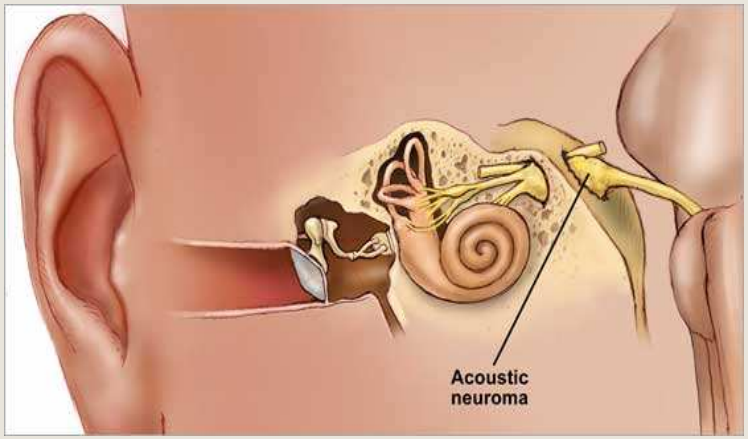
ACOUSTIC NEUROMA
DIAGNOSIS AND MANAGEMENT



Moderators

Dr Ashish Suri

Dr Sumit Sinha

Presented by

Dr Avijit Sarkari

Introduction

- Vestibular schwannoma is the most common tumor occurring in the CP angle (about 85-90%)
- 6 % of all intracranial tumors
- Incidence in US: 10 per million / year
- Peak incidence in 4^{th} to 6^{th} decade
- M:F= 2:3
- 95% Sporadic (unilateral)
- 5% Neurofibromatosis type 2 (bilateral)
- Slow growing tumors = Average 1.8 mm/year (0.2 to 4.0 mm)

Introduction

- Neither Neuroma nor Acoustic (auditory)
- Schwannoma : arising from vestibular nerve Schwann cells at transition zone of the peripheral and central myelin Obersteiner Redlich zone (at the lateral CPA/medial IAC)
- Majority originate within the IAC
- Equal frequency on Superior and Inferior vestibular nerves ????
- Rarely occur on the cochlear division of the 8th CN.
- VS occurs as a result of mutations in a tumor suppressor gene
  - Merlin
  - Located on chromosome 22q12
- VS requires both copies to be mutated
- People with NF2 inherit one mutated gene

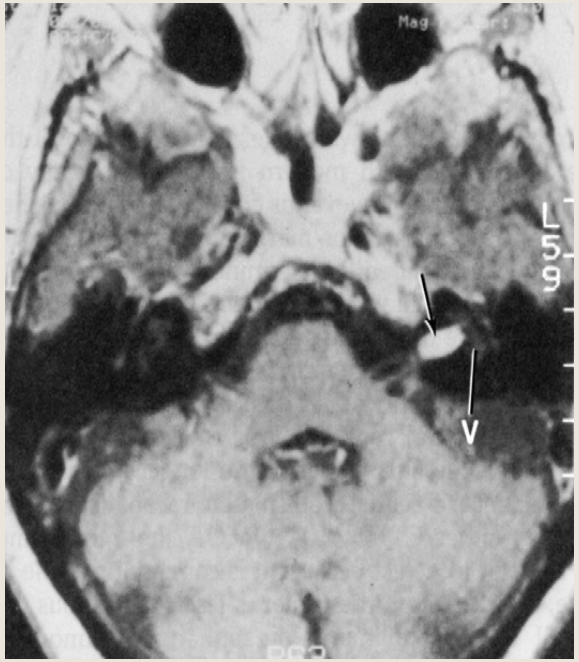
Cerebello-pontine angle

- Superior limb of cerebellopontine fissure
- Inferior limb of cerebellopontine fissure
- Apex-located laterally where superior & inferior limbs meet
- Floor- Middle cerebellar peduncle
- Anterior: posterior surface of temporal bone
- Posterior: anterior surface of the cerebellum
- Medial: lateral surface of brainstem
- Lateral: petrous bone

Cerebello-pontine angle

- Cranial nerves:
  - V
  - VII & VIII
  - IX, X, XI
- Important structures:
  - Flocculus
  - Lateral aperture of 4th ventrical
  - AICA

- Intracanalicular → intracisternal → brain stem compressive



Grading

- *Koos*: (Grade 1-4) upto 1, 2, 3, >3 cm (intracanalicular+cisternal)
- *Ojemann*: (small, med, large)<2, 2-3, >3cm (intracisternal)
- *Samii:* *>3 x 2 cm large, rest small (both intra + extrameatal also T1, T2, T3 AB, T4 AB*
- *Sekhar*: (small, med, large) <2, 2-3.9, >3.9 cm (only intracisternal)

Jackler Staging System

Stage	Tumor Size
Intracanalicular	Tumor confined to IAC
I (small)	< 10 mm
II (medium)	11-25 mm
III (Large)	25-40 mm
IV (Giant)	> 40 mm

What do patients complain of ?

- Hearing loss
  - 95% of patients
  - Most have slowly progressive loss
  - 20% have sudden HL
  - Level of hearing loss is NOT a predictor of size
- Tinnitus
  - 65% of patients
  - Usually constant with a high buzzing pitch
- Disequilibrium
  - 60% of patients
  - Usually well-compensated

Hearing Loss

- Most frequent initial symptom
- Most common symptom ~ 95% AN patients
- Asymmetric SNHL
- High Frequency
- Decreased Speech Discrimination
- Lack of conclusive correlation between tumor size and hearing *

* *Stipkovits EM et al., Am. J. Otology 1998: 19; 834-9*

Pathophysiology of Hearing Loss

- Exact etiology is unknown
- Compressive effect on cochlear nerve
- Vascular occlusion of internal auditory artery

Gardener Robertson Scale

Grade I (good-excellent)	PTA (dB) : 0-30	SD (%) : 70-100
Grade II (serviceable)	PTA (dB) : 31-50	SD (%) : 50-69
Grade III (non-serviceable)	PTA (dB) : 51-90	SD (%) : 5-49
Grade IV (poor)	PTA (dB) : 91-max	SD (%) : 1-4
Grade V (none)	PTA (dB) : not testable	SD (%) : 0

Distribution of Hearing in AN

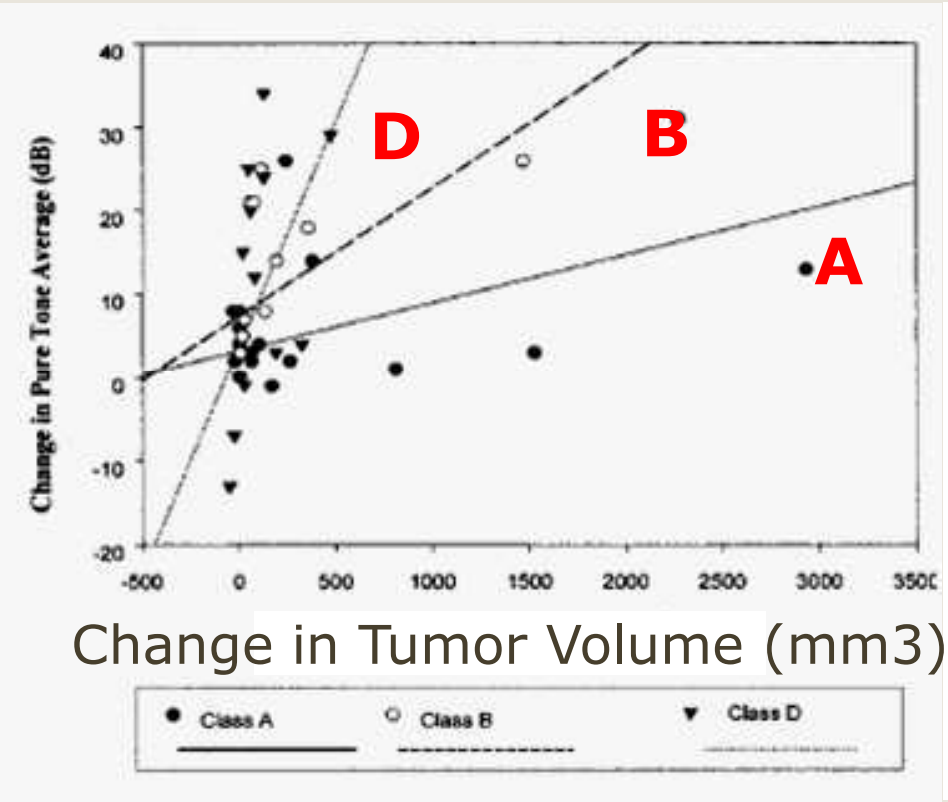
TABLE 1. Hearing on affected side in 190 patients with vestibular schwannoma[a]

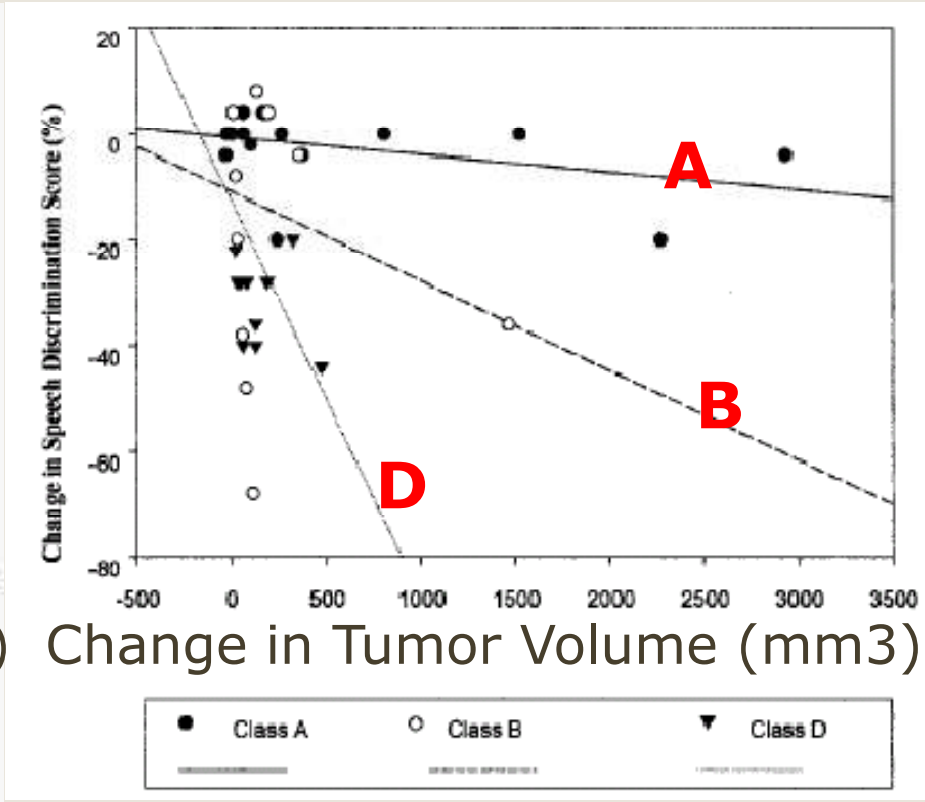
Gardner-Robertson class	Percentage of patients
A	21.1
B	27.9
C	15.3
D	35.8
Total	100.0

[a] Forty-nine percent of patients had serviceable hearing.

Myrseth: Neurosurgery, Volume 59(1).July 2006.67-76

Tumor Growth & Hearing





PTA **SDS**

Massick DD. Laryngoscope 2000: 110; 1843-9

More complaints....

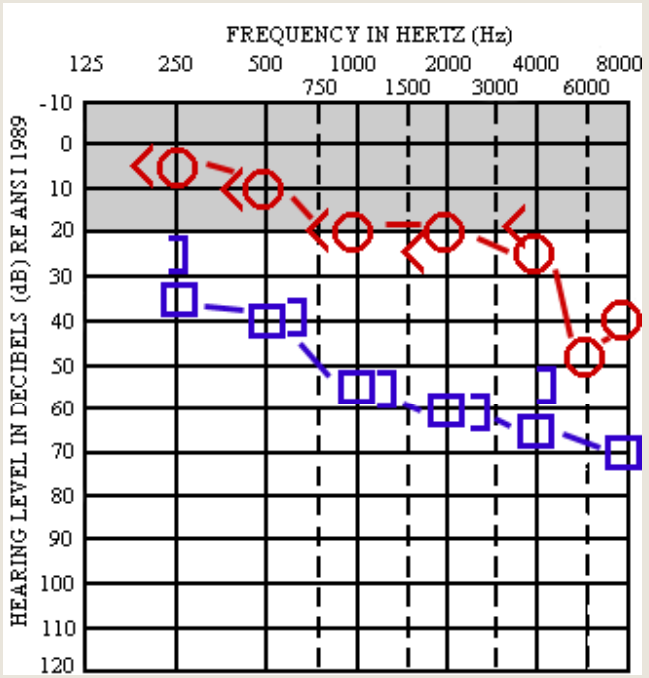
- Facial and trigeminal nerve dysfunction
  - Usually V2 numbness
  - Sensory component of CN VII is usually involved first
    - *Hitselberger sign* – numbness of the posterior EAC
  - Facial weakness or spasm occurs in 17% of patient
- Cerebellar:
  - Wide gait, Falling to side of lesion
- Brainstem:
  - Headache, altered MS, nausea,
  - Visual Loss,
- Other Cranial nerves:
  - IX – dysphagia (large tumors, J F S)
  - X – hoarseness, aspiration (large tumors, J F S)
  - XI – shoulder weakness (large tumors, J F S)

Diagnostic Tests

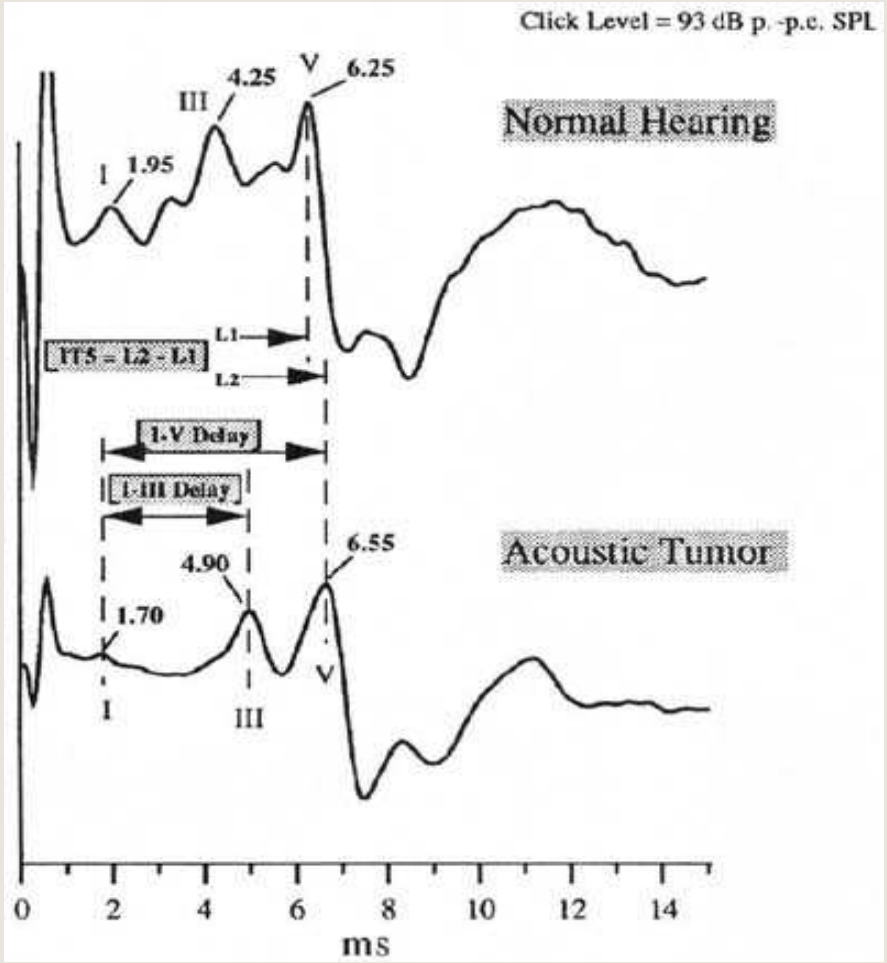
- Audiometric Testing.
- Electrophysiologic Testing.
- Vestibular Testing.-
  - ENG
  - Computerized dynamic posturography.
  - Rotary chair testing.
- CT & MRI.

Audiometric Testing

- Pure-tone testing:
  - SNHL- most commonly high frequency (65%).
  - Normal hearing (5%).
- Speech discrimination:
  - Scores out of proportion with pure-tone thresholds.
  - Some may score well.
  - Rollover phenomenon improve the sensitivity.
- Acoustic reflex thresholds:
  - typically elevated or absent.
  - If present then reflex **decay** measured.
  - The sensitivity is 85% for detecting retrocochlear problem.



BAER: Retrocochlear Pathology



- Most sensitive & specific audiologic test.
- Increased interpeak intervals
  - I-to-III interval of 2.5 ms, III-to-V interval of 2.3 ms, and I-to-V interval of 4.4 ms
- Interaural wave V latency difference (IT5)
  - Greater than 0.2 ms (40-60%).
- Poor waveform morphology i.e. only some of the waves are discernible
- Absent waveform in 20-30%.
- Wave 1 present but all remaining waves are absent in 10-20%.
- Normal in 10-15%.

Fraysse B et al. First International Conf. on Acoustic Neuroma. 1992

BAER: Diagnostic Efficiency

- Generally, Efficiency increases with Size
- Sensitivity: > 90 % for tumor > 3 cm
- False negative Rate:
  - 15 % (Wilson 1992 – 6/40)
    - 33 % (5/15) for Intracanalicular Tumor
- False positive Rate:
  - > 80 % (Jackler 2005)
- Positive predictive value:
  - 15 % (Weiss 1990 – 4/26)
  - 12 % (Walsted 1992 – 23/185)

Cost-Effective Initial Screening for Vestibular Schwannoma: Auditory Brainstem Response or Magnetic Resonance Imaging?

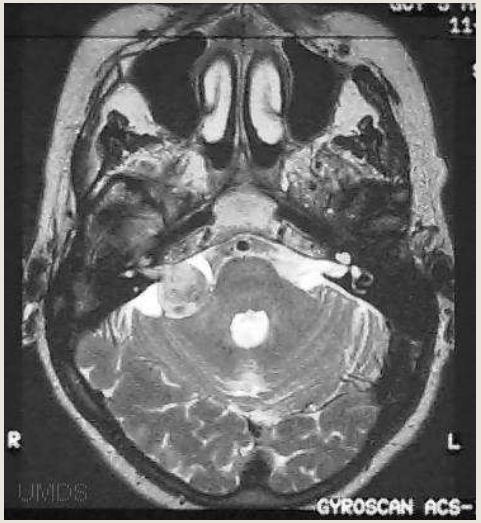
V Rupa, A Job, M George, V Rajshekhar. Dept of ENT & NSx , CMC, Vellore

Otolaryngol Head Neck Surg , June 1, 2003 vol. 128 no. 6 823-828

- 90 patients with asymmetric audiovestibular symptoms, investigated prospectively with both ABR and gadolinum-enhanced magnetic resonance imaging (GdMRI).
- 6 were diagnosed with VS on GdMRI.
- On ABR testing, 4 patients with VS had retrocochlear pathology and 2 with profound sensorineural hearing loss had no responses.
- ABR was found to have a sensitivity of 100% and specificity of 61.9%. A protocol involving screening of all patients with asymmetric audiovestibular symptoms using ABR and only subjecting those patients with no responses or retrocochlear pathology to GdMRI would effect a savings of $1200 for every patient detected to have a VS.
- ***CONCLUSIONS*: Including ABR as the preliminary screen for patients with asymmetric audiovestibular symptoms is a cost-effective strategy.**

MRI is the Gold Standard

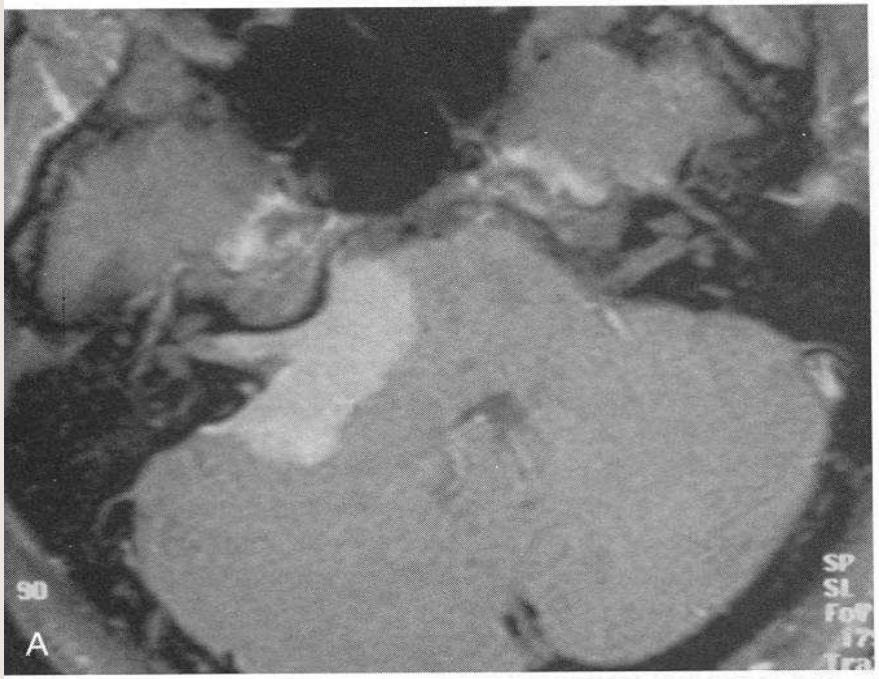
T1 pre-Gad

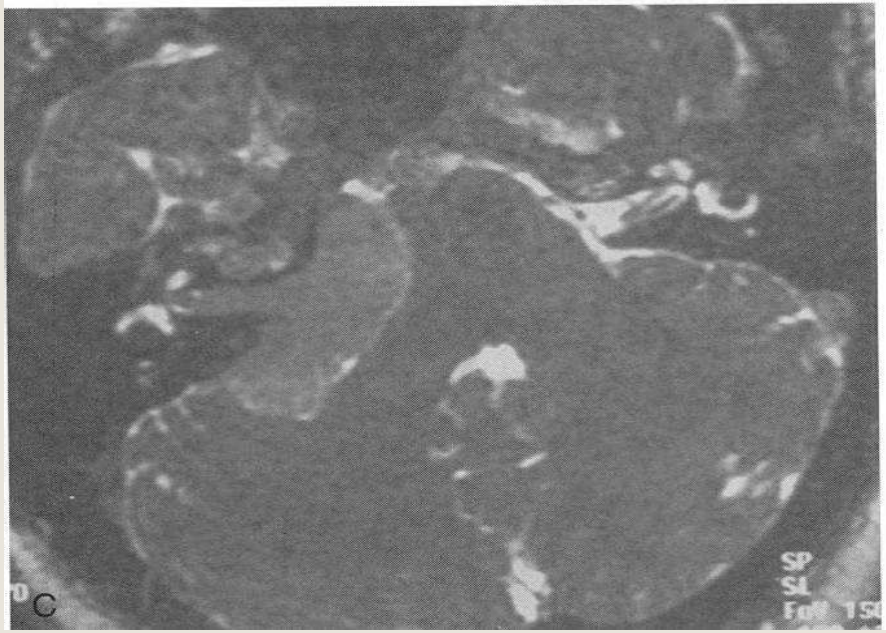


T2

T1 post-Gad

- T1: Isointense to brain, hyperintense to CSF
- T2: Hyperintense to brain, hypointense to CSF
- T1 -Gad: Enhancing





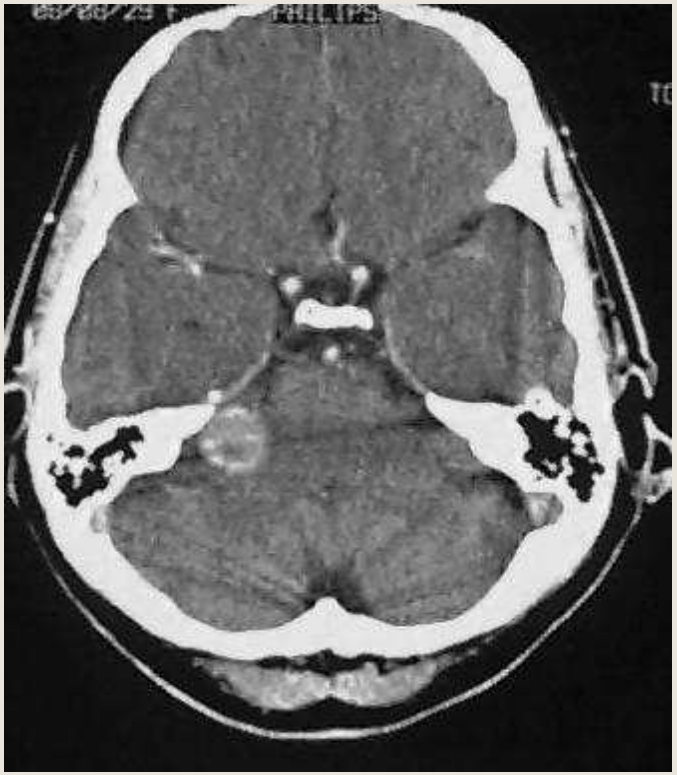
Meningioma Features:

- Arise from surface of petrous bone.
- Obtuse angles to petrous bone.
- Uncommonly involves the IAC.
- Frequently with dural tail.
- Calcifications common.
- Pial vessel flow voids.

MRI characteristics of other lesions

Lesion	T1WI	T2WI	Enhancement	Suggestive feature
Epidermoid	Hypo	Hyper	No	Hyper on DWI
Dermoid	Hyper	Hypo	No	Fat and calcium
Arachnoid cyst	Hypo	Hyper	No	Iso to CSF, hypo on DWI
Anuerysm	Hypo	Hypo	Possible	Well circumscribed hypo- on T2
Cholestrol gran	Hyper	Hyper	No	Hypo- rim on T1 & T2
Chondroma	Hypo	Hyper	Variable	Origin from synchondrosis
Chordoma	Hypo	Hyper	Yes	Intra tm septa

CT Brain with contrast



- **Heterogeneous enhancement on contrast**
- **Indicated in: Contraindication to MRI (metallic implants), claustrophobic patients**
- **May not be able to detect small tumor < 1.5cm**
- **Radiation risks**

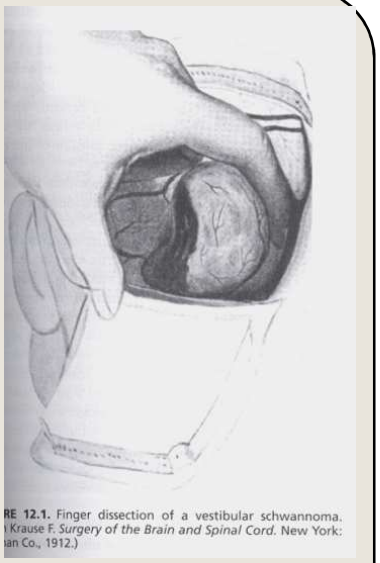
<u>Pre-op Thin cut CT of post fossa</u>
(Samii: Essen In Nsx)

Identify bone destruction
Expansion of IAC
Position of labyrinth-relation to fundus
Position of sigmoid sinus and em vein

Management options - No strict guidelines

- Surgery
- Radiosurgery or Fractionated RT
- Observation (with careful audiologic and radiologic monitoring)
- Management of Preoperative Hydrocephalus –
  - Asymptomatic – Steroids/Intraop EVD
    No special treatment
  - Obviously sick; gross hydrocephalus with symptoms of raised ICT like headache, vomiting, papilloedema – Ventriculoperitoneal shunt

Historical perspective

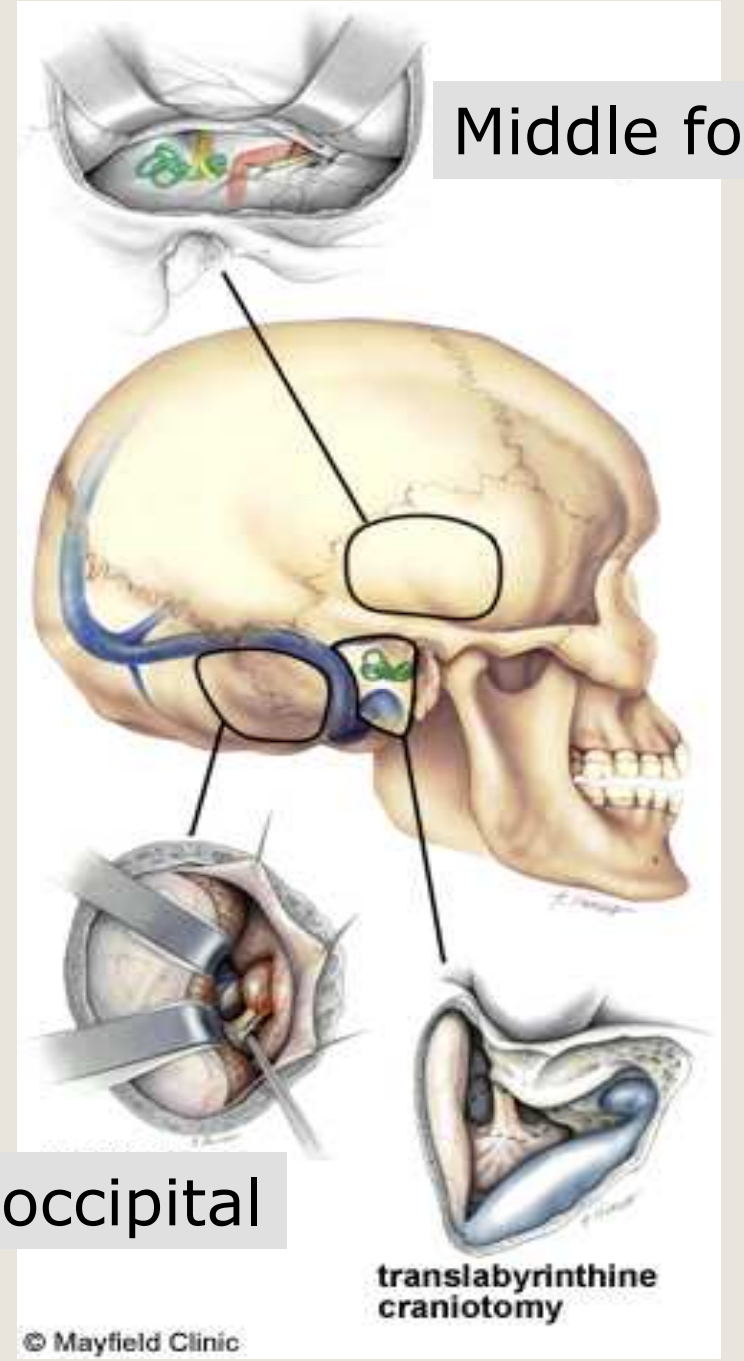


RE 12.1. Finger dissection of a vestibular schwannoma. Krause F. *Surgery of the Brain and Spinal Cord.* New York: ian Co., 1912.)

- **Sandifort-**1777, earliest description of AN
- **Sir Charles Balance**, 1894 – finger enucleation
- **Annadale**-1895, first true AN removal (Cushing)
- **Cushing** 1905- subtotal intracapsular removal
  - Hemostasis: silver clips, bone wax, electrocautery
  - Mortality: 20 % (1917) → 4% (1931)
- **Dandy**, 1925- first total removal unilat SOC (advocated- ventricular tapping, open cisterna magna, resect lateral third cerebellum, unroof IAC for complete resection) . Mortality 10%
- **William House** (1960)
  - Translabyrinthine approach using surgical drill and operating microscope
- **Givre & Olivecrona** – pioneered facial nv. Preservation
- **Rand and Kurze** –1968-cochlear + facial n. preservation
- **Delgado**- intraop VII nv monitoring, 1979

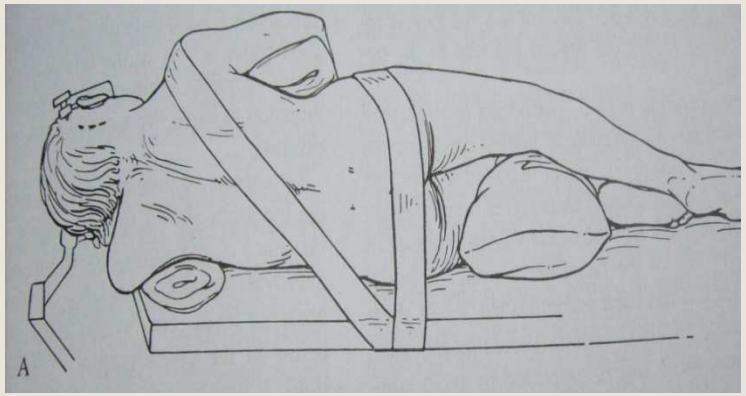
Surgical approaches

- Retromastoid suboccipital transmeatal approach
- Middle fossa approach
- Translabyrinthine approach



Retromastoid suboccipital transmeatal approach

- **Position – Surgeon's preference**
  - Lateral oblique
    - Comfort of the surgeon
    - Excellent visualization of CPA, direct visualization of vessels
    - Ease of tumor removal
    - Prevention of hypotension
    - No concern about air embolism
  - Semisitting/sitting-air embolism, hypotension, surgeon discomfort, but clean field
  - Prone
  - Lateral : BPI
  - Supine oblique: Cervical spondylosis



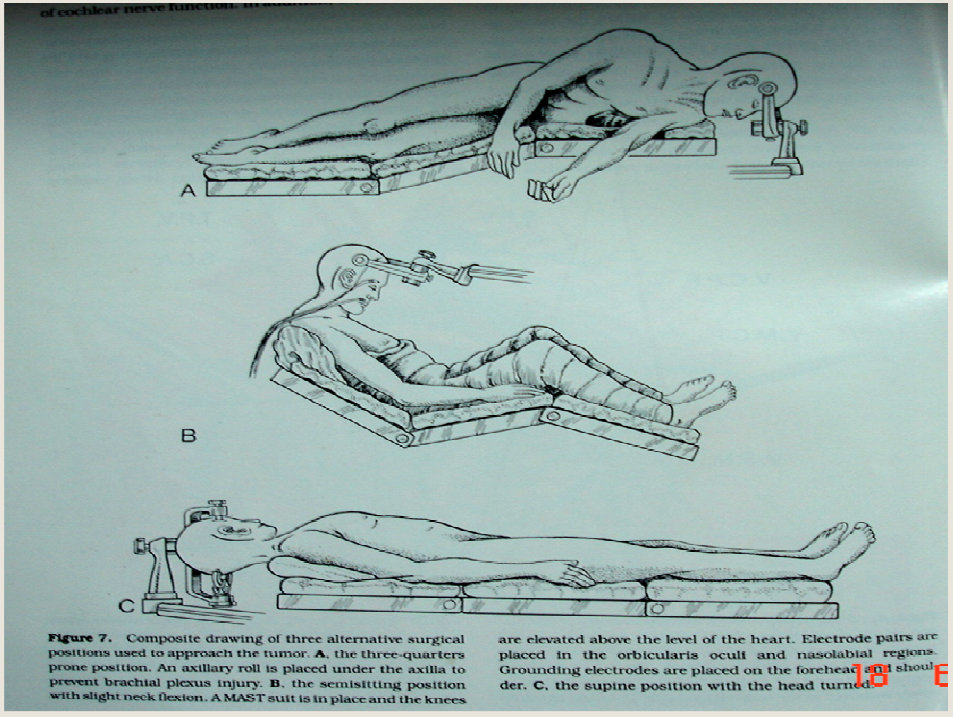
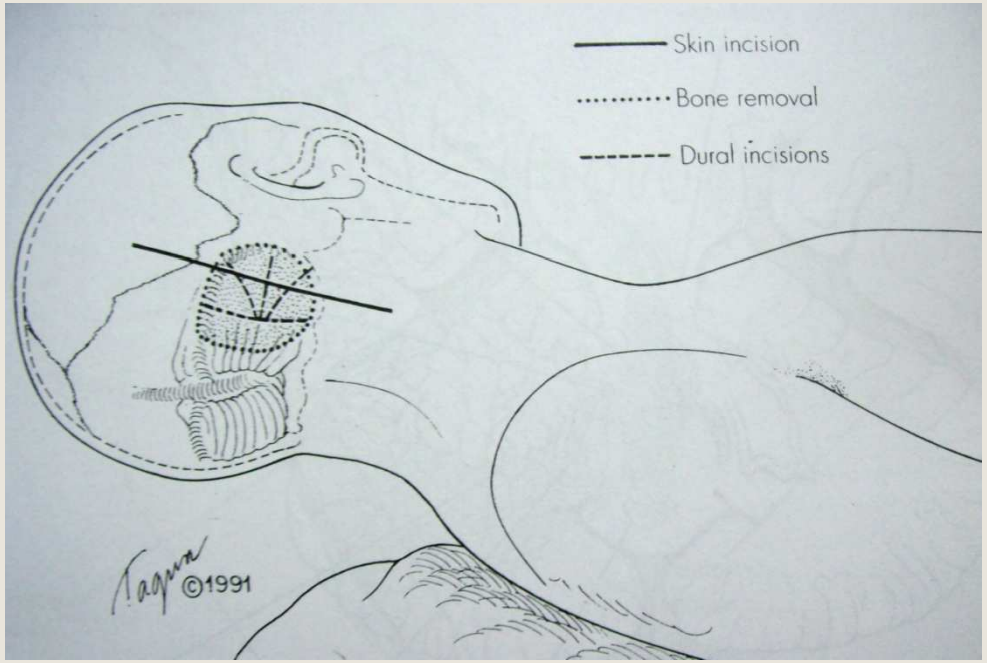


Figure 7. Composite drawing of three alternative surgical positions used to approach the tumor. **A**, the three-quarters prone position. An axillary roll is placed under the axilla to prevent brachial plexus injury. **B**, the semisitting position with slight neck flexion. A MAST suit is in place and the knees are elevated above the level of the heart. Electrode pairs are placed in the orbicularis oculi and nasolabial regions. Grounding electrodes are placed on the forehead and shoulder. **C**, the supine position with the head turned.

Images: MLJ Apuzzo, Brain Surgery, 1993

Retromastoid suboccipital transmeatal approach



- Incision –
  - Vertical linear (1 cm medial to the mastoid process)
  - 'S' / Lazy 'S'
  - Inverted 'J' -shaped/ Hockey-stick
- Anatomical variants-
  - Dolichoectatic VA/Occipital artery
  - Hypoplastic VA

    (20 %)- Avoid extreme flexion

Retromastoid suboccipital transmeatal approach

- Landmarks of Cerebellopontine angle and extent of craniotomy –

Extradural landmarks	**Intradural landmarks**
Transverse sinus superiorly	Tentorium superiorly
Sigmoid sinus & tranverse – sigmoid junction laterally	Petrous bone laterally
Mastoid tip/ digastric groove inferiorly	Flat floor of posterior fossa inferiorly

Retromastoid suboccipital transmeatal approach

- Maintain the arachnoidal plane & dissect it from the tumor superiorly, inferiorly & medially
- Dissect sup pole first in large tm- V^{th} nerve is easily identifiable
  - V nerve- typical flat app. with prominent fascicles, located at the junction b/w the tent & temporal bone
  - Arachnoidal bands b/w tm capsule & the nv. cut by meticulous technique
  - Superior petrosal vein. coagulated, if needed

Retromastoid suboccipital transmeatal approach

- Internal decompression (CUSA/laser/biopsy forceps)
- Push arachnoid with vessels back
- Debulk Tm
- Tumour capsule separates from the arachnoid
- Sharp dissection
- Cut arachnoidal bands b/w tm and cranial nerves, dissect into the cleft b/w the tumor & brainstem
- Identify VII & VIII nerve
- **All the pressure to be placed on the tm capsule while separating it from the cr. nerves & brainstem**

Retromastoid suboccipital transmeatal approach

- Facial N displacement:
  - Ant 70%
  - Sup 10%
  - Inf 13%
  - Post 7%
- Shape:
  - Thin bundle 2/3
  - Splayed over capsule 1/3

- Facial nerve landmarks-

- Lateral end of pontomedullary sulcus, 1-2mm ant to VIII n
- VII n arises 2-3 mm above the most rostral rootlet of IX n
- ***Choroid plexus*** protruding from the foramen of Luschka- VII n lies just anterosuperior
- ***Flocculus***- lies just posterior to the site where VII& VIII nerves join the pontomedullary sulcus
- Silvery white, very shiny vs. dull yellow (Vestibular nv)

RMSOC – Intrameatal part

- Drill the posterior meatal wall
- Continuous irrigation to prevent thermal injury
- Prevent bone dust dissemination in subarachnoid space
- High projection of the jugular bulb, mastoid air cells. Identify neural structures in IAC after opening the dura
- Care taken not to enter the labyrinth
- The dissection along the eighth nerve is done in a **medial to lateral direction**
- **Meticulous dissection to prevent VII n injury as it turns the corner over the medial and ant lip of IAC to enter the subarachnoid space**

Retromastoid suboccipital transmeatal approach

- SUBTOTAL REMOVAL
  - Hearing preservation in large tumors.
  - Very thin 7th nerve with thick adhesions to tm.
  - Elderly debilitated pt with brainstem compression.

Retromastoid suboccipital transmeatal approach

Advantages:

- Good exposure of the CPA cistern
- Good for medial tumors
- Even large Tm
- Facial preservation
- Hearing preservation (50% in tumors <2cm)
- Direct visualization of vessels

Disadvantages:

- Poor exposure of lateral end of the internal auditory canal
- Cerebellar retraction
- CSF leak (7-21%)
- Persistent postop headache

Complications of RMSOC approach

- Post-operative cranial nerve dysfunction – V, VII, VIII, LCN
  - VII nerve paresis- Artificial tears, Lubricant eye gel, Lateral tarsorrhaphy, reanimation
  - LCN paresis- RT feeds/ Feeding gastrostomy
- CSF leak from the wound
  - **Rule out Hydrocephalus**
  - Stitch + Temporary LP drain
- CSF oto-rhinorrhoea
- Meningitis
- Wound infection
- Post-operative hematoma

Complication avoidance

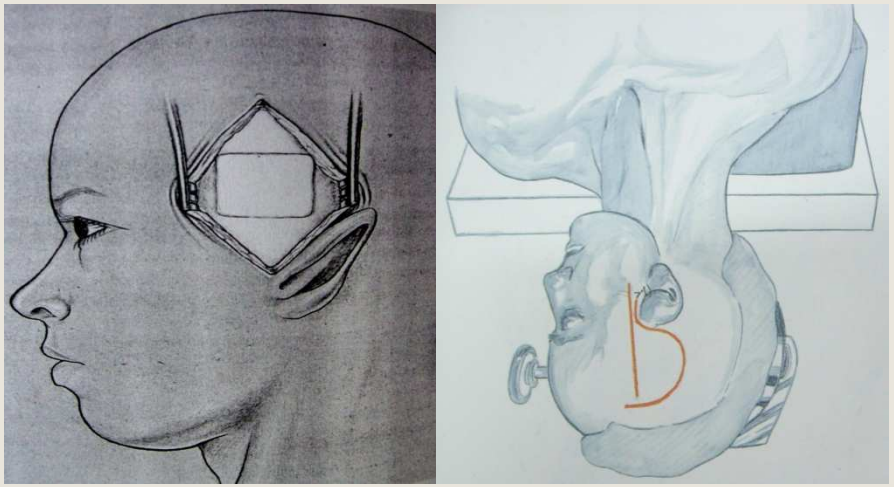
- VIII nerve dysfunction-
  - Intra-op BAER
  - Identify cochlear division at the transverse crest
  - Arachnoid over the cochlear nerve to be preserved
  - Minimal manipulation of the nerve
  - Preservation of auditory artery
- VII nerve dysfunction-
  - Intraoperative monitoring
  - Identify nerve at the nerve root exit zone and at the transverse crest
  - Sharp dissection
  - Minimal manipulation of the nerve
- V nerve dysfunction-
  - Debulk large mass-remove tumor from the nerve, not nerve from the tumor
- LCN dysfunction-
  - Minimal manipulation of the nerve
  - Leave arachnoid over the nerves Intact
  - Protect with gelfoam
  - Sharp dissection

Complication avoidance

- Injury to AICA-
  - Never coagulate any vessel until proximal/distal directions and supply is determined
- CSF otorhinorrhoea (Paradoxical CSF rhinorrhoea)-
  - Knowledge of pneumatized temporal bone and porus
  - Waxing of mastoid air cells
- Cerebellar swelling/infarction-
  - Adequate size of craniotomy
  - CSF to be released from cisterna magna
  - Periodic release of pressure from retractor
  - Craniectomy
  - Lasix/mannitol
- CSF leak from the wound-
  - Clean sharp incision
  - Careful handling of wound edges
  - Meticulous closure of the fascial layers, esp. along the inferior aspect of the wound overlying the mastoid tip, where clear fascial layers are not always present
  - Strict antisepsis, minimizing chances of wound infection

Middle fossa approach

- House, 1961
- Indication: Small intracanalicular tumor, especially in lat part with the aim of facial nerve & hearing preservation.
- Extradural subtemporal approach with microneurosurgical unroofing of IAC



Middle fossa approach

- IAC exposed by following GSPN to the geniculate ganglion
- The bone is then drilled off the arcuate eminence until only a thin layer of bone remains over superior semicircular canal→Posterior boundary of the dissection of IAC
- VII nv. followed from geniculate ganglion to the lateral end of the IAC

Middle fossa approach

- **Advantages-**
  - Extradural dissection
  - Complete exposure of the IAC
  - Avoid blind dissection in lateral IAC
  - Total removal of Tm, even the lat part- good for small tm
  - Hearing preservation (50-70%)
  - No risk of CSF leak

Disadvantages:

- Facial nerve comes first- more manipulation
- Limited access to post fossa, esp. if there is bleeding
- Only small intracanalicular tm
- Elderly patients with thin dura are less tolerant to temporal lobe retraction

Postop Complications:

Bleeding
Stroke
SIADH
CSF leak
Meningitis

Translabyrinthine approach

- Panse, 1904
- House, 1964
- Exposes posterior fossa in the retromeatal trigone (Trautmann's triangle)
    - Sigmoid sinus
    - Jugular bulb
    - Superior petrosal sinus
- Moderate sized tumor⟶ 1-2.5cm
- Auditory function lost

Translabyrinthine approach

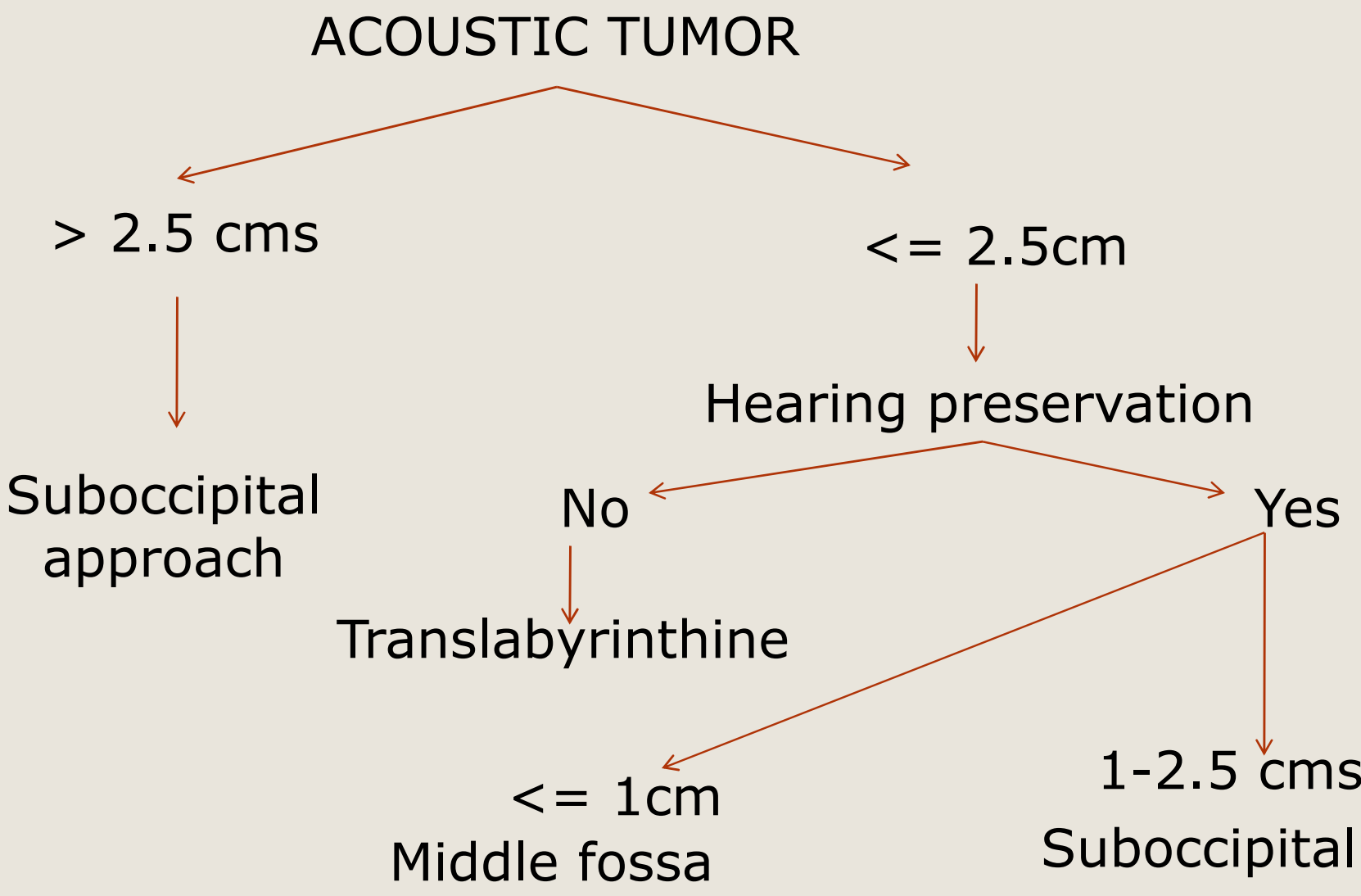
Adv:

- Early identification of facial nv. – 97 % preservation
- Direct approach to the CPA, absence of significant cerebellar retraction
- Short distance b/w surface and tumor
- Excellent exposure of the lateral end of IAC
- Less postop headaches

Disadv:

- Deafness
- Reduces exposure
- **More CSF leak – 27 %**
- Middle ear infection is a CI
- Limited in patients with anterior sigmoid sinuses and high-riding jugular bulbs

Choice of approach



Facial N preservation

- 1st- Cairns 1931
- Olivecrona- 1st to attempt in a large series of patients

- VII nerve monitoring
  - EMG monitoring of ms innervated by VII nerve
  - Displayed on an oscilloscope connected to an audio amplifier
  - Statistically significant difference in anatomical & functional VII nerve preservation
- Facial Motion sensor

- Ebersold (1992): amplitude of CMAP on stimulation of facial N in lateral IAC predicts outcome.
- Others-
  - Intraop change in threshold of stimulation reflects amount of damage.
  - Threshold of stimulation at REZ at brainstem reflects outcome.
- Delayed Facial weakness: edema/inflammation- usually complete recovery by 6 month
- Outcome assessed at one year

House and Brackmann Grading

1- normal

2- close eyelids with min efforts

3- no functional impairment, close eyes with max efforts, obvious synkinesis/ contracture/ hemifacial spasm

4- Normal symmetry and tone at rest, can't close eyes, severe synkinesis etc

5-Asymmetry at rest, decreased/absent nasolabial folds, minimal movt. of eyelids

6- No motion, loss of tone

Outcome

- Ojemann 1993:
- Tm size
- 1cm-100%

  1-2 m- 95%

  2-3cm- 80%

  3-4cm- 60%

  >4 cm- 50-55%

Samii: better results for same size

Microsurgical management of giant acoustic neuromas: An institutional series of 400 cases

Sumit Sinha, B S Sharma, *Asian Journal of Neurosurgery 2008; 11: 47-56*

- *Facial nerve anatomically preserved in 78%, last follow up- 82% patients showed acceptable facial function.*
- *GTR in 24.2%, NTR 47.2% and STR 28.6%.*
- *The preoperative tumor size was not statistically related to the extent of resection.*
- *Meningitis and Chest infection-major causes of morbidity. Mortality 2.2%.*
- *Intraoperative facial nerve monitoring: definite adv in anatomical preservation.*
- *Learning curve of surgeons, large tumor size, preoperative lower cranial nerve involvement, altered sensorium at the time of admission, cystic nature of the tumors and low general condition at the time of surgery were the* ***main factors contributing to morbidity and mortality.***

Hearing preservation: Monitoring

- BAER- monitors pathways central to tm
- ECoG- CAP of auditory nerve monitors pathways distal to tm, cochlear microphonics indicates status of cochlea
- Cochlear N direct recording of CNAP

Elliot and McKissock first reported hearing preservation in 1954

BAER

- Wave V is most prominent – monitored
- Disadv:
  - BEAR unrecordable pre op in 1/3 patients
  - Delay in response of upto 20-60 sec due to signal averaging

ECoG

- Monitors **CAP** of auditory nerve near the cochlea and **cochlear microphonics**, generated from hair cells.
- Adv:
  - Rapid feed back of N1.
  - Not affected by anesthetic agents.
  - Almost always detectable.
- Direct recording of CNAP from nerve- good predictor, but impairs surgical field, 8th nerve must be visible before it can be used

Problems: dislodgement of electrode, fluid in middle ear may block sound transmission.

Recent Advances

- *Direct recording of potentials from cochlear nucleus in lateral recess* (Jannetta et al)
- *Fast BAER response* (10 sec) by using electrodes attached to cerebellar retractor (Samii et al)

- Mech of hearing loss:
  - Direct damage to cochlear nerve
  - Involvement of cochlear nerve by tm
  - Interruption of blood supply to cochlea/nerve
  - Injury to Labyrinth
- Delayed hearing loss:
  - Nerve edema
  - Impairment in vasa- nervosum circulation
  - Increased permeability of endoneural vessels after mech. compression trauma
  - Progressive scarring of IAC with compression of cochlear N or microvasculature

Technical points avoiding cochlear nerve injury

- *Jannetta et al:*
  - Elevate cerebellum, avoid medial retraction !
  - Sharp dissection with scissors (No CUSA/laser/forceps)
  - Alternate dissection in all directions
  - Preserve even small vessels going into the IAC

Hearing preservation: Prognostic factors

- GOOD
  - Small tm (<2cm)
  - Good pre op hearing
  - Lack of lateral tm extension to fundus of IAC
  - Absent caloric response (tm from sup vestibular N.)
- POOR
  - Sudden intra-op loss of potentials is a poor prognostic factor
  - Intraoperative presence of severe adhesions between nerve and tm- m imp. factor : *Moriyama et al JNS 2002*

Results

- *Gormley, Shekhar et al, NS 1997* (179 patients, 5yr FU, 99% complete removal)
- Size Facial(Grade 1/2) Hearing (Grade 1/2)

Size	Facial(Grade 1/2)	Hearing (Grade 1/2)
<2	96%	48%
2-3.9	74%	25%
>4	38%	0

Conservative management

- 2nd radiologic exam in 6 months, then yearly
- If the tumor grows 2-3 mm in the first year, then they will likely need treatment

- Indications
  - Advanced age (> 65 yrs)
  - Short life expectancy (< 10 yrs)
  - Poor general health
  - Small tm with minimal / no symptoms especialy intracanalicular
  - Tm in the only or better hearing ear.
  - NF 2
  - Patient preference
- Contraindications
  - Young patient
  - Healthy patient
  - Symptomatic progression
  - Compression of brainstem structures

Conservative management

- Disadvantages
  - Risk of hearing loss even in non-growing tm.
  - Loss of patient compliance
  - Chances of hearing preservation better in cases short symptom duration

Result of conservative management

- Prospective cohort study of 72 patients
  - Age at presentation: 60.8 years
  - Mean follow-up: 80 months
- Mean tumor size at diagnosis: 9.4 mm
- Mean tumor growth rate: 1 mm/ year
- 87% growth rate < 2 mm/ year
- Tumor growth
  - + : 39 %
  - 0: 42%
  - - : 19%
- No correlation between growth and age, gender, size at presentation, or presenting symptoms
- 32 % failed conservative management

Raut V et a.: Clin Otolaryngol 29:505–514, 2004.

Role of Endoscopy

- Tm<3cm
- 1.5-cm "keyhole" retrosigmoid craniotomies
- 95% tumors were completely removed
- Anatomic preservation of the facial nerve – 100% & of the cochlear nerve in 82% cases
- No LCN paresis
- No death

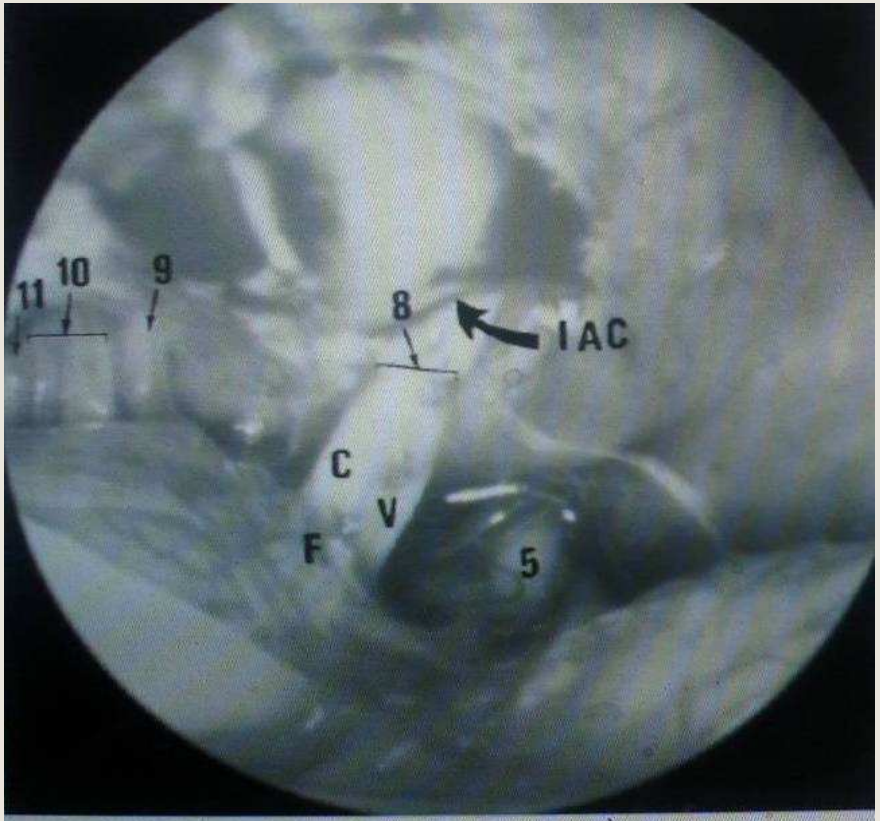


FIG. 1. Inspection of the posterior fossa through a 0° endoscop
C = cochlear nerve; F = cerebellar flocculus; V = vestibular nerv
5, 8, 9, 10, 11 = fifth, eighth, ninth, 10th, and 11th cranial nerves

Kabil MS, Shahinian HK. A series of 112 fully endoscopic resections of vestibular schwannomas. Minim Invasive Neurosurg. 2006 Dec;49(6):362-8.

Quality of life(QOL) after V S surgery

- Factors contributing to QOL-
  - Hearing loss
  - Facial paresis
  - Postoperative ataxia
  - Dysguesia
  - Post-operative headache

Quality of life after acoustic neuroma surgery.

Ann Otol Rhinol Laryngol. 1996 Jun;105(6):423-30.

- 33% required postoperative home help
- Suboccipital surgery-
  - Poorer facial nerve function
  - More reports of pain & incapacity to work
- Translabyrinthine approach-
  - More severe pain, postoperative vertigo
- Larger the tm, more no. of patients unfit to work

Taste dysfuntion in vestibular schwannoma

RN Sahu, S Behari, VK Agarwal, PJ Giri, VK Jain. *Neurology India, Jan- Mar 2008, Vol 56,*

- n= 142.
- Pre-op decreased sensation - 40.8%, Sensory disturbance- 33-45%
- Post-op disturbance- 45.8%
- Improved taste- 6.9 %
- Conclude
  - Dysguesia must be included in pre-op counselling.
  - Dysguesia to be included in facial nerve function assessment.

Facial paralysis and surgical rehabilitation: a quality of life analysis in a cohort of 1,595 patients after acoustic neuroma surgery. Otol Neurotol. 2005 May;26(3):516-21

- 45.5% - experienced worsened facial weakness caused by surgery, and of these, 72% reported that it was permanent.
- 28% felt significantly affected by facial weakness
- The factor most often associated with poor outcome was a large tumor

Quality of life(QOL) after V S surgery

- Treatment for VS results in a significant reduction in QOL.
- Major effect is in the psychological area, with increased rates of emotional distress and impaired social functioning
- Patients with facial weakness are at the greatest risk of a poorer psychological outcome.

Role of Radiosurgery

- Low-morbidity alternative to microsurgery
- Similar long term tm control rate
- Indications-
  - Hearing loss/enlarging tumor in the only hearing ear
  - Functional hearing
  - Residual/recurrent tumor after subtotal removal
  - Patient with a tumor having ≤ 2cm intracranial extension
  - Major medical illness
  - Older patients (>75)
  - Patient's decision
- Contraindications
  - Tumors > 3 cm
  - Prior radiotherapy
  - Tumor compressing brainstem

Role of Radiosurgery

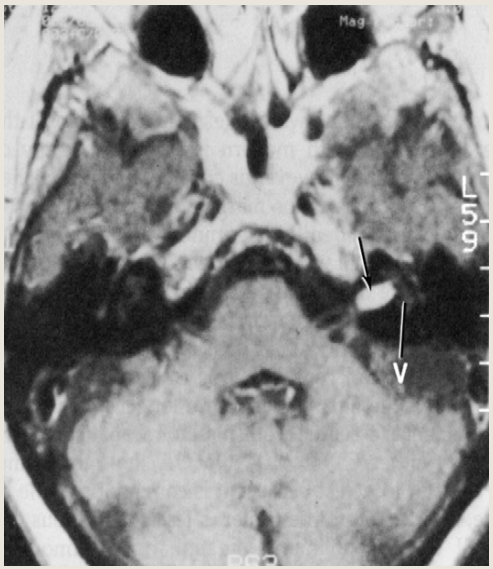
- Multiple iso-centers (6-13) to achieve high degree of conformality
- Prescription doses most commonly used today – 12-13 Gy
- Outcome
  - Local control (non-progression): 94%
  - Transient swelling in the first two years
  - Hearing preservation: 47 – 77% - decreases each year after radiation and stabilizes after 3 years in 50% of patients
- Complications
  - Facial nerve injury: 5 - 17%
  - Trigeminal nerev injury: Numbness 2 - 11%
  - Hearing loss
  - Hydrocephalus: 3%
  - Radiation induced tm

Radiosurgery experience-

Institute	n	Median FU (months)	Median Marginal tm dose	Tm control	V nv complication	VII nv	VIII nv
Pittsburgh	313	24m	13 Gy	98.6%	4 %	0 %	30 %
Osaka	51	60m	12 Gy	92%	2 %	0 %	44 %
Marseille	97	36-108	12-14 Gy	97 %	4%	0%	30%
Jefferson	69	27m	12 Gy	98%	5%	2%	67%
Amsterdam	49	33m	12.5Gy	100%	8%	7%	25 %
AIIMS (10 yrs)	**198**	**27m**	**12 Gy**	**95.50 %**	**0.5 %(1)**	**2%(4)**	

Management decisions

- Depend upon-
  - Size of tumour
  - Symptoms of the patient
  - Age
  - Hearing preservation
  - Patient's wish



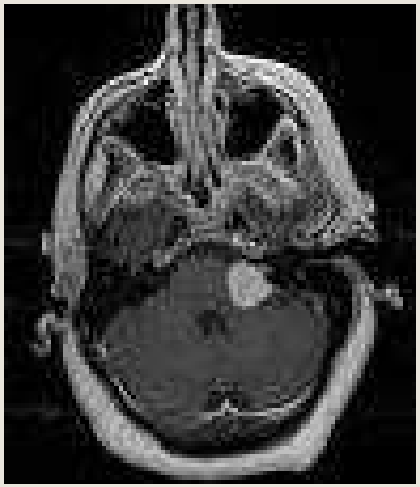
- Intracanalicular tm ≤ **5mm-**
  - ?VS, haemangioma
  - Usually asymptomatic
  - Observation with regular audiologic and radiological monitoring
- Intracanalicular tm **5-10mm-**
  - GK
  - If patient wishes surgery- Middle fossa approach

VESTIBULAR SCHWANNOMA: SURGERY OR GAMMA KNIFE RADIOSURGERY?
A PROSPECTIVE, NONRANDOMIZED STUDY. Neurosurgery. 2009 Feb 4

- Tm with CP angle extension,

  10-25mm-

  - GK
  - Surgery- Suboccipital if hearing salvagable
    - Translabyrinthine if hearing already gone

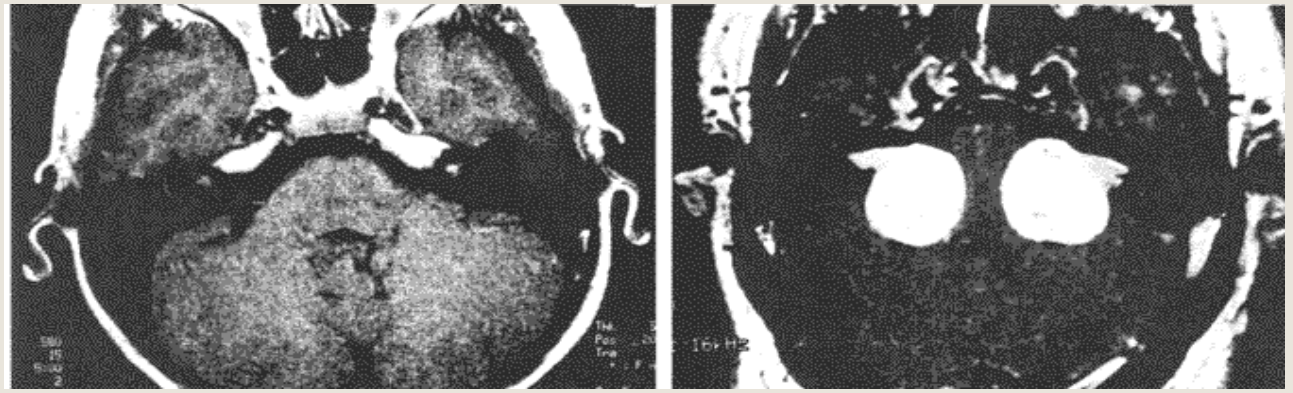


- **25-35mm-**
  - Surgery – Suboccipital
- **35-50mm-**
  - Surgery – Suboccipital – Total microsurgical excision/ Subtotal excision + Post-op GK to small residual

Only hearing ear ?

- Stable hearing – FU with MRI and PTA
- Progressive hearing loss- choice discussed with patient
- Options:
  - GK/fractionated RT
  - Subtotal removal/complete removal with IAC decompression with an attempt to save hearing
  - Translabyrinthine removal and placement of brainstem auditory implant

NF-II



- B/L vestibular schwannoma is the hallmark
- Autosomal Dominant, 22q
- *More difficult to remove surgically (more invasive, higher growth rate)*
- *Young patients, Faster tm growth*
- *Multilobulated tm, arising from multiple cranial nerves*
- *Poor VII and VIII nerve outcomes with both surgery and GK*
- *Goal: decompression of brainstem followed by preservation of facial and auditory function*
- *In general, the larger tm is operated on first*
- If facial paralysis after first surgery → surgery on opposite side is delayed until the nerve recovers or facial reanimation is performed

NF-II

Samii M.Management of vestibular schwannomas (acoustic neuromas): auditory and facial nerve Function after resection of 120 vestibular schwannomas in patients with neurofibromatosis 2.

- Chances of good outcomes are best when surgery is performed early and when there is good preoperative hearing function
- Ideal:- complete of resection with functional cochlear nerve preservation
- Subtotal microsurgical resection with functional cochlear nerve preservation in the last hearing ear

NF II : MARSEILLE GUIDELINES

- Operate worst hearing side first.
- SRS favoured whenever possible.
- One ear deaf: deaf ear 1st especially if tm larger.
- Both ear deaf: larger first

Tumor control and hearing preservation after Gamma Knife radiosurgery for vestibular schwannomas in NF2

MS Sharma, R Singh, SS Kale, D Agarwal, BS Sharma, AK Mahapatra

Journal of Neuro-Oncology Volume 98, Number 2, 265-270

- 1997 to 2008 : 30 patients with 54 VS
- Tumor control rate : 87.5% (33.3% tumor regression)
- Hearing preservation : 66.7%
- One patient : worsening of facial function.
- CONCLUSION: GKS for VS provides satisfactory tumor control and hearing preservation in patients with NF 2

Rehabilitation

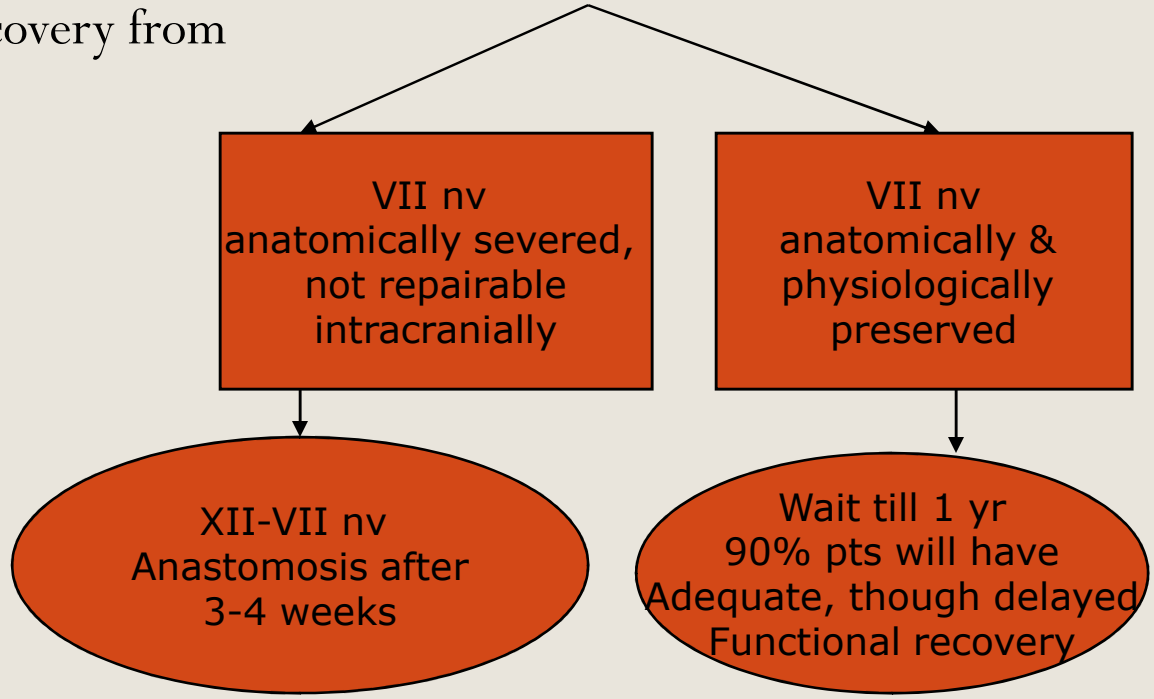
- **Problems after Vestibular Schwannoma Surgery :**

1. Facial paresis
2. Hearing impairment
3. Cerebellar ataxia
4. Lower cranial nerve paresis
5. Post- operative headache
6. Cognitive impairment and depression

Facial Reanimation

- Factors to be considered-
  - Cause & Extent of facial paralysis
  - Duration of facial paralysis
  - Likelihood of recovery from facial paralysis

- <u>Timing of surgery after post-op VII nv paralysis</u>



Facial reanimation procedures

- **Dynamic procedures-**

  improve facial tone & motor function

  - Primary nerve repair
  - Nerve grafting
  - Neuromuscular pedicle grafts
  - Regional ms. Transposition
  - Microvascular muscle transfers

- **Static procedures-**

  - add support and symmetry to the patient's face at rest
  - supplement results of nerve grafting/ dynamic procedures
    - Gold weight implantation in upper eyelid
    - Lower lid ectropion correction

- **Ideal**- directly re-establishing facial nerve continuity
- **Nerve interposition grafting-**
  - Can be performed upto 1 year after injury
  - Results best if performed within 30 days
  - Results poor if performed> 2yrs after injury, consider ms. transposition in such cases
    - Hypoglossal-facial anastomosis
    - Spinal accessory-facial anastomosis
    - Phrenic-facial anastomosis

Hypoglossal-facial anastomosis

- **Classical**
  - Baker & Conley
  - Entire proximal XII nv sutured to distal main trunk of VII nv
  - Improvement in eye closure, facial tone & facial asymmetry
  - Variable degree of tongue paralysis with resultant deglutition dysfunction
- **Modified**
  - May et al
  - *End-to-side(jump)* interposition nerve graft between XII nv and VII nv
  - Preserved tongue function in > 90% cases

Muscle transposition

- Temporalis/ Masseter muscle transposition: (when Vth nerve is preserved)-
- Alone after 2 years or along with jump graft to prevent sagging of facial muscles till graft starts functioning
- **Adv**: immediate results

Static procedures for paralyzed eyelids

- Lateral tarsorrhaphy(? cosmetic concern)
- Gold weight implantation in upper eyelid – to restore eyelid closure
- Palpebral sling placement
- Procedure to correct lower lid ectropion – implant a piece of auricular cartilage in the lower eyelid

  - 1-cm incision at the tarsal-supratarsal fold region, centered just medial to the midpupillary line
  - Subcutaneous pocket
  - 0.9.1,1.1gm weights, 1mmx5mmx10mm

Hearing Rehabilitation

Multichannel auditory brain stem implant (ABI)

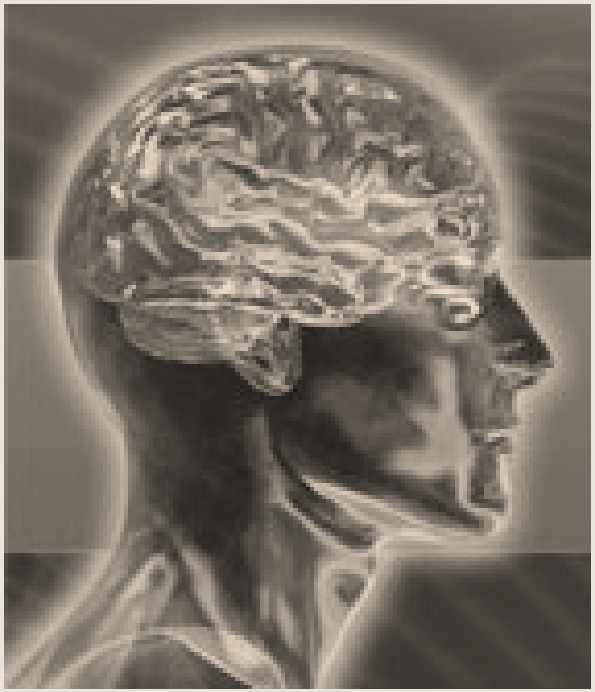
- Indi: NF-2 patients >12yrs
- Tech: direct stimulation of cochlear nucleus
- Multichannel implant placed in lateral recess
- Results: 80% patients can hear sounds
- Most patients can recognize >70% of sentence along with lip reading
- Learning period of 6-12 months

Lower cranial nerve rehabilitation

- RT feeds/ Feeding gastrostomy
- Nurse with intact side down
- Tracheostomy
- Chest physiotherapy
- Deglutition training

Cognition/ depression

- Psychological counseling
- Patient support group- Acoustic neuroma association
- Family support group



Thank you