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 thank 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 former 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
Preface

We were very pleased to be invited to edit the Rhinology Volume of *Scott-Brown's Otolaryngology* but we realized from the outset that it would be a difficult task as there have been many fundamental changes in rhinology over the eight years since the last edition. The introduction of two new volumes has necessitated further alteration in the format of this book.

Five new chapters appear, covering endoscopy of the nose and sinuses, rhinomanometry, the mechanisms and treatment of allergy, sleep apnoea and snoring, and the orbit. This volume has gained, from other volumes, chapters dealings with radiology and the nasopharynx whilst losing congenital deformities of the nose and foreign bodies which are not to be found in the Paediatric Volume.

Acute sinusitis, chronic sinusitis and vasomotor rhinitis were formerly dealt with in three separate chapters. The assessment, differential diagnosis and medical management of these are now combined in a single chapter - 'Rhinitis, sinusitis and associated chest disease' but a separate chapter has been devoted to their surgical management.

We would like to thank all the contributors to this volume who have worked so hard to ensure that their chapters were comprehensive and up-to-date and have consistently returned manuscripts and proofs on time. It has been a great pleasure to work with such a fine team. We are indebted to Alan Kerr for his helpful comments and encouragement and the staff at Butterworths. We would like to thank Dr Hughes of the Pharmacy Department of the Brompton Hospital, together with Peter Zwarts, Librarian, for all their assistance, in addition to Amanda Laman for her secretarial help and finally our wives and families for their forbearance and support.

I. S. Mackay  
T. R. Bull
Chapter 1: Examination of the nose: conditions of the external nose

T. R. Bull

Examination

The exterior of the nose requires no special apparatus for examination. The nasal fossae, however, are not easy to examine without good lighting, special instruments and, occasionally, the topical use of a vasoconstrictor to shrink the nasal mucosa.

A head mirror or head light is necessary for a thorough examination and leaves both hands free for using instruments: a hand-held light is adequate to demonstrate the nasal vestibule. In children, an auriscope with a wide speculum is a useful additional instrument for examining the nasal fossae. A nasal speculum is necessary in adults to dilate the vestibule and elevate the tip of the nose. In children, however, where the use of instruments is best avoided, simple pressure on the tip of the nose will suffice. A child may require to be held firmly during this examination, if, any instrument is needed, for example for removal of a foreign body.

The Thudicum nasal speculum is in common use - several sizes are needed, with blades of varying lengths. The spring action should be gentle and the blades smooth. If the speculum is inserted into the skin-lined, relatively insensitive vestibule and the blades are allowed to spring apart or if the speculum is opened forcibly, it is an uncomfortable experience.

The blades are opened just sufficiently to give an adequate view of the nasal fossae. To see the upper and lower limits of the nose requires angulation of the beam of light, the patient's head and the speculum. The longer bladed St Clair Thomson or Killian speculum is useful for seeing the middle and posterior thirds of the nasal cavities and for lifting turbinates to expose the inferior and middle meatuses, but it can only be used under local or general anaesthesia.

Fibreoptic endoscopy

Fibreoptic endoscopy enables a considerably more thorough examination of the nasal cavities and postnasal space. Local anaesthetic to the inferior meatus enables the narrow endoscope to be inserted along the floor of the nose, and familiarity with the technique enables such features as the openings of the nasolacrimal duct and maxillary sinus along with a clear view of the postnasal space to be achieved as a routine procedure in out-patients.

Examination of infants

When examining a neonate's or infant's nose, the patency of the nasal airway on quiet breathing is assessed and the presence or absence of nasal discharge is noted. The infant is best examined wrapped, with its arms included, in a shawl or blanket and held flat on its back on a bed or table with a nurse to control head movements.
Common nasal conditions requiring investigation in infancy are discharge and an inadequate airway. Slight discharge is common during the first few days of life as the nose is cleared of intrauterine and birth contaminants. A nasal swab is taken and the nose examined to exclude a collection of infected debris or other sources of infection. Suction with a soft rubber-zipped Zöllner aural sucker is a useful technique for clearing the nasal fossae.

A moderate degree of nasal obstruction may interfere with feeding, and severe or complete obstruction, as seen in bilateral choanal atresia, threatens life due to the infant's inability to learn mouth-breathing. If the baby lies quietly breathing with the mouth shut, then the nasal airway is adequate. If the baby is restless and can only take a few sucks at the nipple or teat without drawing its head back and gasping for breath, then an inadequate nasal airway is probable. If mucus is seen bubbling in the nostril, or if clouding with the breath on a bright surface held beneath the nostrils can be demonstrated, then some airway must be present. Otherwise, the patency of the airway must be tested with a blunt-ended soft rubber or plastic catheter to exclude choanal atresia. It should be demonstrated that the catheter passes through the oropharynx where it will cause gagging and can be seen through the open mouth. Suction can be used on withdrawing the catheter to remove any secretions which may have been the cause of the obstruction. Failure of the catheter to pass into the oropharynx requires X-ray examination which may require general anaesthesia. Lateral soft tissue X-rays must be taken first, followed by similar views taken after introducing a radiopaque solution into each nostril cavity separately while the infant lies on his back. Congenital nasal deformities are usually seen in conjunction with hare-lip and cleft palate and involve the alar cartilages. An abnormal fusion of the nasal processes produces a characteristic external deformity.

**Examination of adults**

The external nose is examined by observation and palpation. When assessing injury to the nasal bones and cartilage, it is important not to miss injury to the adjacent structures particularly the eye and cribriform plate. Many different types of swelling and cysts appear on or near the nose. Dermoids are found at the lines of fusion of the nasal processes, and the philtrum of the lip and they also occur over the nasal bones and columella. A moist sinus near the bony cartilaginous junction on the bridge of the nose is seen in dermoid cysts, with a deep extension upwards between the nasal bones which may extend to the anterior cranial fossa. A furuncle often occurs near the tip arising in hair follicles in the vestibule and presenting as a tender red external swelling. Sebaceous cysts may occur on the nose, while swellings near the alae may be dental in origin - either cysts or abscesses.

Acne rosacea, with its butterfly rash over the nose and cheeks, is a common condition of the nasal skin, but virtually any skin disease can affect the nose. In rhinophyma the lower half of the nose enlarges due to hypertrophy of the skin and proliferation of the sebaceous glands. Skin neoplasms found here include epitheliomata and melanomata. Loss of tissue around the edges of the nares may be due to trauma or to healed lupus. Ulceration and inflammation of the skin surrounding the anterior nares is often secondary to discharge from the nose. Herpes simplex ulcers also may present in this site; they are frequently multiple and in most instances there is a history of previous attacks. Skin lesions restricted to the distribution of the maxillary division of the fifth cranial nerve are characteristic of herpes zoster and this can be complicated by secondary infection.
The shape of the nose alters with age. Loss of elasticity of the soft tissue between the columella and the caudal margin of the septal cartilage and the alar and upper lateral cartilages results in a drooping of the tip of the nose, so that the nose appears longer (Parkes and Kamer, 1973). The variations in the shape of the nose related to racial characteristics are also well recognized.

Palpation of the nasal bones and cartilages differentiates a bony deformity from a cartilaginous or soft-tissue swelling. An external deviation of the nasal bones and cartilage is probably associated with a deviated nasal septum.

Destruction of the bony septum, with subsequent saddle deformity, may result from syphilis and of the cartilaginous skeleton from lupus vulgaris. Enlargement of the bony skeleton may be due to a general bone disease, such as Paget's disease, or to a fibro-osseous dysplasia. Cartilaginous enlargement may be due to a chondroma or chondrosarcoma.

X-rays of the nasal bones will demonstrate recent fractures and show the degree of displacement; X-rays may have a medicolegal significance but are of very limited help in the management of nasal fractures.

**Examination of the vestibule of the nose**

The vestibule is the skin-lined anterior compartment of the nose. Its size and shape vary according to age, and from one person to another, but it is pear-shaped in adults with a narrow slit-like upper angle between the septum and ala. The skin contains hair follicles, sebaceous and sweat glands. The vibrissae become well developed in middle-aged and elderly men. Most of the hairs arise from the lateral and medial walls. The skin lining extends further posteriorly on the lateral wall and this side is more flexible than the medial. Insertion of a speculum causes more discomfort on the medial side where the sensitive mucosal surface is nearer the front of the nose and the wall is relatively rigid. The mucocutaneous junction is identifiable by a change of colour to pink and by the moist appearance of the mucosa, due to its surface film of mucus.

Metaplasia of the columnar mucosa to a squamous cell lining may occur in the anterior part of the nasal passages, especially on the septum. This is usually due to frequent rubbing of these areas by the patient, through habit or to relieve irritation. The metaplasia stands out as whitish dry areas on the surrounding normal mucosa and, where these are continuous with the vestibular skin, the mucocutaneous junction is obliterated. Frequent rubbing of the septum produces even more marked changes, sometimes progressing to ulceration and septal perforation. The anterior end of the septal cartilage may become dislocated out of its groove in the maxillary crest and protrude into one or other vestibule. It causes an obvious, and often unsightly, projection into the vestibular lumen and tends to cause nasal obstruction and be subject to trauma. Various industrial dusts and fumes, notably nickel and chrome, tend to be deposited on the vestibular septum and may cause septal perforation.

A nasal speculum may hide more of the vestibule than it reveals. Examination of this region should begin with inspection from several angles, assisted by pressing on the columella and tip of the nose to open up different areas of the vestibule. A short bladed speculum is
inserted just within the anterior nares and, deliberately, each wall and the floor are inspected. The upper angle and the upper lateral wall are particularly difficult to examine and a small mirror often gives the best view of these parts.

Staphylococcal infection of the hair follicles is common and may produce acute inflammation, or the patient may be a symptom-free carrier of bacteria. Swabbing of the nasal vestibules of hospital staff is a routine procedure when a bacteriologist is trying to trace the origin of a ward infection. A vestibular staphylococcal infection may go unnoticed by the patient while infection is transmitted to other parts of the head and neck, causing such problems as furuncles, conjunctivitis and otitis externa.

While many types of skin condition occur in the vestibule, one of the most common is a papilloma which is usually pedunculated. Malignant change in a papilloma is very rare but carcinomata and melanomata do occasionally present in the nasal vestibule.

**Examination of the nasal fossae**

The nasal fossae in an adult are approximately 7.5 cm long and 5 cm high. The airway through them is tortuous and its shape and size depend upon two factors:

1. the configuration of the skeletal elements - septum, lateral wall and turbinate bones
2. the nasal mucous membrane, which is liable to considerable changes in thickness. These changes are particularly marked over the inferior and middle turbinates and depend upon a variety of exogenous and endogenous factors.

The nasal airway is subject, therefore, to wide variation from day to day in any one person.

The area of the nasal fossae visible on anterior rhinoscopy varies considerably, but the anterior part of the septum and floor are always visible, while the area of the lateral wall that can be seen depends upon the size of the anterior end of the inferior turbinate. This is the most conspicuous feature on first inspection of the nasal fossae and patients may attend hospital for advice on this 'tumour' which they have glimpsed in the mirror. A large inferior turbinate which obstructs a satisfactory view of the middle turbinate and middle meatus can be reduced in size by the application of a vasoconstrictor solution, such as topical adrenaline.

When inspecting the nasal fossae, the following should be evaluated: the airway, the septum, the inferior turbinate and meatus, the middle turbinate and meatus, and the floor of the nose.

An examination which follows this routine avoids overlooking a particular site in the nose.

**The airway**

A complaint of nasal obstruction is very common and the range of individual tolerance to this symptom is wide.
For assessment of the airway, each side of the nose is examined separately, and one anterior naris is occluded without deforming the opposite side. A bright surface held beneath the nostrils to compare the area of misting may also demonstrate and compare the airways. If the obstruction is worse on inspiration, the alae nasi may be seen to collapse on to the septum. Sometimes, a previous submucous resection may cause the septum to 'flip' or impinge on inspiration against the lateral wall of the nose. If the obstruction is mainly on expiration, a 'corking' effect in the posterior choana is a possible cause and may be due either to a large posterior end of an inferior turbinate or to ethmoidal or choanal polyps. Occasionally, adhesions are seen between the septum and turbinates - usually as the result of surgical or other trauma.

The septum

The general configuration of the septum is first related to the external shape of the nose. A septal spur on one side usually means a concavity on the other, but sometimes the septum is thickened and bulges into both nasal passages. This occurs as an acute condition in a septal haematoma or abscess. A chronic thickening of the septum may be due to a duplication of the cartilage or to an organized haematoma. The septum is rarely straight and small deformities are often of no clinical significance. It is also remarkable how a marked deviation of the septum found on a routine examination may be unassociated with any nasal symptoms. They should be noted, however, and a drawing is often a useful note. Examination of the septum should exclude perforation and areas of granulation.

The inferior turbinate and meatus

The inferior is the largest turbinate and it is subject to considerable variation in size, due mainly to changes in its submucosal vascular bed. Hypertrophy of the inferior turbinate may occur when the airway is large: the concave aspect of a deviated septum is usually seen to be opposed by a compensatory enlargement of the inferior turbinate. A wide airway results in drying of the mucous film and crusting. The mucosa is thick in chronic and allergic rhinitis and this is an important cause of nasal obstruction. Hypertrophy of the inferior margins of the middle and inferior turbinates and of their posterior ends is common in these conditions.

The inferior meatus is not usually visible unless the inferior turbinate is lifted upwards and inwards, and this requires anaesthesia. Sometimes the anterior opening is very narrow and low down near the floor of the nose, making the introduction of probes and other instruments difficult. If the inferior meatus is visible, the lateral wall is seen to curve laterally in the anterior third. The nasolacrimal duct enters the meatus just below the attachment of the turbinate in this area, but it is rarely visible except with the nasal endoscope.

The middle turbinate and meatus

The middle turbinate, like the inferior one, is subject to variations in size and shape. A large turbinate often contains an air cell, while oedema, hypertrophy or polypoid change in the mucosa are all common. The shape is also influenced by the size of the airway, and physiological compensatory hypertrophy occurs in wide nasal fossae as with the inferior turbinate.
The middle meatus is the main drainage channel for the sinuses and the most likely place to find evidence of sinus disease. Most nasal polyps first appear in this space. While it is possible to view the area in some noses, it is often necessary to use a vasoconstrictor spray to reduce the inferior and middle turbinates for an adequate view. With the meatus exposed, the hiatus semilunaris and bulla ethmoidalis are seen anteriorly and it is possible to cannulate or probe the ostium of the maxillary antrum more posteriorly.

The floor of the nose

Foreign bodies usually lodge between the inferior turbinate and the septum and may be overlooked by failure to examine directly along the floor of the nose. A swelling in this area may extend from the teeth, the palate or the buccalveolar sulcus, for example a nasoalveolar cyst. An ulcer in the floor of the nose should be probed to exclude a communication with the oral cavity. Posteriorly, an antrochoanal polyp or a hypertrophic end of an inferior turbinate may cause obstruction.

The superior turbinate and meatus and the olfactory mucosa are not seen on routine clinical examination. Any polyp or swelling apparently arising from these areas may communicate with the cranial cavity.

Other examination techniques

Probing

A mucosal anaesthetic is usually needed before probing to demonstrate the consistency, mobility and site of attachment of a lesion. Ulcers are probed to detect if the underlying bone is exposed or if there is a sinus or fistula. The septum can be touched to see if the cartilage or bone is missing or to demonstrate a perforation, by passing the probe through and observing it in the other nasal fossa. The grating sensation felt on probing a rhinolith is characteristic.

X-rays

Lateral and anteroposterior X-rays and tomograms are helpful in demonstrating the nasal fossae. Soft-tissue shadows can be seen outlined against the air space, and the size and situation of radiopaque foreign bodies and rhinoliths can be seen. Because the septal cartilages are radiotranslucent, little information about them, and accurate assessment of the airways, are not obtainable by X-ray examination.

Examination under anaesthesia

Under general or local anaesthesia, the nasal passages can be dilated with a speculum, such as Killian's or St Clair Thomson's, and the nasal fossae can be examined to the choanae. The turbinates can be infractured and the blades of the speculum inserted beneath the inferior and middle turbinates, giving good exposure of the meatus.
Conditions of the external nose

The nose and nasal vestibules are covered with skin and may be involved in many generalized skin diseases, while a few dermatological conditions, such as acne rosacea, are especially liable to affect the nasal skin. The vestibules and surrounding skin are liable to infection because this is the main air inlet and patients frequently rub this area, causing minor abrasions and introducing infection on the finger. The hairs of the nasal vestibule are thick and strong; their follicles are a common site of both acute and chronic infection presenting as furuncles, cellulitis or vestibulitis.

The normal nasal flora

Jacobson and Dick (1941) swabbed the nose in 500 consecutive patients admitted to a medical ward who had no nasal symptoms. They found the common organisms to be Staphylococcus albus, diphtheroid bacilli and, occasionally, in apparently normal noses, they also found Staphylococcus aureus and Micrococcus catarrhalis.

The results of nasal swabs taken from staff at The Royal National Throat Nose and Ear Hospital showed the following results (Rees, 1969, personal communication): out of 158 persons examined, 44 grew coagulase-positive staphylococci, and 38 out of 44 organisms cultured were resistant to penicillin. Laryngectomy cases have nasal swabs taken preoperatively as a routine and, in 1969, out of a total of 25 patients, 16 grew no pathogenic organisms, five grew staphylococci, two grew beta-haemolytic streptococci, one grew pneumococci and one grew Proteus sp.

The nose is, therefore, free of pathogens in the majority of normal people but it should be examined as a source of infection in institutional outbreaks. Coagulase-positive staphylococci are the most common pathogens and may cause acute or chronic infection in the vestibule.

Acute infections

The nasal vestibule is a painful site for a furuncle which is probably the commonest acute infection. The infecting organism is nearly always Staph. pyogenes (aureus). It is important to find out the sensitivities of the particular strain of Staphylococcus sp. If the furuncle is pointing or has discharged, this information is easily obtained, but a swab from the surface of the vestibule probably reveals the type of Staphylococcus. The vestibule of the nose may act as a reservoir of staphylococci from which other parts of the body may become infected; alternatively, staphylococci may be transferred from other areas of the body, perhaps from ears, eyes or perianal region to the nasal vestibule, the finger nails acting as the connecting link.

Nasal furuncles are potentially dangerous because the veins of the nose drain on each side into the facial veins, which, like the ophthalmic veins communicate directly with the facial veins and also communicate with them indirectly through the supraorbital veins. The ophthalmic veins pass by way of the superior orbital fissure to the cavernous sinus. Severe nasal inflammatory processes may therefore extend to the venous sinuses of the brain, causing
thrombophlebitis, a condition which may prove fatal despite antibiotics. Squeezing or incision of nasal furuncles is best avoided unless they are definitely pointing. The first indications of cavernous sinus thrombophlebitis as a complication of a nasal furuncle are those of malaise, headache and pyrexia. There may be superficial tenderness along the facial veins, with lid oedema and chemosis of the conjunctiva. Later, there is proptosis with limited eye movement. These local signs are due to the obstruction of the venous return from the ophthalmic veins.

The great majority of nasal furuncles are minor infections, however, and resolve spontaneously by discharging into the vestibule. A topical antibiotic ointment suffices as treatment for the small furuncle and vestibulitis. A severe furuncle with a general reaction requires systemic antibiotics and, because of the potential dangers, close observation to ensure that the infection is showing an early response to treatment.

Recurrent boils suggest either local trauma, which may be self-induced, or an underlying general condition which reduces the patient's resistance to infection. Having excluded local causes, the patient's blood picture should be investigated and diabetes mellitus must be excluded.

Vestibulitis

Vestibulitis is a condition in which the skin of the nasal vestibule becomes excoriated and infected. An eczema of the vestibular skin gives a similar picture. Repeated trauma to the vestibule when the nose is rubbed or cleaned excessively by the patient is a common cause. The projection of a dislocated columellar portion of the cartilaginous septum into the vestibule is one of the most common predisposing factors in vestibulitis. The skin overlying the projecting cartilage is thin, stretched and easily damaged with minimal trauma to the nose. Persistent infected ulceration develops. Advice to avoid unnecessary trauma, with the application of an ointment such as aureomycin, often suffices as treatment, but removal or correction of the projecting cartilage may be necessary.

Unilateral vestibulitis in a child is invariably diagnostic of a foreign body. The offensive discharge from the mucosal irritation of a foreign body causes a secondary vestibulitis. Vestibulitis affecting both nares is usually due to eczema, which may be localized to the nose or may be part of a generalized tendency to eczema. Purulent rhinorrhoea may also cause vestibulitis. The watery rhinorrhoea of nasal allergy or coryza may also cause an excoriation of the vestibular skin.

Impetigo is a contagious skin infection of the superficial layer of the epidermis caused by a pyogenic staphylococcus and occasionally by a streptococcus. Pustular vesicles form and break, with yellow transparent adherent scabs. Impetigo commonly affects the face and nasal vestibules. The condition usually settles with the application of an ointment such as aureomycin.

Erysipelas

This causes an acute inflammation of the skin and subcutaneous tissues of the nose. The skin is raised and deep red in colour, with pain, heat and vesiculation, often accompanied by headache, fever and malaise. The organism is Streptococcus sp, which usually enters
through a small fissure in the skin. Erysipelas in the region of the medial canthus simulates an acute frontal sinusitis, with eye closure from lid oedema. The diagnosis is clear, however, from the sharp margin of the reddened area of skin, the absence of intranasal symptoms and signs, and the normal sinus X-ray. Penicillin is usually curative.

**Herpes simplex and zoster**

Small vesicles forming near the nostrils or lips are not uncommon with a coryza. They are painful and break down, with a watery discharge, to coalesce and form a larger irregular ulcer. The vesicles tend to recur at the same site and are caused by the herpes simplex virus. This virus may remain latent in the skin until local irritation, possibly combined with a period of lowered resistance, causes the virus to become active and the vesicles to develop.

The nasal skin is affected in zoster of the maxillary division of the trigeminal nerve when vesicles involve the cheek, nose, nasal vestibule, nasal mucosa and palate. In ophthalmic herpes, the eye lesion and vesicles in the supratrochlear and supraorbital areas demand most attention, although vesicles also affect the nose in the distribution of the anterior and posterior ethmoidal and external nasal nerves. Pain may precede the eruption and, in the maxillary or mandibular division, may be mistaken for dental or sinus disease. Once the eruption develops, diagnosis is made by the exact limitation of the outbreak to the nerve distribution, and limitation to the midline is characteristic. Secondary infection of the eruption makes diagnosis more difficult but the distribution remains unaltered. Lesions of the palate and nasal mucosa are rarely seen in the vesicular state, and they appear as discrete superficial ulcers which heal in 3-4 days. Treatment is restricted to the skin, with an ointment such as neomycin and hydrocortisone 1%. Postherpetic neuralgia is rare in these cases.

**Chronic infections**

**Lupus vulgaris**

Lupus vulgaris may cause skin involvement of the nose by the tubercle bacillus and occurs more frequently in women than in men. The skin invasion usually begins between the ages of 2 and 15 and occurs in someone who has already been infected by tuberculosis. Reddish-brown papules first appear on the nose, which when pressed with a glass slide, become white, and 'apple-jelly' nodules appear as small brown semi-transparent spots. Borrie (1975) describes two types of lupus involving the nose:

1. A slowly progressive usually non-ulcerative tuberculous infection of the skin. The disease is probably borne by the fingers to the nose and the bacillus enters the deep layers of the skin from a finger scratch. The condition is characterized by miliary tubercles forming lupus nodules in the dermis.

2. An ulcerative type of infection of the skin, which may spread rapidly, and which is nearly always secondarily infected by staphylococci.

In the slowly progressive type of lupus the papules become soft, coalesce, and break down, forming shallow ulcers with undermined edges. Severe scarring and nasal deformity result with nasal obstruction of varying degree caused by contraction of the scar tissue at the
vestibule. A chest X-ray is necessary and evidence of tuberculosis elsewhere in the body is to be excluded. Lupus, before adequate chemotherapy, resulted in extreme nasal deformity with virtually complete destruction of the external nose and closure of the nares. Lupus is now rare, presenting with less conspicuous lesions, and early treatment has eliminated such gross sequelae.

Syphilis

Primary lesions of syphilis may present on the nose and *Treponema pallidum* is demonstrated in the exudate from the ulcer. The primary sore appears 9-90 days after exposure to infection and, if untreated, heals slowly in 3-10 weeks, leaving a thin atrophic scar.

Tertiary syphilis of the nose has the usual characteristic of the gumma. A hard, painless nodule breaks down to leave a deep ulcer with a typical punched out margins and a 'wash leather' base. A gumma heals with scarring and destruction of tissue. A painless chronic inflammatory nodule requires the exclusion of syphilis as a diagnosis.

The nasal vestibule may become involved in congenital syphilis, giving rise to 'snuffles', and late congenital syphilis produces the classic saddle nose with occasional more severe deformity. Gummatous lesions which destroy the nasal septum are in part responsible for the external deformity of the nose and cause large crusted septal perforations.

Other skin conditions

Acne rosacea

Enlarged superficial blood vessels in the skin of the nose and cheeks cause the dusky red colour and shiny surface characteristic of acne rosacea. There is usually secondary hypertrophy of sebaceous glands, and acneiform lesions may be superimposed. The disease is seen more commonly in women at the menopause. In some cases, usually males, there is enormous hypertrophy of the sebaceous glands, leading to the condition of rhinophyma. With gross deformity, surgical excision of the excess skin is necessary, avoiding damage to the underlying cartilage.

Lupus erythematosus

The skin lesion affects the nose and cheeks with a symmetrical butterfly distribution. There are patches of erythema and scaling which slowly become thin atrophic scars. In these areas, there is a stippling caused by filling of the orifices of the sweat glands and hair follicles with horny plugs. The condition is differentiated from lupus vulgaris by the absence of apple-jelly nodules and ulceration, and the presence of stippling. In about 5% of cases, the disease become systemic, with malaise, multiple arthritis and kidney lesions in addition to the skin changes. In these cases, blood examination will show a raised erythrocyte sedimentation rate and gamma-globulins, leucopenia and the presence of typical lupus erythematosus cells - rosettes of leucocytes around nucleoprotein. Sarcoidosis, leprosy, scleroderma, yaws and rhinosporidiosis affecting the nose are described in Chapter 8.
Neoplasms of the skin of the nose and vestibules

Papillomata

Papillomata are common and may require excision on account of their appearance or irritation to the patient.

Rodent ulcers (basal-cell carcinomata)

These are common in the skin covering the nose, especially around the alae nasi where they may be inconspicuous. A small semi-transparent pearly nodule first appears which ulcerates and fails to heal. The ulcer slowly enlarges (so the history is usually long) and causes local destruction of skin, cartilage and bone but either without metastasis to nodes or systemically. The diagnosis should be suspected in any long-standing ulcer on the nose and, if the diagnosis is in doubt, the lesion is biopsied. Radiotherapy or excision is curative in an early lesion.

Squamous-cell carcinoma

This may occur on the skin of the external nose or vestibule. The history is short, unlike that of rodent ulcer. A carcinoma on the external nose usually has an ulcerated centre with rolled everted edges.

The prognosis, with treatment for a small early lesion, before there is lymphatic metastatic spread or involvement of underlying bone and cartilage, is good. Some early neoplasms respond to radiotherapy while more advanced lesions require wide excision with radical neck dissection for cervical metastases. A painless lesion in the vestibule or on the septum is often ignored by the patient or missed by the doctor in the early stages. The prognosis with nasal carcinomata is therefore frequently not good and early metastases in the cervical nodes are seen.

Injury to the nose

Nasal fractures, with or without skin laceration, are common injuries. An external cosmetic deformity, and injury to the septum causing nasal obstruction, are the main relevant aspects of nasal fractures.

Fractures of the nasal bones, like fractures elsewhere, can be simple, with or without displacement, or compound. The fractures may be associated with both skin and mucous membrane laceration, causing epistaxis as well as external bleeding. Nasal bone fractures with associated fracture of the maxillae may involve the nasal sinuses, which fill with blood. It is important to exclude associated fractures of the skull particularly those of the anterior cranial fossa or orbital margin in a case of nasal fracture. The nasal septum is frequently displaced when the nasal bones are fractured, and a septal haematoma may also form, lifting the perichondrium on one or both sides and causing marked nasal obstruction. Pain suggests secondary infection of the haematoma. Management is discussed in Chapter 13.
Chapter 2: Radiology of the nose and paranasal sinuses

P. D. Phelps

Sinuses

Although computerized tomographic scanning is now the best means of imaging the sinuses, cost and rationing of scan time in the UK necessitate a continuing reliance on plain film views. Standard radiographic sinus projections are the commonest imaging examination of the ear, nose and throat, undertaken to confirm or exclude sinusitis in patients with nasal symptoms or to show evidence of neoplasia. Computerized tomography is reserved for problem cases or those where neoplasia is found or suspected.

Plain radiography

Views for the nose

Radiological examination of the nasal bones can best be made by placing a dental film in direct contact with the side of the nose and centring the incident beam horizontally through the nose. This lateral view may be supplemented by a cranio-caudal projection obtained by inserting an occlusal film between the teeth and directing the vertical beam through the nasal bones onto the film. These views are usually requested following facial trauma, to show fractures of the nasal bones as well as lateral shifts and displacements.

Standard sinus views

A specialized skull unit should be used whenever possible to allow accurate positioning and unvarying focus film distances with a moving grid. Pathological processes affecting the paranasal sinuses encroach on the air in the sinuses and are seen on the radiographs as alterations in the translucency of the sinus. Examination in the erect position is desirable to reveal fluid levels which may be present. It is possible to perform erect examinations on modern isocentric skull units, but without the advantages of fixed reference planes (see Volume 1, Chapter 17).

Asymmetry of the paired sinuses will usually result in the smaller sinus appearing more opaque because of its thicker bony walls. This should not be mistaken for a pathological state. It is not uncommon for one frontal sinus to be much smaller than its fellow or even to be absent. Antra differ in size less often, though small differences are not uncommon. Rarely one maxillary antrum fails to develop and in consequence the maxilla looks dense on plain X-ray films. The smaller size of the affected antrum should alert the observer to this possibility.

The following projections allow a good all-round assessment of the paranasal sinuses: the occipitomental, the occipitofrontal, and either or both the lateral and the overshot axial views.
**Occipitomental (or Waters') view**

The patient sits facing the Bucky support with the chin resting against it, the median sagittal plane aligned to the midline. The mouth is supported wide open with a transradiant perspex 'bite block' and the baseline is adjusted to make an angle of 45° with the film. In older patients it may be necessary to angle the central ray of the tube caudally to compensate for an inability to extend the head sufficiently, but if a skull table is used with the object table angled through 20° (or more) forwards towards the patient, the correct position will be consistently maintained, and will always allow the use of a horizontal X-ray beam. The tube is adjusted so that the central ray passes horizontally to the middle at the level of the inferior orbital margins and to the centre of the film.

**Structures demonstrated**

The antra are clearly visible, the frontal sinuses are projected obliquely, though their floors are clearly shown. The ethmoid cells are largely obscured, but a few cells may be seen within the nose and medial to the lamina papyracea on the inner wall of the orbit. The sphenoidal sinuses are visible through the open mouth. If, after examining the film, a fluid level is suspected, its presence or absence can be confirmed by repeating the view with the sagittal plane of the head tilted 20-40° to the side in question.

**Occipitofrontal (or Caldwell) view**

The patient sits or lies prone with the forehead and part of the nose in contact with the Bucky support. The baseline is adjusted perpendicular to the film and the tube is angled 15° caudally. If a skull table is used the object table is angled 15-20° towards the patient, the central ray is directed horizontally to the nasion, and to the centre of the film.

**Structures demonstrated**

The frontal sinuses are clearly shown. The upper parts of the antra are obscured by the petrous bones but their lower parts are visible. The floor of the sella turcica, the crista galli, the nasal septum and middle and inferior turbinates can be seen. The ethmoidal and sphenoidal sinuses are superimposed.

**Overshot axial - submentovertical view**

The patient sits facing the tube. The neck is fully extended, so that the vertex rests against the Bucky support. The baseline should be as near parallel to the film as possible. The central ray is directed 2.5 cm anterior to the intermeatal plane to the midline, from beneath the mandible at an angle of between 90 and 100° to the baseline, and to the centre of the film. Slight over-extension is an advantage as both anterior and posterior walls of the frontal sinuses are then seen behind the dental arch.
Structures demonstrated

The ethmoidal and sphenoidal sinuses are shown free of superimposed cells. The posterior walls of the antra, petrous apices and base of the anterior and middle fossae of the skull are clearly seen. In elderly patients, or in those with short thick necks, it is often difficult to obtain sufficient extension of the head to bring the baseline parallel to the film. If a skull table is available this can be easily countered by angling the object table forwards towards the patient. If not, the central ray of the tube must accordingly be angled cephalically, to compensate for lack of head extension.

Lateral

The patient sits facing the Bucky table and the skull is then rotated into the lateral position. The central ray is directed horizontally to a point behind the outer canthus of the eye and to the centre of the film. When particular interest is directed to the nasopharynx, the patient sits sideways on with the head in the lateral position and the chin protruded. In children under 7 years of age the central ray is directed horizontally to a point immediately posterior to the angle of the mandible. In older children or adults the central ray is directed to a point 2.5 cm in front and 3.75 cm below the superimposed external auditory meatus and to the centre of the film. The focal film distance is increased from the standard 90 cm to reduce magnification.

Structures demonstrated

On the lateral projection the paired sinuses are superimposed on one another, but the extent of pneumatization of the frontal and sphenoidal sinuses can be gauged, especially in their vertical and horizontal directions. The thickness of the soft tissues in the nasopharynx, the uvula, and the extent of the nasopharyngeal airway can be assessed. Enlarged adenoids can be clearly shown. This view is essential when opaque foreign bodies are being sought, or when surgery on the sphenoid bone or transnasal implantation of radioactive isotope seeds into the pituitary gland is contemplated.

Other radiographic examinations

Oblique views

These may be useful to show the posterior ethmoid cells and optic foramina. The right and left sides are examined separately. The patient is positioned to face the Bucky table and the baseline is adjusted perpendicular to the film, the head is then rotated through 35-40° so that the rim of the orbit, the nose and cheek on the side to be examined, are 'flattened' against the Bucky support. The central ray of the tube is angled 10-15° caudally (or the baseline raised 10-15°) to prevent the petrous bones obscuring the ethmoids, and directed to pass through the centre of the orbit and the centre of the film.
Structures demonstrated

This view shows the posterior and middle groups of ethmoidal cells with some superimposition of the sphenoidal sinuses on the side to be examined, as well as the optic foramen and floor of the anterior fossa. The frontal sinuses are shown obliquely. Half the sphenoid bone and a few posterior ethmoid cells of the opposite side are also shown.

Occipitomental views with varying angulation

Overangled or underangled occipitomental views may be used to give tangential views of the roof of the antrum in suspected injuries in that area. The patient is positioned as for a normal occipitomental view and the additional views are taken with the baseline at 60° and 30°, respectively. Increased angulation may be needed in patients in whom the petrous bones overlie the base of the antrum in the standard view.

Panoramic views (orthopantomography)

Panoramic tomography in the plane of the dental arches is used for dental surveys (see Volume 1, Chapter 17), but at the same time the orthopantomograph gives a good display of the lower aspects of the antra which may be used to supplement the plain film demonstration of the sinuses.

Conventional tomography

Hypocycloidal or spiral tomography is preferable as fine bone detail on thin sections can be shown without the streaking produced by the linear mode. Until recently, complex motion tomography was the definitive investigation for most sinus lesions after plain radiographs. It has been replaced almost entirely by high resolution computerized tomography which gives similar sectional imaging, but with the advantage of much better discrimination of soft tissues.

Coronal section tomograms may be useful for elderly patients unable to adopt the chin up or head hanging position needed for coronal computerized tomography. The examination should be performed with the patient prone to restrict the radiation dose to the eyes. Ready comparison of symmetry between the left and right sides can be made. Lateral tomograms in the sagittal plane are also useful because positioning for direct sagittal CT sections is particularly difficult.

Computerized tomography (CT)

The superiority of CT over other methods of imaging the sinuses can be summarized as follows:

1) the bony walls of the sinuses are demonstrated at least as well by CT in the high resolution mode as by conventional radiography and tomography
(2) an excellent anatomical display of soft tissue densities, including fluid levels and polypoid masses, within the normally air-filled cavities of the sinuses, nasal cavity and postnasal space is provided

(3) most important of all, disease extending beyond the bony perimeters of the sinuses into the adjacent soft tissue of the orbit, brain and infratemporal fossa can be imaged.

These applications of CT have disappointed in only one way. While giving an excellent anatomical display, CT generally fails to predict the histological nature of the pathological process, unless there is characteristic calcification within a tumour such as a meningioma or chondroma.

'Tissue characterization' with or without contrast enhancement is almost always unsuccessful if measurement of attenuation values is used, and this was appreciated in the early days of CT (Forbes et al, 1978). Contrast enhancement is required:

(1) if intracranial extension of disease is suspected on clinical grounds or from the anatomical demonstration, particularly on the coronal CT sections

(2) if a particularly vascular tumour such as a meningioma or juvenile nasopharyngeal angiofibroma is suspected; a contrast infusion to ensure that enhancement occurs in the vascular phase is required in such cases (see Volume 1, Chapter 17)

(3) occasionally if an inflammatory process is suspected.

Most American authorities, however, recommend routine use of contrast enhancement for the investigation of the facial area (Hasso, 1984). The present author does not advocate this approach, as intravenous contrast medium usually enhances all soft tissue structures around the sinuses, and is therefore unhelpful, besides adding to the length of the examination, increasing the not inconsiderable radiation dose to the patient, and adding the risk of a systemic reaction to the contrast agent.

The standard plane for CT scanning of the paranasal sinuses is parallel to the infraorbital meatal baseline. This plane is nearly parallel to the planes of the hard palate, the zygomatic arches and much of the orbital roof. The antra are seen in cross-section, the ethmoid and sphenoid sinuses are well demonstrated, as are the anterior and posterior walls of the frontal sinuses. These axial sections are also the best for demonstrating the orbital contents and the nasopharynx.

The axial scans are then reviewed to decide whether further sections are necessary in the coronal plane. Generally, evaluation of structures parallel to the infraorbital baseline or evidence of intracranial extension of the disease process necessitate coronal sections. For both axial and coronal scans, contiguous sections, 5 mm thick, are usual. Degradation of the images by metal fillings in the teeth, especially in the coronal plane, may require repositioning of the patient's head or the angle of the gantry.

The soft tissue structures of the face need to be assessed on a window setting of 400-800 Hounsfield units, while bone detail is best shown at a setting of 1000-3000 Hounsfield
units. Even at the wide window setting, thin plates of bone, especially the lamina papyracea forming part of the medial wall of the orbit, may appear to be dehiscent because of partial volume averaging. Erosion of a thin plate of bone should not be diagnosed on the CT scan unless an adjacent soft tissue mass can be demonstrated, and then the problem of soft tissue silhouetting is added to that of partial volume averaging (see Volume 1, Chapter 17).

The maxillary antra are small at birth and expand progressively during the first decade of life. Occasionally they may remain infantile, and then it can be difficult, on plain films, to distinguish this cause of opacity from the more usual such as chronic infection or fibrous dysplasia. The antra are rarely asymmetrical but the absence of pathological features and presence of a normal ostium can be confirmed by axial CT. Sometimes a small sinus may be double.

**Magnetic resonance (MR)**

The bony margins of the sinuses appear as a plane of absent signal on magnetic resonance scans and this limits the usefulness of the technique for examination of the sinuses. Moreover, the intense signal from the high fat content of bone marrow, as in the basisphenoid and petrous apices and around the frontal sinuses, can be very confusing for the radiologist interpreting the scans. This is particularly so as retained fluid within the sinuses gives a similar intense signal from the high water content.

It is difficult or impossible on a CT scan to differentiate tumour tissue from retained fluid in sinuses where the drainage of a sinus is blocked by obstruction from the tumour. Differentiation on an MR scan is simple and clear. Extension of sinus neoplasia into the cranial cavity is shown very well by magnetic resonance without the need for contrast enhancement and the ability to image in any plane is a considerable advantage.

**Angiography**

The requirements for angiography of the sinuses have become much reduced since the advent of CT and MR imaging. Occasionally, angiography is needed to demonstrate the blood supply of soft tissue lesions of the face, especially in vascular tumours such as juvenile nasopharyngeal angiofibroma, when therapeutic embolization may be carried out as a preoperative procedure.

The important radiological features of some of the commoner conditions affecting the paranasal sinuses will be considered briefly.

**Specific clinical situations**

**Trauma**

Injuries to the face and sinuses are considered in Chapter 14. Fractures are usually demonstrated by conventional radiographic techniques, but tomography and CT are often necessary to show the fracture lines. Fluid levels in sinuses indicating a cerebrospinal fluid fistula are well shown by CT. The improved soft tissue imaging of CT and MR imaging can also be an advantage in trauma cases, especially for showing the state of the orbital contents.
**Inflammatory disease**

Mucosal thickening, a common finding on plain sinus views, is shown far more readily on CT scans. Normal mucosa is too thin to be demonstrated on the scan, but minor degrees of thickening in the absence of relevant symptoms may be dismissed as an incidental finding. If the sinus ostium becomes blocked, a completely opaque sinus will result. An air/fluid level may be observed at an intermediate stage before the obstructed sinus becomes totally opacified.

Chronic obstruction of the ostium or a septate portion of the sinus cavity gives rise to mucocoeles. Proptosis is often the presenting symptom. The diagnosis is often suspected on plain skull radiographs, but CT confirms the benign nature of the lesion and gives an accurate display of its extent (Price and Danziger, 1980). Initially it was hoped that a low CT attenuating reading would help to confirm the diagnosis of mucocoele but such readings have proved extremely variable. Nevertheless, the diagnosis can usually be made from the expansile appearance of the lesion.

The characteristic radiological features of benign nasal polyposis are familiar to otolaryngologists. However, in a proportion of patients the changes are much greater and include widening of the ethmoid labyrinth and nasal cavity, bone thinning and expansion, and mucocoele formation (Lund and Lloyd, 1983). The ethmoid widening and opacity can be shown by plain films or CT.

Most acute sinus infections are successfully treated by antibiotics, but sometimes the inflammatory process spreads beyond the confines of the sinus cavities. The close association with the orbit means that the most frequent complication of acute inflammatory sinus disease is orbital cellulitis with pain, oedema and proptosis. A subperiosteal or orbital abscess is a more serious complication which may require surgical exploration and drainage. The decision regarding surgical intervention is greatly helped by the CT demonstration of the site of the abscess. Patients with orbital cellulitis frequently require external drainage. Frontoethmoidectomy may be required if resolution does not occur or chronic infection is present (Harrison, 1980). Pus between bone and orbital periosteum is well shown by coronal CT. If the infection breaches the periosteum the normal muscle and fat planes rapidly become indistinct and vision deteriorates. Orbital ultrasound is said to be helpful in the differentiation of orbital cellulitis and abscess formation (Goodwin, 1985).

**Postoperative state of the sinuses**

Following surgery on the paranasal sinuses, radiological investigation may be required to determine the presence of recurrent disease. After the Caldwell-Luc operation, the postoperative appearance is opaque on plain sinus views, but CT has shown that this opacification is commonly due to bone thickening (Cable et al, 1981). This would seem to be the result of periosteal stripping followed by the deposition of new bone. The postoperative antrum is often smaller than normal. Although CT differentiates well between bone and soft tissue, unfortunately it is not so satisfactory for excluding residual soft tissue disease such as loculated pus.
Tumours

The CT diagnosis of a tumour requires the presence of a soft-tissue tumour mass. Changes in the adjacent bones are secondary features. Categorizing the bone destruction seen on CT as either aggressive bone destruction or bone remodelling aids differential diagnosis. Bone remodelling or expansion reflects slow growth of the tumour. New bone is laid down on the outer surface of the sinus wall as erosion takes place from the inner wall adjacent to the tumour and differentiation from a mucocoele or expansion by benign simple polyposis may be difficult.

A mass in the middle meatus of the nasal cavity extending into the antrum is highly suggestive of an inverting papilloma (Lund and Lloyd, 1984). Other features of this tumour which have been shown by CT are small areas of calcification within the tumour mass and sclerosis of the sinus walls, although the latter is a non-specific change most frequently seen in chronic sinus infection. More obvious extensive and dense calcification within an expansile mass is a feature of chondroma or chondrosarcoma. This calcification is an important feature in the diagnosis, well demonstrated by CT but not shown by MR. Lloyd and Phelps (1986) have shown significant advantages in entirely different ways for both CT and MR in the diagnosis and assessment of one particular rare benign tumour, namely juvenile angiofibroma. Thirty cases were described, all of whom had bone destruction at the base of the pterygoid lamina. The distinctive radiological features of these tumours are discussed in Volume 6, Chapter 2, and the radiological anatomy and pathology of the infratemporal fossa and parapharyngeal region in Volume 5, Chapter 2.

Malignant sinus neoplasms characteristically produce aggressive bone destruction: the bone is rapidly permeated and destroyed. Such destruction is seen primarily with squamous cell carcinoma which accounts for nearly 80% of all paranasal sinus malignancies (Sone, 1985). Where plain sinus views are the usual primary means of imaging, it is most important for both radiologists and surgeons to look carefully for erosion of the bony margins of the sinuses, especially in the presence of suspicious clinical features.

Obliteration of fascial planes beyond the sinus walls is the most characteristic CT sign for the identification of malignancy. Whenever a soft tissue mass extends beyond the bony confines of a sinus, neoplasia should be ruled out by biopsy. Extension of sinus neoplasia into the orbit or cranial cavity affects the management of the disease. Coronal CT sections with contrast enhancement or equivalent MR views are necessary if initial clinical and radiological examinations indicate extension of disease up to the floor of the cranial cavity.

Bony tumours and bone dysplasia

Osteomata are common benign tumours which produce no symptoms unless they extend beyond the sinus, block the ostium, cause pressure on a nerve or displace other structures. They are commonest in the frontal sinuses and are readily demonstrated by plain radiographs.

Tomography or CT is sometimes required to show the point of attachment to the sinus wall and axial CT gives a good demonstration of the posterior extent. The density of the mass depends on the amount of ivory and cancellous bone present in the tumour. A benign bony
tumour which fills the sinus, with no radiographic features to suggest an osseous lesion, may cause problems in assessment. A large mass in the antrum which appeared to have arisen from the alveolus was histologically diagnosed as a giant-cell reparative granuloma and its true relation to the antrum only revealed at surgery. Other benign tumours of the jaws can produce a similar appearance on CT with destruction of the alveolus and evidence of spread into the soft tissues. Meningiomata may very rarely arise in the sinuses but are more likely to affect the sinuses by extension from within the cranial cavity. CT is the imaging investigation of choice, not only to demonstrate calcification within a tumour mass, but also to show the osseous sclerosis that occurs with meningioma. Nevertheless, differentiation of meningioma en plaque from developmental diseases such as fibrous dysplasia may be difficult. Fibrous dysplasia is either polyostotic or monostotic and usually develops early in life. The skull and facial bones may be affected singly or as part of a more generalized disease. Characteristically there is thickening and expansion of bone but the density of the lesion depends on the amount of fibrous tissue present; when this is high, a ‘ground glass’ appearance is seen. In other cases dense bone predominates and, less commonly, a mixed type with islands of dense bone in a fibrous matrix occurs.

The nasopharynx (postnasal space)

These alternative terms to describe the space behind the posterior choanae of the nose highlight the dilemma of whether the nasopharynx should be considered with the nose or the rest of the pharynx. Traditionally radiological examination of the nasopharynx was by lateral and submentovertical views to show soft tissue masses distorting or obstructing the airway or eroding the base of the skull. Complex motion tomography improves the assessment of these features and helps to demonstrate masses and base of skull erosion, but it has largely been supplanted by CT which is now the optimum means of demonstrating the nasopharynx. Although an excellent demonstration of the air mucosal interface is obtained, it should be remembered that this is better assessed clinically with a mirror or nasopharyngoscope. The characteristic shape of the nasopharynx on axial CT shows torus tubarius as the most prominent landmark. In front is the opening of the eustachian tube and behind the lateral pharyngeal recess, or fossa of Rosenmüller. What is more important, and a great advance in imaging, is the ability of CT to demonstrate the deep tissue planes which lie beneath the mucosa. The axial view of these planes complements the clinical examination because deep extension is the hallmark of malignancy in the nasopharynx. The pterygoid muscles attached to the lateral pterygoid plate and the deglutition muscles can be recognized as well as the fat in the paranasopharyngeal space between them. Further consideration of these parapharyngeal structures is given in Volume 5 on the pharynx. Diseases of the nasopharynx frequently obstruct the eustachian tube, and the subsequent opacity of the middle ear cleft is easily demonstrated by CT.

Malignant tumours of the nasopharynx

Almost all deeply infiltrating lesions of the nasopharynx will prove to be primary neoplasms, usually squamous cell carcinoma. The most important feature on the CT scan is obliteration of the paranasopharyngeal soft tissue planes and in particular the fat in the paranasopharyngeal space. The deep soft-tissue structures are normally very symmetrical so comparison between the two sides is useful. Such symmetry is not so constant in the normal outline of the mucosal surface.
Sometimes carcinomata of the upper aerodigestive tract are not apparent on examination of the mucosal surfaces because the tumour arises either submucosally or within a deep crypt and then extends into the deep tissue planes rather than the lumen. In one series (Mancuso and Hanafee, 1983) there were 19 mucosally inapparent carcinomata of the upper aerodigestive tract out of 160 cases examined. There may or may not be a mass bulging the wall of the nasopharynx but diagnosis can only be made by CT guided biopsy. Endoscopy and biopsy compromise the effectiveness of CT and therefore should be carried out after the CT examination.

Asymmetry of the muscle layers may be due to atrophy after radiotherapy or sometimes to neurogenic muscular atrophy from infiltration by a carcinoma. Occasionally nasopharyngeal carcinomata grow in a more exophytic manner into the lumen of the nasopharynx, but this is usually a feature of lymphomata.

Erosion of the skull base and extension of tumour into the cranial cavity along the carotid artery or through the foramen lacerum can be shown by enhanced CT, although the soft tissue intracranial extension is probably best demonstrated by magnetic resonance.

### Chordoma

Chordomata are predominantly midline tumours. A large soft tissue mass in the postnasal space is associated with destruction of the basisphenoid and sometimes flecks of calcification best shown by CT. There is usually an associated intracranial mass and irregular destruction of adjacent bone depending on the site of origin (Jeans, 1984).

### Summary

Conventional plain radiographs will continue to play a major role in the initial investigation of diseases of the paranasal sinuses, despite the limitations of this technique. Good radiographic method and positioning of the patient are important, as is the ability of the observer to detect early signs of disease such as erosion of the sinus walls. High resolution computerized tomography gives an excellent demonstration of both fine bone detail and soft tissue anatomy on the same sectional picture and is now the investigation of choice. CT can demonstrate a tumour early in the course of the disease and can be used to recognize the exact extent of the lesion for optimal staging prior to therapy. CT plays an important role in follow-up and can be used to show residual or recurrent disease. Magnetic resonance gives better soft tissue imaging in three planes and appears to be better than CT for showing extension of disease into the cranial cavity. However, bony structures are not imaged and therefore MR does not appear likely to replace CT for the investigation of diseases of the paranasal sinuses in the foreseeable future.
Chapter 3: Endoscopy of the nose and sinuses

C. B. Croft

The technique of examination of the nasal cavity and nasopharynx has remained essentially unchanged since the development of otolaryngology as a separate specialty during the nineteenth century. Routine anterior and posterior rhinoscopy, using a speculum, mirrors and light source, are still the standard techniques of inspection and assessment of the nasal airway. The views obtained by these methods are necessarily incomplete and are usually augmented by radiological examination of the nose and paranasal sinuses. The development of endoscopic techniques allows direct examination of these structures and has focused attention on the difficulties of accurate assessment in nasal and sinus disease. Simply stated, there are three major reasons why endoscopy is of vital importance if adequate information is to be obtained about the state of the nasal cavities and paranasal sinuses:

1. certain parts of the nasal cavities are inaccessible to view without the use of endoscopic techniques, that is the superior, middle and the inferior meatus

2. radiographic evidence of the degree and type of sinus pathology has now been shown to be highly suspect; at best there is only a 50-60% correlation between radiographic findings and the actual findings in the sinuses at endoscopy or surgery (Illum, Jeppesen and Langebaeck, 1972; Herberhold, 1973; Pfleiderer, Croft and Lloyd, 1986)

3. examination of the postnasal space via mirror posterior rhinoscopy is impossible in about 20% of patients.

History of endoscopy

In 1853, D'Esormeux demonstrated an alcohol illuminated urethroscope, winning a prize at the Paris Exhibition of that year. Endoscopy was born. The development of electricity followed and, by 1879, Leiter had developed a distally illuminated, water-cooled cystoscope which created great interest. Nitze's success in using this instrument stimulated Zaufal to use a modified cystoscope to examine the eustachian tube orifice pernasally during the 1880s. However, the 'father' of nasal endoscopy was Hirschmann, who used a special endoscope only 4.0 mm in diameter to examine the middle meatus and study the sinus ostia at the turn of the century (Hirschmann, 1903). Hirschmann also examined the maxillary antrum, via a molar tooth socket, for diagnostic purposes, but had been preceded by Reichert in 1902 who published his observations on antroscopy and was the first to perform and describe minor intrasinus manipulations under endoscopic control (Reichert, 1902). However, the work done by these pioneers was not taken up, and perhaps the arrival of apparently adequate diagnostic radiology during the next decades pushed this new technique to one side. By 1925, nasal endoscopy was being described as 'an interesting amusement' and had been largely discarded.

During the 1950s, several developments improved the technology of endoscopy - Hopkins working at Imperial College developed his solid rod lens system and the proximal 'cold light' source. These important innovations allowed much better optical views using the
new telescopes and greatly extended their use. Subsequently, several workers on the Continent have reported on the beneficial results of sinus endoscopy in the diagnosis and treatment of chronic sinus disease. This work has been carried on enthusiastically during the last decade and has resulted in the excellent publications of Buiter (1976) and Draf (1983).

**Indications for endoscopy**

**Nasoendoscopy**

(1) To evaluate inaccessible areas in the nasal cavity, for example the *superior meatus* in cases of anosmia; the author has found patients with polyps and obstructing adhesions *above* the middle turbinate completely unseen on routine anterior rhinoscopy which required surgical lysis and which resulted in a return of the sense of smell.

The *middle meatus* - evaluation of sinus ostia and surroundings for obstructing pathology. This area can only be visualized with difficulty using routine anterior rhinoscopy.

The *inferior meatus* - usually to check on the status of previously created surgical antroscopies, are they patent or obstructed?

(2) To monitor the nasal fossa after resections for tumours, that is radical maxillectomy or more extensive craniofacial resections. It is impossible to examine the cavities of these patients adequately if the hard palate is intact.

(3) To evaluate epistaxis of unknown origin and the nasal fossa in cases of cerebrospinal fluid rhinorhoea. The author has used endoscopy to help localize a difficult cerebrospinal fluid leak to the sphenoid sinus area.

(4) To assess the spread of paranasal sinus tumours within the nasal fossa, with respect to their superior and posterior extremities.

(5) To assess the degree and site of airway obstruction in the nasal cavity. Lesions obstructing the posterior choanae and nasal cavities can best be assessed endoscopically.

(6) To monitor progress in the treatment of nasal and paranasal sinus infections.

**Nasopharyngoscopy**

Hays (1909) described endoscopic examination of the nasopharynx through the mouth - and subsequently many workers have followed this route (Buiter, 1976). The author prefers to examine the nasopharynx pernasally using flexible fibroptic instruments such as the Olympus ENF ‘P’. *(See the section on Technique.)*

**Inaccessible nasopharynx**

To examine the nasopharynx in 15-20% of the population in whom it is inaccessible.
Examination of tumours of the nasopharynx

This technique allows precise documentation of the site and extent of the tumours of the nasopharynx. It is possible to obtain a biopsy under local anaesthesia on an outpatient basis - either by using a flexible fibrescope with a biopsy channel (Olympus ENF-LB) or by inserting a Barts' forceps through the opposite nasal fossa and obtaining a biopsy under direct visual control. This technique allows rather more precise biopsies than those obtained using a mirror and pernasal biopsy under general anaesthetic in the classical manner.

Post-treatment evaluation of the nasopharynx

The majority of patients with nasopharyngeal tumours will receive primary radiotherapy, although a minority, such as those with angiofibromata, will be treated surgically. The problems which arise following radiotherapy treatment are those of mucosa crusting and scarring, which can make indirect mirror examination and interpretation difficult. Direct pernasal examination is most helpful in these difficult cases in differentiating between recurrence and mere crusting with infection.

Functional studies

Examination of the eustachian tube orifice in cases of dysfunction is of paramount interest to the otolaryngologist. Is there benign or malignant obstructing pathology? Or evidence of inflammatory or allergic changes creating the problem? Direct biopsies can be taken which will help in planning treatment.

The major resurgence of interest in direct nasopharyngoscopy followed the work of Pigott (1969) who used this route to study patients with incompetence of the velopharyngeal sphincter, both before and after cleft palate repair. Nasopharyngoscopic monitoring of the velopharyngeal sphincter is now one of the standard methods of diagnosis and assessment at cleft palate clinics and has led to the documentation of microforms of cleft palate (Croft, Shprintzen and Daniller, 1978) which are of particular interest to otolaryngologists because of the dangers of rhinolalia aperta developing in these patients after adenoidectomy. It has also become apparent that there are various patterns of closure of the velopharyngeal sphincter (Croft, Shprintzen and Rakoff, 1981) and that the different components of palatal elevation, lateral pharyngeal and posterior pharyngeal wall motion vary considerably from individual to individual. It is also clear that the posterior wall motion of 'Passavants' ridge occurs in a percentage of normal subjects and is not merely a compensation seen in patients with velopharyngeal insufficiency (Croft, Shprintzen and Rakoff, 1981).

Airway evaluation

There is increasing evidence that airway obstruction in the nasal cavity or nasopharynx may be responsible for the development of snoring with or without sleep apnoea. Although this subject is covered elsewhere (see Chapter 17), the author regards flexible nasopharyngoscopy as mandatory in studying patients with sleep-related breathing problems. This manoeuvre allows documentation of both physical and functional airway obstruction and the reader is referred to the recent paper by Sher et al (1985) which discusses endoscopic examination in combination with the Müller manoeuvre to predict the response of patients to
upper airway surgery such as uvulopalatopharyngoplasty. This would appear to increase the success rate of this type of surgery in relieving obstructive sleep apnoea and snoring.

**Technique**

*Rigid nasoendoscopy*

The author favours the 30° angle viewing 4.00 mm Storz Hopkins rod for evaluation of the nasal fossa. Simple preparation of the nose requires adequate spraying with a solution of 2% lignocaine and 1:2000 adrenaline which allows excellent mucosal reduction and anaesthesia. The wider nasal cavity is then examined with the patient semi-recumbent and the head supported to prevent movement. The telescope is passed posteriorly above the inferior turbinate, below and medial to the middle turbinate. The nasal cavity is examined thoroughly with the middle meatus examined as the telescope passes to the posterior choana. Posteriorly the sphenoethmoidal recess is examined followed by evaluation of the region above the middle turbinate. Occasionally nasal anatomy may dictate the use of the inferior meatus to reach the nasopharynx and this area must be examined in patients who have previously undergone intranasal antrostomy or Caldwell-Luc procedures.

The main contraindication to rigid nasoendoscopy is deformity of the nasal septum which may preclude the use of rigid instrumentation - flexible fibreoptic endoscopy will then be required.

*Flexible nasoendoscopy and nasopharyngoscopy*

The author favours the Olympus ENF-P nasoendoscope which has a 2.9 mm diameter bundle and 90° tip deflection in both directions. Although this instrument is fragile and has no biopsy or suction channel, its uniquely small size allows pain free examination of the airway and it is possible to use it in neonates without difficulty. The ENF-LB has a larger 4.0 mm bundle and integral biopsy channel which is useful for clearing secretions and obtaining endoscopic biopsies. The use of either rigid or flexible instrument requires use of a demisting agent, such as 'Ultrastop' or simply using a little Savlon solution. Warming the rigid telescopes in warm water is perfectly satisfactory but, over the long term, may damage flexible fibreoptic instruments.

Local anaesthesia and mucosal reduction is obtained just as in rigid endoscopy using a 2%/1:2000 lignocaine/adrenaline spray. Occasionally patients require more than this rather simple application and packing the nose with a pledget of cottonwool soaked in the same solution is helpful. This is particularly so in children who may object to having their noses sprayed, but will tolerate a small cottonwool pad.

Flexible nasoendoscopy is performed with the patient sitting and the head supported. The examiner faces the patient and the endoscope tip is passed into the nasal cavity through the controlling fingers of the left hand which rest lightly on the patient's nose. The right hand controls the endoscopic body with the tip deflector control, allowing appropriate adjustments of the angulation of the endoscope within the nasal cavity. The widest part of the nasal cavity is examined first. This is usually the area between the middle and inferior turbinate. Flexible
endoscopes of the type described are very versatile and allow complete examination of the nasal cavity in nearly all cases.

**Sinus endoscopy**

**Uses and indications**

(1) Accurate diagnosis - resolving the dilemma of radiographic accuracy (*see below*):
   - maxillary antrum
   - frontal sinus
   - sphenoid sinus

(2) Suspicious symptoms with negative radiographs, unexplained pain or bleeding

(3) Intrasinus surgery and monitoring the response to therapy

(4) Early diagnosis of malignancy of the paranasal sinuses

(5) Sinus involvement in pathology from surrounding sites.

**Method**

**Endoscopic instruments**

Rigid sinus endoscopy requires the use of rigid telescopes using Hopkins optics. The author uses Storz Hopkins rod telescopes with $0^\circ$, $30^\circ$ and $70^\circ$ angles of view. These solid rod lens telescopes provide excellent contrast and resolution with good depth of field. They allow good photographic documentation of intrasinus pathology.

**Photographic equipment**

(1) Storz photo flash generator - which combines a cold light source, flash and flash generator; the generator has variable settings for flash intensity

(2) Single lens reflex camera (Olympus OM-1) with special endoscopic zoom lens and ring adaptor (Storz)

(3) High speed film - 400 ASA.

**Surgical instruments**

Maxillary or frontal sinus endoscopy requires the:

(1) Storz standard 16.5 cm trochar and cannula - the sphenoid sinus requires an extra long 20 cm trochar cannula, but with a similar diameter

(2) Syringes and 12.0 F suction catheters for irrigation and aspiration of sinus content

(3) Optical biopsy forceps - rigid integral biopsy forceps with an $0^\circ$ miniature Storz Hopkins telescope
(4) St Bartholomew’s ethmoid forceps - up and down biting for biopsies via the antrostomy

(5) Thudichum’s speculum and Hills elevator.

**Technique**

**Maxillary sinoscopy**

The nose is prepared by spraying with 1 mL of a solution of 2% topical lignocaine and 1:2000 adrenaline. This reduces problems with bleeding and mucosal reduction improves access to the favoured inferior meatal route. Under local anaesthesia, a strip of cotton wool soaked in lignocaine and adrenaline is packed into the inferior meatus.

Access is by way of (a) the inferior meatus or (b) the canine fossa. The author favours the inferior meatal route, believing it to be the simplest and therefore the safest of the two routes. The technique is very similar to that used for proof puncture and therefore already very familiar to otolaryngologists.

The 5.0 mm trochar and cannula is inserted under the highest point of the inferior turbinate and the bony wall pierced with a rotating movement of the trochar. It is important not to drive the trochar too far lest the lateral antral wall be traumatized and subsequent bleeding obscure the view of the antrum. The trochar is removed and, using telescopic control, the cannula is gently drawn backwards until the antral mucosa can be seen. This will provide a maximal view of the sinus interior. The antrum is then inspected using 70° and 30° telescopes. The telescopes have either been heated before use or the lens demisted using Ultrastop. It is important that inspection is performed by rotating the telescope and cannula around their axes and that no lateral leverage is used on either cannula or telescope as both bleeding and pain may result.

If bleeding has occurred and the field is obscured, then gentle suction via a No. 12 catheter or suction/irrigation with warm saline will clear the field fairly rapidly. Once a good view of the antrum is obtained, the condition of the mucosa and the extent of any abnormality is assessed. Free pus can be sampled for culture and sensitivities. Any mucosal abnormality can be definitely identified by biopsy and the site, number and patency of the sinus ostia assessed.

The canine fossa route certainly provides a more consistent view of the maxillary sinus ostium; it is approached following injection infiltration of the canine fossa mucosa with 2% lignocaine with 1:200,000 adrenaline. This procedure is best performed with the patient semi-recumbent. The highest part of the canine fossa is palpated and a small stab incision made using a No. 15 Parker knife blade. The trochar and cannula are then inserted using a rotating action into the antero-lateral wall of the antrum. Telescopic examination is then performed by way of the cannula as described.
Frontal sinoscopy

The main indications for endoscopic examination of the frontal sinus are unresolved radiographic abnormalities in symptomatic patients. This examination should be performed on a recumbent patient under general anaesthetic. Precise radiographic documentation of the lateral and anteroposterior dimensions of the frontal sinus is mandatory. The medial eyebrow is infiltrated with an injection of 2% lignocaine and 1:200,000 adrenaline. A 1.5 cm incision is made in the inferior margin of the eyebrow and the floor of the frontal sinus exposed. The periosteum is elevated and, using a drill with a cutting burr (6.0 mm), an inspection hole is made just below the orbital rim - the cannula and telescopes can then be inserted and pathology assessed and biopsies taken as required.

Sphenoid sinoscopy

The sphenoid sinus ostium can be visualized nasoendoscopically as already noted. It is quite possible to open the anterior wall of the sphenoid sinus near the midline under telescopic control, but the face of the sphenoid varies considerably in thickness and, rather than take risks in this vital area, the author prefers to approach the sphenoid via the midline trans-septal route using the operating microscope once the sphenoid rostrum is reached.

Discussion: sinoscopy versus radiology

A major problem which soon confronts the otolaryngologist engaged in sinus endoscopy is the discrepancy which is apparent between radiographic evidence and endoscopic findings.

Accurate diagnosis in cases of chronic sinus disease has always been difficult. It is certainly the case that poor preoperative diagnosis has contributed to many of the unsatisfactory results of sinus surgery and perhaps to its low popularity rating with patients. Efforts to improve diagnostic accuracy with antroscopy soon revealed the problem of poor correlation between X-ray changes and endoscopic findings. Illum, Jeppesen and Langebaeck (1972) and Herberhold (1973) showed only a 62% and 64% positive correlation in their comparative studies of sinus endoscopy and radiology. A recent study by the author (Pfleiderer, Croft and Lloyd, 1986) has confirmed the very poor correlation between what can be deduced from an X-ray plate and what is actually going on inside the maxillary antrum. The authors compared antroscopic and radiographic findings in 115 patients, examining 193 antra. The comparison was effected by correlating endoscopic and radiographic changes as shown in Table 3.1. Thus an 'allergic' mucosa was identified as pale and polypoidal at endoscopy and correlated with an irregular outline of the thickened mucosa seen radiographically. This study found that the radiological and antroscopic diagnosis was identical in 85 out of 193 cases studies, a positive correlation of 44%. However, current radiological techniques produced false positive and false negative findings of 35% and 9%, respectively. Radiographic techniques were particularly poor in identifying inflammatory disease, identifying an infected antrum in only 26% of cases in which the definitive diagnosis was made antroscopically. Furthermore, the antroscopic findings in 29 opaque antra showed that the majority were due to severe mucosal thickening, while a minority were infected and contained pus. Clearly, proper surgical planning in cases of sinus disease requires more objective information than that provided by X-rays and the return from a sinus irrigation.
Table 3.1 Radiological and antroscopic classification of mucosal changes in the maxillary antrum

<table>
<thead>
<tr>
<th>Radiographic</th>
<th>Antroscopic</th>
</tr>
</thead>
<tbody>
<tr>
<td>I Clear</td>
<td>I Normal</td>
</tr>
<tr>
<td>II Cyst-like lesion</td>
<td>II Solitary cyst</td>
</tr>
<tr>
<td>III 'Polypoid' mucosal thickening*</td>
<td>III Allergic mucosal change</td>
</tr>
<tr>
<td>IV 'Straight' mucosal thickening&amp;</td>
<td>IV Infective mucosal change</td>
</tr>
<tr>
<td>V Opaque antrum</td>
<td></td>
</tr>
</tbody>
</table>

* Interpreted as being of allergic origin
& Interpreted as being of infective origin.

In this respect, it is a salutary lesson to inspect the maxillary antrum endoscopically after an apparently adequate sinus wash-out. The antrum may still contain large amounts of purulent material in spite of a clear return from sinus irrigation - certainly antroscopic control of irrigation of a grossly infected antrum is most useful.

Possible reasons for the diagnostic inaccuracy and discrepancies of sinus radiography may include:

1. A narrow lateral recess - the lateral recess contains less air than the main sinus cavity and superimposed bony shadows may create an appearance indistinguishable from mucosal thickening.

2. Basal region - radiographic variation due to differing bone density and contours at the base of the antrum can give rise to false positive and false negative information about the state of the mucosa at the sinus base.

3. Radiographs of maxillary antra which have been the site of previous sinus surgery are notoriously difficult to interpret. Endoscopic evaluation is mandatory to assess the status of the antrum in any meaningful way following surgery.

4. Faults in radiographic technique - cysts in the anterior portion of the antrum or mucosal thickening in the anteroinferior area of the antrum may show up as an opaque antrum in an overtilted occipitomental view.

Management of chronic mucosal disease

The ability to examine the sinus interior and contents and to directly biopsy and examine diseased mucosa presents a distinct advantage in the management of chronic sinus disease. In the presence of infected mucosa, repeated endoscopy can monitor recovery and the response to appropriate medical therapy. Return of the mucosa and vessel pattern to normal with identification of the natural sinus ostium are encouraging signs. Lack of response may suggest that a more radical approach would be justified.
The intrasinus control of simple pathology such as cysts and polyps is greatly facilitated by the use of endoscopic surgery as described. These approaches via the inferior meatus have greatly reduced the number of Caldwell-Luc procedures performed in the author's patients and reduced the number of patients requiring inpatient care and surgery. The socioeconomic effects of these treatment policies are obviously beneficial to all concerned.

**Diagnosis of sinus malignancy**

Antroscopy provides an opportunity to diagnose malignant disease of the upper jaw at an earlier stage than hitherto. The poor and unchanging survival figures for malignant disease of the paranasal sinuses (with five-year survival of around 35%) have not altered much over the last 25 years. Antroscopy does provide the opportunity to diagnose malignancies before they have spread beyond the bony confines of the sinus and while they have a much greater potential for cure. During the recent study of 190 antrosopies (Pfleiderer, Croft and Lloyd, 1986), one case of antral malignancy was discovered.

A further advantage of sinus endoscopy is the ability to fully assess the spread and involvement of tumours arising both in the paranasal sinuses and the nasal fossa. Radiographically opaque sinuses, in such cases, may be due to either direct tumour involvement or to obstruction of the natural ostium by tumour with retained and possibly infected secretions. Proper treatment planning demands precision in assessing the full extent of tumour involvement.

**Complications**

**Vasovagal collapse**

Endoscopic evaluation of the nose and paranasal sinuses under local anaesthesia may occasionally result in collapse. The author has had to deal with one respiratory arrest in several thousand endoscopies and during a recent series of 170 outpatient antrosopies two vasovagal collapses were experienced. Clearly it is important to have full resuscitation facilities available in any unit performing these procedures.

**Epistaxis**

This is usually minor and rarely requiring more than a temporary pack.

**Incomplete examination**

About 4% of endoscopies will be unsuccessful due to the anatomy of the sinus or septum and it may be necessary to adopt the canine fossa route. Bleeding may obscure the view and preclude a satisfactory examination. Management will require lavage and gentle suction to clear the sinus of retained blood.
Chapter 4: Rhinomanometry and nasal challenge

Ronald Eccles

Site of nasal resistance

In adults, the nose contributes two-thirds of the total airway resistance (Ferris, Mead and Opie, 1964; Speizer and Frank, 1964). This nasal resistance can be divided into three components: the nasal vestibule, the nasal valve and the turbinated nasal passage.

The case is presented below that: the nasal vestibule acts as a flow-limiting segment on inspiration and contributes one-third of the nasal resistance; the nasal valve contributes most of the remaining two-thirds of nasal resistance; and the turbinated nasal passage has a minimal contribution to nasal resistance. The three components of nasal resistance are illustrated.

The narrowest point of the nasal passage determines the overall resistance to airflow and this region is often referred to as the 'nasal valve' (Dishoeck, 1965; Bridger, 1970; Haight and Cole, 1983b). However, there is some dispute in the literature as to whether the nasal valve lies in the nasal vestibule or more posteriorly within the bony cavum of the nose.

The site of major resistance to air flow in the nose has been previously described by Dishoeck (1965) as situated at the junction of the upper and lower cartilages (limen nasi), but there is convincing anatomical evidence which indicates that the isthmus nasi at the pyriform aperture is the narrowest region of the nasal airway (Bachman and Legler, 1972).

The change in airway resistance along the nasal passage can be determined by passing a pressure sensing cannula carefully along the passage and determining the pressure-flow relationships during quiet breathing (Bridger and Proctor, 1970). Using this technique, Haight and Cole (1983b) clearly demonstrated that the major site of nasal resistance lies at the anterior end of the inferior turbinate just within the first few millimetres of the bony cavum as shown. The major site of nasal resistance occurs at the entrance of the piriform aperture. This site of maximum airway resistance agrees with the anatomical evidence and places the nasal valve at the isthmus nasi of the piriform aperture.

The nasal valve is the narrowest part of the nasal airway and the aperture is determined by the position of the inferior turbinate which projects forward toward the piriform aperture and the nasal vestibule. The aperture at this nasal valve region is controlled by the state of engorgement of the inferior turbinate which, in its congested state, can cause complete obstruction of the airway. The significance of the erectile properties of the inferior turbinate has been discussed by Haight and Cole (1983b), who found that the anterior end of the turbinate could advance by as much as 5 mm after application of histamine.

The anterior site of the major component of nasal resistance at the isthmus nasi has important surgical implications as it follows that the main turbinated region of the nasal passage offers very little resistance to air flow. Minor corrective surgery, such as the trimming of septal spurs posterior to the nasal valve region, will therefore have little effect on nasal resistance.
The nasal vestibule, because of its compliant walls, is liable to collapse in response to the negative pressure created during inspiration, and in this way the nasal vestibule can limit nasal air flow and act as a flow-limiting segment or Starling resistor. Bridger (1970) described this flow-limiting segment of the nose as lying in the nasal vestibule between the upper lateral cartilages and the septum. Partial collapse of the lateral wall of the nasal vestibule has been shown to occur when ventilation through one nostril reaches around 30 L/min (Bridger and Proctor, 1970; Haight and Cole, 1983b). The flow-limiting effect of the compliant nasal vestibule occurs only on inspiration and expiratory air flows are not limited in this way.

The resistance of the turbinate nasal passage downstream from the nasal vestibule on inspiration does not normally limit peak nasal air flow (Proctor, 1977). This down-stream resistance on inspiration has only a minor contribution to the overall nasal resistance, as can be seen from the slight resistance change over this region.

**Control of nasal resistance**

In the normal subject, nasal resistance to air flow is primarily regulated by the sympathetic nervous system, as sympathetic activity determines the state of engorgement of the venous erectile tissue. In nasal allergy the effects of local mediators must also be taken into consideration and their significance will be discussed later. The venous erectile tissue is particularly well developed over the anterior ends of the inferior and middle turbinates and along the base of the septum.

The mechanism regulating the swelling of the erectile tissue is poorly understood; Cauna and Cauna (1975) described throttle or cushion veins which may control its filling. Wright (1895) and Burnham (1935) suggested that the close apposition of arteries and veins in the bony canals of the turbinates was significant, as arterial dilation would cause compression of the venous plexus draining the erectile tissue and this would lead to swelling of the erectile tissue.

The venous erectile tissue of the nasal mucosa has a dense adrenergic sympathetic innervation (Dahlstrom and Fuxe, 1965) and electrical stimulation of the sympathetic nerves to the nose causes a pronounced vasoconstriction and a marked decrease in the volume of blood held in the mucosa (Angård and Edwall, 1974; Eccles and Wilson, 1974; Malm, 1977). Under normal conditions, there is a continuous sympathetic vasoconstrictor tone to the nasal venous erectile tissue and section or local anaesthesia of the cervical sympathetic nerves which supply the nasal mucosa causes nasal congestion and an increase in nasal resistance to air flow (Beickert, 1951; Stoksted and Thomsen, 1953; Eccles, 1978b). The sympathetic division of the autonomic nervous system has a major role to play in the regulation of nasal resistance as a final effector pathway in various reflexes which will be discussed later.

The parasympathetic innervation of the nasal mucosa is primarily to glandular tissue and stimulation of parasympathetic nerves causes a watery nasal secretion with little effect on venous erectile tissue and nasal resistance (Eccles and Wilson, 1973, 1974). Section of the parasympathetic nerves to the nose as they pass through the nerve of the pterygoid canal (Vidian nerve) has proved successful as a last resort in the treatment of nasal hypersecretion (Golding-Wood, 1973).
The nasal vestibule is surrounded by cartilages which are attached to muscles controlled by the facial nerve. These muscles are important in facial expression as is apparent in facial palsy caused by interruption of the motor pathways in the facial nerve. Activity of the dilator naris muscle and the movement of the alar cartilage can be used as a test of facial nerve integrity in suspected cases of facial nerve damage or compression (Sasaki and Mann, 1976).

Contraction of the dilator naris muscle causes a dilation or flaring of the nostril but, according to Haight and Cole (1983b), this has little effect on nasal resistance to air flow as the major resistance lies at the isthmus nasi at the entrance of the piriform aperture. However, Strohl, O'Cain and Slutsky (1982) report that voluntary flaring of the nostril can cause a 20% reduction in nasal resistance.

The function of the dilator naris muscle and other muscles around the nasal cartilages is probably to stabilize the nasal vestibule and prevent alar collapse during the high negative pressures developed during deep and rapid inspiration. Upper airway negative pressure in the anaesthetized rabbit has been shown to increase the activity of nasal muscles and this observation supports a stabilizing role for nasal muscles (Mathew, 1984). Nasal air flow has also been shown to enhance the activity of the nasal dilator muscle in the cat and this finding suggests that nasal air flow is an important stimulus in the control of upper airway accessory respiratory muscles (Davies and Eccles, 1984). At peak inspiratory nasal air flows, the pressure within the nasal vestibule falls well below atmospheric pressure and, when this suction effect exceeds the supporting tension of the lateral nasal wall, nasal collapse occurs and flow is limited.

**Measurement of nasal resistance**

**Basic principles**

Nasal resistance to air flow is calculated from two measurements: nasal air flow and trans-nasal pressure as shown. Both these parameters are measured by means of differential pressure transducers and this is why the study of nasal pressure and flow is termed ‘rhinomanometry’, since manometry involves the measurement of pressure. Nasal air flow can be measured by means of a pneumotachograph which consists of a gauze resistance inside a cone-shaped tube. The pressure difference across the gauze generated by air flow through the tube is used to measure air flow. Transnasal pressure can be measured by relating the pressure at the posterior nares to that at the entrance of the nostril which will normally be atmospheric pressure or nasal mask pressure.

Nasal resistance to air flow may be calculated from the following equation:

\[
R = \frac{P}{V}
\]

*R* = resistance to air flow, in cmH₂O/L per s or Pa/cm³ per s  
*P* = transnasal pressure, in cmH₂O or Pa  
*V* = nasal air flow, in L/s or cm³/s.
This equation is a compromise which has been generally accepted by rhinologists and it does not take into consideration separate components of laminar and turbulent air flow (Clement, 1984).

Rohrer (1915) described an equation which allowed for both components of laminar and turbulent air flow in the respiratory tract, but his conclusions concerning the significance of laminar air flow in the airways have been shown to be in error (Williams, 1972; Hey and Price, 1982). For the greater part of the respiratory cycle, nasal air flow is turbulent and this turbulence aids in mixing the air and facilitates the exchange of heat and moisture. Nasal air flow shows evidence of turbulence when transnasal pressures exceed 40-80 Pa and therefore laminar air flow would only be found when dynamic pressure and flow values are close to zero (Hey and Price, 1982).

A plot of the dynamic relations of transnasal pressure and flow on an x/y plotter shows a curvilinear relationship as illustrated. Nasal air flow rises with increase of transnasal pressure, but at the higher pressures there is a limitation of flow due to the increased frictional effects of turbulent air flow. The flow-limiting effect of nasal alar collapse is only apparent during rapid or close to maximum inspiratory manoeuvres.

The curvilinear relationship between transnasal pressure and flow means that one cannot simply determine nasal resistance from the slope of the graph, as would be the case with a straight line relationship. The slope of the P/V plot varies along its length and, therefore, it is not possible to describe the curve with a single numerical value for resistance. It is, however, possible to define the resistance at any given sample point along the curve and this has been the solution recommended for a standardized measurement of nasal resistance (Clement, 1984).

The positive or negative pressure at the posterior nares results in air flow through both nasal passages. The right and left nasal air flows are normally asymmetrical due to the nasal cycle (see below) and therefore a single pressure value may relate to two different air flows. It is, therefore, sensible to standardize nasal resistance by measuring both nasal air flows at the same sample pressure point rather than by measuring transnasal pressures at the same sample flow point.

Unilateral nasal air flow measured at a sample pressure point of 150 Pa and bilateral nasal air flow measured at 75 Pa have been recommended as universal standards (Clement, 1984). However, the Asian population cannot always achieve these pressures during normal quiet breathing and the lower sample pressures of 100 and 50 Pa, respectively, are generally accepted for nasal resistance measurements in Japan.

It seems likely that, for clinical determination of nasal resistance, measurement of nasal air flow at a sample pressure will become the standard, as nasal resistance meters produced commercially are already adopting this system. Sample pressure points of 150 Pa and 100 Pa for unilateral resistance determination will probably be used by the majority of rhinologists in Europe and Japan.
Total nasal resistance to air flow can be either determined directly using the posterior method of rhinomanometry (see below) or it can be calculated by combining the two separate values of nasal resistance for the two nasal passages as shown in the formula below:

\[ \frac{1}{R(\text{total})} = \frac{1}{r(\text{left})} + \frac{1}{r(\text{right})} \]

where the reciprocal of total resistance is equal to the sum of the reciprocals of left and right resistance.

Total resistance can also be directly calculated from the separate nasal airflows obtained at a sample pressure, e.g., at 150 Pa:

\[ R_t = \frac{P(150 \text{ Pa})}{V_r + V_l} \]

\( R_t = \) total nasal resistance
\( P = \) transnasal pressure at sample point (150 Pa in this example)
\( V_r = \) right nasal air flow
\( V_l = \) left nasal air flow.

When quoting values for total nasal resistance it should be stated whether the value was obtained by measurement of total nasal air flow using posterior rhinomanometry, or whether the air flows of the nasal passages have been measured separately.

The use of a sample pressure point to determine nasal resistance is a compromise, as the single numerical value calculated as nasal resistance only described one point on the P/V respiratory curve.

It is possible to sample the curve at numerous points and obtain an average resistance value with the aid of a microprocessor, and this technique has particular advantages when comparing the resistance values of different segments of the respiratory tract (Cole, Fastag and Niinima, 1980).

Another approach to defining the P/V curve has been described by Broms, Jonson and Lamm (1979) who found that curves from a large sample of subjects could be arranged in radial order and that the curves only rarely crossed each other. From this arrangement of the curves, it was possible to define nasal resistance in terms of a polar coordinate system. This particular system of determining nasal resistance is now commonly used in Sweden.

**Techniques of rhinomanometry**

The determination of nasal resistance involves the measurement of nasal pressure and flow as described above. Active rhinomanometry involves the generation of nasal air flow and pressure with normal breathing. Passive rhinomanometry involves the generation of nasal air flow and pressure from an external source, such as a fan or pump, to drive air into the nose.

Active rhinomanometry can be divided into anterior and posterior methods according to the siting of the pressure-sensing tube.
In active anterior rhinomanometry, the pressure-sensing tube is normally taped to one nasal passage as shown. The sealed nasal passage acts as an extension of the pressure-sensing tube to measure pressure in the posterior nares. With this method, nasal air flow is measured from one nostril at a time and the pressure-sensing tube is swapped from one side to the other. Therefore, the P/V curves and nasal resistance are determined separately for each nasal passage and the total resistance is then calculated by summing the values as shown in the formulae above.

In active posterior rhinomanometry, the pressure-sensing tube is held in the mouth and detects the posterior nares pressure when the soft palate allows an airway to the mouth as shown. Total nasal air flow can be measured from both nasal passages, or by taping off one nostril; the right and left nasal air flows can be measured separately. Total nasal resistance can be determined directly from the total nasal air flow and transnasal pressure with this method. A disadvantage of this method, when compared with the anterior method, is that not all subjects can obtain an airway around the soft palate into the mouth. With some training of subjects using feedback from a P/V plot on an oscilloscope or monitor, it is possible to obtain satisfactory results from about 80% of subjects. The use of a P/V plot is necessary when performing posterior rhinomanometry as this method is more prone to artefacts than anterior rhinomanometry because of pressure changes in the mouth due to tongue movements. The use of a small cone-shaped oral cannula made from a plastic disposable auroscope cone often helps subjects to obtain an airway to the mouth and also helps prevent blockage of the cannula with the tongue.

Passive rhinomanometry involves the direction of an external flow of air through the nose and out of the mouth as shown. The method may involve either measurement of the driving pressure at a constant flow or measurement of the flow at a constant pressure. Passive rhinomanometry is particularly useful if it is necessary to separate the upper and lower airways for experimental work (Bundgaard, Sybbalo and Widdicombe, 1983; 1984).

Active anterior rhinomanometry, using surgical tape to seal a pressure sensing tube into the nasal passage, is one of the most commonly used methods for clinical determination of nasal resistance (Solow and Greve, 1980) and it will probably become the clinical standard for studies on nasal disease.

The following precautions are recommended when determining nasal resistance to air flow.

(1) The use of a face mask is recommended rather than nasal cannulae as the insertion of a cannula into the nostril is likely to cause distortion of the airway and irritation. The face mask should form a soft airtight seal and should not pull on the cheek as this can distort the nasal vestibule and disturb nasal resistance.

(2) Equipment should be routinely calibrated for pressure and flow measurement. A sloping manometer filled with light paraffin can be used to calibrate the pressure and a rotameter and air cylinder to calibrate flow as shown.

(3) A single determination of nasal resistance is unreliable because of possible airleaks around the mask etc. It is, therefore, more satisfactory to take two sets of, say five, readings.
with a change in mask position between the sets. If the two sets of readings are similar then an air leak is unlikely and the mean value of resistance can be calculated.

Nasal resistance meters are now produced commercially and some of the more modern equipment incorporates a microcomputer to process and store the information from the P/V curves as shown.

**Factors influencing nasal resistance**

Nasal resistance to air flow is primarily determined by the state of congestion of the inferior turbinate and the septal wall at the level of the isthmus nasi. The filling of the venous erectile tissue in the mucosa of the turbinate is controlled by the sympathetic innervation and by the presence of local mediators such as histamine. Apart from this nervous and local mediator control, nasal resistance may be influenced by other complicating factors such as a deviated nasal septum or the presence of nasal polyps.

What is the normal range of nasal resistance in healthy subjects? This question has not been properly answered in the literature because of the confusion regarding methodology and standardization of rhinomanometry. However a total nasal resistance of 2-3 cm H\(_2\)O/L per s is generally accepted for the adult with the resistance of each nasal passage varying between 2 and 8 cmH\(_2\)O/L per s. Nasal resistance is inversely related to age with maximum values in the infant at around 12 cmH\(_2\)O/L per s total resistance (Polgar and Kong, 1965) which declines to the adult value at around 16 years of age and then shows only a slow further decline with increasing age (Saito and Nishihata, 1981; Syballo et al, 1986). Unlike other respiratory parameters, such as vital capacity etc, there is no correlation between total nasal resistance and age, sex or height (Saito and Nishihata, 1981).

**Nasal cycle**

The air flow through the nasal passages is normally asymmetrical and most normal subjects show a regular cyclic change in nasal air flow as shown. A nasal cycle is found in 80% of the population, yet most subjects are completely unaware of any changes in nasal air flow because the total resistance to air flow remains relatively constant due to a reciprocal relationship between the nasal passages (Heetderks, 1927; Stoksted, 1953; Eccles, 1978a, 1982; Hasegawa and Kern, 1978b).

Since the nasal cycle was first described in the scientific literature by Kayser (1895), there have been numerous other studies repeating his observations and extending our understanding of this unusual asymmetry of nasal air flow. However, the functional significance of the nasal cycle is still obscure, apart from the fact that the alteration of nasal air flow allows a rest period from the damaging effects of nasal air flow.

The regular changes in nasal resistance are regulated by changes in sympathetic tone to the nasal venous erectile tissue, with the low resistance side having the greatest sympathetic vasoconstrictor tone (Stoksted and Thomsen, 1953; Eccles, 1978b).

There is some experimental evidence from animal studies that the regular oscillations in sympathetic tone are controlled from the respiratory areas of the brainstem with the control
of nasal resistance being closely integrated with respiratory activity (Bamford and Eccles, 1982; Eccles, 1983).

The regular oscillations in nasal resistance associated with the nasal cycle are not disturbed by the congestion or decongestion of a nasal passage or by occluding one nasal passage (Haight and Cole, 1983a; Bende, 1985). Thus it seems that local stimuli such as air flow in the nasal passage do not influence the central rhythm of the nasal cycle.

**Exercise**

Exercise causes a decrease in nasal resistance to air flow which is directly related to the workload for values between 30 and 120 watts (Richerson and Seebohm, 1968; Dallimore and Eccles, 1977; Hasegawa and Kern, 1978a). The typical effects of exercise on nasal resistance are illustrated. As a means of decongesting the nose, exercise is more potent than topically applied nasal vasoconstrictors as it decongests those areas of the nose which may not be accessible to a nasal spray. The reduction in nasal resistance is probably caused by an increase of sympathetic tone to the nasal erectile tissue, as the response is abolished during blockage of the stellate ganglion. Since exercise can cause a decrease in resistance, it is important that subjects are rested prior to nasal resistance readings.

**Respiration**

An increase in arterial carbon dioxide due to rebreathing or asphyxia causes a pronounced nasal vasoconstriction and a reduction in nasal resistance to air flow. These responses are mediated via the cervical sympathetic nerve and are probably initiated by the stimulatory effects of carbon dioxide on central and peripheral chemoreceptors which initiate a reflex increase in sympathetic tone to the nasal blood vessels (Tatum, 1923; Dallimore and Eccles, 1977; Hasegawa and Kern, 1978a).

A decrease in arterial carbon dioxide due to hyperventilation causes nasal vasodilation and an increase in nasal resistance to air flow, probably by the same arterial chemoreceptor reflex as described above (Tatum, 1927; Dallimore and Eccles, 1977; Hasegawa and Kern, 1978a; Babatola and Eccles, 1986).

Oscillations in nasal resistance with a respiratory rhythm have been described in anaesthetized animals, with a slight decrease in resistance during inspiration (Bamford and Eccles, 1982; Lung et al, 1984).

The regulation of respiration and the control of nasal resistance are closely integrated and studies of anaesthetized cats indicate that the vasomotor control area which regulates nasal resistance is situated within the respiratory areas of the brainstem region (Bamford and Eccles, 1982).

**Posture**

Changes in posture can cause marked changes in nasal resistance due to changes in jugular venous pressure and reflex changes in sympathetic tone to the nose.
The change from erect to supine posture causes an increase in total resistance to air flow which may be explained by an increase in jugular venous pressure (Rundcrantz, 1969; Hasegawa, 1982).

On adoption of the lateral recumbent posture, reflex changes in nasal resistance occur so that the dependent nasal passage congests and the upper nasal passage decongests. This partitioning of nasal air flow ensures that the upper nasal passage is responsible for the major component of the nasal air flow. The changes in nasal air flow are caused by a pressure stimulus to the skin, with the axillary region being the most sensitive area for the initiation of the reflex (Rao and Potdar, 1970; Haight and Cole, 1984; Davies and Eccles, 1985).

Reciprocal changes in nasal resistance may be obtained in the lateral recumbent subject by alternatively lying on one side and then the other, as shown. This postural reflex change in nasal resistance overrides any asymmetry in nasal resistance due to the nasal cycle.

These effects of changes in posture must be taken into consideration when measuring nasal resistance in any posture other than the normal seated upright posture.

**Nasal reflexes**

Mild mechanical or chemical stimulation of the nasal mucosa causes sneezing usually associated with nasal secretion and congestion. The afferent limb of this reflex is the trigeminal nerve with the efferent components including respiratory muscles and the autonomic innervation of the nose (Richardson and Peatfield, 1981). The sneeze has not been studied in as much detail as other respiratory reflexes and there is some controversy as to the efficiency of a sneeze in clearing the nose as in many subjects most of the air flow passes through the mouth (Birch, 1959).

**Air and skin temperature**

Warming or cooling of the skin surface can induce reflex changes in nasal mucosal blood flow and there are several detailed studies on this area which indicate that the nasal mucosa responds in the same way as the skin (Cole, 1954, 1982; Drettner, 1961). Cooling of the skin causes a decrease in nasal mucosal blood flow and warming an increase. Therefore the nasal mucosa has a thermoregulatory role and body heat is conserved or lost to the expired air in order to maintain body homeothermy. In this respect the nasal mucosa responds to body cooling or warming in the same way as the skin.

Although the effects of changes in skin temperature on nasal blood flow are well documented, the effects of changes in the temperature of inspired air on nasal blood flow have not been studied in as much detail and the results of these studies are inconclusive (Drettner, 1961; Cole, 1982).

The temperature of the inspired air may vary independently from skin temperature, for example on a cold day the inspired air may be below freezing point but the skin of a warmly clad person may be near body temperature. In these conditions, the cold inspired air causes congestion of nasal venous erectile tissue and an increase in nasal resistance to air flow (Holmes et al, 1950; Takagi, Proctor and Evering, 1969; Cole, Forsyth and Haight, 1983).
However, the effects of cold inspired air on nasal blood flow are uncertain. The nasal congestion associated with cold inspired air can be overcome by the decongestant effects of exercise and this indicates that it is a vascular congestion rather than oedema which causes the increased resistance to air flow (Cole, Forsyth and Haight, 1983).

The increase in nasal resistance to air flow on inspiration of cold air is caused primarily by nasal congestion, rather than an accumulation of nasal secretion. Nasal hypersecretion may accompany the nasal congestion (Holmes et al, 1950) and this secretory response may be accompanied by the condensation of water from the saturated expired air in the nasal cavity.

**Emotional and psychological responses**

The effects of various emotional and psychological disturbances on nasal function have been studied in detail by Holmes et al (1950). In general, acute and chronic emotional disturbances may result in nasal congestion sometimes associated with hypersecretion. An acute decrease in nasal resistance would be found with any stressful stimuli which caused an increase in sympathetic vasoconstrictor tone or an increased release of adrenaline from the adrenal medulla.

**Sensation of nasal air flow**

The subjective sensation of nasal air flow is very important as regards patient comfort, but it is a parameter which is often overlooked by clinicians treating nasal obstruction. In some patients the objective measurement of nasal resistance determined by rhinomanometry may indicate a normal airway, but the subjective impression of the patient is one of nasal obstruction. In these cases there may be some loss of nasal sensation of air flow, perhaps due to nasal pathology or damage to sensory pathways.

Nasal air-flow sensory receptors have been proposed by several groups (McBride and Whitelaw, 1981; Burrow, Eccles and Jones, 1983), and there is evidence that anaesthesia of these air-flow receptors can lead to disturbance of respiration during sleep (White et al, 1985). Experiments on anaesthetized animals indicate that nasal air-flow receptors are present in the nasal vestibule and that the sensory pathway is via branches of the infraorbital branch of the trigeminal nerve (Davies and Eccles, 1984).

The sensation of nasal air flow is markedly enhanced by aromatics, such as menthol, which are frequently incorporated in preparations used to treat nasal obstruction associated with the common cold; these have a marked effect on the subjective sensation of nasal air flow but do not have any decongestant action (Burrow, Eccles and Jones, 1983). The sensitization or stimulation of nasal air-flow receptors by menthol has been shown to enhance the activity of upper airway accessory respiratory muscles and this action may help to prevent upper airway collapse especially during sleep (Davies and Eccles, 1985).

**Nasal challenge**

Nasal challenge or nasal provocation tests involve the administration of suspected allergens directly into the nose in order to determine the nasal sensitivity of a subject to test
substances. In cases where there is some doubt over the results of skin or blood tests for allergen sensitivity, then nasal challenge may provide valuable diagnostic evidence of sensitivity (Mygind, 1978; Davies et al, 1985; Weeke, Davies and Okuda, 1985).

Although nasal challenge tests have often been used for research purposes in the past, their use is not firmly established in clinical practice because they are time consuming and always carry the risk of an anaphylactic reaction. Nasal challenge can be useful, however, to confirm that a positive skin or blood test is clinically relevant, as a control in immunotherapy, and when bronchial challenge of asthma patients is contraindicated (Mygind, 1978). It has been claimed that allergy can be localized exclusively to the nose and be demonstrated by nasal challenge, but not by skin tests (Huggins and Brostoff, 1975); however, Mygind and Lowenstein (1982) state that nasal challenge in patients with negative skin tests has no place in clinical practice, except perhaps for occupational allergy. The usefulness of nasal challenge and rhinomanometry in studying occupational allergy is supported by the results of other researchers who have found nasal challenge useful in the identification of occupational allergens (Okuda et al, 1982; Gervais, Ghaem and Eloit, 1985).

Bronchial challenge can be readily quantified by measuring forced expiratory volume, FEV₁, and a 20% fall in FEV₁, is generally accepted as a positive response. Unfortunately there is no similar standard for nasal challenge.

**Local and reflex effects**

The tickling sensation, sneezing and hypersecretion caused by allergen challenge are due to stimulation of sensory nerve endings in the nasal mucosa. There is now evidence that histamine is the mediator responsible for these responses as they can be reduced by H₁ antagonists, such as chlorpheniramine. From this evidence it has been proposed that there are histamine H₁-receptors on the sensory nerves supplying the nasal mucosa (Kirkegaard, Secher and Mygind, 1983).

The nasal congestion and increase in nasal resistance caused by allergen challenge is due mainly to the local effects of mediators released from mast cells acting directly on nasal blood vessels. Unilateral antigen challenge causes unilateral nasal congestion without any contralateral congestion (Konno, Togawa and Nishihara, 1982; Haight and Cole, 1983a). Thus, the vascular response to mediators can be explained by their local action, whereas the secretory response depends on a reflex activation of parasympathetic nerves to nasal glands, as shown.

Histamine, when administered into the circulation or applied topically, causes nasal vasodilation and an increase in nasal resistance and there is evidence for both histamine H₁- and H₂-receptors on nasal blood vessels (Hiley, Wilson and Yates, 1978).

Histamine sprayed into the nasal cavity induces symptoms which are very similar to those caused by administration of allergen in the sensitive subject, and this, together with the presence of histamine in the basophilic cells of the nasal mucosa, has implicated histamine as an important mediator of nasal allergy (Mygind, 1982). However, there are important differences between the histamine and allergen provocation tests which indicate that histamine is not the sole mediator of nasal allergy. First, provocation with allergen, but not histamine,
gives rise to local eosinophilia, which can be demonstrated in a nasal smear from 1-3 hours to 1-3 days after provocation, and second allergen provocation, but not histamine, increases nasal reactivity (Connel, 1968).

The allergic response is dependent on a complex soup of mediators whose time course of release and activation varies widely; however, the time course of response can often be divided into early and late phases. The response to allergen challenge in the lungs is often an immediate response occurring in a matter of minutes and a late response occurring after several hours; however, this separation of responses has not been conclusively shown in the nose (Richardson, Rajtora and Penick, 1979; Davies et al, 1985).

**Standardization of nasal challenge**

At the time of writing, there is no generally accepted procedure for nasal challenge and, before any progress can be made in this field, the following variables need to be standardized.

The actual allergen preparation itself can be a source of much variability. Purification of pollen extracts may give a more consistent product, but many active substances may be removed. The initial material itself can vary according to how and when and from where it is collected. The final concentration of allergen on the mucosa and the area of mucosa over which it is spread can also determine the level of allergic response (Mygind, 1978).

The method of administering the allergen varies from one centre to another with nasal drops, nebulized droplets, powders and allergen-soaked filter paper discs all being used (Wihl and Mygind, 1977; Konno, Togawa and Nishihara, 1982; Weeke, Davies and Okuda, 1985). It is difficult to compare responses when such different methods are used.

The severity of the allergic response to nasal challenge may be measured in terms of sneezes, secretion, a change in nasal resistance, the levels of mediators in secretion or by measuring the concentration of albumin in secretion as a marker for changes in mucosal permeability (Borum et al, 1983).

Measurement of nasal resistance to air flow alone does not always provide a comprehensive measure of the allergic response but use of a simple instrument such as an inspiratory peak flow meter can give clinically meaningful results (Weeke, Davies and Okuda, 1985).

Once there is some generally agreed standardization of nasal challenge, then it may provide useful diagnostic information in the clinic, but at present it is still very much a laboratory-based procedure.
Chapter 5: Abnormalities of smell

Ellis Douek

Awareness of both the internal and external environment is obtained by special receptor cells and organs. Much of this internal awareness is related to the maintenance of homeostasis in the body and does not necessarily reach consciousness. Yet, no one will deny the general feelings of ill health which a breakdown in this balance will produce. The external sensors providing information about the external environment are, on the contrary, principally related to conscious perception, but there is little doubt that they also provide an awareness of the environment that is not wholly conscious. This aspect is particularly important in the chemical senses and, although still not clearly defined in the human, ought to be kept in mind when considering the chemical senses.

The chemical senses

It is not possible to separate olfaction from the gustatory sense and the chemical response mediated by the trigeminal nerve. First of all, the nature of the stimuli is similar as the stimuli represent the chemical rather than the physical properties found in hearing and sight. Secondly, they act in combination - for instance, perception of flavours relies on all three, although the more subtle elements depend on the sense of smell.

Olfaction

The olfactory mucosa is limited to a region which includes the upper part of the superior turbinate, a corresponding region of the nasal septum and the roof of the nose which is between these.

The olfactory epithelium consists of receptor cells, supporting cells and basal cells. The receptor cell is a bipolar neuron whose distal process carries cilia which project into the nasal cavity - these cilia are assumed to carry on their surface membrane receptors sensitive to odoriferous molecules. The proximal process, which is long and thin, is invested by the basal cells until it perforates the basement membrane and comes into association with the cytoplasm of the Schwann cells. These slender axons come together in the submucosa to form fasciculi which travel in parallel towards the openings in the cribriform plate of the ethmoid. They end in the olfactory bulb which lies on the intracranial side of this structure.

As well as the epithelium, the olfactory mucosa contains Bowman's glands. Neither the nature of their secretion nor their function is properly understood.

In many vertebrates, including humans, the olfactory mucosa has a yellowish colour. This is due to pigment granules found in the supporting cells as well as in those of Bowman's glands. They consist mainly of complex lipofuchsins, but also of carotenoids, and it is the presence of these carotenoids that suggests a similarity to visual pigment, leading to the use of vitamin A in the treatment of olfactory abnormalities. However, there is little evidence of empirical success with this treatment and the theoretical basis is likely to be erroneous.
The place of olfaction in mammals is worth considering because, although its role in man is limited by comparison with other species such as dogs, some functions may have persisted to a degree which has been overlooked. Other functions can also be trained to a wider use in individuals who have lost another sense such as vision.

Many animals live in an olfactory rather than a visual world and olfaction allows them direction both above and below ground. The sense of smell is closely related to food and is necessary for finding it as well as for correct identification and assessment of its edibility. Sensing other animals, whether as prey or as predators is largely olfactory. Sociobiological functions may also be present in man at a muted and unrecognized level. Mammals use olfaction as a means of territorial marking and of appreciating their mates’ sexual condition. Ranking in animal groups is often related to smells as is bonding between parent and offspring. These functions are in fact a form of communication between animals of the same species and probably have a place in humans at conscious or subconscious levels and may explain the wide use of artificial odorants. The nature of the odorants involved is the subject of important work at present and it seems that in many species a chemical emitted by one animal results in dramatic physiological changes in another. The mode of action, as a type of external hormone, has led to their being given the name ‘pheromone’. Although no evidence has been obtained in humans and the higher primates of a direct effect of olfactory stimuli on the neuroendocrine system, it is possible that a subthalamic olfactory projection may provide access to the part of the brain associated with emotional behaviour. The relation between olfactory symptoms and disturbed psychological and psychiatric patterns has long been known, but the specific mechanism has not been elucidated.

In lower animals pheromone effects are striking. The smell of male urine results in a lengthening of the oestrus cycle in the female of the same species; oestrus can quickly be induced by an odorant in male urine. This is known to be a small androgen-dependent molecule. It is species specific and not only can it synchronize ovulation in a group of females, but it also blocks an early pregnancy from a different male. No pheromonal effects have, however, been shown in man.

**Classification of symptoms**

Patients may simply complain that they have lost the ability to smell. Alternatively complex and bizarre descriptions may be presented which can obscure the real pattern of the abnormality. It is important for a classification to be retained in the mind, and for the patient to be questioned accordingly to avoid the vagueness which often impedes a proper understanding when perceptive problems are considered.

The following arrangement may be found to be useful:

(1) quantitative changes
   (a) decreased sensitivity to smells
      (i) anosmia
      (ii) hyposmia
   (b) increased sensitivity to smells
      (i) hyperosmia
(2) qualitative changes
   (a) peripheral type
      (i) local causes
      (ii) anosmic zones
      (iii) single non-discriminating response
      (iv) essential parosmia
   (b) central type
      (i) illusions
      (ii) hallucinations
      (iii) abnormal sense memory.

Patients will generally present with the symptom of being unable to smell and often they will add that they cannot taste. It is only under close questioning that they may agree that they can smell a little or occasionally. This distinction between anosmia, or a complete loss of smell, and hyposmia, some decrease in the sense of smell, is crucial to the prognosis and management.

It is also important to decide whether the abnormality is bilateral or unilateral, as unilateral changes may occur in intracranial lesions. It has been demonstrated that loss of smell need not be to the same degree for every odorant. For instance, testing olfaction with different odorants in patients suffering from allergic rhinitis will show relatively normal responses to some and none at all to others. These areas have been called 'anosmic zones' (Douek, 1967). There are also a relatively large number of individuals who have specific anosmias to a particular odour which appears to be genetically determined.

Another hyposmic response in fact represents a type of anosmia. This is a single non-discriminating response to odours and means that the patient may experience a fleeting olfactory sensation of slight degree, but that it is the same smell whatever the stimulus. There is no doubt that this symptom suggests an invariably bad prognosis. It occurs commonly in those patients where the olfactory neurons have been damaged during a viral upper respiratory infection.

The causes of decrease in olfactory sensitivity can be divided into two broad groups as shown below.

**Abnormalities of structures not directly related to the olfactory organ**

Structural abnormalities, such as a deviated nasal septum, practically never cause anosmia or hyposmia in isolation, so that septoplasty invariably fails if offered as a treatment.

Allergic or vasomotor rhinitis, on the contrary, commonly causes abnormalities of smell with related problems of taste. Rarely, total anosmia is present, but the typical features of these conditions are a fluctuation in the degree of hyposmia and the presence of residual perception of smell. On testing, differences to different smells can be found creating the anosmic zones which are often present in allergic rhinitis.

Loss of smell in chronic sinusitis and sinusitis of infective origin is not dissimilar to that of nasal allergy and fluctuation is obvious in response to treatment. Atrophic rhinitis is
not a specific cause of loss of smell, but the infected crusts may set up changes producing a diminution in smell. Specific rhinitis as from, for example, syphilis, sarcoidosis, leprosy, scleroma, and tuberculosis, does not cause loss of smell except in so far as obstruction of the airway is produced. The same applies to tumours involving the nose and paranasal sinuses, but in those conditions other nasal symptoms are more apparent and olfaction hardly dominates the picture.

Nasal polyps should be mentioned specifically as they produce quite severe loss of smell. The cause is simple obstruction which can be corrected by removal, but also the specific effects of allergic rhinitis which cannot be corrected.

Misuse of intranasal medication can affect the sense of smell as it produces a rhinitis, but the effects are by no means permanent if properly treated.

Abnormalities of the olfactory organ and its central connections

Damage to the olfactory organ itself occurs rarely. When this happens it is associated with an influenza-like illness and recovery never takes place. On questioning, the patient frequently recognizes the fleeting non-discriminating olfactory response, but the prognosis is inevitably poor.

Tumours of neuro-olfactory origin are very rare but are reported from time to time. Gross appearance is similar to nasal polyps and the diagnosis is histological although they tend to be more vivid in colour than benign polyps.

The effects of ageing on smell have recently received more attention. Interest arose when a study (Chalke and Dewhurst, 1957, 1958) showed that more than 70% of domestic fatal accidents occurred in persons aged more than 75 years, but that an increasing number of accidents were due to gas. It was suggested that one cause was inability to smell gas. Hinchcliffe (1962) reviewed perception in old age and suggested that all sensory modalities including olfaction show an exponential decrease with age, calling this a fundamental 'presbypsychic' law. The most important study (Van Toller, Dodd and Billing, 1985) showed that by using pure, single odorants which could give repeatable results, there was indeed and decrease in sensitivity with age for 10 odorants. There is also a qualitative loss centred on the primary notes of odorants with decrease in discriminating ability. Interestingly, however, anosmia does not appear to be a feature of old age and no differences between male and female and between smokers and non-smokers were detected.

Abnormalities of smell will occur when interference with the central connections of the olfactory organ takes place. This is common in intracranial lesions and also in association with psychological disturbance. These will be considered separately.

**Intracranial lesions**

These may be related to trauma or to tumours and can be found in epilepsy.
Trauma

Anosmia not infrequently follows a head injury and, although severe injuries are more likely to cause anosmia, even minor ones can produce this deficiency. Frontal injuries are more common, but occipital blows are more likely to produce anosmia.

Recovery, if it takes place, does so during the first few weeks after the injury. After that the prognosis is invariably poor. Only a minority of cases are unilateral, but this is always uncertain as complaints are less likely. Decrease in smell without complete loss is uncommon and usually if the patient still complains of loss of smell after about three months, this indicates a total and irrecoverable anosmia. A small number of cases develop parosmia, where smells are severely altered and sometimes are experienced spontaneously. These smells are generally unpleasant and the prognosis is poor often contributing to depression.

The causes of post-traumatic anosmia are still a matter for speculation, but tearing of the fine olfactory fibres as they pass through the canals of the cribriform plate is likely especially in fractures of that region and where there is a cerebrospinal fluid leak. In contrecoup injuries a shearing tear of the olfactory nerves is possible. Compression of the tracts and bulbs from oedema or blood clot must also occur.

Tumours

Direct pressure on the olfactory nerves or tract raises the olfactory threshold but does not appear to prolong olfactory fatigue, whereas supratentorial, intracerebral lesions will do so on the side of the tumour, both effects may be present simultaneously.

Osteomata growing from the inner table of the skull or the paranasal sinuses grow slowly but unilateral loss of smell may appear as the first symptom.

Meningiomata, particularly of the olfactory groove, may also present in this way.

Frontal lobe tumours usually present other symptoms including intellectual deterioration together with visual disturbance and headaches. Occasionally focal convulsions occur. Anosmia can also be a feature. Tumours around the optic chiasma can cause disturbance of smell as well as visual defects. Temporal lobe tumours do not cause anosmia but there may be some impairments and fits with olfactory aura can occur.

Epilepsy

Abnormalities of smell are found in temporal lobe epilepsy. Olfactory aura are rare but when they occur the hallucinations tend to be unpleasant. They have been described as organic in nature, such as a smell of putrefaction or faeces, or chemical such as petrol or ether. Sometimes there is a smell of burning and only occasionally is it pleasant and perfumed. Usually this smell comes from outside the patient and only rarely may the symptom represent the whole seizure. Generally, however, there are other features such as emotional effects - rage and anger or anxiety and fear. Memory changes such as déjà vu may be associated with the olfactory aura as well as motor and sensory phenomena though the most common are gustatory.
Although some patients have described more prolonged olfactory changes after seizures, these do not seem to be permanent.

**Psychogenic disorders**

There is no doubt that the abnormalities of smell are present in psychiatric illness and are also related to powerful emotional changes which verge on disorder.

Abnormal sense memory has proved an interesting phenomenon in many ways. The sense of *déjà vu* which can occur in epileptic aura sometimes involves a smell. Occasionally a sense of *jamais vu* is described when visual, auditory and olfactory experiences appear, surprisingly, never to have been experienced before.

There are many literary allusions to abnormal sense memory, but the most striking is that of Marcel Proust at the beginning of his great work. It starts with the dipping of a small cake, a *madelaine*, into a cup of coffee and the smell experienced by bringing it up to his mouth evokes memories of childhood. These are so detailed that they lead to the 13 volumes of *A la Recherche du Temp Perdu*. Obviously, the author is using this as a literary device, but it represents a well known phenomenon. Although the recall which smell produces cannot be called a disorder, it can sometimes trigger emotions of fear and anxiety which lead to a real disorder.

More common abnormalities are those which can be called illusions or hallucinations. The distinction between the two is not always clear. Illusion may involve alteration of smell, almost invariably for the worse and sometimes carrying a powerful emotional content. Hallucination is the experience of a smell when none is present.

There is no specific abnormality for a specific disorder, but abnormality has been described in schizophrenia as well as in alcoholism, senile dementia and depression. Hallucinations of smell become built into the delusional system of schizophrenia together with auditory hallucinations.

Olfactory hallucinations also occur in depressive illness sometimes associated with delusions of guilt and shame.

There is a group of people who develop a belief that they have a bad smell. This may emanate from the mouth or represent body odour. Although this is a delusion, these patients do not show any obvious sign of psychiatric illness otherwise. They fall nevertheless into a particular group, as the majority are under 30 years of age and they present as shy, embarrassed and withdrawn persons. They are not infrequently referred for an otolaryngological opinion and the diagnosis has to be differentiated from true halitosis. In the latter condition the complaint comes from other people such as the patient's relatives and the individuals themselves state that they are not aware of an abnormal smell at all. This distinction is not always simple as olfactory hallucinations usually give rise to delusions regarding the environment, and the patients will describe the behaviour of other people as responding to their own bad smell. Referral for psychiatric treatment is often resisted and, when accepted, not usually successful.
An interesting historical reference is that of Louis XI of France who is described as always being conscious of a bad smell about him. This may have been associated with paranoid features as his personality was deeply suspicious and cunning, leading to dangerous and fatal repercussions on those around him.

There is a similar, but lesser, disturbance which relates more to taste than smell and which could be confused with it. It seems to affect mainly women between the ages of 40 and 55. It presents principally as a bad taste in the mouth and sometimes gives rise to anxiety as to whether others are aware of it in the form of halitosis. Reassurance here is necessary after exclusion of a dental or gingival problem as the symptoms are usually self-limiting.

Both hysteria and malingering occurs from time to time after nasal operations, and very much less frequently after head injuries and fractures.

**Testing the sense of smell**

Compared with the advances made in testing auditory and visual acuity, quantifying the sense of smell still poses a difficult problem. There are two major difficulties which make quantification difficult. The first is that, as opposed to our knowledge of the nature of sound and of light, the stimulus which produces smell is only partially understood. The second is that it has not been possible, as yet, to devise satisfactory techniques of recording objective responses.

**The nature of the olfactory stimulus**

There is now no doubt that the stimulus producing a sensation of smell and flavour, whether olfactory, trigeminal or gustatory, is chemical in nature and that the olfactory sense depends on odorant molecules.

An odorant may be pure, representing a single molecular type, or it may be a mixture. Naturally occurring odours are usually mixtures and rose oil, for instance, contains more than 400 different types of molecules. The relative number of these molecules decides the particular quality of the oil. The difference between pure chemicals and mixtures makes a considerable difference when testing the sense of smell.

There are some common features of odorant molecules: the molecular weight usually lies between 20 and 300 and the molecules are relatively apolar. Consequently they are relatively less soluble in water and more soluble in both the lipid phase of membranes and in the hydrophobic binding sites on proteins. There is usually a single polar group which gives the molecule a certain orientation.

There is also a definite correlation between the shape and size of the odorant molecules and the quality of their smell. The most important demonstration of this was that of Amoore (1962).
Tests of smell

These are continuously being proposed with an increasing degree of refinement but they fall within certain types.

Threshold tests

These can be performed by dilution in air, in an inert powder or on paper, but nowadays the most commonly used tests are by dilution in liquid. The nature of odorants has developed from complex mixtures to single chemicals. A useful technique is that recently proposed by Van Toller, Dodd and Billing (1985). They have used the following 10 pure odorants:

1. trimethylamine - fishy
2. 1-valeric acid - sweaty, cheesy
3. phenylethanol - rose water
4. 2-acetyl pyrazine - roasted popcorn
5. methone - minty
6. 2-isobutyl-3-methoxy-pyrazine - green pepper
7. 5-alpha-androst-16-en-3-one - urinous
8. acetic acid - vinegar (trigeminal)
9. dodecylmercaptane - petrol-like
10. musk ketone - musky.

All of these were in smelling bottles and musk was on a perfumer's smelling strip.

The kit is portable in 20-mL vials packed in boxes of 10 rows of five bottles each. They were in aqueous solution except for the dodecylmercaptan which was dissolved in diethylphthalate.

The technique used for measuring threshold was that employed by Amoore (1970). This requires that subjects sniff five bottles in each concentration row of the odorant to determine which two contain the odorant. The other three only contain the solvent. It can be called the 'two-out-of-five-forced-choice' method. Subjects first tried sample series and then the test proper is carried out with the record marked on a score sheet.

Odour quality

In Van Toller, Dodd and Billing's test the subjects, using standard concentration, had to place them on a scale of descriptor terms. Two scales were used - one bipolar and one monopolar.

Olfactory spectrogram

This technique attempted to combine a qualitative and quantitative element in a rapid test (Douek, 1967).
The odorants used were based on Amoore's description of the smells of the seven most common (primary) odours: ethereal, camphoraceous, musky, floral, minty, pungent, putrid. The dissolved substances were placed in bottles and the air above the solutions was blasted into the nose by a syringe.

The volumetric quantity required to produce a sensation of smell was recorded on a block graph for each substance. Although the mixtures were complex rather than pure, and the blasting technique introduced many variables, the approach proved valuable in a clinical situation as it compared one nostril with another, one patient with another, and could record the progress of one case. It allowed the recognition of 'anosmic zones' which could recover with treatment in conditions such as allergic rhinitis.

In cases where hysteria or malingering is suspected, it is necessary to test the patient with trigeminal stimuli, such as dilute ammonia, acetic acid or menthol, as often they will deny any sensation.

It is also important to test the sense of taste as they will frequently deny that sensation also.

**Management of abnormalities of smell**

There are few useful forms of treatment and diagnosis should be based on the description below.

**Exclusion of the intracranial lesions**

This includes the history and examination of the cranial nerves as well as the nose. If there is any question of such a lesion computerized axial tomography is required.

**Recognition of sinus disease**

This requires sinus X-rays as well as nasal examination.

**Recognition of peripheral causes**

Commonly this is related to nasal abnormalities such as nasal polyps, chronic sinusitis and allergic rhinitis. Diagnosis is from examination and sinus X-rays.

A particularly important feature in these cases is the presence of residual experiences of smell even though these may be transient.

If the cause of the abnormality is an intracranial lesion there is no useful treatment. If the lesion appears to be peripheral there is a much better prognosis and the treatment should be intensive. Sometimes surgical intervention, such as the excision of polyps or sinus drainage, is necessary as a first step and this often should be followed up by treatment for an underlying allergic rhinitis.
Chapter 6: Rhinitis, sinusitis and associated chest disease

Ian Mackay and Peter Cole

The lining of the nose and paranasal sinuses is continuous and it would be rare for inflammation to affect one without the other. In addition, the symptoms of sinusitis, particularly chronic sinusitis, and rhinitis overlap to a large extent and it is therefore convenient to consider the two together as rhinosinusitis. Similarly, the upper respiratory tract (nose and paranasal passages) and lower respiratory tract (tracheobronchial tree and alveoli) are closely related, not only in anatomy, but also in a number of physiological functions and responses to the environment. Chest disease is therefore frequently associated with rhinosinusitis and both the otolaryngologist and chest physician may see principles of aetiology, pathogenesis, investigation and management of diseases of one tract reflected in the other - with instruction and advantage to each. The aim of this chapter is to consider the aetiology, differential diagnosis and medical treatment of rhinosinusitis with its associated chest diseases.

The complex interrelationship of rhinosinusitis and the chest is well exemplified by the presence of sinusitis in association with bronchiectasis, which was noted many years ago (Quinn and Meyer, 1929), acute exacerbation of chronic bronchitis (Ogilvie, 1941) and recurrent bronchitis. A study of 200 patients with chronic purulent sputum production, most with proven bronchiectasis, revealed a high prevalence (42%) of rhinosinusitis (Cole, 1981b). In studies of college students, asthma occurred in 17-19% of patients with allergic rhinitis and 56-74% of patients with asthma were found to have allergic rhinitis (Settipane, 1984a). Nasal polyposis in association with asthma is a well-known finding and of particular interest is the well-documented triad of asthma, nasal polyps and aspiration intolerance. Nasal polyposis in children raises the possibility of associated cystic fibrosis.

The role of allergy is discussed in detail in Chapter 7 but is briefly discussed here as an underlying aetiological factor. Rhinomanometry, radiology, nasal endoscopy, sinus surgery and complications of sinusitis are each dealt with under separate chapter headings while acute coryza and some of the rarer chronic inflammations are considered in Chapter 8.

Aetiology and differential diagnosis of rhinosinusitis

The symptoms of rhinitis include nasal obstruction and mucoid or purulent rhinorrhoea which may be anterior or, if posterior, often referred to as a postnasal drip. In addition, patients may complain of itching, sneezing, snoring, facial pain and headaches, anosmia and associated ageusia, and epistaxis.

Previous descriptions have classified rhinitis as infective and non-infective, the latter being subdivided into allergic and non-allergic. While this may have the advantage of simplicity, it overlooks the fact that rhinitis is often of multifactorial aetiology with considerable overlap of clinical manifestations. Patients with an underlying allergic problem may develop swelling of the lining of the nose and paranasal sinuses, leading to stasis and subsequent infection, but the presenting symptoms of mucopurulent rhinorrhoea, nasal obstruction and facial pain would not necessarily suggest an underlying allergic aetiology. Conversely, not all patients presenting with purulent mucus in the nasal fossae have infection...
- a smear may reveal this to be due to eosinophils, which is crucial to recognize because the patient will often respond dramatically to topical or systemic corticosteroids. The classification used by the authors is summarized in Table 6.1.

**Table 6.1 Aetiology of rhinitis**

(1) Mechanical (trauma, tumour or foreign body)
(2) Allergy
(3) Mucociliary clearance abnormality
(4) Immunity deficiency
(5) Granulomatous conditions
(6) Autonomic imbalance
(7) Hormonal
(8) Iatrogenic.

**Mechanical obstruction**

This may be due to deviation of the nasal septum, the nose or both, enlargement of the turbinates or bulla ethmoidalis or hypertrophy of the adenoids. Mechanical obstruction, for whatever cause, will impair drainage and may result in rhinosinusitis.

Deviation of the nasal septum not only results in obstruction on the side to which the septum deviates, but compensatory hypertrophy of the turbinates on the opposite side may result in bilateral obstruction. This compensatory hypertrophy may block the frontonasal duct and the ostia draining the ethmoids and maxillary sinuses.

**Tumours**

Tumours of the sinuses, both benign and malignant, may cause mechanical obstruction leading to sinusitis. Harrison (1979) listed pain, nasal obstruction, swelling of the cheek and purulent nasal secretions as the commonest presenting symptoms of carcinoma of the paranasal sinuses and nasal cavity, symptoms which all too often are attributed to 'sinus' by the patient or his general practitioner. Once referred for specialist opinion, the possibility of an underlying malignancy should never be overlooked and, in any case with unilateral symptoms and signs, the index of suspicion should remain high and a biopsy taken at the earliest opportunity.

**Foreign bodies**

Foreign bodies in the nasal cavities, particularly in children, may present with nasal obstruction and rhinorrhoea which is usually mucopurulent, sometimes blood stained and occasionally associated with sneezing. Unilateral mucopurulent rhinorrhoea in children should be assumed to be due to a foreign body until proven otherwise. Foreign bodies may enter the nose through the anterior naris (children inserting buttons, beads and any other small objects which conveniently come to hand); the posterior naris, during vomiting, coughing and regurgitation; penetrating wounds and nasal injury; palatal perforation, as in cleft palate; sequestration of bone *in situ* after trauma; and calcification *in situ* to form a rhinolith.
Some foreign bodies appear to be inert and may remain for many years without producing any signs or symptoms. Should a foreign body become imbedded and surrounded by granulation tissue, it may then act as a nucleus for concretion to occur, becoming coated with calcium and magnesium phosphate and carbonate to form a rhinolith. According to Ransome (1979), this process may occur around an area of inspissated mucopus, or even a blood clot. Rhinoliths usually form near the floor of the nose and are radiopaque.

**Dental**

Dental roots and prosthetic materials used for dental filling may be forced into the antrum causing maxillary sinusitis. The danger of fractured dental root perforating the antrum is increased where there is periapical rarefaction secondary to periapical abscess. Wright (1979) stated that first molar roots are more likely to cause trouble than those of the second molar, despite the fact that the roots of the latter are more closely related to the antral floor. The roots of the first molars diverge more widely, increasing the difficulty of forceps extraction and increasing the chance of root fracture. Roots adjacent to edentulous spaces are most likely to be involved and males are affected more often than females.

Radiological evidence of a dental foreign body can sometimes be difficult to obtain. Standard occipitomental and lateral X-rays of the maxillary antrum have only a limited value. Occlusal films and an orthopantomogram may similarly fail to reveal a root once it has been forced into the sinus. If any doubt remains, sinoscopy should be undertaken.

**Allergy**

Allergy is undoubtedly a common cause of rhinitis, so common that many practitioners regard all forms of rhinitis as being of allergic aetiology. Clearly this is not the case (see Table 6.1).

Strictly speaking, the term 'allergic rhinitis' relates to the immediate immunoglobulin (IgE) antibody-mediated hypersensitivity reaction to specific allergens, most allergens being proteins with a molecular weight between 10,000 and 50,000. In practice, however, many allergens causing rhinitis are complex compounds often consisting of a number of immunogenic molecules called antigens. The term 'allergen extract' is thus used, for example pollen, house dust mite etc, each of which contain many antigens, all or only a few of which may induce the IgE response, leading to symptoms. IgE antibodies evoked by exposure to antigens become attached to the surface of tissue mast cells and basophil leucocytes as cytophilic antibodies. Interaction between the cell-bound IgE antibodies and the respective allergens results in the release of histamine and other mediators from these cells, causing the signs and symptoms of rhinitis.

Other mechanisms may result in release of a complex array of inflammatory mediators, for example non-specific irritants (cold air, fumes and dust), as well as ingested acetylsalicylic acid and certain food preservatives and dyes. Because these latter responses are not necessarily IgE mediated, the terms hyperreactivity or hyperresponsiveness are often used to encompass these forms of provocation.
The prevalence of allergic rhinitis varies markedly from study to study - 0.1% (Schwartz, 1952) to 28% (Malmberg, 1979). Part of the discrepancy is due to the difficulty of making a definite diagnosis in a condition where the symptoms may vary from a 'mild stuffy nose' to severe disability.

Skin tests, although helpful in many cases, can be misleading as studies have revealed an atopic skin test positive rate as high as 35% in healthy populations (Mygind and Lowenstein, 1982), and the skin is also a remote target from the nasal mucous membrane. Nasal smears revealed a prevalence rate of eosinophilia of approximately 25% (Malmberg, 1979), but not all patients with eosinophilia necessarily have allergic rhinitis. The term 'eosinophilic non-allergic rhinitis' (ENR) has been applied to a group of patients exhibiting nasal eosinophilia of at least 25% with no history of allergy and with negative skin tests. This group responds well to topical corticosteroids (Mullarkey, 1984).

Nasal secretions with a very high eosinophil count associated with allergic rhinitis may appear yellow or green and the patients may be diagnosed as having infective rhinosinusitis. Conversely, nasal secretions accompanying acute coryza may be clear and mistakenly assumed to be non-purulent, allergic rhinitis.

The distinction between allergic rhinitis and infective rhinitis is not always as clear-cut as it may first appear: the two may, and often do, occur at the same time, with allergy leading to swelling and inflammation of the nasal mucous membranes causing mechanical obstruction which impedes drainage and clearance from the sinuses and allows 'secondary' bacterial infection.

**Mucociliary clearance abnormality**

The mucociliary system comprises the first line of defence for upper and lower respiratory tracts, trapping and removing inhaled microorganisms, allergens and noxious agents.

The nasal vestibule as far back as the nasal valves, is lined with skin and the olfactory region limited to the superior turbinate and corresponding portion of septum is lined with specialized sensory epithelium; the remainder of the nose and paranasal sinuses is lined with respiratory epithelium.

The epithelium comprises four main cell types: ciliated epithelial cells, non-ciliated epithelial cells, goblet cells and basal cells arranged in a pseudo-stratified pattern on an underlying basement membrane.

The cilia appendages of the ciliated cells are approximately 6 microm long, 0.25 microm in diameter and in the region of 100-200 to each cell. In cross-section, each cilium can be seen to have two central microtubules surrounded by a ring of nine doublet microtubules, protruding from one side of which are inner and outer dynein arms composed of ATPase protein responsible for the energy production required for the beating of the cilium.
Both ciliated and non-ciliated cells have 200-400 microvilli each, the number increasing towards the nasopharynx. These microvilli are approximately one-third of the size of the cilia and have a central core of actin filaments. Microvilli are not capable of active movement and their function is debatable but they probably promote ion and fluid transport between the cells and the periciliary fluid, regulating the composition of the latter and overlying mucus (Petruson, Hansson and Karlsson, 1984).

The goblet cells are unicellular mucus-secreting glands found above the basement membrane. They possess well-developed Golgi apparatus and endoplasmic reticulum of mainly granular type, consistent with high synthetic activity. The distribution and density of goblet cells have been extensively studied by Tos (1982) who found the highest density in the inferior turbinate (11,000 cells/mm²) and lowest in the septum (5,700/mm²), the density in the sinuses being in the mid-range but highest in the maxillary sinuses. Generally, there is a higher density of goblet cells posteriorly near the nasopharynx.

In addition to goblet cells within the epithelium, there are multicellular glands deep to the basement membrane in the lamina propria. The anterior serous nasal glands are not thought to play an important role in man. Small seromucous glands are evenly distributed in the mucosa of the respiratory region (8-9/mm²), although in the paranasal sinuses the density is very much lower (0.06-0.47/mm²) (Tos, 1982).

Nasal secretion is a complex mixture containing material secreted by the goblet cells, seromucous glands and lacrimal glands, material transported across the membrane of epithelial cells equipped with microvilli together with microorganisms and condensed water from expired air. Lucas and Douglas (1934) suggested that mucus had to be arranged in two layers for the mucociliary transport mechanism to be effective; a superficial viscid sheet, the gel layer, moving over underlying serous fluid, the sol layer, which bathes the cilia and microvilli. The cilia beat in the low-viscosity periciliary layer at a frequency of 12-15 Hz with a rapid 'stiff-armed' effective stroke during which claw-like projections from the tips of the cilia (Jeffery and Reid, 1975) engage the thick, viscous gel layer to propel this towards the nasopharynx. During the recovery phase, the cilia bend to return entirely within the thin sol layer in a plane at right angles to the effective beat and sweeping across the surface of the cell.

The transport rate of mucus averages about 6 mm/minute but a wide range is found in normal subjects. Slow clearance rates of less than 1 mm/minute have not been adequately explained but would appear to be related in most cases to variations in physicochemical properties of secretions rather than in the rate of ciliary beating. In a few cases, however, deficient clearance may be attributable to genetically determined abnormalities in ciliary morphology and function. Systemic dehydration and certain air pollutants may also reduce mucus clearance rates (Proctor, 1982) and there is evidence for bacterial products reducing mucus clearance by slowing ciliary beating.

In 1933, Kartagener described a syndrome consisting of bronchiectasis, sinusitis and situs inversus, but it was not until recently that this was associated with a genetically inherited autosomal recessive abnormality of the respiratory tract cilia, usually partial or complete dynein arm deficiency (Afzelius, 1976; Pedersen and Mygind, 1976; Eliasson et al, 1977). In this condition, which was initially termed 'immotile cilia syndrome', there is severe
disturbance of the normal pattern of ciliary beating with a very variable degree of immotility (Rossman et al, 1980) so that the preferred term is now 'primary ciliary dyskinesia'. Mucociliary clearance is profoundly impaired resulting in chronic or recurrent infection. Males are infertile because of the basically similar structure of cilia and the sperm tail, rendering the latter dyskinetic also (Pedersen and Rebbe, 1975). The relationship between primary ciliary dyskinesia and dextrocardia is postulated to be due to random rotation of the archenteron when cilia on embryonic cells are not functioning (Afzelius, 1976). The important implication of this is that only 50% of patients with this condition will be suspected by dextrocardia/situs inversus found on chest X-ray or at examination - therefore, one should not exclude the possibility of primary ciliary dyskinesia simply because dextrocardia is not found, that is one of the requirements for Kartagener's syndrome is not present. These patients may simply present with a life-long history of rhinorrhoea and cough.

While slow clearance may result from dyskinesia of the cilia, it is equally likely to occur with physicochemical abnormalities of the mucus. Patients with Young's syndrome (obstructive azoospermia, sinusitis and bronchiectasis or bronchitis) present with infertility, chronic sinusitis and often cough with purulent sputum - reminiscent of primary ciliary dyskinesia but examination of their cilia reveals that these are normal in structure and function. Examination of the semen reveals azoospermia and exploratory scrototomy reveals normal spermatogenesis, but a hold-up of sperm transport down the genital apparatus at the level of the caput epididymis where the sperm are found in a viscous, lipid-rich fluid, thought possibly to be due to a metabolic abnormality of the cells lining the tract (Young, 1970; Hendry, Parslow and Stendronska, 1983). Similar affection of respiratory mucus may account for the viscid sinus and respiratory secretions in this syndrome.

Young's and Kartagener's syndromes are excellent examples of primarily mucus and primarily ciliary abnormalities respectively.

Far more common, however, are secondary mucociliary transport abnormalities due to, for instance, upper respiratory tract infection - the nasal secretions become thick and mucopurulent and are not cleared in the normal manner. Viral infections of the upper respiratory tract damage the ciliated epithelium thereby reducing mucociliary clearance (Wilson et al, 1987a). Certain bacteria associated with chronic sinus and bronchial sepsis (Haemophilus influenzae, Streptococcus pneumoniae and Pseudomonas aeruginosa) have been shown to release factors slowing and disrupting cilia (Wilson, Roberts and Cole, 1985) and, in the case of P. aeruginosa, these factors have been characterized as the low-molecular-weight pigments pyocyanin and 1-hydroxyphenazine (Wilson et al, 1987b). Such molecules slow mucus transport in animal models and are found in human respiratory secretions during chronic sepsis in amounts sufficient to have a ciliary dyskinetic effect in vivo (Sykes et al, 1987).

**Immunity deficiency**

**The defences**

A variety of defence mechanisms eliminate foreign material breathed into upper and lower respiratory tracts. These can be considered under the headings of those 'resident' in the respiratory tract and those attracted from the systemic circulation when the local mechanisms
fail, and in each case, such defences can be divided into those which are non-specific and those which are immunologically specific (Table 6.2). This is an artificial division because, in practice, there is considerable interaction of the various defences, and such is this 'defence in depth' that it is sometimes very difficult to isolate a single defect as being the sole cause of infective problems in the respiratory tract.

Table 6.2 System of defences of the respiratory tract

Local ('resident') mechanisms

(1) Non-specific
   reflexes (cough, sneeze)
   mucociliary system
   epithelial integrity and lining fluid (anatomical barrier)
   antimicrobial substances in lining fluid (e.g., lysozyme)
   pulmonary macrophage

(2) Specific
   immunoglobulin (secretory IgA, IgE)
   lymphocytes

Systemic ('recruited') mechanisms

(1) Non-specific
   serum factors (e.g., opsonins, complement components)
   granulocyte phagocytes
   mononuclear phagocytes

(2) Specific
   serum immunoglobulins (IgG, IgA, IgM, IgE)
   lymphocytes.

Presentations to otolaryngologist and chest physician

Since the upper and lower respiratory tracts share common defence mechanisms, defects in them will be reflected as infective presentation to either otolaryngologist or chest physician, or both. Since the upper respiratory tract brooks a higher rate of environmental attack, the otolaryngologist may see infections first which will later affect the lung. It is crucial that these are recognized to be due to immunity deficiency (particularly those which are treatable) early, that is by the otolaryngologist, because by the time the patient presents to the chest physician with bronchopulmonary involvement, it is often too late for effective treatment. This is because the delicate architecture of the lung is easily damaged irreparably and, even though an immunity deficiency recognized at that stage may still be reconstituted, the lung damage is already irreversible. Attendant chronic disease and predisposition to further infection occurs, even when the immunity deficiency is treated, because of the distorted anatomy disrupting front line defences such as mucociliary clearance.
In general, there are three principal presentations of infection to the otolaryngologist and chest physician which cause problems in diagnosis and management. First, the acute, overwhelming infection usually due to profound immunity deficiency (for example, panhypogammaglobulinaemia; acquired immune deficiency syndrome, AIDS). Second, recurrent, acute episodes of infection with apparently normal periods intervening. Here, there is a prevalence of immunological abnormality (notably selective IgA deficiency) of between 40 and 70% in the lower respiratory tract (higher in recurrent pneumonia than bronchial infections). Recurrent viral upper respiratory infections usually herald such episodes. Third, the patient with chronic purulent bronchial disease associated with upper respiratory symptoms in over 80% of cases and frank chronic purulent sinusitis in over one-third of cases. Here, paradoxically, there is less than 10% prevalence of immunity deficiency but instead a 'vicious circle' of chronic mucosal damage (see below) which demands a particular attitude to treatment (Cole, 1981a). It is crucial that these three types of presentation should alert the otolaryngologist to the possibility of immunity deficiency or progressive form of disease requiring special approaches to treatment if irreparable damage is not to occur - and affect the lower respiratory tract which may prove fatal.

**Immunity deficiencies**

Patients with systemic immunity deficiency (for example lack of antibodies, panhypogammaglobulinaemia) frequently present with symptoms in the respiratory tract because of the intimate association of this tract with the environment and the potentially harmful agents in it. Because the nose and paranasal sinuses are in the 'front line' of the respiratory tract they encounter greater attack from environmental agents, so it is not surprising that rhinosinusitis is frequently the first presentation of such systemic immunity deficiency.

In a series in the Brompton Hospital Nose Clinic, nine of 250 patients presenting with upper respiratory tract symptoms were found to have significant immunoglobulin deficiency (Mackay et al, 1983). Five of these patients with severe panhypogammaglobulinaemia, who had been referred from chest physicians, gave a history of having initially presented to otolaryngologists with infective upper respiratory tract symptoms before developing irreversible lung disease.

The protective role of IgA in the respiratory tract is far from clear. A number of functions have been suggested, but data to support these roles *in vivo* in man are scanty. It has been estimated from blood transfusion screening, that selective IgA deficiency occurs in one in 500-700 healthy individuals (Bachmann, 1965). However, there is no doubt that some patients with severe, recurrent symptoms in the upper and lower respiratory tracts lack serum and secretory IgA and that there is a significant prevalence of this abnormality in patients presenting with infective rhinosinusitis and bronchial sepsis. Whether this deficiency is causal is doubtful - it may be a marker of other associated deficiency of defences.

Oxelius et al (1981) have shown an association of IgA deficiency with deficiency of certain subclasses of IgG (notably IgG2) in children, and it may be that the subclass deficiency is causal, since IgG2 is a particular important antibody against polysaccharide capsular antigens such as those of the microorganisms *H. influenzae* and pneumococcus. It is important that this is confirmed also for adult infective respiratory disease because,
although IgA deficiency is not routinely replaceable, normal human immunoglobulin replacement therapy can reconstitute IgG2 deficiency. Also, IgG2 subclass deficiency will be not diagnosed by routine quantitation of the total major immunoglobulin classes IgG, IgA and IgM, and requires separate estimation of IgG subclasses.

Despite the great advances in this field over the last few decades, it is still by no means rare to find patients who are unusually susceptible to upper respiratory tract infections and to be unable to identify any defect in defences, systemic disease or environmental exposure likely to offer a reasonable explanation (Andersen and Proctor, 1982).

**Chronic infection without classical immunity deficiency (the vicious circle)**

Over 80% of patients with chronic bronchial sepsis (complaining of daily purulent sputum production) suffer upper respiratory symptoms and over one-third have frank chronic purulent rhinosinusitis. It would appear paradoxical that less then 10% of these patients can be found to have classical immunity deficiency, so why do they have persistent infection?

There are a number of facts about these patients which suggest an answer to this question (Cole, 1984). First, they are mostly young (mean age approximately 40 years); second, the majority are non-smokers; third, they are usually wheezy; fourth, their upper and lower respiratory tracts are colonized by predominantly non-invasive microorganisms (for example unencapsulated *H. influenzae*); fifth, they respond to this colonizing microbial load with an exuberant immunological response in 80% of cases; sixth, they may suffer progressive (sometimes rapidly) disease with gross scarring of the airway/sinus mucosa and, sometimes, death due to ultimate respiratory failure. These facts can be explained by a normal short-lived useful host response to eliminate invading microorganisms becoming subverted into a tissue-damaging chronic inflammatory response.

The scheme depicts the situation in which a normal person disposes of an insult to the upper or lower airways by mucociliary clearance in the majority of instances - an acute, short-lived, controlled, useful inflammatory response removing the few insults not so eliminated.

**Normal elimination of foreign material from the respiratory tract**

('virtuous circle')

Health --> 'Attacker' --> Mucociliary clearance occasionally with an acute, controlled inflammatory response --> Elimination of 'attacker' --> Health.

A predisposing damaging insult (for example virus infection) or underlying disease (cystic fibrosis) provides an ecological niche in which front-line clearance mechanisms are less than perfect. This allows microorganisms to loiter in the sinuses/bronchial tree and the ability of some of them to release molecules directly inhibiting ciliary function (Wilson, Roberts and Cole, 1985) enables these organisms to colonize and stimulate the host to continued attempts at eliminating them. The host response is chronic and inflammatory which, unfortunately, is relatively unselective and damages 'bystander' normal mucosal surfaces and tissues leading to progressive tissue damaging disease. This process, termed a 'vicious circle' (Cole, 1984), is the antithesis of acute invasive infection such as acute sinusitis or pneumonia since the microorganism (which is actively invasive in the latter) is relatively passive and the
tissue damage is mediated largely by host rather than microbe. This distinction is important for treatment as will be seen later.

**The 'vicious circle' of chronic inflammation of the respiratory tract**

Failure to eliminate 'attacker' --> Amplified inflammation:
---> Progressive damage to 'bystander' normal tissue
---> Impaired mucociliary clearance --> Release of microbial cilioinhibitory factors -->
Microbial colonization --> Amplified inflammation.

**Granulomatous conditions**

Granulomatous lesions in the nose may induce nasal obstruction and rhinorrhea mimicking rhinitis due to other causes.

On occasion, nasal symptoms may be the presenting feature of multisystem granulomatous disease. Wegener's granulomatosis, sarcoidosis, Churg-Strauss syndrome and polymorphic reticulosis may all be associated with pulmonary and nasal lesions. The Churg-Strauss syndrome is characterized by the presence of bronchial asthma, eosinophilia and vasculitis with necrotizing granulomata (Olsen et al, 1980). Polymorphic reticulosis is a necrotizing lymphoproliferative lesion with a predilection for the upper respiratory tract and lungs, and locally obstructive lesions are diagnosed histologically by a characteristic angiocentric lymphoid infiltrate (McDonald, De Remee and Kern, 1974).

Granuloma formation is a host response seen in the case of certain infective agents which are not cleared by the primary defences of the respiratory tracts, blastomycosis, histoplasmosis, leprosy, rhinoscleroma, tuberculosis and syphilis, all of which are considered in Chapter 8.

Berylliosis produces a granulomatous response similar to sarcoidosis but a history of occupational exposure will raise suspicion of the diagnosis (McCaffrey and McDonald, 1983).

**Autonomic imbalance**

'Vasomotor rhinitis' is a term utilized when the findings fit no other category of disease. It is a convenient term by which to describe disease of unknown aetiology. The term often means different things to different physicians and surgeons, a shortcoming which makes it almost useless (Connell, 1984). 'Autonomic imbalance' is used in textbooks as a cover for basic ignorance in this area (Mygind and Lowenstein, 1982). Despite this criticism, this heading has been chosen to group together various well-recognized clinical entities which cannot readily be classified under any other headings and which logically can be attributed to imbalance of the components of autonomic innervation of the nose and paranasal sinuses.
Autonomic innervation

Sympathetic

This appears to be regulated by the hypothalamus as electrical stimulation of this area of the brain causes nasal vasoconstriction (Eccles and Lee, 1981). The preganglionic sympathetic fibres originate in the thoracolumbar region of the spinal cord from where they pass to the superior cervical ganglion via the vagosympathetic trunk. Postganglionic fibres pass from the superior cervical ganglion to the plexus around the internal carotid artery then via the deep petrosal nerve (which is joined by the preganglionic parasympathetic greater superficial petrosal nerve) to form the vidian nerve, the nerve of the pterygoid canal. The vidian nerve emerges from the pterygoid canal where it enlarges to form the sphenopalatine ganglion, the sympathetic nerves continue without synapsing to the turbinates. Sympathetic fibres also reach the nose from the carotid plexus via the infraorbital and ethmoidal branches of the trigeminal nerve (Eccles, 1982).

Parasympathetic

The preganglionic parasympathetic fibres originate in the facial nucleus of the brainstem from where they pass to form the greater superficial petrosal nerve. This is joined by the sympathetic fibres of the deep petrosal nerve to make up the vidian nerve which passes through the pterygoid canal to relay in the sphenopalatine ganglion. The postganglionic secretomotor fibres continue to the nasal mucosa.

In addition to the autonomic nerves, the nose is supplied by the olfactory nerve (first cranial nerve) to the olfactory epithelium in the uppermost part of the nose, and by the trigeminal nerve which supplies sensation in the upper part of the nose via the anterior ethmoidal nerve, a branch of the ophthalmic division, and in the lower part of the nose by the sphenopalatine nerve which is a branch of the maxillary division.

The activity of the autonomic nerves is influenced by reflexes which may be initiated in the nasal mucous membranes via the sensory nerve supply. This may involve the central nervous system or short reflex arcs: by either route, they will affect the autonomic balance controlling the secretory and vasomotor innervation of the nasal glands and blood vessels. Stimulation of the sympathetic nerves results in the release of noradrenaline which causes vasoconstriction in the venous erectile tissue. Stimulation of the parasympathetic supply causes the release of acetylcholine, resulting in a watery nasal secretion and vasodilatation. In addition to acetylcholine, other neurotransmitters may be involved; the roles of the vasoactive intestinal polypeptide (VIP) and substance P (SP) have been extensively investigated by Anggard (1981).

Stimulation of the vidian nerve in man has been carried out and the results reported by Rucci et al (1984). Histological changes indicated an increase in secretory activity, an increase in vasodilatation of both the deep venous plexus and the periglandular vascular supply and intense degranulation of mast cells, mainly in the vicinity of the glandular and vascular components.
Emotional rhinitis

It has been proposed (Eccles and Lee, 1981) that prolonged exposure to stress could result in failure of hypothalamic control over the sympathetic innervation leading to autonomic imbalance, causing rhinitis. In addition, increased nasal resistance has been reported in response to hyperventilation (Hasegawa and Kern, 1978), a common finding in patients with emotional rhinitis.

Emotional rhinitis may take many forms. One is entirely hysterical where examination and thorough investigation fail to reveal any abnormality: normal rhinomanometry findings in a patient complaining of nasal obstruction at the time of the test being a good example. In this type, the imagined nasal symptoms are usually associated with a multitude of other unrelated symptoms and the nasal symptoms should be looked upon simply as another 'call for help'. Certain cases of postnasal drip may fall into this category and one can sometimes sympathize with the statement that postnasal drip is a figment of the imagination of the patient or his general practitioner.

Other patients will present with a specific history that certain emotional events (public appearances, arguments, etc), will lead to nasal obstruction or even profuse watery rhinorrhea. The parasympathetic pathway and its central nervous systemic connection provide a network for such an occurrence (Connell, 1984).

Emotional factors affecting the nose were extensively investigated by Holmes et al (1950), who biopsied the nose during different periods of emotional conflict. They found that fear produced a 'sympathetic' response with vasoconstriction, while frustration, humiliation and anxiety resulted in a 'parasympathetic' response with engorgement of the mucous membranes.

Hyperreactivity

Autonomic imbalance with parasympathetic overactivity leads to hyperreactive mucous membranes which will respond to non-specific stimuli such as cold air (Krajina, Harvey and Ogura, 1972). Hyperreactivity, the 'twitchiness' of the mucous membranes, can be demonstrated by their responsiveness to intranasal histamine and methacholine challenge. Connell (1968) found that if symptoms could be produced by provocation with a single allergen, the response to the same provocation 2 days later was significantly more pronounced. He named this phenomenon 'the priming effect'.

In addition to parasympathetic overactivity and allergy, infection (particularly viral infection) may lead to the mucous membranes becoming hyperreactive. Postviral hyperresponsiveness and hypersecretion in the lower respiratory tract is well recognized (Empey et al, 1976).

Clement, Stoap and Kaufman (1985) described nasal hyperreactivity as a condition in which there is an overreactivity of nasal mucosa to stimuli of non-specific endogenous (physical stress, mechanical irritation, endocrine stimulation or poor venous return) or exogenous (thermal or mechanical stimulation, humidity or drug induced) origin. Results of histamine challenges in a group of patients with non-allergic perennial rhinitis were compared
with those obtained in a group of normal controls. They found a slight but significant difference in the two groups, although with considerable overlap.

**Nasobronchial reflex**

Recently, nasal-bronchial interactions and the relationships between innervation and reactivity in nose and lower respiratory tract airways have been the subject of much discussion (the subject of 'Cellular and Neurogenic Mechanisms in Nose and Bronchi', supplement no 128 to the European Journal of Respiratory Diseases, 1983, edited by Mygind, Rasmussen and Mølgaard). In particular, possible reasons for increased nasal reactivity in the common cold, allergic rhinitis and perennial non-allergic rhinitis have been suggested by Borum et al (1983) to be an increased number of mediator cells, increased sensitivity of epithelial nerve receptors ('irritant receptors'), increased reactivity of individual gland cells and increase in number of secretory cells. The consensus opinion at present is that a nasobronchial reflex probably does exist in man, although evidence for this is stronger in animals, and that nasal stimulation may cause not only reflex bronchoconstriction, but also mucus secretion in susceptible subjects, although the exact mechanism in relation to type of nasal stimulation is obscure.

**Neuropeptides**

The respiratory tract possesses neuropeptidergic innervation mediated by the release of such peptides as vasointestinal peptide (VIP), substance P (SP), pancreatic polypeptides (PP) and others.

The Dale principle of one autonomic neuron producing and releasing only one transmitter has been challenged in recent years by the immunochemical demonstration of many peptides with powerful biological action, localized to nerves both in the peripheral and central nervous system, which may be present together with classical transmitters in the same neuron (Hökfelt et al, 1980). Autonomic nerve fibres are richly distributed in the nasal mucous membrane and stimulation of these fibres results in major changes in blood circulation and secretions in this lining. These changes are influenced by reflexes such as those stimulated by nasal irritant receptors and their effects have been described above. Studies of the nasal autonomic innervation in cat, rat and man have shown substance P, vasointestinal polypeptide and pancreatic polypeptide to be present in classical pathways. Substance P is found principally in sensory neurons ending in the spinal trigeminal nucleus, around sphenopalatine ganglion cells, around blood vessels and within the nasal epithelium; vasointestinal polypeptide is found in parasympathetic postganglionic cholinergic neurons innervating blood vessels and glands; pancreatic polypeptide is localized to mucosal artery, innervating noradrenergic ganglion cells in the superior sympathetic ganglia.

The nasal mucosa contains peripheral branches of capsaicin-sensitive, substance P-immunoreactive trigeminal neurons around its vascular supply and within itself. This forms a local axon reflex arc arrangement which can induce local vasodilatation and also increased vascular permeability.

Afferent sensory substance P-containing neurons may activate the efferent parasympathetic neurons, which are vasointestinal polypeptidergic and cholinergic, via central
and/or peripheral mechanisms causing subsequent increased vasodilatory effects and secretion. Vasointestinal polypeptide and pancreatic polypeptide adrenergic nerves are juxtaposed around nasal mucosal arteries so the known inhibitory effect of pancreatic polypeptide on this vasointestinal polypeptide-induced vasodilatation may be a physiological control mechanism (Anggard, Lundberg and Lundblad, 1983).

This complex system may account for a number of conditions seen in the upper and lower respiratory tracts whose pathogenetic mechanisms are obscure. It is possible that increasing understanding of this area will lead to an explanation of, and treatment for, several troublesome conditions affecting the nose and paranasal sinuses (for example non-allergic perennial rhinitis; Kurian et al, 1983).

**Hormonal**

There is considerable clinical and experimental evidence to indicate that both male and female sex hormones affect the nasal mucosa. Rhinitis is often associated with puberty, sexual excitement (‘honeymoon rhinitis’), menstruation and pregnancy (Eccles, 1982).

Rhinitis during pregnancy not uncommonly presents a problem to otolaryngologists who may question the desirability of undertaking investigations such as X-rays at this time. In addition, medication requires careful consideration as the basis of this (certain antibiotics and corticosteroids) may be contraindicated and most practitioners would prefer to postpone any surgical intervention requiring general anaesthesia until after delivery, particularly as the nasal disorder frequently resolves spontaneously at that time. Sorri, Hartikainen-Sorri and Karja (1980), however observed that sinus infection was often associated with this rhinitis. Of approximately 2000 pregnancies, 47 patients were referred with nasal symptoms of which 30 were found to have sinus infection confirmed by antral lavage. It was suggested that this relatively high figure of infection might be related to changes in the immunological status and the high rate of viral infection associated with pregnancy (Vesikari, 1975). However, there is another possibility, that the nasal congestion and inflammation lead to mechanical obstruction and interfere with normal nasal mucociliary clearance and other local defence mechanisms, leading to sinus infection in much the same way as may nasal polyps.

In addition to pregnancy, the occurrence of rhinitis has been reported in women taking oral contraceptives high in oestrogen. Toppozada et al (1984) studied the ultrastructure and histochemical changes in the nasal mucosa of 25 females using the contraceptive pill. Although 15 patients developed no nasal symptoms, changes were found in all 25 similar to those in symptom-free pregnant females - glandular hyperactivity, increased acid mucopolysaccharide content of the ground substance and increased phagocytic activity - while the 10 patients who developed nasal symptoms showed evidence of squamous metaplasia, intraepithelial oedema, glandular hyperplasia, histiocytic proliferation and deposition of fibrous tissue. The histochemical reactions were similar to those of chronic hypertrophic non-allergic rhinitis.

Harrison (1957), in a study of patients suffering from familial haemorrhagic telangiectasia, showed that prolonged oestrogen therapy in men produces metaplasia of the columnar ciliated nasal epithelium to stratified squamous epithelium. Atrophic rhinitis is discussed in Chapter 8, but it is interesting to note that it is more common in females and it
has been one author's clinical experience that it is more common following the menopause. Some patients improve when treated with either systemic hormone replacement therapy or topical hormone creams (conjugated oestrogens - equine). Oestradiol in arachic oil 10.000 units/mL has also been recommended (Weir, 1979).

While atrophic rhinitis appears to be more common in females, excessive watery rhinorrhea has been noted in elderly males. Watson-Williams (1952) described a condition called 'old man's drop': seen in men usually well over 60 years of age with profuse watery discharge tending to hang as a drop at the end of the nostril until wiped away. He treated 23 patients with testosterone propionate tablets (placed beneath the tongue, not sucked) 5 mg daily for 3 weeks and reported that '17 patients were completely relieved'. Eight patients were treated with Stovaine lozenges (amylocaine hydrochloride) having a similar appearance and taste and 'all were quite definite that these were ineffectual'. He also added that the course may need to be repeated once or twice a year.

Thyroid hormones are also known to affect the nasal mucosa, myxoedema being associated with large, boggy, pale turbinates similar to those seen in perennial non-allergic rhinitis and biopsy revealing intense oedema (Connell, 1984).

**Iatrogenic**

**Rhinitis medicamentosa**

The aetiology of this form of rhinitis is the abuse, by overuse, of topical nasal decongestants. The patient, for whatever reason, whether it be a transitory upper respiratory tract infection or a long-standing perennial rhinitis, uses a topically acting vasoconstrictor which results in an immediate improvement in symptoms. In the short term, this causes no problem but if the patient should continue to use this, it is possible for a vicious circle to ensue, whereby decongestion is followed by rebound vasodilatation such that ever increasing doses of vasoconstriction are required more and more frequently to ensure an adequate airway.

Ideally, a topical vasoconstrictor for clinical use should act on the cavernous sinusoids to reduce the swelling of the erectile tissue. Naumann (1961), however, demonstrated that decongestants constrict resistance vessels in addition to the exchange and capacitance vessels, resulting in local ischaemia. Mygind (1979) described the local side-effects of long-term use of topical vasoconstriction under three headings:

1. secondary hyperaemia (rebound) which commences a few hours after administration, more marked with adrenaline than ephedrine and possible due to beta-adrenergic stimulation as both alpha and beta-receptors are stimulated by these substances

2. tachyphylaxis - a phenomenon whereby ever increasing quantities of a drug are required to elicit a given response - in this situation, possibly due to lowered alpha-adrenergic responsiveness of smooth muscle

3. local irritation - increased irritability associated with a sensation of burning and dryness in the nose.
Rijntjes (1982) carried out rhinomanometry and both light- and electron-microscopic examination of biopsies from 20 patients who had used topical vasoconstriction at least once daily for a minimum of 6 months. Rhinomanometry revealed an improved nasal airway in 75% of patients 4-6 months after discontinuing the use of decongestants. It was interesting to note that the normal nasal cycle (alternating patency and obstruction from one side to the other every few hours - see below) was not found in patients with rhinitis medicamentosa and this cycle did not appear to return, even 6 months after discontinuing usage. Scanning-electron microscopy and light microscopy revealed a decrease in the quantity and quality of cilia with metaplasia of the mucous membranes to cuboidal cells. The ultrastructure of the remaining cilia remained unaltered.

Some decongestants appear to be more related to rhinitis medicamentosa than others. Naphazoline caused a marked vasoconstriction of long duration and appears to be incriminated more often than the more recently introduced vasoconstrictors such as xylometazoline and oxymetazoline. Adrenaline has a rapid action of short duration followed by considerable rebound. The action of ephedrine is less intense but longer-acting than adrenaline.

**Aspirin intolerance**

Aspirin intolerance may occasionally present with chronic rhinitis (Settipane, 1984b). More often, however, it accompanies nasal polyposis and asthma. Samter and Beers (1968) described the initial presentation as a profuse watery rhinorrhea developing after aspirin ingestion in the majority of patients during their second or third decade. Initially intermittent, it later becomes perennial, frequently followed by nasal polyps and hyperplastic sinusitis. Urticaria is also common and middle-aged patients often develop bronchial asthma.

The incidence of nasal polyps in asthmatic patients with aspirin intolerance is as high as 36% (Chafee and Settipane, 1974). Frequently, patients in this group are found to be skin test negative but to have eosinophilia in the blood or nasal secretions.

The mechanisms involved in aspirin intolerance are not known, but it is possible that inhibition of the cyclo-oxygenase pathway by aspirin may result in preferential lipo-oxygenase metabolism leading to increased production of leukotrienes and slow reacting substance of anaphylaxis (SRS-A) causing bronchospasm. A similar mechanism involving the arachidonic acid pathway and prostaglandins may be responsible for the production of nasal polyps.

**Drugs acting on the sympathetic nervous system**

Alpha-adrenergic blocking agents used in the treatment of hypertension, such as guanethidine and bretylium tosylate, may result in vasodilatation and nasal obstruction, as may methyldopa and reserpine which deplete sympathetic nerve endings of their catecholamine stores (Ariens, 1967).

There is some evidence that beta-agonists such as isoprenaline cause vasodilatation and nasal obstruction (Hall and Jackson, 1968; Malm, 1974) and that beta-antagonists may inhibit the vasodilatation induced by beta-agonists, suggesting that the nasal beta-receptors are of the beta₂ type (Malm, 1974). The clinical effect on the nose of beta-blockers in man, however, remains uncertain.
Nasal stuffiness may also result from drugs used to cause peripheral vasodilatation in the treatment of migraine and peripheral vascular disease, such as the ergot alkaloids, for example dihydroergotamine mesylate.

**Anticholinesterases**

Drugs which inhibit cholinesterase and potentiate the action of acetylcholine, such as neostigmine used in the treatment of myasthenia gravis, may produce nasal obstruction.

**Surgery**

Overzealous surgery may result in atrophic rhinitis. It has long been thought that excessive removal of the inferior turbinates may result in atrophic rhinitis, although recent studies suggest that this may not be the case (Martinez et al, 1983; Ophir, Shapira and Marshak, 1985).

The cause of primary atrophic rhinitis is not known, although numerous organisms have been incriminated: coccobacilli (Lowenberg, 1894), *Bacillus mucosus* (Abel, 1895), *Coccobacillus foetidus ozaenae*, diphtheroid bacilli and *Klebsiella ozaenae* (Henriksen and Gundersen, 1959). In addition to infective organisms, the possibility of hormonal influences have been discussed above.

Whether the cause be infective, hormonal (as described above) or simply degenerative with age, patients frequently complain of nasal obstruction despite the fact that on examination, the nose is patent - indeed in most cases overpatent - as can be demonstrated by rhinomanometry. Surgical implantation of cartilage partially to obstruct the airway has been successful in some cases, although it is not known why the nose should feel blocked when the airway is patent and the symptoms relieved when the airway is partially obstructed (Connell, 1984).

One possible explanation for the observation that some patients may develop atrophic changes following surgery on the inferior turbinates while others appear not to, is that in certain instances, primary atrophic rhinitis may present with the symptom of nasal blockage and the patient undergo reduction of the turbinates resulting in aggravation of the symptoms. This is one reason why preoperative rhinomanometry should be carried out whenever this condition is suspected.

Septal perforations following submucosal resection of the nasal septum or other trauma such as 'nose picking' may be associated with crusting and bleeding, the latter leading to further crusts sometimes associated with purulent rhinitis. It is important to exclude other possible causes such as malignant tumours, granulomata, chronic infections and exposure to noxious fumes or cocaine.

**Clinical features**

Examination of the nose has been considered in Chapter 1 and endoscopy in Chapter 3. These techniques are therefore not considered here. Suffice it to say that in all cases of rhinitis, the nose must be carefully examined and this may require topical vasoconstriction.
If this is necessary, however, it should be postponed until after nasal mucociliary clearance tests and rhinomanometry have been performed if it is intended that these should be done at the same visit.

**Acute sinusitis**

Externally, there may be slight redness and swelling of the cheek spreading to the lower eyelid from the antrum and the upper eyelid from the frontal sinus. In children, ethmoiditis may be associated with marked swelling and abscess formation medial to and above the inner canthus and swelling of the eyelids.

There may be tenderness over the frontal sinus, although this may be confused with tenderness over the supraorbital nerve which can accompany maxillary sinus infection.

Tenderness to pressure over the floor of the frontal sinus immediately above the inner canthus is usually diagnostic of frontal sinusitis and tapping the supraorbital ridge may cause severe pain. This is, however, highly subjective and many patients will complain of pain in these regions if sufficient pressure is exerted even over a normal sinus. On anterior rhinoscopy, there will be generalized swelling and hyperaemia of the nasal mucous membrane. After decongesting the mucosa, it may be possible to see pus extruding into the middle meatus from the maxillary, anterior ethmoidal and frontal sinuses or into the superior meatus from the sphenoid and posterior ethmoidal sinuses.

On posterior rhinoscopy, pus may be seen in the postnasal space and there will be generalized swelling and redness of the mucous membranes.

**Mechanical obstruction**

Bilateral choanal atresia will present at birth because neonates are obligatory nasal breathers. This presents as a medical emergency and, unless an airway is inserted through the mouth, the child will asphyxiate.

Unilateral choanal atresia is less dramatic, however, and a careful examination needs to be undertaken in any case of total unilateral nasal obstruction presenting early in life. The easiest way to start the examination is by placing a cool stainless steel tongue depressor immediately below the child’s nostril. Warm air exhaled through the nose will cause misting on the spatula. If this occurs on both sides, choanal atresia is excluded. If one side is totally blocked, misting will only occur on the opposite side and choanal atresia along with other causes of nasal obstruction will need to be considered.

Anterior rhinoscopy may reveal other causes of nasal obstruction and it is important to exclude the possibility of a foreign body. Where the index of suspicion is high, examination of the nose and postnasal space with a microscope under general anaesthesia may be necessary particularly in small infants who may not be entirely cooperative.

In any patient with nasal airways obstruction, examination should always start with the external appearance of the nose. No amount of surgery on the septum will improve the airway if the problem is due to external deviation of the nose, even if the patient is not
concerned with his appearance. In assessing the septum, the degree of deviation as well as the site of the deviation is important. Cottle (1960) has named five areas of the septum:

- area 1 is the anterior caudal border where the septum may deviate to one side or other of the columella
- area 2 is the region of the valve
- area 3 is the superior mid-portion of the septum
- area 4 is the inferior mid-portion
- area 5 is the remaining posterior region.

This nomenclature has nothing to do with the anatomy of the nose but is important from a functional point of view. Deviation of a small degree in area 2 will be critical, deviation in area 3 may be associated with external deviation of the nose, whereas a slight degree of deviation in areas 4 or 5 may be less relevant. In assessing the site and degree of deviation, one must also take note of any associated pathology, that is, is there any evidence of vestibulitis due to interference with the normal smooth laminar air flow along the sides of the septum? Are there any polyps posterior to a deviation? Many patients will have a deviated septum without symptoms and rhinomanometry may be helpful in deciding how much the patient's symptoms may or may not be associated with the septal deformity.

Mechanical obstruction may also result from hypertrophy of the turbinates (both middle and inferior), from nasal polyps, from tumours - both benign and malignant - and from adhesions following trauma or previous surgery.

Alar collapse is another cause of inspiratory nasal obstruction. Nearly all patients can cause indrawing of the lower lateral cartilages if they breathe in fast enough and this should only cause symptoms if it occurs at physiological flow rates. If one asks the patient to tilt the head backwards a little, the nostrils can be examined while the patient breathes in and out through the nose. Alar collapse may occur in the elderly due to lack of spring in the lower lateral nasal cartilages associated with loss of tone of the facial muscles, in particular the dilator nares. It may occur following rhinoplasty or in association with other problems, such as deviation of the septum in the valve area or a wide columella. This is considered in further detail in Chapter 15.

**Allergy**

There may be redness and excoriation of the skin of the nose following excessive nose blowing and wiping. The eyes may be red and watery in the typical seasonal rhinoconjunctivitis due to pollen allergy. Concomitant coughing and wheezing characteristic of asthma may be present.

On anterior rhinoscopy, the nasal mucous membranes are usually pale, 'boggy', hypertrophic and wet. It can at times be difficult to differentiate between hypertrophic, pale, swollen, 'polypoidal' turbinates and polyps; although the latter are insensitive when lightly probed and can be displaced, while the turbinates remain fixed.
Posterior rhinoscopy will reveal clear watery secretion and hypertrophy of the posterior ends of the inferior turbinates which, at times, can be bluish in colour and lobulated, thus referred to as mulberry turbinates.

**Abnormalities of mucociliary clearance**

Examination of the nose in patients with primary ciliary dyskinesia usually reveals mucopurulent secretions on the floor of the nasal cavity and in the postnasal space. Polyps are seldom seen. The ears should be carefully examined as a high proportion of these patients will be found to have glue ear (Greenstone et al, 1985).

Patients with Young's syndrome (obstructive azoospermia, bronchiectasis and sinusitis associated with abnormally viscous mucus and slow mucociliary clearance despite normal cilia) tend to have a similar clinical picture to those with primary ciliary dyskinesia, although it is interesting that in both groups, the nasal symptoms seldom cause more than mild inconvenience to the patient, and in primary ciliary dyskinesia have been present since birth.

**Immunity deficiency**

Patients with systemic immunity deficiency often present with the signs of (recurrent) acute or acute on chronic infection with mucopurulent rhinorrhea, hyperaemic and swollen mucous membranes and purulent postnasal drip. Infection may also be present at other sites such as ears, chest and skin.

Acquired immune deficiency syndrome (AIDS) may present with otolaryngological symptoms. There are frequently signs of rhinitis with 'granular' mucosa and purulent discharge not unlike the appearance of sarcoid although with less bleeding and crusting. Patients almost invariably develop oral candidiasis and often otitis externa with occasional otitis media. Kaposi’s sarcoma skin lesions can develop at any site but should be looked for in the mouth as well.

**Granulomatous conditions**

On examination, there is frequently severe crusting and blood clot. Granulation tissue may be obvious, although usually one needs gently to lift away the crusts of dried secretions to reveal the underlying granular mucous membranes. Wegener's granuloma is often associated with a musky, unpleasant odour. Wegener's and non-healing granulomata are dealt with in Chapter 18 and the other chronic inflammations in Chapter 8. Millet seed granulomata of sarcoidosis may be occasionally seen in the conjunctival sac.

**Autonomic imbalance**

As one would expect in an ill-defined group such as this, the appearances on examination are variable.

The mucosa may be moist, pale, bluish and swollen, similar to that seen in allergic rhinitis. The main complaint in this group is often rhinorrhea - 'the runners' - while in the other group - 'the blockers' - the mucosa may be swollen but otherwise relatively normal.
Hormonal

In puberty and pregnancy, the nasal mucosa appears swollen and often hyperaemic, although the latter may be related to abuse with topical vasoconstrictors. In both these groups, the primary symptom is nasal blockage and patients have often resorted to proprietary topical decongestants by the time they seek specialist advice. One should take particular care to exclude secondary infection in rhinitis in pregnancy as almost one half of these patients presenting with symptoms suggesting rhinitis may be found to have sinus infection (Sorri, Hartikainen-Sorri and Karja, 1980). The watery rhinorrhoea seen in elderly men may originate from the anterior serous glands as there may be little evidence of rhinitis affecting the mucous membranes generally. Indeed, in many cases, the mucosa may look if anything a little atrophic rather than hypertrophic.

Atrophic rhinitis which is more common in women and may be oestrogen related, presents with a foul-smelling discharge (ozaena). On examination, the mucosa appears thin and atrophic and the nasal cavities are full of hard crust which often yellow. When these are removed there is frequently an underlying green mucopurulent discharge. The mucosa is friable and bleeds easily. Attempts made by the patient to remove these crusts often cause epistaxis resulting in blood clots collecting in the nasal cavities which then act as foreign bodies, causing further mucopurulent discharge.

Iatrogenic

On anterior rhinoscopy, the mucous membranes in rhinitis medicamentosa appear swollen and red. The bright red swollen mucosa which was commonly seen with abuse of naphazoline (Privine) led to this condition being referred to as 'Privine nose'. This is less of a problem with xylometazoline or oxymetazoline. It is perhaps wrong to assume that all patients developing hypertrophy of the mucosa following decongestion do so because of the effect of the drug, for as Connell (1984) pointed out, most patients who abuse topical nasal drugs started to use them because they already had a nasal problem.

This is not true, however, for those patients who become addicted to cocaine and who are unlikely to have had any nasal symptoms prior to their 'snorting'. Cocaine interferes with the uptake of noradrenaline into nerve endings potentiating the action of adrenaline (Eccles and MacClean, 1977).

Short-term abuse with cocaine leads to rebound and a picture similar to abuse with alpha-adrenergic agonists; long-term abuse however leads to severe necrosis due to ischaemia with loss of mucosal lining and underlying supportive structures. There is often a perforation of the nasal septum with bleeding, crusting and foul-smelling discharge. In severe cases, the destruction may lead to gross distortion and external disfigurement of the nose.

Excessive surgical reduction of the inferior turbinates may cause an overpatent nasal airway with severe crusting and underlying mucopurulent discharge (Martinez et al, 1983; Ophir, Shapira and Marshak, 1985).
Investigations

Sinus X-ray

The technique and overall interpretation of radiology of the nose and paranasal sinuses is of such importance that a separate chapter is devoted to this (Chapter 20. It is important, however, to stress here that all patients with nasal symptoms, particularly if these are of long standing, should undergo sinus X-rays and chest X-rays, the former to exclude any possible bony erosion or other radiological changes suggesting the possibility of underlying sinister pathology and the latter to detect signs of bronchiectasis, Wegener's granulomatosis, dextrocardia or some other related chest disease.

Three standard views of the sinuses are normally requested: occipitomental, occipitofrontal and lateral.

Sinus X-rays may reveal no abnormality, mucosal thickening, fluid levels (in which case, a tilted view will be helpful) or total opacity. However, it is important to remember that these X-rays are not infallible. The shadow of the lip may sometimes appear as mucosal thickening, totally clear X-rays will occasionally be taken of sinuses found to contain pus on wash-out and a rate of 9% false negatives (normal X-rays with positive return) was reported by Phleiderer, Drake-Lee and Lowe (1984). The same authors also reported a very high false negative rate with no return on wash-out in 50% of the sinuses reported as showing a fluid level and 47% of those reported as opaque, but only 12% of those sinuses reported as 'mucosal thickening' gave any return, while other authors reported results ranging from 16% (Hinde, 1950) to 60% (Axelsson et al, 1970) in this latter group.

An opaque antrum on X-ray may on occasion be due to thickening of the bony wall (Proops, 1983), while previous Caldwell-Luc surgery results in scarring of the lining which, in most cases, will lead to great difficulty in interpreting the radiological appearance.

Sinus X-rays will frequently be requested by practitioners other than otolaryngologists and a report of mucosal thickening of the maxillary antra is then usually interpreted as 'sinusitis'. It is important to appreciate that the lining of the sinuses and nose is continuous and that many conditions producing swelling of the nasal mucosa will be associated with mucosal swelling in the antra without it necessarily being attributed to 'infection'. Hayfever, for example, with profuse watery rhinorrhoea may even be associated with fluid levels in the sinuses at times.

All this does not mean however that X-rays of the sinuses are of no value, far from it. All patients with nasal symptoms should be referred for radiological assessment and this at times will result in unexpected information, such as bony erosion at the base of the skull, suggesting a postnasal space carcinoma, or opacity of the antrum with bony erosion from tumours of the sinuses presenting with symptoms of rhinosinusitis, although bony changes may be seen even with benign nasal polyps (Lund and Lloyd, 1983).
Transillumination of the sinuses

A small light is used, covered with a suitable glass cover which can be removed for disinfection. The patient is placed in a darkened room and the light placed centrally in the mouth with the lips pursed around the cover. Any dental plate must be removed prior to the examination. The antra will transilluminate causing an infraorbital crescent of light and glowing pupil. This will be equal on both sides, providing the antra are normal and equal in size. Mucosal thickening, pus or other pathology will prevent normal transillumination. This may, however, be difficult to interpret if both sides are involved. McNeill (1963) found a positive correlation between transillumination and antral lavage in 68% of cases. It is interesting to note, however, that he found this to be 15% less accurate than X-ray examination and that even here, in the antra of three cases out of 25 reported as entirely normal radiologically, pus was found at lavage. Ballantyne and Rowe (1949) reviewed 100 cases and found transillumination was often misleading in the diagnosis.

One instance in which it can be helpful, however, is a large cyst in the maxillary antrum. Here, an X-ray will reveal an opaque antrum but transillumination is brilliantly clear. Transillumination of the frontal sinuses is even more difficult to interpret as these sinuses are so frequently of unequal size.

Ultrasound

A transmitter emits soundwaves of high power and short duration which are reflected back to a receiver from the interface between objects of varying acoustic impedance. A probe, coated with electrode gel, is moved over the antrum and the ‘echo’ of the ultrasonic wave is recorded on an oscilloscope which can be photographed with a polaroid camera to produce a permanent record.

In an air-filled sinus, most of the ultrasound will be reflected from the interface between the anterior wall of the sinus and the air filling that sinus, leading to an early peak followed by a flat graph. Variations in this pattern will occur with fluid filling the sinus which will produce a ‘back wall echo’ - a double peak will suggest a cyst and multiple peaks may be seen with mucosal thickening.

Böckmann et al (1982), in a comparison of radiology with ultrasound investigation in diagnosing maxillary sinus pathology, showed good correlation in 95%. Pfleiderer, Drake-Lee and Lowe (1984), in their study of 108 washouts, however, concluded that ultrasound adds little.

Swabs and antral lavage

Gwaltney and Hayden (1982) reviewed 12 studies reporting the frequency of bacteria cultured from the vestibule of the nose of normal individuals and found 40-100% incidence of Staphylococcus epidermidis and Micrococcus, 25-40% Staph aureus, 90-100% diphertheroids and 1% Gram-negative bacteria. From the posterior nares, Streptococcus pneumoniae was isolated in 15-25%, Haemophilus influenzae in 6-40%, Strep pyogenes in 6%, Staph aureus in 12%, Neisseria meningitidis in 4-27% and Gram-negative bacteria in 13%. If many of these
are regarded as normal commensal flora, one is left in serious doubt as to the usefulness of nasal swab for bacteriology in the treatment of infective rhinitis.

The common cold and other respiratory virus syndromes are caused by viruses such as rhinoviruses, coronavirus, parainfluenza viruses and these may in turn lead to sinusitis. More commonly, however, they lead to swelling of the lining of the nose and sinuses, blockage of the ostia draining the sinuses (particularly the ostia of the maxillary sinus), interference with the clearance mechanism and stasis. These factors favour secondary infection by the multitude of bacteria already present. With chronic infection, the sinus ostia may well become totally blocked leading to a negative pressure and low oxygen concentration; this, combined with an impaired blood supply to the nasal mucosa, may explain the high frequency of anaerobic organisms found, which, in one study (Frederich and Braude, 1974), was as high as 43 out of 62 cases in which bacteria were isolated.

In acute sinusitis, the results of nasal cultures correlated poorly with those obtained by proof puncture (Axelsson and Brorson, 1973). The latter are summarized in Table 6.3 (Gwaltney, 1979).

Table 6.3 Organisms isolated from nasal cultures from patients with acute sinusitis

<table>
<thead>
<tr>
<th>Cases</th>
<th>Microbes</th>
<th>Mean(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bacteria</td>
<td><em>Strep pneumoniae</em></td>
<td>31</td>
</tr>
<tr>
<td></td>
<td><em>H. influenzae</em></td>
<td>21</td>
</tr>
<tr>
<td></td>
<td><em>Strep pneumonia +</em></td>
<td>5</td>
</tr>
<tr>
<td></td>
<td><em>H. influenzae</em></td>
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<tr>
<td></td>
<td>Anaerobic bacteria</td>
<td>6</td>
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<td></td>
<td><em>Staph aureus</em></td>
<td>4</td>
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<tr>
<td></td>
<td><em>Staph pyogenes</em></td>
<td>2</td>
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<tr>
<td></td>
<td><em>N. catarrhalis</em></td>
<td>2</td>
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<tr>
<td></td>
<td>Gram-negative rods</td>
<td>9</td>
</tr>
<tr>
<td>Viruses</td>
<td>Rhinoviruses</td>
<td>15</td>
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<td></td>
<td>Influenza virus</td>
<td>5</td>
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<td></td>
<td>Parainfluenza virus</td>
<td>3</td>
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</tbody>
</table>

In patients with cystic fibrosis and other forms of severe chronic sinusitis and bronchiectasis, *Pseudomonas* is commonly found.

Fungal infection is relatively rare by comparison with viral and bacterial infection. It is known to occur in cases of trauma to the face, poorly controlled diabetes, severely debilitated patients such as those with carcinomatosis and patients who have been treated with immunosuppressive drugs (Berlinger, 1985). It may, however, occur in otherwise healthy patients (Meikle, Yarington and Winterbauer, 1985). The commonest fungus involved is *Aspergillus*. 
In many ways, it is surprising that fungal infection is not seen more often, particularly in view of the large number of patients now treated with topical corticosteroids. In the oropharynx and laryngopharynx, however, candidiasis may be seen in 10% of cases treated for asthma with topical beclomethasone.

Nasal smears

Examination of nasal smears for eosinophils was first reported by Eyermann in 1927. Since then, many authors have reported its usefulness in distinguishing between 'allergic' and 'infective' rhinitis. Mygind (1979) gave an excellent account of the technique and its interpretation but commented that 'this simple technique ... has not been accorded the general use it deserves'. This may be because it is not quite so simple.

First, a nasal smear is taken by scraping the mucous membranes with a cotton applicator; this should be taken from as far posteriorly as practical and as much secretion as possible should be collected. This is smeared on a glass slide and dried in air. It is then covered with 18 drops of May-Grunwald stain and, after 30-45 seconds, 6 drops of Giemsa stain are added and left for 30-45 seconds, after which the smear is placed under running tapwater. It is then quickly decoloured with alcohol before placing it once again under running tapwater. The slide is then ready for microscopy. Marked eosinophilia may be found in allergic seasonal and perennial rhinitis, but may also be found in cases of non-allergic eosinophilic rhinitis. It is useful in distinguishing those patients in whom yellow or greenish secretions are due to a very high count of eosinophils rather than infection and in whom treatment with antibiotics would be unhelpful.

Biopsy of nasal mucosa

This should not be undertaken as a routine investigation. Nasal cilia can be harvested by the brushing technique described below which requires no anaesthesia and is without complications. By contrast, taking even the smallest biopsy from the turbinates can occasionally be followed by very troublesome epistaxis requiring packing and even transfusion.

Under local anaesthesia in the outpatient or 'office' setting, it is impractical to take more than a fairly small biopsy and this is often done with aural cupped granulation forceps. The tiny specimen taken, while putting the patient at possible risk of severe epistaxis, is, however, seldom sufficient for adequate histological examination.

If a biopsy is required to exclude Wegener's granuloma, sarcoidosis or a possible malignancy, it is better to perform this as a formal procedure in a well-equipped operating theatre with topical anaesthesia and vasoconstrictors together with locally injected anaesthetic agents. A reasonable-sized biopsy can then be taken with turbinate trimming scissors and the nose packed with an adequate pack which is left in situ for 24-48 hours.

Prick skin tests for immediate hypersensitivity

Skin tests may be performed either by placing a drop of a solution of the extract to be tested on the skin and gently pricking the epidermis with a lifting action, or by
intracutaneous injection of a much more dilute solution of the extract. The prick test is preferred, however, because it is not painful, easier to interpret, much safer, the solutions are more stable and the procedure is quick and easy to perform. The allergen extracts used are solutions in glycerin of the common allergens such as *Aspergillus fumigatus*, *Alternaria*, grass pollen, cat fur, dog hair, feathers, house dust, *Dermatophagoides pteronyssinus* (house dust mite), milk and egg. After placing a drop of the solution on the skin, the superficial epidermis is very gently pricked with a sterile disposable needle or lancet, using a slight lifting motion and without drawing blood. In allergic patients, a weal and flare develops within 10-15 minutes (immediate); less commonly, late reactions (6 hours) may develop.

In addition to the allergen extracts, a positive histamine control (1 mg/mL) and a negative control (allergen-free stock solution) should be used - the histamine in order to ensure there is no factor interfering with a positive result (for example antihistamine therapy), the negative control in case the patient wheals to trauma alone (for example dermatographism).

If the positive control is negative, it is important to repeat the tests after ensuring that the patient abstains from antihistamines for 48 hours. Some of the recently introduced H1-antagonists such as astemizole, may abolish positive skin tests for up to one month. Systemic corticosteroids however do not affect the result.

**Rhinomanometry**

The importance of this measurement is such that an entire chapter (Chapter 4) has been devoted to it. Undoubtedly, it will form an increasingly important part of the investigation of all patients with nasal symptoms.

However, there are many problems associated with this investigation, not the least being that one is measuring a 'moving target'. As the subject breathes in, the nasal apparatus flattens and the faster the air flows inwards, the more the alar cartilages and upper lateral cartilages are drawn inwards, increasing the nasal resistance. Breathing outwards through the nose, the opposite will happen, causing a decrease in nasal resistance. The nose is designed as an air-conditioning mechanism to warm and humidify as well as to purify the air before it passes to the lungs. As such, it is a very effective organ and can adapt quickly to changes in the environment: in order to warm cold air, more blood passes through the turbinates and vice versa - the environment therefore affects the results of any rhinomanometry; added to this, exercise and posture also alter resistance. Finally, even in the same subject resting and without alteration in posture in a stable environment, the resistance will change: alternating from side to side every 3-4 hours due to the 'nasal cycle'.

Anterior and posterior, and both active and passive forms of rhinomanometry are described. From a clinical point of view, the passive method is of little value as it decreases the resistance of the nose as air passes inwards by 'blowing out' the lower lateral and upper lateral nasal cartilage. This is the opposite effect to that which occurs in a physiological situation and would be of no value in investigating, for example, a possible alar collapse.

The posterior method, which has some advantages, in that it may avoid the necessity of applying measuring device to the anterior part of the nose, is regrettably impossible to
perform in 25% of subjects who are unable to relax the soft palate sufficiently (Mackay, 1979). The anterior active method is the method chosen for routine clinical use and suitable equipment is now commercially available.

In 1983, a committee was set up to report on standardization of rhinomanometry (Clement, 1984). At this meeting, it was decided that the method of choice was the active anterior method, that is 'the measurement of nasal air flow and pressure at the nostrils during respiration'. That pressure is recorded via a tube fixed with adhesive tape to one nostril and flow is measured with a pneumotach via a face mask and recorded during quiet breathing in a sitting position. Values are expressed in SI units, pressure in pascals (Pa) and flow in cm$^3$/s, expressing the resistance at a fixed pressure of 150 Pa.

Rhinomanometry is undertaken both before and after decongesting the nose to reduce the effect of the nasal cycle and indicate both the 'reversibility' of any obstruction (see spirometry pre- and post-bronchodilatation in assessing the reversibility of lower respiratory air flow obstruction for example in asthma) and the relative importance of any septal (or nasal) deviation.

Rhinomanometry has three valuable roles.

**Research**

Undoubtedly, this has so far been its most important function. It remains the only method by which one can objectively assess and compare the results of different medical treatment regimens (for example comparison of two corticosteroid sprays in the treatment of hayfever) or surgical techniques (submucosal diathermy versus trimming of the inferior turbinates).

**Reassurance**

Subjective assessment of nasal patency is often paradoxical. In very cold weather, the nasal turbinates will be hugely dilated with blood, attempting to warm the inspired air - the resistance will increase: despite this, the nose may feel clear. This phenomenon was well illustrated by Eccles and Jones (1983) who demonstrated no change in nasal patency when applying menthol to the nose, despite the fact that this was interpreted as a far better airway by the subject, possibly due to the stimulation of the cold receptors in the nose. The consequence of this is that many patients who in fact have a 'normal' airway may complain of nasal obstruction. Here, the importance of rhinomanometry cannot be overstressed, as to treat these patients for nasal obstruction will never be rewarding and may indeed worsen the situation - particularly if the patient's sensation of obstruction is due to relative insensitivity of the nasal mucosa - leading to lack of appreciation of the air flow or stimulation of cold receptors of the nose. To undertake radical or even partial trimming of the turbinates in rhinitis sicca will only aggravate the problem and even topical corticosteroids may have a deleterious effect in this situation.

Reassurance and explanation both for the patient and his referring doctor will be more helpful and less harmful; rhinomanometry plays an essential role in this.
Reassessment

When rhinomanometry is undertaken as routine in the investigation of all patients with nasal symptoms, decongesting the nose at the time of initial examination will obviously invalidate the test. This means that an adequate view may not be obtained until after decongesting the nose for rhinomanometry. Patients should therefore be re-examined at this stage. If the added information available from the nasal airway studies does not at first appear to correlate with examination findings, further examination may reveal an obstruction posteriorly which might otherwise have been missed; rhinomanometry in these circumstances acts as a useful 'double-check'.

Sinuscopy

Endoscopy of the nose and paranasal sinuses is dealt with in Chapter 3. It remains the only certain way of investigating the contents and appearance of the lining of the paranasal sinuses, as even a sinus wash-out may mislead by giving a clear return, despite the fact that there may be inspissated mucus or crumbly purulent masses present, as in sinusitis caseosa. Draf (1983), reviewing over 1000 cases in which sinuscopy of the paranasal sinuses was undertaken, concluded that in 20% of cases, it was not possible to ascertain the presence of suppurative maxillary sinusitis by means of X-rays and proof puncture alone. Comparative endoscopic examination immediately after standard irrigation of the antrum without visual control showed that complete removal of the purulent matter from the antrum by this means was possible in only a small number of cases. There was surprisingly poor correlation between X-ray and endoscopic investigation - Draf (1983) found complete agreement in only 42%, moderate agreement in 36% and no agreement in 22% of 301 random maxillary sinus endoscopies with similar findings in the frontal and sphenoidal sinuses.

X-rays, transillumination and ultrasound are not infallible, even proof-puncture can be misleading. Sinoscopy remains the only definitive diagnostic procedure. Like proof puncture, however, it is unpleasant whether undertaken via the canine fossa or the inferior meatus and is not without hazard. It is unlikely that it will ever be undertaken on a routine basis on all patients presenting with nasal symptoms.

A full history and examination, combined with radiological investigation and careful follow-up to monitor response to treatment would appear to give sufficient information in the majority of cases, reserving endoscopy of the sinuses for those cases which are either refractory to treatment or where the former investigations indicate it.

Nasal mucociliary clearance

Nasal mucociliary clearance comprises two major components, thick mucus which is beaten away by cilia (appendages at the luminal border of the ciliated epithelial cells). Transport of mucus in the nose may therefore be affected by abnormalities of ciliary beating in periciliary fluid and those of the physicochemical quality and quantity of mucus. A simple measurement of the transport of mucus is therefore valuable in screening for these abnormalities which may underlie or be secondary to nasal disease.
Quinlan et al (1969) developed a method for measuring nasal mucociliary clearance in man; this involved placing a radioisotopically labelled particle on the anterior nasal mucosa and tracing its clearance with a gamma camera. This method, while having research potential, is not practical in the routine clinical situation. Andersen et al (1974) described a technique in which they replaced the labelled particle by saccharin and this has become the standard method for measuring nasal mucociliary clearance. A 0.5 mm particle of saccharin is placed approximately 1 cm behind the anterior border of the inferior turbinate. It is important not to place it too far anteriorly as clearance here is forwards rather than backwards. The time elapsing until the first experience of a sweet taste at the posterior nasopharynx is recorded as the nasal mucociliary clearance time in minutes. If carefully performed, the test is reproducible (Stanley et al, 1984). Patients should be tested in standard environmental conditions and must be instructed not to sniff, eat or drink and to avoid coughing and sneezing if possible. They should be tested in the sitting position with the head about 10° flexed to avoid the particle falling backwards into any postnasal stream, and should not be told the nature of the particle. The particle should be inserted under direct vision to ensure there is no gross mechanical obstruction to the test. If a patient is unable to perceive the correct (sweet) taste after 60 minutes, it is important to test his ability to taste saccharin placed directly on the tongue as, rarely, persons may lack this ability.

The rate of clearance has a wide range in normal persons. Proctor (1982) using the radioisotopically labelled particle method found that in apparently healthy adult subjects under optimal environmental conditions, the result ranged from 1 to over 20 mm/minute. Nevertheless, the saccharin clearance method is simple, inexpensive and useful as a routine investigation to screen gross mucus transport abnormality. In children, dyeing the particle of saccharin can assist in verifying when the child tastes, the dye appearing from the postnasal space on oral examination. In one study, all but two of 30 healthy controls tasted saccharin within 20 minutes while 28 out of 158 patients presenting with mucopurulent rhinitis did not perceive a sweet taste after 60 minutes or more (Mackay et al, 1983). It is wise to repeat the test if it is abnormal, using the opposite nostril. There was a good correlation between the results of tests in normal volunteers performed 2 weeks apart and in opposite nostrils (Stanley et al, 1984) and, in most, the transport time was 30 minutes or less.

If a patient, in the absence of mechanical obstruction, is found consistently to be unable to perceive the taste of saccharin 60 minutes after the test is commenced (innate inability to taste it having been ruled out by testing the taste of it directly on the tongue), mucus transport is grossly abnormal and further tests are required to indicate whether the ciliary or mucus component of transport is responsible.

Ciliary beat frequency

Having found a grossly prolonged mucus transport time by the saccharin test, it is simplest first to ask whether the cilia are capable of beating normally by obtaining strips of ciliated epithelium from the lateral aspect of the inferior turbinate using a fibreoptic bronchoscopy cytology brush. This procedure requires no anaesthesia and can be safely employed at all ages, if necessary serially. The specimens are transferred to buffered saline by agitating the brush in the solution, and mounted on a coverslip-slide preparation sealed with silicone grease. A photometric method (Rutland and Cole, 1980) is then used to measure the beat frequency of cilia which are kept at body temperature on the warm-stage of a phase-
contrast microscope; light is 'gated' and shone through a small area of cilia from below, the effective 'straight arm' beat of the cilium interrupting the light beam which is detected by a photometer. The electrical signal generated is processed by a ciliary beat frequency analyser (Greenstone, Logan-Sinclair and Cole, 1984) and the beating expressed in hertz (beats/s). The normal range in man for nasal cilia is 12-15 Hz, there being a gradient with slower beating more peripherally in the bronchial tree (Rutland, Griffin and Cole, 1982).

Simultaneous with this measurement of ciliary beat frequency, the percentage of cilia which are immotile can be determined by direct vision as, occasionally, ciliary beat frequency may be only at the lower limit of the normal range but the number of cilia beating is low.

The primary, genetically inherited syndrome which was formerly termed 'immotile cilia syndrome' and which is now named 'primary ciliary dyskinesia' (because the cilia are often not completely immotile), initially presents to paediatricians and otolaryngologists and can be diagnosed using this methodology. However, recently, it has been recognized that cilia obtained from sites of purulent infection beat slower than normal (Wilson et al, 1986). Therefore, as this slowing is a secondary effect due to elastase 'leaked' from neutrophils (Smallman, Hill and Stockley, 1984) and toxins released by bacteria (Wilson, Roberts and Cole, 1985; Sykes et al, 1987), it is important to be sure that primary ciliary dyskinesia is only diagnosed from specimens taken from areas which are not grossly purulent. If necessary, infection may need to be treated and the test repeated.

**Blood tests**

Besides routine investigations such as full blood count with differential white cell count, erythrocyte sedimentation rate, urea and electrolytes, and liver function tests to detect underlying disease predisposing to infection, it is crucial to test immunological host defences (Cole, 1985). This can be done by testing serum for deficiency of IgG, IgA and IgM. In this way, panhypogammaglobulinaemia and selective antibody deficiency (for example IgA) will be diagnosed. This is particularly important in the case of rhinosinusitis, first, because hypogammaglobulinaemia is treatable by immunoglobulin replacement injections/infusions; second, because early diagnosis (and treatment) will avoid irreversible damage especially to the lungs; and third, because these conditions usually present first to paediatricians and otolaryngologists (Mackay et al, 1983). Missing such treatable conditions, albeit uncommon, frequently leads to tragic consequences with either death from acute, fulminating infection or development of irreparable bronchiectasis. Rarely, total Ig classes may be normal but subclasses of IgG may be deficient and require treatment (Stanley, Corbo and Cole, 1984), but estimation of such subclasses usually requires blood to be sent to a special centre.

The role of IgA in the respiratory tract is still much in doubt, although most books tend dogmatically to state its primary role in bacterial disease. Certainly, it has an important antiviral role but its antibacterial effect is not well worked out. Patients with local secretory IgA deficiency can usually be detected by their concomitant deficiency of serum IgA, but it is important to remember that local and systemic systems are distinct and a normal serum IgA does not exclude deficiency or absence of secretory IgA. The latter can be directly tested for in special centres (Stanley and Cole, 1985).
Differential diagnosis of facial pain

Many patients presenting with facial pain will believe they have sinusitis. The pain associated with acute sinusitis has been described above and Chapter 20 is devoted to headaches and facial pain. There follows here, however, a brief outline of conditions which may present with pain mimicking sinus disease.

Dental neuralgia

Dental caries developing infection in the dental pulp will be characterized by poorly localized pain. The affected tooth may be sensitive to thermal change but not to percussion until the periodontal membrane is involved. Apical infection will lead to swelling of the cheek which at times may be marked. The swelling in this region may cause opacity of the sinus X-rays which may be confused with that seen in sinusitis, sinoscopy being required to ensure the diagnosis.

Pain may also occur in the region of an extracted tooth, possibly due to damage of one of the branches of the dental nerve, which may continue for weeks or months.

Trigeminal neuralgia

This common condition of unknown aetiology tends to occur mainly in the second half of life. The pain is unilateral and usually starts in one or both of the lower two divisions of the fifth cranial nerve. Paroxysms of severe stabbing pain may occur spontaneously or be precipitated by minimal movement, such as talking or eating, or occasionally by light touch. Attacks may occur intermittently for weeks or months followed by periods of remission.

Migraine

This pain is usually unilateral, there may be prodromal symptoms (including visual disturbances, such as 'fortification' spectra, nausea, vomiting, hemianopia, unilateral paraesthesia and occasionally even dysphasia) followed by the severe and debilitating headache.

A particular and uncommon variety of migraine which may easily be confused with sinus disease is migrainous neuralgia (or cluster headaches). Classically, the patient is awakened every night with severe pain in and around the eye. The pain lasts about 20 minutes and is accompanied by redness and watering of the eye with swelling of the eyelid. The pain is said to be excruciating and the patient usually gets out of bed and paces the floor. The attacks occur in clusters, lasting a few weeks with intervals of as long as 12-18 months.

Temporal arteritis

This usually occurs in patients over the age of 60 years who develop pain and tenderness over a swollen and tender temporal artery.
Nasopharynx tumour

Tumours in this site may remain asymptomatic for many months. Occasionally, facial pain may be the first symptom. If this is accompanied by conductive deafness, immobility of the soft palate and an enlarged lymph node in the neck, the diagnosis becomes more obvious.

Brainstem lesions

Primary and secondary lesions of the pons and medulla, multiple sclerosis, thrombotic lesions and syringobulbia may cause facial pain. Herpetic and postherpetic neuralgia may also cause pain similar to sinusitis.

Psychogenic facial pain

Psychogenic pain may arise as a manifestation of stress in an otherwise fit person, as a symptom of psychiatric illness such as an anxiety neurosis or depression, or as a feature of a 'built-in' personality trait. Such pain can be divided into a number of different symptom complexes including facial arthromyalgia (temporomandibular joint dysfunction or Costen's syndrome), atypical facial pain, atypical odontalgia and oral dysaesthesia. The pain differs from that of well-defined neurological conditions in that it does not correspond to any cranial nerve distribution and is often bilateral (Feinmann and Harris, 1984).

Facial arthromyalgia causes a dull ache in and around the temporomandibular joint with referred otalgia and occasional shooting pains radiating upwards towards the temple and down into the neck. It has been attributed to bruxism, missing teeth, lax joint ligaments and malocclusions, although Thomson (1971) found the incidence of malocclusion no higher in patients with arthromyalgia than in the general population.

Atypical facial pain is localized to the facial bones or tooth bearing part of the jaws rather than the temporomandibular joint or facial muscles. It is subdivided into atypical odontalgia - pain or discomfort in the teeth - and oral dysaesthesia - burning or altered sensation in the tongue, gingiva, lips or denture bearing area without detectable local or systemic pathology.

Treatment of acute rhinosinusitis

The treatment of acute sinusitis is medical. Antibiotics, analgesics and decongestant medication reduce oedema and increase clearance and drainage from the sinuses.

Antibiotics

Bearing in mind the likely bacteria involved (as described above), oral amoxycillin (or if parenteral treatment is given, the less well orally absorbed but cheaper drug ampicillin) should be considered the drug of choice in treating acute sinusitis. It will be effective against *Strep pneumoniae* and the majority of *Haemophilus* spp which are the commonest organisms involved. It will not, however, be effective against *Staph aureus*, beta-lactamase-producing *Haemophilus influenzae*, some anaerobes and some aerobic Gram-negative rods. It seems reasonable to initiate treatment on an empirical basis without resorting to culture, with
ampicillin 500 mg four times daily or amoxycillin 500 mg three times daily for 10 days (Mackay, 1984). If the patient fails to respond, antral lavage should be undertaken and the washings sent for aerobic and anaerobic culture.

In some cases, the dose given above does not appear to be adequate and one might consider increasing the dose to amoxycillin 3 g twice daily. This can be prescribed in sachets and the patients should be instructed to take them before meals.

In patients known to be allergic to penicillin, co-trimoxazole (trimethoprim 80 mg plus sulphamethoxazole 400 mg) two tablets, twice daily has been shown to be effective (Hamory et al, 1979). Although tetracyclines have been used in the past, pneumococci and H. influenzae are not always sensitive. Cefaclor is effective against H. influenzae (including those that are beta-lactamase positive) and Staph aureus.

Decongestants and anti-inflammatory drugs

The key to successful treatment of sinusitis is to maintain clearance/drainage by ensuring that the ostia are kept patent. Once blocked by mucosal swelling the oxygen content within the sinus will decrease, while the carbon dioxide level increases, particularly if purulent secretions are present (Aust and Drettner, 1974). This encourages the growth of anaerobic bacteria and pneumococci which may be facilitating anaerobes. Low oxygen tension, high carbon dioxide tension and lowered pH hampers the bactericidal activities of polymorphonuclear leucocytes. The purulent secretions and bacteria may also affect ciliary function (as previously mentioned) so that normal clearance is rapidly inhibited and inflammatory products may only be cleared by gravity drainage via the ostia, and any decrease in patency impedes this further.

Systemic decongestants do not appear to be as effective as topical preparations (Anggard and Malm, 1983). Oxymetazoline and xylometazoline hydrochloride appear to be effective and have less 'rebound effect' than other topical decongestants. They should not be used for more than a few weeks, however, or there is a risk that the patient may develop rhinitis medicamentosa.

Anti-inflammatory drugs are particularly effective at reducing the mucosal swelling associated with the inflammatory response of infection. A topical spray (Dexa-rhinaspray) containing dexamethasone, neomycin and tramazoline, a decongestant, can be used in conjunction with systemic antibiotics. Sprays, however, are poorly distributed to the majority of the nasal mucosa and this is particularly true in the blocked nose where the best distribution is obtained from a pipette (Mygind, 1979). In addition to this, if there is any remaining activity of the mucociliary system, the effect will be to transport the preparations away from the sinuses. Logically, therefore, drops instilled into the nose in the head downwards position would appear to be the most effective way of decongesting the ostia of the sinuses. The head down and backwards position has been advocated by Mygind, but radiological studies lead us to believe that the head down and forwards position as shown is the most effective and clinical trials support this (Chalton et al, 1985; Wilson et al, 1987c). Patients are instructed to instill two drops of betamethasone into each nostril and remain in the head down and forwards position for 2 minutes. When the nose is particularly blocked, prior application of a nasal decongestant can improve penetrance.
**Analgesics**

The pain of sinusitis can usually be relieved by aspirin and codeine preparations; it is unusual to have to resort to opiates. Special caution should be taken to ensure that the patient is not allergic to aspirin in view of the association between aspirin intolerance and rhinitis.

Surgical treatment is rarely required for acute sinusitis. Occasionally, however, maxillary sinusitis does not respond to conservative measures, in which case lavage may be indicated and, when this is required, it would appear to be beneficial to repeat it at frequent intervals to remove pus, restore ciliary activity and ventilate the sinus. Drettner (1983) has described a method whereby a plastic tube can simply be introduced via the cannula into the antrum. The tube has a 'memory' so that it curls into a spiral shape unless it is straightened out with a trocar inserted along its length; once this is removed, the tube will curl and remain in situ for as long as necessary, allowing repeated and frequent irrigation without new punctures.

The frontal sinus may occasionally require surgical drainage by an incision made immediately below the inner margin of the eyebrow together with an opening made with a burr. A silicone drainage tube can be inserted into the sinus via the wound and sutured in situ.

Sydow, Axelsson and Jensen (1982), compared 27 different modes of treatment for sinusitis. Each group consisted of between 34 and 74 patients. In all, 1320 patients (2039 maxillary sinuses) were treated and assessed both on clinical grounds and radiologically. One group was treated with topical nasal decongestants alone, another with only systemic decongestants, a third group by irrigation of sinuses alone, three groups were treated with antibiotics alone (amoxycillin, pivampicillin and axidocillin) and the remainder with penicillin V, lincomycin, methicillin, doxycycline, spiramycin, ampicillin, cephradine, erythromycin and bacampicillin, all with or without irrigation, nasal decongestants or both. Their conclusion was that the therapeutic outcome differs very little among the groups, although those patients treated with nasal decongestants alone (both topical and systemic) appeared to do least well. It is also interesting to note that Mann et al (1981) reported a 79% spontaneous cure after 2 weeks in patients treated with analgesics alone.

**Treatment of chronic rhinosinusitis**

The principles involved in the treatment of chronic rhinosinusitis are first to attempt to identify and treat the underlying cause and second, if possible, to restore the inflamed mucosal lining to health.

Whatever the underlying cause, logical treatment is required to interrupt the 'vicious circle' (see above) of amplified inflammation which results in progressive tissue damage in the long term. There are a number of points on the vicious circle where treatment may be aimed, notably, at the colonizing microbial flora using antimicrobial agents, at the tissue-damaging inflammatory response to this flora using topical anti-inflammatory agents and at the compromised mucociliary clearance. The latter is perhaps the crux of treatment (Sykes et al, 1986) for if the sinuses can regain normal clearance, this defence mechanism will tend to prevent recurrence of infection in the normal manner. There is a close parallel between
chronic rhinosinusitis and bronchiectasis and similar principles of management apply, the
overriding one being to achieve either normal mucociliary clearance or the less desirable
substitute, artificial drainage, which is gravity assisted postural drainage in the case of
bronchiectasis but surgical antrostomies in sinusitis. Occasionally, the mucosa may be deemed
to be irreversibly damaged and complete removal of the lining will be the only alternative.
This will, however, be replaced with scar tissue which can never restore normal clearance
function and is therefore always vulnerable.

Such an example raises an important practical aspect of treatment of chronic sinusitis
which contrasts with that of the acute disease. Probably, the majority of patients with
established chronic sinusitis have at least some irreparable damage. In practice this commits
most of them to long-term treatment. The advent of safer antimicrobial and anti-inflammatory
therapy is forcing the medical practitioner to abandon the ‘you’ll have to live with it’ attitude
and assume active long-term medical therapy with logical surgical intervention as required.
Although at present it is not possible to help adequately all patients with this condition,
especially those with ‘end stage’ disease, nevertheless, persistence with logical principles of
treatment does improve many so long as it is clearly realized that such therapy must be
actively pursued for long periods of time, often for life.

Inflammation results in swelling of the mucosal lining which quickly blocks the
natural ostia. Secondary bacterial infection then occurs and the first line of treatment will
therefore include both antibiotics and medication to reduce inflammation. Topical
dehcongestants may be effective for short-term use but with chronic infection, long-term
medication is invariably required and the authors therefore prefer to use topical anti-
inflammatory drugs such as betamethasone. Undoubtedly, the most effective way for this to
reach the ostia is to use drops in the head down and forwards position instilling two drops
in this position and maintaining the posture for 2-3 minutes two or three times daily (Wilson
et al, 1987c). This can be combined with either topical antibiotics such as gentamicin or
neomycin or systemic antibiotics such as amoxycillin or cotrimoxazole.

When treatment with this regimen over a period of 3-4 weeks fails, surgical treatment
will be required. Sinus washout may be helpful in the treatment of acute sinusitis before the
mucosa has become too inflamed, but it is unlikely that a single washout will benefit a patient
with chronic mucosal changes. The disadvantage of weekly washouts can be overcome by
using an indwelling tube which can be used for daily lavage, instillation of medication and
aeration. If simple medical treatment fails, it is likely that the problem will be recurrent and
it appears more logical to attempt to provide more permanent drainage by undertaking an
intranasal antrostomy or similar drainage procedure of the other sinuses when they are
involved. The surgical management of this is described in Chapter 11.

**Treatment of the underlying condition**

*Mechanical obstruction*

Surgery to correct a deviated nasal septum will certainly be helpful if this is causing
sufficient obstruction to interfere with normal sinus drainage. This, however, is probably
seldom the case, although septal surgery is certainly important to improve the nasal airway.
Surgical reduction of the turbinates however may radically influence sinus drainage; the inferior turbinates, unless reduced, may obstruct an inferior meatal intranasal antrostomy and the middle turbinates may require careful conservative reduction if these are influencing maxillary and frontoethmoidal drainage.

Allergy

The mechanism and treatment of allergy, particularly allergen avoidance measures, are dealt with in Chapter 7. In some instances, an underlying allergic aetiology may compromise the normal clearance mechanism leading to secondary bacterial infection. In such cases, it will be necessary to treat the inflammatory process first with antibiotics and anti-inflammatory drugs (and possibly surgery) to restore the normal patency to the sinuses. Once this has been achieved, the underlying allergic problem will require maintenance therapy.

The treatment of allergic rhinitis has been revolutionized over the last few years by the introduction of first, topical corticosteroids (beclomethasone, flunisolide and budesonide) and second, non-sedating H1-antagonist antihistamines (terfenadine and astemizole). The latter rarely sedate or potentiate the action of alcohol. Terfenadine must be taken twice daily and is rapidly effective although remains so for a relatively short period (6-12 hours). Astemizole has the advantage of once daily dosage although its maximum effectiveness may not be obtained for a week or more. Skin tests may remain negative for up to 4 weeks from the last dosage. Both are contraindicated in pregnancy.

The combination of topical corticosteroids and systemic H1-antagonists used simultaneously is particularly effective. Occasional dryness and bleeding may occur with topical steroids but no irreversible damage to the mucosa has been reported and, provided they are used in the recommended dosage, they have no systemic side-effects.

Systemic corticosteroids are probably underused in the management of this condition. Because of their well-recognized side-effects, they are not suitable for long-term maintenance therapy but are very useful as a short course to bring the symptoms under control with a view of maintaining any improvement using topical corticosteroids, antihistamines or sodium cromoglycate. The latter is less effective than topical corticosteroids but is an entirely safe medication, is only effective as a prophylactic agent and is more likely to be effective in atopic patients.

The prospect of long-term maintenance therapy does not appeal to some patients and hyposensitization or surgery may be requested. Hyposensitization, more effective in children than adults, is only of proven value in the treatment of single allergen (pollen or house dust mite) disease. It carries, however, considerable risk of anaphylaxis and, as such, is not recommended. Such treatment in atopic patients may worsen existing allergic disease or provoke asthma, and is more likely to provoke anaphylaxis in such patients.

Allergy cannot be 'cured' by surgery. That does not mean that surgery has no place, however. Surgical reduction of the turbinates, correction of deviation of the nose or septum and surgery to improve sinus patency may all play an important role, particularly where nasal obstruction is the patient's main symptom. Some patients may be able to contend with
Abnormalities of mucociliary clearance

These may primarily affect the ciliary or mucus component of clearance (primary ciliary dyskinesia, Young's syndrome and cystic fibrosis in which mucus is abnormally thick and tenacious) or be secondary. Primary syndromes are not 'curable' as yet, but secondary abnormalities (due to changes in mucus viscoelasticity induced by bacterial infection, for example after a viral infection of the upper respiratory tract; due to bacterial product- or neutrophil elastase-inhibition of ciliary function) can be treated in the expectation that in a reasonable proportion of patients the underlying condition will resolve and allow mucus and/or cilia to return to normal. In many cases, the chest is also involved in such disease and requires corresponding treatment (Stanley et al, 1985). At this point, it is appropriate to point out that cigarette smoking reduces mucociliary clearance and ciliary beat frequency in the nose as well as the bronchial tree, so that such habits may adversely affect treatment for secondary clearance abnormalities (Stanley et al, 1986).

With primary ciliary abnormalities such as primary ciliary dyskinesia, and primary mucus problems such as cystic fibrosis and Young's syndrome, as yet no 'cure' can be achieved; this is not to say, however, that symptomatic treatment is not helpful. The authors' experience suggests that these patients can obtain symptomatic relief with topical mucolytics such as alkaline nasal douche which is sniffed through the nose two or three times daily followed by the application of topical corticosteroids. Antral washout will not give lasting relief as it will do nothing to improve the underlying primary problem. If sinus X-rays reveal a fluid level, an antrostomy should be undertaken. As there can be little or no mucociliary transport, a middle meatal antrostomy will not be beneficial and a radical inferior meatal antrostomy, combined with surgical reduction of the inferior turbinate, is indicated to enable adequate drainage by gravity as well as facilitate aeration of the sinus and provide access for douching and, if necessary, topical medication.

Nasal polyps may occur with primary ciliary dyskinesia: in two out of 16 (Greenstone et al, 1985), six out of 15 (Pedersen and Mygind, 1982), 10 out of 33 (Levison et al, 1983). If these do not respond to topical corticosteroids they will require surgical removal.

Glue ear is another common finding in primary ciliary dyskinesia. Myringotomies were performed without success in four out of 16 patients reviewed by Greenstone et al (1985). The ears continued to discharge through the ventilation tube until it was either removed or extruded. Since there is no likelihood of primary ciliary dyskinesia resolving, it would seem unhelpful to advocate insertion of ventilation tubes. Fortunately, the hearing loss is seldom severe and in the authors' series, patients' otological problems did not deteriorate despite the condition having been present for many years in the older patients.

For many patients with primary nasal mucociliary clearance problems, their otolaryngological symptoms present little more than an inconvenience, possibly because their symptoms have been present for most of their life and they accept them as normal. For many, a simple medical regimen will be helpful; in some, surgical procedures to improve sinus drainage may be indicated. The most important aspect, however, should be an awareness of
the condition in the hope that early diagnosis and referral for appropriate chest management may prevent or delay the onset of serious chest disease such as bronchiectasis.

**Immunity deficiency**

Although, as previously described in this chapter, there are a number of immunity deficiency states which usually present initially to otolaryngologists or paediatricians, the condition panhypogammaglobulinaemia (very low IgG, IgA and IgM) is by far the most important to recognize and treat, because irreparable lung damage may be avoided thereby. Although an uncommon condition, missing the diagnosis is disastrous.

Conditions in which IgG and IgM are low or absent are treated by reconstitution with immunoglobulin. The standard therapy until recently was intramuscular injections of normal human immunoglobulin (Lister) given weekly in a dose of approximately 25 mg/kg body weight, varying according to its catabolism by the patient. In practice, this entailed painful, large volume injections usually divided between the buttocks. Nevertheless, patients preferred such discomfort to their infections. Monitoring of serum levels of immunoglobulin was often of limited help and the dose in such patients was regulated according to clinical control of frequency and severity of infections. The addition of fresh frozen plasma infusions at intervals to the regimen was sometimes required in the more seriously affected patient. Untoward effects (for example, anaphylaxis, rashes) were common and unpredictable.

The situation has now changed with the advent of intravenous preparations of immunoglobulin which appear to be less likely to cause untoward effects. Such preparations are given by infusion at less frequent intervals (usually 2-4 weekly) and have the advantage that the dose can be increased without increasing discomfort as occurred with intramuscular preparations. However, the preparations are very expensive and, because they have all been relatively recently introduced, the efficacy of some is not fully known. Opsonization of bacteria for phagocytosis is one important function of immunoglobulin and this can be used as the basis of a laboratory test to compare the efficacy of various intravenous preparations *in vitro* (Munro, Stanley and Cole, 1985).

Selective immunoglobulin A deficiency is not treatable at present (and, as mentioned before, it is debatable whether reconstitution of IgA would affect bacterial infections). However, IgA deficiency seems to be a marker for IgG subclass deficiencies in some cases and the latter are probably treatable by replacement immunoglobulin. At present, the situation is much clearer in children (Oxelius et al, 1981) than in adults.

The complexity of the immunological deficiencies which may be associated with chronic or recurrent rhinosinusitis make it mandatory to refer affected patients to a specialist centre.

**Granulomatous conditions**

Although these conditions will require appropriate systemic medication, topical treatment with alkaline nasal douche to remove crusts and inspissated mucus followed by the instillation of corticosteroid and antibiotic drops (betamethasone and neomycin) in the head down and forwards position, may not only give symptomatic relief but may, at times, control
the local condition, such as sarcoid restricted to the nose, in such a way that the systemic medication can be reduced. The treatment of Wegener's granulomatosis is dealt with in Chapter 18.

**Autonomic imbalance**

**Non-specific rhinitis (nasal hyperactivity, intrinsic rhinitis or 'vasomotor rhinitis')**

Because this condition, or perhaps group of conditions, is so ill understood, it is equally difficult to treat and the authors have found it more helpful from a practical point of view to categorize the treatment according to the patient's 'main complaint', that is obstruction, sneezing and rhinorrhoea.

**Obstruction**

Where nasal obstruction is the main complaint, and particularly where it is the only complaint, surgery would appear to give the best results. An initial trial of topical corticosteroids and antihistamines is reasonable, but the authors in reviewing over 1000 patients, found it beneficial in less than one-half. Prior to undertaking surgical reduction of the turbinates, it is important to confirm that the nose is indeed obstructed as patients' subjective assessment of their nasal airway is poor and rhinomanometry is helpful in establishing which patients are likely to benefit.

**Sneezing**

Topical treatment is less helpful in treating those patients where the main complaint is one of sneezing. Budesonide was shown by Pipkorn (1983) to be slightly preferable to other topical corticosteroids for this condition but, by and large, most patients in this group are not helped and may even find their symptoms are made worse.

Antihistamines offer the best chance of success. Astemizole is useful in that it need only be taken once daily and, because of its pharmacokinetics, if for any reason the medication is omitted for a day or two, it will continue to be effective. Many patients in this group find that their symptoms occur in bouts: they may remain symptom-free for weeks, then experience a severe bout of intractable sneezing for several hours or days. Terfenadine, which is quickly effective, may be more suitable for this group, using it only when required.

**Watery rhinorrhoea**

Non-specific watery rhinorrhoea may respond to antihistamines and topical corticosteroids but when it does not, topical ipratropium offers an effective alternative.

When medical treatment fails, surgical division of the nerve of the pterygoid canal (vidian neurectomy) can be considered. The nerve can be approached via the maxillary sinus (Golding-Wood, 1961); other approaches are transpalatal (Chandra, 1969), trans-septal (Minnis and Morrison, 1971), and transnasal (Kirtane, Prabhu and Karnik, 1984). Complications include diplopia, anaesthesia of the palate and infraorbital region, impairment of lacrimation, blindness, infraorbital neuralgia and sinusitis. Kirtane, Prabhu and Karnik (1984) claimed that
the transnasal approach is the simplest and most direct and reported no serious complications in 247 cases, over 95% having relief from their symptoms of rhinorrhea and sneezing, but no long-term results are reported. Until long-term controlled studies have been carried out, vidian neurectomy is probably best left to those few surgeons who have special experience of it and should be avoided by others.

**Emotional rhinitis**

Whether depression is the cause or result of rhinitis remains debatable. There can be no doubt, however, that many patients with this condition can be successfully treated with imipramine which not only has an antidepressant effect but also acts as an anticholinergic drug. An initial dose of 25 mg three times daily can be increased to a maximum of 200 mg daily if drowsiness is not a problem. It is important to note, however, that it may take 4 weeks or more before the maximum effect is experienced.

For those patients in whom no diagnosis can be made and no objective evidence exists for their symptoms, explanation and reassurance alone are preferable to medication or surgery, which are unlikely to benefit the patient and may later even be blamed for their condition: hence 'I had no trouble with my sinuses until they were washed out'.

**Hormonal**

**Pregnancy**

Nasal obstruction is usually the main complaint in rhinitis associated with pregnancy and, because of pregnancy, it is difficult to treat. Topical nasal decongestants may be helpful but should not be continued long term. The problem of 'rebound' can, to some extent, be alleviated by decongesting one side of the nose one day and the other side the next. Linear diathermy to the surface of the inferior turbinate under local anaesthesia may give more lasting relief but may result in greater nasal obstruction for the first 7-10 days.

Topically acting corticosteroids such as beclomethasone are unlikely to have any significant undesirable systemic effect and may prove beneficial. Antihistamines, particularly astemizole and terfenadine, are specifically contraindicated during pregnancy.

Secondary bacterial infection may be treated with penicillins but tetracyclines should not be used. Acute infection of the maxillary sinuses not responding to medical treatment alone may require antral lavage under local anaesthesia.

**Menopausal women**

Atrophic rhinitis occurring in women after the menopause may respond to topical oestrogen. Manual removal of large crusts is followed by regular douching with alkaline nasal douche after which conjugated oestrogen cream (Premarin) is applied.
Senile rhinorrhoea

This problem affects men more than women and, although testosterone propionate was advocated by Watson-Williams (1952), the authors have no experience with this. Most patients in this group however can be controlled with topical ipratropium (Atrovent).

Iatrogenic

Rhinitis medicamentosa resulting from overuse of topical decongestants is best managed by explanation of the mechanisms of 'rebound'. It is also necessary to look for any other underlying cause for the patient's symptoms, such as allergy. Provided that there are no contraindications, a short course of systemic corticosteroids (prednisolone 10 mg for 5 days reducing to 5 mg for a further 5 days), together with systemic antihistamines (terfenadine 60 mg twice daily for one month) and avoidance of any topical medication may be all that is required. Where there is some other underlying cause, this will require appropriate management and maintenance therapy.

Where this fails, surgical reduction of the inferior turbinates will be required followed by strict avoidance of any decongestants. Partial trimming allows a reasonable airway in the early postoperative stage which can be maintained with an alkaline douche to remove crusts and caked blood clots for the first few weeks.

Concluding remarks

It is hoped that in writing this chapter, the authors have demonstrated the similarities and close relationship between the upper and lower respiratory tract. Knowledge gained from study of disease in one inevitably affects knowledge about the other and is the main reason why otolaryngologists and chest physicians should work in close relationship for mutual benefit and benefit of their patients.
Chapter 7: Mechanisms and treatment of allergic rhinitis

A. B. Kay

Definition and prevalence

Allergic rhinitis is an IgE-mediated hypersensitivity disease of the mucous membranes of the nasal airways characterized by sneezing, nasal blockage and discharge. Conjunctivitis and bronchial constriction often accompany these symptoms. Allergic rhinitis is mostly found in association with exposure to aeroallergens. A similar syndrome is occasionally observed in a minority of susceptible individuals following ingestion of certain foods, but an immunological basis for these reactions is as yet to be established. Allergic rhinitis is either seasonal (for example summer hayfever) or perennial. Perennial rhinitis with severe seasonal exacerbations is common.

The disease is extremely common and affects approximately 10-20% of North Americans and between 10 and 15% of north Europeans. Perennial rhinitis is probably more common in adults than children. There are several excellent reviews on the subject (for example Wassermann, 1982; Norman, 1985).

Immunopathology

Immunoglobulin E

Immunoglobulin E (IgE) (see Geha, 1984) has the unique property of binding reversibly to high affinity receptors on mast cells and basophils. The interaction of antigen with cell-bound IgE initiates the secretion of pharmacologically active substances that cause clinical manifestations of immediate hypersensitivity.

IgE is composed of two heavy chains (epsilon) and two light chains (kappa or lambda). The immunoglobulin classes are depicted diagrammatically. The intact molecule has a molecular weight of 188,000 with the heavy and light chains being of molecular weight 72,500 and 23,000, respectively. The sedimentation coefficient is 11S and the carbohydrate content of the molecule is 12%. IgE is heat labile. Heating for 2 hours at 56°C destroys its capacity to interact with Fc receptors but not its allergen-binding capacity.

IgE receptors with a high affinity have been identified on mast cells and basophils and tentatively termed 'Fc-epsilon_1'. Low affinity receptors, apparently antigenically distinct, are present on subpopulations of T and B lymphocytes, monocytes and macrophages, eosinophils and platelets. The low affinity receptors have been tentatively termed 'Fc-epsilon_2' (Capron and Capron, 1987).

Extensive work in rats has clearly shown that the development of an IgE response is dependent on T lymphocytes, cooperation between T and B lymphocytes, with regulation of IgE synthesis by a number of T-cell-derived soluble factors. It has been suggested that a defective suppressor T-cell function may underlie the atopic state in man, especially as a number of diseases with impaired thymus-dependent immunity have markedly elevated levels of IgE.
It has been shown in the rat that there are IgE potentiating and suppressing factors (Ishizaka, 1984). This isotype-specific regulation of IgE involves the conversion of virgin B cells bearing surface IgM to IgM-IgE double-bearing cells that are the precursors of IgE-forming cells. IgE-bearing memory B cells develop from these precursor cells after antigenic stimulation. These, in turn, form IgE-forming plasma cells. These various steps are regulated by T cells and their products. The scheme, worked out largely in mice and rats, has identified distinct IgE regulatory T cells which bear receptors for IgE. These receptors are shed and have the capacity to bind to IgE receptors on memory B cells. These Fc-epsilon associated binding factors are, if glycosylated, IgE potentiating factors but, if non-glycosylated, they are IgE suppressor factors. Glycosylation is dependent on the IgE regulating the T cell being influenced by either a glycosylation enhancing factor or glycosylation inhibitory factor. These in turn are derived from T-cell subsets with the production of glycosylation enhancing or inhibitory factors being determined largely by the nature of the adjuvant. For instance, aluminium hydroxide favours the secretion of glycosylated IgE helper factor, whereas complete Freund's adjuvant favours the secretion of non-glycosylated IgE suppressor factor.

Comparatively little is known about the regulation of IgE biosynthesis in man, although the data obtained so far are compatible with those from experimental animals. Normal peripheral blood lymphocytes do not synthesize IgE in vitro, whereas lymphocytes from atopic individuals spontaneously synthesize IgE with large amounts being observed during the season of grass or ragweed pollen. It has been shown that IgE-specific helper T cells reside within the CD4 subpopulation of T cells and secrete a glycoprotein which has an affinity for IgE and which enhances IgE synthesis by IgE-bearing cells. Conversely, there appears to be a suppressor T cell which predominantly resides within the CD8 subpopulation of cells which suppresses IgE synthesis. Serum from normal non-allergic individuals contains a low-molecular-weight factor (mol wt of 15,000) which specifically suppresses IgE synthesis. Thus an IgE suppressor factor has been isolated in normal serum and IgE helper factors have been identified in serum and supernatants from cell lines of individuals with high IgE. These observations have clear implications for therapy.

IgE-bearing B cells are usually detectable by the eleventh week of fetal life although IgE production in utero is negligible. At birth, the concentration of cord blood IgE is less than 1 international unit (1 IU = 2.4 nanograms). Over 50% of newborn infants have no detectable IgE. In adults the concentrations peak between 10 and 15 years and then tend to be stable from 15 to approximately 50 years, thereafter decreasing with age. Elevated serum levels are found in association with various metazoan parasitic infections, allergic asthma, hayfever and atopic eczema. IgE levels tend to correlate with the degree of antigen stimulation. For instance, IgE levels in hayfever sufferers rise significantly during the pollen season.

Although IgE is not essential for the maintenance of health, there is evidence to suspect that IgE may play a role in immunity to helminthic parasites. It may also serve in a more general role by facilitating increased permeability to allow the accumulation of IgE antibodies, complement and phagocytic cells to the site of invading foreign antigens.

The 'cross-linking' hypothesis is generally accepted as the means whereby mediator release is initiated (Ishizaka and Ishizaka, 1984). Antigen molecules are thought to interact with two adjacent cell-bound IgE antibody molecules so forming a bridge composed of IgE-antigen-IgE. Mediator release will only occur if the allergen is divalent or polyvalent, since
neither hapten nor Fab fragments of anti-IgE will give rise to histamine release. In addition, histamine release can be triggered by antibodies to the receptor molecule so indicating that IgE antibodies act only as a method of cross-linking the receptor molecules. It is likely that cross-linking the receptors allows the formation of a calcium channel and that calcium influx triggers the events that lead to histamine release. There are other mechanisms for triggering histamine release which bypass the requirement for receptor molecules and cross-linking. These non-IgE triggering release mechanisms include lectins, Compound 48/80, and anaphylatoxins (fragments cleaved from the third, fourth and fifth components of complement). A number of intravenous anaesthetic agents, radiocontrast media and plasma volume expanders are also associated with non-IgE release of mast cell mediators. Their effect is possibly through anaphylatoxin formation through activation of the alternative pathway of complement.

**Mediators**

Mediator release initiated by dimerization of cell membrane-bound IgE molecules by specific antigen with subsequent cross-linkage of cell membrane IgE (Fc) receptors is a secretory process that results in the elaboration of preformed mediators contained within lysosomal granules and membrane-derived lipid mediators. The initial signal is stimulated by phospholipid metabolism and adenylate cyclase activity in the cell membrane so generating di- and monoacylglycerol and arachidonic acid and converts adenosine 5'-triphosphate (ATP) to adenosine cyclic 3':5'-monophosphate (cyclic AMP), respectively. A component of this altered phospholipid metabolism may include phospholipid methylation (Ishizaka and Ishizaka, 1984). However, it is now considered that stimulation of the phosphatidylinositol cycle with alterations in the biophysical properties of the cell membrane is the factor responsible for exposing channels allowing the influx of calcium ions and the progression of the secretory response to the cell cytosol. In both rat and human mast cells, cyclic AMP-dependent transport of ions and water across the perigranular membranes partially solubilize the packaged mediators prior to their extracellular discharge. Diacylglycerol, generated in the membrane, activates protein kinase C which has the capacity to phosphorylate the light chain of myosin with subsequent contraction of thin filaments. With utilization of energy, translocation of the granule to the cell surface occurs where fusion of the perigranular matrix and plasma membrane exposes the granular matrix to the extracellular environment. Mediators are released from the partially solubilized granular matrix at different rates of ion exchange.

In parallel with the events leading to granule secretion, stimulation of membrane phospholipases (C and A2) generates increased membrane levels of lysophospholipids, diacyl- and monoacylglycerol which are membrane fusagens and a substantial amount of polyunsaturated fatty acid, arachidonic acid. Subsequent metabolism of arachidonic acid by the cyclo-oxygenase pathway generates predominantly prostaglandin D2 (PGD2), while the lipo-oxygenase pathway generates leukotriene B4, a chemotactic factor, and leukotrienes C4 and D4 which constitute the biological activity previously recognized as the slow-reacting substance of anaphylaxis (SRS-A). Thus the non-cytotoxic secretory process of the mast cell is similar to that observed in exocrine and endocrine cells, but is unique in the specificity of the immunological stimulus which initiates coupled activation-secretion and in the spectrum of preformed and newly formed generated mediators which characterize the immediate hypersensitivity reaction.
Mast cells have been located interepithelially, although the largest numbers are observed in the submucosa. Basophils are often found in blown secretion from patients with rhinitis. Thus water-soluble allergens which are readily leached from air-borne particles would be free to interact with IgE-sensitized mast cells (and basophils) which, in turn, lead to the release of pharmacological mediators of hypersensitivity. These pharmacological agents include histamine, leukotrienes C₄, D₄, E₄ and B₄, kinin-like activity, prostaglandin E₂ and tosylarginine methyl ester esterase (TAME-esterase) activity (Naclerio et al, 1986). Rapid release of mediators from cells located in the epithelium or found free on the nasal mucosa would lead to increased permeability by the opening up of epithelial tight junctions so allowing allergen access to deeper mast cells. In fact symptoms, that is sneezing, occur within one minute when sensitive patients undergo nasal challenge. A number of mast cell or basophil-derived mediators have been detected in nasal secretions after challenge with appropriate allergen.

It has been shown that basophils are often present in blown secretions from allergic subjects, and mast cells have been observed from scrapings from the same patients. It was concluded that, during the pollen season, basophils migrate through the surface epithelium to the airway lumen whereas mast cells migrate only into the epithelium. The increase in epithelium mediator cells after allergen provocation and during the pollen season was associated with successful immunotherapy and corticosteroid treatment.

Before the importance or otherwise of a biochemical or pharmacological mediator can be established in the pathogenesis of disease, it must be able to evoke the symptoms, be detected in pathological fluids, and specific antagonists should ameliorate or modify the symptoms of the disease.

**Histamine**

Instillation of histamine into the nose produces immediate itching followed by sneezing, nasal discharge and blocking (Mygind, 1982). Thus the symptoms are very similar to those produced by allergens in the sensitized subject. It should be noted that allergen provocation, unlike histamine provocation, results in infiltration of inflammatory cells, particularly eosinophils, and heightened nasal reactivity. Small amounts of histamine can be identified in nasal washings after allergen challenge. Selective H₁ antagonists such as astemizole (Hismanal) inhibit sneezing and watery discharge but not nasal blockage.

Unilateral histamine provocation of the human nose causes a marked homolateral blockage, but only a short-lasting and insignificant challenge of contralateral nasal patency indicating that a direct histamine effect on blood vessels, and not reflex activity, may be important for any persistent nasal blockage in allergic rhinitis. Histamine affects the vascular tube by both H₁ and H₂ receptors resulting in dilatation of some, and constriction of other blood vessels and oedema formation. Histamine has a fairly weak H₂ mediator effect on mucous glands and appears to increase mucous glycoproteins without significantly affecting the total volume of nasal discharge. Thus the inability of ordinary histamines and the new selective H₁ antagonists to deal with nasal blockage in allergic rhinitis could be due to the presence of H₂ receptors in nasal vasculature. Combined use of H₁ and H₂ antagonists in the nose only partially prevents histamine-provoked vascular changes. Thus the effects of histamine are complex. Although there is slight bilateral blockage, there is considerable
bilateral hypersecretion after histamine instillation which is stimulated through H₁ receptors. Thus sneezing and a large part of mucus secretion appears to be reflex mediated.

Reports on the instillation of leukotrienes into the nose have been fragmentary although leukotrienes C₄, D₄, E₄ and LTB₄ have been identified in nasal washings after allergen but not methacholine challenge. The sulphidopeptide leukotrienes cause hypersecretion of mucus as well as constricting smooth muscle. They also dilate the vasculature, but it is unknown which part of the nasal vasculature is affected by these lipid mediators. LTB₄ is a powerful chemoattractant for neutrophils and, to a lesser extent, eosinophils. Studies on the effects of selective leukotriene antagonists in rhinitis have not yet been forthcoming. PGD₂ was observed during the early, but not late, phase reactions suggesting that basophils may be involved in the late phase reactions since PGD₂ is not derived from this cell type (see below). However, aspirin, a cyclo-oxygenase inhibitor which would prevent PGD₂ formation, is of no proven value in the treatment of allergic rhinitis and in an appreciable portion of individuals actually causes rhinitis symptoms. Thus the role of PGD₂ must remain unclear.

High-molecular-weight neutrophil chemotactic activity has also been identified in nasal washings from grass-pollen-sensitive patients challenged with aqueous extracts of pollen antigen. Kinin-like activity has been observed in nasal secretions but the significance of this observation is unclear. An enzyme which hydrolyses TAME-esterase has also been observed.

Late-phase reactions have been most extensively studied in the lower airways, but have also been reported in the nose and skin (Kay et al, 1984). Following the immediate reaction, a proportion of individuals will have a late-phase reaction developing 3-11 hours after the immediate response. The late-phase reaction is generally considered to be the result of infiltrating inflammatory cells since local accumulation of neutrophils, eosinophils and basophils is well documented.

**Other immunoglobulins**

Allergic rhinitis is associated, not only with allergen-specific IgE, but also with the presence of IgG and IgA antibodies in serum from untreated patients (Platts-Mills, 1982). IgE antibodies to a single allergen represent as much as 50% of the total IgE. The majority of sera from patients with hayfever contain less than 0.1 microg of IgE antibody to pollen allergens per mL. Using antigen-binding techniques, it has been possible to show that all sera contain detectable IgE antibody and detectable IgG antibody against the same allergen. The quantity of IgG antibody is on an average only three or four times greater than the IgE antibody. In sera from untreated patients, there is a good correlation between the IgG and IgE antibody responses. Most non-allergic individuals have no detectable IgG or IgE antibody. Generally speaking, none of the non-allergenic proteins in complex mixtures such as house dust induce IgG antibody responses in man. Thus, there is little doubt that, when antibody responses to inhalant allergens occur as a result of an actual exposure, the IgE and IgG antibody responses occur in parallel. It was further shown that IgG and IgA antibodies (in addition to IgE) are produced locally in pollen hayfever. It was suggested that the local response to pollen antigens in which the secreted antibody was produced was from mucosal plasma cells while the IgG and IgE antibodies in serum would derive from local lymph nodes (Platts-Mills, 1982). A possible model of the local immune response to pollen allergens was proposed in
which antibodies produced by plasma cells in the mucosa are predominantly secreted, whereas 
the local lymph nodes are the main site of production of serum IgG and IgE antibodies to 
pollen allergens.

Relationship of immunoglobulin concentrations to symptoms

Most non-allergic persons have negative skin tests and lack both IgG and IgE 
antibodies to inhalant allergens (Platts-Mills, 1982). A proportion of people, however, are 
unaware of any nasal symptoms but have positive skin tests. These asymptomatic people can 
be divided into three groups:

1. negative skin tests and negative serum antibodies
2. negative skin tests and positive serum IgG antibodies
3. positive skin tests and serum IgG and IgE antibodies.

A proportion of the skin test positive patients will develop symptoms in subsequent 
years. Some patients spontaneously recover from hayfever. This has been estimated at a cure 
rate of 8% in 3 years for hayfever. Some patients who have recovered from grass pollen 
hayfever still have positive skin tests and detectable IgE antibodies. Although the evidence 
is still incomplete, it appears that spontaneous cure of human allergic diseases is usually 
allergen specific and involves a gradual reduction in the concentrations of IgG and IgE 
antibodies. In general, it is thought that patients do not develop and IgE response for a second 
time. This observation is not well documented and certainly there are no data available on the 
mechanism by which 'relapse' is prevented. Similarly, there are very few data available on the 
conditions for natural induction of IgE responses in man. while it is clear that symptoms of 
hayfever may not develop until the age of 20 or later, it is not clear at what stage these 
patients develop IgE antibodies. Immigrants to England from Hong Kong who never have 
symptoms in their first summer often develop hayfever within 3 or 4 years. Sensitization to 
laboratory animals can be particularly rapid. Persons who 'change' animals often become 
allergic to a new species within a few months. In addition, several workers who had never 
worked with a laboratory animal before became sensitized to rat urine proteins within 3 
months. The development of IgE antibodies and the onset of symptoms in infants suggests 
that the onset of sensitization was related to viral infections. Possibly inflammation of the 
nasal mucosa allows more allergen entry or acts as an adjuvant and allows the allergic 
response to one allergen to potentiate an antibody response to other allergens. The role of 
short-term sensitizing IgG antibody and subclasses of IgG in the pathogenesis of hayfever and 
related atopic disease remains unclear. At present there remains considerable doubt as to 
whether short-term sensitizing IgG antibodies are responsible for immediate hypersensitivity 
in any human condition. There is also no convincing evidence that IgG4 (or IgA) antibodies 
play a direct role in mediating the symptoms of hayfever, perennial rhinitis or asthma.

The role of T cells in inhalant allergy is unclear, although T cells from allergic rhinitis 
patients will respond in vitro to specific antigen and anti-IgE (Platts-Mills, 1982). It is 
possible that sensitized T cells play a role in recruiting basophils to the nose. Experiments in 
guinea-pigs suggest that basophil accumulation in the skin is, in fact, a form of delayed 
hypersensitivity and can be mediated by the same T cells that mediate delayed 
hypersensitivity. Although several groups have succeeded in measuring IgE produced by 
human lymphocytes in vitro it is doubtful whether an in vitro suppressor phenomenon or T-
cell abnormalities and atopic diseases can be related to the aetiology of the disease or the mechanism of desensitization. In general, T cells from non-allergic individuals do not respond to allergens in vitro.

In hayfever there is a strong correlation between the sensitivity of mediator cells, that is peripheral blood basophils, and the severity of disease during natural pollen exposure. The amount of antigen required to release 50% of histamine from peripheral blood basophils remains fairly constant from season to season in most treated adults. This index of cell sensitivity varies considerably from patient to patient, although the severity of a patient's allergic symptoms during the ragweed season is directly related to the sensitivity of cells to ragweed antigen E. Serum levels of specific IgE antibodies also correlate with the severity of symptoms in untreated ragweed-sensitive patients. Similarly, leucocyte histamine release and serum levels of IgE antibodies also have a close correlation.

The pathology of allergic rhinitis will vary with the length of exposure and accompanying complications. In uncomplicated seasonal hayfever, submucosal oedema and infiltration with eosinophils and, to a lesser extent, neutrophils are the prominent findings. In long-standing perennial rhinitis, epithelial cell damage and thickening of the basement membrane may be observed.

**Allergens**

The term 'allergen' was originally used to describe any substance which could give rise to an altered reaction, that is either an immune or hypersensitivity response. It is now used in a limited fashion and describes only those substances which elicit an allergic response manifest by a hypersensitivity reaction. An 'atopic allergen' is one which gives rise to a type I immediate hypersensitivity reaction mediated by specific IgE antibody.

Extracts of allergen have been used since the early part of the century when Noon (1911) first undertook 'desensitization' using extracts of grass pollen. One of the major problems which has beset allergists ever since this form of treatment was introduced is the problem of clearly defining and standardizing extracts used for diagnosis (that is skin prick tests or the radioallergosorbent test - RAST) and immunotherapy. Even today the situation remains unsatisfactory, but there is considerable development in the field of allergen purification. Furthermore, there has been a rapid increase in the knowledge of the chemistry of allergens due to the development of several biochemical and biological techniques in vitro. Thus, it is now possible in many instances to purify, characterize and standardize allergen extracts fairly precisely.

Naturally occurring allergens have been shown to be proteins or glycoproteins, which are freely soluble and so easily absorbed through mucous membranes and usually have a molecular weight in the range of 10,000-40,000. Contrary to earlier belief, there do not seem to be any particular physicochemical features or characteristics for allergens. Thus neither the molecular size, hydrophobicity nor primary amino acid structure determines whether or not proteins will act as allergens.

The allergen proteins most frequently encountered in organic material and located normally in the human environment are derived from a number of source materials. The ones
most relevant to allergic rhinitis are the aeroallergens, such as pollens from wind-pollinated grasses, trees and weeds; mould spores and myceliae; mite proteins and proteins originating from animal dander, urine, saliva etc. Ingestants such as milk, soy, fish and cereals, eggs and nuts, also serve as source material for food allergens (which are probably of little importance in rhinitis).

Some of the proteins in source materials induce allergy in humans that are of higher frequency than others. These substances are termed major allergens. For instance, it was shown by skin prick testing that ragweed antigen E (RAE) was the major allergen from ragweed in contrast to the minor allergens RA3, RA4 and RA5. Major allergens have been defined as 'allergens that show specific IgE binding in more than 50% of the population allergic to that particular allergen and when they show strong IgE binding in at least 25%'. Minor allergens are those which give rise to IgE binding in less than 10% of patients. Allergens in between these extremes are defined as intermediate allergens. 'Binding' is a relative term, which is used in techniques such as crossed radioimmunolectrophoresis, and refers to the association between allergen and specific IgE antibody.

Major allergens are very readily extracted from source material and are often the most abundant proteins in an extract, although they may only be a small percentage of the dry weight of an allergenic particle. For example, antigen P1 from the faeces of the house dust mite and the major allergens of birch and rye grass pollens are released very rapidly. A notable exception is antigen E of ragweed. Allergens, in general, are remarkably stable to enzymatic biodegradation and chemical denaturation, possibly since they are already subjected naturally to a number of biodegradation processes, both from their own proteolytic enzyme system (for example pollens and fungi), as well as resisting various host defence mechanisms.

A further general characteristic of allergens is their ability to exist in multiple molecular forms which differ only slightly in their pI values, but which carry the same allergenic determinants for combination with corresponding IgE antibodies (iso-allergens). These slight differences in the net charges of polypeptides are dependent, not only on differences in the ratio of acidic to basic amino acid residues, but also on minor structural differences which are, in turn, dependent on the nature of their carbohydrate moieties or, for example, differing degrees of amidation.

**Plant allergens**

**Grasses** (see Hubbard, 1984)

Allergy to grass pollen allergens is by far the most common cause of hayfever in the UK, especially as the climate is most favourable for the production of luscious green growth during a large part of the year (Hubbard, 1984). This abundance of grasses is almost entirely artificial in origin and due to the continual labours of many generations of our ancestors, together with the cumulative action of the grazing and treading of their domestic animals. Under the climatic conditions and on most soils, these artificial grasslands, when removed from the control of man and beast and left to the effects of competition and natural selection, gradually revert to scrub, and in most cases from scrub to forest.
Permanent grassland contains many species of grass. These include perennial rye-grass (*Lolium perenne*), large leaved Timothy-grass (*Phleum pratense*), cocksfoot (*Dactylis glomerata*), meadow fescue (*Festuca pratensis*), tall fescue (*Festuca arundinacea*), common bent (*Agrostis tenuis*), creeping bent (*Agrostis stolonifera*), Yorkshire fog (*Holcus lanatus*), red fescue (*Festuca rubra*), rough meadow-grass (*Poa trivialis*), and meadow fox-tail (*Alopecurus pratensis*). Amenity grassland covers about 850,000 hectares in the UK, or nearly 4% of the land surface.

The turf grasses include velvet bent (*Agrostis canina* subsp *canina*), brown top (*Agrostis tenuis*) and Highland bent (*Agrostis stolonifera*), fine-leaved sheep's fescue (*Festuca tenuifolia*), hard fescue (*Festuca longifolia*), subspecies of red fescue (*Festuca rubra*), chewings fescue (subsp *commutata*), slender creeping red fescue (subsp *litoralis*), strong creeping red fescue (subsp *rubra*), crested dog's tail (*Poa trivialis*), smooth meadow-grass (*Poa pratensis*), small-leaved Timothy grass (*Phleum bertolonii*), large-leaved Timothy-grass (*Phleum pratense*), and perennial rye-grass (*Lolium perenne*).

Grasses are flowering plants (spermatophyta) whose pollens are dispersed by the wind (anemophilous). The flowers are usually perfect, that is they contain both pistils and stamens (Solomon, 1984). The grasses are all angiosperms and monocotyledons. Most of the grasses in the UK flower during May, June and July, the greatest number being in bloom towards the end of June and early in July. Woodland and mountainside species generally flower later than species of the same genera from open situations in the lowlands or from the south. Individual grasses have fairly regular daily flowering periods, although this is affected by the weather as the florets remain closed on dull or wet days. In the majority, flowering takes place in the early morning (mainly 04:00-09:00), in a few about midday, and in others during the afternoon and evening (15:00-19:00). Most species flower only once a day and this lasts for about 4-12 days (usually 7-8 days), depending on the type and size of flower heads. The majority of UK grasses are chasmogamous, their florets opening for the exertion of the anthers and stigma. Obviously only the wind-pollinated, rather than the self-pollinated, grasses are important in allergy.

Pollen grains become air-borne when they fall into an airstream or are scourcd by turbulent eddies (Solomon, 1984). Pollens are projected through a relatively thin lamina boundary layer of still air into the constantly moving lower atmosphere. There are many factors which determine dispersal. These include the strength and proximity of upwind sources, diluting vertical and horizontal crosswinds, prevailing winds, local surface effects, that is vegetation screens, cul-de-sacs; on a warm day pollen-loaded air is higher, thermal inversions are thought to exist and rising air will not ascend further. The effect of thermal inversions is to cause second nightly peaks when the atmosphere cools and the pollen grains subside to the surface. Overall, levels of air-borne pollen are increased by warm, dry, clear conditions and fall during unreasonable cold or wet periods. Pollen grains can travel long distances and have been recovered in trans-Atlantic flights at an altitude of 3000 metres in summertime. However, most wind-borne pollens are lost within a few hundred metres of their source.

It has been found that the development of high concentrations of grass pollen in the air over London, as compared with Liverpool and Glasgow, respectively, is associated with
more rapidly rising values of accumulated temperature. In a study contrasting grass pollen and fungal spores in London and Davos, it was found that an earlier rise and sustained higher values of accumulated temperature were associated with higher counts. Prolonged rain washes the air free of particulate matter and very heavy rain washes pollens, and even the anthers which bear them, to the ground.

It appears that the pollen clouds flowing over British cities are mainly of an exogenous origin from such sources as crops, pastures and woodlands outside (Davies, 1969). The 5 km² of grassland in parks of central London are probably not a significant source of grass pollens. Buildings and open spaces and cities affect the deposition of grass pollens. Winds are deflected over the rooftops and large particles, that is pollen grains, pass into the relatively still air between the buildings and tend to settle quickly to the ground. On hot, sunny days street level temperatures one degree higher than those in the airstreams above the 25 metre high rooftops have been observed, and convection and turbulence will tend to inhibit the settlement of small spores, that is fungi (into the city).

Although no detailed survey has been undertaken into the prevalence of atopic disease in various regions of the UK, hayfever is probably less frequently diagnosed in the north-west and Wales than it is in the east and south-east England. Wales and the northern, north-western and east and west Ridings regions of England are ecologically similar and characterized by mountainous and upland areas with grass, heath and moorland type of vegetation. By contrast, in the other regions of England and Wales there is very little mountain and moorland, and the land is mainly devoted to arable farming or mixed crops with some stock raising. As a consequence, these areas are a greater source of seasonal allergens, particularly grass pollens. Also, because of the west and south-west prevailing winds, spore and pollen clouds over east and south-east regions of England will tend to be denser because of wind collections from the more westerly areas.

Pollen counts of 50 grains/mm³ or more seem to be associated with clinical symptoms in susceptible individuals. However, very sensitive individuals will experience symptoms from concentrations as low as one grain/mm³.

**Structural features in the identification of pollens**

Most pollens are between 12 and 17 microm in diameter and are usually spheres or sphere-like particles (Solomon, 1984). The distinctive surface features depend on the intactness or otherwise of the tectum and other exterior features. Grains can be smooth (psilate) or roughened (scabrate or verrucate). Sometimes they have club-shaped projections (clavate). Apertures in the exine are referred to as pores or furrows if they are elongated. Grain regions are termed 'polar' or 'equatorial'. Anemophalous grains may have one or more pores alone (porate), furrows alone (colpate) or furrows with central pores (colporate).

Determination of pollen output by anemophalous species is not well understood, but appears to include temperature, humidity, rainfall and life intensity. Davies and Smith (1973) were able to estimate the annual start and eventual intensity of grass pollen exposure from measurements of accumulated spring temperatures.
Thus pollen is either distributed by insects or by the wind. Pollens which are insect-borne are derived from plants with flowers which have colour and a scent to attract the insect, and the grains themselves are sticky and adhere to the insect. Wind-borne pollens are lightweight, can be carried long distances, and must be profuse in quantity. Since there is no need to attract insects, an attractive flower is unnecessary.

Allergy to flower pollens probably does not exist and it is dubious as to whether extracts of flower pollen should appear in skin test kits.

In the UK, the pollen seasons of importance to the allergist are: tree pollen allergy in the spring, grass pollens in the summer, and weed pollen allergy in the latter part of the summer and early autumn. In the USA there are also roughly three pollen seasons of importance to the hayfever/asthma sufferer. These are trees, grasses, and weeds, with pollination appearing locally at about the same time. The 'pollen season' extends over a much broader period, although this will of course vary from place to place.

**Fungi**

Fungi are a unique group of organisms and classified as a separate kingdom from plants and animals on the basis of their ability to absorb nutrients and their multinucleate character (Austwick, 1981). Fungi may be unicellular (yeast-like), filamentous (mould-like), or both (dimorphic). Fungi may form reproductive propagules through an asexual production by mitotic nuclear division (for example buds, sporangiospores or conidia) and/or through a sexual process of meiotic division (for example zygospores, ascospores or basidiospores). It is the conidia of several common fungi which are of importance in IgE-mediated allergy. A number of fungi are also associated with extrinsic allergic alveolitis.

All fungi possess a true nucleus (eukaryotic), are incapable of forming all of their nutritional requirements (heterotrophic), and lack chlorophyll (achlorophyllous). It is a fourth feature - the ability to release extracellular enzymes which break down nutrient stores and to absorb the nutrients - which distinguishes fungi from most other living organisms. Thus, animals ingest nutrients, plants synthesize nutrients, and fungi absorb nutrients. A further important characteristic of fungi is their multinucleate structure. In other words, the entire developing body of the organism lacks complete cross-walls (septa). Some have incomplete septa, but in all fungi there is direct communication throughout the whole of the organism.

Most species grow optimally at slightly acid pH and at room temperature (20-25°C), although there are notable exceptions to these rules. Most fungi are strict aerobes, although many yeasts are facultative anaerobes. Most fungi grow at relatively high humidity, although the wall surrounding asexual and sexual propagules inhibits desiccation and permits fungi to survive extreme or prolonged droughts. Conidia become airborne as a result of various changes in the environment. These include wind, rain, light and changes in relative humidity. In an outdoor environment there are two daily peaks of conidial discharge at 03:00-06:00 when the relative humidity is highest, and 15:00-18:00 - the driest, windiest part of the day. There are, of course, seasonal variations.

The number of outdoor air spores may vary from between 200 and 2,000,000 per mm² of air, averaging on a daily basis 10,000-20,000 per mm² with peak concentrations rarely
exceeding 200,000 per mm² for short periods only. Such peaks usually correspond with conditions favouring the formation and liberation of numerous ascospores or basidiospores and seldom occur with conidial fungi. *Cladosporium* sp usually produces the most numerous spore type in the air during daytime but at night it is replaced by spores of *Ascomycetes*, *Basidiomycetes* and *Sporobomomyces* spp. Concentrations in England vary between 3200 and 6500 spores per mm² with peaks up to 240,000 per mm² for short periods. Spores of *Alternaria* sp are the next most abundant by day, but mean daily concentrations may be only 50-150 spores per mm².

The abundance of the night-time air spores has only been appreciated with the use of continuously operating spore traps. These have shown that balistospores of *Sporobolomyces* and ascospores of *Didymella exitialis* may often be abundant, exceeding 200,000 spores per mm² under suitable conditions close to their source.

Rain may cause an initial increase in spore numbers, due to the tap and puff effect but, if prolonged, it washes all spores out of the air. However, it may subsequently stimulate ascospore release. Indoors, the composition of the air spore is greatly influenced by local sources. In the absence of a source, the type of spores present in the air may be similar to outdoors but with smaller numbers. However, in the presence of a dry rot fruitification, up to 360,000 spores per mm² of air have been found and, when mouldy fodders are disturbed, up to 10¹⁰ spores per mm².

The spore types associated with fodders and other stored products depend on the way in which they have been stored, their water content and degree of spontaneous heating. *Actinomycetes* sp associated with farmer's lung, for instance, occurs in hay stored wetter than 35% water content which heats to 50-70°C. The *Aspergillus* species - *A. glaucus*, *A. versicolor*, *A. nidulans* and *A. fumigatus* - predominate in hay stored at 25%, 29%, 31% and 40% water content, respectively.

*Common allergenic fungi* (Salkin and Haines, 1984)

**Alternaria**

These are the most common air-borne fungal propagules. They are characterized by septation in two planes, a narrow projection from one end, and dark pigmentation. However, their morphology is quite variable in respect of size, shape, and number of septa. These asexual structures are usually encountered outdoors from July to September in the late afternoon. Spores of *Alternaria* are found when the atmosphere is warm and dry, have their origins on the surface of vegetation and are especially abundant when cereals are harvested. In the air over London alternaria spore concentrations reach 100 per mm² (or, rarely, 1000).

**Cladosporium**

The kinetia of this fungus are the most common air-borne fungal propagules found outdoors. They are characterized by dark pigmentation, the presence of one or several septa, and linkage in short, branch chains. Spores of *Cladosporium* occur in the atmosphere when the air is warm and dry and, as for *Alternaria*, their origins are on the surface of vegetation.
and when cereals are harvested. Indoors, spores of *Cladosporium* will colonize wallpaper and even painted surfaces. Outdoors the concentrations of spores of *Cladosporium* (and *Alternaria*) rise to a peak in the late summer and early autumn. In the air of London, spore concentrations of *Cladosporium* reach 10,000 per mm³.

**Aspergillus**

The kinetia of this mould are among the most common indoors and can be identified by their relatively small size (frequently less than 4 microm), their uniform round shape and their formation in chains on specialized physicular heads. The morphology of the kinetia developing on specialized types is used to identify the species.

The spores of *Aspergillus* are phialospores, that is they develop from the tip of short, modified hyphacallsterigma (or phylophor), several hundred of which are arranged over the surface of a claviscolen end of an erect hypha (condiophore 4). This is the typical structure of the sporing head of *Aspergillus*. As each sterigma tip expands, so a cross-wall develops below and cuts off the spore. The process is then repeated so that each sterigma bears a single line of spores; on each head is a column of many thousands of spores.

*Aspergillus* sp like *Penicillium* is sometimes called a storage fungus, since both are common causes of rot in stored grains, fruits and vegetables. *Aspergillus*, in particular, will thrive on substances with a low moisture content (12-16%). They are the two moulds most commonly cultured from houses especially basements and dark areas.

**Penicillium**

The kinetia of this mould are also very common indoors. They are characterized by their almost round shape and formation in chains on broom-shaped kinetial heads. They can be isolated all the year round. (Many spores such as those of *Aspergillus* and *Penicillium* cannot be morphologically without colony characteristics.)

Other possible allergens to be considered in IgE-mediated hypersensitivity include *Fusarium* and *Aureobasidium (Pullularia)* spp.

**House dust mite**

House dust mite contains many different agents which may be allergenic in man, each of which may be an important allergen in any particular household (Mosbech, 1985). Apart from the house dust mite, which will be considered in some detail, material derived from domestic animals such as cat saliva are important sources of house dust allergen, as are fungal spores, tree grass and weed pollens, insects (particularly in the USA where cockroach allergy is common), textile fibres, human dander and other agents. It was appreciated in the 1920s that an extract of house dust obtained from vacuum cleaners and certain types of pillows and mattresses gave positive skin prick tests in atopic individuals. It was not until 1964 that clear evidence was presented that pyroglyphid mites (*Dermatophagoides* sp) were a major source of the skin test reactivity in house dust. Voorhorst, Spieksma-Boezeman and Spieksma (1964) demonstrated that allergen accumulated progressively in mite cultures and that allergen
activity of house dust from different parts of the world correlated well with the number of mites.

Mites are more closely related to spiders and scorpions than to insects. They have eight legs in the adult stage and body segmentation is lacking. The primary house dust mites are *D. pteronyssinus* (Dp), *D. farinae* (Df) and *Euroglyphys maynei* (Ep), all belonging to the Pyroglyphidae family. Early mite cultures were grown on human skin scales, horse dander, fishmeal and/or wheatmeal. However, they are not grown on various types of non-allergenic media and skin responses to mite extracts are highly specific. It is now clear that mites are present in many parts of the world and dust mites are regarded as an allergen of major importance in Australia, Japan, Hong Kong, the West Indies, South Africa, the continent of Europe and the USA. In general, it is in the areas with the highest relative humidity that mites thrive; in contrast, areas with long periods of dry weather have poor mite growth. Thus the prevalence of the different mites is related to climate rather than to geographical conditions. In general Dp is found predominately in northern Europe, New Zealand and Australia whereas Df is found largely in North America and central Europe. Ep appears to be more prevalent at higher altitudes.

The female Ep produces 20-40 eggs once or twice during life. They are hatched after 6 days and the total immature life lasts about 25 days. The mature adult lives for about 2 to 3.5 months, in houses, the bedroom is the preferential breeding ground, particularly the bed itself. Here, sufficient food is supplied in the form of human scales and the temperature is optimal. The mattress is the most important habitat from which bed clothes are repopulated after laundry. Dusts from overstuffed furniture, carpest and unlaundered clothing can contain high numbers of house dust mites.

The optimal temperature for growth of Dp is $25^\circ C$ with a relative humidity of 70-80%. (For Df, a temperature of 25-30$^\circ C$ with a 50-60% relative humidity is ideal.) Thus mites are more abundant in humid homes and, in temperate climates, the number of house dust mites increases during the humid summer months, whereas during the heating season when the indoor relative humidity is low, mite numbers decline. The low humidity during the winter seems to be an important factor in the survival of the mite population. Similarly, the lower numbers of mites at high altitudes might be related to a low indoor relative humidity in the regions investigated.

**Allergen purification**

The development of techniques for growing mites on non-ectodermal heat-denatured mammalian material greatly facilitated the purification of mite allergens. It was shown by Chapman and Platts-Mills (1980) that the major allergen from Dp is a glycoprotein with an apparent molecular weight of 24,000 which is freely soluble in aqueous solution. This was designated Dp, antigen P$_1$. By means of crossed radioimmunolectrophoresis, 29 different components derived from Dp have been identified; 44-72% of the IgE against Dp was directed against antigen P$_1$ (which is identical to the antigens termed Dp 42 and Dpt 12).

The major allergen P$_1$ appears to represent as much as 20% of the protein in aqueous extracts of the cultures. Although P$_1$ is present as multiple iso-allergens with a very wide
range of PI values, it migrates as a discrete band on polyacrylamide-gel electrophoresis (PAGE) and this band is coincident with the heaviest protein band seen in the crude extract. Most of the allergen in cultures appears to be associated with mite faeces while mite cuticle and eggs contain negligible quantities. Antigen P₁ elutes very rapidly from dust, from mite cultures and from mite faeces (greater than 90%) in 2 min) while elution from live mites is very slow. Thus the major allergen present in mite cultures is associated with a particle of similar size to pollen grains, elutes very rapidly, has a molecular weight of 24,000 and is abundant 'in aqueous extracts'. These properties are similar to those of other inhalant allergens. By contrast, the mite allergen does not appear to have any chemical properties in common with pollen allergens. (Faeces 10-40 microm in diameter contain 0.1 nanog antigen P₁ per particle, equivalent to 10 mg per emulsed elution.)

Environmental exposure to antigen P₁ has been assessed by measuring the antigen P₁ content of dust extracts and by comparing the distribution of antigen P₁ in air-borne particles. Dust samples from houses in the Harrow area of London contained 100-100,000 nanog antigen P₁ per g of dust. In contrast, the dust samples from schools or hospital wards contained very little allergen (less than 10 nanog). In a particle sizing experiment, using a cascade impactor, it was not possible to detect air-borne allergens in undisturbed rooms. However, when the dust in a room was disturbed, for example by vacuum cleaning, most of the allergen was associated with particles of greater than 10 microm in diameter. Some of these particles were identified as mite faeces by immunodiffusion and electron microscopy. As pointed out by Tovey et al (1979) natural exposure to dust mite allergens is quite different to formal bronchial or nasal provocation testing in that the rate of decomposition, although continuous, is very slow. Thus natural exposure to dust mite allergen would be in the form of a few faecal particles, perhaps 100 per hour, although only 10% of these would be expected to enter the airway.

**Clinical features**

The symptoms of allergic rhinitis vary from minor trivial inconvenience to profuse symptomatology to a point where symptoms adversely affect the quality and enjoyment of life (Wassermann, 1982). Certain individuals, when untreated, are incapacitated for several days.

**Seasonal rhinitis**

(1) The first symptom of the hayfever season is usually sneezing. In severe cases paroxysms of sneezing occur at frequent intervals throughout the day. Sneezing is probably largely due to histamine release acting through reflexes.

(2) Excessive fluid and mucus secretion (rhinorrhea) is believed to be the response of serous and seromucous glands to mast cells/basophil-derived mediators.

(3) Nasal obstruction or blockage is the result of vascular engorgement, that is vasodilatation and oedema.

(4) Itchiness of the nose, eyes, palate are common features.
(5) Tearing, itching and redness of the eyes together with some degree of periorbital oedema is usual in hayfever. Other symptoms include tightness of the chest (sometimes with wheezing) and a burning or raw sensation in the throat.

**Perennial rhinitis**

The symptoms of perennial rhinitis differ slightly from seasonal rhinitis largely as a result of long-standing nasal mucosal inflammation in the untreated situation. Sneezing, itchiness and nasal discharge are prominent, but the rhinorrhea may be more viscous or purulent depending on the degree of cellular recruitment. Conjunctivitis is far less frequent in perennial rhinitis than in allergic rhinitis. Perennial rhinitis is also accompanied by varying degrees of loss of smell (anosmia), loss of taste (ageusia) and symptoms associated with the eustachian tube (hearing defects and ear pain).

On examination, patients with perennial rhinitis or hayfever (during the pollen season) have pale, boggy nasal mucosa covered with thin mucus or mucopurulent secretions. Hyperaemic conjunctiva is noted in hayfever. Examination of the nose is best accomplished with the use of a nasal speculum. The colour of the membrane, the degree of oedema, the presence and types of secretions and tumours or mucosal ulcerations or the presence of polyps should be assessed.

**Laboratory investigations**

Confirmation or otherwise of a diagnosis of allergic rhinitis can be achieved by skin testing or, when appropriate, by the radioallergosorbent test. Skin testing is inexpensive, accurate, rapid and can be undertaken with a wide variety of antigens at a single skin testing session. Skin prick tests are preferred to scratch or intradermal tests because of their reproducibility, low instance of false positive reactions, less risk of systemic anaphylactic reactions, better accuracy (especially with food antigens) and patient preference. It should be remembered that a *positive histamine control* and a *negative diluent control* must always be used. A number of agents interfere with the immediate skin prick test. These include antihistamines and oral sympathomimetic agents. It should be remembered that astemizole (Hismanal), being a recently introduced selective H1 antagonist, has a long half-life and skin testing should be performed at least 28 days after stopping this preparation.

A radioallergosorbent test is indicated under several circumstances. These include the presence of dermatographism or extensive skin disease where skin prick testing is impractical and where, as stated above, the patient has been taking medication that interferes with the interpretation of the skin prick test. The radioallergosorbent test is less sensitive than the skin test and is relatively expensive taking several days to perform and is limited by the fact that in most routine laboratories only a limited number of antigen assays are available. The radioallergosorbent test is more quantitative than skin tests and this may be desirable when patients are being monitored for immunotherapy.

Nasal cytology is sometimes helpful in the differential diagnosis of nasal complaints. Samples may be obtained either by blown secretions or by gentle scrapings of the lateral nasal wall. The material is smeared on to a glass slide, fixed in ethanol and stained with May
Grunwald/Giemsa. The presence of eosinophils, neutrophils, basophils, mast cells, epithelial cells and bacteria should be recorded.

**Diagnosis**

The diagnosis of seasonal hayfever in the UK is usually straightforward and based on a seasonal history of symptoms together with a positive skin prick test to grass and/or tree and weed pollens. Occasionally, patients' symptoms are confined to the tree pollen season and this is confirmed with the appropriate skin tests. Seasonal hayfever in association with weeds or fungal antigens in the absence of pollen hypersensitivity is rare. On the other hand, many highly atopic individuals will have positive skin tests to grass, tree and weed pollens as well as fungal spores and their symptoms will extend sometimes from early April to the end of August.

The diagnosis of perennial rhinitis is sometimes more difficult, although a history of sneezing and rhinorrhoea all the year round together with positive skin prick tests to the house dust mite and/or animal danders is usually sufficient to make the diagnosis. It is important to recognize, however, that several forms of rhinitis may coexist.

In patients whose main symptom is obstruction of the nasal passages and where sneezing and rhinorrhoea are either minimal or absent, a diagnosis of a deviated septum, non-specific hypertrophy of the inferior turbinates, nasal polyps, rhinitis medicamentosa, atrophic rhinitis, or blockage with tumour or foreign body should be excluded. In patients whose primary symptom is posterior rhinorrhoea with sneezing and nasal obstructing being minimal or absent, acute or chronic rhinosinusitis, acute or chronic nasopharyngitis, or neoplasia may be responsible. Patients with sneezing with little obstruction or rhinorrhoea, but with negative skin tests or skin test which do not correlate with a clinical history, may have eosinophilic non-allergic rhinitis or primary nasal mastocytosis.

**Differential diagnosis**

It is important to recognize that several forms of rhinitis may coexist. Other causes of rhinitis apart from allergy include mechanical obstruction, hyper-reactivity to a wide range of non-specific irritants (dust, fumes, odours, strong light, cold air), immune deficiency (causing infection), mucous abnormalities, ciliary dyskinesia (primary or secondary), granuloma, malignancy, hormonal (menstrual, pregnancy), drugs (rhinitis medicamentosa) and emotional factors.

**Irritants**

Many individuals, particularly young women, experience obstruction and rhinorrhoea which is aggravated by a variety of minor environmental stimuli, such as exposure to dusts, fumes, odours, strong light, cold air, spicy foods, changes in humidity. In these patients, sexual arousal or defaecation may be associated with similar symptoms. This condition is a form of nasal hyper-reactivity but is sometimes termed ‘vasomotor instability' or 'vasomotor rhinitis'. In any event it is a diagnosis by exclusion. It should be noted that many of the stimuli may affect patients suffering from other forms of rhinitis, but by definition patients with vasomotor instability have no other primary cause for their reactivity. In some cases
systemically administered drugs caused obstruction and rhinorrhoea. These are usually agents affecting adrenergic regulation and include hypertensive agents (that is alpha-methyldopa) and beta-adrenergic agents and agonists. Hormonal medications and agents such as the contraceptive pill may be causative. Rhinitis of pregnancy is an important example of this problem as are drugs used in the treatment of hypo- and hyper-thyroidism. Obstruction and rhinorrhoea secondary to systemic medications are usually differentiated from rhinitis caused by the topical use of alpha-adrenergic medications capable of irritating the nose. Rhinitis secondary or sprays used for the treatment of other causes of rhinitis is secondary by nature and termed 'rhinitis medicamentosa'.

**Eosinophilic non-allergic rhinitis**

Some patients with perennial nasal symptoms of paroxysms of sneezing, profuse watery rhinorrhoea and pruritus of the nasopharyngeal and conjunctival mucosa have negative prick or intradermal immediate skin tests, negative serum radioallergosorbent tests to common inhalant allergen, and negative methacholine provocation challenges. These patients characteristically have eosinophilia in nasal biopsies with inactivated cilia, damaged nasal epithelium, thickened basement membrane and an oedematous submucosa infiltrated with plasma cells, lymphocytes, eosinophils and neutrophils, and the occasional mast cell. This form of inflammatory rhinitis seems to be responsible for about 15% of non-infectious non-structural chronic rhinitis in adults in certain parts of the USA. In one-third of the patients, there is associated nasal polyposis and sinusitis. Symptoms are aggravated by a variety of non-specific inciting agents such as odours, weather changes and irritants. In general, nasal obstruction and discharge and anosmia are more marked than the episodes of sneezing and conjunctivitic characteristic of allergic rhinitis. Some patients, particularly those with polyps, have exacerbation of symptoms after the ingestion of non-steroidal or anti-inflammatory drugs. The nasal mucosa appears pale and oedematous and is associated with thick yellow mucus. The underlying pathological mechanism is unknown. It is not thought to involve specific IgE although the activation of mast cells by non-immunological mechanisms has been suggested. The condition does not respond to disodium chromoglycate.

**Other conditions**

An uncommon form of chronic rhinitis is nasal mastocytosis in which mast cell are prominent in nasal scrapings. This disorder is most commonly seen in adults and is characterized by nasal obstruction and watery rhinorrhoea.

Atrophic rhinitis is observed particularly in patients who have undergone extensive nasal surgery although the condition was once common in the elderly. The nasal passages feel obstructed and irritated and dry. Infection and bleeding are frequent complications. The turbinates are small, the mucosa thin and there are diminished numbers of mucous glands and goblet cells, and scanty cellular infiltrate except in the presence of secondary infection where neutrophils are abundant.

**Allergy and sinusitis**

The association between allergy and sinusitis is essentially two-fold. First, allergic mechanisms may contribute to obstruction of the sinus ostia and, in that sense, represent a
predisposing factor. Secondly, perennial allergic rhinitis has some of the features of chronic sinusitis, particularly nasal discharge and obstruction.

In sinusitis, obstruction of the sinus ostium produces hypoxia leading to vasodilatation, ciliary dysfunction and mucous gland dysfunction. These cause transudation of fluid, stagnation and the production of viscid fluid, respectively, which in turn all lead to the production of retained thick secretions. If bacteria are carried into the sinuses, ideal situations exist for bacterial multiplication.

Complications

Seasonal rhinitis is sometimes associated with mild bronchial asthma and about 50% of patients with rhinitis have increased non-specific bronchial hyper-reactivity. It has been suggested that patients with rhinitis without symptoms, but who show non-specific bronchial hyper-reactivity in the range of asthmatic subjects, will be those who proceed to develop symptomatic asthma with time. However, the course of seasonal rhinitis is very variable. Some patients will spontaneously remit even after many years of symptoms and hayfever is common in the elderly. In children, rhinitis may predispose to an increased incidence of otitis media, serous otitis and chronic sinusitis. Otitis media and middle ear infections occur more frequently in allergic children. The pathophysiological changes in allergic rhinitis may lead to obstruction of the eustachian tube with dysfunction and middle ear effusion. Serous otitis is not an allergic disease but a frequent complication of nasal allergy particularly in children.

Treatment

The management of hayfever and perennial allergic rhinitis can be considered in terms of allergen avoidance, anti-allergic drugs and hyposensitization. The drugs can be subdivided into those for prevention and those for relief.

Allergen avoidance

Although the concept of allergen avoidance is simple, straightforward and obvious, in practice it is often very difficult to undertake. Generally speaking, it is virtually impossible to avoid exposure to pollens and spores during the appropriate seasons, unless the patient is prepared to travel to areas of the country where air-borne levels are insufficient to cause symptoms. On the other hand, inhaled air can usually be efficiently cleansed of allergens, although maximal efficiency often requires extensive and elaborate devices. Mechanical filters can remove particles of greater than 1-5 microm from the air but must have the capacity to recirculate frequently since multiple passages are required for effective cleaning. Devices such as electrostatic precipitators and high efficiency particulate air filters are particularly efficient. They can clean the air of 99% or more of particles. Individual hayfever helmets can be purchased (the Hincherton hayfever helmet). The device consists of a plastic globe (spaceman's helmet) connected to a small battery-driven air filter. Apart from being somewhat unsightly the helmet heats up in direct sunlight (the greenhouse effect). Hayfever sufferers should obviously avoid prolonged exposure to the countryside and should drive with their car windows closed. Generally speaking, air filtering systems in cars do not filter out pollens. Wearing glasses may prevent pollen settling in the eyes and thus prevent discomfort.
Exposure to mould spores can sometimes be reduced. For instance, patients should avoid barns and mowing grass or raking leaves since high concentrations of mould spores may be found in these situations. Garden sheds and cabins which have been closed for several months may have a high mould spore concentration as will the basement of houses and certain foods and beverages. As with pollens, air conditioning and air filtration systems are the only way of reducing exposure on a 24-hour basis. In theory, avoidance of domestic animals such as cats and dogs is readily manageable, although sensitive patients are often unwilling to accept such obvious advice. It should be remembered that animal danders, particularly cat, dog, guinea-pig, mouse, rabbit etc (allergens derived from) may take 6 weeks or more of repeated cleaning to remove from a house. Most allergists do not allow asthmatic children to continue to live with a domestic animal to which they are sensitive. Similarly, patients with symptoms in association with laboratory animals should continue to avoid working with them unless there are extremely efficient filtering devices to avoid exposure.

The role of allergen avoidance in association with house dust mite remains controversial. One of the problems is that it is still unclear whether the recommendations usually given to patients or parents for cleaning the house, particularly the bedroom are effective at reducing the allergen content of the dust. It has been assumed that the bedroom is the single most important source of allergen exposure, but there is no reason for believing this and, in centrally heated houses, high levels of dust mite allergen are often found in living room floors or furniture. In order to reduce allergen exposure successfully, it may be necessary even to change flooring, furniture and cleaning practices in the whole house. With the development of better assays for the allergen content of dust samples or room air it should be possible to give better advice on methods for cleaning houses. Killing of mites in dwellings by temperature alone is seldom feasible, at least in temperate climates. In colder regions, it is possible to expose mattresses, blankets, pillows and eiderdowns to the necessary -18°C for two days. The same effect could be accomplished by heating to more than 45°C for 2 hours. Humidifiers should be avoided in homes of house dust mite allergic patients. Materials which conceal mites and prevent cleaning are obviously more heavily infected, that is carpets compared to plain floors and mattresses with inner springs or kapok compared with foam mattresses. Unfortunately repeated vacuuming does not remove mites completely from carpeting and vigorous vacuuming of bedding is also relative ineffective at getting rid of mites. Mites are highly resistant to germicides. Gamma-benzene hexachloride (Lindane) kills mature Dermatophagoides pteronyssinus, but at unacceptably high concentrations. Natamycin, a fungicide with less toxicity, appears to have some effect on symptoms in an uncontrolled clinical trial. It acts on fungi which are important for the growth of Dermatophagoides sp.

Thus, the dust avoidance measures can be summarized as follows: smooth, uncluttered easily cleaned surfaces are recommended and bare floors and walls are suggested. Small objects such as toys, books and records should be placed in drawers or closed cabinets. If carpeting is unavoidable low pile types are preferred. Pillows and mattresses should be enclosed in air-tight plastic or fabric encasing. Feather and down pillows should be avoided and synthetic pillows should be replaced every 2-3 years. A dust mask should be worn when cleaning the room.
**Anti-allergic drugs**

The most effective drugs for the relief of symptoms are antihistamine preparations, whereas local corticosteroids and disodium cromoglycate are effective for prevention. Systemic medication with histamine H₁ antagonists lessen itching, sneezing and eye symptoms, but have relatively little effect on nasal blockage. The classic antihistamines cause sedation which many patients find unacceptable. This problem has been largely overcome by the introduction of the selective H₁ antagonist terfenadine (Triludan) and astemizole (Hismanal). These compounds are largely free from the anticholinergic sedative effects of the classical antihistamines. They rarely cause sedation or psychomotor impairment and do not potentiate the effects of alcohol or benzodiazepines. Astemizole has the advantage of a once daily dosage but it takes several days to achieve its full effect. The drug is given as a single daily dose of 10 mg, but a loading dose of 7 days of 20-40 mg is sometimes recommended. Astemizole can cause weight gain. The very long half-life of astemizole (21 days) has caused concern. The recommended dose of terfenadine is 60 mg twice daily. The drug acts fairly rapidly, but there is some evidence that suggests that tachyphylaxis can occur with prolonged usage (that is several days). Neither astemizole nor terfenadine are recommended in pregnancy.

Many patients with relatively mild hayfever find that the older antihistamines, such as chlorpheniramine (Piriton) and azatadine (Optimine), are quite satisfactory, especially when taken at night. There is in fact a bewildering array of classic antihistamines on the market, some of which are combined with decongestants (that is Actifed (triprilodine and pseudoephedrine), Dimotapp (brompheniramine and phenylephrine)). Oral sympathomimetics alone or in combination with an antihistamine are contraindicated since they are less efficient than topical treatment and may cause serious systemic adverse effects. Drugs for relief include corticosteroids and disodium cromoglycate. At the present time four different corticosteroid sprays are available for the treatment of allergic rhinitis. These are beclomethasone (Beconase - as a freon propelled pressurized aerosol, or as an aqueous spray), budesonide (Rhinocort - freon delivery only) and flunisolide (Syntaris - aqueous spray only). These preparations successfully control nasal symptoms and the four preparations available seem to have similar efficacy and are equally acceptable to patients. Twice daily dosage is now advised and, once patients are stabilized, they may be able to reduce the dose to once daily. With the recommended dose local corticosteroid side-effects are minor and systemic effects are not a problem. Effects related to the delivery system have to be considered since freon delivered aerosols may cause drying and crusting of the nasal mucosa and, occasionally, slight bleeding.

Disodium cromoglycate is available for nasal application as an insufflation, a spray or as drops (Rynacrom). The eye drops have the trade name Opticrom. Disodium cromoglycate is only effective if used prophylactically and has to be applied at least four times a day. In seasonal rhinitis it is probably as effective as corticosteroids as a prophylactic agent.

Oral corticosteroids or ACTH are sometimes indicated although symptoms are rarely severe enough to warrant shirking their untoward effects. Short courses of oral prednisolone are effective and can be titrated to individual requirements, whereas injectable depot preparations are expensive and their dosage inflexible.
Ketotifen (Zaditen) causes sedation and weight gain and has no particular advantages. The manufacturer's recommended dose is one tablet twice daily.

**Hyposensitization (immunotherapy)**

This should be considered only in patients who fail to respond adequately to anti-allergic medication. It should be remembered that the adverse effects can be life-threatening and doctors have to be prepared to deal with general anaphylaxis. Patients should be observed for up to one hour after each injection. (Recently a controversial recommendation stated that observation should be continued for 2 hours.) Benefit appears to be dose dependent and maintenance therapy over 3 years of treatment is usually advised. The subject of hyposensitization has been admirably reviewed by Norman (1980) and Adkinson (1982).

Serial injections of pollen extracts were first used by Noon, and Freeman, in 1911, with the aim of eliminating 'the toxic effect' of pollens. Despite its extraordinary popularity, it is a sad fact that 75 years later there is still a considerable area of ignorance regarding this form of treatment. Views range from great enthusiasm to considerable scepticism. The treatment of patients with IgE-dependent allergic disorders by serial injections of allergic extracts is an attempt to provide some protection from natural exposure to the antigens which induce the untoward symptoms. For this reason the term 'immunotherapy' is preferred to desensitization and hyposensitization. (In fact there is little evidence that target cell reactivity is diminished and, therefore, the terms 'desensitization' and 'hyposensitization' are probably inappropriate.) Desensitization is more appropriately applied to situations in which adverse reactions to agents, such as insulin or penicillin, are treated by rapid administration of the agent over a period of hours in order to administer therapeutic amounts of the material in question. In 1954, Frankland and Augustin carried out the first placebo-controlled trial of desensitization employing both crude and partially purified grass pollen extracts in patients with hayfever and seasonal asthma. The active preparation was significantly better than placebo in relieving symptoms of rhinitis and asthma, although it was confirmed that a large proportion of patients (30-40%) had a potent placebo response. Lowell and Franklin (1965) were the first to provide evidence that immunotherapy was specific for the allergen used. Similar evidence in ragweed versus grass pollen sensitivity was provided in the 1970s. Generally speaking there is good evidence that immunotherapy is effective in seasonal rhinitis due to grass pollens, ragweed pollens and mountain cedar pollens and probably also to the house dust mite, although results from house dust mite studies vary considerably. There is reasonable evidence of benefit for allergic asthma, especially in children, but many remain sceptical. The one single absolute indication for immunotherapy is in those at risk from local or general anaphylactic reactions to venoms of Hymenoptera (insect sting allergy).

**Clinical indications**

Specific immunotherapy has no place outside of allergic rhinitis, allergic asthma and sensitivity to Hymenoptera. There is evidence that oral hyposensitization is ineffective and there is no evidence for the use of bacterial vaccine in asthma. There are a limited number of controlled trials of immunotherapy with moulds and animal allergens, but there has been insufficient work in this area to recommend this form of treatment for these conditions.
Selection of patients for immunotherapy

For reasons of time and expense, the decision to embark on a course of immunotherapy needs to be weighed carefully. Patients must have a convincing history that natural exposure to the aero-allergen induces clinically significant allergic symptoms. In certain instances provocational evidence is also required. Detection of IgE antibody specific for the relevant environmental allergen is also essential. Allergic rhinitis (or asthma) in which the symptoms correlate with skin tests or the radioallergosorbent test results and the condition is severe and non-responsive to other treatment, is usually an indication for specific immunotherapy. The requirement for positive skin tests to the relevant allergen is absolute because there is no evidence that desensitization works in patients who do not have positive skin tests.

Finally adequate avoidance measures and/or well-tolerated symptomatic medications should also be established. In general, immunotherapy is restricted to patients with moderate to severe IgE-mediated symptoms attributed to aero-allergens which are unavoidable for more than two months of the year. Immunotherapy may be justified in less sensitive patients with limited sensitivities if drug regimens are either ineffective or poorly tolerated or where for reasons of job or profession (acting, broadcasting etc) total release from symptoms is essential.

What immunotherapy involves

Immunotherapy involves serial subcutaneous injections of immunogenic extracts of relevant allergens administered in progressively increasing doses until a potent immunogenic dose or maximal tolerated dose is achieved. Usually a potent immunogenic dose is one which is significantly greater than that required for a specific IgG response. For most allergenic extracts the immunogenic dose is poorly defined but is probably not reached until the concentrations of 1 to 100 w/v or stronger are employed. Thus sufficient allergen must be administered to induce the immunological changes necessary for successful therapy. Putting it another way, once the decision has been made to use this form of treatment, sufficient quantities have to be administered. Relief of symptoms is related to higher doses and/or prolonged treatment and, therefore, the achievement of a maximum tolerated dose is a therapeutic goal once the commitment to undertake immunotherapy has been made. The maximum tolerated dose is the highest dose which can be administered without producing unacceptable side-effects. Some degree of local discomfort and swelling around the injection site normally accompanies immunogenic doses in highly sensitive patients and, therefore, must often be endured if therapeutic success is to be achieved.

Theories regarding the mode of action of immunotherapy

The way in which immunotherapy prevents allergic symptoms is still incompletely understood. Symptomatic relief can be achieved after 6-20 weeks of intensive treatment provided that sufficient quantities of allergen are administered. During the first year of immunotherapy, skin test reactivity to the allergen in question changes little, if any, and antigen-induced leucocyte histamine release is likewise unaffected. These observations suggest that the cellular reactivity of IgE-laden skin mast cells and circulating basophils have not been
appreciably altered. Thus, immunotherapy does not appear to desensitize the effector cells of the allergic response.

The concentration of circulating IgE antibody, assessed by radioallergosorbent assays or similar techniques, usually increases during the early part of immunotherapy. Thereafter, the levels gradually decline and it may take several years before the serum IgE antibody levels return to pretreatment levels. Thus neutralization or depletion of circulating IgE by injected allergen is also an unlikely explanation of the effects of immunotherapy. Likewise, the induction of immunological tolerance is an unlikely explanation since immunotherapy is associated with large amounts of serum-blocking antibody predominantly IgG. This IgG antibody competitively competes for antigen with cell-bound IgE on basophils and mast cells and may thereby induce a significant shift of the dose responsive to relevant environmental allergens. Although a protective effect of passively effused IgG antibody has been demonstrated in insect allergy, there is as yet no direct evidence which indicates that IgG 'blocking antibody' is principally responsible for symptom amelioration observed in inhalant allergy immunotherapy. Quantitatively blocking antibodies do not correlate very well with the response to treatment and, in some instances, patients who have responded well clinically produce very little IgG antibody. Blocking antibodies also increased in nasal secretions but the rise is far less than in the serum. Calculations of the concentration of allergen and quantities of blocking antibody suggest that binding of allergen by IgG antibody might not be rapid enough to prevent induration with mast cells. The association between IgG antibody levels and symptom amelioration is weak and subject to numerous exceptions indicating that there is not a simple quantitative relationship between IgG antibody and protection from allergic exposure.

When immunotherapy is continued for 3 or more years, additional immunological changes are observed in many patients. Serum IgE antibody begins to decline slowly suggesting possible suppression of IgE antibody resynthesis. However, the evidence that alterations to suppressor cells which control IgE production play a role in human desensitization is very poor. There are no studies that show that a 3-month course of injections lead to significant reduction in serum IgE antibody or in skin tests of sensitivity. In most studies, after 3-6 months of treatment serum IgE antibody has risen slightly. On the other hand there is evidence that long-term immunotherapy is associated with reductions in IgE antibody, reduction in seasonal boost of IgE antibody and, possibly, a long-term decrease in skin sensitivity. Most studies have been too short to fully document these effects. There are a number of T-cell-related effects and many studies have established that the response in vitro of T cells to allergen is changed after desensitization. These changes have been documented in a variety of ways; they include an increased production of macrophage inhibiting factor (MIF), reduced lymphocyte proliferation, increased suppressor cell effects in vitro and changes in the number of suppressor T cells. The interpretation of these findings in vitro is presently unclear and is unlikely to be related to suppression in vivo of IgE since most of the T-cell effects were observed at a time when IgE antibody concentrations were not altered. There are other ways in which T-cell changes might be relevant to the success of immunotherapy. Thus T cells might directly contribute to chronic symptoms, produce agents which affect mediator release and may be involved in basophil chemotaxis.

It was seen nevertheless that complete suppression of IgE antibody would presumably lead to a 'cure' of the disease. For this reason there have been many efforts to accelerate the
decline of IgE antibody by manipulating the immune system with a variety of modified allergens.

**Allergen extracts used in specific immunotherapy**

In general, grass weed and tree pollen extracts are the most reliable and well-standardized extracts for immunotherapy with inhalant allergens (Dreborg, Einarsson and Longbottom, 1986). Mould spore extracts have considerable variability in the quality and standardization of fungal extracts which limits their usage. Similar problems are true for animal epidermal (dander) extracts. In general, animal dander allergy is readily managed by avoidance so that immunotherapy is only rarely indicated, for example in occupational exposures. House dust 'extracts' are highly variable in composition. House dust mite extracts have been used successfully in certain instances. The other pollen subjected to controlled study is mountain cedar (because of standardization problems the *optimum* dose is still difficult to describe but a dose of crude, unaltered extract adequate to ensure symptomatic relief will often be accompanied by a considerable risk of a large local or systemic allergic reactions).

Most studies of house dust sensitivity gave mixed results. The bacterial vaccines have consistently failed to be efficacious in a number of controlled studies of the treatment of asthma related to respiratory infections, a recent study showing occasional bronchial reactivity by immunization with cat dander on very few subjects. Although there have been a large number of placebo controlled, and mostly blind, studies on the effect of injection immunotherapy on house dust mite or house dust allergy, the results have been variable. Two showed no effect at all, the rest showed varying effects ranging from clinical improvement without immunological changes to effects on both immunological variables and symptoms, including bronchial hyper-reactivity. The general unimpressive lack of effect might have been due to the selection of patients, the allergen doses administered, the duration of treatment and the reliability of objectivity of the parameters for assessment.

In the USA, the high prevalence of positive skin tests to a cockroach allergen have been demonstrated in indigenous asthmatic populations. The use of cockroach allergen for immunotherapy is still under evaluation. Substances not recommended for specific immunotherapy include foods, whole body extracts of stinging insects, feathers, synthetic material (for example kapok), bacterial extracts, gums and glues, enzymes (for example subtilis) and occupational allergens.

Aqueous allergen extracts are still the most commonly employed materials for immunotherapy. They are the most economical form of therapy and less likely than precipitated and/or pyridine extract materials to contain allergens that are denatured or lost by processing. Aqueous extracts may be purchased commercially in buffer saline in lyophilized form or 50% glycerin. Potency is usually expressed on a weight per volume basis or in protein nitrogen units (PNU) or in biological units. For many pollen allergens 1 mL of a 1:20 allergen extract is approximately equivalent to 25,000 PNU. A predictable relationship between PNU and weight per volume ceases to exist for non-pollen allergens, especially for foods and fungal extracts where the protein extracted from the given weight of material is highly variable.
Applications of immunotherapy

There is no evidence that mixtures of several allergens are efficacious, although within the 'family' of allergens there is considerable if not complete cross-reactivity. For instance, an extract of Timothy grass is probably as effective as a mixture of several grass pollens but combining, say, Timothy with house dust mite or even Timothy with tree and weed pollens is to be discouraged. In any event, any allergen to which the patient is extremely sensitive should be given separately so as not to eliminate the amount of other allergens given due to recurrent large, local or systemic reactions. Allergens are given subcutaneously once or twice weekly until the maintenance dose has been achieved or the maximal tolerated dose is achieved. The interval between injections is then increased to 2 weeks, and, if clinical improvement continues, the intervals may be 3 or 4 weeks. A suitable extraction concentration for initiation of therapy varies from 1:10,000 (w/v) to 1:25,000 (w/v) depending on the sensitivity of the patient. This is one-tenth or one-hundredth of a biological unit. Twenty to 35 injections are usually required to reach a satisfactory maintenance level of 0.2-0.5 mL of 1:100 (w/v) or 1 mL of 100,000 biological units per mL. In sensitive patients, the dosage schedule may require individual modification because of local or systemic reactions. Once maintenance dose is achieved it is usually continued perennially; however, for pollens it is the practice in the UK to recommend that allergen therapy is discontinued during the pollen season and recommenced the following winter. There appears to be no rationale in this point of view and co-seasonal maintenance injections are given by some practitioners. Sometimes it is necessary to reduce doses temporarily during the major allergen season if reactions to immunotherapy appear to occur more frequently. Some adverse reactions are to be expected in the course of immunotherapy in a highly sensitive patient, but are not predictable by any known immunological or clinical tests. Some practitioners prefer a cluster regimen to minimize the number of attendances at the clinic. Using this regimen maintenance levels could be achieved by week 5. There is some debate as to whether conventional or semi-rush regimens are associated with more or less side-effects. Sometimes a rush regimen is used in which maintenance dose is achieved over a 24-hour period. This is always undertaken under strict supervision on an in-patient basis.

Successful immunotherapy is usually apparent within 6 months after achieving a maintenance level dose. Many patients continue to improve during the second and third year of treatment and, therefore, 3 years' treatment is usually advised for all forms of immunotherapy. Very sensitive patients may respond immunologically and clinically even though the maximum tolerated doses are substantially less than the usual maintenance levels. When immunotherapy is discontinued, symptoms usually return over a period of several years although some patients remain symptom free. Unfortunately, there is no way to predict in advance which patients will benefit most from immunotherapy, although most patients who receive adequate doses can expect to experience fewer symptoms and require less anti-allergic medication. Environmental control should still be instituted. There is no practical way of monitoring immunological variables in patients undergoing immunotherapy for aeroallergens. Allergen sensitivity by skin tests is unlikely to change within the first years of treatment even in the face of a good clinical response. Serum IgE antibody levels determined by radioallergosorbent test and measurements of serum IgG antibodies may be useful in venom immunotherapy. Conjunctival reactivity is sometimes recommended as a useful monitor for efficacy.
Immunotherapy should be discontinued if the patient cannot be advanced to a satisfactory dose level because of persistent adverse reactions or if the patient is poorly compliant and fails to receive injections regularly. If no clinical improvement is observed after 3 years of maintenance, or if the patient is essentially symptom free after two consecutive years, or the patient has been on immunotherapy continuously for 6 or more years, then these are all indications for stopping treatment.

There are many reasons for treatment failure. These include inadequate environmental control and avoidance of allergens particularly in the home, and the development of new sensitivity since the initiation of treatment. The allergen may be administered in insufficient doses or potency of the treatment materials may decline. An incorrect initial diagnosis should also be considered. An inadequate trial period is a possibility as is the masking of partial benefit due to progression of the underlying disease.

**Allergen immunotherapy with modified allergens**

Immunotherapy results in the reduction of symptoms but does not eliminate them, and patient must not have false expectations. During recent years there have been a number of attempts to improve immunotherapy with the use of modified allergens, with the hope of decreasing allergenicity in the side-effects but maintaining immunogenicity.

Some examples of major directions and attempts to improve allergen immunotherapy include water-in-oil emulsion (to reduce rate of allergen absorption), purification of antigens, alteration of antigen to reduce allergenicity but retain immunogenicity (allergoids), preparation of agents that selectively suppress IgE antibody production (antigen:polyethylene glycol). Water-in-oil emulsions were abandoned because of the production of sterile abscesses and the danger of myeloma. Purified antigens are expensive and do not reduce the risk of anaphylactic-type reactions. In Europe, pyridine-extracted aluminium hydroxide-absorbed extracts have largely replaced aqueous extracts during the last decade.

Too little is known about dose regimens and the maximum doses with conventional aqueous, depot and modified extracts. Modified allergens, showing less allergenicity, with retained immunogenicity have less side-effects. There is no evidence that any of these components have a suppressive effect on the ongoing IgE response. Local immunotherapy, that is local nasal and oral administration, has not proved to be effective. Induction of anti-idiotypic antibodies may be a possibility in the future.
Chapter 8: Acute and chronic inflammations of the nose

Neil Weir

Acute infective rhinitis

The term 'acute infective rhinitis' is taken to mean an acute viral or bacterial infection of the nasal mucous membrane. It is exceedingly common and is a manifestation of the common cold, influenzal infections of the upper respiratory tract, the exanthems and certain specific infections. It can also occur as a secondary response to local irritants and trauma.

The common cold (coryza)

Incidence

The common cold is probably the commonest viral infection in man. The incidence is variable but it is estimated that the average young adult has two to three colds a year. Children and young adults are particularly susceptible to rhinovirus infections and women may experience more infections than men, perhaps because they are in closer contact with young children.

Predisposing factors

Climate

Colds occur all the year round but in temperate climates they are more common in winter than in summer. Rhinovirus infections are more prevalent in autumn and spring and coronavirus infections seem to occur mostly from December to March.

Environment, temperature, chills and humidity

Kerr and Lagen (1933-34) exposed groups of susceptible men in the same room, under perfect conditions of humidity, temperature and ventilation, with subjects in the early stages of a cold, and in some they even inoculated cold filtrates into the conjunctival sac without obtaining a single transfer, suggesting that an ideal environment increases resistance.

There is a widespread belief that chilling may precipitate a cold in an individual; however, attempts to demonstrate such an effect experimentally have given negative results (Andrewes, 1950). Chill may act in two ways: by lowering general resistance to infection; and by causing reflex vasoconstriction of the nasal mucous membrane. The normal temperature of the mucous membrane of the nose has been shown to vary between 33 and 34°C. Chilling of the body surface may reduce the temperature of the nasal mucosa by as much as 6°C (Mudd, Goldman and Grant, 1921). The optimal relative humidity of the atmosphere is 45%. A lowering of relative humidity to 15%, as may easily occur when the relatively dry cold outside air in winter is heated indoors by radiators, withdraws more water than the nasal mucosa can supply, and causes drying of the mucous blanket. Excessive humidity is also harmful, as it reduces the evaporation of sweat from the skin and, owing to the high
conductivity of water vapour, a slight lowering of temperature produces severe chilling, with the effects described above.

Hope Simpson (1958a,b) demonstrated a striking correlation between increase in the frequency of colds in a group of families and falls in the temperature of the soil. Humidity also affects the survival of viruses (Tyrrell and Bynde, 1961). Common cold viruses prefer high humidity. Influenza viruses survive better in dry air.

**Immune status**

Local immunity in the nose is primarily the result of concentrations of IgA and IgG, which are normally present in nasal secretions in the ratio of 3:1 compared with 1:5 in serum. IgA is produced in response to local antigen stimulation but does not combine with complement and therefore is unable to lyse bacteria; it is, however, effective as a viral neutralizing substance. IgA is a relatively short-lived antibody the half-life of which has been estimated to be 13 days but, in practice, is more likely to be measured in minutes because, although there are very large numbers of IgA-producing plasma cells in submucosal tissues, it is soon carried away in mucous secretions. It is thus not uncommon to find reinfection with the same virus serotype in consecutive years. Failure to produce secretory IgA occurs in approximately one in 800 persons who live without ill effects. Patients with generalized hypogammaglobulinaemia, however, have frequent infections of the upper respiratory tract (Wilson and Montgomery, 1980).

**Nutrition and vitamin deficiency**

The lowering of resistance by hunger and undernutrition was shown by Cruickshank (1942), who found that the death rate in measles, pertussis, influenza and bronchopneumonia among poor children was five times greater than among those better off. Deficiencies in vitamins A, C and D are said to increase susceptibility to infection but the claim that vitamin C is effective in preventing colds is not supported by controlled trials (Andrewes and Tyrrell, 1965).

**Fatigue, fitness and exercise**

Locke (1937) assessed the fitness of subjects by their oxygen consumption under standard exercise, and found that 64% of those with fitness above 0.6 had only one cold a year, while 80% of those below 0.5 had four colds a year. However, colds very often hit the man who is feeling very fit on his return from holiday (Andrewes and Tyrrell, 1965).

**Nasal obstruction**

Deviation of the nasal septum, hypertrophy of the turbinates, enlarged adenoids, polyps, scars and adhesions all interfere with ventilation and the free passage of air through the nasal chambers, and with the secretion and movement of the mucous blanket, and thus predispose to infection.
**Foci of chronic infection**

Foci of infection in the sinuses, nasopharynx or pharynx, by decreasing tissue resistance, favour infection. The more important of these in children are chronic adenoiditis, tonsillitis and sinusitis and, in adults, chronic sinusitis and tonsillitis.

**The pH of nasal secretion**

A drift to the acid side is associated with few bacteria, while an alkaline drift is associated with many bacteria in the nasal secretion. Rhinoviruses are destroyed by an acid pH (Ketler, Hamparian and Hilleman, 1962).

**General diseases**

Any general disease, but particularly renal, hepatic and blood disorder, diabetes mellitus and tuberculosis, may lower general resistance to colds.

**Causative agents**

**Viruses**

In general it may be said that, in communities, the causative agent of the common cold is ubiquitous, but that infection occurs only when an individual's resistance is lowered, or when is subjected to an overwhelming concentration and virulence of the causative agent. It is generally accepted that the common cold is due to infection with filterable viruses, followed by secondary infection with bacteria.

In spite of the rapid advances in virology and the isolation, identification and even culture of many viruses, it is still uncertain what proportion of respiratory illnesses is caused by them. The viruses responsible for colds are listed in Table 8.1 (Reed, 1981).

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<tr>
<th>Virus</th>
<th>Serotypes</th>
<th>Proportion of colds (%)</th>
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<tr>
<td>Rhinovirus</td>
<td>89 different types, probably more</td>
<td>50</td>
</tr>
<tr>
<td>Coronavirus</td>
<td>3 or more types, and possibly subtypes</td>
<td>15-20</td>
</tr>
<tr>
<td>Influenza</td>
<td>A and B and their subtypes; C</td>
<td>Together</td>
</tr>
<tr>
<td>Parainfluenza</td>
<td>Types 1, 2, 3, 4</td>
<td>about</td>
</tr>
<tr>
<td>Respiratory syncytial</td>
<td>One type</td>
<td>15-20</td>
</tr>
<tr>
<td>Adenovirus</td>
<td>36 types, but only about half of them causing respiratory tract infection</td>
<td>10-20</td>
</tr>
<tr>
<td>Other viruses</td>
<td>Includes some enteroviruses, other known viruses, and perhaps some unknown</td>
<td>10-20</td>
</tr>
</tbody>
</table>
Each of the viruses listed in Table 8.1 can be said to be associated with its own 'typical' clinical effect which, for rhinoviruses and coronaviruses, is the common cold syndrome. Influenza viruses, respiratory syncytial viruses are, however, known to cause more serious infections.

**Rhinoviruses**

These are members of the picornavirus group and are thus biologically related to polioviruses and other enteroviruses, and to foot and mouth disease virus. They are about 25 nm in diameter and incorporate protein subunits in which the antigenic specificity of each of 89 or more types is incorporated. They have an optimal growth temperature of 33°C. Nasal swabs and nasal washings produce the best specimens for culture on human embryonic fibroblasts such as WI-38. Identification of the serotype of a rhinovirus is difficult because of the large range of types. However, antisera can be used against 89 types. A group-reactive serological test for rhinoviruses is not available and therefore retrospective diagnosis of rhinovirus infection cannot easily be established using paired sera collected in the acute and convalescent stages of the illness.

**Coronaviruses**

Coronaviruses were first classified as a distinct group in 1968 but are still not fully understood because of the technical difficulties in isolating them from clinical specimens. They are RNA-containing viruses about 100-120 nm in diameter. Two early isolates were named 229E and OC43. The former can be grown in tissue culture, but the latter needs organ cultures of human embryonic nasal epithelium or trachea. Serological tests available include complement fixation and neutralization tests for 229E, and complement fixation and haemagglutination inhibition for OC43. Coronaviruses cause typical colds. The incubation period is slightly longer than for rhinovirus colds.

**Mode of transmission**

**Droplet and dust**

In talking, sneezing and coughing innumerable infected droplets are sprayed out which fall to the ground at distances of 0.9-1.8 m. Bedmaking, house dust and the manipulation of handkerchiefs also contribute large numbers of airborne particles (Dumbell, Lovelock and Lowbury, 1948).

**Droplet nuclei**

Droplet nuclei are small droplets which evaporate as they fall, and shrink to less than 0.1 mm in diameter. In this form they remain suspended in the air as mist, and drift on the air currents for as long as 2 days, and thus have a much wider range than that of droplets (Wells and Wells, 1936). These will transport viruses, but are too small to carry the larger bacteria.
Contact

The causative organism may be transmitted by kissing, food, fingers and fomites.

Pathology

In the earliest stage of invasion there is transient vasoconstriction. This is followed by a vasodilatation, oedema and increased activity of the seromucinous glands and goblet cells.

Leucocytic infiltration of the tissues follows, with swelling and desquamation of the epithelial cells. The secretion is at first clear, watery and sterile, with a few epithelial and pus cells, but later it becomes coloured and viscid, stiffens on a handkerchief and contains may pus cells and bacteria. The toxins produced in the mucous membrane are swiftly taken up by the lymphatics, and passing through the cervical lymph glands and ducts reach the blood stream. Resolution takes place by a reversal of these processes, and by proliferation of the remaining tissue cells to replace those that have been destroyed.

The lysozyme content is reduced in the early stages. The average pH of the nasal mucus lies between 5.5 and 6.5. During an acute rhinitis the reaction becomes alkaline, and during resolution it returns to neutral.

Bacteriology

Cultures from the normal mucous membrane of the posterior areas of the nose are usually sterile, provided that they are not contaminated from the vestibule and anterior areas. Cultures from the anterior nares show staphylococci in 43% of normal individuals.

In the first 3 days of a common cold the cultures from the posterior areas may be sterile, but after the third day pure or mixed cultures of streptococci, pneumococci, Haemophilus influenzae, or staphylococci are often shown (Tweedie, 1934).

Clinical picture

The course of a cold may be described in four stages.

(1) Prodromal or ischaemic stage. This lasts for a few hours, and represents the stage of local invasion and general nasal ischaemia. The familiar hot, dry or tickling spot is felt at the site of the invasion, while the general nasal airway seem unusually patent.

(2) Early reaction and irritation. The infection, which is at first localized, spreads to the adjacent mucous membranes over the surface and by way of the lymphatics. This process may take a few hours or days. The site of invasion is often the first to recover, while the disease is still active in those areas which have been affected later. The throat is dry and sore on swallowing, and there is sneezing, watery nasal discharge and obstruction. The mucous membrane is red and swollen. General symptoms of mild toxaemia and fever now appear.
(3) Stage of venous stasis and secondary infection. After the second day, the colour of the mucosa becomes dusky, with a bluish tinge, the discharge thickens, diminishes and becomes mucopurulent. The obstruction and toxaemia are at their maximum.

(4) Resolution. The symptoms and signs gradually diminish, and after 5-10 days recovery takes place.

**Complications**

*Nasopharyngitis and pharyngitis*

The nasopharynx and pharynx are invariably infected to some extent in every cold.

*Sinusitis*

Sinusitis is one of the most common complications, but the sinuses are not invaded during the course of an uncomplicated cold.

*Pharyngotympanic salpingitis, otitis media and mastoiditis*

The infection ascends from the nasopharynx, invading the pharyngotympanic tube, middle ear and mastoid cells in sequence. It may be arrested at any point of the ascent.

*Lymphadenitis*

This is usually transient, and affects the retropharyngeal and deep cervical group.

*Tonsillitis*

A mild inflammation usually accompanies a cold, but parenchymatous or follicular tonsillitis is considered as a complication.

*Lower respiratory complications*

Laryngotracheitis, bronchitis, pneumonia and asthma constitute the group of lower respiratory complications.

*Gastroenteritis*

This complication is rare except in infants.

*Other complications*

Nephritis and rheumatism are allergic and toxaemic manifestations.

**Diagnosis**

Other causes of rhinitis should be excluded (see Chapter 6).
Laboratory diagnosis may be made retrospectively by taking acute and convalescent sera and comparing the antibody levels. However, more rapid diagnostic techniques such as immunofluorescence are now available. In the course of an infection large numbers of infected cells are sloughed off into the respiratory secretions. These cells will carry markers or antigens specific for the virus involved and can be recovered by nasal swabs or washings, stained using specific antiviral antisera labelled with a fluorescent dye, and seen by fluorescence microscopy. This technique can now be applied to a range of common respiratory viruses including influenza viruses A and B, parainfluenza viruses 1-3, adenovirus and respiratory syncytial virus.

Prophylaxis and vaccines

Based on the observation that rhinovirus infections may possibly be spread by the manual route, thorough hand washing with soap and water will remove the virus from contaminated hands. Avoidance of fingerling the nostrils and conjunctiva could reduce the chance of self inoculation.

Because of the antigenic diversity of respiratory viruses, with the exception of vaccines for influenza A and B infections, there are no suitable vaccines against the other respiratory viruses. No specific antirhinovirus compound has yet shown sufficient activity in man to merit further development but, nevertheless, work continues in this field.

Wide spectrum antiviral prophylaxis or therapy would have considerable theoretical advantages and the natural antiviral substance, interferon and its inducers, would seem to be an ideal solution. Double-blind placebo-controlled studies using interferon in rhinovirus-infected volunteers have given encouraging results. It is important though to ensure that any form of antiviral therapy should be of very low toxicity before its use in common colds can be justified.

Treatment

There is no known specific treatment for the common cold, but general and local supportive and palliative treatment can mitigate the severity and complications. There are so many different varieties of colds, so many different individual reactions to them, and so many different individual responses to treatment, that no hard and fast therapeutic rules can be laid down.

General treatment is directed to providing the best conditions for rest, both general and local, and at the same time supplying heat and the maximum blood flow to the infected tissues. Unfortunately, the majority of patients are not willing to submit to full-scale treatment for a cold of moderate severity, and modifications must be made, according to the circumstances.

Complete rest, both general and for the upper respiratory tract, necessitates confinement to bed in an even temperature of 18-20°C, with a humidity of 45%.
Heat, both local and general, is provided at first by a hot bath. Inhalations of menthol or tincture of benzoin (BP) (one teaspoonful in a pint of steaming water), may be soothing and will apply heat directly to the mucous membrane of the nose.

Analgesics and antipyretics, such as acetylsalicylic acid, may be valuable for the general malaise, aching and feverishness of the cold. Codeine compounds are more effective as sedatives. Both should be combined with a copious fluid intake.

Antihistamines have not been shown to reduce fully or abolish the symptoms of colds, but they can be particularly effective in the allergic patient who is often unduly susceptible to colds. Antihistamines can be usefully combined with an analgesic.

Alcohol is a sedative which is the chief justification for the faith placed in whiskey as a treatment in the early stages of a cold. It is also a vasodilator and counteracts the discomfort of the peripheral vasoconstriction at that stage.

Local vasoconstrictors should be used sparingly as the excessive use of any vasoconstrictor agent should be avoided on the grounds of interference with ciliary activity, mucosal blood flow and local tissue resistance. Temporary relief may be achieved by using ephedrine 0.5% in isotonic saline. This is particularly helpful in enabling a child to sleep or a baby to suckle.

Antibiotics do not appear to influence the course of a cold and therefore should only be used, and then in full doses, if complications develop such as middle-ear infection, sinusitis, tonsillitis, bronchitis or pneumonia.

Influenzal rhinitis

Influenzal rhinitis occurs in association with an infection by one of the influenza viruses. There are three main groups of virus unrelated antigenically (A, B and C).

Influenza A virus which has undergone several mutations since its discovery in 1933 has been responsible for pandemics of the disease. The original A virus has since been replaced by different strains, A1 (1946) and later A2 (1957). It is indeed unfortunate that the virus is subject to antigenic change for there is little or no cross-immunity and an entire population may find itself susceptible to the 'new' virus. Influenza B and C viruses are less liable to antigenic variation. The virology of these infections and its particular interest to the otolaryngologist have been described in some detail by Dudgeon (1969) and also by Anderson (1969).

The characteristic lesion is a varying degree of necrosis of the ciliated epithelium of the upper respiratory tract (nose and, in some cases, trachea). For a time there may even be replacement by transitional epithelium, and secondary bacterial invasion is inevitable.

In some cases of influenza the rhinitic manifestations are not marked or are overshadowed by tracheal, gastrointestinal, pulmonary or general symptoms, but in others severe coryza is simulated and in some of the pandemics many cases have been complicated by epistaxis.
Preventive treatment by the injection of immunizing vaccines is generally applied to those persons leading institutional lives, where the risk of infection is greater, or to the elderly or infirm, particularly those with chronic pulmonary or heart disease, renal disorders or diabetes, where any complication is likely to be more serious. A recent estimate of the mortality among the elderly in England and Wales, based on calculations of excess mortality, suggests that in each winter between 1967 and 1973 an average of about 11,000 elderly people died directly or indirectly from the effects of influenza (Clifford et al., 1977). Nevertheless, the vaccines are not a panacea, for not only do they have a tendency to toxicity, but the immunity which they confer is transient.

The principal vaccine currently in use (Influvac or Fluvirin) contains inactivated surface antigen influenza virus which in 1986/87 is prepared to cover A/Philippines/2/82 (H3N2), A/Chile/1/83 (H1N1) and B/USSR/100/83.

Specific chemotherapeutic agents have been developed for the prophylaxis and treatment of influenza. Amantadine (Symmetrel) is thought to impair the uncoating of a virus once it has entered the host and may also impair viral penetration of the host cell wall. It is notably effective against influenza A.

Treatment of the established case is along general lines and consists of rest, analgesics and, in severe cases, prophylactic antibiotics. Local nasal treatment is not advocated.

**Rhinitis of the exanthems**

In measles, scarlet fever, pertussis and the enteric group, typhus, smallpox and chicken-pox, an acute rhinitis occurs in the prodromal and early stages. The local condition does not differ from that described in the common cold.

Differential diagnosis depends on the associated specific signs and symptoms. Secondary bacterial rhinitis is common, often very severe and of suppurative type, and complications are more frequent than after the common cold.

**Specific rhinitis**

**Acute nasal diphtheria**

**Definition**

An acute infective rhinitis caused by *Corynebacterium diphtheriae*.

**Clinical picture**

Nasal diphtheria may be primary or secondary to the faucial form. In the latter case it indicates a severe attack. There is often a transient simple rhinitis in the early stage of faucial diphtheria, but no membrane forms and it passes off in a few days.

The acute form differs from the chronic form described later in the short duration, pyrexia and general toxaemia, adenitis and subsequent paralysis. In the UK immunization has
practically eliminated diphtheria but the occasional case might arise from immigrants who have not been immunized (see Chronic diphtheric rhinitis).

**Treatment**

*C. diphtheria* is sensitive to penicillin, and a course of 4 or 5 days' intramuscular and local penicillin should be given in addition to the full doses of the antitoxin intravenously. Antitoxin neutralizes the toxins, while penicillin shortens the disease but does not neutralize the toxins.

There is a tendency for *C. diphtheriae* to persist in the nose for weeks after such an attack. Isolation should be continued until the cultures from three successive daily swabs have been negative.

**Acute syphilis**

The condition of acute syphilis is discussed under the heading of Nasal syphilis.

**Erysipelas**

In erysipelas of the external nose, the nasal mucous membrane may become secondarily infected by the streptococcus from the skin. The infection responds rapidly to penicillin.

**Glanders**

Acute glanders differs from the chronic form described below only in the rapidity of onset and the severity of both local and general manifestations. There is marked fever and prostration, and a pustular rash develops resembling smallpox. The nasal mucosa is greatly swollen, and later ulcers form and may destroy the septum and turbinates. The lymph glands are swollen inflamed, and may suppurate. Death usually follows within a few weeks.

**Diagnosis**

Glanders is most likely to be confused with smallpox, typhus fever, erysipelas, impetigo or syphilis.

**Anthrax**

Primary anthrax of the nose with malignant pustule formation has been described.

**Candidiasis (moniliasis)**

This subject is discussed below.
Gonorrhoea

Rhinitis caused by infection with *Neisseria gonorrhoeae* is certainly rare. Unlike the conjunctivae the nasal mucous membrane has a high resistance to this infection. One or two doubtful cases of purulent rhinitis in infants have been said to be caused by gonorrhoea, but their authenticity has been doubted. The infection responds to penicillin or to co-trimoxazole (Septrin).

Local irritants and trauma

In this group there is a simple catarrhal reaction in the nasal mucous membrane with particularly severe irritation amounting in some cases to actual neuralgic pain in the nose and face. Sneezing and copious watery discharge are important features. The reaction follows immediately on the exposure and persists while that lasts. In most cases it passes off rapidly unless the causative agent has produced some destruction of the epithelium, in which case regeneration and healing may take some days before it is complete. The period of recovery depends on the severity and degree of subsequent secondary infection.

Chronic infective rhinitis

There are many forms of chronic rhinitis and not a little confusion has arisen from the fact that the term has been taken by different authorities to include different conditions. In the present section the accent has been laid on 'infection', and the conditions referred to are either the result of, or associated with, the latter.

Atrophic rhinitis

Atrophic rhinitis is a chronic nasal disease characterized by progressive atrophy of the mucosa and underlying bone of the turbinates and the presence of a viscid secretion which rapidly dries and forms crusts which emit a characteristic foul odour sometimes called ozaena (a stench). There is an abnormal patency of the nasal passages.

Aetiology

The aetiology of atrophic rhinitis is still unknown. In the past numerous organisms have been cited as the cause, among which are *Coccobacillus* (Loewenberg, 1894), *Bacillus mucosus* (Abel, 1895), *Cocobacillus foetidus ozaena*, diphtheroid bacilli, and *Klebsiella ozaenae* (Henriksen and Gundersen, 1959). It is true that these organisms may be found in cultures but there is little evidence that they cause the disease.

Other factors which have been regarded as possible causes are chronic sinusitis, excessive surgical destruction of the nasal mucous membrane, and syphilis.

Atrophic rhinitis usually commences at puberty and is much more common in females than males; thus it is generally accepted that endocrine imbalance may play a part. Heredity is an important factor and there appears to be a racial influence in that the yellow races, Latin races and American Negroes are relatively susceptible, whereas the incidence is low in natives of equatorial Africa. Poor nutrition is undoubtedly a factor in the development of the
condition and Bernat (1965) considers that atrophic rhinitis is an iron-deficiency disease. Recently, immunologists have considered atrophic rhinitis to be an autoimmune disease. Fouad et al (1980) studied cellular immunity in patients with atrophic rhinitis using the leucocyte migration and spontaneous rosette tests in vitro and confirmed that there was an altered cellular reactivity or loss of tolerance to nasal tissues, which they considered might be precipitated primarily by virus infection, malnutrition and/or immunodeficiency which trigger a destructive autoimmune process with the release of antigen(s) of nasal mucosa into the circulation.

Pathology

Most authors agree that there are patches of metaplasia from columnar ciliated to squamous epithelium, that there is a decrease in the number and size of the compound alveolar glands, and that there are dilated capillaries; but some (Taylor and Young, 1961) were unable to demonstrate endarteritis and periarteritis of the terminal arterioles. It is possible, therefore, that there are two types of atrophic rhinitis:

1) type 1, characterized by endarteritis and periarteritis of the terminal arterioles, which is the result of chronic infection and which might benefit from the vasodilator effect of oestrogen therapy

2) type 2, in which there is vasodilatation of the capillaries, which might be made worse with oestrogen therapy.

It seems likely that in the past the majority of cases were of type 1.

Taylor and Young (1961) also found that the endothelial cells lining the dilated capillaries had more cytoplasm than normal capillaries and showed a positive reaction for alkaline phosphatase which suggested the presence of active absorption of bone which is a feature of atrophic rhinitis.

Clinical picture

The presenting symptoms are most commonly nasal obstruction and epistaxis. Anosmia may be present and the patient is often only made aware of the loathsome effluvium surrounding her by the reluctance of others to come within her vicinity. Sometimes the symptoms are mainly pharyngeal and are caused by the pharyngitis sicca which often accompanies the condition or by choking when detached crusts slip from the nasopharynx into the oropharynx.

Clinical examination of the morose and dejected patient confirms the presence of foetor in all but the earliest cases and the nasal cavities are found to be lined with green, yellow and black crusts. Even before the removal of the latter the enormous capacity of the nasal passages is apparent and their detachment reveals a bleeding and ulcerated mucosa and shrivelled turbinates.
Investigations

Before embarking on treatment it is advisable to exclude the presence of sepsis in the paranasal sinuses by radiology, and if necessary by proof puncture. Swabs from the nasal secretions may be cultured, but while of interest, the results are unlikely to be of great value in the management of the case. Serological tests to exclude syphilis are essential as syphilis is certainly the most likely condition to be confused with atrophic rhinitis. The blood picture, serum proteins and iron should also be checked.

Treatment

Conservative

In the first place the patient should be instructed to douche the nose twice daily with an alkaline solution prepared by dissolving in 280 mL (1/2 pint) warm water a teaspoonful of the following powder:

- sodium bicarbonate 28.4 g
- sodium diborate 28.4 g
- sodium chloride 56.6 g.

Regular nasal cleansing is the basis of the conservative treatment in atrophic rhinitis and it may be of some consolation that, if the patient is prepared to carry out this simple treatment with unfailing regularity, freedom from offensive effluvia may almost always be achieved.

Following the removal of crusts by forceps or suction it is customary to apply either 25% glucose in glycerin, which inhibits the proteolytic organisms, oestradiol in arachis oil 10,000 units/mL or Kemicetine antiozaena solution (each mL containing chloramphenicol 90 mg, oestradiol dipropionate 0.54 mg, vitamin D₃ 900 IU, propylene glycol). The use of potassium iodide by mouth with the object of increasing nasal secretion has been suggested. Autogenous vaccines may be given. Sinha, Sardana and Rjvanshi (1977) have reported promising results using tissue therapy with systemic human placental extracts, which gave an 80% improvement in 2 years, and submucosal intranasal injection of human placental extracts which produced 93.3% relief over the same period of time.

Surgical

Numerous attempts to relieve the condition surgically have been made in the past. These include submucous injections of paraffin, and operations aimed at displacing the lateral nasal walls medially (Lautenslager's operation). More recently Teflon strips, polythene and cartilage have been inserted after flaps of mucoperichondrium were raised from the septum or mucoperiosteum from the floor or lateral walls. Wilson (1964) has reported good results from the submucosal injection of a suspension of powdered Teflon in 50% glycerin paste. Chatterji (1980) reports successful results using autogenous medullary (cancellous) bone graft as a single long piece of bone.
Repeated stellate ganglion blocks have been employed with some success by Sharma and Sardana (1966) who advocate cervical sympathectomy or blockade as a possible first line of treatment. Previously, however, autonomic surgery for atrophic rhinitis had proved disappointing.

Encouraging results have been obtained following the closure of one or both nostrils by plastic surgery (Young, 1967). Young's method is to raise folds of skin inside the nostril and suture the folds together with the object of complete interruption of the air flow. After periods varying from months to several years the nostrils have been reopened revealing absence of crusting and normal mucosa. Sinha, Sardana and Rjvanshi (1977) found that bilateral closure was not tolerated by some patients who disliked mouth-breathing and a nasal voice. However, partial nostril closure leaving a 3 mm hole was well tolerated and gave similar results with no recurrence of disease over a 2-year period. Any further increase in size of the hole rapidly decreased their success rate.

**Rhinitis sicca**

Rhinitis sicca is the term often reserved for a dry, mildly atrophic anterior rhinitis which does not progress to the full clinical picture of atrophic rhinitis described above. The causes are not well defined but it is generally recognized that the condition occurs in alcoholism, anaemia, nutritional and constitutional diseases and in those engaged in dry, hot and dusty occupations.

The pathology resembles that of early atrophic rhinitis; indeed some authorities would not distinguish the two as separate entities. There is deficiency and inactivity of the seromucinous glands, metaplasia of the columnar ciliated epithelium to cuboidal or squamous epithelium and deficiency of the mucous blanket. A penetrating ulcer of the anterior part of the cartilaginous septum may be present.

The patient complains of discomfort, irritation and sometimes epistaxis and crusting but the crusts are thin and dry and do not as a rule extend to the posterior part of the nasal cavities as do the crusts of atrophic rhinitis; neither do they emit a characteristic fetor.

Clinical examination reveals a dry, whitish or glazed mucous membrane sometimes accompanied by crusting or complicated by a septal perforation.

As in atrophic rhinitis the patient should be investigated with a view to excluding nutritional deficiencies or local infection.

In treating the disorder all possible causes should be removed and if necessary iron and vitamins administered. Locally, douching with the solution advocated for the treatment of atrophic rhinitis is undoubtedly of value, but the time-honoured treatment with oily drops and sprays is to be deprecated owing to the danger of inhalation lipoid pneumonia and paraffin granuloma. These sinister conditions have been recognized for a number of years and their pathology is clearly described by Spencer (1968).
Rhinitis caseosa

Rhinitis caseosa (nasal cholesteatoma) is a chronic inflammation of the nose associated with the formation of granulation tissue and an accumulation of offensive cheesy material resembling cholesteatoma.

The condition is rare and is usually unilateral, although bilateral involvement has been reported. It is slightly more common in males and can occur at any age. The cause of rhinitis caseous is unknown but numerous theories have been advanced including those of tubercle, syphilis, cholesteatoma and polyp degeneration. The most widely accepted explanation is the theory of suppurative rhinitis complicated by obstruction where rhinitis caseosa is a secondary condition symptomatic of an underlying primary nasal abnormality (for example rhinoliths, deviated nasal septum, inflamed turbinates or polyps) which tends to interfere with the egress of discharge from the nose. There may be coexisting sinus infection.

Microscopical examination of the caseous debris shows keratinous material, numerous organisms and sometimes cholesterol crystals. The lining mucous membrane shows chronic inflammatory changes.

Clinical examination in the early stages merely reveals that one side of the nose is filled with whitish debris but later the bone is invaded, the soft tissues of the face are inflamed and abscesses may form and burst through the skin.

Careful investigation by means of radiology and histological examination is necessary to exclude the presence of coexistent conditions such as sinus infection or malignant disease, and treatment consists of thorough removal of the debris by scooping it out followed by repeated irrigation to ensure its complete removal. Any obstructive lesions should be corrected surgically. Surprisingly perhaps, the prognosis is extremely good provided that care is taken to follow-up the patient and deal with any signs of stagnating discharge.

Gangosa

Gangosa (rhinopharyngitis mutilans; gangraengosa; kaninloma) is a slowly progressive ulceration and necrosis of the palate, nose and pharynx. As a disease it appears to be a separate entity but it may be clinically indistinguishable from tertiary yaws (see Yaws); thus there may arise a certain amount of confusion.

The geographical incidence of the specific form is limited to the pacific Islands, Sri Lanka and equatorial Africa. Gangosa affects males and females of all ages and is associated with dirty and insanitary conditions. It is extremely rare in the white races but has been reported. The cause and mode of spread are unknown; no specific organisms have been found in the tissues or in the discharge.

The disease commences as a small painless nodule in the middle of the palate. This perforates into the nose and spreads intermittently destroying all structures including the nose, palate, orbit and its contents and even the entire face. Pain is absent.
The disease may be steadily progressive, or may be arrested at any stage, the resulting scars resembling those of burns. Most cases survive (Arrowsmith, 1921; Myerson, 1933). Serological tests for syphilis are negative and there is no response to antisyphilitic treatment.

**Nasal syphilis**

Nasal disease secondary to infection with *Treponema pallidum* can occur in every age group from the neonate to the elderly. The disease is no longer common and the signs and symptoms in the early stages may be difficult to elicit, particularly if antibiotics have already been given.

The histological appearances of the syphilitic lesion are characterized by oedema, and infiltration of the stroma with lymphocytes, plasma cells and endothelial cells. The perivascular cuffing by these cells and the endarteritis reduce the lumen of the blood vessels, and result in necrosis and ulceration.

**Primary syphilis**

The lesion of primary syphilis can appear on the external nose or inside the vestibule. It presents as a hard, non-painful ulcerated papule that is often associated with an enlarged rubbery non-tender node some 3-4 weeks after contact. There may be malaise with a pyrexia. The lesion usually disappears spontaneously in 6-10 weeks. It has to be differentiated from malignant neoplasms and furunculosis. Malignant neoplasms are progressive, and occur in the later age groups. Furunculosis is painful and suppuration follows.

The following will be useful in establishing a diagnosis:

1. cultures from the surface of the lesion will be negative

2. smears examined by dark-ground illumination or after staining should show the spirochaete, *Treponema pallidum*

3. serological tests for syphilis may be positive, except in the earliest cases, or in those cases already having antibiotics; serological tests in current use include: (a) Venereal Disease Reference Laboratory (VDRL) tests; (b) *Treponema pallidum* haemagglutination test (TPHA); (c) fluorescent treponemal antibody test (FTA); (d) *Treponema pallidum* immobilization test (TPI).

4. a biopsy may be performed in doubtful cases; the microscopical appearances are characteristic.

Owing to its rarity and the fact that the chancre does not present a typical appearance, the diagnosis is often overlooked and may not be suggested until secondary manifestations are seen. The hardness and painlessness of the nodule, with early and great enlargement of the lymphatic glands, should suggest the diagnosis.
General antisyphilitic treatment, with intramuscular penicillin, should be given at once, and the chancre may be cleansed with 1:2000 solution of perchloride of mercury and the surface smeared with 2% yellow mercuric oxide ointment.

**Secondary syphilis**

The secondary stage of syphilis is the most infectious and symptoms appear 6-10 weeks after inoculation. The most common manifestation is a simple catarrhal rhinitis. Clinically this does not show any special characteristic, except in its persistence. There may be crusting and fissuring of the nasal vestibule.

Secondary syphilis is rarely recognized in the nose, as mucous patches hardly ever occur on such a thin attenuated mucous membrane. The diagnosis is usually suggested by the appearance of other secondary lesions, particularly the development of mucous patches in the pharynx, roseolar or papular rashes, pyrexia and the shotty enlargement of many lymph nodes. The scar of the primary lesion may be visible. Serological tests for syphilis are positive. The response to antisyphilitic treatment is so rapid as to be of diagnostic value.

The condition responds to general antisyphilitic treatment.

**Tertiary syphilis**

This is the stage most commonly encountered in the nose. The pathological lesion is the gumma, invading mucous membrane, periosteum or bone. The bony portion of the septum is the site most commonly attacked. More rarely the lateral nasal wall, frontal sinus, nasal bones or floor of the nose are invaded. Pain and headache (which is always worse at night), swelling and obstruction are the early symptoms. The swelling may be diffuse or localized, and offensive discharge, bleeding and crusting follow, but the pain is then relieved. The olfactory acuity diminishes. In neglected cases, perforation of the affected nasal wall, and collapse of the bony support of the nose may occur. Ultimately there may be severe scarring, and secondary atrophic rhinitis.

The earliest stage of simple swelling is not often seen. Later there is a diffuse or localized submucosal swelling, and infiltration. The surface is red, and may be nodular. The lesion is usually unilateral but, if the septum is involved, the swelling is seen on both sides. Tenderness of the nasal bridge is a characteristic sign. As a rule, when first seen, ulceration has already taken place, and a putrid-smelling discharge is escaping from the crusted surface. The crusts should be removed, and bare bone may be felt with a probe. The margins of the ulcers are irregular, overhanging and indurated.

The following are special aids to diagnosis:

1. there is no shrinkage with vasoconstrictors
2. radiographs show rarefaction of bone, with blurring of the cortical outline
3. serological tests for syphilis are positive in 90% of cases
4. biopsy shows the typical syphilitic histological appearances.
This stage has the following complications and sequelae:

(1) secondary infection with pyogenic organisms
(2) sequestration
(3) perforation of the septum, palate or nasal walls
(4) collapse of the nasal bridge, and deformity of the face
(5) scarring and stenosis of the nasal passages
(6) atrophic rhinitis
(7) intracranial complications from involvement of the meninges.

**Differential diagnosis**

A gumma should be suspected when there is a firm reddened nodular swelling of the bony portion of the septum or nasal wall, with obstruction, nocturnal pain and tenderness of the nasal bridge. Ulceration, fetor and necrosis of bone practically confirm the diagnosis. Serological tests for syphilis are positive. Other blood changes are absent, and the response to treatment is rapid. In all cases of doubt a biopsy should be performed, as the histological appearances in syphilis and in all the conditions given below are characteristic.

*Yaws* differs from syphilis only in its origin in tropical countries, the onset in childhood by extragenital infection, and the gross skin lesions. Serological tests for syphilis are usually positive and the lesions respond to antisyphilitic treatment.

*Lupus vulgaris* affects mainly the anterior cartilaginous portion of the septum and anterior ends of the turbinates. There may be associated nodules in the skin. Apple-jelly nodules may be seen, and there is no special odour.

In *tuberculosis* the course is rapid, and the skin is not affected. Typical signs of tuberculosis may be present in the lungs.

*Sarcoïd* resembles tuberculosis, but does not caseate; nodules appear in the skin and other organs. There is anergy to tuberculin, and the Kveim-Siltzbach skin test is usually positive.

In *atrophic rhinitis* the fetor is characteristically offensive and nauseating. The mucosa does not ulcerate deeply, and there is no bone necrosis.

*Lepra* occurs only in certain countries, is painless and develops very slowly. Nodules may be present in the skin, and deformity is severe in the late stages. Areas of anaesthesia may be present. *Mycobacillus leprae* may be seen in the discharge.

*Scleroma* occurs in patients of Central European, Asian, American and African origin. It is slow, painless and does not ulcerate. Stenosis and adhesions are characteristic. Associated lesions are found in the nasopharynx and larynx.

*Chronic glanders* closely resembles tertiary syphilis, but there is an intermittent pyrexia, and *Loefflerella mallei* may be cultured from the discharge.
Leishmaniasis occurs chiefly in South American countries. It commences as a nodule on the septum, which spreads slowly, destroying cartilage, but not bone. It is followed by fibrosis, and scarring. The histology is characteristic, and the Leishman-Donovan bodies can be identified. Response to tartar emetic is rapid.

Benign neoplasms grow slowly, and are painless. Ulceration and bleeding are rare, except in angioma.

A malignant neoplasm is at first unilateral. It grows steadily and ulcerates superficially, and the surface is hard and friable, and bleeds readily on probing. Radiographs show invasion and destruction of bone.

A sequestrum must be distinguished from a foreign body or a rhinolith by probing. The first is always attached deeply at some point, the second and third can always be moved, if only to a slight extent. When bone necrosis is present, only the silent form of osteomyelitis requires to be excluded. In this the swelling is more diffuse; it is associated with sinusitis, there are general signs of infection and a leucocytosis. Radiographs show the typical worm-eaten appearance of the bone.

A septal perforation caused by a gumma is situated posteriorly on the vomer or ethmoid. When due to rhinitis sicca, trauma, lupus vulgaris, leprosy or chrome ulceration it affects the anterior cartilaginous portion.

Treatment

General treatment

General antisyphilitic treatment is given.

Local treatment

The nasal passages must be cleared of crusts and discharge by copious alkaline douches every morning, and repeated if necessary two or three times a day. Dilute mercuric nitrate ointment should be applied freely to the nasal vestibules.

Sequestra should be removed with great care. The free portion may be removed piecemeal, but any firmly attached portion should be allowed to separate naturally, as avulsion may cause severe haemorrhage or damage adjacent tissues. Gummas respond rapidly to general antisyphilitic treatment, but atrophic rhinitis and deformity may persist after the disease is cured.

Perforations of the palate and deformities of the face may be repaired by plastic surgery.

Hereditary or congenital syphilis

In congenital syphilis, any of the lesions described under the secondary and tertiary forms of syphilis of the nose may occur.
In the infant, 'snuffles' is the most common lesion. This begins about the third week of life, but may appear as late as 3 months after birth. At first it appears as a simple catarrhal rhinitis. In a short time it becomes purulent, with secondary fissuring and excoriation of the nasal vestibule and upper lip. The obstruction may be so severe as to interfere seriously with suckling and nutrition.

Gummatous and destructive lesions occur most commonly at puberty in the 'latent' form of the disease. Mucous membrane, periosteum and bone may all be affected. The resulting ulceration and destruction lead ultimately to atrophy of the mucous membrane, secondary atrophic rhinitis, and sinking of the nasal bridge, producing the saddle-nose deformity. Serological tests for syphilis of the patient and parent are positive; biopsy shows the characteristic syphilitic histological picture.

There may be a prenatal and family history of syphilis, miscarriages or stillbirths. Snuffles should be suspected when a severe rhinitis with excoriation of the nares develops about the third week of life, and interferes with suckling. A common cold infection at this age may often produce a severe rhinitis, but there is usually a definite history of exposure to infection; serological tests for syphilis are negative, and cultures may show virulent pyogenic organisms. When obstruction dominates the picture congenital atresia of the choanae or adenoid hypertrophy must be excluded by sounding the nasal passages with a soft catheter.

In the tertiary form, the diagnosis rests on the presence of other stigmata, particularly Hutchinson's incisors and Moon's molars, interstitial keratitis, corneal opacities, sensorineural deafness and the serological reactions.

**Treatment**

In snuffles the airway must be restored for suckling. The discharge is removed by gentle suction and irrigation and drops of 0.5% ephedrine in normal saline solution should be inserted into the nose, with the head hyperextended, before feeding.

In the tertiary forms simple nasal toilet by syringing with isotonic alkaline douche solution will remove the crusts and discharge, and yellow mercuric oxide ointment may be applied frequently to the nasal vestibules.

In both forms antisypilitic treatment is essential and rapidly arrests the disease, but the destruction and deformity remain.

**Tuberculosis**

Tuberculosis of the nose is very rare. It may be nodular or ulcerative. It affects the cartilaginous portion of the nasal septum, and has been reported on the lateral nasal wall. It may be primary (Havens, 1931) but is usually secondary to tuberculosis of the lungs.

The symptoms are discharge, slight pain and partial obstruction. On examination a bright red nodular thickening, with or without ulceration, is seen on the septum. Tuberculosis follows a relatively rapid course, and ulceration leads to perforation of the septum.
Bacteriological examination of the discharge shows tubercle bacilli, and biopsy will confirm the typical appearance of tuberculosis.

Nasal douches may be used to remove the discharge and crusts. Treatment is with antituberculous drugs (rifampicin, ethambutol, isoniazid, pyrazinamide, streptomycin, PAS) in a planned schedule for at least 6 months.

**Lupus vulgaris**

Lupus vulgaris is an indolent and chronic form of tuberculous infection which affects the skin and mucous membrane.

It is twice as common in females as in males, and is developed most often in early adult life. It is a disease mainly of northern climates, and is rare in the tropics. The mucocutaneous junction of the nasal septum is the most common site of inoculation as this is frequently exposed to trauma in patients who have the habit of picking the nose. The nasal lesion is frequently associated with, or a precursor of, nodules on the face.

Sections of tissue show the characteristic appearance of a tuberculous granuloma. In the centre, at first, there is a collection of reticuloendothelial cells which soon necrose and coalesce. Around this necrotic centre there is a ring of living reticuloendothelial cells, and around this ring are lymphocytes, plasma cells and fibroblasts; scattered throughout the tubercle are found giant cells, with a peripheral arrangement of nuclei.

The early symptoms are those of nasal discharge and obstruction followed by crusting and occasional epistaxis. When the ulceration is established there may be slight fetor and soreness. Ulceration may be followed by fibrosis and contraction, with distortion of the alae nasi. When the turbinates are extensively involved the ciliated epithelium is not renewed and atrophic rhinitis may develop.

The course is very slow, and may last for a lifetime with periods of regression and healing, alternating with periods of active extension, depending to a great extent on the general health of the patient.

The typical early lesion is a reddish firm nodule at the mucocutaneous junction of the nasal septum. In more advanced cases, there may be extensive involvement of the floor of the nose and the turbinates, spreading backwards from the primary site. The surface shows superficial ulcers and crusts. The septum may perforate, but only in the cartilaginous portion, and there is no sinking of the nasal bridge.

If the disease spreads forwards there may be external scarring and distortion of the nasal vestibule, tip and alae nasi, and nodules may be seen in the skin of the face.

Blanching, bacterial examination and biopsy are of use in diagnosis.

(1) To show apple-jelly nodules, the blood is expressed from adjoining tissues by pressure with a glass slide on the skin, or shrinkage with cocaine and adrenaline on the mucous membrane, thus making the pinkish lupus nodules more evident by contrast.
(2) Bacteriological examination of the discharge may show tubercle bacilli.

(3) Biopsy will confirm the typical histological picture. For differential diagnosis, see above under Tertiary syphilis.

**Complications**

(1) Pulmonary tuberculosis develops in a small proportion of cases.

(2) Dacryocystitis, corneal ulceration, nasopharyngeal lupus and lupus of the face may occur.

(3) Atrophic rhinitis may be a sequel.

(4) Epithelioma may develop in the infected tissue.

Sudden increase in size and hardness of one area and, in the elderly patient, an increased tendency to bleed should arouse the suspicion that a malignant change has supervened. A biopsy should be taken, and the tissue examined histologically.

Treatment consists of specific antitubercular therapy and calciferol (vitamin D$_3$) 150,000 units daily for 6-9 months. Plastic repair of deformities of the nose may be required when the disease has been arrested.

*Sarcoidosis (Boeck's sarcoid)*

Sarcoidosis is a chronic systemic disease of unknown cause which is clinically characterized by involvement of virtually every organ with a non-caseating granulomatous inflammation closely resembling tuberculosis without caseation. The tubercle consists of a collection of pale-staining epithelioid cells, sometimes surrounded by a thin layer of lymphocytes. Giant cells are present and, in older lesions, contain asteroid intracytoplasmic inclusion bodies which stain with haematoxylin (Schaumann Bodies). This histological picture is not, however, specific for sarcoidosis as it may be seen in other granulomata, for example tuberculosis, leprosy or berylliosis. Before confirming the diagnosis it is therefore important to exclude these other causes.

Nasal sarcoidosis was first described by Boeck (1905), and confirmed by biopsy by Kistner and Robertson (1938).

**Aetiology**

Two hypotheses have been advanced (Gordon et al, 1976):

(1) that sarcoid is a special form of tuberculosis which is the result of an altered bacillus with an atypical host response. Tuberculosis is known to precede, occur with or follow clinical sarcoidosis. However, tubercle bacilli have been reported in only a few cases.
(2) That an unidentified organism or agent is responsible, for example pine pollen, wood dust, beryllium and silica or tubercle bacilli, *M. leprae*, a protozoon, virus or fungus.

**Incidence**

Sarcoidosis occurs all over the world but is more prevalent in rural south-eastern USA and Scandinavian countries. Coloured races are more affected than white, and females more than males. Nasal sarcoidosis occurs in 3-20% of systemic cases. The median age of onset is 25 years, and 50% of cases occur below the age of 30 years.

**Clinical picture**

Presenting symptoms include nasal discharge, which ranges from serosanguineous to mucopurulent, nasal obstruction and epistaxis. There may be a secondary sinusitis, a result of superadded infection or involvement by the disease. Nasal skin and bone lesions are asymptomatic. There may be a general swelling of the bridge of the nose with discoloration of the overlying skin (Black and Munro, 1966).

Examination of the nasal mucosa may reveal tiny, 1 mm, yellow nodules surrounded by hyperaemic boggy mucosa. Alternatively the mucosa may be dry, ulcerated and covered with crusts. The anterior septum and inferior turbinate are the most commonly involved areas and adhesions may develop between them, resulting in stenosis of the anterior nares. Septal manifestations may arise spontaneously or may appear following septal surgery in the unrecognized case. Nasal skin lesions appear as elevated, yellowish, dry, scaling, discrete nodules or plaques. These may coalesce to form large bluish-red granulomata, separated by normal skin, over the tip, alae, columella or dorsum. Violaceous, diffuse bulbous affliction of the nasal tip area in conjunction with other skin and pulmonary lesions was separately described by Besmer in 1889 as *Lupus pernio* (Gordon et al, 1976) and is a manifestation of chronic multisystem sarcoidosis (James, 1959). Weiss (1960) believed that skin and mucosal lesions are complete, separate independent lesions.

Nasal lesions may be associated with other lesions in the head and neck which may include tonsil, tongue, salivary glands, lacrimal glands, bronchial mucosa, paranasal sinuses, nasopharynx, or larynx. Heerfordt's syndrome describes a transient bilateral facial palsy associated with fever, parotid enlargement and uveal tract disease.

**Diagnosis**

The nasal mucous membrane, particularly in the region of the anterior septum and inferior turbinates, can be easily biopsied and will produce valuable diagnostic material. The histological picture is described above. Culture and stains for acid-fast bacilli and fungi should be negative. There is usually an anergy to the tuberculin skin test, but pulmonary tuberculosis develops during the course of the disease in 10% of cases and tuberculin hypersensitivity then develops. The Kveim-Siltzbach skin test (in which a subcutaneous injection of a suspension from a lesion in another case is followed by the development of a sarcoid nodule) is usually positive in all mucosal cases, and in 75% of patients with active sarcoidosis. It is an invaluable aid in the differential diagnosis of granulomata of the nose.
The radiographic changes in cases of involvement of the nasal bones are characteristic and consist of cystic, punched-out lesions with thinning of the cortex of the bone and a reticular pattern in the medulla (Curtis, 1964). Hilar node involvement is shown in radiographs of the chest, and bone cysts are seen in radiographs of the hands and feet.

Serum and urinary calcium levels should be measured to exclude hypercalcaemia. Serum immunoglobulin, particularly IgG, and the erythrocyte sedimentation rate (ESR) may be raised and a mild anaemia, leucocytopenia and eosinophilia may be present. The serum levels of serum angiotensin converting enzyme, which is secreted by the epithelioid cell granuloma, are evaluated in about 60% of patients with active sarcoidosis (Studdy et al, 1978).

**Differential diagnosis**

The differential diagnosis includes:

(1) chronic irritation and foreign body reaction, as in berylliosis

(2) infectious conditions, such as tuberculosis, actinomycosis, rhinosporidiosis, leprosy, syphilis, glanders, histoplasmosis and blastomycosis

(3) other granulomatous conditions, such as Wegener's granulomatosis

(4) acquired immune deficiency syndrome (AIDS) may present with a similar granulomatous condition in the nose.

**Treatment**

There is no specific therapy. Steroids may be used locally or systemically. Local depot steroids (McKelvie, Gresson and Pokhrel, 1968) produce a marked decrease in the size of mucosal lesions, but there is little reduction in the size of lesions in patients with systemic disease. Topical steroid nasal drops are beneficial. Atrophic rhinitis may develop as a result of the disease or secondary to depot steroids. In a series of 53 patients with sarcoidosis of the upper respiratory tract, James et al (1982) found systemic steroids were necessary in 46% of the patients. When steroids alone are insufficient in the management of chronic fibrotic sarcoidosis they can be combined with either chloroquine (adult oral dose of 250 mg on alternate days for about 9 months) or methotrexate (adult oral dose of 5 mg, once weekly for a course of 3 months). The long-term use of chloroquine carries the risk of occasional development of irreversible retinal damage.

**Chronic diphtheric rhinitis**

Chronic diphtheric rhinitis (fibrinous rhinitis) is an inflammation of the nasal mucous membrane, caused by *Corynebacterium diphtheriae*. Diphtheria is now extremely rare in the UK. More commonly a fibrinous rhinitis may be caused by the pneumococcus, staphylococcus or streptococcus and is seen very occasionally in debilitated children.
All the changes of a severe chronic inflammation are seen and on the surface there is extensive necrosis and defoliation of the epithelium. The area is covered with a membrane of fibrin and entangled cells. The fibres of fibrin extend deeply into the submucosa and this accounts for the tenacity of its adhesion, and for the bleeding when the membrane is removed. The corynebacteria and pneumococci cause the formation of an adherent membrane, but staphylococci and streptococci produce only a superficial membrane which can be stripped off easily.

The local symptoms are obstruction, and discharge which is watery at first and later becomes bloodstained and mucopurulent. The course of the disease is slow, and may go on for 3 months, ending in spontaneous recovery. Paralysis, toxaemia and other general symptoms are absent.

The anterior nares may be excoriated by the acrid discharge. The nasal mucosa is generally congested and swollen, and the inferior turbinates, floor of the nose and sometimes the septum are covered with a greyish adherent membrane. After removing this a raw bleeding surface remains.

Bacteriological examination of the nasal discharge should never be omitted, and if *C. diphtheriae* is present the organism should be tested for virulence.

Treatment consists of systemic antibiotics and nasal toilet. Systemic antitoxin is unnecessary but, in the past, local application of antitoxin has been found beneficial. In all cases the patient should be isolated.

*Rhinoscleroma*

Rhinoscleroma, or scleroma, is a progressive granulomatous disease commencing in the nose and eventually extending into the nasopharynx and oropharynx, the larynx and sometimes the trachea and bronchi (Friedmann, 1966).

Scleroma may occur at any age and in either sex. It is seen mainly in central and south-eastern Europe, North Africa, Pakistan and Indonesia, Central and South America. it may be seen anywhere in the world and people of any race may be affected. There is, in most patients, one common factor - a poor standard of domestic hygiene.

**Pathology**

Although there is still controversy over the precise pathogenesis of scleroma it is now generally agreed that the causative organism is the Gram-negative Frisch bacillus (*Klebsiella rhinoscleromatis*). That this organism was a secondary invader following an initial filterable virus infection is disputed by the work of Fisher and Dimling (1964), who failed to reveal virus-like particles or inclusion bodies on electron microscopy. Steffen and Smith (1961) successfully recovered the organism from the lungs of mice previously inoculated with *K. rhinoscleromatis*, and Sinha, Pandhi and Prakash (1969) isolated it from 60% of their cases. A complement fixation test based on the reaction of the patient's serum with suspensions of *K. rhinoscleromatis* was described by Levine (1951) and re-evaluated by Toppozada et al
Histologically granulomatous tissue infiltrates the submucosa and is characterized by the presence of an accumulation of plasma cells, lymphocytes and eosinophils among which are scattered large foam cells (Mikulicz cells), which have a central nucleus and a vacuolated cytoplasm containing Frisch bacilli and Russell bodies, the latter resembling plasma cells and having an eccentric nucleus and deep eosin-staining cytoplasm.

Friedmann (1963), in an electron microscope study, observed the transformation of plasma cells into Russell bodies. Further ultrastructural work by Toppozada et al (1981) has demonstrated the different stages of distension of the rough endoplasmic reticulum up to the formation of Russell bodies inside the 'reactive' plasma cell, thus supporting the theory of an intracellular formation of Russell bodies. The histochemical studies of Gonzalez-Angulo et al (1965) indicated a high content of mucopolysaccharides around the walls of the *Klebsiella* and inferred that this may be responsible for the protection of the organism against both antibiotic therapy and the patient's own antibodies.

**Clinical picture**

There are three recognized stages of this chronic and progressive disease.

(1) **The atrophic stage.** Changes occur in the mucous membrane of the floor of the nose, septum or turbinates which resemble atrophic rhinitis, with crust formation and foul-smelling discharge.

(2) **Granulation or nodular stage.** Non-ulcerative nodules develop which at first are bluish-red and rubbery and later become paler and harder.

(3) **Cicatrizing stage.** Adhesions and stenosis distort the normal anatomy. The disease may extend to the maxillary sinus (Mossallam and Attia, 1956; Yassin and Safwat, 1966), the lacrimal sac (Badraway, 1962), the nasopharynx, hard palate, trachea and main bronchi. Spread to lymph nodes has been reported but is extremely uncommon and is thought to be prevented by early fibrous tissue deposition blocking the lymphatics (Badraway and El-Shennawy, 1974). Bone may be extensively involved (Badraway, 1966). Malignant change is uncommon but can occur (Yassin and Safwat, 1966).

**Treatment**

Once the diagnosis has been confirmed by biopsy, treatment must be intense and prolonged in order to eradicate the disease completely. Bactericidal antibiotics in large doses are given for a minimum of 4-6 weeks and are continued until two consecutive cultures from biopsy material are proved to be negative (Ssali, 1975). In practice, the most effective antibiotics are streptomycin and tetracycline. Based on an observation by Rizk (1977) that acriflavine solution *in vitro* kill *K. rhinoscleromatis*, Shaer et al (1981) treated 50 patients suffering from rhinoscleroma with local applications of differing concentrations of acriflavine.
solution over an 8-week period. The 2% solution produced a complete cure of the disease in all its stages after 8 weeks, whereas the 5% concentration caused recurrent epistaxis, vestibulitis and an occasional septal perforation, and the 1% solution failed to cure all cases.

Chemotherapy may be combined with surgery to re-establish the airway without causing further atrophic changes. This is most effectively achieved by discrete removal of granulations and dilatation of the airways combined with the insertion of polythene tubes for 6-8 weeks (Ssali, 1975).

In late cases where the disease has been eradicated plastic reconstructive surgery may be required.

**Leprosy**

Leprosy is a chronic granulomatous disease caused by *Mycobacterium leprae*, an acid-fast bacillus morphologically similar to *M. tuberculosis*. Although *M. leprae* cannot yet be cultured on an artificial medium it can, nevertheless, be inoculated into experimental animals, particularly immunologically deficient mice (Rees, 1966), to produce a disease similar to that in man.

Worldwide, 12-15 million people suffer from some form of this disease which is particularly prevalent in India, Central Africa, and Central and South America.

Three main types of leprosy are recognized (Barton and Davey, 1976).

1. **Tuberculoid leprosy** in which solitary lesions cause anaesthetic cutaneous ‘patches’ with involvement of one or more related sensory or motor nerves with possible paralysis of muscles. Cutaneous patches may extend as far as the nasal vestibule but nasal mucosa is not involved bacteriologically or histologically. Isolated cranial nerve palsies (for example, fifth and seventh) may occur.

2. **Lepromatous leprosy**, in which there is diffuse infiltration of skin, nerves and mucosal surfaces. *M. leprae* tends to favour an environment where the temperature is lower than central (core) temperature. Thus cutaneous infiltration on the face is more apparent on the edges of the pinna, chin, nose and brow. Nasal mucosal involvement occurs early in the disease process and is present in 97% of patients with lepromatous leprosy (Barton et al, 1973). The nasal discharge in these patients, who frequently have minimal systemic signs, contains millions of potentially infectious bacilli and therefore suggests that this is the principal route of spread of infection. Most commonly there is crust formation, nasal obstruction and blood-stained discharge. Hyposmia may be demonstrated in over 40% of patients with lepromatous leprosy (Barton, 1974).

3. **Borderline leprosy**. The first two types are probably immunologically stable. Patients with borderline leprosy with poor resistance may develop the lepromatous type or less commonly, as the disease is modified by treatment or as immunity is acquired, the tuberculoid type. Skin lesions are more numerous than in tuberculoid leprosy and are frequently seen around the eyes, nose and mouth. In pure borderline leprosy there is no
involvement of the mucous membranes of the nose, mouth, pharynx or larynx. A conversion to the lepromatous type is indicated by the appearance of mucosal involvement.

**Clinical picture**

With lepromatous leprosy the earliest sign is a nodular thickening of the nasal mucosa which appears paler than normal and often has a yellowish tinge. These isolated nodules most commonly first involve the anterior end of the inferior turbinate. The disease progresses to gross inflammation of the nasal mucosa and severe obstruction and is out of proportion to the almost imperceptible clinical changes of lepromatous leprosy elsewhere in the body. Perforation of the cartilaginous portion of the nasal septum is followed by perichondritis and periostitis of the nasal cartilages, inferior turbinates and anterior nasal spine which leads to the typical nasal deformity. Atrophic rhinitis, fibrotic atresia or stenosis of the airway are typical sequelae.

Mcdougall et al (1974) have made an extensive histological study of biopsies of nasal mucosa in patients suffering from leprosy. They found no bacilli or evidence of leprosy infection in the septum and turbinates of borderline cases. However, in lepromatous leprosy bacilli were found in macrophages, fibroblasts, within the cytoplasm of endothelial cells of blood vessels and lymphatics and within the free lumina of secretory gland acini.

**Diagnosis**

Diagnosis of early and intermediate changes in the nose, pathognomonic of lepromatous leprosy, can often be made in the absence of other manifestations of the disease. The presence of atrophic rhinitis and septal perforations is indicative of late disease with other systemic manifestations (Barton, 1976).

Early diagnosis is essential as the nasal component of the infection results in a highly bacilliferous nasal discharge which is the principal route of transmission of the disease (Davey and Rees, 1974). Confirmation is by microscopy of the nasal discharge for acid-fast bacilli, microscopy of scrapings of the nasal mucosa (the most positive site being the anterior end of the inferior turbinate) for acid-fast bacilli, and histology of the nasal mucosa. Radiographs of the anterior nasal spine frequently show erosion (Møller-Christensen, Bakke and Melsom, 1052) and in a study of sinus radiographs the most constant finding was mucosal thickening of the maxillary antra on the occipitomental view (Barton, 1979).

It should always be remembered that, with the current patterns of migration, cases of leprosy may be seen in countries where it is no longer endemic (Barton and Davey, 1976).

**Treatment**

Dapsone remains the standard drug in the treatment of leprosy and will reduce the bacterial count of nasal discharge to zero or near zero within 2 months; however, there are increasing reports of dapsone resistance. The more expensive drugs, rifampicin (Rifadin) and clofazimine (Lamprene), act more rapidly and reduce the count to zero in 10 days; however, their cost precludes their general usage in developing countries. Local treatment to the nose will help to prevent the external deformity of advanced lepromatous leprosy. Betnovate (1
part) in Unguentum (2 parts) has been used with good results (Baron, 1978, personal communication). In cases of late involvement when the nasal septum has been perforated and atrophic rhinitis is established, careful crust removal is important. The crusts may be softened with a solution of sodium bicarbonate, sodium borate and sodium fluoride 15 g each, dissolved in 500 mL of warm water. After removal of the crusts, the nasal cavities can be painted with a suitable ointment such as Vaseline 1 kg, glycerine 200 g, Vioform 300 g, and crystal violet 5 g (Barton, 1985).

Yaws

Yaws (framboesia) is a disease closely resembling syphilis, if not identical with it, and occurs only in natives of the tropics. It is widespread in Central Africa, Jamaica and the Philippines. The causative organism is Treponema pertenue, which is indistinguishable morphologically from T. pallidum. Transmission of the disease is by direct contact, which is usually extragenital; there is a high incidence in infancy and childhood.

Clinical picture

Primary, secondary and tertiary stages occur as in syphilis. Characteristically yaws affects principally the skin and only rarely the mucous membranes, except at the mucocutaneous junctions. Nasal lesions are very rare and do not differ in appearance from those of syphilis. When very extensive and advanced there may be complete destruction of the nose and palate, involving the whole maxilla, face and pharynx. Clinically this is indistinguishable from true gangosa, but in yaws the serological tests for syphilis are positive, and the lesions respond to antisyphilitic treatment.

Another special form is designated 'goundou'. In this there is a chronic osteitis, forming bilateral rounded swellings of the nasal processes of the maxillae, which may encroach on the orbits and destroy the eyes. In the early months there is pain and serosanguineous nasal discharge.

The lesions in the nose are indistinguishable from syphilis. Some authorities consider that the two diseases are identical, but that their manifestations differ in natives of certain areas of the tropics. Differentiation from syphilis is made on the country of origin, the onset in childhood by extragenital infection, the gross skin lesions, and the fact that it is never congenital and that it does not cause quaternary lesions in the nervous system.

Treatment

The lesions respond rapidly to treatment with a single large dose of long-acting penicillin. Attention must be paid to general nourishment and hygiene.

Chronic glanders

Glanders is a specific inflammatory disease due to infection with Loefflerella mallei which is parasitic in horses, donkeys and mules. The infection is extremely rare in man. It occurs in both acute and chronic forms in grooms and others who handle horses. The infection is transferred directly from the horse to the human through an abrasion of the skin,
or occasionally through the nose or mouth. The incubation period may be a few hours, but
is usually 2-6 days in the acute form. In the chronic form it may be as long as a year.

**Clinical picture**

The disease is usually ushered in with an acute febrile attack, and in some cases a rash
develops which resembles smallpox. After a variable length of time, up to 5 years, during
which the organisms lie latent, subcutaneous and intramuscular abscesses appear and nodules
develop in the skin and in the mucous membrane of the mouth, palate and nose (Robins,
1906). The nodules ulcerate and later heal, and fresh ones appear and pass through the same
stages. The ulceration closely resembles that of tertiary syphilis. In the nose there is also a
severe generalized rhinitis, with tenacious mucopus and crusts lying on a reddened and
scarred mucous membrane.

Throughout the active stages a variable pyrexia of 1 or 2°C is constant, but periods
of complete remission of all signs of the disease are common.

In fatal cases, death is due to toxaemia and pulmonary and intracranial complications.
The duration of the disease may be anything from 6 weeks to 15 years. It has been estimated
that 6% of cases recover (Robins, 1906).

It is very difficult to distinguish the lesions from those of tertiary syphilis, but in
glanders there is usually a characteristic daily intermittent pyrexia, and in syphilis the serology
is positive. In the latter condition there is prompt response to antisyphilitic treatment, and
characteristic papery scars are left after healing.

Any cases diagnosed as tertiary syphilis with a negative serology, and no response to
antisyphilitic treatment, should be suspected as possible cases of chronic glanders. Culture and
isolation of *Loefflerella mallei* are often difficult, but intraperitoneal inoculation of the male
guinea-pig produces inflammatory changes in the tunica vaginalis of the testis (Straus's
reaction). Biopsy may not show any certain points of differentiation.

**Treatment**

The organism is sensitive to the tetracyclines, streptomycin, chloramphenicol and the
sulphonamides.

**Pathogenic fungi and yeasts**

The classification and pathology of these diseases is complex and the reader is referred
to the superb and exhaustive description given by Emmons et al (1977).

**Rhinosporidiosis**

Rhinosporidiosis is a chronic infestation by the fungus *Rhinosporidium seeberi*, which
predominantly affects the mucous membranes of the nose and nasopharynx but occasionally
involves the lips, palate, uvula, maxillary antrum, conjunctiva, lacrimal sac, epiglottis, larynx,
trachea, bronchus, ear, scalp, skin, penis, vulva and vagina. Osteolytic lesions in the bones
of the hands and feet have been reported by Chatterjee et al (1977). However, rhinosporidiosis is usually limited to surface epithelium but may on occasions be widespread with visceral involvement. The disease, which is chronic and is characterized by the formation of papillomatous and polypoid lesions, tends to affect young males and is endemic in many parts of India and Sri Lanka (Satyanarayana, 1966). Very occasionally the disease has been seen in Europeans who have visited India and Sri Lanka, but it is recognized to be rare for the condition to affect people outside these centres although reports of sporadic cases have come from the USA, Brazil, Africa and Europe. The mode of infection is probably by dust from the dung of infected horses and cattle but this has still to be confirmed.

The characteristic lesion is a bleeding polyp. Histologically the polyp has a vascular fibromyxomatous structure. Throughout the tissue are seen round or oval cells containing the sporangium. The walls of these are of thick chitin, but are thinned at one point where the cells will burst, sporulation will take place, and the spores will spread through the lymphatics into the connective tissue, where they develop into the trophic stage and complete the life cycle.

Epistaxis is often the only symptom, but in the early stages there is a viscid nasal discharge, with irritation and partial obstruction. With the development of the characteristic polyps the obstruction gradually increases until it may interfere with swallowing. Constitutional symptoms are rare, and the disease runs a slow course. The polyps may be present for years before the patient seeks advice.

The lesions are friable, in shape and colour resembling a strawberry, with a greyish undersurface studded with sporangia, showing as white dots. When sessile the polyps appear as multiple nodules, or may assume a leaf shape, with rounded or dentate margins. They arise primarily in the vestibule and are usually attached to the septum, but may spread backwards into the nasopharynx, and even hang down into the oropharynx. The nasal mucosa is generally swollen, hyperaemic and covered with copious viscid secretion, containing spores but no pus cells. The lymphatic glands are not affected.

Microscopical examination of the nasal discharge will show spores. Biopsy and histological examination of the polyps reveals the characteristic appearance described above. At first sight carcinoma may be suspected, on account of the friable masses which bleed on contact. The studding of the undersurface with white sporangia should suggest the diagnosis, and this may be confirmed by the special investigations.

Treatment consists of a combination of medical and surgical methods. The former includes the local injection of depot corticosteroids into the polypoidal masses, and in some cases systemic treatment with amphotericin (Fungizone) has been tried. Recently diaminodiphenylsulphone (dapsone) has been shown to be effective in controlling rhinosporidiosis. Growths are removed by wide excision with the cutting diathermy and cautery to the base, as on occasions excessive bleeding may occur.

The phycomycoses

The phycomycoses are a diverse group of mycoses caused by fungi which are traditionally placed in the class Phycomycetes. Although the term is now rejected by the
formal taxonomic system it is retained in medical mycology (Emmons et al, 1977). It has since been recommended to revert to the name *mucormycosis* for those mycoses caused by fungi belonging to the order Mucorales and the name *entomophtharamycosis* proposed for those mycoses caused by the fungi which belong to the order Entomophtharales. Certain members of each order can produce nasal disease of which the two major conditions are.

**Entomophtharamycosis conidiobolae (rhinophycomycosis)**

This disease is caused by *Conidiobolus coronatus* and is manifested as prominent nasal polyps and granulomata in the nasal cavity. Most cases have been seen in Central Africa, India, Brazil and the West Indies. Males are affected more than females. Symptoms consist of nasal obstruction and swelling over the nose and later the cheek and upper lip. Lesions usually begin in the inferior turbinate and spread in the submucosa through the natural ostia to the paranasal sinuses and to the subcutaneous tissues of the face. Histological examination shows a granulomatous reaction with collections of multinucleate giant cells in the centres of which hyphae can be seen. Treatment consists of removal of the tumour masses and systemic amphotericin (Fungizone).

**Orbital and central nervous system mucormycosis (rhinocerebral phycomycosis)**

This condition is a short-term and often rapidly lethal fungal disease in the nose and paranasal sinuses (Groote, 1970). The principal causative fungi are *Mucor circinelloides, Absidia corymbitera, Mucor javanicus* of the family Mucoraceae and order Mucorales. Because *Mortierella* (order Mucorales) and *Basidiobolus* (order Entomophtharales) have been also identified as a cause, the use of the term 'phycomycosis' was recommended by Straatsma, Zimmerman and Gass (1962).

Phycomycetes are ordinarily saprophytic organisms existing in soil, manure, fruits and starchy food. They can be cultured from the human nose and gastrointestinal tract. They become pathogenic when the patient’s general resistance has been altered by metabolic disorders or chemotherapeutic agents. This is most often associated with diabetic ketosis but can be seen with uraemic acidosis, leukaemia, malnutrition; steroid, antmitabolic or antibiotic therapy; and severe burns. The fungus has a remarkable affinity for arteries and by dissecting the internal elastic lamina from the media leads to extensive endothelial damage and thrombosis. Pathologically there is a mixed picture of inflammatory and necrotic changes. Later the veins and lymphatics are involved.

*Mucormycosis* appears in cerebral, pulmonary, ocular, superficial and disseminated forms. Orbital and central nervous system mucormycosis is the most common, and usually commences in the nose and extends by direct extension and intravascular propagation to involve the paranasal sinuses, orbit, cribiform plate, meninges and brain. The most characteristic rhinological finding is a black necrotic turbinate resembling a mass of dried clotted blood. Unilateral gangrene and perforation of the hard and soft palates may occur from involvement of the palatine arteries. Sinus radiographs show thickening of the lining of the sinuses, no fluid levels and spotty destruction of the bony walls.
Early clinical recognition of this potentially fatal disease is essential before irreversible changes occur. The disease is confirmed by biopsy.

Treatment consists of control of the original precipitating condition, heparinization, systemic amphotericin (Fungizone) and local drainage and debridement. Bahadur et al (1983) found long-term use of oral potassium iodide beneficial. The exact mode of action of the drug is unknown but it is believed to have an antifungal property.

**Aspergillosis**

Aspergilloses are infestations, which usually occur in those who handle doves and other small captive birds, in which *Aspergillus fumigatus* and *A. niger* are common, or as secondary infestations during treatment with antibiotics or corticosteroids. Primary paranasal aspergillus granuloma, although rare in the rest of the world, is common in northern Sudan. it can give rise to a pansinusitis and is due to *Aspergillus flavus-oryzae*.

There is a leucocytic and endothelial-cell infiltration, with patchy necrosis and a few giant cells. The symptoms are nasal obstruction, sneezing and watery, mouldy-smelling discharge. On examination the nasal mucous membrane is covered with a greyish (*fumigatus*) or black (*niger*) false membrane. The infection usually also invades the antrum (Tilley, 1915). The course of the disease resembles that of tuberculosis.

It is most likely to be mistaken for diphtheria, syphilis, tuberculosis or atrophic rhinitis; cultural examination of the membrane should determine the diagnosis.

Rarely in debilitated or immunosuppressed patients, acute aspergillosis may become a very aggressive nasal and sinus infection. This results in a relentlessly progressive vasculitis and thrombosis resembling mucormycosis. Extension from the nose and paranasal sinuses can quickly involve the orbit and central nervous system. In these instances the condition can occasionally be fatal.

The specific treatment consists of repeated cleaning and local application of 1% aqueous solution of gentian violet or nystatin. Amphotericin (Fungizone) may be given systemically. When the sinuses are involved, operative clearance should be performed (Adams, 1933).

**Blastomycosis**

Blastomycosis is due to an infection by *Blastomyces dermatidis*, an encapsulated yeast-like fungus, which is practically confined to certain parts of America. The disease starts in the skin, although primary inhalational infection of the lungs may occur in some cases. The nose is rarely affected, but oronasal mucosal involvement is a manifestation of disseminated blastomycosis. The mucosal lesion consists of a papillary hyperplasia with cysts which contain polymorphonuclear leucocytes surrounding the organisms. Regional lymph nodes are not usually affected but dissemination by the blood stream may produce widespread abscesses in the viscera, especially the lungs.

The specific treatment is with amphotericin (Fungizone).
Cryptococcosis

Cryptococcosis is caused by inhalation or ingestion of Cryptococcus neoformans, a yeast-like fungus closely resembling but nevertheless distinct from, Blastomyces dermatidis. The fungus is found in pigeon or other avian excreta and of the fatal fungal infections in the USA is second only to histoplasmosis (Briggs, Barney and Bahu, 1974). There is, however, a worldwide distribution of the infection, which disseminates after pulmonary infestation to almost any tissue but particularly to the brain and meninges to give a chronic meningitis resembling tuberculous meningitis. Isolated lesions may occur in lymph nodes, skin, bone and eye. Nasal involvement is uncommon but external ulceration, nasopharyngitis and pansinusitis have been described. Briggs, Barney and Bahu (1974) described a case of ulceration of the nasal vestibule, biopsy of which revealed focal ulceration of the squamous mucosa and oedematous submucosa containing numerous round-to-oval yeast organisms surrounded by a clear 'halo-like' space caused by the capsule.

Treatment with amphotericin (Fungizone) and flucytosine (Alcobon) can be monitored by a specific slide later agglutination test. Complete resolution can occur.

Actinomycosis

The genus Actinomyces consists of two principal species: A. bovis, the cause of actinomycosis or 'lumpy jaw' in cattle; and A. israelii, the cause of actinomycosis in man.

The anaerobic fungus, Actinomyces israelii, grows in the tissues in the form of colonies composed of branching mycelial threads with clubbed ends - 'ray fungus'. The colonies appear in pus as 'sulphur granules'.

Actinomyces israelii is frequently present as a harmless parasite in the mouths of normal individuals, where it is found around the teeth and in the tonsillar crypts. Trauma is an important predisposing factor in the development of 'cervicofacial' actinomycosis, although the exact conditions necessary to cause an infection are unknown. The infection may originate in a tooth socket and spread to adjacent tissues to produce a large hard, woody mass involving the face, jaw and neck. Softening occurs and multiple sinuses may develop, through which the characteristic pus exudes. The nose is rarely the site of primary infection but nasal actinomycosis has occurred following the implantation of xenologous processed bone for atrophic rhinitis (Thomas, Toohill and Lehman, 1974).

The general symptoms are pyrexia, toxaemia and rarely death. There is extensive tissue destruction and scarring.

Treatment consists of penicillin in large doses for 4-6 weeks and surgical drainage.

Candidiasis (moniliasis)

Candidiasis, commonly known as thrush, is caused by the yeast-like fungus Candida albicans which is a common inhabitant of the skin and oral cavity.
The infection occurs very commonly in the mouth and occasionally in the nose in neglected and marasmic infants and old people. It may occur in epidemic form in institutions and may be seen as a complication following courses of broad-spectrum oral antibiotics, long courses of systemic steroids and in immunosuppressed patients. There is a predisposition to candidiasis in patients suffering from diabetes, tuberculosis and AIDS.

Candidiasis presents as small, discrete, pearly or dirty-white patches in the mucous membrane on a red moist surface. The patches are easily removed without bleeding.

The condition responds to simple cleansing and painting with 1% aqueous solution of gentian violet or the local application of nystatin; alternatively amphotericin (Fungizone) or flucytosine (Alcobon) may be given in severe cases. Any predisposing cause should be sought and corrected.

Histoplasmosis

Histoplasmosis is an infestation due to a yeast-like fungus, *Histoplasma capsulatum*. It is most commonly found in central regions of the USA, but cases have been described throughout the world. Histoplasmosis is a diffuse disease of the reticuloendothelial system and is usually manifest by enlarged spleen, liver and lymph nodes with intestinal ulceration and anaemia. Nasal lesions are rare and may be nodular or infective. Secondary lymphadenitis develops.

The diagnosis is made by biopsy and the histoplasmin skin test which helps to differentiate pulmonary lesions from tuberculosis.

Treatment is with amphotericin (Fungizone).

Sporotrichosis

This is primarily an infection of the skin, usually of the hand, caused by *Sporothrix schenckii*. It very rarely affects the nasal mucous membrane but could be transposed there either directly from a lesion on the hand or by haematogenous spread. Infection follows a prick with a thorn. After a few days a nodule develops which becomes red and tender, finally bursting to discharge viscid yellow mucopus in which organisms may be found. Spread is also by the lymphatics along which secondary nodules develop.

Treatment is with iodides or amphotericin (Fungizone).

Nasopharyngeal leishmaniasis

This condition, sometimes known as American leishmaniasis or espundia, is caused by *Leishmania brasiliensis* as distinct from *L. donovani* - the cause of visceral leishmaniasis or *L. tropica* - cutaneous leishmaniasis. It is found chiefly in South and Central America and is transmitted by the sandfly (*Phlebotomus*).
The Leishman-Donovan bodies, which in the mammalian host do not occur in flagellated form, are approximately oval in shape and 2-6 microm in length with an eccentrically placed vesicular nucleus. They may be demonstrated in the discharge from the ulcers and in the reticuloendothelial cells in the granulomatous tissue.

The site of inoculation is usually on the exposed parts where a papule resembling a chancre develops and ulcerates, later healing and leaving a scar. Polypoid growths may form and there may be extensive destructive lesions involving the soft tissues or cartilage of the nasal septum, mouth, pharynx and larynx. Bone is generally not involved. There may be regional lymphadenitis and in untreated cases death follows from exhaustion.

**Myiasis**

Nasal myiasis, which is not uncommon in hot and humid climates, particularly in India where it is known as peenash, is a demoralizing condition of infestation of the nasal cavities by maggots, the larvae of a fly (genus *Chrysomyia*).

Myiasis can also affect the ear, mouth or larynx and reaches a peak in the months of September to November. It can affect any age, and both sexes equally. The majority of sufferers live in bad hygienic conditions and have a source of offensive decaying material, for example atrophic rhinitis, chronic sinusitis or chronic suppurative otitis media, which provides a suitable environment for the eggs of the fly to hatch into larvae no less than 1.5 cm in length. The eggs may also be deposited in a slight abrasion or crack in mucous membrane.

The entomological aspects of myiasis are well described by Sood, Kakar and Watlal (1976). The patient complains of a diffuse swelling around the nose and eyes, nasal obstruction, epistaxis or the presence of maggots coming out of the nose. Rhinoscopy reveals a congested oedematous mucosa, necrotic material with embedded maggots, ulcerated mucosa or septal perforations. The disease can spread to the paranasal sinuses and via the nasolacrimal duct to the lacrimal sac. In the later stages the nasal bones may be destroyed and death may result from sepsis, meningitis or suicide.

Treatment is general, with antibiotics and supportive therapy, and local, with olive oil or liquid paraffin which stifle the larvae. Maggots are removed piecemeal and the nasal cavity is douched. To prevent further infestation the predisposing conditions of poor hygiene and a source of chronic infection must be removed.
Chapter 9: Nasal polyps

A. B. Drake-Lee

Nasal polyps are an easily recognizable clinical entity. They result from the prolapsed lining of the ethmoid sinuses and block the nose to a variable degree depending on the size. On examination polyps appear as pale bags which arise most commonly from the middle meatus and are relatively insensitive when probed. This helps to differentiate polyps from mucosa of the middle turbinate which may be polypoid. The pale colour is a result of the poor blood supply but, in the presence of repeated trauma and inflammation, they may become reddened. They are usually bilateral and, when unilateral, transitional cell papilloma (Ringert's tumour, inverted papilloma) or malignancy need to be excluded. Antrochoanal polyps are similar in colour to ethmoidal ones, but are a different disease affecting the maxillary sinus, where the lining prolapses by way of the ostium into the nasal fossa and postnasal space.

Simple polyps may arise at any time after the age of 2 years, and if suspected before this they are likely to be meningoceles or encephaloceles and the floor of the anterior cranial fossa should be examined radiographically. However, it is unusual for simple nasal nasal polyps to arise before the age of 10 years and if found may be the presenting complaint of cystic fibrosis (Schwachman et al, 1962). It follows that any child with nasal polyps should have cystic fibrosis excluded by sweat tests. It is much more common for polyps to arise in established cases (Schwachman et al, 1962; Drake-Lee and Pitcher-Willmott, 1982).

Although polyps are a disease of the ethmoid sinuses, the mucosal changes frequently extend further into the nose and into the other paranasal sinuses. The maxillary sinuses are affected more commonly than the frontal and sphenoid sinuses. The extent of these changes may be seen radiographically. The mucosal changes may not be limited to the nose since patients may have coexisting asthma.

There are three factors which may be involved in the pathogenesis of nasal polyps; the mucosal reactions at the cellular level; the relatively poorly developed blood supply of the ethmoid sinuses; and the complex anatomy of the ethmoid labyrinth.

Treatment is a combination of surgery to the polyps and medical therapy with topical corticosteroids. Whatever therapeutic regimen is used, nasal polyps are a chronic condition which is prone to recurrence, in some cases with surprising frequency.

Historical background

The condition was first recognized in India and by 1000 BC curettes had been devised to remove them (Vancil, 1969). Hippocrates (460-370 BC) recognized them as well and devised a method to remove them using a piece of string which was passed through the nose into the nasopharynx. Sponge was attached to the postnasal end and the sponge was then pulled through the nose removing all before it! The word polyp comes from the Greek, although it was subsequently latinized and means many-footed (poly-pous). Snares and forceps similar to those used today were developed in the middle ages. All polypoidal conditions were initially grouped together until histological classification helped to
differentiate them from the neoplastic conditions (Berdal, 1954). Billroth, who described their histological characteristics in the middle of the nineteenth century, still considered them neoplastic. Zuckerkandl understood that they were an inflammatory condition. He also demonstrated that the histological changes in the sinuses were the same as those in polyps.

The first advance in treatment was the introduction of cocaine for local anaesthesia. In addition to the anaesthetic properties, it is also a vasoconstrictor. It is still widely used today, sometimes in combination with a general anaesthetic. More extensive surgery is better performed under general anesthesia. Better illumination has aided surgery. With a more controlled technique of anaesthesia, removal with the aid of a microscope and by endoscopy may be favoured.

Aetiology

There has been a number of different theories put forward for the pathogenesis of nasal polyps. Although a single aetiology would be attractive, this may not be the case, but it appears that patients may be divided clinically into several groups. There are five main theories of pathogenesis: the Bernoulli phenomenon, polysaccharide changes, vasomotor imbalance, infection and allergy. All may contribute to polyp formation, but none can be universally incriminated.

Bernoulli phenomenon

The Bernoulli phenomenon results in a pressure drop next to a constriction. This sucks the mucosa of the ethmoids into the nose. If this were true then the mucosa nearest the nasal valve would always be polypoidal in the normal nose.

Polysaccharide changes

An alteration in the polysaccharides of ground substances has been postulated by Jackson and Arihood (1971), but analysis of polyps has shown them to be oedematous (Taylor, 1963) with little alteration in the collagen. The collagen appears normal on analysis, although it tends to be recently formed.

Vasomotor imbalance

Vasomotor imbalance is implied because the majority of cases are not atopic and no obvious allergen can be found. Patients frequently have a prodromal period of rhinitis prior to occurrence of polyps. Polyps themselves often have a very poor nerve supply and they may be palpated freely and insensitively. Blood vessels are encountered in polyps but they are infrequent and usually comprise capillaries. Larger polyps have little smooth muscle within them. Vasomotor problems may cause polyps but this is conjecture alone.

Infection

The literature of the 1930s and 1940s proposed the concept that there were two types of maxillary sinusitis - purulent and hyperplastic - however, it became increasingly clear that the terms have been used interchangeably. Purulent sinusitis results from infection, usually
by bacteria. The inflammatory changes may extend into the ethmoids and cause the mucosa to become polypoidal. It is encountered today as unilateral disease. Hyperplastic sinusitis is associated with mucus hypersecretion in which organisms may be found and cultured. Infection may, like chronic bronchitis, exacerbate the condition, but it does not cause it. Inappropriate surgery on the maxillary sinus leaves an intranasal antrostomy through which the new mucosa undergoes the same changes and subsequently prolapses through the artificial ostium. Polyps now appear from both the middle and inferior meatus.

The cause of inflammatory reactions is uncertain. There are mucosal changes in the maxillary sinus in the majority; these are labelled sinusitis and it is frequently inferred that the sinuses are 'infected'. Indeed, mucus is washed out and when cultured a significant proportion grow an organism. The commonest organism is the non-capsulated *Haemophilus influenzae* (Majumdar and Bull, 1982). This bacterium is a common commensal in the nose and oropharynx and is frequently cultured from the sputum of patients with chronic bronchitis. In the latter case it exacerbates the condition and it may do so for cases with nasal polyps. Unfortunately, it is difficult to implicate any further relationship because antibiotics have little effect on the course or recurrence of the disease and merely modify the infectivity of the mucus. The actions of corticosteroids allow no place for infection as a primary cause. Corticosteroids improve the condition in over one-half of the cases and, where they do not improve, they certainly do not exacerbate it as expected if infection plays an important role. It may be possible that patients are allergic to bacteria, but no evidence has been found nor any mechanism shown.

**Allergy**

Allergy has been implicated because of three factors: the histological picture where 90% or more of nasal polyps have an eosinophilia; the association with asthma, and the nasal findings which may mimic allergic symptoms and signs. Unfortunately allergy is an imprecise term which is used in a variety of ways. It was originally used by von Pirquet in 1906 to describe the altered host reactivity to an antigen which, in today's terms, is any immune response. It is now used more commonly to mean hypersensitivity. The immune reactions involved cause tissue damage and are mainly mediated by the immunoglobulin IgE which is attached to the mast cell. Degranulation of the mast cell is a rapid event and is complete within 30 minutes.

It is now clear that mast cells may be triggered by other reactions including IgG4, complement activation, some drugs, chemicals and non-specific factors. The resulting degranulation produces similar symptoms whatever the trigger. The mast cell releases preformed elements, histamine, heparin and other vasoactive and chemoatcic factors and generates the metabolites of arachidonic acid, the prostaglandins and leukotrienes, the latter including slow reacting substance of anaphylaxis (SRS-A).

Clinically it is easy to consider symptoms such as attacks of anterior rhinorrhea, sneezing and blockage as allergic when no obvious cause is found and the patients have one or more positive skin tests.
Recent work

Mast cells

Mast cells are a heterogeneous collection of cells and have been divided in animals into two main groups: mucosal and connective tissue types; in addition, circulating basophils may also enter the tissue. Ultrastructural analysis showed that mast cells were degranulated (Cauna et al., 1972) and this has been confirmed, but further comment on the granule morphology and cell structure suggests that the features may not be consistent with those described in the allergic nose (Drake-Lee, Barker and Thurley, 1984). The mast cells are also degranulated on the inferior turbinate of over one-half of the patients. It suggests that mast cell reactions within the nose as a whole may be important in the development of polyps in these cases. Mast cell reactions occurring within the polyps would lead to oedema once the condition has started.

Nasal polyps oedema

Histological examination has demonstrated that polyps are mainly oedematous (Taylor, 1963). The extracellular oedema is easy to extract and has been analysed in many studies (Berdal, 1954; Donovan et al., 1970; Drake-Lee and McLaughlan, 1982). Following removal, the polyps may be coarsely minced and centrifuged and the resulting sera collected and analysed. Matched serum may be taken at the same time. Berdal injected polyp fluid subcutaneously and repeated skin tests. Those patients who had a positive skin test had greater reactions when tested again at the site of injection. Donovan et al showed that the level of IgE was raised in polyp fluid irrespective of the results of skin tests.

Immunoglobulins

Nasal polyp tissue continues to behave as normal respiratory mucosa in some respects and it is able to produce immunoglobulins from the plasma cells present. All immunoglobulins are found in polyp oedema, both IgA and IgE levels tending to be higher in polyp fluids than in sera. The levels of IgG, IgA and IgM are variable and elevated levels probably represent a recent upper respiratory tract infection. IgA is a dimeric immunoglobulin which has a junctional chain and secretory piece which makes it stable in mucus. It is the surface immunoglobulin of the respiratory and gastrointestinal tract. The levels are higher in polyp fluid and so it could be argued that this causes polyps in a manner similar to that advocated for IgE.

Allergen-specific IgE

IgE was discovered as the main immunoglobulin in immediate hypersensitivity (Ishizaka and Ishizaka, 1967) and soon afterwards the radioallergosorbent test (RAST) was developed to detect allergen specific IgE in serum (Wide, Bennich and Johansson, 1967). Mixed grass pollens and house-dust mite are the commonest allergens to cause allergic rhinitis. Only 25% of patients with nasal polyps have positive skin tests to these allergens (Drake-Lee et al., 1984). Since these are the two most commonly positive skin tests in patients with nasal polyps it would seem logical to expect these to be the commonest raised levels of allergen specific IgE. RAST levels in polyp fluid and sera however are raised only
infrequently (John and Merret, 1979). This would suggest that allergic reactions may occur but are infrequently encountered in patients with nasal polyps.

**Free histamine in polyp fluid**

When mast cells degranulate a variety of products is produced of which histamine is the easiest to measure and has been measured in polyp oedema (Drake-Lee and McLaughan, 1982). Levels which are between 100 and 1000 times the serum level are encountered. This would suggest that when mast cells degranulate, local homeostatic mechanisms may be overcome. This occurs most easily in the ethmoid sinuses, partly because of the anatomy which allows the mucosa to prolapse into the nose, and partly because the blood supply is less well developed here and is therefore less able to remove vasoactive compounds. This is a dynamic state so that polyps will vary in size.

**Asthma and nasal polyps**

The association of nasal polyps and asthma has long been accepted and has recently been reviewed (Maloney and Collins, 1977). Around 20-40% of patients with polyps have coexisting asthma and it appears that a similar proportion of adults with asthma have nasal polyps. Unfortunately earlier work suggested that polypectomy caused asthma, but studies were anecdotal and could not be confirmed.

It is the late onset asthma which is associated with nasal polyps rather than the childhood asthma. The incidence of childhood asthma is about 5% of the population and it was 3.5% in a recent study of cases with nasal polyps (Drake-Lee et al, 1984). Asthma usually commenced around the onset of nasal polyps with over one-half of patients developing either polyps or asthma within 5 years of each other. Surgery has little effect on asthma; some patients noticed a subjective improvement in their asthma (Maloney, 1977). It has been suggested that patients with asthma may be a distinct subgroup within the disease because the sex incidence of patients with asthma and polyps is equal whereas polyps usually occur more frequently in males.

**Aspirin hypersensitivity**

Patients with aspirin hypersensitivity, asthma and nasal polyps are a well-recognized subgroup (Samter and Beers, 1968) which occurs in up to 8% of patients with nasal polyps. The mechanism for both aspirin hypersensitivity and asthma is unclear, but it is not an allergic reaction and there is some suggestion that there is an alteration in prostaglandin synthesis (Sczeklik, Gryglewski, Czerniawska-Mysik, 1975).

**Recurrence**

Recurrence of nasal polyps is one of the problems facing every otolaryngologist in the management of these cases. The rate of recurrence is variable. A 2-year study showed that 5% of patients had had five or more previous polypectomies (Drake-Lee et al, 1984). It is difficult to study those factors which are associated with recurrence, but several appear to be important. As might be expected those patients developing polyps at a younger age and associated with a long history of nasal complaints tend to have more severe recurrence.
Patients with severe nasal disease often have more extensive surgery, but this has not been shown to decrease the recurrence rate. Perhaps the single most easily detectable aetiological factor is the association with asthma. Patients with asthma suffer more severe recurrence in general and, if they have aspirin hypersensitivity, then this is increased even more. It is noteworthy that hay fever, childhood asthma, penicillin allergy and multiple positive skin tests, all manifestations of allergic diatheses, are not associated with severe recurrence.

**Age**

Nasal polyps are a disease of adults, although children with cystic fibrosis and occasionally teenagers develop them. The incidence every 10 years is equal between the ages of 30 and 60 years following which the prevalence decreases. Patients who are atopic and asthmatic do not develop polyps any earlier.

**Incidence**

The true incidence is difficult to assess, but it may be inferred from the incidence of asthma since the frequency of nasal polyps in late onset asthmatics is the same as that of late onset asthma in patients with nasal polyps. The incidence of asthma is about 5% with over one-half of the patients developing asthma during childhood. It would be expected that between 1 and 20 per 1000 of the adult population would have nasal polyps one or more time in their life. This would fit with the general prevalence of nasal disease in the UK where about 10% of the population suffers from seasonal allergic rhinitis (hay fever), about 5% suffer from perennial allergic or vasomotor rhinitis and of these only a proportion would develop nasal polyps.

**Sex**

There is a strong male predominance in patients who have polyps; figures range between 2:1 and 4:1 depending on the study. The male predominance is also present in children with cystic fibrosis (even though more males have cystic fibrosis). The sex incidence of patients with asthma and nasal polyps is equal which suggests a different clinical subgroup.

**Racial groups**

Nasal polyps have been reported in all major racial groups although the comparative incidence has not been documented.

**Animals**

The only other animal to have nasal polyps is the chimpanzee and polyps occur infrequently. Although cattle may develop polypoid lesions of the nasal septum and cats suffer with eustachian tube polyps, they are different lesions so it would appear that this disease is virtually confined to man.
Macroscopical features

Polypoidal lesions may arise from the nasal mucosa particularly the middle and inferior turbinate and also from any respiratory mucosa in the nose. The classic and commonest situation for their development is the ethmoid sinuses; they arise from beneath the middle turbinate anteriorly and above the middle turbinate posteriorly. Polyps may occur in the other sinuses, particularly the maxillary antrum and following inappropriate surgery in the antrum, they prolapse through the antrostomy. The colour of nasal polyps varies but it is usually a translucent white bag that blocks the nose to a variable degree. Polyps may become red with repeated trauma and nasal infection and, in the most florid cases, prolapse through the anterior nares. The degree of development varies from side to side and without therapy the size may also vary considerably; up to one-quarter will regress spontaneously without treatment. Polyps are usually multiple.

Histology

Tissue removed from the maxillary sinuses, nasal polyps and the bronchi (from patient from status asthmaticus) are similar. Nasal polyps usually have a respiratory epithelium with ciliated columnar epithelium with goblet cells. If there has been repeated trauma, squamous metaplasia occurs. The gross oedema will give rise to artefacts when polyps are processed for scanning electron microscopy since this process involves the dehydration of material. As the polyp shrinks the surface epithelium is lost to a variable extent and this has been described as 'cobblestones'. There is apparent thickening of the basement membrane which will vary from area to area and polyp to polyp. The submucosal tissue is grossly oedematous and contains few blood vessels, which are mainly capillaries, and the occasional nerve fibre. The cellular infiltrate is mainly plasma cells, small lymphocytes, macrophages, and the most striking feature, an eosinophilia. The eosinophilia may be very variable not only from patient to patient but also between polyps in the same patient. Eosinophils are found in 90% of polyps, the remaining cells being neutrophils. Occasionally, some of the stromal cells may show marked atypia (Friedman and Osbourne, 1982).

Clinical features

Nasal obstruction

Almost without exception all patients suffer from nasal blockage. This is constant, although it will vary with the size and position of polyps. The mildest form of complaint is congestion. Patients will often complain they feel as though they have a cold at times and this may be socially embarrassing. Usually there is some nasal breathing. It may be that, following surgery with the improvement of nasal function with adequate warming and humidification of the air, those patients with asthma notice an improvement in their chest symptoms.

Rhinorrhoea and sneezing

About one-half of patients suffer from attacks of either rhinorrhoea and/or sneezing and this may be helped by surgery when large areas of oedematous mucosa are removed. Patients who suffer with these symptoms will often say that they have hay fever. This is
because the symptoms are the same, but they are perennial and even if intermittent have no obvious triggers.

**Sense of smell**

Partial loss of sense of smell and alterations in taste are common complaints. These do not usually recover following treatment except in some cases treated by corticosteroids, particularly when taken orally when there may be a general improvement in respiratory function.

**Pain**

Although not frequent, pain does occur in patients with polyps and is usually over the bridge of the nose, the forehead and cheeks. It is worse when the nose is congested or when the postnasal drip changes in colour.

**Postnasal drip**

Most patients will complain of some postnasal drip. It is usually white, but may become green or yellow, particularly with maxillary pains or following exacerbation of nasal symptoms. Alteration of the mucus and its hypersecretion is a consequence of inflammation irrespective of cause; since nasal polyps are an inflammatory condition then mucus hypersecretion occurs. A severe eosinophilia may change the colour of the mucus from white to yellow or a yellow/green colour and was called allergic pus in the 1930s. The postnasal drip may improve following surgery or, if the mucus changes to green, may become normal following antibiotic therapy.

**Epistaxis**

This is infrequent and follows extensive clearing of the nose; if it does occur as a major symptom then it indicates a more sinister pathology underlying the nasal lesion.

**Signs**

Patients have a distinctive hyponasal voice and when the blockage is so severe then polyps may be seen externally. The obstructed nasal breathing may produce mouth breathing and occasionally flaring of the alar cartilages. This later sign is usually produced by the polyps themselves. If polyps develop before the nasal and facial bones fuse, hypertelorism will develop in more florid cases: it is seen in children with cystic fibrosis. This may also occur in adults if polyps remain untreated. The intranasal signs have been discussed previously.

**Investigations**

There are no specific haematological, biochemical and immunological investigations that are required apart from those involved in the general examination of cases prior to surgery. Skin tests have been widely used to investigate nasal cases but the incidence of positive skin tests is no greater than expected in the general population (Pepys and Duveen,
Those patients with positive skin tests do not present any earlier nor have more severe occurrence. Cases with hay fever and dust mite allergy should have these treated in their own right. Sweat tests should be performed in children.

Radiology

Plain radiographs of the paranasal sinuses taken by the conventional three views will show the extent of the disease in the nose and paranasal sinuses. Radiotranslucency in the nose, hypertrophy of the turbinates, and deviation of the bony septum may be seen. The ethmoid complex is almost always opaque on the side of the polyps. The maxillary sinus will have changes in most cases with mucosal thickening of a variable degree until the antrum may eventually become opaque. Fluid levels are encountered and may be due to retained secretion alone or purulent material, since blockage of the maxillary ostium by polyps will prevent the migration of mucus from the sinus. Expansion of the ethmoids will be encountered in those with polyps which developed before the bones fused. Bony erosion, although highly suggestive of malignancy, may be found in patients who have polyps and is the result of previous surgery. Previous surgery is generally implicated in the mucocoeles which may develop. They are most commonly frontal or ethmoidal with a rare primary sphenoid mucocoele (Lund and Lloyd, 1983). Plain radiographs of the sinuses should be taken on all patients who present with polyps to determine the extent of the disease, exclude any bone changes, and as a baseline for follow-up.

Treatment

Unfortunately much of the literature on the treatment of polyps is anecdotal and has not been subject to useful trials and scrutiny. It is the author's view that, unless any operation has been shown to be more effective than another, it is best to perform the simplest operation with the best view and with least risk of harm to the patient. Most surgeons today treat polyps surgically, but many polyps are sensitive to corticosteroids, and where polyps are not obstructing the nose completely a preoperative trial of corticosteroids is worthwhile.

Preoperative medical treatment

The proportion of patients sensitive to corticosteroids is uncertain but approximately one-half seem to have some response. It is not possible in most otolaryngological clinics to give the thorough medical examination required before commencing oral corticosteroids, so they should be avoided even though they are more effective than topical preparations.

Betamethasone nose drops, two drops twice daily each side, may be given for one month. The position of administration should be either in the recumbent head dependent posture, or the 'Mecca' position which is kneeling with the head well down and forwards. It allows drops to penetrate more easily into the ethmoid region (Charlton et al, 1985). Alternatively aqueous beclomethasone or flunisolide, two puffs, may be tried (Dingsor et al, 1985). Aqueous preparation are probably more effective than Freon sprays. If patients have congested and polypoidal turbinates this may be the more effective method of obtaining a general cover of the nasal mucosa. Polyps may disappear completely and treatment should be continued for at least 3 months.
**Surgical treatment**

**Anatomical considerations**

The anatomy of the nose and ethmoids has been considered elsewhere but there are several points which should be highlighted. The middle turbinate is the key to nasal surgery. If surgery is performed medially, then the cribriform plate may be breached and the anterior cranial fossa entered. If surgery is lateral then the ethmoid complex may be entered. Extensive intranasal operations on the ethmoids render the middle turbinate unstable, if it is lost or removed it makes subsequent orientation difficult. Since may operations are performed subsequently by junior surgeons the risk of complications is greater.

The eye is the lateral relation of the ethmoids and the lamina papyracea is very thin; it is easy to enter the orbit if ethmoid surgery is vigorous. The simplest complication is herniation of orbital fat, but more extensive surgery may injure the medial rectus anteriorly and the optic nerve posteriorly.

**Preoperative preparation**

The nose should be prepared adequately before surgery to cause vasoconstriction and shrinkage of the mucosa. Drier fields are obtained under local anaesthesia alone. Various methods are used to prepare the nose and each will give a good field if used well. The author prefers to perform nasal surgery under local anaesthesia if possible, but patients should be admitted and sedated with an adequate intramuscular premedication. Surgery should be performed in theatre where all facilities are available. Preparation with cocaine 10% spray is given on the ward 15 minutes before local anaesthesia. This allows a much more thorough local anaesthetic to be given. Cocaine paste 25% is used on wires and the mucosa is painted thoroughly. Nerve blockage is obtained with one wire far back under the middle turbinate to block the sphenopalatine ganglion. The anterior ethmoidal nerves are blocked by a wire between the nasal bones and septum. If the inferior meatus is to be entered a further probe is inserted here as well.

Silver is used for the wires as it is malleable and bends if the patient were to have a vasovagal attack and fall forwards. These attacks are prevented by premedication and performing the local anaesthetic with the patient lying flat. Adequate time should be left for the anaesthetic to take effect. When general anaesthesia is used, an endotracheal tube and throat pack are required in addition as with all nasal surgery.

**Surgery**

There are different views on the type of surgery required for nasal polyps. Many patients will have infrequent recurrence and to advocate an extensive operation initially before the problem of recurrence is evident is illogical. Simple polypectomy is the treatment of choice. Polyps may be removed either by an avulsion or cutting snare or forceps such as Tilley Henckel's. Care must be taken when using forceps not to go either too medially or too far lateral. All polypoidal mucosa should be removed from the ethmoids, the lower border of the middle turbinate and the inferior turbinate.
Although intranasal ethmoidectomy is advocated by some authors, it is neither a complete nor safe operation. It is impossible to remove all the anterior and posterior ethmoid air cells without making the middle turbinate unstable. If this is lost, os is the main intranasal landmark.

External ethmoidectomy is performed through an incision medial to the inner canthus of the eye (Howarth's) or through an incision in the natural skin crease below the infraorbital margin (Patterson's). All the ethmoid cells may be removed once the orbit has been displaced laterally with the lacrimal apparatus and the anterior ethmoidal vessels divided. The anterior ethmoidal vessels provide a clue to the level of the cribriform plate. All the ethmoidal cells should be removed and the sphenoid sinus may be entered if desired. Care should be taken to open the ostium of the frontal sinus widely to prevent mucoceles which are a late complication of surgery. There have been no trials to determine whether external ethmoidectomy prevents recurrence, although it has some advocates (Hughes, 1973).

The mucosal changes may extend into the maxillary sinus and some surgeons advise the Jansen-Horgan procedure. This is the combination of a Caldwell-Luc operation with a posterior ethmoidectomy through the antrum and an intranasal antrostomy and middle ethmoidectomy. It is rarely performed today.

The Caldwell-Luc operation may be undertaken alone for maxillary mucosal disease but, frequently, recurrent polyoid mucosa prolapses through the antrostomy in the inferior meatus. This is exceptionally difficult to treat.

Complications of surgery

The main immediate complication is haemorrhage. This occurs at the time of surgery and is usually minimized if the operation is performed under local anaesthesia. It is often unnecessary to pack the nose when topical vasoconstrictors are used and the patient feels the immediate benefit of the improved airway. Packing will control the haemorrhage in virtually every case particularly when simple polypectomy alone is performed. Slight ooze and serosanguineous discharge occur from the raw areas for the next few postoperative days.

The other principal complications occur from damage to the cribriform plate and the orbit. The anterior cranial fossa may be entered and this may result in cerebrospinal fluid leakage only or, more importantly, meningitis and abscess formation. Complete anosmia is rare since in most cases the olfactory bulb will remain undamaged. The orbit may be entered through the lamina papyracea or through the posterior ethmoid air cells. Damage to the orbital periosteum may cause herniation of periorbital fat, medial rectus palsy, damage to the anterior and posterior ethmoidal arteries and division of the optic nerve.

Recurrence of nasal polyps is not a complication of surgery since it is a feature of the disease.

Postoperative medical care

There is no single approach to care. Some surgeons advocate a 10-day course of decongestants and steam inhalations for all patients who have undergone nasal surgery. The
role of long-term medical treatment with corticosteroid sprays has yet to be fully evaluated, but evidence suggests recurrence may be controlled (Mygind et al, 1975; Deuscht and Drettnner, 1977). There are two main reasons to give corticosteroid sprays - to control this symptoms of rhinitis and to prevent recurrence.

Patients who have rhinitis and polyps do not, as a group, seem more prone to recurrence. Any trial that groups control of these symptoms together with recurrence is open to question. There is only one true test of control and that is the regimen which gives rise to fewer operations. There would seem to be no case for giving all patients who present with nasal polyps, for the first time, long-term steroids until the results of large clinical trials are available. In those patients with established disease, a one-year trial of medical treatment would seem to be appropriate. It is sometimes necessary to combine corticosteroid nose drops with an aqueous-based corticosteroid spray.

Antihistamines have virtually no place in the management of recurrence. The sedation caused by some products is unacceptable and, although non-sedating antihistamines are available, enzyme induction occurs in the liver after 6 weeks and so the effect of the antihistamine is reduced. A short course may be prescribed if the rhinitis is difficult to control.

**Diets**

There has been much enthusiasm recently for treating allergy to foods by exclusion diets. Tartrazine dyes are linked to aspirin pharmacologically and, since a group of patients has aspirin hypersensitivity, it would seem logical to exclude tartrazine dyes from their diet. No controlled trials have been performed, but patients frequently say that they feel better and have fewer nasal symptoms. The treatment does not harm the patient, has no side-effects and may help some of those who want to try to do something themselves to prevent recurrence.

**Antrochoanal polyps**

These polyps arise in the maxillary antrum and prolapse through the ostium of the sinus in the middle meatus. They hang either in the nose or, if larger, into the posterior choana. The choanal part of the polyp may be seen in the oropharynx where it pushes the soft palate downwards and forwards.

Antrochoanal polyps (Killian's polyps) are rare and probably occur in all racial groups. Like benign nasal polyps they are more common in males than females. The onset is usually before 40 years, although they may be found at all ages. The polyps tend to be dumb-bell in shape with a constriction where they pass through the ostium of the sinus. They occur usually either from the left or right maxillary sinus alone, but may rarely be bilateral. Attempts have been made to define from where in the maxillary antrum they arise. The floor and lateral wall are more common, although their site of origin cannot be determined frequently. The polyp is similar in colour to the simple types being pale white or translucent in appearance.

Histology shows a respiratory epithelium over a normal basement membrane. The ultrastructure is grossly oedematous and the cellular infiltrate is similar to ordinary polyps except that there is no eosinophilia. There have been no ultrastructural studies of the polyps.
The commonest symptom is unilateral nasal blockage, although when very large they may cause bilateral blockage. Other nasal symptoms are uncommon except for anterior nasal discharge which is usually mucoid. The polyp may not be visible on anterior rhinoscopy but is usually seen posteriorly, occasionally without the aid of the mirror.

Radiographs of the sinuses may show mucosal thickening or a completely opaque antrum. They are almost never normal on the affected side. The lateral view may show the polyp in the postnasal space.

**Aetiology**

Antrochoanal polyps are an entity of unknown aetiology. They are not associated with allergy, lower respiratory tract disease or sinusitis. Proetz suggested that they may be the result of a faulty development of the maxillary sinus ostium since it is always large. The ostium may be large because of expansion by the polyp but this is unlikely since there is no expansion of the posterior choana by large polyps nor is there any erosion or displacement of the middle turbinate medially.

**Treatment**

There is no medical treatment either preoperatively or postoperatively. Preoperative nasal preparation with a vasoconstrictor is essential. It is necessary to remove both parts of the polyp. There has been debate on the best method of removal. The approach is dictated by the age of the patient. Simple intranasal polypectomy alone will almost always be followed by recurrence. Many of the patients are young and the dentition is incomplete so that a Caldwell-Luc approach is not indicated. An antral wash out may produce straw-coloured fluid and should be performed since it may help in the dissection of the antral mucosa if simple polypectomy is performed. It is often impossible to remove the polyp through the nose, therefore it has to be delivered through the oropharynx. Larger polyps may be difficult to remove because they develop adhesions in the nose which have to be broken by blunt dissection.

A Caldwell-Luc antrostomy is the treatment of choice in adults since recurrence will be reduced. In children, once dentition is complete, simple polypectomy is replaced by the more radical procedure. All the lining of the sinus is removed together with the polyp. It is debatable whether an antrostomy into the inferior meatus is required.
Chapter 10: The nasal septum

David Brain

Injuries of the septum

The anterior part of the nasal septum projects in front of the plane of the pyriform aperture and is frequently damaged when the nose is injured. This may result in a haematoma formation and/or septal deviations.

Septal haematoma

When the septum is subjected to a sharp buckling stress, submucosal blood vessels are frequently torn and, if the mucosa remains intact, this will result in the formation of a haematoma. If the injury is severe enough to fracture the septal cartilage, the blood will often pass through to the other side and produce a bilateral haematoma. The blood mainly accumulates in the subperichondrial layer and this will usually interfere with the vitality of the cartilage which becomes avascular, as it depends on the perichondrium for its nutrition. Avascular cartilage can probably remain viable for 3 days, but after this the chondrocytes die, and absorption of the cartilage follows. Cartilage absorption can occur with alarming rapidity and Fry (1969) has suggested that the process is hastened by enzyme action, probably in the form of one of the tissue collagenases. Small haematomata will not cause this necrosis of cartilage, but may slowly absorb leading to permanent thickening of the septum with gross fibrosis.

Symptoms and signs

The dominant symptom is nasal obstruction, and initially there may be some discomfort. Examination is best made without a speculum, and will reveal a smooth rounded bilateral septal swelling which often extends to the lateral nasal walls causing complete obstruction.

Treatment

Fry (1969) has shown that the early surgical drainage of the haematoma reduces the risk of cartilage necrosis, and is therefore always indicated. A long hemitransfixion incision is made and usually the haematoma will have elevated the perichondrium from the cartilage. Blood is aspirated, together with any necrotic material. The state of the cartilage is carefully assessed and, if a defect is present, it is advisable to support the defect with homograft cartilage (Masing, 1965). This should be cut to a size slightly greater than the perforation and, after insertion, a small drain is inserted into the bottom of the cavity, and the mucosa is replaced and maintained in this position by nasal packing.

Cartilage grafts can be used even if abscess formation has occurred, as Masing (1965), Hellmich (1974) and Vase and Johannessen (1981) have all shown that these grafts take well, and effectively prevent the saddling deformities which otherwise invariably occur. The homograft cartilage can be harvested from patients who have undergone a submucous resection, and can be conveniently stored in 0.1% sodium mercuro-thiosalicylate.
Complications

*External deformity of the nose*

The cartilaginous dorsum of the nose is supported by the septal cartilage and, if this support is lost, dorsal saddling in the supratip area will result. If this type of injury occurs during childhood, it may also affect the development of the whole of the mid-third of the face with resulting maxillary hypoplasia.

*Septal abscess*

A haematoma may easily become infected and this will frequently lead to abscess formation. This complication is commonly associated with an increase in the severity of the pain, together with the usual manifestations of toxaemia, such as pyrexia and a raised pulse rate. The advent of secondary infection makes extensive cartilage necrosis virtually inevitable, and is an even more pressing indication for surgical drainage.

*Septal deviation*

Septal deviations are extremely common, but are not usually severe enough to affect nasal function. The incidence of these deformities is much higher in the leptorrhine type of nose found in the Caucasian races.

*Aetiology*

Many septal deviations are due to direct trauma and this is frequently associated with damage to other parts of the nose such as fractures of the nasal bone.

*The birth moulding theory*

In many patients with septal deviations there is no obvious history of trauma. Gray (1972) explains these cases by means of the birth moulding theory. Abnormal intrauterine posture may result in compression forces acting on the nose and upper jaws (the widest part of the face). Displacement of the septum can result and the nose can be exposed to further torsion forces during parturition. Jeppesen and Windfield (1972) found 29 cases of septal dislocation in 907 newborn infants (3.19%). Dislocations were more common in primipara and when the second stage of labour lasted more than 15 minutes. Dislocations were generally to the right in the case of left occipitoanterior presentations and to the left with right occipitoanterior presentations. Subsequent growth of the nose accentuates these asymmetries.

*Pathological anatomy*

Deformity of the nasal septum can be classified into the following types.

*Spurs*

These are sharp angulations which may occur at the junction of the vomer below, with the septal cartilage and/or ethmoid bone above. This type of deformity is usually the result
of vertical compression forces. Fractures through the septal cartilage may also produce sharp angulations. These fractures heal by fibrous union and the fibrosis extends to the adjacent mucoperichondrium. This increases the difficulty of the flap elevation in this area.

**Deviations**

These lesions are characterized by a more generalized bulge. 'C'- or 'S'-shaped deviations occur which can be either in the vertical or horizontal plane, and they usually involve both the cartilage and the bone.

**Dislocations**

Here the lower border of the septal cartilage is usually displaced from its medial position and projects into one of the nostrils.

Septal deviations are also frequently associated with anatomical abnormalities in adjacent areas.

*The lateral nasal wall*

A compensatory hypertrophy of the turbinates and ethmoidal bulla usually occurs on the side of the septal concavity.

*Maxilla*

The compression forces which are responsible for the septal deviations are often asymmetrical and may also involve the maxilla, producing flattening of the cheek, elevation of the floor of the affected nasal cavity, distortion of the palate and associated orthodontic abnormalities. The maxillary sinus is usually slightly smaller on the affected side.

*The external nasal pyramid*

Anterior septal deviations are often associated with deviations in the external nasal pyramid. Deviations may affect any of the three vertical components of the nose and there are three common types which are listed in order of severity.

*Cartilaginous deviations*

In these cases, the upper bony septum and the bony pyramid are central, but there is a deviation of the cartilaginous septum and vault.

*The C deviation*

In this lesion, there is displacement of the upper bony septum and the pyramid to one side and the whole of the cartilaginous septum and vault to the opposite side.
The S deviation

Here the deviation of the middle third (the upper cartilaginous vault and associated septum) is opposite to that of the upper and lower thirds.

With deviations of the nose, the dominant factor is the position of the septum. Beekhuis (1973) has succinctly summarized this principle with the dictum 'as the septum goes, so goes the nose'. The first step, therefore, in treating the twisted nose is to straighten the septum, and if this objective is not achieved, there is no hope of successfully straightening the external pyramid.

There is therefore a sound pathological basis to the concept of straightening a twisted nose by means of a one-stage septorhinoplasty procedure.

The effects of septal deviations

Only the more severe deviations affect nasal function and therefore require treatment.

Nasal obstruction

This is always found on the side of the deviation and is also often present on the opposite side as a result of the hypertrophic changes in the turbinates.

Mucosal changes

The inspiratory air currents are often abnormally displaced and frequently become concentrated on small areas of nasal mucosa, producing an excessive drying effect. Crusting will then occur, and the separation of the crusts often produces ulceration and bleeding. The protective mucous layer may then be lost and resistance to infection reduced. The mucosa around a septal deviation may become oedematous as a result of Bernoulli's phenomenon, which states that 'when there is a flow of gas through a constriction, it produces a negative pressure'. This negative pressure will, in turn, predispose to mucosal oedema in the affected area thus further increasing the obstruction.

Neurological changes

It is possible that the pressure exerted by septal deviations on adjacent sensory nerves can produce pain. This concept was first elaborated by Sluder (1927) and the resultant condition has been called 'the anterior ethmoidal nerve syndrome' (Shalom, 1963). In addition to their direct neurological effects, reflex changes perhaps may result from septal deformities which affect the nasopulmonary and nasal reflexes.

Symptoms

The symptoms caused by septal deviations are entirely the result of their effects on nasal function. The dominant symptom is nasal obstruction but this is rarely severe enough to cause anosmia. Douek (1974), in a review of many patients suffering from anosmia, never found this symptom to be the consequence of an uncomplicated septal deviation.
Signs

Septal deviations are usually quite obvious on anterior rhinoscopy. It is important first to inspect the nasal vestibule without using a speculum because the blade of this instrument can easily straighten the septum and thus hide a caudal deviation. Local anaesthesia with cocaine may facilitate the inspection of some of the more posterior deviations. Sometimes the patient complains of unilateral nasal obstruction and anterior rhinoscopy will reveal that the septal deviation is to the opposite side. This phenomenon has been called 'paradoxical nasal obstruction' (Arbour and Kern, 1975). These patients have a long-standing, fixed, unilateral nasal obstruction to which they may have become accustomed, and of which they are no longer aware. The mucosal swelling associated with the nasal cycle, results in an additional intermittent nasal obstruction on the wider side of the nose, and this becomes the dominant symptom to be appreciated by the patient.

Septal deviations in the region of the nasal valve area cause the greatest obstruction, because this is at the narrowest part of the nasal cavity. The Cottle test will confirm the fact that the obstruction is in the valve area. In this useful and simple test, the patient pulls the cheek outwards and opens up the internal nares and thus reduces the blockage.

The septum cannot be considered in isolation and it is therefore necessary to perform a careful inspection of the lateral nasal wall to determine the size of the turbinates. Examination must also include the external nasal pyramid, the palate and the teeth as these structures are often also involved to some degree with septal deformities. Whenever sinus complications are suspected, X-rays of the paranasal sinuses are indicated.

Septal deviation in the newborn is sometimes associated with asymmetry of the nostrils, an oblique columella and tip which points in the direction which is opposite to the deviation. The nostril on the affected side may look distinctly flattened. These characteristic features are rarely present, and most cases are diagnosed by anterior rhinoscopy and the use of Gray's struts. These struts are 4 mm wide and 2 mm thick and, after lubrication, are inserted into the nostrils, and then gently pushed backwards along the floor of the nose, hugging the septum. Normally the struts can be introduced for a distance of 4 or 5 cm, but in cases of deviation, a frank obstruction is encountered, usually 1.5-2 cm back from the nostril. This is the most reliable test, and is well tolerated during infancy. Less frequently, the compression test may be positive. In this test the nasal tip is pushed backwards and if there is a septal dislocation, it will collapse against the philtrum of the upper lip.

Indications for submucous operations on the nasal septum

Septal deviations

Cottle has classified septal lesions into three types.

Simple deviations

Here there is a mild deflection of the septum which does not cause obstruction. The majority of Caucasians have this type of septum, and it certainly does not require any surgical treatment.
Obstruction

This is a more severe deviation of the nasal septum which may touch the lateral wall of the nose, but on vasoconstriction the turbinates shrink away from the septum.

Impaction

This is a very marked angulation of the septum with a spur which lies in contact with the lateral nasal wall, even after the application of a vasoconstrictor. Surgical treatment is reserved for some of the obstructing lesions and most of those associated with impaction, the essential indication being a skeletal septal obstruction.

In many patients with septal deviations there is also some generalized mucosal pathology in the form of a perennial rhinitis, and this will not be corrected by septal surgery. Thomas (1978) has shown that the most common cause of poor results following a submucous resection operation is the presence of a coexisting perennial rhinitis. A test that would differentiate between the obstruction caused by a skeletal septal deviation and that as a result of mucosal pathology, would therefore be very helpful in assessing the suitability of patients for septal surgery (Cottle, 1968). Claims that this can be achieved by performing measurements of nasal resistance before and after vasoconstriction with either drugs or following physical exercise have been made by Broms (1982) and by Jessen and Malm (1984). Unfortunately, McCaffrey and Kern (1979) did not find these tests of much help for this purpose.

Closure of septal perforations

Most techniques which have been described for the closure of septal perforations involve the submucous elevation of the flaps for this purpose.

Source of grafting material

Submucous resection of nasal cartilage and, less commonly, vomerine bone, is sometimes required to obtain graft material for such operations as rhinoplasty and tympanoplasty.

To obtain surgical access

Submucous resection of the septum has been advocated as giving the necessary access for the following surgical operations to be performed:

(1) hypophysectomy (Hirsch, 1952)
(2) vidian neurectomy (Minnis and Morrison, 1971).

The development of septal surgery

The study of the history of septal surgery is both interesting and instructive. It clarifies the basic problems encountered in treating septal deviations, and demonstrates the limitations of the various techniques which have been evolved to solve them. During the nineteenth
century, surgeons started tackling these problems by a variety of techniques which have now been completely abandoned. Acute spurs and angulations were removed either by shaving down the convexities (Langenbeck, 1843; Dieffenbach, 1845; Chassaignac, 1851), or by performing a complete removal of the deviation by punch forceps (Rubrecht, 1868). The usual result of these operations was to exchange a septal deviation for a perforation. These techniques are only of historical interest, and there is little doubt that the first major breakthrough in surgical therapy occurred about the turn of the century with the development of the submucous resection operation.

As so often happens, the idea of effecting a submucosal removal of the deviation occurred to several surgeons working independently at the same time. Probably the earliest was Ingalls in 1881, but the names of Killian and Freer are usually associated with the refinement and popularization of the actual procedure (Ingalls, 1882). It was Killian (1904) who described the technique which is most commonly practised today, with a retention of both dorsal and caudal struts of cartilage to prevent any subsequent change in the external shape of the nose. Freer (1902) adopted a much more radical approach as, in his view, the septal cartilage did not contribute to the support of the nasal pyramid and could be completely removed if necessitated by the extent of the pathology. He admitted that 'saddling' of the dorsum did sometimes occur in the supratip region, but said that this was always due to rough surgery, which had damaged or partly removed the upper lateral cartilages.

The submucous resection operation was undoubtedly a great advance and was widely adopted throughout the world. With subsequent experience, however, it was evident that there were certain associated problems. For surgical purposes, the septum can be divided into anterior and posterior parts by a vertical line drawn from the frontal nasal spine to the maxillary nasal spine. Deviations posterior to this line can be easily and effectively treated by the submucous resection technique. The problems occur when using this technique in the anterior part of the septum. All too frequently, the operation was followed by a supratip depression and columellar retraction. To minimize these complications, most surgeons adopted the conservative Killian technique, but retention of dorsal and caudal struts does not ensure complete immunity; in addition, the deviations may be found in the region of the dorsal and caudal struts, and would therefore not be corrected by this operation. These complications occurs much more frequently than is generally realized because they often take many months to develop. Immediate saddling is rare; usually it occurs as a result of scar contraction in the septum. Some surgeons have attempted to solve the problem of scar contraction by replacing all or part of the excised cartilage, while others have avoided producing a large defect in the cartilaginous septum by mobilizing and repositioning the septum in the central position, so that the bulk of the cartilage is retained and is still attached to its mucoperichondrium as part of a compound flap.

The first significant improvement was made by Metzenbaum (1929) in Chicago, using the latter concept. The operation was applicable only to caudal dislocations of the septum without fragmentation of the cartilage and gross fibrosis. He likened the principle to that of a swinging door, but late failures were fairly common. A swinging door has a hinge on one side and free edges on the other three borders. In the Metzenbaum operation, the hinge was effectively produced by the incision at the level of the deviation. There was an existing free border inferiorly and one was produced posteriorly by separation of the cartilage from the vomer. There was not, however, a free border anteriorly where the septum was often tethered.
to displaced upper lateral cartilages and the traction from this source, and also sometimes from the mucoperichondrium which was liberated only on one side above the incision, produced increased tension on the unfreed side during healing, which was prone to cause a recurrence of the deflections. To overcome these problems, Peer, in 1937, completely excised the deviated caudal segment of the cartilage. If possible, he reinserted it as a free graft, but if the tissue was either unsuitable or inadequate, he obtained a similar sized graft resecting cartilage from the central or more posterior part of the quadrilateral cartilage. This operation developed the concept of cartilage excision followed by cartilage replacement. The original Peer operation was extended to include removal of the entire cartilaginous septum. This concept reached its logical conclusion in the Galloway operation (1946). Galloway removed the entire nasal cartilage, and replaced the anterior septum with a single free autograft cut from the excised cartilage. He also described a useful detail of operative technique, in the manner in which he facilitated the placing of the graft with traction sutures.

Afterwards, the graft was held in place with mattress sutures, and the traction sutures were removed. Subsequent experience with this operation showed that it was by no means always successful because:

1. unequal scar contraction between the two septal flaps sometimes led to a recurrence of the deviation
2. absorption of the autograft sometimes occurred leading to saddling of the supratip region
3. the lower end of the graft sometimes immobilized the membranous septum and gave it a rather peculiar and unnatural appearance.

In 1948, Fomon et al endeavoured to solve the first and third of these problems by the use of small autografts. The whole principle of septal removal, followed by septal replacement, has some inherent drawbacks and consequently the alternative solution of mobilization and repositioning of septal cartilage has been revived and further developed. This septoplasty concept, in particular, has been popularized by Cottle and his associates.

More recently, the permanent change in the shape of septal cartilage by morselization has been advocated by Rubin (1983). The deviated cartilage is crushed by a morselizer after the mucosal flaps have been elevated on both sides, and it is claimed that the new flattened shape of the cartilage is retained on a permanent basis.

The principles of septal surgery

From the experience over the last 90 years, it is evident that, from a surgical point of view, the septum can be divided into anterior and posterior segments by a vertical line drawn between the nasal processes of the frontal and maxillary bones. Deviations in the posterior segment can be easily and effectively treated by the classic Killian submucosal resection operation, whereas those in the anterior segments should be treated by a more conservative septoplasty technique.
Anaesthesia for septal operations

Septal surgery can be satisfactorily performed under either local or general anaesthesia. The high quality and ready availability of anaesthetists has resulted in a preference for general anaesthesia in the UK. A general anaesthetic is also invariably required for children and nervous adults. It is necessary to pack the nose about 15-20 minutes before the operation with 1.25 cm (0.5 inch) ribbon gauze which has been soaked in cocaine and adrenaline. This will greatly diminish the amount of bleeding at operation. The postural nerve block technique described by Curtiss (1952) is easily the best of the local anaesthetic methods. It was evolved from the earlier technique of Moffett (1941), but is much simpler and quicker, and is quite as effective. The patient is placed in the Proetz position with the chin and external auditory meatus in the same vertical plane. Then 2 mL of 4% cocaine solution are introduced into each nostril using a special angulated needle. The cocaine gravitates into the superior meatus where it blocks both the ethmoidal and sphenopalatine nerves. The patient is kept in this position for 10 minutes. A small quantity of 2% lignocaine is finally injected into the columnella. This method gives far better results than the older technique of using cocaine and adrenaline packs as it is often difficult to push these packs beyond the septal deviations.

Septoplasty

Septoplasty is an operation which should be performed under direct vision. To achieve this it is necessary to obtain adequate illumination. A satisfactory headlight is therefore essential and can also be supplemented by using an expanding nasal speculum fitted with a light carrier. Bleeding can obscure the operative field, and it is therefore very important to obtain maximum vasoconstriction of the mucosa before making the first incision.

This operation should not be a single standardized procedure, but should be tailored to the needs of the individual patient. For example, if the deviation is confined to the caudal border of the septum anteriorly, there is no need to touch the posterior part of the septum. There are however, certain general principles and these include:

(1) Incision.

(2) Exposure: the cartilaginous and bony septum is exposed by the complete elevation of a mucosal flap on one side only. Contact between the cartilaginous septum and the mucoperichondrial flap on the other side is maintained as much as possible because, in addition to ensuring the viability of cartilage, it also greatly reduces the risks of complications such as haematoma and abscess formation, perforation and over-riding of the different segments of the cartilages.

(3) Mobilization and straightening: the septal cartilage is then freed from all its attachments apart from the mucosal flap on the convex side. Many deviations are maintained by extrinsic forces such as the caudal dislocation of the cartilage from the vomerine groove. Mobilization alone will often correct this type of problem. When deviations are due to intrinsic causes, for example healed fractures, it is necessary to combine mobilization with some direct surgery on the cartilage such as a strip excision of the fracture line. Bony deviations are treated either by fracture and repositioning or by submucous resection of the deviation.
(4) Fixation: the septum is then maintained in its straightened position during the healing phase by sutures and splints.

**Incision**

The incision is best made at the lower border of the septal cartilage as was originally advocated by Freer. A unilateral (hemitransfixion) incision is adequate for a septoplasty and, for the right-handed surgeon, this is usually most conveniently made on the left side. The advantages of this incision have been tabulated by Bernstein (1973a) in the following fashion:

1. The incision is placed in a relatively avascular plane.
2. The mucosal edges here are both thick and tough, thus reducing the risk of tears. If tears do occur, a satisfactory repair is normally quite easily performed.
3. It provides easy access to the whole of the septum, including the caudal septal border, and the region of the anterior nasal spine with its associated premaxillary crest.
4. If the septoplasty is to be combined with a rhinoplasty, it is easy to extend the incision through to the opposite side and thus produce a full transfixion incision. It is important to make the incision as high as possible because a low incision through the membranous septum may be followed by a retraction of the columella. The first step is therefore to displace the columella downwards and to the opposite side by means of traction, exerted with dissecting forceps or a Cottle columellar clamp. The lower border of the septal cartilage will then be plainly visible and the incision made down to the perichondrium, which is incised and the subperichondrial flap elevation then commenced.

**Exposure**

It is usually best to expose the cartilaginous and bony septum by elevating the mucosal flap on the concave side. The difficulties of the flap elevation are partly a result of the anatomy of the various tissue layers and can often be greatly increased by fibrosis and scarring in these layers following previous trauma. The surgical anatomy of this region is of extreme importance and must be clearly understood if mucosal tears are to be avoided. It is easy to elevate the mucosal flaps across both the ethmoid-vomerine suture and the ethmoid-septal cartilage suture, because very few periosteal or perichondrial fibres pass into either of these suture lines. The difficulties of flap elevation occur mainly at the junction of the septal cartilage above, with the anterior nasal spine, premaxillary crest and vomer below. This is because the perichondrium encloses the cartilage in a complete envelope which does not fuse with the periosteum. The periosteum forms another inferior envelope over the adjacent bony septum and may result in a pseudo-joint capsule which can permit a side-to-side movement of the septal cartilage. The subperichondrial plane over the septal cartilage is therefore not continuous with the subperiosteal plane below and the difficulty in uniting these two planes can easily lead to tears. For this reason, most iatrogenic perforations occur along the chondro-vomerine suture, particularly anteriorly because the bony groove is widest here, and the problems are greatest.
As a general principle of flap elevation, it is usually best to leave the most difficult areas to last, since they can then be approached from several directions and under direct vision. A suitable technique for dealing with these problems has been evolved by Cottle et al (1958) who started the elevation over the septal cartilage and worked upwards and backwards always keeping above the chondro vomerine junction. This step in the operation was called the production of the 'anterior tunnel'. Once this had been accomplished, attention was then directed to the posterior end of the incision, and the periosteum over the anterior nasal spine was incised and then elevated backwards on both sides over the premaxillary crest, then the vomer, again keeping below the chondro vomerine suture. These were the so-called 'inferior tunnels. Finally, the most difficult elevation was performed which involved uniting the anterior and inferior tunnels under direct vision using a sharp dissector or knife. This is the so-called 'maxilla-premaxilla' approach of Cottle.

It is unusual for all the steps in this particular technique to be required, but in the really difficult demanding case, and particularly when performing revision surgery, it is the best method of choice.

**Mobilization and straightening**

The first step is to separate the lower border of the septal cartilage from its osseous base. In many cases, this lower border has been dislocated from its osseous groove and there is also a considerable amount of fibrosis which can greatly distort the anatomy. A sharp dissector knife is always required. The lower border of the septal cartilage is encased in a perichondrial envelope, and it is usually possible to continue the subperichondrial elevation downwards over the concave side of the septum, then around its lower border and upwards for a few millimetres in the convex side. When the cartilage has been freed, an attempt is made to reposition it back into the midline where it should rest in its osseous groove. Usually this is impossible because of the excess height of the septal cartilage and it is then necessary to remove a strip of cartilage about 3-4 mm wide from its lower border. This excised cartilage is part of the quadrilateral plate and may be up to 4 cm long. It can make an ideal autograft, should one be required at a later stage in the operation. It should therefore be kept in sterile saline during the rest of the operation in case the need arises. It is usually also necessary to straighten and lower the vomerine crest in order to make a suitable bed to accommodate the septal cartilage. The anterior nasal spine must not be removed. When deviated it can be fractured and repositioned in the midline. If the bony septum is deviated it is sometimes possible in the less severe cases to reposition it in the midline with a heavy elevator after preliminary fracture. However, this technique is inadequate for the angulated spurs which are often encountered at the junction between the ethmoid plate and vomer. Here a vertical incision is made through the septal cartilage, just behind the line joining the nasal process of the frontal and maxillary bone. The mucosal flap is then elevated off the cartilage and bone on the opposite side, and the deviated cartilage and bone may be removed back to the face of the sphenoid. When making this vertical incision through the cartilage, it is important not to make it too anteriorly, as otherwise the nasal dorsum will only be supported by a narrow strip of cartilage, and this can fracture superiorly and lead to a saddling deformity.

If the external nasal pyramid is twisted, it is important to separate the skin and subcutaneous tissue off the underlying upper lateral cartilages. This will allow the skin to be
draped easily over the straightened cartilaginous dorsum without the risk of cutaneous traction on the upper lateral cartilages producing a recurrence of the deviation.

This uncovering of the cartilaginous dorsum is easily performed through the classic intercartilaginous incisions. Usually a 15 blade Bard Parker knife is used to make the incision at the level of the internal nares. Anteriorly, each cartilaginous incision is united with a transfixion incision. A series of opening and closing movements with a pair of Knapp scissors will enable the elevation of the subcutaneous tissues off the cartilaginous dorsum to be easily effected.

The plane of dissection should be directly above the perichondrium as most of the blood vessels are found in the more superficial layers. The upper lateral cartilages are firmly united to the cartilaginous septum. There are often secondary changes in the upper lateral cartilages associated with a twisted nose. When this occurs it is necessary to separate the upper lateral cartilages from the septum and this is best done submucosally. By now the septal cartilage has been fully mobilized and, in the absence of intrinsic deviations, should be easily repositioned in the midline. A careful examination should be made at this stage and the mobility of the septum checked by moving it from side to side with a septal elevator. If there is any reduced mobility, its exact site should be noted and further trimming at this point may be necessary.

Other possible factors include a large turbinate, which will require treatment, the details of which are given later in this chapter.

At times, the mucosa on the narrow side of the nose is too short to allow the septum to return to the midline. This problem can be solved by cutting through the mucosa at the junction of the nasal floor and the septum. There will be a residual dehiscence on the floor of the nose after the septum has been repositioned into the midline, but this will re-epithelialize quite rapidly afterwards. Any residual obstruction is usually the result of intrinsic deviations in the septal cartilage. Old fractures in the cartilage often heal by fibrous union and this may result in severe angulations, which are best treated by the removal of a narrow strip of cartilage along the line of the deviation. This will break the spring of the cartilage which can then usually be repositioned into the midline.

Sometimes the septal lesion is so severe, either because of previous disease (for example, a septal haematoma), or where extremely radical surgery is necessary to correct the deviation, that too little supporting cartilage remains anteriorly to maintain the normal shape of the nasal pyramid and the columella. This is the type of problem that is not infrequently found in the professional boxer. In such a nose, the anterior residual septal cartilage can be supported with a free bone graft, taken preferably from the thinner part of the perpendicular plate of the ethmoid or, if this is impossible, from the rather thicker vomer. A suitable piece of bone can be obtained with a pair of heavy angled Fomon scissors and chisel. It can then be cut to the shape of the dorsocaudal septum, and two holes drilled in it to accommodate the fixation sutures. The bone graft is placed alongside the residual septal cartilage and sutured in position. Bernstein (1973b) has shown that bone is much more satisfactory when used in this support role than cartilage, which frequently become absorbed.
Fixation

At the end of the operation, the septum should be lying freely in the midline and if this objective has not been achieved, neither suturing nor splinting will prevent subsequent failure. If it has been necessary to make multiple incisions in the cartilage, over-riding of the segments can be a problem and this is best corrected by a Wright (1967) suture. Here, a through-and-through mattress suture is used, with one arm passing between the segments of the cartilage, and the other through all three layers of the septum. A figure-of-eight suture, immobilizing the lower border of the septum to the anterior nasal spine, is then inserted. Finally, the septo-collumellar incision is closed with a few sutures. Silastic splints are then inserted into the nose. The nose is packed with 1.25 cm (0.5 inch) ribbon gauze impregnated with bismuth iodoform paraffin paste (BIPP).

The classic submucous resection operation

The Killian incision is most commonly used for this operation. This is in an oblique incision about 5 mm above the caudal border of the septal cartilage. Elevation of the mucosal flap through this incision is usually easier than with the hemitransfixion incision, although this can also be used for this particular operation. The exposure stage is similar to that for septoplasty. Afterwards, an incision is made through the septal cartilage about 1 cm above and parallel to its lower border. The incision should be made through the cartilage, but not through the opposite perichondrium. The mucoperichondrium can then be elevated off the far side of the cartilage through this incision. A pair of angled scissors is introduced and used to cut through the septal cartilage in a direction which is parallel to and at least 1 cm posterior to the nasal dorsum. It is then possible to remove the obstructing cartilage and bone leaving these dorsal and caudal struts of cartilage to maintain the support of the nasal dorsum and columella. The cartilage is removed with Luc's forceps or a Ballenger's swivel knife. Any deviated bone in the region of the vertical plate of the ethmoid is then removed. The next step is to elevate the flaps off the maxillary crest and vomer. The periosteum covering this is not in the same plane of cleavage as the cartilaginous dissection. A separate breakthrough has to be made with a knife or dissector on to the bone to elevate the periosteum. The crest is finally removed with a hammer and gouge or with Jansen-Middleton bone forceps.

If the flap is torn, this does not matter unless there is another tear on the other side exactly opposite, when a septal perforation will inevitably result unless a satisfactory repair is effected. The site of the tear is first reinforced by the introduction of a small autograft of septal cartilage or bone between the flaps and the lacerations are then sutured. Some surgeons routinely replace septal cartilage and bone after performing the classic Killian technique. The almost universal use of central heating in North America tends to produce atrophic changes in the nasal mucosa and Briant (1977, personal communication) considers there is a very definite risk that this is increased after the loss of support of the septal cartilage and bone, and that septal perforations for this reason are by no means uncommon following a perfectly performed Killian-type operation. Therefore, he advocates that the excised cartilage and bone be straightened in a Cottle's crusher and then re-inserted between the flaps.
**Postoperative care**

Packs are removed after 24 hours and splints after 7 days. Antibiotics are not usually required.

**Septal surgery in the growing nose**

Since the turn of the century, it has been widely believed that the nasal septum plays an important role in the development of the facial skeleton and, in particular, the nose. For this reason most surgeons have avoided performing surgical operations on the growing septum for fear of producing some retardation of growth.

Some of the earliest work was undertaken by Hayton (1948), who made a careful study of 31 patients aged between 6 and 14 years who had been treated in Logan Turner's clinic in Edinburgh by the classic Killian operation. In 10 of the patients, there was some broadening of the nose, which was associated with a supratip depression.

Septal surgery performed during childhood carries with it the additional problem that it may interfere with the subsequent growth of the nose. Because of this risk, it was the usual practice to postpone all septal surgery until after the age of 16 years but, more recently, this view has been challenged by Cottle (1951), Jennes (1964), Huizing (1979), and others. Attempts have been made to elucidate this matter by animal experimentation (Ismail, 1964; Hartenstrom, 1970), and by observing the effects of injuries and operations performed during childhood.

Verwoerd, Urbanus and Nijdam (1979), Rhôys-Evans and Brain (1981) and Sarnat and Wexler (1961, 1967) have all shown that removal of cartilage in experimental animals interferes with the subsequent development both of the nose and of the maxilla. Brain and Rock (1983) performed a cephalometric study of 29 adult patients awaiting surgical treatment for injuries which had occurred during childhood, and demonstrated significant differences in nasal and orthodontic development compared with a control population.

A septal abscess occurring during childhood invariably leads to a saddle deformity, and Hayton (1948) showed that this also frequently follows the Killian submucous resection operation. In addition to the saddling of the dorsum, damage to the caudal septum can interfere with the development of the nasal tip. There is, therefore, universal agreement that no operation should be performed on the septum during childhood, which involves radical removal of cartilage. Any surgery performed at this age should be of a very conservative nature, and should be confined to the repositioning of the septum. It may be necessary to incise the cartilage in order to achieve this. Verwoerd considered it advisable to avoid actually performing the operation during either of the two nasal growth spurts. The results of these conservative operations are far from good. The experimental work on rabbits by Rhôys-Evans and Brain (1981) showed that a recurrence of the deviation often occurs and, in clinical practice, it is found that frequently up to 50% of these patients need revision procedures when they reach the age of 16 years.
Reduction of septal dislocation in the newborn

This should be undertaken as early as possible as it becomes increasingly difficult with the passage of time (Metzenbaum, 1936). The present author considers it to be impossible after about the age of 3 months, although Gray (1972) has had successes up to the age of 9 months. No anaesthetic is necessary, and both the instrumentation and technique have been developed by Gray, who inserts a special pair of neonatal nasal forceps in the nose. The middle of the palate is then firmly pressed downward for about 15-20 seconds to pull the septum straight. The septum is then manipulated into the midline.

The septum in rhinoplasty

It is necessary to consider the septum in rhinoplastic surgery because it is of great importance:

(1) in the deviated nose
(2) in reduction rhinoplasty
(3) as a support to the nasal dorsum
(4) as a source of graft material for augmentation
(5) there may be an unrelated septal problem requiring surgical treatment.

The septum is the central supporting strut of the nasal pyramid and it participates in every external nasal deviation. A look at a cross-section of the deviated nose shows that it is impossible to straighten the external pyramid by any combination of osteotomies without straightening the nasal septum, and it is for this reason that the most important skill required to correct a deviated nose is a capacity to deal with difficult septal problems, and this is why the author believes that this type of case is best treated by an otolaryngologist trained in rhinoplastic techniques.

The septum is also of importance in a reduction rhinoplasty. After the removal of a nasal hump, the nose, on cross section, resembles a truncated cone. It is then necessary to narrow the external pyramid, the result of which is that a modest septal deviation, which preoperatively was of no functional importance, will now produce obstruction and unfortunately, it is by no means uncommon for a patient with a functioning normal nose to have a cosmetic rhinoplasty resulting in iatrogenic obstruction.

Ulceration and perforation of the septum

These are usually different stages in the same pathological process and, apart from the traumatic cases, septal perforations are usually preceded by ulceration. The energetic and successful treatment of a septal ulcer will therefore prevent the development of perforation, and this is particularly important in the case of children, as the development of a perforated septum in the growing nose will often retard growth both of the nose and of the mid-third of the face.
**Causes of septal perforations**

(1) Trauma
   - (a) surgical
   - (b) repeated cautery
   - (c) digital trauma ('nose picking')

(2) Malignant disease
   - (a) malignant tumours
   - (b) malignant granuloma (Wegener's)

(3) Chronic infections
   - (a) syphilis
   - (b) tuberculosis

(4) Poisons
   - (a) industrial
   - (b) cocaine addicts

(5) idiopathic.

Apart from syphilis which normally attacks the bony septum, most perforations are found anteriorly, in the septal cartilage. Unfortunately, most are iatrogenic in origin and usually occur as a complication of septal surgery, particularly when the Killian submucous resection technique is used. Although the septoplasty procedure does not give complete immunity against this complication, perforations are a rarity following this operation. Perforations result from mucoperichondrial tears, particularly when they are bilateral and overlapping. Gross post-traumatic fibrosis increases the risk, and the site of the perforation is usually along the line of the chondrovestibular suture where the anatomy of perichondrial and periosteal layers also increases the difficulty of flap elevation. When mishaps of this kind occur during a submucous resection operation, every effort should be made to prevent this complication by inserting a bony or cartilaginous autograft between the torn flaps and also by closing the tears with catgut sutures.

Repeated cautery of the septum can lead to perforations. The risk is much greater when both sides of the septum are cauterized at one sitting, and it is therefore wise to have an interval of 3-4 weeks between the two treatments. In the author's experience, patients who suffer from Osler's disease are lucky to escape this complication.

Septal perforations are sometimes occupational in origin and the commonest such cause is penetration of the nasal mucosa by one of the hexavalent forms of chromium. In addition to its role in plating processes, this metal is used in certain tanning, dyeing and photographic processes. Workers engaged in the manufacture of dichromates are particularly at risk. Other causes include exposure to anhydrous sodium carbonate (soda ash), arsenic and its compounds, organic compounds of mercury, particularly mercury fulminate, alkaline dusts such as soap powders, hydrofluoric acid and fluorides, capsaicin, the pungent active principle of capsicum (chillies), vanadium, dimethyl sulphate, cocaine and other drugs taken as snuff, copper salts (rarely), and lime (rarely).

The incidence of chrome perforation among platers has been greatly reduced by the use of exhaust ventilation and seromists. The highest incidence of chrome perforation is found in chemical workers engaged in the production of chromates. In one such factory in the UK,
236 out of 480 workers had chrome perforations. There was no obvious relationship between the incidence of the perforation and the length of exposure. There are suspicions that at least some of these perforations were self-inflicted, because the monetary award for this occupational disease is often considered to far outweigh the resulting minimal disability.

**Symptoms and signs**

Apart from the traumatic causes, septal perforations are usually preceded by ulceration. There are often four well-marked stages, starting with redness and congestion of the mucosa producing irritation and rhinorrhea. Shortly afterwards the mucosa becomes blanched and anaemic; later it undergoes necrosis as revealed by the development of tough adherent crusts over the affected area. Finally, the crusting extends into the substance of the cartilage and a perforation results. Septal perforations are quite often asymptomatic, but the development of large crusts may cause obstruction and the separation of these crusts may lead to bleeding. Patients not infrequently complain of abnormal dryness in the nose, and sometimes of a dull discomfort over the bony dorsum. The passage of respiratory air often produces a whistling noise. Crusting problems are usually much worse when there is any interference with the normal respiratory air currents, as may occur with such obstructive lesions as septal deviations behind the perforations.

Brain (1980) has shown, in a series of 69 septal perforations, the 62.4% were completely free from any symptoms. The two main factors found to affect the function of the nose and produce symptoms, were the size and position of the perforation. The larger the perforation, the more likely it was to cause problems (*Table 10.1*). This relationship is of considerable importance, since the 1922 edition of Coate's text book actually advocates the surgical enlargement of perforations as a form of treatment. This operation can lead to intractable problems and should never be performed. The only symptom found more often in the case of small perforations was a 'whistling' noise during inspiration.

**Table 10.1 Symptoms in relation to size of perforation**

(1) Small - troublesome in 2 out of 20 (10%)
(2) Medium - troublesome in 11 out of 33 (33.3%)
(3) Large - troublesome in 13 out of 16 (81.25%)

The position of the perforation is the second major factor, and undoubtedly the more anterior the lesion the more likely it is to cause symptoms (*Table 10.2*). The state of the residual nasal mucosa is also of importance. Some cases of perforation are due to malignant disease or to a midline granuloma and may have been heavily irradiated, and this does often lead to a severe atrophic rhinitis which greatly increases the crusting problem.

**Table 10.2 Symptoms in relation to position of perforation**

Area 1 - present in 1 out of 1 (10%)
Area 2 - present in 4 out of 8 (50%)
Area 3 - present in 5 out of 42 (11.9%)
Area 4 and area 5 - all perforations present in these areas also extended anterior to involve other areas
Diagnosis

The history is of importance in the diagnosis of traumatic and occupational cases. When the edge of the lesion looks raised or hypertrophic, a biopsy should be performed to exclude malignancy. A biopsy is also essential in suspected cases of Wegener's granuloma. Serological tests for syphilis should always be performed if the lesion is involving the bony septum, and the erythrocyte sedimentation rate is invariably raised in cases of Wegener's granuloma; this can be a useful confirmatory diagnostic test for this condition, together with the biopsy.

Treatment

The first objective in the management of septal ulcers and perforations is to cure the causative disease process. Conditions such as malignant tumours, malignant granuloma, and chronic infections are discussed in other chapters. In the occupational cases, it will be necessary to obtain the cooperation of the industrial medical officer to prevent further exposure to the toxic agent. Most recent cases have occurred when the exhaust ventilation system in the chrome plant has become defective.

The second objective is to encourage natural healing of the lesion and, if this does not occur, to consider performing a surgical repair. The patient must be told to treat his nose with great care, and to avoid traumatizing actions such as vigorous blowing and nose picking. The patient should also apply some Cicatrin cream on the tip of the little finger to the lesion, twice daily. This treatment will heal most ulcers, although the original area will often permanently remain white, dry and scarred.

Perforations never heal spontaneously, but fortunately most do not cause symptoms, and therefore do not require any treatment. Crusting and bleeding are the main problems associated with the more troublesome minority. Less severe cases can be satisfactorily controlled by the use of a nasal douche (Collunarium alkalinus), but should this prove to be inadequate, the closure of the perforation, either by filling it with an obturator, or by means of a surgical operation, will have to be considered. Obturators are a simple, safe, and reliable method of closing almost any septal perforation. This method can be used to close large perforations, and the author has managed defects up to 4 cm in diameter by this technique.

Cooperation with a specialist in dental prosthetics is essential. The obturators are constructed from Silastic and are made from an impression of the perforation. The results of this form of treatment have been recorded by Brain (1980) for both medium (Table 10.3), and large (Table 10.4) perforations.

Table 10.3 Results of obturator closure: medium-sized perforations

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<tr>
<td>Good</td>
<td>50%</td>
<td></td>
</tr>
<tr>
<td>Fair</td>
<td>41.7%</td>
<td></td>
</tr>
<tr>
<td>Poor</td>
<td>8.3%</td>
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</tbody>
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The poor result was in 1 patient with severe atrophic rhinitis.
Table 10.4 Results of obturator closure: large perforations

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<tbody>
<tr>
<td>Good</td>
<td>50%</td>
</tr>
<tr>
<td>Fair</td>
<td>12.5%</td>
</tr>
<tr>
<td>Poor</td>
<td>37.5%</td>
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The worst results are obtained in patients with the largest perforations. None of the small perforations was troublesome enough to require closure. An obturator will reliably close almost any perforation, and eliminate the crusting which occurs around its edges. What it does not do is to replace the normal functioning septal mucosa. In the smaller and medium-sized perforations, the compensatory capacity of the residual mucosa is sufficient to overcome this deficiency, whereas this is not the case with nearly 40% of the larger perforations. This is a very unfortunate finding, because the difficulties in effectively closing a septal perforation surgically are directly proportional to the size of the perforation.

Another problem with surgery is the fact that, not infrequently, an unsuccessful operation actually enlarges the perforation and can make the patient considerably worse. It has been the author's policy to close all troublesome septal perforations with obturators, and only to consider surgical closure as a secondary treatment when the obturator fails to reduce dramatically the symptoms.

If the services of a specialist in dental prosthetics are not available it may be possible to close the perforation with prefabricated Silastic buttons, which have been popularized by Kern, Facer and McDonald (1977). The retention of this type of device is, however, far less satisfactory.

The operative closure of septal perforations is a rather unsatisfactory chapter in the subject of nasal surgery. It is extremely difficult to close surgically any perforation larger than 2 cm in diameter. Masing (1965) has shown that the partial anterior closure of larger perforations does frequently greatly reduce the symptoms.

These operations are technically difficult to perform, and it is therefore important to effect an adequate exposure of the perforation and this is not generally possible with the usual endonasal approach. When the upper limit of the perforation is 2 cm or less from the nasal floor, it is usually possible to obtain a reasonably good exposure by making an incision through the alar facial crease. Higher perforations are best reached through the external rhinoplasty approach, which has been popularized by Goodman and Strelzow (1978).

Perforations can be closed either by grafts or by flaps. Good results have been obtained by Fairbanks and Fairbanks (1973), using temporalis fascial grafts. An alternative graft is the three-layer composite graft from the pinna, using the technique popularized by Walter (1969). Free grafts taken from the turbinates have been employed to close small septal perforations (Ismail, 1964; Seiffert, 1967), but unfortunately the amount of tissue available is extremely limited, and it is impossible to close a defect which is 1 cm or more in diameter with this technique. These small perforations rarely need treatment and should this be necessary, they usually do well with obturators.
Flaps, on the whole, are a more reliable method of closing the septal defects. The mucosal flaps can be cut from the septum, the inner surface of the upper lip, or the lateral nasal wall. Septal mucosal flaps are rarely satisfactory as the mucosa around the perforation is usually thin and atrophic. Cartilage has often been extensively removed and the normal tissue planes have often been completely obliterated with fibrous tissue. The amount of the mucosa available for closure is inversely proportional to the diameter of the perforation. A buccal flap from the inner margin of the upper lip can be brought through a stab incision in the floor of the nose and used to close septal perforations.

The maximum width of the flap is, however, only 2 cm and this limits the size of perforation which can be treated by this technique, popularized by Tardy (1973). Masing (H., 1978, personal communication) has successfully used a vestibuloconchal flap from the lateral wall of the nasal cavity, which is elevated anteriorly and then swung medially to be sutured to the anterior edge of the septal perforation. Six weeks later the pedicle of the flap is divided and then rotated into the posterior part of the perforation, where it is sutured in place.

The enlarged turbinate

Many cases of nasal obstruction are the result of enlargement of the turbinates. There is almost always a compensatory structural hypertrophy of the inferior turbinate on the concave side of the septal deviation, and there are many patients complaining of nasal blockage with a fairly central septum, in which the obstruction is entirely due to turbinate hypertrophy. The inferior turbinate contains by far the greatest quantity of erectile tissue and this is the structure which is usually most severely involved.

Most of the bilateral cases are the result of some form of perennial rhinitis which may be either of the allergic or non-allergic type. Frequently, the problem is compounded by the development of a rhinitis medicamentosa due to self-treatment with topical decongestants.

Medical treatment

The first step in the management of these patients is to identify and treat the cause of the enlargement. Many cases are a consequence of perennial rhinitis and often medical treatment is extremely effective in controlling the discharge and sneezing, but is far less successful in relieving the blockage. Despite this, however, it is extremely important to treat the cause otherwise the results of surgical treatment of the condition are likely to be far from permanent.

The only medical treatment which appears to reduce the size of the turbinates is the administration of corticosteroids. The author has never achieved any success with drugs such as pseudoephedrine hydrochloride. The corticosteroids are best given by mouth and a 7-day course of prednisolone, starting with 35 mg on the first day and then reducing the dose by 5 mg on each successive day, is recommended. This treatment will frequently open up the nose and it is then possible to maintain the beneficial effect by the use of topical steroid sprays which are now able to adequately penetrate the nasal cavities. The injection of corticosteroids into the nasal mucosa is only mentioned to be condemned as, in the literature, there are at least three cases of blindness following these injections (Plate and Asboe, 1981).
Surgical treatment

Anatomically the whole turbinate is usually thickest in its middle third, although the bone is thicker anteriorly. Posteriorly the turbinate usually diverges away from the septum and in many cases where there is no mulberry hypertrophy of the posterior end, it may be possible to leave this segment.

Numerous surgical techniques have been described and advocated for the treatment of these patients, and they basically involve either altering the position of the inferior turbinate or reducing its bulk.

Changing the position of the inferior turbinate

(1) Lateral outfracture of the inferior turbinate.

(2) A submucous resection of the inferior turbinate. This can achieve a reduction in the bulk as well as change in position of the turbinate.

Reducing the bulk of the inferior turbinate

(1) Cautery
(2) Submucosal diathermy
(3) Cryosurgery
(4) The laser
(5) Submucous resection of the inferior turbinate
(6) Turbinectomy.

Lateral outfracture of the inferior turbinate can produce a modest but definite improvement in the airway, and its main indication is probably when dealing with the slight to moderate structural enlargement of the inferior turbinate, which is so often associated with the deviated septum. This minor procedure can then be usefully combined with the septal surgery.

Linear cautery of the inferior turbinate can produce a contracting scar which in turn will reduce the size of the turbinate. It is a fairly successful treatment for the slight to moderate cases of hypertrophy and is free from major complications. About 60% of patients obtain an improvement in their airway, although, of the successful cases, at least 10% develop some recurrence of the blockage which will require treatment at a later stage. David (1985, personal communication) has shown that it is necessary to protect the septum with a sheet of Silastic while the cautery is being performed to prevent adhesions which are likely to develop in 20-30% of all cases.

Submucosal diathermy also produces fibrosis and works in a similar fashion. This treatment has been popularized by Groves (1976) and has the advantage that the maximal effect is on the erectile tissue and not on the surface of the mucosa.

Some quite serious complications have been reported including devitalization of the bones which will then slowly separate as a sequestrum, leading to irritating crusting and
bleeding for a period of up to 2 or 3 months. Very severe nose bleeds have occasionally followed this procedure, usually in the form of a secondary haemorrhage.

Cryosurgery can produce necrosis of the turbinate mucosa thus reducing its size, and this treatment has been strongly advocated by Bicknell (1979). Secondary haemorrhage can, however, be a major complication of this operation and the present author has known of at least two cases requiring multiple transfusions and repeated packing.

More recently the argon laser has been developed as a form of treatment for the enlarged turbinate (Soldatov, 1985).

In the more severe cases, submucous resection of the inferior turbinates can be quite an effective procedure. Removal of the bone not only reduces the bulk of the organ, but also its skeletal support and the residual mucosa then falls away downwards and laterally, thus further opening the airway. Unfortunately, the inferior turbinate bone is very irregular and it is impossible to perform a blunt elevation of the overlying mucosa. It is vital, therefore, to use a sharp elevator, such as the Cottle knife, if one is to avoid tearing the mucosa into shreds and it is very important to try not to mobilize the bone before the mucosal elevation is complete, otherwise it becomes much more difficult. For this reason it is best to start with the elevation of the lateral flap, in case this occurs, as it is always easier to perform the medial elevation afterwards.

For the really severe case, turbinectomy is the only effective treatment. For many years it was widely taught that ozaena was a common and incurable complication of this operation. These fears appear to be groundless and Courtiss, Goldwyn and O'Brien (1978) were unable to find, in the world literature, one single documented case of ozaena following turbinectomy. The turbinate does have an extremely rich blood supply, especially posteriorly, and bleeding can be a problem, but this can often be reduced by applying a large pair of Birkett artery forceps over the base of the inferior turbinate for 30 seconds before cutting through the crushed tissue with a pair of turbinectomy scissors. The operation is facilitated by preliminary in-fracture of the turbinate.
Chapter 11: Surgical management of sinusitis

Valerie J. Lund

The majority of surgical procedures in the treatment of sinusitis were originally described in a pre-antibiotic era when rapid surgical intervention was often necessary to avert disaster. Today, although the operations remain the same, the indications for their use and the relative frequency with which they are required have altered. Acute sinusitis is now seldom a killer but chronic rhinosinusitis is common. Surgery for sinusitis aims to drain purulent secretions either by way of the natural ostium or more usually by the creation of an alternative drainage pathway which may be temporary or permanent. In so doing, complications are avoided and the sinus lining is given the opportunity to recover. When irreversible damage to the mucous membrane is deemed to have occurred, it may be removed completely and an attempt made to obliterate the sinus.

Embryological considerations (Schaeffer, 1920)

The maxillary sinus is first recognizable as a shallow groove expanding laterally from the infundibulum in the fourth intrauterine month. Extension laterally to reach the lateral cartilaginous plate is followed by absorption and expansion so that, at birth, the sinus has commenced invasion of the maxilla with its lower border about 4 mm above the nasal floor. Expansion and pneumatization continue until 8-9 years of age when the floors of the sinus and nasal cavity are roughly equal and the sinus 2 x 2 x 3 cm in dimension. Growth continues at the rate of 2-3 mm/year until the adult stage is reached when the sinus floor is usually lower than the nasal cavity (0.5-10 mm).

The ethmoids arise from preformed furrows between folds which develop on the lateral wall of the nose and are discernible in the fourth intrauterine month. The cells are primarily evaginations of nasal mucosa which grow into the lateral ethmoidal masses and, by further growth of the sacs and absorption of bone, become established as the cellular labyrinth which is well-pneumatized at birth. Up to the age of 6 years they grow slowly, but thereafter more quickly to reach their permanent shape by puberty.

The frontal sinus is absent at birth and only becomes obvious at 6-12 months of age. It develops:

(1) by direct extension of the whole frontal recess

(2) from one or more of the anterior group of ethmoidal cells arising in the frontal furrows associated with the frontal recess

(3) occasionally from the ventral extremity of the infundibulum ethmoidale by direct extension or from one of its cellular outgrowths.

Development often occurs from a combination of these origins but, in many instances, the frontal sinus is embryologically an anterior ethmoidal cell which has grown sufficiently far into the frontal region to be topographically a separate sinus. Extension into the horizontal portion of the frontal bone occurs first, followed by extension between the tables of the
vertical portion by simultaneous sinus growth and resorption of cancellous bone. This invasion
is inconstant resulting in variation in thickness of the posterior wall and, although dehiscence
in the bone of the walls are uncommon, the lamina papyracea and sinus floor are often thin.
The sinus is usually fully developed by the age of 15 years.

The sphenoid is recognizable from the third intrauterine month as an invagination of
the sphenoethmoidal recess. At birth it is 0.5 x 2 x 2 mm high and becomes fully aerated by
the age of eight years.

As a consequence of this differential development, young children are potentially
vulnerable to ethmoiditis, while later, the maxillary sinus becomes the most common primary
site of infection, and frontal sinusitis is rare before puberty.

The maxillary sinus

Applied anatomy

The average dimensions of the adult maxillary sinus are 25-35 mm (breadth), 36-45
mm (height), and 38-45 mm (length) (Eckert-Mobius, 1954), with the natural ostium opening
high on the lateral wall in the posterior infundibulum of the middle meatus.

Operations on the maxillary sinus must be examined in the light of certain anatomical
considerations. The inferior meatus lies under the inferior concha and extends downwards to
the floor of the nasal cavity. It is the largest of the three meatus and extends almost the entire
length of the lateral wall of the nose. It is deepest at the junction of its anterior and middle
thirds and the lower orifice of the nasolacrimal duct is found at this level (Gray's Anatomy,
1973). The inferior turbinate is attached in an arc with a maximum height range of 1.6-2.3
cm (mean 1.92 cm), 1.6 cm along the bony lateral wall. This constitutes the superior limit to
any surgery in the inferior meatus. The nasolacrimal duct opens at or just anterior to the most
cephalic portion of the inferior meatus, under the genu of the turbinate.

The thickness and quality of the bone alters within the meatus, with a gradual change
from compact to lamellar bone superior to inferiorly and anterior to posteriorly so that the
thinnest bone lies in the superior central portion of the meatus where it can be most easily
perforated. The change in bone thickness inferiorly makes it progressively more difficult to
cut down to the floor of the nose, while the thickness and decreasing meatal height preclude
anterior surgical extension. This, together with the constant position of the nasolacrimal duct
orifice, ensure that the duct opening is rarely damaged during surgery.

The blood supply of the inferior meatus derives primarily from the lateral
sphenopalatine artery, which may be regarded as the terminal part of the maxillary artery. It
passes through the sphenopalatine foramen into the cavity of the nose at the posterior part of
the superior meatus. Here it gives off its posterior lateral nasal branches which ramify over
the conchae and meatus, anastomosing with the ethmoidal vessels and the nasal branches of
A constant vessel arises from the lateral sphenopalatine artery, entering the inferior meatus
posteriorly, and running superior to inferiorly at between 4 and 5 cm along the bony lateral
wall. It then descends below the level of the palate, rising again very anteriorly on the lateral
wall. The surgeon's desire to avoid damage to this vessel together with changes in bone thickness and inferior meatal height limit the posterior extent of any surgical procedure.

The anterior superior alveolar nerve is derived from the infraorbital nerve and contributes to the superior dental plexus. The nerve is brought very constantly to the level of the anterior attachment of the inferior turbinate. At this point, branches pass inwards to the nasal chamber, inferior turbinate and meatus. A nasal branch which passes through a minute canal in the lateral wall of the inferior meatus supplies the mucous membrane of the anterior part of the lateral wall as high as the ostium of the maxillary sinus and the floor of the nasal cavity communicating with branches of the pterygopalatine ganglion (Gray's Anatomy, 1973). The distribution of this nerve demonstrates how changes in dental sensation can occur by direct trauma during inferior meatal surgery.

An explanation for paraesthesia following Caldwell-Luc procedures is readily apparent from an examination of the anterior surface of the maxilla. This displays a number of slight elevations overlying the roots of the upper teeth. Above and lateral to that of the canine tooth is the canine fossa in which the levator anguli oris is attached and which marks the natural point of entry into the maxillary sinus during a Caldwell-Luc procedure. Above the fossa is the infraorbital foramen, the anterior end of the infraorbital canal transmitting the infraorbital nerves and vessels.

The infraorbital nerves and vessels travel to the foramen initially in a sulcus which extends from the lateral portion of the orbital apex. Fifteen millimetres from the orbital margin, the infraorbital structures enter the canal which represents a constant thinning in the bone and which may be dehiscent. The orbital floor is thinnest along and medial to the sulcus, often 0.5-1 mm thick, affording a natural point of weakness between sinus and orbit (Lang, 1981).

**Historical review**

The first clear indication of the existence of the paranasal sinuses was provided by Berenger del Carpi, anatomist and surgeon at Bologna in the early sixteenth century (Wright, 1914). Fallopius (1600) referred to the maxillary sinus and suggested that the sinuses were absent in children until they reached maturity. Apart from the quotation: 'In a person having a painful spot in the head, with intense headaches, pus or fluid running from the nose removes the disease' (Hippocrates, 5th century BC), which may be inferred as describing sinus infections, the maxillary sinus and associated suppuration were not properly described until 1651 when they were reported by Nathaniel Highmore and for some time the sinus was referred to by this eponym.

The treatment of maxillary sinusitis by opening and irrigating the sinus via various routes has a long and varied history. Highmore himself advocated decompression by thrusting a silver bodkin through an empty tooth socket. Many of the earliest writers such as Cowper (1707) and Meibomius (1718) recommended irrigation through the alveolar tooth margin after moral tooth extraction, while Lamorier (1743) and Desault (1798) preferred the canine fossa approach. John Hunter (1835) was one of the first proponents of the intranasal approach and Zuckerkandl (1893) initially advocated perforation of the middle meatus, but later abandoned the technique because of the potential for orbital damage. The first description of the inferior
meatal antrostomy was probably by Gooch in 1770 (Cordes, 1905), but routine puncture of the inferior meatus was not common until advocated by Krause (1887), Mickulicz (1887) and Lichtwitz (1890). They used needle, trocar and stylette respectively. Thus began the first attempts at diagnosis by proof puncture followed by treatment by irrigation. In 1890 Lichtwitz invented the cannula which accompanied the perforating needle.

In his definitive paper on intranasal antrostomies Mikulicz (1887) described all the anatomical and physiological pitfalls of the operation. He realized that any perforation tended to close and felt that the cavity must be kept open to drain purulent secretions. Consequently, he recommended an antrostomy 5-10 mm in height, 20 mm in length and made flush with the floor of the nose.

Shortly after the introduction of the inferior meatal antrostomy, it was superceded by a more radical procedure described in 1893 by the American, Caldwell, in 1894 by Spicer from England and in 1897 by Luc from France, which differed from that advocated by Lamorier and Desault in that a counter-opening into the nose was included. Enthusiasm for the Caldwell-Luc procedure as the primary treatment of choice lasted throughout the early part of this century but, in the 1920s, a more conservative approach prevailed which meant that antral washout was succeeded by intranasal antrostomy via the inferior meatus and the Caldwell-Luc procedure only performed if that failed. This approach was supported by influential names such as Parker and Colledge (1921), Mackenzie (1927), St Clair Thomson (1926) and Negus (1958) and was facilitated by the advent of antibiotics.

An examination of the operative figures between 1950 and 1985 suggests that this trend is continuing despite a decline in the numbers of inferior meatal antrostomies from 1950 to mid-1960s. Since then the increase in antrostomies has occurred at a time when the Caldwell-Luc procedure has been decreasing in popularity and may be assumed to be at the expense of the more radical procedure. Furthermore, analysis of total numbers of operations suggests that this increase is genuine and not due to an overall increase in operating.

**Antral lavage**

**Indications**

Antral lavage has been used both in the diagnosis and treatment of sinusitis. Its diagnostic role has been to clarify radiological appearances by proof puncture. Therapeutically it is used in the treatment of acute and subacute maxillary sinusitis and pansinusitis which has failed to respond to conservative medication.

Antral puncture is usually performed through the inferior meatus. Sounding of the middle meatus to find the natural ostium is fraught with difficulties, not least the inability to find the ostium in 15-20% of cases (Van Alyea, 1936). The potential for orbital damage and the possibility of trauma and subsequent scarring of the ostium have precluded its use. Perforation of the anterior wall through the canine fossa for the introduction of a sinuscope represents a new facet to diagnosis as a more accurate form of 'proof puncture' (Chapter 3) and also offers a route for antral lavage.
Antral washouts may be performed at regular intervals, although opinion varies on the number and frequency of the puncture and on its efficacy, with some surgeons preferring to perform an inferior meatal antrostomy in the presence of pus and an obstructed ostium. An alternative method (Goode, 1970) is the insertion of an indwelling catheter through which daily irrigation can be performed until the quantity and quality of secretion improves.

### Table 11.1 Operations on the maxillary sinus and frontoethmoid region

#### Operations on the maxillary sinus

<table>
<thead>
<tr>
<th>Conservative</th>
<th>Radical</th>
</tr>
</thead>
<tbody>
<tr>
<td>antral washout</td>
<td>Caldwell-Luc</td>
</tr>
<tr>
<td>intranasal antrostomy</td>
<td>Denker</td>
</tr>
<tr>
<td></td>
<td>Canfield</td>
</tr>
<tr>
<td></td>
<td>obliteration - McNeill</td>
</tr>
</tbody>
</table>

#### Operations on the frontoethmoid region

<table>
<thead>
<tr>
<th>Conservative</th>
<th>Radical</th>
</tr>
</thead>
<tbody>
<tr>
<td>trephination of frontal</td>
<td>external frontoethmoidectomy</td>
</tr>
<tr>
<td>intranasal ethmoidectomy</td>
<td>Lynch-Howarth</td>
</tr>
<tr>
<td>transantral ethmoidectomy</td>
<td>Patterson</td>
</tr>
<tr>
<td>Jansen-Horgan</td>
<td>Killian</td>
</tr>
<tr>
<td></td>
<td>osteoplastic flap - Macbeth</td>
</tr>
<tr>
<td></td>
<td>flap with obliteration - Goodale and Montgomery</td>
</tr>
<tr>
<td></td>
<td>sinusectomy - Riedel.</td>
</tr>
</tbody>
</table>

### Contraindications

The proximity of the orbital floor and teeth in the small maxillary sinus of children under the age of 3 years makes antral puncture hazardous and is, therefore, rarely performed. Similarly in the underdeveloped maxilla with thick bony walls, puncture may be technically difficult. Antral puncture and lavage is inadvisable in febrile acute maxillary sinusitis untreated by antibiotics because of the risk of osteomyelitis and septicaemia. In the presence of trauma which may have disrupted the orbital floor, antral washout is contraindicated and, if drainage of haematoma is deemed necessary, inferior meatal antrostomy is safer.

### Anaesthesia

Antral washout can be performed under local or general anaesthesia.
Local anaesthesia

The nasal cavities are first sprayed with 10% cocaine and 1:1000 adrenaline solution and left for 3-4 minutes. This leads to shrinkage of the mucosa and facilitates insertion of cotton wool into the inferior meatus and drainage from the middle meatus through the natural ostium. Pledgets of cotton wool soaked in 10% cocaine and 1:1000 adrenaline solution can be placed along the inferior meatus and left for a further 4 minutes. Alternatively, 25% cocaine paste on silver wire wool carriers or Tumarkin wires can be placed in the nasal cavity, ideally one at the genu of the inferior turbinate and one approximated to the sphenopalatine ganglion at the posterior end of the middle meatus and turbinate.

Cocaine may cause adverse side-effects. Gastric absorption is more rapid than that from nasal mucosa so excessive cocaine trickling down the nasopharynx should be avoided. The maximal dose of cocaine for an adult is usually between 100 and 200 mg or up to 3 mg/kg. Malleable silver wires should be used in the nose, so that if the patient collapses this does not result in damage from wires being pushed through the cribriform plate.

General anaesthesia

This is rarely required for antral washout alone unless dealing with children or anxious adults. A cuffed oral endotracheal tube is employed and haemostasis and access facilitated by additional local anaesthetic vasoconstricting agents such as 25% cocaine paste, which can be spread with a cotton wool bud in the surgical field preferably with ECG monitoring. Moffat's solution can be instilled into the nose several minutes prior to surgery. This is composed of 4% cocaine solution, and 1% sodium bicarbonate in equal parts to 1/4 parts 1:1000 adrenaline.

Surgical technique

With the patient seated comfortably, the wool carriers or pledgets are removed and the inferior meatus visualized using a Thudicum speculum. A Tilley Lichtwitz trocar and cannula is used for puncture and it is advisable to check that the instruments match, engaging smoothly and with a sharp trocar end protruding 3 mm from the cannula. This is passed under the attachment of the inferior turbinate up to the genu where it will naturally come to rest. The instruments are held with the body of the trocar in the palm of the hand and the index finger running along its shaft so movement is controlled. Holding the patient's head steady, the trocar is directed towards the tragus of the homolateral ear.

Moderate pressure accompanied by a gentle boring action is usually sufficient to perforate the inferior meatal wall at its thinnest point. The trocar is advanced until it abuts the opposite antral wall and then withdrawn several millimetres. The trocar is then removed. The patient now leans forwards, holding a bowl beneath the chin to collect washings and is instructed to breathe through the mouth and to remark on any discomfort as lavage proceeds. The washout is performed using a Higginson syringe and sterile normal saline or water at 37°C. As fluid is flushed into the sinus the majority returns via the anterior nares, but any running posteriorly readily runs out of the mouth into the bowl. Washings can be inspected for the presence of mucus and sent for bacteriological and cytological examination. It may
be preferable to aspirate with an empty syringe before attachment of the Higginson apparatus to obtain an undiluted specimen.

If the procedure is performed under general anaesthesia, the patient may be placed in the tonsil position with a Boyle-Davis gag in place or with a head tilt of 15° and a throat pack. In either case, lavage is achieved with an ordinary hypodermic syringe containing 5-10 mL of fluid which is introduced and then aspirated to avoid unnecessary overflow into the nasopharynx. If the natural ostium is occluded, drainage may be facilitated by the introduction of a second cannula alongside the first. Excessive pressure should never be used. Care should also be taken not to introduce air during the procedure as fatal air embolus may result (MacNab Jones, 1976).

If a purulent washout is obtained, lavage should continue until it is clear. If the washout is initially clear, instillation should continue as mucoid material may require loosening. Following adequate lavage the cannula is withdrawn and the patient warned that fluid may continue to drain for the next few hours.

Complications

Mild haemorrhage may occur from the puncture site which can be stopped with 1 cm vaseline ribbon gauze packing for 12-24 hours during which time the patient must remain in hospital.

Incorrect positioning of the cannula should not occur if the technique described is followed. However, occasionally the anterior wall is breached leading to pain and swelling of the cheek. This is rapidly noticed in the conscious patient but, under general anaesthesia, requires observation and palpation. Similarly, perforation of the orbital floor leads to immediate pain. Under general anaesthesia, bulging of the orbital contents may be observed and for this reason the eyes are always left untaped and the upper lids gently lifted by an assistant. In the presence of a dehiscent infraorbital canal, even a correctly placed cannula can produce this complication. Excessive zeal on cannula introduction can also lead to perforation of the lateral or posterolateral wall, but is rare with controlled insertion. In all these circumstances, the procedure is abandoned and antibiotics given.

Inferior meatal antrostomy

Indications

This operation has been used in many forms of maxillary sinusitis, but is probably most successful in acute, recurrent or subacute sinusitis which has failed to respond to conservative medication, and usually one or two antral washouts. Although most commonly used in chronic sinusitis - by 78% of British otolaryngologists (Lund, 1986) - its success depends upon a return to normal of reversibly damaged mucous membrane by a combination of aeration and gravitational drainage. The degree of reversibility of the damaged mucosa will, therefore, determine the efficacy of the procedure.
Anaesthesia

Although the operation can be performed under local anaesthesia, general anaesthesia with cuffed oral endotracheal tube and pharyngeal pack is preferable. The use of a topical anaesthetic agent such as cocaine paste or Moffat's solution is useful in preparing the nasal mucosa prior to surgery.

Surgical technique

Many variations in technique are described and many alternative instruments may be used for each stage of the procedure.

The patient is prepared and towelled in a 15° head-up position and, under headlight illumination, the inferior turbinate is elevated with a Hill elevator. This instrument is then used to perforate the inferior meatus at the highest point under the genu of the turbinate. Enlargement is performed posteriorly with Grunwald nasal turbinate forceps, anteriorly with Seymour Jones antrum forceps and superiorly and inferiorly with a Hayek antrum punch forceps, either up- or down-cutting. The common use of the Ostrom forceps to cut anteriorly is not recommended as this instrument was designed for use in the middle meatus (Ostrom, 1913) and is vulnerable to breakage if used on the considerably thicker bone of the inferior meatus. Using an illuminated Killian speculum or headlight, the operation can be performed under direct vision.

Anatomical constraints limit the size of the antrostomy but, ideally, at least 2 cm x 1 cm windows should be fashioned to ensure long-term patency. Care should be taken to lower the inferior rim as much as possible to minimize the inevitable sump which results between floor of nasal cavity and that of the maxillary sinus. The edge of the antrostomy should be as smooth as possible with removal of free bone fragments and mucosal tags which predispose to infection and premature closure. Injudicious rasping of the edge is also not recommended. While discrete polyps can be removed via the antrostomy, blindly curetting with a Mackie curette is unwise. A sinuscope may be inserted via the antrostomy to inspect the antrum.

The use of rubber drains and packing into the antrum through the antrostomy is usually unnecessary, although a vaseline pack in the nasal cavity may be used for 24 hours. The inferior turbinate should be re-positioned at the end of surgery, although some surgeons prefer to trim a small anterior portion. The creation of a mucosal flap employing microsurgical techniques has been described (Reynolds and Brandow, 1975) but is of doubtful value. Suction cleaning and saline douching may be used postoperatively.

Complications

Examination of the anatomy of the inferior meatus readily demonstrates the potential complications of the operation. If the antrostomy is extended too far posteriorly the inferior meatal branch of the lateral sphenopalatine artery is encountered resulting in significant haemorrhage. Ordinarily haemorrhage from the antrostomy edge is not a problem.
Anterior extension may damage branches of the anterior superior alveolar nerve plexus leading to altered dental sensation, the incidence of which may have been underestimated in the past. Damage to the nasolacrimal duct orifice is fortunately rare due to its position and the quality of surrounding bone.

After initial fashioning all antrostomies undergo some circumferential closure due to healing, on average 0.4 cm. However, complete closure may be anticipated if an antrostomy is made 1 cm x 1 cm or less.

**Middle meatal antrostomy**

This operation has been advocated in the past (Lavelle and Spencer Harrison, 1969) but the potential for orbital damage and scarring of the natural ostium outweigh its possible physiological advantages. While the work of Hilding (1941) and Proetz (1941) demonstrated movement of ink particles by cilial streaming towards the natural ostium irrespective of an antrostomy, the situation with damaged cilia and thick tenacious mucopus is clearly different and there is no doubt that secretions do drain through patent inferior meatal antrostomies.

**Caldwell-Luc procedure**

**Indications**

The operation aims to remove the irreversibly damaged mucosal lining of the maxillary sinus and facilitate aeration and gravitational drainage via an inferior meatal antrostomy. It is, therefore, reserved for chronic maxillary sinusitis and is most usually performed after the failure of conservative medication, antral washout and intranasal antrostomy. The cavity becomes partially obliterated by fibrous tissue which may confuse subsequent interpretation of sinus X-rays (Noyek and Zizmor, 1976).

In addition to its role in the treatment of maxillary sinusitis, there are a number of other situations in which this procedure is of value:

1. removal of foreign bodies, such as a root of a molar or premolar tooth or dental amalgam
2. inspection and biopsy of antral malignancy
3. closure of oroantral fistula
4. surgery for dental cysts involving the antrum
5. as part of the approach to the pterygomaxillary fissure and sphenopalatine fossa
6. removal of recurrent antrochoanal polyps
7. elevation and stabilization of orbital floor fractures (McNab Jones, 1976).
Contraindications

It is rarely performed in children, as damage to the secondary dentition may result.

Anaesthesia

While the operation may be performed under local anaesthesia (using a maxillary nerve block) it is most commonly carried out under general anaesthesia with a cuffed oral endotracheal tube and pharyngeal pack. The use of a topical vasoconstrictor agent within the inferior meatus and injection of 1:200,000 adrenaline into the gingivolabial sulcus and soft tissues of the canine fossa is recommended.

Technique

The use of the headlight or illuminated speculum is usual with the patient positioned with 15° of head flexion. An incision is made down to bone in the gum margin, 3 mm above and parallel to the gingivolabial fold from the posterior edge of the lateral incisor to the first or second molar tooth (3-4 cm). It is advisable that the incision does not lie directly over the opening in the anterior face of the maxilla to lessen the risk of fistula.

The mucoperiosteal flap is then dissected superiorly with a periosteal elevator to expose the anterior wall of the sinus, taking care to avoid damage to the infraorbital nerve arising from the foramen just below the orbital rim. Retraction throughout the procedure should be as gentle as possible to avoid soft tissue swelling and paraesthesia.

The anterior wall is opened in the canine fossa where the bone is relatively thin. A 5 mm Jenkins gouge or drill may be used to make the initial entry and bone removed circumferentially with Hajek or Kerrison punch forceps to produce a circular opening approximately 1.5 cm in diameter. Inferior extension which may lead to damage of the teeth and their neural supply and lateral extension which may result in haemorrhage from the anterolateral terminal branches of the sphenopalatine artery should be avoided. Bleeding from the bony edge can be controlled by crushing the bone with punch forceps or diathermy.

The mucosa is then incised, the sinus inspected and the mucosa removed by careful elevation and dissection. Bleeding can be troublesome until all mucosa is removed and the success of the procedure depends upon complete exenteration of all diseased lining so that an uninfected fibrous membrane lines the sinus postoperatively. Particular attention should be paid to the inferolateral angle and roof which may be difficult to visualize.

A large inferior meatal antrostomy (2 x 1 cm) is fashioned as previously described, though the use of an inferiorly based mucosal flap is usually unnecessary. Packing the inferior meatus for 24 hours and even occasionally the antrum via the antrostomy is necessitated by significant haemorrhage though care must be taken on its removal to ensure no strands are caught on the bone of the edge of the antrostomy. Suturing of the buccal incision is recommended with absorbable suture material (to decrease the risk of fistula formation and obliteration of the labio-alveolar sulcus) but should be sufficiently loose to allow drainage of blood.
The patient should be advised against overenthusiastic blowing of the nose for at least a week and should replace upper dentures within 24 hours to avoid obliteration of the labio-alveolar sulcus.

Complications

Pain and soft tissue swelling are minimized by attention to surgical technique. Haemorrhage can occur from both the anterior bony wall or inferior meatal antrostomy but is usually controlled by packing.

Paraesthesia due to damage of the infraorbital nerve may be temporary or permanent but should be avoided by careful dissection and retraction. Damage to the teeth apices and their innervation may lead to alteration in dental sensation and very rarely devitalization and discoloration of the tooth.

Oroantral fistula may occasionally occur particularly if care is not taken with the siting of the incision. The fistula may be temporary or permanent requiring surgical intervention later. Osteomyelitis of the maxillary bone is more a theoretical than a real complication these days.

Modifications of the Caldwell-Luc operation

Denker's procedure (1906)

This allows access to the nasal cavity and maxillary sinus simultaneously by continuing the incision medially to the frenulum. It is in other respects very similar to the Caldwell-Luc operation.

Canfield's operation (1908)

An intranasal incision is made just behind the vestibule. The periosteum is elevated laterally over the edge of the pyriform aperture and into the canine fossa. This anterior angle of the maxillary sinus is chiselled off to expose the antral contents and then the opening is continued posteriorly into an intranasal antrostomy.

Maxillary sinus obliteration (McNeill, 1966)

The variable success of the intranasal antrostomy and Caldwell-Luc operations and the interest in obliterative procedures in the frontal sinus led McNeill to describe an operation of maxillary sinus obliteration based on experimental work in cats.

Through a sublabial approach, an inverted U-shaped flap is drilled in the bone of the anterior maxillary sinus. The mucous membrane of the sinus is removed completely and the exposed bony surface burred with a drill. The cavity is then filled with abdominal fat. Considerable success was claimed for the procedure, but it is rarely performed today.
The frontoethmoidosphenoidal complex

Applied anatomy

The skull contains only one ethmoid bone in the form of a cross which provides the central support for the anterior cranial fossa. The crista galli protrudes above the horizontal bar and the perpendicular plate below. At either end of the horizontal bar are the lateral air cell cavities or labyrinths. The whole bone is 2.5 x 2.5 x 2 cm in the adult, and is pyramidal in shape with the apex pointing anteriorly and the base abutting the sphenoid so that the posterior wall of the last ethmoidal cell forms the anterior face of the sphenoid (Ritter, 1982).

A plane of bone, the basal lamella, traverses the entire labyrinth, medially giving attachment to the middle turbinate and laterally abutting the lamina papyracea. The lamina papyracea forms the lateral wall of the air cells and part of the medial wall of the orbit and is extremely thin. Superiorly, the thin fovea ethmoidalis bone separates brain from air cells.

The middle turbinate lies inferomedial to the ethmoid mass. The anterior end is broad and often contains an air cell, the posterior end is narrow with the bone attached to the basal lamella. The middle turbinate and its basal lamella divide the ethmoid into anterior cells (2-8) draining mainly into the anterior infundibulum of the middle meatus and the posterior cells (1-7) draining into the superior meatus. Another important anatomical relationship, apparent in the coronal section, is that of the middle turbinate to the floor of the orbit which lies level with the midpoint of the turbinate.

In an anterior coronal section, the middle turbinate lies inferior and medial to the air cells but as one progresses posteriorly, this relationship changes so that the middle turbinate comes to lie more inferiorly and less medially with the result that the bone lateralizes as one progresses backwards. Because of the alteration in the relative position of the middle turbinate it may not always be a reliable landmark and it has been suggested (Ritter, 1978) that the bone of the middle meatus itself is preferable as staying medial to this ensures the safety of the lamina papyracea. It should be further realized that all ethmoidal cells except the most posterior lie medial to the medial wall of the maxillary sinus. While anterior contact between antrum and ethmoidal block is just a few millimetres, the larger posterior ethmoidal cells are in contact with more of the medial superior surface of the maxillary sinus.

On coronal sections it is evident that the lamina papyracea in the vertical plane is in line with the medial wall of the maxillary sinus. Of significant clinical relevance is the way in which the lamina papyracea curves 2-3 mm medially as it courses from the apex of the orbit anteriorly rendering the orbital contents more vulnerable than is appreciated. In addition, the solid bone of the greater wing of the sphenoid can be pneumatized by ethmoid cells.

Bearing in mind this anatomy, it becomes evident that it is impossible to exenterate all cells from below without jeopardizing the orbit, rendering both intranasal and transantral approaches intrinsically inadequate for complete ethmoidal exenteration. Pathologically, the thickness of the medial wall provides little anatomical barrier to infections of the ethmoid with the resultant clinical complications of orbital cellulitis, orbital abscess, optic neuritis and cavernous sinus thrombosis (Jarrett and Gutman, 1969; Schram, Curtin and Kennerdell, 1982).
The applied anatomy relevant to the external ethmoidectomy approach requires separate consideration. The subcutaneous tissues are very vascular and significant bleeding often occurs from branches of the supratrochlear artery and angular vessels. A definite single cut down to bone through the soft tissues will expose and facilitate clamping of these bleeding vessels.

The periosteum should be incised and elevated with care, particularly where it is adherent to the frontonasal and frontoethmoidal sutures to avoid damage to the trochlea. Similarly, when dissecting periosteum from the medial orbital wall, care should be taken as tearing the periosteum leads to troublesome prolapse of orbital fat into the surgical field.

The anterior and posterior ethmoidal vessels are encountered as periosteal dissection continues and provide important landmarks to the level of the cribiform plate and dura of the anterior cranial fossa. The posterior ethmoidal artery, in addition, lies 3-8 mm anterior to the optic nerve in its foramen. The rule of 24-12-6 may be applied to the medial wall of the orbit, representing respectively, the average distance in millimetres from the anterior lacrimal crest to the anterior ethmoidal foramen, from anterior to posterior ethmoidal foramen and from posterior ethmoidal foramen to optic canal (Rontal, Rontal and Guilford, 1979). The situation can, however, be very variable, with 16% of patients having no anterior ethmoidal foramen and 30% multiple ethmoidal foramina (Harrison, 1981).

Once the ethmoidal system is opened, it should be remembered that the middle turbinate, while not infallible, still represents an important landmark particularly to the cribiform plate; removal of the turbinate makes revision procedures particularly hazardous. It is, therefore, recommended that it is retained at least until the end of the operation. The close proximity of the cribiform plate makes it vulnerable to damage (both iatrogenic and pathological) leading to cerebrospinal fluid fistula. In addition, tubes of dura on fibres of the olfactory nerve running to the superior part of the lateral wall of the nose can be damaged leading to a similar complication.

The frontonasal duct is frequently damaged by disease and surgery in this region. To ensure patency and decrease the potential for subsequent mucocoele formation, it is advisable routinely to open into the frontal sinus and remove the medial part of the sinus floor and, in certain circumstances, to create an artificial frontonasal duct with Silastic tubing.

The frontal sinus is radiologically recognizable in 50% of subjects by the age of 6 years and fully developed by 15 years of age although 1-2% remain undeveloped (Schaeffer, 1920). Greatest variation in size occurs in the frontal sinus with a range of 28.95 ± 8.62 mm breadth, and 20.5 ± 7.74 mm in sagittal length, but maximum dimensions of 49 mm and 45.5 mm respectively have been reported in one study (Lang, 1981). The sinus is often divided into two parts, a transverse part which grows into the orbital roof and a vertical part which develops upwards within the squamous portion of the frontal bone and comes to lie in the front of the floor of the anterior cranial fossa. Separate pneumatization of the horizontal portion is often overlooked in operative procedures owing to its depth from the frontal region.

The sinus may be compartmentalized by incomplete bony septa, but a complete intersinus septum is usually present though often situated in a paramedian position.
Supernumerary sinuses are extremely common and normally drain independently by separate ostia communicating with the frontal region of the middle meatus. In addition, not infrequently, anterior ethmoidal cells encroach on the floor of the frontal sinus ballooning up into the lumen. They may arrange themselves in tiers and encroach upon the frontonasal duct. Finally, diverticula can bud off from the sinus at an early stage, remaining in communication with the parent sinus but again easily overlooked at surgery.

Considerable variation exists in the manner by which the sinus communicates with the nasal cavity and is directly related to the embryology. When it arises as a direct extension of the whole frontal recess, it may open by an ostium into the anterior part of the middle meatus. When the sinus arises from one of the furrows or from one of the cells of the infundibulum, a frontonasal duct is present. Consequently, the sinus may drain via the duct alone, separate from the infundibulum ethmoidale or partly by both routes. The length, diameter and tortuosity of the duct varies considerably and in those with a long serpentine course and encroached upon by a neighbouring ethmoidal cell, the slightest swelling of the mucosa will cause occlusion. In its terminal portion, other structures may impinge such as an asymmetrical nasal septum, enlargement of the uncinate process and ethmoidal bulla or a large middle turbinate.

Venous drainage is normally along the frontonasal duct mucoperiosteum to the veins of the nasal cavity but, in addition, veins from the frontal sinus, in the vicinity of the trochlear fossa pass through small bony foramina into the orbit, connecting with the superior ophthalmic venous system.

The relative thickness of the bony walls determines where empyemas of the sinus most frequently rupture. This tends to be the floor and lower anterior wall just above the medial canthus, although in large sinuses, rupture can occur high on the forehead.

The sphenoid, in addition to its main cavity, which in the adult measures $14 \times 14 \times 12$ mm, may also have diverticula. This may lead to pneumatization of both the greater and lesser wings and even of the pterygoid plates. As a consequence, a variable amount of pituitary fossa is exposed to the sinus lumen and in large sellar sinuses the internal carotid and adjacent nerves form obvious grooves with only a thin intervening layer of bone. even when the sinus is small, the superior wall is usually the thinnest (1 mm) rendering the optic chiasma vulnerable to penetration of the walls with surgical instruments. The posterior wall, although thin, can usually be directly visualized while the inferior wall is relatively thick.

The ostium is located one-half to one-third the distance up the face of the sphenoid, usually 2-5 mm from the dura and the same distance from the midline. The intersinus septum is always thin and often asymmetrically placed leading to unequal cavities.

**Historical review**

From the outset, operations on the frontoethmoidal complex fell into two groups (Macbeth, 1954): those designed to enhance drainage while preserving facial contour; and those aimed at eradicating irreversibly diseased mucosa disregarding cosmesis. As with the maxillary sinus, initial surgical treatment of infection concentrated on opening and draining the frontal sinus by trephination (Wells, 1870; Ogston, 1884). Ogston described a midline
vertical incision, using a trephine about 1 cm in diameter, followed by enlargement with a chisel, removal of the mucous membrane and insertion of a drainage tube into the nose. A more extreme procedure was suggested by Riedel in 1898 in which the inferior and anterior walls were removed leading to severe cosmetic deformity.

To overcome some of these problems, Killian (1903) proposed removal of the anterior and inferior walls but preservation of supraorbital bridge. Later he extended dissection to include the frontal process of the maxilla to facilitate drainage through the anterior ethmoidal cells and this procedure remained popular during the beginning of the twentieth century. An alternative approach was described by Lothrop (1915) who suggested circumventing the frontonasal duct on the diseased side by removing the intersinus septum, thereby converting the frontal sinus into one single cavity draining into the nose.

The Killian procedure was criticized by the advocates of 'external ethmoidectomy' as it left a 'dead space' (Howarth, 1921), the frontonasal duct region frequently closed and the ridge of bone often sequestrated (McNally and Stuart, 1954). Jansen (1902) had proposed removal of the frontal sinus floor, but with preservation of the anterior wall. His operation included exenteration of the adjoining ethmosphenoid system and was modified and popularized by Howarth (1921) in the UK and Lynch (1921) in the USA. Their operation which included removal of all frontal sinus mucous membrane, dealing with any intranasal obstruction and antral disease with the creation of an enlarged frontonasal drainage via an ethmoidectomy, dominated the first half of the twentieth century.

Mosher (1913) is credited with the first description of an intranasal ethmoidectomy, based on careful anatomical studies. Since then discussion has centred around preservation or removal of the middle turbinate. Skillern, following Mosher, removed the turbinate to enhance ventilation (1928), whereas Pratt (1925) advocated its preservation, a view which most practitioners have endorsed, although dissatisfaction with the procedure and its dangers have led to increasing use of the external approach (Eichel, 1979).

Smith (1934), Sewall (1935) and Negus (1947) added their support for the external approach, although some differences existed between the methods described. Both Lynch and Smith insisted on the importance of removing all mucous membrane, while Howarth and Negus suggested that mucosa should be preserved where possible. Lynch, Sewall and Smith advocated removal of all the sinus floor; Howarth initially supported this but on later review (1936) felt it was not essential, a view which Negus endorsed (1947). Lynch and Negus recommended the concomitant treatment of antral disease while little mention of this is made by Howarth, nor did he consider sphenoidal drainage, which was felt to be important by Lynch, Sewall and Smith. Finally, Lynch suggested that exenteration of the anterior ethmoidal cells would provide adequate frontonasal drainage; Sewall suggested a mucoperiosteal flap from the middle turbinate while Smith, Howarth and Negus preferred a split skin graft and rubber tubing as a stent.

There followed further attempts to preserve the frontonasal duct with exogenous material such as tantalum (Goodale, 1945) or acrylic obturators (Erich and New, 1947) with more recent interest in the mucoperiosteal flap (Ogura, Watson and Jurcina, 1960; Baron, Dedo and Henry, 1973). However, a high recurrence rate of problems led Boyden (1952) to conclude that the efforts to preserve the frontonasal duct were the chief cause of failure and
resulted in the emergence of the osteoplastic flap as the definitive operation for chronic frontal sinus problems.

Originally described by Schonborn (1894), Briegar (1895) and Winckler (1904), the operation was championed by Gibson and Walker (1951) and Macbeth (1954) in the UK and Bergera and Itoiz (1958) and Goodale and Montgomery (1958) in the USA. Macbeth believed that obliteration of the sinus, once it was stripped of its mucosa would occur naturally with fibrous tissue and bone as supported by the experimental work of Samoilenko (1913), Walsh (1943), and McNeill (1966). However, without complete removal of all the mucous membrane, obliteration might be incomplete and consequently oblitative procedures were devised using both exogenous and endogenous material. The use of exogenous material was largely unsuccessful (Schenck, 1975) and it was superceded by the use of endogenous substances such as abdominal fat (Goodale and Montgomery, 1958; Bergara and Itoiz, 1958). It is worth noting that Kuhnt described obliteration of the frontal sinus in 1895 and referred to Runge who performed a similar procedure 100 years earlier!

The decreasing incidence of chronic frontal sinusitis, the over-estimation of recurrence associated with the Lynch-Howarth technique and undoubted cosmetic problems of the osteoplastic flap together with the risk of infection both at the primary and donor fat site in oblitative procedure has led to a swing back to the Lynch-Howart operation in the UK, while the osteoplastic flap continues to be popular in North America (Rubin, Lund and Salmon, 1986).

**Intranasal ethmoidectomy**

**Indications**

The usual indication for this procedure is chronic sinusitis in association with nasal polyposis. It provides an inadequate approach for complete exenteration of the ethmoid complex but may be a preliminary procedure for more extensive external surgery. The operation aims to restore normal function while maintaining normal nasal anatomy by exteriorizing the ethmoid labyrinth into the superior nasal vault.

**Contraindications**

The procedure has become less popular, with fewer exponents familiar with the surgical anatomy which in turn leads to less usage. It is difficult to teach and is particularly hazardous in the presence of previous ethmoidal surgery. It should be very carefully considered when operating in the region of the only seeing eye.

**Surgical technique**

The operation is best performed under general anaesthesia with topical vasoconstrictors to achieve haemostasis. A reversed Trendelenburg position is preferred.

If the operation is being performed for nasal polyposis, it is better to attempt to clear the polyps intranasally to gain access. Similarly it may be necessary to out-fracture the inferior turbinate, and in-fracture and remove the first centimetre of the middle turbinate.
Using a long-bladed Killian speculum, the ethmoids may be entered just lateral to the anterior tip of the medial wall of the middle turbinate and the area cleared superiorly using a Tilley Henckel forceps, until the hard white bone of the fovea ethmoidalis is seen. The forceps must always be used with the opening blade directed medially, away from the lateral wall.

The position of the lamina papyracea may be determined by placing the forceps intranasally and a finger on the outer side of the lacrimal bone at the medial canthus of the eye. The ethmoidal labyrinth may then be exenterated, using the middle turbinate as the medial boundary and working with back-biting forceps towards the ascending process of the maxilla. The middle turbinate should be preserved as a landmark for as long as possible. Adrenaline-soaked ribbon gauze may be used during the procedure to maintain adequate haemostasis.

The surgeon should constantly watch for the appearance of yellow orbital fat in the surgical field and should consistently avoid directing any instrument laterally when 5 cm or greater from the inferior alar rim of the nasal vestibule to avoid damage to the optic nerve. The sphenoid may be entered by this approach. Displaced turbinates may be repositioned and the nose packed.

In summary the following rules should be applied to surgery in this area (Jafek, 1985):

1. Avoid the cribriform plate by not going superior to the medial canthal ligament
2. Avoid the orbit by not going lateral to the medial canthal ligament
3. Always curette downwards, anteriorly and medially
4. Examine all material removed for the presence of orbital fat
5. Operate only under direct vision.

Complications

Injury to the lamina papyracea may lead to haemorrhage. This in turn may lead to intraorbital but extraperiosteal bleeding with anterior tracking producing a periorbital haematoma (Harrison, 1981), erroneously considered by some as the hallmark of a successful operation. Posterior tracking of the haematoma leads to proptosis and visual loss, necessitating removal of nasal packing or exploration (Leopold, Kellman and Gould, 1980). Direct injury to the orbital periosteum may lead to fat prolapsing into the surgical cavity which can be packed with gelatin sponge.

Dural injury via the cribriform plate can lead to cerebrospinal fluid leak which, if noticed peroperatively, can be packed with fascia lata or muscle and Whitehead's varnish pack, treated with appropriate antibiotics and the patient nursed sitting up. If this is unsuccessful, it is possible to explore the area and use a septal mucosal flap to plug the dehiscence.

Despite precautions, blindness and meningitis are possible consequences of the operation and careful monitoring of the patient must be instituted postoperatively.
Transantral ethmoidectomy - Jansen Horgan procedure (1902, 1926)

Indications

The operation is used in combination with the Caldwell-Luc procedure for chronic inflammation and infection affecting the maxillary sinus and ethmoidal cells. It has also been used as a route for orbital decompression in 'malignant' exophthalmos.

Contraindications

The operation is only of use when complete exenteration of the ethmoids is not required.

Surgical technique

After performing a routine Caldwell-Luc approach, the posterior cells are opened through the antrum by pushing a closed Tilley Henckel forceps upward, medially and posteriorly at the upper and inner angle of the antrum, towards the opposite parietal eminence. If the natural ostium of the maxillary sinus can be located, the ethmoidal bulla lies immediately superior to it. The opening is enlarged with punch forceps and the posterior and middle cells exenterated employing the usual precautions with regard to the cribriform plate and orbit. The anterior wall of the sphenoid can be identified and opened. The anterior and agger nasi cells can only be reached safely if the operation is combined with an intranasal ethmoidectomy.

Complications

These are similar to those for an intranasal ethmoidectomy, that is haemorrhage and orbital trauma.

External frontoethmoidectomy

Indications

By combining a transorbital with a transnasal approach, this procedure offers access, illumination and perception of depth which obviates many of the disadvantages inherent in less 'open' operations. It aims to convert the ethmoid labyrinth into a single cavity and is useful in a number of circumstances:

(1) chronic infection unresponsive to conservative medication

(2) complications of acute ethmoiditis such as orbital cellulitis in which it is a useful approach for decompression and drainage

(3) recurrent polyposis, especially when previous intranasal surgery has been employed destroying useful landmarks
(4) in combination with the surgical exploration of the frontal sinus in mucocoele formation

(5) as a means of access in ethmoidal artery ligation for epistaxis, transtemoidal hypophysectomy, dacryocystorhinostomy, repair of cerebrospinal fluid leaks and decompression of malignant exophthalmos

(6) it has no place alone in the definitive oncological treatment of sinus malignancy except for occasional diagnostic purposes and has been superceded by the craniofacial operation in this area.

**Lynch-Howarth procedure**

The operation is best performed under general anaesthesia via an oral tube, with pharyngeal pack and the application of topical vasoconstrictors. The patient lies in the reversed Trendelenburg position, with 15° head flexion. A temporary tarsorrhaphy should be performed initially to protect the eye.

The incision is made slightly curved medial and concave towards the medial canthus of the eye, straight down to bone. The incision may be extended under the eyebrow to facilitate access to the frontal sinus. Bleeding from the angular vessels is often encountered. The periosteum is elevated with care to reveal the nasal process of the maxilla, frontal bone and medial wall of orbit. The lacrimal sac is elevated and displaced laterally.

Dissection continues posteriorly to reveal the anterior ethmoidal vessels which are ligated with sutures or neurosurgical clips or coagulated with bipolar diathermy. The posterior ethmoidal vessels approximate to the posterior limit of the ethmoidal cells and are also ligated. Dissection is aided by the use of 1 cm ribbon gauze soaked in topical adrenaline and retraction is best performed with a malleable copper probe.

The thin medial wall of the orbit is perforated with ease, exposing the ethmoidal cells which may be progressively exenterated under direct vision, bearing in mind the level of the cribriform plate. Thus the middle turbinate and middle meatus can be defined both intranasally and externally. It is important that exenteration continues up to and including the sphenoid and that all diseased mucous membrane is removed. Similarly it is important to open into the frontal sinus, the medial floor of which will be approached via the anterior ethmoidal cells. The amount of frontal sinus floor removed will depend on the access required and the extent of disease in the frontal sinus. The diseased lining mucosa should be removed completely, bearing in mind the variable anatomy of the sinus.

It is important to establish and maintain patency of the frontonasal region and to this end a fenestrated Silastic tube, 1 cm in diameter, is placed from the frontal sinus, through the ethmoidal region to open in the nasal cavity. The length of time that the tube must be left *in situ* has yet to be established but 3-5 months would seem reasonable and allows the establishment of a permanent patent channel. Attempts to reconstruct the frontonasal region by the use of mucoperiosteal flaps and split skin grafts are unnecessary, fraught with failure and obviated by this technique.
The periosteum, subcutaneous tissues and skin are sutured carefully with catgut and silk and a pressure dressing applied for 24 hours. The skin sutures can be removed at 4-5 days.

**Complications**

Problems may result from the incision itself, including oedema and infection, paraesthesia of the skin, damage to the medial palpebral ligament and webbing of the wound. This cosmetic deformity has been made much of in the American literature, but careful placement of the incision renders it a minor problem (Rubin, Lund and Salmon, 1986).

Haemorrhage can occur per- or postoperatively associated with retraction of the ethmoidal vessels before adequate haemostasis is achieved but it is usually self-limiting.

Dural exposure, either surgically or by the pathology itself is not uncommon, but any evidence of a cerebrospinal fluid leak should be treated with the appropriate antibiotic and closure of the defect primarily with fascia lata, tissue glue, Gelfoam or a septal mucosal flap.

Significant damage to the periorbita should be repaired immediately to avoid prolapse of orbital fat into the surgical field. Periorbital swelling is minimized with a pressure dressing and any resultant epiphora and diplopia are usually transient. Diplopia may also result if the globe is decompressed surgically after accommodating to long-term displacement. The eye should be washed with saline at the end of the procedure to remove blood, and Chloromycetin ointment instilled to prevent conjunctivitis.

Serious visual loss is unusual but can obviously result if the globe is injured and is a theoretical complication of sudden decompression after long-standing displacement, and may be treated with prophylactic use of steroids.

Failure to maintain the patency of the frontonasal duct may be associated with subsequent mucocoele formation (Schenck, 1975) and the original disease process may recur, which has led to the operation being supplanted by the osteoplastic flap in the USA.

**Transorbital ethmoidectomy**

**Patterson's operation (1939)**

The indications for this operation are similar to those for the Lynch-Howarth procedure but, in addition, it allows access to the orbital floor which is of use in orbital trauma or decompression and for transethmoidal hypophysectomy.

The incision, 2 cm long, is made in the natural crease line, one finger's breadth below the inferior orbital margin. The orbicularis muscle is split and the periosteum incised and elevated to the orbital margin. Once again, meticulous care is needed to avoid tearing the periosteum, and the origin of the inferior oblique muscle (except in 9% of patients where this muscle is intraperiosteal) is eventually revealed and, medially, the lacrimal sac (Harrison, 1981). As dissection advances, this gap opens to reveal the orbital floor and medial wall as far posteriorly as the posterior ethmoidal foraminae. Mobilization of the lacrimal sac provides
access to the anterior ethmoidal cells and removal of the lamina papyracea is performed under direct vision. This is continued until the compact bone of the sphenoid surrounding the optic canal is seen, when further removal of ethmoidal cells is carried out transnasally. Again it is important to open into the sphenoid sinus and the frontal sinus may also be entered through this approach though with less ease than with the Lynch-Howarth operation.

Complications are similar to those for the Lynch-Howarth procedure with recurrence of disease and transient epiphora associated with oedema of the orbicularis fibres being the commonest.

The frontal sinus

**Frontal sinus washout**

**Indications**

This procedure is performed when acute suppurative frontal sinusitis has failed to respond to antibiotics (oral and parenteral), decongestants and topical vasoconstrictors. Following the surgical principle of draining pus under pressure, it aims to avoid mucosal necrosis, osteomyelitis and intracranial complications. It should be done only after adequate antibiotic treatment has been instituted but should not be delayed if the situation is deteriorating or fails to resolve rapidly.

**Contraindications**

Radiological examination to establish the existence and extent of the frontal sinus should precede any surgery in this region.

**Surgical technique**

The operation is performed under general anaesthesia, in the reverse Trendelenburg position, with an oral tube and pharyngeal pack. A temporary tarsorrhaphy protects the eye. An incision is made 1 cm below the medial end of the eyebrow, straight down to bone. The sinus is usually entered with ease, using a small gouge and hammer or drill and the purulent contents released. The entry hole is enlarged with Citelli or Hayek punch forceps to allow adequate visualization of the sinus and a drainage tube is inserted and sutured in place.

In the presence of a pansinusitis, which is commonly the case, antral lavage may be indicated and, postoperatively, intravenous antibiotics should be continued in combination with regular frontal sinus washout. When the washout return is clear, and fluid begins to appear in the nose, suggesting restored functioning of the frontonasal duct, the drainage tube can be removed. Failure of normal drainage to be re-established may necessitate further surgical intervention such as external frontoethmoidectomy.
Complications

Careful placing of the incision avoids damage to the trochlea, supraorbital and supratrochlear nerves. Care should also be taken with anteriorly placed dura in sinuses which may have dehiscent bony walls.

Osteoplastic flap procedure

Indications

The procedure is primarily designed for conditions of chronic suppuration which have failed to respond to all other means of treatment. It has been used as an approach for osteomata, for the repair of trauma to this region, and in the treatment of frontoethmoidal mucocoeles.

Contraindications

If the ethmoids are extensively involved, it can be difficult to gain adequate access to this region via the osteoplastic flap approach alone.

Preoperative preparation and surgical technique

X-rays of the frontal region are taken to determine the extent of the sinuses and the pathological changes. From the X-ray, a template can be made to be used during the operation. Silastic sheeting may be used which can be sterilized preoperatively. Bacteriological cultures from the nose should be taken and the appropriate prophylactic antibiotics given with premedication.

The skin should be shaved 4 cm back from the hairline and prepared with aqueous Hibitane. If obliteration is to be performed, the abdomen must also be prepared. The operation is performed under general anaesthesia, in the reversed Trendelenburg position. The head should be positioned so that the plane of the forehead is horizontal. Temporary tarsorrhaphies are performed and infiltration with lignocaine and adrenaline is helpful.

A coronal incision is made, through the skin, subcutaneous tissue and frontalis muscle, behind the hairline, but taking care not to incise the periosteum, and the flap elevated inferiorly in the plane between the frontalis muscle and the periosteum down to the supraorbital rims and glabella. Neurosurgical clips are useful for haemostasis and the incision is extended inferiorly to a point just anterior to the root of the helix to provide wide exposure.

The sterilized template is then placed over the frontal sinuses, aligning the supraorbital rims exactly and the superior and lateral margins of the sinus are marked with methylene blue. The periosteum may then be incised along this line down to bone. It is advisable to elevate the periosteum for 2-3 mm on each side of the incision to facilitate closure.

Using a fissure burr or oscillating saw, a cut is made round the outline, cutting just inside the line to ensure the incision is within the limits of the sinus and bevelling it obliquely to prevent the bone falling in on the replacement. The entire margin is cut around, including
the supraorbital rims and glabella. It is often necessary to cut through the intersinus septum with a chisel to free the anterior wall which can then be prised down and forwards, so that the osteoplastic flap is hinged along the floor of the frontal sinus just posterior to the supraorbital rim.

All diseased tissue can be removed and the mucosa stripped completely. The bone should then be burred to remove all traces of mucous membrane and the last vestiges inverted into the frontonasal duct to obliterate it. If an attempt is to be made to obliterate the sinus, fat from the left lower quadrant of the anterior abdominal wall is removed and must be handled with care to avoid trauma. (The right side is avoided so that it cannot be mistaken for an appendicectomy scar at a later date.) The abdominal wound is closed in layers after achieving haemostasis. The fat is placed in the sinus cavity and should fill it completely.

The bony flap is replaced, and the periosteal layer repaired meticulously to avoid cosmetic deformity. The skin is sutured in two layers and a pressure dressing applied for 24 hours. An alternative incision can be made just superior to the eyebrows and connected across the glabella. This is suitable when the sinus is small, and on no account must the eyebrows be shaved. The preceding description applies to the operation for bilateral disease, but it is possible to open one sinus alone by cutting parallel and lateral to the intersinus septum, the position of which can be determined from radiography. It is, however, rarely employed.

**Complications**

Cosmetic problems associated with the incision and repair are not uncommon and haematoma collection under the flap may occur. Frontal bossing, bony depression and nasal skin necrosis have been reported (Sessions et al, 1972; Ward and Bauknight, 1973; Schenck, 1975; Hardy and Montgomery, 1976) and may necessitate subsequent cranioplasty and bone grafting. In middle-aged men with receding hairlines even the best scar may become obvious with time. Osteomyelitis can develop in the frontal bone flap and both the primary and donor site wounds can be infected. The dura may be torn if the template is incorrectly drawn or copied leading to a cerebrospinal fluid leak and, as with all operations for sinusitis, the possibility of recurrence of the original pathology has been reported as high as 25% (Schenck, 1975).

**Radical frontal sinus procedures**

Fortunately there is now little place for the more radical frontal sinus operations. The removal of the anterior sinus wall and floor in sinusectomy as described by Riedel (1898) leaves considerable disfigurement, particularly when the sinuses are large and, although attempts have been made to correct this with acrylic implants (Ritter, 1978; Barton, 1980), these are often unsuccessful due to rejection in the presence of continued infection.

**Sphenoid sinus operations**

The sphenoid sinus is rarely infected alone. As part of a pansinusitis it can be drained intranasally, directly or via an intranasal ethmoidectomy. Direct cannulation of the sphenoid ostium can be done with a sphenoid cannula (10 cm length) introduced along the nasal septum towards the end of the middle turbinate, making approximately a 30° angle with the floor of
the nose. By gentle manipulation it may be possible to enter the ostium and irrigate carefully with warm saline. More usually the sphenoid is opened during an external frontoethmoidectomy. The most frequent requirement for sphenoid sinus exposure is in the approaches to hypophysectomy (Chapter 21).

**Choice of treatment in sinusitis**

A wide range of procedures is available in the treatment of sinusitis and the choice of operation must be tailored to the requirements of the individual patient and the experience of the surgeon. In acute sinusitis when conservative measures fail, the surgical principle of drainage of pus is implemented to achieve rapid resolution and avoid serious complications. It is when the condition is recurrent or persistent that the selection of a surgical approach is more difficult. The multiplicity of these approaches indicates the intrinsic inadequacies inherent in the treatment of chronic sinusitis and while the progression through increasingly more radical procedures is not to be deprecated, it may be that our failure to deal adequately with chronic sinusitis is due to the use of operations which depend on a return to normal of irreversibly damage mucosa.

In the case of maxillary sinusitis, after an adequate course of broad-spectrum antibiotics and decongestants, antral lavage is often effective. How often this should be repeated before proceeding to further surgery is debatable and there has been a move towards earlier surgical intervention as compared with the once traditional 'weekly washout'. To establish better drainage other procedures such as submucous resection of the nasal septum and removal of nasal polyps, must also be considered.

The purpose of the inferior meatal antrostomy is presumed to be a combination of aeration and gravitational drainage which facilitates a return to normal or reversibly damaged mucous membrane. The operation should, therefore, be implemented at an early stage. To achieve long-term patency of the antrostomy, it is necessary to aim at a 2 x 1 cm window in an adult with careful attention to technique, but it is difficult to evaluate the clinical effectiveness of the procedure as many patients undergo cyclical improvement and deterioration of their symptoms without any appreciable alteration in antrostomy size. Indeed, it may not matter if the antrostomy does close.

The problem lies in the inability to quantify accurately the degree of mucosal damage and, therefore, the decision to implement a Caldwell-Luc operation is often an arbitrary one, usually based on clinical failure of the antrostomy.

In infections of the ethmoid and sphenoid sinuses, cannulation and lavage are not recommended. In frontal sinusitis, there is a place for external trephination but instrumentation of the frontonasal duct is best avoided as it commonly produces stenosis. Concomitant involvement of the antrum often compounds these situations and must be treated at the same time.

The intranasal and transantral approaches to the ethmoids combine an inability to perform total exenteration of the cells with significant potential hazard and require considerable surgical expertise. As fewer of these operations are performed, this expertise is increasingly difficult to obtain. In 1969, Davison reported on 100 consecutive cases and
attributed his improved results to his greater familiarity with the anatomy, a view which has been supported by Freedman and Kern (1979). In opening the ethmoids sufficiently to relieve obstruction, and allowing drainage and ventilation, it may not be necessary to remove every vestige of diseased mucosa and exenterate every cell. In aiming merely to decrease the incidence of recurrence rather than eliminate it, these operations have a role especially in polypoid disease of the antroethmoidal complex. It is, therefore, important to preserve the middle turbinate as a future landmark in these circumstances. However, there are many surgeons today who would agree with Mosher’s remark (1929) that 'intranasal ethmoidectomy is the blindest and most dangerous in all surgery'.

The external frontoethmoidectomy offers excellent access and minimal postoperative complications. Although it also requires a thorough understanding of the anatomy, it is the treatment of choice in any established condition. The success of the procedure is dependent upon adequate removal of the frontal sinus floor, removal of the entire mucosal lining, complete exenteration of the ethmoid cells and opening into the sphenoid, and the establishment of a large permanent drainage channel into the nasal cavity.

The need for radical frontal sinus surgery is decreasing and while the osteoplastic flap still has a role for the most intrasigent disease, it is rarely implemented and offers few advantages over a radical external frontoethmoidectomy.

**Sinusitis in children**

Because of the development of the sinuses, sinusitis is unlikely to be a clinical problem before 2-3 years of age. Acute infections, particularly of the ethmoids, may result from any upper respiratory tract infection, chronic sinusitis may accompany recurrent adenotonsillitis. Cleft palate deformities, choanal atresia, foreign bodies, tumours and allergy may all predispose to infection, although the teeth are rarely a source of problems, as the deciduous teeth are separated by the buds of the permanent dentition.

Interpretation of X-rays can be difficult due to the size of the sinuses and symptoms of nasal obstruction, mucopurulent discharge and cough may not be directly related to sinusitis. However, antral puncture may be indicated after failure of medical treatment, usually in conjunction with adenoidecetomy and tonsillectomy and always under general anaesthesia.

Cannulation and daily washouts are not tolerated well by young children and it is occasionally necessary to resort to an inferior meatal antrostomy. Anatomical restrictions and continuing bone growth result in early closure of the holes. Care must be exercised to avoid damage to the teeth roots and for this reason the Caldwell-Luc operation is not recommended. While decompression of orbital cellulitis resulting from acute ethmoiditis is sometimes necessary, surgical intervention in the frontoethmoidal region is rarely indicated.
Chapter 12: The complications of sinusitis

B. H. Pickard

In most cases sinusitis is uncomplicated and the effects are contained within the sinus cavity. It is the purpose of this chapter to draw attention to the effects of the spread of infection beyond the sinus wall and thus to influence the clinician first, by guiding his treatment in order to prevent the subsequent spread of disease, and second, by making him aware of the sinus origin of these complications when he sees them.

First are the systemic effects of sinusitis: pain, malaise, anxiety and pyrexia. Second are the effects on the nose of an infected discharge from an open sinusitis. Third, in a closed sinusitis, the disease may extend beyond the normal confines of the sinus, forward into the face, laterally into the orbit, upward and backward into the cranial cavity.

The type and activity of the infection will dictate whether an acute spreading infection such as orbital cellulitis or meningitis and encephalitis occurs or whether an abscess forms within the orbit or cranium, extradurally, subdurally or within the brain. Finally, infection may spread within the skull bones to produce an osteomyelitis.

Anatomy

The paranasal air sinuses consist of an ordered group of cavities within the bones of the skull. They are approximately symmetrical although the frontal sinuses in most skulls differ on the two sides and the sphenoid is seldom symmetrical. These discrete sinuses, in many places separated from each other by a thin sheet of bone, form a continuous system from the frontal sinus in the forehead through the ethmoid to the sphenoid below the sella turcica. The lower margin of the ethmoid labyrinth abuts on the maxillary sinus below.

The maxillary sinus (antrum of Highmore) is the largest and contains up to 15 mL of air in the adult. Mostly in the maxilla, it extends laterally into the body of the zygoma. It is shaped like a triangular pyramid with the base upward to form the orbital floor which is incomplete at its central thin part where it is traversed by the groove and canal of the infraorbital nerve. The medial wall is shared with the nasal cavity and is penetrated by one or two openings joining the sinus to the hiatus semilunaris in the middle meatus of the nose.

The anterior wall underlies the cheek and is thinnest at the canine fossa above which is the foramen through which the infraorbital nerve emerges. The posterolateral wall is attached to the pterygoid process laterally, forms the anterior wall of the infratemporal fossa and is closely related to the maxillary artery in its third part before it gives rise to the sphenopalatine and greater palatine branches which supply the posterior part of the nasal cavity.

The ethmoid sinuses, varying in number from three to 20 on each side, are closely packed together into the space between the medial wall of the orbit and the upper part of the nasal cavity. Above, they reach higher than the cribriform plate extending into the frontal bone each side of the ethmoid notch of the frontal bone. Below and laterally the lower cells are in contact with the upper medial recess of the maxillary sinus. Below and medially the
labyrinth is related to the middle meatus and middle turbinate which may contain a separate sinus.

The anterior and middle groups drain into the middle meatus and the posterior group into the superior meatus. It is not easy to define and distinguish these groups.

The most posterior sinus is the sphenoid which lies behind the most posterior ethmoid, completing the sequence posteriorly. If small, the sinus is situated in front of the sella in contact with that of the opposite side. If large on one or both sides, the cavity is in close proximity to the whole outer surface of the sella turcica which appears as a hemispherical prominence protruding into the cavity. The structures closely related to the sinus are the hypophysis (pituitary), the optic nerves and chiasma, the internal carotid arteries and the cavernous sinuses. Each sinus has a single opening into the sphenoethmoid recess.

A fully developed frontal sinus is L-shaped in sagittal section and shows a vertical part extending into the forehead with a scalloped upper and lateral margin. The horizontal part spreads into the anterior and medial part of the roof of the orbit.

In some skulls the horizontal part is absent or replaced by an anterior ethmoid sinus. Throughout, the frontal sinus is closely related to the anterior cranial fossa being separated by the inner table, or dural plate, of frontal bone. Each sinus communicates with the anterior end of the hiatus semilunaris by the frontonasal duct. The frontal sinus is not present at birth but can be recognized from the age of 5 years. Commonly asymmetrical the sinus is sometimes absent on one or both sides.

**General effects of sinusitis**

**Pain**

In acute closed sinusitis the pain may be severe and the nasal effects minimal. The pain is felt at the sinus or referred to other areas supplied by the trigeminal nerve. Frontal sinus pain may be felt in the forehead, the orbit or as headache.

Maxillary sinus pain is felt in the situation of the sinus, in the upper teeth or above and lateral to the sinus. Although facial swelling may occur with maxillary sinusitis, it is uncommon.

Facial pain is more widespread in acute pansinusitis. In chronic closed sinusitis the pain is much less and extension of disease may present signs before the sinusitis is suspected.

**Systemic effects**

The acute inflammation of 200 cm² of vascular mucous membrane by active organisms will allow sufficient toxic absorption to give general effects of prostration, malaise, pyrexia and tachycardia.
Nasal effects of sinusitis

Although the nose has been infected at the start of an acute sinusitis, in a closed sinusitis the nasal mucous membrane may return to normal giving neither signs nor symptoms of sinusitis. On the other hand, in an open sinusitis the escape of discharge into the nose will perpetuate a rhinitis. This rhinitis may be simple, hypertrophic or at worst atrophic.

The atrophic rhinitis complicating chronic sinusitis may present a difficult clinical problem if surgery is required to control the sinusitis since any surgery is likely to aggravate the atrophic changes and increase the crusting.

Mucocoele

In the course of an acute infection, chronic infection or as the result of injury, the opening of one sinus may be permanently stenosed. As a result, even after subsidence of the infection the pressure in the sinus cavity will increase as a result of continued secretion. A gradual expansion will result.

The stenosis of the ostium is most likely to occur in the frontal sinus where the narrow duct, approximately 2 mm wide, passes for a distance of 5-10 mm through the ethmoid bone in close relationship to the anterior ethmoid sinuses.

As the sinus expands with pressure, erosion of the bony walls occurs and the thinnest section - the orbital wall of the horizontal part - will most often (90%) give way first and the mucocoele will continue to expand into the orbit. If the cavity remains uninfected it may enlarge slowly without pain and considerable displacement of the eye may occur before the condition is recognized.

The differential diagnosis lies between a sinus enlargement and other causes of ocular displacement such as dys Hormonal exophthalmos, orbital tumours and pseudotumours. Treatment is by immediate surgical exploration and the restoration of a permanent passage into the middle meatus of the nose.

When the displaced bone has been removed it is usual for the orbital contents and displaced periosteum to fall back into their correct position, but if displacement has been prolonged and extreme, the restoration may be incomplete.

In one in 10 of the frontal sinus mucocoeles the anterior wall will erode first and the swelling appear on the forehead. In a very small number expansion will occur through the posterior wall or dural plate, and subsequent expansion will invade the anterior cranial fossa. The picture may elude detection if the only signs are those of frontal lobe compression.

In children, a mucocoele from an ethmoid sinus is more common and will expand into the orbit as the thin lateral wall of the sinus (lamina papyracea) is expanded. The picture is very similar to that of frontal sinus expansion but with less downward displacement of the eye. In a young person the return of the displaced orbital soft tissue is more rapid.
If the orbital contents do not return sufficiently closely to their correct position, not only is the result unsightly but diplopia may persist. It may be necessary to correct the non-alignment by surgery to re-site the anterior attachment of the extraocular muscles.

**Expansion of the maxillary sinus**

The anatomy is such that sufficient obstruction of the ostia to permit expansion of the maxillary sinus by retained secretions is rare, but it is sometimes found in caseating sinusitis, cholesteatoma or as a result of facial fractures.

The expansion will occur by erosion or displacement of the two thin areas of the sinus wall, namely the canine fossa and the floor of the orbit through or close to the canal and groove of the infraorbital nerve. If the displacement is in the canine fossa, there is a gradual filling of the lower hollow part of the cheek with little or no pain. The swelling may develop over a period of months or years and be unnoticed by the patient. An upward expansion will present as a protrusion of the orbital contents, swelling of the lower eyelid and an upward and forward displacement of the eye.

Occasionally, simple drainage of the mucocoele by intranasal antrostomy may be sufficient. If there has been much displacement of bone a Caldwell-Luc operation will allow access to this displaced bone in order to restore the bony contours of the sinus to their normal position. If the upward bony displacement is too great to be replaced from below, a skin incision must be made along the inferior margin of the orbit. From this exposure the orbital periosteum is elevated. If possible all the bone is pressed down into its normal position but some may have to be removed. Care must be taken to identify and preserve the infraorbital nerve.

If the sinus has been expanded by cholesteatoma or caseating sinusitis the removal of all the lining is necessary to reduce the risk of recurrence.

**Spread of infection**

**Pharyngeal and laryngeal complications of sinusitis**

In open or discharging sinusitis the discharge is carried backward in the nasal cavity and descends through the nasopharynx into the pharynx. The mucous membrane will become infected and show a simple pharyngitis which is unresponsive to treatment. Spread of infection through the lining to invade the subepithelial lymphoid tissue will produce a granular pharyngitis with visible nodules as the lymphoid masses enlarge. Alternatively a dry atrophic pharyngitis may result.

In younger persons a chronic or recurring tonsillitis may be the first evidence of sinusitis. Further downward spread will give laryngitis or tracheitis and bronchitis but the relationship of sinusitis to bronchiectasis is not cause and effect but related underlying pathology.
**Dental complications**

The roots of the upper teeth, particularly the second premolar and first molar may lie in close proximity to a well-formed maxillary sinus. The tooth roots which project into the antrum are then covered by a thin layer of bone resembling the fingers of a glove. Extraction of one such tooth, even with the greatest care and after recognition of the special anatomical problems may result in an oroantral fistula, which can be recognized by the passage of air if the patient attempts a Valsalva manoeuvre which, on this occasion, means an attempt to exhale against the pinched nostrils with the mouth open.

The tooth socket should immediately be cleared of any tooth or bone fragments and a dental plate provided which will protect against the access of food. If the maxillary sinus is infected, either before the extraction or as a result of access of food and organisms via the fistula there is a risk of persistence of the fistula.

After 4-6 weeks of failure to heal, a surgical repair should be attempted. The sinus mucosa, which by now will have prolapsed into the fistula to form a polyp, should be excised along with the lining of the opening. A flap of buccal mucosa with the underlying periosteum can be raised, and with both ends attached, the centre is advanced medially and sutured across the hole. The only unfortunate result is a local loss of buccal sulcus which may make denture fitting difficult.

The alternative is a pedicle flap of palatal mucosa and periosteum, attached posteriorly to include the greater palatine artery. The bony edge of the fistula is reduced in order that the flap may lie without kinking. The free anterior end of the flap is turned laterally across the hole and sutured in place.

After repair the site should again be immediately protected by a dental plate.

**Local spread of infection**

**Swelling of the face**

Cellulitis may arise from frontal, ethmoid or maxillary sinusitis as the infection spreads in the subcutaneous tissues. There is mild pyrexia and some pain but the swelling may be localized or widespread over one half of the face, reaching from the mandible to the hairline and back to the ear.

Although the diagnosis of cellulitis may be clear, it is often difficult to establish the site of origin of the infection as a similar picture is given by spread from the teeth, skin pustule, lymph nodes, salivary glands or the outer ear.

The diagnosis of sinusitis as the cause is obscured by the overlying soft tissue swelling which gives an appearance of loss of transradiance on the usual X-ray views of the sinuses. Further examination of the films will show that the opacity extends beyond the limits of the sinus.
Abscess as an extension of chronic sinusitis is sometimes visible in the face. Direct forward extension through the thick anterior wall of the frontal sinus is rarer, but an abscess beside the bridge of the nose above the medial canthus is more common.

Ethmoidal sinusitis may give rise to an abscess below the medial canthus resembling an abscess of the lacrimal sac. An abscess of the cheek is rare in simple maxillary sinusitis but more likely in destructive or caseating sinusitis.

Surgical treatment of these abscesses is necessary once suppuration has occurred. The sinus itself must be explored and free drainage established for the abscess, either through the sinus cavity or by a separate drain. In attempting the latter it must be remembered that there is an area of osteitis between the sinus cavity and the abscess which, if left, may perpetuate the condition.

**Orbital extension**

The maxillary, ethmoid and frontal sinuses have extensive bony walls in common with the orbit.

**Extraperiosteal abscess**

In a chronic empyema of the sinus pus may seep through a natural opening in the bone, such as the canals for the anterior ethmoid or infraorbital vessels, or through eroded bone to form an extraperiosteal abscess within the orbital cavity.

If this abscess forms on the medial wall of the orbit the eye will be displaced forward, laterally and downward, whereas an abscess on the floor will displace the eye forward and upward. If the subperiosteal abscess is anterior, arising from the frontal or an anterior ethmoid sinus, a soft tissue swelling can be felt behind or above the medial canthus.

Antibiotic treatment combined with drainage of the abscess and the causal sinus is imperative to prevent damage to the orbital contents by further extension.

**Orbital cellulitis**

An acute infection can spread through the orbital periosteum to enter the orbital fat, wherein it can extend between the orbital muscles, nerves and blood vessels.

If an orbital cellulitis has developed the eyelids will be swollen but not red. On forcing open the eyelids the eye will be seen to be proptosed, the conjunctiva swollen and engorged and the eye movements restricted. The most worrying indication of the extent of the infection is a decrease in vision. This may be partial or complete and is sometimes permanent.

It is usually possible to arrest the damage to the orbital contents by high doses of antibiotics. An abscess may form and its position should be established by orbital scanning before a decision is made jointly with the ophthalmologist as to the method of drainage.
Cellulitis of the eyelids

An acute ethmoid sinusitis, usually in a young person, can spread forwards and laterally into the eyelids. There is soft tissue swelling with redness of the skin as the cellulitis develops in front of the orbital septum which is a fibrous layer running from the orbital margin to the tarsal plates.

The condition is dramatic, frightening and painful but much less dangerous than orbital cellulitis. When the eyelids are pulled apart the eye is seen to be in the normal position, the conjunctiva is normal, eye movements and vision are unaffected.

Cellulitis will usually resolve on medical treatment but occasionally an abscess will remain. In an older patient this complication is an indication for subsequent surgical treatment of the sinus as erosion of the bony wall is the common route of infection. In a child the underlying sinus disease will usually resolve completely without surgical intervention.

Intracranial complications of sinusitis

Despite the progress in medical treatment of sinus and ear disease, spread of infection from sinusitis and otitis media is the commonest cause of intracranial sepsis. The cavities of the frontal, ethmoid and sphenoid sinuses are closely related to and separated by a thin wall of bone from the anterior cranial fossa. The roofs of the ethmoid cells extend upward into the frontal bone each side of the ethmoid notch containing the cribriform plate and crista galli. Infection may spread toward the meninges by erosion of bone, by pressure within a sinus, by osteitis of the wall of a sinus or through the veins in the bone.

Meningitis and encephalitis

An acute infection entering the anterior cranial fossa will produce a localized or general meningitis. Further invasion will cause an encephalitis.

The clinical picture will change. The patient will become ill, the pyrexia will increase and the pain which was at first localized to the sinus will spread to a more generalized headache with vomiting, pains in the back and stiffness of the neck. Consciousness will be affected as the condition advances with clouding and later unconsciousness.

The cervical rigidity, head retraction and positive Kernig's sign will indicate the diagnosis, although ocular signs, papilloedema and decreased pupil reaction may not occur in the first few days. The bacterium responsible can only be established by lumbar puncture.

Treatment is energetic with heavy doses of a suitable antibiotic. When the intracranial infection has been controlled, the sinus responsible should be explored and permanent drainage established. In rare cases where the response to treatment is slow or if the condition, having improved, relapses, the sinus should be drained before the intracranial infection has been arrested.
Extradural abscess

Gradual erosion of bone by a chronic sinusitis, pyocele or infected mucocoele, may allow pus to collect between the dura mater and the bony skull wall.

The clinical picture may change little and gradually, the emphasis moving from sinus symptoms to a more general headache. An extradural abscess should be suspected if the symptomatic response to proper drainage of an infected sinus is not immediate and complete.

Treatment is by aspiration with instilling of an appropriate antibiotic. Rarely drainage or excision are needed if response is complete. The sinus responsible must be explored and drained.

Subdural abscess

Further inward extension of suppuration will produce an abscess between the dura and the brain. The clinical manifestations are more dramatic, there are pyrexia, fits and localizing neurological signs. Immediate treatment is by aspiration of the abscess and antibiotics are necessary to prevent permanent damage to the underlying brain.

Intracerebral abscess

As the frontal lobe of the brain is invaded the clinical picture will change. The patient may not appear severely ill at first and, if an abscess forms gradually in the brain tissue, the early signs may be drowsiness and apathy. Localizing signs do not appear early in a frontal lobe abscess but fits and papilloedema will appear as the abscess develops. Treatment is by a combination of antibiotics and aspiration of the abscess in the first instance with drainage of the sinus at a later stage unless there is rapid refilling of the abscess.

Cavernous sinus thrombosis

The venous cavernous sinuses lie on each side of the body of the sphenoid bone within which are the two sphenoid sinuses. The internal carotid artery, third and fifth nerves (especially the trochlear, ophthalmic and maxillary divisions) lie in the walls of the venous sinus. Tributaries of the cavernous sinus are the ophthalmic vein, various cerebral veins, sphenoparietal and central retinal veins.

Cavernous sinus thrombosis can result from infection in the orbit, on the face or in the paranasal sinuses. The illness appears suddenly with a sharp increase in temperature, pain in the eyes, proptosis and swelling of the conjunctiva and eyelids. The signs are usually bilateral as the thrombosis spreads to the second venous sinus.

Treatment is by antibiotics to control the local infection and pyaemia, and by anticoagulants to arrest the spread of thrombosis, but it still remains a dangerous condition.
Osteomyelitis

The cancellous portion of the vertical part of the frontal bone contains a large volume of bone marrow in the adult skull. In the infant, the body of maxilla contains bone marrow as well as the developing maxillary sinus. Both these sites can harbour an osteomyelitis secondary to sinusitis, the maxilla in the first year of life and the frontal bone in an adolescent or adult after full development of the frontal sinus.

Osteomyelitis of the maxilla in a child will present as a painful gradual swelling of the cheek and lower eyelid, with pyrexia and malaise. The infection usually subsides on medical treatment without pointing or discharge and surgical drainage is rarely necessary.

Osteomyelitis of the frontal bone is more extensive and dangerous. The area of potentially infected bone is greater and the proximity to the anterior cranial fossa allows intracranial spread.

As infection spreads from the frontal sinus into the frontal bone, pain does not increase immediately, but as the typical puffy tumour (Potts) appears there is severe pain. Treatment is energetic and medical, but surgical drainage and the removal of sequestra may be needed later. The final result may be a visible deformity of the forehead.

Barotrauma

This subject is covered in Volume 1, Chapter 7 but the specific aspects relating to the sinuses merit repetition.

Symptoms

In a healthy person the pressure changes in commercial flying are of little consequence. However, if there is any obstruction of the ostia, problems may occur. At first there is a mild discomfort with or without a sense of heaviness over the affected sinus and, whereas a Valsalva manoeuvre will usually relieve the ear symptoms at this stage, it is valueless in sinus barotrauma.

This discomfort will develop into pain which reaches its maximum when the aircraft lands. The pain will gradually subside over a period of hours or days as the low pressure is relieved by the development of an effusion in the sinus.

The acute condition can be relieved by a further ascent and sometimes will not recur if descent is more gradual.

In the rapid descent of unpressurized fighter aircraft the pain was sometimes sufficiently severe as to make it impossible for the pilot to maintain control.

Treatment

The frontal sinus is most often involved as a consequence of the anatomy of the frontonasal duct. Treatment is medical including analgesics for the pain and medication for
any underlying infection or allergic condition which may have predisposed to the obstruction. If the maxillary sinus is involved the acute pain may be relieved by antrum puncture.

**Prevention**

The main problem lies in the treatment of aircrew or passengers who repeatedly suffer sinus barotrauma after flight. The first step is to identify any anatomical or pathological disorder in the nose which would obstruct air entry into the affected sinus or sinuses. If such a disorder is treated the future attacks may be averted.

In rare cases, an antrostomy may be needed to bypass the natural ostium of a maxillary sinus which is inadequate. It is never justified to explore and drain the frontal sinus to prevent barotrauma. A prolonged abstinence from flying would be preferable.
Chapter 13: The fractured nose

A. G. D. Maran

Epidemiology

According to Fry (1967), fracture of the nose is the commonest fracture in humans. This chapter refers to fracture of the nasal bone but this is only half of the problem of the fractured nose. If fractures of the quadrilateral cartilage and the bone are taken in conjunction, then it must be the commonest fracture but no figures exist to confirm this.

It is certainly more common in the Caucasian than in the Asian or African. In the Caucasian, the nose is the most prominent feature of the face in the anteroposterior direction and is at risk in any fall or facial injury. Furthermore, it is reasonably easily diagnosed because deviation, saddling or swelling are easily noticeable in a nose of this shape. In the African or the Asian, with the squat nose, then deviation and saddling are not so easily noticed and fractures may be missed. This is also the case in children under the age of 5 years where the nose does not form a prominent part of the face. In children, the nose may be fractured and not noticed clinically but the fracture alters the growth pattern of the nose and the nose thereafter grows 'squint' (Pirsig and Lehmann, 1975; Grymer, Gutierrez and Stoksted, 1985).

The nose alone is fractured by low velocity trauma. If it is fractured by high velocity trauma then it is usually accompanied by other facial fractures usually of the Le Fort type I or type II (Le Fort, 1901).

Any nasal injury may be associated with neck or skull damage. The essential whiplash nature of the injury that fractures a nose must always be suspected of having caused neck trauma.

Causes

Four main causes of nasal trauma are: personal assault, sports injuries, personal accidents and road traffic accidents.

The commonest cause is probably assault. In this group the people most affected are, of course, young males. Although many of these patients will come to the hospital spontaneously, a number will come because of pending legal action and it is essential that accurate documentation is made on the first visit with these patients.

The types of sports that cause nasal trauma are football, especially when two players go to head a high ball; rugby, usually due to elbow or fist injuries; racquet sports such as squash where the racquet during the back or forward swing can hit an opponent's nose, or karate. Although it is stated that fractured nose in boxing is rare (calculated by the British Boxing Board of Control to be two out of 4350 bouts), this just applies to nasal bones. It would, indeed, take a blow of enormous frontal force to break the nasal bones with a large 170-gram (6-oz) boxing glove. The area of contact is far too large. What is very common in boxing, however, is damage to the nasal septum.
Under the heading of personal accidents come such events as falls in the home and elderly patients, in particular, will present in this fashion together with the traditional walking into doors or other objects. It is occasionally seen in the spectators at ice hockey matches and has been reported in golf spectators.

The force of injury in a road traffic accident is usually high velocity, so other facial fractures are also caused. It is possible, however, to have isolated nasal damage from the nose hitting the dashboard or steering wheel at a relatively low velocity.

Special attention needs to be given to women and children admitted with nasal trauma. More care than usual should be given to taking the history of events leading up to the nasal trauma in these patients, because of the increasing frequency of wife beating and child abuse.

**Pathogenesis**

If the central part of the face is traumatized then five things can happen depending on the velocity and the direction of the blow.

*No fracture*

With a low velocity blow, the septal cartilage will be deviated to the side and may be pushed back as far as to touch the cheek, but unless it is split between its two fixed points, which are the end of the nasal bone and the maxillary spine, then its inherent elasticity will bring it back to the mid-position and the worst that the patient will suffer is some nasal congestion due to oedema of the septal mucosa.

*Class 1 fracture*

If the blow is of greater severity then a class 1 fracture will result. The nasal bone, although one rather short bone, is in two distinct parts. The distal half is thin and easily broken from the proximal part which is joined to the frontal bone and becomes very thick. A class 1 fracture, therefore, either breaks the quadrilateral cartilage between its two fixed points or takes with it a depression or displacement of the thin distal part of one of the nasal bones. To cause this fracture the direction of the blow must be frontal or frontolateral. The vertical fracture that occurs in the nasal septum was first described by Chevallet and still bears his name. The distal segment of the nasal bone is attached to the upper lateral cartilage which stops it falling into the nasal cavity.

In children these fractures can occur in a greenstick variety and so at puberty, when a growth spurt occurs in the nose, the nose can apparently grow in a squint fashion (Pirsig and Lehmann, 1975). There may also be reduced growth of the nasal bone giving a ski-slope deformity of the nose. The depression of the distal part of the nasal bone may also not be noticed at the initial injury because of the recoil due to the underlying upper lateral cartilage. It is only if the fractured distal segment becomes impacted under the remaining nasal bone that the fracture becomes obvious.
Class 2 fractures

These are fractures of the nasal bone, including the frontal process of the maxilla, as well as the structures involving in a class 1 fracture. This fracture needs a fine degree of medium velocity trauma. There has to be quite a difference between the trauma that causes a class 1 fracture and a class 2 fracture and, if there is enough force to break the thick root of the nasal bone and the maxillary processes of the frontal bone, then it is likely that the fracture will spread and become a Le Fort type II or III fracture. The importance of a class 2 fracture, however, is that it indicates that the ethmoid labyrinth has not collapsed or been compressed. It is recognized by the fact that the patient does not have gross depression of the the nasal bones, although there will be a high deviation. It is nearly always due to lateral trauma and, if the same velocity of trauma were applied from a frontal direction, then it is almost certain that the fracture would have become a Le Fort type fracture.

The nasal bones are attached to the perpendicular plate of the ethmoid. If there is any degree of deviation of the nasal bones visible other than a slight depression of a distal fragment of a nasal bone then, by definition, the perpendicular plate of the ethmoid must also be fractured. It has been shown on several occasions that this fracture takes on a special shape which is governed by the lines of tension present in a structure with the shape of the nose (Mayell, 1973; Harrison, 1979; Murray and Maran, 1980). It is a C-shaped fracture which begins just under the tip of the nasal bones in the quadrilateral cartilage and extends posteriorly and caudally through the perpendicular plate of the ethmoid and, at the anterior border of the vomer, turns anteriorly to run along the lower part of the perpendicular plate of the ethmoid and through the quadrilateral cartilage just above its junction with the maxillary groove. This latter cartilaginous fracture has, on occasion, been called the Jarjavay fracture and has occasionally been described as a dislocation of the nasal septum from the maxillary spine. It is in reality, however, merely an extension of the C-shaped fracture that occurs in deviation of the nasal bones accompanied by fracture of the perpendicular plate of the ethmoid.

This has important implications for treatment because mere replacement of the nasal bones may be successful but, in over 40% of cases, there will be redisplacement of the nasal bones due to overlapping of the fractured ends of the perpendicular plate of the ethmoid and the quadrilateral cartilage. The tension in this overlap will drag the nasal bones back to their original fractured position (Murray and Maran, 1980). Although more will be said about this under Treatment, it is essential to excise the overlapping fragments, before manipulating the bones back so that they heal in their correct position.

Class 3 fractures

A class 3 fracture indicates that the velocity of the trauma has been even greater. It means that the fracture has extended to include the ethmoid labyrinth. The three classes of fracture indicate the three areas of the nose with varying strengths. The class 1 fracture indicates a rather weak but elastic area, the class 2 fracture indicates the main buttress and strength of the nasal skeleton and the class 3 fracture indicates that the fracture has extended back into another weak, but this time rigid, part of the nasal skeleton. If the ethmoid labyrinth, which consists of anything from 4 to 12 air cells, is fractured, then it virtually telescopes on itself taking with it the bony nasal skeleton. The perpendicular plate of the
ethmoid is also rotated thus pulling the quadrilateral cartilage backwards giving the patient a pig-like appearance with forward facing nostrils and saddling. There is apparent widening of the space between the eyes, described as telecanthus and the medial ligament which is attached to the lacrimal crest is often disrupted allowing even further telecanthus. The lacrimal sac and the nasolacrimal duct may be damaged in this injury leading to subsequent tearing and the nasal bones may become impacted under the frontal bone.

This is one of the most difficult nasal reductions and can be likened to the repair of a shattered egg shell. It carries with it many other complications which will be discussed later.

Le Fort type II and III fractures

These are discussed in Chapter 14.

Although most of these fractures are closed, if the velocity is high enough or if the trauma is due to a sharp instrument, they may be compound. They are, however, all compound when viewed from the interior of the nose and the consequence of an open fracture with regard to the nasal bone and associated structures does not carry with it the same possibly serious consequences of a similar opening in a fracture involving a long bone.

Complications

Deviation of the nose

Although this is primarily a cosmetic problem, it carries with it probable nasal obstruction. This nasal obstruction may be present to a greater or lesser extent, however, and many patients with grossly deviated noses do not complain of obstruction while others with mild deviations complain bitterly. The surgeon must learn to differentiate between those who are too shy to talk about the cosmetic abnormality and sublimate it into functional nasal problems, and those who have a truly functional nasal problem. This to a large extent decides the type of nasal operation required.

There will be some nasal obstruction with either of the septal fractures, namely the Chevallet or the Jarjavay. Both will carry some degree of deviation of the lower part of the nose, but it may not be obvious on frontal viewing. It is nearly always obvious on viewing the tip from below. The caudal end of the septum very often presents itself in one or other nostril with consequent asymmetry. If the fracture was created in the early years of life then there may be associated distorted growth of the lower and upper lateral cartilages (Pirsig, 1986). Greater degrees of trauma can cause reduplication of septal fragments. A spur is basically either a Jarjavay fracture if occupying the whole length of the floor of the septum, a reduplication of cartilage if involving a moderate amount of the septum or an isolated septal fragment if localized.

The concept of dislocation of the septum is incorrect. It is intuitively attractive to explain the presence of the caudal end of the quadrilateral cartilage in the nose and a bony cartilaginous disjunction along the floor as the quadrilateral cartilage having exited from the maxillary spine, but this in fact is incorrect. Operations on these patients show that it is always a fracture along the bony cartilaginous junction and there is still cartilage present in
the maxillary crest. It is, in fact, mechanically impossible to extract the cartilage from the
maxillary crest in view of the complicated cross ties between the periosteum and
perichondrium in this area.

Deviation of the nasal bones can recur after manipulation if the nasal bones are pulled
back by the overlapping fragments of the fractured perpendicular plate of the ethmoid. Patients
with nasal deviation can present later if the original fracture was missed due to
swelling or if the patient did not attend primarily. Another reason for persistence of nasalone deviation is the clinician believing an X-ray that is reported as showing no fracture.

A false impression of nasal deviation can be given by a depressed distal fragment of
nasal bone. It is essential that this is diagnosed or else undue or overenthusiastic attention can
be applied to the rest of the nasal skeleton which is really quite normal.

The effect of a greenstick fracture was pointed out earlier; with growth, it will cause
a deviated nose either in the bony or cartilaginous segment.

**Bleeding**

Nearly every patient who has trauma to the nose will bleed due to mucosal lacerations.
Particularly troublesome bleeding can occur with fractures of the perpendicular plate of the
ethmoid when the long sphenopalatine artery may be caught up in the fractured segments and
not allowed to constrict. A similar situation can occur in class 3 fractures where the anterior
ethmoidal artery may be held open by pieces of the fractured ethmoidal labyrinth. Both of
these situations, if the bleeding persists in spite of packing, demand an open exploration and
arterial ligation.

**Saddling**

When blood is in contact with cartilage for any period of time, cartilaginous absorption
results. If the septal cartilage is fractured, reduplicated or absorbed, then there is loss of height
(Huizing, 1986). This may not be immediately obvious. The area of cartilage loss will fill
with fibrous tissue which will then contract in the avascular phase and pull on the remaining
cartilaginous pieces of the septum. This pulls the upper lateral cartilages down between their
fixed points between the lower lateral cartilage and the undersurface of the nasal bone. The
patient then presents with a dip between the lower lateral cartilages and the nasal bone.

A greater degree of saddling is found in class 3 fractures. When the ethmoid labyrinth
is shattered, then the nasal bones are pushed backwards underneath the frontal bone and the
perpendicular plate of the ethmoid rotated upwards carrying with it the quadrilateral cartilage.
The patient then presents with a ski-slope nose and hypertelorism.

The lower lateral cartilages are always supported on the maxillary bone and the caudal
end of the quadrilateral cartilage. Although they may be rotated upwards, showing the nostrils
from the frontal view, their height is usually preserved thus exaggerating any saddling.
**Cerebrospinal fluid leak**

It is unlikely in a class 1 and class 2 fracture that there will be an associated cerebrospinal fluid leak, but any clear discharge from the nose must be regarded with suspicion and tested accordingly for the presence of sugar. In a class 3 fracture, it is quite likely that the fracture line could extend through the fovea ethmoidalis thus involving the dura, giving a cerebrospinal fluid leak. A greater degree of class 3 trauma may involve the sphenoid, but this would be exceedingly rare.

Perhaps the commonest site of a cerebrospinal fluid leak is a fracture involving the cribriform plate in the class 3 variety. Here the dura extends along the olfactory nerves as they exit through the pits in the olfactory plate, and a minor degree of trauma is enough to produce a cerebrospinal fluid leak. Although a cerebrospinal fluid leak usually seals spontaneously and does not require operative intervention, it can create a preformed tract and recurrent meningitis may be a later problem.

**Orbital complications**

Any central facial trauma involving fracture of the nasal bones will result in leakage of blood into the lax periorbital space giving a haematoma. It is essential not to accept this in every case as the common complication of a fractured nose, because it may mean that there is an associated fracture of the floor of the orbit. Class 3 fractures are the type that affect the orbital apparatus. The medial orbital ligament can be detached from the lacrimal crests, causing further hypertelorism and also diplopia. Damage to the lacrimal sac and the nasolacrimal duct will result in tearing and dacryocystitis. It would take very severe injury to create a fracture line that went back to the optic foramen to cause blindness, but it must be remembered that the posterior ethmoid cells is only 1-3 mm away from the optic foramen, although this latter foramen is surrounded by very thick bone.

**Healing**

Skull bones form in membrane and, in general, heal by fibrous union rather than calcification. Nasal bones heal by calcification and are in this respect odd. The assessment of a healed nasal fracture is, however, often complicated by the calcification of the subperiosteal haematoma that often accompanies a nasal fracture. It is possible that a nasal hump for which the patient requests cosmetic rhinoplasty later in life is a calcified subperiosteal haematoma from a minor degree of trauma in early life. A depressed distal fragment of the nasal bone can distort the upper lateral cartilage. Distortions of the upper lateral cartilage due to trauma or growth abnormalities are exceedingly rare and usually reflect anomalies of the bone or the quadrilateral cartilage.

Cartilage heals by fibrous union. Cartilage that is absorbed by blood will, over a period of 18 months, have an effect on the attached cartilages and in this way the lower lateral cartilage and upper lateral cartilages may be distorted.

When dealing with nasal fractures, one has to be aware of the concept of dynamic healing. Scar tissue takes a full 18 months to mature and, while there is a 90% result at 3
months, a final result cannot be estimated before 18 months and indeed the result may gradually get worse over this period.

An example of this is nasal saddling. Some degree of nasal saddling and retraction of the nasolabial angle is inevitable after the submucous resection operation but it is seldom seen until up to a year after such a procedure. The same applies to saddling due to cartilage absorption after trauma, which takes up to a year to occur.

Greenstick fracturing in childhood leads to abnormal growth which is evident during growth spurts (Pirsig, 1986).

Clinical features of nasal trauma

History

The patient may present with no immediate past history of trauma but the nose is deviated. These patients have had trauma in the past and have not recognized it at the time. This can apply not only to deviations but also to saddling and hump formation. The only cause for a deviated nasal septum in the absence of trauma is an asymmetrical face. Although when one talks about trauma one thinks of adult trauma, there has been a great deal of work done on the effects of forceps delivery on the nasal skeleton and the immediate septal abnormalities due to fracture and distortion found after such deliveries.

When the patient does give an immediate past history of trauma then there are a number of points to establish. The first is the direction and the degree of velocity of the trauma. This gives an estimate of the fractures that are possible. Secondly, one has to get the patient to give some description of the shape of his nose prior to the trauma and also his breathing capacity through each nostril. Often this is impossible and asking the patient to bring photographs to the clinic at the next visit is largely a waste of time because, unless the photographs are taken as for rhinoplasty, then one can tell little from holiday snap shots.

The patient who has had fairly severe trauma may have epistaxis, a cerebrospinal fluid leak, diplopia, tearing or telecanthus.

It is essential to make good notes of these injuries because, although few of them come to law, those that do are expected to have a fairly accurate description of the findings and the circumstances. With the law in mind one should also remember to ask the patient about neck pain, limitation of neck movement and root symptoms down the arm, since every facial injury should be regarded as a potential neck injury.

Examination

This is very often difficult because of nasal swelling. There is little to be lost by carrying out immediate first aid and asking the patient to return to the clinic again 5-7 days later. At the first visit, however, it is essential to assess the severity of the injury and also other associated injuries. It is essential to notice whether or not there is cerebrospinal fluid leak and to get any bleeding under control. The patient can be given nose drops to help the
recovery of nasal airways but, if there is any suspicion of septal haematoma, then an incision under local anaesthesia should be performed, followed by drainage if necessary.

**Radiology**

By and large, X-rays for nasal fractures are a waste of time and there is no place for routine radiology in simple nasal injuries. Vascular markings look just like fractures and patients may be taken to theatre on the basis of a radiologist's report which relates to a vascular marking. The importance of X-rays is to rule out more extensive fractures than are assessed clinically and also to make sure that the neck is not damaged.

Radiographs may also show that there is blood in the maxillary antrum and, in these cases, it is essential to make sure that there is not a concomitant fracture of an antral wall. If a class 3 fracture is suspected, then tomography is essential to assess the damage to the ethmoid labyrinth and anterior cranial fossa. If the patient has a number of other facial injuries, then it is best to record these by photography.

**Nasal endoscopy**

If there is sufficient airway then nasal endoscopy should be carried out. This is done with a 70° telescope after vasoconstricting the nose. In particular, one looks for compounding of the fracture of the nasal bones into the nose and the state of the perpendicular plate of the ethmoid.

**Treatment**

**No treatment**

About half the patients who come to an otolaryngology outpatient department as an aftermath of nasal trauma, have no fracture either of the nasal septum or the nasal bone. They may have some oedema but what they require is documentation, examination, reassurance and vasoconstrictor nose drops for the subsequent swelling.

Some patients may become regular attenders with nasal trauma. Their septum may be badly deviated and the nasal bones deviated from previous trauma. These patients should probably not be offered treatment for the acute injury and it should be explained to them that a corrective septorhinoplasty is required, but this would not withstand further trauma as much as their present broken nose. Most patients will accept this situation.

A special category is created by sportsmen who wish to continue in their chosen sport or who have long-standing nasal deformity. These patients should be told that a septorhinoplasty would certainly not strengthen their nose and that further nasal trauma was going to occur. They should be advised to return for a corrective septorhinoplasty when they have retired from active participation in their sport.
Class 1 fractures

These are the fractures of the nasal septum and the distal segment of the nasal bone. Simple manipulation and packing may well be sufficient in these patients. This is usually performed under general anaesthesia with an endotracheal tube and a pharyngeal pack. Elevation of the nasal bone with accurate packing in the vault of the nose should be successful. If manipulation of the Chevallet fracture is not successful, then an open reduction can be done relatively quickly and easily by realigning the fractured segments and stitching the fractured area making sure the knot is to the side that was concave. Packing on the side that was obstructed by the fracture should be left in for at least 3 days. Much attention has been given to the optimum time to carry out simple manipulation. It does not seem to matter so long as it is performed within the first 2 weeks after the injury but thereafter osteotomy may be required.

Class 2 fractures

It has been pointed out earlier, that about half of these fractures redisplace because of overlapping segments of the fractured plate of the ethmoid and the posterior part of the nose as part of the C-shaped fracture that accompanies severe deviations (Murray and Maran, 1980). The manipulation of the nasal bone here should be accompanied by an open reduction of the septal deformity. This takes the form of a Killian incision in the nose with elevation of septal fragments. The C-shaped fracture is easily seen and the overlapping segments excised. The nose is then packed, a plaster-of-Paris splint applied and the packing and splinting removed after 3 days.

Class 3 fractures

The problem with this fracture is to pull the depressed nasal bones out of the face and support them when the normal bony support of the medial wall of the orbit has disintegrated. The septum also requires attention because the rotation of the vertical plate of the ethmoid carries with it the quadrilateral cartilage which gives the patient a pig-like appearance, with the nostrils facing anteriorly. A third problem lies in impaction of the nasal bones under the frontal bone.

These problems all require open reduction. It may be necessary to make an incision over the nasofrontal angle in order to move the nasal bone from under the frontal bone. Nothing can be done about the disintegrated medial wall of the orbit because the bone is so thin, but the nasal bones can be held forwards over wires in the hope that they will attach themselves once again to the frontal bone and that, with replacement of the septum, the upper lateral cartilages will give them support lower in the nose. The wires are 26 gauge and are passed under the nasal bones with straight cutting needles and are tied on the outside over silicon or lead plates. It is better to do this open reduction as soon as possible after the injury but it can be done at any stage. The manipulation of the fractured nasal bone may have to be augmented with osteotomies if the delay is more than 3 weeks (Brain, 1986).

The septum is dealt with as before with a Killian excision and septal exploration. The aim of this operation is to pull the septal cartilage forwards and downwards and it is essential to do all of this surgery as soon as possible because, on leaving for some days, the nasal skin
shrinks and it is impossible thereafter to get increase of length of the nose without adding in extra skin by grafting.

**Treatment of complications of nasal trauma**

**Bleeding**

The bleeding after nasal trauma is either due to mucosal lacerations which should stop within 24 hours with nasal packing or, if it persists and is profuse, then it is due either to an open long sphenopalatine artery or an anterior ethmoid artery. The site of the bleeding should be identified and, if it is from the long sphenopalatine, then open septal exploration should be carried out and, when the artery is freed from the fractured segments, it usually stops with packing.

If the anterior ethmoid artery is continuing to bleed after an ethmoidal fracture, then it needs to be clipped with an external ethmoid approach.

There is no place for postnasal packing in the treatment of bleeding after nasal trauma because the bleeding almost never comes from the artery of Zuckerkandl at the posterior end of the turbinate.

**Cerebrospinal fluid leaks**

It is very seldom that cerebrospinal fluid leaks after nasal trauma require surgical repair. It is only if a bone fragment has been inserted in the dural tear that the leak will continue. In this case it is better to approach the area through a frontal craniotomy. Any approach to the cribriform plate through this area, however, carries with it certain permanent anosmia and thus it is a procedure that should be avoided if at all possible. The importance of a cerebrospinal fluid leak is the possible complication of meningitis, either sooner or later. The patient should be observed for meningism in the immediate post-trauma period and, after discharge, the medical advisers should be appropriately warned to take meningitis type symptoms seriously. The place of routine prophylactic antibiotics is still uncertain.

**Orbital complications**

Telecanthus is improved by correcting the height of a flattened nasal dorsum. If the medial ligaments are torn from the lacrimal crest, then they have to be wired together again and it is best done with 24 gauge wire. There is little that can be done as an immediate reconstruction of the lacrimal sac and nasolacrimal duct. If the patient develops tearing after the trauma and subsequent correction has settled, then a dacryocystorhinostomy should be on offer.

**Treatment for late complications of nasal trauma**

**The deviated nose**

This may be due to deviation of the nasal septum, the nasal bones, or both. It is essential that the nose be 'read' properly so that the correct procedure can be carried out. It
is very seldom that anything needs to be done to the upper lateral cartilages or lower lateral cartilages as these usually reflect displacement due to the quadrilateral cartilage or the nasal bones.

A standard septoplasty approach would be used for the deviated septum. A hemitransfixion incision is made, flaps are elevated and the septal damage inspected. There will almost certainly be a Jarjavay fracture which will look like a ‘spur’. This deviated cartilage should be excised. The cartilage that is still in the maxillary crest should not be removed but the rest of the cartilage should be placed on it.

Scoring fresh nasal cartilage, as described by Fry (1966) does cause it to bend to the other side due to the release of elastic forces within the cartilage. This, however, does not apply in cartilage that has been traumatized. Scoring scarred cartilage will never cause it to move and, if there is a Chevallet fracture, then it is best dealt with by excision of the fractured segments and appropriate stitching.

It is important to judge what the patient hopes to achieve by surgery after trauma. If he/she wants an airway more than cosmetic improvement, then consideration should be given to an inferior turbinectomy on each side.

A medial and lateral osteotomy, as is performed for cosmetic rhinoplasty, is not applicable to bones that have healed after trauma. Any attempt at osteotomy on previously fractured bones will result in an irregular fracture line running along the old fracture line. There will be no control over the osteotomy as there would be in cosmetic rhinoplasty. This is why the triple osteotomy has gained favour. A complete medial, intermediate and lateral osteotomy is performed and this results in the nasal bones virtually being shattered (Mackay, 1986). It releases all of its scarred forces, however, and allows it to be manipulated and reformed in the midline. Appropriate packing and fixation completes the procedures.

The saddle nose

Cartilaginous saddles

There are many ways to improve the appearance of a nose that has suffered a saddle deformity as a result of cartilage absorption secondary to trauma. Minor degrees of saddling can be dealt with by the insertion of small pieces of diced homograft cartilage via an intercartilaginous incision.

Most patients, however, who have a noticeable cartilaginous saddle, should have either a Silastic stent or a piece of rib cartilage inserted into the appropriate area and fashioned to the appropriate shape via an external rhinoplasty approach.

Although heightening the cartilaginous segment of the nose will reduce the broadening effect that usually goes with saddling as a result of splaying of the upper lateral cartilages, some cases may benefit from one of the various cartilaginous swing techniques applied to the upper lateral cartilage (Sear, 1977).
Bony saddling

This is usually the result of a class 3 fracture and is the result of failure to extract the nasal bones from under the frontal bone or a malunion of the nasal bone with resultant depression into the space left by the fractured ethmoid labyrinth.

Again an external rhinoplasty approach is to be preferred and either the dorsum of the nasal bones is freshened or osteotomies are performed. It needs a raw bone surface for a bone graft to take and in these cases an iliac crest bone graft is preferable to a Silastic stent.
Chapter 14: Fractures of the facial skeleton and facial asymmetry

Michael Gleeson

Aetiology

Fractures of the mandible and middle third of the facial skeleton are most commonly the result of road traffic accidents or physical combat (Hagan and Huelke, 1961; van Hoof, Merkx and Stekelenburg, 1977; Starkhammar and Olofsson, 1982; Brook and Wood, 1983). In recent years the introduction of compulsory seat belt legislation has decreased the incidence of these injuries in the UK (Price, 1983; Steele and Little, 1983). In children, falls, accidents while playing and sports injuries are the major causes of facial fractures (Fortunato, Fielding and Guernsey, 1982). The possibility of non-accidental injury should never be overlooked (Rowe, 1969; Hall, 1972). Fractures resulting from complicated dental extractions and pathological lesions of the jaws are seen in all age groups.

Primary care of maxillofacial injuries

Maxillofacial fractures can endanger the airway and are frequently associated with brain damage (Hoffman, 1976). Although a patient with a fracture of the middle third presents an alarmingly dramatic picture the first considerations are to:

1. ensure an adequate airway and ventilation
2. look for major abdominal or other injury if the patient is exsanguinated or in shock
3. assess the level of consciousness.

Detailed examination and definitive treatment of the facial injury must take second place to these requirements.

The establishment of a safe airway and effective ventilation is the most essential primary measure to prevent hypoxia and retention of carbon dioxide, as these are the most important factors in the development of post-traumatic cerebral oedema and damage.

The need for endotracheal intubation should be carefully considered in both the conscious and comatose patient. Posterior displacement of the tongue secondary to mandibular fractures, or posteroinferior displacement of the maxilla with middle third injuries, directly compromises the airway. It may be necessary to reduce a middle third fracture immediately by hooking the fingers of one hand around the posterior margin of the patient's hard palate and pulling the displaced jaw forward. Even those patients with a good airway initially may deteriorate to a very critical state as oropharyngeal oedema develops. Paradoxical chest movement due to a flail segment is a further indication for primary intubation. In conscious patients the loss of voice, dysphagia, surgical emphysema of the neck and pain on palpation of the thyroid cartilages suggests a fracture of the larynx. Increasing stridor with these symptoms and signs is a mandate for immediate tracheostomy.

Haemorrhage into the airway from either the nose or mouth can be torrential. It usually ceases spontaneously but must be controlled by nasal packs, balloons, catheters, sutures or arterial ligature. Super-selective arteriography and embolization is very effective
and indicated in severe late onset bleeds (Mehrotra et al, 1984). Shock is rarely the result of haemorrhage from a facial fracture and, when present, is likely to be due to rupture of abdominal viscera, intrathoracic injury, fractures of the limbs or extensive soft tissue lacerations. The source of blood loss must be recognized, controlled and replaced appropriately and promptly, otherwise hypotension further compromises the patient's cerebral state.

Only after such complications have been controlled should assessment of the head injury be undertaken. The level of consciousness is the most useful criterion. Regularly repeated accurate documentation of spontaneous speech, response to command and reaction to painful stimuli provides a clinically useful scale which reflects improvement or deterioration of the cerebral state (Teasdale and Jennet, 1974). Alcohol is frequently a factor in the acquisition of these injuries and it should be remembered that it may be contributing to the depression of the level of consciousness. Facial swelling may make examination of the eyes exceptionally difficult. However, pupil size and reaction to light must be recorded. Dilatation of a pupil at some interval after injury is highly suggestive of an ipsilateral expanding supratentorial lesion. Urgent reduction of pressure by either medical or surgical means is essential in such cases (Garfield, 1972). Subdural pressure monitoring is a sophisticated method of detecting changes in intracranial pressure before the traditional clinical signs develop. It is available in most neurosurgical centres and facilitates patient management.

Absence of limb movements in an acute head injury indicates primary cerebral damage. Such patients will have a diminished level of consciousness. The presence of a mono-, para- or quadriplegia in a relatively alert patient is highly suggestive of a plexus or cord lesion for which immediate immobilization should be instituted.

The cranium must be palpated very carefully to detect depressed fractures. Scalp lacerations are possible source of intracranial infection and should never be probed in case the venous sinuses are damaged. Lacerations can be satisfactorily managed by irrigation with saline and dry dressings. Cerebrospinal fluid rhinorrhoea and otorrhoea may be present and, in the acute situation, can be very difficult to detect, especially when mixed with blood. It may be distinguished at this stage by comparing the discharge with blood escaping from other sites. As the blood flow diminishes due to clotting the escape of cerebrospinal fluid becomes more obvious. If present, prophylactic antibiotics should be administered. Nasal intubation should be avoided, when possible, if the cerebrospinal fluid leak is from the nose.

Tetanus prophylaxis should be administered to any patient who has not been immunized within 3 years of the accident; 0.5 mL adsorbed tetanus toxoid intramuscularly is the recommended booster dose for those immunized between 3 and 10 years before their injury. A supplementary 250 units of human antitetanic globulin should be given at a different site in the non-immunized patient and to those who have not received toxoid for over 10 years.

Clinical examination of facial injuries

There is rarely any urgent need to reduce and fix facial fractures. Indeed the optimal time for definitive treatment is between the fifth and eighth days post-trauma. This allows
sufficient time to assess the fracture, make splints if required and improve the general medical condition of the patient. Some complications such as orbital haematoma require urgent treatment and even if initially absent may develop before or after reduction. The clinical condition of the patient on admission can preclude full radiological examination and unavoidably some emergency room films lack sufficient definition. There is therefore no substitute for careful clinical extra- and intraoral examination. The following is an accepted approach to the clinical examination of these injuries.

**Eyes**

On admission look for penetrating injuries, corneal abrasions, dislocation of the lens and lacerations involving the lacrimal apparatus. Inspect any subconjunctival haemorrhage and determine its posterior limit, the discs for signs of retinal damage and ischaemia, light reflexes, external eye movements and direction of maximal diplopia if present. An assessment of ocular level should be made by comparing the position of the pupils.

Lack of patient cooperation and periorbital oedema limits the accuracy of the ophthalmic examination. Only gross displacements of the globe will be obvious initially. As oedema subsides such deformities become more evident. In patients with diplopia a forced duction test may then be appropriate to determine the presence of significantly herniated orbital contents. Avulsion of the medial canthal ligament produces an abnormal slant of the palpebral fissure. In such injuries lateral traction of the eyelid fails to produce tension at the medial canthus.

**Nose**

The examination of injuries limited to the nose has already been discussed in Chapter 13. Suffice it to mention two salient features. First, persistent cerebrospinal fluid leaks may only be shown by tipping the patient head and face down. Second, unilateral epistaxis in the absence of a direct nasal injury suggests a fracture involving the maxillary antrum.

**Middle third of the face**

The face should be inspected with particular attention to the distribution of oedema and ecchymoses. Disproportionate lengthening secondary to posterior displacement of Le Fort fractures, although masked by swelling in the acute phase, becomes apparent during the first week as the oedema subsides. The bony contour of the face should be palpated to detect step deformities and surgical emphysema. Sensory testing to determine the distribution of any deficit and evaluation of facial nerve function must be recorded.

**Mandible**

The bony outline of the lower jaw should be felt for step deformity. Careful observation of the symmetry of jaw movement on opening, while placing the little finger of each hand in the patient's external auditory canals, ascertains temporomandibular joint function. Any sensory deficit of the lower lip should be noted.
Mucosa and dentition

The state of the dentition is particularly relevant to the management of these injuries. Missing or fractured teeth should be recorded as they may have been inhaled or dislodged into the soft tissues. The health of the residual teeth is also important in planning methods of fixation. A general inspection of the dental arches for asymmetry is essential. Segments of the dental arches should be gently manipulated to elicit any abnormal mobility or crepitus. The occlusion is frequently subtly or grossly deranged, but may need expert dental assessment for its detection. Blood-stained saliva is a further indication of a fracture which is compound into the oral cavity. The mucosa should be examined for lacerations and haematoma of the buccal and lingual sulci, and of the palate. If the patient is edentulous, the dentures can be of immense assistance, even if broken. They should be obtained or retrieved and any missing fragments accounted for.

Signs of mandibular fractures

Body, angle and symphysis fractures

(1) Step deformity palpable externally or intraorally.
(2) Asymmetry of the lower dental arch and derangement of the occlusion.
(3) Pain, paradoxical movement and crepitus on distraction of the fractured segments.
(4) Haematoma in the buccal sulcus or floor of the mouth.
(5) Blood-stained saliva.
(6) Anaesthesia in the distribution of the mental nerve.

Condyle fractures

(1) Tenderness over the temporomandibular joint.
(2) Trismus.
(3) Deviation of the jaw towards the injured side on opening the mouth.
(4) Inability to move the mandible to the side opposite the injury.
(5) Deviation of the jaw to the fractured side at rest with anterior open bite secondary to gagging of the occlusion on the molar teeth in fracture dislocation.
(6) Symmetrical anterior open bite in bilateral fractures of the neck of the condyles.

Signs of middle third fractures

These injuries are classified into central and lateral types, although in clinical practice these are frequently combined.

Central middle third fractures

(1) Epistaxis.
(2) Circumorbital ecchymosis (panda facies).
(3) Facial oedema.
(4) Surgical emphysema.
(5) Lengthening of the face.
(6) Oral respiration.
(7) Infraorbital nerve sensory deficit.
(8) Anterior open bite (Le Fort II and III).
(9) Haematoma at the junction of the hard and soft palate.
(10) Floating palate and teeth (Le Fort I).

Lateral middle third fractures

Many of the features seen in fractures of the central middle third are present also in the lateral variety, for example circumorbital ecchymosis, facial oedema and surgical emphysema. In addition the following may be seen:

(1) Subconjunctival haematoma.
(2) Proptosis.
(3) Alteration of the ocular level.
(4) Increase in interpupillary distance.
(5) Limitation or absence of external eye movements.
(6) Diminished visual acuity.
(7) Step deformity of the orbital margin.
(8) Epiphora.
(9) Limitation of mandibular movement with depressed arch fractures.
(10) Flattening of the cheek.
(11) Step deformity of the zygomatic buttress on intraoral examination.
(12) Haematoma of the buccal sulcus.

Radiological evaluation of maxillofacial fractures

All patients must have chest, cervical spine and supine lateral skull radiographs taken. In this way significant chest and spinal fractures will be recognized at the outset. Their treatment may take precedence over further radiographical examination or contraindicate the neck extension required for other facial views. Fluid levels in the sinuses or air within the cranium indicates the presence of a cerebrospinal fluid leak which might otherwise be unrecognized.

In each case a standard facial series should be taken. The entire mandible can be demonstrated by a 10° posteroanterior view, an orthopantomogram or panellipse and lower occlusal films. The facial bones are best seen with a 30° occipitomental, a 30° anteroposterior (Towne) and submentovertical views, while the orbits are more clearly projected on a 45° occipitomental radiograph. Further films and projections may be necessary after this screen, for example orbital tomograms and temporomandibular joint films.

Computerized tomography has added a further dimension to the documentation of maxillofacial injuries. Fracture lines are more clearly seen, using bone windows, than with conventional films. Furthermore, the data can be reformatted in any plane. The only drawback of this method of examination is that it must await establishment of a secure airway and requires patient cooperation.
Principles of treatment

The general principles of management of fractures of the long bones apply equally to the facial skeleton. The fractured segments must be accurately reduced, securely immobilized and maintained free of infection for a period of time sufficient to allow bony union.

Facial fractures differ to some extent from others in that nearly all are compound either directly into the mouth or nose, or indirectly into the mouth through the periodontal ligament. It may not be in the best interests of the patient to effect closure by extraction of those teeth involved in the fracture, as their continued presence may be required for fixation or for guidance of the jaws into a functional position. Experience has shown that this compromise between potential infection and accurate reduction is acceptable.

The viability of small fractured segments of the jaws is far better than elsewhere in the body. In general they should be retained as their preservation aids the restoration of facial contours and sequestrum formation is uncommon.

Both closed and open techniques of reduction are practised. All methods aim to fix the fractured part to the nearest superior structure in continuity with the base of the skull. Precise restoration of the occlusion registers the correct functional position of the jaws and the dentition forms an additional splint. The period of fixation is variable but ranges from 10 days for a condylar fracture to 6 weeks for angle or body fractures of the mandible and Le Fort fractures.

Fractures of the mandible

Surgical anatomy

There are several patterns and combinations of fractures recognized in the mandible. Each is determined by the magnitude of the impact, the direction of the blow, the age of the patient, state of his jaws and condition of his dentition (Hodgson, 1967; Huelke and Harger, 1969). Stability or displacement of the fragments is determined mainly by the action of the attached musculature and the plane of the fracture line.

The weakest part of the mandible is the subcondylar region and it is therefore the most common site to fracture. The angle is the next most frequent region, followed by the body, lateral chin and symphysis. Single and multiple fractures occur with equal frequency (Hagan and Huelke, 1961). The most usual combinations of mandibular fractures are bilateral subcondylar fractures, fractures of the body and opposite angle and fractures of the body with the contralateral condyle. Bilateral body, bilateral angle and comminuted fractures are comparatively rare.

The subcondylar region is protected by the zygomatic arch and is therefore usually fractured as the result of an indirect force delivered either to the chin or contralateral mandibular body. Such fractures are rarely grossly displaced. Anteromedial rotation of the condyle secondary to the pull of the attached lateral pterygoid muscle is usual. Fracture dislocations of the joint posteriorly and centrally into the middle cranial fossa are also occasionally seen (Lindahl, 1977).
Displacement of angle fractures is determined by two factors. First the masseter, medial pterygoid and temporalis muscles pull the posterior segment medially upward and forward. The second factor is the direction of the fracture line in the vertical and horizontal planes. Fractures running forward from lingual to buccal resist medial displacement of the posterior fragment and are called *vertically favourable*. Those running in the opposite direction are more easily displaced lingually and are therefore *vertically unfavourable*. Similarly fractures which run from the superior border of the mandible forward to the inferior margin resist upward displacement and are termed *horizontally favourable*. Those running in the opposite direction are more easily displaced and are called *horizontally unfavourable*. In patients with third molar teeth the fracture line invariably runs through either the socket or crypt.

Body fractures are mainly sustained in the first molar or canine regions. Like all fractures through tooth-bearing areas they are usually compound into the mouth. The tendency of the posterior fragment to upward displacement is counteracted, to some extent, by the attachment of the mylohyoid muscle. However, the action of this muscle encourages medial displacement of the posterior fragment.

Displacement of anterior fractures is governed by the extrinsic muscles of the tongue. The part of the mandible bearing the genial tubercles is pulled lingual to the other. Fractures of the ramus and coronoid process are stabilized by the splinting action of the masseter and medial pterygoid muscles and therefore minimally displaced. Conversely multiple or comminuted fractures are usually grossly displaced.

*Closed reduction techniques*

**Intermaxillary fixation**

There are several modifications of interdental wiring techniques, termed intermaxillary fixation. In a cooperative, dentate patient wiring can be undertaken under local anaesthesia. This method of fixation has much to commend it. Simplicity apart, of most significance is the fact that the conscious patient has control of his airway throughout the procedure and is less likely to vomit in the immediate postoperative period.

**Method**

Soft, 0.35 mm diameter stainless steel wire is work hardened by stretching a further 10% of its length. Small loops are made in the wire and fixed to groups of teeth in both jaws by encircling the free ends of the wires around the necks of the teeth before twisting their ends together. These anchorage points are subsequently linked and bound tightly together by elastic or wire so that the dentition comes together in centric occlusion.

In the partially dentate patient preformed arch bars or sectional, silver cap splints can be used to link the dentition into a functional unit. Intermaxillary fixation is then applied by means of wire or elastic bands. In the immediate postoperative period intermaxillary fixation is safer with elastic bands than wire as they can be cut or removed faster and more easily in the event of airway obstruction.
The addition of anchorage hooks to the dentures of an edentulous patient makes these highly suitable as splints to hold the mandible and maxilla together in the correct relationship. These Gunning-type splints are retained by peralveolar wiring through the maxilla and circumferential wires around the mandible. Placement of the wires is effected by the use of an awl.

The above methods of fixation are only applicable to fractures of the tooth-bearing parts of the mandible. They will not provide adequate immobilization for fractures of the condyle, ramus or for some unfavourable fractures of the angle or posterior body. In these instances other methods of fixation should be employed.

**External pin fixation**

External pin fixation still finds favour in particular situations. It can be employed to advantage in cases with combined fractures of both the mandible and maxilla, those with gross comminution or bone loss, for example pathological fractures, gun shot wounds and in cases with atrophic edentulous jaws or osteomyelitis.

**Method**

Pairs of pins linked by cross struts are screwed into the main fragments of the mandible. These are then connected and secured by a universal joint and bar assembly so that the fragments are maintained in the correct relationship.

The biphase appliance is a modification of external pin fixation. Specially designed screws, with threads at both ends, are inserted into the bone fragments and connected by a temporary metal bar. A permanent plastic connector is fashioned from cold cure acrylic resin, adapted over the free ends of the pins and secured by nuts. This system is tolerated extremely well by the tissues for prolonged periods. It is therefore particularly useful for patients in whom bone grafting is required particularly for avulsive trauma and resection of malignant tissue.

**Open reduction techniques**

**Transosseous wiring**

Direct transosseous wiring is a satisfactory and simple method of fixation. It has particular advantages in controlling the edentulous posterior fragment, comminuted fractures which are compound externally and in multiple fractures for stabilizing the lower border of the mandible.

**Method**

Holes are drilled with a No 6 rose-head burr either side of the fracture line, taking great care not to damage either the marginal branch of the facial nerve in gaining access to the mandible or the inferior dental nerve when drilling the holes. Stainless steel wire 0.5 mm in diameter is passed through these holes and tightened to approximate the fractured segments.
The reduction can be further secured by a second wire inserted through the same drill holes but tied in a figure-of-eight across the lower border.

In the past every method of plating and pinning familiar to orthopaedic surgeons has been employed to stabilize fractures of the jaws, for example Kirschner wires (Vero, 1968), Steinmann's pins, titanium mesh, titanium plates (Battersby, 1967; Frost, El-Attar and Moos, 1983), nylon straps and bone staples (Williams, 1985). Although available in most hospitals, and therefore convenient for use, they have all been superseded by the development of compression techniques. The degree of fixation achieved by these methods is unrivalled. Furthermore, compression reduction and fixation accelerates and materially alters the histological pattern of bone healing (Becker, 1974; Champy et al, 1978).

**Compression plates**

The plates are available in several lengths ranging from 3 cm to 7 cm. In one end are placed circular fixation holes by which one side of the fracture is secured with screws. At the other end is an oblong hole and an eccentrically countersunk compression hole. The insertion and tightening of a screw in the compression hole forces the fragments together along the plane determined by the screw in the oblong hole. No other form of fixation is normally required, but it has been found beneficial by some to place the jaws in intermaxillary fixation until the soft tissues have healed.

Oblique fractures are not always suitable for compression plating. In such cases compression can be achieved with lag screws which engage the lingual plate through a bucally prepared hole. Tightening the screws draws the fragments together. Neither compression plates nor lag screws need to be removed unless they become infected.

Compression plates are particularly useful when prolonged immobilization of the jaws would be better avoided in such patients as epileptics, the aged and in body fractures associated with fractures around the temporomandibular joint. Plating is only contraindicated in cases with gross contamination or wounds that will not close.

**Management of condylar fractures**

Fortunately, the degree of displacement is not significant in the majority. The ultimate aim is to produce a functional result either by creation of a pseudoarthrosis or bony reunion of the condyle. Much controversy exists over the correct method of treatment. The fragments can be plated or wired directly but this is not a simple procedure and places the main trunk of the facial nerve at considerable risk of surgical trauma. However, intermaxillary fixation for 10 days is usually sufficient to establish a functional jaw relationship. This makes the patient more comfortable while local muscle spasm and pain subside and is followed by graduated mobilization of the joint.
Fractures of the middle third

Central middle third

Surgical anatomy

These injuries are usually the result of a blow to the front of the face. It is traditional to divide them into alveolar and Le Fort I, II, and III, as most follow these lines of weakness (Le Fort, 1901). This division is of considerable practical significance as it establishes the precise level from which the fractured segment can be suspended and to which structures it may be secured.

Alveolar

This is a fracture through the alveolar process only. It may or may not contain teeth.

Le Fort I (Guerin)

This type of fracture runs above the floor of the nasal cavity, through the nasal septum, maxillary sinuses and inferior parts of the medial and lateral pterygoid plates.

Le Fort II

This is a fracture which runs from the floor of the maxillary sinuses superiorly to the infraorbital margin, through the zygomaticomaxillary suture. In the orbit it passes across the lacrimal bone to the nasion. The infraorbital nerve is often damaged by involvement in this fracture.

Le Fort III

This represents a disconnection of the facial skeleton from the cranial base. The fracture traverses the medial wall of the orbit to the superior orbital fissure and exits across the greater wing of the sphenoid and zygomatic bone to the zygomaticofrontal suture. Posteriorly it runs inferior to the optic foramen, across the lesser wing of the sphenoid to the pterygomaxillary fissure and sphenopalatine foramen. The arch of the zygoma is also broken.

Displacement of all these fractures is the result of the initial impact and tends to be backwards and downwards along the base of the skull. This imparts a characteristic dish face deformity to the patient. All are compound to the nose or paranasal sinuses and some breach the dura. A few are associated with a paramedian split of the palate.

External fixation techniques

External frames tend to be bulky and unsightly. They interfere with the patients visual field and sleep and cannot be used if there is a possibility that a craniotomy will be required or if the patient is likely to have an epileptic fit. External fixation has therefore diminished in popularity since the advent of plating systems which overcome these problems. It is, however, still occasionally used for fractures with gross anteroposterior instability, for
example combined Le Fort and condylar fractures. In these circumstances it usually complements other open methods of fixation.

Disimpaction and reduction of the maxilla may require some force. Either Walsham's or Rowe's forceps can be used to grip the maxillary segment, one blade being placed in the nose and the other on the palate. The maxilla is then gently rocked laterally and forward into its correct position. Occasionally it is impacted behind a fractured malar bone and is quite impossible to reposition until the latter has been reduced. The mandibular dentition is the most accurate guide to the correct, functional position of the maxilla on the cranial base, assuming that the mandible itself is or has been rendered intact. Intermaxillary fixation is therefore applied after reduction of the maxilla in these cases.

The first type of external fixation used was that of a plaster head cap and metal outrigger. It had several disadvantages, not the least of which were instability and patient discomfort. It has been superseded by designs incorporating skeletal pins, for example Box frame, Levant frame and Royal Berkshire halo (Levant, Gardner-Berry and Snow, 1969; Mackenzie and Ray, 1970; Levant, Cook and MacFarlane, 1973; Georgiade et al, 1981). All these devices provide a stable frame attached to the cranium at several points. The maxillary alveolar ridge or dentition is then firmly attached to this frame. These frames are simple to apply using Toller or Moule skeletal pins and universal joints. They are comfortable and well tolerated by the patient.

Facial transfixion is rarely used today, although it is simple and effective. It is particularly suited to Le Fort II fractures in which the zygomatic complex is intact and for those cases where external frames are contraindicated due to skull fracture. Remarkable stability can be achieved by driving a Kirschner wire through the zygomatic bone, across the facial skeleton in a plane inferior and parallel to the inferior orbital rim to exit via the opposite zygoma. The ends are protected by corks or trimmed short. No anaesthesia is required for their removal (Vero, 1968).

**Internal skeletal fixation**

The simplest method of fixation is that of internal wire suspension (Adams, 1942; Kufner, 1970). The fractured maxillary segment may be suspended from various points of the craniofacial skeleton which depend only on the level of the fracture, for example zygomatic arches, orbital rims, forehead, piriform apertures. Thus a Le Fort III may only be suspended from the frontal bone, whereas a Le Fort I can be suspended from any of the above points. This form of suspension is not ideal as it may exert a backward force on the fractured segment and thus encourage the relapse of the displacement. To avoid this, modifications using steel implants have been developed (Stoll, Schilli and Joos, 1983).

Direct wiring can be used as easily for maxillary fractures as for those of the mandible. It permits accurate alignment of the reduced fragments through simple surgical approaches. The technique is identical to that used in the mandible and similarly the wires need not be removed. Miniaturized compression plates serve exactly the same purpose and are far superior to wiring in terms of stability.
**Lateral middle third**

**Surgical anatomy**

The body and processes of the zygomatic bone constitute the lateral middle third. Blows to this part of the face are common and may cause either a depressed fracture of the entire zygomatic bone or a fracture of the arch.

Depressed fractures of the zygomatic bone are sometimes called tripod fractures because the bone breaks in three places - frontozygomatic suture, the infraorbital rim and the zygomatic buttress. These fractures are classified according to their rotation about vertical and horizontal axes. The vertical axis runs between the frontozygomatic suture and the first molar tooth, while the horizontal axis is in the plane of the zygomatic arch. Fractures may therefore be rotated medially or laterally about these planes. In severe injuries the bone is dislocated or comminuted and, in all, there is disruption of the orbital floor.

Two factors govern the degree and type of displacement. First the direction and site of the impact relative to the axes of the zygomatic bone and second, the pulls of the masseter and integrity of the fascial attachments.

The arch tends to break at its weakest point which lies just posterior to the zygomaticotemporal suture. Displacement is usually in a medial direction and can produce trismus by interfering with the coronoid process and temporalis muscle. If the temporalis and masseteric fascia is disrupted the arch tends to collapse inferiorly.

Blows to this aspect of the facial skeleton do not always break the zygomatic bone. A sudden impact with the globe may cause fracture of the orbital floor alone, producing a 'blow-out'. The external eye movements are then frequently restricted by herniation or incarceration of the orbital contents through this defect.

**Management**

Many fractures of the zygomatic complex will not require reduction (Rowe, 1985). Indeed, there is a very real risk of iatrogenic blindness following treatment which should be considered in all those with minimal deformity and impaired vision on the contralateral side (Ord, 1981).

Some patterns of fracture are likely to be stable immediately after reduction, for example medially displaced fractures of the arch, fractures rotated about the vertical axis. Others are unstable and require additional fixation, for example inferiorly displaced arch fractures and fractures rotated about the horizontal axis. Stability after reduction depends to a great extent on the integrity of the periosteum covering the bone and that of its fascial and muscular attachments.

Unlike other facial fractures reduction of the zygomatic complex can only be performed by open techniques. The restitutional force can be applied in either a direct or indirect fashion. The classical method is that described by Gillies, Kilner and Stone (1927) using the temporal fossa approach. A skin incision is made just behind the hair line...
anterosuperior to the pinna. The incision is developed through the temporal fascia so that an
elevator of the Bristow type can be passed deep to it on the surface of the temporalis muscle
to lie behind the body of the zygomatic bone. Controlled elevation can then be applied to the
bone which usually snaps back into place. Great care should be taken to avoid undue pressure
on the parietal bone in this manoeuvre. The Rowe elevator avoids this by incorporating a
hinged lifting arm of the same length as the elevator. Furthermore, the precise location of the
tip of the elevator can be gauged by the position of the lifting handle relative to the orbit.
This is also the preferred method for the reduction of a medially displaced fracture of the
arch.

Elevation of the body of the zygoma has also been described through a direct,
transcutaneous approach using a hook (Poswillo, 1976) and through the buccal sulcus using
a periosteal elevator (Balasubramaniam, 1967; Quinn, 1977). Temporary fixation may be
applied either by packing the maxillary antrum with bismuth-iodine-paraffin paste gauze
through a Caldwell-Luc approach, by a silicone wedge supporting the lateral antral wall
(Gorman, 1980) or a Foley catheter inserted through an intranasal antrostomy.

Fractures found to be unstable following reduction need to be wired or plated using
a direct approach. Stabilization across both the fronto-zygomatic suture and the infraorbital
margin is then obtained. The zygomatic arch can be reassembled in a similar fashion, but it
is also wise to drape the periosteum of the arch to the temporalis fascia in order to prevent
subsequent inferior relapse.

In a few cases the floor of the orbit together with some of the orbital contents
collapses into the antrum. This produces severe limitation of the movement of the globe and
permanent alteration of its position. This so-called 'orbital blow-out' fracture, may be an
isolated injury or the result of a direct blow to the eye ball itself. More commonly, herniation
of the orbital contents into the maxillary antrum is part of a Le Fort II, III, or zygomatic
complex fracture.

The decision to intervene operatively can be very difficult, as minor degrees of blow-
out are not always attended by subsequent diplopia and nearly all orbital injuries cause
diplopia in the acute phase (Crumley et al, 1977; Steidler, Cook and Reade, 1980). It is
sensible to explore and provide support for the orbital floor during the acute phase in some
circumstances, for example gross herniation of orbital contents. Certainly it adds little to the
procedure of transosseous wiring of the orbital rim and similarly an antral pack can serve the
dual role of providing temporary fixation for a tripod fracture while reducing the blow-out.
In other cases, it is better to adopt a conservative approach by observing eye movements at
regular intervals until it becomes clear that fixation or limitation of movement has developed
(van Herk and Hovinga, 1973; Bartkowski and Krzystkowa, 1982). In cases where doubt
exists the antrum can be opened to inspect the defect. Fibreoptic inspection using a sinuscope
is a simpler way to obtain the same information (Westphal and Kreidler, 1977).

There is a wide choice of methods for repairing the orbital floor or medial wall.
Transconjunctival and subciliary approaches are described and both have their advocates
(Tessier, 1973; Converse et al, 1973; Wray et al, 1977). The inexperienced may find that the
transconjunctival route gives them inadequate access and that a lateral canthotomy is required.
The operative field is rarely a problem with the subciliary route, although postoperative
ectropion, either transient or permanent, can follow. In most, gentle freeing of the incarcerated contents together with reduction of the malar fracture will suffice. More significant deficiencies need to be grafted with bone or supported by Silastic. Occasionally these may need to be supported by an antral pack.

It is essential that the retinal blood vessels, visual acuity or pupillary reflexes are carefully monitored in the immediate postoperative period as the central retinal artery may be compromised by retrobulbar haemorrhage. Immediate evacuation of haematoma must be undertaken if the sight is to be preserved. The clinical signs of this painful complication are increasing proptosis and intraocular pressure, an afferent pupillary defect, pallor of the optic disc and diminishing visual acuity.

**Postoperative care**

Careful attention should be paid to oral hygiene. The nursing staff must swab the mouth three times a day with diluted hydrogen peroxide followed by aqueous chlorhexidine (Corsodyl mouthwash). A hygienist should visit regularly to attend to local inflammation of the gingivae. The intermaxillary fixation needs to be regularly checked, wires or bands replaced as necessary and the free ends carefully buried between the teeth or protected with wax so that they do not traumatize the soft tissues. In due course damaged teeth, having served their purpose of fixation require restoration or removal and missing teeth may need to be replaced.

**Cerebrospinal fluid rhinorrhoea**

Cerebrospinal fluid rhinorrhoea is classified into aetiological categories under two main subdivisions, traumatic and non-traumatic (Ommaya et al, 1968, *Table 14.2*). Most cases are produced by accidental or iatrogenic trauma. Non-traumatic cerebrospinal fluid rhinorrhoea is important to consider and recognize but rare.

It has been reported that between 2% and 9% of head injuries are complicated by cerebrospinal fluid rhinorrhoea and, in those with fractures involving the paranasal sinuses, this incidence increases to 25% (Lewing, 1954; Raaf, 1967; Charles and Snell, 1979). Most of these involve the anterior cranial fossa and allow cerebrospinal fluid to leak through the cribriform plate or roof of the ethmoids where the dura is attached firmly and easily torn. In others the cerebrospinal fluid passes through a breach in the posterior wall of the frontal sinus (Calcaterra, 1980).

Cerebrospinal fluid leaks from fracture or congenital defects of the temporal bone may also present as rhinorrhoea if the tympanic membrane is intact. The cerebrospinal fluid then escapes via the eustachian tube into the nasopharynx. Non-traumatic cerebrospinal fluid rhinorrhoea is usually secondary to intracranial tumour or hydrocephalus (Ommaya, 1964).

Pituitary tumours are the most common neoplasm to produce a leak but cases associated with nasopharyngeal carcinomata and acoustic neuromata have been reported. Despite local disease, for example pituitary adenomata, which might be expected to predispose to leakage from the sphenoid sinus, this entity is rare. Almost all fistulae arise in the anterior cranial fossa (Kaufman, 1969). Escape of cerebrospinal fluid from the middle cranial fossa
directly to the nose may result from the persistence of the craniopharyngeal canals (Lowman, Robinson and McAllister, 1966; Hooper, 1971). Leakage from this area is more often a complication of trans-sphenoidal surgery, the implantation of yttrium seeds (Bateman, 1966) or secondary to tumour erosion (Norsa, 1953).

**Symptoms**

Nasal discharge is the most prominent symptom. It may be provoked or increased by physical work or change in posture. Some patients are aware of a persistent salty taste in their mouths and others may be troubled by a continuous headache secondary to low pressure.

**Diagnosis and localization**

Cerebrospinal fluid is usually clear with specific gravity of 1.004-1008. Unlike other pure nasal secretions it contains glucose. The clinical diagnosis depends on the measurement of the glucose content which must be undertaken carefully to ensure that the result is consistent with concurrently drawn lumbar cerebrospinal fluid. Glucose oxidase papers (Clinistix) are not reliable for this purpose (Gadeholt, 1964). Examination of the nose and nasopharynx with a flexible nasopharyngoscope may help localize the side from which cerebrospinal fluid is leaking (von Haacke and Croft, 1983). It is usually necessary to introduce markers or dyes, for example indigo carmine, methylene blue, fluorescein (Kirchner and Proud, 1960) or radioactive tracers into the lumbar theca and then, subsequently, to measure their relative concentrations in patties placed at the sinus ostea in the lateral wall of the nose. Aseptic meningitis has been reported following these procedures.

Plain radiographs are seldom helpful in localizing the origin of cerebrospinal fluid rhinorrhoea, although polytomography will often demonstrate the defect either directly or indirectly through changes in the adjacent paranasal sinuses or their linings (Charles and Snell, 1979). High resolution CT scanning is very useful in traumatic cases and even more so for the investigation of non-traumatic cases where an underlying tumour or hydrocephalus must be found or excluded (von Haacke and Croft, 1983). It should be remembered that cerebrospinal fluid rhinorrhoea in association with congenital malformations may be arising from multiple sites. The correction of these cases is frequently problematic.

**Treatment**

The majority of traumatic cerebrospinal fluid leaks heal without surgical intervention. Until the leak ceases the patient is at a significant risk of developing pneumococcal meningitis. All should be given adequate antibiotic prophylaxis, instructed to avoid nose blowing and kept quietly in hospital. For adults, orally administered penicillin 500 mg four times daily and sulphadimidine 500 mg four times daily provide adequate antibiotic cover. Leech and Paterson (1973) considered that repair should be undertaken if the cerebrospinal fluid rhinorrhoea persisted for longer than 7 days as the protection afforded by long-term antibiotic prophylaxis diminished after that period. Earlier repair offers no better protection against meningitis and carries with it the morbidity of a surgical procedure to close a defect that may heal spontaneously.
The first successful intradural repair, using autogenous fascia lata, was reported by Dandy in 1926. Until recently, variations of this technique were accepted as the standard approach to this problem. It is now recognized that craniotomy is attended by a significant morbidity, usually followed by anosmia and not universally successful, failure rates of up to 27% being reported (Ray and Bergland, 1967; Calcaterra, 1980).

The first extracranial approach to repair a defect of the cribriform plate was reported by Dohlman in 1948. Leaks from the frontal sinus, cribriform plate and sphenoid sinus may be tackled by an external ethmoidectomy approach using fascia or a mucosal flap from the nasal septum as described by Hirsch in 1952.

The trans-septal route employed for hypophysectomy may also be used to control cerebrospinal fluid rhinorrhoea from this region (Calcaterra and Rand, 1976). This has the advantage of avoiding any facial incision and allows the sphenoid to be packed with fascia and a free muscle graft without any open communication with the nasal cavity. In all these cases lumbar cerebrospinal fluid drainage is advisable for a few days postoperatively in order to maintain a constant low pressure on the closure.

It is thought that high pressure leaks act as a safety valve limiting the potential damage of persistent raised intracranial pressure. The repair of these should, therefore, only be undertaken after the cause of the high pressure has been dealt with or a suitable shunt inserted.

**Facial asymmetry**

No face is perfectly symmetrical. A degree of facial asymmetry adds character and sometimes enhances an individual's appearance. The rapid development of asymmetrical features is a common presentation of neoplastic, cystic and inflammatory lesions of both the facial skeleton and soft tissues. It is alarming for patients and on occasion prompts them to consult a surgeon at a relatively early stage, even before the onset of pain, anaesthesia or functional deficits. Other asymmetries, frequently subtle at the outset, continue to develop for a more prolonged period of time, sometimes spanning several decades. These conditions are usually genetic disorders or malformations. It is therefore reasonable to outline these disorders on a chronological basis according to the age at which they first manifest, detailing only those not dealt with elsewhere in the text.

**Facial asymmetry of childhood**

Dental infections, paranasal sinus sepsis and viral parotitis are the most common causes of acute facial asymmetry in this age group. The diagnosis of these conditions is rarely problematic.

**First and second branchial arch syndromes**

A wide spectrum of deformities is seen in patients with these syndromes. They are thought to be caused by a haemorrhage arising in the anastomosis which precedes the formation of the stapedial arterial stem (Poswillo, 1973). A number of synonyms have therefore been applied to these conditions, for example necrotic facial dysplasia,
otomandibular dysostosis, craniofacial microsomia, lateral facial dysplasia and hemifacial microsomia.

Although almost invariably unilateral in presentation some bilateral cases have been recorded. Even in these there is considerable difference in expression between the two sides. The most common abnormalities are hypoplasia of the external and middle ear, mandibular ramus and facial expression, parotid gland and occasionally the facial nerve. These defects often appear mild at birth but progress to severe asymmetrical deformities with growth. Some are amenable to surgical correction (Murray, Kaban and Mulliken, 1984).

A number of recognized syndromes exist in which branchial arch dysplasia is but one component. Most of these syndromes are symmetrical.

**Goldenhaar's syndrome**

This consists of hemifacial microsomia, clefts of the lips and palate, epibulbar dermoids, vertebral, cardiac and renal abnormalities. A few of these patients are mentally retarded. The degree of expression is variable and the mode of inheritance far from clear.

**Hemifacial hypertrophy (Curtois' syndrome, Steiner's syndrome)**

There is no recognized basis for this deformity which affects both the hard and soft tissues of the face and jaws. In some there may be total body hemihypertrophy, while in others the enlargement is limited to the face. The degree of distortion is very variable. In some it is mild while in others it is monstrous. Nearly all are evident at birth, become accentuated at puberty and stabilize when active growth stops.

**Klippel-Trenaunay-Weber syndrome (angio-osteohypertrophy syndrome)**

In this condition facial hemihypertrophy is seen in association with a segmentally distributed angiomatous naevus, most commonly in the second trigeminal dermatome.

**Facial asymmetry secondary to osteomyelitis or trauma to the temporomandibular joint**

This is a rare cause of progressive facial asymmetry which is secondary to ankylosis of the temporomandibular joint and retardation in growth of the mandible (Souyris, Moncarz and Rey, 1983). Infection may spread to the joint from localized osteomyelitis of the jaw or follow a generalized septicaemia. In the past this was most often seen after scarlet fever, typhoid, pneumonia, influenza and measles.

**Facial asymmetry of adolescence**

**Fibrous dysplasia**

In this condition areas of bone are replaced by fibrous tissue and become enlarged. These lesions may be monostotic or polyostotic. The polyostotic variety tends to be unilateral in distribution. Enlargement of the facial bones is painless, usually noticed in adolescence and
continues after somatic growth has ceased. These lesions may cause derangement of the
dentition, protrusion of the eyes, obliteration of the sinuses and nasal passages. Foraminal
encroachment can result in deafness and blindness. When the long bones are affected they
become bowed and sometimes fracture repeatedly.

Polyostotic fibrous dysplasia when associated with cutaneous pigmentation and
endocrine disorders is known as Albright's syndrome. The degree of pigmentation is
proportional to the extent of bone involvement. Precocious puberty is the most frequently
associated endocrine disorder but hyperthyroidism, diabetes mellitus and acromegaly have all
been recorded. Skeletal growth in these children is rapid, but as the epiphyses tend to fuse
early the resulting adult stature is short.

**Progressive hemifacial atrophy (Parry-Rhomberg syndrome)**

Unilateral progressive wasting of some or all of the facial tissues in this uncommon
condition commences in the second decade. Atrophy of the subcutaneous fat and muscle
proceeds together with underdevelopment of the facial bones. In some, intracerebral
calcification has been seen and is thought to be associated with haemangiomas. Reflex
asymmetry, impaired sensory function and optic atrophy may also be present.

**Neurofibromatosis (von Recklinghausen's disease)**

This disease is transmitted in an autosomal dominant fashion but rarely becomes
clinically obvious before puberty. The tumours in this condition are usually multiple, smooth,
and rounded, and may attain a considerable size, thereby producing craniofacial distortion.
Any cutaneous, visceral or cranial nerve can be affected. Cutaneous pigmentation, café-au-lait
spots, is present in these patients. Later sarcomatous change in the neurofibromata is well
recognized.

**Cysts of the jaws**

Many types of cyst arise within the jaws. Some emanate from odontogenic tissue, for
example primordial, dentigerous and periodontal cysts. Others develop from sequestrate
fissural epithelium, for example globulomaxillary and median cysts. Although swelling limited
to the jaws, displacement of adjacent teeth, infection or fracture is the most common mode
of presentation, some may achieve considerable proportions so slowly and asymptotically
that facial asymmetry is the first sign.

**Facial asymmetry of adult life**

In this age group, facial asymmetries are more usually the result of sinus mucocoeles,
osteomyelitis, benign salivary tumours or tumours of the jaws. In this later category
ameloblastoma, Ewing's tumour, osteosarcoma and fibrosarcoma should be considered as they
afflict a younger age group than other oral neoplasms.
Facial asymmetry of old age

Malignant salivary tumours, sinus neoplasms and carcinomata of the oral cavity are the predominant causes of facial asymmetry. These conditions produce asymmetry of the face relatively rapidly and are described elsewhere.

Paget's disease (osteitis deformans)

The bony enlargement seen in this disease is not always symmetrical. The facial bones are frequently affected early in the clinical course which may extend for decades. Foraminal encroachment may produce deafness and blindness. Sarcomatous change in the affected bones is a well recognized late complication of this disorder.
Chapter 15: Rhinoplasty

T. R. Bull and I. S. Mackay

A knowledge of nasal plastic surgery is necessary for otolaryngologists as variations in the external nasal shape are frequently linked with septal deformities. Correction of the airway associated with rhinoplasty forms a considerable part of nasal surgery.

In recent decades, a standard technique for rhinoplasty has evolved. The surgical steps present little technical difficulty, but their correct application is far from easy and rhinoplasty is a branch of surgery where errors related to lack of judgement are very obvious. Septoplasty aims to reposition the septum to the midline with minimal excision of the cartilaginous and bony components.

In almost all cases, correction of the septum can be combined with rhinoplasty in one operation. When the septal or external nasal deformity is gross, however, the patient should be warned that two steps may be necessary. A surgical anachronism is a standard submucosal resection to be followed at a later date by a rhinoplasty. A standard surgical step in rhinoplasty involves some degree of separation of the upper lateral cartilages from the septum. With inadequate support in the midline following removal of the septal cartilage, the division of the upper laterals from the remaining septum is likely to result in collapse of the nasal dorsum with saddling of the nose. A rhinoplasty after a standard submucosal resection is, therefore, compromised and a satisfactory result may not be achieved.

Analysis and selection of patients

A sense of aesthetics and common sense are necessary qualities to combine with sound surgical technique if satisfactory rhinoplasty results are to be achieved with minimal postoperative problems. The main cause of patient dissatisfaction following rhinoplasty is a failure of the surgeon to understand the patient's wishes. It is important for the patient to have a clear and realistic idea of the limitations of surgery. Good preoperative photographs are essential and the changes possible with surgery can be demonstrated to the patient on the photographs. Photographs also give the surgeon an excellent concept of the changes which can be achieved and the correct surgical steps which must be applied. In postoperative analysis photographs once again are extremely helpful.

The surgical approach to rhinoplasty should be influenced by the sex of the patient. In the female, one is operating for 'beauty'; in the male, the common complaint about the nose is that it gives rise to ridicule and comment and the operation is to eliminate these unsettling factors. Also, in the male, rhinoplasty may be carried out if a change in a particular racial appearance is required. Occasions, however, in which 'beauty' is involved for males are obviously less common.

In a male, particularly, a complaint about nasal deformity may be the presenting symptom of a profound psychiatric disturbance. It is important to detect these disturbed personalities in the preoperative assessment. Eight surgeons have been killed by disenchanted and mentally disturbed patients after rhinoplasty. The well-adjusted patient is invariably specific in the dislike about the shape of the nose. A complaint that the nose is too large,
deviated or has a hump, which links with the examination findings, is usually a clearcut case for rhinoplasty without psychological problems. The disturbed patient, however, is non-specific and evasive in the complaints about the nasal shape. A preoperative psychiatric assessment is necessary for those in whom the surgeon is doubtful about the patient's exact wishes and motives.

Care must be taken not to 'over-operate': the changes with rhinoplasty should be definite but subtle in most cases, and over-dramatic change may well predispose to a surgical or 'operated' appearance.

The age of the patient also has specific relevance for rhinoplasty. It is usually not advisable to operate under the age of 15 or 16 years, for the septum and the nasal bones may continue to grow or the shape continue to change. In older patients, a less radical alteration should be the aim. A gross nasal reduction in the more elderly not only tends to appear unnatural, but the elasticity of the skin is less and may not 'take up', leaving folds over the dorsum of the nose.

The height of the patient is another factor to be considered before rhinoplasty, in that a small nose may well be suitable for a small patient but frequently, and particularly in the male, a small nose on a tall patient - especially if the nasal tip is over-rotated - looks unnatural.

The moderate nasal deformity is probably the easiest to correct. In correcting a minimal deformity, extreme accuracy is necessary to achieve the desired result. Gross nasal deformity, which appears to present as an easy surgical challenge, may also be difficult. A natural-looking nose following a gross nasal reduction is not easy to achieve.

Finally, it is important to assess the nasal skin texture before rhinoplasty. With thick skin, changes or reductions made in the underlying cartilage and bone are not so obvious when the thick 'blanket' of skin is redraped. In thin skin, however, minimal change is apparent and any irregularity or asymmetry in reduction may present an obvious deformity. Telangiectasia of the skin may also be more obvious after a rhinoplasty and tethering of the skin to the bone or cartilage is more common with thin, ageing or atrophic skin.

Surgical anatomy of the nose

Approximately one-third of the supporting structure of the nose is made up of bone, the remaining two-thirds being cartilaginous. This may, however, vary considerably.

The bony skeleton

The two nasal bones project from the nasal process of the frontal bone superiorly and from the frontal process of the maxilla laterally. These bones are supported in the midline by bony nasal septum - the perpendicular plate of the ethmoid which is continuous with the vomer inferiorly and the cartilaginous septum anteriorly. The nasion is the depression of the profile at the root of the nose where the nasal bones meet the bossed glabella of the frontal bones.
The floor of the nose is formed posteriorly by the palatine bone and anteriorly by the maxilla. These paired structures both fuse in the midline to form a crest which supports the bony and cartilaginous septum. Anteriorly, the crest of the maxilla protrudes forwards as the nasal spine. If this projects too far, it can make the nasolabial angle more obtuse and give the appearance of a tethered and short upper lip.

**Cartilages of the nose**

The mid-third of the nose is formed by the cartilaginous septum and the paired upper lateral nasal cartilages. The upper laterals are triangular in shape and are overlapped superiorly by the nasal bones and frontal process of the maxilla and overlapped inferiorly by the lower lateral nasal cartilages; the groove thus formed is known as the limen nasi and is the landmark for the 'intercartilaginous incision'. It is important to note that the upper laterals lie below the nasal bones superiorly, and it is easy to disarticulate these structures when attempting to elevate the skin and periosteum overlying the nasal bones, leading to an unsightly deformity.

The lower lateral nasal cartilages are also known as the alar cartilages and form the lower third of the nose. These two cartilages are made up of a medial crus, which forms the columella, and a lateral crus, which provides the framework for the tip of the nose, preventing alar collapse on inspiration.

The lower margin of the lateral crus does not follow up the margin of the nostril but ascends away from it as it travels laterally, that is the margin of the lower lateral cartilages may be 1 cm from the margin of the nostril laterally, but only 1 mm distance medially. The fact that these two margins do not run parallel is of utmost importance surgically, whether attempting to follow the rim of the cartilage for a 'rim incision' or perhaps, even more significantly, when leaving 3-4 mm of cartilage undisturbed in a 'cartilage-splitting incision'.

The highest point of the lower lateral cartilage is referred to as the dome. This is usually at the junction of the medial and lateral crus but may, on occasions, be at a variable distance along the lateral crus. A 'facet' is found in most patients lying between the columella and lateral rim of the nostril. This facet is also referred to as the 'soft triangle'; it is not backed by cartilage and surgical interference at this point should be avoided as postoperative notching may occur which is impossible to correct.

Lying between the upper and lower lateral cartilages are several variable cartilages referred to collectively as the sesamoid cartilages.

The columella is formed by the caudal end of the septum, the nasal spine and the medial crura of the lower lateral cartilages. The nasolabial angle is that angle formed between the lip and the columella and is normally about 90° in males and a little greater in females.

**Muscles of the nose**

The procerus can be considered as a continuation of the frontalis muscle. Contraction of this muscle shortens the nose, pulling it upwards. Many surgeons in the past have removed a small portion of the procerus in an attempt to deepen the frontonasal angle. Since this is
usually replaced by scar tissue, it is seldom helpful. Laterally, the alar fibres of the nasalis and levator labii superioris shorten the nose and dilate the nostrils while the transverse fibres of the nasalis muscle compresses and contracts the nostril. Inferiorly, the paired depressor septi nasi muscles pass from the bone of the maxilla above the incisor teeth to the septum and alar cartilages and depresses the tip of the nose. In some patients, excessive activity of this muscle can cause the tip of the nose to move excessively while talking and division of this muscle may prevent this. Other facial muscles have an indirect effect on movement of the nose. All these muscles are innervated by the seventh cranial nerve.

**Anaesthesia**

In the majority of cases, the authors prefer to undertake surgery under general anaesthesia with the airway protected by a cuffed endotracheal tube and bleeding controlled by both topical application and infiltration with a vasoconstrictor. If this is undertaken in a careful and thorough manner as described below, bleeding is minimal. Controlled hypotensive anaesthesia results in excellent operating conditions, but involves risks over and above a standard anaesthetic.

If a general anaesthetic is to be given, the patient will need to be admitted a reasonable period prior to surgery. A full history must be taken and thorough general examination undertaken, together with any relevant preoperative investigations to ensure that there are no contraindications to general anaesthesia.

The patient must have nothing to eat or drink for a minimum of 4 hours preoperatively and is premedicated in the usual way. As the nose is to be infiltrated with vasoconstrictors, the anaesthetists may wish to avoid halothane and use a beta-blocking agent, provided this is not contraindicated, for example by asthma. The patient is intubated with a cuffed endotracheal tube which is inflated and the pharynx then packed with moist ribbon gauze. It is important to ensure that the tube is firmly fixed in the midline with strapping to avoid any asymmetry of the face. The patient should be monitored with an electrocardiographic (ECG) recorder throughout.

The patient should be placed on the operating table in the supine position with the head on a head-ring and the table rotated to about 15-30° in the head-up position. The operating lights are then arranged with one directly above and a satellite directed at the nose from the foot of the table. A headlight or fibreoptically illuminated instruments will also be required, particularly if any septal work is required.

In addition to topical application of vasoconstrictors to the nasal mucous membranes, the nose may be injected with 1% lignocaine containing 1:80,000 adrenaline. Care must be exercised while undertaking this, with a small bleb injected at a time, using a maximum of 6 mL. The following sites should be injected:
1. between the upper and lower lateral cartilages
2. along the nasal septum
3. laterally, towards the infraorbital nerve
4. along the site of the lateral osteotomy
5. the columella
6. the lower margin of the lower lateral cartilages.
It is important to wait about 10 minutes for the vasoconstrictors to have maximum effect.

The osteocartilaginous vault

A prominent dorsal nasal hump is the commonest cause of patient dissatisfaction with the appearance of their nose. In theory, the steps taken to 'dehump' and 'infracture' the nose are not difficult; nonetheless, it is all too easy for the final result to be 'unfavourable'.

A secondary hump or 'pollybeak' will result from inadequate removal of cartilage in the supratip region, while excessive removal of bony hump, particularly in males, can produce a most unsatisfactory appearance and incomplete lateral osteotomies may prevent proper infracture of the nasal bones, leading to an 'open-roof' deformity.

How much hump to remove? This is a question which cannot easily be answered as it will depend on the thickness of the skin, the amount of cartilaginous hump relative to bony hump, the width of the hump and angulation of the tip of the nose, as well as the inclination of the nasofrontal angle. The aim, however, should be to reduce the nose in harmony with the remaining facial features, remembering that it is always better to remove too little than too much.

There is much debate as to whether it is better to reduce the hump before the tip or remodel the tip before the hump. Those in favour of undertaking tip-plasty first will argue that one cannot judge how far to reduce the hump until the tip has been corrected; on the other hand, it can be argued that the degree of tip modification will depend on the new profile following correction of the osteocartilaginous vault. Another point in favour of the profile alignment first, is that this requires the introduction of relatively large and cumbersome saws and osteotomies which may more easily damage the lower lateral cartilage or tear the delicate flaps in the vestibular skin following tip-plasty. Those in favour of leaving the bony work until last will also argue that this causes most bleeding and should not be done until all the finer work is complete. The authors have no special preference, although they tend to undertake profile correction first, followed by tip-plasty except possibly in those cases with a short columella, where projection of the tip can be seen to be the main aim following which little, if any, hump reduction is required.

The first step after suitably arranging the patient in the head-up position, cleaning the nose and infiltrating with vasoconstrictors, will be to trim the vibrissae of the nasal vestibule as these not only get in the way of surgery but also tend to cake with blood clot afterwards, causing discomfort.

An incision is made along the sulcus formed by the lower lateral cartilage overlapping the upper lateral cartilage - the limen nasi. This intercartilaginous incision is continued medially to become continuous with the transfixion incision which separates the columella from the caudal border of the septum. Some surgeons will include a small sliver of cartilage in the columella (high transfixion incision) to prevent contracture of the scar pulling the tip of the nose downwards, contributing to a 'pollybeak'.
A 'hemitransfixion incision' is a contradiction in terms and may relate to two quite separate procedures: either a short incision which completely transfixes the columella but extends only a short distance down towards the nasal spine, or an incision which does not completely transfix the columella since the incision may be made on the right but not extend through to the left, while still being complete because it exposes the whole of the caudal end of the septum. The former incision is thought by some to reduce the likelihood of a polybeak deformity due to scar contracture pulling down on the tip of the nose. The latter is particularly useful where extensive septal work is to be undertaken, and the surgeon will find it easier to elevate the mucosa if one side remains intact to act as a retractor.

The skin overlying the upper lateral cartilages is then elevated up to and a little beyond the osteocartilaginous junction of the nasal bones and upper lateral nasal cartilages. One then attempts to elevate the skin and periosteum overlying the nasal bones, being careful not to disarticulate the upper laterals from the undersurface of the pyriform aperture. The elevation is continued up towards the glabella. Lateral elevations should not be continued too far, particularly if multiple osteotomies are to be undertaken when the comminuted fractured bones will rely on the overlying skin and underlying mucosa to splint them in position. If multiple osteotomies are not required, the elevation may be continued a little more laterally; wider elevation may be helpful in 'redraping' the skin if a radical reduction is to be undertaken.

The upper lateral nasal cartilages are then divided from the nasal septum, keeping as close as possible to the nasal septum to prevent the formation of a 'T-shaped' structure which may be difficult to lower at a later stage. Occasionally patients may have a 'V-shaped' deformity where the upper lateral nasal cartilages and nasal septum join, which can be confusing unless the problem is recognized.

The upper lateral nasal cartilages and septum are then lowered, each by the same amount, up to the nasal bones using Foman scissors and the hump reduction completed with an osteotome or chisel. The bony and cartilaginous hump can then be removed with a large artery forceps, but should always be pushed further into the nose prior to withdrawal to detach any adhesions remaining between mucous membrane, periosteum and bone.

An alternative method is to lower the osteocartilaginous hump using a Bull's nasal saw which has a single fine blade, allowing the operator to remove even a small hump and a rounded blunt end to prevent damage to the overlying skin.

The nasal bones can be reduced further with a rasp although cartilage must be removed with a scalpel or scissors. Once a satisfactory profile has been achieved, the lateral walls of the nose will need to be infractured to close the flat 'open' bridge. In order to achieve this, lateral and medial osteotomies will be required. If these are curved in towards each other, these may suffice; if not, a superior osteotomy may be additionally required.

The techniques for undertaking the lateral osteotomy differ greatly. Few surgeons now use the saw as this requires elevation of a tunnel of skin and periosteum laterally along the line of the proposed osteotomy, which leads to marked swelling postoperatively, is laborious, and may cause bone dust which can become infected. A small 2-3 mm osteotome may be introduced intranasally at the lateral aspect of the pyriform aperture, via sublabial incision or
through a tiny stab incision directly through the skin. Whichever method is used, it is important that this osteotomy should be as low as possible to prevent a 'step' deformity which can often be felt and may be seen postoperatively. Once the osteotomies have been performed, the nasal bones can be infractured.

The height of the septum and lower lateral cartilages should then be rechecked. Ideally, the upper laterals and septum should be lowered equally. It is preferable to remove a little too much cartilage from the supratip region rather than too little as a secondary hump or 'pollybeak' deformity is the commonest cause of secondary revision although, in some cases, this may well result from hypertrophic scar tissue rather than inadequate removal of cartilage.

Reducing the caudal end of the septum will not, in itself, shorten the nose - to achieve this will require surgery to the lower lateral cartilages or tip-plasty in addition to often minimal reduction of the septum. Excessive removal of caudal septal cartilage simply results in columellar retraction or an excessively obtuse nasolabial angle.

Nasal tip rhinoplasty

In the early days of rhinoplasty, the finer surgery of the nasal tip was relatively ignored and more attention was given to the steps of hump removal and nasal bone infracture with lowering of the septum and upper lateral cartilages. In the last two decades, more thought and more elaborate techniques for the alar cartilages have been developed. Finer, more subtle and predictable changes in nasal tip anatomy can be achieved. Nonetheless, the more elaborate tip techniques have the disadvantages of producing more problems with tip asymmetries and deformities if wrongly applied or incorrectly carried out. Nasal tip deformities, furthermore, are probably more conspicuous than any other site on the nose or face.

One of the more common problems in the nasal tip is a bulbosity or fullness over the upper or cephalic aspect of the alar cartilages. Excision of the cephalic aspect of the alar cartilage not only achieves narrowing of this area, but makes some rotation of the nasal tip possible by a 'visoring' of the caudal rim of the alar cartilage into the cephalic defect.

Access to the cephalic portion of the alar cartilage is commonly achieved by a retrograde dissection via the intercartilaginous incision. With this approach, however, it is not easy to achieve an accurate and symmetrical excision of cartilage and a cartilage-splitting incision and delivery of the cephalic portion of the cartilage is preferred. Whichever approach or technique is applied to the cartilage, it is important to preserve the underlying vestibular skin. If an excess of vestibular skin is excised, pinching and alar collapse may follow. This problem, unsatisfactory from the point of view of both airway and cosmesis, is also probable if there is a failure to preserve a sufficient rim of alar cartilage to support the lateral wall of the nose. About 3-4 mm of cartilage is necessary to maintain the alae in a lateral position and prevent this collapse, but in a thin-walled nose with thin skin and rather lax cartilage, more lateral crus may be necessary. The texture of the skin and cartilage, therefore, may well determine the probability of this lateral alar collapse; with relatively thick skin and strong cartilage, a rim of 2 mm will suffice for support. If, however, skin and cartilage are removed from the lateral wall of the vestibule and removed to excess, then the pinching or 'knock-
kneel'd tip deformity will result. It is also important to excise equal portions of alar cartilage from each side to preserve tip symmetry and this is particularly necessary when the overlying skin is thin and the underlying cartilage is relatively strong and resilient.

Although the cartilage-splitting incision allows delivery of the upper aspect of the alar cartilage, this approach will not suffice when more radical changes are needed in tip anatomy. A rim incision with complete delivery of cartilage is necessary in these cases.

The rim of alar cartilage does not follow the margin of the nasal vestibule. The alar cartilage is placed obliquely and although, medially, the medial crus forms the rim of the vestibule, the lateral crus is up to 5 mm or more above the margin of the nostril. The rim incision and full delivery of the alar cartilage is necessary to correct bifid of flat nasal tip deformities. It is also necessary if lengthening of the columella is needed, and may be required to reduce a marked tip projection. Any radical change in tip anatomy which may be needed in such gross deformities as the cleft-nose nasal tip, also require a full delivery of the nasal cartilages. The rim incision and delivery of the alar cartilage gives an almost complete exposure of the lateral crus, the dome and the medial crus of the cartilage. If the cephalic half or two-thirds of the alar cartilage is removed (with preservation of vestibular skin), rotation or visoring of the remaining rim of cartilage achieves marked tip rotation. In most instances, it is better to avoid dividing the lower lateral cartilage at the dome or elsewhere for, although incision of cartilage in itself will alter tip anatomy as the crura spring apart, irregularity or sharp edges will result. This problem is particularly likely with resilient cartilage underlying thin skin. Symmetry of any cartilage incision or excision and the avoidance of any sharp edges is, therefore, important in these cases. It may be necessary however, to divide the alar cartilages separating the lateral and medial crus completely from one another to increase tip projection. In the technique described by Goldman (1965) - which is one of the better established techniques for increasing tip projection - the alar cartilages are divided lateral to the dome; the medial crura, plus the medial portion of the lateral crura are then sutured back-to-back. Cartilage grafts and struts are also advocated for insertion between the medial crura and are another way to increase tip projection and lengthen the columella. Recently, Sheen (1978) has advocated the case of cartilage grafts to give increased tip projection and a more pleasing tip anatomy. These grafts certainly have a proven place in revision rhinoplasty. In these cases, an excess of cartilage has almost certainly been excised at a previous operation or operations and replaced with fibrous tissue. Cartilage replacement is often clearly needed. In primary standard rhinoplasty, however, the use of nasal tip grafts or adjunct grafts is not usually necessary.

The saddle nose

The cartilaginous septum and maxillary bony crest form the main supports of the lower two-thirds of the nasal dorsum. If there is insufficient cartilage to give support, either due to absence or fibrosis of the cartilaginous part of the septum, nasal saddling to various degrees will result. Nasal saddling is, therefore, commonly seen after septal haematomata, following septal surgery or injury and, if the haematomata becomes infected, nasal collapse is almost inevitable. Patients should of course be warned prior to drainage of a septal abscess or haematoma that some degree of nasal saddling may occur. Immediate grafting is advocated by some (Huizing, 1986) but, in most instances, grafting of the dorsum is deferred until the degree of saddling is evident. Loss of septal support for the nasal dorsum, although resulting
mainly from trauma, may follow many of the chronic inflammatory conditions which involve cartilage such as sarcoidosis, tuberculosis, polychondritis and syphilis. Malignant granuloma may also damage septal cartilage and lead to nasal to nasal dorsum collapse; some degree of saddling may also be a familial or racial characteristic.

When considering management of a nasal saddle, one’s first thought tends to be of a suitable graft material. It should be remembered, however, that many small saddles are accentuated by a nasal hump and simple removal of the hump suffices to solve the saddle defect resulting in a smaller nose which may be a bonus, particularly in the female patient. In some instances, it may be possible to remove a dorsal hump and use this to graft the saddle. With more severe saddling, however, an implant is required to restore an acceptable nasal contour. The problem is to select from the great number of alternatives the most acceptable and reliable long-term graft. When the saddle is due to loss of cartilaginous support, a ‘soft’ cartilaginous graft is preferable to the harder implants such as bone and synthetics. When the saddle defect involves the entire dorsum, that is both bone and cartilage, one of the harder implants is probably to be preferred.

Iliac crest bone grafts are still widely used and advocated in many standard texts, although their long-term survival is doubtful. When the saddling involves the bony skeleton of the nose, iliac crest grafts have a place but they are usually unsatisfactory for the cartilaginous saddle, producing a rather rigid and wide unnatural appearance. The donor site gives some considerable pain initially and produces a scar.

Autografts are probably the most useful graft material for the majority of nasal saddle defects. For the small saddles, sufficient cartilage may be obtained from the nasal septum. Usually, however, the cause of the saddle defect makes the likelihood of sufficient residual septal cartilage being available improbable. The bone of the septum from the vomer and ethmoid are thin and less satisfactory to fashion than the cartilage. Rib cartilage may be needed for the large saddle defects, but the harvesting risks of pneumothorax, postoperative pain and scar are among other disadvantages. Rib cartilages may twist, whatever manoeuvres are taken to avoid this troublesome long-term possibility.

Aural conchal cartilage is the most useful graft for the majority of saddle defects and can be obtained with minimal deformity, if necessary, from both ears. The shape of the conchal cartilage lends itself well to restoring good contour to saddle deformity. Although this cartilage gives many good results, it is a viable graft and change in shape and bulk may occur, so, as with all implants, it is not totally reliable. It is arguable whether this graft should be used for saddling related to chronic inflammatory diseases such as sarcoid, relapsing polychondritis or with malignant granuloma. It is not always possible to be certain that the disease is quiescent and the implanted graft may become involved in the inflammatory process and cause a complex deformity with the cartilage involved in the chronic inflammatory process.

Irradiated preserved cartilage is also advocated and impressive reports are available (McGlynn and Sharpe, 1981). Synthetic implants have a place in the management of nasal saddling although there are those who oppose their use with almost religious fervor. It is curious that, for chin augmentation, synthetic implants are almost universally used with little in the way of criticism. The thick overlying soft tissue present over the chin implant,
however, makes extrusion unlikely and conceals minor displacement. Synthetic implants underlying the nasal dorsal skin are close to the nasal cavity and to the exterior. Even with minimal infection, therefore, extrusion of the graft either externally or through a sinus inside the nose is possible. The firm synthetic implants may also move and their asymmetry of outline is obvious and aesthetically unacceptable. It is not true, however, to say that most synthetic nasal implants extrude. Long-term follow-up of cases show that this is not evident if certain precautions are taken (Mackay and Bull, 1983). An implant inserted through a midline vertical columella incision into a pocket underlying the dorsum of the nose with few or no other rhinoplasty steps undertaken concurrently will almost certainly remain in situ. Telangiectasia of the skin may occur and displacement from the midline position gives rise to asymmetry. The synthetic meshes, such as Supramid, also act as a satisfactory filling material in the nose and good results have been demonstrated (Beekhuis, 1975). These materials, however, cannot easily be fashioned or shaped. If infection does occur, the extrusion of solid implants results in a return to the status quo and surgical removal is also simple. With the mesh implants, however, complete removal may be difficult or impossible. The same problem of surgical removal arises with an infected bone or cartilage graft.

Firm synthetic implants are useful in saddling involving the nasal bones and cartilage and are particularly acceptable in some cases where a saddle appearance is an unwanted racial characteristic. With thin or scarred skin, synthetic implants are better avoided and any skin blanching overlying the implant at the time of insertion is to be avoided for the skin may later break down at this site.

There is probably no single ideal implant for all saddle deformities, but conchal cartilage grafts suffice and are possibly the most suitable graft at present to select for the moderate cartilaginous saddles. However, there is insufficient ear cartilage for gross saddle defects and, in this situation, a synthetic, rib or iliac crest graft is required.

**The deviated nose**

Correcting a deviated nose is one of the more difficult procedures in rhinoplasty. This is in part due to the fact that two-thirds of the nose is made up of cartilage which, unlike bone, does not 'stay put' but tends to spring back to its former position. In many cases, extensive septal surgery will be required which can considerably complicate any rhinoplasty procedure; lastly any inequality or asymmetry of the bony side walls may not be corrected by the usual medial and lateral osteotomies. Regrettably, although the procedures required to correct a deviation may be complex, this is seldom fully appreciated by the patient, whose attitude can often be summarized by the statement: "I don't want the shape changed, just straighten it!" As with all rhinoplasty surgery, it is well worth spending more time explaining the limitations than the expectations of any proposed surgery and this is particularly true of the deviated nose, when it should be carefully explained that no guarantee can be given that the nose will be perfectly straight but that certainly one hopes to achieve an improvement.

As long ago as 1845, Dieffenbach advocated division of the upper lateral nasal cartilages from the septum, the nose being held in its new position with bandages. In 1889, Trendelenburg was performing sophisticated procedures to correct the deviated nose by undertaking endonasal lateral osteotomies, percutaneous superior osteotomies and dividing the septum from the nasal crest using a fine osteotome. Joseph described a technique in 1907
whereby bilateral lateral osteotomies were undertaken in addition to removing a triangular wedge of bone from the 'long side'. A similar 'wedge-technique' together with an asymmetrical hump reduction was described by Foman in 1936 and later modified in 1960 (Foman, 1936, 1960). Cottle's description in 1960 of a high lateral osteotomy on the broad side and low osteotomy on the short side, together with a septoplasty and 'push down', although apparently effective, was criticized on the basis that it could lead to a 'step deformity'.

The techniques required to correct a deviated nose may be considered under four headings:

1. correction of the septum and upper lateral cartilages
2. dealing with deviation and asymmetry of the nasal bones
3. the nasal tip
4. augmentation, that is filling a depression to give an appearance of straightening the nose.

The last of these would not be suitable on its own if there is any functional element, but is a useful supplementary technique used in conjunction with the former three and may occasionally be all that is required to correct a purely aesthetic deformity.

**The nasal septum and upper lateral cartilages**

The septum is dealt with more fully in Chapter 10. When the septum, although deviated, is itself reasonably straight, that it, it may bend at one particular point only or it is straight within the nose but both nose and septum deviate to one side of the face, then the so-called 'septoplasty technique' can be usefully employed. A mucoperichondrial flap is elevated from both sides of the nasal septum or completely on one side with superior and inferior tunnels on the other. If mucoperichondrium is left attached to one side, it may reduce the chance of resorption of cartilage at a later date although, in practice, this seems rarely to occur. Leaving the flaps attached one side but not on the other, however, may lead to unequal scarring and later contracture which it is felt by some could lead to further deviation of the nose and septum at a later date. The septum is then separated from each upper lateral nasal cartilage above, divided at the 'bend' posteriorly, repositioned into the maxillary crest below, or the crest itself repositioned following an osteotomy if it is not in the midline and, finally, the septum repocketed anteriorly into a slot incised behind the columella.

Skin overlying the upper lateral nasal cartilages and nasal bones is elevated in the normal way via an intercartilaginous incision and, following detachment of the upper laterals from the septum, any inequality can be corrected by lowering the 'long' side and possibly even augmenting the 'short' side with a cartilage graft.

Septoplasty techniques are less satisfactory for dealing with deviations which are maximal at the valve area, where there is generalized 'ballooning' of the septum towards one side or where there is gross buckling and distortion of the nasal septum. In these cases, it may be necessary to undertake a submucosal resection of as large a portion of septal cartilage as possible which can then be repositioned and replaced as a free graft, or it may need to be completely replaced with an autograft from the ear or a homograft of banked septal cartilage. In these cases, there is a serious risk of supratip depression and saddling developing at a later
date and, when possible, it is better not to separate the upper lateral cartilage attachment to the septum. This will, however, make it impossible to correct any inequality of the upper laterals and more difficult to correct any deviation of the middle third of the nose. Very occasionally, it may be necessary to stage the procedure in these circumstances.

The septum may, in some instances, be dislocated to one side of the columella anteriorly, but the remaining septum may be in a satisfactory position. Providing there is adequate support for the columella, which can readily be checked by pressing a finger in the columella, then any displaced cartilage must be surplus to requirement and may be excised together with its covering mucous membranes, provided this is excised parallel with the septum to ensure that no useful cartilage is removed. If the cartilage is excised at a right-angle to the septum, too much caudal border may be removed leading to unsightly columella retraction.

**Deviation of the upper third - nasal bones**

Correction of the bony upper third should not in theory present too much of a problem, as it should be possible to refracture, manipulate and immobilize the nasal bones into any desired position. In practice, however, residual deviation in this area is not uncommon and this may, in many cases, be due to the fact that medial, lateral and superior osteotomies will not correct the deformity.

Trauma to one side of the nose will often result in a curved deformity and, if one considers the nasal bones individually, each can be considered as 'canoe-shaped' or 'banana-shaped' - medial, lateral and superior osteotomies will simply result in two fragments, each still with the same curved shape.

Tardy and Denneny (1984) describe the use of vertical intermediate osteotomies using a 2 or 3 mm osteotome endonasally. While this certainly results in greater mobilization of the fragments, it may still not be ideal because a transverse or horizontal intermediate osteotomy is more logical. This is difficult to perform endonasally, but can easily be performed using a percutaneous technique.

The nose is prepared in the normal way; a small stab incision is then made in the midpoint of the desired osteotomy. Through this single incision, the osteotome is then moved up and down along the proposed line scoring the periosteum, and then tapped through in three or four positions to complete the osteotomy. The same technique can be used for the lateral and superior osteotomies. A medial osteotomy is undertaken in the normal manner. Following this, the nasal bones are infractured or outfractured into the desired position. It is important to note that skin and mucoperiosteum must not be widely elevated as these are required to splint the relatively small fragments of nasal bones. Intermediate osteotomies, whether longitudinal or transverse, must be completed before any other osteotomies. There may be further comminution of the nasal bones, but this need not be a disadvantage and, indeed, was the basis of a technique described by Kazanjian and Converse (1972) who advocated covering the nose with cottonwool, protecting the eye with wadding and refracturing the nose with a strong blow from a mallet.
**Nasal tip**

The cartilage-splitting technique described above can at times be used to correct a tip deformity by an asymmetrical reduction, either removing more cartilage from one side than the other or reducing the tip only on the side to which the nose deviates. Occasionally, a tip can be reduced and the cartilage removed used to augment the other side.

Where the tip not only deviates to one side but also requires projection, the technique described by Goldman may be useful. A rim incision is performed and the overlying skin elevated. Skin and cartilage are then incised at a point some 2 mm lateral to the dome; the two sides are then sutured back-to-back in such a way that they can not only project the tip but also twist it back to the midline.

**Augmentation**

In addition to augmentation of the tip, as described above, it may at times also be useful to consider augmentation on the concave side of the nose over the upper lateral cartilage and nasal bones. It is always wise to consider the need for this carefully at the preoperative planning stage as, following infiltration and elevation of the skin together with a certain amount of inevitable oedema of the tissues during surgery, the need for augmentation after undertaking various other manoeuvres can be difficult to visualize. One can frequently be disappointed with long-term results in a case where a graft was considered preoperatively, but deemed to be unnecessary at the time of surgery.

Autografts are preferable to homografts and it is usually not difficult to find a suitable piece of cartilage. It is often necessary to remove cartilage from the nasal septum while correcting a deviated nose, and this makes ideal graft material. Where this is not available, however, conchal cartilage from the ear is equally suitable.

Whenever possible, these grafts should be positioned into a pocket which has been fashioned to exact size of the graft to prevent the latter being displaced. While this is highly desirable, it is not always practical; for example, if it has been necessary to elevate the skin over the dorsum to remove a hump, it is then impossible to fashion any pocket in the supratip region. In these circumstances, it is possible to secure the graft with 4-0 plain catgut sutured through the skin and secured with Steri-strip.

**Deviation associated with gross deformity of the nose**

Where there is gross deviation, particularly if this is associated with severe septal deformity, as with the cleft-lip nose, an external rhinoplasty approach should be considered. This allows excellent exposure of the septum, together with direct visualization of the nasal anatomy. It is, however, simply an approach and does not in itself offer an advantage other than improved exposure and access. The principles outlined above will still need to be followed in order to correct the deformity.
Alar collapse

Alar collapse is the phenomenon whereby the alae nasi collapse inwards on inspiration in a valve-like manner, causing nasal obstruction. It is most common in elderly patients with a drooping tip, loss of elasticity of the cartilage and atrophy of the dilator muscles. It may occur after rhinoplasty, if too much lower lateral nasal cartilage has been excised. Loss of vestibular skin may result in gross scarring and contracture in the valve area.

Conductance is proportional to the fourth power of the diameter of a vessel and a very small change in the width of the nostril will markedly affect the flow. As air passes inwards through the nose, it will produce a Venturi effect, tending to draw the nostrils inwards, as this occurs, the resistance will increase tending to cause further indrawing until the nostrils eventually close off completely. In the normal subject, this will occur if inspiration through the nose is sufficiently forceful and a diagnosis of alar collapse should be reserved for those patients unable to achieve inspiration at physiological flow rates.

While a relatively small decrease in the diameter of the airway may greatly increase resistance, the converse is also true and a relatively minor alteration of the nasal septum in the valve area or narrowing of the columella may be sufficient to prevent the initiation of this phenomenon. One can test the relative significance of a wide columella by gently squeezing it between the two limbs of a Thudicum speculum. If this improves situation, it is a simple matter to excise an ellipse of skin to include the herniating portion of the medial crus of the lower lateral cartilage. The skin is then gently undermined and closed with sutures.

In addition to narrowing caused by the medial crus, the lateral crus may at times herniate inwards, causing obstruction; this cannot be corrected by simple excision, however, but needs to be dissected free and repositioned into a new pocket slightly more lateral and superior to its former position.

Apart from these supplementary techniques, the methods used to correct this problem fall into three groups:

1. prostheses
2. modification of existing cartilage
3. grafting additional material.

Prosthetic devices

Many prostheses have been tried over the years from quills and reeds to the silver wire alae nasi dilator used by Clement Francis at The Metropolitan Ear, Nose and Throat Hospital. Not one of them has, however, found lasting favour. More recently, Davenport, Brain and Hunt (1981) have reported greater success using acrylic materials. A mould is made with silicone putty which is then cast in clear acrylic resin through which a hole is drilled to provide an airway.
Modification of existing cartilage

In some cases, alar collapse is due to slit-like narrowsness between the septum and the upper lateral nasal cartilages. Walter (1976b) has described a technique whereby the valve can be enlarged by dividing the upper laterals from the septum and covering the defect with a flap from the upper border of the lower lateral cartilage.

The skin is elevated over the upper lateral nasal cartilages which are then divided, together with the underlying mucosa, from the septum on both sides. A flap of cartilage and vestibular skin is then fashioned, based on the dome and retaining a 2-3 mm strip of cartilage along the lower border undisturbed. This flap is then rotated upwards to fill the defect between the septum and lower lateral cartilages.

Rettinger and Masing (1981) noted that the medial crus and lateral crus of the lower lateral cartilages lie in the same plane, in many patients with alar collapse, and this is particularly true in the elderly patient with a drooping tip. They also observed the tension lines in a plastic model under polarized light. By rotating the two limbs of the model in opposite directions, they noticed that these tension lines were distributed over a larger surface increasing the stability of the system. These principles were applied to the problem and the lateral border of the lower lateral cartilage was rotated upwards into a more cephalic position.

A rim incision is performed and the lateral crus dissected free of both overlying skin and underlying vestibular skin until the whole of the lateral crus remains attached only at the dome. A pocket is developed above the upper lateral and sesamoid cartilages and the lateral crus rotated upwards by means of a traction suture. The cartilage can then be maintained in its new position either with a mattress suture tied over Teflon foil or with fibrin glue.

Rettinger and Masing reported an initial success rate of 18 out of 19 patients with a minimum follow-up period of 6 months and pre- and postoperative rhinomanometry confirmed this improvement in the four patients in whom this was performed.

Cartilage grafts

In some cases, there is insufficient cartilage to reposition; this is particularly true following excessive excision of cartilage during rhinoplasty. In other cases, although present, the cartilage appears to have atrophied and lost all its elasticity such that repositioning is unlikely to be beneficial. In these cases, it is necessary graft new cartilage either from the septum or from the conchal cartilage of the ear, which is probably better as the latter is suitably curved and elastic. Where support alone is needed, a cartilage graft will suffice; if vestibular lining is needed in addition to support, a composite graft will be required.

Cartilage graft

An ellipse of cartilage is taken from the concha of the ear and inserted into a pocket prepared at the site where support is lacking. A small incision is made in the vestibular skin and a pocket fashioned with curved pointed scissors - the size of which should exactly match that of the graft to prevent the graft wandering into the wrong position. The pocket is then closed with a few fine sutures.
Composite cartilage graft

Alar collapse with stenosis may follow rhinoplasty, although it should not occur if the principles outlined here are followed; in particular, vestibular skin should not be excised. In addition to surgery, other trauma and in particular burns, sometimes following cautery for epistaxis, may result in this deformity.

Aural conchal composite grafts offer the most effective method to correct this difficult problem. Although composite grafts were described many years ago (Konig, 1902), it is due to Claus Walter's more recent innovative work that the wider application of these grafts has been developed (Walter, 1976a).

For the more minimal stenosis involving the apex of the vestibule alone, two separate elliptical composite grafts may be used. The vestibular skin is incised and undermined, on either side, but no skin is removed; the composite graft is then sutured into the defect. When stenosis is associated with saddling, a large composite graft can be used, with a small strip of skin removed from the midline of the graft where it 'bridges' the septum. This can provide cartilage to fill the saddle, cartilage to prevent alar collapse and skin to replace the stenosed vestibule as a simple procedure. The external rhinoplasty approach may facilitate the securing of these grafts.

External rhinoplasty

The standard rhinoplasty techniques, evolved from Joseph's and Roes' original operations, involve no external incisions on the nose or face. The technique is, however, to some extent blind and certainly placing intranasal sutures or accurate securing of grafts is not always easy via the standard intranasal incisions of rhinoplasty.

Rethi (1956) demonstrated that an excellent exposure and access to all the nasal structures could be achieved with elevation of the nasal skin via a transverse columella incision linking the cartilage rim incisions. The transverse incision across the columella is, however, invariably ugly and conspicuous and almost impossible to revise effectively. In 1974, Goodman of Toronto demonstrated that an inverted 'V'-shaped incision resulted in an almost imperceptible scar. Vertical columellar incisions are barely perceptible whereas transverse incisions are very obvious.

This small change in the incision led to a resurgence of interest in the external or 'open' rhinoplasty operation particularly in the USA, where it is now widely practised. It is not possible at present to say how acceptable this technique will become for routine rhinoplasty, for the standard technique is so well tried and proven that it will be difficult to displace. In certain gross deformities, marked deviations and revision techniques, the external rhinoplasty has the advantage of demonstrating the anatomy clearly so that correction can be better controlled and sutures accurately placed. For the insertion of composite or intranasal grafts and possibly access for repair of a septal perforation, the external rhinoplasty approach should be considered. It also forms an excellent access for excision, with minimal scarring, for lesions and swellings under the nasal dorsum which may hitherto have been approached by a direct incision through nasal skin.
Dressings, splints and plasters

Prior to the application of any dressings, one should ensure that any cartilage-splitting incisions have been sutured and that mucoperichondrial flaps are replaced and secured. It is not usually necessary to suture the intercartilaginous incisions.

Splints

Silastic splints may be required to immobilize the nasal septum and help to maintain this in the midline. These should be thick enough to provide rigid support, of sufficient length to support the entire length of the septum, but should not be too wide as they may otherwise cause pain due to pressure between the septum and the middle turbinate. Splints must be secured with a transfixion suture securing the two sides and not sutured one to the other across the front of the columella, as this is not only annoying to the patient but may cut into their skin leading to a permanent scar which is difficult to correct.

It is normally an easy matter to cut the suture and slide the splints out approximately one week later.

Packs

The nose will normally need to be lightly packed with a double layer of Telfa dressing, or a small vaseline pack, although the latter should be applied carefully as any vaseline entering the wound may cause a granuloma. Packing is removed at 12-24 hours.

The nose is strapped with 0.5 inch (13 mm) Steri-strips, moulding the nose in the desired position ensuring that the tip is held well up, approximating the transfixion incision and keeping the tip of the nose suitably pinched in. It is important to keep the supratip well strapped down to prevent any haematoma forming which may contribute to a pollybeak. One should avoid leaving any small area of skin unstrapped as it is possible for this to herniate through, leading to skin necrosis and ugly scarring.

External fixation

Many different materials have been used; plastics have the advantage that they are light and easy to keep clean; various designs of metal splints are available which are easy to apply but may expand with swelling of the nose, leading to a loose fit unless they are squeezed tight again a few days postoperatively. Plaster of Paris, although one of the oldest, remains popular with many surgeons and the authors use a splint cut from 6-8 layers of plaster of Paris bandages. This is dipped into warm water, squeezed out in gauze, applied over the nose and secured with a further layer of strapping.

Whether or not this external splint should cross the nasion up onto the forehead, remains debatable. Some feel that, as there may be postoperative oedema and swelling at the root of the nose, the plaster can be lifted up in such a way that there is no longer adequate support of the nasal bones where it is most needed. Conversely, it can be difficult to secure the upper end of a splint which is applied to the nose alone, and continuing the plaster onto the forehead not only facilitates this but may help to keep the nose in the midline. One
compromise is to apply a half-moon-shaped splint of 6-8 layers of plaster to the nose with an additional spur of 6-8 layers placed along the dorsum onto the forehead leaving a small gap under the splint at the nasion.

**Bolster**

Half a gauze eye-pad strapped under the nose makes a suitable bolster; this can be changed as necessary until the internal packing is removed the following day, whereafter bolsters should be discouraged and as much air as possible should be allowed to get to and, if possible, up the nose.

**Postoperative care**

**Recovery**

If surgery is performed under general anaesthesia, a great deal is asked of the anaesthetist. Not only should the patient remain totally unconscious throughout the period of surgery but no sooner is the surgeon finished, the patient is required to be completely awake, conscious and cooperative. If too drowsy, they may well inhale blood, particularly when nursed in the optimal head-up position, itself aimed at reducing bleeding and swelling. If confused or thrashing about, the patients may displace the dressing and splints or this may occur as a result of attempting to restrain them. Fortunately, this surgery is not associated with much pain, this being particularly true if the nose is infiltrated with vasoconstrictors combined with local anaesthetic agents (for example lignocaine). Large doses of postoperative analgesia are therefore not required and, combined with careful preoperative medication and modern general anaesthetic agents, the ideal requirements can be achieved. Normal recovery procedures should be observed and, once fully conscious, the patient is transferred to the ward.

**Ward**

On the ward, the patient should be nursed in the sitting position for as much time as possible. Temperature, blood pressure and pulse should be observed as well as other signs of bleeding, that is swallowing, although severe bleeding at this stage is very rare.

**Antibiotics**

Routine prophylactic antibiotics are not required; they should be considered, however, in patients with splints which are to be left in situ for one week and in any case in which implants have been used, whether these be allografts, autografts or homografts. The commonest infecting organism is Staphylococcus sp. and, since this is very often penicillin resistant, an antibiotic which is specific for this should be prescribed.

**Complications of rhinoplasty**

Complications and problems arise with rhinoplasty when the standard techniques are carried out to excess or incorrectly. Excess removal of tissue or over-correction at the time of rhinoplasty is the commonest cause of the now well-recognized unnatural postoperative
rhinoplasty appearance. A nose with a natural non-operated appearance is the aim of rhinoplasty; surgical judgement along with a clear understanding of what the patient wants and expects are essential preoperative requisites.

A small removal of bony hump is usually all that is required and either a saw with a narrow single (rather than wide double-cutting) edge or an osteotome is the correct instrument. The wide saw encourages the surgical removal of an excess of tissue and this, coupled with too little excision of the lower cartilaginous portion, is the commonest cause of the pollybeak appearance. If an excessive portion of nasal bone is removed, and then to achieve alignment an equally large portion of septal or upper lateral cartilage is removed from the nasal dorsum, a ski-slope appearance of the nose results. These excessive changes are particularly unsatisfactory in the more elderly patient where the skin may not take up, and where gross change in nasal contour is to be avoided as it frequently appears unnatural.

Excision of the caudal angle of the septum is a standard step in rhinoplasty but it is unnecessary if the nasolabial angle is preoperatively over 90-100°. An excessively 'open' nasolabial angle produces a 'pig-snout' appearance and many potential rhinoplasty patients are now aware of this surgical appearance and will make a specific request for this to be avoided.

Excessive removal of lateral crus of the alar cartilage has already been mentioned as a step predisposing to pinching with collapse of the lateral wall of the nasal vestibule, resulting in a 'knock-kneed' nasal tip appearance.

Failure to obtain a good lateral osteotomy of the nasal bones with infracture results in a wide nasal bridge which is sometimes termed the 'open-roofed' deformity. The saw is the most ineffective and traumatic instrument for the lateral osteotomy and its use predisposes to this deformity: an osteotome is to be preferred.

If asymmetrical excision of the alar cartilage is carried out and the rim of the cartilage remaining is too narrow, particularly near the dome, 'bossing' or asymmetry of the nasal tip becomes obvious. If the medial and lateral crura are divided when the overlying nasal tip skin is thin, irregularities or highlights will show. Asymmetrical division of the cartilages, or division on one side only, is particularly likely to predispose to this unsatisfactory appearance. Morcelizing techniques of the tip cartilages are also at risk of producing tip irregularities with thin overlying skin.

Intranasal complications of rhinoplasty also occur. The standard rhinoplasty technique correctly carried out does not prejudice the nasal airway. Over 500 patients assessed postoperatively at The Royal National Throat, Nose and Ear Hospital had no complaint of nasal obstruction following rhinoplasty. Nonetheless, vestibular stenosis following revision rhinoplasty or excessive incisions inside the nasal vestibule, particularly if coupled with excision of vestibular skin, will lead to stenosis and narrowing of the airway which may be very severe.

Failure to excise the caudal portion of the upper lateral cartilage may result in a projection of this cartilage into the vestibule with an area of vestibulitis.
Synechiae should not occur but trauma to the lateral nasal wall skin and mucosa, if coupled with damage to the septal mucosa, may predispose to this and careful use of instruments is a relevant factor. Excessive lowering of the upper lateral cartilage along its dorsal margin also predisposes to webbing of the nasal valve with nasal obstruction.

Rhinoplasty and septoplasty, however, are operations which, if correctly applied, give excellent results achieving both an improvement in nasal airway and an acceptable and natural improvement in the external nasal shape.
Chapter 16: Epistaxis

O. H. Shaheen

History

Epistaxis is mentioned in medical literature dating back to very early times. Hippocrates (fifth century BC) was probably the first to appreciate that pressure on the alae nasi was an effective method of controlling nose bleeds, although in some cases he resorted to nasal packing and the application of cold fomentations to the shaved head. He regarded the complaint as being primarily of young persons, and was the first to describe vicarious menstruation.

Ali Ibn Rabban Al-Tabiri (AD 850) devoted a chapter of his massive work *The Paradise of Wisdome* to epistaxis. In it he wrote: 'The complaint of nose bleeding is due to swelling of a vein and its rupture, or perhaps a reduction in the force which confines the blood within.' He implied that some of the medications inserted into the nose owed their efficacy as much to their temperature as to their pharmacological properties.

Morgagni (1769) recognized 'the extremely turgid blood vessels about that part where the alae nasi are formed with the bone, about a finger's breadth more or less from the bottom of the nostril'. He was reported to have stopped nose bleeds by introducing his finger and 'pressing that part whereupon the blood ceased to flow, so that it was not even discharged by the posterior nostril into the fauces'. Morgagni drew his inspiration from his former teacher Valsalva and for this reason Little's area is referred to as 'Locus Valsalvae' in Italian circles. Morgagni's records also contain the suggestion, previously entertained by Valsalva, that nasal haemorrhage might be arterial in origin for it was his practice to 'syringe the nose with cold water and to apply the spirit of wine, especially to contract the mouths of swollen arteries'.

Mahomed (1880-1881) who pioneered the development of the sphygmomanometer stated that 'the frequency with which severe epistaxis occurs in old people with high arterial pressure is striking and for them very fortunate for if their noses did not bleed their brains would'. In 1879, Little published his case reports in the *Hospital Gazette* (Rainey, 1952) in which he identified the site of bleeding as being at the caudal end of the septum, and a year later Kiesselbach made similar observations. However, even after the introduction of modern histological methods, investigations into the mechanism of epistaxis were few and relatively uninformative, so that until recently very little was known about the pathology of nasal blood vessels.

The first attempts at arterial ligation were in 1868 (Bartlett and McKittrick, 1917) when Pilz of Breslau tied the common carotid artery, and it was much later that external carotid ligations were performed for the control of nose bleeds. Seiffert (1928) introduced ligation of the internal maxillary artery via a transantral approach and Goodyear (1937) was the first to tie the anterior ethmoidal artery.
Vascular anatomy of the nose

Textbook descriptions of the vascular anatomy of the human nose are based on Zuckerkandl's original and comprehensive studies of the subject (1982). The nose is vascularized by the internal and external carotid arteries via their respective branches, there being a confluence of the two systems, particularly at the caudal end of the septum where a number of arteries anastomose with each other (Little's area).

With the exception of Little's area, the middle turbinate has for a long time been regarded by clinicians as the dividing line between the internal and external carotid distributions, with a corresponding imaginary line of demarcation at the same level on the nasal septum (Weddell et al, 1946). This landmark has served as a guide in deciding which of the two areas is responsible for the epistaxis, and has allegedly helped the surgeon to decide which artery to ligate in severe cases of epistaxis.

The dividing line between the two carotid distributions may not, however, coincide exactly with the level of the middle turbinate. The work of Zuckerkandl (1892) and Burnham (1935) indicates that the blood supply to the turbinate is derived exclusively from the external carotid artery and that anastomosis between the two carotid distributions occurs above and anterior to its attachment to the lateral nasal wall, and not within it. They also described an artery to the superior turbinate and meatus, with a corresponding vessel on the septum, both of which originate from the nasopalatine branch of the sphenopalatine artery (external carotid).

Shaheen (1967) confirmed the presence of a branch from the nasopalatine artery supplying the superior meatus, turbinate and corresponding septum by X-raying the excised nasal fossae of cadavers which had been previously injected with barium-gelatin mixtures. It would therefore seem that the area designated as receiving blood from the internal carotid artery is smaller than previously supposed. Certainly the gross disproportion between the diameters of the anterior ethmoidal artery and the sphenopalatine at their points of entry into the nose would corroborate this view. The surgeon who lacerates the anterior ethmoidal artery in an external ethmoidectomy rarely has difficulty with haemorrhage; similarly those who deliberately ligate this vessel for epistaxis are always impressed by its small size. By contrast the terminal segment of the internal maxillary artery is a much larger vessel. The calibre of the posterior ethmoidal artery is also small, so that its contribution to the nasal blood supply is unlikely to be significant even if it varies reciprocally in size with the anterior ethmoidal vessel as suggested by Batson (1935).

It is noteworthy that the anterior ethmoidal artery was found to be absent unilaterally in 14% of cadaver dissections, and bilaterally in 2.5% of cases, the canal being either imperforate or filled with fibrous tissues or nerves (Shaheen, 1967).

This supports the contention that these vessels contribute very little to the blood supply of the nose, even if a somewhat larger posterior ethmoidal artery is found doubling for the missing anterior vessel and running a similar course to it, as sometimes happens. In this connection, the surgeon who sets out to ligate the ethmoidal vessels should be aware that, when the anterior vessel is missing, the posterior ethmoidal artery may arise directly from the circle of Willis and may, therefore, not be encountered in the orbit at all. This arrangement
conforms much more to the state of affairs in early embryonic life when the posterior ethmoidal artery is the dominant vessel of the nose, dwarfing not only the anterior ethmoidal artery but the nasopalatine vessel as well (Shaheen, 1967).

Burnham (1935) in his description of the anatomy of the lateral nasal wall claimed that the arteries to the inferior and middle turbinates and their respective branches lay partly embedded in the bone of these structures. In the case of the inferior turbinate, he found the bony canals containing the branches of the inferior turbinate artery extended along the central three-fifths of the bone. The middle turbinate artery and its branches were protected by a bony covering in the posterior half of the concha. Thus a considerable segment of both of these arteries and their branches is unlikely to give rise to epistaxis even if rupture occurs. By the time the arteries have emerged from their bony channels to lie beneath the mucous membrane, they will have diminished considerably in size.

Ogura and Senturia (1949) found in a series of patients with epistaxis that the bleeding point arose on the lateral wall in 28 out of 88 cases, and other authors have similarly implicated the lateral wall as a common site for bleeding. Shaheen (1967), on the other hand, was unable to find any cases of bleeding from the lateral wall of the nose in 117 cases, and his anatomical dissections and serial sections of the nose confirmed the findings by Burnham (1935).

The vast majority of patients who suffer from arterial epistaxis bleed from the nasal septum, and chiefly from the area where anastomosis of the nasopalatine, greater palatine, anterior ethmoidal, and coronary arteries takes place. This plexus was originally described by James Little and it is important to note that bleeding from it is arterial in origin, and not venous as some reports suggest. The venous bleeding, which is common in young persons, arises from the vein which lies immediately behind the columella at the anterior edge of Little's area. It runs vertically downwards and crosses the floor of the nose obliquely before joining the venous plexus on the lateral wall of the nose.

The dynamics of the nasal circulation depend to a large extent on the presence of arterioarterial anastomoses between the various arteries which contribute to the vascular supply of the nose. The branches of the anterior and posterior ethmoidal arteries join in a series of arcades in the upper one-third of the nose and the branches of the sphenopalatine artery anastomose with those of the ethmoidal arteries above the level of the middle turbinate. Opposing heads of pressure meet in the anastomoses with a sharp interface between the two, which can be displaced by dropping the pressure in one or other of the opposing systems. Shaheen (1967) demonstrated, by means of dye injections into the carotid vessels of live humans, that the dispersion of dye in the nasal mucous membrane could be affected by dropping the pressure in the system not being injected. For instance, dye injected into the internal carotid artery failed to appear in the nose, confirming the poor circulation of the ethmoidal vessels, but when the external carotid was occluded at the time of injection the entire upper half of the nose was suffused with dye from above downwards. The rapidity with which such dye displacement takes place, confirms the importance of the arterioarterial anastomoses in the nose.
The importance of possible anastomoses across the midline must also not be overlooked, either at the nasopharyngeal end or between the two anterior ethmoidal arteries at the crista galli.

These observations could well explain the many documented reports of failed ligation in which surgeons assumed, probably incorrectly, that they had tied the wrong vessel simply because bleeding has not stopped after ligation.

The arteriovenous anastomoses which are present at the anterior end of the inferior turbinate and septum at a microscopical level are probably of little importance in the aetiology and persistence of epistaxis, but their precise role is as yet far from clear.

Clinical manifestations

The prevalence of epistaxis in random samples of the population was found in one study to be between 10 and 12% (Shaheen, 1967). The age distribution shows an increase in frequency between the ages of 15 and 25 years, and later from 45 to 65 years, but with little difference between the sexes.

In only a small number of cases can epistaxis be attributed to a well-defined primary cause, such as a blood dyscrasia, a blood vessel abnormality, or local nasal pathology. In the majority of cases bleeding arises from an artery or a vein without any obvious abnormality to account for it; hence the terms 'spontaneous' or 'idiopathic epistaxis' which have been coined to cover this, the commonest category of epistaxis.

Certain contributory factors may be implicated in the onset of bleeding in cases of so-called 'spontaneous epistaxis', such as nose blowing, sneezing, coughing, straining, pregnancy, coryza and sinusitis. They all share one thing in common, namely a sudden rise in vascular pressure.

Venous epistaxis from the retrocolumellar vein tends to occur in subjects under the age of 35, whereas arterial epistaxis occurs in the older age groups. The duration of bleeding, as might be expected is short-lived in venous epistaxis, and quite prolonged in bleeding of arterial origin (Table 16.1).

Table 16.1 The striking difference in duration between venous and arterial epistaxis

<table>
<thead>
<tr>
<th></th>
<th>10 min or under</th>
<th>Over 10 min and under 2 h</th>
<th>Over 24 h</th>
</tr>
</thead>
<tbody>
<tr>
<td>Subjects over 35 years</td>
<td>10</td>
<td>18</td>
<td>44</td>
</tr>
<tr>
<td>Subjects under 35 years</td>
<td>28</td>
<td>13</td>
<td>4</td>
</tr>
</tbody>
</table>

Furthermore, there is an inverse relationship between the frequency and duration of epistaxis, the more severe arterial haemorrhages recurring rarely more than once or twice. No correlation can be established between the prevalence of epistaxis in random samples of the population, and their blood pressure status, although there is some correlation between the
severity of epistaxis and the degree of vessel wall disease as judged by retinoscopy (Shaheen, 1967). The finding of a high proportion of subjects with high blood pressures in hospital practice (Table 16.2) signifies, not that hypertension causes epistaxis, but rather that patients with higher blood pressures have more severe or persistent bleeding and are therefore eligible for hospitalization.

Table 16.2 The blood pressure distribution of subjects attending hospital with epistaxis. Pressure adjusted to a standard reference age for both sexes

<table>
<thead>
<tr>
<th>Diastolic pressures (mmHg)</th>
<th>Number of subjects</th>
</tr>
</thead>
<tbody>
<tr>
<td>65</td>
<td>2</td>
</tr>
<tr>
<td>70</td>
<td>0</td>
</tr>
<tr>
<td>75</td>
<td>3</td>
</tr>
<tr>
<td>80</td>
<td>7</td>
</tr>
<tr>
<td>85</td>
<td>7</td>
</tr>
<tr>
<td>90</td>
<td>5</td>
</tr>
<tr>
<td>95</td>
<td>8</td>
</tr>
<tr>
<td>100</td>
<td>10</td>
</tr>
<tr>
<td>105</td>
<td>7</td>
</tr>
<tr>
<td>110</td>
<td>5</td>
</tr>
<tr>
<td>115</td>
<td>6</td>
</tr>
<tr>
<td>120</td>
<td>6</td>
</tr>
<tr>
<td>125</td>
<td>3</td>
</tr>
<tr>
<td>130</td>
<td>1</td>
</tr>
<tr>
<td>135</td>
<td>0</td>
</tr>
<tr>
<td>140</td>
<td>2</td>
</tr>
</tbody>
</table>

The pathology of nasal arteries

Examination of the medium and smaller nasal arteries of persons dying in middle and old age has shown that these are subject to a progressive replacement of the muscle tissue in the tunica media by collagen (Shaheen, 1967). This change varies from interstitial fibrosis to almost complete replacement of the muscle by scar tissue. It seems that persons giving a history of epistaxis exhibited the more severe changes, but this is not to say that these changes are necessarily responsible for vessel rupture. They could, however, account for the lengthy duration of arterial haemorrhages, presumably because of a failure of the vessel to contract down in the absence of sufficient muscle in the tunica media.

It is also apparent that larger vessels of the calibre of the maxillary artery are prone to calcification (Mönkenberg’s sclerosis). The resulting lack of elasticity could well contribute to the pathogenesis of small vessel rupture by the creation of a local systolic hypertension.

The precise mechanism of bleeding is thought to be a dissecting aneurysm of the nasopalatine artery or one of its branches, but the factors initiating the process have, so far, not been identified.
It is also a mystery why bleeding should occur from the retrocolumellar vein in young subjects. Careful inspection of the site shortly after a bleed sometimes reveals a tiny area of local ballooning overlying the vein, and this could possibly signify an area of vessel wall weakening, perhaps as a result of localized ischaemia.

**Clinical management of spontaneous epistaxis**

*The young person with recurrent bleeding*

After taking a careful history to establish that bleeding is not secondary to systemic disease, the nose is examined for signs of recent bleeding and for local abnormalities. In the absence of any obvious local disease, attention is turned to the septum which will often reveal an engorged vein at the anterior end of Little's area just behind the columella. If bleeding has been quite recent, a microaneurysm may be seen in the mucosa overlying the vein. Topical anaesthesia with 5% cocaine followed by cauterization with a silver nitrate stick will suffice to control most cases of epistaxis.

Some cases are particularly obstinate and require more than one application of a caustic agent, in which case it may be more effective to use trichloroacetic acid. Great care must be taken to ensure that none of the acid comes into contact with the nasal vestibule, as this will leave a particularly painful burn. There are some patients who bleed in spite of seemingly adequate attempts at cauterization, and the best policy is to coagulate the offending vessel with diathermy under general anaesthesia. Galvanocautery under local anaesthesia is not to be recommended in children, and even adults find the experience unpleasant. The sight of the heated filament, the sensation of heat within the nose, and the smell of charred flesh are off-putting to all but the hardiest individual.

**Treatment of epistaxis in the young**

Pinching the nostrils is the time-honoured method of stopping venous bleeding from the caudal end of the septum. Once bleeding has ceased, the nose can be cocainized and cauterized, although the vessel may bleed during the process of applying the caustic agent. Perseverance is required until bleeding finally stops.

**Treatment of recurrent epistaxis in older subjects**

Epistaxis in older persons does not recur with the same frequency as it does in younger people. Some patients only have the one major bleed, and when examined afterwards there may be very little to see. In such cases, there is nothing to be gained by cauterizing Little's area, unless it is certain that bleeding previously originated from this part of the septum.

Assessment of the cardiovascular system is important, however, and the patient should be referred to a physician if any abnormality such as hypertension is discovered. This is not so much to prevent further epistaxis, as to protect the individual from the harmful effects of the raised blood pressure.
Management of epistaxis in the elderly

Observations of the patient's pulse, blood pressure, and general condition are made in order to gauge the extent of blood loss. Estimation of the packed-cell volume in conjunction with the haemoglobin will also guide the clinician as to the need for replacing lost blood. At the same time the bleeding and clotting times and platelet count should be investigated to exclude a blood dyscrasia.

The nose is examined, preferably with the patient sitting upright in a chair. A plastic cover is draped around the neck and a bowl placed in the hands. Inspection of the nose may show a spurting blood vessel on the nasal septum, but usually the site of bleeding is smothered in blood. Cocainizing the nose with a 5% spray serves two purposes, namely to allow the introduction of a catheter so that the blood can be sucked away, and to stop bleeding by vasoconstriction. This gives the examiner the opportunity of locating the site of bleeding, but if this has stopped, suspect areas can be gently rubbed with orange sticks loaded with cotton wool in an attempt to cause further bleeding.

The bleeding point is cauterized if accessible, but quite frequently it is situated far back on the septum or behind a spur so that cauterization is technically impossible.

If the bleeding persists, the nose should be packed, preferably with ribbon gauze medicated with a suitable antiseptic such as bismuth-iodine-paraffine (BIPP). This can be left undisturbed for several days without fear of the patient developing complications. The old-fashioned method of controlling epistaxis by sitting the patient up with a cork between his teeth (Trotter's method) and allowing him to bleed until he becomes hypotensive is to be deprecated. Death from coronary thrombosis secondary to hypotension is a well-recognized complication of epistaxis and regularly appears in the Registrar-General's mortality statistics. It must be emphasized that old patients with poor hearts and circulations do not tolerate severe and prolonged blood loss.

Some clinicians prefer to insert an inflatable balloon in preference to ribbon gauze which is abrasive to the nasal mucosa. However, inflatable balloons will not adapt themselves readily to the irregular contours of the nose, and are therefore less reliable in the control of epistaxis.

After two to three days the packing can be removed, and in most instances bleeding will have stopped completely. If recurrent bleeding occurs, however, the pack will have to be re-inserted. Patients in this category, together with those suffering from prolonged epistaxis and those who are generally unfit, should always be hospitalized. They should be nursed sitting up and sedated to allay their anxiety and lower their blood pressure. The choice of sedative is a matter of individual preference, but opiates such as Omnopon (morphine) are popular, although they may sometimes cause vomiting. Diazepam by injection is also effective. If bleeding persists in spite of adequate packing, serious consideration must be given to the need for arterial ligation. There is some evidence to suggest that submucous resection of the septum may be helpful when bleeding originates from behind a prominent septal spur.
Postnasal packing may be helpful in those cases where anterior packing alone has failed to control the bleeding. It is particularly indicated in the control of haemorrhage following adenoidecsection. It is normally undertaken under general anaesthesia but can be accomplished under local anaesthetic in the cooperative patient. A small catheter is passed through each nostril from anteriorly backwards into the oropharynx. These are then drawn out through the mouth, and tied to two tapes which are secured to the pack. The catheters and attached tapes are then pulled forward through the nose, and tied across a bolster or dental roll which protects the columella. A piece of thread, previously attached to the lower edge of the pack is brought out through the mouth and secured to the cheek with adhesive tape. The anterior nasal cavities can then be packed with BIPP. The patient should be covered with a suitable broad-spectrum antibiotic and, after the bleeding has been controlled for a few days, the anterior packing removed. After cutting the tapes knotted across the columella, the pack can be removed through the open mouth, by pulling downwards on the lower central thread.

**Arterial ligation**

The patient who continues to bleed every time the pack is removed or who keeps on bleeding with the pack in situ will generally have to be transfused. If, over a period of 4-5 days, bleeding has not stopped, arterial ligation should be performed. In the absence of definite knowledge about the whereabouts of the bleeding point, it is reasonable to interrupt the external carotid system, since this supplies as much as 90% of the nasal mucosa. Bleeding from the ethmoidal region is in fact very uncommon and is rarely of a severity to merit arterial ligation, in spite of the occasional report describing severe ethmoidal bleeding.

Although interruption of the internal maxillary artery has become fashionable, it is by no means certain that this is necessarily more effective than ligation of the external carotid artery. Being nearer to the source of bleeding, the drop in blood pressure in that part of the nose supplied by the maxillary artery is greater than after tying the external carotid. From this, dividing the maxillary artery should, in theory, be more effective, but, as mentioned previously, the drop in pressure almost certainly encourages the displacement of blood from other areas of the nose through arterial anastomoses, with the possibility of bleeding persisting. Pearson, Mackenzie and Goodman (1969) also pointed out that continued haemorrhage after maxillary artery ligation could result from retrograde blood flow by way of arterio-arterial anastomoses between branches of the maxillary artery in the pterygopalatine fossa. Such distortions of flow would by-pass the interrupted segment of the maxillary artery thereby contributing to the persistence of haemorrhage. The descending palatine artery was particularly singled out by them as being a possible source of retrograde bleeding into the final portion of the maxillary artery, the moral being that not only should the main trunk be ligated as close to the nose as possible, but that as many branches as possible should be interrupted. By contrast, ligation of the external carotid artery does not produce quite the same blood pressure reduction distally and is, therefore, less effective in controlling bleeding, although blood flow from the ethmoidal to the nasopalatine areas may, in fact, be less. If bleeding persists after ligation of the maxillary artery, it is logical to proceed to interruption of the anterior ethmoidal artery with the prospect of arresting the bleeding permanently. The addition of ethmoidal artery ligation to external carotid ligation for persistent haemorrhage is less likely to be as effective, since the cause of the persistent bleeding in this case is probably inadequate drop of pressure in the distal external carotid, rather than displacement of blood from one area of the nose to another. In cases of hypertension, it would be
reasonable to ligate the maxillary and anterior ethmoidal arteries empirically at the same sitting.

**Ligation of the internal maxillary artery**

This operation is usually performed under general anaesthesia. A sublabial incision is made and then as large an opening as possible is made in the anterior antral wall without compromising the infraorbital nerve. The thin posterior bony wall of the antrum is shattered gently with a gouge and removed piecemeal with punch forceps to reveal the underlying periosteum on the posterior wall of the maxilla. This is incised horizontally from side to side and the fat of the pterygopalatine fossa teased out with long straight artery forceps until the tortuous maxillary artery is seen. The artery is divided between clips as close to the sphenopalatine foramen as possible and clips are placed on any large adjacent branches. The creation of an antrostomy is optional.

**Ligation of the anterior ethmoidal artery**

This is performed through an external ethmoidectomy incision. After ligating branches of the angular vein, the incision is continued down to the periosteum which is then incised in the line of the incision. The periosteum is elevated posteriorly, first off the lacrimal fossa, then the lamina papryacea of the ethmoid. The medial orbital periosteum is retracted laterally together with the lacrimal sac and held out of the way by a self-retaining retractor (Talbot). The artery is identified as a funnelling of orbital periosteum into the ethmoid labyrinth at the junction of the medial and superior walls of the orbit about half-way back from the orbital margin. It is coagulated and divided, and the incision closed without drainage.

**Other methods of treatment**

On the assumption that the application of cold stimuli to the nose is likely to reduce or arrest epistaxis, the use of cryotherapy has been advocated to control intractable bleeding (Hicks and Norris, 1983).

More recently therapeutic embolization under selective angiographic control has been carried out with apparent satisfaction (Vanwyck, Vinuela and Heeneman, 1982).

**Unusual causes of epistaxis**

**Osler's disease**

This is a familial hereditary complaint in which sufferers develop prominent telangiectatic formations recognized as red spots on the lips and the mucous membrane of the mouth, especially the tongue, as well as telangiectases on the face and in the nose. The defects in the nose are liable to cause severe epistaxis, and bleeding is rarely from one site alone. The condition may be complicated by the presence of lesions in the gut which may bleed, or arteriovenous malformations in the lungs.

Harrison (1957) has shown that high doses of oestrogen will lessen the frequency and severity of nose bleeding, probably by inducing a squamous metaplasia of the nasal mucous
membrane. However, this form of treatment is inappropriate in male patients, and could well be dangerous if prescribed over a long period of time.

Saunders (1963) advocated excising the mucous membrane of the anterior part of the septum and lateral nasal wall and its replacement by a split-skin graft which is laid on the perichondrium and sewn into position. This treatment is temporarily effective but bleeding usually recurs months or years later. The carbon dioxide laser has been used in recent years to destroy the lesions which are visible in the anterior part of the nasal fossa and, if necessary, access may be improved by resorting to lateral alar incisions. The lesions nevertheless gradually make their reappearance and bleeding eventually recurs. Radiotherapy is also successful for a time in controlling the nose bleeds.

Haemorrhage in this condition may sometimes be unusually severe and may require repeat transfusions to keep up with the blood loss. In some patients the disease runs a relatively mild course, whereas in others it may become increasingly debilitating, with the development of chronic anaemia and hypertension.

**Bleeding diatheses**

Epistaxis may be a manifestation of a clotting defect, increased fragility of capillaries, or a deficiency in platelets. A history of prolonged bleeding after trauma or dental extraction is suggestive or, alternatively, bruising or bleeding into joints. When suspected, the patient should be stripped and a search made for signs of purpura, bruising and swollen joints; a Hess's test and platelet count are carried out, and the bleeding and clotting times are measured. If required, specific tests for deficiencies of the factors responsible for coagulation can be performed to rule out such diseases as Christmas disease and haemophilia. In elderly subjects Waldenström's macroglobulinaemia should be excluded.

The treatment depends to a large extent on the individual cause of the blood dyscrasia, but in the short term, blood transfusion may be necessary. Particular caution should be exercised in packing the nose of patients with bleeding diathesis as clumsy handling will result in trauma to the mucous membranes, leading to further haemorrhage and nasal packing in such cases may well do more harm than good.

**Nasopharyngeal angiofibroma**

This arises in male adolescents and is thought, by some, to be a vascular malformation. It is characterized, not infrequently, by severe haemorrhages resulting in anaemia, local sepsis, and debility. The diagnosis is confirmed by arteriography and the correct treatment is surgical removal.
Chapter 17: Sleep-related breathing disorders

Richard L. Goode and Andrew R. Swanston

The last few years have seen a relative explosion in learned papers dealing with sleeprelated breathing disorders. The knowledge that a number of significant physiological abnormalities may be attributed to sleep disorder is not, however, new. Recent interest has been stirred with understanding of the causes of these disorders and new approaches to treatment.

The sleep apnoea syndromes are a common cause of excessive daytime sleepiness and may lead to pulmonary hypertension, systemic hypertension, cardiac arrhythmias, mental dysfunction, heart failure and sudden death. The otolaryngologist is increasingly involved in the multidisciplinary approach to assessment and treatment of the obstructive and mixed forms of sleep apnoea; it is important for the modern otolaryngologist to be familiar with the diagnosis and treatment of sleep apnoea syndromes. Snoring is no longer to be considered a rather humorous, frivolous sign of no clinical significance (Robin, 1968), a status accorded for many years. Snoring is a common feature of sleep apnoea syndromes and recent literature has concentrated on it as a possible symptom or sign of a more sinister disorder (Guilleminault and Dement, 1978; Morton, 1986). Incidence figures for snoring state that 53% of an adult male population will snore intermittently and 31% will snore regularly; for women the figures given are 38% snoring intermittently and 19% regularly (Lugaresi, Coccagna and Cirignotta, 1978). Snoring increases with age and up to 60% of men and 40% of women between the ages of 60 and 65 will snore; in the 30-35 year age group the percentages are only 20% for men and 5% for women (Lugaresi et al, 1982). Most people in this large population will be subjectively undisturbed by their snoring yet current work at The Royal National Throat, Nose and Ear Hospital in London, and Stanford Medical Center, California, supports the view that patients with 'heroic' snoring (snoring that occurs in any body position and can be heard in the next room through a closed door), even without daytime sleepiness, may have an occult form of sleep apnoea syndrome and require treatment for medical as well as social reasons.

Before discussing sleep apnoea syndromes, some definitions should be given:

**Apnoea** - cessation of air flow at the nostrils and mouth for at least 10 seconds.

**Apnoea subtypes** - apnoea can be subdivided into:
- **central** - no air flow or respiratory effort
- **obstructive** - no air flow despite respiratory effort
- **mixed** - a combination of central and obstructive apnoea.

**Hypopnoea** - a fall in the average tidal volume by more than 50%.

**Apnoea index** - the apnoea index equals apnoeas and hypopnoeas per hour of sleep.

**Sleep apnoea syndrome** - the diagnosis is made when the apnoea index equals or exceeds 5 episodes per hour or 30 over 7 hours. It must be classified as central, obstructive or mixed.

Since night-time sleep is interrupted by episodes of apnoea, the patient is sleepy the next day (excessive daytime somnolence); in fact this is so much so that he/she may fall
asleep while driving to work, or fall asleep at inappropriate moments during the working day. The obstructive form of sleep apnoea occurs almost exclusively in men (96%), and is regularly accompanied by loud snoring (100%) and abnormal movements during sleep (kicking of legs, slapping of arms, sitting up, falling out of bed and struggling for breath) (Guilleminault and Dement, 1978). Intellectual deterioration, with inattention and difficulty in concentrating, occurs in 78% while 48% have personality changes; 42% note impotence while 30% have intermittent nocturnal enuresis, and 36% morning headaches. Systemic hypertension (systolic pressure 150-210 mmHg, diastolic 95-120 mmHg) is present in 52%. Eighty per cent of the subjects are overweight. Interestingly enough, only 78% of this group with documented obstructive sleep apnoea have a chief complaint of excessive daytime somnolence - the remainder are referred primarily for loud snoring and apnoeic episodes during sleep, usually noted by the spouse and present for many years.

The diagnosis of sleep apnoea is confirmed by nocturnal polysomnogram. This study consists of simultaneous measurement during sleep of eye movement (electro-oculogram (EOG) or electronystagmogram (ENG)), electrocardiogram (ECG), chest/abdominal respiratory movements, nasal and oral air flow, oxygen saturation (using an ear-lobe or finger oximeter) and often a chin electromyogram (EMG). The study is ideally performed overnight in a sleep laboratory.

The EEG allows sleep to be staged and periods of rapid eye movement (REM) and non-REM sleep are identified by EOG. REM sleep usually occurs between 03:00 and 06:00 in normal individuals and is often accompanied by dreaming. Sleep apnoea is more common during REM sleep. The Holter monitor electrocardiogram shows the presence of bradycardia and tachycardia plus any other arrhythmia. The measurement of chest/abdominal movements and oral/nasal air flow is essential for documenting the length and number of apnoeas and hypopnoeas, as well as distinguishing central from obstructive apnoea. The oximeter documents any desaturation that may occur during sleep, while the chin EMG gives an index of the amount of mylohyoid muscle tone, so that the stage of sleep can be determined. Additional measurements can be made; body movements can be recorded with a vibration transducer and snoring documented with a tape recorder.

The nocturnal polysomnogram as outlined above is both time-consuming and expensive; full technical support is required and 'laboratory conditions' are necessary to standardize data collection. In a clinical sense full polysomnographic assessment is beyond the means of most departments of otolaryngology. The microprocessor has provided cheaper and more convenient devices which are becoming increasingly available for the purposes of sleep screening. The Vitalog, similar in use to the Holter monitor, can be taken home by the patient and worn overnight (Miles and Rule, 1986). The latest oximeters can similarly be worn in the home situation and are simple enough to be patient-operated - these store up to 8 hours of data, covering the sleep period, and can be used to display trends of oxygen desaturation by simple connection to a microcomputer. Devices such as these should put the diagnosis of sleep apnoea syndrome well within the reach of most departments and could be used to screen for the more 'at risk' patients who would benefit from further investigation and treatment.

Central sleep apnoea presents in a different way to obstructive sleep apnoea syndrome. Insomnia and depression were more frequent symptoms in one series (Guilleminault and
Dement, 1978) and excessive daytime somnolence may not be the most frequent complaint; 80% of the patients were male and the average age at presentation was 45 years.

Common causes of sleepiness must be evaluated. 'Expected sleepiness' may relate to lifestyle or the working of shifts; the taking of various drugs can lead to undue somnolence. Other causes include the narcolepsy - cataplexy syndrome (Mendelson, Gillin and Wyatt, 1977), which is commonly confused with sleep apnoea syndrome - the two conditions may coexist; periodic movements of sleep (nocturnal myoclonus) may be implicated. Central and obstructive sleep apnoea must be distinguished from these other possible causes of excessive daytime somnolence. Idiopathic hypersomnolence is a diagnosis of exclusion.

Narcolepsy is a sleep disorder characterized by attacks of sleep with sudden onset and short duration, around 15 minutes, that may occur at any time. The attacks are usually one to several hours apart and after an attack the patient feels refreshed. The onset of the disease is usually between 10 and 20 years of age; both sexes are equally affected. Narcolepsy is commonly associated with episodes of cataplexy - a sudden loss of tone in the major striated muscles producing total or partial collapse. These episodes may last from seconds up to 30 minutes. Sleep paralysis, a paralysis of striated muscles that occurs at the onset of sleep, and hypnogogic hallucinations (auditory, visual or tactile hallucinations that occur at sleep onset) are also seen in association with narcolepsy. Difficulties with sleep at night are seen. The diagnosis is confirmed by the presence of REM-onset sleep during a daytime sleep study, since daytime sleep does not normally begin with rapid eye movements.

Sleep apnoea syndromes may obviously coexist with conditions which contribute to the symptomatology. Chronic obstructive airways disease, pulmonary fibrosis, and heart disease with congestive cardiac failure may all mimic sleep apnoea syndromes and should be differentiated by appropriate history taking and relevant investigations. Cerebral tumours, hypoglycaemia, severe anaemia and hypothyroidism may all cause excessive daytime somnolence; depression is another common cause.

Pickwickian syndrome is a combination of excessive daytime somnolence, morbid obesity and right heart failure - alveolar hypoventilation produces an elevated $P_{CO_2}$, depressed $P_{O_2}$ and the patients show a marked tendency to oropharyngeal collapse during sleep. A polysomnogram is necessary to determine the role of obstructive sleep apnoea syndrome in these patients.

Conditions producing obstructive sleep apnoea syndrome must be looked for and treatment possibilities evaluated. Any condition which narrows the oropharynx, hypopharynx or supraglottic larynx can produce obstructive sleep apnoea syndrome. This includes hypognathic mandible, hypertrophic tonsillar tissue, long soft palate and uvula, large tongue (as in acromegaly and hypothyroidism) and tumours of the pharynx, supraglottic larynx, tongue base and neck. Less common as a sole cause, but possibly contributing to obstructive sleep apnoea syndrome, is nasal or nasopharyngeal obstruction due to nasal septal deviation, large adenoids, allergy or tumour.

Drugs which may contribute to or cause sleep apnoea syndromes include alcohol, sleeping pills, tranquillizers, sedatives, anti-epileptic drugs and antihistamines. Withdrawal of the drug should be effected when evaluating the patient.
The evaluation of obstructive sleep apnoea syndrome requires, therefore, careful history taking and examination - nose, nasopharynx, oral cavity, oropharynx, hypopharynx, larynx and neck must be inspected to rule out abnormal enlargements and narrowing as well as obstructing tumours. Further investigation will now commonly include haemoglobin (anaemia), haematocrit (polycythaemia in chronic oxygen deprivation), high kilovoltage neck imaging in normal respiration and phonation (palatal and faucial relations), fibreoptic nasoendoscopy and rhinomanometry (assessment of obstruction to nasal air flow).

The severity of sleep apnoea syndromes can be gauged by polysomnography. The higher the apnoea index, the worse the condition. Oxygen desaturation greater than 50%, or long runs of either bradycardia or tachycardia, suggest a severe state, as do ventricular arrhythmias. Pulmonary hypertension or right heart failure as a result of sleep apnoea syndrome obviously denotes a severe form of the disease. Patients with coronary artery disease, chronic obstructive, fibrotic pulmonary disease, or morbid obesity may start with a low oxygen saturation, elevated carbon dioxide, pulmonary hypertension and a tendency for cardiac arrhythmias, so that what would normally be a mild case of sleep apnoea syndrome may be severe in these individuals. Sudden death in sleep apnoeics is not rare and has been noted in both infant and adult populations (Guilleminault et al, 1984).

No firm classification is accepted at the time of writing. Each case must be judged individually for severity; there are no hard and fast rules for guidance - a situation not unfamiliar to the practising clinician in most areas of surgery. Severity and type of sleep apnoea syndrome determine treatment possibilities and it is necessary to seek some classification. Surgical management options for obstructive sleep apnoea syndrome are discussed below and indications for the various types of surgical management available are listed.

Investigations for obstructive sleep apnoea syndrome

Fibreoptic endoscopy

The flexible nasoendoscope is routinely used as an outpatient facility in many departments of otolaryngology. Nasoendoscopy during sleep can provide valuable information as to the site of obstruction in obstructive sleep apnoea syndrome (Guilleminault et al, 1978; Rojewski et al, 1982). Nocturnal nasoendoscopy is chiefly a research tool used to site the level of obstruction visually; the Müller manoeuvre, performed on the awake patient, provides similar information. The nasoendoscope is passed to the postnatal space and the patient is instructed to attempt a snore with the mouth closed - the principal site of obstruction can be seen directly (Sher et al, 1985).

Imaging

A lateral cephalometric X-ray of the head and neck, performed by a competent technician, may be useful in determining the site of obstruction (Riley et al, 1983). A lateral high kilovoltage neck radiograph, taken in phonation, is also useful in demonstrating velopharyngeal sphincter action and the relevant dimensions of this important region. A hypopharyngeal site of obstruction is suggested by a dimension of less than 10 mm, measured from the base of the tongue to the posterior pharyngeal wall.
Computerized tomographic (CT) scanning at the level of the oro- and hypopharynx can be useful, particularly if the patient falls asleep during the examination (not so unlikely as it may sound when the patient is suffering true excessive daytime somnolence) (Haponik et al, 1982).

In some centres fluoroscopy may also be of use and its proponents make strong claims for its use in establishing the exact site of obstruction - a most necessary preliminary to successful management, particularly of the surgical type (Smith et al, 1978; Suratt et al, 1983).

**Pulmonary function testing**

Hypoventilation must be fully evaluated when assessing an obstructive sleep apnoea sufferer. The measurement of flow-volume loops has been suggested as a test for obstructive sleep apnoea during the day (Haponik et al, 1981; Sanders et al, 1981); however, results obtained on awake patients do not regularly diagnose obstructive sleep apnoea syndrome during sleep, even when the tests are performed with the patient supine (Riley et al, 1983; Tammelin et al, 1983). The tests can be helpful if positive for upper airway obstruction. Cardiopulmonary disorders with upset of ventilatory function can be assessed with blood gas studies. As a general rule, pulmonary function and blood gas studies obtained while awake do not correlate with studies obtained during sleep (Garay et al, 1981). In many centres an in-dwelling catheter is placed prior to sleep, so that arterial oxygen saturation (SaO₂) levels as indicated by ear-lobe oximetry can be matched with blood gas values; this should become unnecessary with reliable oximetry and is obviously unacceptable as a routine clinical tool.

**Sleep latency test**

This test measures the time taken by a subject to fall asleep and is monitored by an EEG. It gives information on the degree of excessive daytime somnolence experienced by a subject and can be used for assessment purposes both before and after therapy - it does offer an objective method for measuring response to therapy (Orr and Moran, 1985). Sleep apnoea syndrome patients have a short sleep latency time, usually less than 4 minutes; normal values fall in the 6-15 minute range. Multiple sleep latency tests consist of evaluations over a 48-hour period, with a polysomnogram each night and six sleep latency tests per day at 2-hour intervals. Tests of mental function are performed throughout. Patients with obstructive sleep apnoea syndrome tend to have shorter sleep latency tests earlier in the day and longer ones later in the day. Normal subjects tend to have longer sleep latency tests at the beginning and end of the day with the shortest times in the middle of the day.

**Sleep apnoea in children and infants** (see Volume 6, Chapter 23)

Obstruction of the upper airway may arise from multiple factors, both congenital and acquired. Infants with large tonsils and adenoids, Pierre Robin syndrome, retrognathia or Crouzon's disease may present with obstructive sleep apnoea syndrome. Cleft palate repair with a pharyngeal flap may also initiate the syndrome. Sleep apnoea syndrome has been linked with sudden infant death syndrome (Guilleminault et al, 1984). Central and obstructive factors must be fully evaluated in the management of these children and preoperative assessment must include chest radiology, ECG and full blood count - also be alert to the risks
from secondary effects of prolonged sleep apnoea syndrome (polycythaemia, cardiomegaly, pulmonary hypertension, etc).

**Treatment**

The main emphasis in this section will be on surgical treatment, now usually performed by the otolaryngologist/head and neck surgeon. As in all areas of surgical management, treatment chosen will depend upon the cause and severity of the condition.

**Non-surgical treatment**

Medical means are used for the treatment of central sleep apnoea and most of the drugs used can improve obstructive sleep apnoea as well. Weight loss for the obese is often helpful (Harman, Wynne and Block, 1982) and the use of a tricyclic antidepressant, such as protriptyline, has also produced some improvements (Clark et al, 1979). It is necessary to be aware of the increased potential for cardiac arrhythmias with tricyclic agents; there is an increased incidence of arrhythmia in the sleep apnoea syndrome sufferer. Monitoring is required and protriptyline may be given in one 20 mg dose at night and increased to 30 mg if symptoms persist - the side-effects of tachycardia, urinary retention and dry mouth may occur (a result of the anticholinergic action).

L-Tryptophan has been successfully used in patients with mild central sleep apnoea syndrome (Schmidt and Jackson, 1982). The usual dose is between 3 g and 5 g taken orally 30 minutes before bedtime.

Patients with hypoventilation and decreased ventilatory drive may benefit from medroxy-progesterone acetate, a respiratory stimulant. Sleep apnoea syndrome sufferers, including those with obesity, have found this preparation of use (Strohl et al, 1981); it is of most use in mild cases with an apnoea index below 50 and oxygen saturation above 60%. An average daily dose by mouth is between 60 mg and 120 mg.

Other drugs used include acetazolamide (White et al, 1982) and naloxone (Atkinson, Surath and Wilhoit, 1983), without conclusive results. All drug therapy plans should exclude central nervous system depressants (Guilleminault et al, 1982), alcohol being a major culprit.

High altitudes may exacerbate the condition of a sleep apnoea syndrome patient. Low flow oxygen 1-3 L/min) can be helpful at night - particularly where hypoxia is implicated in the aetiology of the apnoea (Martin et al, 1982). Oxygen therapy is contraindicated if hypoxia is helping to initiate the breathing response.

Continuous positive airway pressure (CPAP) has proved most effective in the management of obstructive sleep apnoea syndrome (Sanders, Moore and Eveslage, 1983). Obese individuals, with a mixed apnoeic picture, often benefit from this therapy - surgery has an increased risk in this group since the problem is not entirely obstructive (Rapoport et al, 1982). The equipment for nocturnal use is now commercially available and it provides a tight-fitting nasal mask, connected by tubing to a quiet pump; the mask is worn during sleep and the patient quickly comes to rely on the improved quality of sleep achieved.
Similar improvements can be achieved by the use of prosthetic devices; many have been patented. They may be indicated in selected cases where tongue retention in a forward position seems particularly desirable and the patient can tolerate the device (Cartwright and Samelson, 1982). It is estimated that over 300 anti-snoring devices have received patents in the USA during this century (Garfield, 1983).

**Surgical treatment**

**General anaesthesia for obstructive sleep apnoea syndrome patients**

These patients require special attention from the anaesthetist since they may pose several problems. They are often obese and possess the short, thick neck for which anaesthetic intubation is often difficult. Mask ventilation can be inadequate since hypopharyngeal structures behave under anaesthetic as they do under sleep conditions, collapsing inwards; pharyngeal tubing and mandibular elevation may help but rapid intubation under these adverse conditions may be required. Due to the common combination of central sleep apnoea, obstructive sleep apnoea and chronic obstructive airways disease, the use of high concentration oxygen plus positive pressure ventilation, which blows off carbon dioxide, may depress the ventilatory drive reducing or abolishing spontaneous respiration, even in a relatively light anaesthetic state. Paralysing agents routinely used for intubation may aggravate this tendency.

Immediate postoperative problems may occur in the recovery area since the ventilatory drive may remain depressed, due to the high PO$_2$ and low PCO$_2$ following anaesthesia. Mask ventilation may present similar problems to those encountered during the induction period, with the added problems of postoperative oedema and bleeding. Severe hypoxia is a risk since these patients are often used to a relatively low PO$_2$ under normal circumstances, due to accompanying pulmonary or cardiac disorders, and they need just a short time to reach a dangerously hypoxic level. Increase in pulmonary compliance is often seen, adding to the difficulty.

The following rules of thumb are suggested for general anaesthesia in the obstructive sleep apnoea syndrome:

1. Intubations in sleep apnoea syndrome patients may be difficult and conventional intubation techniques may not apply; fibreoptic 'rail-roading' of the endotracheal tube should be considered, or even awake nasal intubation;

2. Preoperative sedation is considered undesirable - despite some suggestions from recent research that REM-sleep blocking premedication is safe in children (B. Donaghue, 1987, personal communication);

3. Paralysing agents should be avoided during intubation;

4. Nasopharyngeal tubes should be checked for length (nares to epiglottic tip) and inserted for airway maintenance during induction; simple oral airway maintenance during induction; simple oral airways may not give sufficient anterior tongue displacement for an adequate airway; the same is true for mandible elevation or dislocation;
(5) ear-lobe oximetry is useful as a monitor of haemoglobin SaO₂ and is simple to use during induction of anaesthesia, extubation and in the postoperative period;

(6) facilities should be available for tracheostomy;

(7) extubation should not be performed until the patient is fully awake;

(8) narcotics and hypnotics should be used with extra caution in these patients, both before and after surgical procedures; respiration must be carefully observed;

(9) careful postoperative monitoring is essential, the nasopharyngeal tube being left in place where adequacy of the airway is in doubt;

(10) selected cases may benefit from the preoperative administration of steroids and 48-hour, postoperative humidification of inspired air is helpful; steroids may be given in reducing dosage over 5 days.

Nasal surgery

Selected patients from a population of obstructive sleep apnoea syndrome sufferers may benefit from nasal surgery of one form or another; nasal obstruction is rarely the primary cause however. Correction of a deviated nasal septum may give both subjective improvement and a diminution in the number and duration of obstructive sleep apnoea episodes (Simmons and Hill, 1974; Heimer et al, 1983).

Correction of the deviated nasal septum could help by:

(1) improving reflex mechanisms: upper airway obstruction could lead to apnoea via disturbed reflex mechanisms (trigeminally or vagally mediated), which normally act to preserve airway patency in the presence of negative pressure in the upper airway (Mathew, Abn-Osba and Thach, 1982);

(2) direct mechanical effects: obstruction of the upper airway causes an increase in the pressure gradient for air flows and the pressures within the upper airway become more negative relative to atmospheric pressure - leading to secondary collapse in the hypopharyngeal area, snoring and, possibly, sleep apnoea syndrome.

When nasal obstruction is present and symptomatic it should be treated. If surgical treatment is required for a deviated nasal septum, nasal polyps, hypertrophic turbinates or enlarged adenoids, then it should be performed to attain improvement of the symptomatic nasal obstruction - and as an additional benefit it may improve, to an unpredictable degree, the symptoms of obstructive sleep apnoea syndrome, including snoring.

Uvulopalatopharyngoplasty

This operation involves resection of about 1.5 cm of the free, posterior border of the soft palate and includes the uvula, posterior tonsillar pillars and part of the posterior pharyngeal wall medial to the pillars each side - the amount of tissue taken varies with the
amount of 'redundant', loose, mucous membrane present (Simmons, Guilleminault and Silvestri, 1983). The operation is performed with a tonsillectomy if tonsils are present. It has been about 50% successful in the correction of obstructive sleep apnoea syndrome (attenuated apnoeas tend to persist) and 95% successful in relieving snoring. Subjective improvement is usually marked and believed to be related to the less frequent/shorter duration apnoeas induced in most subjects; there are also beneficial secondary social results for the sufferer.

Fujita et al (1981) described the surgical technique in the USA, which is a modification of a snoring operation reported by Ikematsu (1964). Variations of the surgical technique have been reported by Simmons (Simmons, Guilleminault and Silvestri, 1983) and Hernandez (1982).

Despite continued experience with this procedure and well-documented series (Fujita et al, 1985; Katsantonis et al, 1985), the criteria for 'success' are still vague and the parameters for prediction of surgical outcome uncertain. The authors feel that a 'successful operation' should reduce the apnoea index to less than 20 and maintain a haemoglobin SaO2 greater than 80%. With these two demands in mind, the present authors' indications for the operation are:

1. oxygen saturation which drops below 80%;
2. apnoea index worse than 20;
3. significant daytime sleepiness;
4. heroic snoring - producing marital or social problems;
5. significant cardiac arrhythmias during sleep.

Two or more of any of these indications is a reason to consider this operation.

The history is an important consideration. Excessive daytime somnolence leading to loss of job, or accidents with machinery, is clearly significant. The authors consider it justifiable to perform the operation for 'heroic' snoring alone, but recommend a preoperative sleep screening/Vitalog study to rule out concomitant obstructive sleep apnoea syndrome (with full polysomnography in suspects). If obstructive sleep apnoea syndrome is diagnosed then close follow-up must be ensured; postoperative polysomnography will demonstrate the degree of control achieved and indicate the need for any further therapy.

Severe obstructive sleep apnoea syndrome still requires tracheostomy for immediate management; these patients show haemoglobin SaO2 below 50% on a repeated basis and/or significant, sleep-induced, cardiac arrhythmias (ventricular tachycardia, long runs of ventricular extrasystoles or bradycardia below 30 beats/minute). Uvulopalatopharyngoplasty is now often combined with tracheostomy in the hope that the obstructive sleep apnoea syndrome will be corrected by it, thus enabling early reversal of the tracheostomy. This combined procedure minimizes worry over further airway deterioration subsequent to uvulopalatopharyngoplasty alone (postoperative oedema or bleeding) and can be converted to permanent tracheostomy if decannulation fails. Most patients with obstructive sleep apnoea
syndrome undergoing tracheostomy and uvulopalatopharyngoplasty have a contributing disorder which produces daytime hypoventilation; they have abnormal SaO₂ levels while awake.

The relative contribution of the various components of uvulopalatopharyngoplasty in a successful outcome is not yet known. It would seem that the palate resection is the part of the operation which does the most good in the majority of patients without tonsils, particularly for the relief of troublesome snoring. Scarring and contracture may well play a part in preventing collapse at this level, the main obstructive component. More experience with the technique, under properly controlled conditions, will be necessary before an answer to this question is found. Identification of the sleep apnoea syndrome patient most likely to benefit from the procedure is still fraught with difficulty and there are, as yet, no clear guidelines. The uvulopalatopharyngoplasty is not effective for obstruction at the level of the hypopharynx, and these patients should be identified from the physical examination and subsequent investigations.

The presence of retrognathia, Angle class II malocclusion or macroglossia, producing difficulty in visualization of the larynx on indirect mirror examination, should raise the suspicion that the level of obstruction is in the hypopharynx. Obese patients, with short, fat necks should also be examined with hypopharyngeal obstruction in mind - the large amount of fat in the neck tends to sag inwards during sleep, as tonus in the pharyngeal musculature diminishes. A depressed, malformed hyoid may also contribute to the obstruction.

There are many subjects with obstructive sleep apnoea syndrome, where the site(s) of obstruction cannot be determined by simple physical examination; a multidisciplinary approach must be used and the otolaryngologist can contribute significantly with the manoeuvres and investigations previously described.

**Tracheostomy**

This ancient operation in the repertoire of the otolaryngologist still provides the standard by which all surgical procedures for obstructive sleep apnoea syndrome must be judged. It is invariably successful but has the distinct disadvantage of a hole in the neck. The tracheostomy tube is valved during the working day, to enable normal speech, and left open at night. Variations on the theme have been tried, but removal of the tracheostomy generally leads to a return of obstructive sleep apnoea syndrome; some cases tried in conjunction with uvulopalatopharyngoplasty give cause for optimism.

General anaesthesia is preferred for this procedure and a cuffed tracheostomy tube is inserted in adults. The type of tube chosen for permanent use is at the discretion of the individual surgeon. Fenestration of the tube is desirable and the patient is taught self-management at an early stage. The Montgomery (1980) tracheostomy tube requires no ties and has several advantages over conventional tracheostomy tubes, in patients with necks of normal diameter. It does not have a cuff, a disadvantage in the early postoperative period in case of bleeding or need for positive pressure ventilation. Permanence of the tracheostomy will be facilitated by skin flaps, such as those turned inward to line the tract as described by Fee and Ward (1977).
An extra-long tracheostomy tube may be required in particularly obese patients; it is wise to have this tube available at the start of the procedure. Positioning of the tube tip after insertion must be carefully checked, since there is a real risk of insertion into the right main bronchus. Chest X-ray or fibreoptic bronchoscopy will confirm the correct location. It is necessary to assess the tube daily for a while to avert the problem of it slipping into a lower position. Obese patients with thick, fat necks heal slowly after tracheostomy. Antibiotic prophylaxis is recommended in these patients.

Other surgical procedures for obstructive sleep apnoea syndrome

Mandibular advancement

Malocclusion and retrognathia are surgically correctable; sagittal split osteotomies will correct the abnormality and pull the tongue forward, eliminating obstructive sleep apnoea syndrome due to these deformities (Bear and Priest, 1980).

Genial tubercle advancement

The genioglossus muscle attaches to the genial tubercle on the inner aspect of the anterior mandible; advancement of this structure should pull the tongue forward, even in the absence of retrognathia (Riley et al, 1986). The lower one-third of the anterior mandible, with attached genioglossus and digastric muscles, is wedged out between the mental foramina below the roots of the incisor teeth. Exposure is obtained via an incision in the gingivolabial sulcus. The wedge of bone is tapered so that the back edge is larger than the front. The bony wedge is pulled anteriorly so that the back of the tapered bone wedge lies on the anterior edge of the mandible, advancing the genial tubercle and attached muscles by the thickness of the mandibular body. The tapered resection prevents the bone wedge from slipping backwards; it is held in the new position with stainless steel wires. The operation can be combined with section of the hyoid depressor muscles (sternohyoid, thyrohyoid and omohyoid) on the lower aspect of the hyoid bone; hyoid suspension minimizes relapse.

Hyoid expansion

In some patients with obstructive sleep apnoea syndrome, inspiratory collapse of the hypopharyngeal lumen is observed; this is the likely mechanism of airway obstruction. Expansion hyoidoplasty was developed as an alternative to tracheostomy, the original work being carried out in dogs and with human cadaver studies (Patton, Ogura and Thawley, 1984). The hyoid is sectioned to give three portions, cutting the bone at the lesser cornu on each side. The three pieces are then attached to a curved piece of stainless steel, so that the side portions are pushed outwards, expanding the hypopharynx. The operation may prove useful in patients with symptomatic superior hypopharyngeal-base-of-tongue inspiratory collapse resistant to medical therapy.

Hyoid suspension

There are various materials available for suspension of the hyoid bone. Freeze-dried fascia has been used (R.G.) after cutting the hyoid on both sides at the lesser cornu and detaching the infrahyoid muscles. Two holes were drilled in the lower aspect of the anterior
mandible and the hyoid pulled up to within 1.5 cm of the lower border of the mandible using the fascial strips. Uvulopalatopharyngoplasty and tracheostomy were performed at the same time, the latter being reversed soon afterwards. The procedure is similar to laryngeal suspension performed as part of the reconstruction after base-of-tongue and supraglottic laryngeal resection (Goode, 1976). It may be combined with advancement of the genial tubercle (Riley et al, 1986).

**Base of tongue resection**

In many cases of obstructive sleep apnoea syndrome, a large tongue seems to be the cause of obstruction; limited tongue base resection has been developed in response to this observation; simultaneous uvulopalatopharyngoplasty and tracheostomy may be advocated. A tracheostomy may be reversed later if adequate clearance of the obstruction is obtained. There are two routes.

*Transoral*

This is the route of preference since it avoids an external incision and the problems that go with it; however, the very size of the tongue base itself may prevent adequate access. The portion for removal is usually between the foramen caecum and the valleculae, particularly in the midline. Horizontal wedge resection is currently under evaluation (R.G.) and this involves wedge removal from the middle portion of the tongue base. The wedge measures 2-3 cm in each plane and the wound is closed with 2-0 chromic catgut sutures. Care is taken not to transect the lingual vessels, or hypoglossal nerves which lie on the lateral aspect of the tongue base. In some cases, a vertical wedge excision may be preferable, but the vertical excision tends to narrow the hypopharynx at this level.

*External*

Obstruction to the transoral route indicates an external approach. A horizontal incision is made at the level of the hyoid and extended inwards, over the top of the hyoid bone, to reach the valleculae - similar to the approach used for laryngectomy.

A tracheostomy with cuffed tracheostomy tube is routinely performed with tongue-based resections, to avoid postoperative airway obstruction secondary to oedema and/or bleeding. Selected cases of obstructive sleep apnoea syndrome have demonstrable improvement following this procedure - but the indications for this operation are still not clear.

Further experience is needed with all of these listed procedures to enable a valid assessment of their place, if any, in the management of obstructive sleep apnoea syndrome. Effective surgical management, based on the site of obstruction, would seem to be a realistic goal. The authors have not yet achieved the diagnostic expertise that will allow the knowledge of whether nasal surgery, uvulopalatopharyngoplasty, tracheostomy, genial tubercle advancement, hyoid suspension, resection of the tongue base or some combination of these operations is the management of choice in idiopathic obstructive sleep apnoea syndrome. It is certain, however, that the modern otolaryngologist will continue to play a major role in both the assessment and treatment of patients with sleep-related breathing disorders.
Chapter 18: Non-healing granulomata and tumours of the nose and sinuses

A. D. Cheesman

The non-healing granulomata and tumours of the nose and sinuses both present with nasal obstruction and epistaxis. A tumour-like mass is seen on nasal inspection and the diagnosis is usually made by biopsy.

Non-healing granulomata

Many chronic inflammations of the nose are characterized by the formation of granulation tissue infiltrated by chronic inflammatory cells, namely granulomata. Most of these granulomata are the result of a specific infectious organism and are termed 'specific granulomata'. In others, the aetiology is less clear and they are termed 'non-specific granulomata' or more familiarly to the otolaryngologist as the 'midline non-healing granulomata'.

Generally, the correct diagnosis can be established by either histological or microbiological examination. Table 18.1 lists some of the more common types of nasal granulomata. The specific granulomata are discussed in Chapter 8.

Table 18.1 Nasal granuloma

Specific
- Syphilis
- Tuberculosis
- Lupus vulgaris
- Leprosy
- Sarcoidosis
- Rhinosporidiosis
- Mucormycosis
- Aspergillosis
- Histoplasmosis
- Blastomycosis
- Sporotrichosis
- Leishmaniasis

Non-specific
- Wegener's granulomatosis
- Lethal midline granuloma (midfacial lymphoma)

Non-specific granulomata

These lesions, more frequently called the non-healing midline granulomata of the nose, have for many years, been the cause of considerable confusion to both clinicians and pathologists. There have been many apparently different clinical entities described with detailed but non-specific histological appearances. Fortunately, over the last decade both
clinical and pathological studies have clarified the situation and most clinicians recognize two main groups: Wegener's granulomatosis and the lethal midline granuloma (nasal lymphoma). Both groups have an appropriate therapeutic regimen and, provided the correct diagnosis is made early, the prognosis has been dramatically improved.

**Wegener's granulomatosis**

Wegener's granulomatosis is a systemic disease of unknown aetiology. It may present to the otolaryngologist at various sites in the head and neck, but primarily it involves the upper and lower respiratory tracts and the kidneys. Other parts of the body are less commonly involved. It is distinguished histologically from polyarteritis nodosa by the typical formation of granulomata.

Wegener's description of rhinogenic granulomatosis in 1939 was a classic paper in which he described both the clinical and pathological features of the condition. The essential histological features are: necrotizing granulomata of the upper and lower respiratory tracts; focal necrotizing glomerulonephritis of the kidneys; and systemic vasculitis.

**Clinical features**

The original concept of the condition was of a fulminating disease leading to early death from renal failure. The patient often presents with a persistent 'cold', complicated by a blood-stained nasal discharge. Nasal examination at this stage reveals thickening of the mucosa with some ulceration and crust formation. The appearances are similar to atrophic rhinitis, but the patient is obviously unwell. Biopsy of the nasal granulations may be reported on as non-specific chronic granulation by an inexperienced pathologist, but careful examination of multiple biopsies will generally demonstrate the diagnostic epithelioid necrotizing granulomata, fibrinoid necrosis and focal vasculitis. However, the rapid clinical deterioration with evidence of systemic involvement of both the lungs and kidneys confirms the diagnosis. The chest X-ray shows localized areas of infarction which may proceed to cavity formation. Urinalysis will often show red cells, casts and proteinuria, and tests of renal function will demonstrate a decreased creatinine clearance; the erythrocyte sedimentation rate is raised. Untreated, the patient rapidly progresses to renal failure and death within six months.

However, it is now recognized that there is a more benign natural history with one or other of the main sites being primarily involved with little obvious involvement of the other sites. Careful investigation will generally confirm the diagnosis by demonstrating decreased renal or pulmonary function. This modified presentation, when primarily involving the nose, is the usual source of confusion to the otolaryngologist. The patient complains of nasal obstruction, crusting and occasional epistaxis, and the usual initial diagnosis is atrophic rhinitis. Sinus X-rays will show a thickened lining membrane, and culture of the nasal secretions often grows *Staphylococcus aureus*. In such cases the true diagnosis will only be obvious on investigation and it is important to obtain an erythrocyte sedimentation rate, urinanalysis, creatinine clearance and chest X-ray. The nasal biopsy is important and should be referred to an experienced head and neck pathologist for an opinion. The need for a confirmatory renal biopsy in such cases is best discussed with the renal physician, for early diagnosis and treatment will prevent the development of the crucial renal failure.
Other sites in the head and neck, occasionally involved either by direct spread or as separate entities, are the middle ear, eyes and orbit, the palate and oral cavity. Systemically the skin, joints, heart and central nervous system may be involved.

Treatment

The essential aim is to control the renal involvement as the usual cause of death is renal failure. High doses of steroids (40-60 mg/day) often result in a rapid clinical improvement, but long-term control or cure depends on the use of the cytotoxic drugs, azathioprine or cyclophosphamide. Cyclophosphamide, a widely used alkylating agent, is the drug of choice in North America (used in doses of 2 mg/kg per day). Its main side-effects are well documented, but the occurrence of haemorrhagic cystitis may be confusing in Wegener's granulomatosis. With the long-term use of drugs necessary in Wegener's granulomatosis, sterility, particularly in males, is a problem. In the UK, the drug of choice has been the antimetabolite, azathioprine (3 mg/kg per day). This drug is used widely as an immunosuppressant in transplant surgery. Its main side-effects on the liver and bone marrow are close related and, with the control of the condition, the dosage can usually be decreased. The variation of the dosage requires considerable experience, and is best monitored by the clinical improvement and by the fall in the erythrocyte sedimentation rate. In particularly severe cases, both drugs can be used concurrently with benefit. Long-term use of the cytotoxics beyond the period of active disease is probably necessary to prevent relapse. In dealing with relapses, it is important to exclude concurrent sinus infection in an immunosuppressed patient with disordered nasal anatomy and physiology. Opportunistic fungal infections are not uncommon. After a prolonged period of inactive disease some patients are able to stop all medication, but continued close supervision is necessary. Management of the nasal cavities during the active phase requires regular irrigations and the use of glucose-in-glycerin nose drops to reduce crusting. Sinus drainage surgery is necessary if there is an associated sinus infection. Correction of the common saddle deformity is best left until control of the disease is well established.

Non-healing midline granuloma

The title of this condition is satisfactory for current clinical usage, but with a better understanding of the underlying pathology a more suitable term will arise in due course. Clinically, the condition can readily be differentiated from Wegener's granulomatosis - the slow progressive destruction of the nose and midfacial region by an apparent chronic inflammatory response is much greater than seen in the latter. There is remarkably little systemic disturbance and no evidence of pulmonary or renal involvement, death eventually following intercurrent infection or cachexia. Originally described by McBride in 1897, the condition has been of interest and confusion to both clinicians and pathologists over the years with a variety of aetiologies being suggested. More recently the consensus of opinion has been that the condition is probably a malignant lymphoma (Kassell, Echevarria and Guzzo, 1969), the variable clinical picture being the result of different degrees of immunological control in individual patients. Harrison (1974), on clinical grounds, used radiotherapy with success and, in 1977, Michaels and Gregory found common histological features suggestive of lymphoma in a group of patients with this disorder. The features of widespread necrosis with atypical cells were termed 'necrosis with atypical cellular exudate' by them and were considered to be consistent with a histiocytic lymphoma. Furthermore, four of their cases had
similar lymph node metastases confirming their view that this was a malignant lymphoma. More recently Ishii et al (1982), using immunofluorescent techniques, demonstrated that the cell-surface markers of the malignant cells in lethal midline granuloma had the same cell-surface phenotype as is usually found in human peripheral T cells. Consequently, they concluded that lethal midline granuloma was a nasal T-cell lymphoma. T-cell lymphomata tend to develop in such extralymphatic sites as the skin and nasal mucosa. They also found the histological appearance to be consistent with T-cell lymphoma.

**Treatment**

The condition responds well to local radiotherapy, consistent with the histological diagnosis of lymphoma. Initially low non-curative doses appeared satisfactory for control, but with experience, relapses became common and now full lymphoma curative doses are used to the midfacial region and regional lymph nodes. The use of steroids and cytotoxics has been completely without success, supporting the different aetiology to Wegener's granulomatosis. Following curative radiotherapy, surgical debridement and reconstruction can be used to minimize the deformity, or a nasal prosthesis can be worn.

In conclusion, most workers now consider that non-healing granuloma should be subdivided into two categories: the well-accepted Wegener's granulomatosis, and the malignant nasal lymphoma. This latter group contains the polymorphic reticuloses. Whether there is a third small group of 'idiopathic' midline granulomata of non-lymphoma origin is becoming increasingly uncertain and, with full experienced pathological investigation, this possibility will probably disappear completely.

**Nasal tumours**

A wide variety of tumours of different histological type are found in the nasal cavities and paranasal sinuses. The more common types are listed in Table 18.2. Benign tumours are not uncommon, but malignant tumours are rare constituting less than 1% of all malignancies (3% of head and neck tumours). The presenting symptomatology of all tumours is similar and in some cases histological examination is necessary to decide whether any particular tumour is malignant.

**Benign tumours**

**Papilloma**

Squamous papillomata of the skin of the nasal vestibule and anterior septum are quite common. Usually they are treated in the outpatient department by cautery or cryosurgery. However, if they recur they should be excised for histological examination, as the early case of squamous carcinoma of this area is readily curable whereas the missed case is often incurable.

Transitional cell papilloma or inverted papilloma was originally described by Ringertz (1938), and its potential for malignancy was discussed by Osborn (1970) and Hyams (1971. The diagnosis is made histologically, the deep invaginations of the epithelium into the stroma being the typical features. They are usually found unilaterally, and there is a male
predominance of 5:1. They may be present at any age, but are found most commonly in the fifth decade. Treatment is by surgical removal (radiotherapy is not indicated even for recurrences). Local intranasal removal tends to be followed by recurrence, probably due to the difficulty in seeing the extent of the involved mucosa on the complex anatomy of the lateral nasal wall. In those cases where repeated or rapid recurrence occurs, removal by a lateral rhinotomy approach is more effective in preventing recurrence. The important feature of these tumours is their tendency to undergo malignant change, in about 2-5% of cases. It must also be remembered that the papilloma may be present simultaneously with carcinoma in about 5-10% of cases; consequently careful follow-up and examination of all material removed surgically is important.

**Table 18.2 Tumours of nose and paranasal sinuses**

<table>
<thead>
<tr>
<th>Benign</th>
<th>Malignant</th>
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<tbody>
<tr>
<td>Epithelial</td>
<td>Squamous cell carcinoma</td>
</tr>
<tr>
<td>Adenoma</td>
<td>Adenocarcinoma</td>
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<tr>
<td>Papilloma</td>
<td>Anaplastic carcinoma</td>
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<td></td>
<td>Transitional cell carcinoma</td>
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<td></td>
<td>Malignant melanoma</td>
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<td></td>
<td>Salivary gland tumours</td>
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<tr>
<td></td>
<td>Adenoid cystic carcinoma</td>
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<td></td>
<td>Mucoepidermoid</td>
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<td></td>
<td>Malignant pleomorphic</td>
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<td>Aesthesioneuroblastoma</td>
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<table>
<thead>
<tr>
<th>Non-epithelial</th>
<th>Fibrosarcoma</th>
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<tbody>
<tr>
<td>Fibroma</td>
<td>Angiosarcoma</td>
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<tr>
<td>Haemangioma</td>
<td>Haemangiopericytoma</td>
</tr>
<tr>
<td>Nasal glioma</td>
<td>Meningioma</td>
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<tr>
<td>Neurilemmoma</td>
<td>Chondrosarcoma</td>
</tr>
<tr>
<td>Chondroma</td>
<td>Osteogenic sarcoma</td>
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<tr>
<td>Osteoma</td>
<td>Lymphosarcoma</td>
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<tr>
<td></td>
<td>Rhabdomyosarcoma</td>
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<td></td>
<td>Plasmacytoma</td>
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<td>Chordoma</td>
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<table>
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<tr>
<th>Odontogenic tumours</th>
<th>Fibro-osseous tumours</th>
</tr>
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<tbody>
<tr>
<td>Haemangioma</td>
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</table>

Haemangioma may be found anywhere in the nasal cavities, but commonly are found on the anterior part of the septum, where they are called the 'bleeding polyp of the septum'. They are probably not true tumours but vascular malformations (Osborn, 1959). Recurrence is common unless the base of the polyp is excised from the septum.
Fibroma

Simple fibromata are occasionally seen as single firm polyps in the nose, but do not grow to any size.

Neurofibroma

Neurofibromata involving any of the nerves inside the nose may grow to quite a large size and a lateral rhinotomy is often necessary for complete excision.

Osteoma and other osseous tumours

Harrison (1984) reviewed this group of tumours and emphasized the need to correlate the clinical features with both radiological and histopathological findings if one is to understand the natural history of each tumour and plan its management. The benign osteomata are the commonest tumours in this group and are often found as an incidental finding in the frontal sinus on X-ray. The majority are asymptomatic and do not progress rapidly. However, they tend to occur at an earlier age in Arabs and often grow to quite large sizes in this ethnic group. Surgical excision is indicated where they cause symptoms and demonstrate an increase in size. Complete removal is essential and the base attachment must be included. As the base is often in contact with the underlying dura, a craniofacial type of approach is sometimes indicated (Cousins, Lund and Cheesman, 1987).

Fibrous dysplasia, originally described by von Recklinghausen in 1891, is now divided into two types - the multiple polyostotic lesions of Albright’s syndrome and the monostotic lesions more familiar to the otolaryngologist involving the bones of the skull. The management of the monostotic foci is discussed fully in Chapter 14, and consists of limited surgery to reduce the cosmetic defect. Radiotherapy may cause malignant change and is contraindicated.

Tumours and cysts of dental origin

Ameloblastoma and dental cysts are mentioned in Chapter 14.

Malignant tumours

Cancer of the nasal cavities or paranasal sinuses is a highly lethal condition and particularly unpleasant by its obvious nature both to the patient and the family. The results in the past have been unsatisfactory with a 30% overall 5-year survival. Frazell and Lewis (1963) commented that the unsatisfactory results could be attributed to a number of factors: the disease was invariably advanced on presentation; the complex anatomy of the region and close relationship to the orbit and skull base; and the reluctance of surgeon and radiotherapist to treat aggressively for fear of increasing the natural mutilation of the disease. Hopefully, over the last two decades there has been an improvement in this situation.

The rarity of these tumours, which constitute less than 1% of all malignancies (3% of head and neck tumours), means that many primary physicians will not see a single case in the whole of their professional careers. Their consequent relative unawareness of the condition
and the similarity of the symptoms with the more common inflammations of the upper respiratory tract results in a failure to think of the true diagnosis before the tumour extends beyond the bony margins of the sinus. The average delay between first noticeable symptom and diagnosis is 6 months. Unfortunately, this situation is unlikely to improve in the UK due to the scarcity of primary physicians with extra training in otolaryngology. Hopefully, in other countries with a higher proportion of otolaryngologists, earlier presentation will become more common. The greater availability of computerized tomographic (CT) scanning is also likely to result in the earlier detection of these tumours in those patients whose symptoms take them to other specialties such as neurology and ophthalmology.

There is an increasing tendency to refer these cases to major centres, and this often results in better management. The surgeon and radiotherapist being more experienced are more able to provide the very individual treatment regimen that is so often necessary with these tumours. The use of CT scanning enables the precise delineation of the tumour extent, and careful planning of both the radiotherapy and subsequent surgical resection.

**Aetiology**

The upper jaw is one of the few sites in the head and neck where a definite aetiology has been established for some tumour types.

Adenocarcinoma of the nasal cavity and sinuses is known to be common among woodworkers (Acheson et al, 1968). Esme Hadfield in her Hunterian Lecture (1970) showed the incidence of adenocarcinoma to be 10 times greater in High Wycombe compared with the rest of Buckinghamshire, and Acheson et al (1982) showed the skilled furniture maker, particularly the machinists, had a cumulative risk of at least 1 in 120 during their industrial lifetime of acquiring the disease (similar to carcinoma of the bronchus). The occupational risk was recognized by the Government and, in 1969, in the UK, adenocarcinoma of the nasal sinus in woodworkers in the furniture industry became a prescribed disease under the 1959 National Insurance (Prescribed Diseases) Regulations and it is the duty of the doctor to acquaint patients of their rights. The particular type of wood machined also appears to be significant, the African mahogany being the most dangerous. It is interesting to note that this wood is often used in fires by the Bantu tribesmen of South Africa who have the highest incidence of upper jaw cancer in the world, although in those cases, squamous cell carcinoma is the more common type.

Barton (1977) discussed the role of nickel as a carcinogen in squamous cell carcinoma in nickel workers. In Norway, the modification of the industrial process and a screening programme among the workers has resulted in a decline in incidence.

**Pathology**

A large variety of different tumour types has been described in the upper jaw and Table 18.2 lists the more common types. The most common histological type is squamous cell carcinoma, present in 80% of cases.

The primary site is not always easy to determine with several different sinuses commonly involved by the time the patient presents. The majority (60%) of tumours appear
to be of antral origin, 30% arise in the nasal cavities, and the remaining 10% arise from the ethmoids. Primary frontal and sphenoid tumours are very rare.

Palpable cervical lymphadenopathy is present in about 15% of cases on presentation. This small figure is because the lymphatic drainage of the paranasal sinuses is to the retropharyngeal nodes and thence to the lower deep cervical chain. Consequently, the early involved nodes are not easily palpated in any area of the neck.

Presentation

The presentation of each particular case depends on the primary site, the direction and extent of spread. Nasal cavity tumours present with the nasal symptoms of obstruction and epistaxis. Ethmoidal tumours also present with nasal symptoms, but also may have early orbital symptoms such as proptosis and epiphora, with diplopia being a late symptom. Frontal sinus tumours tend to present solely with orbital symptoms. Sphenoid sinus tumours generally present late to neurologists with neurological symptoms.

It is instructive to look at the potential presentation of antral tumours. Tumours within the antral cavity are unlikely to present early unless fortuitously they involve the infraorbital nerve giving a change in facial sensation, or alternatively bleed giving rise to epistaxis. Any epistaxis in an elderly patient who is not hypertensive requires radiological investigation, but the sinus X-rays are best deferred for 7-14 days to allow resolution of any inflammation associated with nasal packing. When the tumour breaches the antral walls, definite signs and symptoms become more obvious, their exact nature depending on the particular wall eroded.

Invasion of the nasal cavity leads to nasal obstruction and epistaxis and the tumour is often clearly seen. Less commonly, the tumour causes ethmoidal polyposis and apparently normal nasal polyps are seen; hence it is essential to examine histologically all material removed from the nose. Inferior spread to involve the palate and alveolus may result in presentation to the dentist with either an ill-fitting denture or loose tooth. Frank ulceration of the palate is a late symptom. Anterolateral spread into the soft tissues of the face may result in epiphora by involving the lacrimal sac. Facial swelling and disordered sensation are more common. Anterior spread is more likely to result in palpable cervical lymphadenopathy. Posterior spread into the infratemporal fossa and skull base may cause less obvious symptomatology, loss of trigeminal function, and trismus occurring from involvement of the pterygoid muscles. Spread into the nasopharynx may result in deafness as a result of eustachian tube dysfunction. Superior spread into the orbit causes early proptosis by increasing the volume of the orbital contents, direct involvement of the nerves and muscles occurring late.

Investigation

The objectives of investigation are to obtain a histological diagnosis and to determine the extent of the tumour. A biopsy may be readily obtained from tumour presenting in the nasal or oral cavities. If the tumour is within the antrum, a biopsy is best obtained by an intranasal antrostomy which will also provide drainage during radiotherapy. Biopsy by a Caldwell-Luc approach was previously recommended because it allowed better visualization of the tumour within the sinus and also palpation of the antral walls. This is more
appropriately done by CT scanning, and there is always a potential danger of removing the bony barrier between tumour and the facial soft tissues, even if subsequent radiotherapy 'sterilized' the area. In making the biopsy good representative samples must be taken, and any overlying necrotic tissue must be previously removed.

The extent of the disease is best determined radiologically and the modern CT scanner has revolutionized the assessment, both by demonstrating the full posterior-superior spread of the tumour, and also by showing where a more conservative approach is possible, such as around the orbit. Unfortunately, the full potential of CT scanning is not always realized particularly in departments where these tumours are seen uncommonly. The section of the most appropriate tomographic cuts and the variation in window settings are important to demonstrate the extent of both bone and soft tissue involvement. In the absence of an experienced radiologist, these are best determined at the time of the scan by discussion between the clinician and radiographer. Scans performed by a radiographer alone, and subsequently sent for later reporting by a radiologist, rarely give the necessary information. Axial cuts are readily obtained on all machines, but the present coronal and sagittal reconstructions offered by some machines are unsatisfactory for the detail required, and true coronal cuts obtained by putting the patient through the scanner in the submentovertical position are much better. The use of contrast is of particular value in assessing soft tissue involvement.

Magnetic resonance (MR) imaging is becoming more widely available; this allows scanning in three planes and, with increasing experience, promises to be able to distinguish more accurately between tumour spread and a mucocoele in adjacent sinuses.

Angiography apart from vascular tumours has little benefit to offer.

Nasoendoscopy may be valuable in early tumours, but with the tumours usually encountered the associated bleeding and mucocoele formation makes the procedure difficult, and the information obtained is generally inferior to radiology.

Classification

The classification of tumours is considered an essential feature of cancer management. It enables the individual clinician to plan an appropriate method of treatment based on experience, and also allows comparison between different treatment regimens. Unfortunately, the complex anatomy and late presentation with extensive disease involving more than one site has made classification difficult, different systems have been suggested and there has been a failure to agree on a standard UICC (International Union Against Cancer) classification. The first practical classification was proposed by Sebileau (1906) and he divided the upper jaw into three regions: suprastructure, mesostructure and infrastructure. Lederman (1970) adapted Sebileau’s classification to the TNM system, but unfortunately it has not found wide acceptance. Harrison (1978) has discussed in detail the problems with the various classifications. The main objection to all systems is the doubt that each case can be accurately recorded as to site and extension. With the advent of CT scanning, this objection has been largely overcome and it is to be hoped that a universally acceptable classification will be found. At the present time the only accepted classification is the American Joint Committee on Cancer (AJCC) system for the maxillary antrum.
Treatment

There is no widespread agreement on treatment regimens for upper jaw cancer. This is partly because of the relative rarity of the tumours, which means few clinicians develop a wide experience, but is more because of the lack of a standard system of classification which prevents meaningful comparisons between different centres.

The wide variation in tumour extent on presentation, and the differing response depending on histological type, means that each case must be considered separately, and the management planned on general principles.

The condition is naturally mutilating and death tends to be delayed and unpleasant; consequently many patients require some form of active treatment, if only for palliation. No single modality of treatment has been shown to be successful (Robin and Powell, 1981) and in most centres a combination of radiotherapy and surgery is used. Failure to achieve cure is generally due to local disease and, despite many different drug combinations, the use of systemic chemotherapy has not shown any significant benefit. Local chemotherapy with the intra-arterial infusion of cytotoxics has been disappointing apart from control of pain, and this can usually be more easily achieved by other means. More recently the use of topical 5-fluourouracil cream in conjunction with low dose irradiation and subradical surgery has shown good results (Kneght et al, 1985). The present author, however, has had little success with this modality in palliation following radical surgery, and more evidence is needed before widespread use can be recommended.

Early tumours localized to the primary site can probably be cured by either radiotherapy or surgery, but for the vast majority of cases full dose radiotherapy followed at an interval of 4-6 weeks by radical surgery is the recommended approach. Preoperative radiotherapy probably reduces the viability of the tumour, both at the primary site and within the lymphatic drainage area not encompassed by any subsequent surgery. With the modern sources available, the irradiation is concentrated in the tumour area sparing the skin, ensuring little subsequent surgical complication. Some clinicians advocate primary surgery and postoperative radiotherapy in the belief that primary debulking helps the radiotherapy; it also allows the residual areas of tumour to be identified histologically. This latter approach is of value in slow growing tumours, such as the adenoid cystic carcinoma, adenocarcinoma, and chondrosarcoma.

Combined therapy is also indicated for palliation because modern rehabilitation methods ensure little disability even following maxillectomy. Involvement of the facial skin is not a contraindication to treatment, and in practice many such cases do well. The involved area is best excised and repaired with a rotation flap of skin from either the forehead or cervical region, and occasionally microvascular free flaps have a part to play. The use of an on-lay prosthesis does much to restore the patient's confidence and may allow the return to a normal life.

Ideally, an operation should be planned to encompass the tumour in each individual patient, and over the years many different operations have been described. In practice there are two major procedures - lateral rhinotomy and total maxillectomy - and both can be extended by craniofacial resections for extensive tumours.
The basic technique of each operation is well described in the standard textbooks of operative surgery, but many points require special emphasis to ensure good tumour clearance and rapid rehabilitation. The selection of the operation depends on the preoperative assessment, but generally if the palate or zygoma is involved a total maxillectomy is indicated; in most other tumours a lateral rhinotomy will give good tumour clearance and requires little rehabilitation.

Skilled anaesthesia is essential; topical anaesthesia of the nasal mucosa with Moffet's solution and hypotensive general anaesthesia are of considerable benefit to the surgeon.

Maxillectomy

Soft tissue approach

The maxilla is best exposed by the Weber-Fergusson incision. The transverse limb should be placed close to the lid margin to prevent postoperative oedema of the lower lid and, in the medial canthal region where the potential for skin loss as a result of radiotherapy is greatest, it is helpful to curve the incision forward over the nasal bones for additional support postoperatively. A midline upper lip incision, if closed in three layers, is cosmetically more acceptable than a stepped incision. The mucosal incision along the midline of the hard palate swings laterally at the junction with the soft palate passing behind the maxillary tuberosity and then round the alveolus anteriorly. The facial skin flap is raised and all the soft tissue incisions are gently dissected free of the bone to allow the subsequent osteotomies.

Osteotomies

The maxilla is freed from the skull by osteotomies through the frontal process of the maxilla, through the body of the zygoma, through the midline of the palate, and the pterygoid plates need to be free posteriorly. The palatal osteotomy is placed in the floor of the nasal cavity and made either with a fissure burr or Gigli saw. The pterygoid plates are best separated from the maxilla with a curved osteotome, and subsequently dissected free from the muscles. The final two osteotomies are made with a fissure burr: medially through the ethmoid cells and frontal process of the maxilla after dividing the lacrimal sac; laterally the osteotomy is made through the body of the zygoma, except for those laterally placed tumours where the zygoma needs to be included in the resection, then the osteotomy is made in the lateral orbital wall below Whitnall's tubercle and through the zygomatic arch.

The remaining bony attachments are the posterior ethmoid cells and posterior antral roof, and these break readily on mobilizing the maxilla. The remaining soft tissue attachments are freed with Mayo scissors, and the maxilla removed. Bleeding from the internal maxillary artery is controlled initially by packing and then by application of a Ligaclip.

Completion of resection

Following removal of the maxilla, further tissue must be removed to ensure complete tumour clearance and promote drainage from the remaining sinuses. The ethmoid cells should be exenterated, and both the sphenoid and frontal sinuses opened widely. If there is obvious involvement of the orbital periosteum, orbital exenteration is generally indicated. The support
of the globe is complex and virtually all the medial and inferior orbital walls can be removed without the eye sinking. However, laterally, removal of Whitnall's tubercle gives considerable drooping of the eye which is best corrected by transposing the temporalis muscle medially. Orbital exenteration is achieved by an extraperiosteal dissection and transection of the muscle cone at the apex with Mayo scissors. Bleeding from the ophthalmic artery stops with local pressure. Following orbital exenteration, the eyelids are preserved but the lid margins and tarsal plates are excised to give a smooth skin lined cavity which accepts an on-lay prosthesis satisfactorily.

Postoperative spread into the pterygoid muscles is best managed by an alternative craniofacial procedure as further dissection after maxillectomy is complicated by venous bleeding from the pterygoid plexus.

Rehabilitation

Careful rehabilitation ensures minimal cosmetic and functional defect following maxillectomy. Healing of the bony cavity is fairly rapid, but it is advantageous to apply a split-skin graft to the back of the face flap. After resuturing the facial incision, the cavity should be immediately fitted with an obturator. An initial cover plate should have been constructed preoperatively to fit the palate. This is then built up with gutta percha to fill the cavity and to restore the normal facial contours. The main problem with this type of prosthesis is its weight, and help in retention is necessary. Retention is aided medially by creating a ledge on the floor of the nasal cavity by resecting the inferior part of the septum, and laterally a mucosal ledge can be made by suturing the anterior margin of the soft palate to the lateral labial mucosa over a short distance. If the prosthesis still tends to fall into the mouth, it can be secured by a circumzygomatic wire.

The primary prosthesis is generally changed after 10 days, and progressively more sophisticated prostheses can be made over the next 4-8 weeks. The final prosthesis should be no more problem than a bulky upper denture.

Lateral rhinotomy

This approach gives good access to the nasal cavities, the ethmoids, the nasopharynx and sphenoid, and also to the pterygopalatine fossa. For more extensive tumours, an en bloc resection can be achieved by combining this operation with an anterior craniofacial approach.

The incision is cosmetically very acceptable as it passes along the lateral border of the nose and around the alar margin. The upper end should start just above the level of the medial canthus. The upper lateral cartilage is freed from the nasal bones at the pyriform opening and the soft tissue flap is elevated from the frontal wall of the maxilla and nasal bones. The orbital periosteum is elevated as for an external ethmoidectomy, and the lower part of the lacrimal sac is exposed by nibbling away the anterior lacrimal crest. The orbital contents can then be completely freed medially by dividing the sac low down, and also by freeing the insertion of the inferior oblique tendon and trochlea by sharp dissection from the orbital rim. Access to the anterior nasal cavities can be increased by removing the nasal bones with little cosmetic defect. However, it is more usual to include the lateral nasal wall and ethmoid complex in the resection. The bone is freed by osteotomies cut with a fissure burr - first
through the anterior wall of the maxilla just lateral to the medial antral wall. This osteotomy starts at the orbital rim lateral to the lacrimal fossa and extends inferiorly to the level of the nasal floor. Further osteotomies are made:

(1) along the lower border of the lateral nasal wall in the inferior meatus, and
(2) through the lamina papyracea just below the anterior ethmoid artery and also across the medial orbital floor.

Finally the upper osteotomy is continued forward through the frontal process of the maxilla and nasal bone. This frees the whole block of the lateral nasal wall and ethmoid complex, apart from their posterior attachments just in front of the optic and sphenopalatine foramen. In this region the bone is very thin and easily fractured by elevating the block medially. Virtually all the mucosa of the nose can be included as a cuff with the main specimen, the posterior and antral mucosal attachments being freed by scissors. The view obtained following the removal of this main block of tissue is excellent and the excision can be extended into the sphenoid and frontal sinuses or alternatively into the pterygopalatine fossa. At the completion of the procedure, the operative cavity is packed with a Whitehead's varnish pack for 7-10 days.

Other surgical procedures

**Palatal fenestration**

This operation was originally designed for the implantation of radium into the maxillary antrum, and was claimed to allow good postoperative visualization of the cavity. However, with the alveolus left intact the view is very limited and with modern prostheses the operation has no advantage over the classical maxillectomy.

**Anterior craniofacial resection**

Involvement of the cribriform plate region has long been known as one of the major reasons for failure to control ethmoidal neoplasms; some surgeons even saw it as a contraindication to surgery. Smith, Klopp and Williams (1954) described a surgical approach to this region which was subsequently developed by Ketcham et al (1973) and Clifford (1977). The present author's team has been using a modified craniofacial technique for 10 years (Cheesman, Lund and Howard, 1986) and feels that it must be considered the ideal procedure for most ethmoidal neoplasms. The operation uses a lateral rhinotomy approach for anterior access. This is supplemented by a small midline 'window' craniotomy giving access to the floor of the anterior cranial fossa. After shrinkage of the brain with controlled hyperventilation to reduce end-tidal PCO₂ to 22 mmHg, the dura is elevated from the roof of the ethmoids and cribriform plate and the area is encompassed with a cranial osteotomy. This osteotomy, in conjunction with those of the lateral rhinotomy, allows the en bloc resection of both ethmoid complexes. Involved dura can be excised and repaired with fascia lata. Involved brain can be excised, but cure is unlikely to result at this late stage, although palliation is excellent. The window craniotomy is wired back in place and the soft tissue is closed with remarkably little cosmetic defect. The combined approach not only gives excellent visualization of the ethmoid region, but readily allows extensions of the resection into the sphenoid, the orbit, the pterygopalatine fossa and the skull base centrally. The initial results
of this type of surgery in all series have seen a doubling of the cure rate for ethmoidal
tumours to about a 60% 5-year cure rate.

**Lateral craniofacial resections**

The routine use of CT scanning shows that many antral tumours extend posteriorly to
involve the infratemporal fossa. In the past, attempts to clear the infratemporal fossa anteriorly
have not been very successful mainly due to venous bleeding from the pterygoid plexus. A
middle fossa extradural approach to the foramen rotundum allows the roof of the
infratemporal fossa to be freed and an *en bloc* resection of the medial infratemporal fossa is
possible as part of a classic maxillectomy. The combination of this approach with an anterior
fossa resection allows the *en bloc* removal of the orbit for extensive tumours involving the
orbit.

**Orbital exenteration**

Attempts to preserve the orbital contents and reduce mutilation have, in the past, often
resulted in orbital recurrence, requiring a second operation and decreased cure rate. Ketcham
et al (1973) clearly showed that orbital exenteration for involvement of the orbital periosteum
doubled the cure rate from 32% to 62% even in more advanced tumours. Consequently, it was
the author's initial practice to remove the orbital contents if the orbital periosteum was
involved with tumour. Histological examination however showed that the tumour rarely
penetrated the orbital periosteum to involve the orbital fat. More recently, the author's team
has practised resection of the orbital periosteum and, in the absence of involvement of the
underlying orbital fat on frozen section, has preserved the orbital contents. This has been
achieved with little disturbance of ocular function, and of 20 cases there has only been orbital
recurrence in two cases; both had successful subsequent orbital exenteration. Medial defects
in the orbital periosteum are repaired with split-skin grafts, but inferior resections must be
repaired with fascia to prevent prolapse of the globe.

**Special problems**

**Malignant melanoma**

Malignant melanoma of the nasal mucosa is very rare, about 1% of all malignant
melanomata. Usually they have a pigmented appearance, but amelanotic tumours are not
uncommon, and are often diagnosed as anaplastic carcinomata unless the intracytoplasmic
pigment is sought. They respond poorly to radiotherapy; chemotherapy is equally
unsuccessful, and may aggravate the situation by altering the patient's immune status.
Consequently, wide surgical excision is used, but the success of the operation probably
depends more on the patient's immune competence. Virtually the whole of the nasal mucosa
can be removed by a lateral rhinotomy approach and experience indicates that there is nothing
to be gained by a full craniofacial resection. Lymph node metastases in the neck are usually
isolated and can be removed individually, there being no evidence to support the use of a
radical neck dissection. Local recurrences can often be controlled for many years with no
active treatment and symptomatic debulking can be achieved by use of the laser or cryoprobe.
Death from melanomatosis often follows some mild immunological challenge such as
influenza.
Aesthesioneuroblastoma (olfactory neuroblastoma)

This malignant tumour is derived from the neuroectoderm. It may present to the rhinologist as a nasal tumour or alternatively to the neurosurgeon as an anterior fossa mass. Many of these tumours produce vasoactive hormones, and urinary assays of the metabolites, dopamine and 3-methoxy-4-hydroxymandelic acid have been used to monitor recurrence. The realization that this tumour is distributed on both sides of the cribriform plate and requires a craniofacial resection has resulted in an improved prognosis. For an early tumour, craniofacial resection offers the chance of complete cure, although at this time adjunctive curative radiotherapy is still recommended.

Adenoid cystic carcinoma

These tumours have a propensity to spread along the perineural spaces, and recurrences often occur at distant sites along the course of nerves supplying the primary area, many years after apparently successful local cure. They are not radiocurable, but do respond to radiotherapy. A combination of surgery and radiotherapy is indicated for these cases, surgery being used first to remove the bulk of the tumour and to indicate the likely areas of perineural spread. In the young patient, long-term cure is sought by attempting to include the involved nerves as far centrally as possible. In terms of an antral lesion, this entails a lateral craniofacial approach dividing the maxillary nerve at the foramen rotundum before it enters the wall of the cavernous sinus. If tumour is found within the nerve at this level, it is best controlled with radioactive implants, and iodine-125 seeds, with their long half-life, are a promising source for this type of tumour.

Adenocarcinoma

The adenocarcinomata or wood-workers' cancer generally involves the ethmoid sinuses. It is a relatively slow growing tumour and rarely metastasizes. It is said to be less radiosensitive than the squamous cell carcinoma and long-term cure rates are poor (less than 20% over 5 years). The initial results of craniofacial resections for these tumours in the ethmoid are encouraging and the current policy is to initially debulk the tumour intranasally. This is followed by a full course of radiotherapy and then 6 weeks later by a planned craniofacial resection of the ethmoids.

Tumours of the nasal cavity

These tumours are more common in males and, although 5% are bilateral, there is a predominant involvement of the right side of the nose, possibly related to the trauma of nose picking. The lateral wall is most commonly involved, 50% on the turbinates and, with decreasing frequency, the septum, vestibule, posterior choana and floor. Squamous cell carcinoma is the commonest histological type. As a group the do better than the paranasal sinuses with a 50% 5-year survival. For most sites either radiotherapy or surgery is used, but tumours of the vestibule and septum are best treated surgically. Tumours in these sites look innocuous, but there is a real danger of spread along the tissue planes of the cartilaginous nose and also into the upper lip. Local radical excision with careful histological control of the specimen is essential and, if the tumour is found near excision margins, postoperative radiotherapy should be given. If the tumour is restricted to one side of a cartilage plane local
resection is possible, but if it breaches the cartilage boundary, cartilaginous rhinectomy is necessary. Prosthesis rehabilitation should be used initially until tumour recurrence can be excluded. Delayed nasal reconstruction is possible, but many patients prefer a prosthesis which can be cosmetically very acceptable.
Chapter 19: Nasopharynx (the postnasal space)

C. T. Chew

The human nasopharynx is mainly derived from the primitive pharynx. It represents the nasal portion of the pharynx behind the nasal cavity and above the free border of the soft palate. It has also been called the postnasal space and epipharynx, with criticisms as to the proper terminology. The propositions of postnasal space and epipharynx are mere descriptive anatomical concepts. The concept that there is an anterior 'nasal' and posterior 'pharyngeal' component is supported by embryological, morphological and functional considerations (Kanagasuntheram, Wong and Chan, 1969; Leela, Kanagasuntheram and Khoo, 1974).

Morphological and histological studies show that the anterior portion proximal to the tubal orifice resembles the nasal cavity morphologically, while the posterior portion possesses features resembling the oropharynx. The junctional zone is the belt along the tubal orifice where the first and third pharyngeal arches meet.

Innervation studies show that the portion proximal to the tubal orifice is innervated by the maxillary division of the trigeminal (V) nerve, and that posterior to the tubal orifice by the glossopharyngeal (IX) nerve.

Functional studies with contrast and cinefluorography reveal structural differences between the two components. Contractility is observed only in the posterior portion.

Surgical anatomy

The average dimensions of the nasopharynx in the adult are 4 cm high, 4 cm wide and 3 cm in length. The posterior wall is about 8 cm from the pyriform aperture along the floor of the nose.

The anterior wall is formed by the choanal orifice and the posterior margin of the nasal septum.

The floor is formed by the upper surface of the soft palate, which occupies the anterior two-thirds, and by the nasopharyngeal isthmus.

The roof and posterior wall form a continuous sloping surface bounded by the body of the sphenoid, the basiocciput and the first two cervical vertebrae to the level of the soft palate. The upper portion of the posterior wall lies in front of the anterior arch of the atlas with a mass of lymphoid tissue embedded in the mucous membrane (nasopharyngeal tonsil or adenoid). The prevertebral fascia and muscles separate the adenoid from the vertebrae.

The lateral wall is dominated by the pharyngeal orifice of the eustachian tube. Located in the middle of the wall, it is about 1.5 cm equidistant from the roof, posterior wall, choana and the floor. The tubal elevation (torus tubarius), created by the elastic cartilage of the tube, is particularly prominent in its upper and posterior lip. Behind the posterior margin of the torus, between it and the posterior wall, lies the lateral pharyngeal recess or the fossa of
Rosenmüller. Aggregates of lymphoid tissue of variable sizes around the tubal orifice and part of the recess are collectively called the tubal tonsil.

The fossa of Rosenmüller is situated at a corner between the lateral and dorsal walls. Although not obvious in infants, the recess can measure up to 1.5 cm in depth in adults. More often than not it appears as a cleft, trabeculated at times and recedes posterolaterally to an apex near to the edge of the carotid canal opening. It opens into the nasopharynx at a point below the foramen lacerum.

Anatomical relations of the fossa of Rosenmüller are:

- Anteriorly: eustachian tube and levator palatini
- Posteriorly: pharyngeal wall mucosa overlying the pharyngobasilar fascia and retropharyngeal space, containing the lateral-retropharyngeal lymph node of Rouvière
- Medially: nasopharyngeal cavity
- Superiorly: foramen lacerum and floor of carotid canal
- Posterolateral (apex): carotid canal opening and petrous apex posteriorly, foramen ovale and spinosum laterally
- Laterally: tensor palatine and the mandibular nerve, and the prestyloid compartment of the parapharyngeal space. The fossa forms the medial border of the most superior part of the parapharyngeal space.

As the superior constrictor does not reach the base of skull, a lateral gap (sinus of Morgagni) is created. This gap is bridged only by the pharyngobasilar fascia. Through this, the eustachian tube with its two muscles, one on each side, enters the nasopharynx. Along the inferior border of the two muscles the fossa of Rosenmüller is separated from the parapharyngeal space by mucosa and pharyngobasilar fascia. Thus tumours can easily infiltrate and breach this area to spread into the parapharyngeal space.

**Epithelial lining of the nasopharynx**

The nasopharyngeal mucosa is thrown into numerous folds and crypts. The actual surface area is approximately 50 cm² in the adult. During fetal life there is a gradual transition of the respiratory ciliated epithelium to squamous type in the lower and posterior part of the nasopharynx. True squamous metaplasia occurs only in postnatal life and is completed by about 10 years of age. About 60% of the total epithelial surface is lined by stratified squamous epithelium. The mucosa abutting the choanae and immediate nasopharyngeal roof is completely lined by ciliated epithelium. Patches of squamous and ciliated epithelium, intermingling with islets of transitional or intermediate types, cover the rest of the roof and lateral walls. The posterior wall is dominated by squamous epithelium. The nasopharyngeal mucosa differs from the rest of the upper respiratory tract in that the subepithelial connective tissue is rich in lymphoid tissue. It consists of numerous small lymphocytes, plasmacytes, reticular cells and fibroblasts. This 'lymphoepithelium' together with aggregates of lymphoid tissue and the tonsils constitute Waldeyer's ring.
Adenoid and nasopharyngeal lymphoid tissue

Anatomically, the adenoid tissue is lined by epithelium which is thrown into numerous folds separating the lymphoid follicles. There are also deep crypts similar to those in the palatine tonsil. The lymphoid tissue consists of both T and B lymphocytes, with the latter predominating. The adenoid is poorly developed at birth and is not visible on X-ray in infants under the age of 1 month but is clinically identifiable by the fourth month. It is radiologically demonstrable in only 50% of infants under 6 months, and in all infants by the age of 6 months (Capitanio and Kirkpatrick, 1970). Radiologically, the adenoid appears as a soft tissue shadow mass on the roof and posterior nasopharyngeal wall immediately below the pituitary fossa.

By the age of 2 years, hypertrophy and hyperplasia of the adenoid occurs. Rapid growth occurs from 3 to 5 years with a consequent decrease in the nasopharyngeal airway. After that the adenoid size remains relatively constant while the nasopharynx increases in size (Jeans et al, 1981). Involvement of the adenoid occurs after puberty; however, the lymphoid tissue persists into old age.

The adenoid is of clinical importance. Any diminution in size or its absence could indicate an underlying immunodeficiency, for example familial hypogammaglobulinaemia and Wiskott-Aldrich syndrome. The presence of a nasopharyngeal mass in infants under the age of 1 month should raise the suspicion of a tumour such as encephalocele, as the adenoids are not detectable at this age.

Pharyngeal hypophysis

The anterior pituitary gland is formed by a median ectodermal upgrowth from an invagination (Rathke's pouch) of the stomatodeum immediately in front of the buccopharyngeal membrane. This upgrowth migrates cranially through the mesenchymal tissue, which later forms the body of the sphenoid, to rest in the anlage of sella turcica. Its original course is identified by the craniopharyngeal canal. Breaks may occur in the bucco-hypophyseal stalk to form accessory or aberrant endocrine tissues in the body of the sphenoid (Boyd, 1956). Remnants of the stalk persist in the nasopharyngeal roof to form the pharyngeal hypophysis which is a tiny elongated body of tissue in the mucoperiosteum underlying the posterior vomerosphenoidal articulation. Histologically the pharyngeal hypophysis contains chromophobe cells similar to those in the pituitary. Its functional role is not clear but it has been observed to undergo hypertrophy in women over the age of 50 years (McGrath, 1971). The pharyngeal and ectopic hypophyseal tissue may give rise to chromophobe adenoma in the nasopharynx or sphenoid (without sellar enlargement or involvement).

The pharyngeal bursa

This structure is often confused with Rathke's pouch. When present, it appears as a median sac-like depression in the posterior nasopharyngeal wall just above the upper fibres of the superior constrictor (Dorrance, 1931). It may extend upwards to the tubercle of the occiput. Inflammation of the pharyngeal bursa is known as Thornwaldt's bursitis.
Tumours of the nasopharynx

Many types of tumours, including rare, primitive ones, have been described in the nasopharynx (Table 19.1). Only a few nasopharyngeal tumours, for example hairy polyps, have characteristic macroscopic appearances. Preliminary biopsy is often required for histodiagnosis before starting treatment. The histodiagnosis of some tumours can be difficult. Of all the tumours, carcinoma is the most common. It is unique in its epidemiology and racial predisposition, with distinctive immunogenetics influencing its prognosis and survival.

Table 19.1 WHO classification of tumours of the nasopharynx (Shamugaratnam and Sobin, 1978)

I Epithelial tumours

(a) Benign
(1) Squamous cell papilloma
(2) Oxyphilic adenoma (oncocytoma)
(3) Pleomorphic adenoma
(4) Others

(b) Malignant
(1) Nasopharyngeal carcinoma
(2) Adenocarcinoma
(3) Adenocystic carcinoma
(4) Others

II Soft tissue tumours

(a) Benign
(1) Juvenile angiofibroma
(2) Neurofibroma
(3) Neurilemmoma (schwannoma)
(4) Paraganglioma (chemodectoma)
(5) Others

(b) Malignant
(1) Fibrosarcoma
(2) Rhabdomyosarcoma
(3) Neurogenic sarcoma
(4) Others

III Tumours of bone and cartilage

IV Tumours of lymphoid and haematopoietic tissues

Malignant lymphomata
V Miscellaneous tumours

(a) Benign
   (1) Teratoma
       solid
cystic (dermoid cyst)
   (2) Pituitary adenoma
   (3) Meningioma

(b) Malignant
   (1) Malignant melanoma
   (2) Chordoma
   (3) Craniopharyngioma
   (4) Others

VI Secondary tumours

VII Unclassified tumours

VII Tumour-like lesions

(1) Pseudoepitheliomatous hyperplasia
(2) Oncocytic metaplasia and hyperplasia
(3) Cysts
(4) Angiogranuloma
(5) Fibromatosis
(6) Amyloid deposits
(7) Infective granuloma
(8) Benign lymphoid hyperplasia
(9) Lethal midline granuloma (Stewart's)
(10) Wegener's granulomatosis.

Situated at the skull base with close proximity to the surrounding head and neck spaces, the nasopharynx is a clinical blind spot in many aspects. Tumours arising here may masquerade their symptoms to regions other than the primary site. This has often led to delayed diagnosis and treatment.

It is important to keep in mind that in all painless head and neck lumps, malignancy must be suspected and a primary tumour in the nasopharynx can elude exhaustive investigations, including computerized tomographic study and biopsy. If a nasopharyngeal carcinoma is suspected continued searching is worthwhile, because very often the elusive answer may lie beneath the apparently normal mucosa.

Nasopharyngeal cancer

besides the 'lymphoepithelium', the nasopharyngeal wall also contains glandular and connective tissues surrounded by bones and cartilage of the skull base. A wide variety of
malignant tumours may originate in the nasopharynx from the many types of tissue elements present there (Table 19.2).

**Table 19.2 Types of malignant nasopharyngeal tumours**

1. Epithelial  
   - Nasopharyngeal carcinoma, adenocarcinoma, adenoid cystic carcinoma, others
2. Lymphoid and haematopoietic  
   - Malignant lymphoma, Hodgkin's disease, Burkitt's lymphoma, plasmacytoma
3. Bone and cartilage  
   - Chondrosarcoma, osteosarcoma
4. Soft tissue  
   - Fibrosarcoma, rhabdomyosarcoma, others
5. Miscellaneous  
   - Malignant melanoma, chordoma, craniopharyngioma, others.

The relative proportion of cancer types in the nasopharynx varies in different countries. Nasopharyngeal carcinoma is the most common form irrespective of geography and race. It constitutes more than 90% of all nasopharyngeal cancers in most countries. In populations and countries with a high incidence of nasopharyngeal cancer, for example south-east Asia, nasopharyngeal carcinoma predominates over other types of cancer, so much so that the ratio is approximately 99:1 (Shamugaratnam et al, 1979).

**Epidemiology of nasopharyngeal cancer**

**Geography and race**

Nasopharyngeal carcinoma has a distinctive epidemiological pattern. Its incidence among the Chinese and other south-east Asians is about 10 to 50 times higher than that of other countries. This cancer is not strictly associated with the Mongoloid race per se, as the northern Chinese, Koreans, and Japanese all have a low incidence.

The highest incidence (age-standardized rate (ASR) of 15-30/100,000 males) occurs in southern China, Hong Kong, south-east-Asian Chinese and emigrant Chinese elsewhere. Moderately elevated incidences (ASR 5-15/100,000 males) are found among other south-east Asian races (Malays, Indonesians, Kadazans, Thais and Filipinos), Eskimos and some North Africans, Malta, Tunisia, Algeria and the Sudan have much lower incidences than the Asian countries but are still appreciably higher than those in America and Europe. Low incidence (ASR 1 or less per 100,000 males) is present in the rest of the world.

**Geographical and migrant variations in Chinese nasopharyngeal carcinoma**

Descriptions of neck growths with eventual death were documented in the ancient Chinese medical literature. These descriptions were most probably those of nasopharyngeal carcinoma. In China, remarkable geographical variations in incidence are observed. It is highest in the south and declines towards the north. When and where the southern Chinese emigrate, they retain their high risk of nasopharyngeal carcinoma. Their incidence is appreciably higher than that of the indigenous population. The incidence of nasopharyngeal
carcinoma among Chinese born in the USA is about 20 times higher than that of the Caucasians; but is significantly lower - about one-half - than that of Chinese born in China and those in Singapore. However, in Singapore, there is no significant difference in risk between Chinese born in China and those in Singapore. It is interesting to note that the incidence of nasopharyngeal carcinoma among the Cantonese (Chinese of Guandong origin) in Singapore (age-standardized rate per 100,000 males is 29.4, females 10.8) is very close to that of Hong Kong, where more than 90% of the population are Cantonese.

Racial differences

There is a variation of incidence of nasopharyngeal carcinoma among different ethnic groups in countries with multiracial populations, for example Hawaii and Singapore. In Singapore, the incidence of nasopharyngeal carcinoma is highest among the Chinese, intermediate among Malays and lowest among the Indians. The Indians, basically a low-risk group, have not shown any increase in the incidence of nasopharyngeal cancer despite residing in a country with high incidence. This contrasts with the Eskimos in Alaska who live in a low-risk country but have a high incidence, approximately 15 times that of the general US population (Lanier et al, 1976).

Even among the souther Chinese, there is a marked variation in the incidence of nasopharyngeal cancer among the dialects or specific community groups. The rate of nasopharyngeal carcinoma among the Cantonese is approximately twice that of the other two major groups (Shamugaratnam, 1978).

Sex

Nasopharyngeal carcinoma is more common in males with the age-standardized male:female ratio between 2-3:1.

Age

The plateau age distribution curve

The age-incidence rate curve of nasopharyngeal carcinoma is different from other forms of cancer. It begins to rise at the end of the second decade of life and reaches a peak in the fourth decade, and then stays at a plateau. This contrasts sharply with the other leading epithelial cancers of the lung and oesophagus. The shape of the age distribution curve suggests exposure to carcinogens early in life and/or an interaction of viral or environmental agents with susceptible genes. The absence of a progressive increase in older age suggests reduced exposure or susceptibility with age.

Bimodal age distribution

In Chinese, nasopharyngeal carcinoma is rare below the age of 15 years. However, in certain low-risk populations, a second peak is observed in the age distribution curve. In Tunisia, 15% of patients with nasopharyngeal carcinoma are below the age of 16 years. There is also a high proportion of nasopharyngeal carcinoma in patients below 20 years of age in other low-risk countries such as India (Bombay), Uganda, the Sudan, among US blacks and
in the Kadazans (high-risk group) in east Malaysia. This bimodal distribution suggests the influence of different aetiological factors or variations in host response.

**Environmental factors**

The aetiology of nasopharyngeal carcinoma remains obscure. A susceptible genetic constitution clearly plays a part and some environmental cofactors are equally important. The significance of environmental factors is supported by the following observations:

1. Epidemiological data on geographical clustering in southern China and Chinese emigrant populations
2. The age-incidence rate curve in the high-risk population
3. Time trend: the high risk for the disease among the Chinese in southern China, Hong Kong and Singapore have virtually remained unchanged for the last 50 years. The incidence of the disease in the second and third generation of Chinese born in the USA has declined when compared with that of their forefathers and relations in the East. The difference can be partly attributed to the change in the environment and life-style. On the other hand, the environmental change is not profound in south-east Asia. The Chinese have retained their 'micro-environment' preserving the oriental life-style especially with regard to food and customs. It is therefore likely that any associated environmental risk factor(s) is closely linked to the traditional rather than the modern life-style of the southern Chinese. Besides the Epstein-Barr virus, a variety of inhaled and ingested agents have been proposed as the aetiology (Table 19.4). Some of these propositions are supported by findings from controlled studies and laboratory evidence, but most are inconclusive.

**Table 19.4 Various environmental agents/factors implicated in the aetiology of nasopharyngeal carcinoma**

<table>
<thead>
<tr>
<th>Agent/factor</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epstein-Barr virus</td>
<td>Raised antibody, Viral genome in tumour cells</td>
</tr>
<tr>
<td>Chemical</td>
<td></td>
</tr>
<tr>
<td>Tobacco</td>
<td>Cigarette smoking</td>
</tr>
<tr>
<td>Drugs</td>
<td>Chinese herbal medicine</td>
</tr>
<tr>
<td>Plant products</td>
<td>Epstein-Barr virus activating properties/cofactors</td>
</tr>
<tr>
<td>Diet</td>
<td>Salted fish, Nitrosamines</td>
</tr>
<tr>
<td>Cooking habits</td>
<td>Household smoke and fumes</td>
</tr>
<tr>
<td>Religious practices</td>
<td>Incense and joss stick smoke</td>
</tr>
<tr>
<td>Occupation</td>
<td>Industrial fumes and chemicals</td>
</tr>
<tr>
<td>Others</td>
<td>Socioeconomic status, Previous otolaryngological ailments, Weaning habits, Nutritional deficiencies, Metals (arsenic, chromium, nickel).</td>
</tr>
</tbody>
</table>
Household smoke and cigarette smoking

Exposure to smoke has long been suggested as a risk factor following observations on indoor cooking in chimney-free homes in southern China. There appears to be no relationship between nasopharyngeal carcinoma and household smoke. The boat-people of south China who cook in the open air have a higher incidence of nasopharyngeal carcinoma, and the women who are more exposed to smoke from firewood used in cooking have a lower risk than men. The geographical distribution of nasopharyngeal carcinoma bears no relationship to the pattern of cigarette consumption or the incidence of lung cancer.

Occupation

Exposure to nickel, chromium and radioactive metals has been associated with cancer of the nose and paranasal sinuses but not with nasopharyngeal carcinoma. Inhalation of chemical fumes in certain occupations may explain the occurrence of nasopharyngeal carcinoma in industrialized countries with a low incidence. There is recent evidence to suggest a relationship between chemicals and activation of Epstein-Barr virus. A remarkably high detection rate of nasopharyngeal carcinoma was reported in a chemical factory during an Epstein-Barr virus serological survey in Wuchou City, China (Zeng et al, 1985).

Ingestants

Salted fish had been proposed as an important aetiological factor in the southern Chinese population. Ungutted salted marine fish contains an appreciable amount of volatile nitrosamine, principally N-nitrosodimethylamine and N-nitrosodiethylamine. These are known to induce squamous cell carcinoma and adenocarcinoma in the nasal and paranasal cavities in experimental animals (Huang et al, 1978). However, this does not satisfactorily explain the male predominance of nasopharyngeal carcinoma. Previous controlled studies relating the incidence of nasopharyngeal carcinoma to that of consumption of salted fish in California (Henderson et al, 1976) and Malaysia (Armstrong, Kannan Kutty and Armstrong, 1978) did not show any significant correlation, although a recent follow-up report on the same Malaysian study indicated childhood salted fish consumption a risk factor (Armstrong et al, 1983).

Histopathology

Nasopharyngeal carcinoma arises from the crypts and squamous or respiratory epithelium lining the wall. It may be preceded by squamous metaplasia. There has been considerable disagreement over the histological classification of nasopharyngeal carcinoma. Terminologies such as lymphoepithelioma, undifferentiated carcinoma, non-keratinizing carcinoma, transitional cell carcinoma and anaplastic carcinoma have been used to describe the poorly differentiated carcinoma that commonly occurs in the nasopharynx. The term 'lymphoepithelioma' is used to describe non-keratinizing and undifferentiated nasopharyngeal carcinoma in which numerous lymphocytes are found among the tumour cells. The lymphoid elements are not neoplastic. According to the WHO Classification (Shamugaratnam and Sobin, 1978), three histological types are recognized on the basis of their light microscopic appearances:
(1) squamous cell carcinoma
   (a) well differentiated
   (b) moderately differentiated
   (c) poorly differentiated
(2) non-keratinizing carcinoma
(3) undifferentiated carcinoma.

**Squamous differentiation**

All histological subtypes of nasopharyngeal carcinoma consistently show ultrastructural and immunohistochemical evidences of squamous differentiation. These include the non-keratinizing and undifferentiated carcinomata with no evidence of squamous differentiation on light microscopy. Therefore they may be considered variants of the squamous carcinoma. The most common subtype seen in high-risk countries is the undifferentiated type.

**Clinical-pathological significance**

*Cervical lymph node metastases*

Nasopharyngeal carcinoma and its metastases often display fairly characteristic cytological and histological features which enable a presumptive diagnosis of their origin to be made. This would then alert an unsuspecting surgeon to search for the primary tumour after excising a lymph node, without prior examination by an otolaryngologist.

**Clinical presentations of nasopharyngeal carcinoma**

**Macroscopic appearances**

Nasopharyngeal carcinoma has no characteristic macroscopic features. The lesion may appear ulcerative and by infiltrative or be a more exuberant polypoid type of tumour. Inflammation of the 'lymphoepithelioma' may mimic tumour appearance. Very early preclinical and infiltrative carcinoma retains a relatively normal mucosal appearance, and the diagnosis is based on histopathology.

**Anatomical sites of origin**

Primary tumour distribution is found in the following order of frequency:

(1) lateral wall
(2) superior-posterior wall
(3) more than one wall
(4) anterior wall and floor.

More than 80% of the tumours are unilateral. The right and left sides are equally affected. Most of the tumours arise from the lateral wall, especially the fossa of Rosenmüller and around the eustachian cushion.
**Symptomatology**

The marked invasive and metastatic powers of the nasopharyngeal carcinoma are responsible for the symptomatology. From the primary site the tumour may spread in one or more of the following directions:

1. anteriorly to the nasal cavity and paranasal sinuses, pterygopalatine fossa and apex of orbit
2. posteriorly to the retropharyngeal space and node of Rouvière, destroying the lateral mass of atlas
3. laterally into the parapharyngeal space
   a. prestyloid compartment: with involvement of the mandibular nerve, pterygoid muscles and infiltration of the deep lobe of parotid
   b. poststyloid compartment: with vascular compression of the carotid sheath and vessels, and invasion of the last four cranial nerves and cervical sympathetic nerves
4. superiorly through the sphenoid body and sinus involving the parasellar structures and optic nerve, petrous apex and foramen lacerum, spreading along the carotid canal into the cavernous sinus involving nerves III, IV, V, and VI
5. inferiorly to the oral cavity and retrotonsillar regions.

**Clinical features**

Most patients have multiple symptoms which are insidious in onset and are sometimes disregarded by the patients and doctors. The main symptoms are cervical lymphadenopathy (60%), epistaxis and nasorespiratory symptoms (40%), audiological symptoms (tinnitus, otalgia, deafness) (30%), neurological symptoms (headache, cranial nerve palsies) (20%), and metastases which may be local (paranasal sinus, parapharyngeal space, infratemporal fossa, orbit and parotid) or distant (spine, lung and liver).

**Cervical lymphadenopathy**

Nasopharyngeal carcinoma has a tendency for early lymphatic spread. The lateral retropharyngeal lymph node (of Rouvière) is the first lymphatic filter and is not palpable. The common first palpable node is the jugulodigastric and/or the apical node under the sternomastoid. Contralateral lymph node metastasis is not uncommon.

The parotid gland and lymph nodes can be involved if the parapharyngeal space is breached. Secondary deposits may mimic a primary parotid tumour. Parotidectomy has been erroneously performed as a result.
**Epistaxis and nasorespiratory symptoms**

Epistaxis as primary presenting symptom is unusual; it is more commonly seen in advanced nasopharyngeal carcinoma with or without skull base erosion or postradiation infection. Blood-stained nasal mucus and saliva on hawking are more frequently seen. Erosion into the maxillary antrum mimics sinusitis. The blood-stained rhinorrhoea from nasopharyngeal carcinoma may masquerade as primary maxillary cancer. Complete nasal obstruction is a late presentation; should it occur in the early stage of the disease, it is often due to superimposed infection. Ozaena occurs as a result of tumour necrosis and is typical of advanced nasopharyngeal carcinoma.

**Tinnitus and aural symptoms**

Serous otitis media with tinnitus is not an uncommon presentation of nasopharyngeal carcinoma and the primary tumour may be insignificant in the peritubal region. Adult Chinese patients with unresolving serous otitis media have to be presumed to have nasopharyngeal carcinoma until proven otherwise.

**Neurological palsies**

All the cranial nerves, either singly or in groups, can be affected by nasopharyngeal carcinoma through tumour invasion or compression. The most frequently involved cranial nerves are V, VI, IX and X, accounting for 50% of all palsies (Khor et al, 1975). Nerves IX and X are invariably involved together and are the most common group to be affected. The nerves to the ocular muscles (III, IV and VI) are the next group commonly affected. Isolated single cranial nerve palsy is common with nerves V and VI.

**Pain and headache**

Pain is an ominous symptom in nasopharyngeal carcinoma. Severe pain with headache is the hallmark of terminal disease. It signifies tumour erosion to the skull base and surrounding structures. Sepsis, particularly in sphenoidal sinusitis, also produces severe headache. If accompanied by trismus, the disease is in an advanced stage for the tumour has involved the pterygopalatine fossa and the pterygoid muscles.

Atypical facial pain or unexplained headache in the absence of obvious clinical findings in the nasopharynx may be a presenting symptom of nasopharyngeal carcinoma. The nasopharynx can appear deceptively normal when the carcinoma has in fact spread extensively outwards by submucosal infiltration. In such situations, a computerized tomography (CT) scan of the nasopharynx and base of skull is most helpful in delineating the outlying tumour extension and skull base erosion. The small nasopharyngeal primary may just represent the tip of the 'tumour iceberg'.

**Distant metastases**

The incidence rate of distant metastases is about 30%, of which skeletal metastases account for more than one-half. The thoracolumbar spine is the most common site of involvement followed by the lung and liver. Distant metastases indicate a grave prognosis
with a median survival of 3 months; 90% of patients die within one year of diagnosis of the first metastasis. In a study of 352 consecutive cases of nasopharyngeal cancer treated with radiotherapy (Khor et al, 1978), 60% of patients who manifested distant metastases had no evidence of recurrent disease in the nasopharynx or cervical nodes. This implies that a significant proportion of patients probably have occult metastases at the time of initial diagnosis.

Nasopharyngeal examination

Examination of the nasopharynx can be problematic. Although anatomically the width of the nasopharynx is 4 cm, the 'functional channel' is only about 2 cm (Khoo, Chia and Naplon, 1967). Posterior rhinoscopy is also restricted by the pharyngeal reflex, patient cooperation and inability to open the mouth. Furthermore, the mirror may only give an 'edge-on' view of the fossa of Rosenmüller due to the latter's posterolateral inclination. Nevertheless, mirror examination is still the quickest way to assess the nasopharynx, sometimes under anaesthesia. In this procedure, the patient is placed in the tonsillectomy position with a Boyle-Davis gag. Two polythene tubes or catheters, inserted naso-orally, retract the soft palate forward. Transoral mirror examination is then performed with a mirror that has been dipped in diluted cetrimide - this effectively demists the mirror. Biopsy is performed via the nose with the help of the mirror view or Yankauer speculum transorally.

Although a transoral nasopharyngoscope provides a panoramic view under magnification, it has not eliminated the limiting factors of posterior rhinoscopy. With the introduction of transnasal fibreoptic nasopharyngoscope, close-up end-on viewing of the nasopharynx has been possible. Any tiny growth which escapes detection with routine mirror examination can be identified. Biopsy can also be performed under direct visual guidance.

Nasopharyngeal biopsy

Methods

(1) transnasal
   (a) blind
   (b) posterior mirror rhinoscopy
   (c) endoscopy
(2) transoral
   (a) Yankauer speculum
   (b) rigid endoscopy.

Transnasal blind biopsy

The slim Hildyard postnasal biopsy forceps is preferred for routine nasopharyngeal biopsy. Specimens from its 3 mm diameter cup are more than adequate for tissue histopathology. In this method, the forceps are inserted along the nasal floor, slightly angulated upwards and laterally to bite at the posterolateral nasopharyngeal wall. It causes little discomfort to the patient. Clinically positive nasopharyngeal carcinomata are often diagnosed at the first biopsy. However, small or anteriorly placed tumours (about 10%) may be missed by this method.
Flexible fibreoptic nasopharyngoscope

This is the most useful and versatile endoscope for nasopharyngeal and upper aerodigestive system examination. Before the procedure, the nose is first anaesthetized with 5% cocaine spray (or 4% lignocaine if patient is allergic to cocaine). Two orange sticks (one for each nostril) with cotton pledgets soaked with 5% cocaine are inserted along the floor to the nasopharynx. With this method, anaesthesia of the nasopharynx is often achieved. However, one must be aware of a common pitfall, that is the failure to keep the pledget sticks in the nasopharynx. This may occur as a result of the soft palate propelling the pledget sticks away from the nasopharyngeal wall. This reflex action usually stops once the nasopharynx is anaesthetized. An induction time of 10 minutes is allowed before the endoscopic procedure. The flexible scope is then inserted transnasally. It gives a good view of the nasal floor, the walls of the nasopharynx and the fossa of Rosenmüller. On passing it below the soft palate, pharyngolaryngoscopy can be performed.

Flexible endoscopic biopsy

The more recent nasopharyngoscopes are equipped with biopsy forceps (introduced through the suction channel). However, the biopsy specimen is tiny and not always suitable for immunohistological study. An improvised method using an older flexible nasopharyngoscope (Olympus ENF-P) and Hildyard forceps is described.

The flexible nasopharyngoscope is first introduced through the nose contralateral to the nasopharyngeal tumour. Its tip is directed towards the tumour. As the nasopharyngoscope is rather short (total length of scope 45 cm and diameter of insertion tube 4 mm), it can be steadied with one hand once it is in the nasopharynx. The Hildyard biopsy forceps are then inserted along the nasal floor on the side of the tumour into the nasopharynx. In fact, in most cases, it is small enough to be introduced through the same nostrils as the endoscope. The position of the biopsy forceps can be checked by the scope.

The advantages of this method include:

1. Tiny tumours in any quadrant including the difficult fossa of Rosenmüller can be biopsied accurately

2. The usual problem of postbiopsy bleeding obscuring the vision is avoided as the scope is far away from the biopsy site

3. It is also a reliable method to detect and biopsy postradiation tumour recurrence beneath the necrotic scab that may persist long after radiotherapy

4. It obviates the need for diagnostic nasopharyngeal curettage. This latter procedure should be discouraged, as it is almost impossible to curette the lateral nasopharyngeal wall, where most tumours primarily occur.
Stage-classification of nasopharyngeal carcinoma

Few staging systems of head and neck cancers have encountered more controversy than that of nasopharyngeal carcinoma. To date there are no fewer than 10 different classifications.

1952 Geist and Portman
1962 UICC (modified 1974 and 1978)
1965 Chinese classification (Shanghai)
1970 J. H. C. Ho (Hong Kong, modified 1978)
1971 Chen and Fletcher (M. D. Anderson Hospital, Houston)
1975 German work group of clinical oncology (Cologne)
1976 American joint committee (modified 1978)
1977 Kyoto symposium (Japan)
1979 Changsha conference (China)
1981 Guangzhou stage-classification

None has gained general acceptance. Recent classifications are modifications of the older systems. The proposed Guangzhou stage-classification (19891) is one such modification to be evaluated. A synopsis of the stage-classifications in use is given in Appendix 19.1.

Treatment

Radiotherapy

Radiotherapy is the definitive treatment for nasopharyngeal carcinoma and its regional nodal metastases. Complications such as radiation myelitis, brainstem damage, optic atrophy and retinitis are rare. However, mucositis, xerostomia, dental caries and radiation serous otitis media are some of the sequelae of the treatment. Pretreatment dental clearance and treatment of oral sepsis is mandatory. It is a prophylactic measure against postradiation radionecrosis following dental extraction.

Chemotherapy has been used to supplement radiotherapy for advanced cervical nodal metastases as well as to treat visceral metastases. However, the result is disappointing.

Results

Results of treatment vary with the stage of disease and the age of the patient. Comparison of treatment results between different centres is made difficult by the lack of a generally accepted stage-classification. The accepted overall 5-year survival rate is 30-40% with megavoltage radiation therapy.

Role of surgery

Surgery plays a minor role in the treatment of nasopharyngeal carcinoma. It is restricted to obtaining a biopsy and inspection of the nasopharynx, for example palatal fenestration, in selected patients. This procedure is seldom performed nowadays following the
introduction of fibreoptic scopes. Radioresistant nodes may be removed by radical neck dissection.

**Immunology of nasopharyngeal carcinoma**

**General cell-mediated immunity in nasopharyngeal carcinoma patients**

Impaired T-cell functions are found in more than one-half of newly diagnosed and untreated patients with nasopharyngeal carcinoma. This can be demonstrated *in vivo* by the Mantoux test and *in vitro* by the phytohaemagglutinin response of lymphocytes (Chan et al, 1978). Similar impairment is also observed in treated patients who are in remission. Cell-mediated immunity against Epstein-Barr virus is still present in patients with nasopharyngeal cancer as demonstrable by the lymphocyte transformation assay against Epstein-Barr virion antigens, even though as a group they seem to have a lower response than control populations (Chan, Chew and Kunaratnam, 1979). Impaired viral specific T-cell immunity and increased suppressor T-cell activity in patients with nasopharyngeal cancer suggest immunosuppression. Antigen overload has been suggested to be the cause of the immunosuppression.

**Epstein-Barr virus and its association with nasopharyngeal carcinoma**

This is one of the herpes viruses. Its lymphotropic action is restricted to the B lymphocytes which are found in abundance in the 'lymphoepithelium'. Epstein-Barr virus primary infection takes place in childhood and is always accompanied by seroconversion and harbouring of the virus in a dormant state for life. The virus may be reactivated with raised serological titres in immunosuppressive states. Among human cancers, only Burkitt's lymphoma and nasopharyngeal carcinoma are closely associated with Epstein-Barr virus. These two cancers have different seroepidemiological backgrounds. Nasopharyngeal carcinoma is an epithelial tumour and is not related to the endemicity of the Epstein-Barr virus. Decades elapse between primary infection (< 5 years of age in southern Chinese) and the occurrence of nasopharyngeal carcinoma (peak around the fourth decade). Its association with Epstein-Barr virus is supported by:

1. the presence of a humoral immune response, in patients with nasopharyngeal carcinoma against Epstein-Barr virus-determined antigens, including the structural antigens such as the viral capsid antigen (VCA), early antigen (EA) and nuclear antigen (EBNA)

2. the presence of Epstein-Barr viral markers, DNA and nuclear antigen, in nasopharyngeal carcinoma tumour cells (Wolf, Hausen and Becker, 1973). However, Epstein-Barr viral particles have yet to be observed in nasopharyngeal carcinoma cells from biopsy samples.

**Aetiological role of Epstein-Barr virus in nasopharyngeal carcinoma**

More than 90% of patients with nasopharyngeal carcinoma have elevated antibody titres to Epstein-Barr virus-determined antigens compared with controls of the same ethnic group and geographical location. Of the many types of nasopharyngeal carcinoma, only the undifferentiated/poorly differentiated forms consistently express, irrespective of race or geographical location, the Epstein-Barr nuclear antigen which is an indicator of the presence
of Epstein-Barr virus genome (Desganges et al, 1975; Andersson-Anvert et al, 1977). Moderately to well-differentiated nasopharyngeal carcinomata are devoid of Epstein-Barr virus DNA or nuclear antigen. Like the normal population, this patient group does not have elevated antibody to Epstein-Barr virus antigens. Other aerodigestive tract carcinomata have so far failed to produce Epstein-Barr virus markers with the exception of undifferentiated carcinoma of the nasal fossa (Huang et al, 1978a).

It is not clear at present how the viral DNA becomes associated with the epithelial carcinoma cells or when the epithelial cells are infected by the virus, whether it is before or after the malignant change (passenger virus), or as a result of impaired host immunity. After all, only the B lymphocytes are known to have receptors for Epstein-Barr virus. The aetiological role of the virus in nasopharyngeal carcinoma is still controversial.

**Epstein-Barr serological markers**

In comparison to patients with other head and neck carcinomata, patients with nasopharyngeal carcinoma have a broader spectrum and higher geometric mean titres of a series of Epstein-Barr virus antibodies (Henle et al, 19970). Important Epstein-Barr virus-related antibodies in nasopharyngeal carcinoma are:

1. IgA and IgG to viral capsid antigen
2. IgA and IgG to early antigen
3. antibody to nuclear antigen
4. antibody-dependent cellular cytotoxicity antibodies.

These are of clinical importance in evaluating a patient with nasopharyngeal carcinoma for the stage of the disease at the time of diagnosis, the effect of and response to therapy, and the clinical course and survival.

Antibodies against viral capsid antigen, early antigen and nuclear antigen are the most useful in clinical practice and their titres correlate well with each other. The IgA response to Epstein-Barr virus antigens in nasopharyngeal carcinoma is unique and characteristic of patients with nasopharyngeal carcinoma. Antibody-dependent cellular cytotoxicity, a process known to be effective in the destruction of viral-infected cells, appears to act on Epstein-barr virus-induced membrane antigens. It is capable of destroying the infected cells. The antibody-dependent cellular cytotoxicity antibodies titre may represent a functional immune response against tumour cells *in vivo*.

**Seroimmunological index in the diagnosis of nasopharyngeal carcinoma**

Immunoglobulins IgA/VCA, IgG/VCA, and IgA/EA, IgG/EA are useful diagnostic markers of nasopharyngeal carcinoma. Their titres are related to the tumour load, and geometric mean titres increase with advancing stage of the disease in untreated patients. The diagnostic titres for viral capsid antigen and early antigen antibodies are:

<table>
<thead>
<tr>
<th></th>
<th>IgA</th>
<th>IgG</th>
</tr>
</thead>
<tbody>
<tr>
<td>VCA</td>
<td>1/10</td>
<td>1/640</td>
</tr>
<tr>
<td>EA</td>
<td>1/5</td>
<td>1/80</td>
</tr>
</tbody>
</table>
IgA/VA has the highest sensitivity with a nasopharyngeal carcinoma detection rate of 95%, followed by IgA/EA which is highly specific for nasopharyngeal carcinoma with almost no false positives. IgG/VCA has the least discriminatory value as a primary serological diagnostic indicator of nasopharyngeal carcinoma (Ho et al, 1981).

**Clinical course and survival**

The titres of IgA/VCA and IgA/EA are useful clinical indices for the follow-up of nasopharyngeal carcinoma patients after treatment. They decline to a low level after successful treatment (Henle et al, 1977). An upsurge of viral capsid antigen, early antigen and nuclear antigen antibodies would indicate clinical recurrence and/or metastasis. It may be useful to be aware of the occurrence of an occult tumour and this would necessitate careful evaluation of the patient.

The geometric mean titres of early antigen, viral capsid antigen and nuclear antigen (taken at the time of diagnosis) were significantly higher in the patients who died (within 4 years) from the disease compared with those who survived. Antibody-dependent cellular cytotoxicity antibody titre was highest in long-term survivors while viral capsid antigen and early antigen antibody titres showed a progressively inverse relationship to survival. The geometric mean titres of early, viral capsid and nuclear antigens increase stepwise with disease stage but decline towards the end-stage. Antibody-dependent cellular cytotoxicity antibody clearly demonstrates its value as a biological titre in determining the survival of patients (data from Chan et al, 1979).

**Prognostic serological markers**

The prognostic markers of nasopharyngeal carcinoma include specific Epstein-Barr virus antibody titres. The Epstein-Barr virus titres are dependent on the histological types, the availability (load) of various Epstein-Barr virus antigens and host immune competence. The titres may not be elevated in the early and end stages of the disease nor in cases with intracranial extension without significant lymph node involvement.

(1) Prognosis and survival is inversely proportional to the geometric mean titres of viral capsid antigen and early antigen antibodies.

(2) Good prognosis is indicated by a high antibody dependent cellular cytotoxicity antibody titre. Its titre appears to be independent of the disease stage. This suggests that a parameter independent of the tumour load is involved.

**Other clinical applications**

**Screening for nasopharyngeal carcinoma in high risk populations**

IgA/VCA is of practical value in serological screening for nasopharyngeal carcinoma in endemic regions. A large-scale seroepidemiological survey has been conducted in Guangzi Autonomous Region, China since 1978. A total of 148029 normal subjects over 30 years old were tested for IgA/VCA. A total of 3533 cases (2.4%) were found to be positive and from this group 55 cases of nasopharyngeal carcinoma were detected. Thirty-two more cases of
nasopharyngeal carcinoma were diagnosed in subsequent follow-up. The period between the
detection of raised IgA/VCA and the clinical onset of stage I nasopharyngeal carcinoma
ranged from 8 to 30 months (mean 13). The detection rate of the screened IgA/VCA-positive
population was estimated to be more than 80 times the annual nasopharyngeal carcinoma
incidence of the general population of comparable age (Zeng et al. 1983). This indicates
positively the existence of subclinical and early nasopharyngeal carcinoma in the otherwise
asymptomatic IgA/VCA-positive individuals. The long subclinical period (months or even
years) with raised IgA/VCA probably indicates the slow tumour growth with ample time for
the Epstein-Barr virus antigens to stimulate the immune system. As there is no diagnostic
macroscopic appearance of nasopharyngeal carcinoma, it is likely that tumour in the early
stage is indistinguishable from the normal 'lymphoepithelium' and lymphoid aggregations
commonly seen in the nasopharynx. A raised IgA/VCA titre identifies these high risk
individuals for further clinical and immunohistological evaluation of the nasopharynx
(Desgranges et al. 1982).

**Differential diagnosis of nasopharyngeal carcinoma**

Morphologically nasopharyngeal carcinoma can be confused with lymphoma especially
in the low risk population.

**Occult primary tumour with cervical metastases**

The nasopharynx is still the most frequent site of an occult primary tumour in the head
and neck with cervical metastases. In certain clinical situations where the primary tumour is
difficult to detect, a positive Epstein-Barr virus IgA serology, and positive immunohistological
markers on the metastatic tumour tissue serve as an adjunct to pathological identification
(Coates et al, 1978). Multiple nasopharyngeal biopsies under endoscopic vision are indicated
if the Epstein-Barr virus serological markers are positive. Should the serology be negative and
other head and neck regions are clear, an enlarged lymph node should be excised *in toto*
with the capsule intact. Fresh lymph node tissue is sent for identification of nuclear antigen and
DNA in the tumour cells. If nuclear antigen is demonstrated the primary tumour is most likely
to be a nasopharyngeal carcinoma. Nuclear antigen has so far not been demonstrated in other
carcinomata of the head and neck except some parotid gland tumours reported in Eskimos
(Saemundsen et al. 1982) and, recently, thymic carcinoma (Leyvraz et al, 1985). One must
be aware of the fact that a small primary nasopharyngeal carcinoma is known to give rise to
over tumour deposits in the parotid gland.

Indiscriminate excision biopsy of lymph nodes should be discouraged. Such biopsy
offers little in the clinical management if the primary tumour is still untreated. It would
further compromise the prognosis in nasopharyngeal carcinoma and increases the morbidity
to radiation should the wound break down as a result of tumour seedings.

**Immunogenetics of nasopharyngeal carcinoma**

Patients with nasopharyngeal carcinoma have shown a prominent genetic susceptibility
to this cancer. This is evident by the following observations:
(1) high risk among southern Chinese population

(2) differential high risk in emigrant Chinese in comparison to the indigenous population

(3) family clustering of nasopharyngeal carcinoma in Chinese

(4) elevated risk in people having genetic admixture with Chinese

(5) low risk in other racial groups despite living in high-risk countries, for example Indians in Singapore.

Genetic markers in nasopharyngeal carcinoma

Patients with nasopharyngeal carcinoma among the Chinese are found in a genetically distinct subpopulation. HLA is the only genetic system so far shown to have strong association with this cancer.

**HLA (histocompatibility locus antigen)**

The major histocompatibility gene complex on the short arm of chromosome 6 comprises six recognized loci called HLA-A, -B, -C, -DR, -DQ and -DS. There are at least 18 recognized distinct alleles at the HLA-A locus and 32 distinct alleles at HLA-B locus. Each allele determines a product (antigen).

**Haplotype**

Because of their close linkage, the combination of alleles at each locus on a single chromosome is usually inherited as a unit referred to as haplotype (for example A2-BW46). Since one chromosome is inherited from each parent, every person possesses two HLA haplotypes (for example A2-BW46, A11-B40).

Example: AB x CD ==> AC - AD - BC - BD

Paternal haplotype A: A2-BW46 B: A11-B40
Maternal haplotype C: A2-B40 D: AW19-B17

**Linkage disequilibrium**

The alleles for HLA vary in frequency and presence among different ethnic groups. The linkage pattern also differs between different human populations. In the general population, certain alleles of one locus tend to be associated with that of another, with a frequency far exceeding that expected if the two genes were segregated independently and separately. This is called linkage disequilibrium and is observed in Chinese patients with nasopharyngeal carcinoma (A2-BW46, AW19-B17).
**HLA and nasopharyngeal carcinoma in Chinese**

*History of Sin 2 antigen*

An oriental B antigen was discovered with high frequency among Singapore Chinese patients with nasopharyngeal carcinoma in a pilot study in 1974. It was designated Singapore-2 (Sin 2) (Simons et al, 1976; Simons and Day, 1977). Meanwhile, an independent study in the USA also found an HLA-antigen (designated HS) occurring with high frequency among the Chinese Cantonese patients with nasopharyngeal carcinoma in California (Payne, Radvany and Grumet, 1975). It is now known that Sin 2 and HS are identical, and it is designated BW46 by the WHO committee on leucocyte nomenclature. There are not three well established associations between HLA and nasopharyngeal carcinoma involving A2, BW46 and B17. Subsequent studies in newly diagnosed Chinese patients with nasopharyngeal carcinoma showed HLA associations with haplotype A2-BW46 and AW19-B17 (Chan and Simons, 1977; Chan et al, 1981) *(Table 19.5)*.

**Table 19.5 Summary of HLA types and their relationship to survival pattern and clinical behaviour of nasopharyngeal carcinoma**

<table>
<thead>
<tr>
<th>HLA pattern</th>
<th>Clinical behaviour and survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>AW19-B17</td>
<td>Short-term survivals&lt;br&gt;Mostly young onset &lt; 30 years&lt;br&gt;Poor CMI, PHA and Mantoux&lt;br&gt;High VCA/EA titres, low ADCC titre&lt;br&gt;Most die within 2 years from onset</td>
</tr>
<tr>
<td>A2-BW46</td>
<td>Intermediate term survivals&lt;br&gt;Older onset &gt; 30 years</td>
</tr>
<tr>
<td>A2 without BW46 or B17</td>
<td>Long-term survivals (40% 5-year survival)&lt;br&gt;Low VCA/EA titres, high ADCC titres.</td>
</tr>
</tbody>
</table>

**Differential HLA frequency distribution**

Differential frequency distributions of HLA antigen are seen among the newly diagnosed Chinese patients with nasopharyngeal carcinoma with regard to the age of onset of the disease. They are as follows:

1. BW46 is confined to older patients (> 30 years old)
2. B17/BW58 is associated with both young and old patients but particularly with younger patients (< 30 years old)
3. B11 and B13 are associated with decreased risk (B13 is associated with younger patients).
**Haplotype distribution and relative risk**

The association of a particular disease with a particular HLA antigen is quantitated by calculating the relative risk. This can be defined as the chance an individual with the disease-associated HLA-antigen has of developing the disease compared with an individual who lacks the antigens.

**Relative risk**

<table>
<thead>
<tr>
<th>HLA</th>
<th>Relative Risk</th>
</tr>
</thead>
<tbody>
<tr>
<td>A2</td>
<td>1.5</td>
</tr>
<tr>
<td>BW46</td>
<td>1.9</td>
</tr>
<tr>
<td>B17</td>
<td>2.1</td>
</tr>
<tr>
<td>Haplotype A2-BW46</td>
<td>3.4</td>
</tr>
<tr>
<td>AW19-B17</td>
<td>2.2</td>
</tr>
</tbody>
</table>

Haplotype or joint occurrence of AW19-B17 and A2-BW46 is associated with a higher risk than B17 or BW46 alone (Chan et al, 1983). This suggests that the risk is associated with haplotype rather than individual antigens. These two pairs are known to be in linkage disequilibrium in the general Chinese population.

Despite the occurrence of 50 or more alleles at the HLA-A and -B loci, only a very limited number emerged associated with nasopharyngeal carcinoma. A11 is rarely found with B17 or BW46, while the association of A2 with BW46 in patients with nasopharyngeal carcinoma is stronger than in control patients. No other stronger linkage is seen than the AW19-B17 association. The different HLA association with varying age of onset suggests that there is heterogenicity within the Chinese population with nasopharyngeal carcinoma. Of the racial groups, there are certain similar HLA findings among Malay and Chinese patients with nasopharyngeal carcinoma - the association with B17 (Chan et al, 1985).

BW46 is not found in the Caucasian population. It does not occur commonly nor is it in linkage with HLA-A2 in non-Chinese Asian populations. The HLA gene association is the most convincing evidence for the role of genetics in the aetiology of nasopharyngeal carcinoma.

**Surgical approaches to the nasopharynx**

There are many surgical approaches to the nasopharynx. The various extracranial approaches include:

1. transnasal-maxillary (transnasal and transantral)
2. transpalatal
3. sublabial mid-facial degloving approach
4. others (transpharyngeal, transmandibular, transcervical, infratemporal fossa approach).

The many surgical approaches attest to the difficulty of surgery in the nasopharyngeal region. More than one surgical approach is often needed to provide adequate and optimal exposure. The transnasal and transantral approaches provide good access only for removing
tumours in the maxilloethmoid region. Often they are combined as a transnasal-maxillary approach (resecting the maxilla and lateral nasal wall) to provide a reasonable access to the nasopharynx. With the sublabial mid-facial degloving technique, the nasopharynx is accessible from both sides of the face. Other approaches for example, transmandibular (adopted for palatectomy), transcervical (for high cervical and disc surgery), transpharyngeal (through the floor of the pharynx above the hyoid) and infratemporal fossa approach (radical skull base surgical procedure transecting the auditory canal, zygoma and petrosectomy) are seldom employed nowadays.

Tumours that are limited to the nasopharynx can be removed by the standard transpalatal approach. Such tumours include minor salivary gland tumours (pleomorphic adenoma, mucoepidermoid tumours), haemangioma, inverted papilloma, melanoma and rhinosporidiosis. A combined intra- and extracranial approach may be required for tumours that demonstrate notable intracranial extension, for example craniopharyngioma. The selection of an appropriate surgical approach depends upon the extent of the tumour and which structures and spaces are involved. This can be illustrated in the surgery of angiofibroma. Postoperative haemostasis can be achieved by postnasal packing and/or a Foley catheter (inflated with 20-30 mL of water) brought out through the nose. The advantage of using a Foley catheter is that should bleeding recur on removing the pressure, the balloon can be re-inflated.

Angiofibroma

The angiofibroma is a benign yet biologically aggressive tumour. It originates almost exclusively from the posterior nasal and nasopharyngeal region in adolescent males. Thus it has been known as juvenile angiofibroma, although cases have been reported in older adults and in females as well. The usual clinical behaviour of the tumour is one of expansive growth with a potential for intracranial extension. Histologically, angiofibroma is composed of fibrous connective tissue interspersed with variable proportions of endothelium-lined spaces. A preponderance of fibrous stroma may indeed allow surgical removal with relative ease in some cases. However, even with advances in surgical and arterial embolization technique, the intraoperative blood loss is still a major concern. The capacity for spontaneous regression of angiofibroma at sexual maturity is doubtful.

Incidence and age

Angiofibroma is a relatively rare tumour and the age of onset is in the second decade. The reported incidence ranges from 1/5000 to 1/50.000 of otolaryngological patients in different countries (Table 19.6). Over a 30-year period (1949-1979), the Head and Neck Service of the Sloan-Kettering Memorial Cancer Centre in New York reported only 31 male patients between the ages of 11 and 21 (median age 14 years) (Witt, Shah and Sternberg, 1983).

Pathogenesis

The tissue of origin remains unknown in angiofibroma. Various theories have been proposed. The fibroblastic theory suggests abnormal growth or response of the connective tissues such as the embryonic occipital plate (chondrocartilage between the body of the
sphenoid and the basiocciput) prior to its ossification at the age of 25 years (Bensch, 1878),
the ventral periosteum of the posterior nasopharyngeal wall (Ringertz, 1938) or the fascia
basalis from the fusion of the pharyngeal aponeurosis and the buccopharyngeal fascia near
the base of the skull (Brunner, 1942).

Table 19.6 Incidence of angiofibroma

<table>
<thead>
<tr>
<th>Author</th>
<th>Country</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Handousa, Farid and Elwi (1954)</td>
<td>Egypt</td>
<td>1/50,000</td>
</tr>
<tr>
<td>Harma (1959)</td>
<td>Finland</td>
<td>1/6000</td>
</tr>
<tr>
<td>Bhatia, Mishra and Prakash (1967)</td>
<td>India (Lucknow)</td>
<td>92 cases in 27 years</td>
</tr>
<tr>
<td>Witt, Shah and Sternberg (1984)</td>
<td>USA (New York)</td>
<td>31 cases in 30 years</td>
</tr>
</tbody>
</table>

The second theory suggests that angiofibromata are hormone-dependent tumours (based
on their predilection to occur in an age group undergoing endocrine changes) and that the
tumours occur as a result of either oestrogen or androgen stimulation due to oestrogen-
androgen imbalance (Martin, Ehrlich and Abels, 1948). Recent documentation of androgen
receptors in these tumours has lent some support to this theory (Lee et al, 1980). The third
theory is that of hamartomatous origin. Osborne (1959) observed some morphological
similarity between the anomalous vessels in the angiofibroma and that of the nasal erectile
tissue in the lamina propria of the choana and nasopharynx. Hamartomatous proliferation of
such vascular or aberrant erectile tissue may be medicated under hormonal influence (Schiff,
1959).

Clinical features

Signs and symptoms

The most common presenting symptoms are nasal obstruction and epistaxis. Less
common symptoms include tinnitus, eustachian dysfunction with conductive loss, facial
swelling, proptosis and diplopia. Clinically, the tumour appears as a reddish-purple nodular
mass on one side of the posterior nares and nasopharynx. It may fill the nasopharynx
completely, displacing the soft palate forward. The tumour can be examined quite easily by
posterior rhinoscopy or transnasally with a flexible nasopharyngoscope.

Site of origin

Angiofibromata have a broad base. They originate from the posterolateral wall of the
nasal cavity and the adjoining superolateral nasopharyngeal wall. The sphenopalatine foramen
is always involved. After radiation, tumours often involute back towards this region (Sessions

Tumour spread

Angiofibromata often grow and extend along natural foramina and fissures, displacing
and distorting the adjacent structures. Larger tumours, however, may erode bone. As they
expand, collateral blood supplies develop. The tumours spread laterally from the
sphenopalatine foramen to the pterygopalatine fossa through the pterygomaxillary fissure.
From this narrow fossa they eventually expand into the infratemporal fossa and the cheek.
They can also extend along the inferior orbital fissure, across the apex of the orbit into the superior orbital fissure. Continued tumour expansion causes pressure erosion of the base of the pterygoid plate and greater wing of the sphenoid. This brings the tumour against the dura of the middle cranial fossa.

Medially, the tumour fills the nasopharynx and distorts the nasal septum, turbinates and the soft palate. It may erode into the posterior ethmoidal and sphenoidal sinuses, allowing direct extension of tumour into the orbit, cavernous sinus and the parasellar region.

**Diagnosis**

The patient's age, sex, symptoms and physical findings are often diagnostic of angiofibroma and allow differentiation from other more common nasopharyngeal tumours, for example nasopharyngeal carcinoma. Nasopharyngeal carcinoma is usually ulcerative and infiltrative with early lymphatic spread. Occasional difficulty arises in clinical diagnosis in a young patient presenting with polypoidal nasopharyngeal carcinoma which may mimic an angiofibroma in symptoms, signs and radiological features. Diagnostic biopsy may be needed prior to planning invasive diagnostic procedures. However, biopsy can cause uncontrollable epistaxis if the tumour is well vascularized. In typical cases of angiofibroma, radiological and angiographic investigations are sufficient to obviate the need for pretreatment biopsy (Sessions et al, 1976).

**Radiological diagnosis**

Radiological findings of the juvenile angiofibroma include:

1. Nasopharyngeal soft tissue mass
2. Widening of the pterygopalatine fissure (anterior bowing of the posterior wall of the maxillary antrum and posterior bowing of the pterygoid plate) is the classical sign in early angiofibroma - however, it is not pathognomonic; similar radiological features have also been observed in schwannomata, fibrous dysplasia and nasopharyngeal cancer (Schaffer et al, 1978; Som et al, 1981)
3. Enlargement of the superior orbital fissure in patients with proptosis
4. Distortion of the nasal septum, erosion and opacification of the paranasal sinuses.

Computerized tomographic (axial and coronal) studies with contrast delineate the tumour and its extension. They show, in detail, the bone and soft tissue of the skull base which is hitherto obscure on plain X-rays. Contrast enhancement differentiate sinus opacification due to tumour invasion from opacification as a result of ostial obstruction.

**Angiographic anatomy and therapeutic embolization**

The angiographic features of angiofibroma are consistent. In the arterial phase there is rapid filling of increased numbers of dilated vessels, followed by the characteristic dense homogeneous blush. Subtraction technique gives excellent visualization of the tumour, and
is useful in detecting the pre-existing extra-intracranial anastomoses. The major arterial supply is always from the ipsilateral internal maxillary artery (Roberson et al, 1979). Collateral blood supplies may come from the ascending pharyngeal artery, contralateral internal maxillary artery and branches of the internal carotid system. Large tumours with intracranial extension are likely to receive major collaterals from the internal carotid system. Such collaterals may also occur in patients whose external carotid arteries have been ligated. Thus angiographic study is of particular importance in managing these groups of patients.

Preoperative therapeutic embolization of the major feeding vessels is a major asset in reducing intraoperative blood loss. Embolization with Gelfoam thromboses the vascular bed, whereas in ligation, collaterals open very quickly. Postembolization fever and facial pain are the two common sequelae of this procedure. One complication is the escape of emboli into the intracranial circulation due to reflux or via unrecognized external-internal carotid anastomoses (Lasjaunis, 1980).

Treatment

Surgery is the treatment of choice for angiofibroma and radiotherapy is generally reserved for unresectable lesions. Primary treatment with radiotherapy has not gained general acceptance despite comparable results (Cummings, 1980; Cummings et al, 1984). The latent effects of radiation on facial skeletal growth and the potential to cause sarcomatous change have dissuaded many from considering this mode of treatment. Cryosurgery, sclerotherapy and electrocoagulation are seldom employed except for treating small accessible recurrences. Androgens or oestrogens, as definitive treatment or as adjuncts in inoperable and recurrent tumours, have produced variable results.

Surgical approaches

More than one surgical approach is often needed for complete removal of an angiofibroma. It may recur if not completely removed. Angiofibromata with intracranial extension should be removed with a combined intracranial approach, particularly when the main blood supply is from the internal carotid artery.

Transnasal-maxillary approach

Tumour in the maxillary antrum and anterior part of the nose may be removed through the sublabial incision. Preoperative ligation or clipping of the internal maxillary artery can be carried out transantrally. Denker's extension of the Caldwell-Luc procedure, resecting bone from the face of the maxilla and lateral nasal wall provides fair access. Exposure is limited, however, by the soft tissue of the lip and nose. The use of lateral rhinotomy or Weber-Fergusson incision improves the exposure but is limited by its unilaterality and mid-facial scarring. Digital manipulation of the lateral tumour extension in the pterygopalatine fossa is made possible by extending the sublabial incision to the maxillary tuberosity. It often needs to be combined with the transpalatal approach to deliver the tumour completely.
**Transpalatal approach**

This approach exposes the nasopharynx and allows extensions into the sphenoidal and posterior nasal fossa. There are many variations to the palatal incision (Wilson, 1957). The U-shaped incision is preferred as it can be extended around the tuberosity of the maxilla to join the sublabial incision to reach the pterygopalatine fossa. In this procedure, after the mucoperiosteal flap is elevated (preserving the greater palatine neurovascular pedicle), bone from the posterior hard palate is removed. Once the tumour has been completely exposed and mobilized, it is removed with the mucoperiosteum of the nasopharynx.

**Sublabial mid-facial degloving approach**

This procedure is essentially a bilateral extended sublabial and transnasal-maxillary approach (Casson, Bonarmo and Converse, 1974; Conley and Price, 1979). It obviates visible scarring and allows adequate exposure to the nasal complex, nasopharynx and mid-third of the face. The initial gingivolabial incision is across the midline from one maxillary tuberosity to the other. The soft tissue on both sides of the face is then elevated subperiosteally up to the infraorbital foramina. The infraorbital nerves are exposed and preserved. Routine intercartilaginous incisions are used, separating the soft tissue of the nose from the upper lateral cartilage as in rhinoplasty. A transfixing septal incision then separates the cartilaginous septum from the medial crura of the alar cartilage. Finally, an incision along the pyriform aperture connects the circumferential septal-vestibular incisions to the sublabial incision. This allows total mid-facial degloving up to the root of the nose and infraorbital foramen. The necessary bone is then resected from the maxilla, the antrum and the lateral nasal wall to provide access to the nasopharyngeal region. This approach also allows the pterygopalatine and infratemporal fossae to be reached. One of its postoperative complications is vestibular stenosis.

**Tumours of lymphoid and haemotopoietic tissue**

These tumours possess no gross characteristics which allow differentiation from the epithelial tumours of the nasopharynx. The symptomatology is similar to that of other invasive tumours occurring in this region. Epistaxis and nasal blockage are the usual presenting symptoms. Pain may signify pressure or invasion of adjacent structures. Biopsy is needed for a definitive diagnosis. Patients with no involvement of regional lymph nodes may have disseminated disease at the time of diagnosis. Management requires a multidisciplinary approach involving the radiotherapist and medical oncologist.

**Malignant lymphoma**

About 25% of malignant lymphomata are of extranodal origin, with Waldeyer's ring, second only to the stomach, as the most common site of involvement (Freeman, Berg and Cutler, 1972). Within the Waldeyer's ring, the faucial tonsil is the most frequent site of lymphomatous involvement, followed by the nasopharynx. In a review of the world literature from 1935 to 1969, Banfi et al (1970) reported malignant lymphoma to represent between 1% (in south-east Asia) to 43% (in Europe) of all nasopharyngeal tumours. The low incidence in the east Asian population is related to the prevalence of nasopharyngeal carcinoma.
Diagnosis and treatment

Advances in immunopathology using tissue markers and monoclonal antisera have further classified lymphomata into various immunological types (Parker, 1979). The normal or reactive lymphocyte population is heterogeneous whereas a malignant, non-Hodgkin's lymphoma is a clone of lymphocytes carrying specific surface markers. These markers enable the non-Hodgkin's lymphoma to be identified and typed as being B or T cell. Hodgkin's disease has no monoclonal marker pattern and is based on a morphological diagnosis. Staging follows the definitive diagnosis. In localized disease, radiation alone is the treatment of choice. For systemic disease, chemotherapy alone or in combination with radiation is the preferred treatment (Jacobs, Weiss and Hoppe, 1986).

Plasmacytoma

Plasmacytic dyscrasia occurs in the bone marrow (medullary) and any structure containing reticulo-endothelial tissues (extramedullary plasmacytoma). They are histologically similar. The extramedullary plasmacytoma occurs most commonly in the head and neck region and has a predilection for the upper aerodigestive tract, especially the nasal sinuses and the nasopharynx (Batsakis and Fries, 1964). It can be solitary or multiple in form. It is relatively rare and the incidence compared to multiple myeloma is 1:40 (Pahor, 1977). Males predominate in the ratio 3:1 and the peak incidence is in the fifth decade.

Diagnosis and treatment

Once the histological diagnosis is confirmed, it is necessary to exclude multiple and systemic involvement. Investigations would include radiological skeletal survey, haematological evaluation, bone marrow trephine biopsy and aspirate, and immunoglobulin electrophoresis study. Solitary plasmacytoma in the head and neck is generally treated by excision (depending on the site and accessibility) or local radiotherapy. The clinical course of plasmacytoma of the nasopharyngeal or upper aerodigestive tract is unpredictable (Booth, Cheesman and Vincenti, 1973). Despite treatment, it may recur or eventually evolve into a systemic form after a variable latent period. Hence long-term follow-up and surveillance is necessary.

Paediatric nasopharyngeal tumours

Paediatric nasopharyngeal tumours are rare. They cause respiratory obstruction, and create problems in diagnosis and management. Besides adenoid hypertrophy (which is unusual in early infancy) and antrochoanal polyp, the differential diagnoses of a nasopharyngeal mass include:

1. teratoid: dermoids, teratoma, epignathi
2. neuroectodermal: encephalocele, brain heterotopia, meningioma
3. dysontogenetic: chordoma, craniopharyngioma
4. miscellaneous: cysts, haemangioma, hamartoma, rhabdomyosarcoma.
**Teratoid tumours**

Most of the tumours arise from the midline or lateral wall of the nasopharynx and may be attacked to the palate. Females outnumber males by 6:1. In contrast to dermoids, teratomata of the nasopharynx are recognized later in infancy (Foxwell and Kelham, 1958).

**Dermoids or hairy polyps**

This is the commonest variety. They probably arise from inclusion errors during the fusion of the lateral palatine process. They are often pedunculated and covered by hairy skin containing dermal glands. Occasionally the main tumour mass is connected to an intracranial portion through a perforation in the skull base. Histologically, they are bidermal with fibroadipose tissue, bone, cartilage and fragments of striated muscles.

**Teratomata**

These are more complex than the dermoids in structure. Histologically, they are tridermal with nervous tissue. They are frequently associated with deformities of the skull (anencephalia, hemicrania and palatal fissures). Teratomata grow aggressively and, in this aspect, are true neoplasms. Unlike teratomata elsewhere in the body, nasopharyngeal teratomata have not been reported to undergo malignant degeneration (Willis, 1968).

**Epignathi**

This is the least common variety. It consists of the well-formed organs and limbs of a parasitic fetus. Highly developed teratoid tumours are much rarer in the head and neck; they often result in stillbirths.

**Basal encephalocoeles and brain heterotopia**

The human nasopharynx is closely related to the embryonic development of the neural tube. The juxtaposition of the nasopharynx and the prosencephalon may further account for the very rare nasopharyngeal neuroectodermal tumours, for example basal brain heterotopia. Generally encephalocoeles occur in approximately 1/4000 births and less than 10% are of the basal type (Blumenfeld and Skolnik, 1965). Among the basal encephalocoeles, the sphenopharyngeal type is the most common. Within the nose and nasopharynx it can cause obstruction and deform the upper airway. The intranasal sac may be mistaken for a nasal polyp. It must be differentiated from nasal glioma and brain heterotopia.

**Chordoma and craniopharyngioma**

Chordoma is a slow-growing tumour of low malignancy. The craniocervical form occurs along the embryonic craniocervical axis of the notochord bar - the clivus, nasopharynx and first two cervical vertebrae. It erodes bone extensively with displacement of the surrounding structures, making complete surgical removal difficult (Mabrey, 1935; Batsakis and Kittleson, 1963). This tumour is not very sensitive to radiotherapy.
Few cases of craniopharyngioma in the nasopharynx have been described (Johnson, 1962). It is probably derived from remnants of the Rathke's pouch and the craniopharyngeal canal. Devoid of a definite capsule, the tumour proper is soft with multiple septa separating the cystic spaces. Calcification may be present. Clinically, the intracranial portion of the tumour may cause increased intracranial pressure, endocrine disturbances and retarded sexual development. Surgical decompression may be required to relieve the raised intracranial pressure. Complete removal of the main cyst necessitates a subfrontal approach and can be difficult (Matson and Crigler, 1969).

**Nasopharyngeal cysts**

Nasopharyngeal cysts occur in the roof and the lateral wall. They include the Rathke's pouch cyst, Thornwaldt's cyst from the pharyngeal bursa and branchial cleft cyst (on the lateral wall) (Taylor and Burwell, 1954).

**Symptomatology**

The clinical picture will depend on the size, nature and site of the tumour. Choanal obstruction may give rise to snuffling and rhinorrhoea. Long pedunculated tumours may cause intermittent attacks of coughing, apnoea and dysphagia. Sessile tumours may block the nasopharyngeal airway completely and distort the palate, impeding mouth breathing and feeding. Nasopharyngeal obstruction is often dramatic in the first few months of life as infants are obligate nose breathers (Moss, 1965; Swift and Emery, 1973). The risk of asphyxia and difficulty in feeding is greater in nasopharyngeal tumours than in bilateral choanal atresia. Any intracranial communication of the tumour always predisposes the infant to the threat of cerebrospinal fluid rhinorrhoea and ascending meningitis.

**Radiological investigations**

Radiological investigations include plain X-ray tomogram and computerized tomographic study.

Plain lateral skull X-ray may show a soft tissue mass obstructing the nasopharynx and displacing the soft palate anteroinferiorly. Adenoid hypertrophy in early infancy is very unusual. A choanogram may outline the attachment of the nasopharyngeal mass and demonstrate the patency of the choana. A tomogram of the base of skull may be necessary to exclude intracranial extension particularly through the sella turcica and sphenoooccipital synchondrosis. The most useful investigation is the CT scan. It provides information on the nature of the tumour, its site of origin and both its intra- and extracranial extension.

**Surgical management**

Large teratoid masses may cause acute respiratory obstruction and need immediate surgical removal. The more common pedunculated 'hairy polyp' can be removed easily by a snare. In less urgent cases of nasopharyngeal tumour, endoscopic assessment under anaesthesia and careful biopsy is required to establish the diagnosis before definitive treatment. Laryngoscopy and bronchoscopy should also be carried out to exclude other causes of upper airway obstruction. The transpalatal approach is used to remove sessile tumours.
Intracranial communication is uncommon but must be excluded prior to any surgery. The existence of such a communication may necessitate combined intra- and extracranial surgery.

Appendix 19.1 Stage classification of nasopharyngeal carcinoma currently in use

(1) UICC TNM Classification

UICC (1978)

(a) Anatomical regions and sites

Posterior-superior wall: extends from the level of the junction of the hard and soft palates to the base of the skull.

Lateral wall: including the fossa of Rosenmüller.

Inferior wall: consists of the surface of the soft palate.

Note: The margin of the choanal orifices including the posterior margin of the nasal septum is included with the nasal fossae.

(b) TNM pretreatment categories

\[ T: \text{ primary tumour} \]

Tis  pre-invasive carcinoma (carcinoma in situ)
T0  no evidence of primary tumour
T1  tumour confined to one site (including tumour identified from positive biopsy)
T2  tumour involving two sites
T3  tumour with extension to nasal cavity and/or oropharynx
T4  tumour with extension to base of skull and/or involving cranial nerves
Tx  the minimum requirements to assess the primary tumour cannot be met

\[ N: \text{ regional lymph nodes} \]

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

\[ M: \text{ distant metastases} \]

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.
(c) Stage-grouping (1978)

Similar to AJC (1978) stage-grouping.

(2) The American Joint Committee for Cancer Staging and End-Results Reporting Stage Classification

AJC/TNM Stage Classification (1978)

Anatomy

The anterior limit of the nasopharynx is the choana, through which it is continuous with the nasal cavity. Its roof is attached to the base of skull and slopes downward to become continuous with the posterior pharyngeal wall. The lateral wall is composed of the torus tubarius, the eustachian tube orifice, and that portion of the mucosa of the fossa of Rosenmüller extending up to its apex and junction with the roof. The inferior limit of the nasopharynx is level with the plane of the hard palate.

Anatomical site

Posterior-superior wall (vault).
Lateral wall.

TNM categories

\[ T: \text{ primary tumour} \]

\[ T_0 \text{ no evidence of primary tumour} \]
\[ T_{is} \text{ carcinoma in situ} \]
\[ T_1 \text{ tumour confined to one site of nasopharynx or no tumour visible (positive biopsy only)} \]
\[ T_2 \text{ tumour involving two sites (both postero-superior and lateral walls)} \]
\[ T_3 \text{ extension of tumour into nasal cavity or oropharynx} \]
\[ T_4 \text{ tumour invasion of skull or cranial nerve involvement, or both} \]

\[ N: \text{ cervical lymph nodes (midline nodes are considered as homolateral nodes)} \]

\[ N_x \text{ nodes cannot be assessed} \]
\[ N_0 \text{ no clinically positive node} \]
\[ N_1 \text{ single clinically positive homolateral node 3 cm or less in diameter} \]
\[ N_2 \text{ single clinically positive homolateral node more than 3 cm but not more than 6 cm in diameter or multiple clinically positive homolateral nodes, none more than 6 cm in diameter} \]
\[ N_3 \text{ massive homolateral node(s), bilateral nodes or contralateral node(s)} \]
$M$: distant metastasis

Mx not assessed
M0 no (known) distant metastasis
M1 distant metastasis present

Stage grouping

<table>
<thead>
<tr>
<th>Stage</th>
<th>T</th>
<th>N</th>
<th>M</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>T1</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>II</td>
<td>T2</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>III</td>
<td>T3</td>
<td>N0</td>
<td>M0</td>
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<td></td>
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<td>N1</td>
<td>M0</td>
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<td>T1-4</td>
<td>N2-3</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T1-4</td>
<td>N0-3</td>
<td>M1</td>
</tr>
</tbody>
</table>

(3) Ho's classification (Ho, 1978)

TNM categories

Tumour

T1 tumour confined to the nasopharynx (space behind the choanal orifices and nasal septum and above the posterior margin of the soft palate in its resting position)
T2 tumour extending to the nasal fossa, oropharynx or adjacent muscles or nerves below the base of the skull
T3 tumour extending beyond T2 limits and subclassified as follows:
  T3a bone involvement below the base of the skull (including floor of sphenoid sinus)
  T3b involvement of base of skull (including the lateral and posterior walls of sphenoid sinus)
  T3c involvement of cranial nerve(s)
  T3d involvement of orbit, laryngopharynx (hypopharynx) or infratemporal fossa

N: regional lymph nodes

N0 no palpable nodes (excluding nodes thought to be benign)
N1 node(s) wholly in the upper cervical level, bounded below by the neck crease extending laterally and backwards from or just below the thyroid notch (laryngeal prominence)
N2 palpable node(s) between the crease and the supraclavicular fossa, the upper limit being a line joining the upper margin of the sternal end of the clavicle and apex of an angle formed by the lateral surface of the neck and the superior margin of the trapezius
N3 palpable node(s) in the supraclavicular fossa and/or skin involvement in the form of carcinoma en cuirasse or satellite nodules above the clavicles
Stage-grouping

I tumour confined to the nasopharynx (T1 N0)

II tumour extending to nasal fossa, oropharynx or adjacent muscles or nerves below the base of the skull (T2) and/or N1 involvement (T1 N1, T2 N0 and T2 N1)

III tumour extending beyond T2 limits or with bone involvement (T3) and/or N2 involvement (T1-2 N2, T3 N0-1)

IV N3 involvement, irrespective of the stage of the primary tumour (T1-3 N3)

V haematogenous metastasis and/or involvement of the skin or lymph nod(s) below the clavicle (T1-3 N0-3 M1).
Chapter 20: Headache and facial pain

K. J. Zilkha

Headache

It is probable that nearly everyone has had a headache at some time. It is only when the single attack is exceptional in some way, or if there are repeated attacks, that the patient has to seek the advice of a doctor. There are features in the timing, quality and the situation of the headache which may help in its explanation. The severity of the headache by itself is not a good indicator of its cause.

Pain is such a subjective symptom that, at best, the degree of discomfort which is associated with it is only one feature. If it is of sudden onset, and particularly if associated with neck stiffness, one is justified in considering subarachnoid haemorrhage as a likely cause. A ruptured intracranial aneurysm, or less commonly an arteriovenous malformation, may be the underlying pathology. The circumstances of the pain at the onset can be of material significance. A history of trauma, or unusual effort, or infection may be relevant. Many patients recall an injury, which later proves to be irrelevant. Conversely, many patients with a subdural haematoma and headache do not remember the knock on the head. The drinker who expects a hangover headache, but does not remember the head injury, is a good example of this.

Early morning headache, particularly headache which interrupts sleep, is usually more serious, and typically is a feature of raised intracranial pressure. Aggravation by change in posture, such as bending down, or by coughing and sneezing can be expected. It is, however, when it becomes a particular feature of the headache that it may underline its seriousness. It is worth enquiring as to whether the pain is relieved by lying down, as nearly all patients with cluster headache and many with migraine, find that this aggravates the situation.

A throbbing quality is usually associated with increased vascularity, as in a feverish headache or that associated with infection. A sharp pain may indicate that the cause is local, and affects trigeminal, occipital, or upper cervical roots.

The site of the pain can be of diagnostic help; it may be related to ear infection, dental abscess or sinusitis. The finding of localized tenderness is another pointer to the diagnosis.

There may be unusual circumstances, such as the headache which may commence during sexual intercourse. It is very uncommon but so severe and frightening that a vascular event is suspected. It can be reproduced on subsequent similar occasions - and naturally can cause anxiety. Usually there is no evidence of a subarachnoid haemorrhage, and the condition is self-limiting. The male is the usual sufferer, but on two occasions in the author's experience, the female partner experienced the headache. The explanation seems to be the position of the head and neck during lovemaking.

Single attacks of headache demand an intensive search for the cause. Repeated attacks have the advantage of establishing a pattern, with more clues and information about the incidence.
Examination may reveal evidence of raised intracranial pressure with optic disc swelling, meningeal irritation with neck stiffness, other focal neurological signs, raised blood pressure or tenderness over the superficial temporal artery. One has to be aware that optic neuritis may also show disc swelling and be associated with headache or orbital pain. Usually it is possible to discover the accompanying central scotoma, and of course there is marked impairment of visual acuity. Neck stiffness may accompany acute febrile illness, or any severe occipital and neck pain, and may be unrelated to the presence of blood or inflammatory change in the meninges. It is generally recognized that hypertension is not usually accompanied by headache; it may, however, be the presenting symptom of rapidly rising blood pressure as in toxæmia of pregnancy, renal failure or phaeochromocytoma. Examination of the fundus may show the expected arterial changes with haemorrhages and exudates. Over the age of 60, one always suspects temporal arteritis. There may not be the expected tenderness over the superficial temporal artery and the sedimentation rate may not show the big rise which is customary. Temporal artery biopsy and a trial of steroids are desirable to avoid the very real risk of sudden onset of blindness.

Glaucoma may be responsible for an acute onset of headache and measurement of the intraocular pressure is essential to exclude it.

**Migraine**

It is not unusual for the first attack of migraine to raise alarm and anxiety. The pain may be accompanied by visual disturbance, motor and sensory symptoms, confusion and vomiting. The circumstances and subsequent total recovery may give a clue to the diagnosis. Subsequent attacks will be less alarming, though still requiring management and treatment. The expectation of unilateral pain with visual disturbance and nausea and vomiting is only true for classical migraine. Many more suffer from common migraine, with repeated attacks of headache which need not be unilateral and in some instances may be bilateral with every episode, and not necessarily accompanied by other symptoms (Friedman, 1962; Lance, 1982). Sufferers from classical migraine may also complain at other times of simple headaches with or without nausea. Hemiplegic migraine, associated with unilateral numbness, tingling and perhaps weakness, is usually more incapacitating. Vertebrobasilar migraine is also accompanied by vertigo, ataxia and often slurring of speech. It is rewarding to study the role of trigger factors and the relationship of the headaches to the menstrual cycle, certain foods, stressful situations, and conditions at work. The differential diagnosis from muscle contraction headache can be particularly difficult.

**Ergotamine-dependent headache**

Ergotamine has long had a reputation of being an effective drug for the treatment of acute migraine, preferably taken very early in the attack. It is very tempting for sufferers to take ergotamine as soon as they think an attack is coming on. This may lead to regular medication with ergotamine in tablet or suppository form, amounting to 3-5 mg weekly. What usually follows is that the headaches become more frequent, occurring many times a week. They no longer resemble the original migraine attacks, although an actual migraine may supervene. In the majority, they are simple, severe headaches which apparently respond to ergotamine. The treatment is to stop the ergotamine. Invariably, there is a severe attack of headache within a week of stopping, but provided no recourse to ergotamine is made, the
vicious cycle of headache and ergotamine therapy is finally broken (Rowsell, Neytan and Wilkinson, 1973; Legg, 1976).

**Chronic paroxysmal hemicrania**

This is a relatively rare form of headache with recurrent unilateral attacks only responding to indomethacin. It is much more common in men, and there are no other distinguishing features. After a successful trial of indomethacin, the condition usually improves and it is then possible to withdraw the medication gradually over a period of a few months.

**Cerebral tumour**

It is no exaggeration to say that many sufferers from headaches are seeking reassurance that it is not caused by a cerebral tumour. That a primary or secondary tumour is a rare cause of headache is, in itself, not reassuring to the individual. It is very unusual for a tumour to give rise to headache early in the history, though raised intracranial pressure may cause early morning headache with nausea, vomiting and papilloedema.

Localization of the headache is usually a poor indication of the site of the tumour, with the exception of a rapidly expanding growth. It is generally accepted that the pain is due to stretching and distortion of pain fibres associated with the meninges and larger vessels. The presence of other symptoms such as somnolence, intellectual impairment, disturbances of the senses of smell or vision adds to the significance of the headache. Epilepsy of late onset is another important clue. These symptoms also help with the localization of the tumour. Exacerbation of the headache by change in posture, such as bending, coughing or sneezing is not specific to cerebral tumour or raised intracranial pressure. Localized tenderness is worth looking for, and palpation of the head may be useful aid to the diagnosis. An intensification of the headache may indicate haemorrhage or cystic degeneration. CT scanning has revolutionized the investigation of brain tumours. One word of warning: when a tumour is strongly suspected, a lumbar puncture is inadvisable.

**Benign intracranial hypertension**

This is an uncommon condition frequently associated with headache and usually affecting overweight women. There is a marked rise in the intracranial pressure with papilloedema and visual symptoms and signs. CT scanning of the brain reveals normal or small ventricles, suggesting the diagnosis. Lumbar puncture is safe and gives the only accurate indication of the degree of rise in intracranial pressure, and the subsequent course of the condition (Boddie, Banna and Bradley, 1974).

**Cerebral aneurysm**

A sudden severe pain may be the dramatic presentation of a ruptured cerebral aneurysm with a subarachnoid haemorrhage. It is quite common for a second episode to occur before the significance of the first is appreciated. It is not possible to localize the site of the aneurysm along the circle of Willis or the adjoining vessels, by the situation of the headache. It is often frontal and later may be accompanied by pain in the neck, exacerbated by head and
neck movement. Associated symptoms, or signs, such as a complete third nerve palsy admirably localize the aneurysm, in this instance along the posterior communicating or basilar arteries. The aneurysm may be silent until the moment of rupture though there are many instances when the unilateral pain has been present for weeks or months. It is usually persistent, and localized to one or other orbit and is presumably associated with expansion of the aneurysm. Sadly, CT scanning may fail to reveal the aneurysm and it is the author's usual practice to recommend digital subtraction angiography to try and establish whether an aneurysm is present. It is safer than carotid and vertebral angiography.

Temporal arteritis

Although the clinical picture is well recognized, the headache may not be at all severe, and may indeed be absent. It is not uncommon for the patient to appear depressed, or indeed to be so as a consequence of the headache. If the condition is suspected, it is really not safe to await the result of the artery biopsy. Immediate treatment with steroids is recommended. The diagnostic histological criteria persist for a few days after starting steroid therapy.

Meningitis

Acute bacterial meningitis is dramatic in onset with high fever, severe headache and generalized symptoms of the infection. Drowsiness and neck stiffness suggest the diagnosis. Subarachnoid haemorrhage may also be associated with fever, but generally not above 38°C. Symptoms and neurological signs, of cerebral involvement usually indicate a meningoencephalitis or cerebral abscess. Viral meningitis is now probably more common than bacterial meningitis - but it is imperative to perform a lumbar puncture to aid the diagnosis. Tuberculosis with or without the presence of cerebral tuberculomata, remains an important cause of meningitis. The onset is often insidious over several weeks, accompanied only by a low grade fever, headache and malaise. CT scanning is not generally available and often it is this examination which demonstrates the presence of a single tuberculoma, frequently in the posterior fossa or there may be multiple lesions. Neck stiffness is still a cardinal sign of any meningal infection, together with a positive Kernig's sign. Examination of the cerebrospinal fluid should distinguish the various causes of meningitis. Carcinomatous and other malignant invasion of the meninges can also present dramatically with headache and perhaps vomiting and neck stiffness. A low sugar content in the cerebrospinal fluid is still in keeping with the presence of mononuclear cells and a raised protein level. It is the demonstration of malignant cells in the centrifuged deposit which makes the diagnosis.

Cough headache

Any headache, from whatever cause, can be expected to be aggravated by coughing. Typically, headache associated with raised intracranial pressure is exacerbated by coughing, sneezing, straining at stool, as well as bending. There is a form of headache which is precipitated by coughing, and with no other accompanying symptoms or signs, which is benign and with a limited natural history of weeks or months. The pain is very severe, if brief, and only comes on with coughing. It improves slowly over the course of many months and, perhaps surprisingly, a more rapid recovery can follow lumbar air encephalography (Symonds, 1956). CT scanning shows a normal ventricular system.
Cluster headache

Described by Harris (1926) as periodic migrainous neuralgia, the clinical syndrome of unilateral severe pain with a blocked nostril and redness of the eye, occurring daily in bouts, has now come to be generally recognized as cluster headache, after the description by Kunkle et al (1952). There is usually a remarkable regularity in the timing of the attacks, by day or night. The pain is intense, and may last an hour or more. Many sufferers admit to knocking their head against the wall, and to feeling suicidal. Attacks occur every six months or a year, and usually last a few weeks. Missing out one year naturally gives hope of a more lasting remission, only to be disappointed. It is very rare in women, and the author's youngest sufferer was 21 years of age. Rarely, both sides of the head can be affected at different times. Symonds (1952) advocated the use of ergotamine tartrate by injection for the prevention of attacks. Other forms of ergotamine tartrate preparations have also been used including an effervescent tablet and by inhalation. Lithium (Ekbom, 1981) has also been used with success. Inhalation of oxygen may also cut short an attack (Kudrow, 1981).

A comparison between migraine and cluster headache is shown in Table 20.1.

Table 20.1 Comparison of migraine and cluster headache

<table>
<thead>
<tr>
<th></th>
<th>Migraine</th>
<th>Cluster headache</th>
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<tbody>
<tr>
<td>Onset</td>
<td>Adolescence</td>
<td>Adult life</td>
</tr>
<tr>
<td>Sex incidence</td>
<td>Female:male, 2:1</td>
<td>Male:female, 10:1</td>
</tr>
<tr>
<td>Family history</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Eyes</td>
<td>Visual aura common</td>
<td>One eye waters</td>
</tr>
<tr>
<td>Nausea and vomiting</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Nature of pain</td>
<td>Pulsating, hemicranial</td>
<td>Constant pain in region of one eye</td>
</tr>
<tr>
<td>Patient's reaction</td>
<td>Lies down</td>
<td>Walks about</td>
</tr>
<tr>
<td>Duration</td>
<td>3-12 hours</td>
<td>Less than 3 hours</td>
</tr>
<tr>
<td>Frequency</td>
<td>1-8 per month</td>
<td>Daily, often at night</td>
</tr>
<tr>
<td>Menstruation</td>
<td>Related</td>
<td>Not related</td>
</tr>
<tr>
<td>Prolonged remissions</td>
<td>Rare</td>
<td>Common</td>
</tr>
</tbody>
</table>

Muscle contraction (tension) headache

The characteristics of headache are never less defined than when associated with muscle tension. Described as 'band-like', 'a tightness', 'a heavy weight on top', it is usually bilateral, insidious in onset, frequent and often associated with other symptoms of tension. Although vomiting is rare, it may be accompanied by nausea, particularly following heavy doses of medication. Although when typical it can easily be distinguished from attacks of classical migraine, it is not unusual to encounter difficulty in differentiating it from common migraine. Both causes can be present, and many sufferers learn to differentiate between the two.
**Temporomandibular joint dysfunction**

Dental malocclusion and arthritic changes in the temporomandibular joints are relatively common. It may present with otalgia and pain radiating up towards the temples, along the jaw and upper neck or localized to the joint. It is not always related to mastication, and can present without localized tenderness and with no indication of a dental problem. It would be relatively easy to diagnose if the pain followed the use of new and ill-fitting dentures, with trismus and pain on chewing. Dental examination and study of jaw movement will help, but more detailed study of the bite and X-rays of the joints may be necessary.

**Cervical spondylosis**

It has been estimated that 60% of people over the age of 40 have some degree of spondylosis. The usual presentation with neck ache, and pain radiating down one or other arm is frequently inconsistent with the severity of the radiological changes. Whilst even relatively moderate changes can be associated with pain, more severe arthritic changes can be completely pain free. Headache is an uncommon presentation and is more likely to be associated with muscle contraction, itself related to the neck symptoms. Occasionally a high cervical lesion at C2 is associated with pain over the occipital region.

**Depression**

Any severe pain, persisting relentlessly, will be associated with depression. Both reactive and endogenous depression can in turn be associated with headache. A detailed history of the onset and which symptom came first, the headache or the depression, is usually inadequate. Personal problems at home or at work are all too common. The headache can be quite persistent and poor sleep, loss of appetite and loss of affect confirm the possible diagnosis. It is essential to carry out a full neurological examination, and to be aware of any change in the characteristics of the pain. Depression can coexist with other causes and antidepressant therapy can be expected to help with the depression and the headache. Certain occupations demand attention to detail and the tasks are necessarily repetitive. Sewing machinists are one such group. They work long hours at home, are paid per item, and develop quite severe headache. They invariably deny any depressive symptoms, but respond quickly to antidepressant treatment. Patients prescribed monoamine oxidase inhibitors should always be warned not to eat foods rich in amines. Blackwell and Mabbitt (1965) identified tyramine in cheese as the cause of the headache and hypertensive reaction which may follow.

**Facial pain**

As with headaches, facial pain may be a manifestation of many and various conditions. The characteristics of the pain, its severity, situation, radiation, precipitation and associated symptoms may give a clue to the cause. Detailed clinical examination may reveal dental tenderness, or impaired facial sensation.
Infection

Sinusitis

This is considered more fully in Chapter 6. It is an obvious cause if it is associated with localized tenderness, mucopurulent rhinorrhoea and preceded by a history of coryza. It may be associated with a generalized headache.

Dental infections

Pain on chewing, with associated localized tenderness, may be the only abnormal finding. Radiological evidence is helpful. A persistent root infection may escape detection for some time. The pain may radiate along the jaw or be referred to the ear.

Ear and parotid infection

Otalgia or pain over the mastoid area, associated with otorrhoea, may suggest the diagnosis. Parotitis may present with headache and tenderness in the preauricular region and it may be several days before the swelling of the gland makes the diagnosis more obvious.

Herpes zoster

The pain may precede the appearance of the vesicles, usually in the distribution of the ophthalmic or maxillary branches. Ocular complications are common with persistent sensory impairment. Postherpetic neuralgia is much more common in those over the age of 60. Analgesics, antidepressant therapy and transcutaneous nerve stimulation may all be needed in what can be a severe, protracted and painful condition.

Ocular causes

Refractive errors and ocular palsies may be associated with pain in the eye as well as headache. Inflammatory lesions such as iritis or uveitis are also painful. The prompt diagnosis and treatment of glaucoma is rewarding in the early relief of pain and prevention of visual impairment.

Dental occlusion and temporomandibular joint dysfunction

Despite early orthodontic treatment many still have problems with malocclusion in adult life. Pain and tenderness along the jaw, and the examination of the bite, may suggest the diagnosis. A prosthesis may both improve alignment and relieve pain. Early arthritic changes in the temporomandibular joints may be responsible for unilateral or bilateral pain. It is usually aggravated by tension and anxiety and is often relieved by efforts at relaxation and the use of mild tranquilizers.

Salivary calculi

These usually present with pain during and after meals, and there may be a tender swelling of the gland. Commonly involving the submandibular gland and its duct, the pain
can be reproduced by a suitable stimulus such as sucking a lemon. Radiological examination is necessary to demonstrate the offending calculi.

**Trigeminal nerve lesions**

Pain in the face may be accompanied by sensory impairment in one or more of the three branches, ophthalmic, maxillary and mandibular, and motor weakness of the jaw, involving the temporalis, masseter and pterygoid muscles. Involvement of the ophthalmic division is usually accompanied by an impaired or absent corneal reflex. It is usually for the motor defect to accompany sensory changes in the mandibular distribution. Although tumours in the cerebellopontine angle commonly involve the trigeminal nerve, or its central pathway in the brainstem, it is rare for them to present with pain. It is more likely for the patient to complain of unpleasant numbness or altered feeling. The commonest such tumour, an acoustic neuromia, usually presents with deafness. As well as sensorineural hearing loss, and a possible complaint of giddiness and tinnitus, there is usually trigeminal involvement, and there may be an early facial weakness and cerebellar symptoms and signs. Other tumours in the region, a fifth nerve neurofibroma, and a meningioma in the region of the petrous apex, are more likely to present with isolated trigeminal nerve involvement. A nasopharyngeal carcinoma may present with facial pain and sensory impairment. In this instance, there is conductive deafness. Other tumours, in the pituitary region, sphenoid sinus or at the base of the skull, usually present with other cranial nerve lesions. Glioma of the brainstem may involve lower cranial nerves and is accompanied by long tract signs. Paget's disease of the skull may present with facial pain, as well as headache. Unlike the pain in trigeminal neuralgia, it is usually persistent and without exacerbation by trigger factors. X-rays of the skull, tomography, CT scanning and magnetic resonance imaging may all be necessary to establish the cause.

**Trigeminal neuralgia (tic douloureux)**

The pain is usually described as sharp, needle-like or piercing. It comes on in paroxysms which may be very brief or last many minutes. Any movement of the face, including talking, eating and laughing, or touch as in shaving or applying makeup, may trigger off a paroxysm. The pain is very severe with accompanying tears in the eyes. Once the paroxysm subsides, there is a dull ache, which lingers on and serves as a reminder of the previous pain and a warning of the next one. It may affect one or more of the divisions of the trigeminal nerve, commonly the second and third. A sufferer can always localize the trigger area although reluctant to demonstrate it. The condition is much more frequent over the age of 60. Neurological examination, including motor and sensory function of the trigeminal nerve, is normal. Presentation at an earlier age, and with neurological signs, may be due to multiple sclerosis, acoustic neuromia or meningioma near the petrous bone. Radiological, neurophysiological investigation and examination of the cerebrospinal fluid may be necessary to establish the diagnosis. It is not unusual for sufferers to present to a dentist. Dental extractions are common, and some patients are rendered edentulous. Occasionally dental treatment precedes the onset of pain. Although the cause of trigeminal neuralgia is not established, aberrant vessels in the proximity of the trigeminal nerve have been noted during the course of posterior fossa exploration and root section for the relief of pain. Attacks may last weeks or months at a time, with long periods of freedom. There is usually a progressive increase in the severity and duration of the pain, over the next few years. Carbamazepine
relieves the pain in two-thirds of cases, in doses of 100-200 mg three or four times daily (Campbell, Graham and Zilkha, 1966). Side-effects, particularly drowsiness or giddiness, may restrict the dosage. Poor response or no response, as in a third of cases, justifies a trial of phenytoin, in doses of 100-300 mg daily. It is, however, less effective than carbamazepine. Failure of adequate relief of the pain by medication leads to consideration of other measures. The choice lies between phenol injection of the trigeminal nerve, and stereotactic thermocoagulation.

**Trigeminal neuropathy**

The term was first used by Spillane and Wells (1959) to describe persistent sensory disturbance in the trigeminal distribution, with no obvious pathology. Pain may be present. In the author's experience there is usually previous dental treatment and the condition remains localized and non-progressive.

**Glossopharyngeal neuralgia**

The pain is very similar to trigeminal neuralgia, but localized to the lower jaw, in front of the ear, in the ear, or in the throat. It may radiate down the neck. It is much less common than trigeminal neuralgia and typically the pain is triggered by swallowing rather than chewing. Carbamazepine is usually effective, but if side-effects prevent its long-term use, then avulsion of the glossopharyngeal nerve may be necessary.

**Migraine variants**

Usually considered as a unilateral headache, the pain may be periorbital or in the face. The aura may be absent and there may be little or no nausea. Often it is the presence of precipitating factors such as menstruation or dietary triggers, which may suggest the diagnosis.

**Atypical facial pain**

This is not an uncommon presentation of depression. The pain is usually severe and persistent, with only a few of the features of the underlying depression, including interrupted sleep and loss of affect. It responds to antidepressant treatment quite quickly.

**Causalgia**

Typically this is described as a burning or searing pain, which may be accompanied by trophic skin changes. With the increase in head and facial injuries, this condition is becoming more frequent. It may be directly related to the site of the injury or be associated with partial nerve damage. The diagnosis is made on the history. Sympathetic block, or sympathectomy, may be necessary for the relief of pain (Rasmussen and Freedman, 1946).
Chapter 21: Trans-sphenoidal hypophysectomy

R. A. Williams

Operations on the pituitary gland are now nearly always for the removal of pituitary tumours. This form of pituitary surgery is not a complete hypophysectomy, as ideally the normal pituitary is not removed. However, the term 'hypophysectomy' is too widely used and accepted to suggest any redefinition.

Pituitary operations are performed 'from below', which is trans-sphenoidally, or 'from above', which involves a craniotomy. The sphenoid sinus has been approached in a variety of ways, but two have emerged as the most satisfactory. The transthoroid method is used by otolaryngologists; it has the advantage of a wider access and exposure, but the disadvantage of a facial scar. The sublabial trans-septal route is mostly used by neurosurgeons, and there are some centres where otolaryngologists and neurosurgeons operate together.

History

The leading text books of 100 years ago described the pituitary gland as surgically inaccessible. Horsley (1906) was the first to decompress, by the transcranial route, a pituitary tumour which was causing blindness, but did not record this in print for some years. The threat of blindness was then the indication for operation. A number of surgeons tried modifications of the transcranial route, both intradurally and extradurally. Schloffer (1906, 1907) for example made an external incision across the eyebrows and then down to join a lateral rhinotomy. However, these operations seriously damaged nasal function and produced unacceptable scars on the face. In 1909 Cushing started his sublabial trans-septal operation. At about the same time, Hirsch (1911a,b) described an operation through the nose which was later modified to become much the same as the Cushing operation. Cushing (1932) continued with a large series of 159 trans-septal operations and 88 transfrontal operations. The 5-year recurrence rate was 35% from below and 13% from above. He therefore returned mainly to the transfrontal route in 1928. His overall mortality was only 5.8%. His name stands out as a great pioneer of pituitary surgery.

Chiari performed the first transthoroid trans-sphenoid operation in 1912, and this approach was taken-up later, in the 1950s, by a number of otolaryngologists who were removing the pituitary gland as part of the treatment for carcinoma of the breast and prostate. Angell-James (1967a,b), Bateman (1962, 1963), Briant (1964, 1968) and Williams (1978) had large series of operations at that time and it was their experience that reintroduced the transthoroid operations for pituitary tumours. Other approaches from below, such as the transantral operation described by Hamberger et al (1959, 1960, 1961) and the transpalatal operation by Trible and Morse (1965) have not generally been continued.

Indications

Surgery was initially the only method of dealing with pituitary tumours. Later radiotherapy by external beam with X-rays (Jenkins, 1972) or protons (Kjellberg et al, 1968; Lawrence, 1963, 1973) and by implantation of radioactive gold or yttrium (Wright et al, 1970) became effective and safe. Both surgery and radiotherapy were at first non-selective, just
destructive. Microdissection to remove the neoplastic portions and leave the normal parts was introduced, and the idea had great promise. However, for reasons discussed later, this is not always so successful. Radiotherapy is not selective and does not completely destroy a tumour leaving the normal gland intact, especially in the case of well differentiated neoplasms, but it does have a useful and effective role to play sometimes. The effectiveness of a number of drugs on pituitary function has revolutionized the treatment of various pituitary tumours. It is now possible to block certain selected pituitary and hypothalamic secretions and to shrink large pituitary tumours medically. However, the management of pituitary tumours today still requires the use of surgery, radiotherapy, and medication. It should be for the endocrinologist to decide how best to treat each case.

**Removal of normal pituitary gland**

Hypophysectomy for carcinoma of the breast and prostate with secondary metastases is now only rarely indicated. Before effective chemotherapy it was certainly a worthwhile procedure, so long as the time interval between the original diagnosis and the appearance of secondary spread was more than 2 years and that these were in bone only. Those with liver disease did uniformly badly. Dramatic remissions were achieved in only two out of three patients operated on, even when these indications were right. This operation is still occasionally worthwhile, especially in carcinoma of the prostate with secondary tumours in the bone with pain, where it may be possible to obtain a remission of a year or two after all other treatments have been exhausted. Diabetic retinopathy, if at the proliferative stage, can be arrested by removing the pituitary; however, laser surgery has made this indication redundant. In theory there are other hormone-dependent tumours which might be affected by hypophysectomy, but this has not met with any success especially in the case of malignant melanoma.

**Pituitary tumours**

The indications for the removal of pituitary tumours may be for their local effects or for medical reasons. Locally, if a pituitary tumour enlarges upwards, it will result in bitemporal hemianopia and restriction of the visual fields leading to total blindness. There will of course be the intracranial pressure effects of headache and papilloedema. When these tumours extend laterally into the cavernous sinus they will affect the cranial nerves III, IV and VI. Large downward extensions may present in the nasopharynx and sometimes cause cerebrospinal fluid leaks and recurrent meningitis. It may be necessary therefore to decompress, if not completely remove, large pituitary tumours for their serious local effects.

**Medical indications for the removal of pituitary tumours**

**Acromegaly and gigantism**

If it is possible to remove the tumour completely by trans-sphenoid surgery, this is now the most effective treatment for acromegaly. There are large series reported by Hardy (1973, 1975, 1978, 1979) and Williams (1974, 1975) indicating nearly 80% cute, that is reduction of growth hormone to normal, by surgery. High energy radiotherapy and especially proton beam therapy are certainly partially effective treatments for tumours causing acromegaly. So far medical treatment for acromegaly has met with limited success. It is
sometimes possible to shrink these tumours and lower the growth hormone to a variable extent but the results of medical treatment are not predictable. Maybe in the future the medical treatment of acromegaly will become as effective as it is with prolactin-secreting tumours.

Cushing’s disease and Nelson’s syndrome

Cushing's syndrome may be caused by a pituitary tumour, adrenal tumour, ectopic ACTH-producing tumour, and the administration of steroids or certain other drugs. The diagnosis of the cause of Cushing's syndrome is not always straightforward. However, the endocrinologist may ask the surgeon to treat Cushing's syndrome by removing a pituitary tumour. These tumours may be microtumours not apparent on plain X-ray but showing on computerized tomography (CT) scans, or they may be large invasive tumours. Usually the tumour is small and provides the possibility of a successful microdissection with removal of the tumour and sparing the normal pituitary. Large invasive tumours are very difficult to cure surgically or in any other way. If an adrenalectomy has been performed for Cushing's syndrome primarily due to a small pituitary tumour, then some months or years later the pituitary tumour is liable to grow. The result is high adrenocorticotropic hormone (ACTH) and melanocyte-stimulating hormone (MSH) levels with generalized pigmentation as well as the local problems associated with the development of the pituitary tumour - this is Nelson's syndrome (1958, 1960). The results of trans-sphenoid surgery for Cushing's disease can be highly satisfactory with a complete cure of a very serious medical condition. These patients may, however, be extremely unwell on first presentation and it may be advisable to block the production of cortisol medically for some weeks or months until the patient is fit for operation.

Prolactinoma

Before the discovery of prolactin, these tumours were included with chromophobe tumours and it was thought that they did not have any hormonal effects. Prolactin-secreting tumours as their name implies actually secrete the hormone directly in large quantities. However, any suprasellar tumour interfering with the control of prolactin can cause hyperprolactinaemia, but the prolactin level in this case is not as high as with a prolactin-secreting tumour. In women, hyperprolactinaemia mainly causes amenorrhoea and infertility, but sometimes galactorrhoea as well. In men, hyperprolactinaemia may cause impotence, gynaecomastia and skin changes. Before medical treatment for prolactinoma became so effective, surgery was widely used, especially in the USA and Canada. This is where microdissection first came into its own and pressure was put on the surgeon by the endocrinologist to remove the tumour to allow a woman to become pregnant, but not to remove all the normal pituitary. The results of microdissection were sometimes most satisfactory but they depended on the size of the adenoma. Hardy (1979) had a cure rate of 90% with non-invasive tumours of less than 10 mm size, dropping to zero for large invasive tumours. A microadenoma with a piece of normal pituitary attached, which was thought to have been completely removed is shown. However, with special staining it was possible to show that in other parts of this pituitary there were abnormal clumps of prolactin-secreting cells which could have grown into another microadenoma. Prolactin-secreting tumours are now generally treated medically and it is only when this medical treatment cannot be tolerated
by the patient that surgery is occasionally necessary. Even large tumours causing pressure effects will shrink with medical treatment.

**Chromophobe tumours (adenoma and germinoma) in children**

These tumours do not have any direct hormone effects but they may raise the prolactin level indirectly by pressure on the prolactin controlling mechanism. A germinoma is a rare pituitary tumour occurring in childhood and pituitary surgery is sometimes necessary to make the diagnosis so that the patient can then be treated with radiotherapy.

**Other pituitary tumours**

There are other rare pituitary tumours producing a mixture of hormones and these may have to be treated surgically.

**Craniopharyngioma and cordoma**

These tumours may occasionally require trans-sphenoid surgery.

**Contraindications**

It is not safe to proceed with trans-sphenoid hypophysectomy in the presence of nasal or sinus infection, and this should be cleared up first. The removal of a normal pituitary for secondary carcinomatosis of the breast or prostate is rarely indicated now, but is contraindicated if the time between the diagnosis of the primary lesion and the appearance of secondary deposits is less than 2 years. The liver may contain secondary deposits; if there is evidence of this clinically by jaundice, an enlarged liver, or biochemically, patients are not helped by hypophysectomy. The platelet count may be reduced by bone deposits, radiotherapy or chemotherapy and, if it is below 30 x 10⁹/litre troublesome bleeding can be expected. Although this may be overcome by platelet transfusions, it usually means that there will not be any worthwhile recovery. Partial pneumatization or non-pneumatization of the sphenoid is frequently quoted as contraindication to trans-sphenoid hypophysectomy. This is not the case. It is not difficult to pneumatize the sphenoid with a drill. The soft cancellous bone can be removed until the more compact wall of the sphenoid is encountered. Once the shape of the pituitary fossa has been identified the operation can proceed normally. Upward extensions of more than a few millimetres anteriorly or 1 cm posteriorly are not accessible from below, and this is usually a contraindication to a trans-sphenoid operation. The exception is a dumbbell tumour, needing surgery from below and above. It has been found better to stage these operation, starting trans-sphenoidally.

**Preoperative investigations**

**Medical**

Medical investigations include measuring all pituitary hormone levels and often an oral or intravenous glucose tolerance test with measurements of serum growth hormone and insulin levels, as well as blood sugars. These tests are usually arranged by the referring endocrinologist.
Surgical

A full blood count is necessary, including platelets, if there is any question of bone marrow disease or toxicity. Serum electrolytes are also required, because in some conditions, such as Cushing's disease, the potassium levels may be dangerously low for general anaesthesia. Liver function tests are indicated in patients with secondary carcinomatosis. It is safer to have two units of blood cross-matched.

Although blood transfusion is hardly ever required, it may be vital if there is serious bleeding from the cavernous sinus or carotid artery.

Imaging

Routine skull X-rays will show the outline of the pituitary fossa, the pneumatization of the sphenoid, and the size of the frontal sinuses. Sinus X-rays should also be taken to exclude infection. For pituitary tumours it is necessary to show the outline of the upper part of the gland, and the first method for doing this was by an air encephalogram. However, this investigation was not entirely without risk and has now been completely abandoned in favour of CT scans. High quality CT scans with sagittal reconstructions show the whole pituitary gland very satisfactorily. The degree of upward extension is important when deciding whether trans-sphenoid surgery is appropriate and an empty sella with the dura dipping right down into a large fossa is easily seen on a CT scan. A microadenoma of the gland can also be identified. Hardy (1979) has shown that growth-hormone-secreting tumours are more often in the lateral parts of the fossa inferiorly; prolactin-secreting tumours in the lateral part of the gland superiorly; and ACTH-secreting tumours more often central. The author has not found that this can be relied upon. Neurosurgeons prefer to have arteriography of the internal carotid arteries to show their position. There is certainly one occasion where this can be most informative, and that is if a tumour has been treated medically and has apparently regressed. The carotid arteries can then move towards the midline and the space between them may be only a few millimetres, with the residual tumour below. Although the carotid arteries can be identified positively during surgery it is helpful to know where to expect to find them.

Visual fields

Visual field defects begin to appear when the tumour extends upwards to the optic chiasma; this is an important sign as it usually means the trans-sphenoid operation is not indicated and that surgery is better performed from above. It is good practice to have visual fields routinely recorded and to compare them with any subsequent tests, in case for example there is a recurrence of the tumour.

Drugs

Adequate doses of steroids must be given to cover the operation and the postoperative period. For example a basic regimen of steroid cover would be: prednisolone 2 mg hourly intravenously for 24 hours and then prednisone orally 15 mg per day for 5 days, gradually reducing to a maintenance level over the next 2 weeks. However, the details of how this steroid cover is arranged are not critical. Experience has shown that if trans-sphenoid surgery is covered by prophylactic antibiotics, infection is extremely rare. A pack will normally be
left in the nose for about 9 days and to prevent infection the combination of a broad-spectrum antibiotic and an antibiotic which diffuses easily into the cerebrospinal fluid is effective. These antibiotics should be continued until the pack is removed. Desmopressin (DDAVP) should be available for the first few postoperative days to treat diabetes insipidus. Water intoxication is however more dangerous than diabetes insipidus, so this drug should only be given when necessary. When the specific gravity of the urine is low and the urinary output exceeds 500 mL/h for 3 consecutive hours, this is a reasonable indication for the injection of desmopressin 2 microg. Also, a total output of more than 5 litres in 24 hours may indicate that the diabetes insipidus should be treated. Some patients may have been told that they may become thirsty and because of this they drink so much that they can simulate a diabetes insipidus.

**Anaesthesia**

Intubation during anaesthesia may be difficult because of the large tongue in patients with acromegaly, and large laryngoscopes and long endotracheal tubes should be available. Whether the ventilation is spontaneous or controlled is for the anaesthetist to decide, providing that the venous pressure can be kept low. Air embolism does not seem to be a problem because under direct vision it is possible to see if blood is coming from the cavernous sinus or if air is entering it, and the ventilation pressure can be adjusted accordingly. In the author's experience it is not necessary to monitor the neck veins with Doppler probes for air embolism. Hypotension can be most helpful during the dissection of the pituitary. However, in the early stages of the operation it is better to have a normal blood pressure so that all the superficial bleeding can be thoroughly controlled to avoid a subcutaneous haematoma.

**The operation**

**Transethmoid approach**

The operating table is tipped about 25° head-up, and the neck slightly flexed to face the surgeon who stands on the right hand side of the patient. It may be necessary to perform a submucous resection of the nasal septum for access to the right side of the nose. An external incision is made, curved round the medial side of the orbit. The incision is deepened towards the nose medially, so that the lacrimal sac is avoided. Superiorly the supratrochlear nerve is also avoided by straightening the upper 1 cm of the incision. The incision is deepened to the bone by dividing the periosteum, which is then separated from the bone and dissected back past the orbital rim until the anterior ethmoidal artery is exposed. This artery runs through the frontoethmoidal suture and represents the upper limit of the roof of the ethmoids. The artery is sealed above and below by diathermy and is divided; there is usually no bleeding but there may be a small extravasation of fat. The dissection continues to the posterior ethmoidal artery which is left as a landmark. The lacrimal sac is lifted out of its groove and mobilized to avoid tension, when the retractor is inserted. A Luongo retractor is then placed in position. The paper plate of the ethmoid is removed up to the anterior ethmoidal artery and if necessary back as far as the posterior ethmoidal artery; the orbital rim is taken away with a drill or gouge. The frontonasal duct must not be opened widely in acromegalic patients for the soft tissues may later prolapse and obstruct the duct. This removal continues downwards as far as the posterior edge of the lacrimal fossa. A complete
external ethmoidectomy is performed. It is convenient to use forceps either through the external opening or through the nose lateral to the middle turbinate. It is not necessary to remove the middle turbinate unless a wide access is required for the larger tumours, when this step is indicated. The sphenoid sinus is opened through the posterior ethmoid cell and usually the right sphenoid is entered first. The position of the intersphenoid septum can be seen on the submentovertical X-ray of the skull, and it is nearly always necessary to remove this septum. The opening into the sphenoid sinus is widened and the rostrum of the vomer is removed either with back biting Ostrum's forceps or a drill, until full exposure of the front of the pituitary fossa is achieved.

**Trans-septal approach (Cushing's)**

A gingival incision is made. The periosteum is elevated with the mucosa to expose the mucosa of the pyriform opening and floor of the nose. The dissection is continued to the front of the nasal septum. Firm retraction is needed to elevate the upper lip and tip of the nose, using small right-angled retractors. The mucosa is separated from the floor of the nose, to both sides and from the nasal septal cartilage. The bony opening of the front of the nose is thus exposed, and can then be enlarged inferolaterally with a bone punch or drill. This is not always necessary, and may cause temporary or occasionally permanent denervation of the incisor teeth. A submucous resection of the nasal septum is then performed, holding the flaps apart with a large Killian's speculum. A Hardy bivalve speculum is inserted. The anterior wall of the sphenoid sinus is opened with a gouge or drill and removed laterally as far as possible. The pituitary fossa is not identified as easily from this angle as from the ethmoids, especially if the pneumatization is not full. From this point the operation is the same whichever approach is used, except that with the transethmoid approach straight instruments can be used, but with the sublabial approach angled instruments are better. This is because the microscope view and all instrumentation are through the same relatively small opening trans-septally.

**Opening the pituitary fossa and dissection**

A Zeiss operating microscope with a 300 mm objective lens is positioned, viewing through the external incision where a transethmoid approach has been used. The bone over the pituitary fossa is drilled away using a small cutting Burr and this does not damage the dura. A layer of bone is removed extending above to the top of the sphenoid sinus, sideways until the bone thickens, and the full width of the fossa is reached, and downwards to the floor. The floor is not removed, as this is needed to support the muscle plug at the end of the operation. Large tumours often erode through the bone and sometimes through the dura presenting in the sphenoid sinus. At this stage the front of the pituitary fossa may be seen as a pulsating sheet of dura. The carotid arteries are occasionally exposed laterally and can be identified by their thicker appearance. The cavernous and intercavernous sinuses can usually be seen through the dura and the incision made to avoid them. Bleeding from the cavernous sinuses can be a problem but is lessened by good anaesthetic technique and adequate head-up position. The author has never found it necessary to abandon an operation because of bleeding. The venous sinuses are less obvious with expanding tumours than with small normal-sized pituitary glands. If there is an almost 'empty sella', care must be taken to enter the gland or tumour and not the subarachnoid space. If possible a cruciate incision is made, but if there is insufficient space between the superior and inferior intercavernous sinuses, a
transverse incision is adequate. A diathermy incision helps to seal the two layers of dura, but some surgeons prefer a knife.

When the gland is opened a tumour will extrude whereas a normal-sized gland just presents itself. Angell-James dissectors are especially shaped to separate the floor and sides of a normal gland and are useful for microdissection. Where a large tumour is present the normal pituitary is not easy to find, and probably enough of it resides in the diaphragm where the stalk emerges. It is best therefore to remove all the tumour that can be found but not to clean the diaphragm too thoroughly, if tumour does not appear to be attached to it. Where there is some upward extension, and when the tumour has been decompressed the diaphragm may well come down into view. Downward extensions can be followed easily and removed; they may even extend out into the basisphenoid and occiput, and can be drilled away under direct vision. Lateral extensions are more difficult and it is often not possible to see completely the lateral extent of a tumour. Dissection therefore has to be blind. For this situation Hardy curettes are useful but great care has to be taken not to curette too firmly out of sight, or serious bleeding can result. For microtumoures, microdissection is often possible. This involves removing the tumour and leaving an adequate quantity of normal pituitary gland behind. The tumour can usually be identified as a separate adenoma appearing different from the normal gland. Experience is required to be confident about this identification but the tumour may be whiter and of a softer consistency. A plane of cleavage may appear, but it is advisable to take some adjacent normal pituitary for it has been shown histologically that, if a cleavage plane is followed, some tumour may be left behind. If there is doubt in identifying the tumour from the normal gland small pieces may be sent for frozen section. Cerebrospinal fluid will escape if the diaphragm is breached. This can be sucked away until pressure is lowered so the flow will then stop, and the operation can proceed.

Insertion of a piece of muscle into the fossa stops bleeding and escape of cerebrospinal fluid. It also supports the diaphragm and prevents postoperative headache due to stretching. This muscle should be placed in the fossa lightly rather than packed lightly. A large plug may extrude. The muscle can then be covered with Sterispon to prevent adherence of the pack. A 1.25 cm (half-inch) ribbon gauze pack is inserted into the sphenoid sinus, coming out through the right side of the nose. More packing should be placed in the nose to keep the sphenoid pack in position. The sphenoid pack stays in for 9 days but the nasal packs can be removed sooner especially with a Cushing's approach, depending on the stability of the sphenoid pack. With the transethmoidal operation the pack should fill the ethmoid and not bulge into the orbit. Only the skin requires suturing. Padding and bandaging the eye for 12 hours helps to reduce postoperative swelling, but care must be taken not to apply the bandages too tightly and compress the eye.

**Postoperative care**

The patient's temperature often rises during the first 12 hours but this does not necessarily indicate infection and it is probably due to some hypothalamic disturbance. From the second to about the eighth postoperative day the temperature is often subnormal for the same reason. After the nasal packs are removed on the ninth day the treatment is the same as for any other intranasal operation. It is sometimes necessary to remove crusts from the nose, but with saline nose sniffs for 2 or 3 weeks the nose usually becomes clean and healed. Looking into the nose the pulsations of the pituitary may be seen for up to one month.
postoperatively. Medically, steroids ought to be continued until the normal pituitary function has been tested, usually about 6 weeks after operation. The preoperative tests are repeated to assess the effect of the operation and whether it is necessary to continue with steroids and/or thyroxine. After surgery for large pituitary tumours about 25% of patients may have some pituitary hormone deficit and require replacement therapy.

**Complications**

Some patients develop a superficial haematoma or black eye but this is outside the orbital periosteum. It may look alarming for a few days but usually settles within 5 days. If the eye becomes completely closed it is important to test the vision and eye movements to exclude excessive intraorbital pressure. Temporary diabetes insipidus occurs in up to one-third of patients. Cerebrospinal fluid leaks are rare but if they occur following removal of the nasal packs the treatment should be conservative, with the continuation of the antibiotics. The leakage usually settles within 3 weeks. Only if the muscle pack has come out completely would it be necessary to take the patient back to the operating theatre and insert another muscle plug. In the author’s experience this has only occurred once in over 300 tumours operated on and the reason was that the patient decided to restore himself to his normal health. It was on the fiftieth press-up on the tenth postoperative day that the cerebrospinal fluid started to leak. This was followed by meningitis. No other case of meningitis has been encountered in this series and it is generally a very rare complication. Frontal sinusitis occurs especially in acromegalic patients if the frontonasal duct has been opened too widely. With proper operative technique this should be avoided but if it does occur, an operation some months or years later to reopen the frontonasal duct may be necessary. In experienced hands the mortality of this operation is less than 1%.

**Conclusion**

Trans-sphenoid pituitary surgery is a most effective way of treating some pituitary tumours which extend downwards into the sphenoid sinus. Neurosurgeons usually approach the sphenoid via the nasal septum, but otolaryngologists use an external ethmoidectomy approach. CT scans have made it possible to localize microtumours which can often be dissected leaving the normal pituitary intact. The overall management of pituitary tumours should be by an endocrinologist, with a surgeon who can operate trans-sphenoidally, a neurosurgeon and a radiotherapist. The treatment of pituitary tumours should be restricted to centers where these specialists are available.
Chapter 22: The orbit

Valerie J. Lund

The orbit is an area of considerable interest to the otolaryngologist, but one into which he or she may stray with trepidation. It may be involved in pathology arising in the nose and paranasal sinuses and surgical approaches and procedures can involve this area both intentionally and accidentally. However, armed with a clear understanding of the anatomy and careful clinical and radiological assessment, it is an area in which the otolaryngologist should be confident to operate.

Applied surgical anatomy
(Wolff, 1976; Doxanas and Anderson, 1984)

The importance of the anatomy of the orbit lies mainly in its relationships, rather than its contents, with the anterior cranial fossa lying superiorly, the nasal cavity and ethmoid labyrinth medially, the maxillary antrum inferiorly and the infratemporal and middle cranial fossae laterally. It is described as being a roughly quadrilateral pyramid, with its base directed forwards, laterally and slightly downwards. The average volume of the orbital cavity is 26 mL, 70% of which is occupied in normal individuals by retrobulbar and peribulbar structures. The orbit is a fixed bony cavity and an increase of orbital volume of only 4 mL produces 6 mm of proptosis (Gorman, 1978).

The orbital margin provides a protective boundary for the globe as it is stronger than the orbital walls. If the orbit is struck with a round object which diffuses its impact, the orbital rim will withstand considerable force. However, compression of the orbital contents will produce a 'blow-out' fracture of the inferior or medial walls (Smith and Regan, 1957; Dodrick et al, 1971; Rumelt and Ernest, 1972; Mirsky and Saunders, 1979).

The zygoma itself frequently withstands direct blows, but fractures at sites of potential weakness, namely at the zygomaticofrontal and zygomaticomaxillary sutures. Clinically this fracture is evident by depression or flattening of the orbital rim, inferior deviation of the lateral canthus, localized step deformity of the inferior rim at the zygomaticomaxillary suture, tenderness over the zygomatic arch, pain with mastication and ecchymosis of the buccal mucosa. Varying degrees of floor fracture and displacement may be associated with tripod fractures and because of the significant functional and cosmetic deformity which can result, it is important to diagnose and repair them early.

Superior wall

The roof is composed of the orbital plate of the frontal bone, behind which is the lesser wing of the sphenoid. The bone is thin (generally less than 3 mm) except in the sphenoid area and continues to thin with age so dehiscences may appear. The extent of frontal and ethmoid sinus invasion is variable and may go as far as the zygomatic process or optic foramen which can be surrounded by ethmoidal cells. The superior orbital notch lies about 5 mm from the orbital margin in a parasagittal plane, which connects the mental foramen with the infraorbital foramen.
Incisions must be placed to avoid the superior orbital notch and the levator palpebrae superioris muscle. This is done by planning the incision as high as possible, dissecting at right-angles to the skin down to the superior orbital rim. The superior wall is encountered during frontal sinus trephination, frontoethmoidectomy, orbital decompression, exploration of fractures, lacrimal gland excision and orbital exenteration.

**Medial wall**

This wall is of the most significance to the otolaryngologist. Four bones are united by vertical sutures:

1. the frontal process of the maxilla
2. the lacrimal bone
3. the orbital plate of the ethmoid
4. a small portion of the body of the sphenoid.

The rule of 24-12-6 may be applied to the medial wall of the orbit, representing respectively, the average distance in millimetres from the anterior lacrimal crest to the anterior ethmoidal foramen, from anterior to posterior ethmoidal foramen and from posterior ethmoidal to optic canal (Rontal, Rontal and Guildford, 1979). The situation can, however, be very variable, with 16% of patients having no anterior ethmoidal foramen, 30% multiple ethmoidal foramina (Harrison, 1981), and in 4.6% the ethmoidal foramina are absent bilaterally (Shaheen, 1967). In addition, the level of the cribriform plate is variable so that the anterior ethmoidal foramen can only be taken as an approximate guide.

The thin medial wall is a poor anatomic barrier to infections of the paranasal sinuses with the potential complications of orbital cellulitis and abscess, optic neuritis and cavernous sinus thrombosis. An understanding of the anatomy of this region is the key to a number of operations:

1. ethmoidal vessel ligation
2. exploration of medial wall fracture
3. external frontoethmoidectomy
4. orbital decompression
5. transthymosphenoidectomy and hypophysectomy
6. closure of cerebrospinal fluid leaks
7. lateral rhinotomy.

**Inferior wall**

The floor is composed of three bones:

1. the large orbital plate of maxilla
2. the zygomatic orbital plate anterolaterally
3. the orbital process of palatine bone.

The infraorbital foramen is in line with the superior orbital notch, lying half-way along the rim and continues as the infraorbital canal. Anterior, and occasionally the middle, superior
alveolar nerves are given off from the canal which, if damaged, can lead to denervation of
the upper dentition (Harrison, 1971).

Lateral to the nasolacrimal canal is a pit marking the origin of the inferior oblique
muscle which is the only extrinsic muscle to take origin from the front of the orbit and is
encountered in Patterson’s external ethmoidectomy. In 9% the origin of the muscle is
intraperiosteal with no bony attachment which facilitates the operation. Its relationship to the
bony opening of the lacrimal sac is also variable and may be up to 5 mm distant. The lateral
portion of the orbital floor is safer to explore than the medial since the inferior rectus and
inferior oblique muscle are more medially located within the orbit. Thus medial floor blow-
out fractures are more likely to cause entrapment (Gozum, 1976).

The distance between the infraorbital foramen and the optic canal also varies
considerably with an average distance of 46 mm. The posterior wall of the maxilla is about
25 mm from this foramen, which is of importance when contemplating a transantral orbital
decompression. More often it is encountered in orbital fractures or maxillectomy.

**Lateral wall**

This is composed of:

1. the orbital surface of the greater wing of the sphenoid posteriorly and
2. the orbital surface of the zygoma anteriorly.

It may be encountered during orbital decompression, infratemporal fossa surgery,
exploration of fractures and modified craniofacial resections involving lateral orbitotomy. In
intraorbital procedures the danger of damage to the superior orbital fissure or the optic nerve
is minimal. Through the superior orbital fissure at the orbital apex pass cranial nerves III, IV,
VI, the ophthalmic branch of V, and the ophthalmic vein. The fissure is found to be no closer
than 28 mm from the frontal zygomatic suture at the rim. Due to the curved nature of the
orbit and the limited access to this area, it would be difficult and unnecessary to proceed to
this depth during any intraorbital procedure. Likewise, the optic nerve lies 8 mm behind the
medial edge of the fissure and would also be under minimal danger from this approach.
Therefore, as long as a distance of 25 mm from the frontal zygomatic suture is maintained,
safe dissection can be carried out on the lateral orbital wall (Rontal, Rontal and Guildford,
1979).

**Changes with age**

Exploration of the orbit in children is fortunately rare. Growth of the orbits occurs
with the development of the facial skeleton. Initially the orbital fissures are large, the orbital
index high and the volume great so that little change occurs in overall size after 7 years of
age. The infraorbital fissure is present at birth, but the groove may remain dehiscent for some
years and also reaches adult size by 7 years.

Bone begins to resorb with age so that defects may result allowing contact between
orbital periosteum and dura, and the inferior orbital fissure may enlarge. Little discernible
difference can be seen between the sexes.
**Periorbita**

The importance of the orbital periosteum lies in its ability to protect the orbital contents and resist the spread of infection and tumour. It is adherent to the orbital margins, sutures, foramina, fissures and lacrimal fossa and is continuous with dura through the superior orbital fissure, optic canal and ethmoidal canals. It is fixed to the posterior lacrimal crest enclosing the lacrimal fossa and traverses the duct as far as the inferior meatus. It must, therefore, be dissected from its attachments with care, at the very least to avoid troublesome prolapse of fat into the operative field.

The extremities of the tarsal plates in the lids are attached to the orbital margin by strong fibrous structures - the palpebral ligaments. Only the medial ligament is important since its deep portion is attached behind the lacrimal sac. Care must be taken to reflect the whole ligament with attached skin during the incision for external ethmoidectomy.

Fascia bulbi (Tenon's capsule) is a thin membrane surrounding the globe from cornea to optic nerve. Inferiorly it is thickened to form the suspensory ligament of Lockwood, the importance of which becomes evident after total maxillectomy (Manson et al, 1985).

**Radiographic evaluation of the orbit**
(Lloyd, 1975; Bilaniuk and Zimmerman, 1980; Dutton, 1984)

In addition to clinical evaluation, important information is provided by radiological assessment of this area. Techniques available include:

1. plain X-ray
2. hypocycloidal tomography
3. computerized tomography
4. magnetic resonance imaging
5. orbital venography
6. carotid angiography
7. ultrasound.

The last three techniques have become less popular with the advent of computerized tomography and magnetic resonance imaging.

Plain X-rays should include an occipitofrontal posteroanterior view and an occipitomental posteroanterior view, both of which were designed to evaluate the paranasal sinuses and with which the otolaryngologist is familiar. In addition, a lateral view, submentovertical view and oblique apical projections of the optic canals are helpful. Plain X-rays of the orbit of patients with proptosis will reveal abnormalities in up to 33%, of which 21% are diagnostic (Lloyd, personal communication). Evidence of increased soft tissue density in the orbit and adjacent structures must be sought and variations in size and shape of bony outlines noted. Tissue emphysema may be apparent, occurring in up to 50% of blow-out fractures (Lloyd, 1966) and evidence of bony dehiscences and destruction may be seen. Occasionally orbital calcification and hyperostosis may be noted, which are always of pathological significance.
Hypocycloidal tomography still has an important role in orbital assessment and may demonstrate abnormalities not readily visualized on plain X-ray or may better demonstrate these abnormalities. It is routinely performed in coronal, lateral and axial planes and to visualize the optic canals a 39° oblique projection may also be requested. However, to a large extent this technique has been superseded by computerized tomography. Bilaniuk and Zimmerman (1980) have gone so far as to say that it is the only diagnostic modality that allows an accurate assessment of the degree of orbital involvement by paranasal sinus disease.

Computerized tomography allows simultaneous examination of bony structures and associated soft tissues with the orbital fat acting as a natural contrast medium and should always be performed in axial and coronal planes. Excellent detail of orbital structures is provided by this technique combined with a highly accurate assessment of paranasal sinus pathology (Lund, Howard and Lloyd, 1983).

More recently, magnetic resonance imaging has been evaluated in this area and is proving to be an important investigative technique (Lloyd et al, 1987).

Orbital trauma

A sense organ enclosed within an incomplete bony box is inevitably vulnerable to trauma. Accidental trauma can occur in association with facial injury or as a complication of sinus surgery.

Accidental surgical invasion

This can occur during antral lavage, the risk being clearly greatest if a middle meatal route is chosen, and when the procedure is performed under general anaesthesia. An incorrect angle of entry through the inferior meatus can lead to penetration of the orbital floor or, in the presence of a dehiscent infraorbital canal, excessive pressure of lavage may affect the orbital contents. For this reason the eyes should always be uncovered during the procedure and observed closely so that the operation can be abandoned at the first sign of proptosis.

Orbital damage is a constant hazard in intranasal ethmoidectomy. With poorly developed sphenoidal sinuses, the posterior ethmoidal cells extend backwards in intimate relation to the optic nerve so it is often impossible to exenterate completely the ethmoids by this route without jeopardy to the eye. Careful attention to technique with preservation of the middle turbinate for as long as possible is mandatory. The medial canthal ligament forms the lateral boundary and a constant watch must be kept for the appearance of orbital fat, indicative of a breach in the orbital peristeum.

Although Freedman and Kern (1979) reported a 2.8% complication rate in 1000 cases undergoing intranasal ethmoidectomy of which less than 1% were orbital, there is no doubt that this is a difficult operation both to perform well and to teach. Injury to the lamina papyracea may lead to haemorrhage which can result in intraorbital but extraperiosteal bleeding. With anterior tracking this produces a periorbital haematoma.

Eichel (1979) accepted a 25% rate of orbital ecchymosis in his series and haemorrhage was the commonest complication reported by Freedman and Kern (1979) but to regard it as
the sign of a 'successful' operation is to be deprecated. Anterior tracking of blood may cause little harm, but any posterior accumulation may cause proptosis and visual loss, necessitating removal of nasal packing or exploration (Leopold, Kellman and Gould, 1980). Periosteal damage leading to prolapse of fat, which can be packed with gelatin sponge, is especially likely if previous surgery has been performed, but is obviously best avoided as it may also be associated with damage to the medial rectus muscle.

**Facial trauma involving the orbit**

An early awareness and assessment of orbital function is important to avoid further damage. Fractures of the middle third of the face may involve the orbit or it may be traumatized alone, resulting in 'blow-out' fractures. The zygoma itself is thick but its three attachments to the skull are not, so displacement of the malar complex is often associated with fractures of the orbital floor.

Early signs and symptoms of a 'blow-out' of the orbital floor are restriction of extraocular mobility (particularly in upward gaze), infraorbital anaesthesia (including the canine teeth), orbital swelling and ecchymosis. The patient with a nasoethmoid complex fracture will also present with periorbital ecchymoses, probably secondary to ethmoid artery damage, flattening of the nasal root and widening of the intercanthal distance. Traumatic telecanthus does not always occur but should be excluded and may be difficult to establish when oedema obscures the medial palpebral angle. An alternative is to measure the interpupillary width which is normally twice the intercanthal distance. The medial canthal ligament can be further tested by traction on the lashes which should make lid margin taut. If this does not occur, disruption of the medial ligament has probably occurred.

It is obviously important to carefully assess visual acuity, the pupils, and lid and ocular movement. Surgical emphysema may be palpable and globe displacement indicates retrobulbar haemorrhage in the case of exophthalmos or orbital floor displacement if enophthalmos is present. Epiphora is not a reliable sign of nasolacrimal damage which may need separate assessment once initial oedema has settled (Holt and Holt, 1985). The possibility of cerebral spinal fluid rhinorrhoea should never be overlooked.

Radiological examination is important in determining the site and extent of bony trauma. Waters occipitomental and Caldwell occipitofrontal views will demonstrate orbital fractures in 70% of cases, but false positives are common and tomography is helpful if a 'blow-out' fracture is suspected which will show the classic 'tear-drop' sign in the antrum roof. In these cases and to assess the cribriform plate region, computerized tomography scanning is frequently indicated.

Therapeutic success must be judged in terms of preservation of binocular vision, the prevention or resolution of enophthalmos and the restoration of ocular mobility. Sacks and Friedland (1979) found the commonest presenting signs or symptoms in 100 patients with orbital floor fracture to be hyperaesthesia, infraorbital rim stepping, periorbital swelling and diplopia. Some symptoms such as oedema and hyperaesthesia will often resolve spontaneously, but considerable controversy surrounds the indications and timing of orbital floor exploration.
If there is no evidence of extraocular muscle movement restriction and no diplopia, despite radiographic evidence of a fracture, exploration may be unnecessary (Putterman, Stevens and Urist, 1974). However, combinations of diplopia and enophthalmos usually require immediate exploration once other causes of diplopia such as haematoma of the inferior rectus muscle have been eliminated by forced duction testing. Enophthalmos alone only causes diplopia in extremes of gaze but is unacceptable cosmetically. It may be masked initially by oedema or haematoma so, unless there is any loss of visual acuity, it is advisable to wait 7-10 days before deciding to operate. This is particularly pertinent when one considers the risk of visual loss associated with exploration. In 72 patients with orbital floor fracture who underwent repair, six lost vision as a result of infection or haemorrhage (Nicholson and Guzak, 1971), although it must be remembered that these patients represent a ‘high-risk’ group of whom 40% have associated ocular complications such as retinal artery damage. It is possible for late enophthalmos to develop due to secondary atrophy of orbital fat.

Surgical management depends on the extent of injury. Collapse of the anterior wall of the maxilla or orbital floor requires intra-antral manipulation, followed by Whitehead's varnish packing to stabilize bony fragments. The use of a Foley catheter to support the orbital floor can result in uneven elevation. Reconstruction of the orbital floor is indicated in enophthalmos, prolapse of orbital fat or with comminuted, hinged or inferiorly displaced bone. A transverse lower lid incision along the malar line gains exposure of the orbital periosteum which can be carefully elevated. The defect can be repaired with a number of substances including bone, tantalum, Teflon and silicone, but Silastic sheeting seems to be well tolerated and most successful. When a synthetic floor implant is used, antral packing is unnecessary and undesirable.

Fractures of the lamina papyracea are seen more frequently in complex midfacial fractures and can be associated with cerebrospinal fluid leak, lacrimal damage, visual loss and severe epistaxis from the anterior ethmoidal artery. In addition, there may be downward displacement of the medial canthus, subcutaneous or subconjunctival emphysema and limitation of lateral movement. If there is evidence of posterolateral displacement of the lacrimal bone and its attached medial canthal ligament, an open repair is indicated to reattach the bone with wires. Bilateral traumatic telecanthus requires bilateral exploration and wiring of the fractured bone. Damage to the nasolacrimal apparatus can be dealt with by splinting using Silastic tubes. In these circumstances early exploration produces the best results as late reconstruction can be difficult. Late complications associated with these injuries include one eye being lower (usually due to inadequate interosseous wiring and reattachment of Lockwood's ligament), diplopia, enophthalmos and occasional dacryocystitis (Reynolds, 1978).

A number of operations are available for access to or for specific procedures on the medial wall of the orbit. These may be considered in the context of particular pathologies which impinge on the orbit.

**Frontoethmoidal mucocoeles**

These mucus-filled cysts occur within the frontoethmoidal complex and most frequently present initially to the ophthalmic surgeon. Their capacity for expansion and erosion of bone results in encroachment on the orbit via the floor of the floor of the frontal sinus or orbital plate of the ethmoid. As a consequence, proptosis was the commonest...
presenting symptom in virtually all 98 patients seen between 1962 and 1986 (Harrison, personal communication). The degree of proptosis can vary from 1 to 17 mm and because it is slowly progressive, unless infection supervenes, diplopia only occurs in 65% and at extremes of gaze. In addition to proptosis there is concomitant displacement of the globe laterally and in the case of frontal mucocoeles, inferiorly. Ocular mobility is usually decreased in upward gaze due to the presence of a mass in the upper inner quadrant which often has a characteristic 'egg-shell' crackling sensation on palpation.

The radiological appearances are characteristic with evidence of general loss of translucency, loss of scalloping and definition of the frontal sinus margin and supraorbital depression and/or erosion. Computerized tomographic scanning may give additional information particularly when the ethmoidal complex is involved (Lloyd, Bartram and Stanley, 1974).

The aetiology of the condition has been attributed to a combination of obstruction of the frontonasal duct and inflammation, and this has important implications on the operative management of the condition. There is considerable evidence that a number of bone resorbing substances are produced by the mucocoele lining, including PGE₂ and collagenases (Lund, 1986). As a consequence it is important to remove all lining mucosa, the only exception being in areas where erosion of the posterior wall has occurred exposing dura (38%; Harrison, 1980).

The presumed frontonasal obstruction must be overcome by creation of a new channel by exenteration of the anterior ethmoid and insertion of 1 cm indwelling fenestrated Silastic tube for 3-5 months. The inter-sinus septum is intact in 95% of normal subjects, so an additional alternative drainage route should be created by routine perforation of the septum at surgery. All these requirements can be satisfied by the external frontoethmoidectomy (Lynch-Howarth approach) (see Chapter 11). A recurrence rate of 4% can be expected using this procedure, which compares favourably with that quoted for the osteoplastic flap (3-25%) (Zonis, Montgomery, and Goodale, 1966; Bosley, 1972; Sessions et al, 1972; Bordley and Bosley, 1973; Schenck, 1975; Hardy and Montgomery, 1976).

In the immediate postoperative period proptosis and ocular mobility improve, although diplopia may be initially worse due to sudden decompression. However, this can be expected to improve in the long term in the vast majority (96%; Rubin, Lund and Salmon, 1985). While blindness may be a theoretical consideration from rapid decompression, it has not been encountered and most patients (97%) experience no persistent change in visual acuity. Problems resulting from medial canthal ligament or lacrimal sac damage occur extremely rarely.

Osteomata

Osteomata arising in the frontoethmoidal region may encroach on orbital contents. Their origin may be difficult to determine if large, although they are presumed to arise at junctional points of membranous and cartilaginous bone and are composed of a cancellous core with varying amounts of surrounding dense compact bone. In a series of 23 patients (Atallah and Jay, 1981), 10 presented with ocular symptoms of whom six had proptosis.
Osteomata are readily visualized by plain sinus X-ray and are occasionally incidental findings which, if asymptomatic and quiescent, require no treatment.

However, in the presence of symptoms and when associated with mucocoele formation (5%; Lund, 1986), an external frontoethmoidectomy approach is indicated. In the larger osteomata, arising from the lateral ethmoidal complex, a lateral rhinotomy may be required to provide sufficient access, although the pedicle is often narrow. Occasionally large osteomata arising in the maxillary sinus have necessitated partial maxillectomy and, in two cases, craniofacial resection has been performed for anterior cranial fossa invasion (Cousins, Lund and Cheesman, 1987). Adequate access is important to allow complete removal which will result in cure.

Infection

The availability of antibiotics has considerably altered the frequency with which orbital complications result from sinusitis but, if they occur, the consequences are serious. In 1969, Jarrett and Gutman reported the figures of the pre-antibiotic era in which 20% of patients with orbital infection completely lost vision, 20% died and 10% had permanent visual damage. Embryological considerations determine that sinusitis in the child under 5 years is confined to the maxillary and ethmoid sinuses, while the frontal sinus is the commonest source of orbital cellulitis in adults. In children relatively minor upper respiratory tract infections can suddenly result in orbital complications, whereas in adults the situation may be more chronic, associated with inadequate oral antibiotic therapy or occasionally conversion of a mucocoele to a pyocoele.

Congenital dehiscences or open suture lines in the child, at the lamina papyracea, ethmomaxillary suture or in the orbital wall of the maxilla offer preformed channels and, in general, the bone is softer and more diploic in a child, facilitating spread. The orbital periosteum can easily be detached from the smooth surface of the bone by abscess formation.

The complex venous drainage and absence of valves in ophthalmic veins allows direct communication between the cavernous sinus, orbit and pterygoid plexus. Consequently spread of infection along suture lines can result, by direct invasion of bone or retrograde thrombophlebitis, leading to 159 children developing orbital infection out of 6770 with sinusitis (Fearon, Edwards and Bird, 1979). In 1937, Hubert classified orbital complications of sinusitis into five groups:

1. Inflammatory oedema of the eyelids with or without oedema of orbital contents
2. Subperiosteal abscesses with
   a. Oedema of the lids or
   b. Spread of pus to the lids
3. Abscess of orbital tissues
4. Mild and severe orbital cellulitis with phlebitis of ophthalmic veins
5. Cavernous sinus thrombosis.

Rarely, superior orbital fissure involvement leads to ophthalmoplegia, anaesthesia of the ophthalmic division of the trigeminal nerve and in combination with optic nerve damage an 'orbital apex syndrome' results.
Orbital cellulitis presents with oedema of the lids and chemosis with varying degrees of proptosis and globe displacement. Visual acuity may be difficult to assess due to lid swelling. Rapid administration of intravenous antibiotics must be instituted or the situation will progress with abscess formation and ultimately compression of the central retinal artery. Constant careful monitoring of visual acuity (in particular of colour vision) and ocular mobility will rapidly indicate when surgical intervention is appropriate. While the majority respond, antral lavage or frontal trephination may be indicated at a later date. Spontaneous discharge may occur through the upper lid from the frontal sinus or near the medial canthus from the ethmoid.

Failure to improve or any clinical deterioration requires external drainage with insertion of a drain and possibly irrigation often in combination with antral lavage. Forty-eight hours of irrigation suffice without the danger of actually introducing further infection. Frontoethmoidectomy may be indicated if this treatment fails to produce dramatic resolution, when the abscess is not superficial or there is clinical evidence of visual deterioration. Adequate elevation of the orbital peristome posteriorly is necessary to locate the pus and removal of the orbital plate of the ethmoid allows intranasal drainage.

The potential severity of orbital complications of sinusitis should not be underestimated and may constitute one of the surgical emergencies facing the otolaryngologist.

**Orbital decompression**

The otolaryngologist with an interest in head and neck surgery may occasionally be called upon to decompress orbital contents. Malignant exophthalmos is the commonest indication, although pseudotumours, Wegener's granulomatosis and the palliation of metastatic deposits have also been reported (Harrison, 1980; Sobol, Druck and Wolf, 1980).

In malignant exophthalmos characterized by hypertrophy of orbital muscle and fat, the thyroid metabolism must be treated first and is often followed by steroid therapy. Failure of this to result in permanent improvement necessitates surgical decompression. In 1911, Dollinger first described orbital decompression by removal of the lateral wall of the orbit (attributed to Kronlein) allowing swollen orbital contents to herniate into the temporal fossa. Two decades later, Naffziger (1931) published an account of decompression into the anterior cranial fossa via a transcranial route.

Sewall (1936) first described the use of the paranasal sinus cavities for decompression by removing the medial orbital wall, while Hirsch (1950) later reported inferior decompression into the maxillary antrum. Ogura (1978) advocated a combination of these two methods, but this technique is difficult due to limited access and Harrison (1981) has suggested that the Patterson's transorbital approach is a safer and more effective alternative (see Chapter 11). This allows removal of the entire orbital wall medial to the infraorbital nerve and can if necessary be combined with a lateral orbitotomy to remove the lateral wall and floor lateral to the nerve.
Lateral rhinotomy

The virtues of this surgical approach to the medial wall of the orbit have been more recently appreciated (Harrison, 1977; Schram and Myers, 1978). This procedure has undergone little modification since it was first described in 1902 by Moure, Professor of Otolaryngology in Bordeaux, offering a combination of excellent access and postoperative cosmesis. It allows inspection and resection of the entire lateral nasal cavity, septum and most of the medial orbital wall and is, therefore, applicable to the management of a variety of pathologies but most notably that of malignant melanoma (Lund, 1982), angiofibroma (Harrison, 1986), and inverted papilloma.

The incision should reach superiorly to just below the medial palpebral ligament, thus avoiding detachment of this important structure. Elevation of the skin above this point is easy and allows further bone removal if necessary. The incision then follows the nasomaxillary groove, curving round the ala to enter the nose. Detachment of the nose from the pyramidal opening allows the nasal framework to be swung away, exposing surrounding bone. The underlying pathology will determine how much of this bone is removed, but can include frontonasal process, anterior wall of maxilla as far laterally as a vertical plane through the inferior orbital foramen, lacrimal fossa and lamina papyracea, preserving the infraorbital rim and the complete lateral wall of the nose with nasal septum. No matter how large the underlying bone defect, primary closure presents no problem with good cosmetic result. Resection is inevitably limited superiorly by the cribriform plate but this can at least be reached in safety, with no risk to vision or extraocular musculature. If there is any doubt as to the patency of the nasolacrimal apparatus, the lacrimal sac can be opened and the lining everted.

Management of the orbit in antroethmoidal neoplasia

The orbit is threatened by a variety of neoplasms originating in adjacent structures, because of its anatomical relationship to skin, skull, palate, salivary glands, nasal glands, nasal cavity and, in particular, the antroethmoidal complex. The importance of the eye, both as a sensory organ and for its aesthetic value, makes the management of this area particularly pertinent to both surgeon and patient.

Preservation of orbital contents must be based on the unemotional decision that it will not jeopardize prognosis and that the eye will have aesthetic and functional capabilities if preserved. This is of particular relevance if postoperative radiotherapy is contemplated which may result in cataract, glaucoma and loss of vision.

Invasion of the orbit from the anterior ethmoidal complex occurs early and in a number of ways. Visual symptoms of epiphora, proptosis and diplopia are commonly reported in a series of patients with malignant tumours of the nasal cavity and paranasal sinus (Lund, 1983; Weber and Stanton, 1984). Invasion commonly occurs through the thin or dehiscent bone of the lamina papyracea or through the region of the inferior orbital canal, both areas readily accessible to surgery. Distinction must be made between intraorbital spread which is extraperiosteal and that in which the periosteum is breached as this has important implications on management.
Of more sinister prognostic importance is invasion of the posterior medial wall from the posterior ethmoids to involve the optic canal directly or infiltration of the retrobulbar structures from below, with spread in either case via the orbital apex to the middle cranial fossa. High resolution computerized tomographic scanning in axial and coronal planes with wide window width settings offers important preoperative assessment (Lund, Howard and Lloyd, 1983) to which magnetic resonance imaging is now contributing.

Finally, in the case of adenoid cystic carcinoma, in addition to direct and perineural spread, the possibility of embolic phenomena must be considered as demonstrated in a section of optic nerve taken some distance from an ethmoidal tumour (Howard and Lund, 1985).

The presence of proptosis and limitation of ocular movement usually indicates extensive invasion which is often associated with involvement of the cribriform plate and pterygopalatine fossa. Orbital involvement like anterior cranial fossa invasion was once considered to indicate a poor prognosis, but it is failure to control local disease which has escaped through the orbital apex, which is more important than the actual point of entry into the orbit. In Harrison's (1978) series at least 50% of patients required orbital exenteration. However, the palliative role of orbital exenteration is also of importance as it prevents painful proptosis and, in combination with total maxillectomy, allows control of residual tumour by laser or cryosurgery.

Before the advent of the craniofacial procedure, total maxillectomy and orbital exenteration in combination with radiotherapy was the treatment of choice for most tumours arising in this area. Any evidence of anterior cranial fossa extension would now indicate craniofacial resection.

When total maxillectomy is performed with orbital exenteration, the lids may be preserved as they are not implicated in lymphatic dissemination nor usually in direct tumour spread. Removal of the lash margins and tarsal plates with suturing together of the lids to close the orbital defect results in a layer of thin, well-vascularized elastic skin which lines the empty socket, a situation which is only compromised by overzealous preoperative radiotherapy. The eventual use of a high quality prosthesis supported by spectacle frames precludes the need for reconstructive techniques which are not only difficult but may compromise cure (Conley and Baker, 1979).

In the absence of orbital involvement with total maxillectomy, the globe is left supported by the suspensory ligament of Lockwood. There may be some prolapse, often several weeks later, and a proportion of these patients require reinforcement of the area with nylon mesh or Silastic sheeting.

Accurate evaluation of orbital involvement is possible with the craniofacial procedure (see Chapter 180. The osteotomies in the floor of the anterior cranial fossa encompass the cribriform plate and orbital roofs laterally and extend backwards to the jugum, stopping just anterior to the optic chiasma. The orbit is entered to ensure adequate removal of the medial wall and anteriorly the osteotomies are completed with a fissure bur through the frontonasal duct, uniting intra- and extracranial cuts.
In cases in which the medial bony wall of the orbit has been breached but the periosteum is intact, the compromised area of periosteum can often be resected and grafted with a split-skin graft, preserving the globe and its musculature. Contraction of the graft results in remarkably little disturbance of ocular function. Ketcham et al (1973) believed that their attempts to conserve orbital contents in 24 patients required re-evaluation as the survival figures in this group were 32% compared with 30 patients in whom unilateral orbital exenteration had been performed and whose survival was 62%. However, in a recent series of 60 patients (Cheesman, Lund and Howard, 1986), resection of involved periosteum and frozen section control of adjacent orbital contents (with the possibility of future surgery if tumour recurs in this area) has not been associated with a poorer prognosis. Seventy-six per cent of patients who had orbital exenteration are dead compared with 38% of those in whom orbital periosteum alone was removed. This may, however, merely reflect extensive disease with an associated poorer prognosis.

Occasionally tumours of the orbit itself such as meningiomata may become sufficiently large to impinge on the sinuses necessitating total maxillectomy and orbital exenteration. Similarly infratemporal fossa disease (for example neurolemmomata) can extend into the infraorbital fissure or breach the inferior and lateral walls requiring lateral orbitotomy in combination with an infratemporal resection. Finally, large neglected basal cell carcinomata of the surrounding orbital skin occasionally present to the head and neck surgeon who may need to employ rotation, myocutaneous or free microvascular flaps for reconstruction.