Acoustic Neuroma Surgery: Retrosigmoid Techniques

13

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The last century has seen great strides in the accurate diagnosis and microsurgical management of acoustic neuroma (AN), with improvements in mortality rate and preservation of both facial nerve function and hearing [1]. Acoustic neuromas were among the earliest intracranial lesions to be anatomically localized on the basis of symptoms [2, 3]. The first reported surgical attempt was by Charles McBurney, who opened the suboccipital plate with a chisel in 1881 but was forced to abort the case following excessive cerebella swelling [4]. Early surgical attempts were heroic interventions of last resort in moribund patients and were associated with surgical mortality rates of up to 78% [5]. With developments in surgical technique and sterility, Harvey Cushing reported a mortality rate of 4% in 1931 [6]. Walter Dandy further advanced the field using ventriculographic and pneumoencephalographic imaging and a unilateral suboccipital craniotomy [7, 8]. With such advancements, complete tumor excision became more commonplace and rates of anatomic preservation of the facial nerve approached 65% in 1941 **[9–11]**.

William House introduced the operating microscope to acoustic neuroma surgery in 1961 and advocated that each operation be performed by a team of a neurosurgeon and neuro-otologist [12]. Elliott and McKissock in 1954 were the

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first to report hearing preservation following a retrosigmoid (RS) resection of an AN [13]. Subsequently, surgeons have focused on the extent of resection and avoidance of facial weakness and hearing loss—factors critical to a patient's choice among management options of clinical and radiographic monitoring, three surgical approaches, and stereo-tactic irradiation. This chapter will focus on the indications, predictive factors, classification, microsurgical technique, and outcomes for a retrosigmoid approach to an AN resection.

Pathology and Pathophysiology

The pathophysiology of ANs in relation to hearing is comprehensively explored in Chap. 12, while the biology and genetics are covered in Chap. 9. Briefly, mechanisms of cranial nerve dysfunction, including hearing loss, can be categorized as compressive, infiltrative, ischemic, or a combination of these. Although the vast majority of ANs arises from the vestibular divisions of the eighth cranial nerve, infiltration of the cochlear nerve is common even in cases with small tumors, good preoperative hearing, and unremarkable intraoperative appearance [14, 15].

Preservation of the cranial nerves requires a functional, anatomically continuous nerve with an adequate vascular supply. Tumor exposure, cerebellar retraction, or dissection of the tumor from adjacent normal structures can disrupt a nerve's continuity, function, or vascularity [16]. Sekiya and Moller demonstrated in a primate model that avulsion of the internal auditory artery in the cerebellopontine angle (CPA) could result in hearing loss [17]. In canine models, either mechanical nerve distortion or vasospasm from vascular manipulation alters brainstem auditory potentials [18, 19] and produces demyelination and thrombosis of the vasa nervorum [19]. The occasional spontaneous recovery of cochlear nerve function weeks to months after its loss during

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surgery may represent resolution of a neural conduction defect caused by ischemia, mechanical retraction, or a combination of both [16].

The cranial nerves, particularly the cochlear nerve, can be injured by traction on the nerve from sustained cerebellar retraction, which may be required in a retrosigmoid approach to large tumors. Traction injury to the nerve fibers occurs at mechanically weak sections, such as the Obersteiner–Redlich zone of transition from Schwann cell sheath to glial cell coverage, which lacks the reinforcing endoneurium of the distal nerve [20]. Furthermore, in the case of the cochlear nerve, the fragile small fibers located laterally at the modiolus are prone to avulsion from the base of the cochlea, evident intraoperatively as sudden loss or prolonged latency of wave V of the auditory brainstem response (ABR) despite preservation of wave I following cerebellar retraction [21].

Meticulous surgical technique is required at all times. Sharp dissection can partially or completely divide the nerve. Blunt dissection can stretch, shear, or avulse vital nerve components or blood vessels. Electrocauterization can cause thermal injury to the cochlear or facial nerve or its blood supply and should be avoided in its proximity. Preservation of the vascularity of a cranial nerve is often key to maintenance of function. Drilling away the posterior wall of the internal auditory canal (IAC) in the retrosigmoid approach can inadvertently damage the inner ear either directly or by thermal conduction. Opening into the bony labyrinth can also compromise hearing, an outcome prevented in some cases by early recognition and closure of the opening with bone wax, particularly if the fenestration occurs at the convexity of a semicircular canal. Hearing preservation is much less likely if either the cochlea or vestibule is transgressed.

ABR findings can help differentiate true neural injury from cochlear injury. In cochlear nerve injury, wave I of the ABR is preserved, but injury to the cochlear nerve itself affects all waves of the ABR. Isolated cochlear injury is confirmed postoperatively by the ability to activate the cochlear nerve electrically by stimulating the promontory despite clinical deafness [22].

Investigation

Audiometry

The quality of a patient's hearing is a major consideration in choice of treatment of an acoustic neuroma. Given the high dependence of postoperative hearing on preoperative hearing, outcomes for all treatment strategies, both interventional and observational, are stratified according to preoperative hearing level [23, 24]. Thus, accurate preoperative assessment of hearing is critical.

A patient's hearing quality is usually described in terms of thresholds for hearing pure tones and accuracy in speech discrimination. The unit of measurement for sound pressure is the decibel (dB), which is based on a logarithmic ratio. In pure-tone audiometry, the pure-tone average (PTA) is the mean threshold for sound detection (dB) at the octave frequencies of 250, 500, 1000, 2000, 4000, and 8000 Hz. Occasionally, interoctave frequencies of 3000 Hz and 6000 Hz are also used. Zero decibel is the lowest amplitude of sound detected by an ideal ear. Normal thresholds fall between 0 and 25 dB for all frequencies. The standard audiogram represents a graph of the perception threshold (dB) as a function of frequencies (Hz) tested. The frequencies most needed for speech lie between 500 and 3000 Hz. One hearing classification system based on dB level includes normal hearing (0-25 dB) and mild (25-40 dB), moderate (40-60 dB), severe (60-80 dB), and profound (>80 dB) hearing loss.

Speech audiometry evaluates the relative clarity or "usefulness" of the patient's hearing of speech. Word recognition is tested using a standardized list of 25–50 single-syllable words "phonetically balanced" to represent the relative frequency of sounds in the language being tested. The word recognition score (WRS) is the percentage of words the patient is able to repeat correctly.

The combination of PTA and speech reception threshold is highly informative about the usefulness of a patient's speech, the etiology of hearing loss, and potential therapeutic interventions. For example, patients with poor pure-tone thresholds but relatively preserved word recognition should respond well to hearing aids because they can still process amplified speech in a meaningful way. However, patients with favorable pure-tone thresholds but poor word discrimination may not benefit from amplification because of perceived distortion. This is often the case in patients with neural hearing losses caused by retrocochlear pathology such as an AN; the resulting disordered firing of the cochlear nerve both raises perception thresholds and disproportionately limits understanding by impairing sound processing. Traditionally, a WRS higher than 50% is thought to be required for effective use of hearing aids. A simplistic WRS model includes class 1 (100-70%), class 2 (69-50%), class 3 (49–1%), and class 4 (0%) word recognition [25, 26].

Classification

The classification scheme of Gardner and Robertson, which combines PTA and WRS, was used by many early studies of acoustic neuromas (Table 13.1) [27]. It has been supplanted by a scheme developed by the American Academy of

Tab	le 13.1	Gardner–Ro	bertson	hearing of	classification
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Grade ^a	Description	PTA or SRT (dB) ^b	WRS
Ι	Good	0–30	70-100
II	Serviceable	31-50	50-69
III	Nonserviceable	51-90	5-49
IV	Poor	>91-max loss	1-4
V	None	No response	No response

PTA pure-tone average, SRT speech reception threshold, WRS word recognition score

^a If PTA/SRT score and WRS do not qualify for the same class, use the class appropriate for poorer of the two scores

^b Use better score of either PTA or SRT

Table 13.2
American Academy of Otolaryngology-Head and Neck

Surgery (AAO-HNS)
Image: Comparison of the second se

Class ^a	Description	PTA or SRT (dB) ^b	WRS
А	Good	0–30	70-100
В	Serviceable	31-50	50-69
С	Nonserviceable	>51	>50
D	Poor	>51	<50

PTA pure-tone average, SRT speech reception threshold, WRS word recognition score

^a If PTA/SRT score and WRS do not qualify for the same class, use the class appropriate for poorer of the two scores

^b Use better score of either PTA or SRT

Otolaryngology-Head and Neck Surgery (AAO-HNS) (Table 13.2) [28]. AAO-HNS class A and B hearing correspond to Gardner–Robertson grade I and II hearing. However, the AAO-HNS class C and D place a greater emphasis on the WRS and thus provide greater insight into a patient's potential to benefit from hearing aids.

More recently, guidelines elucidating the minimum standard for reporting hearing loss have been published by the AAO-HNS hearing committee in an attempt to improve data comparison between studies and enable pooling of data for meta-analysis [29]. These guidelines recommend the use of preintervention and postintervention scattergrams, which plot WRS along the *x*-axis and PTA along the *y*-axis, enabling a granular display of hearing outcomes at the individual patient level. Importantly, the PTA is calculated using 0.5-, 1-, 2-, and 3-kHz air conduction thresholds, and the WRS is presented at up to 40 dB sensation level of maximum comfortable loudness [29].

Definition of Success

Comparisons of hearing outcomes from managing ANs have long been confounded by investigators' inadequate characterization of initial hearing [23], use of different hearing classification systems, and employment of varying definitions of useful hearing and, thus, of rates of successful hearing preservation. This discrepancy has been recognized in both the otolaryngology and neurosurgical literature. In 2012, the hearing committee of the AAO-HNS produced an updated set of reporting standards, which have been outlined

updated set of reporting standards, which have been outlined in the previous section [29]. More recent consistent use of current classification systems utilizing PTA and WRS has facilitated more meaningful analysis of outcome. For vestibular schwannoma management, the 2018 guide-

lines of the Congress of Neurological Surgeons propose that useful (or serviceable) hearing be defined as a WRS of greater than 50% and a PTA or speech response threshold of less than 50 dB, which is equivalent to AAO-HNS class A or B and Gardner-Robertson score of grade I or II [23]. However, these scales must be used cautiously. The AN patient who has a good WRS in the quiet may still complain of substantial impairment of speech understanding in noise. Furthermore, the usefulness of a specific level of hearing in a tumor-affected ear also depends on the quality of hearing in the contralateral ear. In general, if hearing in the affected ear has perception thresholds in the speech frequencies more than 30 dB above or if WRS is more than 30% below those of the contralateral ear, hearing in the affected ear contributes little to the patient's speech comprehension. This 30/30 criterion for useful hearing is used by clinicians who counsel patients about treatment options and expectations. In the future, it may be more appropriate to present data according to a change in PTA and WRS over time, with use of visual aids such as scattergrams, and to include metrics that have improved correlation with the real-world impact of hearing loss, such as speech recognition in noise and associated quality-of-life surveys [30].

Auditory Brainstem Responses

ABRs are the most sensitive and specific audiologic tests for the diagnosis of ANs and were used extensively prior to magnetic resonance imaging (MRI). Among patients with documented ANs, 20–30% have lost all ipsilateral ABR waves, 10–20% have only wave I, 40–60% have all waves but the latency of wave V is increased, and 10–15% have normal waveforms [31]. However, the technique suffers from a rate of false-negatives of approximately 15%, but this rate can range from 33% for intracanalicular tumors to 4% for larger lesions [31–33]. Rates of false-positives (an abnormal ABR when no AN is present) are much higher, exceeding 80% in some series [34–36].

ABRs also may be prognostic for hearing preservation. One study of 286 patients correlated preserved hearing with lower mean interwave V latencies (0.51 vs 0.7 ms for those with no postoperative hearing) and absolute wave V latencies (5.35 vs 5.96 ms) on preoperative ABR [37]. Another study of 107 patients found that rates of hearing preservation were significantly higher if the preoperative ABR had good morphology (63% vs 48% in those with poor ABR morphology) and a wave III (66.7% vs 33.3% with no wave III) [38]. Matthies and Samii classified preoperative ABRs into five types: B1–B5. Types B1 and B2 contained waves I, III, and V with variable latencies [39, 40]. Patients with a wave III (types B1–B2) had a higher rate of hearing preservation than patients without wave III (types B3–B5). Aihara and colleagues found that an interaural difference of wave V latency (IT5) of less than 1.12 ms was prognostic of useful postoperative hearing [41].

Otoacoustic Emissions

Some hearing loss from an AN or its treatment involves loss of cochlear function, some of which may reflect disruption of its vascular supply [42]. Otoacoustic emissions (OAEs) emanate from the cochlea's outer hair cells. Preserved OAEs may indicate preserved cochlear function and encourage hearing preservation strategies. Although several studies have examined OAEs in AN patients, only a few patients both lack an ABR and yet have intact OAEs that meet the criteria predictive of potential hearing preservation [43–45]. Ferber-Viart and colleagues found that OAEs were a significant predictor of hearing preservation, but Brackmann and colleagues failed to identify a significant correlation. Further studies are needed to fully evaluate the role of OAEs in AN surgery [37, 38]. Another study found that preoperative transient otoacoustic emission was a favorable prognostic indicator of preservation of useful hearing preservation after surgery [46].

Vestibular Testing

Electronystagmography (ENG) is frequently abnormal in AN patients. The caloric response stimulates the lateral semicircular canal, which is innervated by the superior vestibular nerve (SVN). An absent caloric response may indicate injury to the superior vestibular nerve (SVN) by a tumor originating from either vestibular nerve. Ninety-eight percent of patients with an AN originating from the superior vestibular nerve show a reduced caloric response compared with 60% of those with a tumor from the inferior vestibular nerve (IVN) [47]. Furthermore, those with an AN arising from the SVN had significantly less postoperative hearing loss, likely because the SVN is less intimately related anatomically with the cochlear nerve and the internal auditory artery than is the IVN [36, 48, 49]. Three recent studies have failed to demonstrate ENG as a significant prognostic factor in hearing preservation, likely because ENG is not specific for the nerve of origin [37, 50, 51]. Therefore, we do not routinely order caloric testing.

Radiology

Imaging of the CPA and the AN is covered comprehensively in Chap. 3. The discussion here focuses on imaging characteristics important to hearing conservation microsurgery.

MRI Screening: When to Do It?

Whether all patients with otherwise unexplained asymmetrical hearing loss should undergo MRI screening for a potential AN is controversial. In a retrospective cohort comparison study of more than 400 patients with asymmetrical hearing loss, Gimsing and colleagues found an interaural asymmetry of perception threshold of greater than 15 dB at two contiguous frequencies (between 2000 and 8000 Hz), an interaural asymmetry of WRS of greater than 20%, unilateral deafness, an interaural asymmetry of perception threshold of greater than 20 dB at two contiguous frequencies, or unilateral tinnitus that had the highest sensitivity for identifying an AN. Another retrospective study of more than 200 patients found an interaural asymmetry of greater than 15 dB at 3000 Hz provided the highest positive likelihood ratio (2.91) for the presence of an AN [52, 53]. Guidelines of the Congress of Neurological Surgeons recommend that greater than 10 dB asymmetry at two or more contiguous frequencies or greater than 15 dB at any single frequency warrants MRI [54].

Imaging Characteristics

Contrast-enhanced MRI is the imaging modality of choice for ANs. Key features can confirm the expected diagnosis, guide the choice of approach, and help assess risks of complications. For instance, far-lateral extension of a tumor in the IAC raises concern that all of the tumor may not be removed by a retrosigmoid exposure without increased risk of hearing loss [55]. In most retrosigmoid approaches, exposure of the lateral third of the canal risks injury to the otic capsule and thereby reduces the chances of hearing preservation [56–58].

In a cadaveric study, high-resolution computed tomography (CT)-based frameless navigation (with or without endoscope) further facilitated lateral access; whether outcomes in patients improve remains to be shown [59]. Gerganv and colleagues found that a shorter distance between the lateral tumor margin and fundus significantly correlated with worse hearing outcomes [60]. Another study found incomplete obliteration of the IAC to be a positive predictor of serviceable hearing after surgery [61]. Lateral intracanalicular extension of tumor can also challenge a middle fossa approach in which the lateral 25% of the IAC may be obscured by the overhang of the transverse crest [62].

Anterior extension, including erosion of the anterior bony wall of the IAC, is unfavorable for facial nerve outcome and likely associated with significant tumor compression of the cochlear nerve and possibly hearing loss [63]. Similarly, tumor prolapsed laterally into the cochlear modiolus eliminates the chance of hearing preservation.

As previously mentioned, the origin of the tumor from the superior or inferior vestibular nerve is significant for preserving both hearing and facial nerve function [48]. The intimate relationship between inferior vestibular nerve tumors and the cochlear nerve and internal auditory artery reduces the likelihood of hearing conservation [64]. These tumors also tend to deflect the facial nerve superiorly, leaving it in a less favorable position for a middle fossa approach. We routinely use coronal MRI to determine the tumor's location relative to the transverse crest because this relationship has practical implications for selecting a surgical approach. The optimal MRI sequence for visualizing cranial nerves is a high-resolution T2-weighted MRI; however, improved definition of nerves utilizing tractography is an area under active investigation [55, 65, 66].

The size and location of the tumor are major considerations in the choice of surgical approach. Intracanalicular tumors can be managed via either the middle fossa or retrosigmoid approaches. The middle fossa approach probably provides the best chance of hearing conservation in small tumors. However, it often requires significant manipulation of the facial nerve situated between the surgeon and the tumor, carrying higher risk of facial nerve dysfunction, particularly for a tumor from the SVN [62, 67].

In tumors with a CPA component of 0-15 mm in diameter, the middle fossa approach is associated with a relatively high rate of transient facial nerve dysfunction, but long-term results are similar to those of the retrosigmoid approach [67, 68]. In tumors with CPA components of 10–18 mm in diameter, the hearing conservation rate via the middle fossa approach was only 34% compared with 63% for tumors with less than 10 mm extension into the CPA, while long-term facial nerve outcomes were worse [69]. Similarly, a metaanalysis of surgical approach for an AN found that hearing preservation rates were similar for middle fossa and retrosigmoid approaches to tumors more than 1.5 cm in diameter, but facial nerve dysfunction was significantly higher with the middle fossa approach [67]. Informed patient participation in the choice of approach is essential because different patients may weigh the relative importance of hearing and facial function differently.

In patients with serviceable hearing and tumors with 10–25 mm diameter extension into the CPA, a retrosigmoid approach is preferred if the lateral third of the IAC is free of tumor. Hearing preservation rates are low in tumors with a

CPA extension greater than 25 mm [70]. Yet, it is still reasonable to attempt hearing conservation via the retrosigmoid approach in these cases, particularly if the patient has excellent preoperative hearing and the extension of the tumor into the IAC is limited.

Complications

The complication profile associated with the retrosigmoid approaches will be considered here with a focus on hearing preservation. In contemporary acoustic neuroma surgery, facial nerve injury is uncommon, and the risk of permanent severe or total paralysis is below 10%. This risk is greater for large tumors [67]. Facial nerve outcomes of translabyrinthine and retrosigmoid approaches are generally comparable, although a meta-analysis suggested that a retrosigmoid approach results in better facial nerve outcomes than translabyrinthine or middle fossa approaches for tumors greater than 3 cm and significantly better outcomes than the middle fossa approach for intracanicular tumors [67]. Others have found that the middle fossa approach has a higher incidence of transient weakness for tumors with less than 10 mm extension into the CPA and of permanent weakness for tumors with 10-18 mm CPA extension [68, 69]. Therefore, if hearing preservation is to be attempted, we prefer the retrosigmoid approach for all tumors with more than 10 mm extension into the CPA.

Persistent postoperative headache can be a significant morbidity. Headache is more common with the retrosigmoid approach; in one study, postoperative headache was 3.8 times higher after a retrosigmoid than after a translabyrinthine approach. It may persist for 6 months after surgery [32, 67, 71]. Its cause is not completely clear. The risk of headache associated with postoperative aseptic meningitis can be reduced by limiting dissemination of and thoroughly removing intradural bone dust that results from drilling open the IAC. Replacement of the suboccipital bone plate and a curvilinear incision have been advocated to reduce postoperative headaches [48, 72–81].

Retraction of the cerebellum during the retrosigmoid approach can injure it; encephalomalacia in the lateral 1–2 cm of the hemisphere is sometimes seen on T2-weighted MRI after surgery. Most patients have no symptoms. If the injury extends more deeply, a prolonged ataxia may result.

In our experience, efforts to spare the cochlear nerve in hearing preservation approaches increase operative time and the risks of postoperative vestibular dysfunction and tumor recurrence. However, a study of more than 700 patients found that the middle fossa approach was associated with a higher risk of recurrence than retrosigmoid and translabyrinthine approaches whose risks were similar [76]. The increased vestibular dysfunction likely reflects abnormal signals from vestibular nerve remnants, which may slow vestibular compensation. Tumors can recur from a small fragment left in the fundus [77]. The chances of recurrence are higher with the middle fossa approach than with the other two. The relative risk of recurrence after retrosigmoid and translabyrinthine approaches is controversial. Recurrence after a retrosigmoid approach may be more common when dissection of the lateral third of the IAC is blinded by the preserved otic capsule [56–58, 62, 76]. Although the endoscope is routinely used in some centers to inspect the distal IAC for residual tumor, there is minimal evidence that this reduces recurrence [78, 79]. In our experience, it is often difficult to use angled endoscopes in such a small area without risking injury to the facial nerve and difficult to discern tumor from nerve and in the fundus.

Operative Techniques

Choice among operative approaches should consider numerous factors, including whether hearing preservation is to be attempted, the size of the tumor, its radiological characteristics, potential complications, and patient preferences. The comparison of operative strategies is considered in detail in Chap. 5. The focus of this chapter will be the surgical nuances of the retrosigmoid approach.

Retrosigmoid Approach

The retrosigmoid approach takes a suboccipital intradural route between the posterior petrous face and the lateral cerebellum to the CPA and IAC. It is perhaps the most versatile of all approaches to the CPA as it may be used both in hearing preservation procedures and for large tumors in which hearing preservation is not a consideration [67]. The following steps are critical.

Patient Position and Monitoring

After general anesthesia is induced, arterial and bladder catheters are inserted. A prophylactic antibiotic (cefuroxime 2 g, intravenous [IV]) is typically given. Electrodes for monitoring cranial nerves (V, VII, IX, XI) and earphones and electrodes for monitoring ABR are placed (when ABR is being monitored). Care should be taken to isolate the external auditory canal and insure that the sterilizing solution does not compromise hearing assessment.

The patient is placed in the supine-lateral position, and the ipsilateral shoulder is elevated on a folded blanket. The head is turned away from the side of the lesion, ideally 20° beyond lateral, while the neck is flexed 20°, and the vertex is angled inferiorly 10° to place the retromastoid region uppermost in the surgical field. Some surgeons use rigid head fixation (e.g., a Mayfield head holder), but it is unnecessary unless navigation is to be used. The left lower quadrant of the abdomen is prepared in a sterile fashion and draped in anticipation of harvesting a fat graft. The surgeon stands or sits at the head of the operative table. To confirm awareness of the surgical plan by the entire team, a "team time-out" is performed prior to the incision.

Incision

The retromastoid region is shaved, prepared, and draped in sterile fashion. The incision is designed to expose bone overlying retrosigmoid dura from the origin of the sigmoid sinus from the transverse sinus to just above the jugular bulb. The course of the transverse sinus is approximated by a horizontal depression in the skull, immediately above the superior occipital line, extending laterally from just above the inion to the asterion, just above and posterior to the top of the pinna. The course of the sigmoid sinus can be approximated by the vertical prominence of the posterior aspect of the mastoid superior to the digastric groove. A 6-cm vertical incision is marked parallel and 1 cm posteromedial to the vertical prominence from 2 cm above to 4 cm below the horizontal depression (Fig. 13.1). Curving the ends of the incision slightly can be useful in patients with bulky necks by enabling greater retraction of the scalp flap. Before the marked line is incised, it is injected with local anesthetic (lidocaine 1% with 1/100,000 epinephrine).



Fig. 13.1 The incision is made in the retromastoid region. Placing the incision in a relatively anterior position minimizes trauma to the nuchal musculature and the occipital nerve. (Reproduced from Jackler RK [90] with permission, copyright © 2007 RK Jackler, MD)

Soft Tissue Dissection

The incision extends through skin, galea, and suboccipital fascia and muscle down to the bone. Inferiorly, special care is taken to avoid injuring a vertebral artery passing anomalously above the foramen magnum. To minimize devascularization and facilitate closure, we advocate minimal use of the monopolar cautery until the muscle layer is reached. The soft tissue is elevated from underlying bone to expose the posterior mastoid anterolaterally and 3 cm of suboccipital bone posteromedially from just above the level of the transverse sinus to below the suboccipital convexity. Ideally, a periosteal elevator is used to minimize muscle trauma and thermal injury. The Apfelbaum modification of a suboccipital self-retaining retractor is placed.

Craniotomy

A single burr hole is drilled just medial and inferior to the asterion, which overlies the transition of the transverse sinus to the sigmoid sinus, a point approximated by the intersection of the vertical retromastoid line and transverse depression. The dura is carefully cleared from the bone using a Penfield dissector #3. A craniotomy 3-4 cm high and 2-3 cm wide (depending on the size of the tumor) is opened immediately inferior to the transverse sinus and posterior to the sigmoid sinus. Residual bone covering the posterior aspect of the sigmoid sinus is drilled away to increase the anterolateral exposure (Fig. 13.2). Doing so usually requires isolation, coagulation, and division of a prominent emissary vein entering the midportion of the sigmoid sinus. Particularly for large tumors, bone removal should extend below the convexity of the suboccipital bone to facilitate access to the cisterna magna. The margins of the craniotomy should be coated with bone wax, particularly occluding any opened mastoid air cells.

Dural Opening

The dural incision runs from the superolateral corner to the midline of the craniotomy. It proceeds inferiorly in a vertical line before turning inferolaterally to the inferior-lateral corner, thereby creating a rhomboid-shaped flap based anteriorly. Tack-up sutures pull the flap taut anterolaterally, partially rotating the posterior margin of the sigmoid sinus forward. Alternatively, a posteriorly based dural incision can be made to allow the flap to be held under the retractor (Fig. 13.3). An additional incision from the inferoposterior corner of the durotomy to the inferoposterior corner of the craniotomy frees an inferior triangle of dura, which can be retracted inferiorly to provide access to the cisterna magna. Prompt elevation of the cerebellar tonsil (using a Teflon-coated retractor) and opening of the arachnoid of the cisterna magna (using a No. 11 blade) under direct vision with a microscope permit drainage of cerebrospinal fluid and decompression of the posterior fossa-a maneuver particularly important with large tumors.



Fig. 13.2 A single burr hole is drilled just medial and inferior to the asterion. A craniotomy is opened immediately inferior to the transverse sinus and posterior to the sigmoid sinus. Any residual bone covering the posterior aspect of the sigmoid sinus is removed using a combination of rongeur and drill to increase the anterolateral exposure. *SS* sigmoid sinus, *TS* transverse sinus. (Reproduced from Jackler RK [90] with permission, copyright © 2007 RK Jackler, MD)



Fig. 13.3 The dura is incised about 5 mm from the edge of the craniotomy to facilitate its suture closure. Relaxing incisions are created to define small dural flaps, which are tacked up with small sutures. (Reproduced from Jackler RK [90] with permission, copyright © 2007 RK Jackler, MD)

Retraction

The approach to the tumor at the meatus and in the CPA is along the anterolateral surface of the cerebellar hemisphere and middle cerebellar peduncle. Arachnoid of the posterior aspect of the central CPA cistern is incised to allow a 5/8-in.wide retractor blade to be positioned so that the lateral cerebellar hemisphere can be elevated from the posterior petrous face. AdapticTM, a nonadherent material, is placed between the cerebellum and the blade to protect the cerebellum and enhance hemostasis. The retractor is positioned just dorsal to the interface of the posterolateral convexity of the tumor with the middle cerebellar peduncle. The arachnoid just superior to the nerves of the jugular foramen is divided to allow their relaxation inferiorly, away from the inferior pole of the tumor. A similar division of arachnoid superiorly allows the superior pole of the tumor to be separated from the petrosal vein, which should be preserved (Fig. 13.4).

Identification of Nerves at Brainstem

Early identification of the eighth and seventh cranial nerves proximally at the brainstem is helpful to their preservation. If the tumor is large enough to completely obscure the seventh and eighth cranial nerves at the brainstem, it must first be partially debulked as described below. In other cases, the proximal nerves can be found beneath the inferior pole of the tumor. A tuft of choroid plexus at the foramen of Luschka lying just inferior to the flocculus is often a helpful landmark [64]. It lies just posterior to the origin of the ninth cranial nerve from the brainstem. The entry of the eighth cranial nerve into the brain-



Fig. 13.4 An Apfelbaum retractor is used as the base of a retractor arm that supports the malleable blade. The blade is positioned over the lateral aspect of the cerebellum over a strip of AdapticTM. The lateral lobe of the cerebellum and the flocculus is then elevated off the tumor to expose the posterior aspect of its extracanalicular portion. The petrosal vein (also known as Dandy's vein) should be preserved. If needed for large tumors, it may be controlled with bipolar cauterization. (Reproduced from Jackler RK [90] with permission, copyright © 2007 RK Jackler, MD)

stem is about 4 mm superior and 2 mm posterior to the origin of the ninth cranial nerve. The seventh cranial nerve exits the brainstem at a point in line with the origins of the glossopharyngeal and vagal nerves, 2 mm anterior and inferior to the entrance of the eighth cranial nerve [80].

Electrophysiologic stimulation can confirm the identity of the nerve. The main stem of the anterior inferior cerebellar artery (AICA) usually passes laterally below the facial and vestibular nerves at the brainstem, but it can pass above or, rarely, between them [81]. Usually a moderately sized vein of the pontomedullary sulcus, sometimes accompanied by a twig of the rostral branch of the AICA, passes between them. Aggressive bipolar coagulation in the area should be avoided lest either nerve be injured.

The facial nerve almost always passes anterior to the tumor. Its course can be estimated from the initial direction of the nerve along the brainstem, but its subsequent path to the meatus cannot be reliably predicted as inferior (immediately lateral beneath the lower pole), intermediate (obliquely across the midportion of the tumor), or superior (up along the brainstem and then lateral to the anterior part of the upper pole). It rarely passes through the tumor and almost never lies posterior to the tumor [81]. Nonetheless, electrophysiologic simulation should always be performed prior to incising the pseudocapsule to exclude a possible posterior location, during general debulking to exclude potentially injurious penetration of the anterior pseudocapsule, and when tracing the nerve during its dissection from the pseudocapsule.

The cochlear nerve typically passes along the anterior aspect of the lower third of the tumor. Its preservation is best attempted by dissecting tumor away from any uninvolved nerve at the inferior pole. Often the cochlear component is not distinct from residual uninvolved vestibular nerve throughout the dissection. Instead, the tumor's smooth surface—evident as it is separated from the chalice of expanded uninvolved nerves—serves to reassure that the cochlear nerve, passing even more anterior, is being preserved. Such a strategy is also more likely to preserve the critical microvascular supply to the nerve and inner ear.

Removal of CPA Tumor

The arachnoid covering the posterolateral aspect of the tumor is swept posteriorly from the petrous face back over the tumor to the cerebellum. Preservation of this arachnoid plane permits extra-arachnoidal resection of tumor and greatly facilitates dissection of the tumor pseudocapsule from cerebellum, middle cerebellar peduncle, brainstem, and cranial nerves.

Removal of a CPA tumor begins with internal debulking. Risk of injury to the facial and cochlear nerves is minimized by entry into the posterior aspect of the tumor after electrophysiological screening for the facial nerve. The pseudocapsule is incised after bipolar coagulation and after the tumor within it is morselized and removed. This intratumoral debulking relaxes the pseudocapsule, encourages its separation from the facial nerve, and permits rotation of more of the tumor into direct surgical access without excessive manipulation of the facial nerve. Iterative internal tumor debulking, dissection of the pseudocapsule away from uninvolved nerves, and trimming freed tumor reduce the tumor to a thin plaque along the facial and cochlear nerves.

The larger the tumor, the greater is the risk of traumatic or ischemic injury to the cranial nerves or brainstem [82]. In this circumstance, the facial and cochlear nerves are likely to be elongated and attenuated and more vulnerable to injury. Therefore, dissection must be meticulous. The facial nerve is particularly vulnerable when it takes a long superior course along the brainstem before turning back inferiorly and laterally to cross to the meatus. Occasionally, a small plaque of tumor wedged at the apex of this hairpin turn must be left to avoid injuring the nerve.

Larger tumors are more likely to compress the brainstem and breach its arachnoidal protection. On preoperative T2-weighted MRI, this scenario is often apparent as brainstem edema. However, the brainstem's surface is usually remarkably tolerant of careful microdissection of the tumor's pseudocapsule. Such dissection must be performed with great care to avoid diverging from the surface of the tumor into neural tissue. This risk is greatest at points where the tumor attaches to the brainstem, usually corresponding to small arteries or veins bridging between tumor and brainstem [83]. Veins leaving the tumor and arteries branching solely to tumor should be isolated from the brainstem, coagulated, and divided. Bleeding caused by inadvertent rupture of such small vessels often stops with time and gentle pressure. Consequently, patience is preferable to aggressive efforts at coagulation, which might injure the brainstem or nerves [84]. Any attachment to larger, more proximal branches of the AICA and, with much larger tumors, to the superior cerebellar, posterior inferior cerebellar, basilar, and vertebral arteries must be identified and carefully freed [85]. Incorporation of such an artery within the tumor is another indication for leaving a small plaque of residual tumor.

Opening the Posterior Wall of the IAC

The IAC portion of the tumor can be exposed by drilling away the bone from its posterior wall. The IAC can be drilled early after cerebellar retraction or later after the CPA component of tumor has been retracted. When possible, we prefer to drill the IAC early, before dissection of arachnoid planes in the CPA. Doing so helps minimize the spread of bone dust into the cistern and may reduce the incidence of aseptic meningitis and postoperative headache. The definitive identification of the facial nerve in the IAC may also help during subsequent dissection of the CPA component. With larger tumors, the CPA component may be debulked to obtain sufficient access to the posterior petrous face prior to IAC drilling.

The location of the porus acusticus can be palpated with a ball hook, as can that of the operculum endolymphatic sac, which represents the origin of the vestibular aqueduct. The axis of the IAC extends from the porus to just superior to the operculum. The dura over the petrous face is carefully incised along this line to avoid cutting the endolymphatic sac. Superior and inferior dural flaps are retracted to expose the bone posterior to the IAC (Fig. 13.5). The superior flap is



Fig. 13.5 To expose the IAC component of the tumor, the posterior osseous wall of the canal must be opened. With larger tumors, this aspect of the procedure is performed after the CPA component is debulked. To expose the bone overlying the IAC, dural flaps are elevated anteriorly and posteriorly. Preserving the dural flaps provides a purchase for suture closure of the defect at the end of the procedure. Before

bone removal, Gelfoam[®] (G) is placed in the posterior fossa to confine the spread of bone dust within the subarachnoid space. 10 = vagus nerve; 9 = glossopharyngeal nerve; ES = endolymphatic sac; 5 = trigeminal nerve. (Reproduced from Jackler RK [90] with permission, copyright © 2007 RK Jackler, MD)



Fig. 13.6 (a) A cutting bur is used to rapidly drill a trough to the level of the IAC dura. (b) Diamond burs are then used to excavate troughs anterior and posterior to the IAC. Approximately two-thirds of its circumference is exposed to allow the IAC to be in high relief. It is particu-

larly important to funnel the porus acusticus widely to avoid overhangs that might obscure the tumor-facial nerve interface as it angulates sharply into the CPA. (Reproduced from Jackler RK [90] with permission, copyright © 2007 RK Jackler, MD)

elevated to the tentorium. Doing so often entails sacrifice of the subarcuate artery, a branch of the AICA. This sacrifice is generally well tolerated as this artery is usually an end artery into the surrounding bone [64]. The inferior flap is elevated off the endolymphatic sac and to the superior margin of the jugular foramen. Gelfoam[®] is packed around the tumor to limit the spread of bone dust.

Bone removal commences with a 3 mm cutting bur at the porus to identify the posteromedial dura of the IAC. A 3-mm and then a 2-mm diamond bur is used to define the IAC further. The endolymphatic aqueduct is a useful landmark for the lateral extent of safe bone removal. Drilling through the endolymphatic aqueduct risks injury to the underlying crus commune and subsequent irreversible inner ear damage.

Troughs are drilled superior and inferior to the medial IAC to facilitate tumor exposure and resection. The jugular bulb may be immediately inferior to the IAC and, in some cases, may overlap its posterior face, which may obstruct exposure in an anatomic variant called a high-riding jugular bulb. The surgeon should review the patient's preoperative images to be alert to this possibility and to avoid injury to the jugular bulb. The dura of the IAC is opened along its axis with a No. 11 blade or a myringotomy knife, and superior and inferior flaps are created (Fig. 13.6). Given the variability of the facial nerve's course, the nerve stimulator should be used before the dura is incised to prevent inadvertent sharp injury to the nerve.

Removal of Intracanalicular Tumor

Once the dura of the IAC is incised and the contents of the canal are exposed, tumor removal can begin. The nerves are identified using the landmarks and features discussed above. Given the posterior exposure of the IAC, the vestibular

nerves and tumor are encountered first. At the lateral end of the canal's opening, the facial nerve is sought just anterior to the superior vestibular nerve. The plane between the facial nerve and tumor is developed, and the cochlear nerve is identified just anterior to the inferior vestibular nerve. Careful debulking of tumor allows additional neural exposure. The portion of the vestibular nerve of tumor origin continuous with tumor is divided and dissected from the remainder of that nerve as well as from the other vestibular nerve, facial nerve, and cochlear nerve. Special effort must be made to verify removal of all lateral tumor extending deeply toward the fundus. This far-lateral dissection must sometimes be performed without the benefit of direct visualization; a small endoscope may permit a more definitive view of the distal canal. Small, cupped micro-instruments passed gently along the facial nerve are used to palpate osseous landmarks (such as the transverse crest) and to retrieve any residual tumor.

The initial dissection within the IAC usually proceeds laterally to medially. However, as noted below, the direction of dissection is not as important as its delicacy. Critical issues include avoiding traction on tumor tissue or on any nerve that might result in stretch of cochlear or facial fibers anchored at the meatus (commonly) or fundus as well as identifying and preserving the internal auditory artery. The dissection is continued through the meatus and along the superior petrous face to join that from the CPA. Once the removal of the tumor is complete, the facial nerve can be stimulated to ensure functionality; stimulus at 0.1 mA predicts good facial nerve outcome [86].

Closure

Closure begins with verification of hemostasis with the patient's blood pressure at normal levels and with a Valsalva

maneuver. Irrigation of the microscopic field flushes debris from the subarachnoid space. The drilled petrous surfaces of the IAC are coated with bone wax to obliterate any opened mastoid air cells. Subcutaneous fat from the abdomen is placed over the nerves in the IAC and covered by a piece of Surgicel[®] to hold it in place. Sometimes, a single suture (4-0 nylon) can be placed to appose the dural flaps of the posterior petrous face over the fat graft. The cerebellar retractor is removed, and the cortex of the hemisphere is inspected for bleeding. The dura is closed in watertight fashion, using a graft or muscle patch as needed. The margins of the craniotomy are again waxed. The bone plate is secured with titanium miniplates. Suboccipital muscle and fascia and galea flaps are sutured, and the skin is stapled.

Microsurgical Dissection

Microsurgical dissection of the tumor can be difficult, and certain challenges are common to all approaches. Removal of tumor remnant from the proximal acousticofacial bundle represents one such challenge. The remaining nerves are often compressed and distorted into a shape that resembles a tulip or wine chalice surrounding the tumor remnant. Usually, the remaining vestibular nerve is posterior, the cochlear nerve is more inferior, and the facial nerve is anterior. The tumor can often be separated from the more substantial facial nerve before it is dissected from the cochlear nerve. Preservation of any uninvolved vestibular nerve often helps during dissection. The margin of the tumor is not a true capsule. Rather, it is a pseudocapsule formed by peripheral tumor cells arranged more compactly and more tangentially to the tumor surface compared with central tumor cells [87]. Because peripheral tumor cells can adhere to attenuated nerves, this final stage of dissection must be extremely delicate.

Often, partial rotation of the tumor and nerves can optimize the orientation of the dissection plane. Turning the plane to enable multiple approaches for dissection can help. The direction of dissection is less important than minimizing the traction on the nerves. Fine blunt dissection along a cleavage plane with a small disk dissector is useful. This maneuver can stabilize the nerve, while gentle traction of the tumor is maintained with a small suction. A broad plane of dissection is always desirable and is best maintained by sweeping the dissector delicately over as much of the surface of the tumor interface as possible. When the correct plane is maintained, the tumor's pseudocapsule appears smooth.

Loss of the correct plane can result in tiny remnants of tumor against nerve. These tumor plaques often infiltrate where fine vessels bridge tumor and nerve. They can become progressively thicker if dissection continues in a false plane. The correct plane can be regained either by sharply dividing the inciting attachment and elevating the plaque or by dissecting at another edge of the tumor. Remaining nerves should be inspected closely for residual tumor fragments. Small bleeding points along the facial or cochlear nerves should not be coagulated; thrombin-impregnated Gelfoam[®] pledgets, gentle pressure, and patience are preferable.

Intraoperative Monitoring

Intraoperative monitoring of cranial nerve function was introduced by Delgado and colleagues in the 1970s and is now standard procedure in most operating rooms [88, 89]. While it has primarily been aimed at helping identify and preserve cranial nerves, there is increasing interest in using it as a electroprognostic marker for early postoperative counseling of patients and in timing facial reanimation [90, 91]. The recent guidelines for AN surgery released by the Congress of Neurological Surgeons have recommended the use of intraoperative eighth cranial nerve monitoring in AN surgery [91]. This chapter specifically focuses on monitoring for hearing preservation. The first and most common method is recording of ABRs. The measurement of direct cochlear nerve action potentials (CNAPs), electrocochleography (ECoG), and evoked OAEs are other potential options.

Intraoperative ABR recording uses headphones within the ear canal to deliver a repetitive click stimulus to the ear. Electrodes over the mastoid and scalp then measure the electrical response of the inner ear, cochlear nerve, and brainstem. By averaging the response over time, distinct waves (I–V) can be recorded, providing information on the integrity of the auditory pathway. Despite the use of high stimulus rates, more than 1 min may be needed to obtain a reproducible waveform. A stimulus intensity of 95 dB is used to maintain an adequate signal-to-noise ratio. An ABR from the contralateral ear serves as a control and as a monitor for generalized effects such as anesthesia and temperature [92]. Clinically, the amplitude and latency of waves I, III, and V are monitored. Although each wave is generated from numerous sources, it is useful to consider wave I as arising from the distal eighth cranial nerve, wave III from the superior olivary complex, and wave V from the inferior colliculus [93]. Harper found significant improvement in hearing preservation rates for small tumors (less than 11 mm) using monitoring, with the presence of waves I and V being a positive predictor variable (with 67% likelihood of useful hearing preservation) [94].

In a review of intraoperative ABR changes in 201 patients undergoing AN resection, the risk of deafness associated with temporary loss of either wave I, III, or V was 11–14%. The risk associated with permanent loss of any of these waves was 65–78% [39]. The disappearance of waves I and III usually preceded the disappearance of wave V. The disappearance of wave III was the earliest and most sensitive sign. Neu and colleagues classified intraoperative ABR into four prognostic patterns [95]. Hearing was preserved in all patients with a stable wave V (pattern 1), whereas all patients with an abrupt loss of ABR (pattern 2) lost hearing. An irreversible loss of either wave I or wave V (pattern 3) was associated with eventual postoperative hearing loss. Patients with reversible ABR changes (pattern 4) had variable outcomes. Despite correlations between intraoperative ABR changes and postoperative hearing, some surgeons believe that changes in the ABR lack specificity. A major deficiency is that the lengthy acquisition times may not give sufficient warning to enable surgeons to take corrective action if the nerve is in danger.

The direct measurement of CNAPs using an electrode adjacent to the cochlear nerve in the operative field allows reproducible waveform averages to be obtained within seconds, compared with as long as 1 min for ABR averaging [92]. Both monopolar and bipolar electrodes are placed at the root entry zone of the eighth cranial nerve or more distally in the IAC. Amplitude and latency variability are measured continually during surgical maneuvers that put the nerve at risk. Advantages of CNAP over conventional ABR include near real-time feedback to the surgeon, easier identification of waveforms, and reliable responses even when conventional ABR is lost or deformed [96]. In one study, CNAP waveforms could be recorded in 92% of patients, whereas ABR could be obtained in only 48% of patients undergoing hearing conservation surgery [97]. The first positive peak (N1) in the CNAP waveform is generated by the cochlear nerve, and the latency is similar to that of wave II on the ABR [98]. Either decreased amplitude or increased latency of N1 can signify injury to the eighth cranial nerve [99, 100]. At the end of surgery, the presence of the N1 waveform in the CNAP is prognostically significant. In one review, no patient lacking N1 had postoperative hearing, whereas 79% of patients with N1 had measurable hearing [101]. Piccirillo and colleagues found that patients with tumors smaller than 1.5 cm and normal preoperative hearing were more likely to have AAO-HNS class A hearing; however the presence of CNAP did not ensure a good hearing outcome [102].

The primary disadvantage of the CNAP is that it reflects the integrity of the cochlear nerve only to the point where the electrode is placed. It does not give information about the integrity of the auditory pathway downstream, as does the ABR. With large tumors, the root entry zone of the cochlear nerve may be inaccessible until some tumor has been removed. In cases where the root entry zone is inaccessible, the electrode can be placed extradurally against the bone of the IAC and adjacent to the cochlear nerve [103]. CNAP recordings are highly dependent on electrode position; therefore, care must be taken during manipulations that might displace the electrode. With care, the electrode and wire can be placed outside the path of microsurgical instruments. ECoG, another near-field technique, also can be used to record cochlear microphonic (CM) potential and summating potential (SP). CM potentials are generated by cochlear outer hair cells, and the SP represents a depolarization of the hair cells. The SP is a presynaptic response, whereas the CNAP is a postsynaptic response [92]. When ECoG and CNAP are used together, the site of damage can be localized to the cochlea or the eighth cranial nerve. However, use of ECoG alone is limited. ECoG potentials can persist for some time despite complete division of the eighth cranial nerve [104].

Several studies have compared the utility of these various intraoperative monitoring techniques. Battista and colleagues retrospectively reviewed 66 patients who underwent either ABR, ECoG, or CNAP monitoring during hearing conservation surgery [105]. Postoperatively, they found serviceable hearing in 24% of patients with ABR monitoring, 17% with ECoG monitoring, and 40% with CNAP monitoring. However, these differences did not reach statistical significance. In a review of 77 patients by Danner and colleagues, CNAP monitoring was associated with a significantly higher rate of measurable postoperative hearing than ABR (64% vs 41%) and was highest in tumors smaller than 1.5 cm [106]. However, when only serviceable hearing was considered. there was no statistically significant difference (43% vs 27%). Colletti and Fiorino found that patients monitored with ABR and CNAP had a significantly better postoperative PTA than those monitored with ABR alone (54.1 dB vs 82.5 dB) [107]. Unfortunately, data regarding the WRS or percentage of patients with serviceable hearing were not given.

Facial nerve monitoring is almost universal in AN microsurgical procedures. Lenarz and Ernest reported improved facial nerve function in both immediate and long-term (1 year) outcomes, particularly in larger tumors (>1.5 cm in diameter) with 87% of monitored patients having a House– Brackmann grade I–III immediate result compared to 74% of unmonitored patients [108]. A retrospective analysis in patients undergoing both translabyrinthine and retrosigmoid approaches found similar results in 121 patients [109]. In view of these findings, recent guidelines from the Congress of Neurological Surgeons have recommended the use of facial nerve monitoring [91].

Hearing Results

The success of hearing preservation surgery varies widely, and confounding factors and bias must be considered when comparing different surgical approaches and techniques (Table 13.3). Guidelines from the Congress of Neurological Surgeons found that the probability of maintaining serviceable hearing for small to medium tumors (<2 cm in diameter)

Table 1	3.3	Results	of	hearing	preservation	studies
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	Number of patients			
	with			AAO-HNS
	serviceable		Tumor size	Class A + B ^b
Study	hearing	Approach	(cm) ^a	No (%)
Glasscock et al. [112]	136	38 MF, 98 RS	<1.5	37 (27)
Brookes and Woo [113]	17	RS	<1.0	9 (53)
Arriaga et al. [110]	26	RS	Mean = 1.66	14 (54)
	34	MF	Mean = 0.72	24 (71)
Slattery et al. [50]	143	MF	Mean = 1.2	74 (52)
Irving et al. [68]	25	MF	IC	11 (44)
	20	MF	0.1-1.0	12 (60)
	5	MF	1.1-2.0	1 (20)
	17	RS	IC	2 (12)
	12	RS	0.1-1.0	3 (25)
	21	RS	1.1-2.0	3 (14)
Satar et al. [69]	104	MF	IC-0.9	57 (62)
	47	MF	1-1.8	15 (33)
Rohit et al. [114]	107	59 MF, 48 RS	<1.5	34 (32)
Arts et al. [111]	62	MF	0.3–1.8	45 (73)
Grayeli et al. [115]	44	RS/MF	<1.5	25 (57)
Quist et al. [116]— immediate	49	MF	NR	27 (55)
Quist et al. [116]—5- year follow-up	16	MF	NR	12 (75)
Sughrue	702	RS	NR	330 (47)
Yamakami	36	RS	<1.5	26 (72)
et al. [118]				
Mazzoni et al. [119]	189	RS	>3.2	47 (25)
Di Maio et al. [120]	28	RS	>3.0	6 (21%)
Hilton et al. [121]	78	MF	NR	51 (65%)
Maw et al. [122]	33	RS	<3.0	38%
Chee et al. [123]	126	RS	<2.0	43 (34%)
Lee [124]	59	RS	<3.0	11 (19)
Kaylie et al. [125]	27	RS	<4.0	8 (29)
Ferber-Viart [38]	86	RS	>4.0	47 (55)
Gormley [126]	69	RS	<3.9 cm	38%
Post et al. [127]	46	RS	0.9–4.0	18 (39%)

Table 13.3 (continued)
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	Number of patients with serviceable		Tumor size	AAO-HNS Class A + B ^b
Study	hearing	Approach	(cm) ^a	No (%)
Rowed [128]	26	RS	IC	50%
Samii [129]	16	RS	IC	56%
Colletti [107]	25	RS	0.4–1.2	57 (%) (A, B, C)
Nonaka [130]	170	RS	<2.0	82.8%
Sameshima [131]	82	RS	<1.5	73.2%
Sanna et al. [132]	107	RS		54.2%
				Hannover Class ^c (H1 + H2)
Samii and Matthies [133]	29	RS	T1 ^d	6 (21)
	96	RS	T2 ^d	25 (26)
	249	RS	T3 ^d	39 (16)
				Gardner– Robertson Grade I + II
Cohen et al. [134]	128	RS	<0.5	32 (37) ^e
			0.6–1.0	32 (34) ^e
			1.1–1.5	38 (24) ^e
			>1.5	26 (11) ^e
Dornhoffer et al. [51]	65	MF	<0.5	39 (60) ^e
	11	MF	0.5 - 1.0	7 (64) ^e
	17	MF	1.0-1.5	8 (47) ^e
Betchen et al. [135]	142	RS	$0.4 - 4.0^{f}$	43 (30) ^e
Rowed and Nedzelski [128]	26	RS	IC	13 (50) ^g
	68	RS	0.4-1.5	20 (29) ^g
Lin [136]	113	RS	<2.0	30 (27), 18 (16) at 9.5 years
Goel et al. [137]	42	RS	>2.5	13 (31)
Fischer et al. [138]	22	RS	>3.0 to <1.0	12 (55)

IC intracanalicular, MF middle fossa, RS retrosigmoid

Note: Selected studies using the middle fossa approach are included in the table for comparison

Modified with permission from Jackler RK and Driscoll CLW [139] ^a Tumor size includes posterior fossa component except when

indicated

^b AAO-HNS classification system

^c New Hannover classification system

 d T1 = intrameatal; T2 = intrameatal and extrameatal; T3 = filling the cerebellopontine angle

° Pure-tone average <50 dB and word recognition >50%

 $^{\rm f}$ Size range of tumors with preserved hearing

^g Pure-tone average <50 dB and word recognition >60%

following microsurgical resection was between 25% and 50% at 2, 5, and 10 years postoperatively [23]. For patients with AAO-HNS class A or Gardner–Robertson grade I, 2-year and 5-year probabilities of serviceable hearing were 50–75%, dropping to 25–50% at 10 years [23].

Factors with a heavy impact on hearing outcome postoperatively include preoperative serviceable hearing, size (particularly less than 15 mm in diameter), and a distal internal auditory cerebrospinal fluid cap, while age and sex were not strong predictors [23]. Other factors that confound comparison of results among series include differing restrictions on tumor location, surgical approaches, metrics of hearing results, classification systems, and definitions of success.

The highest rates of hearing preservation have generally been reported with small tumors treated via the middle fossa approach [67–69, 110, 111]. In the most favorable conditions, the rate of preservation of useful hearing surpasses 50%. The middle fossa approach, however, has three disadvantages. Firstly, exposure of the CPA component of the tumor is limited. Secondly, in contrast to the translabyrinthine approach, the lateral IAC may require blind dissection. Thirdly, compared with other approaches, the facial nerve is at increased risk of permanent palsy if the cisternal tumoral component is more than 1.0 cm in diameter [62, 67, 69].

Overall hearing preservation rates via the retrosigmoid approach tend to be lower than those after a middle fossa approach for smaller tumors (less than 15 mm in diameter) [67]. However, a direct comparison controlling for tumor size and preoperative hearing status is difficult. In one series, as many as 25% of patients retained serviceable hearing after the retrosigmoid approach for tumors less than 20 mm in diameter [68]. Hearing preservation rates are diminished when the cisternal tumoral component is more than 20 mm in diameter [128, 133, 134, 140].

Which Approach?

A recent review found hearing preservation rates of 18.9– 77% with the middle fossa approach with a facial nerve preservation rate of 50–86%. The retrosigmoid approach also had excellent hearing preservation rates of between 11% and 68% with a higher facial preservation rate (59–98.7%) [25, 130, 131, 133, 141–145]. Recent consensus guidelines found either approach was reasonable for hearing preservation [146]. When choosing the approach for a particular patient, overall success rates are not as important as individual prognostic factors. For example, a patient with a small tumor, minimal IAC involvement, excellent preoperative hearing, and a normal ABR will likely have a 50% chance of retaining hearing regardless of whether the middle fossa or retrosigmoid approach is used. Colletti and colleagues found that tumors less than 3 mm from the IAC fundus had higher preservation rates with a middle fossa approach while, for those with more medial location, the middle fossa approach was not superior to a retrosigmoid approach [147]. Conversely, patients lacking these favorable characteristics will likely have poor results. Given that only a small fraction of patients with ANs are candidates for hearing preservation and the probability of success is limited, one can estimate that only 5% of patients with ANs will have useful hearing in the tumor ear after surgery. In comparison, a recent review of the literature found similar facial nerve preservation rates for retrosigmoid (36–95%) and translabyrinthine (29–89%) approaches [146].

Attempts at hearing preservation surgery are encouraged for patients with good preoperative hearing (class A or B) and tumors less than 20 mm in diameter [146]. Using a retrosigmoid approach, Sameshima found a hearing preservation rate of 73.2% in tumors less than 15 mm in diameter, and Nonaka and colleagues reported hearing preservation rates of approximately 83% for tumors less than 20 mm in diameter [130, 131]. At a mean follow-up of 18 months, Grayeli and colleagues found a hearing preservation rate of 57% in patients who presented with serviceable hearing [115]. An attempt at hearing preservation may be warranted for even large tumors. In patients with tumors larger than 30 mm in diameter and serviceable hearing preoperatively, Di Maio and colleagues found that 21% had serviceable hearing following RS surgery [120].

Follow-Up and Long-Term Outcomes

Imaging

Protocols for postoperative imaging stipulate different intervals between scans and lengths of follow-up depending on tumor, patient, and surgical factors. In a study of 299 patients for whom gross total resection of tumor was achieved, Bennett and colleagues found just 3 patients with nodular enhancement on MRI at 1 and 5 years; 2 of these were the only patients who developed recurrence [148]. Similarly, low recurrence rates were found in a translabyrinthine series by Tysome and colleagues: of 314 patients, 97% had no recurrence at 2 years, while 8 had linear enhancement at 2 years, none of whom progressed over 5–15 years [149]. One patient with nodular enhancement at 2 years had tumor progression. In a study of 50 patients, Arlt and colleagues found that 2 of 22 patients had recurrence after gross total resection at approximately 3.5 years, while 9 of 28 patients had recurrence after subtotal resection [150]. Recent guidelines recommend that baseline MRI be obtained within the first year following surgery, with annual or biannual imaging for at least 5 years [55]. If the patient develops nodular enhancement, more frequent imaging is indicated [55].

Hearing

Even after initially successful hearing preservation surgery, both pure-tone thresholds and speech discrimination can deteriorate over time. In 14 of 25 patients operated on via the middle fossa route, the average speech reception threshold loss was 12 dB, and the average loss of speech discrimination was 25% over a mean follow-up of 8.1 years [49]. A different study assessed preservation of serviceable hearing in 35 patients with Gardner-Robertson grade I and II hearing after a retrosigmoid approach over an average of 7 years. Overall, 30 patients (86%) maintained serviceable hearing, 5 patients (14%) dropped to class 3 or 4, and 3 patients (9%) increased from a class 3 into the serviceable range [135]. Similar rates of long-term hearing deterioration, ranging from 22% to 36% over 5 years, have been reported by others [79, 151]. Another retrospective study found 27% of patients had serviceable hearing in the immediate postoperative period, but this number dropped to 16% over 10 years of follow-up [136]. In contrast, in a study of patients under 40 years of age, Sughrue and colleagues found that, if patients had preserved hearing postoperatively, no patient progressed to nonserviceable hearing even after 10 years [152]. Similarly, in a study of 15 patients who had preserved hearing postoperatively. Yamakami and colleagues found that 12 patients (80%) maintained serviceable hearing after 7 years [118]. Another study found that, for tumors less than 20 mm in diameter and serviceable hearing prior to surgery, rates of continued serviceable hearing at 2, 5, and 10 years are 47%, 45%, and 43%, respectively [23].

Long-Term Risk of Recurrence

In a study of 299 patients who underwent gross total resection, Bennett and colleagues found only 2 patients (0.67%) developed recurrence [148]. Similarly, another study found only 1 patient of 314 developed recurrence following gross total resection [149]. However, in 203 patients, Carlson and colleagues found that subtotal resection increased future recurrence 16-fold [153]. Bloch and colleagues found that the recurrence rate of near total and subtotal resection over 3 years in 79 patients was 3% and 32%, respectively [154]. Another study of 20 subtotal resected tumors found only 1 recurrence over a mean follow-up of 5 years [155]. A recent study on a patient with unilateral AN who underwent a retrosigmoid approach found that there was a significant recurrence rate in subtotal resected ANs, with a recurrence-free survival rate at 5, 10, 15, and 20 years of 93%, 78%, 68%, and 51%, respectively [156]. Even in gross total resection, recurrence-free survival for 5, 10, 15, and 20 years was 96%, 82%, 73%, and 56%, respectively, while subtotal resection had 5-, 10-, and 15-year recurrence-free survival of 47%, 17%, and 8%, respectively [156]. A recent report evaluating large ANs (>2.5 cm), where residual tumor was treated with radiation, found that the likelihood of regrowth was three

times higher in subtotal resection and radiation when compared to gross total or near total resection, with similar facial nerve outcomes between the groups [157]. Overall, there appears to be a low risk of recurrence over the first 5 years following gross total resection. Because the risk is higher after subtotal resection, patients with incomplete tumor removal should be followed for decades.

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