

# Acoustic Neuroma: Etiology, Presentation, and Treatment

Paul Kongkham, B.Sc. (OT1)

## Abstract

Acoustic neuromas (vestibular schwannomas) are common, benign tumours of the posterior cranial fossa, arising from the neurilemmal sheath of the vestibulocochlear nerve. The signs and symptoms associated with these tumours are defined by the proximity of the neoplasm to other structures in the cerebellopontine angle. Advances in neurological imaging now permit earlier diagnosis of acoustic neuromas. Surgical innovations such as the intraoperative microscope, cranial nerve monitoring, improvements in microsurgical methodology, and advances in radiotherapy have reduced the morbidity and mortality associated with the treatment of these tumours. This review provides a description of the clinical neuroanatomy of acoustic neuromas and the current treatment strategies available.

## Introduction

Acoustic neuromas are benign Schwann cell tumours that arise from the vestibulocochlear nerve (CN VIII) in the posterior cranial fossa. These neoplasms account for approximately 10% of all primary intracranial tumours and are the most common tumour of the cerebellopontine angle (80%), with an incidence of approximately 1 per 100 000.<sup>1-5</sup> Although acoustic neuromas most often occur unilaterally (> 95%) and produce an onset of symptomatology during middle age, bilateral tumours occur in approximately 5% of patients and are pathognomonic for neurofibromatosis type-2 (NF-2).<sup>6-7</sup> Acoustic tumours frequently arise from the superior division of the vestibular portion of CN VIII, proximal to the porous acousticus, in a region of the neurilemmal sheath known as the Obersteiner-Redlich zone (the junction between central and peripheral myelin).<sup>8</sup> Although the rate of tumour expansion is variable (between 1-10 mm per

year), the majority of acoustic neuromas grow approximately 1-2 mm annually.<sup>7-8</sup>

## Genetics

Deletion of the *NF-2* tumour suppressor gene on the long arm of chromosome 22 is thought to contribute to the initiation of acoustic tumour development.<sup>7-9</sup> This deletion may arise as a somatic or germline mutation, the origin of which predisposes the individual to the development of unilateral or bilateral tumours, respectively. Although mutation of the *NF-2* gene is thought to be the early genetic event that initiates tumourigenesis, neoplastic growth appears to depend upon additional mitogens such as nerve growth factor (NGF), platelet-derived growth factor (PDGF), basic fibroblast growth factor (bFGF), and transforming growth factor beta 1 (TGF -  $\beta$ 1).<sup>7, 9-11</sup> Curiously, the known tumour suppressor genes *p53*, *adenomatous polyposis coli* (APC), and *NF-1* do not appear to be important in the development of acoustic tumours.<sup>12</sup>

## Signs and Symptoms

The size of the acoustic neuroma usually determines its symptomatology, as growing tumours can compress adjacent cranial nerves (CN) as well as the brainstem and cerebellum (Table 1).<sup>13</sup> In particular, the acoustic neuroma may impinge on the contents of the internal auditory canal, including CN VII, CN VIII and the internal auditory artery.<sup>14</sup> The majority of tumours will produce some form of otologic dysfunction by the time of diagnosis, with the most common symptoms experienced early in this disease being hearing loss, tinnitus, and dysequilibrium (all related to compression of CN VIII).<sup>1, 14-18</sup>

Progressive, unilateral hearing loss occurs almost invariably (~ 98%) in patients with acoustic tumours and commonly man-

**Table 1**  
**Significant Surgical Anatomy for Acoustic Neuromas, Organized by Cisterns**

CISTERN	LOCATION	CN	ARTERIAL STRUCTURES	SIGNIFICANT VENOUS STRUCTURES
Cerebello-medullary	Lateral to brainstem, caudal to pontomedullary sulcus	IX, X, XI	Vertebral arteries PICA	
Cerebellopontine	Lateral to brainstem, along surface of pons and cerebellum	V, VI, VII, VIII	SCA AICA	Superior Petrosal Vein
Premedullary	Anterior to medulla, between the medulla and clivus	XII	Anterior Spinal Arteries	
Prepontine	Anterior to pons, rostral to premedullary cistern		Basilar Artery	

*CN - cranial nerve, PICA - posterior inferior cerebellar artery, SCA - superior cerebellar artery, AICA - anterior inferior cerebellar artery.*

ifests as a loss of high frequency tones and a reduced ability to discriminate speech.<sup>1, 14, 17, 18</sup> Patients often complain of difficulty carrying on telephone conversations and on average, report reduced hearing for a period of 4 years prior to their diagnosis of acoustic neuroma.<sup>1, 14, 18</sup> Acute hearing loss occurs in a minority of individuals (~ 10%), primarily in younger patients with large tumours.<sup>1, 14, 17, 18</sup> The precise mechanism that underlies hearing loss is uncertain, but may be related to direct compression of CN VIII, reduction in the blood supply to CN VIII or to the cochlea itself. However, because neither the severity of hearing loss nor the reduced ability to discriminate speech correlate with tumour size, the etiology of these neurological deficits is likely multifactorial.<sup>17</sup> Tinnitus is experienced by approximately 70% of patients with acoustic neuromas and is thought to be caused by neural or vascular compression.<sup>1, 14, 17, 18</sup> The tinnitus is often persistent and high-pitched, occurring ipsilateral to the tumour.<sup>14</sup> Dysequilibrium is the third most common symptom produced by acoustic tumours, with an incidence that correlates positively with tumour size.<sup>1, 14, 17, 18</sup> However, if tumour growth is slow, the ipsilateral vestibular deficit may be sufficiently gradual to allow compensation by contralateral vestibular input preventing true vertigo.<sup>14</sup>

Acoustic tumours that extend into the cerebellopontine angle frequently cause anterior displacement of the facial nerve (CN VII), compressing it against the anterior surface of the tumour capsule (Figure 1).<sup>8, 14</sup> Signs and symptoms caused by compression of CN VII include facial weakness and alterations in taste sensation.<sup>1</sup> Further extension of the tumour rostrally can displace the trigeminal nerve (CN V), compressing it between the tumour mass and the tentorium cerebelli. Signs and symptoms produced by compression of CN V include facial hypesthesia/anesthesia (often in the distribution of the second division of CN V), otalgia, and ipsilateral reduction or loss of the corneal reflex.<sup>8, 14, 17, 18</sup> Rarely, compression of CN V or VII results in symptoms that mimic microvascular compression syndromes such as tic douloureux or tic convulsif, complicating the clinical presentation of acoustic neuromas.<sup>14, 15, 18</sup> Caudal extension of the

acoustic tumour occurs less frequently, but may displace the glossopharyngeal (CN IX), vagus (CN X) and accessory nerves (CN XI), producing dysarthria, dysphagia, and hoarseness.<sup>8, 14, 17</sup>

Extensive tumour enlargement leads to compression of the brainstem and cerebellum.<sup>14</sup> Cerebellar compression contributes to the dysequilibrium experienced by patients with an acoustic neuroma and may also result in an ataxic gait and limb incoordination.<sup>14</sup> Brainstem compression may cause respiratory depression and long tract signs such as hemiparesis.<sup>8, 14</sup> Compression of the fourth ventricle with obstruction of cerebrospinal fluid drainage can cause non-communicating hydrocephalus and elevated intracranial pressure that may manifest as headache, nausea, vomiting, reduced visual acuity, diplopia, or obtundation.<sup>8, 14, 16, 17</sup> Without intervention, tonsillar herniation secondary to posterior fossa mass effect may ensue.<sup>14</sup>

### Diagnosis

Diagnosis of acoustic neuroma requires a thorough history, otological, and neurological examinations. Otologic findings are

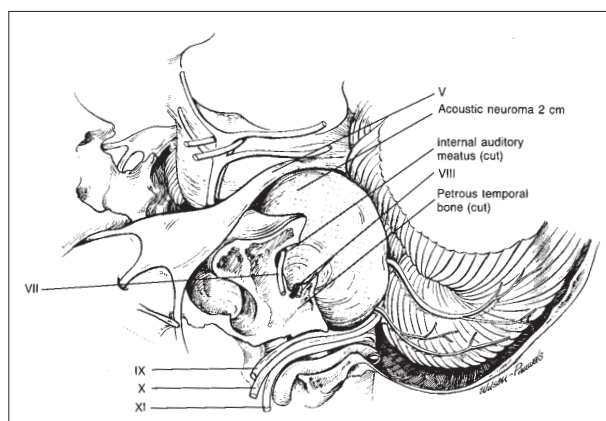


Figure 1. Schematic of a large acoustic neuroma within the left cerebellopontine angle, depicting displacement of adjacent cranial nerves, the cerebellum and brainstem.

consistent with a sensorineural hearing loss and may include a unilateral reduction in hearing, an ipsilateral positive Rinne test and a Weber test lateralizing opposite to the ear with the hearing deficit. On neurological examination, the cranial nerve signs and symptoms described previously may indicate the presence of a cerebellopontine angle lesion. Audiometric testing (i.e. pure tone audiograms, speech discrimination) can provide a baseline measurement to aid in the choice of treatment and in evaluating post-treatment hearing function. In patients with an acoustic neuroma, pure tone audiograms often show a sensorineural loss of high tones.<sup>8, 14, 17</sup> Poor speech discrimination suggests that the lesion is retrocochlear, with severity often related to the extent of auditory nerve fiber loss.<sup>14</sup> At present, the most reliable, non-radiologic test for acoustic neuroma is the evaluation of brainstem auditory evoked potentials, which measure the changes in electroencephalographic responses to auditory stimuli.<sup>14</sup> Additionally, caloric testing evaluates vestibular function by stimulating the vestibular apparatus with water and measuring the duration of the resulting nystagmus. Specifically, this tests the function of the horizontal semicircular canal, which is innervated by the superior division of the vestibular nerve.<sup>14</sup>

When acoustic neuroma is highly suspected, initial radiologic evaluation may be more cost-effective than audiometric or electrophysiologic testing.<sup>20, 21</sup> Thin-slice axial plane magnetic resonance imaging (MRI) with gadolinium enhancement shows acoustic tumours as round/oval enhancements centred on the internal auditory canal. This procedure has a sensitivity of 98%, a low false positive rate, and can reveal lesions with a diameter of only a few millimeters (Figure 2).<sup>6, 8</sup> Computed tomography (CT) may also show widening of the internal auditory canal due to bony erosion by the tumour.

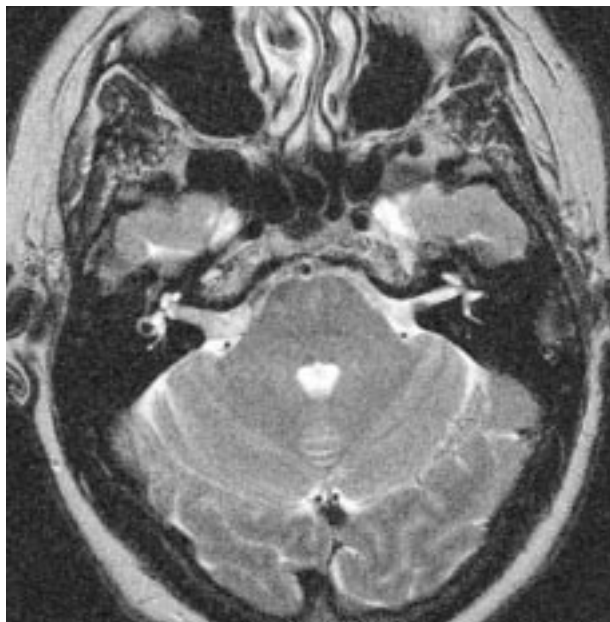


Figure 2. An axial T2 - weighted MRI without contrast, revealing a right-sided acoustic neuroma extending from within the internal auditory canal into the cerebellopontine angle.

## Treatment

The choice of management strategy is dependent upon several factors, including the patient's general health, symptomatology, tumour size and growth rate, hearing function, past attempts at treatment, and the presence of bilateral tumours.<sup>6, 8, 18, 22, 23, 24</sup> Three strategies are used to treat acoustic neuromas: expectant management, radiotherapy and microsurgery. Expectant management involves monitoring the patient for clinical or radiological signs of tumour progression. Serial audiometry and imaging (CT or MRI) is performed annually until tumour growth plateaus. This is a reasonable alternative for patients with asymptomatic tumours, with a tumour in the only-hearing ear, with high operative risks, or who refuse surgical intervention.<sup>25, 26</sup> Microsurgical or radiotherapeutic intervention is advised if the rate of growth exceeds 2 mm per year.<sup>8</sup>

Radiotherapy can be effective at halting tumour growth and reducing the size of small or medium-sized intracanalicular tumours.<sup>27</sup> Up to 98% of patients undergoing radiotherapy may not require further surgery.<sup>29</sup> Two forms of radiotherapy are used to manage acoustic neuromas: Gamma Knife and Linear Accelerator (LINAC). Gamma Knife radiotherapy involves stereotactically-guided irradiation of the tumour using multiple [<sup>60</sup>Co] sources that surround the patient's head.<sup>29</sup> This treatment allows for tumour control rates of up to 95%.<sup>27</sup> LINAC also delivers photons derived from a single [<sup>60</sup>Co] source, thus providing a high dose at the tumour site with minimal peripheral radiation by rotating the source about the patient's head.<sup>29</sup> However, irradiation of irregularly-shaped targets is more difficult with LINAC, and there is a higher risk of injury to surrounding structures. Recently, the approach of fractionation, delivering the total dose of radiation over several days, has been applied to LINAC methods and has been shown to reduce the incidence of post-treatment cranial neuropathies when compared to single-dose LINAC.<sup>29</sup> While radiotherapy may preclude the need for surgical intervention, it increases the risk of trigeminal neuropathy, hydrocephalus, and exposure of normal brain tissue to radiation with unknown risks of inducing secondary tumour formation.

Microsurgical resection is considered the standard treatment modality for acoustic neuromas.<sup>6, 8, 17</sup> Three surgical approaches, translabyrinthine, suboccipital, and middle fossa, are commonly used. The translabyrinthine method involves drilling through the mastoid portion of the temporal bone and the vestibule between the external auditory canal and the sigmoid sinus.<sup>8, 17</sup> This method allows for early identification and protection of CN VII, avoidance of cerebellar retraction, a lower risk of injury to caudal cranial nerves, visualization of the anterior brainstem, and good access to the tumour bed to control any postoperative complications that may arise.<sup>6, 8, 17, 23</sup> However, this approach also results in obligatory iatrogenic hearing loss in the affected ear, potentially longer operating time, and difficulty visualizing the brainstem with large tumours.<sup>6, 8, 17</sup>

The suboccipital method involves performing a craniotomy in

the angle of the transverse and sigmoid dural sinuses, and approaching the tumour via the angle between the cerebellum (medially) and the petrous temporal bone (laterally). This approach permits good tumour exposure (regardless of size), excellent control of bleeding (especially around the brainstem), and the possibility of preserving both CN VII and CN VIII (cochlear portion) function.<sup>6, 8, 17, 30</sup> Disadvantages include late identification of CN VII, the risk of cerebellar injury due to retraction pressure, and increased difficulty in removing tumour from the internal auditory canal without violation of the vestibule during drilling.<sup>8, 17</sup> Use of an endoscope during this procedure may overcome some of these disadvantages by allowing better visualization of residual tumour at the lateral end of the internal auditory canal as well as exposed petrous air cells not seen with the operating microscope.<sup>31, 32</sup>

Of these surgical approaches of acoustic neuroma resection, the least commonly used is via the middle fossa. This involves accessing the tumour from above after retracting the temporal lobe rostrally, and drilling through the roof of the internal auditory canal. Advantages of this approach include good exposure of small intracranial tumours and the possibility of hearing preservation.<sup>6, 8, 17</sup> However, there is limited exposure available via this method, and there may be an increased risk of facial nerve palsy and injury to the temporal lobe secondary to retraction.<sup>8, 17</sup>

The frequency of usage of the suboccipital approach versus the translabyrinthine approach are now approximately equal (50% versus 45% respectively), while the middle fossa approach is used in only 3% of cases.<sup>17</sup> Several criteria have been developed as aids in deciding the best surgical approach for patients with tumours of varying severity.<sup>18, 22, 23</sup> Discrepancies between these criteria stem from differences in their definitions of serviceable hearing, tumour size classification, and surgeon preference for a particular approach. One system currently in use groups patients into the following categories: elderly patients (over age 70), patients with small tumours (2.5 cm or less) and serviceable hearing, patients with small tumours and poor hearing, and patients with large tumours (3 cm or more) regardless of their hearing status.<sup>22</sup> The suboccipital approach is used to resect small tumours in patients with serviceable hearing. The translabyrinthine approach is used to treat all large tumours and small tumours in patients with poor hearing.<sup>22</sup> In the elderly, small asymptomatic tumours are managed expectantly with yearly CT scans and clinical exams.<sup>22</sup> If their tumours are causing symptomatic hydrocephalus, surgery is performed to insert a cerebrospinal fluid shunting device.<sup>22</sup> Large tumours with progressive symptoms in the elderly are treated surgically according to the same criteria indicated above.<sup>22</sup>

The surgical treatment of acoustic neuromas is still fraught with complications. A common complication is the development of cerebrospinal fluid (CSF) fistulae, which occurs in up to 35% of cases and manifests as CSF rhinorrhea, otorrhea, or surgical site drainage.<sup>8, 18, 31, 33, 34</sup> The majority of these fistulae resolve with

subarachnoid lumbar drainage, although some require surgical closure.<sup>18</sup> Complications related to cranial nerve injury are common. Of these, palsies of CN VII are often the most distressing for patients.<sup>6</sup> Despite the fact that the majority of patients with acoustic neuromas have normal facial nerve function preoperatively, up to 44% have impaired facial function postoperatively.<sup>17</sup> In addition to possible disfiguring facial weakness, facial palsies increase the risk of exposure keratitis and corneal ulceration, and care must be taken to protect the eye from these sequelae. Injuries to CN VIII are also common.<sup>33</sup> The vestibular portion of the nerve is rarely spared in surgery and postoperative vertigo, dizziness, and nausea and vomiting are common. In addition, the cochlear portion is frequently injured, with up to 75% of patients exhibiting a decline in hearing function postoperatively.<sup>17</sup>

## Summary

Acoustic neuromas are common tumours of the posterior cranial fossa. Despite being histologically benign, these tumours cause significant morbidity as a result of localized growth, with resultant compression and displacement of adjacent neural structures. Without medical intervention, severe neurological compromise due to brainstem compression and cerebellar herniation may result. Improvements in diagnostic ability and innovations in both surgical and non-surgical forms of management have led to earlier and more effective treatment of these tumours with an emphasis on reducing the associated morbidity to the patient. However, significant morbidity is still associated with the management of this disease. Therefore, patients must be informed of all treatment options and given realistic appraisals of expected functional outcomes for each modality in order to decide on the most appropriate course of management.

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