Schwannomas of Cranial Nerves

SURGICAL ANATOMY of Cerebellopontine Angle Tumors

1. Surgical Anatomy of Cerebellopontine Angle:
   a. Vascular structures within cerebellopontine angle:
      i. Nerves:
         1. External auditory canal:
            a. Temporal bone:
   b. Temporal bone:

2. VESTIBULAR SCHWANNOMA (S. ACOUSTIC NEURONOMA):
   a. Etiology:
   b. Pathology:
   c. Clinical Features:
   d. Diagnosis:
   e. Audiologic assessment:
   f. Vestibular testing:
   g. Imaging:
   h. Treatment Algorithm:
   i. Treatment - Surgery:
      i. Approach:
      ii. Preoperative:
      iii. Intra-operative monitoring:
   j. Translabyrinthine approach:
   k. Retrosigmoid approach:
   l. Middle cranial fossa approach:
   m. Tumor resection:
   n. Postoperative:
   o. Complications:
   p. Outcomes:
   q. Treatment - Stereotactic Radiotherapy:
   r. Fractionated:
   s. SRs:
   t. Indications:
   u. Methodology:
   v. Stereids:
   w. Follow Up:
   x. Results:
   y. SRs after Failed microsurgery:
   z. Microsurgery after Failed SRs:
   {.
   a. Repeat SRs:
   b. Case discussions:
   c. Treatment - Medications:
   d. Prognosis:
   e. Natural History:

3. FACIAL SCHWANNOMA:
   a. Clinical features:
   b. Diagnosis:
   c. Differential Diagnosis:
   d. Treatment:
   e. TRIGEMINAL SCHWANNOMA:
   f. HYPOGLOSSAL SCHWANNOMA:

CEREBELLOPONTINE ANGLE TUMORS

1. VESTIBULAR SCHWANNOMA (80%)
2. MENINGIOMA (20%)
3. OTHER RARE CASES:
   1) EPIDERMIDIS (1%)
   2) OTHER SCHWANNOMAS - TRIGEMINAL (< 8% of intracranial schwannomas), FACIAL NERVE (extremely rare)
   3) VASCULAR TUMOR
   4) EPIDERMOID
   5) METASTASES

Clinically - Cerebellopontine Angle Syndrome: see p. CNT 2

SURGICAL ANATOMY of Cerebellopontine Angle

- cerebellopontine angle is space filled with CSF
- medial boundary - brain stem
- roof and posterior boundary - cerebellum
- lateral boundary - posterior surface of temporal bone
- floor - formed by lower cranial nerves (IX-XII)
- flocculus of cerebellum may lie within cerebellopontine angle and may be closely associated with CN7 and CN8
- CN7: enters superior petrosal or transverse sinus between torcular and point at which greater petrosal sinus joins travaein of Labbé
- CN8: exits brain stem through series of small rootlets anterior to posterior boundary of brain stem before turning back on itself toward posterior surface of brain stem. AICA can thus be anterior to posterior, it first follows ventral surface of brain stem, but within cerebellopontine angle it takes long loop laterally to porus acusticus. In 15-20% of cases, AICA actually passes into lumen of internal auditory canal before turning back on itself toward posterior surface of brain stem. AICA can thus be divided into 3 different segments - premeatal, meatal, postmeatal
- main branch of AICA 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 meatal segment of AICA:
   1) small perforating arteries supply blood to brain stem
   2) subarachnoidal artery passes through subarachnoidal fossa into posterior surface of temporal bone
   3) internal auditory artery (labyrinthine artery)
- CN7 and CN8 receive blood supply from small branches of AICA.

Two veins must be kept in mind during surgical procedures on cerebellopontine angle:
1. Petrosal vein (of Dandy) - carries venous blood from cerebellum and lateral brain stem to great petrosal sinus. It is encountered in area of CN7 anterior to porus acusticus. The petrosal vein often carries enough venous blood that its obstruction can lead to venous infarction and cerebellar edema.
2. Vein of Labbé (anatomy is quite variable) - carries returning venous blood from inferior and lateral surface of temporal lobe; enters superior petrosal or transverse sinus between transversal and point at which greater petrosal sinus joins transverse sinus.
Nerves

Facial nerve leaves brain stem anterior to foramen of Luschka. If so leaves brain stem, fibers are sheathed in oligodendroglia. Within few millimeters of leaving brain stem, however, nerve loses oligodendroglia and becomes ensheathed by Schwann cells. It passes directly across cerebellopontine angle for about 2 cm, accompanied by CN9. It consistently enters internal auditory canal by crossing anterior superior margin of porus acusticus.

Vestibulocochlear nerve arises from brain stem slightly posterior to CN7.

CN9 remains sheathed in oligodendroglia for approximately 15 mm (almost to point at which it passes into internal auditory canal). It has longest oligodendroglial investment of any peripheral nerve. The junction between oligodendroglia and Schwann cells thus occurs just medial to porus acusticus. Because acoustic neuromas arise from Schwann cells, they arise most commonly within most lateral portions of cerebellopontine angle or internal auditory canal.

Nervus intermedialis (nerve of Weigert) leaves brain stem together with CN9.

1) at some point within cerebellopontine angle, nervus intermedius crosses over to become associated with CN7. It may do so as several separate rosettes. At what point nervus intermedius crosses to become associated with CN7 is considerably variable (in 22% individuals, it is adherent to CN8 for ≥ 4 mm). As CN8 and CN7 reach porus acusticus they pass into it together with nervus intermedius and, sometimes, loop of AICA.

Internal Auditory Canal

• ≈ 8.5 mm (5.5-10.5 mm) in length, lined with dura, and filled with CSF.
• medial end (porus acusticus) is oval in shape.
• lateral end (facialis or lumen cerebelli) is complicated structure - divided into superior and inferior half by bony transverse crest.

1) transverse crest is further subdivided into anterior and posterior segment by vertical bony crest, often referred to as Bill’s Bar 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 foramen, which marks point at which CN7 leaves internal auditory canal and enters its labyrinthine segment. Because most lateral portion of internal auditory canal is 4.5 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 cochlea and anterior end of superior semicircular canal.

2) inferior portion of foramen is single oval-shaped space, anterior portion of which is occupied by stylomastoid foramen (stylo mastoidea foramen) filled with small openings to accommodate terminal branches of cochlear nerve. The posterior portion is filled with macula cribrosa 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 tegmen tympani and greater petrosal sinus attach to medial edge of superior surface of temporal bone.
• arcuate eminence represents most superior portion of superior semicircular canal, which often rises slightly higher than plane of superior surface of temporal bone. It is often difficult to identify, especially in well- pneumatized temporal bone.
• geniculate ganglion generally lies within substance of temporal bone just medial to and few millimeters anterior to head of malloefs. The geniculate ganglion may be sitting right on surface of temporal bone with no bony covering, or alternatively, it may be several millimeters beneath superior surface of bone. The head of malloefs is generally easy to identify if thin bone of tegmen is removed so as to enter into middle ear space from above. In difficult surgical situations, head of malloefs can be used to identify geniculate ganglion. The geniculate ganglion gives off greater superficial petrosal nerve, which courses anteriorly and empties 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 the area. It can then be followed retrograde to geniculate ganglion.
• 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 lies foramen ovale, which accommodates third (mandibular) division of CN5. The horizontal portion of carotid canal courses through anterior temporal bone medial to foramen spinosum and foramen ovale.
• cochlea cannot be identified from surface appearance of superior temporal 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

• rirris: 0-17.1 cases per 100,000 population.
• 7% of all intracranial tumors.
• patients are 40-60 years of age.
• 95% are sporadic. 5% are associated with NF2.
• bilateral tumors are pathognomonic for neurofibromatosis-II (tumors occur earlier).

PATHOLOGY

about schwannoma — see p. Onc60 >>

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

tumor originates at point 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 (≤ 1 mm/year) - can grow to substantial size before clinical symptoms (neurostructures are compressed). + 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 Systemic Review and Evidence-Based Guidelines for Vestibular Schwanommas (2017)

Level 3 recommendation: What is the prognostic significance of KL-6 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.

Neural Atria, LLC

Oncology: schwannomas of cranial nerves. Onc62 (2)
Acoustic neuroma with displaced facial and cochlear nerves (nerves we are trying to preserve):

**Clinical Features**

**Clinical stages**: otologic → neurologic → neurosurgical

N.B. many small tumors never become clinically apparent!

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**
   - 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.
   - **CNS** and later **CN7** are affected - test **CORNEAL REFLEX**!
     N.B. facial weakness (10% - 5-10% patients) vs. hyposthesia of posterior external auditory canal (skin served by n. intermedius of CN7) - early sign - 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.
AUDIOMETRIC ASSESSMENT

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

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

2. ACOUTIC REFLEX TESTING - acoustic reflex decay.

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

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 audiogram: 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 8000 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 tinnitus (either purely unilateral tinnitus or bilateral tinnitus with subjective asymmetry) - this practice is low yielding in terms of vestibular schwannoma diagnosis (< 1%).

N.B. asymmetric tinnitus 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 classifications Haines and EB (Gardner-Robertson hearing classifications) grading of hearing.

VESTIBULAR TESTING

CALORIC TESTING - marked vestibular hypoactivity (canal 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/FLAIR or DTR) may be used to augment visualization of the facial nerve course as part of preoperative evaluation.
- adults with cystic vestibular schwannomas should be counseled that their tumors may more often be associated with rapid growth, lower rates of complete resection, and facial nerve outcome 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 (2012)

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

a) postradiation 3D T1 MPRAGE (magnetization prepared rapid acquisition gradient echo).

b) high-resolution T2 (including constructive interference in steady state [CISS] or fast imaging employing steady-state acquisition [FIESTA] sequences).

- MRs 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 - postradiation 3-D T1 MPRAGE – nodular enhancement considered suspicious for recurrence.

- for gross total resection - postradiation 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 (2012)

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

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.
2. In patients with bilateral vestibular schwannomas, growth rate may increase after resection of the contralateral tumor - more frequent imaging may be indicated, based on the nonoperated tumor’s historical rate of growth. Careful consideration should be given to whether contrast is necessary in followup studies or if high-resolution T2 (CISS or FIESTA) MRI may be adequate instead.

3. Contrast MRI – tumor in right internal auditory canal (arrow):
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 Intracranial Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

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

Small intracranial 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/vertibuloocochlear 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 radiosurgery – 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 slowing 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 reverses 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/nu and epidermal growth factor receptor (EGFR) pathways

Level 3 recommendation: ERLOTINIB is not recommended.

Level 3 recommendation: EBOTRILUM is not recommended.

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

Treatment Hearing preservation CN7 preservation

Chemotherapy 85% 90-100%

Surgery lower
data demonstrates that consistent and durable hearing preservation in sporadic VSs remains an elusive goal. Most patients eventually develop nonserviceable hearing as a result of disease or treatment.

• Auditory 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):

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 ½ 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).
  - if facial paralysis with no recovery within few months → surgical reintervention (another cranial nerve<bits further downstream. CN7 is usually anteriorly to tumor (so nerve is hidden).

• tumor blood supply is from dura of porus acusticus.

Approach selection

1) patient has no useful hearing:

a) retrosigmoid approach (preferred)

b) retrosigmoid approach (for tumors with significant inferior extension)

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 electrocorticography favors hearing conservation (ENG tests horizontal semicircular canal; may be interrputed 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).

- tumor diameter ≥ 1.5-2.0 cm - hearing is difficult to conserve.

3) anatomic variations that make translabyrinthine approach difficult / impossible:
- high-rising 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. If this sinus has to operate, reduced / absent flow in contralateral sinus (injury to remaining sinus = catastrophic venous infarction).
- contracted sigmoid sinus (small lumen for tumor removal + often associated with suppurative otitis media).

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

- Level 3 recommendation: surgery to improve long term outcomes.
- Level 3 recommendation: surgery for patients without preoperative tinnitus, 30% of patients experience a decrease in tinnitus. Of patients without preoperative tinnitus, 30% developed it in the immediate postoperative period.

- Level 3 recommendation: surgery for patients with large (≥4 cm) tumors. Injuries of the nerves intermedius are underestimated because this nerve is rarely 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. Factors such as the presence of excess cerebrospinal fluid or blood, the stimulation voltage used to direct decision making regarding the data averaging that is required to assess changes in hearing, may occur after surgery. In various studies, successful hearing preservation rate was 2-57% using the retrosigmoid approach and 32-68% using the middle fossa approach have been reported.

Tinnitus becomes worse in 60-20% of individuals after tumor removal. In the majority of individuals, tinnitus remains unchanged in approximately 25-60% of cases, experience a decrease in tinnitus. Of patients without preoperative tinnitus, 30-50% developed it in the immediate postoperative period.

Intra-operative Monitoring

of facial and vestibulocochlear nerve → dramatic reduction in morbidity.

- facial nerve – BAEPs
- vestibulocochlear nerve – ECOG, and direct CNAPs.

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

- Level 3 recommendation: perioperative NICE definition (or with the addition of hyperventilation) should be considered to improve postoperative outcomes.

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

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

Radiosurgery 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-III) 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 80% at 6 months. Facial nerve function (grades I-II) can be preserved in only 40-50% of patients with large (≥4 cm) tumors. Injuries of the nerves intermedius are underestimated because this nerve is rarely assessed preoperatively.

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Preoperative

- Intra-operative monitoring

- Level 3 recommendation: facial nerve monitoring must be routinely utilized during surgery to improve long-term 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 anatomic facial nerve does not reliably predict poor long-term facial nerve function (cannot be used to direct decision-making regarding the need for early reinervatation procedures).

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

The overall facial nerve function is difficult to conserve.

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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) Incidence of CSF leak.
5) Allows retraction of CN7 continua (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 (compared with retrosigmoid approach) of inferior portions of cerebellopontine angle, cranial nerves, temporal bone anterior to porus acusticus.
3) Fat graft (from abdomen) 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.

Source of picture: Frank H. Netter “Clinical Symposia”; Ciba Pharmaceutical Company; Saunders

RETROSIGMOID APPROACH

Advantages
1) can be applied to all acoustic tumors* - for operations that sacrifice hearing and operations that attempt to conserve hearing (even for relatively large tumors).
2) best wide-field (panoramic) visualization of posterior fossa - especially helpful when displacement of nerves is not predictable.

The retrosigmoid approach provides the best widefield visualization of the posterior fossa. The time needed for tumor exposure is minimal. The inferior petrosal sinuses are not involved in this approach. The advantage is greatest when the petrous bone anterior to the far posterior surface of the temporal bone anterior to the porus acousticus is more clearly observed than in the translabyrinthine approach. Panoramic visualization is especially helpful when displacement of nerves is unpredictable. 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 cerebellopontine resection or resection (→ edema, hemotoma, infarction).
3) Incidence of CSF leak
4) severe protracted postoperative headache – can be diminished with intraoperative steps:
   - avoid contaminating CSF and subarachnoid space with bone dust;
   - bone flap is replaced and any residual bony defect is eliminated with methylmethacrylate or hydroxyapatite (elimates direct attachment of posterior cervical musculature to dura).
5) tumor is between surgeon and CN7

The retrosigmoid approach may require cerebellopontine resection or resection, which can lead to the development of postoperative edema, hemotoma, 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.

Operative details
1) surgery is done in T-shape, atachialoid incised, and cerebellum is retracted medially to expose cerebellopontine angle (cerebellum frequently falls away from posterior surface of temporal bone; hyperventilation, steroids, and intraoperative mannitol reduce ICP – easy surgical exposure with limited amount of retraction. Nonetheless, gentle cerebellar retraction is frequently necessary).
2) drill off posterior wall of IAC (endolymphatic sac is the landmark how posteriorly we can drill).
3) once adequate exposure has been obtained, tumor is clearly visualized along with brain stem and lower cranial nerves is not predictable.
4) debulking of tumor is next step and must be carefully performed so as to maintain anterior portions of capsule if injury to cranial nerve VII and/or VIII is to be avoided; incise tumor capsule longitudinally (along long axis of CN7 and VII) and with max care so as not to injure labyrinth.
5) once tumor has been substantially debulked, posterior wall of internal auditory canal is removed:
   - dissect dura from posterior surface of petrous bone
   - high-speed drill (irrigating drill, diamond drill bit)
   - bone dust should be prevented from entering subarachnoid space during this phase of procedure. One probable cause for severe and untreatable postoperative headache is spillage of bone dust into subarachnoid space during tumor removal. Carefully place Surgifloc, pieces of Gelfoam, Telfa pads, and/or cottonoid strips around operative site so that bone dust from drilling adheres to it 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 common 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 canal, 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 canal wall.
   - length of internal auditory canal varies considerably from individual to individual, and knowing exactly how much posterior wall canal 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 or auditory nerve (if hearing is to be saved) while at same time increasing chances of leaving tumor at fundus. Use of preoperative angled endoscopes has been of proven, dissection from medial to lateral is thought to be less traumatic to both cochlear nerve and to vascular supply of inner ear. The vestibular nerves are generally sacrificed, 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 cranial nerves VII and VIII. As tumor capsule is carefully removed from brain stem, root entry zone of cranial nerve VII can be identified. The capsule is then carefully removed from facial nerve with as little trauma as possible.
face nerve monitor is great help in this portion of dissection. Variety of techniques 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 disadvantages. Most importantly, it requires summing large number of repetitions in order to extract response from background noise. Consequently, delay is seen between 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 not achieved.

once tumor removal is complete and hemostasis is absolute, dura is closed and craniotomy is repaired — see p. Op300 –>

middel cranial fossa approach

Advantages

1) fully exposes lateral third of internal auditory canal without sacrificing hearing (some surgeons reach tumor by extending 0-5 cm into middle cranial fossa)

2) extradural.

3) risk of dural laceration becomes increasingly more likely as elevation continued anteriorly to foramen ovale if additional exposure is desired. Dural elevation should proceed from posterior to anterior to avoid injury to cranial nerves.

4) very limited exposure of posterior fossa, also may leave temporal lobe.

5) technically difficult.

6) postoperative trigeminal neuralgia (related to manipulation and/or injury to temporalis muscle).

The facial nerve generally courses along the anterior superior portion of the tumor. Depending on whether the tumor is of superior or inferior vestibular nerve origin, the facial nerve may have to be dissected and displaced to obtain access to the tumor. This may make the nerve more vulnerable to injury because cranial nerves are in close proximity to the tumor. 

If this area can be well visualized, then tumor removal is generally both successful and relatively straightforward. Larger tumors frequently have facial nerve splayed out over anterior superior portion of the tumor. Tumor removal begins, as with other approaches, by carefully debulking. Once tumor is well exposed, then facial nerve can be stimulated with low stimulus intensities, chances of good postoperative facial nerve function increase. If required, tumor removal is completed and hemostasis is absolute, then true lateral (parking lot) approach is employed.

Once geniculate ganglion has been completely exposed, labyrinthine portion of nerve can be identified and followed medially and inferriorly into internal auditory canal. Remember that labyrinthine portion of nerve takes markedly vertical course as it moves from lateral geniculate ganglion to proximal fundus of internal auditory canal, which lies 2 or more millimeters deep to geniculate ganglion. Some surgeons prefer to identify internal auditory canal medially. Once medial end of canal is completely identified, they follow canal laterally to fundus of internal auditory canal.

Operative details

Position: head must be in true lateral position. In young individuals with supple neck, this can often be accomplished by turning patient's head toward side of tumor, but neck mobilization in elderly patients or concern exists that forced head turning will limit posterior fossa circulation or aggregate cervical spine dislocation, true lateral positioning should be used.

Exposure: must be centered over vertically oriented line that passes 1 cm anterior to external auditory meatus. This is most easily accomplished through linear incision. A posteriorly based U-shaped or curvilinear incision is used if concern exists about hair loss and/or scar contracture. Depending upon incision used, temporalis muscle is incised or incised inferriorly. A temporal craniotomy (approximately 3 x 3 cm) is performed just above its base at root of zygoma. The dura is draped up from bone to middle cranial fossa, and osmotic diuretics, head elevation, hyperventilation, and steroids are used to limit cerebral edema.

note that must be removed is indicated in yellow (tumor is in orange).

• once geniculate ganglion has been completely exposed, labyrinthine portion of nerve can be identified and followed medially and inferriorly into internal auditory canal. Remember that labyrinthine portion of nerve takes markedly vertical course as it moves from lateral geniculate ganglion to proximal fundus of internal auditory canal, which lies 2 or more millimeters deep to geniculate ganglion. Some surgeons prefer to identify internal auditory canal medially. Once medial end of canal is completely identified, they follow canal laterally to fundus of internal auditory canal.

• internal auditory canal should be skeletonized approximately 270°. The most difficult area to expose is in way during tumor removal (CN7 courses anterior superior portion of tumor) - tempoitary postoperative paresis is more common.

• temporal lobe must be retracted → temporal lobe injury (especially troublesome with dominant side).

• tumor must be removed is indicated in yellow (tumor is in orange)
TUMOR RESECTION

- may leave 1 mm “tumor carpet” on CN7 to avoid nerve injury.
- if tumor extends to EAC (impacted tumor) - CN7 is compressed and high risk of injury.
- CN8 fibers fan out into modiolus – it takes very minimal force to disrupt those fibers – no lateral- to-medial motion while resection, i.e. all sweeping motions must be very gentle medial-to-lateral.
- if need only to debulk tumor (so get off brainstem to allow radiation) – high risk of bleeding.

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.
  - palliative treatment for persistent vertigo after vestibular schwannoma removal - trans tympanic cisternotomy injection to kill remaining vestibular input so brain will no longer receive weird signals from malfunctioning vestibular apparatus.
- discharge on 3–4th postoperative day.

Subtotal resection – wait 1.5–3 mos, then SRS

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 it). see above for MRI requirements

- N.B. distinguishing tumor recurrence from postoperative inflammation (both are MRI enhancing) can be quite difficult. recurrences tend to be globular while inflammatory enhancement tends to be linear.
- recurrence rate is ≤ 5–10% (vast majority of recurrences follow subcapsular 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 - labyrinthine artery (*failure to conserve hearing) and branches to facial nerve (postoperative facial nerve weakness).
   - may be very intimately associated with tumor (esp. larger tumors).

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

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

4. Occlusion of vein of Labbe:
   - severe edema (→ brain herniation) and temporal lobe infarction;
   - vein of Labbe 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 unshanced CT while operating room is prepared.
     - if deterioration is rapid, forge CT and take patient directly back to operating theatre.
     - if deterioration is very rapid (lack 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 → 10th postoperative day → brainstem compression.
   - rotating shaft of surgeon’s biter is often culprit (surgeons usually look past shaft to head of biter 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. 17
   - prophylactic peroperative steroids are widely used (but unequivocal evidence for effectiveness is lacking).
   - 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.
   - 80–90% cases have short delay for few hours to week (vasospasm, vascular injury, traction, nerve edema, 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).

8. Transient abnormality of CSF-resorption → mild transient postoperative hydrocephalus.

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

10. Aseptic meningitis: corticosteroids are extremely helpful.

11. CSF leak through:
   - about management – see p. 564
   a) wound
   b) eustachian tube.

12. Severe postoperative headache - see retrosigmoid approach (disadvantages)
   - H: high-dose NSAIDS, aggressive manipulative physical therapy.

13. Tinnitus worsening (6-20% patients)
OUTCOMES
Radiosurgery Practice guideline (2006) – outcomes by surgery approach >>

CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2012)
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 hearing at baseline - Level 3 recommendation:
- moderately high probability (> 50-75%) of hearing preservation immediately following surgery, at 2 yr.
- at 5 yr.
- and at 10 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 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 radiosensitive and because of proximity with brain stem.

FRACTIONATED
- better hearing preservation (than single-session SRS)
- 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 p. Onc26 >
- can be performed using the Gamma Knife®, modified LINACs or the proton beam.

CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)
There are no studies that compare radiosurgery equipment: Gamma Knife vs LINAC vs proton beam.

INTRAOPERATIVE
N.B. brainstem compression is not improved by radiation – use surgery instead!
1) new / residual / recurrent tumor
2) intracranial 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) older 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)

*Kooy INTRAOPERATIVE tumors should be offered microsurgery as first management.

CNS Systematic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)
Intracranial small tumors (< 2 cm) without evidence of radiographic progression and no tinnitus - Level 3 recommendation: observation is recommended - does not have a negative impact on tumor growth or hearing preservation compared to SRS.

METODOLOGY
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(12-14 Gy) or 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 past patients with contralateral 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 (4–8 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 in order to identify the goal of radiosurgery in an individual patient and the tolerance of the surrounding structures.
- the fall off on cochlea, semicircular canal and brainstem is checked to keep them below tolerance level.

STEROIDS
- there is no consensus on the use of corticosteroids on the day of radiosurgery.
- some do not use steroids at all before, during or after radiosurgery.
- some give IV 40 mg of MEFHYLEDUREEDONE 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: MRT 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.

- MRI 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 ventriculoperitoneal shunting).
  - by 12 months, follow-up MRI shows loss of central tumor enhancement (indicates favorable response).
  - 4-6% of tumors show pseudoprogression (tumor swelling) for 6-12 months – face may twitch – it is temporary.
  - by 2 years, partial involution of tumor is noted.
  - stereotactic radiosurgery 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 (> 95%); high probability (> 75%–79%) of hearing preservation at 2 and 5 yrs moderately low probability (> 25%-50%) of hearing preservation at 10 yrs.

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 1 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
- moderately low probability (> 25-50%) of hearing preservation at 10 yrs.
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:
  - 98% at 6 years (6% of tumors initially increased 1-2 mm during the first 6-12 months and lost central enhancement, thereafter regressing);
  - 95% at 10 years;
  - at 10-15 yrs: 73% reduction in tumor volume, 25.5% no further change, 3% delayed resection, 0.8% required management of hydrocephalus.

- 6-year actuarial rates for preservation (12-13 Gy):
  - facial nerve function > 99% o trigeminal nerve function 95.6 ± 1.8%
  - useful hearing 78.6 ± 5.1%.

- 13 Gy tumor margin dose: 0% risk of new facial weakness and 3.1% 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.

Survey of patients:

<table>
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<th>Issue</th>
<th>Resection1</th>
<th>GK1</th>
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</thead>
<tbody>
<tr>
<td># of cases</td>
<td>32</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>+ ve reccomendation</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 Balance Problems</td>
<td>78%</td>
<td>7%</td>
</tr>
<tr>
<td>New-onset tinnitus</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!
SRS does not improve tinnitus, balance, hearing.

SRS may be used even 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 524-530.

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 can be repeated 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. Christian Iorio-Morin et al. Repeat Stereotactic Radiosurgery for Progressive or Recurrent Vestibular Schwannomas. Neurosurgery, Volume 85, Issue 4, October 2019, Pages 535–542.

SRS is better option (vs. microsurgery) – risk of CN7 injury 0.2% (vs. 15-20%), better hearing preservation.
CNS Systemic Review and Evidence-Based Guidelines for Vestibular Schwannomas (2017)

**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 Subset of patients with vestibular schwannomas):**
- 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 classification with variable cut-points
3) nongrowth of the tumor.

Tumor size at the time of diagnosis, age, and sex do not predict future development of nonserviceable hearing during observation.

**FACIAL SCHWANNOMA**

- uncommon lesion (< 1.2% of temporal bone tumors; 0.9% of schwannomas of cerebellarpetine 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 multilicentricity (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 stylo mastoid 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 (paresis, 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 intracranialular CN7 TUMOR and VESTIBULAR SCHWANNOMA is often impossible.

**BIPSY** usually results in facial paralysis; pathologic diagnosis is made only intraoperatively.

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

Axial CT - large, circumferential, osseous area in labyrinthine segment of facial nerve canal just anterior to cochlear capsule (small arrowshead); 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 via facial nerve (e.g. parotid mucoepidermoid carcinoma, squamous cell skin carcinoma)

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).

Nerve repair:
  a) smaller tumors – primary anastomosis.
  b) larger tumors – interposition (cable) graft - greater auricular or sural nerve.

N.B. best expected functional result rarely exceeds House-Brackmann grade III paresis!

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.
- DISADVANTAGES - patient has good nerve function, and tumor removal nearly always destroys residual nerve function + grafting transected nerve never results in facial function better than House-Brackmann grade III/VI.

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

HYPOGLOSSAL SCHWANNOMA
Contrast TI-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) - widening and high signal of right side of tongue (arrowheads) indicates atrophy and fatty replacement.
Schwannomas of Cranial Nerves

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