Schwannomas of Cranial Nerves

Last updated: August 25, 2019

CEREBELLOPONTINE ANGLE TUMORS

1. Surgical Anatomy of Cerebellopontine Angle:
   - Vascular structures within cerebellopontine angle:
   - Nerves:
     - Internal auditory canal
   - Temporal bone:

2. Vestibular Schwannoma (80%)

3. Other rare cases:
   - Meningioma (20%)

Other rare cases:

1. VESTIBULAR SCHWANNOMA (80%)
   - Other schwannomas - trigeminal (< 8% of intracranial schwannomas)
   - Facial nerve (extremely rare)
   - Vascular tumor
   - Lipoma
   - Epidermoid

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2. Meningioma (20%)

3. Other rare cases:
   - 1) Schwanmomas (1?)
   - 2) Other schwannomas - trigeminal (< 8% of intracranial schwannomas), facial nerve (extremely rare)
   - 3) Vascular tumor
   - 4) Lipoma
   - 5) Metastases

Clinically - Cerebellopontine Angle Syndrome. see p. CNT >>

SURGICAL ANATOMY OF Cerebellopontine Angle

- cerebellum and lateral brain stem
- temporal bone
- roof and posterior boundary - cerebellum
- lateral boundary - posterior surface of temporal bone
- facial nerve - formed by lower cranial nerves (IX-XII)
- diencephalic region - thalamus, hypothalamus, basal ganglia
- main branch of ACA passes over cranial nerves VII and VIII in only 10% cases; remainder of time, it either passes below VII and VIII cranial nerves or, in 25-50% of individuals, actually passes between them.
- three branches that regularly arise from mesial segment of ACA: 1) small perforating arteries supply blood to brain stem.
- subarachnoid spaces: 1) between substantia and pia mater, 2) between pia mater and dura mater
- CN7 and CN8 receive blood supply from small branches of ACA.

VESSULAR STRUCTURES WITHIN CEREBELLOPONTINE ANGLE

1. Arteries:
   - most important vascular structure is ACA; its arises most commonly as single trunk from basilar artery but can arise as 2 separate branches. In rare cases, it originates as branch of PCA. As ACA moves from anterior to posterior, it first passes ventral surface of brain stem, but within cerebellopontine angle it takes shape and courses through diencephalic region and mesial segment of ACA to reach lateral brain stem.
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   - CN7 and CN8 receive blood supply from small branches of ACA.

2. Veins:
   - Must be kept in mind during surgical procedures on cerebellopontine angle: 1) Petrobasal vein of Dandy brings venous blood from cerebellum and lateral brain stem to greater petrosal sinus. It is encountered in area of CN7 anterior to pons acusticus. The petrosal vein often carries enough venous blood that its obstruction can lead to venous infarction and cerebellar edema.
   - Veins of Labbe (anterior is quite variable) carries returning venous blood from inferior and lateral surface of temporal lobe, enters superior petrosal or transverse sinus between temporal and occipital lobes at which greater petrosal sinus joins transverse sinus.
Facial nerve leaves brain stem anterior to foramen of Luschka.

- as it leaves brain stem, fibers are sheathed in oligodendroglia. Within few millimeters of leaving brain stem, however, nerve loses oligodendroglia and becomes sheathed by Schwann cells. It passes directly across cerebellopontine angle for about 2.0 cm, accompanied by CNS. It consistently enters internal auditory canal by crossing anterior superior margin of porus acousticus.

Vestibulocochlear nerve arises from brain stem slightly posterior to CN7.

- CNS remains sheathed in oligodendroglia for approximately 15 mm (almost at point to which it passes into internal auditory canal) but loses its oligodendroglial investment of any peripheral nerve. The junction between oligodendroglia and Schwann cells thus occurs just medial to porus acousticus. Because acoustic neuromas arise from Schwann cells, they arise most commonly within only lateral portions of cerebellopontine angle or internal auditory canal.

Nerves intermedius (nerve of Webersberg) leaves brain stem together with CNS.

- at some point within cerebellopontine angle, nerves intermedius cross over to become associated with CN7. It may do so as several separate rootlets. At what point nerves intermedius cross over to become associated with CN7 is considerably variable (in 22% individuals, it is adherent to CN8 for > 34 mm). As CNS and CN7 reach porus acousticus they pass into to together with nerves intermedius and, sometimes, loop of AJCA. 

INTERNAL AUDITORY CANAL

- ≈ 8.5 mm (5.5-10.5 mm) in length, lined with dura, and filled with CSF.

- medial end (porus acousticus) is oval in shape.

- lateral portion of foramen spinsum or lana cribrosa is complicated structure - divided into superior and inferior half by bony transverse crest.

1) upper half is further subdivided into anterior and posterior segment by vertical bony crest, often referred to as BURIC S. After William House, who popularized its importance as surgical landmark. The vertical crest separates macula cribrosa superior, series of very small openings through which terminal fibers of vestibular nerve pass in order to reach cupula of superior semicircular canal, from macula foramina, which marks point at which CN7 leaves internal auditory canal and enters its labyrinthine segment. Because most lateral portion of internal auditory canal is 4.8 mm deep to geniculate ganglion, labyrinthine segment of CN7 must take fairly vertically oriented course upward to reach it. The labyrinthine segment may be less than millimeter wide as it passes between cochlear and anterior end superior semicircular canal.

2) inferior portion of foramen is single oval-shaped space, anterior portion of which is occupied by rounded depression (tractus spiralis foraminosus) filled with small openings to accommodate terminal branches of cochlear nerve. The posterior portion is filled with macula crista inferior, through which pass terminal ends of inferior vestibular nerve.

TEMPORAL BONE

- laterally, irregular superior surface of temporal bone transitions relatively smoothly to temporal squama.

- free edge of tentorium and greater petrosal vein attaches to medially edge of superior surface of temporal bone. arterial meningeal artery and associated veins pass. A few millimeters anterior and lateral to foramen spinosum is identified in this area. It can then be followed retrograde to erups through superior surface of temporal bone at facial hiatus.

- facial hiatus is 4.8 mm anterior to geniculate ganglion. The greater superficial petrosal nerve can be identified in this area. It can then be followed retrograde to macula crista inferior.

- centimeter or so lateral to greater superficial petrosal nerve lies foramen spinosum, through which middle meningeal artery and associated veins pass. A few millimeters anterior and lateral to foramen spinosum is foramen ovale, which accommodates third (mandibular) division of trigeminal nerve. The horizontal portion of carotid canal courses through anterior temporal bone medial to foramen spinosum and foramen ovale.

- cochlea can be identified as anterior appearance of canal bone. It lies just anterior to labyrinthine segment of facial nerve but is deep to geniculate ganglion.

VESTIBULAR SCHWANNOMA (s. ACOUSTIC NEUROMA)

- benign intracranial extra-axial tumor that arises from Schwann cell sheath investing either vestibular or cochlear nerve.

N.B. it is schwannoma (not neuroma)

EPIDEMIOLOGY

- prevalence 0.7-1.0 cases per 100,000 population.

- 7% of all intracranial tumors.

- patients are 40-60 years of age.

- 5% are sporadic.

- bilateral tumors are pathognomonic for neurofibromatosis-II (tumors occur earlier).

PATHOLOGY

- vestibular division : auditory division = 3-20 : 1

- tumor originaes at pont where nerve acquires its reticulin and Schwann cell investment (in internal auditory foramen) - local erosion of internal auditory meatus.

- angle between cerebellum and pons grows slowly (2 mm/year) - can grow to substantial size before clinical symptoms (neurostructures can grow without compression).

- once tumor has grown sufficiently large, it continues growth by eroding bone or by spilling out into cerebellopontine angle (angle is relatively empty - tumors can continue to grow until 3-4 cm in size before they come in contact with important structures).

CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannoma (2017) Level 3 recommendation. What is the prognostic significance of KI-67 labeling index, proliferating cell nuclear antigen labeling index, degree of vascular endothelial growth factor expression, Antoni A vs B histologic patterns, mitotic figures, or other light microscopic features - no recommendation can be made due to a lack of adequate data.
Acoustic neuroma with displaced facial and cochlear nerves (nerves we are trying to preserve):

CLINICAL FEATURES

1. Early symptoms - ipsilateral neural hearing loss & tinnitus
   - hearing loss is in speech frequencies (earliest complaint - difficulties hearing conversations on telephone).
   - 3-5% patients have normal hearing at time of diagnosis.
   - mechanisms of hearing loss:
     a) direct injury to cochlear nerve → progressive hearing loss (85-95%)
     b) interruption of cochlear blood supply (labyrinthine artery compression by tumor) → sudden and fluctuating hearing losses (5-15%) - may improve spontaneously or in response to steroids!
     c) secretion of toxic substances by tumor cells (e.g. TNF-α)
   N.B. any asymmetric sensorineural hearing loss requires that acoustic neuroma be ruled out!
   unilateral tinnitus alone is sufficient reason to evaluate for acoustic tumor!

2. Dizziness & unsteadiness
   N.B. tumor grows slowly - central compensatory mechanisms can prevent or minimize vertigo - true vertigo is rare!!!
   - patients should be cautioned about underwater activities because of increased risks for disorientation!

3. Compression of cranial nerves as tumor grows large, it projects from internal auditory canal into cerebellopontine angle.
   • CN5 and later CN7 are affected - test CORNEAL REFLEX!
     N.B. facial weakness is rare (only 5-10% patients**) vs. hypesthesia of posterior external auditory canal (skin served by n. intermedius of CN7) - it is speculated that sensory fibers are more susceptible to pressure than motor axons
     *facial weakness associated with small or medium-size tumor should raise suspicion that it is not acoustic neuroma!
   • patients often are unaware of facial hypoesthesia.
   • less often, CN9 and CN10 are affected.

4. Compression of neuraxis
   • large tumor (≥ 4.0 cm) begins to compress cerebellum and pons → ataxia, hydrocephalus.

Source: Neurosurg Focus © 2013 American Association of Neurological Surgeons
## Diagnosis

**Audiologic Assessment**

differentiation from sensory hearing loss - see p. Ear32 >>

1. **Auditory** (to all patients to establish baseline) – severe impairment of speech discrimination, absent recruitment, marked tone decay (pathologic adaptation).


3. **BAER** (detects tumor before CT, but misses tumors in patients with excellent hearing) - increased latency of 3rd waveform, absence of waveforms, etc. see p. Ear30>>

N.B. in modern neuroimaging era, audiologic testing is no longer used for diagnosis, but provides pretreatment baseline?

### CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

#### Level 3 recommendation

- Screening MRI (for vestibular schwannomas) is recommended for adults with asymmetric audiograms:
  - a) ≥ 10 dB of interaural difference at ≥ 2 contiguous frequencies
  - b) ≥ 15 dB at 1 frequency

N.B. selectively screening patients with ≥ 15 dB of interaural difference at 3000 Hz alone may minimize the incidence of negative MRIs.

### CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

#### Level 3 recommendation

- Screening MRI (for vestibular schwannomas) is recommended for adults with asymmetric tumors (either purely unilateral tumors or bilateral tumors with subjective asymmetry) - this practice is low yielding in terms of vestibular schwannoma diagnosis (< 1%).

N.B. asymmetric tumors is a relatively unreliable screening tool for VS.

### CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

#### Level 3 recommendation

- Screening MRI (for vestibular schwannomas) is recommended for adults with sudden sensorineural hearing loss - this practice is low yielding in terms of vestibular schwannoma diagnosis (< 3%).

N.B. SSHL is better screening tool than tinnitus.

### AAO-HNS (American Academy of Otolaryngology-Head and Neck Surgery hearing classification)

- Class I (Gardner-Robertson hearing classification) grade of hearing.

### Vestibular Testing

#### Caloric Testing - marked vestibular hyporeactivity (circular paresis).

### Imaging

### CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

#### Initial Preoperative Evaluation - Level 3 recommendation:

- high-resolution T2 plus contrast-enhanced T1 MRI.
- standard T1, T2, FLAIR, and DWI sequences obtained in axial, coronal, and sagittal plane may be used for detection of vestibular schwannomas.
- adults with proven or suspected vestibular schwannomas by imaging - T2-weighted MRI (CISS/FIESTA or DTI) may be used to augment visualization of the facial nerve course as part of preoperative evaluation.
- adults with vestibular schwannomas should be counselled that their tumors may more often be associated with rapid growth, lower rates of complete resection, and facial nerve outcomes that may be inferior in the immediate postoperative period but similar to noncystic schwannomas over time.
- degree of lateral internal auditory canal involvement by tumor adversely affects facial nerve and hearing outcomes.

### CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

#### Surveillance (for growth of a vestibular schwannoma) - Level 3 recommendation – either:

- postcontrast 3D T1 MPRAGE (magnetization prepared rapid acquisition gradient echo).
- high-resolution T2 (including constructive interference in steady state [CISS] or fast imaging employing steady-state acquisition [FIESTA] sequences)
- MRIs should be obtained annually for 5 yr, with interval lengthening thereafter with tumor stability.

### CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

#### Postoperative Evaluation - Level 2 recommendation:

- postcontrast 3-D T1 MPRAGE – nodular enhancement considered suspicious for recurrence.
- for gross total resection – postoperative MRI to document the surgical impression may occur as late as 1 yr after surgery.
- for near-total / subtotal resection - more frequent surveillance scans are suggested; annual MRI scans may be reasonable for 5 yr. Imaging followup should be adjusted accordingly for continued surveillance if any change in nodular enhancement is demonstrated

### CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

#### Vestibular schwannomas associated with NF2 - Level 3 recommendation - should be imaged similar to sporadic schwannomas with the following cavers:

1. More frequent imaging may be adopted - because of a more variable growth rate, then annual imaging may ensue once the growth rate is established.
1. **X-ray** – enlarged internal auditory canal (“trumpet-shaped”).

2. **MRI** with gadolinium enhancement – definitive diagnostic test!!!
   - tumor has intense enhancement!
   - can demonstrate 1-2 mm tumors (CT may miss tumors < 1.1.5 cm).
   - if MRI is contraindicated but suspicion is high → air-contrast cisternography (high sensitivity - can detect relatively small intracanalicular tumors).

3. **Gas CT cisternography** (injected intrathecal gas is maneuvered to outline CN8) can demonstrate small tumors within IAC that are typically missed on routine CT, invasive and less sensitive than MRI.

**Kooos grading:**
- stage I – small intracanalicular tumor
- stage II – small tumor with protrusion into cerebellopontine angle and no contact with brainstem
- stage III – tumor occupying cerebellopontine cistern with no brainstem displacement
- stage IV – large tumor with brainstem and cranial nerve displacement

**Classification depending on location and brainstem compression:**
- Type A — intracanalicular.
- Type B — cerebellopontine angle.
- Type C — compressing brainstem.
- Type D — deviation of fourth ventricle.

**MRI** – mass impinging upon cerebellum from cerebellopontine angle:

Magnified view:
TREATMENT ALGORITHM

A) surgical excision – mainstay of treatment (esp. for larger tumors! ← may require adjuvant radiotherapy), most cost effective for patients < 45 yrs (vs. > 45 yrs – SRS)

Dade Lunsford: surgery only for symptomatic mass effect on brainstem!

CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

Small intracanalicular tumors (< 1.5 cm) - insufficient data to support a firm recommendation that surgery be the primary treatment.

Small intracanalicular tumors (< 1.5 cm) and good preoperative hearing - Level 3 recommendation: hearing preservation surgery via the middle fossa or the retrosigmoid approach may be attempted.

Patients with NF2 - insufficient evidence that surgical resection should be the initial treatment.

There is insufficient evidence that a multidisciplinary team (neurosurgeon + neurologist) provides superior outcomes compared to either subspecialist working alone.

* complete resection and facial/vestibulocochlear nerve preservation

Does a subtotal surgical resection of a VS followed by stereotactic radiosurgery (SRS) to the residual tumor provide comparable hearing and FN preservation to patients who undergo a complete surgical resection?

There is insufficient evidence if subtotal resection followed by SRS provides comparable hearing and CN7 preservation vs. complete surgical resection.

There is insufficient evidence to support either surgical resection or SRS for treatment of preoperative balance problems.

Level 3 recommendation: surgical resection may be used to better relieve trigeminal neuralgia than SRS.

Level 3 recommendation: microsurgical resection after SRS - increased likelihood of a subtotal resection and decreased CN7 function.

B) stereotactic radiotherapy – better (than surgery) preservation of CN5, CN7, hearing

N.B. radiosurgery is still a "hearing preservation approach" (patient is counseled that likelihood of hearing loss is similar to continued observation).

Indications and CNS guidelines >>

C) careful serial observation (15-40% ultimately require therapeutic intervention): see above for MRI requirements (sequences and timing) >>

1) short life expectancy (elderly patients, severe medical conditions)
2) small tumors (esp. if hearing is good)
3) tumor on side of only hearing ear or only seeing eye

N.B. during observation period, most (> 70%) patients who are initially eligible for hearing conservation surgery lose their eligibility!

Dade Lunsford: in 4.4 yrs tumor volume doubles – observation only for elderly!

CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

Patients with NF2 without surgical options:

Level 3 recommendation: BEVACIZUMAB is recommended in order to radiographically reduce the size or prolong tumor stability, to improve hearing or prolong time to hearing loss.

- bevacizumab has made the most progress and appears to be a viable treatment option for patients with NF2 and growing tumors or loss of hearing. In these patients, bevacizumab recovers some useful hearing function and results in tumor reduction; however, the effect is ultimately lost with time succumbing to the natural tendency of the tumor to grow.

Level 3 recommendation: LAPATINIB may be considered.

- dual tyrosine kinase inhibitor which interrupts the HER2/neu and epidermal growth factor receptor (EGFR) pathways

Level 3 recommendation: ERLOSTINIB is not recommended.

Level 3 recommendation: ENVERTILIMUS is not recommended

Any patient undergoing observation - Level 3 recommendation: ASPIRIN may be considered (to augment inflammatory response)

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data demonstrates that consistent and durable hearing preservation in sporadic VSs remains an elusive goal. Most patients eventually develop nonoserveable hearing as a result of disease or treatment. Radiofrequency brainstem implants (ABI) are used successfully in hearing loss secondary to vestibular schwannomas.

Radiosurgery Practice Guideline for Vestibular Schwannomas (Guideline Report 84-06, original guideline 2006):

Algorithm for management of incident VS

- 

TREATMENT - SURGERY

see MRI features important for surgery planning and outcomes

Approach Main approaches:
1) retrosigmoid (suboccipital) approach: for small tumors with minimal extension into IAC (less than 1 of proximal IAC with predominantly CPA component) when patient desires hearing preservation
2) translabyrinthine approach
3) middle cranial fossa approach: for small intrameatal tumors ≤ 1.5 cm

- subtotal resection → adjuvant radiotherapy, see below

facial nerve function preservation is the most important goal: fortunately CN7 is in continuity at end of most resections (any postoperative paresis tends to be temporary)
  - if facial nerve is divided during surgery, it is sutured together (or nerve graft is placed between stumps)
  - facial paralysis with no recovery within few months → surgical reinnervation
- tumor blood supply is from dura of porus acusticus.
- CN7 is usually anterior to tumor (so nerve is hidden).

Approach selection
1) patient has no useful hearing:
   a) translabyrinthine approach (preferred)
   b) retrosigmoid approach (for tumors with significant inferior extension)
   - *30/70 rule: individuals with pure-tone average > 50 dB and speech discrimination < 50% do not have useful or salvageable hearing; other surgeons use 30/70 rule.
2) patient has useful hearing:
   a) retrosigmoid approach (for tumors that have significant volume medial to porus acusticus).
b) middle cranial fossa approach (for tumor within lateral portion of internal acoustic canal, esp. small intracanalicular tumors):  
- normal preoperative BAER favors hearing conservation.  
- abnormal electroneurogram favors hearing conservation (ENG tests horizontal semicircular canals, which are innervated by superior vestibular nerve; normal ENG = superior vestibular nerve is normal - tumor must have originated from inferior vestibular nerve, which is directly adjacent to cochlear nerve).

3) anatomic variations that make translabyrinthine approach difficult/impossible:  
- high-riding jugular bulb (above level of inferior internal auditory canal).  
- anteriorly placed sigmoid sinus (distance between sigmoid sinus and external auditory canal few millimeters or less - limited space for surgeon to operate).  
- reduced/absent flow in contralateral sinus (injury to remaining sinus → catastrophic venous infarction).  
- contracted semicircular mastoid (little room for tumor removal + often associated with supplicative otitis media).

CNS Systemic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2012)  
What surgical approaches are best for long-term function and facial nerve preservation:  
- serviceable hearing is present - insufficient evidence to support the superiority of either the middle fossa or the retrosigmoid approach.  
- serviceable hearing is not present - insufficient evidence to support the superiority of either the retrosigmoid approach or translabyrinthine approach.

CNS Systemic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)  
Size of tumor - Level 3 recommendation: Patients with larger tumor size should be counseled about the greater than average risk of loss of serviceable hearing.

Endoscopic assistance - Level 3 recommendation: surgeon may choose it in order to aid in visualization.

Radiotherapy Practice Guideline for Vestibular Schwannomas (Guideline Report #4-06, original guideline 2006)  
Preservation of facial function continues to improve. However, facial nerve outcomes vary according to tumor size and operator experience. When tumors are smaller than 1.5 cm, good facial nerve function can be expected (House-Brackmann grades I-II) in more than 90% of patients at Centers of Excellence. Only 3-2.6% of patients with this size tumor have poor outcomes (House-Brackmann grades III-IV). In addition to tumor size, preoperative electroneurologic testing can help predict postoperative outcome, although this testing is not commonly used. The overall facial nerve preservation rate is 90% if the facial nerve function (grades I-III) can be preserved in only 40-50% of patients with large (>4 cm) tumors. Injuries of the nerve intermixed with this nerve has been assessed preoperatively. The ability to preserve hearing has increased substantially over the last couple of decades. Depending on criteria used for successful hearing conservation, hearing preservation has been reported in 30-80% of patients following cochlear nerve monitoring surgery. Meta-analysis performed by Gardiner and Robinson showed an overall average success rate of about 35%. Delayed hearing deterioration may occur after surgery in 30-50% of patients who originally had successful hearing preservation. In various series, serviceable hearing preservation rate is 57% using the retrosigmoid approach and 32-68% using the middle fossa approach have been reported.

Tinnitus becomes worse in 6-20% of individuals after tumor removal. In the majority of patients, tinnitus remains unchanged or improves by approximately 25-50% of patients experience a decrease in tinnitus. Of patients without preoperative tinnitus, 30-50% developed it in the immediate postoperative period.

Preoperative  
Level 3 recommendation: peripherally induced ENG (or with the addition of HYDROXYETHYL STARCHS) should be considered to improve postoperative outcomes.

Level 3 recommendation: preoperative vestibular rehabilitation is recommended to aid in postoperative mobility after surgery.

Level 3 recommendation: preoperative ENTABILATION ablation of the vestibular apparatus should be considered to improve postoperative mobility after surgery.

Iatrogenic Facial Nerve Injury

CNS Systemic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)  
Level 3 recommendation: intraoperative CO2 laser is not recommended during surgery to improve postoperative facial nerve function.

Level 3 recommendation: intraoperative facial nerve monitoring can be used to accurately predict long-term facial nerve function after surgery.  
- the presence of favorable testing reliably portends a good long-term facial nerve outcome.  
- the absence of favorable testing in the setting of an anatomically intact facial nerve does not reliably predict poor long-term outcome (function cannot be used to direct decision-making regarding the need for early reinervating procedures).

N.B. poor intraoperative EMG electrical response of the anatomically intact facial nerve should not be used as a reliable predictor of poor long-term facial nerve function

CNS Systemic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)  
Patients with measurably hearing and tumors < 1.5 cm - Level 3 recommendation: intraoperative cochlear nerve monitoring should be used during surgery when hearing preservation is attempted. Insufficient evidence if direct monitoring of the eighth cranial nerve is superior to far-field auditory brainstem responses.

- Level 3 evidence supports the use of intraoperative cochlear nerve monitoring in hearing preservation VS surgery. The most common method employed was auditory brainstem response. The responses and the interpretation of results.
- The presence or characteristics of Wave I and V, as well as the cochlear compound action potential (CAP), were the most useful parameters discussed. The biggest challenge with neuromonitoring of the cochlear nerve involves the technical aspects and delayed feedback. Auditory brainstem responses are plagued by the challenge of getting the right due to the data gap that is required to assess changes in function. To circumvent this, direct cochlear nerve monitoring has been used instead. The technical requirements and challenges of performing direct cochlear nerve monitoring, however, were made apparent in various reports. They range from the inability to place electrodes at the nerve root exit zone prior to tumor resection, to the difficulty in keeping the probes in place throughout the duration of surgery or securing the probe without causing iatrogenic damage to the nerve. Finally, factors such as the presence of excess cerebrospinal fluid or blood, the stimulation voltage used to elicit responses, or the interference of electrocorticogram stimuli all have been reported to alter responses and the interpretation of results.

Intra-operative Monitoring

of facial and vestibulocochlear nerve - drastic reduction in morbidity.  
- facial nerve – EBAF  
- vestibulocochlear nerve – BAEF, ECOG, and deep CNAPs.

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- Level 3 evidence supports the use of intraoperative cochlear nerve monitoring in hearing preservation VS surgery. The most common method employed was auditory brainstem response.
- The responses and the interpretation of results.
- The presence or characteristics of Wave I and V, as well as the cochlear compound action potential (CAP), were the most useful parameters discussed. The biggest challenge with neuromonitoring of the cochlear nerve involves the technical aspects and delayed feedback. Auditory brainstem responses are plagued by the challenge of getting the right due to the data gap that is required to assess changes in function. To circumvent this, direct cochlear nerve monitoring has been used instead. The technical requirements and challenges of performing direct cochlear nerve monitoring, however, were made apparent in various reports. They range from the inability to place electrodes at the nerve root exit zone prior to tumor resection, to the difficulty in keeping the probes in place throughout the duration of surgery or securing the probe without causing iatrogenic damage to the nerve. Finally, factors such as the presence of excess cerebrospinal fluid or blood, the stimulation voltage used to elicit responses, or the interference of electrocorticogram stimuli all have been reported to alter responses and the interpretation of results.
**TRANSLABYRINTHINE APPROACH**


**Advantages**

1. **Best view** of lateral brain stem facing acoustic tumor.
2. Fundus and lateral end of internal auditory canal are **completely exposed**; CN7 can be identified at location where it is undistorted by tumor.
3. **Cerebellar retraction** is almost never necessary.
4. **Juxtaposition of CSF leak**.
5. **Allows resection of CN7 continuity** (if CN7 has been divided or sacrificed) by rerouting CN7 and performing primary anastomosis (interposition graft can sometimes be avoided).

The translabyrinthine approach provides the best view of the lateral brain stem facing the vestibular schwannoma. • Retraction of the cerebellum is not needed. • The fundus and lateral end of the internal auditory canal are completely exposed; the facial nerve can be identified at a location where it is undistorted by tumor growth and compressed into the labyrinthine segment, decreasing the risk of delayed postoperative facial nerve palsy. • Possible opening of the tentorium in case of an extra large tumor that may extend upward. • Safe control of the lateral surface of the pons. Improved surgical comfort. • No need for a seated or lateral positioning of the patient.

**Disadvantages**

1. Complete **hearing sacrifice** is unavoidable.
2. **Worse visualization** (comparing with retrosigmoid approach) of inferior portions of cerebellopontine angle, cranial nerves, temporal bone anterior to porus acusticus.
3. **Fat graft** (from abdomen) is required.
4. Sigmoid sinus is more vulnerable to injury (bleeding is difficult to control).
5. High jugular bulb or anteriorly placed sigmoid sinus can substantially compromise space available for tumor removal.

Hearing sacrifice is complete and unavoidable. • The inferior portions of the cerebellopontine angle and cranial nerves are not as well visualized as they are in the retrosigmoid approach. The temporal bone anterior to the porus acusticus is also less well visualized. • A tissue graft (fat, fascia, homograft or synthetic dura) is required. Some surgeons prefer hydroxyapatite cement. • The sigmoid sinus may be more vulnerable to injury. • Need to extend the approach in case of contract mastoid or high jugular bulb position. • The third portion of the facial nerve may be vulnerable to injury during the approach. • The approach requires additional time.

**Operative details**

- Small incision behind ear.
- Petrous bone is gradually removed with high-speed drill until facial nerve is identified and exposed to point where it can be separated from tumor and protected.
- Bone that must be removed is indicated in yellow (tumor is in orange).

- Dura of posterior fossa is seen easily - can be opened to access to intradural component of tumor.
- Large bone and dural defects are closed by autogenous free fat graft.

**RETROSIGMOID APPROACH**

The retrosigmoid approach provides the best widefield visualization of the posterior fossa. The time needed for tumor exposure is shorter. The inferior portions of the cerebellopontine angle are frequently exposed to the posterior surface of the temporal bone anterior to the porus acusticus are more clearly observed than in transmastoid or translabyrinthine approach. Panoramic visualization is especially helpful when displacement of nerves is not predictable, especially when combined with microsurgical techniques. • Hearing conservation surgery can be attempted even for relatively large tumors through the retrosigmoid approach. Destruction of the labyrinth is not required as part of the retrosigmoid approach.

Disadvantages

1) Highest incidence of tumor recurrence / persistence.
2) May require cerebellar retraction or resection (<== edema, hematomata, infarction).
3) Incidence of CSF leak.
4) Severe protracted postoperative headache – can be diminished with intraoperative monitoring techniques and use of hydroxyapatite or methylmethacrylate (eliminates direct attachment of posterior fossa dura to bone).

Advantages

1) conquest of nerves is not predictable.
2) panographic visualization is especially helpful when displacement of nerves is not predictable, especially when combined with microsurgical techniques. • Hearing conservation surgery can be attempted even for relatively large tumors through the retrosigmoid approach. Destruction of the labyrinth is not required as part of the retrosigmoid approach.

Operative details

- operation that fully exploits surgeon's microsurgical skill.
- monitoring – bilateral BAER, ipsilateral facial nerve EMG
- patient position
  a) supine turn head toward contralateral shoulder
  b) true lateral position (Scalp/Ear) permits occiput to be rotated little more bit laterally – superoingly – slightly more direct view of internal auditory canal.

RETROSIGMOID EARRING – see p. Op300
Bone that must be removed is indicated in yellow (tumor is in orange):

The retrosigmoid approach may require cerebellar retraction or resection, which can lead to the development of postoperative edema, hematomata, infarction and bleeding. • The retrosigmoid approach is associated with a greater likelihood of protracted postoperative headache. • The highest incidence of tumor recurrence or incomplete resection occurs with retrosigmoid approaches due to poor control of the fundus of the internal auditory canal. • It may be difficult to perform a facial-to-facial nerve graft repair from this approach.

- dura is opened in T-shape, atrachedoid incised, and cerebellum is retracted medially to expose cerebellopontine angle (cerebellopontine angle frequently falls away from posterior surface of temporal bone; hyperventilation, steroidal antiinflammatory medication, and early cerebellar retraction with limited amount of retraction. Nonetheless, gentle cerebellar retraction is frequently necessary).
- drill off posterior wall of LAC (endolymphatic sac is the landmark how posteriorly we can drill).
- once adequate exposure has been obtained, tumor is clearly visualized along with brain stem and lower cranial nerves. However, cranial nerves V and VIII are rarely observed because they are almost always already retracted forward and lateral. The superior surface of tumor is visualized.
- debulking of tumor is next step and must be carefully performed so as to maintain anterior portions of capsule if injury to cranial nerves VII and VIII is to be avoided, meize tumor capsule longitudinally (along long axis of CN7) and debulk with pituitary rongeur.
- once tumor has been substantially debulked, posterior wall of internal auditory canal is removed:
  - discute dura from posterior surface of petrous
  - high-speed drill (irrigating drill, diamond drill bit)
  - bone dust should be prevented from entering subarachnoid space during this phase of procedure. The only visible cause for severe and untreatable postoperative headache is spillage of bone dust into subarachnoid space during tumor removal. Carefully place Surgicel, pieces of Gelfoam. Telfa pads, and/or cottomoid strips around operative site so that bone dust from drilling adhes to them and is removed as they are removed.
  - great care must be taken to avoid injuring labyrinth while removing posterior wall of internal auditory canal. Portions of labyrinth quite commonly are medial to lateral end of internal auditory canal. Although no single anatomic landmark is completely reliable for prevention of injury to labyrinth, singular nerve and its capsule, vestibular aqueduct, and cochlear aqueduct are all used as important surgical landmarks. Careful measurements taken from preoperative CT scans can provide useful information during drilling of posterior wall canal
  - length of internal auditory canal varies considerable from individual to individual, and knowing exactly how much posterior canal wall needs to be removed to adequately expose tumor can help limit inadvertent injury to labyrinth. Blind extraction of tumor from internal auditory canal without removing posterior wall prevents significant risks of injury to facial and/or auditory nerve (if hearing is to be saved) while at same time increasing chances of leaving tumor at fundus. Use of intraoperative angled endoscopes has been of considerable help in performing this phase of procedure.
  - once internal auditory canal is exposed, dura is opened and tumor is removed from it. Although never proven, disconnection from medial to lateral is thought to be less traumatic to both cochlear nerve and to vascular supply of tumor capsule. These nerves are usually detached, and unless hearing is to be preserved, cochlear nerve is sacrificed as well. Eventually, surgeon is left with anterior portions of capsule adhered to brain stem and posterior wall of internal auditory canal. Tumors as capsule is carefully removed from brain stem to entry zone of cranial nerve VII can be identified. The capsule is then carefully removed from facial nerve with as little trauma as possible.
  - facial nerve monitor is placed in this portion of dissection. Technique of various has been used to monitor cochlear nerve when hearing preservation is desired. The most commonly used method is intraoperative ABR audiometry, but it has number of drawbacks. Most importantly, it requires summing large number of repetitions in order to extract response from background noise. Consequently, delay occurs between surgical manipulations and ABR changes. Direct cochlear nerve monitoring offers advantage of real-time feedback, but fully satisfactory method of placing and securing electrode still is lacking.
  - once tumor removal is complete and hemostasis is absolute, dura is closed and craniotomy defect is repaired – see p. Op300.

MIDDLE CRANIAL FOSSA APPROACH

Advantages

1) fully exposes lateral third of internal auditory canal without sacrificing hearing (some surgeons reach tumors that extend 0.5-1.0 cm into cerebellopontine angle)

-S.N.B. better hearing preservation than retrosigmoid!
It is the only procedure that fully exposes the lateral third of the internal auditory canal without dural laceration.

**Subtotal resection** may be injured during the elevation of the temporal dura.

**trismus related to manipulation and/or injury to the temporalis muscle.**

**The temporal lobe** may be splayed out over anterior superior portions of the petrous bone. A temporal emissary (approximately 3 cm by 3 cm) is performed with its base at root of zygoma. The dura is elevated up from floor of middle cranial fossa, and osseous dura, head elevation, hyperventilation, and steroids are used to limit cerebral edema.

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**dura of temporal lobe** is then elevated off superior surface of temporal bone. The anterior extent of such elevation usually comes from the anterior clinoid, but middle dural elevation can be divided horizontally if elevation continued anteriorly to foramen ovale if additional exposure is desired. Dural elevation should proceed from posterior to anterior to avoid injury to exposed superficial petrosal nerve or geniculate ganglion. Bleeding from veins associated with middle meningeal artery is often quite brisk but can generally be controlled with Surgicel packing. Medial dissection continues to free edge of temporal lobe.

**superior petrosal sinus** is at posterior surface of temporal bone but not always at its superior edge. Care must be taken to avoid injuring it. If inadvertent injury occurs, bleeding can generally be controlled with wound pressure, Surgicel packing, electrocautery, or hemoclips. When extended middle cranial fossa approach is employed (for tumors that extend centimeter or more outside porus acusticus into cerebellopontine angle), superior petrosal sinus is deliberately divided between clips and tentonism is opened.

**dura** is tightly that integrity of facial nerve is being compromised. If greater superficial petrosal nerve cannot be located and no other landmarks are available, middle ear space can be entered from petrotympanic fissure, and then it can be detected easily, arcuate eminence is extremely helpful landmark. Careful drilling can often identify blue line of superior canal within it. Because most difficult exposure to achieve during middle fossa surgery is lateral posterior end of internal auditory canal, perform dissection as close to superior semicircular canal as possible. The greater superficial petrosal nerve is generally easy to visualize and can be followed retrogradely to geniculate ganglion. It lies less than 1 cm directly medial to foramen spinosum. Once area of geniculate is identified, superior petrosal sinus is deliberately divided. Care must be taken to avoid injury to exposed greater superficial petrosal nerve or geniculate ganglion. Bleeding from veins associated with middle meningeal artery is often quite brisk but can generally be controlled with Surgicel packing. Medial dissection continues to free edge of temporal lobe.

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**POSTOPERATIVE**

**ICU overnight (rapid intervention if ICP increases or bleeding occur).**

**vestibular rehabilitation** begins on first postoperative day and continues twice daily throughout hospital stay.

**MRI** is of superior or inferior vestibular nerve origin, the facial nerve may have to be retracted → temporal lobe injury

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**Follow-up MRI** (completeness of tumor removal) within 3-6 months → at 5 years → at 10 years (if all are normal, further imaging is performed only if clinical circumstances require so).
N.B. distinguishing tumor recurrence from postoperative inflammation (both are MRI enhancing) can be quite difficult; recurrences tend to be global while inflammatory enhancement tends to be linear.

- recurrence rate is ≤ 5-10% (vast majority of recurrences follow subtotal removal - tumor left in lateral end of internal auditory canal where intraoperative visualization is so difficult).

**COMPLICATIONS**

1. Injury to AICA (occurs very rarely).
   - AICA may be loosely attached to tumor capsule, but separating it from tumor is generally fairly easy.
   - sacrificing AICA itself has variable consequences (can lead to devastating neurologic injury or death).
   - AICA branches most vulnerable to injury (Robertson hearing classification) grade I hearing at 5 yr, and at 10 yr.

2. Occlusion of sigmoid sinus
   - If contralateral venous outflow is patent and communication through tectorial is good, complete occlusion may go entirely unnoticed.
   - occlusion of dominant sinus can result in catastrophic ICP↑, venous infarction, and even death.

3. Occlusion of petrosal vein of Dandy (single large venous tract in some patients; series of large veins in others) → edema, temporal lobe or brain stem infarction.

4. Occlusion of vein of Labbé (→ severe edema (→ brain herniation) and temporal lobe infarction;
   - vein of Labbé is not directly in field during acoustic tumor surgery, but injury to superior petrosal sinus may result in vein obliteration.

5. Hemorrhage into posterior fossa → brainstem compression → death within minutes.

   - Rapid neurologic deterioration in first 24 postoperative hours → suspect posterior fossa hemorrhage:
     - if time permits, rapid unenhanced CT while operating room is prepared.
     - if deterioration is rapid, forfe CT and take patient directly back to operating theatre.
     - if deterioration is very rapid (with loss of consciousness, decerebrate posturing, and signs of imminent death), open wound at bedside (posterior fossa decompression) prior to emergent translocation to operating room.

6. Injury to cerebellum (incidence dramatically diminished in recent decades) → edema → obstruction of 4th ventricle, brainstem compression.

   - Rotating shaft of surgeon’s burr is often culprit (surgeons usually look past shaft to head of burr to control bone removal; shaft is often outside surgical field of view).

   - other causes - compression and retraction, alteration of arterial inflow, venous engorgement.

   - treatment - osmotic diuretics, hyperventilation, steroids (resection of involved cerebellar hemisphere part may be required).

7. Postoperative facial paralysis (sometimes unavoidable). about management - see p. CN7

   - prophylactic perioperative steroids are widely used (but unequivocal evidence for effectiveness is lack).
   - tumor < 1.5 cm - good facial nerve function can be expected (House-Brackmann grade I-II) in > 90% of patients.
   - patients with poor facial nerve function (House-Brackmann V-VI) at time of discharge have 25% chance of recovery of normal function.

8. 30-50% cases have onset delayed for few hours ÷ week (vasospasm, vascular injury, traction, nerve stretching, even viral reactivation have been proposed):
   - risk factors for delayed facial palsy: gross total tumor resection, retrosigmoid approach
   - vast majority make complete and total recoveries within 1 month (steroids, antivirals make no difference in recovery).


10. Bacterial meningitis → immediate LP (after CT has excluded significant hydrocephalus - could lead to herniation) → antibiotics immediately.

11. Aseptic meningitis; corticosteroids are extremely helpful.

12. CSF leak through:
   - about management – see p. S64 >>
   - a) wound
   - b) eustachian tube.

13. Severe postoperative headache: may be treated nonoperatively (disadvantages)

   - H: high-dose NSAIDs, aggressive manipulative physical therapy.

14. Tinnitus (6-20% patients)

15. Vertigo – improves over several days; it is predicted if patient never had in a history the episode of severe vertigo (if patient had it in the past and it went away, it most likely means that vestibular system is already nonviable); prophylaxis – never leave one division of vestibular nerve intact (i.e. if superior division got damaged, then cut also inferior division).

**OUTCOMES**


CSS Somatic Review: and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

The overall probability of maintaining serviceable hearing following microsurgical resection of small to medium-sized sporadic vestibular schwannomas - Level 3 recommendation: moderately low probability (25-50%) of hearing preservation immediately following surgery, at 2 yr, at 5 yr, and at 10 yr.

**Subset of patients with AAO-HNS (American Academy of Otolaryngology-Head and Neck Surgery hearing classification) class A or GR (Gardner-Robertson hearing classification) grade I at baseline - Level 3 recommendation**:

- moderately high probability (50-75%) of hearing preservation immediately following surgery, at 2 yr, at 5 yr.
- moderately low probability (25-50%) of hearing preservation at 10 yrs.
The most consistent prognostic features associated with maintenance of serviceable hearing - Level 3 recommendation:

1) Good preoperative word recognition
2) Pure tone thresholds with variable cut-off points
3) Smaller tumor size commonly less than 1 cm
4) Presence of a distal internal auditory canal cerebrospinal fluid fundal cap.

Age and sex are not strong predictors of hearing preservation outcome.

TREATMENT - STEREOTACTIC RADIOTHERAPY

N.B. Acoustic neuromas are not treated with traditional external beam radiotherapy because they are radioresistant and because of proximity with brainstem.

FRACTIONATED
- Better hearing preservation (than single-session SRS)
- Fewer cranial nerve complications!
- Fewer cranial nerve complications!!!
- For LINAC, dose is divided into 3-5 sessions, typically prescribed to the 80% isodose line, using a total mean dose ranging up to 17 Gy.

SRS

Benign tumor as a target for SRS – see Onc26 >
- Can be performed using the Gamma Knife®, modified LINACs or the proton beam.

INDICATIONS
N.B. Brainstem compression is not improved by radiation – use surgery instead!
1) New / residual / recurrent tumor
2) Intracanalicular tumors and small to medium-size tumors without brainstem compression* and without signs of hydrocephalus (if hydrocephalus is present in old or infirm patients, a shunting procedure should be considered in addition to SRS).
3) Oldest patients (conservative treatment may be indicated because of slow growth)
4) Other significant medical problems
5) Only-hearing ear
6) Bilateral tumors (treat larger tumor when symptoms absolutely require)

*Koos IV tumors should be offered microsurgery as first management.

METHODOLOGY

CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2012)

CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

Dose - Level 3 recommendation: There is no difference in radiographic control using different doses - it is recommended that for single fraction SRS ≤ 13 Gy be used to facilitate hearing preservation and minimize new onset or worsening of preexisting cranial nerve deficits.

Number of fractions - no recommendations: There is no difference in radiographic control and clinical outcome using single or multiple fractions.

13±2±3 Gy to 50% or other isodose line that conforms to the tumor margin (historically, 16-20 Gy)

- Lower radiosurgery doses may be a better for patients with bilateral NF2 vestibular schwannomas or patients with contra-lateral deafness from other causes, for whom hearing preservation may be more critical.

- Complete coverage of the tumor and preservation of brainstem, facial, cochlear and trigeminal nerve function is given priority during dose planning.

- N.B. success depends upon high conformity to the tumor margin!

- Planning includes outlining of tumor volume, use of multiple isocenters, beam weighting and use of plug patterns.

- Combination of small beam diameter (4- and 8-mm) collimators; for large tumors, 16 mm collimators are also used.
• series of 4 mm isocenters are used to create a tapered isodose plan to conform to the intracranial portion of the tumor.
• facial-cochlear nerve complex generally courses along the anterior margin and anterior-inferior side of the tumor - the dose plan should by highly conformal in this region.
• treatment isodose, maximum dose and dose to the margin are jointly decided by a neurosurgeon, radiation oncologist, medical physicist and, in some centers, a neurotologist after considering the goal of radiosurgery in an individual patient and the tolerance of the surrounding structures.
• the fall off on cochlea, semicircular canal and brainstem are checked to keep them below tolerance level.

STRESSORS
• there is no consensus on the use of corticosteroids on the day of radiosurgery.
• some do not use steroids at all before, at during or after radiosurgery.
• some give IV 40 mg of methylprednisolone at the onset or conclusion of the procedure. – Cleveland Clinic: 1 dose of methylprednisolone at the end of procedure; Rx of Medrol Dosepak PRN (if patient develops symptoms)
• at other centers, 6 mg of dexamethasone is given immediately before dose delivery and is repeated every three hours for the duration of the treatment.

FOLLOW UP
CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)
Level 3 recommendation: follow-up imaging should be obtained at intervals after SRS to detect recurrence. No recommendations can be given regarding the interval of these studies.

Progression of tumor after SRS – Level 3 recommendation: SRS can be safely and effectively performed as a retreatment.

Level 3 recommendation: patients should be informed that there is minimal risk of malignant transformation of vestibular schwannomas after SRS.

NF2 – Level 3 recommendation: SRS is a treatment option for tumors that are enlarging and/or causing hearing loss.

MRIs at 6 months, 12 months, and 2, 4, 8 and 16 years.
- little change is detected over first 6 months.
- during initial 12 months, tumor may exhibit initial edema, but this declines over time (H: steroids or venulocapilloternal shunting).
- by 12 months, follow-up MRI shows loss of central tumor enhancement (indicates favorable response).
- 4–6% of tumors show postsurgical progression (tumor swelling) for 6–12 months – face may twitch – it is temporary.
- by 2 years, partial involution of tumor is noted.
- stereotactic radiation could completely eradicate tumor, but in most patients, residual tumor is noted on MRI (remains stable for long periods = local control).
- all patients who have some preserved hearing are advised to obtain audiological tests (PTA and SDS) near the time of their MRI followups.

RESULTS
Recent reports suggest a tumor control rate of 93–100% after SRS; pre-radiosurgery hearing can be preserved in 60–70% of patients (higher preservation rates for smaller tumors); facial and trigeminal nerve function can be preserved in > 95% of patients. Multiple meta-analyses reveal that from the patients’ perspective, SRS provides a more desirable outcome than microsurgery!

CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)
Overall probability of maintaining serviceable hearing following SRS – Level 3 recommendation: moderately high probability (> 50–75%) of hearing preservation at 2 yr and 5 yr moderately low probability (> 25–50%) of hearing preservation at 10 yr.

Subset of patients with AAO-HNS (American Academy of Otolaryngology-Head and Neck Surgery hearing classification) class A or GR (Gardner-Robertson hearing classification) grade I hearing at baseline - Level 3 recommendation: high probability (> 75–100%) of hearing preservation at 2 yr moderately high probability (> 50–75%) of hearing preservation at 5 yrs moderately low probability (> 25–50%) of hearing preservation at 10 yrs.

AAO-HNS Gardner-Robertson

Class A

<table>
<thead>
<tr>
<th>Discrimination</th>
<th>Class I (good)</th>
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<tbody>
<tr>
<td>PTA ≤ 30 dB</td>
<td>Discrimination 70–100 %</td>
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Class B

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<th>Class II (serviceable)</th>
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<tbody>
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<td>PTA ≤ 50 dB</td>
<td>Discrimination &gt; 50–69 %</td>
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Class C

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<th>Discrimination</th>
<th>Class III (unserviceable)</th>
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<tr>
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<td>Discrimination ≤ 50 %</td>
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Any PTA

<table>
<thead>
<tr>
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<th>Class IV (poor)</th>
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<tbody>
<tr>
<td>Class IV (poor)</td>
<td>Discrimination 1–4 %</td>
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</tbody>
</table>

Class V (none)

| PTA > 91 dB | Discrimination 0 % |

PNT none

CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)
The most consistent prognostic features associated with maintenance of serviceable hearing - Level 3 recommendation:
1) good preoperative word recognition
2) pure tone thresholds with variable cut-points
3) smaller tumor size
4) marginal tumor dose ≤ 12 Gy
5) cochlear dose ≤ 4 Gy

Age and sex are not strong predictors of hearing preservation outcome.

Dade Lunsford:
- tumor control.
SCHWANNOMAS OF CRANIAL NERVES

- **6-year actuarial rates for preservation (12-13 Gy):**
  - Facial nerve function > 99%
  - Trigeminal nerve function 95.6 ± 1.8%
  - Facial nerve preservation 100%

- **13 Gy tumor margin dose** → 0% risk of new facial weakness and 3.1% risk of facial numbness (5-year actuarial rates); margin dose of > 14 Gy → 2.5% risk of new onset facial weakness and a 3.9% risk of facial numbness (5-year actuarial rates).

- Prior to 1991, tumor margin dose was 18-20 Gy preserving: 79% facial function at 5 yrs, 73% trigeminal function at 5 yrs.

- Unlike microsurgery, immediate hearing loss is uncommon after SRS; if hearing impairment is noted, it occurs gradually over 6 to 24 months.

- Unlike the solitary sporadic tumors that tend to displace the cochlear nerve, tumors associated with NF2 tend to form nodular clusters that engulf or even infiltrate the cochlear nerve – microsurgical resection is difficult but preservation of serviceable hearing in NF2 is an attainable goal with modern SRS.

- **Survey of patients:**

<table>
<thead>
<tr>
<th>Issue</th>
<th>Resection1</th>
<th>G K2</th>
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<tr>
<td># of cases</td>
<td>39</td>
<td>11</td>
</tr>
<tr>
<td>Year of study</td>
<td>2001</td>
<td>1998</td>
</tr>
<tr>
<td>No change in employment</td>
<td>68</td>
<td>69</td>
</tr>
<tr>
<td>– ye reccoeaenztion</td>
<td>74</td>
<td>95</td>
</tr>
<tr>
<td>Mean tumor size</td>
<td>25 mm</td>
<td>22 mm</td>
</tr>
<tr>
<td>New Balaeae Problems</td>
<td>78</td>
<td>7%</td>
</tr>
<tr>
<td>New-onset tinius</td>
<td>57</td>
<td>3%</td>
</tr>
<tr>
<td>New facial dysfunction</td>
<td>63</td>
<td>10%</td>
</tr>
</tbody>
</table>

- N.B. radiation **does not preserve hearing significantly better** than properly conducted surgery!

- **Dade Lunsford:** SRS has better hearing preservation for patients < 60 yo; incidental tumor → OK to watch a little bit but counsel for early SRS.

- SRS does not improve tinnitus, balance, hearing.

- **Tumor size:** SRS may be tried for < 4 cm tumors if minimally symptomatic (Koos grade 4); Iorio-Morin et al. Safety and Efficacy of Gamma Knife Radiosurgery for the Management of Koos Grade 4 Vestibular Schwannomas. Neurosurgery: April 2016 - Volume 78 - Issue 4 - p 521–530

  - Tumor control rates 95% and 92% at 2 and 10 years, respectively; serviceable hearing preservation rates 89% and 49% at 2 and 5 years, respectively; facial nerve preservation 100%

- Cystic tumors respond well to SRS (myth busted).

- **Radiosurgical treatment carries 0.1-0.01% risk of malignant transformation.**

- **SRS AFTER FAILED MICROSURGERY:**

  - Achieves tumor control rate 93-94%.

- **Microsurgery after radiosurgery:**

  - Wait at least 6 months after SRS to document continuous tumor growth.

  - Microsurgical resection is generally more difficult after radiation (significant adhesions between CN7 and tumor → facial nerve injury).

  - Dade Lunsford and several other experts: surgery is not more difficult after SRS.

- **SRS repeates if first SRS failed (usually repeat SRS is fractionated):**

- Can be delivered safely without an increased risk of complications as compared to the initial treatment.
TREATMENT - MEDICATIONS
- There are currently no pharmacologic treatments approved by FDA.
- Only drug that has shown promise is clinical trials is BETAVACUZAN, but it works in about 50% of patients.

PROGNOSIS

NATURAL HISTORY
- Mean tumor growth 1-2 mm/year for “stable” tumors (2/3 of all tumors); 50% of patients will maintain hearing during observation for approximately 5 years.
- Mean tumor growth 2-4 mm/year for those tumors that “grow”
  N.B. Some tumors may exhibit exceptional growth that exceeds 18 mm/year!

The overall probability of maintaining serviceable hearing with conservative observation of vestibular schwannomas - Level 3 recommendation:
- High probability (> 75-100%) of hearing preservation at 2 yr
- Moderately high probability (> 50-75%) of hearing preservation at 5 yr.
- Moderately low probability (> 25-50%) of hearing preservation at 10 yr.

Subset of patients with AAO-HNS (American Academy of Otolaryngology-Head and Neck Surgery hearing classification) class A or GR (Gardner-Robertson hearing classification) grade I hearing at baseline - Level 3 recommendation:
- High probability (> 75-100%) of hearing preservation at 2 yrs
- Moderately high probability (> 50-75%) of hearing preservation at 5 yrs
- Insufficient data on hearing preservation at 10 yrs.

The most consistent prognostic features associated with maintenance of serviceable hearing - Level 3 recommendation:
1) Good preoperative word recognition
2) Pure tone thresholds with variable cut-points
3) Nongrowth of the tumor.

FACIAL SCHWANNOMA
- Uncommon lesion (≈ 1.2% of temporal bone tumors; 0.9% of schwannomas of cerebellopontine angle).
- 5% CN7 palsies are caused by facial nerve tumors.
- Identified along entire course of CN7 (intratemporal much more common than intracranial).
- Small percentage display unusual multicentricity (multiple discrete intraneural connections – “string of pearls”).
- Grow at slower rate than acoustic neuroma.
- Tend to grow longitudinally (along lumen of fallopian canal) - may prolapse into middle ear and out of stylomastoid foramen.
- Otic capsule erosion is present in 30% patients.
- 50% fibers must degenerate before clinical signs of palsy are detected.

CLINICAL FEATURES
- Slowly progressive (> 3 weeks) CN7 dysfunction (paresthesia, often preceded by facial twitching).
- Small tumor can become symptomatic if it arises within narrow bony canal (e.g. labyrinthine segment), while more proximal tumor within cerebellopontine angle can become quite large before causing symptoms.
- Classic presentation - recurrent increasingly severe acute paralytic episodes with partial / complete recovery.
- Other possible symptoms - N.B. Hearing loss is found in 69% patients!
  - Conductive hearing loss (impinging upon stapes and oval window - tumors in horizontal segment).
  - Sensorineural hearing loss (cochlear invasion - tumors proximal to geniculate ganglion, in labyrinthine segment).
  - Vertigo (from labyrinthine fistula).

DIAGNOSIS
OTOLOGIC EXAM
- Otoscopy - mass behind drum (29%).
- Pure tone audiogram.
- Stapedial reflexes (not always reliable indicators of distal CN7 function).
- Serial electroneurography - important for surgery timing. See below.
**CN7 action potential** (valid method for assessing CN7 function waveform) – frequently no dysfunction (CN7 schwannomas are extremely slow growing).

**HIGH-RESOLUTION, THIN-CUT CT** of temporal bone is superior to MRI.

**MRI** - small tumors; hypointense on T1 / hyperintense on T2; marked enhancement.

**enhancing enlargement along large segment of facial nerve** is highly suggestive.

**radiographically distinguishing between intracanalicular CN7 tumor and vestibular schwannoma** is often impossible.

**Biopsies** usually result in facial paralysis; pathologic diagnosis is made only intraoperatively.

**Photography** of resting and dynamic facial nerve function (serial assessment of deteriorating facial nerve paresis) is postoperative comparison.

Axial CT - large, circumferential, osteolytic area in labyrinthine segment of facial nerve canal (just anterior to cochlear capsule (small arrowheads)); bone changes appear to be more expansive than destructive; adjacent old surgical defect (large arrowhead).

**DIFFERENTIAL DIAGNOSIS**

1. Hemangioma of temporal bone
2. Malignant schwannoma
3. Meningioma (e.g. intrinsic to geniculate ganglion and in intratemporal segment of facial nerve)
4. Middle ear carcinoma
5. Hematogenous metastasis
6. Direct spread of neoplastic disease into temporal bone

**TREATMENT** - primarily SURGICAL.

Approach: a) no useful hearing and tumor in internal acoustic canal → translabyrinthine or transtemporal approach.
b) useful hearing is present → hearing-sparing approach (e.g. middle fossa approach).
c) residual / recurrent intracanalicular tumor → translabyrinthine approach is preferred (allows complete tumor removal).

b) larger tumors - interposition (cable) graft - greater auricular or sural nerve.

When to remove? EARLY REMOVAL.

- ADVANTAGES - tumor is completely resected, least likelihood of injuring adjacent structures, maximum number of surviving healthy neurons for grafting nerve.
- DISADVANTAGE - patient has good nerve function, and tumor removal nearly always destroys residual nerve function + grafted transected nerve never results in facial function better than House-Brackmann grade III/IV.

Many surgeons delay surgery until CN7 function has deteriorated beyond grade III/IV:
- surgical intervention is initiated once CN7 function has deteriorated to point in which expected result would be nearly equivalent to current CN7 function (documented with electroneuromography).
- intervention should not be delayed past point where motor endplates have atrophied.

**TRIGEMINAL SCHWANNOMA**

- usually arise from root or ganglion; less commonly, proximal division of CN5.
- occupy middle fossa; 50% extend into posterior fossa (may occupy both middle and posterior fossa - dumbbell shape).
- clinical features:
  1) numbness in trigeminal distribution (= 100%)
  2) trigeminal neuralgia (not common).
  3) mild weakness of mastication (< 50%)
  4) extension into cavernous sinus → oculomotor dysfunction (CN6 palsy may precede CN5 dysfunction!).
  5) extension into posterior fossa → CN7 & CN8 dysfunction, cerebellar and pyramidal tract signs.

**CT with bone windows** - erosion of petrous apex, foramen ovale, or foramen rotundum.

**treatment - surgical resection** via subtemporal, frontal, or suboccipital approaches (cure or long-term control).

A. Coronal contrast T1 - MRI - trigeminal tumor that has extended through foramen ovale (arrowhead).
B. Coronal CT with bone windows - widening of foramen ovale by tumor (arrowhead).

**PHOTOGRAPHY** of resting and dynamic facial nerve function (serial assessment of deteriorating facial nerve paresis) postoperative comparison.

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5. Hematogenous metastasis
6. Direct spread of neoplastic disease into temporal bone via facial nerve (e.g. parotid macroepidermoid carcinoma, squamous cell skin carcinoma)

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HYPOGLOSSAL SCHWANNOMA

Contrast T1-MRI:
Axial image (A) - lobulated, enhancing tumor causes expansion of right hypoglossal canal (arrow indicates normal-sized left hypoglossal canal); tumor has large cystic intracranial component which compresses medulla oblongata (arrowheads), and smaller solid extracranial component.

Coronal image (B) - wasting and high signal of right side of tongue (arrowheads) indicates atrophy and fatty replacement.

BIBLIOGRAPHY for ch. “Neuro-Oncology” → follow this LINK >>