Chapter 16: Neuroanatomy and applied neurophysiology for the otolaryngologist

John Philip Patten

Although this chapter is included in the basic science volume, the applied aspect is of such importance in differential diagnosis of diseases affecting neuro-otolaryngological function that some licence has been taken to indicate, whenever possible, the significance of both anatomy and physiology in disease states and differential diagnosis.

In some instances, gross anatomy is of great importance and in others, complex central connections require detailed elaboration to illustrate and explain clinical disorders. The following account is always biased in the direction of practical applications, and information of limited or dubious clinical importance has been excluded.

The cranial nerves fall into three major groups on the basis of both functional and gross anatomical similarities, and they share common anatomical relationships and pathology. Differing patterns of involvement within these groups allow very accurate differential diagnoses to be advanced, based on both the sequencing and ultimate extent of damage in these groups. The advent of computer-assisted tomography, which until recently was available only in slice format, has added a remarkable dimension to our ability to confirm or refute a clinically based diagnosis in this hitherto investigational no man's land. The recent ability to reconstruct slice scans in both sagittal and coronal planes has further transformed diagnostic accuracy. Interpretation still requires a very good grasp of gross anatomical relationships of the intracranial and extracranial courses of the cranial nerves, and these features will form the bulk of this chapter. The groupings are:

(1) cranial nerves I, II, III, IV and VI and the final distribution of the cervical sympathetic nerve

(2) cranial nerves V, VII and VIII

(3) cranial nerves IX, X, XI and XII and the cervical components of the sympathetic chain.

The influences of cerebellar, pyramidal, extrapyramidal and corticobulbar dysfunction on these nerves, and peripheral evidences of disordered brainstem function, will be detailed at the end of the chapter, or where appropriate.

**Group one**

In the first group, the close relationship between the olfactory and optic nerves and the varying relationships between the three nerves supplying the extraocular muscles are considered. The relationships of the first division of the fifth nerve, which traverses the orbit, to these structures, must also be noted, although the detailed anatomy of this nerve is dealt with in group two.
The olfactory nerve (I)

Anatomy

The olfactory epithelium lies in the olfactory cleft which occupies the upper 10 mm of the nasal septum, the roof of the nasal cavity and down the lateral wall towards the origin of the superior concha. In man its total surface area is some 5 cm² and it is a yellowish colour. In other species, increasing pigmentation is associated with increased sensitivity to odours. The mucosa is bated in a lipid-rich secretion from the epithelial Bowman's glands, indicating that lipid solubility may be a critical factor in odour detection. The olfactory receptor cells lie on the basal epithelium and extend vertically to the surface, from which the terminal enlargement protrudes and gives rise to 8-20 olfactory cilia. Although these have the 9+2 fibril arrangement of mobile cilia in other areas, they are thought to be non-motile and form a dense mat of fibrils lying on the surface of the epithelium. Pinocytic vacuoles have been demonstrated in the terminal enlargement of the receptor cells, but their functional significance is uncertain (Fitzgerald, 1985).

The receptor cells are derived from ectoderm and are unique in being replaced from stem cells every 30 days. They also enter the central nervous system (CNS) as non-myelinated axons without synapsing. These axons become grouped and ensheathed by Schwann cells forming some 20 fasciculi which, invested by pia and arachnoid mater, pass through the orifices of the cribriform plate to enter the olfactory bulbs, lying each side of the crista galli in the floor of the anterior cranial fossa. These axons synapse with dendrites of the large mitral cells in the olfactory glomeruli and each glomerulus receives axons from a wide area of the epithelium - there appears to be no functional grouping of axons. This allows a relatively small number of receptor cells to distinguish a large number of different odours. The axons of the mitral cells form the bulk of the olfactory tract, but centrifugal axons of uncertain origin pass to the olfactory bulb and undoubtedly modify activity in the olfactory glomeruli, perhaps by both inhibitory and facilitatory action.

The olfactory tracts pass posteriorly and slightly laterally crossing the floor of the anterior cranial fossa and the optic nerves, and immediately above the optic chiasm, just in front of the anterior perforated substance, each divides into medial, intermediate and lateral olfactory striae.

The termination of the medial striae is uncertain. Many fibres decussate to the opposite medial striae and these may become the centrifugal fibres of the opposite olfactory tract, having both facilitatory and inhibitory effects on the opposite olfactory bulb. The intermediate striae terminate in the olfactory tubercle, but the latter's further functional anatomy is unknown. The lateral olfactory striae synapse with neurons in the lateral anterior perforated substance, the lateral olfactory gyrus, the prepyriform cortex and the medial group of amygdaloid nuclei - a group of tissues which, in man, represents the primary olfactory cortex. These are the only sensory pathways in man that do not relay in the thalamus. The distribution in the limbic system then contributes to both the pleasurable and unpleasant consequences of odour detection at a conscious level, and the appropriate autonomic responses by way of the hypothalamus. This is related to activity in a secondary olfactory cortex in the entorhinal complex including the uncus and a tertiary olfactory cortex in the posterior orbitofrontal cortex. Descending pathways from these areas enter the pontine reticular
formation in the brainstem, and mediate reflex activity such as salivation (Tanabe, Iino and Takagi, 1975).

**Physiology**

The receptor proteins lie in the olfactory cilia, and it is likely that many different types of protein are involved. A smell must be volatile to enter the cavity, actively sucked into the area of the olfactory epithelium by sniffing, to create turbulent flow in the nasal passages, and also lipid soluble to facilitate access to the fluid-bathed cilia.

Once stimulated, the activity in the neuron is difficult to study. Attempts at single fibre analysis are technically almost impossible and such studies as are available demonstrate no similarities in evoked potentials between similar groups of substances or stimuli. There is considerable evidence that some odours inhibit as well as excite, in addition to which an anatomical arrangement allows not only local inhibition and excitation but crossed and possibly centrally mediated control by both lateral and negative feedback mechanisms. This enables the human being to identify some 3000 different odours. The central pathways clearly allow for further discrimination and perhaps clarification of odour recognition. A contrast can be found in the remarkable process of adaptation, by means of which continuous exposure to an unpleasant smell diminishes perception to such a degree that the smell no longer registers.

Several theories exist which seek to explain odour appreciation. One theory is based on receptor site configuration but it seems unlikely that sufficient variation in shape exists to explain the full range of odours. Furthermore, the lack of structural similarity between chemicals that smell the same makes this explanation improbable as a sole mechanism (Amoore, 1963). A second theory has been proposed which is based partly on structural chemical considerations combined with molecular vibration, and some support for this can be found in the fact that there is a similarity of smell between chemicals with a similar frequency of vibration but a different chemical structure (Wright, 1964). The most acceptable theory, however, suggests that a dissolved molecule of specific size and shape is adsorbed on to and penetrates the receptor membrane, leaving a temporary hole, which allows local depolarization of a size, rate and duration proportional to the molecule characteristics. Even this cannot explain all the features of olfaction and it is probable that a combination of all three possibilities is involved (Davies and Taylor, 1959).

**Applied anatomy and physiology**

Of immediate otolaryngological concern are simple mechanical factors interfering with access of the odour to the receptors, with simple airway obstruction, complicated by oedema or drying up of the mucosa as the most common cause of trouble. Mechanical destruction or blockage of the nasal passages by pathology ranging from allergic rhinitis to complex vascular diseases such as Wegener's granulomatosis can occur. Simple polyps, deviated nasal septum and foreign bodies all have similar effects.

Many drugs and generalized medical conditions that can damage or interfere with a highly metabolically active tissue, with a 30-day turnover rate, can also affect smell. These include generalized metabolic disorders such as renal failure, hepatic failure, endocrine disorders, including diabetes, and influenza. Drugs affecting membrane moistness
(antihistamines), cell turnover (antibiotics, antimetabolites) and cell function (anti-inflammatory agents, antithyroid drugs) may all affect both smell and taste (Schiffman, 1983).

Traumatic lesions of the olfactory fasciculi are caused by the shearing effect of brain movement when the head decelerates during a head injury. This complicates some 30% of serious head injuries, particularly where immediate anteroposterior forces are applied to the head, so that a fall squarely on the occiput is especially likely to result in this complication. In such cases, little or no recovery can be anticipated. Severe injury of this type may also tear the arachnoid cuffs and lead to cerebrospinal fluid rhinorrhea with a significant risk of subsequent meningitis.

Experimental evidence is available that viral infections may gain access to the meninges by means of the same route, even in the absence of prior injury, with herpes simplex encephalitis being a notable example. In the latter condition, the initial localization of the infection to the anterior temporal lobes gives support to this theory of aetiology, the virus presumably gaining access along the olfactory tract, although there is no definite evidence that this is the case (Johnson and Mims, 1978).

Inside the skull, tumours of the olfactory groove, notably meningiomata, will produce unilateral anosmia, usually unrecognized by the patient. On account of the local anatomy, progressive visual loss in the same eye will follow - also often unrecognized by the patient. It is very important to test the sense of smell in any patient with suddenly discovered loss of vision in one eye.

At central level, disorders of smell appreciation are not recognized. Most patients who complain of a constant awful smell sensation or altered smell appreciation are suffering from a depressive or psychotic illness. The most identifiable centrally based disorder is uncinate epilepsy in which an epileptic event originating in the temporal lobe is preceded by the production of hallucinatory phenomena embracing sensation of unpleasant smell and, occasionally, unpleasant taste. These olfactory hallucinations are characterized by being both unpleasant and of extremely short duration, usually only a matter of seconds, often insufficient to enable the patient to identify the odour as other than unpleasant - burning rubber or rotting rubbish being the commonest descriptions volunteered.

Considerable degeneration of the olfactory glomeruli occurs with age. Olfaction is the first sensory modality to be impaired with age and is possibly responsible for decreasing appetite and interest in food in the elderly (Schiffman, 1979).

The optic nerve (II)

The orbit is entirely surrounded by structures of otolaryngological significance, and only the lateral border is relatively safe from possible infection or invasive pathology. The frontal ethmoid, maxillary sinuses and the lateral wall of the nose bound the orbit superiorly, inferiorly and medially and are all prone to infection or malignant pathology.

The optic nerve enters the orbit through a tight canal - the optic foramen. The nerve is a direct extension of the brain and is invested with glial derived tissue to the back of the globe, consisting of three membranes. The inner pial sheath invests the nerve and sends septae
into the nerve itself, dividing the nerve into a bundle of fasciculi. The intermediate arachnoid sheath is very delicate, with a potential subarachnoid space inside it and a subdural space outside. These are covered by a thick extension of the dura which merges with the sclera at the back of the globe. These membranes form a direct means of communication with the intracranial space and are responsible for the transmission of raised intracranial pressure to the optic disc causing papilloedema, although the exact mechanism of the disc swelling remains uncertain.

The myelinated fibres of the optic nerve are derived from the rods and cones of the retina. As these cell processes form the most superficial layer of the retina, they are normally non-myelinated until they enter the disc. Occasional patches of myelination of these fibres as they cross the retina produce a characteristic fundal appearance and a field defect which is unnoticed by the patient, in the same way as there is unawareness of the normal blind spot. The important papillomacular fibres conveying macular vision lie in the medial part of the nerve, assuming their central position in the nerve only at the optic foramen. In spite of this anatomy, extrinsic compression of the nerve in the orbit and the canal specifically affects these fibres, producing a central scotoma rather than a defect spreading in from the periphery, as might be expected purely on anatomical grounds.

There are 1.2 million fibres in each optic nerve, just over half of which decussate in the optic chiasm. The fibres which cross are the fibres from the nasal retina, covering the temporal half field, and enter the contralateral optic tract. The temporal half fibres (the nasal field) pass into the ipsilateral optic tract.

Lesions in the orbit tend to produce mechanical displacement of the globe with proptosis and diplopia. The optic nerve itself is remarkably resistant to damage by pressure and displacement in the orbit, although an infective process may be more damaging by vascular mechanisms (Forrest, 1949; Font and Perry, 1976).

Lesions in the optic canal, however, readily cause visual disturbance and a central scotoma is often the first evidence of a lesion, followed by extraocular nerve palsies and, very much later, proptosis. Meningiomata or neurofibromata of the optic nerve sheath are perhaps the most common tumours in the posterior orbit, but neoplastic infiltration from the paranasal sinuses and nasopharynx can occur and metastatic spread from remote sites such as the prostate or suprarenal is well recognized. In general, the rate of development of the signs and the presence or absence of pain will indicate the likely diagnosis (Takashi, 1956).

Involvement of the optic nerve at the intracranial part of the optic foramen may produce bilateral visual problems. The inferior nasal fibres of the opposite optic nerve not only cross in the chiasm but sweep forwards into the optic nerve before turning sharply and heading posteriorly. They can be damaged by a lesion just anterior to the chiasm. A meningioma of the tuberculum sellae is the lesion most likely to be responsible. This can produce a blind eye, an upper temporal field defect in the other eye (called a junctional scotoma) and, if large, can cause loss of smell on the side of the blind eye and papilloedema in the opposite eye; disc swelling in the blind eye is prevented by the compressing lesion. This condition is the famous, but extremely rare, Foster-Kennedy syndrome.
The optic chiasm itself lies more posteriorly than is generally appreciated - it lies above and behind the pituitary gland, not on the groove in front of the pituitary fossa seen on the skull. Pathology in the pituitary region includes not only pituitary tumours but neoplasia arising in the ethmoid and sphenoid sinus, mucocele of the sphenoid sinus and a variety of aneurysms around the circle of Willis or arising from the great vessels themselves. The importance of excluding vascular anomalies or aneurysms before a transnasal approach to the pituitary fossa is perhaps even more important than in the days of the frontal approach when unrecognized aneurysms were encountered with occasional fatal results. Lesions extending up from the pituitary region damage the underside of the chiasm anteriorly. This produces a bitemporal hemianopia which comes down from the upper temporal field (the lower fibres derived from lower retinal cells, therefore upper field), although the field defect is rarely appreciated by the patient at this stage or, indeed, occasionally even when complete. When testing the temporal fields, particular attention should be paid to the upper temporal field to avoid missing a junctional scotoma (see previous section) or a developing bitemporal hemianopia. In contrast, lesions damaging the chiasm from above and behind tend to affect the lower fields first; these include craniopharyngioma, hypothalamic tumours and a dilated third ventricle.

Because there are many situations in which the visual fields are of help in otolaryngological diagnosis, it is worth describing simple field examination at the bedside. Carefully examined fields, using a red and white hatpin, should be as accurate as screen testing and should take only a few minutes. The examiner should sit in front of the patient (in the traditional otolaryngological position) about 1 m (3 ft) from the patient. The patient should cover one eye. A white 5 mm hatpin, preferably mounted on the handle of a tendon hammer, should be brought into the patient's field of vision on four arcs, upper and lower temporal and upper and lower nasal, respectively. If all are seen at the periphery, no field cut is likely. The pin should then be brought across from the temporal field on a horizontal meridian, with the patient keeping the examiner's pupil in view. The blind spot should be detected without difficulty and can be compared with that of the examiner, with both parties loosing the object in the same area. Following across into the nasal field, any small scotoma will be indicated by the pin disappearing again. The size and shape of the scotoma can then be readily explored and even a small scotoma can be easily confirmed by this technique. At a more sophisticated level, the very earliest evidence of a field defect can be found with the red pin. Care must be taken not to mistake the normal loss of brightness of a red object in the temporal half field for an indication of an early field defect.

Differential diagnosis of the painful red eye (Sergott, 1983)

Otolaryngologist are often involved in cases where blurred vision and diplopia occur in the setting of an inflamed, proptosed eye, and they may well be the first doctors to see the patient. Diagnosis falls into four main groups of disorders - inflammatory, vascular, infective and neoplastic.
**Inflammatory causes**

**Acute thyroid exophthalmos**

The eye is often injected with chemosis. Lid lag is especially noticeable on downward gaze. There may be diplopia caused by globe displacement, although paralysis of the superior and lateral rectus muscles is not uncommon. The condition is usually unilateral. Vision may be threatened and high dose steroids may be of value in treatment. A computerized tomographic (CT) scan will show swelling of the extraocular muscles.

**Pseudotumour of the orbit**

This is an immunologically based inflammatory disorder affecting all tissues in the orbit. It can complicate sarcoid, systemic lupus erythematosus (SLE), tuberculosis, Wegener's granulomatosis, polyarteritis nodosa or the Tolosa-Hunt syndrome. Proptosis, pain and diplopia, associated with a very high sedimentation rate, might all seem to indicate infection. As steroids will be indicated, urgent exclusion of infective disease in the paranasal sinuses is vital. CT scans show normal extraocular muscles in the midst of oedematous orbital contents. The condition occurs in two main age groups: between 10 and 30 years and in the over 60s.

**Vascular causes**

**Acute caroticocavernous fistula**

This condition usually follows known trauma but occasionally an aneurysmal dilatation of the carotid may rupture into the cavernous sinus, producing acute pulsatile exophthalmos with marked arterial pulsation in the fundal veins. Carotid ligation or embolization is the procedure of choice.

**Cavernous haemangioma**

This produces a gradual exophthalmos with proptosis aggravated by bending or straining. There is usually no diplopia or field defect and little pain.

**Infective causes**

Local infections can readily spread into the orbit. Small boils on the nose, eyelids or face had lethal potential in the preantibiotic era. Paranasal sinus infection, especially of the ethmoids, can easily extend directly into the orbit and frontal sinusitis, usually causing oedema of the eyelid and ptosis. In the diabetic patient, all these infections carry even greater risk and additional specific problems such as mucormycosis and other rare fungal infections. The first vesicles of herpes zoster ophthalmicus usually erupt in the eyebrow after several days of severe pain and the acute red eye and oedematous lids may be mistaken for bacterial infection until the vesicles appear.
**Neoplastic causes**

Any primary or secondary neoplasm may involve the orbit, the latter by direct extension or from remote sites. Usually, chemosis and injection are not marked. In the elderly, pseudotumour of the orbit can be a presenting symptom of lymphoma and, as always, the importance of a general physical examination must be emphasized.

The benign primary orbital tumours which are most often seen are lipomata, angiomata and haemangiomata. Less frequently, fibromata, myxomata and leiomyomata may be encountered.

Malignant primary orbital tumours are usually rhabdomyosarcomata which are locally invasive and normally occur in childhood. It is rare for fibrosarcoma, myxosarcoma, liposarcoma, chondrosarcoma, osteogenic sarcoma and haemangioendothelioma to occur. Lacrimal gland tumours of variable malignancy do occur and they tend to be locally invasive through the roof of the orbit into the intracranial cavity.

Metastatic tumours in the orbit are, in 50% of cases, caused by carcinoma of the breast. Tumours originating in the lung and kidney account for the rest. Malignant melanoma has been reported but is hard to distinguish from a primary melanoma of the ciliary body, or retina. In children with neuroblastoma, orbital metastases occur in 20-50% of cases (Fanarier, Saraccco and Blane, 1972).

**Pupillary abnormalities**

As the main determinant of pupil size is the incident light, it is appropriate, at this stage, to discuss the major pupillary abnormalities.

In a blind eye, assuming that the cause of blindness has not simultaneously damaged the iris mechanism, the pupil will dilate or constrict in proportion to the light falling on the unaffected eye. The direct light reaction will be absent but the consensual light reflex from the opposite eye will be intact. No consensual reflex in the normal eye will be seen when the affected eye is stimulated. This is quite a useful check for non-organic claimed loss of vision in one eye.

In acute retrobulbar neuritis, the pupil reaction may be incomplete and the pupil may dilate in spite of a constant light source (pupillary escape phenomenon). In a patient with eye pain, aggravated by movement with blurred vision, this Marcus-Gunn pupil reaction is strongly indicative of demyelinating disease. The postulated mechanism is a decrease in fibres conveying light sensation.

In third nerve lesions, damage to the efferent pupilloconstrictor fibres will produce a fixed dilated pupil even though the patient perceives light normally. Incomplete lesions may merely cause a slightly dilated pupil with a sluggish reaction - an important stage in the evolution of a third nerve palsy in a patient who is deteriorating following a head injury. A useful clue in a conscious patient with a third nerve lesion is the almost constant accompanying ptosis of varying degree, followed by diplopia caused by paralysis of the superior rectus muscle (see next main section). Argyll Robertson pupils resulting from
meningovascular syphilis have become a great rarity. This is a small pupil, usually irregular, that does not react to light but does react to accommodation (Loewenfeld, 1969).

A sympathetic nerve lesion (Horner's syndrome) will be detected by only the most alert clinician. On account of the loss of the less important pupillodilator fibres, a slightly smaller pupil is found showing a normal light reaction; this is because the light reflex pathway mechanisms are unaffected. A modest and variable degree of ptosis will occur which rarely goes lower than the edge of the pupil. As the cervical sympathetic pathway courses in and out of otolaryngological territory, a full understanding of the syndrome is essential to the otolaryngologist (see also section on cranial nerves IX, X, XI and XII) (Jaffe, 1950).

A Holmes-Adie (myotonic) pupil may present as severe eye pain because the pupil fails to constrict in bright light. The affected pupil may be larger or smaller than the other, depending on whether the incident light produces a slower constriction or a slower dilatation of the affected pupil. If the light reaction is very slow, definite constriction followed by slow dilatation may best be demonstrated by maintained forced convergence for about one minute. If the patient sits in a dark room before entering the clinic, the pupil will stay very large, but if the patient enters from a bright sunlit room, the affected pupil may at first be smaller than the normal pupil (Loewenfeld and Thompson, 1967).

The nerve supply to the extraocular muscles

The three nerves supplying the extraocular muscles and controlling eye movements have complex central control mechanisms and run peripheral courses that render them vulnerable, both individually and as a group, to a wide range of surgical and medical disorders. They are of special interest to otolaryngologists because of their involvement in local neoplastic disease and in infective processes originating in paranasal sinuses, nose and nasopharynx.

The oculomotor nerve (III)

The third nerve exits from the brainstem in the interpeduncular fossa and runs forwards and slightly downwards in the subarachnoid space diverging towards the roof of the cavernous sinus. In its distal subarachnoid course, it runs parallel to the posterior communicating artery, hence its unique susceptibility to damage by aneurysms which commonly arise at either end of this vessel. It enters the roof and then the lateral wall of the cavernous sinus in between the two layers of dura, dividing into two branches before entering the superior orbital fissure. In the wall of the sinus, it picks up sympathetic fibres from the plexus on the carotid artery, and additional parasympathetic fibres from the ophthalmic division of the fifth nerve.

The superior ramus supplies the levator palpebrae superioris and the superior rectus muscle. The inferior ramus supplies the medial and inferior recti and the inferior oblique, and it carries the sympathetic and parasympathetic elements to the ciliary ganglion by way of the branch to the inferior oblique.

The anatomy of the pupillary fibres in the nerve itself is of great significance. The fibres appear to lie dorsolaterally in the periphery of the nerve. They are thought to have a
blood supply derived from the pial plexus on the surface of the nerve, the core being supplied by a vasa nervorum. If this latter vessel is occluded by vascular disease (diabetes, arteriosclerosis, arteritis), the peripheral pupillary fibres are spared. Conversely, if the nerve is damaged from without by a surgical lesion (aneurysm, tumour, abscess), the pupillary fibres are readily involved. In third nerve lesions, the involvement or otherwise of the pupil is a major diagnostic pointer. Pain tends to be a feature of surgical lesions; therefore, a painful onset of a third nerve lesion with pupil involvement is almost certain to indicate a compressive lesion. No pain and a spared pupil is almost certain to indicate a medical cause (Wray, 1983).

Central anatomy

The controversial anatomical features of the oculomotor nucleus are beyond the scope of the present text. The nucleus is shaped like an inverted V, straddling the midline. The lateral nuclear columns supply the eyelid and the four extraocular muscles, the superior, medial and inferior rectus and the inferior oblique. The midline nuclei have mainly parasympathetic function, especially the upper midline Eddinger-Westphal nucleus which is the main central control mechanism for pupil size. The fasciculi of the third nerve fan out and traverse the red nucleus and substantia nigra and then converge to form the main nerve trunk as it emerges just lateral to the midline in the interpeduncular fossa.

The trochlear nerve (IV)

The fourth nerve is unique in two ways. It arises from the dorsal aspect of the brainstem at the level of the inferior colliculus and decussates in the superior medullary velum, so that the right nucleus supplies the left superior oblique muscle and vice versa. It also has the longest intracranial course of any cranial nerve and is very slender, both of which are properties that possibly protect it from damage by external pressure around the brainstem and in the subarachnoid space. It enters the wall of the cavernous sinus beneath the third nerve, but crosses it to reach a higher position as it enters the superior orbital fissure to supply the superior oblique muscle. The nerve is almost never damaged in isolation in cavernous sinus lesions, the third and sixth nerves being much more vulnerable. Vascular lesions of the nerve caused by diabetes are probably the commonest cause of pure fourth nerve lesions. Of particular importance to the otolaryngologist is the small fibrocartilaginous loop attached to the trochlear fossa in the upper medial orbit, through which the muscle tendon passes. Accidental or surgical trauma easily damages the tendon in this region and produces an apparent fourth nerve palsy (Burger, Kalvin and Smith, 1970).

The abducent nerve (VI)

The sixth nerve arises from the pontomedullary junction, the most medial of the three nerves arising from this groove, and ascends on the front of the pons, angles forwards across the top of the petrous bone to enter the cavernous sinus in which it lies free in close relationship to the intracavernous portion of the carotid artery. The long subarachnoid and meningeal course of the nerve renders it particularly susceptible to damage in acute and chronic meningitis and any meningeal process, including remote or direct spread of malignancy. Its angulated entry into the cavernous sinus renders it vulnerable to stretch when the brainstem is pushed downwards by raised supratentorial pressure causing false localizing
sixth nerve palsies, which nearly always become bilateral. The nerve may be involved in inflammation of the petrous bone secondary to otitis media. This is often combined with severe pain in the fifth nerve territory and loss of hearing, a condition referred to as Gradenigo's syndrome. Inflammatory disease of the cavernous sinus and aneurysmal dilatation of the carotid siphon are particularly likely to involve the sixth nerve early on. The third and fourth nerves are almost always involved at a later stage by way of the same process. Nerve trunk infarction caused by diabetes, arteritis and arteriosclerosis also occurs exactly as for the third and fourth nerves, as discussed previously. Intracranially, both cholesteatomata and acoustic neuromata may involve the nerve, but this is a relatively rare occurrence. As it enters the orbit, the nerve occupies a lateral position in order to reach its single muscle, the lateral rectus. At this point, it is particularly susceptible to damage by carcinoma infiltrating the orbit through the inferior orbital fissure from the nasopharynx (Rucker, 1966).

Central anatomy

The nucleus of the sixth nerve lies in the floor of the fourth ventricle just lateral to the midline. The fibres of the facial nerve sweep round it. Although derived from the same nuclear column as the third and fourth nerve nuclei, it has migrated during the massive enlargement of the pons, but remains intimately linked by the medial longitudinal bundle discussed in the following. The fasciculi of the nerve have to traverse the whole depth of the pons to reach the point of emergence on the pontomedullary junction. It lies in close relationship to the medial lemniscus and corticospinal pathways.

Central mechanisms of nerves III, IV and VI

The central control mechanisms for eye movement comprise a complex group of pathways which adjust eye position to movement and posture, mainly by vestibular and extrapyramidal pathways.

There are two forms of voluntarily controlled eye movements:

1. visual pursuit where a specific target is fixed and followed, using parietal gaze centres closely integrated with the adjacent visual cortex

2. the ability to select a new target and relocate vision to suit by way of frontal gaze centres more allied to direct pyramidal motor pathway mechanisms.

Damage in either of these areas causes conjugate gaze palsies.

At brainstem level, the need to integrate eye movements mediated by three different cranial nerves, widely spaced in the brainstem, requires complex and extremely rapidly conducting internuclear pathways. The most critical of these is the medial longitudinal fasciculus. Damage in this pathway causes internuclear ophthalmoplegias, with disconjugate gaze palsies. The cortical influences have final relays bilaterally in the brainstem in the pons - the lateral gaze centres. There are also four gaze centres in the midbrain, two on each side; that is one to look up and one to look down. Eye movements occur in saccades, a series of little jerk movements without overshoot or undershoot, until the new position is reached. This is achieved by rapid bursts at 1000 cycles/s by cells in the gaze centres. These bursts are
initiated by voluntary information from the frontal eye fields by way of the anterior limb of the internal capsule. Automatic movements, such as occur in reading, are closely allied to visual information relayed by way of the optic tract without projection to the visual cortex. Feedback from stretch receptors in the ocular muscles is also of great importance in this type of movement. Tracking movements are controlled mainly by the superior colliculi, once the object to be followed has been located, using stereo-optic control. Vergence mechanisms require the voluntary frontal eye fields to work in conjunction with the parietal cortex with simultaneous inhibition of those brainstem mechanisms which normally prevent convergence and divergence. Only those animals with binocular stereoscopic vision have the need to converge to focus close objects.

**Parietal lobe lesions**

Poor object following or pursuit gaze problems are often difficult to demonstrate clinically as lesions in these areas also tend to cause a hemianopic field defect, so that the following movement is lost as the object moves into the blind half field. If, however, the object is kept in sight, a full range of pursuit movement is usually achieved. A tendency to ignore objects on one side (an attention defect) may be related to the inability of the eye to scan peripherally as a result of lack of sensory input.

**Frontal lobe lesions**

An irritative lesion such as a tumour or abscess in the frontal pole will drive the eyes away from the lesion. A right frontal tumour, therefore, will often cause a fit with movement of the head and eyes to the left-hand side before the patient loses consciousness. A destructive lesion, such as surgical extirpation or a cerebrovascular accident, will allow the eyes to gaze preferentially towards the side of the lesion because of the unopposed push from the intact side.

**Midbrain lesions**

Midbrain visual mechanisms are concerned mainly with upward and downward gaze. The classical lesion causing Parinaud's syndrome is a pineal tumour damaging the superior colliculus and the region of the posterior commissure. This also blocks the light reflex relays producing fixed dilated pupils, impaired upward gaze and loss of convergence. Lesions of the inferior colliculus impair downward gaze. In some instances, the ineffectual movements of the extraocular muscles in an attempt to achieve upward and downward gaze may pull the eyeball in and out of the socket, producing retractive nystagmus.

Lesions affecting the thalamic nuclei, both structural and pharmacological, may cause fixed deviations of upward or downward gaze (an oculogyric crisis). Sometimes divergence with one eye up and one eye down (skew deviation), with see-saw nystagmus on attempted lateral eye movement, may occur with lesions in this area. A haemorrhage between the third nerve nuclei produces a divergent squint, with both eyes at the extremes of lateral gaze with intact upward and downward gaze limited only by mechanical factors at this extreme position.
**Disorders affecting the midbrain**

Anteriorly, aneurysms of the upper basilar artery or a tortuous basilar artery (basilar ectasia) may damage and distort the emergent third nerves. Posteriorly, pineal tumours, distortion and dilatation of the posterior end of the third ventricle, resulting from aqueduct stenosis, cause Parinaud's syndrome. Infiltration of the superior medullary velum by direct spread of a medulloblastoma may cause bilateral fourth nerve lesions, and impaired downward gaze. Intrinsic lesions, resulting from vascular occlusion, haemorrhage, demyelinating disease and tumour, cause anterior internuclear ophthalmoplegia, that is a divergent squint with loss of convergence.

There are three named vascular syndromes of the midbrain which are caused by combinations of third nerve lesions and local pathway damage.

**Nothnagel's syndrome:** a third nerve lesion with ipsilateral ataxia resulting from infarction of the superior cerebellar peduncle.

**Benedikt's syndrome:** a third nerve lesion with contralateral cerebellar movement disorder resulting from a lesion of the red nucleus.

**Weber's syndrome:** a third nerve lesion with contralateral hemiparesis resulting from a lesion of the basis pedunculi.

**Pontine lesions**

The pontine lateral gaze centres are often damaged by vascular lesions and demyelinating disease. This results in loss of gaze to the same side as the lesion, as their descending pathways have already decussated. In a drowsy or unconscious patient, this will result in the eyes deviating towards the good side.

**Disorders affecting the pons**

Anteriorly, because of their long meningeal course, the sixth nerves are often involved in bacterial, fungal or malignant meningitis. Pontine tumours may involve the nerve nuclei or fascicular fibres and the posterior internuclear pathways. These tumours usually occur in children or adults with neurofibromatosis. Tumours blocking or infiltrating the fourth ventricle cause headache and vomiting as a consequence of cerebrospinal fluid pathway block, and sixth nerve palsies in consequence of stretching by raised intracranial pressure. If the sixth nerve palsy is caused by direct tumour infiltration, the seventh nerve should also be involved. These tumours include ependymoma, medulloblastoma, cerebellar astrocytoma or haemangioblastoma. Conditions such as multiple sclerosis, haemorrhage and infarction, metabolic disorders (vitamin B deficiency), drug intoxication and fluid balance disturbance may all cause either a conjugate gaze palsy if damaging the lateral pons, or an internuclear ophthalmoplegia with nystagmus if the lesion is in the central pons. Vascular occlusive lesions tend to cause unilateral internuclear ophthalmoplegia, as the lesion extends only to the midline. There are numerous named vascular syndromes of the pons which are a consequence of a variety of combinations of damage to the sixth and seventh nuclei and their fasciculi, and to the sensory, motor and cerebellar pathway. There is no special advantage in learning these
by heart, but the named syndromes include those of Millard Gubler, Foville, Grenet, Raymond-Cestan, Marie-Foix and Gasperini (Loeb, 1962). As a cautionary note, any hint of variability in diplopia should always raise the possibility of myasthenia gravis. If combined with variable dysarthria or swallowing difficulty, a brainstem lesion may be incorrectly suspected. This is a very difficult diagnostic trap into which even experienced neurologists may fall.

**Internuclear lesions**

Internuclear lesions are caused by multiple sclerosis (bilateral) or vascular disease (strictly unilateral unless haemorrhagic). In these instances, the lateral gaze centre is intact and abducts the ipsilateral eye normally - the relay to the opposite third nerve nucleus is blocked and the inward looking eye cannot adduct to match it. With a bilateral lesion, neither eye adducts while the abducting eye moves normally, and shows marked nystagmus. This picture is almost diagnostic of multiple sclerosis. The integrity of the upper brainstem can be demonstrated by intact vertical gaze and convergence, unless the lesion affecting these pathways actually lies between the third nerve nuclei.

**Nystagmus**

A detailed account of nystagmus is given in Chapter 4. From a simplistic neurological point of view, it is a less valuable physical sign than is often believed. The differentiation into the various types - jerk, pendular, rotatory etc - is often less easy to make than is suggested in most descriptions. Ultimately, a breakdown in the vestibular mechanisms as they affect the smoothness and stability of eye movements, is being witnessed. Weak support from vestibular mechanisms will lead to poor maintenance of gaze (slow phase) and a quick restorative movement (the jerk phase) which is the feature used to define the direction of nystagmus. This is maximal when looking away from the side of vestibular lesion, be it in the end organ, the eighth nerve or the vestibular nuclear connections. A controlling influence over vertical eye movements is also apparent in the phenomenon of vertical nystagmus which occurs with a structural or metabolic lesion of the brainstem. It is important to note that vertical nystagmus means vertical displacement of the eyes and not side-to-side nystagmus which is also seen when attempting upward and downward gaze. As defined, vertical nystagmus always indicates brainstem damage. Another feature of brainstem disease is jelly nystagmus, which is probably a consequence of the failure of inhibitory 'pause' neurons which normally stop the 'burst' neurons from producing visible little saccades.

Cerebellar lesions, especially those affecting the flocculonodular lobes, cause nystagmus as a result of the loss of the stabilizing effect of input from head posture receptors. In general, the fast phase of cerebellar nystagmus is towards the side of a cerebellar lesion.

**Group two**

The second major grouping of cranial nerves includes those lying in the cerebellopontine angle. The medial extent of the angle is defined by the sixth nerve, the upper extent by the fifth nerve and the lower extent by the ninth nerve. The seventh and eighth nerves pass in close proximity across the subarachnoid space to enter the internal auditory canal at the start of their long intraosseous courses.
The trigeminal nerve (V)

The trigeminal nerve is the largest cranial nerve. It arises from the middle of the pons and passes forwards and laterally across the subarachnoid space. Its large ganglion lies over the tip of the petrous bone where the nerve divides into its three divisions.

The ophthalmic nerve (V₁)

The first division of the fifth nerve lies below the sixth nerve in the lateral wall of the cavernous sinus and is liable to damage by the same pathologies. Because of its extensive sensory distribution, severe pain in the forehead, nose and scalp, back as far as the vertex may result from such damage.

The nerve divides into three branches as it enters the superior orbital fissure.

(1) The lacrimal nerve runs along the lateral rectus muscle to the lacrimal gland. It supplies the skin over the lateral eyelid and brow. It picks up secretomotor fibres from the zygomatico-temporal nerve which it conveys to the lacrimal gland. In the skin it receives proprioceptive filaments from the facial nerve.

(2) The frontal nerve divides into two nerves, the supratrochlear and supraorbital nerves, which supply the skin of the forehead and scalp to the vertex. They are liable to damage by minor injuries over the brow, and a causalgic syndrome may follow local trauma.

(3) The nasociliary nerve has the important autonomic and cutaneous functions:

(a) The main trunk traverses the orbit and enters the anterior ethmoidal foramen into the intracranial cavity, runs across the cribriform plate and exits from the skull through a slit in the crista galli to enter the nose. It supplies the mucosa of the nasal cavity and emerges at the lower end of the nasal bone to supply the skin over the tip of the nose, ala and vestibule.

(b) In the orbit, the nasociliary nerve gives off branches to the ciliary ganglion and two or three long ciliary nerves which carry the pupillodilator sympathetic fibres, and convey sensation from the cornea. This is of cardinal importance to the protection of the very delicate cornea.

(c) The infratrochlear branch is given off just behind the anterior ethmoidal foramen and lies on the medial wall of the orbit. It supplies the skin of the upper medial eyelid and upper side of the nose.

The corneal reflex

It is essential that otolaryngologists know how to elicit this reflex correctly. The afferent limb of the reflex is by way of the nasociliary nerve as previously described, and the efferent limb is via the facial nerve. A pointed wisp of cotton wool should be used. The patient should be asked to look upwards, whereupon, while resting the hand on the patient's cheek, the wisp should be applied to the lower cornea; care must be taken not to bring it into
vision or a blink reflex will result. The patient will flinch, the eyeball will roll up and the eye will attempt to close. Even if the seventh nerve is paralysed, the eyeball will roll up and the discomfort will be felt. The opposite eyelid will also close as this is a consensual reflex. Absence of the corneal reflex is often the first clinical evidence of fifth nerve damage.

The maxillary nerve (V2)

The middle branch of the fifth nerve ganglion lies in the extreme lower lateral wall of the cavernous sinus and exists by way of the foramen rotundum. It passes through the pterygopalatine fossa and enters the floor of the orbit by way of the inferior orbital fissure. At first, it lies in a groove in the orbital floor and then enters the short canal and exits on to the face by way of the infraorbital foramen. It then supplies the skin of the cheek, midlateral nose and lateral part of the alar, lower eyelid and the mucous membranes of the cheek and upper lip. In its course, it gives off the following branches:

1. meningeal branches to the floor of the meningeal fossa
2. two branches to the sphenopalatine ganglion conveying the secretomotor fibres destined for the lacrimal gland
3. the zygomatic nerve which lies in the floor of the orbit and divides into the zygomaticotemporal nerve (secretomotor to the lacrimal gland and cutaneous sensation to the temporal area) and the zygomaticofacial nerve which, after penetrating the zygomatic bone, supplies cutaneous sensation to the prominence of the cheek
4. the three alveolar nerves which supply the teeth, gums and adjacent palate by way of the superior dental plexus; the anterior superior branch is the largest and supplies not only the incisor and canine teeth, but also the lateral nasal wall, nasal septum, the lower eyelid and the skin of the upper lip.

The pterygopalatine (sphenopalatine) ganglion

This very large ganglion is suspended from the maxillary division, deep in the pterygopalatine fossa. It receives its main connection from the nerve of the pterygoid canal. This carries preganglionic parasympathetic fibres from the nervus intermedius (seventh nerve) and sympathetic elements from the middle meningeal artery. Both groups of fibres are then relayed by way of their complex course to the lacrimal gland. The main outflow, however, is by way of the orbital, palatine, nasal and pharyngeal nerves to the mucous membranes of the orbit, nasal passages, pharynx, palate and upper gums.

The mandibular nerve (V3)

This is the largest branch of the fifth nerve and includes the main motor component of the nerve. It exits from the skull by way of the foramen ovale; the main sensory trunk is joined by the much smaller motor root, in Meckel's cave, just outside the skull. A meningeal branch re-enters the skull with the middle meningeal artery through the foramen spinosum and supplies the lateral, middle and anterior cranial fossae. A small branch, the nerve to the medial pterygoid, supplies the medial pterygoid, tensor tympani and tensor veli palatini.
The main nerve then divides into anterior and posterior trunks. The anterior trunk conveys the bulk of the motor root to supply the masseter, temporalis and the lateral pterygoid. The main branch of the anterior trunk is the buccal nerve which merges with the buccal branches of the facial nerve to supply the skin over the buccinator, the mucous membranes of the cheek and the posterior part of the buccal surface of the gum.

The posterior trunk is mainly sensory and divides into three main nerves.

(1) The auriculotemporal nerve passes behind the temporomandibular joint to join the facial nerve with which it is distributed to the skin over the tragus, helix, auditory meatus and tympanic membrane, and, by way of superficial temporal branches, to the skin over temporalis. It also conveys secretomotor fibres to the parotid gland and fibres derived from the tympanic branch of the glossopharyngeal nerve by way of the otic ganglion (see the following).

(2) The lingual nerve supplies sensation to the presulcal tongue, the floor of the mouth and lower gums. It carries the taste fibres of the chorda tympani to the mucous membranes of the tongue. It also conveys secretomotor fibres from the submandibular ganglion to the sublingual and anterior lingual glands. It communicates with the hypoglossal nerve.

(3) The inferior alveolar (dental) nerve enters the mandibular canal running forwards in the mandible to re-emerge on the chin at the mental foramen dividing into the incisive and mental branches, supplying the skin and mucous membrane of the lower lip, jaw, incisor and canine teeth. The motor component of the posterior trunk leaves the inferior alveolar nerve, just before it enters the mandibular canal, as the mylohyoid nerve supplying mylohyoid and the anterior belly of digastric.

Central mechanisms of the fifth nerve

The central anatomy of the fifth nerve is very complicated. The small motor nucleus lies in a mid-position in the upper lateral pons opposite the nerve root. It receives bilateral supranuclear innervation by way of corticobulbar fibres leaving the main pyramidal pathways at the same level. Direct connections from proprioceptive fibres in the main sensory nucleus allow a simple stretch reflex for mastication to operate. The jaw jerk tests the integrity of this pathway and, if greatly enhanced, indicates a bilateral upper motor neuron lesion above midpontine level, the highest stretch reflex that can be elicited (McIntyre and Robertson, 1959).

The sensory nucleus is very extensive. The cell bodies of the sensory fibres lie in the gasserian ganglion at the petrous apex. At least 50% of the fibres do not enter the main sensory nucleus but are concerned with reflex activity. The other fibres form ascending and descending branches. The ascending fibres enter the mesencephalic nucleus of the fifth nerve. Their subsequent course and function is not understood. The descending fibres convey pain and temperature sensation and form a synapse in the nucleus of the descending tract of the fifth nerve which lies parallel to the descending tract and extends as low as C2 cord level. The sensory fibres derived from the facial, glossopharyngeal and vagus nerves all end in the same tract and are relayed in the nucleus. The secondary ascending pathways swings across the brainstem, ventral to the central canal to form the secondary ascending tract of the fifth
nerve which is closely associated with the medial lemniscus, adding sensation derived from the face to that of the arm and leg. In the decussation, these fibres are very vulnerable to damage by midline lesions, such as syringomyelia and syringobulbia, producing a sensory deficit extending forwards from the back of the head.

Clinical aspects of the fifth cranial nerve

Damage to the fifth cranial nerve is very important to the otolaryngologist. Branches of the nerve and its associated ganglia lie in areas often involved by otolaryngological disease, especially oropharyngeal and nasopharyngeal neoplasms.

Involvement of the motor root of the fifth nerve is quite rare as it seems to be resistant to pressure or distortion. If damaged, the wasting of the masseter is usually easy to see and palpate on teeth clenching. The pterygoids are tested by attempted jaw opening against resistance, the jaw deviating towards the paralysed side.

Painless or painful loss of sensation over any part of the face, but particularly \( V_2 \), is a very ominous finding and malignant disease in the antrum or nasopharynx is the most likely pathology. Repeated examination and biopsy of the nasopharynx is vital in such cases to establish the cause. Involvement of \( V_1 \) is usually painful and nearly always accompanied by extraocular nerve palsies. It is most commonly caused by lesions in and around the cavernous sinus, but may also be involved by malignant disease entering the orbit by way of the inferior orbital fissure.

Nasopharyngeal tumours most commonly arise in the fossa of Rosenmüller or near to the eustachian tube and are commonly anaplastic squamous cell carcinomata. Tumours originating in the maxillary antrum or ethmoids are usually squamous cell or adenocarcinomata. Forty per cent of such tumours present as neurological problems. In 70% of cases the fifth nerve is involved; in 50% of cases nerves III, IV and VI are involved. Visual pathways are affected in 8.5% of cases, and the lower cranial nerves in 10%. The favourite routes of entry into the skull are through the inferior orbital fissure or by way of the foramen lacerum with the carotid artery (Godtfredsen, 1944).

The \( V_2 \) division runs across the mouth of the eustachian tube and the fossa of Rosenmüller, through the orbital floor just above the antrum and on to the face. Nasopharyngeal and antral carcinomata are particularly likely to damage this division and seem to cause loss of sensation more frequently than pain. The surface branches of both \( V_1 \) and \( V_2 \) are easily damaged by blunt trauma around the orbit and cheek, or divided by lacerations. \( V_3 \) is involved in oropharyngeal, tonsillar and mandibular tumours; and, as noted previously, painless numbness over the chin may be the presenting symptom, rather than pain.

Trigeminal sensory neuropathy is a very rare condition in which painless numbness over the fifth nerve territory progressively develops, usually starting in the second division and becoming bilateral. Only the passage of time and failure to demonstrate a responsible lesion allow this diagnosis to be entertained (Spillane and Wells, 1959).

The sensory root of the fifth nerve is very sensitive to distortion and pressure, and loss of the corneal reflex is an important early sign of a lesion in the cerebellopontine angle. It
is rare for extensive loss of sensation over the face to be the presenting symptom of an acoustic neuroma.

**Trigeminal neuralgia**

Trigeminal neuralgia is probably the most painful condition known, in contrast to the cause, which would appear to be minor ageing changes in the nerve or minor irritation by adjacent arteries. From a practical anatomical point of view, the very strict localization of the pain into fifth nerve territory is vital. There is no such entity as atypical trigeminal neuralgia and it is not acceptable to allow the pain to radiate behind the ear, on to the neck, or across the midline, and the exact distribution is the linch-pin of diagnosis. The pain usually occurs in two characteristic distributions. The first runs from the lower canine tooth along the lower jaw to just in front of the ear and sometimes round into the upper jaw, that is it involved both V₃ and V₂. The second less frequent type runs from the upper incisor or canine, up the side or inside the nose and encircles the eye, involving both V₂ and V₁. It is probably this spread over two divisions that makes simple peripheral branch section unsuccessful in managing the condition, although triggering can occasionally be reduced. Although it is claimed that transient sensory deficit may follow a spasm of pain, any evidence of sensory loss, impaired corneal reflex or fifth nerve motor weakness, should invalidate the diagnosis. Although trigeminal neuralgia may complicate multiple sclerosis, it is very rare as a presenting symptom of the latter disease. The condition is dealt with in greater detail in Volume 4.

**Herpes zoster ophthalmicus**

Most patients with this condition develop severe pain in the distribution of V₁. The pain lasts 4-5 days. During this time, the diagnosis of ruptured aneurysm, cranial arteritis or acute frontal sinusitis may all have to be seriously considered. The vesicles usually appear in the medial part of the eyebrow. They then involve the entire distribution of the nerve branch. Severe chemosis of the eye and extraocular nerve palsies may further complicate the picture.

**Aneurysmal dilatation of the carotid artery**

This is the other major condition in the elderly that can cause very severe pain in a V₁ distribution, with chemosis, extraocular nerve palsies and even blindness. The onset is usually very sudden, and the condition typically occurs in elderly females with long-standing hypertension.

**The facial nerve (VII)**

The seventh nerve is primarily motor to the muscles of facial expression. It also conveys the important taste fibres from the tongue by way of the chorda tympani and taste from the palate by way of the nerve of the pterygoid canal. A small but clinically important cutaneous supply to the skin of the external ear is mediated in fibres carried by way of the vagus. These sensory fibres are contained in a separate trunk, the nervus intermedius, which runs with the eighth nerve rather than the seventh nerve in the subarachnoid space. The cell bodies of the sensory root lie in the geniculate ganglion. The nervus intermedius also carries preganglionic parasympathetic secretomotor fibres to the submandibular and sublingual
salivary glands. These fibres originate in the superior salivatory nucleus. Several important branches arise from the intrapetrous part of the nerve.

(1) The greater petrosal nerve arises from the geniculate ganglion. It carries taste fibres from the palate, and it conveys preganglionic parasympathetic fibres to the pterygopalatine ganglion, and, by way of the zygomaticotemporal and lacrimal nerves, to the lacrimal gland. It is joined by the deep petrosal nerve (derived from the sympathetic plexus on the carotid artery) to form the nerve of the pterygoid canal.

(2) A branch from the ganglion joins the lesser petrosal nerve and thence the otic ganglion. This conveys secretomotor fibres to the parotid gland which reach it by way of the auriculotemporal nerve. It also carries sympathetic fibres derived from the carotid artery to the blood vessels of the gland.

(3) A small twig, the nerve to stapedius, arises 6 mm above the stylomastoid foramen.

(4) The chorda tympani arises at the same level and runs forward across the middle ear and enters a canal in the petrotympanic fissure, grooves the spine of the sphenoid and joins the lingual branch of the fifth nerve with which it is distributed to the presulcal part of the tongue.

(5) At the stylomastoid foramen, twigs join both the vagus and glossopharyngeal nerve.

(6) The posterior auricular nerve supplies the muscles of the ear and occipital belly of occipitofrontalis.

(7) The branches to the muscles of facial expression are from above downwards - namely the temporal, zygomatic, buccal, mandibular and cervical - passing through the parotid gland.

(8) Cutaneous fibres are distributed with the auricular branch of the vagus supplying the skin on both sides of the auricle and part of the external auditory canal and tympanic membrane.

The submandibular gland

The submandibular ganglion lies on the lingual nerve. Its preganglionic fibres are derived from the superior salivatory nucleus, and reach it by way of the facial nerve, chorda tympani and lingual nerve. These fibres are secretomotor to the submandibular and sublingual glands. The sympathetic components are derived from the plexus on the facial artery and pass uninterrupted through the ganglion to the blood vessels of the same glands.

The central connections of the facial nerve

The nucleus lies in a deep position in the central pons. The dorsal part of the nucleus receives bilateral supranuclear innervation, whereas the lower part of the nucleus receives mainly contralateral supranuclear innervation. This has important consequence for the clinical
varieties of seventh nerve lesions. The nucleus is closely related to the fifth nerve and this provides a suitable arrangement for the important corneal reflex and its own reflex activity by way of the nucleus of the tractus solitarius. The fascicular course of the nerve is unusual in that the fibres course towards the floor of the fourth ventricle, wrap around the nucleus of the sixth nerve, producing a visible enlargement on the floor of the ventricle (the facial colliculus), and then retrace their course across the entire depth of the pons to exit at the pontomedullary junction. This complex arrangement is thought to result from the migration of the nucleus from an original position in the floor of the fourth ventricle to achieve its close relationship to the nucleus of the fifth nerve and the nucleus of the tractus solitarius.

**Taste mechanisms**

Taste is mediated by means of taste buds - some 50 cells arranged in a pear-like cluster. These are found on the tongue, undersurface of the palate, palatoglossal folds, posterior wall of the pharynx, posterior surface of the epiglottis and the upper third of the oesophagus. They are most numerous on the lateral tongue and decrease in number with age by about 1% per annum. Each taste bud opens on the surface of the mucous membrane as a pore. The buds are found in the vallate, fungiform and foliate papillae. The lifespan of these cells, which are renewed from epithelial cells surrounding the bud, is about 10 days. They are, therefore, very vulnerable to factors inhibiting rapid cell turnover.

Two main receptor cells have been identified, although they are possibly different types of the same cell. Some receptor cells have receptor sites for afferent neurons and small presynaptic vesicles; others contain larger vesicles and have more definite ciliary processes at their tip, just inside the pore. There is evidence of considerable cross-innervation of taste buds which may indicate inhibitory and facilitatory control similar to that seen in the smell receptors. It is thought that patterns of taste over a wide area of receptors are critical in perceiving different tastes, rather than specific receptors being responsible for specific tastes.

The neural connections of the taste receptor cells are the unipolar processes of cells in the geniculate ganglion of the seventh nerve, the inferior ganglion of the ninth nerve and the inferior ganglion of the tenth nerve. The central processes of these cells form the tractus solitarius and they synapse in the adjacent nucleus of the tract. These fibres then ascend in the medial lemniscus to the opposite nucleus ventralis posterior medialis of the thalamus. The final pathway is by way of the internal capsule to the sensory cortex and insula. Some information from the pons relays to the hypothalamus for autonomic reflex purposes. The anatomy of the peripheral taste pathways is complex and, for practical purposes, the supply of the anterior two-thirds of the tongue is mediated by means of the chorda tympani but distributed in the lingual branch of the mandibular division of the fifth nerve. The facial nerve, by way of the greater petrosal nerve and the nerve of the pterygoid canal, also conveys sensation from the taste buds on the palate, through the middle and posterior palatine nerves. Taste sensation from the vallate papillae, pharyngeal tongue and palatoglossal folds is conveyed by fibres carried in the ninth nerve. Taste sensation from the lowest part of the tongue, epiglottis and hypopharynx is carried by the vagus by way of its superior laryngeal branch.

Free nerve endings of the fifth nerve are also widespread, conveying somatic sensation from these areas. Furthermore, they undoubtedly contribute to the perception of extremely
strong stimuli, such as curry powder, carbonated drinks and acid substances; modifications of this pattern of gustatory and simple physical stimuli can alter taste sensation, heightening the unpleasant features of such highly flavoured compounds. It is clear that taste mechanisms are rather more complex than the generally accepted permutations of sweet, bitter, salt and sour. Parallel smell appreciation adds savour to taste. Patients with loss of smell describe all food as tasting like cardboard and only highly spiced or flavoured foods make any impact, often not a pleasant one. Once again, adaptation plays a role. The modification of fruit juice flavours by the previous use of mint toothpaste is a universally appreciated phenomenon. Because of the vital role of smell in taste appreciation, and the frequent simultaneous impairment of smell, it is difficult to isolate specific taste disorders. For example, in Bell's palsy, patients identify tastes as having a metallic flavour in spite of the lesion being strictly unilateral and with no impairment of smell. Chemicals and systemic diseases that modify taste and smell are listed in the previous section on smell.

**Clinical disorders of the seventh nerve**

The seventh nerve is frequently damaged by diseases of otolaryngological origin inside the skull, in the petrous bone and in the parotid gland (Tschiassny, 1953).

For reasons noted previously, a cortical lesion affecting the seventh nerve function, such as a vascular lesion or tumour in the motor strip, will cause weakness maximal in the lower face which is in particular forehead movement and eye closure, will be relatively unaffected, on account of bilateral supranuclear innervation. This is an upper motor neuron facial weakness, and in many instances is more apparent during spontaneous smiling and speaking than during deliberate attempts to move the face to command.

Lesions affecting the whole facial nucleus or peripheral part of the nerve should cause total weakness. In some instances, if weakness is more marked in the lower face - which may occur in the early or recovery phase of a simple Bell's palsy - an upper motor neuron lesion may occur and mimic a lower motor neuron lesion. These difficulties are being stressed because the distinction is of immense diagnostic importance and mistakes are easily made.

Lesions in the brainstem usually also involve the sixth nerve on account of the intimate anatomical relationship, and long tract signs may also be detected on careful examination.

Where the nerve crosses the subarachnoid space and enters the auditory foramen, it lies in very close relationship with the eighth nerve; this is the cerebellopontine angle. An acoustic neuroma is the most frequent lesion found in this area. Acoustic neuromata, although grossly distorting the seventh nerve, very rarely present as a seventh nerve palsy. If there is clinical evidence of a cerebellopontine angle lesion, and if the seventh nerve is involved, alternative pathology is likely (Thomsen, 1976). Permanent damage following surgical removal of an acoustic neuroma is very common.

In the facial canal, the nerve is liable to ischaemic damage and this is the probable mechanism of Bell's palsy in which the nerve is thought to be damaged by the inflammatory response to an antecedent viral infection. In nearly all cases, very severe pain in the ear occurs in the 24 hours before the onset of the Bell's palsy. It is particularly severe and
persistent if herpes zoster is responsible (Ramsay Hunt syndrome). The pain and local swelling may suggest bacterial infection until the vesicles appear 3-4 days later. The facial paralysis is usually complete on the second day and includes occipitofrontalis and platysma. Hearing distortion caused by paralysis of the stapedius, and impaired taste resulting from involvement of chorda tympani, do not always occur, and in mild cases the lower half of the face may be more severely affected than the upper half, as discussed previously (Taverner, 1955).

Seventy-five per cent of patients make a good recovery over 3-6 weeks, with or without treatment. Twenty per cent make an acceptable but slow recovery, complicated by the development of facial synkinesis. This is a consequence of nerve sprouting with subsequent loss of fine control which can turn a smile into a snarl and eye closure into a distorted grimace. Five per cent of cases make little or no recovery and may ultimately require plastic surgical repair. In some cases, aberrant regeneration may lead to lacrimation instead of salivation on eating, so-called crocodile tears (Chorobski, 1951). It is most important that patients with Bell's palsy are not told that they have had a small stroke. Exclusion of underlying hypertension, diabetes, sarcoidosis and inflammatory arterial disease is important.

Middle ear infection especially if associated with cholesteatoma carries a considerable risk of damaging the nerve by similar mechanisms. There is a 1% risk of damage to the facial nerve during mastoid surgery. Fractures through the petrous bones are often complicated by facial nerve palsy. Those of immediate onset are usually caused by nerve laceration. Those of delayed onset, usually 2-3 days after trauma, are caused by oedema and carry an excellent prognosis. Trauma to the nerve as it emerges from the stylomastoid foramen is a well recognized complication of forceps delivery.

Benign hemifacial spasm can occur in either sex and at any age, but seems to be more common in elderly hypertensive females. Since the advent of scanning, a surprising number of underlying lesions have been found in this condition, such as cholesteatoma, acoustic neuromata, meningiomata, or aneurysms of the basilar artery. Many regard CT scanning as a necessary investigation. The symptoms consist of a constant flickering and twitching of the facial muscles. This usually starts around the eye producing involuntary winking and later extends to involve the mouth. It is usually worse in company but continues 24 hours a day. The condition may respond to carbamazepine (Tegretol) but, if the patient's age and condition allow, posterior fossa exploration to identify vascular irritation by a small vessel and exclude other lesions is indicated (Ehni and Woltman, 1945).

Clinical testing of the seventh nerve

A standard sequence of movements should be tested. Wrinkling the forehead, followed by forced eye closure will usually reveal weakness in the upper half of the face. The ability to flare the nostrils and wrinkle the nose should be tested, followed by a forcible showing of the teeth and an attempt to blow out the cheeks. Eversion of the lower lip is difficult to achieve but it is a means of testing the perioral muscles and does produce striking contraction in platysma. It should take only about 30 seconds to perform these tests. Hearing loss is not usually detected by simple clinical testing, although the patient may report distorted hearing.
In the same way, formal tasting of taste with standard test flavours may be carried out but often the patient's own perception of altered taste will be adequate for diagnostic purposes.

Whenever the seventh nerve is damaged, it is important to exclude coexistent fifth nerve damage, in particular the presence of the corneal reflex. Not only will this exclude a simple Bell’s palsy, but the considerable danger to an unprotected and anaesthetic cornea will be identified. Eye movements should be carefully tested to exclude a sixth nerve lesion, which would indicate brainstem damage. Simple clinical tests of hearing should also be performed, particularly if the corneal reflex is depressed, as simultaneous involvement of these three nerves would indicate a lesion in the cerebellopontine angle. It should be remembered that herpes zoster may affect several cranial nerves simultaneously and can cause severe pain which, when accompanied by multiple cranial nerve palsies, can present a very difficult diagnostic situation until the vesicles appear.

The vestibulocochlear nerve (VIII)

The anatomy and physiology of the specialized end organs of the eighth nerve are discussed in the first four chapters of this volume. Therefore, discussion here is confined to the role of hearing impairment and balance disorders in the diagnosis of neurological disease.

Because of the anatomical proximity of the seventh and eighth nerves, simultaneous involvement under all circumstances would seem likely. In reality, such damage is quite unusual, with the exception of acute traumatic lesions of the petrous bone in which both nerves are lacerated simultaneously. These peculiarities are of considerable clinical importance.

The cerebellopontine angle syndrome

The most frequent tumour found in the angle is an acoustic neuroma. Although arising on the vestibular division of the nerve, growth is usually so insidious that a purely vestibular presentation is extremely unusual. Gradual and often unrecognized impairment of hearing is the rule. Similarly, the seventh nerve may become grossly distorted by facial hemispasm or weakness as a presenting symptom is also unusual. In contrast, minimal pressure on the fifth nerve root as the tumour extends upwards, or perhaps stretching of the fifth nerve root as the pons is displaced, regularly produces impairment of the corneal reflex. In spite of this, frank pain or numbness in the face is also very unusual. An acoustic nerve tumour may obviously present as facial hemispasm, facial weakness, facial numbness or a trigeminal neuralgia-like syndrome. In all such instances, however, a cholesteatoma or meningioma in the cerebellopontine angle is a more likely diagnosis. If the clinical picture has evolved extremely rapidly, either metastatic carcinoma or lymphoma could be responsible. Less frequently, and usually in a younger age group, pontine glioma or cerebellar medulloblastoma can extend into the cerebellopontine angle to produce the typical combination of nerve lesions. The age of the patient, usually between 5 and 15 years, should provide a strong clue to these diagnostic possibilities.
Balance disorders and vertigo

Unsteadiness is a common symptom for referral to otolaryngological or neurological clinics. The most important consideration is that of establishing what the patient means by the complaints of being 'off balance', 'giddy', or 'dizzy'. Often, close questioning will reveal that the patient means 'light headed', 'floaty' or 'woozy', all non-specific symptoms usually resulting from anxiety. The patient will not have the illusion of his own movement or that of his surroundings, which is an indication of true vertigo and, therefore, a justification for extensive otoneurological investigation to define the cause.

Disorders of balance, such as Ménière's disease, vestibular neuronitis and benign positional vertigo, will be discussed in detail elsewhere. The frequency with which vertigo occurs as a symptom in migraine attacks also deserves a mention. A feature of all these situations is that the attacks are episodic or provoked by change of position.

Disorders of balance caused by structural organic disease in the central nervous system tend to produce both continuing difficulty with balance and non-stop vertigo. The most frequent causes are multiple sclerosis and cerebral vascular accidents, affecting vestibular and cerebellar connections in the brainstem. Pure cerebellar lesions are less likely to produce vertigo unless they distort the brainstem. They usually produce impaired coordination or a tendency to veer to one side while walking, rather than the drunken reeling with vertigo which is seen in patients with brainstem lesions.

For further discussion, readers are referred to the first four chapters of this volume and later volumes in the series.

Group three

The final group of cranial nerves are not only anatomically bunched at their major exit, the jugular foramen, but share common nuclear origins. They also have peripheral cross-connections for final distribution that make for poor physiological distinction of function as well as complex anatomy. Only the hypoglossal nerve, with its discrete nuclear origin and separate hypoglossal canal, can be discussed in isolation. Even then, its peripheral course brings it into close anatomical relationship with the other three nerves.

The glossopharyngeal nerve (IX)

The glossopharyngeal nerve has sensory, motor and autonomic components. The sensory ganglion cells lie in the superior and inferior ganglia of the nerve, and the central processes pass to the nucleus of the tractus solitarius, conveying taste sensation, and to the nucleus of the spinal tract of the fifth nerve conveying somatic sensation. The motor nucleus is the upper part of the nucleus ambiguus which receives bilateral supranuclear innervation from corticobulbar fibres. This nucleus supplies the stylopharyngeus. The autonomic parasympathetic fibres arise in the inferior salivatory nucleus. These fibres reach the lesser petrosal nerve by way of the tympanic branch and relay in the otic ganglion. The postganglionic fibres are distributed to the parotid gland by way of the auriculotemporal nerve.
The glossopharyngeal nerve emerges from the brainstem in line with the vagus and accessory nerves and exits from the skull by way of the jugular foramen. It descends between the jugular vein and carotid artery picking up sympathetic fibres from the carotid plexus as it loops forwards and medially to reach the soft tissues of the oropharynx, posterior tongue and palate. In its course, it gives off the tympanic (Jacobson's) nerve, conveying the secretomotor fibres for the parotid gland to the otic ganglion by way of the tympanic plexus and lesser petrosal nerve. An important nerve, the carotid branch, innervates the carotid body and carotid sinus conveying, respectively, chemoreceptor and stretch reflex information centrally for respiratory and circulatory reflex function. The final branches are the pharyngeal tonsillar and lingual branches, conveying general sensation and taste from the appropriate areas.

The otic ganglion

The otic ganglion lies just below the foramen ovale, attached to the mandibular nerve but functionally conveying information from the glossopharyngeal nerve. The parasympathetic fibres relay in it and supply the parotid gland by way of the auriculotemporal nerves. Sympathetic fibres from the middle meningeal artery pass through the ganglion and are distributed to the blood vessels of the parotid gland in the same nerve.

Glossopharyngeal neuralgia

This is a rare condition occurring at about one-tenth the frequency of trigeminal neuralgia. It consists of excruciatingly severe pain in the palate, throat and external auditory canal, locations demonstrating the somatic sensory distribution of the nerve. The pain has the typical burning, electric shock quality of neuralgia and is mainly triggered by swallowing. The incidence of underlying lesions inside the skull is though to be very much higher than in trigeminal neuralgia. Both phenytoin and carbamazepine may control the pain. CT scanning would seem a wise precaution in all instances, but small lesions may be missed and intracranial root exploration is necessary if medical treatment fails. Peripheral glossopharyngeal section has little to commend it, and can seriously interfere with normal swallowing mechanisms (Ekbom and Westerberg, 1966).

The vagus nerve (X)

The vagus nerve (the wanderer) is the most widely distributed cranial nerve, hence only those aspects essential to otolaryngologists will be detailed.

The central connections are similar to the ninth nerve.

(1) The dorsal nucleus of the vagus contains motor and sensory components. The motor fibres are general visceral efferent to the smooth muscle of the bronchi, heart, oesophagus, stomach and intestine. The sensory fibres are general visceral afferent originating in the oesophagus and upper bowel with cell bodies in the superior and inferior vagal ganglia.

(2) The nucleus ambiguus gives origin to fibres controlling the striated muscle of the pharynx and intrinsic muscles of the larynx. It has a bilateral supranuclear innervation.
(3) The nucleus of the tractus solitarius is shared with the glossopharyngeal nerve and receives fibres from the taste buds of the epiglottis and vallecula.

(4) The spinal nucleus of the fifth nerve receives general somatic afferent fibres from the pharynx and larynx.

Because of these extensive nuclear connections, multiple rootlets emerge from the brainstem and form a flat cord which enters the jugular foramen. The superior and inferior ganglia lie both in the foramen and just below it, identical to the glossopharyngeal nerve. Both ganglia make connections with the accessory and hypoglossal nerves and the sympathetic plexus on the carotid artery. Below the inferior ganglion, the cranial root of the accessory nerve merges into the vagus nerve which distributes its fibres to the pharynx and larynx.

The vagal branches of practical importance are as follows.

(1) A meningeal branch supplying the dura of the posterior fossa is given off in the jugular foramen.

(2) The auricular branch arises from the superior ganglion. It is joined by a branch from the glossopharyngeal and is distributed to the skin of the external ear with the branch of the facial nerve. These fibres all enter the nucleus of the descending tract of the fifth nerve.

(3) The pharyngeal branch arises just above the inferior ganglion and distributes the accessory components to the pharyngeal plexus, supplying the pharynx and palate.

(4) The superior laryngeal nerve comes off the inferior ganglion and divides into two branches. The internal laryngeal nerve which supplies sensation to the mucous membrane of the larynx and proprioceptive information from the neuromuscular spindles and stretch receptors of the larynx; and the external laryngeal nerve which supplies the cricothyroid and contributes to the pharyngeal plexus, and which is of considerable importance in speech mechanisms.

(5) The recurrent laryngeal nerve has differing courses on each side. On the right it loops under the subclavian artery and on the left under the aortic arch. On both sides it then ascends on the side of the trachea. It supplies all the muscles of the larynx, except the cricothyroid, and carries sensory fibres from the mucous membranes and stretch receptors of the larynx.

The spinal accessory nerve (XI)

The cranial part of this nerve is a detached portion of the vagus and the spinal part is motor to the sternocleidomastoid and trapezius.

The cranial portion arises from the lower part of the nucleus ambiguus and a small component from the dorsal efferent nucleus of the vagus. The nerve rootlets emerge in line with the vagus, are joined by the ascending spinal component and then run laterally to enter the jugular foramen. The cranial portion merges with the vagus at the level of the inferior
vagal ganglion and is then distributed in the pharyngeal and recurrent laryngeal branches of the vagus. These fibres probably supply the muscles of the soft palate.

The spinal root arises from the ventral horn cells from C1 to C5. The fibres emerge from the cord laterally between the anterior and posterior spinal nerve roots to form a separate nerve trunk ascending into the skull through the foramen magnum. It then exits from the skull by way of the jugular foramen in the same dural sheath as the vagus. It runs posteriorly as soon as it emerges to supply the sternocleidomastoid and the upper part of trapezius, and it receives a major contribution from branches of the anterior roots of C3 and C4 to form a plexus which supplies the cervical musculature. Surgical evidence suggests that these root components make important contributions, as upper cervical root section is required to denervate completely the sternocleidomastoid and trapezius. The peripheral portion of the nerve is easily damaged in lymph node biopsy and other operations in the posterior triangle of the neck (Eisen and Betrand, 1972).

The accessory nerve is unusual in that clinical evidence indicates that its supranuclear innervation is ipsilateral. In hemiparetic vascular lesions, the weakness in sternocleidomastoid is on the same side as the lesion. In epileptic fits originating in the frontal pole, the head turns away from the side of the lesion, that is the ipsilateral sternocleidomastoid is contracting. If there is a failure to recognize this distribution, the symptoms may seem to indicate that a patient with a left hemiparesis has a right accessory nerve lesion, and hence a lower brainstem lesion, rather than a simple capsular cerebrovascular accident. This is an easy mistake to make.

The hypoglossal nerve (XII)

The hypoglossal nerve arises from a nuclear column in the floor of the fourth ventricle derived from the same cell groups as the nuclei of nerves III, IV and VI. In the same way as nerves III and VI, the fascicular fibres have to traverse the full sagittal diameter of the medulla to exit from the ventral surface of the medulla between the pyramid and olive. The numerous rootlets combine and become two main fasciculi with their own dural sleeves, and exit by way of the hypoglossal canal just below the jugular foramen. The nerve, therefore, emerges deep to the other structures and has to course downwards and anteriorly to emerge between the jugular vein and carotid artery, cross the inferior vagal ganglion and then course upwards and anteriorly on the hyoglossus, distributing branches to all the muscles of the tongue. It receives sympathetic fibres from the superior cervical ganglion, some fibres from the vagus and the motor roots of C1 and C2 by way of the ansa cervicalis. Numerous filaments connect to and are distributed with the lingual nerve.

Fibres derived from the hypoglossal nucleus supply the styloglossus, hyoglossus, geniohyoid and genioglossus. The fibres derived from the C1 components are distributed to the sternohyoid, sternothyroid, omohyoid, thyrohyoid and geniohyoid. Although a twelfth nerve lesion paralyses one side of the tongue as its most obvious feature, the larynx is also pulled across to the opposite side on swallowing, in consequence of the failure of hyoid elevation on the paralysed side.

The supranuclear innervation of the hypoglossal nucleus is usually bilateral but can be mainly contralateral. The nerve is particularly vulnerable to surgical trauma in operations
on the neck for malignant disease and during carotid endarterectomy (Dehn and Taylor, 1983). Paralysis following central venous catheterization has also been reported (Whittet and Boscoe, 1984).

**The cervical sympathetic nerve**

Horner's syndrome is caused by damage to the cervical sympathetic nerve and is one of the most frequently missed physical signs in medicine. In the present context, its detection is of vital importance.

The cervical sympathetic nerve originates in the ipsilateral hypothalamus, runs through the entire dorsolateral brainstem to the cervical spinal cord at T1 level. The fibres leave the cord, by way of the ventral root, join the sympathetic chain and ascend through the various ganglia to end as a plexus on the carotid artery, on which they re-enter the intracranial cavity. It supplies sympathetic fibres to all the cranial nerves innervating the pupil, glands and blood vessels of the head and neck.

There are said to be several external evidences of Horner's syndrome which are as follows:

1. enophthalmos is rarely visible and of dubious authenticity
2. loss of sweating over the face and forehead is rarely noted unless specifically tested by warming the patient
3. ptosis of the eyelid may be very subtle and somewhat variable, as the nerve endings become sensitized to circulating adrenaline caused by denervation hypersensitivity; the lid rarely drops lower than the edge of the pupil
4. pupilloconstriction, or more accurately a failure of pupillodilatation, leads to an entirely normally reactive pupil to light and accommodation, but through a smaller range
5. congenital Horner's syndrome can occur and is associated with failure of pigmentation of the affected iris which remains blue.

**Causes of Horner's syndrome**

The causes of Horner's syndrome are as follows:

1. lesions in the dorsolateral brainstem, especially vascular lesions in the medulla, multiple sclerosis at any level, or pontine glioma
2. lesions in the central cervical cord, syringomyelia, ependymoma, glioma or traumatic damage
3. lesions of T1 roots, apical carcinoma of the lung, cervical rib, aortic aneurysm or avulsion of the lower brachial plexus
(4) lesions of the sympathetic chain in the neck, thyroid carcinoma, thyroid surgery, neoplastic lesions, local trauma, accidental surgical damage or surgical extirpation for various vascular syndromes of the arm

(5) lesions of the carotid plexus, carotid artery surgery, carotid artery thrombosis, migrainous spasm, local neoplastic destruction in the skull base or involvement by aneurysm or malignancy in the region of the carotid siphon.

**Clinical evaluation of the last four cranial nerves**

Multiple involvement of the last four cranial nerves is extremely common, so that the symptoms and signs of individual nerve lesions can be difficult to isolate, both from looking at the history and on examination. Disorders of swallowing, speaking, coughing and pain syndromes are the usual presenting symptoms.

A glossopharyngeal nerve lesion will cause impaired taste sensation over the posterior third of the tongue, but this is usually asymptomatic, and impossible to test. The loss of somatic sensation over the palate and oropharynx will cause impaired swallowing reflexes as the initial stimulus to deglutition is the arrival of the bolus against the palate. This will lead to occasional choking on food and fluids. Pain in the throat and ear may occur with sensory fibre irritation and true glossopharyngeal neuralgia may result. This characteristically will be triggered by swallowing.

Sensation over the palate should be tested by touching the palate with an orange stick and if sensation appears blunted, this can be confirmed using a long hatpin. A further check can be made by touching the posterior pharyngeal wall while the patient says, 'Ah', to elevate the palate.

A vagal nerve lesion at brainstem or jugular foramen level will affect the palate and vocal cords. Unilateral weakness of the palate causes nasal speech and a tendency for food to come back up the nose. Vocal cord paralysis will cause a hoarse soft voice and prevent explosive coughing. The failure adequately to protect the airway, as swallowing is initiated, leads to spluttering of food and fluids, with secondary regurgitation through the nasal passages. Pain in the ear may result from irritation of the sensory fibres in the nerve. In a peripheral recurrent laryngeal nerve lesion, the palate will not be affected, but the voice symptoms will be similar and a tendency to choke on fluids will still be seen. It has recently been recognized that the external laryngeal nerve and its supplied muscle, the cricothyroid, has a greater effect on speech than previously realized. Damage causes more severe and lasting speech problems than results from a recurrent laryngeal nerve lesion (Kark et al, 1984).

The integrity of the vagus can be assessed by the patient's voice, ability to cough, direct inspection of the palate and indirect or fibreoptic laryngoscopy.

An accessory nerve lesion is really a spinal root lesions as the cranial part of the nerve is distributed with the vagus. Weakness and wasting of the sternocleidomastoid and the upper part of trapezius is readily demonstrable, provided that it is carefully sought. In the same way as with Horner's syndrome, this is a physical finding that is easily missed.
A hypoglossal nerve lesion produces paralysis of the intrinsic musculature of the tongue on the same side. This causes surprisingly little disability and is often discovered accidentally by the patient or his dentist. Once recognized, some slight difficulty with chewing may become apparent.

On examination at rest, the affected side of the tongue will be shrivelled and fasciculating. On attempted tongue protrusion, the tongue will deviate towards the affected side.

**Clinical involvement of nerves IX, X, XI, XII and cervical sympathetic nerves**

As the last four cranial nerves lie in close proximity to one another inside the skull and even closer to one another outside the skull, multiple involvements are the rule and a variety of named syndromes have been reported. Of immediate practical importance are four major anatomical features.

(1) The proximity in the brainstem of the nucleus of IX, X and XI to the spinothalamic tract and descending cervical sympathetic produces multiple nerve involvement, sparing XII and affecting spinothalamic sensation on the opposite side of the body and associated with an ipsilateral Horner's syndrome.

(2) The twelfth nerve lies in a different brainstem vascular territory and the nerve emerges lateral to the pyramid. A vascular lesion of this area will produce a twelfth nerve lesion with contralateral hemiplegia and contralateral impairment of posture sense and touch. As the nerve exits through a separate foramen, it is often spared by a lesion involving the jugular foramen structures from above.

(3) Outside the skull, all four nerves lie so close together that the twelfth nerve is less likely to be spared and is often the first structure affected by lesions infiltrating the area from the oropharynx. Any mass in this region is often palpable.

(4) As the cervical sympathetic has ascended into the region, its involvement (provided that there is no evidence of a brainstem lesion) is certain evidence of an external jugular foramen lesion.

**The named syndromes**

Eponymous syndromes have been applied to almost every conceivable permutation of nerve and tract involvement affecting the last four cranial nerves. Those terms in regular use are as follows:

*Vernet's syndrome* (of the internal jugular foramen) is characterized by the involvement of nerves IX, X and XI only (an identical syndrome has been attributed to Schmidt).

*Avellis' syndrome* (of the brainstem) involves nerve X and the contralateral spinothalamic tract (loss of pain and temperature only).
Tapia's syndrome involves nerves X and XII. It is difficult to see how this syndrome occurs at either brainstem or peripheral level in view of the anatomical reasons above. Presumably this would be a chance association.

Jackson's syndrome is characterized by the involvement of nerves X, XI and XII. Again, it is difficult to see how this combination could occur on any logical basis; presumably, as a consequence of a chance peripheral combination of lesions.

Collet-Sicard syndrome (of the posterior lacero-condylar space). This is basically an external jugular foramen syndrome involving nerves IX, X, XI and XII but sparing the cervical sympathetic nerve.

Villaret's syndrome (of the posterior retropharyngeal space). This is involvement of nerves IX, X, XI and XII and the cervical sympathetic nerve and is diagnostic of an external jugular foramen syndrome.

Wallenberg's syndrome (infarction of the dorsolateral medulla). This consists of lesions of nerves IX and X, the cervical sympathetic nerve, contralateral spinothalamic loss in the limbs, ipsilateral spinothalamic loss over the face and severe vertigo, vomiting and hiccoughs.

Causes (excluding cerebrovascular accidents affecting brainstem)

Intracranial lesions

Neuromata of nerve XII, less frequently of nerves IX, X and XI, and rarely an acoustic neuroma may extend down into the internal jugular foramen.
Meningioma of the lateral recess.
Cholesteatoma (particularly likely to affect VII and IX).
Meningitis (especially malignant or chronic).
Fracture of the skull base.

Extracranial lesions

Thrombosis of the jugular bulb.
Metastatic tumour in the carotid sheath lymph nodes.
Retropharyngeal abscess or neoplasm.
Glomus jugulare tumour. This may start externally and erode through the petrous bone, or start within the vein in the petrous bone and erode through the skull base.

The presenting symptoms in these cases may be the following:

(1) persistent occipital headache, often resembling a migraine
(2) persistent otalgia, which may be aggravated by swallowing
(3) hoarse voice, pain in the throat or persistent sore throat
(4) difficulty in swallowing, choking or nasal regurgitation.
Computerized tomographic scanning has revolutionized the investigation of these syndromes. Previously, plain skull films, tomography, and carotid angiography were used and often failed to establish a diagnosis. CT scanning will reveal very early evidence of skull base erosion or infiltration by tumour.

**Bulbar palsy**

The differential diagnosis of lower cranial nerve lesions includes those conditions destroying motor nuclei in the brainstem. Poliomyelitis has, fortunately, become a condition of the past, and the commonest cause now is motor neuron disease. The presenting symptoms consists of a tendency to cough and splutter, initially on fluids but then extending to include all consistencies. Nasal regurgitation and aspiration are common. Speech becomes progressively unintelligible and the patient typically arrives in the clinic clutching handkerchief to his mouth, with a written list of complaints. In the early stages, poor palatal movements, poor tongue movements and weakness of jaw closure and opening may be found, but the symmetry of involvement may make it difficult to identify mild disability. Fasciculation may be seen in the tongue and facial muscles, or palpated in the masseter. Long tract signs are important and a brisk jaw jerk, increased reflexes and extensor plantars would provide strong supporting evidence for the diagnosis. Myasthenia gravis of the bulbar type is the most important differential diagnosis. Although variability ought to be the hallmark of this disorder, continuing disability occasionally produces a confusing picture. This is further compounded by the occurrence of myasthenia gravis in this particular form in elderly males - the same group who tend to develop motor neuron disease.

**Pseudobulbar palsy**

It has been noted in earlier discussion that certain motor cranial nerve nuclei have equal bilateral upper motor neuron innervation; only the part of the facial nerve nucleus controlling the lower face shows a major variation, as it has mainly contralateral supranuclear innervation. Both the palate and tongue are sometimes visibly affected by upper motor neuron lesions, suggesting a variable pattern of supranuclear innervation. The accessory nerve is unique in having mainly ipsilateral supranuclear innervation. The significance of these variations is in the occurrence of pseudobulbar palsy. This is usually consequent on vascular disease but occasionally occurs in motor neuron disease and in the Steele-Richardson syndrome. These latter conditions produce symmetrical bilateral supranuclear degeneration. In vascular disease, a unilateral lesion will usually cause little or no dysfunction of the lower cranial nerves (Willoughby and Anderson, 1984). An upper motor neuron facial weakness and ipsilateral weakness of sternocleidomastoid and upper trapezius may be detected. A transient weakness of the palate and tongue may be detected on careful examination in the early hours following the incident. Some time later, a stroke on the opposite side will deprive the lower cranial nerves of the residual 50% of their supranuclear innervation. This will result in acute inability to speak and swallow, and is often accompanied by severe emotional liability. In stroke-related disease, these problems will always be of acute onset. In degenerative disease, such as the Steele-Richardson syndrome or motor neuron disease of the upper motor neuron type, the onset is insidious.
Extrapyramidal disease

Fine control of articulation, swallowing and the facial movements associated with speech are all achieved by extrapyramidal mechanisms.

Parkinson's disease

The loss of spontaneous facial expression and infrequent blinking constitute two of the cardinal features of this disease. In the later stages, hypophonic, tachyphemic speech is characteristic, with the short, sharp whispered phrases being virtually unintelligible. The act of chewing food is extremely laboured and the patient may seem to lack the will to initiate swallowing. If to this is added the slowness of cutting up and transporting food to the mouth, then the occasional cachectic state of terminal parkinsonian patients is easy to understand. The apparent sialorrhoea of Parkinson's disease actually represents a decreased swallowing rate with a normal production of saliva; it is not a consequence of excessive secretion.

Choreiform syndromes

Choreiform movements of the tongue, palate and mouth conspire to produce spluttering, slurred, explosive speech. This may be seen in Sydenham's chorea as a transient phenomenon, but it constitutes a severe and progressively disabling problem in Huntington's chorea.

Dyskinetic syndromes

The so-called buccal-lingual-masticatory syndrome is usually seen as a complication of prolonged neuroleptic therapy but can occur in mental subnormality and dementia. In these conditions, the movements do not seem to interfere with speech or swallowing as the movements subside while speaking and eating. They are mainly a feature when the patient is at rest.

The oromandibular syndrome (Meige's syndrome) with slow dystonic opening of the jaw and mouth in association with tongue protrusion and blepharospasm, usually occurs without neuroleptic provocation. In this condition, attempts to talk and eat, if anything, aggravate the movements.

Another possibly related dystonic syndrome is spasmodic dysphonia, a disorder characterized by choking of the voice while speaking, normally as a result of a laryngeal spasm, especially on initial vowel sounds. The patient can usually whisper, hum and sing normally. During the choking phase, spasms in the face and neck muscles, and blepharospasm may be observed (Bicknell, Greenhouse and Pesch, 1968).

Cerebellar disorders

Dysarthria is a feature of generalized cerebellar disease. It typically consists of a slurred spluttering type of dysarthria as breathing mechanisms are desynchronized with speech. There is also incoordination of tongue, palatal and facial movements. Inherited cerebellar degeneration and multiple sclerosis are the commonest causes, although in the latter
condition the disability is compounded by coexistent spastic dysarthria, producing the typical scanning dysarthria of the disease. Cerebellar neoplasms rarely seem to produce definite speech disturbances.

Conclusion

Although much clinical material has been included in this chapter to emphasize the salient features of the anatomy and physiology of the cranial nerves in the clinical situation, the coverage is by no means comprehensive. It is hoped that the clinical physiology of cranial nerve function included here will enable the reader to perform a competent clinical examination of the cranial nerves in those otolaryngological conditions in which there is a high probability of anatomical damage occurring to these structures.