Chapter 21: Acoustic tumours

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History

The fascination that acoustic neuromata have always held for the otolaryngologist originates not only from his conviction that the early diagnosis of the condition at the 'otological' stage is his prerogative, but also that their expeditious surgical removal is greatly enhanced by his traditional knowledge of temporal bone and facial nerve anatomy, and of microsurgical technique. In a sense, the internal auditory meatus is a meeting point between the territories of the neurosurgeon and the otologist, and like other such disputed zones is fraught with danger and pitfalls for the unwary. Where surgical disciplines do abut, there has tended to be a danger in the past for both to deal less than adequately with problems of management, and certainly in the case of lesions of the cerebellopontine angle, it has become increasingly apparent over the past two decades that the high ideals of early diagnosis, total tumour removal, low mortality and morbidity, preservation of the facial nerve and occasionally useful hearing, can only come about if otologist and neurosurgeon work together for the good of the patient.

It would appear that the first case of acoustic neuroma to be fully documented was that described by Sandifort of Leyden, in 1777, in an article entitled 'De duro quodam corpusculo, nerver auditorio adhaerente' in which he described the post-mortem finding of a small firm tumour of the auditory nerve, emerging from the internal auditory meatus and compressing the medulla in a patient who had complained of deafness. Sir Charles Bell, in 1830, provided one of the earliest clinical descriptions of the progressive symptomatology of a patient who was referred to him as a case of tic douloureux, and went on to develop deafness, dizziness and facial paralysis before dying of apparent brainstem compression and raised intracranial pressure. At post-mortem, a semicystic tumour, the size of a pigeon's egg, was found in the cerebellopontine angle, indenting the pons, extending into the internal meatus and involving the fifth and seventh cranial nerves. This description is of particular interest for Bell's own excellent illustration of the post-mortem specimen.

Throughout the nineteenth century there appeared an increasing number of clinicopathological descriptions of what, despite somewhat ambiguous histopathology, were certainly acoustic neuromata, (for example Cruveilhier, 1835; Toynbee, 1853; Stevens, 1879; Oppenheim, 1890) and the reader is referred to the review by Harvey Cushing (1917) for further details of this fascinating period. Ballance is usually credited with the first successful removal of an acoustic neuroma in 1892, but Cushing felt that his case was more likely to have been a meningioma and attributed the honour to a Scot, Annandale of Edinburgh in 1895 - 'a brilliant surgical result, the first recorded'. Generally, however, the mortality and morbidity of early surgical series were dauntingly high, due to late presentation, poor anaesthesia and instrumentation, haemorrhage, and above all the feeling that these tumours could be enucleated rapidly with the finger, a manoeuvre that inevitably resulted in serious bleeding from the anterior inferior cerebellar artery, the importance of which vessel was not appreciated. Indeed, it is significant that Ballance, in 1908, expressed the view that the surgical results might be improved if that artery could be ligated prior to removal of the tumour.
The first attempts at surgical removal were by way of the unilateral suboccipital approach of Krause (1903), that particular writer reporting an operative mortality of 83.8%. In a visionary article in 1904, Panse proposed that an approach through the labyrinth might allow removal of an acoustic neuroma as large as a hen's egg. He defined the anatomical limits of that exposure, the lateral sinus, the jugular bulb, the carotid artery and the temporal lobe but felt that the facial nerve must inevitably be sacrificed. However, he suggested that in certain tumours of this region the facial nerve could be re-routed after being mobilized from the geniculate ganglion to the stylomastoid foramen, thus anticipating Fisch by three-quarters of a century. The translabyrinthine approach appears to have been first employed, in 1911, by Kummel in Heidelberg (Marx, 1913) and Quix (1912) in Utrecht, but failed to find widespread acceptance, Ballance dismissing it as 'objectionable for obvious reasons'. In 1917, Harvey Cushing, in his monograph 'Tumours of the nervus acusticus and the syndrome of the cerebellopontine angle', described his bilateral suboccipital approach to the posterior fossa which allowed not only a wide decompression, but also the possibility of exploring both sides in cases in which there was doubt as to the side of the lesion. He recommended a subtotal intracapsular removal, and was able to reduce the operative mortality by 1931 to 4%, this despite the fact that the tumours were almost always very large with hydrocephalus, brainstem compression and failing vision. Dandy (1925), however, was strongly in favour of a total removal via a unilateral suboccipital approach.

The surgical results of these two great American neurosurgeons were certainly an encouraging improvement on those of their predecessors, but despite that and the invaluable contribution of Atkinson (1949) in clarifying the importance of the anterior inferior cerebellar artery, there remained a certain reluctance on the part of neurosurgeons to embark on this type of surgery unless the tumour was so large as to be causing pressure effects on the brainstem, or raised intracranial pressure. Many patients were in effect told that their tumours were too small for surgery and to return when they had grown larger! It was to this unique surgical anomaly that William House directed his attention in the early 1960s, developing first the middle fossa, and shortly afterwards the translabyrinthine approach to the internal auditory meatus. The great improvement in results achieved by his group, in terms of both mortality and morbidity, particularly to the facial nerve, is largely the consequence of the policy of early diagnosis and surgery and owes much to advances in diagnostic techniques in the fields of both radiology and audiology and to the evolution of microsurgery as an essential skill for both otologist and neurosurgeon. Rand and Kurze (1965) were among the first neurosurgeons to apply microsurgical techniques to acoustic neuroma removal via the suboccipital transmeatal route, an approach which is now enjoying renewed popularity as surgeons seek to preserve hearing in selected cases. In recent years, there have been rapid changes in the audiological and radiological assessment of acoustic neuroma suspects with the emergence of brainstem electric response audiometry, and computerized tomography (CT) and magnetic resonance (MR) promising to bring the ideal of early diagnosis close to reality.

**Anatomy**

The cerebellopontine angle is a triangular area bounded *laterally* by the medial portion of the posterior surface of the temporal bone, *medially* by the edge of the pons and *posteriorly* by the anterior surface of the cerebellar hemisphere and the flocculus, and is part of the lateral medullary cistern. *Superiorly* it is limited by the trigeminal nerve as it crosses the petrous apex and by the edge of the tentorium. Its *inferior* limit is formed by the lower cranial nerves.
(IX, X, XI) as they enter the jugular foramen, and by the hypoglossal nerve. It contains one
important artery, the anterior inferior cerebellar, and two cranial nerves, the facial and
vestibulocochlear, as they pass from their points of origin at the pontomedullary junction
towards the internal auditory meatus.

The internal meatus (Latin meatus, a coming together) is a passage through the petrous
bone leading from the posterior surface of the temporal bone to the medial wall of the
vestibule. It has a porus, or inlet, medially with a sharply defined crescentic posterior lip and
a rather poorly demarcated anterior edge, a canal proper which is roughly cylindrical and a
fundus laterally which abuts the medial wall of the vestibule. The lateral wall of the meatus
presents several features of great surgical importance. It is divided into superior and inferior
halves by the falciform crest. The upper compartment is further separated into an anterior area
for the facial nerve and a posterior area for the superior vestibular nerve, by a sharp vertical
ridge of bone known as 'Bill's bar' after William House. The lower half also comprises two
areas, anteriorly the tractus spiralis foraminosus through which the spiralling fibres of the
cochlear nerve pass, and posteriorly the rather smaller area for the inferior vestibular nerve
supplying the saccule. The singular nerve, a branch of the inferior vestibular nerve supplying
the ampulla of the posterior semicircular canal, passes through a small canal on the floor of
the meatus, about 1 mm from the fundus.

The seventh and eighth nerves leave the brainstem at the region of the pontomedullary
junction, at which point they are closely related to each other and here it is impossible to
make out the individual components of the vestibulocochlear nerve. As they pass laterally
separation between them becomes more apparent, and at the level of the porus, four individual
nerves can be identified, the facial and nervus intermedius anterosuperiorly, the superior
vestibular posterosuperiorly, the cochlear anteroinferiorly, and the inferior vestibular
posteroinferiorly. The anastomosis between the facial and vestibulocochlear nerves is referred
to in Chapter 19.

The anterior inferior cerebellar artery usually arises from the basilar artery as a single
trunk, and in the cerebellopontine angle forms a loop which has an intimate, but somewhat
variable, relationship with the facial and audiovestibular nerves and with the internal auditory
meatus; on occasion the artery may in fact loop right into the meatus. The main branches of
the anterior inferior cerebellar artery are the internal auditory and subarcuate arteries, and
these tend to tether the anterior inferior cerebellar artery to the posterior surface of the
temporal bone. It is important to realize that this is a region of considerable anatomical
variation, and for further details the reader is referred to the excellent post-mortem studies of

The most important venous channels in this region are the jugular bulb which sweeps
up below the internal meatus, but which may if high be a posterior relation of the meatus, and
the superior petrosal sinus running in the line of attachment of the tentorium to the petrous
ridge, and draining the petrosal vein anteriorly.

The relationship of the meninges to the internal meatus and its contents is of
considerable surgical importance. The dura of the posterior surface of the temporal bone is
firmly adherent round the porus where it merges with the periosteal lining of the meatus. The
pia-arachnoid, on the other hand, continues into the meatus investing the nerves in individual
or common sheaths, and blending with the neurilemma. The subarachnoid space therefore extends laterally to the fundus of the meatus.

Pathology

Site of origin

The term acoustic neuroma seems to be fixed indelibly in the English literature, and yet on semantic as well as pathological grounds this is clearly inappropriate. As Schuknecht (1974) has pointed out these tumours have, over the years, been variously described as neuromata, neurilemmomata, neurofibromata, and perineural fibroblastomata, and yet none of these terms accurately describes their histological origin. It would seem that they arise from the Schwann cells which envelop the distal portion of the eighth nerve from the point at which the neuroglial elements cease. This zone, the glial-neurilemmal junction, may be a zone of cellular instability and is thought by some authorities to be the likely site at which neoplastic change occurs. This view does not find favour with Schuknecht (1974) who stated that schwannomata may arise at any point between the glial-Schwann cell junction and the cribrose area. It is, however, clear that, because this junctional area usually lies within the internal auditory meatus, most tumours take origin within that canal, an observation first made by Henschen in 1912. In an important minority of instances the glial-neurilemmal junction is situated more medially, in the cerebellopontine angle and tumours arising here may reach considerable size before presenting, a fact which may be partly explained by Schuknecht's observation that the glial-Schwann cell junction of the cochlear nerve is situated more medially than that of the vestibular. Furthermore, it appears to be the experience of most surgeons in this field that the superior vestibular is the most common nerve of origin of these neoplasms, with only rare involvement of the cochlear (Nager, 1964). It would appear, however, that for the foreseeable future, common usage will decree that the term 'acoustic neuroma' will take precedence over vestibular schwannoma'.

The different pathological features of von Recklinghausen's disease are considered later in this chapter, together with the unique problems of management of that condition.

Pattern of growth

As the tumour grows within the internal meatus, it causes progressive but slow destruction of the vestibular nerve and produces pressure effects on the adjacent cochlear and facial nerves. Because the tumour arises in the nerve sheath, it tends to compress the neurons rather than to infiltrate them, and thus creates a possible plane of surgical dissection between tumour and nerve, although Neely (1981) and Luetje et al (1983) have drawn attention to the fact that this plane may be more apparent than real, and that histological involvement of the facial and cochlear nerves may occur. The tumour is invested in a layer of arachnoid, and as it expands, a double layer is created, which covers the whole tumour and separates it from adjacent structures (Di Tullio, Malkasian and Rand, 1978). At a fairly early stage of growth, the tumour causes an increase in cerebrospinal fluid protein, which in turn may cause some degree of arachnoiditis, often leading to the development of an arachnoid cyst in association with the tumour. The tumour may be small with a large arachnoid cyst and vice versa. Erosion of the bony walls of the internal meatus occurs, particularly at the porus; occasionally tumour can break through the roof of the meatus into a suprameatal air cell system and
continue to grow within the petrous bone, and this may make surgical identification of the facial nerve difficult.

As expansion continues in a medial direction, the cerebellopontine angle is entered, and because this is a large relatively empty space, growth proceeds quite silently. During this time, the facial nerve is becoming increasingly attenuated over the surface of the tumour, and displaced by it, usually in a sharply anterior direction at the porus, but occasionally posteriorly. The tumour also displaces the anterior inferior cerebellar artery, and develops a blood supply from it, although Perneczky (1981) maintains that most of the tumour blood supply is from meningeal vessels. Lye, Elstow and Weiss (1984) and Lye (personal communication, 1986), described an endothelial cell stimulating angiogenic factor (ESAF) which is responsible for new blood vessel formation in a variety of intracranial neoplasms, including acoustic neuromata.

The anterior inferior cerebellar artery and the facial nerve, although often considerably displaced by the tumour, remain separated from it by the double arachnoid layer referred to above. Rarely, an inframeatal loop of anterior inferior cerebellar artery may become compressed within the meatus by a relatively small tumour with resultant ischaemic effects on the cerebellum. When the tumour is about 2 cm in diameter, its upper pole makes contact with the trigeminal nerve as it crosses the petrous apex to enter the cave of Meckel, and compresses it against the pons and midbrain. The lower pole of the tumour displaces the ninth, tenth and eleventh nerves, but these seem relatively resistant to pressure and stretching. Brainstem and cerebellar involvement follow, and quite marked degrees of brainstem shift may occur, to the extent that ultimately, contralateral false localizing signs may be seen. (Cushing's bilateral suboccipital approach was developed partly because of the problems of correct lateralization.) In addition, the tumour may pass ventral to the brainstem and may ultimately reach almost to the opposite temporal bone. Because of the size of the cerebellopontine angle, the slow rate of growth of the tumour, and the capacity of the brain to tolerate quite a striking degree of shift, if slowly applied, the posterior fossa can accommodate a surprisingly large mass before serious changes in cerebrospinal fluid hydrodynamics occur, but a stage is reached when all the 'slack' in the system has been taken up, and hydrocephalus with papilloedema, and brainstem embarrassment may follow quite rapidly, especially if there is a sudden bleed into the tumour. Hydrocephalus may be either internal, from distortion of the aqueduct, or external from obstruction to flow in the lateral medullary cisterns.

Effects on the inner ear

Degenerative changes in the cellular structures of the inner ear, and biochemical alterations in the inner ear fluids secondary to the presence of an acoustic neuroma in the internal meatus have been frequently described (De Moura, Hayden and Connor, 1969a; Schuknecht, 1974), and may account for the fact that in many cases of acoustic neuroma the audiological picture may appear to be cochlear, or exhibit mixed cochlear and retrocochlear features. Suga and Lindsay (1976) postulated that cochlear changes may result from interference with the arterial blood supply of the inner ear from pressure of tumour on branches of the internal auditory artery. They pointed out, however, that the venous drainage of the inner ear is mainly by way of the canals of the cochlear and vestibular aqueducts, rather than via the internal meatus, and that venous backpressure on the cochlea from meatal
obstruction is unlikely to be responsible for inner ear changes as suggested by Brunner (1925) and Watkyn-Thomas (1939). Degeneration is more commonly seen in the cochlea than in the otolith organs or in the semicircular canals. There may be atrophy of the organ of Corti, most frequently seen in the basal turn, but occasionally widespread or complete. Vacuolization of the stria vascularis has been frequently reported, notably by Suga and Lindsay (1976), who observed that quite extensive strial damage could be associated with surprisingly good preservation of the organ of Corti, and of the endolymphatic spaces of the cochlea and vestibule, leading them to conclude that only a small amount of normal stria is necessary for the maintenance of the normal volume of endolymph in the inner ear. The other notable change reported has been in the spiral ganglion, the cells of which may be extensively or totally lost.

The occurrence of high protein levels in the endolymph was first described by Dix and Hallpike (1950) and has been the subject of many subsequent studies (Silverstein and Schuknecht, 1966). An exudate may even be seen in the perilymphatic spaces of the cochlea. Several attempts have been made to identify the protein by electrophoresis, in samples of perilymph taken at the time of translabyrinthine surgery, but the main problem appears to be in obtaining a sample free from contamination with blood. O'Connor et al (1982) were unable to identify a protein pattern specific to acoustic neuroma. Palva and Raumio (1982) carried out immunodiffusion tests using anti-cerebrospinal fluid, anti-tumour antiserum pooled from five patients with acoustic neuromata and were able to demonstrate cerebrospinal fluid and tumour specific proteins in the perilymph. They suggested that cerebrospinal fluid proteins could enter through the cochlear aqueduct, and tumour protein through small channels in the cribose area. The search to identify an immunological marker in the bloodstream has, as yet, met with little success, although Rasmussen, Thomsen and Tos (1981) were able to demonstrate cell-mediated immunity against acoustic neuroma in four out of 11 patients before surgery, as well as in one normal control (out of 16), using the leucocyte inhibition capillary tube technique. Anniko, Arndt and Noren (1981) grew acoustic neuroma cells in organ culture and demonstrated that they were highly radioresistant.

Rare instances are reported of tiny unsuspected and asymptomatic neuromata wholly confined within the cochlea or vestibule, and found by chance at post-mortem, as in the case of Johnsson and Kingsley (1981) who described a small tumour of 1.5 mm diameter within the scala tympani seeming to have originated in the distal process of the cochlear neuron. Such a tumour could well have given rise to a Ménière-like syndrome. Thomsen and Jorgensen (1973) reported a case of an intracochlear neuroma which was seen to originate in the spiral ganglion. Storrs (1974) presented two cases in which an acoustic neuroma presented as a middle ear tumour. Intracochlear neurofibromatosis is a well recognized feature of von Recklinghausen's disease (Linthicum, 1972).

**Gross appearance of the tumour**

The typical acoustic neuroma is a firm, well encapsulated tumour with a somewhat nodular surface which tends to mould itself to the contours of the cerebellopontine angle. There is a relatively well-defined plane of separation between the tumour and the arachnoid, but in places it may be rather firmly adherent to its surroundings, particularly in the proximity of branches of the anterior inferior cerebellar artery, making safe removal at times difficult or occasionally impossible. The medial pole of the tumour usually displaces the brainstem
before it, but on occasion may almost appear to infiltrate into the brainstem, and may enter the fourth ventricle through the foramen of Luschka. The interior of the tumour is usually rather softer than the capsule and, although there is considerable variation from one tumour to another and in different parts of the same tumour, the consistency generally resembles that of a grape. The cut surface is rather variegated, with grey, yellow and purplish areas. Cyst formation within the substance of the tumour is common, and in some instances these cysts may constitute the main bulk of the tumour (Hitselberger and House, 1968). They contain serous yellow or haemorrhagic fluid which may be cerebrospinal fluid. Spontaneous haemorrhage into the tumour is not uncommon and in a large tumour may cause a sudden dangerous increase in intracranial pressure. Calcification is seen occasionally, usually in quite small patches, but Thomsen, Klinken and Tos (1984) described an acoustic neuroma which was almost totally calcified.

**Histological appearance**

Microscopically the neoplastic cells show two characteristic patterns, the Antoni types A and B, thoroughly described by Antoni in 1920. In the Antoni A, or fasciculated type, there is an orderly arrangement of parallel cells with dark staining fusiform nuclei arranged in bundles or whorls separated from each other by areas of relatively acellular fibrous tissue. The term 'pallisading' is applied to describe this appearance. In the more common Antoni type B, or reticular pattern, there is a looser reticular arrangement with fewer cellular elements and a more disorderly arrangement of nuclei. Areas of degeneration may be seen, the result, according to Hitselberger and House (1968), of the tumour outgrowing its blood supply. There are also pale tumour cells containing lipid, giving a general rather foamy appearance, and responsible for the yellow colour of the tumour. This picture has been referred to by Nager (1969) as Antoni B, subgroup 1. His subgroup 2 describes an appearance in which there is a relative paucity of cells with transformation of tumour tissue into a hyaline substance. All of these histological variants may coexist in the same tumour. Malignant change in an acoustic neuroma is rare, but Schuknecht (1974) described and illustrated one case, a 9-year-old girl, in whom this did occur.

**Clinical presentation**

Acoustic neuromata are not common, but their true incidence is difficult to ascertain with any degree of accuracy. All of the frequently reported early estimates were based on post-mortem studies, usually of unsuspected cases, and these are fallacious for two important reasons. First, such a study will inevitably select an aged population, and second, they are usually not consecutive. The estimate of 2.4% by Hardy and Crowe (1936) is based on a non-consecutive series of post-mortem studies, and is therefore likely to be too high, and the famous Witmaack collection of 1720 temporal bones was collected over no less a period than 37 years (Tos and Thomsen, 1984). Perhaps the most realistic epidemiological estimates comes from Tos and Thomsen (1984), who calculated a diagnosis rate for symptomatic tumours of one per 100 000 of population per year, although these authors emphasized that this figure understated the true incidence of the condition, because of missed diagnosis. These tumours have their greatest incidence in the fourth, fifth and sixth decades, and there appears to be a slight bias towards women. Apart from the hereditary factors in von Recklinghausen's disease, little is known at present about their cause.
The evolution of the clinicopathological picture in patients with eighth nerve tumours may be considered in five stages.

1. 'Otological' stage in which the changes are confined to the audiovestibular and, to a limited extent, facial nerves. This stage includes all intrameatal lesions, and extrameatal tumours up to about 2 cm.

2. Stage of trigeminal nerve involvement, suggesting a diameter of more than 2 cm.

3. Stage of brainstem and cerebellar compression, with for example ataxia, direction changing nystagmus and long tract signs, and of involvement of the lower cranial nerves.

4. Stage of rising intracranial pressure, with failing vision, headache and vomiting.

5. Terminal stage, with severe disturbance of the vital brainstem centres, and tonsillar herniation.

There may also be another group of patients, in long-term institutional care, because of behavioural or personality changes, who, if examined with CT scanning, would be found to be harbouring large posterior fossa lesions, and who may be transformed and rehabilitated following successful surgery.

**Otological stage**

**Deafness and tinnitus**

The commonest symptoms are unilateral hearing loss and tinnitus, which occur in over 90% of patients. The deafness is usually gradual in onset and slowly progressive over a period varying from as little as a few months to 20 years or more, but averaging about 2 years (King, Gibson and Morrison, 1976). The patient may volunteer the information that his ability to discriminate speech seems disproportionately poor, especially when conversing on the telephone. In perhaps 10% of cases the hearing loss is sudden and may be profound, due presumably to a vascular accident to the cochlea. Nedzelski and Dufour (1975) estimated that 3% of sudden 'idiopathic' deafness cases turn out to be due to acoustic neuroma. The presence of a clinically silent tumour may render the cochlea more sensitive to other damaging influences, particularly acoustic trauma, and a unilateral 4 kHz dip, appearing suddenly and perhaps only transiently after a relatively brief period of noise exposure may be the first indication of the presence of an acoustic neuroma. At times there may be a fluctuating low frequency hearing loss which, if accompanied by attacks of vertigo, may lead one to suspect a diagnosis of Ménière's disease. In particular, variations in speech discrimination may be seen. The tinnitus has no particular diagnostic features, except that it is non-pulsatile, and usually commences at about the same time as, or precedes, the deafness. Occasionally one will encounter the patient with acuity which is clinically and audiometrically normal, and with no tinnitus who, nevertheless, insists that there is 'something the matter with the hearing' in
one ear. It is worth remembering that there are more subtle aspects of hearing other than those which are measurable on hearing tests, and to take such a complaint seriously.

**Imbalance**

The slowly growing tumour destroys the vestibular nerve from which it arises so gradually that the central nervous system is able to compensate for the unilateral loss of peripheral input so that severe disturbances of equilibrium are the exception. Many patients may suffer a total loss of caloric response on the affected side without ever experiencing any dysequilibrium, and others may complain of no more than slight imbalance or lightheadedness on change of head or body position, especially in the dark. A minority of patients, 30% in the series of Hitselberger and House (1968), suffer from true rotatory vertigo. Many of that group experience a prolonged episode of acute labyrinthise failure, lasting for a few days or more, to which a diagnosis of labyrinthitis or vestibular neuritis may be attached, and which probably has a vascular cause similar to the sudden loss of cochlear function alluded to previously. A small number of patients have recurrent attacks which seem identical to those of Ménière's disease, but a carefully taken history, particularly with respect to the duration, temporal pattern and associated features of the attacks should allow this diagnosis to be excluded with confidence in most instances. The author has encountered a small number of patients whose only vestibular symptom was a Tullio phenomenon brought about by the noise of traffic.

**Facial nerve involvement**

Although the facial nerve is compressed and may be considerably attenuated by the expanding tumour, obvious facial weakness is uncommon. This is because motor neurons, as elsewhere in the body, are more resistant to pressure than sensory fibres. Minor degrees of weakness not apparent to the patient may be detectable on close examination, but if a severe facial weakness occurs in association with other features of a cerebellopontine angle syndrome, the cause is more likely to be a menigioma or primary cholesteatoma than an acoustic neuroma. Facial tic is surprisingly rare, but may occur. Pain, pressure or numbness around the ear are common complaints and may be due to involvement of the sensory branch of the facial nerve. Nervus intermedius involvement is frequently manifested by altered lacrimation, the patient complaining of either a dry irritating eye, or of excessive tearing, and less commonly by alterations in the sensation of taste, with cachoguesia at times. Thomsen and Zilstorff (1975), employing a simple test of the nasolacrimal reflex, found evidence of nervus intermedius involvement in 85% of 125 patients with an acoustic neuroma, an incidence higher than that of trigeminal nerve symptoms, and concluded that apart from audiovestibular findings, a defective nasolacrimal reflex was the most significant clinical evidence of cerebellopontine angle pathology.

**Trigeminal nerve involvement**

The earliest sensory change, occurring when the tumour has reached 2-2.5 cm is nearly always in the cornea, and may result in a feeling of irritation in the eye especially if there is coexisting alteration in tear production. With further growth of the tumour, pain, tingling or numbness may be felt in any or all of the three divisions of the nerve, and occasionally typical trigeminal neuralgia may occur (Bell, 1830). There may also be altered thermal
sensation with a feeling of cold on the face or on the edge of the tongue. There is usually an interval of about 2 years between the first audiovestibular presentation and the appearance of trigeminal signs and symptoms (King, Gibson and Morrison, 1976), but in approximately 5% of patients facial numbness is the initial symptom, particularly when the tumour arises medially.

**Brainstem and cerebellar involvement**

As the tumour enlarges still further, more evidence of neurological involvement appears, with ataxia of the ipsilateral upper and lower limbs presenting as clumsiness due to dysmetria, dyssynergia and dysdiadochokinesia, and with disturbances of gait, the patient tending to lean or stagger to the side of the lesion. Intention tremor may develop and it is important to differentiate it from that of Parkinson's disease which decreases during voluntary movement. Direction changing horizontal nystagmus, vertical nystagmus and rotatory nystagmus are all evidence of involvement of the central vestibular pathways, and although often violent, this is not usually associated with severe imbalance. Clinical involvement of the lower cranial nerves is not frequent, but if present implies the presence of a large tumour. Sterkers has recorded unilateral pharyngeal pain in one case and recurrent laryngeal nerve palsy in another (Portmann et al, 1975).

**Increasing intracranial pressure**

As the intracranial pressure starts to increase, headache becomes more severe and although generalized, is usually worst in the suboccipital region and in the upper neck, and is often associated with nausea and vomiting. The patient may adopt a peculiar head posture, with the neck flexed, a manoeuvre which increases the volume of the cisterna magna by 5-10 mL. There may also be titubation, a rhythmic side-to-side or nodding movement of the head caused by extreme cerebellar distortion. Failing vision due to papilloedema may, even today, be the mode of initial presentation, the earlier otological and neurological symptoms having been ignored by the patient, or worse, by his doctor. Occasionally, however, a cerebellopontine angle lesion can reach this stage with a minimum of symptoms. A patient may present with raised intracranial pressure, no localizing signs, dementia from hydrocephalus and a tremor attributed to Parkinson's disease. Alternatively, vomiting from raised intracranial pressure may be regarded as being of gastrointestinal origin (King, Gibson and Morrison, 1976).

**Terminal stage**

The terminal events in the history are related to failure of the vital centres in the brainstem.

**Examination**

General examination may reveal the presence of cutaneous lesions suggestive of von Recklinghausen's disease, multiple neurofibromata and café-au-lait blemishes. Minor manifestations may not be apparent in the fully clad outpatient, but he should be questioned about the presence of such lesions and if necessary treated to a full examination.
Ears

The tympanic membranes will be expected to be normal in most cases, but they must nevertheless be examined. Chronic middle ear disease can coexist with an acoustic neuroma and even if it is inactive, may present problems to the surgeon by limiting his access to the internal meatus through a sclerotic and acellular petrous bone. Furthermore, there are other causes of a cerebellopontine angle syndrome apart from an acoustic neuroma, and evidence of primary cholesteatoma or glomus jugulare tumour may be apparent on otoscopy. Tuning fork tests will usually confirm a unilateral sensorineural hearing loss.

Cranial nerves

These merit the closest scrutiny, in particular the fifth and seventh.

Trigeminal nerve

All three sensory divisions of the trigeminal nerve should be tested for pin-prick and fine touch, not forgetting to include the tongue, and bearing in mind the fact that the cutaneous branches of the cervical plexus extend up over the angle of the mandible. The most important area for sensory loss is the cornea, which is usually the first to be involved by an expanding lesion in the cerebellopontine angle. The reflex is elicited by stroking the cornea lightly with a wisp of cotton wool, remembering first to remove any contact lenses. Motor function is only rarely impaired, but can be checked by asking the patient to clench the teeth.

Facial nerve

Testing of the facial nerve requires some care. Severe facial weakness is uncommon unless the tumour is very large. All that may be apparent is a slight impairment in the patient's ability to bury the eye-lashes on the affected side when screwing the eyes up tightly. Minor degrees of weakness are more likely to be seen during involuntary movement of the face. Throughout the interview, the examiner should be observing the patient's face and may notice the occasional slight delay in the blink on one side. This may be confirmed by testing the blink reflex by means of a well regulated tap on the forehead with the finger. The cutaneous branch of the facial nerve may be tested by touching the skin of the posterosuperior aspect of the external auditory meatus with the tip of a needle. Loss of sensation (Hitselberger's sign) may occur while the tumour is still confined within the internal meatus, but not all clinicians find this a very reliable test (Portmann et al, 1975). Function of the nervus intermedius is evaluated by testing for lacrimation and taste on the anterior two-thirds of the tongue. Lacrimation may be assessed by carrying out Schirmer's test, in which short strips of filter paper are hooked over the lower eyelid for half a minute. This is a useful test, but there may be a slight theoretical criticism in those patients who have reduced corneal sensation and thus an unequal stimulus to tear production.

A more accurate though more time consuming test may be that of the nasolacrimal reflex as described by Thomsen and Zilstorff (1975). This involves blowing a stream of saturated benzene fumes, 500 mL/minute, into the nostril for 30 seconds directed towards the olfactory area, with Schirmer's paper in the eye. The paper is left in place for a further 30 seconds, before removal, and the lacrimation measured in millimetres. Unfortunately both
sides cannot be tested at the same time, and an interval of 10 minutes is recommended between tests. A difference of 20% between the two sides is considered significant. An elevation in the taste threshold on the anterior two-thirds of the tongue is best measured by electrogustometry, a difference of more than 20 µA between the two sides being considered significant (Pulec and house, 1964).

The other general visceral function served by the nervus intermedius is saliva production in the submandibular gland. There is a submandibular salivary flow test (Magielski and Blatt, 1958), but it is rarely used.

Examination of palatal and pharyngeal sensation and mobility is important, and abnormalities of either may indicate pressure on the glossopharyngeal or vagus nerves by the lower pole of a large tumour.

**Eyes**

The eye is a most fruitful source of information to the neuro-otologist who should be as conversant with the use of the ophthalmoscope as with the otoscope. As stated previously, there are still a surprising number of patients who first present to hospital at the stage of increasing intracranial pressure, and failing vision, and the otologist should be able to recognize not only florid papilloedema, but also the earlier changes of venous congestion and loss of venous pulsation.

Nystagmus is a very common and important sign, its pattern changing at different stages in the growth of the tumour, thus providing useful information about its size. When the tumour is small, fine first degree vestibular nystagmus to the contralateral side may be observed, particularly if optic fixation is abolished by using Frenzel's glasses. As the mass enlarges it comes into contact with the brainstem, producing rather complex changes in the central vestibular connections. Dix and Hallpike (1966) suggested that as the mass made contact with the ipsilateral vestibular nucleus, there was an increase in this fine contralateral nystagmus. Later as the cerebellar connections are involved, the nystagmus becomes direction changing, that is beating to the right on rightward gaze and beating to the left on leftward gaze. The nystagmus to the contralateral side remains fine, rapid and of low amplitude, whereas the nystagmus to the side of the lesion is more coarse and of higher amplitude. The former remains enhanceable when visual fixation is inhibited, whereas this is not true of the latter. The term Brun's nystagmus is often applied to this pattern of direction changing nystagmus. When cerebellar involvement is even more marked, other patterns such as vertical nystagmus, rotatory nystagmus and rebound nystagmus may be seen (Hood, Kayan and Leech, 1973).

The other reflex eye movement that may be disturbed by a large mass in the posterior fossa is the smooth pursuit reflex (Nedzelski, 1983). This is a low velocity tracking movement that allows the eyes to follow accurately an oscillating target such as a finger at frequencies up to about 1-1.5 Hz. At greater frequencies, the eyes cannot follow the target smoothly and the previously clean sinusoidal movement becomes contaminated with small rapid saccadic jumps. 'Saccadic pursuit' is also seen at normal frequencies if there is interruption of the brainstem pathways that subserve it. The reflex can be rapidly assessed by asking the patient...
to follow with his eyes the examiner's finger as it moves slowly from side to side, or it can be recorded graphically using electronystagmography.

**Investigation and diagnosis**

The aim of investigation is to diagnose acoustic nerve tumours while they are at the otological stage, and if possible while they are still confined within the internal auditory meatus. This is an ideal which we are still some way from achieving, although considerable advances have been made over the last 30 years. It is, however, more important today than at any time in the past, that early diagnosis is made, because of the evolution of the surgical techniques to enable safe total removal with preservation of the facial nerve in the majority of instances, and of the cochlear nerve in a small number. The surgical nihilism embodied in the writing of Pennybacker and Cairns in 1950 is a thing of the past: 'While in general we welcome early diagnosis, it is sometimes possible to make the diagnosis long before there is any indication for operation'.

There are two prerequisites: a high index of suspicion on the part of the otologist; and a programme of investigations, both audiovestibular and radiological, which can be relied on to be both sensitive and specific. In effect, every case of unilateral sensorineural hearing loss, or tinnitus, and every case of unilateral vestibular hypofunction, should be investigated to exclude the diagnosis of an acoustic neuroma. Since acoustic neuromata represent only a small minority of cases of unilateral sensorineural deafness, logistic and economic factors may start to become important, as highly sophisticated but expensive imaging techniques such as CT and MR have become available. These imaging techniques have become so refined, that in a society freed from financial constraints, all suspects would proceed straight from pure-tone audiometry to MR. In some parts of the world, that state of affairs may not be far away. More usually, however, suspects are selected for CT or MR through an investigative filter of audiovestibular tests and 'conventional' X-rays, and the exact strategy that is followed again depends upon the facilities available in individual centres.

**Audiovestibular investigation**

Traditional audiometric techniques evolved throughout the 1960s and 1970s as it was discovered that there were differences in certain functions of the ear and hearing depending upon whether the lesion was situated in the cochlea or the auditory nerve. The tests developed during that time are still, in various combinations, the mainstay of initial investigation, although one or two, such as Békésy audiometry and the short increment sensitivity index (SISI) test, are employed less frequently now than perhaps 10 years ago.

The functions traditionally studied are:

1. pure-tone threshold
2. speech discrimination
3. loudness recruitment
4. auditory adaptation.
Pure-tone threshold

There is no characteristic curve for the pure-tone audiogram. Many patients have a high frequency loss, others a flat loss, some a mid-frequency notch; a salutary proportion have a low-tone loss similar to that in Ménière's disease, and a small but disconcerting number may be normal. A total or subtotal hearing loss is present in 16% which makes the assessment of other functions impossible (Johnson, 1968).

Speech discrimination

The cochlear nerve does not require a large population of intact neurons to transmit relatively simple pure tone messages. Speech, however basic, demands a disproportionately greater number of healthy neurons, capable of coping with the complex coding involved, particularly of temporal patterns. For this reason the typical finding in the patient with a neural lesion is of a speech discrimination score which is much worse than one would expect from consideration of the pure-tone threshold, and worse than in a patient with the same degree of cochlear deafness (Schuknecht and Woellner, 1955). Hood and Poole (1971) studied a series of patients with Ménière's disease and defined the broad limits within which the speech audiograms of subjects with cochlear deafness might be expected to fall. By comparing the results from a series of patients with acoustic neuroma with these limits, they found that most of the tumour results fell outside the limits, but that unfortunately, there was some overlap and some of the neural lesions did have ‘cochlear’ speech patterns. There have been many modifications of the technique from different centres, and there is a wide variation in the percentage of positive retrocochlear results from different series. Sheehy and Inzer (1976) found that speech audiometry yielded no positive results in a series of 24 patients, Hirsch and Anderson (1980a) found it of value in 45% of patients, whereas in the series of King, Gibson and Morrison (1976), 78% of tumour cases gave retrocochlear results. Even these authors warned against placing too much reliance on the test. Rudge (1983) pointed out that one of the problems of speech audiometry is the lack of standardization of technique. Taylor (personal communication, 1986) has recently re-emphasized the fact that the score a subject may achieve on speech audiometry is very dependent upon the time and care that the audiometrician applies to the test.

Loudness recruitment

This phenomenon was first described by Fowler in 1936, but was thought by him to have a neural basis. It was Dix, Hallpike and Hood (1948) who opened the door of neuro- otological diagnosis when they reported to the Royal Society of Medicine in London their finding that total recruitment, as assessed by Fowler's alternate binaural loudness test (ABLB) occurred in every one out of 30 patients with Ménière's disease, and was absent in 14 out of 20 cases of neural pathology (in other six cases it was present but incomplete). For the first time, a simply performed test was available which allowed differentiation of neural from cochlear lesions. This observation has, for most investigators, stood the test of time, despite the assertion of Jerger and Jerger (1974) that the concept behind it had in a sense retarded subsequent progress. Hirsch and Anderson (1980a), however, found it to be positive in 64% of acoustic neuromata, Thomsen and Terkildsen (1975) in 77% of all tumours, and King, Gibson and Morrison (1976) in 90% of large tumours. These authors pointed out that the phenomenon of ‘decrement’ may be seen in some cases of acoustic neuroma, that is the
sensation of loudness grows more slowly in the affected ear than the normal ear. The fact that full recruitment, a supposed end-organ phenomenon, may be seen in a number of cases of acoustic neuroma was said by Dix and Hood (1953), writing some years after their original observation, to be 'connected with hair cell changes resulting from occlusion of the cochlear blood supply'. There is again no doubt that good test technique influences the accuracy of the results (Simmons and Dixon, 1966). It is suggested that the procedure is, if possible, carried out at more than one frequency; not uncommonly a hearing loss that appears to be recruiting or even over-recruiting at one frequency will be found not to be so at another, even if separated by as little as 250 Hz.

Other tests of recruitment include Reger's monaural balance test, and the loudness discomfort level (LDL). Both tests were evolved to try to differentiate cochlear from retrocochlear lesions in situations in which the contralateral ear was not normal for comparison, but neither is commonly performed today. Another test which for years was a standard part of the audiological battery, but which has fallen from favour, is the short increment sensitivity index test.

Auditory adaptation

This term describes the observation that a sound presented to the ear at a level just greater than threshold, will become inaudible after a short period of time, the length of which has a predictable value in normal ears. In ears with cochlear deafness, the values are similar to those in normal subjects, but with neural pathology, the speed of this adaptation is classically greatly increased. This phenomenon forms the basis of Carhart's tone decay test (1957). Hirsch and Anderson (1980a) found that it gave positive results in 53% of patients, King, Gibson and Morrison (1976) in 80%, and Johnson (1977) in only 40%. The main criticism of the test is the very high number of 'false positive' results, that is patients with cochlear lesions who have 'retrocochlear' tone decay.

The other great diagnostic tool of the 1960s and 1970s was the Békésy self-recording audiometer, which compared threshold values for pulsed and continues tones. Patterns or response were identified for cochlear, retrocochlear and non-organic hearing loss (Jerger, 1960). In addition to providing information about threshold, the test identified the presence of abnormal adaptation, and also, in the view of some workers, evidence of recruitment. The problem was, as Jerger himself pointed out (Jerger and Jerger, 1974), that both the Békésy and Carhart tests studied phenomena close to the auditory threshold, and it became clear that more information was to be gained from suprathreshold studies, particularly those of the stapedius reflex.

It is apparent from the preceding account that no single test had the reliability or even uniformity from centre to centre to allow a confident diagnosis of acoustic neuroma, and even if employed en masse the tests were still capable of missing an appreciable number of cases. It is remarkable how few proven cases of retrocochlear pathology in any series have a full complement of retrocochlear features on every audiological test. Accuracy of audiological diagnosis has increased dramatically however with advent of two more recent techniques: stapedius reflex studies and electric response audiometry.
Stapedius reflex measurements

There are two parameters of the stapedius reflex which have proved of great value in the identification of retrocochlear pathology, the reflex threshold itself, and its rate of decay. Although the activity of the middle ear muscles has been studied since the last century, it was Metz (1946, 1952) who first studied their role in clinical cases. The use of the test became widespread when the original mechanoacoustic bridges were superseded by the simpler electroacoustic equipment. Following the observations of Anderson, Barr and Wedenburg (1970), many studies have confirmed that in the presence of retrocochlear pathology, the stapedius reflex threshold is elevated above normal levels, whereas in cochlear deafness the threshold is usually normal. They defined significant elevation as 95 dB HL at 250, 500, 1000, 2000 and 3000 Hz, and 100 dB HL at 1500 Hz and stated that for the threshold to be abnormal it must be significantly raised at four out of the six test frequencies (250 Hz - 3 kHz). Subsequent work has suggested that these thresholds are too low and result in a large number of 'false positives'. Chiveralls et al (1976) introduced a concept of 'reflex asymmetry', and proposed that a difference in the reflex threshold between the two ears of more than 15 dB should be regarded as abnormal.

Not infrequently the reflexes are absent at some or all frequencies; in many instances this is presumably because the threshold is beyond the maximum output of the audiometer. Hirsch and Anderson (1980b) found that in the majority of cases of acoustic neuroma, the elevation of the reflex threshold was greater in the higher than the lower frequencies. They also emphasized that the test should be carried out using contralateral recording. There is a possibility that absence of a reflex on ipsilateral recording could be due to interference with the efferent limb by pressure on the facial nerve from a lesion in the cerebellopontine angle. Gibson (1981) reported significant elevation of the stapedius reflex threshold in 75% of small and medium tumours and in 90% of large tumours, and this is fairly typical of the general experience.

Stapedius reflex decay is the decline in amplitude of the reflex on prolonged stimulation, and in individuals with neural pathology the rate at which the decay occurs is increased. Pathological decay is judged to be present if the response amplitude declines by more than 50% in 5 seconds at 500 Hz and at 1 kHz. At higher test frequencies, decay of this magnitude may be normal. The cause of pathological decay is unclear. Rudge (1983) suggested that impaired conduction in the cochlear nerve may result from demyelination, impaired blood supply or some abnormality of axoplasmic flow. It may well be related to the fact that demyelinated fibres cannot sustain a train of electrical impulses. It is clear, however, that the phenomenon arises in the eighth nerve and not the seventh, because it occurs on both ipsilateral and contralateral recording. Hirsch and Anderson (1980b) regarded abnormal stapedius reflex decay as a more specifically retrocochlear finding than elevation of the threshold. They considered the interrelation between the stapedius reflex threshold and decay abnormalities and concluded that an elevated stapedius reflex threshold with normal stapedius reflex decay was a relatively poor indicator of the presence of a lesion in the cerebellopontine angle, whereas abnormal stapedius reflex decay with or even without elevation of the stapedius reflex threshold was highly significant. These writers concluded that the probability of encountering normal stapedius reflex characteristics in a case of a tumour of the cerebellopontine angle seems very slight.
The value of the combined test of stapedius reflex threshold and decay has been confirmed by many subsequent studies (King, Gibson and Morrison, 1976; Chiveralls, 1977), and it has taken its place as one of the simplest and most reliable techniques available to the clinician. One shortcoming of the test, as Flood et al (1984) pointed out, is that the acoustic reflex will be abolished even by a cochlear hearing loss of greater than 60-75 dB, and the coexistence of middle ear disease renders the test impossible. One further parameter of the reflex which has received some attention is its latency. Clemis and Sarno (1980) felt that in patients with an eighth nerve disorder, there was significant delay in the onset of the acoustic reflex compared with the normal contralateral side, but this was not confirmed by Jerger and Hayes (1983), who felt that the concept of latency measurement should be abandoned.

Electric response audiometry

The advent of electric response audiometry has been one of the most important events in the history of audiological medicine and neuro-otology. In the investigation of patients with unilateral sensorineural deafness, two tests are of particular importance; electrocochleography and brainstem electric response audiometry. The main role of electrocochleography in neuro-otological diagnosis seems to be in the identification of endolymphatic hydrops (Gibson, Moffat and Ramsden, 1977), but brainstem electric response audiometry has emerged as the single most reliable audiological indicator of retrocochlear pathology, and has gone far towards superseding traditional psychoacoustic tests. The detailed test procedure has been described elsewhere (Volume 2). In brief, a sound wave entering the cochlea is transduced into an electric potential which is transmitted via the eighth nerve to the brainstem, where it passes through a series of relay stations on the way to the higher auditory centres. This process is associated with a sequence of electric phenomena, which can be detected by scalp electrodes and after a process of 'time-domain averaging', displayed on an oscilloscope screen. Within the first 7 milliseconds following acoustic stimulation, a series of five negative deflections appears. Their sites of origin are thought to be as follows:

\[
\begin{align*}
N_I &\quad \text{cochlear nerve} \\
N_{II} &\quad \text{cochlear nucleus} \\
N_{III} &\quad \text{superior olivary complex} \\
N_{IV} &\quad \text{lateral lemniscus} \\
N_V &\quad \text{inferior colliculus}. \\
\end{align*}
\]

In normal individuals, the latency of these responses is very predictable and reproducible, not only from person to person, but from test to test in the same person. The early identification of these potentials was largely the result of the work of Jewett, Romano and Williston (1970), while Selters and Brackmann (1977) were the first to explore the possible application of the technique to the detection of lesions of the eighth nerve. They argued that any delay in electrical transmission in the nerve, caused for example by a tumour, would be passed on to all subsequent points in the auditory chain, and would be detectable in latency delays in wave V, which by virtue of its magnitude has proved the most convenient for study. They found the interaural latency difference of wave V to be superior to the absolute latency of wave V for the detection of acoustic tumours. They used the terms 'T_v' to identify the latency of wave V, and 'IT_v' for the interaural difference, and regarded the upper limit of normal for IT_v to be 0.2 ms and correctly identified 91% of tumours using this criterion. Terkildsen and Thomsen (1983) preferred an IT_v of 0.3 ms.
Apart from delay in wave V, two other abnormalities may be seen in certain tumour cases. In some patients, there may be no recognizable response despite adequate levels of hearing, and this may be due to a loss of the synchrony of neural discharge necessary to produce an identifiable waveform. The other effect sometimes seen is a slight delay in the contralateral wave V in patients with large tumours causing distortion of the brainstem and of the contralateral auditory pathways.

The main disadvantage of the test is that it can only be relied upon to produce consistent waveforms at hearing levels better than 75 dB. At higher levels, therefore, there will be an increasing number of 'false positive' results. However, the chances of a subject with a normal brainstem electric response having an acoustic neuroma are very slight, certainly less than 5%, and this test is now well established as the single most reliable audiological screening test for the condition (Selters and Brackmann, 1977; Glasscock et al, 1979; Terkildsen and Thomsen, 1983).

Gibson and Beagley (1976) and Morrison, Gibson and Beagley (1976) have reported on the electrocochleographic abnormalities in cases of acoustic neuroma, and described three typical findings - broadening of the eighth nerve action potential, good preservation of the cochlear microphonic, and preservation of the action potential at stimulus intensities that are inaudible to the patient. Unfortunately, as these authors pointed out, the broadening of the action potential waveform is not pathognomonic of retrocochlear disease and a very similar waveform is commonly seen in Ménière's disease and other hydropic disorders. In addition, cochlear microphonic measurements are notoriously variable and hard to standardize. Because of these objections, and the simplicity and reliability of brainstem electric response audiometry, electrocochleography has not gained widespread acceptance in the assessment of retrocochlear pathology.

**Caloric testing**

The traditional bithermal caloric test of Hallpike and Fitzgerald remains an invaluable aid to diagnosis, with a significant canal paresis in well over 90% of patients with an acoustic neuroma, although King, Gibson and Morrison (1976) found that in smaller tumours of less than 2 cm diameter, the test gave normal responses in 20% of instances. It is a fair observation that the pattern of abnormality does not allow one to differentiate between acoustic neuroma and other causes of unilateral vestibular hypofunction such as Ménière's disease. What is highly suggestive, however, is the finding of an absent or considerably reduced response without any dramatic history of vertigo, and this should always increase the clinician's suspicions. The end point of caloric-induced nystagmus may be more obvious if Frenzel's glasses or electronystagmographic techniques are employed.

**Radiological investigation**

Although high quality plain X-rays of the temporal bones, particularly the transorbital view, can reveal pathological changes in the internal auditory canals in a large number of instances, there is no doubt that the proportion of abnormalities detected is increased by the use of tomography (Harner and Reese, 1984). Valvassori (1969), by studying anatomical specimens as well as tomograms, established the limits of normal variation of certain parameters of internal meatus anatomy, and defined three important measurements: vertical
diameter of the internal canal - in over 90% of normal individuals the difference in the vertical diameter between the two sides measured at the same point did not exceed 1 mm; length of the posterior wall of the canal - in over 90% of normal individuals the difference between the two sides did not exceed 2 mm; crista falciformis was always located at or above the midpoint of the vertical diameter of the internal canal. In 90% of normal individuals the difference in position on the two sides, measured from the crista to the superior and inferior walls did not exceed 1 mm. The canal is considered abnormal when there is enlargement of 2 mm or more of any portion, compared with the opposite side, when there is shortening of the posterior wall by at least 3 mm compared with the normal side, and when there is displacement of the crista by at least 2 mm compared with the normal side. Using these criteria the diagnosis of acoustic neuroma was definite in 78% of cases and suggestive in a further 13%. When studying tomograms of the internal auditory meatus, it is important to appreciate that there are variations in the normal canal. Thomsen et al (1981) found that in 70% of normal canals, the shape of the internal meatus was straight, that is the upper and lower walls were parallel to each other, in 14% it was narrow medially, in 14% it was oval and in 2% narrow laterally.

Computerized tomography has proved the greatest radiological advance of recent years in the detection of acoustic neuromata, and as each new generation of scanner emerges, with software programmes that allow coronal and three-dimensional reconstructions, so ever smaller lesions can be diagnosed with confidence. Whereas with the early models, tumours smaller than about 2 cm could not be demonstrated, the current situation is that in most instances an extension of 0.5 cm into the posterior fossa can be readily demonstrated. It is, of course, essential to administer an intravenous contrast agent which is taken up by the vasculature of the tumour and enhances the image. In the event of a negative result, a small volume of intrathecal air may be introduced, and this may demonstrate not only a small intrameatal tumour, but also detail of the related neurovascular structures. Computerized tomography also provides the surgeon with information about the state of the ventricular system and any shift in the brainstem that may have occurred. The technique has largely replaced the traditional air encephalography and myodil meatography, but angiography still has a place in occasional instances, if doubt exists as to the true nature of the lesion in question.

Magnetic resonance

Where available, MR is now an established alternative to CT in the detection of posterior fossa lesions (Jenkins and Isherwood, 1986). The theoretical basis of the technique is discussed in Volume 1. The advantages over conventional imaging methods are a high intrinsic contrast between tissues, an absence of bone artefacts, an ability to image directly in the coronal, sagittal and transverse planes, and an avoidance of ionizing radiation. In the specific case of an acoustic neuroma, there is a high contrast between neural structures, the surrounding temporal bone and cerebrospinal fluid. Large tumours can be more readily identified than with contrast enhanced CT scanning (Kingsley et al, 1985), and in particular the intracanalicular extent and the associated brainstem distortion are more distinctly demonstrated. Small tumours totally confined within the meatus may be seen very clearly, because of the contrast between tumour and bone, and their detection may be improved by the use of contrast enhancement using gadolinium-diethylenetriamin-penta-acetic acid (Gd-
DTPA) (Curati et al, 1986). At the present time, the higher costs of MR compared with CT precludes its routine use, but its future potential seems enormous.

**Diagnostic screening for acoustic neuromata**

As pointed out earlier in this chapter, logistic and economic factors decree that there must be some sort of screening strategy that selects those patients for CT or MR that are most likely to yield positive results, because in the final analysis, the preoperative diagnosis of acoustic neuroma must be made on the basis of positive radiological findings. However suggestive they may be, audiovestibular tests are no more than a means of allowing the clinician to decide which patients to investigate further, and are not in themselves an indication for surgery. There is no single test short of a full CT scan, with or without contrast, or MR which will allow the condition to be diagnosed. It is important that the conventional audiological tests should be carried out with skill and care, and in many otolaryngology departments throughout the world they will be the only options available to the clinician. Ideally, however, the most efficient routine screening programme is, in the experience of the author, a combination of three tests:

1. brainstem electric response audiometry
2. tomography of the internal auditory meatus, applying the criteria of Valvassori (1969)
3. bithermal caloric test of Hallpike and Fitzgerald.

This test battery effectively eliminates all 'false negative' results. In other words, if a patient with a suspected tumour produces normal results from all three tests, his chances of harbouring an acoustic neuroma are virtually nil. Terkildsen and Thomsen (1983) using a very similar strategy suggested that if two or more of these tests gave positive results the patient should be submitted to CT scanning, with air-cisternography if necessary. If only one test gave a positive result, they recommend re-screening after one year. Using this approach, they claimed not to have missed any cases of acoustic neuroma. Mafee et al (1985) also recommended the use of brainstem electric response audiometry and caloric tests in the selection of patients for CT scanning.

**Differential diagnosis**

Acoustic neuroma is by far the most common lesion of the cerebellopontine angle. A meningioma is the next most frequently encountered condition in this region. The majority of posterior fossa meningiomas arise on the posterior surface of the temporal bone and may be differentiated from an acoustic neuroma on CT scanning by the fact that they may not be centred on the internal auditory meatus, and indeed may not be associated with expansion. Furthermore, they are usually clearly evident on the unenhanced scan, whereas an acoustic neuroma requires intravenous contrast for clear visualization. Meningiomata may be seen in association with acoustic neuromata in von Recklinghausen's disease.

Next in the order of frequency are primary cholesteatomata arising from congenital epithelial remnants within the temporal bone. These typically present with facial weakness as
an early sign, and have a characteristic appearance on petrous tomography with a widespread area of destruction bounded by a scalloped edge (Fisch, 1978). Arachnoid cysts of the posterior fossa may occur in the cerebellopontine angle. The usual hypotheses to explain their development include congenital malformation, infection (adhesive arachnoiditis), trauma, increased intraventricular pressure and embryonic rests (Little, Gomez and MacCarty, 1973). They are also commonly described in association with acoustic neuromata, indeed they may be a very large cyst associated with quite a small tumour. The cysts are characteristically thin walled and appear to develop between the layers of the arachnoid. They contain clear fluid identical to cerebrospinal fluid. Vascular causes of a cerebellopontine angle syndrome include basilar artery aneurysm or ectasia (Gibson and Wallace, 1975), von Hippel-Lindau syndrome, or compression of the eighth nerve by a loop of the anterior inferior cerebellar artery. Other neoplastic causes include pontine glioma, in which there are progressive brainstem and cerebellar signs without obvious temporal bone changes, cerebellopontine angle lipoma, which is rare and very difficult to remove totally (Rosenbloom et al, 1985; Pensak et al, 1986), and a secondary deposit.

The commonest condition to be distinguished from acoustic neuroma is Ménière's disease. An acoustic neuroma does not, however, usually cause the typical intermittent severe vertigo of Ménière's disease; it usually produces a retrocochlear picture on conventional audiometry, is not associated with a positive response to glycerol dehydration, and is characterized by unique brainstem electric response audiometry and tomographic changes. Nevertheless, the occasional tumour case presents with a history typical of Ménière's disease, and with a fluctuating low frequency hearing loss with good speech discrimination, full recruitment, normal tone decay and normal stapedius reflex functions. It is desirable for the vigilant clinician to subject all such cases to brainstem electric response audiometry, petrous tomography and caloric testing.

**Surgical management**

The transformation in the prognosis in acoustic neuroma surgery over the last 25 years, is largely as a result of the universal adoption of the operating microscope by both otologists and neurosurgeons. Mortality has dropped to 3% or less, and functional preservation of the facial nerve has proved possible in the great majority of cases (Lye et al, 1982). Furthermore, on a limited number of occasions, it may be feasible to conserve useful hearing. There is no doubt that better results are achieved with small tumours than large. Morrison and King (1982) reported no mortality for small tumours, rising to 2% when the tumours were large. As regards the facial nerve, the same authors reported functional preservation in 100% of small, in 80% of medium-sized, but in only 20% of large tumours. In nearly every instance, therefore, it is desirable to remove an acoustic neuroma at the earliest possible opportunity.

There are certain exceptions to this rule. A small tumour in an aged patient or in one who is a poor anaesthetic risk can be monitored by check CT scanning at annual or biannual intervals. There is also a good case for withholding surgery as long as possible in the patient with an acoustic neuroma in his only hearing ear; this situation is most commonly encountered on the second side of patients with bilateral tumours. In the great majority of cases, however, surgical removal should be total, but in the elderly patient with a large tumour it is often sufficient to carry out a partial or intracapsular removal thus shortening the operation and minimizing the risk of damage to the brainstem or facial nerve. The residual
tumour can then be observed at regular intervals by scanning. Silverstein and Norrell (1982) who have particular experience of this problem, by virtue of the popularity of Florida with the elderly, reported the case of an 83-year-old woman who had a radical intracapsular resection of a large tumour, and was alive and well 10 years later with no apparent increase in the size of the residual mass. There are also occasions when the surgeon has to terminate the operation on the advice of the anaesthetist before total removal has been achieved, usually because of changes in the vital signs occurring when attempting to remove the last portion of capsule from the brainstem. In that event, it is usually possible to complete the removal at a second stage operation.

There are several different surgical approaches, and over the years there have been many modifications and combinations of approaches devised. The main requirement of any approach is access, which must be sufficient to ensure total tumour removal, and to allow the surgeon control over any possible bleeding in the posterior fossa, but with a minimum of trauma to the brainstem and cerebellum. The second requirement is a reliable means of identifying the facial nerve early in the operation, preferably within the internal meatus. There are basically three routes to the cerebellopontine angle - the middle fossa, translabyrinthine and posterior fossa approaches - each with its advantages and disadvantages.

The middle fossa approach offers the possibility of total removal with preservation of hearing of small tumours confined to the meatus. Most of the dissection is extradural, and facial identification at the lateral end of the meatus is unequivocal. The main disadvantage of this approach is the limited access to the posterior fossa, which not only limits the size of tumour that can be removed but also restricts the ability of the surgeon to deal with bleeding in the posterior fossa either at surgery or postoperatively. A bleed from the anterior inferior cerebellar artery would be very difficult to control from the middle fossa. The other disadvantage is that the facial nerve, although easily recognized may be traumatized during tumour removal because it lies between the surgeon and the tumour. The risk of temporal lobe epilepsy from prolonged retraction, although often quoted appears slight.

The translabyrinthine approach developed by William House is the most direct route to the cerebellopontine angle, and allows early and reliable identification of the facial nerve at the lateral end of the internal meatus. Cerebellar retraction is minimal, and as a result the patients tend to have a speedier postoperative recovery. A further important advantage is that in the event of postoperative haemorrhage, the postauricular incision can be rapidly re-opened, the fat packing removed and the situation immediately rectified (House, 1977). Critics of the approach claim that the access is cramped and that it is difficult to remove a large tumour through it, and that, furthermore, the control of posterior fossa bleeding is less secure than with the retrosigmoid approach. One self-evident objection to the technique is the inevitable loss of residual hearing, but Brackmann (1982) pointed out that so far as is known, no single patient out of 1400 operated on for unilateral acoustic neuroma by the House group developed a serious loss of hearing in the opposite ear.

The posterior fossa or retrosigmoid or suboccipital transmeatal approach is the traditional neurosurgical procedure (Di Tullio, Malkasian and Rand, 1978), evolved by Dandy from the original Krause operation. It is the preferred route in cases where preservation of hearing is the aim (Smith, 1982). The good exposure it affords certainly makes it very suitable for the removal of the larger tumours, but it is also favoured by many for the removal
of all acoustic neuromata regardless of size (Welch and Dawes, 1985). The disadvantages include the necessity for cerebellar retraction, although the most skilled proponents of the technique are able to minimize this; there is now never any need to resect part of the cerebellum. Perhaps the greatest criticism (Brackmann, 1982) is that it is difficult and, in some cases impossible, to visualize the lateral end of the internal meatus without opening the labyrinth, so there is the risk that a small fragment of tumour could be left in that site.

Other modifications of approach that have been employed at various times, and which may still have an application include the translabyrinthine - transtentorial operation of Morrison and King (1973), the combined suboccipital-petrosal approach (trans-sigmoid approach) described by House, in which the standard translabyrinthine dissection is extended posteriorly over the divided sigmoid sinus to include that portion of the skull removed in a suboccipital approach, and the retrolabyrinthine approach, which allows partial removal through the mastoid with preservation of the labyrinth.

The questions as to choice of technique and whether acoustic neuromata should be removed by otologists or neurosurgeons have occasioned much, often vitriolic, debate over the years. The real answer is that there is more than one way to achieve the same goal. Ultimately, the most important consideration is, who does the job best in a particular area? In the best hands, there is very little to choose between the results of translabyrinthine and posterior fossa removal. In the most successful series, there is close collaboration between otologist and neurosurgeon, each of whom brings his own expertise to the team, and in many centres each partner learns the skills of the other so that both are totally familiar with all steps in the operation. This then allows the surgeons to change over during the operation and overcomes the problems of fatigue (and hunger). Familiarity with all approaches is desirable and confers a degree of flexibility to the team.

Apart from the operating microscope, other technical advances are constantly appearing, all of which, in a small way, help the surgeon to achieve better results. These include the cavitron ultrasonic surgical aspirator (CUSA) (Epstein, 1983), which reduces the operating time particularly with large tumours, and the CO₂ or argon laser (Glasscock, Jackson and Whitaker, 1981; Smith, 1982). Moller and Jannetta (1984a) have refined the technique of peroperative monitoring of the electromyographic responses from the facial muscles so that an acoustic warning is given when the facial nerve is stimulated. Similarly, the same authors (Moller and Jannetta, 1984b) and Ojemann et al (1984), attempting to preserve hearing, carried out peroperative recording of the electrocochleographic and brainstem electric response from transtympanic and scalp electrodes and found that changes in these potentials provided the surgeon with an early warning of possible damage to the hearing.

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A full description of the anaesthetic considerations involved in the management of these cases is inappropriate in this chapter. Suffice it to say that the services of a skilled and experienced neuroanaesthetist are of paramount importance.
Surgical techniques

Middle fossa approach

The incisions begins in front of the ear at the level of the zygomatic arch and curves gently upwards and backwards to the superior temporal line. The temporalis muscle may be divided in a linear manner to expose the squamous temporal bone, and a 4-cm square craniectomy cut, the lower edge of which is on a level with the upper surface of the petrous bone. Approximately two-thirds of the bone flap should be in front of the external meatus and one-third behind it. Alternatively, the flap may be left attached to a pedicle of temporalis muscle and turned inferiorly. The dura is then gently elevated first to the arcuate eminence, and then anteriorly and medially until the superior petrosal sinus is reached and the middle meningeal artery is exposed as it enters the skull through the foramen spinosum.

The greater superficial petrosal nerve is found about 1 cm behind and slightly lateral to the artery and, using a diamond burr, is followed to the geniculate ganglion which may, in fact, be immediately under the dura without any bony covering. The facial nerve is then traced backwards and medially to the meatus, passing deeply between the cochlea and the superior semicircular canal, neither of which should be opened. As it leaves the meatus, the facial nerve is separated from the superior vestibular nerve by a very obvious vertical crest of bone - 'Bill's bar'. The meatus is exposed from its lateral to medial ends through as wide a bony trough as possible and the dura is incised along the posterior wall of the meatus, that is away from the facial nerve. After positive identification, the tumour is carefully dissected off the facial nerve, taking care to minimize the manipulation of both it and the cochlear nerve which is to some extent protected by the facial nerve. Both superior and inferior vestibular nerves must be totally ablated as the tumour is removed as otherwise there is a risk of postoperative imbalance.

If hearing is to be conserved, care must be taken to avoid damage to the internal auditory artery. Division of the superior petrosal sinus may occasionally be necessary to facilitate removal of a tumour extending into the posterior fossa. After careful haemostasis, the meatus is sealed using fascia or muscle, the middle fossa dura is allowed to sink back over the defect, and the wound is closed in layers, replacing the bone.

Translabyrinthine approach

The incision is an extension of the standard postauricular wound, the upper end reaching as far as the line of the anterior wall of the external meatus, and the lower limit being a point about 2 cm behind the tip of the mastoid. A superiorly-based periosteal flap is preserved in continuity with the lower edge of the temporalis muscle for use in eventual wound closure. An extended mastoidectomy is carried out, removing the bone over the lateral sinus, middle fossa dura and superior petrosal sinus until it is 'eggshell-thin'. The facial nerve is identified at the second genu and its vertical portion is skeletonized. A total labyrinthectomy is then performed taking particular care to avoid trauma to the facial nerve as the inferior crus of the posterior canal is followed into the vestibule, a point at which the drill will be immediately medial to the nerve. The subarcuate artery is encountered under the superior semicircular canal and after labyrinthectomy is the only feature in a dense triangular wall of bone separating the surgeon from the internal meatus. Bleeding from the artery is a
The meatus is now skeletonized superiorly, posteriorly and inferiorly by careful removal of more bone during the process of which the endolymphatic duct and sac will be seen and destroyed. Above the meatus, dissection may be greatly facilitated if there is a well-developed air cell system.

The anatomical limit above the meatus is the middle fossa dura with the superior petrosal sinus. Posteriorly, bone is removed from a wedge-shaped area between the internal meatus and the posterior fossa and, eventually, when the porus is reached, continuity between the meatus and the posterior fossa can be demonstrated. Inferior to the meatus the limiting factor for bone removal is the jugular bulb, and the other structure encountered in this region is the cochlear aqueduct. Bone is finally picked off the dura of the posterior fossa and the internal meatus.

The facial nerve may now be identified at the lateral end of the meatus separated by 'Bill's bar' from the superior vestibular fibres which may be seen entering the vestibule. The intrameatal portion of the tumour can then be separated from the facial nerve. The posterior fossa is then opened by cutting a laterally based U-shaped dural flap, the main bulk of the tumour is exposed, and an intracapsular removal performed, using the House-Urban rotary dissector, or the CUSA. The remaining capsule is then removed by careful dissection in the plane between the tumour and the arachnoid taking particular care to avoid damage to the branches of the anterior inferior cerebellar artery and the lower cranial nerves which are in the arachnoid layer and to the brainstem and trigeminal nerve. The previously identified facial nerve is traced medially. At the porus, it turns sharply forwards and may become very thin and hard to follow but, by careful adherence to the arachnoid plane, it is usually possible to preserve it. If the nerve is known to have been sacrificed, an immediate repair should be effected using a graft from the cervical plexus or sural nerve with re-routing of the horizontal portion of the facial nerve into the meatus if necessary (Barrs, Brackmann and Hitselberger, 1984; Samii, 1984). After tumour removal is complete, the middle ear should be packed with muscle, and the cavity with 1 cm strips of abdominal fat, held in place with the periosteal flap. These measures minimize the risks of a cerebrospinal fluid fistula.

Suboccipital transmeatal approach

The patient is positioned in the reclining lateral 'park bench' position and an S-shaped retromastoid incision is made from the level of the upper edge of the pinna to the spine of C2, taking care to avoid the vertebral artery. After separation of the muscular attachments, the craniectomy is carried out, the limits of which are the transverse sinus superiorly, the foramen magnum inferiorly and the sigmoid sinus laterally. It is important that the occipital bone be removed as far laterally as possible so that the surgeon's line of vision is along the back of the petrous bone, thus minimizing the amount of cerebellar retraction necessary. If the tumour is very large it may be necessary to remove the arch of the atlas. The dura is opened through a triradiate incision and cerebellar retractors gently introduced protecting the underlying cerebellum with patties. The intracranial segment of the tumour is debulked intracapsularly, as described above, taking care to identify and protect the lower nerves, and the trigeminal nerve, as well as the anterior inferior cerebellar artery. A laterally based dural flap is raised over the internal meatus, and the posterior wall of the meatus carefully drilled off, exposing the intrameatal portion of tumour which protects the facial and cochlear nerves in the anterior half of the meatus. If hearing preservation is intended, the labyrinth must not

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be entered, but if this is unimportant there is no doubt that extending the dissection as far laterally as possible increases the chances of total tumour removal. The transverse crest and 'Bill's bar' are identified and the tumour gently separated from the facial nerve using Malis or Yasargil's dissectors. The operation is completed in the manner outlined above, and before closing the wound the internal meatus should be sealed with a muscle plug, and the mastoid air cells inspected for any possible cerebrospinal fluid leak which should be sealed with bone wax.

**Postoperative care**

The patient is returned from theatre to an intensive care unit for regular neurological observations and cardiac monitoring. A nasogastric tube is inserted to minimize the risk of aspiration, particularly if there has been trauma to the lower cranial nerves, and the patient should be catheterized. There will be an intravenous line, but over-hydration must be avoided. The patient will be on broad-spectrum antibiotics, and possibly steroids if there has been any threat of brain swelling. The cornea should be protected until it is obvious that facial function is satisfactory. As a temporary measure a simple silk stitch in the upper lid taped on to the cheek will suffice, but if the weakness is marked, and the Bell's phenomenon poor, and particularly if there is coexisting corneal anaesthesia, a formal lateral tarsorrhaphy should be performed. Tracheostomy may occasionally be necessary if aspiration and swallowing disturbances are a problem. Cerebrospinal fluid fistula is a dangerous condition which cannot be allowed to persist. It may settle spontaneously or with repeated lumbar punctures, but if in doubt, it is advisable to return the patient to theatre for repair.

As recovery proceeds, the patient is mobilized quickly, and is usually ready to go home by about the tenth postoperative day. Occasionally, in the case of a large tumour, postoperative hydrocephalus may be seen and this may require treatment by daily lumbar puncture or possibly ventriculoperitoneal shunting. Imbalance from loss of vestibular function is not usually a major problem because, in most instances, vestibular function has already been considerably reduced prior to surgery. The great majority of patients are fully rehabilitated to their previous levels of activity, and should be able to play golf, jog, cycle, and sail normally. Tinnitus is a surprisingly rare long-term complaint. One of the least appreciated problem areas is the eye in patients who have suffered partial or total facial weakness. Not only may the loss of the protective blink reflex expose the eye to the risk of foreign bodies, but there may be subtle changes in the physical properties of the tear film with resultant pain, grittiness, blurring, dryness or watering. If there is coexisting loss of corneal sensation, neurotrophic changes may occur with the risk of corneal ulceration. It is a clinical problem which demands close ophthalmological attention and possible tarsorrhaphy.

One of the most difficult problems concerns the non-recovering facial nerve. If the nerve is known to have been severed immediate repair is indicated if possible. If it is not possible, an immediate hypoglosso-facial anastomosis should be carried out. The problem arises when the nerve is thought to be partially preserved but no recovery ensues over a long period of observation. Recovery from a degenerated nerve will usually be apparent by one year after surgery. If no recovery is visible at 2 years, a hypoglosso-facial anastomosis should be carried out.
**Hearing preservation**

In a minority of patients with small tumours it is possible to preserve useful hearing by carrying out a middle fossa or posterior fossa removal. While this may seem a laudable objective, the facts must be looked at very critically. First, except in generalized neurofibromatosis the disease is almost invariably unilateral, so that most patients have normal or good hearing in the contralateral ear. Second, emphasis must be placed on the concept of 'useful' hearing, minimum criteria for which should be a 50 dB pure-tone threshold, and 50% speech discrimination score. Only a minority of patients will satisfy these criteria preoperatively, and certainly any postoperative result worse than these levels will not be appreciated by the patient. Furthermore, there is the problem of how results of surgery are reported. If a centre reports '40% hearing preservation' this does not mean that 40% of all patients with acoustic neuromata retain useful hearing, but that 40% of those patients in the series in whom preservation of hearing has been attempted, have retained hearing; this may be nearer 4% of the series as a whole. On the other hand, one has to remember that the risks of leaving a tumour fragment in the lateral end of the meatus are increased if hearing preservation is attempted. In considering an attempt at hearing preservation, the surgeon should always ask himself whether the object of the exercise is the good of the patient or his own self-satisfaction!

**Bilateral lesions**

Bilateral acoustic tumours pose a particularly challenging problem to the otologist (Hughes et al, 1982). They differ in many important respects from unilateral lesions, and merit special consideration. Bilateral tumours may occur as a part of a central form of von Recklinghausen's disease, the commoner characteristics of which are generalized cutaneous neurofibromata, café-au-lait spots, skeletal abnormalities, mental retardation and certain intracranial tumours, of which the most frequently encountered is acoustic neuroma; others include neurofibromata on other cranial nerves, meningiomata, gliomata and ganglioneuromata. von Recklinghausen's disease is an hereditary autosomal dominant condition, with a positive family history in only 50% of cases, suggesting either incomplete penetrance, or a high incidence of spontaneous mutation. On the other hand, a high percentage of bilateral tumour cases occurs in patients without evidence of von Recklinghausen's disease, and some authorities would regard it as a different genetic disorder, notably Moyes (1968), who described a series of 14 members of four generations of one family, all with bilateral tumours, but with scanty or absent evidence of von Recklinghausen's disease. Morrison (personal communication, 1986), however, stated that a careful examination of all unilateral acoustic neuroma patients will reveal some signs of von Recklinghausen's disease in about 10%.

The clinical picture differs from the unilateral lesion. The patients are usually younger, often in their teens or early twenties, with bilateral, symmetrical or asymmetrical hearing loss. The hearing is often remarkably well preserved, even in the presence of very large tumours, and there may be a cochlear picture on conventional audiological testing (Linthicum, 1972), but brainstem electric response audiometry will nearly always reveal bilateral wave V delay. Bilateral loss of vestibular function may lead to ataxia which is worse in the dark, rather than true vertigo, and caloric tests indicate hypofunction on both sides. There may be neurological abnormalities associated with other intracranial or spinal tumours, or with hydrocephalus.
Plain or tomographic radiographs may show bilateral widening of the internal auditory canals, but they may be normal even in the presence of a large tumour if it arises medially in the cerebellopontine angle. Computerized tomography in addition to confirming the presence of bilateral eighth nerve tumours, may show up other cranial nerve lesions.

Pathology

There has been much debate as to the true histological nature of the tumour in von Recklinghausen's disease, the House group maintaining that it is a neurofibroma and not a schwannoma (Linthicum and Brackmann, 1980). This distinction is of some importance in considering the effect on the cochlear nerve. A schwannoma will tend to compress the nerve, causing deafness, whereas a neurofibroma infiltrates the nerve, spreading its fibres apart without compressing them, thus conserving good hearing. De Moura, Hayden and Conner (1969b) on the other hand stated that it is impossible to differentiate between these two groups of tumours by light microscopy. The other interesting feature in von Recklinghausen's disease is the growth of tumour into the cochlea, so-called 'intracochlear neurofibromatosis', which may in part explain the cochlear audiological picture often seen. The rate of tumour growth may differ from unilateral tumours. Whereas many bilateral lesions are very large on presentation, there is no doubt that, in some instances, the growth rate seems very slow indeed, allowing a conservative approach to management.

Surgical management

The surgeon caring for these unfortunate patients has a difficult task, his decisions taking account of many factors - tumour size, level of hearing, age of the patient, and neurosurgical considerations. If one tumour is large and associated with poor hearing, and the other is small and associated with good hearing, then clearly total removal of the large tumour is the first step. What, however, is the correct management of the second side? One school would argue that early surgery should be carried out when the tumour is still small and there is some chance of preserving the hearing (Hughes et al, 1982), whereas others would preserve the second tumour (and with it the hearing) until such times as rising intracranial pressure necessitated surgery, and then carry out a subtotal removal in the hope of preserving some useful hearing (Morrison, 1975). The surgeon's view as to the feasibility of preserving hearing is clearly influenced by whether he believes the lesion to be a schwannoma, in which case there ought to be a plane of dissection between the cochlear nerve and the tumour, or a neurofibroma, in which case the nerve is in theory unsavable. It appears from reported cases of total tumour removal with hearing preservation, that in some instances, at least, there cannot have been infiltration of the cochlear nerve by tumour. The difficulties of management of bilateral eighth nerve lesions are well reviewed by Hughes et al (1982).