facial nerve is often displaced by the tumour, either inwards, where there is little resistance from the deep component of the parotid fascia, or upwards or downwards, depending on the relationship of the nerve to the tumour.

The facial nerve emerges from the fallopian canal and runs anteriorly, inferiorly and laterally to enter the posteromedial surface of the gland. The segment of the nerve which lies in the interval between the stylomastoid foramen and the parotid is extremely short, but is the
ideal location for finding the facial nerve before the parotidectomy proper gets under way. It is best found by searching in the tympanomastoid sulcus which is formed by the edge of the bony external meatus on the one hand and the anterior face of the mastoid process on the other. The nerve emerges from the stylomastoid foramen some 3-4 mm deep to the outer edge of the bony external canal (Conley, 1978).

The second most reliable landmark for finding the nerve is the posterior belly of the digastric muscle which lies just inferior to it and allows a similar trajectory. The styloid process is a useful landmark, but to depend on it for finding the nerve is to court trouble since it lies medial and anterior to the nerve's point of emergence from the mastoid. The posterior auricular artery frequently bleeds during the process of looking for the nerve, since it lies below and just lateral to the facial nerve.

The cartilaginous pointer described by Conley (1978) is an artificially created landmark formed by posterior traction of the external auditory canal. The backward pull on the cartilage causes the meatus to assume the shape of a cornucopia, the curved extremity of which allegedly points to the position of the facial nerve. Of all the landmarks mentioned, this is probably the least reliable since it very much depends on the configuration of the cartilaginous meatus.

The facial nerve divides into upper and lower divisions about 1 cm beyond this point of entry into the parotid, and each then diverges sharply from the other.

The upper division proceeds upwards forwards and very much outwards towards the zygomatic arch and gives off temporal, upper zygomatic, lower zygomatic and buccal branches. This division is almost invariably stouter than the lower division and can therefore withstand more handling. But in elderly or obese individuals, its branches are often tortuous, a feature which makes them liable to be damaged if efforts are neglected to keep the tissues constantly on the stretch while the nerve is being dissected. The pattern of branching in both upper and lower divisions is variable, both in terms of the number of branches and their point of origin. The lower zygomatic branch is, however, constant in one respect, in that it almost invariably lies just above the parotid duct, a point to be remembered when attempting to deal with duct stenoses or stones.

The lower division passes downwards and forwards but lacks the outward inclination of the upper division and, by comparison, therefore lies deeper. It gives off buccal branches, a mandibular and cervical branch and it thins progressively as it becomes more peripheral. Occasionally, a buccal branch may arise from the bifurcation itself.

The thinness of the branches of the lower division, and the mandibular branch in particular, makes paralysis of the depressor anguli oris a common complication of parotidectomy. The very fine interlacing nerve fibres between one branch and another could well explain why the facial nerve will withstand more than a modest degree of handling at operation and yet still recover. These communicating fibres are often absent between the lowest buccal and mandibular branches, hence the propensity to paralysis of the muscle supplied by the latter.
The mandibular branch invariably emerges from the tail of the parotid gland immediately anterior to the retromandibular vein and then passes downwards and forwards on the outer aspect of the deep cervical fascia to enter the submandibular triangle. The proximity of the mandibular branch of the facial nerve to the retromandibular vein at their point of emergence from the tail of the parotid, provides an alternative method for locating the nerve if the usual method is for any reason not possible. By working backwards along the nerve, the two divisions, the other branches, and the main trunk can be found in turn.

The retromandibular vein lies in the deep lobe of the gland immediately medial to the facial nerve and its branches, although very occasionally it may be superficial to it. It gives off small tributaries which pass outwards between the nerve branches and which may be a source of troublesome bleeding when dissecting out the nerve. Deep to it lies the continuation of the external carotid artery which gives off, in turn, transverse facial, internal maxillary and superficial temporal branches as it proceeds superiorly within the deep lobe of the gland. These branches with their venous counterparts must be ligated when the deep of the gland is removed.

The submandibular gland

The gland lies in the triangle of that name, covering the mylohyoid and hyoglossus muscles, and overlapping the inferior margins of the triangle, namely the anterior and posterior bellies of the digastric and their common tendon. It is itself overlapped by the horizontal ramus of the mandible which forms the upper margin of the triangle. Inferiorly, the gland approaches the greater horn of the hyoid which serves as a useful landmark when siting the incision for the operation to remove the gland.

Although ovoid in appearance when viewed from a lateral standpoint, the gland is in fact U-shaped in sagittal section, possessing a large outer component which lies outside the mylohyoid muscle, and a smaller inner component giving origin to the duct on the inner aspect of this muscle. The common stalk joining these two parts curves round the posterior free edge of the mylohyoid. In operations to remove the gland, retraction of the mylohyoid forwards will facilitate exposure of the deep part of the gland and the duct.

The gland is invested in a loose fine capsule which is derived from the overlying deep cervical fascia. If surgical dissection of the gland is limited to a plane within this capsule, any important structure lying outside it will not come to any harm. The deep cervical fascia provides a further external protection since the mandibular branch of the facial nerve and its subsidiary branches lie plastered to its outer aspect and never make contact with the gland or its capsule.

However, it must be appreciated that the most inferior of the branches of the mandibular nerve lies close to the lower border of the submandibular gland and could be damaged when gaining entry to the plane of surgical dissection at the beginning of the operation.

The hypoglossal nerve with its venae committantes lies on the hyoglossus, but is separated from the deep aspect of the gland by a potential space. Escape of disease from the
deep aspect of the gland would be the only likely circumstance to put the nerve at risk when removing benign tumours.

The lingual nerve arches gently downwards just above the deep part of the submandibular gland to which it is attached by a ganglionic connection, alongside which is a small blood vessel. It subsequently passes below the duct then round its outer aspect in the form of a broad loop before heading for the mucosa of the tongue. It is at risk when the deep part of the gland is being mobilized.

**The sublingual gland**

Also ovoid in shape and about 2 cm in size it lies below the submandibular duct between the genioglossus on the one hand, and the mandible and mylohyoid on the other. About half its ducts drain into Wharton's duct and the remainder directly onto the sublingual papilla.

**Incidence of salivary tumours**

Although the geographical incidence varies somewhat from one location to the next, salivary tumours are in general uncommon. In a population of 2.8 million living in an area comprising Liverpool, Merseyside, North Wales and the Isle of Man, the incidence of neoplasms of all complexions was of the order of 1.1 per 100,000 persons. This can be compared with a slightly higher incidence of 1.5 per 100,000 Caucasians in the USA, rising to 1.6 for non-white males and 2.5 non-white North American females. The highest incidence would appear to be among the Eskimos in whom the majority of tumours are of the malignant kind (Evans and Cruickshank, 1970).

Salivary tumours represent about 3% of all neoplasms. Approximately 80% are located in the parotid, 10% in the submandibular gland, the remainder being distributed between the sublingual gland and the countless minor salivary glands (Snow, 1979). This last group comprises the innumerable tiny submucosal serous and mucinous glands to be found in the oral cavity, nose, sinuses, postnasal space, oropharynx, larynx and trachea.

Benign tumours are more common than malignant, although the ratio will vary from one anatomical site to the next. In the parotid, for instance, 80% of tumours are benign whereas in the submandibular gland this drops to 60% and in the oral cavity malignant tumours may well outnumber the benign.

Among the white population of Europe and the USA the sex distribution is about equal, but among non-whites in North America and Africa, women outnumber men and if the parotid is considered in isolation, there is a preponderance of tumours in women (Evans and Cruickshank, 1970).

Benign tumours of the major salivary glands may occur in children but are exceedingly rare. Although fractionally more common in adolescence, they are nevertheless still infrequent by comparison with the vast majority of patients who present in the age range 30-70 years, the average being about 45.
Aetiology

Little is known about the aetiology of salivary tumours and much of what has been written is speculative.

The inoculation of newborn mice with polyoma virus is reported to provoke the formation of salivary neoplasms, although there is nothing to suggest that this is the mechanism in man. Hydrocarbons implanted experimentally into the salivary glands of rats and guinea-pigs are also known to result in tumour formation.

The evidence that low dose radiation may induce tumours in man is much more convincing and is derived from studies of people exposed to the effects of the atomic bomb at Hiroshima. In this group of individuals, the incidence of tumours at all sites is significantly greater than that expected in persons who have never been previously exposed to radiation (Ju, 1968; Takeichi, Hirose and Yamamoto, 1976).

Surgical pathology

Approximately 80% of all salivary tumours arise in the parotid gland, the remainder being distributed between the submandibular, sublingual and minor salivary glands. Eighty per cent of tumours in the parotid gland are benign, whereas in the submandibular the figure drops to 60% and at other sites to something just under 50%.

Of the benign tumours which reside in the parotid, about 80% are pleomorphic adenomata and the remainder a conglomeration of monomorphic adenomata, Warthin's tumours otherwise known as cystadenomata or papillary cystadenomata lymphomatosum, oxyphil adenomata or oncocytomata, and vascular and lymphatic swellings.

In the submandibular gland and at other sites, the only benign tumour to occur with any degree of frequency is the pleomorphic adenoma or mixed tumour.

Benign tumours in the parotid seem to occur most commonly in the lower posterior part of the gland, namely in that portion which fills the retromandibular sulcus. They may present less commonly in the preauricular region or even further forwards over the masseter, or alternatively in the deep lobe. Tumours at these sites seem to have a slightly greater tendency to become malignant and should therefore always be viewed with a modicum of suspicion.

There is no obvious site of election in the submandibular gland for mixed tumour, and more often than not the tumour is found at operation to be larger than previously suspected, virtually replacing the entire gland. Benign tumours are rare in the sublingual gland, but not at all that uncommon in the hard and soft palates where they are nearly always pleomorphic adenomata. At sites other than the parotid, the distinction between benign and malignant is not always easy to make on clinical grounds, and a relatively common alternative which may be confused with mixed tumour is adenoid cystic carcinoma.

There is a group of tumours in which both clinical and histological characteristics are difficult to reconcile with the picture of malignancy and yet their progression ultimately
proves to be overtly nefarious. Included in this group are the acinic cell tumours and some mucoepidermoid tumours.

**Vascular and lymphatic malformations**

Although not tumours in the accepted sense of the word, vascular and lymphatic malformations are very much benign surgical swellings and merit inclusion in this section.

The vascular type differs from the lymphatic in certain specific respects. Such tumours tend to be more obviously confined to an anatomical compartment by comparison with lymphangiomata and, although lacking a proper capsule, their limits are relatively easy to define. The larger the vascular spaces within the swelling, the greater the tendency for there to be significant feeding vessels originating from recognizable local arteries.

Haemangiomata are to be found in the parotid, and may make their appearance in that gland at birth. The congenital variety presents as large bilateral bluish spongy swellings which tend to swell when the infant cries, coughs, or is placed horizontally. They are often associated with haemangiomata elsewhere, notably on the lips and in the subglottic compartment of the larynx. There is some evidence that this type of haemangioma will atrophy as the infant grows, the age of two usually being considered as the turning point (Williams, 1975). Other parotid haemangiomata may appear later, in which case atrophy cannot be expected and surgery is required.

Vascular malformations are also seen in the submandibular and sublingual glands and may then present beneath the oral mucosa.

Lymphatic malformations are by contrast no respecters of anatomical compartments. They are diffuse, occupy large tracts of tissue and cross from one plane to another. They lack a precise boundary, infiltrate muscles, glands, and completely engulf nerves and vessels.

Lymphangiomata of the parotid, for example, surround the facial nerve, infiltrate the temporalis muscle and fascia, spread downwards into the sternomastoid, and forwards and downwards into the submandibular gland.

Those which are primarily related to the submandibular gland invade the mylohyoid, the muscles of the tongue, and even the intraoral mucous membrane.

They are difficult to remove because of their indeterminate boundaries, their far-flung and deep-seated extensions, and their tendency to engulf vital structures. It is always advisable to dissect out important nerves beyond the periphery of the malformation and then to follow them into the swelling itself.

Sometimes these malformations appear to be an admixture of vascular and lymphatic tissue which bleeds excessively during their removal.
**Pleomorphic adenoma**

This is the commonest of all benign tumours and is characterized by slow growth and a clinically benign course. It is essentially an epithelial tumour of complex morphology, possessing epithelial and myoepithelial elements arranged in a variety of patterns and embedded in a mucopolysaccharide stroma. Its capsule is the result of fibrosis of the surrounding salivary parenchyma which is compressed by the tumour, and is referred to as a false capsule. Since the capsule is formed in response to expansion by the neoplasm, it is frequently incomplete, and tumour may be seen projecting through the dehiscences as small bosselations which contact the surrounding gland (Eneroth, 1964).

These projections are sometimes seen in histological sections as small outlying foci of tumour, seemingly separated from the parent tumour by normal glandular tissue, and it is this appearance which prompted the belief at one time that mixed tumours were multifocal in origin. This view was eventually rejected when serial sections demonstrated continuity of the excrescences with the main body of the tumour.

Mixed tumours are often soft in consistency, almost with a myxomatous appearance, and may be the seat of cystic or haemorrhagic degeneration, features which make them susceptible to rupture when handled too enthusiastically.

Their lack of a complete capsule and their softness are compelling reasons for removing them with as wide a margin as possible, and this is generally possible at all the sites from which these tumours arise. The operation of enucleation, which has not been largely abandoned in favour of parotidectomy, resulted in an unacceptably high rate of recurrence, often as high as 40% over a 25- or 30-year period. But, some mixed tumours of the parotid cannot be removed adequately with a satisfactory surrounding margin. Very large tumours replacing the parotid parenchyma, or those sitting on the facial nerve or external auditory canal, are just such instances and the operation to remove them can only be described as a compromise between a parotidectomy and enucleation.

Interestingly, those pleomorphic adenomata which originate from the deep lobe of the parotid and occupy the parapharyngeal space do not commonly recur after surgery in spite of the fact that the operation to remove them is little better than a grand enucleation. The reason for this could well be their tendency to acquire a thicker and more complete capsule than is usually the case at other sites.

The concept of removal with a margin is eminently practicable when mixed tumours arise in the submandibular gland, since it takes many years for the neoplasm to break through the capsule of the gland. It is also feasible when the site of origin is the hard or soft palate, although a surgically created fistula may be necessary to ensure adequate removal.

A recurrent mixed tumour is to be feared, since it presence not a discrete mass but a multiplicity of nodules. In the case of the parotid, it is common to find nodules in the previous scar, subcutaneous tissue, both superficial and deep parotid parenchyma, the sheath of the facial nerve and the perichondrium of the external meatus. Further surgical attempts are often fruitless given the widespread nature of the condition, and may well cause damage to the facial nerve.
Pleomorphic adenomata may, after several years' growth, become overtly malignant. It is estimated that the incidence of malignancy is approximately 6%, although the factor which more directly determines this likelihood is the age of the tumour.

**Warthin's tumour**

This is a tumour primarily, although not exclusively, of men, seen generally in middle and old age. Curiously, it is nearly always found in heavily built or obese individuals with short fat necks and prominent jowls. It is occasionally bilateral and often more than one tumour is found in any given gland.

Ovoid in shape, it is rather like a lymph node and is characteristically situated in the tail of the parotid gland.

Histologically, it is made up of areas of lymphoid tissue intermingling with cystic spaces lined by a tall tubular or papillary epithelium. The presence of lymphoid tissue within the tumour makes it susceptible to inflammation, often secondary to upper respiratory tract infections, with enlargement of the tumour, pain and tenderness. The clinical features therefore include fluctuation in size and intermittent pain. Rarely, the inflammatory reaction is so severe as to cause the swelling to enlarge massively and to ulcerate, even to the extent that a malignancy may be suspected.

Removal with a margin such as one would practise for a mixed tumour will generally suffice. When cut across and viewed by the naked eye, the fresh specimen is often found to contain a viscous chocolate-like fluid. Recurrence after surgery is very rare.

**Oxyphil adenoma (oncocytoma)**

This is a rare benign tumour composed of large pleomorphic eosinophilic cells replete with mitochondria not unlike the oncocytes seen in ageing normal salivary glands. It nearly always arises from the outer part of the parotid gland, but there are reports of it developing in the submandibular gland and oral cavity. It may exceptionally become malignant.

**Benign lymphoepithelial lesion**

This is considered by some to be a solid variant of Warthin's tumour, and by others not to be a tumour at all. As its name implies, it is composed of lymphoid and epithelial components which form a well-defined tumour-like mass indistinguishable from any other benign tumour. It may surround nerve fibres and thus appear malignant, but a frozen section usually settles the issue.

It too has a malignant counterpart which is rare.

**Symptoms**

Patients with benign salivary tumours generally complain of little more than the presence of a swelling, whose growth is so slow as to be barely perceptible from one year to the next. A sudden increase in size strongly suggests malignant transformation, although
infection of a Warthin's tumour may be one explanation. Similarly, pain must be regarded as unusual in benign tumours unless infection or haemorrhage have occurred in a cyst.

Pressure from a benign tumour never causes facial paralysis, even when the nerve is engulfed by the growth as in benign lymphoepithelial hyperplasia. Its presence signifies either malignancy, tuberculosis, or sarcoidosis. Enlargement of the subparotid nodes is a frequent accidental finding during operations for the removal of benign tumours and is invariably due to non-specific reactive hyperplasia.

Examination

The precise anatomical site of the lesion must be defined during the clinical examination to establish whether a swelling is likely to have arisen from a salivary gland or not. Swellings in the retromandibular sulcus, the immediate preauricular region, and over the masseter are, in most cases, of parotid origin. Lumps in the submandibular triangle either arise in the salivary gland or turn out to be enlarged lymph nodes. Non-ulcerative swellings of the oral cavity, especially on the hard and soft palates, but also in the floor of the mouth or palatofacial region, are likely to be benign salivary tumours. In the last case, the tumour is likely to have arisen from the deep lobe of the parotid or close to that structure and to have occupied the parapharyngeal space. Exceptionally, the tumour arises from the fauces and grows backwards and laterally into the parapharyngeal space.

Vascular or lymphatic swellings are softer and spongier than most benign tumours and may present with a blue or purplish tinge, while exceptionally the larger variety may produce a vascular hum audible by auscultation.

In the parotid gland pleomorphic adenomata present as round, firm, reasonably well demarcated tumours, with a tendency to nodularity as they grow. Their site of election is between the ascending ramus of the mandible anteriorly, and the mastoid process and sternomastoid posteriorly, towards the tail of the gland.

Occasionally they arise in the immediate preauricular region, where they tend to be small and, less commonly, still further forward.

Recurrent mixed tumours present as multiple nodules, or as a nodular thickening, although what is generally palpable represents a fraction of the full extent of the disease, much of which is at the microscopical level.

Warthin's tumours, by contrast, lie almost invariably in the lower pole of the gland, are ovoid in shape, and vary in consistency between soft and firm, depending on whether they have been exposed to previous inflammation or not. They may well be bilateral.

It is often difficult to distinguish between a tumour arising within the submandibular gland or an enlarged node close to the gland or on its outer surface. Bimanual palpation is essential to differentiate the two, since a node lying on the outer surface of the salivary gland is unlikely to be adequately palpated by a finger in the mouth, whereas a tumour of the gland itself is more readily compressible bimanually. Pleomorphic adenomata of the submandibular
gland are usually large, quite hard and nodular, but may be confused with a slowly growing malignancy such as an adenoid cystic carcinoma.

Tumours of the palate are often fusiform, firm to hard and nodular. Again the distinction between mixed tumour and adenoid cystic carcinoma may be difficult to make. Apart from the obvious difference in the growth rate, the latter tumour is often discoloured by telangiectases or bleeding into the tumour.

For parapharyngeal masses presenting in the faucial region, bimanual palpation is required to elicit the classical signs of ballottement between a finger in the mouth and a hand on the neck. If the tumour only occupies a small segment of the deep lobe of the parotid, there will be little external fullness of this gland, but if it is large and part of the deep lobe, the parotid is visibly and palpably displaced outwards.

**Investigations**

There are only three useful investigations in benign salivary tumours: sialography, computerized or magnetic scanning, and fine needle aspiration.

Sialography is of limited value. Benign tumours of some size appear as punched out areas against a background of contrast-filled ducts and acini. Definition of the precise anatomical site is often difficult, but the distinction between inflammatory disease and neoplasm is easier to make.

If the outline of the area which fails to fill with dye is ragged, rather than clean-cut, the possibility that the swelling may be malignant must be considered. Disadvantages of sialography are the difficulty of duct cannulation in many patients, and inadequate or excessive filling leading to acinar rupture, both of which may well confuse the picture.

Before the introduction of computerized scanning, isotope imaging with technetium was practised for a time, but the information gained was so imprecise as to be valueless. It was soon superseded by CT scanning which indicates the site and size of the lesion and, by contrast with sialography, is easy and painless to perform. In conjunction with sialography, it optimizes definition and is especially helpful in establishing the configuration and outline of parapharyngeal space tumours.

Magnetic resonance imaging may well help in the investigation of salivary tumours since it is above all others the ideal investigation for assessing soft tissue neoplasms.

Fine needle aspiration has now finally come to stay. There is little doubt it has a great deal to offer in the hands of a person practised in the interpretation of small samples of tissue. However, evaluation of the findings may be difficult if the aspirate has been contaminated with blood, or even misleading if sampling of the swelling has been haphazard. Under these circumstances, a negative diagnosis for malignancy should never be considered as final.

Whereas open biopsy or large needle biopsy carries the danger of implanting seedlings into the biopsy track, fine needle aspiration appears not to have this disadvantage.
Surgical treatment

Partial parotidectomy

This is the operation most often performed for benign tumours, since most tumours of the parotid gland lie superficial to the facial nerve.

The patient is placed in the supine position with the neck extended and the head turned away from the surgeon. Tilting the table in a head-up position facilitates the surgery by reducing the venous pressure. The towels should be so arranged that the corner of the eye and mouth are accessible for inspection.

The incision begins in the preauricular crease and descends to the point where the lobule joins the skin of the face, at which point it inclines backwards at first, then downwards and forwards into a cervical skin crease well below the mandible. A skin flap is elevated to uncover the area which the parotid occupies. The posterior extremity of the gland is separated from the external auditory canal, the mastoid process and sternomastoid muscle so that sufficient room is created to find the surgical landmarks for the facial nerve. Using the tympanomastoid sulcus as the starting point, the nerve is found and followed into the parotid gland.

That part of the parotid gland which lies superficial to the facial nerve is then peeled off the nerve together with the enclosed tumour. The wound is then drained and the incision closed in layers. Tumours of the deep lobe of the parotid and parapharyngeal space are generally removed, either by total conservative parotidectomy, a submandibular approach or a transmandibular technique (Shaheen, 1986).

Removal of the submandibular gland

The position of the patient is the same as for parotidectomy. A horizontal skin incision is made just above the hyoid bone preferably in a skin crease. The incision is deepened through subcutaneous fat, platysma, and deep cervical fascia until the surgical capsule at the lower limit of the gland is reached. This is then opened along the lower border of the gland and a large superior flap elevated in the plane between the surgical capsule and the gland. The facial vessels are ligated both at the inferolateral corner of the gland and at its upper border.

The gland is mobilized until it is pedicled on its deep part, whereupon the mylohyoid muscle is retracted forwards to permit separation of the gland from the lingual nerve. Finally, the duct is severed and the wound closed in layers with drainage (Shaheen, 1986).

Removal of the sublingual gland

This may be performed intraorally after injecting an adequate volume of a vasoconstrictor solution immediately beneath the mucous membrane to lessen bleeding.

An incision medial to the line of Wharton's duct provides access to the gland, but care has to be taken to avoid injuring the lingual nerve which winds round the duct. With the
submandibular duct and nerve retracted medially out of the way, the gland and tumour may be removed.

**Removal of a pleomorphic adenoma from the hard palate**

The patient should be placed in the supine position with a sandbag placed beneath the shoulders and the head fully extended. The table should be in the Trendelenburg position so that, with a Boyle-Davis gag separating the jaws, a clear view of the operation site can be obtained. Illumination with a headlight and adequate suction are imperative.

The incision through the mucous membrane down to the bone of the hard palate is best made with the cutting diathermy to lessen the bleeding, and should be sited a short distance away from the tumour to ensure an adequate surrounding margin of normal tissue.

Enucleation of the tumour by dissection from the underlying bone is not safe unless the bone is drilled away virtually through its whole thickness. The safest approach is to fenestrate the palate so that the tumour is cut out *en bloc*. In either case, provision should be made for the wearing of a protective denture, with or without an obturator, depending on the exigencies of the case.

**Recurrent pleomorphic adenoma**

Recurrence of this tumour occurs when it is enucleated without a margin of healthy tissue or when it is breached accidentally during the course of an operation to remove it, or deliberated as with a biopsy. Spillage of tumour is apt to occur when the tumour is particularly large and tense, or when it is awkwardly situated. Examples of the latter are large tumours impacted between the parotid and the mastoid, deep-seated growths, or tumours sitting intimately on the facial nerve.

As previously mentioned, recurrence presents as a multiplicity of nodules scattered, not only within the parotid parenchyma, but also in the overlying tissues.

If the initial operation which led to the recurrence was simply an enucleation, salvage surgery in the form of a superficial or even total conservative parotidectomy is generally feasible. If, however, the first operation was a parotidectomy with exposure of the nerve, any subsequent attempt to excise residual disease without compromising the facial nerve is difficult to say the least. Under such circumstances, it may be possible to find the nerve in the mastoid, but even then the process of following it through the sea of collagen and neoplastic nodules which is so typical of recurrent mixed tumour is complicated and hazardous. More often than not, the nerve and its branches disappear from view into the surrounding fibrous tissue so that further dissection is technically impossible without damaging the nerve. Given this sort of scenario and the fact that neoplastic nodules are commonly embedded within the sheath of the facial nerve, the surgeon might be forgiven for opting for a radical parotidectomy.

There is, however, an alternative strategy which offers the possibility of preventing recurrence in high risk cases and of dealing with it once it has become established.
It has been common knowledge for many years that postoperative radiotherapy following a limited operation such as enucleation will reduce the recurrence rate of mixed tumours to that expected after parotidectomy, namely about 3-4% (Rafla, 1970).

The rationale for this is that the operation serves to remove the bulk of the disease, leaving scattered microscopical residues which are more readily amenable to sterilization, notwithstanding the known relative radioresistance of mixed tumours.

The application of this philosophy to the management of those patients undergoing adequate surgery by present-day standards, but who are deemed to be at risk, should minimize the risk of recurrence still further. Such cases would include patients with tumours sitting on the facial nerve, those stuck to the meatus, or those which have ruptured during removal.

In practice, the irradiation of such patients has proved to be satisfactory and has eliminated recurrences altogether.

The same approach can be adopted for patients who have an established recurrence. Here the approach is to remove all macroscopical disease by total conservative parotidectomy if this procedure is feasible and to follow on with radiotherapy (Shaheen, 1976).

In those patients in whom the integrity of every branch of the facial nerve is impossible to safeguard, the surgeon cuts his losses by resorting to a total parotidectomy, with every attempt being made to preserve as much of the nerve as possible. Deficits are restored by immediate nerve grafting, and surgery is followed some weeks later by radiotherapy.

Generally, between 4000 and 5000 cGy are given by lateral field and preferably by linear accelerator. The results of treating established recurrences by this method have been equally rewarding, recurrence having been abolished.

This naturally raises the ethical question of whether it is proper to irradiate patients with benign disease. There are indeed isolated reports of patients developing cancers of the parotid gland after radiotherapy, but many of these date back to the era when low dosage therapy was used in the management of lymphoid tissue hypertrophy in childhood. Most radiotherapists with whom the matter can be discussed are sceptical about the likelihood or a properly administered course of irradiation eventually causing cancer of the parotid, although it is conceivable that adjacent areas such as the postnasal space might be at risk.

The pros and cons of the issue will doubtless continue to be debated, but in the final analysis the decision whether to use radiotherapy or not has to be a personal one.
Complications of surgery

Parotidectomy

Haematoma

This is fairly common in view of the dead space resulting from the removal of a substantial segment of glandular tissue. A large suction drain sited away from the facial nerve together with adequate pressure from a suitable dressing will minimize this complication.

Facial weakness

This is generally temporary if it occurs after an uneventful operation. It usually affects the mandibular and frontal branches of the facial nerve and is more prone to occur in the elderly, in those with slender as opposed to stout facial nerves, and when there has been an unusual degree of trauma in the vicinity of the nerve. Pressure or traction on the nerve, constant suction on its surface, excessive dryness of its sheath, and heat from the coagulating diathermy are all factors contributing to facial weakness.

Anaesthesia

Anaesthesia of the lower half of the pinna and preauricular skin results from division of the great auricular nerve and is therefore an inevitable sequel of parotidectomy. The area of numbness gradually diminishes in size and ultimately becomes confined to the lobule of the pinna. Sensation in the surrounding skin rarely returns to complete normality and some degree of hyperaesthesia is usual. In a few patients, the degree of increased sensitivity assumes distressing proportions and is attributable to the presence of an amputation neuroma of the great auricular nerve. Excision of the neuroma and alcohol injection into the cut end of the nerve will resolve the problem by substituting complete anaesthesia for hyperaesthesia.

Fistula formation

This is very uncommon and probably results from an overproduction of saliva in the remaining glandular tissue. A pressure bandage and continuous suction suffice to dry up the wound and permit normal hearing.

Gustatory sweating (Frey's syndrome)

This condition is though to be due to misrouting of parasympathetic secretomotor fibres into cutaneous nerves during the healing phase following parotidectomy. The precise origin of these fibres is uncertain, having been thought to have come by way of the otic ganglion and to have accompanied the auriculotemporal nerve to its final destination int he parotid. There is some reason to suppose that this is not the only source of secretomotor fibres and that some may come by way of the facial nerve itself.

Most patients develop some degree of Frey's syndrome, but in many the condition is so mild as to be barely noticeable. The complaint is of sweating in the preauricular and subparotid regions at meal times, although many patients imagine at first that it is an escape
of liquid through a small opening in the surgical scar. It may on occasion be so bad as to be a severe social embarrassment.

Contrary to reports in the literature, tympanic neurectomy has not been successful in controlling the condition and the effectiveness of other techniques, such as the interposition of fascial grafts between skin and the underlying parotid bed, is also doubtful.

More recently, pilocarpine cream 1% and aluminium hydrochloride solution have been applied with some success (Shaheen, 1984).

**Submandibular gland excision**

**Paralysis of the depressor anguli oris**

This is due to damage to the marginal mandibular branch of the facial nerve and is best avoided by gaining access to the right plane of dissection, well away from the nerve, from the very outset.

Attempts to dissect out the mandibular branch are likely to do more harm than good, and if the nerve is badly traumatized, the likelihood of recovery is remote.

**Damage to the hypoglossal and lingual nerves**

Both these complications are uncommon and generally speaking cannot be rectified (Shaheen, 1984).
Chapter 20: Malignant salivary tumours

Michael Gleeson

Malignant neoplasms arising in salivary tissue are uncommon. While fewer than 3% of all neoplasms originate in salivary glands (OPCS Cancer Statistics, 1983), at least 75% of these are benign. Most salivary gland tumours are pleomorphic adenomata which, despite their benign nature, pose considerable management problems as they recur locally if inadequately resected, ruptured or biopsied and also because of their potential to undergo carcinomatous change. The malignant character of some salivary tumours has only recently been appreciated as their natural history must be measured in decades rather than years. The slow growth pattern of these tumours does not lessen their malignant nature. Despite treatment they often recur or manifest as metastatic disease at a relatively late stage and thus exert a considerable morbidity and mortality.

Normal structure and pathological classification

Normal salivary tissue has a complex structure consisting of secretory cells, which may be either serous or mucous, arranged in acini and drained by a duct system. The major glands - the parotid, submandibular and sublingual - are composed of collections of acini of differing character which secrete into a complex duct system. Around the acini lie myoepithelial cells which display features common to both epithelial and smooth muscle cells. Their probable function is to propel the contents of the acini along the duct system. While the parotid gland is predominantly serous, the submandibular and sublingual glands produce mucoid secretions. There are numerous collections of salivary acini scattered throughout the oral mucosa which are referred to as the minor glands.

The histopathology of salivary neoplasms poses many problems to the pathologist. Like most other tumours they are rarely homogeneous in structure, many present a variety of patterns and some are so uncommon that few pathologists have the opportunity to acquire expertise in their diagnosis.

A widely recognized system of classification has been established (Thackray and Sobin, 1972) which divides these lesions into four sub-groups: epithelial, non-epithelial, unclassified tumours and a group of conditions that may give rise to diffuse or discrete salivary enlargement (Table 20.1). Epithelial tumours predominate in all series.

Distribution

Salivary tumours are more common in the parotid gland than in any of the other glands. The frequency of malignant tumours varies according to the site, being relatively lower in the parotid than elsewhere (Table 20.2). Malignancy is far more frequent in the submandibular, sublingual and minor glands. Furthermore, even within the minor glands groups there is much variation in the proportion of frankly malignant tumours, the lip being the least common site, while those tumours arising in the sublingual glands, albeit rare, are nearly always malignant (Table 20.3). The most common sites for tumours of the minor glands are the palate, lip and buccal mucosa.
Benign tumours (the mono- and pleomorphic adenomata) outnumber malignant tumours at all sites except in the sublingual glands. The available data show that the relative incidence of the more common varieties of malignant neoplasm differs widely between the major and minor glands (Table 20.4). Thus mucoepidermoid tumours and adenoid cystic carcinomata are the most common neoplasms overall.

Table 20.1 WHO histological classification of salivary gland neoplasms

<table>
<thead>
<tr>
<th>Epithelial tumours</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Adenomata</td>
<td></td>
</tr>
<tr>
<td>pleomorphic</td>
<td></td>
</tr>
<tr>
<td>monomorphic</td>
<td></td>
</tr>
<tr>
<td>adenolymphoma</td>
<td></td>
</tr>
<tr>
<td>oxyphilic adenoma</td>
<td></td>
</tr>
<tr>
<td>others</td>
<td></td>
</tr>
<tr>
<td>Mucoepidermoid tumour</td>
<td></td>
</tr>
<tr>
<td>Acinic cell tumour</td>
<td></td>
</tr>
<tr>
<td>Carcinomata</td>
<td></td>
</tr>
<tr>
<td>adenoid cystic carcinoma</td>
<td></td>
</tr>
<tr>
<td>adenocarcinoma</td>
<td></td>
</tr>
<tr>
<td>epidermoid carcinoma</td>
<td></td>
</tr>
<tr>
<td>undifferentiated carcinoma</td>
<td></td>
</tr>
<tr>
<td>carcinoma ex pleomorphic adenoma</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Non-epithelial tumours</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Haemangioma</td>
<td></td>
</tr>
<tr>
<td>Lymphangioma</td>
<td></td>
</tr>
<tr>
<td>Neurofibroma</td>
<td></td>
</tr>
<tr>
<td>Lymphoma</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Unclassified tumours</th>
<th></th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Allied conditions</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign epithelial lesion</td>
<td></td>
</tr>
<tr>
<td>Sialosis</td>
<td></td>
</tr>
<tr>
<td>Oncocytosis.</td>
<td></td>
</tr>
</tbody>
</table>

Malignant salivary gland tumours are more frequent in women than men and have a peak age of incidence in the seventh decade. Adenomata and non-epithelial tumours present at a much younger age and, indeed, many have been reported in infancy. Even discounting congenital anomalies, the salivary tumours arising in infancy have a different distribution from those of adult life (Table 20.5). Pleomorphic adenoma still predominates but, in infants, the mucoepidermoid tumour is the most common malignancy (Byers, Piorkowski and Luna, 1984).
Epidemiology

The reported incidence of salivary gland tumours is variable, for example, 2.5 per million in Norway, 7.5 per million in Sweden and 15 per million in Caucasians living in the USA (Dorn and Cutler, 1959; Soder, 1973). The developing nations would appear to experience a similar incidence (Davies, Dodge and Burkitt, 1964; Loke, 1967).

Table 20.2 Frequency (%) of all primary epithelial salivary gland tumours and malignancy (%) analysed by site

<table>
<thead>
<tr>
<th>Site</th>
<th>Absolute no.</th>
<th>Frequency (%)</th>
<th>Malignant (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parotid</td>
<td>1756</td>
<td>72.9</td>
<td>14.7</td>
</tr>
<tr>
<td>Submandibular</td>
<td>257</td>
<td>10.7</td>
<td>37.0</td>
</tr>
<tr>
<td>Sublingual</td>
<td>7</td>
<td>0.3</td>
<td>85.7</td>
</tr>
<tr>
<td>Minor (oropharyngeal) glands</td>
<td>336</td>
<td>14.0</td>
<td>46.4</td>
</tr>
<tr>
<td>Unknown</td>
<td>54</td>
<td>2.2</td>
<td>0.0</td>
</tr>
<tr>
<td>Total</td>
<td>2410</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

The peak incidence of salivary neoplasms is in the sixth and seventh decades for both men and women. In the major series, salivary tumours have been found to be slightly more common in females than males. This tendency, while present throughout life, becomes most marked in the eighth and ninth decades when there is a female predominance in the population and ratios of 1.6:1 and 1.9:1 are recorded respectively.

Table 20.3 Distribution of salivary tumours in minor glands and malignancy (%)

<table>
<thead>
<tr>
<th>Site</th>
<th>Absolute no.</th>
<th>% of total</th>
<th>Malignant (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Palate</td>
<td>183</td>
<td>54.3</td>
<td>47.0</td>
</tr>
<tr>
<td>Lip</td>
<td>71</td>
<td>21.1</td>
<td>26.7</td>
</tr>
<tr>
<td>Buccal mucosa</td>
<td>38</td>
<td>11.3</td>
<td>50.0</td>
</tr>
<tr>
<td>Tongue</td>
<td>12</td>
<td>3.6</td>
<td>92.0</td>
</tr>
<tr>
<td>Pharynx</td>
<td>12</td>
<td>3.6</td>
<td>50.0</td>
</tr>
<tr>
<td>Tonsil</td>
<td>6</td>
<td>1.8</td>
<td>50.0</td>
</tr>
<tr>
<td>Retromolar</td>
<td>5</td>
<td>1.5</td>
<td>50.0</td>
</tr>
<tr>
<td>Alveolar ridge</td>
<td>5</td>
<td>1.5</td>
<td>100.0</td>
</tr>
<tr>
<td>Tuberosity</td>
<td>3</td>
<td>0.9</td>
<td>100.0</td>
</tr>
<tr>
<td>Ethmoid</td>
<td>1</td>
<td>0.3</td>
<td>100.0</td>
</tr>
<tr>
<td>Total</td>
<td>336</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Predisposing and associated factors

Several possible predisposing factors have been postulated including race, diet, occupation, Epstein-Barr virus, etc (Lanier et al, 1976; Lennox et al, 1978; Saemundsen et
al, 1982; Spitz et al, 1984). Only previous radiation has been shown convincingly to have any significant influence. In a study of survivors of the Hiroshima atomic bomb, Takeichi, Hirose and Yamamoto (1976) found that the incidence of benign and malignant salivary gland tumours was 2.6 times higher than that of a comparable non-exposed population, while that of malignant tumours was 10 times greater. This risk of salivary malignancy was higher in those closes to the hypocentre of the explosion and in those returning early to the city. As with similarly induced thyroid neoplasms there would appear to be a latent period of 15-25 years after exposure before development of the tumour. In a later study of the same population it was established that this increased susceptibility was shared by both the parotid and submandibular glands (Takeichi et al, 1983). Patients given low dosage radiotherapy to the tonsil and nasopharyngeal area for benign conditions have also shown identical trends (Shore-Freedman et al, 1983).

**Table 20.4 Frequency (%) of primary epithelial salivary gland tumours by site**

<table>
<thead>
<tr>
<th>Tumour type</th>
<th>Parotid</th>
<th>Submand</th>
<th>Sublingual</th>
<th>Minor</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Benign</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Plasmacytoma</td>
<td>63.3</td>
<td>59.5</td>
<td>0.0</td>
<td>42.9</td>
</tr>
<tr>
<td>Adenolymphoma</td>
<td>14.0</td>
<td>0.8</td>
<td>0.0</td>
<td>0.0</td>
</tr>
<tr>
<td>Oxyphil adenoma</td>
<td>0.9</td>
<td>0.4</td>
<td>0.0</td>
<td>0.0</td>
</tr>
<tr>
<td>Other monomorphics</td>
<td>7.1</td>
<td>1.9</td>
<td>14.2</td>
<td>11.0</td>
</tr>
<tr>
<td><strong>Malignant</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mucoepidermoid ca</td>
<td>1.5</td>
<td>1.6</td>
<td>0.0</td>
<td>8.9</td>
</tr>
<tr>
<td>Acinic cell tumour</td>
<td>2.5</td>
<td>0.4</td>
<td>0.0</td>
<td>1.8</td>
</tr>
<tr>
<td>Adenoid cystic ca</td>
<td>2.0</td>
<td>16.8</td>
<td>28.6</td>
<td>13.1</td>
</tr>
<tr>
<td>Adenocarcinoma</td>
<td>2.6</td>
<td>5.0</td>
<td>14.2</td>
<td>12.2</td>
</tr>
<tr>
<td>Epidermoid ca</td>
<td>1.1</td>
<td>1.9</td>
<td>0.0</td>
<td>1.2</td>
</tr>
<tr>
<td>Undifferentiated ca</td>
<td>1.8</td>
<td>3.9</td>
<td>14.2</td>
<td>2.1</td>
</tr>
<tr>
<td>Ca ex pleom ad</td>
<td>3.2</td>
<td>7.8</td>
<td>28.6</td>
<td>7.1</td>
</tr>
<tr>
<td><strong>Total no of cases</strong></td>
<td>1756</td>
<td>257</td>
<td>7</td>
<td>336</td>
</tr>
</tbody>
</table>

Some authors have found that patients with salivary tumours are more likely to develop a second primary tumour at any site. A link with hormone-dependent tumours has been suggested as the breast is structurally similar to salivary tissue. Women are more likely to develop a later breast tumour, but in men skin cancer is the only growth with a significantly increased incidence, despite early claims suggesting an association with prostatic carcinoma (Prior and Waterhouse, 1977; Abbey et al, 1984; Spitz et al, 1985).

**Histopathology and natural history of the common tumour types**

**Mucoepidermoid tumour**

Mucoepidermoid tumour is the most common salivary neoplasm to arise in childhood but only the second most common salivary malignancy overall (see table 20.5). The parotid
and minor glands are the most commonly affected sites, and of the latter group, the glands in the palate predominate.

Table 20.5 Distribution of juvenile salivary tumours by site and type

<table>
<thead>
<tr>
<th>Tumour type</th>
<th>Parotid</th>
<th>Submand</th>
<th>Subling</th>
<th>Intraoral</th>
<th>Lip</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pleomorphic</td>
<td>26 (55%)</td>
<td>14 (29.8%)</td>
<td>0</td>
<td>4 (8.5%)</td>
<td>0</td>
<td>3 (6.4%)</td>
</tr>
<tr>
<td>Warthin's</td>
<td>1 (100%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Monomorphic</td>
<td>1 (100%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Mucoepiderm</td>
<td>3 (50%)</td>
<td>2 (33%)</td>
<td>1 (17%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Acinic cell</td>
<td>1 (100%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Adenoid cystoma</td>
<td>1 (100%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Undifferentiated</td>
<td>1 (33%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Unclassified</td>
<td>1 (100%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Others</td>
<td>9 (90%)</td>
<td>1 (10%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>44 (61%)</td>
<td>17 (24%)</td>
<td>6 (8%)</td>
<td>3 (3.5%)</td>
<td>3 (3.5%)</td>
<td>3 (3.5%)</td>
</tr>
</tbody>
</table>

Mucoepidermoid tumours grow slowly and recur locally. Lymph node metastases arise in up to 30% of patients and are present ab initio in 15%. Metastases to the lungs, bones and brain develop in approximately 15%. It has been claimed that recurrence and survival rates are strongly influenced by the histological grading of the tumour and its size at presentation. Low grade tumours are said to be compatible with an 80% 15-year determinate cure rate, while a comparable figure for high grade tumours is reported to be only 33% (Spiro et al, 1978). However, even the most apparently benign tumours can prove to be unexpectedly invasive. It is this feature of the mucoepidermoid tumour that has caused it to be called a carcinoma in some areas of the world.

Mucoepidermoid tumour is composed of two distinct cell types: epidermoid cells and mucous cells, both of which may show varying degrees of differentiation. Mucus is secreted into the stroma of the tumour giving rise to a partially cystic structure. This neoplasm has been graded according to its cellular content. Tumours are designated high grade if 90% or more of their area is made up of tumour cells and less than 10% from intracystic spaces. Low grade tumours display a reversal of this ratio (Evans, 1984). Increased mitotic activity is not necessarily seen even in high grade tumours - a feature partially responsible for the unpredictable nature of this particular neoplasm.

Low grade tumours should be managed by local resection and prolonged follow-up. High grade lesions require more radical resection with adjunctive radiotherapy. Local recurrence normally appears within 12 months of the primary procedure.

**Acinic cell tumour**

Acinic cell tumour accounts for 2.5% of all salivary gland tumours and predominantly affects the parotid gland, where it is said, a very small proportion are found to be bilateral. Like adenoid cystic carcinoma this tumour is very slow growing, a feature that belies its
malignant potential. Local recurrence or the development of metastases after a prolonged disease-free interval are common. This tumour exerts a mortality many years after other salivary neoplasms would have been considered cured. Determinate survival rates of 85%, 65% and 50% for 5, 10 and 15 years respectively, have been reported (Batsakis et al, 1979; Perzin and LiVolsi, 1979; Hickman, Cawson and Duffy, 1984).

Microscopically, a solid pattern is usually seen with varying degrees of organization into an acinar arrangement. Microcystic, papillary-cystic, follicular and poorly differentiated patterns are also recognized, but are rare. Tumours are often not homogeneous and some are multifocal. The constituent cells contain zymogen granules, thus bearing a close resemblance to the serous salivary gland cell.

The management of this tumour is excision of the gland with conservation of all uninvolved nerves. Since nodal disease is uncommon and develops in no more than 10%, elective neck dissection is not indicated.

Carcinomata

Adenoid cystic carcinoma

Adenoid cystic carcinoma is the most common malignant salivary neoplasm. As with the other tumours, its distribution varies between sites. It is far more common in the submandibular, sublingual and minor glands, where it constitutes 16%, 28% and 13% of all neoplasms respectively. However, in the parotid gland only 2% of tumours are of this type (see Table 20.4).

Adenoid cystic carcinoma grows slowly and insidiously with a characteristic propensity for perineural infiltration and spread along the haversian systems and neural canals of bones. This pattern is reflected in the local recurrence and cumulative mortality rates for this tumour, namely 5-year survival - 62%; 10-year survival - 39%; 15-year survival - 26%; and 20-year survival - 21% (Spiro, Huvos and Strong, 1974). Nodal involvement is normally the result of direct spread of tumour rather than by lymphatic spread. Metastases develop late in the disease and are usually pulmonary (Shannon, Allen and Marsh, 1976). Local recurrence is common and appears in at least 50% of patients, but even multiple local recurrences or distant metastases are compatible with prolonged survival (Conley and Dingman, 1974). Indeed, while 33% of patients die within one year of the recognition of their metastatic disease, 20% of those with pulmonary secondaries survive with this increased tumour load for longer than 5 years.

Four histological patterns can be recognized, namely cribriform, basaloid or solid, cylindromatous, and tubular (Batsakis and Regezi, 1979). Type and grade of tumour appear to affect survival. The more solid types are reported to have the worst prognosis, display greater cellular atypia and, thus, are of higher grade than the tubular and cribriform varieties. These latter types are usually better differentiated, of lower grade and are reported to impart a superior survival rate (Perzin, Gullane and Clairmont, 1978; Szanto et al, 1984).

Clinically, adenoid cystic carcinoma usually presents as a painless mass or a submucosal swelling. It is usually non-ulcerated unless it is traumatized. Facial palsy develops
in up to 20% of parotid tumours and palpable lymph node involvement or direct invasion of adjacent tissues is found in 15%.

The management of this tumour is the widest possible excision followed by radiotherapy to improve local control. For a parotid tumour this implies total parotidectomy. The facial nerve is conserved only if there is no evidence of invasion by tumour. Subtotal petrosectomy has been advocated for patients with facial nerve invasion. It is essential to appreciate that the tumour spreads perineurally both distally and proximally. Great care must therefore be given to excision of neural spread in both directions. Part of the mandible and the contents of the infratemporal fossa must also be resected in some of these patients. A radical neck dissection is only indicated for those patients with obvious nodal disease or those in whom a large soft tissue cuff must be removed.

Submandibular disease is best managed by a monobloc resection of the gland encompassing the lingual, hypoglossal and marginal mandibular nerves, together with a suprahyoid nodal clearance. Palatal tumours require wide local resection to include the floor of the maxillary sinus at least. Perhaps it is because this is a relatively simple surgical procedure that disease at this site enjoys slightly better survival rates (Eneroth, Hjertman and Moberger, 1968).

Recurrent tumour is best dealt with by surgery followed by radiotherapy if possible. Advanced and incurable disease is sometimes palliated by radiotherapy which, in a few, makes salvage surgery feasible (Simpson, Thawley and Matsuba, 1984; Matsuba et al, 1984).

**Adenocarcinoma**

This is an uncommon tumour which, by virtue of the overall higher incidence of parotid growths, it most frequently found at that site. However, it constitutes only 25-4% of all parotid neoplasms and forms a much larger proportion of the tumours in other glands - 5% of submandibular tumours and 12-14% of sublingual and minor gland neoplasms (see Table 20.4).

Several histological patterns of adenocarcinoma are now recognized, non-mucin producing types being the most common. Mucinous, papillary, trabecular, clear cell and sebaceous variants are also seen and are more frequent in the minor glands. As a group they display a wide structural range, but have in common neoplastic duct or tubule formation and, by definition, lack any evidence of pre-existing pleomorphic adenoma. Clinicopathological surveys do not implicate any particular histological pattern with increased or decreased survival. The grade of the neoplasm as judged by mitotic rate, cellular pleomorphism and stromal invasion correlates better than the clinical stage and outcome. Most of the high grade tumours are the non-mucin producing type, while the rarer variants are usually of a lower grade. The high grade neoplasms tend to present with locally advanced disease, often with nodal involvement, and are reported to have significantly poorer prognosis (Spiro, Huvos and Strong, 1982).

Clinically, 80% present as masses, half of which are fixed; 85% have been present for less than 5 years, but only 35% for less than 12 months. About 20% are painful but, of those arising in the parotid gland, only 5% produce a facial palsy. Nearly 20% of patients have
nodal disease at presentation and a further 10% develop it later. Failure to control disease is usually manifest as local or regional recurrence but distant metastasis, usually to the lungs, is also common. The clinical management of this tumour is similar to that of the adenoid cystic carcinoma.

**Rare epithelial neoplasia**

Of these only the undifferentiated carcinoma and squamous cell carcinoma are sufficiently frequent to deserve mention.

Undifferentiated carcinoma usually presents in middle age and constitutes 1.8% of all epithelial tumours. These neoplasms can be subdivided into small cell and large cell types, the small cell variety being twice as common as the large cell type (Nagao et al, 1982). Some of the lesions included in this group could be very poorly differentiated adenocarcinoma or epidermoid carcinoma. The acinar arrangement of very poorly differentiated adenocarcinoma can be demonstrated with monoclonal antibodies, thus facilitating a distinction between the groups (Heyderman et al, 1985). Such distinction is essentially academic as this disease is very aggressive and requires radical surgery combined with postoperative radiotherapy to achieve a 5-year survival rate of 20-30%.

Squamous cell carcinoma of the salivary glands is very unusual but represents 1.1% of all salivary epithelial neoplasms. It tends to be restricted to the elderly. From a clinical standpoint, it is essential to establish that such a lesion does not represent a metastasis from a distant or regional site.

**Malignant mixed tumour**

Rapid growth of a pleomorphic adenoma, the development of facial palsy, or the onset of pain, all suggest malignant change. Malignant mixed tumour is a very rare entity and is in fact a collective term for three or more main tumour types. Almost all are adenocarcinomata or undifferentiated carcinomata which are clearly seen to arise close to, or within, a pleomorphic adenoma. This subtype is termed 'carcinoma ex pleomorphic adenoma'. It is an aggressive disease with reported 5-year survival rates of 50%, and all patients died in whom the carcinoma extended more than 8 mm beyond the residual benign tumour, its capsule or invaded bone (Tortoledo, Luna and Batsakis, 1984). Perhaps this is a pessimistic assessment of this disease as many more may have been missed due to the limitations of processing large pleomorphic tumours. Radical local resection and radiotherapy offer the best chance of control.

The other subtypes of malignant mixed tumour are exceptionally rare. A highly malignant, biphasic neoplasm composed of both epithelial and mesenchymal elements - a carcinosarcoma - is occasionally found. Another type of tumour is completely indistinguishable from a benign pleomorphic adenoma but is found to metastasize to the lungs and bones (Batsakis, 1982). By definition, the diagnosis of this neoplasm is made at a time when therapeutic options are very limited. Indeed, experience with this tumour is anecdotal.
Malignant non-epithelial tumours: lymphomata

Lymphomata comprise 40% of non-epithelial tumours and constitute the majority of the malignant lesions in this category. Non-Hodgkin's lymphomata predominate, grade 1 tumours being commoner than grade 2. Three-quarters of these lymphomata are found in the parotid gland, and it is therefore possible that they arise in lymph nodes normally present within the gland. However, for staging purposes they are categorized as an extranodal site. Most lymphomata arise between the fifth and seventh decades of life and some are found in association with a benign lymphoepithelial lesion. There is little evidence to suggest that Sjögren's syndrome predisposes to the development of a primary salivary lymphoma.

Clinically, these lesions are firm and rapidly enlarging, and most give little more than a 6-month history. Pain and facial palsy are not prominent features, but the development of regional lymph node involvement is to be expected in those with a delayed presentation.

In some instances, diagnostic suspicion may be so high as to justify open biopsy of a regionally involved node (Watkin, MacLennan and Hobsley, 1984), however, in view of the comparative rarity of these lesions this practice cannot be recommended for the inexperienced surgeon. The clinical stage and histological type of the lymphoma dictate the treatment regimen. Median survival for patients with this disease is 4 years. However, unlike other salivary tumours, advanced clinical stage does not appear to influence prognosis (Gleeson, Bennett and Cawson, 1986).

Management of malignant salivary tumours

Very few salivary tumours have specific features which are pathognomonic. Open biopsy is almost totally contraindicated in these lesions, the exception being neoplasms of the minor glands. Tumours spilt by biopsy will seed the area and result in the development of multiple recurrences within the biopsy scar. However, the precise histological diagnosis can only be made from tissue obtained at the operation. Despite this, a strong suspicion of malignancy can be aroused by the clinical history, examination and preoperative investigations.

Most salivary neoplasms arise as painless masses which grow slowly. Rapid growth or acceleration of growth in a pre-existing lesion is a strong indication of malignancy, as is pain. It is almost impossible to distinguish by palpation alone whether a salivary mass is benign, malignant or inflammatory. Very few salivary tumours, including those that arise in the minor glands, present as ulcers from the outset. Facial, lingual and hypoglossal nerve palsies or evidence of infratemporal fossa infiltration (trismus) denote malignancy, but are uncommon. Similarly, enlarged regional lymph nodes or distant metastases, although indicative of malignancy are a most uncommon presentation.

Despite frequent initial uncertainty about the character of the neoplasm it can always be clinically staged. Significant features for staging purposes are tumour size, local extension or fixation, neural involvement, nodal disease and distant metastases. The criteria for both the TNM and clinical staging systems are shown in Table 20.6 (Levitt et al, 1981).
The preoperative and intraoperative diagnosis of these lesions relies upon the interpretation of radiographs, aspiration cytology and frozen section histopathology, all considered in the context of the clinical findings.

**Table 20.6 TNM classification system for salivary tumours**

<p>| | | |</p>
<table>
<thead>
<tr>
<th></th>
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<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Tx</td>
<td>Recurrence / unstageable.</td>
<td></td>
</tr>
<tr>
<td>T0</td>
<td>No evidence of primary tumour.</td>
<td></td>
</tr>
<tr>
<td>T1</td>
<td>0.1-2.0 cm in diameter, without significant local extension.</td>
<td></td>
</tr>
<tr>
<td>T2</td>
<td>2.1-4.0 cm in diameter, without significant local extension.</td>
<td></td>
</tr>
<tr>
<td>T3</td>
<td>4.1-6.0 cm in diameter, without significant local extension.</td>
<td></td>
</tr>
<tr>
<td>T4a</td>
<td>&gt; 6 cm in diameter, without significant local extension.</td>
<td></td>
</tr>
<tr>
<td>T4b</td>
<td>Any size with significant local extension.</td>
<td></td>
</tr>
<tr>
<td>N0</td>
<td>No evidence of palpable regional nodes, including palpable but not suspicious regional nodes.</td>
<td></td>
</tr>
<tr>
<td>N1</td>
<td>Evidence of regional node involvement, including palpable and suspicious regional nodes.</td>
<td></td>
</tr>
<tr>
<td>Nx</td>
<td>No assessment of regional nodes.</td>
<td></td>
</tr>
<tr>
<td>M0</td>
<td>No distant metastases.</td>
<td></td>
</tr>
<tr>
<td>M1</td>
<td>Evidence of distant metastases.</td>
<td></td>
</tr>
</tbody>
</table>

**Clinical staging**

<table>
<thead>
<tr>
<th>Stage 1</th>
<th>T1</th>
<th>N0</th>
<th>M0</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>T2</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>Stage 2</td>
<td>T3</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>Stage 3</td>
<td>T1</td>
<td>N1</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T2</td>
<td>N1</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T4</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>Stage 4</td>
<td>T3</td>
<td>N1</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T4</td>
<td>N1</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T</td>
<td>N</td>
<td>M1</td>
</tr>
</tbody>
</table>

**Radiological evaluation of malignant salivary disease**

Computerized tomography (CT) and magnetic resonance (MR) imaging have rendered other methods of demonstrating malignant salivary disease obsolete, for example, contrast sialography and isotope studies (McGahan, Walter and Bernstein, 1984). The precise relation of the tumour to major structures - the external carotid artery, facial nerve and retromandibular vein - can be identified in parotid disease. Careful scrutiny of the integrity of the fibrofatty planes that demarcate the major glands from their surrounding structures gives valuable information about the tumour margin and its regional extension. Obliteration of this plane and obvious infiltration of the surrounding muscles indicate malignancy. The general consistency of the mass is also significant. Non-homogeneous tumours are more likely to be malignant than those displaying an homogeneous structure. Coronal sections of the
facial canal may show expansion secondary to tumour infiltration in those cases with a facial palsy. However, tumour spread will always be far more proximal than the image indicates.

Initial experience with MR imaging is encouraging. Imaging of salivary tumours by this method appears to be equal, if not superior, to CT (Schaefer et al., 1985). There are also many advantages to be exploited with MR. Extensive dental fillings impeded CT imaging in standard planes, but MR imaging does not suffer from metallic artefact, does not require iodine contrast and it gives superior information about vascular tumours. In addition, coronal and sagittal projections can be formatted without changing the patient's position.

The role of biopsy in the management of suspected salivary malignancy

Open biopsy of lesions in the major glands is totally contraindicated. Seeding of the area with neoplastic cells at the time of biopsy is inevitable, leading to local recurrence. Open biopsy as a prelude to treatment is only justified when the tumour arises in a minor gland, for example, the palate. In these sites the overlying and adjacent mucosa should be removed as part of the definitive resection and therefore the extent and outcome of the procedure remains uninfluenced. This is not feasible in a major gland where the overlying skin is normally preserved and only has to be excised if it is infiltrated by tumour or if previous biopsy has been performed.

The concern about tumour spillage at the time of diagnostic biopsy has been overcome by fine needle (22 gauge) aspiration biopsy. Seeding of tumour by this technique has not been documented. Accurate interpretation of smears requires considerable experience and skill on the part of the cytologist. At best a diagnostic accuracy of 85% can be achieved, but only by a few individuals (Eneroth, Franzen and Zajicek, 1967; Sismanis et al., 1981), and this degree of cytological expertise is rare. It is prudent therefore to consider such information in conjunction with that obtained by perioperative frozen section biopsy.

However, even frozen section analysis of salivary tumour tissue is difficult. Accurate histopathological diagnosis of neoplastic salivary tissue is not often achieved and false negative (malignant called benign) rates of 5-12% are commonplace (Hillel and Fee, 1983; Wheelis and Yarrington, 1984).

It is evident that, in the majority of cases, histological diagnosis will prove impossible before surgery and that in a minority the definitive diagnosis will change from benign to malignant on examination of paraffin sections. The surgeon should be aware of this possibility and not compromise his surgical technique when the clinical findings conflict with the frozen section or cytological diagnosis.

Surgical management of malignant salivary disease

The high incidence of local recurrence following simple excision encouraged a more aggressive approach to the management of neoplastic salivary disease. Radical surgery, the removal of the entire gland, and more recently supraradical surgery, that is resection en bloc of the gland with its surrounding tissues and lymphatic drainage field, have been advocated for some tumours regardless of their stage (Conley and Dingman, 1974). There is little to
suggest that these larger operations improve survival unless there is preoperative evidence of neural infiltration or lymph node metastases.

**Parotid gland**

The operative details of parotidectomy are described in Chapter 19. The fundamental principles of surgery for malignant parotid disease are adequate resection of tumour together with the branches or main trunk of the facial nerve where these are either directly involved or potentially so. Frozen section confirmation of tumour clearance in the ends of the resected facial nerve is a prerequisite before any attempt at reconstruction. There should be no hesitation to combine parotidectomy with removal of the mandibular ramus, infiltrated masticatory muscles and maxillary tuberosity in order to obtain a monobloc clearance. In some cases it will be necessary to perform a subtotal petrosectomy or mastoidectomy for tumour removal or to facilitate facial nerve repair.

If the facial nerve must be resected as many of its peripheral branches as possible should be identified at the outset, particularly those supplying the sphincters of the eye and mouth. The main trunk can be prepared at a site free of disease within the temporal bone. Bifurcated or multiple cable grafts can then be interposed to restore continuity. Suitable graft material may be obtained from the greater auricular nerve, sural nerve or cervical plexus. Meticulous attention to the accurate approximation and stabilization of the nerve and graft is essential for a good functional result (Fisch, 1974).

Neck dissection is only indicated if there is palpable neck disease, strong evidence of lymph node involvement on CT examination, or to facilitate myocutaneous flap repair of the defect. The definitive operation must include excision of the scar and overlying skin or mucosa if the patient has previously undergone open biopsy or incomplete removal of the tumour.

**Submandibular gland**

It is unusual for a submandibular salivary neoplasm to present as a mass associated with a palsy of the hypoglossal nerve, lingual nerve or mandibular branch of the facial nerve. More commonly suspicion is aroused by the consistency of the growth which tends to be hard and craggy when malignant. It is also unusual for a tumour to present as duct obstruction with a history of recurrent pain and swelling at meal times. In these cases a radiograph of the floor of the mouth will fail to reveal a calculus and therefore help to distinguish between a tumour and chronic sialadenitis.

The principles governing preoperative biopsy hold as well for the submandibular gland as they do for the parotid. Fine needle aspiration and frozen section diagnosis may indicate malignancy, in which case the resection should include all nerves in close proximity to the tumour together with all suprahyoid lymph nodes.

The technique of removal of the submandibular gland is described in Chapter 19.
Minor salivary gland tumours

Approximately 85% of these lesions arise in either the palate, lip or buccal mucosa. At these sites the differential diagnosis is from a retention cyst or squamous cell carcinoma. The clinical distinction of a cyst by palpation is usually easy.

Resection with an adequate cuff of soft tissue can be achieved in most cases. The commonest site for one of these tumours is the hard palate which may be treated by partial maxillectomy. Since the overlying mucosa is removed in continuity with the tumour it is permissible to biopsy lesions of the minor glands.

Great care must be taken in the assessment of tumours arising in the tonsillar and lateral oropharyngeal region. The clinically unwary may mistake a deep lobe of parotid tumour for one arising in a minor gland. Deep lobe tumours must be removed by an external approach and previous biopsy increases the complications.

The role of radiotherapy

Radiotherapy should only be used as an adjunct to surgery in the treatment of malignant salivary tumours. The sole exception to this rule is that of early stage lymphomata which can be cured by radiation alone. There is a good case for reserving radiotherapy for specific types of tumour. It is certainly indicated for any tumour that shows locally aggressive features, for example, perineural invasion, extensive soft tissue infiltration, lymph node spread and extranodal extension. It should also be given to those in whom the presence of residual disease is suspected after surgery, those whose disease lies close to a preserved facial nerve and in all who have undergone surgery for recurrent disease. It is claimed that with this management policy local recurrence rates are diminished. It is hard to prove these claims in diseases which are uncommon and run a very prolonged course. The evidence in favour of this policy is convincing only for adenoid cystic carcinoma (Jackson, Luna and Byers, 1983; Simpson, Thawley and Matsuba, 1984). Until such data become available patients should not be denied possible benefit, a local dose of 6000 cGy being the accepted norm. However, radiotherapy should be withheld from some patients, for example, those with low grade and stage tumours that have been adequately excised, and non-aggressive tumours in young patients in whom the possibility of inducing a subsequent neoplasm is high.

Radiotherapy also has a role in the palliation of inoperable tumours. Pain can be alleviated to an extent and tumour progress retarded. Complete regression of tumour has been reported with fast neutron therapy but, to date, the experience with this modality has not been uniform (Catterall, Blake and Rampling, 1984).

The role of chemotherapy

Whether chemotherapy has a part to play in the management of these tumours has yet to be determined. Complete and partial responses to the administration of both single and multiple agents have been reported and either type of regimen would seem to be equally effective. Cisplatin, 5-fluorouracil and doxorubicin (Adriamycin) have all been claimed to be useful. No study to date has been able to determine whether chemotherapy prolongs survival
time, but it is clear that in a large proportion of inoperable cases regression of disease and pain can be achieved, albeit temporarily (Suen and Johns, 1982).

**Prognosis of salivary malignancy**

Survival times vary widely according to many factors, namely histological type of salivary tumour, grade and stage of disease, age and sex of the patient, method of diagnosis and type of treatment administered. Of these it is suggested that the stage of disease at presentation correlates the best with ultimate outcome, as perhaps would be expected. Unfortunately, the data from which survival figures have been calculated are based on clinical material collected over decades. During this time considerable advances in diagnosis, operative technique and radiotherapy have been made, all of which influence the validity and accuracy of any conclusions. As a guide to the natural behaviour of the epithelial tumours irrespective of their treatment, grade and stage, the figures in Table 20.7 are the compilation of all reliable series collected between 1964 and 1982 based on 2298 malignant salivary tumours. From this can be seen that the malignant mixed tumour is the least favourable with a collective 5-year survival rate similar to that of carcinoma of the breast, while the acinic cell tumours has the best prognosis (Hickman, Cawson and Duffy, 1984).

**Table 20.7 Prognosis of specific types of salivary gland tumours**

<table>
<thead>
<tr>
<th>Tumour type</th>
<th>No. cases</th>
<th>5-year survival (%)</th>
<th>10-year survival (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acinic cell tumour</td>
<td>101</td>
<td>82</td>
<td>67.6</td>
</tr>
<tr>
<td>Mucoepidermoid tumour</td>
<td>749</td>
<td>70.7</td>
<td>50</td>
</tr>
<tr>
<td>Adenoid cystic carcinoma</td>
<td>1065</td>
<td>62.4</td>
<td>38.9</td>
</tr>
<tr>
<td>Malignant mixed tumour</td>
<td>383</td>
<td>55.7</td>
<td>31</td>
</tr>
</tbody>
</table>
Chapter 21: Parapharyngeal space tumours

Hans Heeneman

Anatomy

The parapharyngeal space is a potential space located on both sides of and parallel to the naso- and oropharynx. It is filled with loose connective tissues, arteries, veins, lymph glands, nerves, muscles, tendons, glomus bodies and salivary gland tissue. It has roughly the shape of an inverted five-sided pyramid with its base towards the sphenoid bone and its apex directed inferiorly towards the minor horn of the hyoid bone (Heeneman, Gilbert and Rood, 1979).

Much of the confusion concerning head and neck spaces in general, and the parapharyngeal space in particular, can be explained by the inconsistent nomenclature relating to these anatomical entities. The parapharyngeal space is known in the literature under the names of pterygomaxillary space, pharyngomaxillary space (Mosher, 1929), lateral pharyngeal space and pterygopharyngeal space (Coller and Yglesias, 1935). The term 'parapharyngeal space' seems most appropriate and is commonly used in the current literature.

Anteriorly, the space is bounded by the pterygoid muscles with their interpterygoid fascia and those fasciae directed laterally towards the buccinator muscle and angle of the mandible. Laterally, the space is limited by the ascending ramus of the mandible, while posterolaterally the deep lobe of the parotid gland and retromandibular fossa can be identified. Medially, there is the pharynx with the tonsillar fossa inferiorly and eustachian tube superiorly. The posterior border is limited by the cervical spine covered by prevertebral muscles and fascia.

Tumours originating within the parapharyngeal space exert pressure on neighbouring structures and expand towards those areas that offer the least resistance, namely the naso- and oropharynx medially, the upper neck between the tail of the parotid and the submandibular gland inferiorly, and the retromandibular fossa posterolaterally.

The retropharyngeal space is connected with the parapharyngeal space in an area just medial to the carotid sheath and its contents. Situated at the junction of these two spaces is the superior lateral lymph node (node of Rouvière, 1927) normally draining the nasopharynx, upper oropharynx and sinuses. The retropharyngeal space provides a pathway towards the mediastinum (Lincoln's highway), while anteriorly and laterally there are connections to other spaces located about the oral cavity and salivary glands.

The parapharyngeal space is divided into anterior (prestyloid) and posterior (poststyloid) compartments by the styloid process and the three muscles and two tendons attached to it (Riolan's bouquet). Table 21.1 summarizes the structures lying in these compartments.

Of special interest is the deep lobe of the parotid gland between the mandible laterally, the medial pterygoid muscle medially and the masseter muscle anteriorly. The sternocleidomastoid and posterior belly of the digastric muscles as well as those structures...
arising from the styloid process are in a posterior position. Patey and Thackray (1956-157) coined the term 'stylomandibular tunnel' formed by the posterior margin of the ascending ramus of the mandible anteriorly, the styloid process and stylomandibular ligament behind and the base of the skull above. This tunnel resists pressure so that tumours of the deep lobe of the parotid gland assume a dumb-bell shape as they grow.

Table 21.1 Structures located in the compartments of the parapharyngeal space and in the retropharyngeal space

<table>
<thead>
<tr>
<th></th>
<th>Prestyloid</th>
<th>Poststyloid</th>
<th>Medial (retropharyngeal)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Arteries</strong></td>
<td>Maxillary</td>
<td>Internal carotid</td>
<td>-</td>
</tr>
<tr>
<td><strong>Veins</strong></td>
<td>-</td>
<td>Internal jugular</td>
<td>-</td>
</tr>
<tr>
<td><strong>Nerves</strong></td>
<td>Inferior alveolar</td>
<td>IX, X, XI, XII</td>
<td>-</td>
</tr>
<tr>
<td></td>
<td>Lingual</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Auriculotemporal</td>
<td>Cervical sympathetic chain</td>
<td></td>
</tr>
<tr>
<td><strong>Lymph nodes</strong></td>
<td>-</td>
<td>++</td>
<td>+++ (Rouvière)</td>
</tr>
<tr>
<td><strong>Glomus bodies</strong></td>
<td>-</td>
<td>++</td>
<td>-</td>
</tr>
</tbody>
</table>

Pathology

Tumours of the parapharyngeal space originate from the various types of cells and tissues present within this space, by direct extension from neighbouring structures or through lymphogenous or haematogenous spread. Numerous primary benign and malignant tumours have been described, the group of salivary gland tumours being most prevalent (50%) followed by the neurogenic tumours (30%). The remaining 20% is made up of a wide variety of tumours, the list of which is getting longer as the limitations of light microscopy, with its inherent shortcomings in distinguishing the relationships of the various structures to one another, are overcome by electron microscopy techniques and special stains to identify cell structures. Tumours originating outside the parapharyngeal space arise from surrounding structures such as parotid gland, mandible and maxilla, nasopharynx, tonsils, sinuses and intracranial cavity.

The parapharyngeal space is richly endowed with lymphatics that drain the sinuses, pharynx and thyroid gland (Robbins and Woodson, 1985). The nodes of the upper deep cervical chain are located in this space. They are connected superiorly to the node of Rouvière, while inferiorly they flow towards the jugulodigastric node, the node most commonly involved with metastatic disease from head and neck tumours. This node lies just outside and inferior to the parapharyngeal space.

While lymph usually flows away from a tumour towards the big veins, under certain conditions the flow is in the opposite direction, for example, when distal lymph channels are obstructed by tumour emboli, previous irradiation or previous surgery. This explains why the parapharyngeal space can be involved secondarily by primary tumours from the oral cavity and elsewhere after previous treatment.
Salivary gland tumours

Two types of salivary gland tumours occur in the parapharyngeal space. The first type arises de novo from salivary gland tissue in the parapharyngeal space and has no connection with the parotid gland. As the tumour grows it pushes a compressed layer of fibroadipose tissue ahead of itself, which might be visible on the newer higher resolution type of computerized tomography (CT) scans. The second type is the 5% of parotid tumours that arise within the deep lobe of the parotid gland (Conley and Clairmont, 1978) and it is this type that has an intimate relationship with the gland. The histology and incidence of the tumours of the deep lobe are similar to tumours originating in the superficial lobe of the gland; most are benign pleomorphic adenomata while the most common malignant tumour is adenoid cystic carcinoma (Hanna et al, 1968; Work and Gates, 1969; Johns, 1977). Discussion of the classification of salivary gland tumours and their biological behaviour is described elsewhere (see Chapters 19 and 20).

Neurogenic tumours

Nerve sheath tumours

Schwannomata

All peripheral motor and sensory axons are covered by Schwann cells throughout their length. Tumours arising from these cells are called schwannomata and are the most common neurogenic tumours in the parapharyngeal space. They are also more common in the head and neck than elsewhere in the body. The terms neurinoma, neurilemmoma and neuroma should no longer be used. Schwannomata are generally solitary and are seen in relation to nerve trunks in that they can readily be dissected from the nerve of origin, most often the vagus nerve.

On cutting the tumour the heterogeneous surface can be seen with solid areas, cysts, haemorrhage, and yellow spots representing areas of lipid deposition.

Microscopic examination reveals that the tumour is composed of alternating areas, some organized and compact (Antoni type A) and some of loosely arranged, relatively acellular tissue (Antoni type B). Occasionally, one pattern may predominate, but the tumours usually are composed of both. The cells are spindle shaped, with elongated and ovoid nuclei. The term Verocay bodies refers to a special arrangement of the cells in an interwoven pattern, producing pallisades with fibrillary zones. There may be macrophages containing lipid and deposition of haemosiderin. Occasionally lymphocytic infiltration around blood vessels is seen. There may be pleomorphism and irregularity of the nuclei with mitotic figures not necessarily associated with malignancy. Axons are found in the capsule and not within the bulk of the tumour.

Neurofibromata

Neurofibromata arise from the Schwann cell and from the perineurium, which is composed of specialized cells called perineurial fibroblasts. In marked contrast to schwannomata, neurofibromata encompass their nerve of origin. It is virtually impossible to
dissect the tumour away from the parent nerve without sacrificing the nerve. These tumours often originate from small subcutaneous peripheral sensory nerve trunks. They are usually solitary although they can be multiple, and are not necessarily associated with neurofibromatosis.

The cut surface of a neurofibroma is usually homogeneous, grey, frequently gelatinous and without haemorrhage and cyst formation. Microscopically, the cells are spindle shaped and are arranged in groups. Special stains show many axons throughout the tumour.

Plexiform neurofibromata appear as multiple nodules or fusiform swellings along the course of the nerve of origin, frequently a cranial nerve. These tumours are characteristic of neurofibromatosis, and under the microscope they show patterns reminiscent of both schwannomata and neurofibromata.

**Malignant schwannomata**

A malignant schwannoma is a malignant neoplasm of nerve sheath origin that infiltrates locally and also metastasizes. The tumours are rare in the general population but occur with higher frequency in patients with neurofibromatosis. Microscopically they are whorls of spindle cells with interlacing fascicles. There is pleomorphism, hyperchromatism, increased cellularity and frequent mitoses. There may be focal areas of chondroid, osteoid or even rhabdomyoblastic metaplasia present. This tumour is known in the literature as fibromyxosarcoma of nerve, fibrosarcoma or myxosarcoma of nerve sheath, neurilemmal sarcoma or secondary malignant neuroma. These terms should no longer be used.

**Nerve cell tumours**

The terms neuroblastoma and ganglioneuroma indicate the cell of origin. Although these tumours usually occur in the adrenal medulla, they occasionally develop along peripheral nerves elsewhere in the body including the head and neck. They may produce catecholamines (Glenner and Grimley, 1974).

**Paragangliomata**

The parapharyngeal space contains vagal bodies closely associated with the ganglion nodosum of the vagus nerve. These bodies contain clusters of chemoreceptor cells and make up a part of the chemoreceptor or glomus system of the body (Lore, 1973). The system consists of minute extra-adrenal paraganglia located along major arteries such as the carotid, pulmonary and mesenteric arteries. Tumours originating from glomus bodies are called glomus tumours, chemodectomata or paragangliomata. At one time the tumours were divided into chromaffin or non-chromaffin, according to the presence of a brown reaction product after the application of dichromate solution, the brown stain representing oxidation products of catecholamines. These findings appear to be inconsistent and today the tumours are classified by site without other qualifying names. In the head and neck, glomus tumours are seen in the jugular bulb, in the crotch of the carotid artery, in the supraglottic larynx where they originate from the glomus laryngicum superior (Greenway and Heeneman, 1975), and from the vagal body (glomus intravagale). Tumours originating from these extramedullary receptor cells rarely produce catecholamines.
Paragangliomata have a familial tendency. They occur more often in females and are often multicentric particularly in people living at high altitude (Heeneman and Maran, 1979). They are generally encapsulated but may be ill defined, with a variegated, rubbery consistency, and are intimately bound to associated vessels and nerves. The cut surface may be smooth and homogeneous and is often yellow to grey with areas of necrosis and haemorrhage. Under the microscope the typical tumour shows reproduction of the architecture of the normal paranglia. The cells are organized into groups, cords, ribbons, and rosettes and have a uniform polygonal outline, with eosinophilic cytoplasm. Nuclei may be hyperchromic and bizarre. Mitoses are unusual and the cells can be shown to contain brown, granular pigment which represents the catecholamines. There is an alveolar pattern, with clusters of cells surrounded by a delicate fibrovascular stroma (Zellballen). Reticulum staining will show the cell mass surrounded by the stroma.

Histologically, paragangliomata are benign. It is difficult to determine malignancy on histological grounds alone. Correlation with clinical findings such as rapid growth, fixation, bone destruction and local invasion is essential. Since these tumours are often multicentric it is sometimes hard to distinguish between metastases and multiple foci. Malignancy occurs in less than 10% of cases (Conley, 1965).

Other tumours

Sarcomata are rare in the parapharyngeal space and take on features of sarcomata found elsewhere. Their identification depends on careful electron microscopic analysis. Involvement of the parapharyngeal space by osteogenic chondrosarcoma is secondary, resulting from expansionary growth of these tumours when they originate from neighbouring structures, for example, base of the skull, maxilla, mandible, and eustachian tube.

A neck mass is the initial manifestation in 30% of lymphomata and involvement of the lymph nodes located in the parapharyngeal space is not rare. Occasionally a primary lymphoma with no involvement of other neck nodes is encountered.

Table 21.2 indicates other primary tumours that may be found in the parapharyngeal space as encountered in the literature. The biological behaviour of these tumours is the same as tumours of similar histology found elsewhere in the head and neck.

Diagnosis

Most tumours of the parapharyngeal space are benign, slow growing and produce symptoms by exerting pressure on neighbouring structures and those within the space. The malignant tumours also invade the surrounding bones: maxilla, mandible, pterygoid plates and base of skull. Often symptoms are long-standing and have been treated unsuccessfully until the tumour becomes manifest. Sometimes the diagnosis is made incidentally during a routine head and neck examination by inspection and palpation of the submandibular trigone, retromandibular fossa and pharynx. Parapharyngeal space tumours expand to these areas as they do not offer the same resistance as surrounding bony structures. Bimanual ballottement of a tumour in the lateral pharyngeal wall differentiates a parapharyngeal space growth from a primary tumour in the soft palate and also provides information as to extent and degree of fixation.
Hearing loss resulting from middle ear effusion caused by pressure on the eustachian tube is the most common symptom. Symptoms of a dull or neuralgia-like pain or a chronic sore throat are next in frequency. Although more common in malignant tumours these symptoms do not necessarily indicate malignancy.

Motor nerve palsies in the distribution areas of the vagus, spinal accessory and hypoglossal nerves are caused by direct pressure on the nerve. When there is ipsilateral vocal cord paralysis, shoulder weakness and a deviation of the tongue, then the jugular foramen (Vernet) syndrome is complete. Horner's syndrome has been reported. Sensory deficits of the mandibular division of the trigeminal nerve or its branches are caused by perineural invasion invariably due to adenoid cystic carcinoma.

Trismus and narrowing of the pharynx leading to airway obstruction, speech defects and dysphagia can be explained by mass effect.

Once a primary parapharyngeal space tumour is suspected and a careful examination of the upper aerodigestive tract has ruled out a primary tumour then an accurate radiological evaluation is indicated. Plain sinus films confirm the presence and size of most soft tissue tumours and demonstrate bone erosion. A chest X-ray to determine the presence of metastases of involvement of the chest cavity with malignant lymphoma is part of the preoperative examination.

The new generation of CT scanners with better resolution power not only accurately delineate the extent of the tumour, but also give information as to whether it arises from the deep lobe of the parotid or de novo. In the latter case there may be a lucent line representing the compressed layer of fibroadipose tissue between the tumour and the deep lobe of the parotid (Som, Biller and Lawson, 1981). Computerized tomographic scanning may indicate the degree and extent of bone erosion. Where there is any question of erosion, particularly in important areas such as the base of skull region, tomography may provide further information.

Enhancement indicates increased vascularization and, as most parotid tumours are relatively avascular, most parapharyngeal space tumours are hypodense. The most common enhancing tumour is the schwannoma followed by the paraganglioma. Haemangioma and tumours with haemorrhagic degeneration also enhance. When there is enhancement, carotid arteriography is indicated not only to determine the degree of vascularization, but also to identify the feeding vessels of the tumour, to detect a vascular abnormality and to determine whether there is displacement of the carotid arterial system. Tumours arising in the deep lobe of the parotid when entering the prestyloid compartment of the parapharyngeal space displace the internal carotid artery posteromedially. The site of neurogenic tumours is less constant and the displacement is variable. Widening of the carotid bifurcation (so-called lyre sign) is seen in carotid body tumours.

Magnetic resonance (MR) imaging may well become an important tool in the preoperative evaluation of parapharyngeal space tumours.

Experienced histopathologists may be able to arrive at a reasonably accurate histological diagnosis in about 80% of cases on the basis of material supplied by a fine needle
biopsy. Only when a tissue diagnosis is required before definitive treatment (for example, in tumours of advanced size or when malignancy is suspected necessitating wide extirpative surgery and/or irradiation) should an incisional biopsy be performed. This should preferably be carried out by an external approach rather than intraorally in view of the risk of seeding of tumour and uncontrollable bleeding.

The method of choice in arriving at a proper histological diagnosis is an excisional biopsy.

**Treatment**

**Surgical management**

The management of tumours of the parapharyngeal space is primarily surgical. The surgical approach to the space is transoral, transparotid, via the upper neck or the retromandibular fossa, transpharyngeal, or by any combination of the above. Exposure can be enhanced by mandibulotomy (Bass, 1982), which is reported to be necessary in fewer than 10% of cases (Stell, Mansfield and Stoney, 1985).

Transoral delivery of the tumour through an incision in the lateral pharyngeal wall is unsafe and to be discouraged unless the tumour is superficial or less than 2 cm on palpation, in which it is hard to identify on a CT scan. Attempted removal of larger tumours causes undue pressure on the capsule with an inherent danger of rupture and seeding. Immediate control of the carotid artery and its branches in cases of massive haemorrhage is impossible.

Deep lobe tumours of the parotid that have not yet entered the stylomandibular tunnel and therefore do not involve the parapharyngeal space, are best dealt with through the transparotid or retromandibular approach. After formal removal of the normal outer lobe of the parotid, the facial nerve and its branches are often found to be stretched by the tumour depending on its size. These branches must be dissected free from the tumour which is readily done if it is benign and has a capsule. Adherence of the tumour to the facial nerve and its branches may indicate malignancy, but it is advisable not to take the nerve or any one of its main branches without histological confirmation and without prior consent from the patient. If the pathologist is unable to supply a tissue diagnosis on frozen section, then it is prudent to abandon the operation at this time, close the wound and return a few days later when the results of permanent sections are available and the patient has been duly informed about the likelihood of postoperative facial nerve dysfunction.

It should be remembered that with deep lobe tumours, a formal en bloc removal with a cuff of healthy tissue as is achieved with outer lobe tumours of the parotid gland, is not possible. The tumour can be dissected free from the deeper tissues by blunt dissection and, depending on its location, can be delivered from between the cervicofacial and cervicoparietal division of the facial nerve (transparotid) or posteroinferior to the main stem and cervicofacial division (retromandibular).

Large parotid tumours involving the parapharyngeal space are palpable in the retromandibular fossa, and when large enough, also in the lateral wall of the pharynx, pushing it medially. Superficial parotidectomy allows for identification of the main stem of the facial
nerve and the branches in greatest danger of being unduly stretched during tumour removal. The tumour can be exposed and bluntly dissected by pulling the mandible forward, cutting the stylomandibular ligament and fracturing the styloid process. If necessary, additional exposure can be obtained by performing an osteotomy at the level of the angle of the mandible, preferably angulated to promote postoperative stabilization. The inferior alveolar nerve is severed causing anaesthesia of the ipsilateral lower lip and chin, a major disadvantage of this technique.

A tumour of the parapharyngeal space presenting as a mass in the upper neck, between the tail of the parotid and the submandibular gland, is probably not a deep lobe tumour but rather a neurogenic one. These presentations are best approached through a horizontal neck incision and dissection upward toward the skull base. Additional exposure can be obtained by removal of the submandibular gland and dissection of the tail of the parotid. The great vessels are readily seen and controlled if necessary.

When the tumour is very large, malignant, or extends way up towards the base of skull area and maximum exposure is required for vessel control and adequate tumour removal, the parapharyngeal space is opened up through a transpharyngeal approach. A median lip splitting incision is extended laterally towards the anterior border of the sternocleidomastoid muscle 3 cm below the inferior margin of the mandible. After clearing the submandibular trigone, the external and internal carotid arteries are identified, and tapes placed around these vessels for control. A stepped mandibulotomy in the area of the symphysis or just anterior to the ipsilateral mental foramen to preserve sensory innervation to the chin and lower lip is carried out. The mucosa of the floor of the mouth is incised lateral to the submandibular duct and 2 cm parallel to the mandible to preserve sufficient mucosa for closure. The incision is extended upward along the anterior tonsillar pillar to the level of the upper pole of the tonsil. The parapharyngeal space is exposed by swinging the mandible outward after division of the muscles of the floor of the mouth, taking care not to injure the lingual and hypoglossal nerves. Further soft tissue dissection upward will provide a reasonably good exposure to the base of skull and tumours in that area.

Paragangliomata require special consideration because of their vascularity. Proximal and, if possible, distal control of major arteries has to be established. In about 10% of cases major vessels will have to be resected with the tumour and preoperative consultation with a vascular and/or neurosurgeon is strongly advised. Preparations will have to be made for potentially massive intraoperative bleeding, particularly in those cases when adequate distal control at the skull base cannot be readily achieved. In these cases the internal carotid artery cannot be grafted.

Glomus tumours often adhere to surrounding tissues including vessels and base of skull. Sharp dissection and good exposure are required. The transpharyngeal approach is the safest for these tumours.

Smaller carotid body tumours lying in the upper neck in the crotch of the carotid artery are best exposed through a large horizontal neck incision over the tumour. They can be readily dissected using the adventitia as the plane of resection.
Larger tumours not only grow between the bifurcation but also surround it forming a readily dissectable line where the two sides meet. This line invariably lies posterolateral to the carotid artery system, and because the tumour is here at its thinnest, it is the easiest area to dissect and identify the adventitial plane of dissection. Massive tumours must be divided to identify the carotid artery bifurcation and to find the adventitial dissection plane.

Paragangliomata are usually benign, slow-growing tumours causing minimal morbidity unless they are large enough to cause pressure symptoms. The risks of surgical resection and postoperative complications, particularly neurological deficits, are high and many elderly patients may die from other causes before treatment for these tumours is necessary. Tumours of the glomus system in patients who live in high altitude areas rarely require resection.

An ipsilateral neck dissection is indicated when the tumour in the parapharyngeal space is malignant and neck nodes are palpable. There is no scientific support for the necessity of leaving the specimen containing primary tumour attached to the neck specimen.

Postoperative care

After surgery to the parapharyngeal space most patients can be routinely extubated immediately. After posterior mandibulotomy when postoperative pharyngeal oedema is to be expected, keeping the endotracheal tube in place for a period of 24 hours followed by supervised extubation is invariably adequate. With the transpharyngeal approach, when the floor of the mouth is incised, a tracheostomy is safest and therefore mandatory.

Suction drainage is required. Antibiotic coverage to prevent wound infection is not necessary so long as the pharynx is not opened. Only then is it advisable to cover the patient with third generation cephalosporins for a period of 3 days, starting one day before operation.

With good knowledge of the anatomy and with sound surgical techniques, the postoperative complications are kept to a minimum. The major problems are related to sensory and motor deficits in the distribution area of those nerves removed or injured during the operation. Overstretching of the facial nerve will cause a temporary paresis, most commonly of the submandibular division. Ipsilateral vocal cord paralysis is to be expected following removal of schwannomata of the most frequently involved nerve, the vagus. Glossopharyngeal and hypoglossal nerve palsies and Horner's syndrome are not rare following the removal of paragangliomata.

Cerebrovascular accidents, particularly in the elderly, may result from surgery for paragangliomata after manipulation of the common and internal carotid arteries or after grafting procedures.

The incidence of salivary fistulae should not be any higher than seen following parotidectomy for tumours of the superficial lobe and certainly not as frequent as after surgery for chronic parotitis. Frey's syndrome (gustatory sweating) is troublesome in about 10% of cases requiring some form of management. Again, the incidence is no higher than in parotidectomy for other reasons.
The role of radiotherapy

Anatomical relations prevent *en bloc* resection of malignant tumours with a wide margin of healthy tissue. Even if the tumour has been removed grossly, it is prudent to treat patients with high grade malignant tumours with a full course of postoperative radiation to the primary site and the ipsilateral neck. Radiotherapy as primary treatment for malignant tumours should be used in patients who appear to have unresectable tumours and should be offered to those patients who refuse surgery. Stage I malignant lymphoma also requires radiotherapy.

A beneficial response of paragangliomata to radiotherapy is explained by intimal thickening of the feeding vessels leading to a decrease in size of these vessels. Diminished tumour opacification and decreased venous shunting is sometimes noted (Myers et al, 1971; Handel et al, 1977). This method has a place in the treatment of elderly patients with large tumours or in patients who refuse surgery.

Malignant neurogenic tumours are radiosensitive but cures have been recorded only with neuroblastomata.
Chapter 22: The oesophagus in otolaryngology

M. J. Drakeley

To be able to eat a meal that has been specifically and lovingly prepared is one of life's most satisfying pleasures; certainly the intake of food is an essential necessity of life. Any interference with the ability to swallow quickly leads to the patient consulting the general practitioner. The patient may then be referred to a gastroenterologist, a cardiothoracic surgeon or to an otolaryngologist. It is essential, therefore, that specialists in all disciplines are familiar with the conditions affecting the oesophagus.

Anatomy and physiology

The oesophagus is that part of the alimentary tract which connects the pharynx to the stomach and therefore has a cervical portion, an intrathoracic portion and a short intra-abdominal portion. The function of the oesophagus is the rapid and satisfying transfer of food in one direction and, less pleasant, gastric gases and contents in the other.

A detailed account of the anatomy and physiology of the oesophagus is given in Volume 1, Chapters 10 and 11. It is sufficient here to say that a knowledge of the act of swallowing goes a long way towards the understanding of the symptoms of disorders of the oesophagus.

The act of swallowing is initiated voluntarily and is completed, thereafter, by a sequence of orderly reflexes. Swallowing may be considered in three phases: oral, pharyngeal and oesophageal. In the oral phase food is chewed, mixed with saliva and then propelled backwards by the tongue into the pharynx. This initiates a reflex inhibition of breathing and the closure of the openings into the nasopharynx, oral cavity and larynx, preventing regurgitation into the nose and aspiration into the larynx and bronchial tree. The pharyngeal phase is quite short. Relaxation of the cricopharyngeal muscle or upper oesophageal sphincter is critically timed to allow the food bolus to pass from the pharynx into the oesophagus. Almost immediately the cricopharyngeus contracts and an orderly primary peristaltic contraction of the oesophagus propels the bolus down towards the stomach. Each peristaltic contraction is preceded by a zone of relaxation. The lower gastro-oesophageal sphincter mechanism - there is no anatomical muscle present - relaxes and allows the food to enter the stomach. Should the oesophagus not be cleared, then a secondary peristaltic wave will complete the process.

Reflux of gastric contents back into the oesophagus is prevented by a combination of the physiological tone of the lower oesophagus, the oesophagogastric angle, the pinch-cock effect of the diaphragm, the negative intrathoracic pressure in association with the slight positive intra-abdominal pressure and perhaps the mucosal folds at the oesophagogastric junction.

With this brief insight into the physiology of the act of swallowing, some of the symptoms of oesophageal disease can now be considered.
**Symptoms of disease of the oesophagus**

Although conditions of the oesophagus are being considered, it should not be forgotten that almost any disease or abnormality of the tongue, pharynx or oesophagus can cause problems with swallowing. Swallowing can be inhibited by pain, neuromuscular dysfunction or mechanical obstruction, thus oesophageal disease may be indicated by one or more of the following symptoms.

*Dysphagia*

It is essential, when a patient complains of difficulty in swallowing, that a distinction is made between those who can initiate the act of swallowing, those who are reluctant to swallow because of inflammation of the throat or because of the embarrassing effects of laryngeal incompetence, and thirdly, those who can swallow properly but the food is either slow to go down, sticks completely or only passes into the stomach after the patient drinks a glass of water.

The question of how long the patient has had the problem should always be asked, for if it has been present for a number of years the cause is likely to be of a benign rather than a malignant nature. Malignant conditions tend to produce symptoms within weeks, although the occasional patient, reluctant to consult his doctor, may put up with his symptoms for a year or more before seeking help.

The changing nature of the diet which the patient can take will also give some indication of the nature and progression of the problem. When sphincters are weak, fluids are more difficult to direct along the proper channels than solids. True dysphagia with its implication of neuromuscular or mechanical obstruction is worse with solids. Precise localization of the site of obstruction is often unreliable, the patient invariably suggesting a level higher than it actually is.

*Regurgitation*

A distinction should be made between the forceful vomiting of gastric contents and the gentle regurgitation of undigested food. The painless regurgitation of undigested food indicates a dilated viscus above an obstruction - seen in its most marked form in achalasia or cardiospasm. Regurgitation due to lesions in the upper oesophagus happens during a meal; indeed the emptying of a pharyngeal diverticulum temporarily relieves the dysphagia caused by the distension of the pouch with food.

Acid regurgitation from the stomach into the throat is associated with a bitter taste - waterbrash. Patients may also feel a burning sensation in the precordium or back. Heartburn is due to a failure of the lower oesophageal sphincteric mechanism to prevent the acid gastric contents from entering the oesophagus when the patient bends down or tries to lie flat in bed. It is frequently associated with the sliding type of hiatus hernia. The volume of regurgitated contents can help in the diagnosis. The dilated oesophagus in achalasia may contain as much as 2 or 3 litres of material, whereas in cases of pharyngo-oesophageal diverticula or carcinoma the volume will be very much less.
Pain

When caused by oesophageal disease, pain may be felt in the epigastrium, in the substernal region, in the left subscapular region of the back, or in the root of the neck. If the pain is associated with, or aggravated by swallowing, its significance is readily appreciated. If dysphagia is minimal or absent, then oesophageal pain may wrongly be attributed to disease of the heart, lungs or stomach. Acid fluids cause ulcerated lesions to burn, so a patient with a hidden pharyngo-oesophageal neoplasm will avoid fruit juices because it aggravates his pain. The spasmodic pain associated with neuromuscular and obstructive lesions is more likely to be elicited by the ingestion of solid food.

Bleeding

Gross haemorrhage from the oesophagus is not often seen. It is usually due to varices. Some patients with neoplasms or inflammatory conditions may vomit or regurgitate a little blood-stained fluid. Fresh blood appearing in the throat may arise from lesions near the upper end of the oesophagus, but is more likely to come from the nose or pharynx.

Respiratory symptoms

Recurrent attacks of bronchitis or bronchopneumonia as a result of laryngeal spill-over or tracheo-oesophageal fistula frequently complicate disease of the oesophagus. Progressive deterioration of pulmonary function may be such that the underlying cause may be missed altogether. Occasionally, cancers of the bronchus may invade the oesophagus, giving rise to dysphagia or even, in advanced cases, broncho-oesophageal fistulae.

Hoarseness, whether due to vocal cord paralysis, simple laryngitis or to the simple weakness of cachexia should, in the absence of a bronchial tumour, arouse suspicion of an oesophageal lesion.

General symptoms

Loss of weight is the most important of the generalized symptoms. It occurs rapidly in the presence of oesophageal obstruction. General malaise and tiredness due to anaemia are frequently observed in oesophageal disease.

Physical examination

As with all patients, the examination of a patient with suspected oesophageal disease should include a physical examination of the chest, abdomen, cardiovascular and nervous systems. Evidence of wasting, weight loss and dehydration should be noted. Inspection and palpation of the neck may reveal enlargement of the thyroid gland. Palpable lymph nodes suggest inflammatory or neoplastic conditions arising in the pharynx or oesophagus. The absence of laryngeal crepitus against the vertebral column may be seen in some cases of carcinoma of the postcricoid region or the upper oesophagus.

Inspection of the mouth and tongue may reveal evidence of anaemic such as glossitis, atrophy of the lingual papillae and cracks or fissures at the corners of the mouth. These
features are frequently seen in the Paterson-Kelly (Plummer-Vinson) syndrome, as are brittle spoon-shaped finger nails. An enlarged spleen may be palpable in patients with this syndrome. Enlargement of the liver may also be noted on abdominal examination, which may be due to metastatic involvement from carcinoma of the oesophagus or stomach. In the early stages of portal cirrhosis, the liver, before it begins to contract, may be palpable. Other manifestations which should be looked for are spider naevi around the neck and upper chest, palmar erythema and, in the advanced stages when bleeding from oesophageal varices occurs, the superficial veins radiation from the umbilicus enlarge to form the caput medusae. Progressive ascites also occurs, as it may with malignant disease of the oesophagus involving the liver.

Mirror examination of the pharynx may reveal palatal or vocal cord paralysis, ulceration or tumour. Even when these areas appear normal, the pooling of saliva in the pyriform fossae arouses suspicion of a lesion at a lower level.

**Diagnostic aids**

Accurate diagnosis of oesophageal disease requires a variety of diagnostic techniques which include various radiographic techniques, endoscopy and measurement of intraluminal pressures of the oesophagus and pH changes at different intraluminal levels.

**Radiological techniques**

Radiological techniques include routine plain films of the chest and neck and fluoroscopic screening with contrast media (see Chapter 2).

*Posteroanterior views* of the chest and mediastinum may show pulmonary fibrosis and tumour enlargement or displacement of the mediastinum, aortic aneurysm, and the shape and the size of the heart. Enlargement of the left atrium in mitral valve disease can result in dysphagia, due to compression of the oesophagus.

*Lateral soft tissue views* of the neck to show outlines of the larynx, trachea, vertebral column, the vertical band of soft tissue representing the postcricoid region and the cervical portion of the oesophagus are especially useful. Widening of the retro-oesophageal soft tissue may indicate the presence of a tumour or perhaps cellulitis. Surgical emphysema suggests a perforation of the oesophagus which may be spontaneous or more commonly, iatrogenic, following endoscopy or reconstructive surgery. A fluid level within an abscess cavity or a persistent gas bubble in the upper oesophageal lumen are also significant abnormalities seen in this projection.

*Fluoroscopic screening* permits the radiologist to observe the passage of a bolus of contrast medium from the pharynx to the stomach. Cine photography or video recordings are especially useful in studies of upper oesophageal and pharyngeal abnormalities where movement is extremely rapid. The best contrast medium is a water suspension of barium sulphate, although a water-soluble contrast medium is advisable when perforation is suspected. Radiography is not the best way to demonstrate the presence or absence of significant or pathological oesophageal reflux, especially if manoeuvres such as abdominal compression or the Valsalva test are required to provoke it. Thus other more sensitive tests should be used to confirm the diagnosis of suspected gastro-oesophageal reflux.
Endoscopy

Endoscopy - the techniques of which are given in Chapter 3 - permits direct inspection of the lumen of the oesophagus and of the oesophagogastric junction. It confirms the information obtained from radiographic and physiological studies. It enables verification of the nature of oesophageal pathology to be made, either by biopsy or from cytological studies of smears or washings. It is also useful in localizing the site of bleeding.

Although it is true to say that endoscopic studies are an essential part of the investigation of oesophageal disorders, they should not be performed by the occasional endoscopist. The original oesophagoscopy were rigid instruments which were difficult and dangerous to use. Oesophageal perforation, particularly in the cervical region, occurred in as many as one in 100 examinations. The introduction of the flexible fibrescope has considerably simplified the endoscopic examination of the oesophagus and has reduced the incidence of perforation. Many believe that rigid endoscopes should be reserved for the removal of foreign bodies.

Some endoscopists will not perform an oesophagoscopy without there being a previous barium swallow to indicate the site of any possible obstruction. This in turn helps to reduce the incidence of oesophageal perforation.

Manometry

The measurement of pressures within the lumen of the oesophagus is an important and reliable technique in the evaluation of the oesophageal disorders, particularly those characterized by abnormalities of oesophageal motility. Manometric studies are performed using saline-filled tubes, usually a constantly infused system. The pressure changes recorded graphically are usually made with the tube assembly in the stomach, in the high pressure zone and in the body of the oesophagus.

Manometric measurements are of use in determining the location of the lower oesophageal sphincter mechanism and also the abnormal segments encountered in achalasia and diffuse spasm of the oesophagus.

pH measurement

The pH reflux test is especially sensitive in confirming the presence or absence of gastro-oesophageal reflux. In this test a small volume of 0.1M hydrochloric acid is instilled into the stomach. A special pH electrode is placed in the oesophagus and the pH recorded while the patient performs various manoeuvres such as coughing, deep breathing and the Valsalva test in various positions. A fall in pH to below 4.0 is taken as indicative of the presence of gastro-oesophageal reflux.

The introduction of the technique of 24-hour pH monitoring has proved especially helpful in patients with complicated diagnostic problems. The refinement of technique which permits pH measurement in the patient's home or working environment rather than in the laboratory, has proved even more useful.
Miscellaneous studies

These include the acid clearing test, the acid perfusion test, the measurement of the potential difference between the inside and outside of the stomach and oesophagus, and technetium scanning.

The acid clearing test measures the ability of the oesophagus to clear regurgitated gastric contents. A prolonged acid clearing test indicates a poor prognosis in terms of the development of oesophagitis and its sequelae. The acid perfusion test is of limited value and was used as a means of differentiating pain of oesophageal origin from that of angina. Measurement of the potential difference between the inside and outside of the stomach and oesophagus has been found useful in identifying the junction between the squamous and columnar epithelium of the oesophageal mucosa. The change in potential difference does not accurately define the gastro-oesophageal junction.

Technetium-99m is used in the diagnosis of oesophageal disease, particularly in attempts to identify mucosal changes in patients with a Barrett or gastric-lined oesophagus and to diagnose oesophageal tumours. Some workers consider technetium scanning a safe non-invasive method not only to detect but also to quantify gastro-oesophageal reflux.

Oesophageal motility disturbances

For convenience disturbances of motility can be considered as affecting:

(1) the upper oesophageal sphincter;

(2) the body of the oesophagus and the lower oesophageal sphincter.

Upper oesophageal sphincter

Oropharyngeal dysphagia, that is cervical oesophageal dysphagia, is caused by a variety of conditions - cerebrovascular accident, Parkinson's disease, bulbar poliomyelitis and multiple sclerosis. Muscular diseases, conditions such as muscular dystrophy, myasthenia gravis and the inflammatory dermatomyositis also affect the upper sphincter. These disturbances of motility are discussed in Chapter 10 and pharyngeal diverticula, another important cause of cervical dysphagia, in Chapter 14.

Body of the oesophagus and lower sphincter

Motility disturbances of the body of the oesophagus and the lower sphincter may be due to achalasia or cardiospasm, diffuse spasm, or to a hypotensive lower oesophageal sphincter as seen in hiatus hernia and scleroderma, and also to miscellaneous conditions such as myasthenia gravis, cerebrovascular accidents, Parkinson's disease, muscular dystrophy, and diabetic and alcohol neuropathies.
Achalasia of the cardia

The aetiology of this condition is unknown. It is characterized by the absence of peristalsis in the body of the oesophagus, a high resting pressure at the lower oesophageal sphincter, and the failure of the sphincter to relax in response to swallowing. It is thought to have a neurogenic cause supported by the absence or reduction in the number of ganglion cells in Auerbach's plexus. Obstruction to swallowing is the commonest and perhaps the earliest symptom. Frequently the patient has most difficulty with cold food, and solids pass more readily than liquids. This latter feature contrasts with the symptoms of a malignant or benign stricture. Regurgitation occurs in nearly 70% of patients particularly at night, can give rise to aspiration and respiratory symptoms. Pain is infrequent.

Few patients lose weight with this condition. It is more frequently seen in women than men and in patients aged between 30 and 60 years, and often these patients are psychologically disturbed. It may occur in children.

A serious complication of achalasia, even in treated patients, is the development of carcinoma of the oesophagus. Therefore all patients should be kept under review for life. The diagnosis of achalasia is confirmed radiologically and by motility and manometric studies. Endoscopy should also be performed to distinguish early achalasia from carcinoma and benign strictures, which can complicate the condition.

Barium swallow X-rays show a dilated oesophagus with a narrowed rat-tail lower end. In advanced cases the oesophagus may contain 2-3 litres of fluid and food debris. Manometry shows absent peristalsis in the whole length of the oesophagus, feeble or absent contractions, a normal or elevated resting pressure of the lower oesophageal sphincter and failure of the sphincter to relax during swallowing when a higher pressure will be recorded.

Treatment

Attempts to relieve the symptoms of patients with achalasia by eliminating the distal oesophageal obstruction by drugs or by forced dilatation, have largely been unsuccessful in the long term. Nitrates or nitroglycerines have been used and some patients have been taught to pass their own bougies. Surgical treatment with the modified Heller myotomy has proved to give much better relief of symptoms than forced dilatation and should therefore be the treatment of choice in all but those who are unfit for operation. It carries a lower mortality, the incidence of perforation is considerably less, and most patients are improved. The approach is via a left posterolateral thoracotomy. Controversy rages between surgeons as to whether or not an antireflux procedure should be undertaken after the myotomy has been carried out, as reflux oesophagitis has been reported postoperatively.

Diffuse spasm

Diffuse spasm differs in that there is normal sphincteric relaxation whether the pressure is normal or elevated. In cases of diffuse spasm of the oesophagus, an extended myotomy is frequently carried out, the myotomy including the lower sphincter if it is shown by manometry to be hypertensive. The length of the myotomy is governed by the extent of
the disease as defined by the manometric studies. The incision may extend up to the aortic arch or even higher in some cases. Oesophageal diverticula if present can be excised.

Perforation of the oesophagus

Perforation or rupture of the oesophagus, in spite of modern therapy, still carries a high mortality and morbidity. Early recognition and treatment should ensure a favourable outcome in most cases.

Although perforation or rupture may occur spontaneously, the commonest cause is instrumental, either by the rigid oesophagoscope or by bouginage. The advent of the flexible fibroptic oesophagoscope has considerably reduced the number of oesophageal perforations. Instrumental perforations may occur at any level, but most are seen in the upper oesophagus especially just above the upper sphincter and also as a result of impingement of the oesophagoscope on the bodies or on osteophytes of the cervical vertebrae. The next most common site both for instrumental and foreign body perforations is the lower oesophagus as it narrows to pass through the hiatus. Perforations of the middle third and abdominal section of the oesophagus are infrequently seen unless benign or malignant strictures have necessitated dilatation. The use of the YAG laser in the cutting out of a passage through inoperable tumors may also result in perforation of the oesophagus.

Postemetic rupture usually occurs at the lower end of the oesophagus but can, on rare occasions, occur at the upper end or even in the middle. Perforation occurs as a longitudinal tear of all layers of the wall of the oesophagus just above the diaphragm. It is thought that the sudden onset of pressure in the oesophagus rather than the actual pressure is the chief contributory factor. The fact that it occurs in adults rather than in children suggests that the strength of the oesophagus is greater in children. Typically postemetic ruptures are seen after ingestion of a heavy meal, or following a beer drinking session followed by retching and vomiting. Other conditions implicated in non-instrumental perforation of the oesophagus include stress associated with neurological disease, or after burns or operations away from the oesophagus. Postoperative leaks may be seen after reconstructive operations on the oesophagus. Injuries to the cervical oesophagus may be seen in suicidal or malicious throat cuts, or gunshot wounds.

The consequences of perforation are due to the contamination of the mediastinal tissues and pleural cavities with digestive juices, food and bacteria which gives rise to cellulitis and suppuration.

The symptoms of perforation depend largely on the site of perforation and the extent of inflammatory reaction. Pain, fever and difficulty in swallowing and flexing of the neck are frequent early features of cervical oesophageal rupture. Surgical emphysema or crepitation may vary in extent but is a common feature. In cases of thoracic oesophageal perforation, cervical surgical emphysema may be present but without tenderness. Cardiorespiratory embarrassment may be a feature and a pneumothorax may also result from perforation of the oesophagus. X-rays of the chest and neck are most useful in making the diagnosis. Indeed, all patients who complain of pain or tenderness after endoscopy should undergo further X-rays. Air in the mediastinal tissue spaces and widening of the mediastinum and retrovisceral space are frequently seen. Pleural effusion with or without pneumothorax may present and
occasionally air may be seen under the diaphragm should the abdominal portion of the oesophagus be ruptured. Abscess formation may also be revealed in long-standing cases and the author has even seen an infected pericardial effusion some 3 weeks after endoscopic perforation. Barium studies are useful in determining the site of perforation and detecting underlying pathology.

Treatment

Early diagnosis of perforation of the oesophagus is essential, mortality being as high as 50% or more if treatment is delayed for over 24 hours after injury. As soon as the diagnosis of perforation is made the patient should not be allowed to take food or fluid by mouth, a central venous catheter should be inserted for parenteral feeding and also for measurement of the central venous pressure. Suitable broad spectrum antibiotics are also given intravenously to minimize mediastinal infection. The site of perforation, the clinical state of the patient at the time of diagnosis and underlying oesophageal pathology influence the management of perforations of the oesophagus. Instrumental perforations of the cervical oesophagus, if recognized early, can be managed by simple conservative measures but, if suppuration has developed, surgical drainage of the retro-oesophageal space and/or the mediastinum may be necessary. Suppuration extending down to the level of the fourth thoracic vertebra can be reached through a cervical incision made along the anterior border of the sternomastoid muscle. This together with parenteral feeding and antibiotics may be all that is required. Sometimes mediastinal abscesses need to be drained via the chest. Complete healing of the oesophagus may take as long as 2 months and mobilization of the patient may be speeded up by the establishment of a feeding gastrostomy or jejunostomy. Some operators have encouraged the healing process by the local endoscopic application of a solution of 20% sodium hydroxide repeated at weekly intervals.

Oral feeding can be resumed when there is no radiological evidence of a leak and there is no discharge from the chest.

Management of perforation of the thoracic oesophagus depends upon the original pathology and the time after injury. Perforation of the thoracic oesophagus is usually more serious than that of the cervical oesophagus. Conservative management in these circumstances is rarely successful and active treatment is necessary. Simple repair of the perforation with drainage of the pleural cavity is suitable when the diagnosis is made early - within 6 hours - and there is minimal soiling of the mediastinum and pleural cavity and no underlying pathology in the oesophagus.

Excision of the oesophagus containing the perforation, stricture or tumor, followed by reconstruction of the alimentary tract is also suitable in cases diagnosed early. Some surgeons prefer to perform the proximal anastomosis in the neck rather than in the thorax, as the consequences of anastomotic leak are less. In the case of delayed presentation or diagnosis, it may not be possible to undertake the major reconstructive surgery until a much later date. In these cases the damaged oesophagus can be excised, pleural drainage, cervical oesophagostomy, closure of the cardiac end of the stomach and a feeding gastrostomy or jejunostomy are all carried out as the first stage with reconstruction, utilizing colon or stomach, being carried out at a later date.
Spontaneous postemetic perforations of the lower oesophagus are best debrided and closed surgically as soon as possible. The suture line may be reinforced by pleura or by using the fundus of the stomach as a patch (Thal). In cases of delayed diagnosis, management comprises simple drainage together with parenteral feeding and antibiotics, but mortality is high, death resulting from sepsicaemia and its sequelae of hepato renal failure and disseminated intravascular coagulation. Similarly anastomotic leaks developing after initial repair are difficult to deal with, hence the two stage forms of treatment.

**Foreign bodies in the oesophagus**

The variety of foreign bodies lodging in the oesophagus is legion, but coins, bones, open safety pins and lumps of meat are by far the most common to be encountered. The site of impaction may be at the upper oesophageal sphincter (cricopharyngeus muscle), at the level of the arch of the aorta or at the level of the hiatus. Pathological causes include benign and malignant strictures.

The incidence of foreign bodies rises in old people who frequently get lumps of meat stuck in the oesophagus by a combination of failing to chew properly - particularly if they are edentulous or wear ill-fitting dentures - or if there is lack of effective propulsion by the oesophageal musculature. Dentures themselves may be swallowed, particularly those that are partial or ill fitting especially when the patient is asleep or under the influence of alcohol.

Impaction of blunt objects usually causes total dysphagia, discomfort and excessive salivation whereas sharp objects may puncture the oesophagus thus giving rise to mediastinitis. The patient is often able to point to the exact site particularly if the foreign body is lodged in the upper part of the oesophagus. When the foreign body is in the middle or lower third, localization is not so accurate and pain is referred to the back or behind the sternum. The pain is of a sharp, cutting nature and is worse when the patient attempts to swallow and occurs at the same site every time the patient swallows. Dysphagia is nearly always present and is due to the size of the foreign body or to inflammatory reaction and spasm caused by its presence. At first there may only be slight difficulty in swallowing, but later the difficulty becomes more pronounced. There is also regurgitation of food, saliva and mucus in the later stages. When the obstruction is complete there may be overflow of oesophageal contents, giving rise to respiratory symptoms suggesting the possibility that the foreign body is in the bronchial tree.

**Examination**

The patient must be observed during the act of swallowing. Palpation of the neck may reveal a tender swelling in the lower part, medial to the sternomastoid, due to the inflammatory reaction around the foreign body by an abscess, and if the oesophagus or pharynx has been perforated surgical emphysema may be felt.

The mouth, pharynx, tonsillar region and the base of the tongue must be examined carefully and with a good light. A small sharp fish bone may lodge in the base of the tongue or the crypt of the tonsil, even if the patient is convinced that the bone has been swallowed.
Each part of the pharyngeal wall must also be examined carefully to find the tip of a fish bone that has become embedded in the tissues. The tissues may well appear bruised and inflamed. If the patient is able to localize the pain to one side, then the foreign body or the laceration caused by its passage must be above the cricoid level and should be visible in a mirror. Most foreign bodies which can be seen in the mirror can be removed without the need for a general anaesthetic. If the patient holds his own tongue the surgeon can, with the aid of his mirror, guide the forceps down the throat, grasp and withdraw the foreign body. He must not carry out a 'blind grab'. If a foreign body cannot be seen in the laryngeal mirror, other investigations are necessary.

All patients with a history of swallowing a foreign body should have posteroanterior chest and lateral neck X-rays taken and if the foreign body is not revealed, the neck, thorax and abdomen should be screened, using a little barium. The opaque medium may be held up and actually outline the foreign body or the column of barium may be split. If the barium does not pass or cannot be washed down with water, then the presence of a foreign body is to be strongly suspected.

The patient should be kept under observation until he is completely symptom free. Abrasion of the oesophageal mucosa will give some pain and cause dysphagia but usually these symptoms disappear rapidly. Increasing pain and dysphagia indicate the presence of a foreign body or some more serious damage to the oesophageal wall. In these circumstances or if there is any doubt about the presence of a foreign body, an oesophagoscopy is imperative.

Ideally endoscopy should be carried out as soon as possible after the radiological examination because the foreign body may have moved. Obviously, in children, small oesophagoscopy or even an anterior commissure laryngoscope may be sufficient if the foreign body is impacted in the upper oesophagus. Oesophagoscopy should be performed under general anaesthesia, the anaesthetist taking care not to pass the endotracheal tube blindly, thus damaging the oesophagus or causing the foreign body to perforate the oesophageal wall. The oesophagoscope should be passed slowly because it is easy to override the foreign body such as a coin, tablets or pieces of meat, especially if it has become coated with mucus. The oesophagoscope is passed as near as possible to the foreign body making sure that it is not pushed further downwards. Grasping alligator-type forceps are passed and the foreign body is pulled up through the oesophagoscopy or until it imjinges on the end of the oesophagoscope which is then removed. Care must be taken passing the cricopharyngeus as the foreign body could be dislodged sometimes to be found in the pharynx from which it can be removed using McGill's forceps.

Meat and soft foreign bodies are extracted piecemeal with forceps. Sharp objects such as bones, safety pins, razor blades and dentures present special problems for the operator. The oesophagoscope is passed until the foreign body is seen and the position of the sharp edge noted. The object of the exercise is to remove the foreign body without damaging the oesophagus.

Mucus, food debris and any barium left after the X-ray examination are removed carefully by suction. If the end of the foreign body is buried in the oesophageal wall, manipulation is necessary to release it so that it can be withdrawn. Any bleeding may be
controlled with adrenaline-soaked gauze pledgets. Small bones may be extracted through the oesophagoscope but large ones should be withdrawn abutted to the end of the scope. Razor blades may have to be broken or bent in half to facilitate their removal. An open safety pin presents little problem if the point is downwards. However, if the point is facing upwards great difficulty is often experienced in removing it. If it is not possible to turn the pin in the oesophagus, it may be possible to manipulate the point of the pin into the lumen of the oesophagoscope and remove it that way. Endoscopic shears have been used and some operators attempt to close the pin with Clerf's or similar forceps. Should all else fail then the pin could be pushed into the stomach and either be rotated and extracted or even be allowed to pass onwards and in the fullness of time be passed per rectum.

Dentures in the oesophagus present many hazards, especially partially dentures with sharp points and hooks. During endoscopy the hook should be manipulated into the lumen of the oesophagoscope before extraction. If the denture is impacted it may have to be divided within the oesophagus.

After removal of the foreign body, it is essential that the oesophagoscope is repeated to verify the presence or absence of any pathology such as benign or malignant strictures at or beyond the level of the foreign body, or injury to the oesophagus. The examination may have to be repeated some days later when the inflammatory reaction has subsided. In this instance, the patient should be treated for periesophagitis or perforation with intravenous feeding and antibiotics.

Most foreign bodies which lodge in the oesophagus can be removed endoscopically but some have to be extracted at operation - these include dentures and razor blades. If the foreign body has been impacted for some time, it may erode through the oesophageal wall and form a periesophageal abscess which will have to be drained and the foreign body removed.

**Hiatus hernia**

Hiatus hernia may be defined as a displacement of the stomach up through the oesophageal opening (hiatus) of the diaphragm into the lower mediastinum.

Two main types of hernia are recognized - sliding and paraoesophageal. In the sliding hiatus hernia the gastro-oesophageal junction, normally below the diaphragm, ascends into the chest. The consequence of this is gastro-oesophageal reflux because the oesophagus is not kept closed by the lower sphincteric mechanism. It should be remembered that not all patients with reflux necessarily have a hernia.

In the paraoesophageal hernia the lower end of the oesophagus and the gastro-oesophageal junction retain their normal anatomical positions below the diaphragm. However, a portion of the stomach protrudes up alongside the oesophagus through an enlarged hiatus. These patients do not suffer from reflux.

A mixed type of hernia may be encountered in which components of both sliding and paraoesophageal herniae are to be found, that is the lower oesophagus and gastro-oesophageal
junction slide up into the chest and, at the same time, part of the stomach passes up alongside the oesophagus through the enlarged hiatus.

**Clinical presentation**

The main symptoms of patients with sliding hiatus hernia are due to reflux and oesophagitis - heartburn on bending or on lying flat in bed, retrosternal discomfort, nausea and occasionally vomiting. Dysphagia may be due to muscular spasm or motility disorders, but is usually associated with the presence of a stricture.

Patients with paraoesophageal hernia do not experience heartburn and oesophagitis unless a mixed form of hernia is present. These patients' symptoms are due to anaemia, abdominal discomfort, dyspnoea and pseudoangina, the 'angina' indicating possible acute obstruction or impaction. In patients with reflux the possibility of regurgitation into the pharynx and aspiration into the larynx and bronchial tree should be remembered.

**Diagnosis**

The diagnosis of hiatus hernia is confirmed radiologically and by manometric and pH studies, as well as by endoscopic examination. Paraoesophageal herniae may be diagnosed on a lateral chest X-ray, the gastric air shadow being seen behind the heart. Fluoroscopic studies using contrast media such as barium also help to confirm the diagnosis.

Oesophagoscopy is desirable not only to determine the level of the gastro-oesophageal junction (the smooth pink oesophageal mucosa changes to the rugal gastric pattern), but also to observe the degree of oesophagitis. The terms mild, moderate and severe should be avoided in favour of a more specific grading such as:

- grade 1 - mucosal erythema;
- grade 2 - erythema plus superficial ulceration;
- grade 3 - ulceration and a dilatable stricture;
- grade 4 - ulceration and an irreversible stricture.

A biopsy of the oesophageal mucosa should be taken to confirm the presence of oesophagitis, to diagnose the so-called gastric lined or Barrett's oesophagus with its columnar epithelium, and to exclude malignancy.

**Treatment**

Many patients have a sliding hiatus hernia without experiencing any symptoms of gastro-oesophageal reflux; they need no treatment.

Symptomatic patients require treatment designed to reduce reflux and to lower the acidity of the gastric contents to minimize its effect on the oesophageal mucosa.
Medical treatment

Medical treatment consists of a weight reducing diet, avoidance of alcohol and tobacco together with drugs such as antacids, antispasmodics and H₂-receptor antagonists, used either on their own or in combination. Patients should not wear tight belts or corsets and they should, if possible, avoid bending. At night they should sleep with the head of the bed raised on blocks. The tendency is to slip off at any extra pillows that may be used, unless, of course, they are placed under the mattress.

Surgery is indicated in the treatment of sliding hiatus hernia only when medical treatment has failed to control the patient's symptoms within 6 months to a year. On the other hand, surgery is the treatment for paraoesophageal herniae if complications are to be prevented.

Age should not be considered a contraindication to surgical repair if the symptoms interfere with the patient's quality of life.

Surgical repair

The aim of any operation to repair a hiatus hernia should be to restore the gastro-oesophageal junction to its rightful anatomical place below the diaphragm while, at the same time, preventing gastro-oesophageal reflux.

The operation should be designed in such a manner that the pupils can readily reproduce the results of their masters. It is true to say that many operations have been described and none has proved to be ideal and we are still awaiting the perfect solution to the control of gastro-oesophageal reflux.

Operations which have stood the test of time include the Belsey mark IV, the Nissen fundoplication and the Hill antireflux procedure.

More recently, a Collis gastroplasty, in which the oesophagus is lengthened by the construction, in continuity, of a tube from the fundus of the stomach, coupled with an antireflux procedure, has found favour with many surgeons including Pearson whose name is now associated with the procedure.

The mark IV operation is performed only through a left thoracotomy incision and the Hill repair via the abdomen. The Nissen fundoplication and the Pearson procedure can be performed through either an abdominal or thoracotomy incision, as also can the Thal gastric patch plus an antireflux procedure.

A likely cause of failure of operations to cure symptoms of reflux is insufficient mobilization of the oesophagus, a reason why many surgeons prefer the thoracic approach. Oesophagectomy and reconstruction with stomach or preferably an isoperistaltic segment of the left colon or jejunum is reserved for patients who have had multiple previous procedures, especially if the oesophagus has become an unmanageable fibrous tube, or has been damaged during re-operation and resection.
The principles of the Belsey mark IV are adequate mobilization of the oesophagus, even as far as the arch of the aorta, and adequate cleaning of the gastro-oesophageal junction. Sutures are placed to close the hiatus before the antireflux procedure is constructed. The antireflux procedure is a 270° wrap of the gastro-oesophageal junction. This is achieved in two layers of mattress sutures, the first being placed in the stomach and oesophageal wall 2 cm above the oesophagogastric junction and back again. The second row of mattress sutures incorporates the central tendon of the diaphragm, that is diaphragm, stomach, oesophagus and back again. When this final layer is tied down the gastro-oesophageal junction is pulled below the diaphragm. Usually three mattress sutures are employed in each layer. Finally the hiatus is closed by closing the sutures already in position in such a way as to avoid obstructing the cardia.

The Nissen fundoplication is usually performed via the abdomen and also requires careful mobilization of the oesophagus. Its chief aim is the restoration of the normal function of the lower oesophageal sphincter mechanism rather than an anatomical correction. The plication is achieved by a 360° wrap of the lower oesophagus by approximating the anterior and posterior fundal folds in front of the oesophagus. The hiatus should also be narrowed to prevent the stomach passing through into the chest.

The original operation has undergone many modifications since its inception and has even been performed in the chest when the oesophagus was short.

Unfortunately there is a recurrence rate after all the operations for repair of hiatus hernia of 10-15% but in some series it may be as high as 75%. However, symptoms may be less severe and therefore more easily controlled with antacids, etc. Only in the last resort should oesophagectomy and reconstruction be necessary.

**Strictures**

In this section strictures associated with reflux oesophagitis are considered along with those due to the ingestion of caustic substances. Malignant strictures will be discussed in the section on malignant tumours.

**Reflux strictures**

The commonest type of oesophageal stricture is that secondary to the reflux of acid or alkali, of which acid oesophagitis is probably the most important. Reflux oesophagitis, if not overcome, can develop into chronic inflammation with ulceration or fibrous scarring which gives rise to stricture formation and shortening of the oesophagus or both. Strictures secondary to reflux can lie at the oesophagogastric junction or higher at the junction between squamous and columnar epithelium - this level may be as high as the arch of the aorta, as seen in the gastric lined or Barrett's oesophagus. Third, a long stricture involving the lower one-third or half of the oesophagus may develop. Possible causes of this long stricture are postpartum vomiting and postoperative nasogastric intubation and suction.
**Symptoms**

*Dysphagia* is the main symptom, typically in patients who have previously suffered symptoms of reflux, such as heartburn and discomfort associated with hiatus hernia, which tend to subside or disappear with the formation of the stricture.

*Loss of weight* is common with reflux strictures.

*Anaemia* and pain may be present, especially if a chronic ulcer is present.

**Investigations**

*Chest X-ray and barium swallow* are important investigations, as in all cases of dysphagia, to outline the stricture and any mediastinal lesion. It could be argued that manometric and pH studies are irrelevant in the presence of fibrous oesophageal strictures but, occasionally, strictures can occur in conditions with motor dysfunction such as achalasia.

*Endoscopy* should be performed in all cases of stricture to exclude neoplasm and to determine the level and severity of the stricture, and the presence of inflammation of ulceration of the oesophageal mucosa. In all examinations a mucosal biopsy of the stricture and, if possible, mucosa above and below it should be taken.

**Treatment**

The initial treatment of peptic reflux strictures is bouginage via the mouth followed by intensive medical therapy to neutralize the acid contents of the stomach which are more easily able to pass into the oesophagus. Bougies such as Hurst or Maloney mercury bougies or the Chevalier Jackson bougie may be passed using the rigid Negus oesophagoscope. The Eder Peustow type of dilators are passed over a fine guide wire placed in position with a fibreoptic oesophagoscope.

In both methods the risk of perforation, and the importance of its early recognition cannot be overemphasized.

Should bouginage and medical therapy fail then surgical treatment may be necessary to relieve symptoms of reflux strictures. These operations are those already described for the repair of hiatus hernia. The stricture must be dilated before the operation and perhaps once or twice afterwards before the dysphagia is completely relieved.

**Corrosive strictures**

The swallowing of corrosives is usually accidental in children and suicidal in adults. Strong solutions of caustic alkali are now rare household commodities. Because of this and precautionary warnings, labelling and safety devices used by manufacturers, these strictures are less frequently seen, while strong acids such as sulphuric, nitric and hydrochloric are only used in industry.
The greatest damage when these poisons are swallowed is seen in the mouth and in the lower third of the oesophagus. It is the injury to the oesophagus which determines the long-term future of the patient. The immediate survival hinges on acid-base equilibrium and renal function, upon the incidence of laryngeal oedema and the development of bronchopneumonia, especially in corrosives which emit fumes such as strong ammonia, hydrochloric acid and foaming nitric acid.

Immediate treatment is directed towards the treatment of shock and pain, and to the neutralization of the corrosive by the appropriate weak solution of acid or alkali. Parenteral feeding and intravenous antibiotics are begun as soon as possible. Careful regulation of the intravenous fluid and electrolytes is essential to preserve renal function.

A full evaluation of the injury by radiology and endoscopy should be carried out, although there is some controversy as to the value of early endoscopy.

A late result of ingestion of a corrosive burn is stricture formation of the oesophagus, the incidence of which may be reduced by the administration of corticosteroids. If the stricture is short and not severe and there is no ulceration or perforation, it can often be dilated. However, in a severely damaged oesophagus, the stricture is tight, fibrotic and elongated. In such cases resection of the affected segment and reconstruction using colon, jejunum or stomach are necessary.

**Postoperative strictures**

These can occur following operations on the oesophagus and other parts of the alimentary tract in which a nasogastric tube has been used. Strictures following oesophageal operations usually appear at the site of the anastomosis.

Those strictures occurring within a few weeks or months after operation are usually due to infection at the suture line and excessive granulation tissue. Stricture may be caused by reflux, local inflammation by tablets, particularly potassium salts and especially in patients with a recurrence of a previously resected neoplasm. Dysphagia following vagotomy and pyloroplasty is not infrequent. If this persists a stricture may develop possibly due to reflux and trauma at the lower end of the oesophagus.

Oesophageal webs or rings are fibrous membrane projections into the lumen producing an obstructive lesion. These are of two types:

1. the cervical web at the pharyngo-oesophageal junction which is more commonly seen in women. The patient complains of dysphagia, which if associated with glossitis and anaemia, is called the Paterson-Kelly or Plummer-Vinson syndrome.

2. lower oesophageal web or Shatskis ring can also cause dysphagia.
'Drug strictures'

Several tables can cause oesophagitis and subsequent ulceration and fibrosis if they are lodged in the oesophagus for any length of time. These are the various preparations of potassium, given in conjunction with diuretics, and antiarthritic drugs.

Increasing cases of oesophageal monilia are being reported with the increasing use of immunosuppressive therapy, steroids and antibiotics. The symptoms are those of painful dysphagia which may be relieved by the oral administration of nystatin.

**Diverticula of the oesophagus**

Upper oesophageal diverticula protrude between the oblique and horizontal fibres of the inferior constrictor muscle of the pharynx - the dehiscence of Killian. Strictly speaking this is a pharyngeal diverticulum or pouch and as such is discussed in Chapter 14.

Mid-oesophageal diverticula are of the traction type; most cause no symptoms and are only discovered on barium studies for unrelated conditions. Low oesophageal diverticula are frequently referred to as paraoesophageal diverticula and are usually associated with other oesophageal conditions such as hiatus hernia and diffuse oesophageal spasm. Inflammation can cause dysphagia and pain which are symptoms of the associated conditions and are indications for surgical treatment. Treatment consists of a long segment myotomy to overcome the motility disorder together with repair of the associated hiatus hernia. Diverticulotomy is not recommended unless the diverticulum has a very narrow neck. Occasionally, it may be necessary to excise that part of the oesophagus containing the diverticulum followed by reconstruction.

**Neoplasms of the oesophagus**

**Benign neoplasms**

Benign tumours of the oesophagus are rare accounting for less than 10% of oesophageal neoplasms. They tend to occur in younger age groups and symptoms are usually of longer duration than with malignant tumours.

**Leiomyoma**

Smooth muscle tumours account for two-thirds of the benign tumours and are seen more frequently in men than in women. Although they may occur anywhere in the oesophagus, they are more commonly seen in the lower third, usually they are solitary lesions, although multiple tumours have been reported. Symptoms depend upon the size of the tumour. dysphagia and a feeling of pressure or fullness substernally are the usual complaints. Radiographically an ovoid sharply demarcated filling defect may be seen.

Endoscopically tumours may be missed because they lie beneath the mucosa but, with care, the bulge into the oesophageal lumen will be observed. Biopsy of the suspected area should be avoided as it may complicate surgical enucleation of the tumour. Enucleation is performed through a left or right thoracotomy depending on the level of the tumour. A
A longitudinal incision is made in the muscular wall of the oesophagus without injuring the intact mucosa. Should the leiomyoma encircle the oesophagus or the gastro-oesophageal junction, oesophagogastrectomy may be necessary for its removal.

**Pedunculated tumours or polyps**

A number of polypoid oesophageal tumours have been described including mucosal polyps, fibromata, lipomata and haemangiomata. Dysphagia is a common feature together with regurgitation and weight loss. Very rarely a pedunculated tumour may be regurgitated into the mouth.

Surgical removal is the treatment of choice, although some pedunculated tumours have been removed via the oesophagoscope. The tumour is approached from the opposite side of the pedicle so that it can be readily seen through a longitudinal incision through the oesophageal wall and removed. The defect in the oesophageal wall is then closed in two layers.

**Oesophageal cysts**

These cysts form the second commonest group of benign neoplasms and represent intramural embryonic rests. They may be lined with either columnar or stratified squamous epithelium. These, too, like leiomyomata can be enucleated from the muscle wall.

Duplication of the oesophagus is a less common abnormality consisting of a tube composed of muscular and submucosal layers with a squamous epithelial lining running parallel to the oesophagus. Although the muscle layers of the duplication and real oesophagus may intermingle, the mucosae remain separate and so can usually be removed without the need for oesophageal resection.

**Malignant tumours**

The aetiology of oesophageal cancer is unknown. It is predominantly a disease of men between the ages of 50 and 70 years. There is a particularly high incidence in certain areas of China, Japan, USSR and South Africa. Epidemiological studies have identified risk factors such as cigarette smoking and a high consumption of alcohol. The ingestion of nitrosamines is carcinogenic for the oesophagus and may explain the higher incidence in the Bantus of South Africa.

Malignant tumours often develop in an abnormal oesophagus, as seen in achalasia, or the columnar or gastric-lined oesophagus, when it has been damaged by corrosive agents. The male members of certain families with tylosis - a thickening of the skin on the palms of the hands and soles of the feet - develop carcinoma of the oesophagus. The female members of the family are the genetic carriers. Squamous cell carcinoma is by far the commonest malignant tumour of the body of the oesophagus. Primary adenocarcinoma of the oesophagus is relatively rare if one excludes the adenocarcinoma at the oesophagogastric junction which is almost invariably a tumour of the stomach extending upwards to involve the oesophagus. Other rare tumours include fibrosarcoma, leiomyosarcoma and primary malignant melanoma.
Oesophageal tumours spread by local extension within the wall and lumen and invade neighbouring structures such as the trachea, the left main bronchus, aorta and the pericardium. Lymphatic spread via a submucosal network ensures spread well beyond the limits of the perioesophageal nodes. Thus cervical tumours pass to the cervical nodes along the jugular veins and also to the supraclavicular group of nodes. Thoracic lesions spread to the local nodes and also to the supraclavicular nodes and to the subdiaphragmatic nodes around the coeliac axis of the aorta. Tumours at the cardia may spread upwards to the mediastinal nodes and also to nodes around the coeliac axis and mesentery. Spread through the blood vessels may result in metastatic deposits in the liver, lungs and bone.

**Clinical features**

The most common symptom of carcinoma of the oesophagus is dysphagia. Difficulty is first encountered with solids and progresses quite quickly to difficulty swallowing liquids and saliva. Weight loss and weakness follow. In high lesions aspiration pneumonia may occur.

Early diagnosis is essential if invasion of adjacent structures is to be prevented. Any patient over the age of 35 years complaining of pain or difficulty in swallowing should have barium studies and oesophagoscopy to exclude a malignant lesion.

Barium studies show an irregular mucosal pattern with narrowing of the lumen without the proximal dilatation seen in benign strictures. The diagnosis is confirmed by oesophagoscopy not only to determine the histological type but to establish the exact level and extent of the tumour. Lesions arising in the cervical and upper portion of the oesophagus should also be investigated by bronchoscopy to exclude involvement of the trachea and main bronchi. Indeed, some lesions prove to be primary bronchial neoplasms which have spread to the oesophagus.

**Treatment**

The aim of any treatment of patients with carcinoma of the oesophagus should be the relief of dysphagia, permitting the intake of a normal diet, improving the quality of life and, at the same time, effecting a cure of the disease.

Long-term survival after all forms of treatment of carcinoma of the oesophagus is disappointingly low and controversy reigns over the merits of otherwise of the various forms of treatment available. Some employ surgery alone, where others consider a cure so unlikely that they irradiate all patients in spite of complications such as radiation pneumonitis and postradiation stricture - a condition that is very difficult to treat. As a compromise some people prefer a combination of radiotherapy and surgery. As yet chemotherapy, whether used alone or in combination with surgery or radiotherapy, has little value in the treatment of oesophageal cancer. Intracavity radiotherapy, with caesium-137 instead of radium or gold, has been revived with some effect albeit over an all too brief a period. Palliation by intubation may allow the patient to take a soft or liquid diet, but does not cure the disease, nor does the use of the neodymium YAG laser which is used to 'burn' a passage through the tumour.
Surgery

Several operations in one or more stages have been used over the years in the management of carcinoma of the oesophagus. For example, the Wookey operation for cancer of the cervical oesophagus which combines resection with the formation of skin tubes to reconstruct the alimentary tract.

Resection of the oesophagus with the use of stomach, jejunum or colon to restore the continuity of the alimentary tract, is most frequently used today, although use of microsurgical techniques has revived interest in free jejunal grafts for pharyngo-oesophageal and cervical oesophageal tumours.

Oesophagogastrectomy

Oesophagogastrectomy with oesophagogastrostomy is widely used for tumours of the oesophagus at all levels and carries a relatively low hospital mortality rate. For tumours of the lower oesophagus and cardia, a left thoracoabdominal incision provides a good exposure to permit mobilization of the stomach, resection and an intrathoracic anastomosis. Some surgeons, however, prefer a left thoracotomy with a radial incision of the diaphragm rather than the more extensive thoracoabdominal incision. For lesions requiring an anastomosis at or above the arch of the aorta, some surgeons, rather than perform the anastomosis anterior to the aorta, employ a combined abdominal and right thoracic approach. When resection is undertaken for cervical lesions, either of these two approaches may be employed for the mobilization of the stomach, the oesophagogastrostomy being performed through a separate cervical incision. A thoracotomy may be avoided by freeing the oesophagus by blunt dissection from the neck and abdomen, resecting it and pulling the freed stomach through the posterior mediastinum into the neck in order to anastomose it to the proximal oesophagus. First described by Grey Turner this operation, revived by Orringer, should only be performed by experienced operators. It is not a minor procedure. In all cases careful mobilization of the stomach is essential in order that the blood supply may be preserved. The stomach is freed from the omentum and mesocolon preserving the right gastroepiploic artery. The short gastric vessels are divided, as is the left gastric artery at its origin from the coeliac axis. Mobilization is considerably enhanced by the use of 'stapling guns' which clip and cut the vessels in one movement. If necessary the spleen and the tail of the pancreas may be included in the resected specimen. To minimize the postvagotomy effect, some surgeons perform a pyloromyotomy or pyloroplasty.

Stapling devices may again be used to divide the stomach at a suitable level before carrying out an end-to-side anastomosis between the stomach and the proximal oesophagus. A variety of anastomotic techniques has been employed together with a variety of suture materials, ranging from a two layer anastomosis with an inner layer of continuous chromic catgut and an outer layer of interrupted silk sutures, to the use of special anastomotic stapling guns, employing two circles of stainless steel wire staples. This latter technique requires a small gastrostomy incision, although the author overcomes this by inserting the instrument through the incision across the pylorus, before closing it as for a pyloroplasty.

A nasogastric tube may be inserted for gastric decompression and intravenous feeding for 4-5 days if necessary, to allow the anastomosis to heal before oral feeding is resumed.
Colon interposition

Initially the right colon supported by the middle colic artery was preferred as it permitted the creation of an isoperistaltic anastomosis, its chief disadvantages being its bulk and discrepancy in size compared with the oesophagus. Furthermore, its blood supply at the ileocaecal region may be defective, and it normally propels fluid rather than solid materials. The use of the transverse colon supplied by the middle colic artery is technically easier but usually it was used in an antiperistaltic manner with far from satisfactory results.

Reconstruction with isoperistaltic segments of the left colon and splenic flexure supplied by the left colic artery has proved to be a most suitable alternative to the right colon. The left colon is more used to the propulsion of solid material and its calibre more closely resembles the oesophagus. It is less bulky than the right colon and its marginal artery permits a linear interposition without kinking. In all cases, care must be taken not to twist the vascular pedicle. The route taken between the stomach and neck may be subcutaneous, substernal or through the posterior mediastinum. In all instances it is essential to avoid any redundancy of the colon within the chest. The colon therefore should be anchored to the margins of the hiatus.

Jejunal interposition and free jejunal grafts

Two techniques for reconstruction have been devised. One is a long vascular pedicle to support a segment of proximal jejunum for isoperistaltic interposition, the limiting factor being the jejunal blood supply. The second is microvascular surgery to permit the use of free jejunal grafts for pharyngo-oesophageal lesions.

Results of surgery

The operative (hospital) mortality in resection of oesophageal cancers depends upon selection, and the experience of the surgeon. In experienced hands operative mortality should be less than 10%. Long-term survival will depend to a degree on the extent of the disease, up to 40% of those without lymph node involvement enjoying 5-year survival.

Treatment of unresectable carcinomata

There is no doubt that irrespective of long-term survival, surgical resection offers the patient the best palliation of symptoms due to carcinoma of the oesophagus. However, approximately 40% of patients referred for surgery are unsuitable for resection because of their poor general condition, the presence of multiple metastatic deposits or involvement of other intrathoracic structures. If, at operation, the tumor is found not to be resectable it can be bypassed by anastomosing the unaffected proximal oesophagus to the stomach or to a loop of jejunum. Failing this, oesophageal intubation may be performed.

Oesophageal intubation

There are two main types of oesophageal tubes, those which can be introduced through the mouth and pushed down through the tumour and those which have to be pulled down through the tumour into the stomach. The Celestin and Mousseau Barbin tubes are pulled
through the tumour via a small gastrostomy incision, cut to a suitable length and then firmly fixed in position. The author prefers to cut side holes in the tube to facilitate drainage through the tube and to prevent blockage of the tube should gastric mucosa abut over the end of the tube. Soutar's and Livingstone tubes are push-through types which can ride up as they have no flange, unlike the Atkinson tube, and the new type of Celestin tube. Tubes are not really suitable for lesions above the level of the arch of the aorta because of regurgitation and aspiration.

Most patients take a soft or liquid diet followed by a fizzy drink to keep the tube clean. Frequently the patient takes too large a piece of food and the tube becomes blocked requiring it to be unblocked or replaced. The insertion of the push-through tubes necessitates dilatation of the oesophagus which can cause splitting or perforation of the mucosa. This, in a patient already considered unsuitable for resection, usually signifies the death of the patient within a few days from mediastinitis and pneumonia.

**Laser**

The neodymium YAG laser is now being used to vaporise tumour tissue either to ablate the neoplasm or to produce palliation. Care should be taken in the selection of patients for such treatment so that they are not denied a potentially curative resection. High oesophageal lesions are particularly suitable for treatment by laser as are recurrences at anastomotic sites of previously resected tumour.

**Postoperative care**

Special attention in the postoperative period following oesophageal surgery should be given to fluid and nutritional requirements. As many patients are elderly with cardiac and renal dysfunction, they should not be overloaded with fluid. Many patients with severe obstruction suffer from hypoproteinaemia and are particularly deficient in albumin, a feature which may be masked by dehydration. Strict fluid balance charts should be kept to record input and fluid loss via the chest drain, nasogastric tube and urinary catheter. A total of approximately 40 mL fluid/kg body weight per 24 hours should cover most circumstances.

Intravenous feeding or parenteral feeding is necessary for patients who are unable to eat and should take care of the patients calorific and protein requirements. Also electrolytes such as potassium chloride can be added to one of the many proprietary preparations now available. One should aim for 168-189 kL (40-45 kcal)/kg of bodyweight and a protein equivalent of 0.2-0.25 g nitrogen/kg of body weight in 24 hours. Daily urea and electrolytes, blood sugar and protein estimations are therefore necessary for any patient on parenteral feeding. The time of resumption of feeding after reconstructive oesophageal surgery varies from 48 hours to 10 days. Thereafter there should be a gradual change from intravenous to oral fluids, at the same time retaining the calorific intake to maintain the postoperative recovery of the patient.

**Physiotherapy**

Special attention to avoid retention of secretions and aspiration pneumonia should be given to all patients who have had oesophageal surgery. Early mobilization will prevent
cardiopulmonary complications, deep venous thrombosis and emboli, and also frozen shoulders.

**Nasogastric tubes**

A nasogastric tube, if used, should not be spiggotted or connected to a closed system. Its function is twofold:

1. to prevent distension of the stomach with air and putting strain on the new anastomosis;
2. to permit aspiration of excess gastric secretion until the ileus has cleared.

Initially the tube should be aspirated hourly and then every 4-6 hours. The tube should be securely fixed with three pieces of tape to the nose to prevent its being pulled out. Should the tube be pulled out it should not be replaced for fear of perforating the anastomosis. If there is excessive regurgitation of gastric contents the tube could be replaced with the judicial use of the rigid oesophagoscope.

**Drugs**

Generally speaking the only drugs required in the postoperative period are analgesics and antibiotics. If complications occur the appropriate drugs may be necessary.

**Complications of oesophageal surgery**

Early complications such as *postoperative shock and haemorrhage* can follow any major operation. As reconstructive operations on the oesophagus are often lengthy, hypovolaemia is probably the commonest cause of postoperative shock.

Bleeding may be evident in the drainage bottles, but sometimes these can become blocked concealing the haemorrhage. Bleeding can also occur in the abdomen and also into the alimentary tract itself.

Serious cardiac complications such as myocardial infarction occasionally complicate major surgery. Abnormalities or rhythm such as atrial fibrillation may be seen. Correction of any deficiency of potassium should be carried out before digitalization is commenced. Late onset of atrial fibrillation may indicate a breakdown of the new anastomosis. Some surgeons advocate routine postoperative artificial ventilation for 24 hours but many do not find it necessary. Retention of secretions or inhalation of gastric contents may also give rise to respiratory difficulties requiring bronchial toilet via a bronchoscope. Pre-existing respiratory disease such as emphysema and chronic bronchitis may also give postoperative problems, particularly if the patient is also a heavy smoker. Good physiotherapy with or without a minitracheostomy should help the patient over his or her difficulties together with the appropriate antibiotics prescribed according to bacteriological culture and sensitivity reports. Pulmonary emboli occurring around the tenth postoperative day can be avoided if deep venous thrombosis is detected and treated early.
Complications arising from the alimentary tract include distension of the stomach, intestines and ileus. Decompression is frequently necessary either via the nasogastric tube already in situ or by the passage of a flatus tube. Patients may suffer from malnutrition following reconstructive surgery, some are reluctant to eat solid food again and have to be helped over the psychological barriers. Often patients are not able to take a normal meal and have to be given small amounts more frequently. Lack of appetite may be due to nausea, reflux or even to the absence of the stomach which normally produces a feeling of hunger. Drugs to stimulate the appetite may help, otherwise some form of enteral feeding may be necessary. A feeding gastrostomy or jejunostomy should be avoided, a fine nasogastric tube being the best way to feed the patient in such circumstances.

**Infection**

Infection, in spite of antibiotic cover, can be a problem after oesophageal surgery. Chest infection, urinary tract infection and even wound sepsis are frequent. Empyema is rare unless there has been an anastomotic leak as in a subphrenic abscess. General septicaemia can occur in these patients from any site, but infected intravenous cannulae must always be considered.

**Anastomotic breakdown**

Anastomotic breakdown is the most serious of all postoperative complications following reconstructive oesophageal surgery. Prevention is better than cure. Therefore the surgeon should do his utmost to prevent devascularization of the stomach, crushing of the oesophagus and placing the anastomosis where tumour is still present. Often malnutrition, infection and abscess formation contribute to the breakdown. Sometimes if the blood supply has been compromised, the gastric suture line, rather than the anastomosis, may break down or the stomach wall may necrose and rupture. Fortunately in experienced hands this complication is uncommon, but awareness of its possibility and prompt diagnosis and treatment will save some patients.

Complications due to fistulae frequently occur 7-10 days after operation. A slight fever or the onset of atrial fibrillation may be early indications of trouble. A chest X-ray when compared with earlier films may show pleural changes or even a hydropneumothorax. The diagnosis may be confirmed, if a pleural drain is in situ, by giving the patient a weak solution of methylene blue dye to drink, the dye soon appearing in the drainage bottle. If the tube has been removed further X-ray screening with contrast media, such as gastrograffin, may demonstrate, not only the site of the leak, but also its size.

Small leaks may be treated conservatively by giving nil by mouth, intravenous antibiotics, parenteral or enteral administration of a high energy and high protein nutrition, and drainage of the pleural cavity. Large leaks or total anastomotic breakdown should be treated by surgery to refashion the anastomosis or to close the fistula. This is often very difficult because of oedema and infection and it may be necessary to excise the stomach and oesophagus performing a cervical oesphagogostomy and a feeding jejunostomy. Should the patient survive the reconstruction, utilization of an isoperistaltic segment of the left colon can be undertaken 3-6 months later.
Conclusions

The oesophagus should be treated with great respect and only surgeons who have received full training in oesophageal surgery in all its aspects should undertake operations on the oesophagus. Usually one has a single chance to get things right, seldom two and rarely three.
Chapter 23: Lower respiratory conditions in otolaryngology

Christopher C. Evans

Several otolaryngological conditions are aetiologically associated with respiratory disorders, and respiratory disease is a common cause of morbidity and mortality in the UK. Thus the principal conditions discussed are nasal and bronchial atopy, sinusitis and bronchiectasis, carcinoma of the bronchus and pulmonary metastases, malignant pleural effusions and postoperative respiratory complications. Some granulomata such as tuberculosis, sarcoidosis and Wegener's granulomatosis may involve the upper and lower respiratory tract and chronic bronchitis and emphysema may influence the treatment options of the otorhinolaryngologist. This chapter is written by a thoracic physician who regrets the historical anatomical barrier imposed by the larynx, which has separated the two disciplines.

Rhinitis, nasal polyps and bronchial asthma

Hippocrates recognized the association between nasal polyps and bronchial asthma, and pollen allergy can produce seasonal allergic rhinitis and seasonal asthma in atopic subjects. The reported incidence of asthma in patients with nasal polyps has ranged from 3% to 72%, while the incidence of nasal polyps in bronchial asthma has varied in reported series from 23% to 42%. Moloney and Collins (1977) calculated that a patient with nasal polyps or with bronchial asthma has a 25% chance of developing the other disorder.

Asthma and rhinitis can be divided into atopic or non-atopic groups according to skin prick testing and IgE levels. The atopic triad of allergic rhinitis, eczema and asthma is seen in up to 70% of all asthmatics. Atopy occurs in up to 30% of the population however, and is detected in all racial groups being maximal in the third decade. Only a proportion of atopic individuals ever develop nasal, skin or respiratory symptoms and it has been demonstrated that skin prick positive students are more likely to suffer from rhinitis and asthma than skin prick negative students after 3 years of follow-up (Hagy and Settipane, 1971). The common allergens by which the atopic status is determined are house dust mite and house dust, grass and tree pollens, animal danders and moulds, especially Aspergillus fumigatus. In 30% of asthmatics no extrinsic cause can be identified and these patients are skin prick negative with normal IgE levels and negative radioallergosorbent (RAST) values. They do however have raised sputum and blood eosinophil counts which are a marker of asthma and not of atopy. Such intrinsic asthmatics may also have rhinitis which, like their asthma, is perennial (Lessof, 1981).

Special mention must be made of bronchial asthma in association with nasal polyps and aspirin sensitivity. This affects up to 5% of all asthmatics who are sensitive to aspirin and other analgesics, as well as the azo dye, tartrazine, found in many food stuffs and drugs. It is thought that the salicylates and other analgesics alter the homeostasis of arachidonic acid mediators such as prostaglandins and leukotrienes, as well as platelet aggregating factor (PAF), thus precipitating bronchospasm (Szczylik, Gryglewski and Czerniawska-Mysik, 1975).

The histological changes in the nasal and bronchial mucosa of patients with rhinitis, polyps and asthma are similar and show thickening of the basement membrane, eosinophilic
infiltration, oedema and epithelial hyperplasia. In addition to atopy, possible aetiological factors common to all three disorders include bacterial and viral infections, drugs such as analgesics and beta blockers, autonomic dysfunction and exercise when cold dry air is inhaled. Occupational causes of bronchial asthma are being increasingly recognized, but the role of food allergy is not yet clearly established.

The effect of nasal polypectomy on asthma is controversial. Deterioration, improvement and the initial appearance of asthma have all been noted after polypectomy, and such changes may merely reflect the spontaneous fluctuation in asthma.

Clinical features

Bronchial asthma is characterized by attacks of wheezing breathlessness which often disturb sleep, are often most severe at breakfast time and limit exercise during the day time. In children, a persistent cough and a tentative diagnosis of recurrent bronchitis should suggest a diagnosis of asthma. In severe attacks, which can be fatal, progressive deterioration occurs which limits the victim first to his house, then his bed and finally interferes with eating, drinking and speech. Approximately 2000 people die annually from bronchial asthma in the UK and most have been breathless for several days before their final illness.

Management

There is a necessity for patients, their relatives and their medical advisers to appreciate the severity of an attack of asthma, and this is most effectively and economically achieved by obtaining a measurement of the peak expiratory flow rate. All general practitioners who visit a patient with asthma should have such an instrument, and in hospital patients, an estimate of the peak expiratory flow rate should be made regularly, not only on medical wards, but especially on surgical units. Regular monitoring of patients with asthma, which may demonstrate a deterioration in function, should alert the doctor and the patient to institute more effective treatment and seek assistance from physician colleagues (Clarke and Godfrey, 1983).

The treatment of asthma is not normally curative. Known allergens such as house dust mite, grass pollens and pet danders should be avoided if possible and strict measures to reduce house dust mite counts in the mattress can be beneficial. These can include the use of synthetic duvets, pillows, sheets and blankets, covering the mattress with plastic covers which should be damp dusted and scrupulous vacuuming of the bedroom carpet or its replacement with linoleum.

Although immunotherapy with hyposensitization may be effective in hay fever, there is no evidence to show that such procedures have any value in the management of bronchial asthma.

The principal drugs used to relieve an acute attack of asthma are the beta-adrenergic or sympathomimetic bronchodilators. Most mild to moderate attacks of asthma respond rapidly to the aerosol administration of a selective beta₂-stimulant such as salbutamol, terbutaline, fenoterol or rimiterol. These agents produce few side-effects apart from tremor when inhaled and their speed of action is prompt when compared to oral administration.
Patients should be instructed in their use, their technique checked regularly, and they should be warned against excessive use. When a patient with asthma fails to respond to the usual dose of inhaled bronchodilator, he should be advised to seek further medical advice. Oral preparations are given to children who cannot operate aerosol inhalers and intravenous injections of salbutamol or terbutaline are of value in severe asthma when bronchospasm prevents the use of the aerosol. Aqueous aerosols or respirator solutions of salbutamol or terbutaline can be administered for 15-20 minutes from a nebulizer in both hospital and domiciliary practice, and are useful in the initial treatment of severe or worsening asthma.

Xanthine bronchodilators such as aminophylline and theophylline in sustained release preparations provide effective background bronchodilatation with twice daily oral administration and a slow intravenous injection of aminophylline is still used by many physicians to relieve acute severe asthma. When this is given, it is vital that the dose is adjusted according to the plasma concentration, if oral xanthine preparations have been previously prescribed.

Prophylactic treatment of asthma requires the regular administration of steroids or cromoglycate. Inhaled steroids such as beclomethasone or budesonide, and sodium cromoglycate inhaled from the spinhaler or aerosol must be taken regularly. Sodium cromoglycate has few side-effects; inhaled steroids may cause hoarseness and oral candidiasis. This can be treated with oral fungicides.

Oral steroids may be required for prophylaxis when control of airflow obstruction has not been achieved using maximum inhaled doses and a short high dose, quickly reducing course of prednisolone is used in acute asthma.

Acute severe asthma, formerly called status asthmaticus, should be treated in hospital by experts using oxygen, intravenous hydrocortisone, nebulized salbutamol and possibly intravenous aminophylline. Such patients require careful monitoring of pulse, blood pressure, peak expiratory flow and blood gas tensions, and the decision to transfer the patient to an intensive care unit should be made early rather than late. Intermittent positive pressure ventilation should only rarely be necessary.

**Sinusitis and bronchiectasis**

Upper respiratory tract infection accounts for up to 50% of lost working days per year from illness in the UK and infection of the paranasal sinuses develops in a significant number. While most paranasal sinus infection results from nasal disease such as deviated septum or polyps, it must be remembered that the columnar mucociliary lining is in continuity from the nose down to the terminal bronchiole. Our understanding of the interrelationship between the upper and lower airways has been advanced recently and sinusitis with or without polyps is frequently detected in patients with bronchiectasis.

**Clinical features**

Bronchiectasis is characterized by chronic dilatation of one or more bronchi. The disorder is recognized by recurrent cough with large volumes of infected sputum which may be complicated by haemoptysis, airways obstruction, or pneumonia and pleurisy. Bronchial
obstruction by mucus plug, foreign body or tumour will result in bronchiectasis if left untreated, and when accompanied by infection, structural distortion of the bronchial tree with pulmonary fibrosis will develop. Many cases of bronchiectasis used to follow tuberculosis, whooping cough and measles, but the incidence of these primary disorders is falling. In health, the bronchial tree is protected by the nasal filter, the cough reflex and normal function of the mucociliary escalator containing immunoglobulins. Congenital and acquired disorders of these functional elements lead to bronchiectasis and, in many cases, sinusitis and structural abnormalities of the paranasal sinuses coexist. Thus bronchiectasis is a constant feature of patients with the autosomal recessive disorder of cystic fibrosis, who frequently have sinusitis and nasal polyposis showing a lymphocytic infiltration. In Kartagener's syndrome and other inherited disorders of ciliary malfunction, a defect in the microstructure of the tubules of the cilia and sperm leads to male infertility, bronchiectasis, situs inversus with dextrocardia and sinusitis or absent frontal sinuses.

In congenital disorders of immunoglobulin synthesis such as agammaglobulinaemia or combined hereditary immunoglobulin and T-cell functional defects such as ataxia telangiectasia, bronchiectasis and sinusitis coexist. Immunoglobulin A is the functional immunoglobulin of sputum, and it may be reduced or disappear following chemotherapy and radiation treatment for Hodgkin's disease and allied disorders (Berdal et al, 1976).

In all these conditions characterized by sinobronchitis, the common infecting bacteria are \textit{Streptococcus pneumoniae}, \textit{Haemophilus influenzae}, \textit{Staphylococcus} spp. and \textit{Pseudomonas} spp. Of special note is the fungus \textit{Aspergillus fumigatus} which has a special predilection for the upper and lower respiratory passages in subjects with impaired mucociliary function. Proximal bronchiectasis may occur in allergic bronchopulmonary aspergillosis characterized by asthma, fleeting eosinophilic pulmonary shadows, positive skin prick tests to \textit{Aspergillus}, eosinophilia and aspergillus precipitins in the serum.

\textit{Treatment}

The sinuses and bronchiectasis require simultaneous and coordinated management. Bronchiectasis is principally controlled medically with postural drainage and self-physiotherapy, appropriate antibiotics for superadded infection and bronchodilators for concomitant airways obstruction. Surgery for bronchiectasis is rare nowadays. Previous surgical results have often been unsatisfactory as a consequence of the diffuse nature of the condition, leading to recurrent symptoms after surgery. However, if the bronchiectasis is demonstrably localized by bilateral bronchography and if pulmonary function is good, then localized resection may be considered, particularly after a spell of failed medical treatment. In some patients with immunodeficiency, substitution therapy with periodic gamma globulin injections may be beneficial.

\textbf{Chronic bronchitis and emphysema}

These disorders are the commonest cause of loss of work time in the UK and they are much more common in men. They frequently cause postponement or cancellation by the anaesthetist of elective surgery. Chronic bronchitis is defined as a persistent cough with sputum for more than 3 months per year for 2 consecutive years. In contrast, emphysema is defined as a dilatation of the air passages beyond the terminal bronchiole and is a pathological
entity associated with destruction of the gas exchanging area of the lung. Both conditions usually exist together and are characterized by airflow obstruction with breathlessness, wheeze on forced expiration and physiological evidence of expiratory airways obstruction. The disorders, often referred to as chronic obstructive airways disease, are seen in smokers from major industrial conurbations. Acute infective exacerbations are a common cause of hospital admission in the winter. Patients have symptoms for decades with a progressive decrease in exercise tolerance, so that they become breathless at rest. The onset of right-sided cardiac failure heralds cor pulmonale caused by hypoxia. Death is associated with respiratory and cardiac failure and may follow elective surgery.

It is essential that a measure of respiratory function, such as the forced expiratory volume in one second and forced vital capacity and their ratio, is made in all patients with a history of chronic obstructive bronchitis about to undergo surgery. Blood gases may also be required to assess alveolar ventilation in these patients. It is important to realize that elective surgery should not be cancelled until all reversible features have been attended to. All patients should be encouraged to stop smoking. Antibiotics should be given for infected sputum, diuretics for cardiac failure and fluid retention, nebulized bronchodilators such as sympathomimetics (see Asthma treatment) and anticholinergics such as ipratropium bromide and physiotherapy for sputum retention. Especially in the summer months and after a careful medical assessment, many patients can negotiate surgery, who otherwise may be rejected or develop postoperative respiratory failure caused by sputum retention, painful respiratory excursions, or the injudicious prescription of respiratory depressants given to relieve pain.

Postoperative dyspnoea

Postoperative pulmonary complications may be anticipated in subjects suffering from the common cold or acute bronchitis undergoing surgery and the operation should be postponed until recovery is complete. In patients with chronic bronchitis and emphysema, bronchial asthma, bronchiectasis and lung fibrosis, intensive physiotherapy should be given before and after surgery. The anaesthetic and postoperative analgesia, retained secretions and impaired respiratory excursions caused by pain are likely to aggravate these conditions and may lead to postoperative atelectasis, pneumonia and bronchospasm.

Atelectasis

Atelectasis is the principal cause of dyspnoea within the first 24 hours after operation. Tachypnoea, tachycardia, restlessness and cyanosis develop and the physical signs are deviation of the trachea and mediastinum towards the affected side, impaired percussion note and diminished or absent air entry on that side. Only when bronchial obstruction is incomplete with alveolar collapse will there be bronchial breath sounds. The chest radiograph may show basal linear opacities, lobar or complete lung collapse. Treatment consists of physiotherapy and when sputum has been obtained, a broad-spectrum antibiotic such as amoxycillin or trimethoprim should be prescribed. Bronchoscopy should be considered when physiotherapy has failed to result in any air entering a pulmonary segment. In elderly patients with collapse, the increased work of breathing may lead to respiratory and cardiac failure, and this can be avoided by timely endotracheal intubation, tracheal toilet and, if necessary, intermittent positive pressure ventilation.
**Pneumonia**

In patients with pre-existing respiratory disorders such as chronic bronchitis and emphysema, deterioration may be detected on the second or third postoperative day by the onset of severe breathlessness, wheezing, fever, cough, tachycardia and signs of consolidation (bronchial breathing and whispering pectoriloquy). Pneumonia has developed and this should be confirmed by a chest radiograph. It is usually caused by infection with *Streptococcus pneumoniae* or *Haemophilus influenzae*. Treatment with broad-spectrum antibiotics, to which the organisms are likely to be sensitive is indicated.

**Pulmonary embolus**

A pulmonary embolus usually results from a deep vein thrombosis in the legs and less commonly from a pelvic venous thrombosis. Venous stasis, hypercoagulability and local injury to the vein may all be found postoperatively, especially in those with cardiac failure and with a poor peripheral arterial supply.

A massive pulmonary embolus may cause cardiac arrest, chest pains simulating a myocardial infarction, acute dyspnoea, syncope or faintness and a smaller embolus is more likely to produce a pulmonary infarction with dyspnoea, pleuritic chest pain and haemoptysis. These patients are usually cyanosed, shocked with a tachycardia, thready pulse and prominent jugular venous pulsation. There may be a triple rhythm on cardiac auscultation. The diagnosis should be considered on any postoperative day since preoperative immobilization may have produced venous stasis. The electrocardiogram may show the S1, Q3, T3 wave pattern which consists of a deep S wave in lead 1 and a Q wave and inverted T wave in lead 3. The T waves may also be inverted in leads V1-V4 and a right bundle branch block pattern may develop. A normal electrocardiogram does not rule out the diagnosis, nor does a normal chest radiograph. Nevertheless, chest radiographic changes are frequently detected such as linear atelectasis, a basal effusion, a raised hemidiaphragm, a wedge-shaped infarction or oligaemic areas. The definite diagnosis is made by pulmonary angiography, but ventilation perfusion lung scanning is usually available and provides adequate diagnostic information. Treatment consists of intravenous heparin pump therapy for at least 5 days, to be followed by oral anticoagulants. Nowadays thrombolytic therapy and surgical embolectomy are rarely required.

**Aspiration pneumonia and the adult respiratory distress syndrome**

Inhalation of vomit containing gastric contents leads to severe lung damage and occurs especially in children and the elderly and when emergency anaesthesia is given without gastric intubation.

If aspiration is suspected, the head should be lowered while the trachea and bronchi are lavaged with saline. Usually the inhalation is not detected and 3-5 hours after operation the patient deteriorates with cyanosis, tachypnoea and tachycardia. Crackles and wheezes are heard and arterial blood gases reveal profound hypoxia (low oxygen tension) and probably hypocapnia (low carbon dioxide tension) secondary to tachypnoea. The chest radiograph shows patchy areas of alveolar shadowing, more often right sided, but not confined to the lower zones. After the acute episode the patient may improve, but progressive deterioration may ensue over the succeeding days with increasing stiffness of the lungs requiring ever
increasing inspired oxygen tensions to combat hypoxaemia. The adult respiratory distress syndrome has developed. Patients should be transferred to the intensive care unit for oxygen therapy, antibiotics, intravenous hydrocortisone and if hypoxia cannot be relieved, intermittent positive pressure ventilation often with positive end expiratory pressure (PEEP). Even so the prognosis is poor. The adult respiratory distress syndrome may also be caused by shock and over-hydration, sepsis and endotoxaemia, oxygen toxicity, fat and thrombus embolism and extensive atelectasis.

Elderly or intoxicated adults can choke while chewing a large bolus of meat. This may impact in the larynx and lead to asphyxia. It should be relieved by the Heimlich manoeuvre, where sudden direct pressure is applied to the patient's stomach by the attendant encircling his arms around the victim with expulsion of the bolus.

**Inhaled foreign bodies**

The otolaryngologist, the chest surgeon, or the chest physician may be called upon to deal with an inhaled foreign body depending on its type, location, and available resources and expertise. The foreign bodies may be divided into exogenous or endogenous, organic or inorganic. Most are aspirated into the right lobe in the conscious patient for anatomical reasons. In the unconscious patient, they may be inhaled into the apical segment of the right lower lobe or the posterior segment of the right upper lobe, and again less frequently on the left.

**Exogenous foreign bodies**

Holinger and Holinger (1978) have reported their experience in over 2000 patients with laryngeal, tracheal and bronchial foreign bodies. Of 534 of these patients seen between 1961 and 1975, 76% were children below 4 years of age, 18% were aged between 4 and 14, and only 6% were over 14 years of age. Peanuts, popcorn or seeds were present in 60%; hardware, pins and pens in 23%; dental objects in 3%; and a miscellany in 14%. Vegetable matter, such as a peanut, excites a rapid, severe chemical bronchitis and is more noxious than inorganic non-vegetable matter.

**Clinical picture**

Episodes of choking, gagging and cyanosis are followed by unilateral wheezing if the object lodges in the bronchus, or bilateral wheezing if in the trachea. Thereafter there may be a symptomless interval varying from hours to months depending on the nature of the foreign body, location and degree of bronchial obstruction.

Vegetable matter excites an early reaction leading to purulent bronchitis. Any foreign body may lead to obstructive emphysema seen on the expiration chest X-ray or to collapse with complete bronchial occlusion.

**Diagnosis**

The clinical history is usually suggestive but may be absent in infants who inhale tiny plastic toys, for instance, which are not radiopaque. Unilateral wheezing is highly suggestive,
particularly when associated with decreased chest movement, impaired percussion note, and reduced breath sounds distal to the foreign body.

Chest X-ray on expiration is essential, particularly if the foreign body is radiolucent and cannot therefore be seen. Bronchoscopy is mandatory.

**Removal of foreign bodies from the tracheobronchial tree**

Foreign bodies should be promptly removed since only 2-4% are coughed out spontaneously. In small children (below the age of 10 years) general anaesthesia with rigid open-tube bronchoscopy is indicated. This should be performed by a bronchoscopist expert using the variety of special forceps (see Volume 6, Chapter 29) now available. Thoracotomy with bronchotomy or segmental resection is rarely justified for removal of an endobronchial foreign body.

In older patients (over the age of 10 years), in whom the larynx and trachea are larger, fibreoptic bronchoscopy under local anaesthesia may be used. Rapid strides have been made in the past few years with this technique and Cunanan (1978) has reported his experience in 300 cases of which 89% were performed with a fibreoptic bronchoscope alone, the remaining 11% being carried out with a rigid bronchoscope. He was able to remove the same range of foreign bodies as above with special forceps. He mentioned a significant drop in mortality and morbidity from 12% in the previous 5 years to 1% in the last 5 years on switching to the fibreoptic bronchoscope. He attributed this fall in part to the avoidance of general anaesthesia in patients with complicating illnesses. Again it must be stressed that the bronchoscopist must be experienced and have the appropriate forceps. A point in favour of fibreoptic bronchoscopy is that the technique is simple enough to allow visual inspection to assess the situation without necessarily attempting immediate removal.

**Endogenous foreign bodies**

Saliva and mucopus may be inhaled by patients in coma from whatever cause and in those with neuromuscular disorders such as a bulbar palsy. In some such cases, a cuffed endotracheal tube with a floppy, low-pressure cuff released at regular intervals may be indicated. Releasing the cuff pressure periodically avoids tracheal trauma and subsequent stenosis. In protracted cases, tracheostomy may be necessary.

During ear, nose and throat surgery, blood and debris may be aspirated into the lungs, although modern anaesthesia with tracheal intubation has largely eradicated this complication.

**Tumours of the tracheobronchial tree**

Nearly all tumours of the tracheobronchial tree are malignant and bronchial carcinoma is the commonest neoplasm in the UK. Benign tumours such as hamartoma, chondroma or lipomata are rare and tumours of low malignancy such as bronchial carcinoid and cylindroma are unusual.
**Carcinoma of the bronchus**

Most cases occur in men over the age of 50 years but the mortality in women is rising with the increased prevalence of women smokers. The evidence incriminating cigarette smoking as the important aetiological factor throughout the world is overwhelming in both retrospective and prospective studies. Occupational hazards such as exposure to asbestos, radioactive materials, nickel and industrial arsenic are recognized risk factors.

Two-thirds of all patients present with respiratory symptoms including cough, haemoptysis and unresolved pneumonia. About 25% present with evidence of metastases or non-metastatic extra thoracic manifestations such as endocrine disorders (Cushing’s syndrome, inappropriate ADH secretion), neurological disorders (encephalopathy, neuropathy, myopathy), and constitutional symptoms such as weight loss, finger clubbing, hypertrophic pulmonary osteoarthropathy or thrombophlebitis. Five per cent of patients are symptom free when the lesion is detected on a routine chest radiograph.

Of special interest to the otorhinolaryngologist is the presentation of a carcinoma of the bronchus with a hoarse voice as a result of the left recurrent laryngeal nerve palsy, and a paralysed left vocal cord, malignant glands in the neck with or without a Horner's syndrome, superior vena caval obstruction with mediastinal invasion and stridor caused by narrowing of the trachea or main bronchi. The diagnosis is suggested by the history and radiological appearances. It is confirmed by histology, usually obtained by sputum cytology or bronchoscopy. Fibreoptic bronchoscopy employing biopsy and brushings yields positive results in about four out of five cases. In peripheral lesions out of range of even the fiberoptic bronchoscope, percutaneous needle aspiration is the diagnostic option of choice for lesions greater than 2 cm in diameter. For smaller lesions the diagnosis may only be made at thoracotomy.

When a diagnosis of bronchial carcinoma is suspected, referral to a thoracic surgeon or physician is advisable. In patients without metastases who have adequate pulmonary function, surgical resection by lobectomy or pneumonectomy offers the best chance of cure with 5-year survival figures for squamous carcinoma and adenocarcinoma in excess of 30%. It should be appreciated, however, that only 5% of all patients who present actually survive 5 years. Radiotherapy and chemotherapy rarely produce cure. Good palliation of haemoptysis, cough, dyspnoea and bone pain may result from radiotherapy, and intravenous chemotherapy regimens using multiple agents are significantly extending survival in those patients with small cell carcinoma. Laser therapy via the bronchoscope is a useful palliative procedure in controlling haemoptysis, stridor and severe breathlessness in patients with endotracheal and endobronchial tumours obstructing the main airways.

**Bronchial adenoma**

This tumour can become malignant and metastasize. Nevertheless, 75% survival at 15 years has been described in the largest reported series to date (Lawson et al, 1976).

Haemoptysis is the commonest presentation and bronchial carcinoma is usually suspected. At bronchoscopy a haemorrhagic cherry-like tumour is visualized, the histology of which reveals bronchial carcinoid or adenoid cystic carcinoma.
Depending on the site, lobectomy or pneumonectomy will be required and follow-up should be prolonged because of the late development of local recurrence which may be amenable to further surgical resection.

Metastatic pulmonary and pleural disease

The lungs and pleura are possible secondary sites of metastatic involvement of primary tumours originating in the head and neck. Pulmonary involvement by secondary deposits usually presents with cough and breathlessness. Crackles may be heard and the chest radiograph will reveal either multiple unequal irregular rounded opacities or linear reticular shadows indicative of carcinomatosis lymphangitis. Transbronchial lung biopsy via the flexible fibreoptic bronchoscope will be required to confirm the diagnosis. Chemotherapy may be given according to the site of the primary tumour and prednisolone may alleviate dyspnoea temporarily.

Malignant pleural effusions usually present with breathlessness and constitutional symptoms such as weight loss and malaise. Only in about 25% of patients is pain on the affected side a leading symptom. The effusion, which is an exudate, is often blood stained, should be tapped, and at the initial aspiration pleural biopsies should be performed to obtain histological proof concerning the nature of the effusion. Pleural fluid cytology and cytogenetics may also confirm the malignant nature of the fluid. The effusion should be tapped to dryness. If the fluid reaccumulates rapidly, pleurodesis should be attempted using tetracycline or bleomycin into the pleural cavity. After the instillation, the patient should be tipped into several positions over 2 hours in order to distribute the sclerosing agent as uniformly as possible over the whole pleural surface.

In a patient in good overall condition with a malignant recurrent pleural effusion, pleurectomy with decortication may be considered.

Granulomata involving upper and lower respiratory tracts

Tuberculosis, sarcoidosis and Wegener's granulomatosis all have characteristic clinical and histopathological appearances and may be diagnosed by the otorhinolaryngologist or the thoracic physician.

Tuberculosis

Although pulmonary and constitutional symptoms predominate in post primary pulmonary tuberculosis, the initial presentation may be with a hoarse voice as a consequence of laryngeal involvement. Swabs, biopsy with culture as well as histopathology are indicated and such patients, who are usually sputum smear positive, should be isolated, referred to a thoracic physician and the diagnosis notified to the environmental health officer.

Cervical tuberculous lymph glands are still removed and sent only for histopathology, thus denying the microbiologist the opportunity to culture the organism and determine its sensitivity to antituberculous drugs. All lymph nodes removed from the neck should be sent for culture. In adults, tuberculosis is especially seen in Asians and Africans. It is also seen in children and the elderly of all races, as well as in patients receiving immunosuppressants
and others suffering from debilitating disorders such as neoplasia, diabetes mellitus, and alcoholism. Initial treatment consists of at least 2 months of combination therapy with bactericidal drugs including rifampicin, isoniazid and pyrazinamide and followed by a further 7 months of at least two of these. Because of side-effects these drugs should be administered and supervised by a thoracic physician with an expertise in tuberculosis, who will also arrange follow-up and contact tracing.

**Sarcoidosis**

This disorder of unknown aetiology commonly presents with erythema nodosum and bilateral hilar lymphadenopathy. Eighty per cent of such patients show no signs of disease nor recurrence after 2 years’ observation without any specific therapy. A systemic disorder, sarcoidosis can also present with symptoms referable to many body systems such as the skin, the eye, the cardiovascular system, the central nervous system, the endocrine system, the respiratory system and others (Table 23.1). Of special interest to the otorhinolaryngologist is the patient with sarcoidosis who presents with, or develops symptoms and signs such as cervical lymph nodes, parotid enlargement with a dry mouth, nasal obstruction and lupus pernio, epiglottic or vocal cord infiltration, or endobronchial sarcoidosis with stridor. Because of the multisystemic nature of this condition, it is wise to refer patients with sarcoidosis to a general or thoracic physician for an overall assessment. The value of diagnostic procedures such as biopsy, a negative Mantoux test (66%) and a positive Kveim biopsy revealing non-caseating epithelioid granuloma (80%) should be considered and treatment with steroids must be carefully evaluated for specific facets of the disorder, which may show serious deterioration if not treated.

**Table 23.1 Clinical manifestations in 537 patients with sarcoidosis reported from London**

<table>
<thead>
<tr>
<th>Clinical manifestations</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lungs and hilar nodes</td>
<td>84</td>
</tr>
<tr>
<td>Erythema nodosum</td>
<td>31</td>
</tr>
<tr>
<td>Peripheral lymph nodes</td>
<td>29</td>
</tr>
<tr>
<td>Eyes</td>
<td>27</td>
</tr>
<tr>
<td>Skin</td>
<td>25</td>
</tr>
<tr>
<td>Spleen</td>
<td>12</td>
</tr>
<tr>
<td>Central nervous system</td>
<td>7</td>
</tr>
<tr>
<td>Parotid</td>
<td>6</td>
</tr>
<tr>
<td>Bones</td>
<td>4</td>
</tr>
<tr>
<td>Others</td>
<td>1</td>
</tr>
</tbody>
</table>

**Wegener's granulomatosis**

The otorhinolaryngologist will be well aware of this rare but potentially fatal disorder which may involve the nose, paranasal sinuses, middle ear and larynx and, in many cases, pathological changes develop in the kidneys, lungs and joints. Symptoms suggesting lower respiratory tract involvement are cough, haemoptysis and pleuritic chest pain, and the chest radiograph shows single or multiple rounded opacities which may cavitate and may
spontaneously heal. The diagnosis in the lung or the kidneys requires adequate biopsy which may not be made from the nasal lesions. Therapy with immunosuppressant drugs such as prednisolone and cyclophosphamide is effective in pulmonary Wegener's granulomatosis and may induce remission so that treatment can be withdrawn.

The otorhinolaryngologist should be aware of the acquired immune deficiency syndrome and obstructive sleep apnoea syndrome which often present to the thoracic physician and are dealt with in chapter 5.
Chapter 24: The management of terminal cancer

The treatment of pain due to malignancy of the head and neck

J. B. Miles

Malignancy of the head and neck is often associated with severe pain. This probably relates to the embryological concentration of sense organs at the head end - that with which the animal confronts his environment. Radical resection, with reconstruction, is sometimes the primary treatment and often also proves effective in relieving pain. However, more commonly treatment consists of histological verification by biopsy followed by radiotherapy. There is no doubt that radiotherapy can relieve pain, but the likelihood of recurrence after such treatment is high and this is usually recognized by the return of the original pain.

With the realization that pain due to recurrence of malignancy in the head and neck is not easy to relieve, it is becoming recognized that secondary or 'salvage' surgery, although not necessarily curative, may be the most appropriate and effective way of relieving the pain (Miles, 1984). Malignancy of the head and neck, particularly when recurrent, extends beyond the specialized boundaries of otorhinolaryngology, ophthalmology, dentistry and neurosurgery, and 'salvage' surgery may be more effective if undertaken by specialist surgeons, in combination.

In recommending such surgery there may be a fine balance between pain relief, with perhaps improved survival, and the possible mutilation and psychological insult of further and massive surgery. If the latter is deemed excessive or the tumour physically unresectable, then treatment should become empiric, directed at the symptomatic relief of pain.

Surgical resection

Basal or squamous cell carcinoma of the scalp

Such cancers are usually not painful until they have invaded the underlying bone and even then the penetration usually has to be complete and to involve the dura for the symptoms to become severe. Radiology may indicate invasion of the bone but will not accurately quantify the extent. Even computerized tomographic (CT) scanning, while more accurately defining whether total bone thickness invasion has occurred, will not easily delineate the extent and degree of infiltration of the dura mater. The only safe way to manage such a situation is to anticipate dural involvement, being prepared for wide dural resection and grafting. The presence of a neurosurgeon at such a procedure would seem advisable, especially if the bone overlies treacherous intracranial structures such as venous sinuses. The dura may be reconstructed with pericranium, usually readily available when large areas of scalp are being mobilized to compensate for the resected tumour, or by lyophilized homologous cadaveric dura.

Cancers of the paranasal sinuses

These cancers are almost always painful and highly malignant. Although some illustrate major sensitivity to radiotherapy by shrinkage (and relief of pain), block resection
is usually considered advisable particularly when the response to radiotherapy has been limited or there is frank evidence of recurrence. Computerized tomographic scanning will illustrate substantial intracranial extension, as notoriously occurs with adenoid cystic carcinoma, and the much less common neuroblastoma, but then only if the radiologist is aware of the need to attenuate the pictures for intracranial tissue as well as bone. Iodine enhancement is also extremely valuable in delineating intracranial tumour from brain tissue. If scanning illustrates the cancer extending up to the base of skull, one must expect involvement of the bone, the need for resection of involved bone and the possibility of needing to resect the dura.

If there is evidence of intracranial spread then the lateral rhinotomy incision is easily extended superiorly in a curvilinear manner to allow an appropriate frontal craniotomy. Following resection of all the visible tumour the dura can be repaired by an on-lay graft of pericranium or lyophilized dura. It is sufficient merely to lay down the graft without stitching, covering it with a haemostatic sheet and allow the brain to descend on it. If the base of skull resection is sufficiently large to suggest the need for supporting this graft, then strips of the outer table of the skull are easily resected and used to bridge the gap below the dura and its on-lay graft.

If there is minimal or no intracranial extension then it is possible to resect the affected bone and the involved dura using the lateral rhinotomy approach and to repair the dural defect, from below, by an underlay lyophilized dural graft. The graft is made sufficiently larger than the defect to tuck in peripherally between the normal dura and the skull base. The cavity is then packed in a routine manner.

Less commonly, more benign tumours such as fibroma, meningioma and chordoma involve the same areas and provoke pain, and are best treated by the same combined surgical block resection. Sometimes the extent of the tumour makes it more appropriately treated by two-stage procedures.

**Cancers of the middle and external ear**

Again, biopsy followed by radiotherapy is the usual primary treatment for these tumours and rarely radical petrosectomy. Severe pain is invariable and recurrence is common. If the tumour appears limited to a resectable area and, in practice, this means lateral to the carotid canal and foramen ovale, then early radical petrosectomy should be considered.

Computerized tomographic scanning is essential, as the deeper tumours can easily extend intracranially, and again the emphasis must be made that the attenuation of the CT scanner for intracranial extension must be that for intracranial contents and not simply bone. The author has encountered, at surgery, an intracranial abscess extending up from the diseased petrous that had not been delineated by scanning with attenuation only for bony tissues. The general condition of the patient and the natural history of the tumour growth must be taken into consideration in deciding on such radical surgery. The practical and cosmetic defects of resecting the ear, and manipulation of the scalp or musculocutaneous flaps, must be explained together with the risks to lower cranial nerves, some of which, such as the seventh nerve, will already have been affected. Despite the substantial nature of this procedure, worthwhile results have been seen particularly regarding pain relief (Stell and Miles, 1986). There was even an
improved survival when the cancer arose from the external ear but not when its origin was in the middle ear.

The technical details of petrosectomy are described in Volume 3, Chapter 22.

*Cancers of the mouth*

Recurrent cancers of the tongue and floor of the mouth can equally prove amenable to major resection of most of the tongue and all of the mandible, with reconstruction by musculocutaneous flaps. The particularly distressing pain of recurrent carcinoma of the base of the tongue is often well palliated by this procedure.

*Malignancy of the cervical spine*

Metastatic cancers are much more common than primary bone tumours in the cervical spine. As with all bone tumours a background constant pain, particularly at night, and probably relating to expansion of the bone with periosteal stretching, can occur. However, in the cervical spine, easily the most mobile part of the skeletal spine, movement related pain, and pain on weight-bearing, are a particular feature and relate to instability. Often the nerve roots are compressed by the collapsing vertebral bodies and radiating pain results. Instability can be recognized from the patient's complaint and is often verified radiographically. Treatment must be directed to establishing stabilization. While radiotherapy can undoubtedly relieve the constant periosteal bone pain it can achieve little when instability is the problem and can possibly aggravate the situation.

Simple measures such as the use of a collar are likely to be of help only when a patient has constant pain with slight weight-bearing aggravation. When the instability is more florid, even the substantial hard collars with breast plate support are rarely effective. The option of skull fixation by halo, with pillars to a chest cast, is seen by most patients as incompatible with an acceptable quality of life.

As most metastatic cancers occur in the vertebral bodies rather than the rest of the vertebrae (Kakulas et al, 1980), and as the main weight-bearing is through the vertebral bodies, then surgical stabilizing procedures are best directed at the diseased vertebral body. In the cervical spine, the anterior approach to the vertebral bodies, as used for cervical disc resection, is familiar to neurosurgeons. The resected vertebral body can be replaced by a bone graft or by a synthetic prosthesis with or without internal fixation. When the vertebral bodies involved by malignancy are considered too extensive to be resected, then a posterior approach will allow internal fixation by rods or wires and alleviate the symptoms. This procedure is, however, less efficient and more painful than the anterior approach.

*Empiric treatment of the pain*

*Medical treatment*

Medical treatment, where it is effective and not associated with intolerable side-effects, is clearly always preferred to surgical manoeuvres. Simple analgesics, particularly anti-inflammatory preparations, either non-steroidal or steroidal, have a part to play especially
when bone is involved. Narcotic medication, being more effective in the sense of a stronger analgesic, and being less of a concern from the abuse standpoint in patients with malignancy and limited life expectancy, is likely to be introduced into the treatment early.

The major recent interest in the management of terminal states, and the development of the hospice movement, has illustrated both the efficiency and appropriateness of regular and adequate morphine administration for pain associated with malignancy (Twycross, 1985). The very common association of nausea and the obligatory association of constipation with such treatment does mean there is a need for prophylactic antiemetic and laxative medication. Still, some people cannot tolerate these side-effects and/or complain bitterly of a 'drugged' mental state associated with high morphine medication.

Recognition of the specific receptors for opiates in brain and spinal cord have led to attempts to achieve morphine analgesia, while minimizing side-effects, by delivering the morphine directly to the central nervous system. Extradural (epidural), and intrathecal, spinal morphine application does relieve pain using very small doses (Wang, Nauss and Thomas, 1979). Implantable delivery systems for chronic administration of morphine have been developed, some simple one-shot percutaneous injection systems and some more complicated multiple dose and powered systems, although the latter appear to be prohibitively expensive (Poletti et al, 1981). Unfortunately these systems still have a risk of respiratory depression and those applied to the cervical spine would seem more vulnerable in this respect.

Morphine administered into the ventricles of the brain is perhaps even more effective as an analgesic. Using dosages of less than 1 mg analgesia can often last days after a single injection. In suitable cases this method of administration can prove effective, although training is clearly required and optimally a member of the family performs the percutaneous puncture of the scalp reservoir rather than leaving the administration to visiting nurses. This method of application is effective for pain due to malignancy anywhere in the body, but is perhaps more appropriate to that involving the head and neck where other options are limited (Lenzi et al, 1985). With the undoubted risk of catastrophic ventricular infection almost certainly correlating with the number of percutaneous punctures of the reservoir, this technique would seem justifiable only when life expectation is limited to weeks or at the most months.

Destructive procedures to the nervous system

Peripheral nerve destruction

Structures of the head and neck are innervated by the trigeminal nerve, the glossopharyngeal nerve, the vagus nerve and the upper four cervical nerves. The trigeminal nerve supplies a great area of skin on the face and scalp, the linings of the mouth and anterior tongue, and also the dura of the anterior and middle fossae. Peripheral nerve destruction is a quick method of denervating and relieving pain but unfortunately, in the head and neck, the malignancy tends to extend beyond the confines of a single nerve and it is therefore relatively rarely possible to provide enough pain relief by a single nerve destruction.

Trigeminal denervation is best achieved by the percutaneous cannulation of the foramen ovale when the ganglion or the sensory root can be destroyed by alcohol, phenol injection or, more commonly of late, by radiofrequency coagulation. As long as the foramen
ovale is radiographically definable this procedure can be undertaken, even in the presence of extensive tumours.

Glossopharyngeal nerve destruction is necessary when a malignancy involves the base of the tongue, the tonsillar region, eustachian tube and the middle ear and can also be achieved percutaneously by radiofrequency coagulation (Broggi and Siegfried, 1979). The medial neural notch of the jugular foramen needs to be radiographically defined and even then it is by no means certain that an effective nerve destruction will be achieved, but certainly it is worth trying. It is quite possible that coagulation in this region will also achieve some destruction of the upper vagus nerve fibers, which is desirable when one recognizes that the tenth nerve may also be involved in the innervation of the tumour area.

It is possible to undertake a surgical peripheral neurotomy by a posterior fossa approach and at one sitting to section the trigeminal nerve, the glossopharyngeal nerve and the upper vagus nerve fibres, and modern microsurgical technique makes this a relatively simple and safe procedure. Personal training and expertise would seem to determine whether a surgical or a percutaneous radiofrequency is used.

In the past, surgical division of the upper cervical sensory roots has also been undertaken but this procedure is very painful and current practice would suggest that percutaneous phenol neurolysis is to be preferred.

**Trigeminal sensory nucleus and tract destruction**

That part of the trigeminal nucleus and tract that descends to the second cervical level in the spinal cord predominantly subserves the sensation of pain. Therefore, destruction of these structures would provide in the head an equivalent of anterolateral cordotomy in the rest of the body, namely ipsilateral pain relief with loss of pinprick and temperature sensation but retention of other sensory modalities. This highly desirable ambition has not proved easily attainable in practice with, in particular, a distressingly high incidence of complications in the form of dysaesthetic sensations.

More recently, a technique involving undercutting of the dorsal root entry zones on the spinal cord is being applied to head and neck pains, the line of destruction being conducted from C3 up to the obex on the medulla. It remains to be seen whether this will prove superior to previous techniques.

**Thalamotomy**

Destruction of the medial thalamic sensory nuclei (central median and intralaminar) in many ways provides the most satisfactory form of pain relief, that is, immediate clearance of pain without recognizable sensory loss or psychological change. It is thought that the polysynaptic high threshold pathways subserving pathological pain pass to the medial thalamus, while the pathways subserving general sensation relay in the ventrolateral nucleus before proceeding to the sensory cortex. Unfortunately, in order to achieve accurate focal destruction of these nuclei a complex technique of stereotaxic surgery is necessary, involving fixation of a metal frame to the patient's head. The procedure has to be undertaken under local anaesthetic in order to ensure against adverse effects involving mistargeting and in order to
achieve sufficient destruction gradually, by incremental techniques, without excessive destruction resulting in morbidity. This technique, therefore, undoubtedly frightens patients. The resultant pain relief also tends to be limited in time and while one might expect, with surety, 3-6 months effect, to achieve more than one year is uncommon. However, the median survival of these patients is 10 weeks so that the use of such a technique is definitely appropriate.

Recent improvements in the basic stereotaxic technique with the use of computer aids in theatre, and even more the incorporation of CT scanning targeting, must lead to a justifiable resurgence in the practice of stereotaxic thalamotomy for cancer pain. Again, while this technique can be effective for pain at any level on the opposite side of the body, anterolateral cordotomy of the spinal cord would seem more appropriate for anything below the neck, while thalamotomy would appear to be more appropriate, and therefore reserved, for contralateral cancer pain of the head and neck (Spiegel and Wycis, 1966).

**Pituitary destruction**

Pituitary destruction by craniotomy was first practised by Luft and Olivecrona in 1952, in an attempt to suppress cancer growth (and pain) in tumours thought to be sensitive to gonadotrophins. Even then, it was clear that the pain relief and tumour regression were not achieved equally and also that on some occasions the pain relief was immediate. Increasingly, less threatening means were devised for destroying the pituitary gland, many using a percutaneous trans-sphenoidal approach (Miles, 1985). Moricca (1974) popularized a technique apparently first described by Greco, Sbaragni and Cammili (1957) by which, simply injecting alcohol into the pituitary gland suppressed some tumour growth and greatly relieved pain. This technique has been widely practised and the results described (Lipton et al, 1978). It has been possible to relieve pain in around 75% of cases and in over 40% the pain relief was total. Unfortunately, the duration of pain relief tends to be limited to weeks or months although periods extending over a year have been recorded. The technique is not without risk (Lipton et al, 1978). Although it is known that injury to the hypothalamus can occur due to spread of the alcohol up to the pituitary stalk and even that electrical stimulation of the pituitary gland itself can provide short-term relief of pain (Yanagida et al, 1984), a convincing explanation is still wanting (Miles, 1985).

Pituitary destruction can undoubtedly provide pain relief in patients unresponsive to other techniques so it would seem justifiable to continue its use. As we have no evidence to suggest that any one technique is better than another, then there would seem every reason to use that which is associated with least morbidity. At present that technique is probably radiofrequency coagulation by the trans-sphenoidal approach.

Among the patients treated in the Centre for Pain Relief, Liverpool, eight have had pain due to malignancy of the head or neck and their responsiveness to this technique has been the same as that with malignant pain elsewhere. Four patients had total relief of pain, two partial and two no pain relief. Again the pain relief tended to last only months.
**Electrical stimulation of the brain**

Studies into electro-anaesthesia showed that when the deep central grey areas surrounding the third ventricle (paraventricular grey) and the midbrain aqueduct (periaqueductal grey) were stimulated electrically in animals a state of surgical anaesthesia could be induced (Reynolds, 1969). Using the technological achievements of cardiac pacemaking and dorsal column spinal stimulation, chronic deep brain stimulation was achieved using implants activated from an external transmitter (Richardson and Akil, 1979). This technique has been used for non-malignant and malignant persistent pain and certainly has proved effective in a proportion of each. Stimulating these deep brain structures usually provokes no sensation, but within 30 minutes pain is gradually relieved, particularly when contralateral, but bilateral lower body pain can be relieved from unilateral stimulation (Richardson, 1979).

The implantation requires stereotaxic techniques with the unpleasantness previously described, a period of trial stimulation by a wire exiting from the scalp and, if the trial proves effective, a second operation, requiring general anaesthetic, for conversion of the percutaneous stage to that of a permanent implantation. The patient then induces activity in the implant by the local application of a transmitter to a buried receiver and the pain relief can last hours before further treatment is required.

The effect is probably mediated through the release of endogenous analgesics (Hosobuchi, 1980) and as such there is a tendency for the development of tolerance. The technique as a whole is somewhat complicated for the average debilitated patient with advanced cancer. Meyerson, Boethius and Carlsson (1979) have used the percutaneous stage as a permanent means which obviates some of the complications and certainly some of the expense, as each complete stimulator set currently costs £3000.

In many ways the repeated need to stimulate is a constant reminder that the cause for the pain persists and this suggests that deep brain stimulation is less suitable for the patient with advanced cancer than an ablative technique, if that can be achieved expeditiously and if it can be made to last.

**Conclusion**

Throughout the world pain clinics have proliferated resulting in efficient and prompt pain relief in many instances. Knowledge, methods and techniques have rapidly increased with the interest in pain and much more is therefore available for patients whose pain relates to malignancy in the head and neck. There is still a need to remember the virtues of surgical resection in relieving some pains and an ever increasing need and responsibility for communication between the specialists dealing with malignancy in this complex area.
Chemotherapy in terminal head and neck cancer

Randall P. Morton and E. B. Dorman

The general comments in this section will relate mainly to squamous carcinomata which comprise the great majority of head and neck cancers. Other epithelial tumours (adenocarcinoma, salivary gland cancer, mucosal melanoma) are less common, with a different pathogenesis and clinical behaviour pattern. Therefore these tumours will be considered separately.

Lymphomata and sarcomata comprise two other special groups for which chemotherapy has a particular role and it is not appropriate to discuss them further here.

Patients eligible for palliation will have advanced primary or recurrent disease which is deemed unsuitable for curative therapy, that is by radical radiotherapy or surgery. Recurrence may be at the site of the previously treated primary, in the regional nodes, or in distant organs, usually lung, liver or bone.

The aim of palliation and how palliative chemotherapy may be used in head and neck cancer patients will be discussed below.

Palliative chemotherapy: general principles

Palliation aims to improve the patient's quality of life without expecting to cure the disease. This is achieved by suppressing symptoms, but not necessarily prolonging survival in the process. However, since most of the major symptoms are due to the presence of tumour, effective palliation is only achieved by reduction of tumour size, which in turn is likely to prolong survival.

Clinical studies of chemotherapy are usually in the form of phase II or phase III trials. In a phase II study the end-point is the response of the tumour. A phase III trial is a much larger, controlled trial carried out to determine whether the activity of a drug noted in a phase II study can be confirmed, whether any new activity or adverse effects arise, and whether the drug has any effect on survival; the end-point is the death of the patient.

A favourable tumour response does not always equate to improved long-term survival. Three regimens were compared with a no treatment control group in the phase III trial. Median survival is extended by up to 63 days, depending on the regimen used. By 6 months a different pattern has emerged, with improved survival shown only in the single agent group, cisplatin (cisplatinum).

Quantitative assessment of tumour response is in fact often difficult, sometimes impossible, and always unreliable. Moreover, a tumour response does not necessarily equate with benefit to the patient. Survival time and morbidity are undoubtedly important for patients with a limited time to live, but increased survival may prolong, rather than relieve, the suffering, and treatment itself may cause side-effects without necessarily conferring any symptomatic relief.
The success of palliation in recurrent disease, as distinct from advanced primary
disease, is especially difficult to assess because the quality of life in patients treated for head
and neck cancer is usually compromised before any recurrence appears (Morton et al, 1985).

**Patient profile and natural history**

Patients are usually in a poor general and nutritional condition, with a performance
status of less than 60 on the Karnofsky scale *(Table 24.1)*. They are often dependent on
alcohol and many have chronic lung disease. Impaired renal and liver function may preclude
the use of some or all chemotherapeutic agents, or demand reduced dosages.

**Table 24.1 Definition of performance status in patients with malignancy**

*Karnofsky*

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>100</td>
<td>Normal; no complaints, no evidence of disease.</td>
</tr>
<tr>
<td>90</td>
<td>Normal activity; minor signs and symptoms of disease.</td>
</tr>
<tr>
<td>80</td>
<td>Normal activity with some effort; some signs and symptoms.</td>
</tr>
<tr>
<td>70</td>
<td>Cares for self; unable to carry out normal activity or work.</td>
</tr>
<tr>
<td>60</td>
<td>Requires occasional assistance; but able to care for most needs.</td>
</tr>
<tr>
<td>50</td>
<td>Requires considerable assistance and frequent medical care.</td>
</tr>
<tr>
<td>40</td>
<td>Disabled; special care and assistance needed.</td>
</tr>
<tr>
<td>30</td>
<td>Severely disabled.</td>
</tr>
<tr>
<td>20</td>
<td>Very sick; hospitalization needed; active supportive therapy necessary.</td>
</tr>
<tr>
<td>10</td>
<td>Moribund; fatal processes progressing rapidly.</td>
</tr>
<tr>
<td>0</td>
<td>Death.</td>
</tr>
</tbody>
</table>

*WHO/ECOG*

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Able to carry out all normal activity without restriction.</td>
</tr>
<tr>
<td>1</td>
<td>Restricted in strenuous activity but ambulatory and able to do light work.</td>
</tr>
</tbody>
</table>
| 2     | Ambulatory and able to self-care but unable to work; mobile for most of waking
hours. |
| 3     | Limited self-care; confined to bed or chair for most of waking hours. |
| 4     | Completely disabled; unable to self-care; confined to bed. |

If left untreated, patients with terminal head and neck cancer die in an exponential
fashion, with a median survival time of about 3 months (Stell, Morton and Singh, 1983). Simple measures such as nasogastric feeding tubes and tracheostomies are frequently
necessary to keep the patient comfortable without prolonging survival greatly.

Most patients require pain relief, and their quality of life can only be described as poor. The discomfort and difficulty with breathing, speaking and swallowing is often matched
by the cosmetic deformity of previous surgery, or by the tumour itself. A high rate of
depression, disability and discomfort has been reported in patients treated for head and neck
cancer (Morton et al, 1984).
Dissemination of head and neck cancer is often present by the time of death, but relatively few patients die because of their distant metastases. Most deaths seem to be related to the effects of recurrent or residual disease in the head and neck. Despite the poor quality of life, almost all patients wish to live as long as possible, but do not want to suffer pain or too much indignity. Chemotherapy is useful as an adjunct to the general care of terminal disease. It gives the patient a feeling that he is being purposefully managed, and his psyche will improve if he observes a tumour response. It is obviously not a palliation panacea.

Chemotherapy theory

See also Volume 1, Chapters 21 and 22.

Tumour cells are killed or attenuated by chemotherapy as a result of biochemical activity. The activity of a drug depends on its pharmacokinetic properties such as inactivation, binding to carrier protein, and excretion. The degree of 'response' that a tumour exhibits depends mainly on the growth fraction (the proportion of tumour cells actively engaged in cell replication). The proliferative phase of a cell cycle renders the cell especially vulnerable because biosynthesis of essential molecules is taking place.

Most of the cells that are not proliferating are 'resting', but retain the potential for replication. These resting cells are generally less vulnerable to chemotherapy.

Tumour destruction is though to follow first-order kinetics, in that a given treatment should kill a constant fraction of vulnerable cells, and not a fixed number. Most tumours follow a Gompertzian growth curve in which the growth fraction is large initially, but progressively decreases as the tumour increases in size. The tumour growth rate increases steadily to a maximum of 35-37% of total tumour volume, then progressively diminishes as the growth fraction continues to decrease. In other words, a large bulky tumour will have a relatively small proportion of tumour cells susceptible to chemotherapy.

Tumour sensitivity to the different chemotherapeutic agents varies. There are several drugs known to be active in head and neck cancer (Table 24.2). Attempts to identify tumours most likely to respond by measuring the proportion of cells in mitosis ('mitotic index'), or by radioactive labelling of those cells undergoing DNA synthesis ('labelling index'), have been generally inconsistent and unreliable.

Theoretically, the tumour mass should first be reduced (for example, by surgery), thereby increasing the growth fraction and rendering the tumour more susceptible to chemotherapy. Also, the combination of several chemotherapeutic agents should circumvent drug resistance through: (a) sequential blockade; (b) concurrent blockade; (c) complementary inhibition; (d) combination of individually active agents providing a more highly selective killing effect; or (e) kinetically based regimens (in which drugs are administered according to their activity in the various parts of the cell cycle). The kinetically based regimens have been popular in the past. They theoretically synchronize the cells entering a new cycle by arresting them in one phase using a phase-specific agent (for example, vincristine, which affects mitosis). Then, when most of the surviving cells are thought to be in the S phase, an S phase-specific agent, such as methotrexate, is given.
Table 24.2 Common chemotherapeutic agents in head and neck cancer, and the usual response rate reported when the single agent is used

<table>
<thead>
<tr>
<th>Agent</th>
<th>Type</th>
<th>Response rate (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cisplatin</td>
<td>Random synthetic</td>
<td>25-60</td>
</tr>
<tr>
<td>Methotrexate</td>
<td>Antimetabolite</td>
<td>20-60</td>
</tr>
<tr>
<td>Cyclophosphamide</td>
<td>Alkylating agent</td>
<td>20-40</td>
</tr>
<tr>
<td>Epirubicin</td>
<td>Antibiotic</td>
<td>30</td>
</tr>
<tr>
<td>Bleomycin</td>
<td>Antibiotic</td>
<td>15-25</td>
</tr>
<tr>
<td>Dacarbazine</td>
<td>Random synthetic</td>
<td>20</td>
</tr>
<tr>
<td>Doxorubicin</td>
<td>Antibiotic</td>
<td>15</td>
</tr>
<tr>
<td>5-Fluorouracil</td>
<td>Antimetabolite</td>
<td>15</td>
</tr>
<tr>
<td>Vincristine</td>
<td>Plant alkaloid</td>
<td>5</td>
</tr>
</tbody>
</table>

An approach to treatment

If the patient is not suffering from the presence or effect of the tumour, no active therapy need be given immediately. Instead, chemotherapy can be withheld and sample, relatively non-toxic and non-invasive measures may be sufficient. High-dose steroids provide a simple form of treatment which can palliate very effectively by causing euphoria, pain relief and reduction of oedema. Alternatively, cytotoxic chemotherapy can be considered if the patient specifically seeks treatment with a view to prolongation of survival, or if he is suffering from the effects of local invasion, and cannot be reasonably managed by medication, simple surgery or palliative radiotherapy.

The treatment will depend on the type of tumour and the patient's age, renal and hepatic function, haematological profile, and nutritional status. Combination chemotherapy can cause added toxicity.

Other forms of therapy include biological response modifiers such as BCG, levamisole/thymosin, and retinoids, but these are not commonly used in advanced disease. Indomethacin, a prostaglandin synthesis inhibitor, has been used alone in patients with advanced and metastatic disease and produced a response rate of 20% (Hirsch et al, 1983).

The patient must be warned about unwanted side-effects and the possibility that the treatment may be ineffective.

The assessment of benefit from chemotherapy for recurrent disease is very difficult, because of the compromised quality of life in patients who have been previously treated for head and neck cancer. It is hard to know how much discomfort is attributable to previous treatment and how much to recurrent tumour. It follows that antitumour therapy may be effective in reducing tumour size without materially improving the patient's 'quality of life'.

Survival time remains the most tangible method of assessing results; tumour response is impossible to quantify accurately, and 'quality of life' is an even more elusive parameter. A proper approach will take cognisance of all these factors, but the fact remains that for practical purposes effective palliation in terminal disease is an art (Shaw, 1985).
Contraindications to chemotherapy

Absolute contraindications are few and are generally very obvious. Chemotherapy must not be given if the patient is very frail or debilitated. If the patient is in renal, cardiac, or liver failure, or if there is severe myelosuppression, chemotherapy must be withheld until the problem is corrected. Relative contraindications refer to the likelihood of unacceptable side-effects, or failure of the treatment to provide benefit. Complications and side-effects are usually related to drug toxicity, and will be more severe if the excretion or inactivation of the agent is impaired. Table 24.3 details the major side-effects and the principal method of elimination of the active agent for the main drugs used in head and neck cancer.

**Table 24.3 Major pharmacokinetic properties and toxic effects of the principal chemotherapeutic agents in head and neck cancer**

<table>
<thead>
<tr>
<th>Agent</th>
<th>Metabolism</th>
<th>Dose-limiting toxicity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cisplatin</td>
<td>Protein bound; hepatic metabolism, renal excretion</td>
<td>Nephrotoxic</td>
</tr>
<tr>
<td>Methotrexate,</td>
<td>95% protein bound; liver and enterohepatic circulation, renal excretion</td>
<td>Nephrotoxic, myelosuppression, mucositis</td>
</tr>
<tr>
<td>5-fluorouracil</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bleomycin,</td>
<td>Conjugation in liver; mainly biliary excretion; some renal excretion</td>
<td>Pulmonary fibrosis, cardiomyopathy, mucositis</td>
</tr>
<tr>
<td>doxorubicin,</td>
<td></td>
<td></td>
</tr>
<tr>
<td>epirubicin</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cyclophosphamide,</td>
<td>Hepatic metabolism, renal excretion</td>
<td>Nephrotoxic, myelosuppression, urothelial damage.</td>
</tr>
<tr>
<td>ifosfamide</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Failure to provide benefit is difficult to predict. The factors known to be associated with prolongation of survival are age, performance status, lymph node status, tumour response and type of agent used (Morton et al, 1985).

If all aspects are considered together with the patient's wishes it is likely that very few patients with advanced stage disease will actually be submitted to any high dose single-agent or complicated multiple-agent regimen (Shaw, 1985).

**Nutritional factors**

Most head and neck cancer patients are noted to be very thin with a considerable amount of muscle wasting when they die. The weight loss may be due to:

1. decreased intake, either from anorexia or as a result of dysphagia;
2. decreased absorption of ingested food;
3. increased loss of nutrients, for example, from fistulae;
4. possible changes in resting metabolism and energy expenditure.
The anorexia can be caused by changes in taste and smell, increased production of lactate and ketones, tumour toxins, psychological factors related to the patient's perception of his disease, the effect of chemotherapy, and various non-specific manifestations of disease.

Tumour production of small peptides and other metabolites is thought to alter host enzyme function, disrupting normal metabolism of the patient. There is an increase in glucose/lactate metabolism (Cori cycle) and body protein turnover, with decreased muscle protein synthesis. Hypoalbuminaemia is a common finding and important because many chemotherapeutic agents are protein bound.

Clearly, then, the patient with advanced cancer has a compromised metabolic and nutritional state and has nutritional requirements that are different from the normal patient. Enteral nutritional support is often required, but parenteral or enteral hyperalimentation to improve the patient's nutritional status, although frequently given (Morton, 1984), is rarely justified in the authors' opinion.

Treatment regimens

Squamous tumours

Methotrexate and cisplatin are both effective as single agents in head and neck cancer, and both can be given in low-dose or high-dose regimens. Ifosfamide and 5-fluorouracil are two other agents which have recently been thought to be effective in combination with cisplatin or methotrexate. There is no evidence, however, that multiple agents confer any additional benefit over the use of the single most effective agent from the combination (see below). Despite this, multiple-agent regimens continue to be popular, the most frequently used being the kinetically based regimen of vincristine, bleomycin and methotrexate (VBM).

Methotrexate

Methotrexate for palliation is usually given every 2-3 weeks as an intravenous bolus of 40 mg/m² of body surface area. It can also be given orally as an outpatient regimen, for example, 50 mg/week.

5-Fluorouracil

Given intravenously, doses vary from 500 mg/week to 1000 mg/m² per 24 hours for 4 days every 4 weeks or so.

Ifosfamide

This agent is given at doses ranging from 1-2 g/m² per 24 hours intravenously over 3-5 days. It is always given with mesna 1-2 g/m² per 24 hours intravenously to protect the urothelium.
Cisplatin

Dose schedules for cisplatin vary between 20 mg/m² per 24 hours intravenously for 4-5 days and 120 mg/m² intravenously over about 8 hours. There are usually 4 weeks between treatments. Any regimen containing cisplatin should include appropriate pre- and posthydration, a high salt load (0.9% saline) and antiemetic medication.

Other

The combination of vincristine-doxorubicin (Adriamycin)-cyclophosphamide (VAC) has been used with success in two patients with recurrent nasopharyngeal carcinoma following radical radiotherapy (Haines et al, 1985). Both tumours were reportedly lymphoepitheliomata (not confirmed with electron microscopy or Epstein-Barr virus antibody titres). If this experience can be repeated by others it will be the first squamous carcinoma of the head and neck to be controlled by chemotherapy when palliation only would normally be given.

Other epithelial tumours

Malignant salivary tumours represent only 5% of all head and neck malignancies but chemotherapy has been used for advanced and recurrent disease. With both single agents and combination regimens tumour responses in up to 24% of patients are recorded, with a median duration of response of up to 6 months. Many of these tumours are slow growing and occur in patients whose general condition is often better than that found with squamous carcinoma. Chemotherapy could therefore be better tolerated.

Occasional responses have been recorded usually to 5-fluorouracil and doxorubicin (Alberts et al, 1981; Creagan et al, 1983; Robustelli et al, 1984).

Mucosal melanoma in the head and neck is rare, most often seen in the oral cavity and nasal fossa, representing 0.2-8% of all malignant melanomata. Primary treatment usually involves radical surgery, but chemotherapy has been used for recurrent or advanced tumours. The most effective drug seems to be dacarbazine, with a response rate of 20%. Bleomycin, vincristine, lomustine, cisplatin and dactinomycin have also been used (Berthelson et al, 1984), but most patients die within one or 2 years despite treatment.

Results of palliative chemotherapy

Survival

A controlled, randomized trial has shown that cisplatin does indeed prolong survival in patients with advanced or recurrent squamous carcinomata (Morton et al, 1985), the prolongation of median survival being 3 months. Another prospective, randomized study of survival in a similar group of patients has shown that there is no difference between methotrexate and cisplatin when used as single agents (Hong et al, 1983). It has also been shown in randomized trials (Drelichman, Cummings and Al-sarraf, 1983; Morton et al, 1985) that the addition of other agents does not prolong survival further.
**Tumour response**

Survival is prolonged in patients in whom a partial or complete tumour response is observed. This is to be expected; however, tumour response is a difficult parameter to assess accurately in anything less than a complete remission (McElwain, 1979; Watson, 1981). A 'partial' response is generally defined as the reduction in tumour size by 50% or more in the perpendicular bidimensional diameters of all observed lesions (Watson, 1981).

Anyone who has diligently tried to apply this definition will appreciate how difficult it is to assess response accurately in many patients.

If there is a complete tumour response, further treatment (surgery or radiotherapy) may control the disease. The overall chance of control of tumour in patients considered for palliative chemotherapy is probably about 1%.

**Side-effects**

The recording of side-effects is important in assessing overall benefit to the patient. The nature and degree of side-effects and toxicity vary with the agent used and the dosage given (see Table 24.3). Toxicity may be acute/subacute (observed during therapy or within 1-2 weeks after treatment) or chronic. Chronic toxicity is usually less severe and develops in a particular organ site or system.

The acute symptoms most disturbing to patients are severe nausea and vomiting (common with cisplatin) and alopecia (most common after bleomycin). Major dose-limiting toxicities are nephrotoxicity (cisplatin, methotrexate), myelosuppression (methotrexate, 5-fluorouracil), mucositis (methotrexate) and pulmonary fibrosis (bleomycin). Patients who experience severe symptoms are unlikely to submit to further courses of chemotherapy.

**Palliation**

The success of the chemotherapy in palliating the patient will depend not only on the effects on the tumour bulk and patient survival, but also on the physical and psychological well-being of the patient. The physical status can be maintained by careful and judicious use of medication and thoughtful dietary management. The psychological condition of the patient will depend to a large extent on the attitude and assistance of close relatives and all the hospital staff that the patient encounters. This amounts to a powerful placebo effect, the benefit of which must not be ignored, whether or not chemotherapy is given.

**Summary**

Patients with terminal head and neck cancer usually have a depressed psyche with an impaired quality of life and a poor general condition. They require special nutritional support and may not tolerate chemotherapy well. It is not known if response and prolongation of life reflect benefit to the patient. Palliative chemotherapy is expensive and toxic, and the possible benefits are limited. Cisplatin or methotrexate are the most effective agents, and the addition of other drugs does not carry any additional clinical advantage. Much care and thought is required before a patient is offered palliative chemotherapy.
The care of the dying

T. S. West

Terminal care for patients with malignant disease will need as much careful planning and coordination as did the earlier attempts to cure. It should be based on the tripod of symptom control, good communication and family support - factors of particular relevance to patient suffering from advanced cancer of the head and neck. For these patients uncontrolled pain and difficulties in feeding, breathing and speaking (Shedd, Shedd and Shedd, 1980), leading to isolation, are the major factors that have to be faced if any quality of life is to be maintained (Moore, 1978).

Symptom control

Pain

Although only about two-thirds of patients with far-advanced cancer experience significant pain in the weeks or months prior to death (Twycross and Lack, 1983), the fear of pain is a major problem. This element of fear enhances many other symptoms, particularly when it is reinforced by unrelieved chronic pain. The management of chronic pain differs in some important ways from pain control in acute situations (Table 24.4).

<table>
<thead>
<tr>
<th>Acute</th>
<th>Chronic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aim</td>
<td>Pain relief</td>
</tr>
<tr>
<td>Sedation</td>
<td>Often desirable</td>
</tr>
<tr>
<td>Desired duration of effect</td>
<td>2-4 hours</td>
</tr>
<tr>
<td>Timing</td>
<td>As required (on demand)</td>
</tr>
<tr>
<td>Dose</td>
<td>Usually standard</td>
</tr>
<tr>
<td>Route</td>
<td>Injection</td>
</tr>
<tr>
<td>Adjuvant medication</td>
<td>Uncommon</td>
</tr>
</tbody>
</table>

The successful use of analgesics in controlling the chronic pain of terminal disease depends on an appropriate drug being given in the correct dose at regular intervals.

Appropriate drug

Mild analgesics, such as soluble aspirin or paracetamol, may be useful in the early stages, but there is no advantage in withholding the narcotics when they are needed. Morphine made up in a simple mixture is most commonly prescribed. If mild analgesia is needed 5-10 mg of morphine mixture 4-hourly may be sufficient. This can be increased as needed to gain control of the pain. More than 90 mg/4-hourly is hardly ever called for. Slow release morphine prescribed 12-hourly and giving the same total dose in the 24 hours as the morphine mixture can be useful. If swallowing is a problem, phenazocine sublingually or oxycodone suppositories in equivalent doses are useful alternatives (Table 24.5). There is no place for intravenous bolus administration.
Table 24.5 Useful strong analgesics and their equivalents

<table>
<thead>
<tr>
<th>Name</th>
<th>Dose interval (h)</th>
<th>Tablet dose (mg)</th>
<th>Morphine equivalent (mg)</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diamorphine</td>
<td>4</td>
<td>10</td>
<td>10/15</td>
<td>Identical to morphine</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(but elixir usually) More soluble</td>
</tr>
<tr>
<td>Phenazocine</td>
<td>8</td>
<td>5</td>
<td>20</td>
<td>Useful, alternative to morphine</td>
</tr>
<tr>
<td>(Narphen)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dextromoramid</td>
<td>2</td>
<td>5</td>
<td>15 (peak effect) (and 10)</td>
<td>Too short acting for regular use</td>
</tr>
<tr>
<td>(Palfium)</td>
<td></td>
<td></td>
<td></td>
<td>Good for 'breakthrough' pain</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Can be used sublingually</td>
</tr>
<tr>
<td>Oxycodone</td>
<td>8</td>
<td>30</td>
<td>15</td>
<td>Useful when swallowing is a problem</td>
</tr>
<tr>
<td>(Proladone)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Dosage

The correct dose of an analgesic is the lowest dose which controls the physical component of a patient's pain. Assessment of this dose depends on estimating a patient's need and then observing the effect of the chosen dose. Observations by ward staff and the patient's family are relevant and will also include reports on the patient's morale and mental state. But pain in cancer may be:

1. caused by the cancer itself;
2. caused by treatment;
3. associated with debilitating disease;
4. unrelated to either the disease or treatment.

It is important to determine the underlying mechanism of pain(s) in far-advanced cancer as the correct choice of analgesic, adjuvant medication or non-medical treatment is often dependent on the cause.

For example, bone pain will respond to a non-steroidal anti-inflammatory drug or to radiotherapy better than to an increase in narcotic medication; pain due to muscle spasm may be relieved by diazepam or physiotherapy; nerve compression pain may need the addition of a corticosteroid to the narcotic analgesic while superficial dysaesthetic pain sometimes responds to amitriptyline but intermittent stabbing pain may be alleviated by carbamazepine (Saunders, 1984).

Interval

Although extra medication should always be available, there is no place for 'on demand' prescribing in the control of the chronic pain. Drugs should be prescribed and given regularly and intervals between doses should depend on the half-life of the particular drug.
**Dysphagia**

Difficulties in swallowing and the fear of choking are common and very distressing to both patient and family; the wish to eat is almost equalled by the need to feed. A patient's swallowing should be observed in an attempt to pinpoint where the mechanism is failing. The consistency of solid or fluid that the patient finds easiest and most difficult to swallow should be noted and a suitable diet then imaginatively designed. Local causes of dysphagia, in particular candida infection, should be looked for and treated.

The best position for the patient (sitting up or semi-recumbent) and for his head and neck during meal times should be discovered. Gastric reflux can be a problem and may be controlled by raising the head of the bed and by prescribing alginates and antacids such as Gaviscon or Asilone. Excessive salivation may respond to drugs that primarily produce a dry mouth, for example, atropine 0.3 mg three times daily or sublingual hyoscine, or do so as a secondary effect, for example, the phenothiazines or, if indicated, an antidepressant.

Occasionally, when the dysphagia is due to interference with the neuromuscular mechanism of swallowing caused by inflammation and oedema splinting muscles and/or interrupting nerve pathways, the use of steroids (dexamethasone 8 mg daily) may produce a most gratifying improvement in the condition (Carter, Pittam and Tanner, 1982) and perhaps, surprisingly, does not cause an unacceptable increase in appetite. A 5-day trial, carefully assessed, should be considered and, if unsuccessful, can be stopped with no side-effects. Steroids may also help in reducing the oedema surrounding an oesophageal block due to tumour.

The basic problem of nutrition usually remains. The feelings of hunger and thirst are both diminished by regular narcotics - a useful side-effect. Meticulous mouth-care and the use of ice cubes, flavoured for example with whiskey or mango juice, alleviate the dryness of mouth that is often the patient's real complaint. Vomiting and constipation must and can be controlled (Butcher, 1983).

As the disease progresses and the patient becomes inevitably weaker and more cachectic, the doctor must be ready to discuss the problems of nutrition with the patient, his family and the team who are caring for him. The pros and cons of instituting tube or parenteral feeding or of performing a gastrosomy during the terminal period may need to be repeatedly reassessed. A cervical pharyngostomy is probably a more acceptable procedure. Almost always the decision will be correctly reached that at this stage such a manoeuvre is not inappropriate and that quality of life will be best maintained by the attention to the person rather than to his loss of weight.

**Dyspnoea**

If the patient has a tracheostomy this will, of course, need proper care often by the patient himself. If a patient is short of breath the cause must be found. Difficulty in breathing and the feeling of breathlessness need to be distinguished. The fear of suffocating can lead to sensation of severe breathlessness. The use of correctly prescribed regular morphine, which does not depress the respiratory centre (Walsh, 1984), and/or diazepam, together with the
sense of security provided by a competent team will take away some of the fears that produce the feeling of breathlessness.

Glucocorticosteroids may help to relieve bronchospasm and may also reduce tumour oedema compressing trachea or bronchus. Excess fluid in the lungs or throat can be dried up by hyoscine given by injection or sublingually. If the dyspnoea is exacerbated by infection, this can be treated if the patient's general condition makes this still appropriate. However, for any asymptomatic pneumonia at the very end of life, antibiotics may well not be the treatment of choice. Appropriate discussion with the care givers, the family and sometimes with the patient himself then allow decisions to be made to give the patient regular morphine to take away the feeling of breathlessness and regular hyoscine to dry up the secretions. Suction and oxygen should be immediately available but they are both frightening and antisocial and, if the patient's medication and sense of security are good enough, it is remarkable how seldom that are needed.

Other symptoms

Fungating lesions

Even in the terminal stage, fungating lesions may respond well to one or two fractions of radiotherapy. The site, rapidity of growth and tendency to bleed need to be noted so that the radiotherapist can give an informed opinion on the possible efficacy of treatment. Fungating lesions may become infected and may smell.

Sinus formation

This can occur and may be distressing and difficult to manage. An attempt should be made to distinguish a sinus between the mouth and the skin and one between the pharynx or oesophagus and the skin. Discharge may then be reduced by lessening the production of saliva or by adjusting the consistency of the diet. A barrier cream may help to prevent excoriation of the skin. Sinuses may become infected.

Infection

Infections are likely to occur. If the infection is producing increased oedema it may be worth trying to control this with broad-spectrum antibiotics and perhaps corticosteroids.

Smell

Smells can be difficult to control. If the smell is due to infection by an anaerobic organism, metronidazole is indicated. It can be given by suppository. Frequent wound toilet is necessary and an air purifier can help.

Toilet

Meticulous wound toilet and mouth-care are fundamental for patients with infected lesions with or without fungation, sinus formation or smell. The frequency and skill with which lesions are cleaned and dressed are more important than what is used. Toothpaste on
a tooth-brush (unless the mouth is painful) can be the most effective way of keeping the mouth clean. Thrush should be expected to occur and must be controlled. If the dressing is painful dextromoramide (Palfium) is an appropriate short-acting analgesic to give half an hour before the dressing. It can be absorbed sublingually.

**Constipation**

Constipation should be prevented. If a patient is on regular narcotics he should also be on a regular aperient but suppositories, enemas and even manual removals may still be necessary. Impaction of faeces must *never* be allowed to occur.

**Pressure areas**

Pressure areas in a dying and cachectic patient should be prevented by good nursing care which demands adequate staffing. Management of bowel and bladder problems help to prevent pressure sores.

*Psychosocial factors in symptom control*

Patients with cancer of the head and neck often have a history of alcoholism, heavy smoking, family conflicts, unstable work histories and a lack of close interpersonal relationships (Cote, 1978). Such people are not easy to be in sympathy with and now that they are faced with a particularly frightening disease, their family and the professional team may have to make a particular effort to overcome barriers of resentment and even of revulsion.

Although no drugs can take the place of staff and family support, people who need comparatively large regular doses of analgesia for their pain and physical distress may also need regular medication for their very understandable mental anxiety and anguish. When morphine is first begun, a weak phenothiazine such as prochlorperazine should also be prescribed to prevent nausea or vomiting. Although this can often be stopped after a few days, in patients with cancer of the head and neck it may need to be continued, and if the patient is under increasing mental stress can be changed for a stronger drug (chlorpromazine, methotrimeprazine (Nozinan)).

The use of the narcotics for the physical components and of the phenothiazines for the mental components of pain is a balancing act and they need to be kept in some sort of proportion to each other. When the correct dose of each has been ascertained, the syringe driver can be used successfully if the patient cannot swallow or is vomiting. Antiemetic and anticholinergic drugs can also be given concurrently. Drugs commonly used in the syringe driver as St Cristopher's Hospice, London, include:

1. diamorphine;
2. methotrimeprazine;
3. hyoscine;
4. cyclizine;
5. haloperidol (Oliver, 1985).
Clinical depression is surprisingly uncommon, but if it is properly diagnosed, an antidepressant drug is useful and will certainly give a good night’s sleep as well as dry up secretions. There is also increasing evidence that the tricyclic antidepressants can produce an analgesic effect (Walsh, 1986), particularly when nerves are invaded.

**Communication**

*The professional team*

Inability to communicate is frustrating when one is with friends, but it is frightening when one is among strangers. Patients with progressive cancer of the head and neck should ideally be cared for by the team they first met before either the diagnosis of malignancy was made or radical surgery embarked on. Continuity of care establishes a mutual trust that can overcome any breakdown in communication and should be planned for from the outset of the disease.

However, staff who have already successfully cared for people with communication problems can approach a new patient with a degree of unembarrassed confidence that may produce immediate improvements in the situation. The patient relaxes and the ability to communicate improves. If the patient is disfigured or the lesion smells, staff have to train themselves to look past these barriers to the real person they hope to reach. By taking the patient's hand, asking permission to sit on his bed and then, while talking, establishing eye contact with him important statements concerning friendliness, good manners and the willingness to face facts are made.

Calling in a speech therapist and the introduction of mechanical aids to communication may be considered even at this late stage. If either prove to be of no help yet another failure has to be coped with.

Throughout the course of the disease the doctor should be ready to answer questions and also be available for discussion (Miller, 1977). The statement, 'My patients never ask me ...' is properly countered with 'I wonder why not ...?'

Questions may arise for no obvious reason, but there are times of crisis, like the first evidence of disease, confirmation of diagnosis, the appearance of secondary spread, the stopping of active treatment and sometimes the death of a fellow patient, when the patient (and his family) may want to talk but may need particular help in initiating the conversation. Sometimes the patient will confide in another member of staff that he is worried about some aspect of his condition. This can be a way of letting the doctor know that more is wanted.

To the question 'Is it cancer, Doctor? Am I going to get better?' it is important not to lie if for no other reason than this lie cannot be sustained ('You don't go to radiotherapy ten times without knowing what's going on'). But it is also important not to give either the stark truth or to confuse the answer in medical jargon. When such a hard question is asked, it is proper to return the ball to the patient's court and allow him the chance to play it in his own style. 'Have you been suspecting that?' allows time for the patient and doctor to look at each other. The way he responds will help to set the pace of the dialogue.
The doctor should only answer one question at a time, watching to see the effect of his reply and checking that his words have been understood. It is wise to find out what the patient understands by the word 'cancer' particularly if other words such as 'tumour', 'growth' or 'abscess' are being used.

When one point has been properly considered the patient should be encouraged to restate the next question: 'You asked if you were going to get better ...?' The response to such a seemingly tentative probe will help the doctor to decide how far he should now move into discussing available therapy and the possible advantages and consequences. With each new piece of information given the doctor must pause, decide if the patient has understood and again allow him either to call a halt or to ask for further information.

If the spouse (or other important person) is not present at such an interview this should be noted and arrangements put in hand for them to be seen. (Failure to keep the appointment will suggest a family in need of help.) In any case it is helpful to make an appointment to see the patient again in a few days' time in order to answer questions that may have come up after the first shock has worn off.

It can also be helpful to have one of the nursing team present. She will help monitor the interview and can check afterwards that there has not been any serious misunderstanding.

The patient's general practitioner can usefully be informed of the situation. Although they themselves are often difficult to contact, general practitioners usually have available and intelligent secretaries.

The patient

When talking to people about cancer it should be remembered that they have four major fears:

1. choking;
2. suffocating;
3. running out of pain-killers;
4. isolation.

If these fears are verbalized, reassurance that the patient will not be left to choke or suffocate, that the pain-killers will not run out and that, whatever happens, the patient will not be abandoned can be honestly given.

There remains the hard question 'Will I die?'. The plain answer is, of course, 'Yes'. But this is a time to take a broader view than just one of life or death. Dying and the fears that go with it may need to be discussed but the matter of living, as fully as possible, should also be on the agenda. This is not a time to take away all hope but rather a time for unfinished business to be completed. Often there is a goal to be aimed for - an anniversary or a reconciliation - which can introduce a positive element into an almost impossible situation.
Sometimes this is also the time for the doctor either to share his own belief that death is not the end or else to try to discover if the patient would like an appropriate minister of religion contacted.

The family

It may have been decided to see the family separately. Often it is right to do so. But statements such as 'Don't tell him doctor, he'd give up if he knew', then have to be dealt with. Such a request has to be taken seriously, but it has to be carefully explained that the patient is not a child (in fact children are often more realistic than adults) and has a right to know more about his own condition; that he will not be told more than he wants and needs to know; that if the doctor does find himself telling the patient more he will not let down the family (and their careful lying) in the process. It can also be stated that, if the matter is properly timed, people do not often give up; rather on learning of mortality they have the opportunity to sum up all that life has been and has meant to them (Saunders, 1984).

When the family (or, indeed the patient) ask 'How long?', no figure should be given in the answer. 'Months rather than years' or 'weeks rather than months' or even 'days rather than weeks' give some indication of the time-scale that the family must work on. But 'the doctors said he had six months' is unhelpful - even if this is what the family heard rather than what the doctor actually said. If the patient dies before the appointed date the family may be unprepared; if he survives after it both he and they have to face a difficult 'in-limbo' situation when the final curtain has failed to come down.

When people ask 'How long?' often the real question is 'How? - what is the manner of death?' Few people have seen a person die and their image is of a dreadful media-type death. Reassurance that death is almost always an increase in weakness and sleepiness, leading to a failure of all the bodily mechanisms; that pain, choking and suffocating, if they occur, can and will be instantly coped with and that if the family cannot be with the patient themselves he will not be left alone, will help. If they appear exceptionally anxious it is worth remembering to ask if anyone else in the family died of cancer - and if so that the death was like.

At such a meeting with the family, children should not be excluded. Children are realists and in practice it is often the child who asks the question that the adults do not dare to voice. After such a meeting, in order to avoid family secrets further isolating the patient, it is important to discuss with the family how much they are going to share with him.

Support of the family

Efforts to improve communication between the patient, the professional team and the family will inevitably lead to involvement with the family. It is likely that they will need support for they, as well as the patient, need to make drastic psychosocial adjustments.

Communication

It is important to obtain a family profile, to discover something of the strengths and weaknesses that a family is bringing to this current crisis and to learn something of their
previous character and behaviour patterns (Earnshaw-Smith, 1982). Drawing a family tree and noting in particular past losses and how the family have coped with them point up those who are in most need of help and those who may provide it.

If family members have never communicated well, it is unlikely that things will change radically at this stage. But if communication has broken down, the presence of an unbiased outsider can act as a catalyst. It is a privilege to sit in with a family, one of whom is soon to die, and by little more than one's presence to help them look each other in the eye, touch each other and laugh as well as cry.

**Family meetings**

Getting the family together should have been attempted at the onset of the disease. Often this is not achieved and when there is no family the team will find themselves providing this aspect of care.

When a family can be assembled to meet the medical, nursing and social work team they will feel supported rather than threatened as it becomes obvious that the two sides have a common objective and are working out how to form themselves into one united team. Such an exercise also avoids the problems that arise when family members are given information separately.

Among matters that may need to be brought out into the open and discussed are guilt from the past and fears for the future, the difficulties that a change in body image can produce in the patient and in the family around him, anger that this outrage should have been allowed 'by God'. At this point it may be helpful to ask the family if they would like to discuss matters with the chaplain.

The need to feed and the need to let go, and - if it has not already been decided - where the patient should die may also need to be talked through.

The right place for a patient to die will depend on his wishes, the likely mode of death, the wishes and availability of the family and the local resources, whether they be based on the home, the hospital or a hospice.

If the decision is made for the patient to die at home it must be made clear who is in charge. The general practitioner should be the one to organize local services, including nursing services like those provided by Macmillan teams or, at night, the Marie Curie nurses. Should the home situation break down, re-admission must be immediate and easy. If a local hospice is available and considered appropriate, liaison should be early rather than late in the planning.

Whether hospital, home or hospice is decided on at the place where the patient (and family) will be most appropriately cared for at each stage, it must be clear who is responsible. This can only happen if there is good communication between all the services available (Saunders, 1984).
There can be no absolute answers to these questions and again uncertainty remains perhaps the hardest factor of all for the cancer patient and his family (as well as the professional team) to contend with.

By looking at these matters together, some correct decisions will be arrived at and a climate of truth can be established in which the family members have a chance to say 'sorry', to say 'thank you' and to say 'goodbye'. Such expressions of feeling made verbally or non-verbally, are important for the dying patient; they are even more important for the family that have to continue to live.

The management of the final phase

It may be as hard for the professional team as it was for the patient and family to accept that the time has come for no further aggressive therapy. While there are still plans for good symptom control, for improving communication and for mobilizing family support, the team are not stating that nothing more can be done.

The time will come, unless a catastrophic incident intervenes, when the patient can be said to be dying. Recognizing this time is a matter of experience as well as observation. Often the nursing staff are far more realistic than their medical colleagues and it is they who will collect and prepare the family as well as know what comfort and even suggest what medication the patient most needs.

At this stage most patients need their medication by injection. It can be important to explain to relatives (and sometimes staff) that the medicine now being given by injection is an equivalent dose to that given by mouth, and that the deterioration in the patient's condition is due to the disease process and not to the drugs.

This is also the time to review all the patient's medication. Steroids, diuretics and antibiotics are examples of drugs that will probably no longer be appropriate, as the deteriorations they were prescribed to prevent are now inevitably occurring.

The main symptoms to distress a patient dying with cancer of the head and neck are:

(1) pain;
(2) breathlessness;
(3) restlessness;
(4) accumulation of secretions.

All these are problems that the patient and those around him have been facing for some time.

Although there is never any point in withholding necessary narcotics, pain does not usually increase markedly over the last 24 hours. If the patient's symptoms had already been reasonably well controlled, a simple increase of the analgesic and continuing it regularly will not only bring the pain under control again, but will also help to cope with any increase in breathlessness or restlessness.
Increasing the dose of phenothiazine (or changing, for example, from chlorpromazine to methotrimeprazine) will help combat mental distress, while diazepam can be useful if physical restlessness is disturbing the patient and those sitting with him. Accumulation of secretions and the 'death rattle' can often be controlled by an injection of hyoscine 0.4-0.6 mg. Hyoscine, when combined with a narcotic and a phenothiazine, is also a most effective sedative and tranquilizer.

If a haemorrhage is feared, diamorphine and a phenothiazine in a slightly higher dose, together with hyoscine, should be prescribed in anticipation and be immediately available.

Terminal pneumonia is usually a peaceful and almost natural way to die. Distressing symptoms like copious phlegm may be treated with hyoscine. Sputum that is difficult to expectorate may respond to a mucolytic. A distressing pleurisy should be treated with an increase in analgesic medication. Carefully monitored physiotherapy may have a part to play.

Isolation can be the most distressing complication of all. If the family are unable, for whatever reason, to be with the patient during his final hours, every effort should be made for one of the team to sit with the patient, holding the hand and demonstrating that just because he is now dying he has not been abandoned.

**Bereavement**

Family support should have been organized during the terminal illness. By getting to know something of the strengths and needs of individual family members, the professional team will have made some progress in identifying those who will need organized bereavement follow-up.

In hospital or in a hospice, an important occasion for assessing such a need is the day after the death of the patient when the family come back for the certificate, the possessions and to view the body. This visit should be taken most seriously and a member of the team should be available, if the family wish it, to spend time with them, living through the past days or sometimes years, and making some estimate of the family's ability to cope with all that lies ahead.

The patient's general practitioner should always be telephoned by the ward doctor and informed of the patient's death. If it is considered the family are at risk he/she can be alerted.

If bereavement counselling is needed, and trained people are available, this should be organized as a continuation of the work of the original caring team (Yorkstone, 1981).

**Conclusion**

In such ways as those prescribed above, the professional team caring for patients who cannot be cured will find enough job satisfaction to continue their work of skilful and effective care.