**T. T. King, W. P. R. Gibson and A. W. Morrison: Tumours of the VIII Nerve**

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Tumours of the VIII nerve are not common, comprising only about 8 per cent of any large neurosurgical series of intracranial tumours, yet have always excited the interest of neurosurgeons to an almost disproportionate degree. Their benign nature (apparent in postmortem specimens), their striking mode of evolution, and the characteristic nature of their clinical picture have no doubt contributed to this.

Early attempts at total removal were attended by a mortality rate as high as 75 per cent (Tooth, 1913). Over a number of years, a standard technique for dealing with these tumours was evolved, from subtotal removal and decompression (Cushing, 1917) to total removal after gutting of the tumour (Dandy, 1925) with gradually improving results. In the last decade new operative approaches and new technical aids, particularly the operating microscope, have contributed to an improvement in the results, but there is no doubt that the early otological diagnosis of small tumours has been the most significant single advance in the treatment of this difficult lesion.

## Pathology

The majority of eight nerve tumours are sporadic in occurrence and unilateral. They are one and a half times more common in women than in men, and occur over a wide age-span with the greatest incidence in the fourth and fifth decades.

Bilateral cases, whether familial or not, are often associated with multiple meningiomas and are probably always a manifestation of von Recklinghausen's disease, although the histology of bilateral tumours is not exceptional. The number of central manifestations of von Recklinghausen's disease (that is spinal root and cranial nerve tumours) tends to vary inversely with the quantity of peripheral and cutaneous signs although eight nerve tumours do sometimes occur in the presence of florid cutaneous lesions. It has been said that familial bilateral eight nerve tumours, which have been known to affect as many as 14 members of one family in four generations, are different from von Recklinghausen's disease, but this is not an established fact.

The Schwann cell sheath from which the tumour is derived does not commence until the nerve has reached the porus acusticus internus, and hence it is to be expected that the tumour would arise within the internal acoustic meatus. Surgical experience of small tumours, which always completely fill the meatus, supports this view and systematic postmortem studies leading to the finding of microscopic tumours are also in accord. It is possible, however, that some tumours do arise a little more medially, which might explain the atypical clinical course sometimes seen.

The nerve of origin is nearly always the superior vestibular but occasionally it is the inferior vestibular or cochlear. The rate of growth of the tumour is not known for certain but the great variety in duration of the deafness in large tumours (usually in the order of 2 years but

sometimes as long as 15 years or more) suggests that different tumours grow at different rates. Morrison (1975) has suggested an average annual growth rate of 0.5 cm.

As the tumour expands in the internal acoustic meatus it obliterates its subarachnoid space and dilates its lumen, even when still confined to this area. In time the tumour extends into the cerebellopontine angle through the porus acusticus. Hullay (1958) asserted that the tumour may expand extradurally, that is within the petrous bone, but there must be some doubt as to whether the intrapetrous neurinomas really are eight nerve tumours. They may have arisen from the facial nerve or within the cochlea itself. Extracisternal expansion is also described, but the great majority of eight nerve tumours are characteristically intracisternal, that is to say they lie within the subarachnoid space and on reaching the brainstem are immediately applied to the pia mater without an intervening layer of arachnoid membrane.

The exact direction in which maximal tumour extension occurs seems to vary from case to case but the first point of contact with the pons is almost always very close to the origin of the trigeminal nerve, which is progressively stretched over the upper and medial pole with expansion of the tumour. The upper pole reaches the tentorium and the petrosal vein and may pass through the tentorial notch. Medially, the pons is indented and rotated towards the opposite side, while backward growth results in indentation of the middle cerebellar peduncle and cerebellum. Inferiorly, the ninth, 10th, and 11th nerves are progressively stretched around the lower pole which may reach the foramen magnum. The subarachnoid space on the lateral side of the tumour may be sequestrated and converted into a cyst containing yellow proteinaceous fluid.

The tumour may reach almost 3 cm in diameter and produce a considerable dent in the pons with no clinical signs other than vestibular and cochlear. Tumours presenting with neurological disabilities are usually 3 cm or more in diameter. In the remainder of this discussion small tumours are those confined to the internal acoustic meatus, medium-sized tumours are those up to 3 cm in diameter and unassociated with clinical signs other than vestibular and cochlear, and large tumours are greater than 3 cm in diameter and associated with some clinical combination of cranial nerve and brainstem involvement, raised intracranial pressue, and a positive brainscan.

## Clinical Features

In the great majority of cases the first symptoms are auditory and vestibular. Although deafness may be associated with tinnitus the patient may take little notice of it. Even with careful otological screening there wil always be some patients who present with large tumours because following the appearance of eight nerve symptoms there is a period, variable in length but usually about 2 years, during which the tumour increases in size without adding to the patient's symptoms. When further symptoms do appear, the tumour is quite large and in contact with the brainstem. Neurological rather than otological signs always mean a fairly large tumour.

Transitory unsteadiness on sudden movement of the head is the common vestibular symptom. The incidence of true vertigo is between 25 per cent and 66 per cent. The order in which other neurological symptoms and signs develop was said by Cushing (1917) to be characteristic, namely suboccipital discomfort, incoordination and instability of cerebellar origin, evidence of impairment of adjacent cranial nerves, raised intracranial pressure, and finally dysarthria, dysphasia, and decerbrate attacks with respiratory failure terminating the illness.

Although the sequence makes diagnosis very easy, it is by no means constant the presenting picture may be unusual. For example if deafness is slight and unrecognized the patient may present with raised intracranial pressure and no localizing signs, dementia from hydrocephalus, or vomiting regarded as being of gastrointestinal origin. It seems that suboccipital discomfort is not very common (although complaints of pain and numbness around the ear may be elicited with careful questioning), and the trigeminal involvement is the most common symptom to follow deafness.

## Trigeminal Nerve

In about 5 per cent of cases facial numbnessis the inaugural symptom, the patient having failed to notice any previous loss of hearing. With large tumours, trigeminal symptoms are present in about 60 per cent of cases and trigeminal signs in 90 per cent. Trigeminal neuralgia is occasionally seen as an early symptom.

## Facial Nerve

This is always closely applied to the tumour over a distance dependent upon the size of the mass, but clinically severe facial weakness is very much unusual and total paralysis is only occasionally seen. Most commonly a mild weakness is manifested only by ipsilateral delay in blinking and normal function is usual even with large tumours. Hemifacial spasm has been reported on occasions.

Disturbance of taste from involvement of the nervus intermedius is best determined by electrogustometry, a difference of over 20 microA between the two sides of the tongue anteriorly being considered significant. Alteration in lacrimation on the affected side may be measured with a strip of litmus paper hooked under the lower lid for one minute, when the length of paper dampened is a measure of tear secretion (the Schirmer test).

## Other Cranial Nerves

Diplopia, when present, is usually due to a sixth nerve palsy. The ninth, 10th, and 11th nerves are not often clinically involved although they are frequently stretched around the lower pole of the tumour. Hypoglossal involvement is a rarity.

## Cerebellar and Brainstem Signs

These are common in large tumours and consist principally of a disturbance of gait, varying from slight to so severe as to render the patient incapable of walking. Ataxia in the upper limbs is not often a complaint, nor is it well developed on examination. Persistent nystagmus is a reliable indication of distortion of the brainstem and can be taken as denoting a large tumour, even if the patient has no signs other than deafness. It is present in over 90 per cent of large tumours, central in character, usually horizontal, and slower and coarser on looking to the side of the lesion. Bulbar signs and decerebrate spasms are such late symptoms that they are rarely seen in ordinary practice nowadays.

## Raised Intracranial Pressure

The frequency of this finding is a direct index of the lateness of diagnosis. Papilloedema may lead to blindness and in Cushing's (1917) series all 30 patients had it: two were blind, six almost so, and only five had normal vision. In a series of 60 cases treated at this hospital the incidence of papilloedema in large tumours was only 38 per cent, and the overall incidence in tumours of all sizes 14 per cent against an incidence of about 70 per cent in large neurosurgical series.

## CSF Protein Measurement

The CSF protein is usually elevated above 100 mg/100 mL in large tumours but is normal in intracanalicular growths and also in a considerable number of tumours of intermediate size.

## Audiological Examinations

Although audiometric tests have a most important role in early diagnosis no single test alone is dianostic. The majority of tumours are atypical in showing deafness of a non-recruiting nature, tone decay, poor speech discrimination, and characteristic findings on electrocohleography. Some cases show few of these audiological signs and almost every patient does have some atypical features. Only by considering the results from a number of audiological test can a tentative diagnosis be reached and even then radiographic evidence is required before surgery can be undertaken in small and medium-sized tumours.

## Pure Tone Threshold Audiometry

It is usually as a result of this test that the diagnosis is first suspected. Comparison of the air-conduction and the bone-conduction thresholds reveals a sensorineural deafness. The shape of the audiogram is variable. Sometimes the majority of patients is so deaf that no measurements are possible. Audiometric curve may have a downward slope, flat curve, ascending and even U-shaped. The majority have a downward sloping curve.

## Speech Audiometry

Classically, speech discrimination with eight nerve tumours is far worse than would be predicted from the pure tone audiogram. The patient often complains that he is unable to understand a telephone conversation with the affected ear. Some workers wrongly consider poor speech discrimination to be the most diagnostic of audiometric tests.

## Absence of Recruitment

The loudness recruitment phenomenon is a usual aspect of cochlear pathology whereby the sensation of loudness grows faster in the deafened ear than in the normal ear. There are several different audiological tests currently in use which demonstrate this phenomenon. These include:

1. The alternate binaural loudness balance test of Fowler which compares the hearing for pure tones between the two ears at varying intensities.

2. The monaural loudness balance test of Reger which compares the hearing of two separate tone frequencies within the same ear.

3. The short increment sensitivity index (SISI) which measures the patient's ability to detect minute increases of intensity at levels 20 dB above his hearing threshold.

4. The loudness discomfort test which measures the stimulus intensity that becomes so loud as to be uncomfortable for the patient. Some authorities prefer to use a "most comfortable loudness test" which gives similar results.

In these and like audiometric tests patients with sensory deafness almost invariably show recruitment while normal subjects and those with conductive hearing loss do not. Ninety per cent of patients with large eight nerve tumours show no recruitment and some exhibit "decruitment" when the sensation of loudness grows more slowly in the affected ear. Absence of recruitment alone is not diagnostic of a neural lesion since the presence of a conductive element must first be excluded. Some 10 per cent of large tumour cases show recruitment and the percentage rises with smaller tumours.

## Tone Decay

Eight nerve tumours classically exhibit temporary threshold drift or tone decay. This means that the patient's hearing for tones deteriorates with time and that sound appears to fade and become less intense. The most commonly used test is Carhart's which measures in decibels the intensity by which a pure tone signal has to be raised to remain audible. Over 20 dB decay in under 3 minutes is suggestive of a neural lesion. A small amount of tone decay is common in purely cochlear forms of deafness, particularly at higher frequencies, and this overlaps with the level of tone decay seen in the less typical tumour cases.

## Békésy Audiometry

In this test, the signal frequency is swept automatically from low to high while the intensity of the signal is controlled by the patient so that it remains just audible. Interrupted (pulsed) and continuous tones are used, and the intensity as determined by the patient is recorded automatically on a graph against the frequency. The results of this test indicate a mixture of the hearing threshold, recruitment, and tone decay. According to the amount of tone decay present, the continuous tracing either falls abruptly away from the pulsed tracing or runs below it. Jerger (1960) analysed a large number of these tracings and found that the former finding was almost pathognomonic of a neural lesion. When the continuous tracing runs below the pulsed tracing throughout the recording it indicates some tone decay even at low frequencies, and this finding is more common in neural than sensory lesions. In our series 24 per cent of the patients with tumours failed to show either of these characteristic traces.

Fixed frequency Békésy recordings provide a sophisticated and accurate method of measuring tone decay at various frequencies.

## Acoustic Reflex Measurements

Provided that the middle ear mechanism and the facial nerves have normal function, it is possible to record the movement of the intratympanic muscles in response to acoustic stimulation. In man only the stapedius muscle responds to sounds, unless they are of sufficient intensity to cause a startle reaction in which case the tensor tympani muscle may also contract. This reflex provides the most reliable audiometric screening test for the detection of eight nerve tumours.

1. *Acoustic reflex threshold measurements.* In a normally hearing subject the stapedius muscle contracts when a pure tone is delivered to either ear at approximately 75 dB above the subject's hearing threshold for that sound. This difference is reduced in patients with recruiting forms of deafness so that, for example, a patient with a cochlear hearing loss of 60 dB may yield an acoustic reflex at 95 dB - a gap of only 35 dB. Patients with eight nerve tumours rarely produce the reflex at less than 75 dB above their hearing threshold and even moderate deafness may render the reflex unobtainable at the maximal output of the audiometer (120-125 dB). Since testing for the acoustic reflex is comparatively quick and simple, it provides a very handy objective method of screening patients with sensorineural deafness for neural lesions. In a series of 60 patients with known tumours tested at this hospital 94 per cent exhibited this phenomenon. False positives, however, are common especially if middle ear pathology is not properly excluded.

2. *Acoustic reflex decay measurements.* In normal subjects and patients with cochlear types of deafness the stapedius muscle remains contracted throughout long durations of acoustic stimulation. No relaxation of the muscle is normally detected when 500 Hz and 1000 Hz tones are used over 15-second intervals although higher frequencies may produce some decay after a few seconds. Anderson found rapid decay at 500 Hz and 1000 Hz and the reflex contraction was

generally halved within the first 5 seconds. The best way to demonstrate decay is to use an XY plotter but since this is such an absolute test it is clinically quite sufficient to measure the time taken for the needle on the balance meter to decay to half its reading.

Standard test criteria are required.

It appears that when the acoustic reflex is present and does not decay an eight nerve tumour is unlikely.

## Electrocochleography

This test involves measurement, usually by an averaging computer, of the electrical output of the cochlea. The largest potentials are obtained by piercing the tympanic membrane so that the point of the needle electrode lies close to the round window. The shape of the action potential was altered in 80-89 per cent of cases in one series. Using stimulus intensities that were below the subjective hearing threshold in 40 per cent of patients it was possible to note preservation of end-organ potentials. In general, large clear microphonic potentials were seen. These findings suggest that the cochlear function is more or less preserved and the cause of the deafness lies more medially along the auditory tract.

## Vestibular Investigations

The Hallpike-Fitzgerald method is most commonly used. In our series, all the large tumours have shown absence or diminution of the caloric reaction on the affected side with only one exception. The results of the test are less predictable in smaller tumours and 20 per cent of out patients with tumours under 2 cm in diameter have had an equal duration of nystagmus with each ear.

## Electro-Oculography (Electronystagmography)

This technique depends on the finding that the eye acts as a dipole and a small deflection of the eyeball results in a change of the electrical axis. This change may be recorded by surface electrodes placed around the orbit. Nystagmus due to vestibular (end-organ) lesions may be suppressed by optic fixation. Use of Frenzel's glasses is often sufficient to reveal the latent nystagmus but electro-oculography has the added advantage that nystagmus may be recorded even in total darkness or with the eyelids closed. In cases of eight nerve tumour, the recordings vary in a complex manner according to whether the effect of the tumour is directed peripherally or more centrally. In 55 per cent of patients with small and medium-sized tumours latent nystagmus has been demonstrated by these techniques.

## Radiological Investigations

### Plain X-Rays

These provide the most valuable screening tests for eight nerve tumours. Enlargement of the internal acoustic meatus was demonstrated in 84.6 per cent of Olivecrona's large series (1967) and our own material shows a similar figurs. It is not, however, universally present. It is best shown by linear or hypocycloidal tomography. On the anteroposterior views abnormality of the meatus is suggested by erosion of the wall, widening of over 2 mm compared with the diameter of the corresponding point in the opposite meatus, and shortening of the posterior wall by more than 3 mm (Valvassori, 1969). Some asymmetry of the meatuses may be normal but marked trumpeting of the medial end suggests a large tumour and erosion of the petrous apex always indicates that the tumour is of considerable size.

### Pneumoencephalography

When this is combined with tomography in the posteroanterior direction, small tumours protruding into the cerebellopontine angle can be well demonstrated and the extent of large tumours can be seen if the air passes over their surface. Distortion of the fourth ventricle always indicates a large tumour. There are some risks attached to pneumoencephalography in large tumours, although in our experience it is safe if the patient does not have papilloedema. It does not demonstrate intracanalicular tumours.

### Positive-Contrast Posterior Fossa Cisternography

In this procedure iophendylate (Myodil) is injected into the lumbar sac and screened into the cerebellopontine angle. If sizeable amounts (6-8 mL) are used, large tumours can be demonstrated but not so well as with air. The main use of the technique is to demonstrate non-filling of the meatus owing to an intracanalicular tumour and this may be done with a small quantity of Myodil (1.5 mL). The extent to which the tumour protrudes beyond the internal acoustic meatus may be underestimated with this technique which usually needs to be combined with pneumoencephalography.

### Vertebral Arteriography

This is popular in some centres because it allows investigation of large tumours without disturbance of the intracranial hydrodynamics. However, it is of no value in diagnosing small tumours and, in our opinion, is inferior to pneumoencephalography with tomography for showing large tumours.

### EMI-Scanning

The place of this in the diagnosis of eight nerve tumours is not yet clear. It seems certain to displace all other methods in the diagnosis of large tumours but the minimum size below

which tumours will be missed is unknown.

## Diagnosis

Early diagnosis can only be made by an otologist but late diagnosis is easy in most cases, although it may be difficult in atypical ones. Since the problems of early and late diagnosis are somewhat different they are considered separately.

### Early

Here the patient has only deafness and the problem is how far to carry the investigations. The demonstration of an enlarged internal acoustic meatus in the presence of deafness, even if the neuro-otological studies are uncharacteristic, is good evidence for proceeding to contrast studies. Suggestive otological tests in the presence of radiologically normal internal acoustic meatus are a more difficult matter and one has to decide on either early radiological investigation, or observation and repeated testing with emphasis on the former.

### Late

If the diagnosis is clinically clear, and especially if it is backed by X-ray evidence, operation is indicated and contrast radiological studies are unnecessary. The advent of isotope scanning and EMI-scanning has reinforced confidence in this approach. Atypical cases may not be easy to diagnose and special radiological investigations may be necessary. It is worth emphasizing that quite large tumours may have no symptoms other than deafness and may be ENT referrals. In this group the brainscan has been particularly useful in warning of the true extent of the tumour.

## Treatment

An eight nerve tumour is a benign progressive lesion that advances at a variable rate but leads ultimately to severe symptoms, notably headaches, progressive loss of vision, dementia, and ataxia of such a degree that the patient is finally bed-ridden and dies under rather distressing circumstances.

Total removal of large tumours, however, has been an operation attended even in the best hands by an appreciable mortality rate in the order of 10-15 per cent and by the risk of severe postoperative disabilities, notably ataxia and multiple cranial nerve palsies, so that in less skilled hands the results have at times been alarming. Block and Nathanson (1963) reported a mortality rate of 31 per cent with 42 per cent of the survivors being totally disabled. The difficulties of the operation are due to the intimate relationship of the tumour with the brainstem and its blood system, either of which may be damaged by removal of the adjacent portion of the lesion. Even before the era of neuro-otological diagnosis many of these patients had few symptoms other than deafness - perhaps only a little numbness of the face and slight unsteadiness of gait on turning the head. The hazards of the operation contrasted strongly with the mildness of the clinical

picture at the time of diagnosis, and even with good surgical results the facial nerve was usually destroyed.

These two facts produced two differing responses. Pennybacker and Cairns (1950) recommended that surgery should only be undertaken when forced by the patient's clinical condition. However, many other surgeon's continued to carry out total removal, while complaining that their bad results were due to the lateness of the diagnosis and the size of the tumours they were offered. Early diagnosis has in some respects accentuated this problem, for in small and medium-sized tumours the operation is now virtually prophylactic and it is absolutely essential to have extremely low mortality and morbidity.

Although adding to the responsibilities of the surgeon, early diagnosis has greatly reduced the risks of the operation and made preservation of the facial nerve with good postoperative function a reasonable aim of treatment.

It has been argued that intracanalicular tumours should not be operated on at all, since they may never become big enough to cause trouble. While this may be the case, intracanalicular tumours have not been shown to be a peculiar species that does not grow, and if one has been diagnosed the only method so far of keeping an eye on the patient has been to repeat the Myodil studies at intervals - an unsatisfactory course. To wait until the patient develops aby symptoms other than deafness is to delay until the tumour is of substantial size. It is possible that the EMI-scanner will allow a waiting policy since it offers the possibility of noting increase in size without submitting the patient to further contrast studies. Otherwise, surgery seems to be the reasonable course to adopt, since mortality and morbidity in this group are very low.

## Standard Neurosurgical Techniques for Large Tumours

These all involve a posterior fossa craniotomy, usually unilateral, and exposure of the tumour by retraction or resection of the cerebellar hemisphere.

**Partial versus total removal.** Partial removal was introduced by Cushing (1917) in reaction to the catastrophic mortality of early attempts at total removal. It enables the facial nerve to be preserved and avoids the dangerous part of total removal, that is the dissection of the tumour from the brainstem. Although prolonged survival can result there is a high risk of recurrence within a few years, and several authors reviewing Cushing's and Olivercrona's series were not impressed by the quality of life after this operation. It should be reserved for old or infirm patients who are judged unfit for a prolonged procedure.

**Total removal and facial nerve preservation.** Neurosurgeons have made total removal with low mortality their principal aim and accepted that removal of the intrameatal portion of the tumour usually involves destruction of the facial nerve. Several authors have succeeded in preserving the facial nerve in a proportion of cases, usually by opening the internal acoustic meatus with a drill and identifying the nerve at that point. However, it is extremely difficult to obtain good function in the facial nerve after total removal of a large tumour by any technique.

## Transtemporal Bone Technique

In 1964 House published a monograph based on the removal of 53 eight nerve tumours by an approach through the temporal bone, with either a very limited exposure of the internal meatus through the middle fossa or a wider exposure through the mastoid and labyrinth. The two special points contained in this monograph were that eight nerve tumours could frequently be diagnosed when quite small and that facial nerve preservation could then be an important aim in treatment. The translabyrinthine approach was a revival of a technique suggested in 1904 by Panse and carried out in 1912 by Quix and later by others, but allowed to lapse having been condemned by Cushing (1921). The intrusion of an otologist into one of the most prized areas of neurosurgery caused a great deal of resentment which was largely directed against the techniques used.

The exponents of the techniques say that they allow the facial nerve to be reliably located at a fixed point within the bone, which is a considerable aid in the further dissection of the tumour from the nerve, and that since the approach is through the temporal bone there is less retraction and damage to the cerebellum, which lessens ataxia postoperatively. Furthermore, with a middle fossa approach it is occasionally possible to preserve some hearing in the case of very small tumours.

The neurosurgical argument, which has not always been put dispassionately, is that the exposure is so restricted that it is insufficient for the removal of a large tumour. It can, however, be extended either backwards into the posterior fossa by ligation of the sigmoid sinus or upwasrds into the middle fossa by division of the superior petrosal sinus. Neurosurgeons also contend that House has placed preservation of the facial nerve above the more important aim of total removal.

The truth is that House's most important contribution was his demonstration of the practicability of early diagnosis, and his enormous series cointains far more small and medium-sized tumours than were previously seen by neurosurgeons. This has stimulated both neurosurgeons and otologists so that the former, while adhering to the routine neurosurgical approach, have benefited by being offered smaller tumours. The use of the operating microscope has also increased the delicacy and gentleness of the operation and these two factors alone would by themselves have brought about an improvement in results.

Our own experience in the use of the translabyrinthine techniques is such that we believe that they have a good deal more advantage that are usually allowed by neurosurgeons, because the approach is more direct although the field is narrow and there is always a risk of CSF leak through the middle ear and of infection postoperatively. When early diagnosis is made and the surgeon uses some form of optical magnification good results can be obtained and the facial nerve reliably preserved by either method, but it cannot be sufficiently emphasized that early diagnosis and treatment require the close cooperation of the otologist and the neurosurgeon. The techniques of middle fossa and translabyrinthine approaches to the internal acoustic meatus have been well described by Portmann et al (1975).

**Summary**

In the last 10 years considerable advances have been made in the diagnosis of eight nerve tumours so that small tumours are now presenting for surgery. New approaches to the internal acoustic meatus through the petrous bone have been developed as a result of the introduction of technical aids - notably operating microscopes and high speed drills. Small tumours may now be removed with low morbidity and mortality and a high expectation of preserving the facial nerve. Large tumours continue to present a formidable challenge, and preservation of the facial nerve with total removal of the tumour remains difficult.