Acoustic Neuroma

Presenter: Tim Makeham

Mentor: Sean Flanagan

Definition

Tumours arising from the Schwann cells most commonly of the vestibular portion of the eighth cranial nerve.

Expression of Schwann cell characteristics is common (expression of S-100 antigen and melanocytic differentiation)

Incidence

8% intracranial tumours. 80-90% of CPA tumours.

Rare in children without NF

Incidence of symptomatic AN 1 per 100,000 person-years.

May affect 0.07% of population (rate of incidental finding study of 24,000 MRI) to 0.7% of the population (autopsy study)

Median age at Dx 50yo

Unilateral in 90% - No preponderance for left or right.

Bilateral AN is limited to patients with Autosomal Dominant NF 2 (96% of patients with NF2 have an acoustic neuroma with a median age of onset <30yrs

NF2 incidence is 1:40 000.

Aeitiology / Risk factors for developing AN

1. 95% are spontaneuous
2. 90% of Schwannoma affect CN8 thought to be due to exposure to loud noise – demonstrated in 2 case controlled studies
3. NF2 – tumour suppressor gene on Ch22 which encodes "Merlin" cytoskeletal protein (all sporadic AN have inactivating mutations and 60% of NF2 patients have inactivating mutations).
4. History of Parathyroid adenoma
5. Exposure to radiofrequency energy may be a risk but this has not been conclusively demonstrated.

Genetics of Acoustic Neuroma and NF2

1. **Classification of Neurofibromatosis**

Neurofibromatosis type 1	
Characteristics	Autosomal dominant disorder that presents as a patient with adrenal tumours (94% of patents) in association with neurofibromas, café au lait spots, axillary freckling, optic pathway tumours and iris hamatomas It is part of a broader group of phaeochromocytoma and paraganglioma associated syndromes. (Others are: MEN2, VHL(Von Hippel-Lidau), and Paraganglioma syndromes 1, 3, 4.) *VHL is hemangioblastoma of the retina, cerebellum, spinal cord and phaeochromocytoma* Malignancy occurs in 12% of patients Nervous system tumours include: • Neuroma • Schwannoma • Meningioma • Optic glioma
Genetic locus	17q11.2 (NF 1) – 57 exons. GTPase activating protein.
Relation to Acoustic Neuroma	Patients with NF1 have intra and extra cranial tumours. <5% develop unilateral Acoustic neuroma.

Neurofibromatoses type 2	
Charateristic	Autosomal dominant condition which is charaterised by Schwannoma, Glioma, Ependymoma, meningioma. It can be thought of as a central form of neurofibramotoses.
Genetic Locus	22q12 – Merlin a cytoskeletal protein and Tumour suppressor gene

Werner syndrome: MEN 1: characteristic nervous system tumours are pituitary adenomas and malignant schwannoma.

2. **Definition Criteria of NF2**
    i. Central, multisystem genetic disorder associated with the formation of multiple intracranial and spinal tumours, most typically bilateral vestibular schwannoma
    ii. Abnormality in the development of neural crest cells
    iii. Hyperplasia, neoplasia, and dysplasias of the neuroectodermal elements and their supporting structures
3. **Diagnositic Criteria for NF 2- Manchester criteria**

    1. Bilateral vestibular schwannomas

    2. First-degree relative with NF-2 and

        1. unilateral vestibular schwannoma or

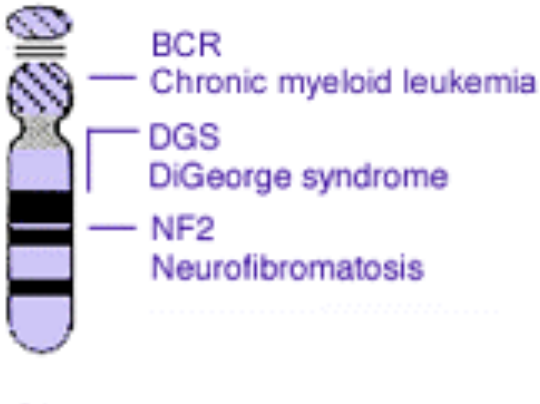
2. any 2 of meningioma, schwannoma, glioma, neurofibroma, juvenile posterior subcapsular lens opacity

3.Unilateral vestibular schwannoma and any 2 of meningioma, schwannoma, glioma, neurofibroma, posterior subcapsular lens opacities

4.Multiple meningiomas (≥ 2) and

1. unilateral vestibular schwannoma or
2. any two of schwannoma, glioma, neurofibroma, cataract

4. Genetics of NF2 and Acoustic neuroma:



i.

ii. Autosomal Dominant disorder affecting Tumor Suppressor gene encoding for the protein Merlin on long arm of Ch 22

iii. Manifestation of disease depends on mutation in both copies of Chromosome 22

iv. Chance of inheriting the disease is 50 %

v. Manifestations depend in part on the type of mutation:

- Protein truncating mutation – more severe, not much protein, receptor block
- Missense mutation – mild slight alteration in protein product (1 AA or base pair)
- Mosaicism

5. Genetic Counselling for NF2

i. Where genetic abnormality is known direct screening of the mutation can be performed using PCR techniques. It allows prenatal diagnosis.

ii. Where the exact abnormality is not known indirect methods can be used. Techniques include.

- Linkage analysis - highly polymorphic markers known to be within or close to the NF2 gene
- Track which one of the chromosome 22 pair carries the NF2 mutation
- Requires that two or more affected individuals be available for testing to track the gene in that specific family

iii.

6. Difference NF1 vs NF2

i. Diagnostic criteria for NF1

- Six or more café-au-lait macules the greatest diameter of which is >5mm in prepubertal patients, and >15mm in post pubertal patients
- Freckling in the inguinal or axillary region

- 2 or more neurofibromas of any type or one plexiform neurofibroma
- 2 or more leisch nodules in the iris
- optic glioma
- distinctive osseous lesion such as sphenoid dysplasia or pseudoarthrits
- A first degree relative with NF1 diagnosed by the above criteria

7. Clinical syndromes and manifestations

i. Wishart
- Early onset, rapid growth, intracranial tumours other than AN

ii. Gardner
- Later onset, slow growth, usually isolated VS

iii. Segmental NF2
- somatic mosaicism
- mutation occurs during embryogenesis rather than in the germline DNA
- only a portion of the patient's cells carry the mutation
- 25% of NF2 cases among patients whose parents did not have NF2
- bilateral vestibular schwannomas if mutation occurred early in embryogenesis
- If late in development - atypical presentation of segmental NF2 with a unilateral vestibular schwannoma and an ipsilateral, additional intracranial tumor, such as a meningioma
- Unlike the traditional forms of NF2, the risk of passing NF2 caused by mosaicism to future offspring is low, still possible in germ cell line

8. Specific management considerations for AN in NF2:

Role of radio surgery is controversial as there is an increased risk of treatment failure with GKR (Gamma Knife)

Rate of tumour control with GKR 50% with a greater risk of Hearing loss and FNP (Radiation therapy may work by disrupting the tumors vascular supply and NF2 tumours may have a greater capacity to express VEGF).

<u>Small AN</u>

Hearing preservation surgery

<u>Medium sized AN</u>

Manage conservately as surgery risks – total deafness, bilateral FNP, oscillopsia

Preservation surgery only if tumours are detected at an early stage and are small

<u>Bilateral large or growing AN</u>

Choice on which ear to operate on initially depends on: ABR, MRI,

If hearing is lost but cochlear nerve is preserved – cochlear implant can be considered

Alternative is for a Auditory – Brainstem – Implant

<u>Cochlear Implant</u>

- Long-term hearing outcomes in six patients who have received a CI 5/6 were able to use the telephone at 5y follow up

Cochlear Implantation in Neurofibromatosis Patients Laryngoscope 117: June 2007 Welling et al

ABI

- 160 patients have received an ABI with surface electrodes - few obtain any open-set speech understanding, although lip reading cues and awareness of environmental sound is noteworthy
- ABI good for NF2 patients whose cochlear nerves have been sectioned during tumor removal or for patients with intact cochlear nerves that do not respond with promontory stimulation.
- Multichannel penetrating electrode holds promise

Auditory Brainstem Implantation in NF2 Laryngoscope 114: December 2004 Kanowitz et al

Currently, CI performance exceeds that of ABI for most NF2 patients

Location

Origionate in the Schwann cells of the superior and inferior vestibular nerves at the transition (or Obersteiner – Redlich Zone). This is the junction of the peripheral and central myelination. The location of the zone is in the lateral CPA or medial IAC.

Therefore, these tumours arise in the IAC most commonly, but can arise in the CPA.

They are most likely to arise in the VN as the Vestibular ganglion in the IAC has the highest concentration of Scwann cells

With in the cranial vault the most common location is the CPA second most common is the trigeminal nerve

Acoustic neuroma affects the superior and inferior vestibular nerves with equal frequence. Only rarely affects the cochlear portion.

Classification

Classification by aeitiology:

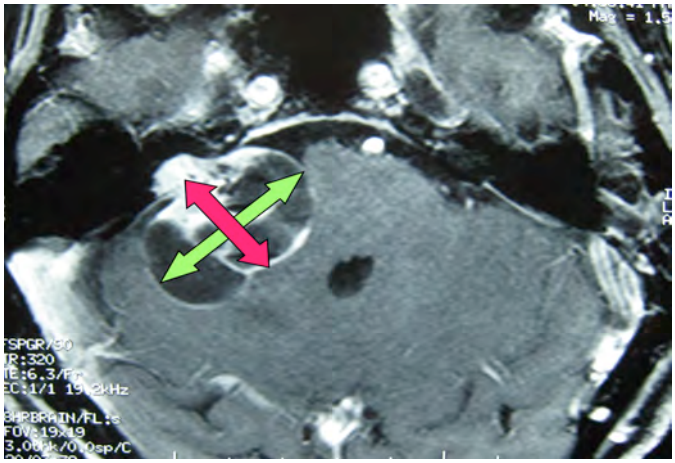
1. Sporadic
    a. Most common type, always unilateral, occurs later in life
2. NF-2 associated Acoustic Neuroma
    a. Rare, austosomal dominant disorder characterized by bilateral AN, other CN neuromas, Meningioma's, Juvenile Cataract
3. Younger age at presentation.

Classification by extent of tumour:

1. Intracanalicular
2. Intracisternal
3. Brainstem compressive

Tumour size by the AAO-HNS

- Determine max AP and ML size
  (parallel and perpendicular to post face of petrous bone)
- Square root of product AP x ML



Jackler stage	Tumour Size
Intracanalicular	Confined to IAC
Stage 1 (small)	<10 mm
Stage 2 (medium)	11-25 mm
Stage 3 (large)	25-40 mm
Stage 4 (giant)	>40 mm

Macroscopic appearance

Well circumscribed encapsulated masses that are attached to the nerve but can be separated from the nerve.

Tumours are firm, grey masses that may have areas of cystic and xanthomatous change.

Microscopic appearance

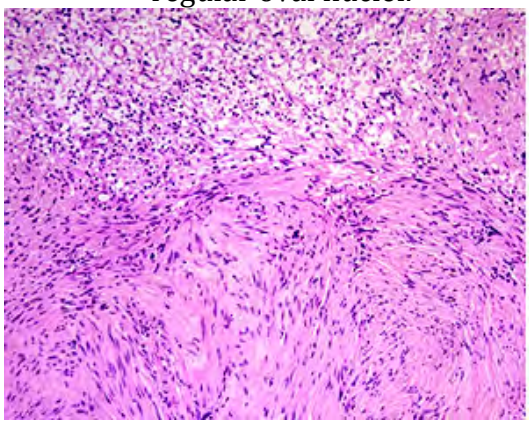
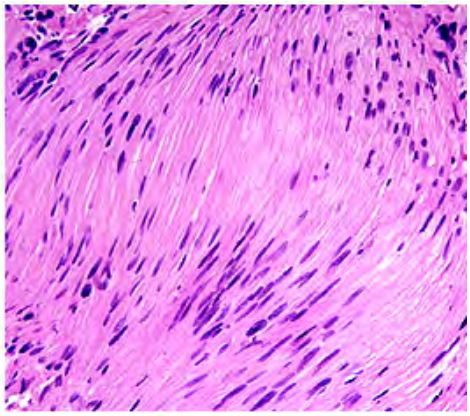
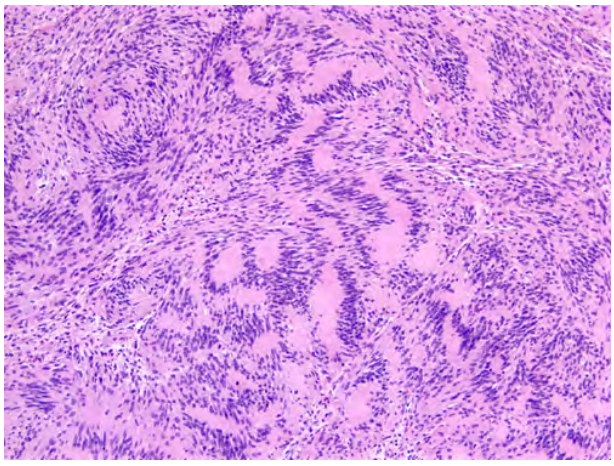
There are two growth patterns:

1. Antoni A pattern of growth –
   - Elongated cells with cytoplasmic processes that are arranged into fascicles in areas of high cellularity with little stromal matrix
   - **Verocay bodies** - these are "nuclear – free zones" that lie between regions of nuclear palisading.
2. Antoni B pattern of growth
   - Less densely cellular with a loose meshwork of cells.
   - There are regions of myxoid change and microcysts

Tumours exhibit a mixture of these two patterns of growth.

The cytology of the individual cells is the same in both regions:

- elongated cell cytoplasm
- regular oval nuclei.

There are some features of degeneration present – nuclear pleomorphism, xanthomatous change

Electron microscopy shows basement membrane deposition encasing single cells. And long – spacing collagen

Malignant degeneration to fibrosarcoma is rare (6 case reports)

Immunohistochemistry

1. Silver stains (for nerve axons) demonstrate that axons are largely excluded from the tumour – ie. The tumour displaces the nerve.
   Axons may become trapped in the capsule

2. S-100 positive – reflects Schwann cell origin

Prognostic factors and prognosis

NF2, Incomplete resection.

Natural History of Acoustic Neuroma's

The natural history of AN is unpredictable and No predictive factors of tumour growth have be identified.

*** Small tumour size and advancing age are NOT predictors of tumour growth

47% of AN demonstrate growth when followed by serial MRI

Rate of growth: ranges from 0.1 to 0.23 cm/yr

LESS THAN 1/3 (30%) of untreated AN grow greater than 0.2cm/yr.

Tumours >2.0cm are more likely to grow.

The pattern of growth can be established over the first 3 years of observation. – slow growing tumours tend to remain slow growing tumours.

Growth rate in patients with NF2 is greater:

- Average growth rate 0.4cm/yr
- More likely to invade the surrounding facial or cochlear nerve.

TABLE VII.
Literature Review of Nonsurgical Patients.

Study	Year	N	Mean Age (y)	Years of Follow-up	Growth Pattern			Growth Rate (mm/y)	Intervention (%)
					Growth (%)	No Growth (%)	Regression (%)		
Present Study	1999	80	69.7	4.4	57.8	34.6	7.7	0.91	7.5
Fucci[29]	1998	119	65	2.5	30	67	3	1.2	18.5
Selesnick[30]	1998	571	64	3.0	54	46	0		
Yamamoto[35]	1998	13	—	1.6	54	46	0	—	62
Glasscock[34]	1997	34		2.4	56	44	0	2.9	
Charabi[31]	1995	104	58.8	3.4	74	18	8	2.4	34
Wiet[28]	1995	53	66	2.2	40	60	0	1.6	—
Strasnick[27]	1994	51	68	2.6	69	31	0	1.1	24
Rosenberg[33]	1993	23	73	4.3	56	38	6	0.6	13
Silverstein[45]	1993	20	73	4.7	40	30	30	—	5
Bederson[26]	1991	70	57	2.2	53	41	6	1.6	12.9
Valvassori[24]	1989	35	—	—	57	43	1	—	—
Thomsen[25]	1988	21	59	4.2	14.3	85.7	0	—	14.3
Nedzelski[32]	1986	17	69	4.3	—	—	—	2.2	26
Silverstein[20]	1985	7	71	1.9	57	43	0	2.0	14
Wazen[19]	1985	4	75	3.4	75	25	0	2.0	0
Zollner[22]	1985	3	54	1.1	66	33	0	2	66

MANAGMENT

History / presentation

Presenting symptoms:

- Sensorineural Hearing Loss 95%
- Tinnitus 50% - 70%
- Balance Disturbance 10% but present in 50% of patients

- Trigeminal Symptoms 20%
- Headache <20%

Additional symptoms include:

- Sudden onset hearing loss – present in 5-15%
- Facial numbness 25% (10% at presentation)
- Facial weakness 10% . RARELY a presenting symptom (only in less than 2% of AN patients) - If present consider:
    - Facial nerve neuroma/ haemangioma
    - Menignioma
    - AVM
    - Cholesteatoma / CPA dermoid

Examination

Specific signs relating to acoustic neuroma and its regional extension

1. Brun's nystagmus
    1. no spontaneous nystagmus in primary position
    2. asymmetry exists at extremes of lateral gaze:
        i. low-frequency, large-amplitude nystagmus when looking to side of lesion
        ii. high-frequency, small-amplitude nystagmus when looking other way
2. Hitzelberger's sign = posterior ear canal numbness due to CN VII loss
3. Rhomberg (Sharpened Rhomberg = 1 foot in front of other) = cerebellar
4. Fikuda test = turns 90 degrees to side of lesion, 50 steps marching on spot
5. Mawson's mattress test = stand on mattress with eyes closed & lights out
    - fall over - if no function, scream when hit mattress
    - if compensated function, scream before hit mattress

In general examination should be methodical and include:

- Ocular and ocular movement examinaton
- Ear examination
- Tuning fork
- Neurological examination examining for cerebellar and vestibular assessment. Gait, FNT, HTT, Head impulse test, Rhomberg, Unteberger/Fikuda, (JPS to exclude peripheral cause)

Investigations

- **PTA**

  Hearing loss in AN patients is asymmetrical and charactersed by: (Johnson)

  - 66% have High tone lose
  - 10% have Low tone loss
  - 10% have FLAT audiograms

  Criteria for suspecting and acoustic neuroma on basis of audiological assessment:

  - Asymmetric SNHL: (inter-test variability = 5 dB)
    - 10 dB difference at 2 frequencies

    - OR > 15dB difference at 1 frequency
- Speech discrimination disproportionately decreased c.f. PTA
- Non-serviceable hearing
    - = 50/50 rule (PTA > 50dB & speech discrimination < 50%)
    - OR 30/70 rule

- **Speech discrimination**

  72% of patients have abnormal discrimination (defined as <62%)

  Lower rates (45%) of abnormal speech discrimination reported by (Hirsch and Anderson)

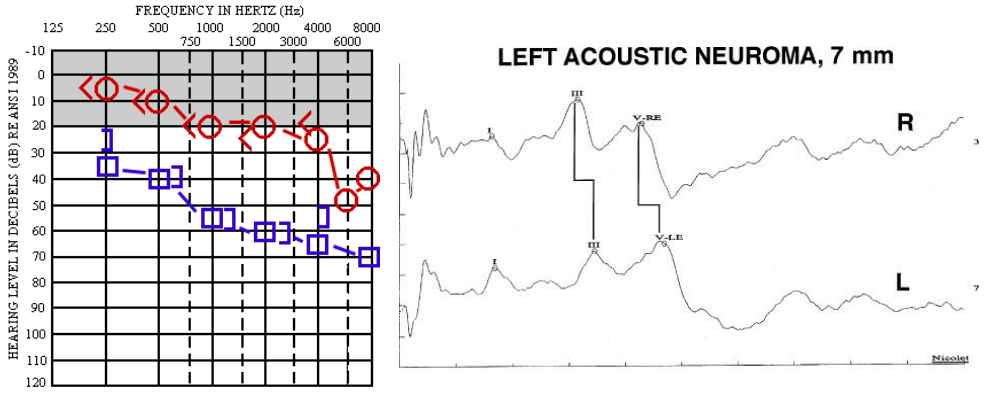
  12% have Normal speech discrimination and PTA

- **Acoustic Reflex Decay**

  Sensitivity up to 85%

- **ABR**

  Test Measures: Interaural V wave latency



Most reliable and reproducible for auditory test for acoustic neuroma. Reliability of test depends on the size of the tumour. It <u>is limited for the detection of small tumours.</u> Abnormal is a difference of >0.2ms in wave I-V latency between ears or I-V latency greater than 6ms

Tumour size	Sensitivity	Specificity
>2cm in diameter	100%	90%
1.1 – 2.0cm	98%	90%
< 1.0cm	89%	90%
Over all	93%	90%

The diagnositic efficiency of the test is reduced when hearing loss is greater than 80dB at 4kHz.

- **ENG**

  Nystagmus may be seen on physical examination and when present is Horizontal

  Vestibular testing has a limited role in acoustic neuroma (preserved ENG in known AN suggests inferior vestibular nerve origon)

- **Radiological**

  Plain radiography - not used

  CT scanning:

  Role: not generally used for the detection of acoustic neuroma's

  Appearance of AN on CT:

  - If calcified
  - Large and extend to the CPA or efface the brain stem
  - Enhancing lesions – but may not be seen in small lesions that are obscured by the bone of the IAC.

  MRI scanning

  Best seen on a contrast enchanced T1. Other sequences are a fast spin echo.

Table 25-3
COMPARISON OF SALIENT CT AND MR IMAGING FEATURES OF THE THREE MOST COMMON CPA LESIONS

	Acoustic Schwannoma	Meningioma	Epidermoid Tumor
Location	Centered to IAC	Posterior petrous wall most eccentric to IAC	Anterolateral or posterolateral to brainstem
Bone changes	Most enlarging IAC	Occasional hyperostosis	Occasional erosion
Shape	Spherical or ovoid, occasionally lobulated, acute bone tumor angle	Hemispherical, rarely plaque-like, may herniate, obtuse bone tumor angle	Variable with tendency to dumbell into middle fossa or contralateral CPA
Density	Mostly isodense, a few slightly hypodense or hyperdense	Isodense or mostly slightly hyperdense, some calcified	Mostly about CSF density, rarely denser than brain, occasional peripheral calcification
CT enhancement	Moderate to marked, often with inhomogeneous enhancement	Marked and homogeneous	Nonenhancing
Intensity T_1W image	CSF < M ≤ Gr	CSF < M ≤ Gr	CSF ≤ M < Gr
Intensity T_2W image	≤CSF	Variable	≤CSF
MR enhanced	Marked, some ''cystic''	Moderate	Nonenhancing

T_1W, T1-weighted; *T_2W*, T2-weighted; *CSF*, cerebrospinal fluid; *M*, mass; *IAC*, internal auditory canal; *CPA*, cerebellopontine angle; *GR*, gray matter.

MRI + gadolinium (best technique)

- detects down to 1 – 2 mm tumours
- enhancement of CN VII in IAC is <u>always</u> abnormal,
  - <u>Reason:</u> as no perineural plexus
  - <u>BUT</u> may normally enhance at & distal to geniculate ganglion

Fast spin echo MRI:

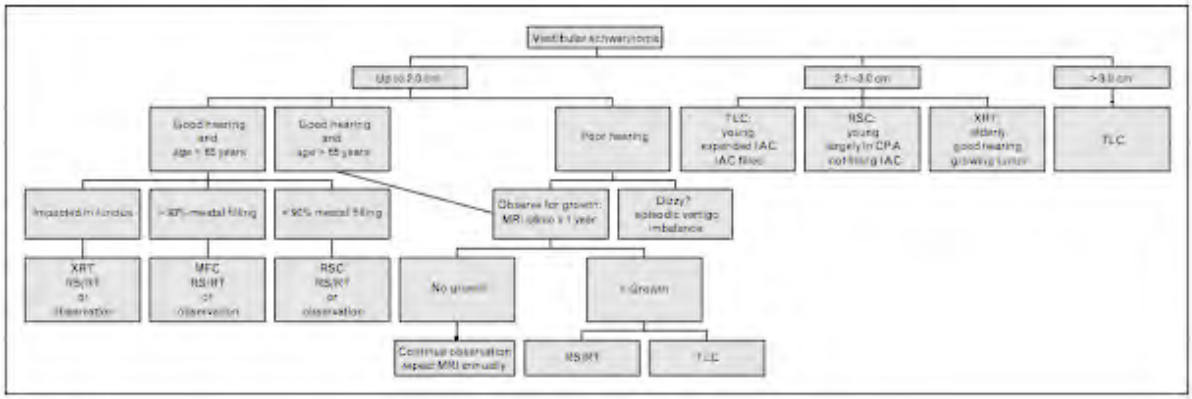
- no gadolinium needed
- cheaper

- through IAM only – thus, not for intra-axial / intra-temporal pathology
- can see intra-labyrinthine tumours
- Evaluates:
  - cochlea & cochlear nerve)
  - labyrinth) good for Paed SNHL
  - vestibular aqueduct & endolymphatic sac)

TREATMENT

Treatment strategies for patients with Sporadic AN and those forming part of NF2 should be considered separately. The general approach to treatment is summarized in the following table from the 2006 article in Current Opinions:

Figure 1 Proposed management algorithm for vestibular schwannoma



TLC, translabyrinthine craniotomy approach for removal; IAC internal auditory canal; RSC, retrosigmoid craniotomy approach for removal; CPA cerebellopontine angle; XRT, x ray therapy; MRI, magnetic resonance imaging; RS/RT, radiosurgery or radiotherapy; MFC, middle fossa craniotomy approach for tumor removal.

Treatment decision depends on :

- **Patient factors**: Age, medical comorbidity, hearing status in contralateral ear, Patient preference / perception of risks
- **Disease factors**: Size of tumour, Bilateral AN, Presence of cystic degeneration, compression of surrounding structures
- **Institution factors**: Availability and experience of MRI / LINAC; Experience with different operative approaches and philosophy of department/institution.

Broad – guiding principles to management:

- Complete clinical assessment including History, examination and investigation with T1 post contrast MRI, PTA, and Speech discrimination
- <u>Tumour size</u>
  - Determine measurement by the greatest diameter taken through the midportion of IAC and including intracanilicular portion
- Conservative treatment

- Hearing preservation surgery
    - Vestibular AN – where size is <2cm, and hearing
    - Better than 50dB with speech discrimination better than 50% (AAO-HNS A or B)
    - Size and PTAvg are only two to significantly correlate with hearing preservation
    - Additional factors: size and shape of tumour, institutional preference, patient preference, surgical expertise.
- Specific considerations for NF2 and bilateral AN:
- Surgery should be performed in the setting of
    - Tertiary institution with appropriate neurosurgical and ICU Support
    - Intraoperative facial nerve monitoring.

Conservative treatment

Indications:	Patient factors: • Elderly patient • Medically unwell patient • Tumor affecting better hearing ear • Reliability of patient to comply with long term follow up Disease factors: • Small tumor (<1.5cm) • Minimal symptoms • Slow growth rate (<0.2cm /yr)
Frequency of scans	MRI – T1 (fat suppressed) with contrast / T2 / STIR • Second scan at 6 months • If no change then annual MRI
Duration:	Scans need to be performed indefinitely on an annual basis If minimal change after 5 – 7 years and patient elderly • Perform CT with contrast every 18 months – 2 years
Indications for surgery	Operate if: • Tumour grows > 1.5cm

"Radio-surgery"

Goals of Rx	Control of tumour growth with preservation of Cranial nerve function
General aspects	Growth control achieved by DNA damage to Schwann cells and to the supporting vasculature Evidence of radiation apoptosis has been reported Apoptosis may be associated with tumour shrinkage: • in the majority of cases, the vestibular schwannoma displays size reduction over time • there are two caveats to these reported findings, however: tumour is still present and over time could theoretically undergo revascularization and regrowth • there are no long-term follow-up studies (>10 years) reporting solely on results of the newer low-dose radiotherapy regimens (20 Gy maximal and 12–13 Gy

	marginal dose) • in fact, regrowth of vestibular schwannomas after many years of quiescence has been reported in cases requiring surgical salvage Tumour will remain after RT and may increase by 1-2mm in the 6 months after treatment Vestibular symptoms may not resolve Hearing may decline in a delayed fashion after treatment Small risk of malignant transformation
Treatment options	1. LINAC – Linear accelerator 2. GKR - Gamma Knife = 201 Co^{60} sources on a sphere aimed at target ▪ 90% stop growing after RTH ▪ Single session treatment of 12Gy 3. Fractionated stereotactic multisession radiotherapy
Indications	Patient: Elderly / Medically unwell Disease: • Tumour volume up to $8cm^3$ or size <2cm • Rapidly growing tumour
Contra-indications	Abslolute: Brainstem compression, Large tumours Relative: Tumours with cystic change
Advantages:	Non-invasive GKR has decreased the dosage amount Rate of tumour control with GKR according to radiation dose: • 100% with 35Gy • 96% with 12Gy • 97% with 13Gy Complication rate is proportional to radiation exposure. Rate of FNP related to dose: • 38% with 35Gy • 1% with 12 – 13Gy
Side effects:	Can recur after 2 years Makes surgery more difficult *Trigeminal Neuralgia:* 3 – 5% of patients with GKR *Hemifacial spasm:* Post treatment: *Hydrocephalus, cerebral oedema delayed peritumoural cyst enlargement, brainstem infarct.* Hearing: 50% - in AAO HNS class A or B Facial nerve: delayed paresis in 20 – 30% (usually temporary) Sarcomatous change: may occure Hydrocephalus: 5% Trend to poorer QOL with radio Sx

Surgical excision

Translabyrinthine	
Indications	• Small growing tumours < 2cm, with NO serviceable hearing: - unable to use telephone, OR - 30dB 4 tone average; 70% speech discrimination • large tumours > 2cm • NF 2 patients where hearing preservation is not an option and concurrent ABI is being planned.
Advantages	• best facial nerve outcome • best access for tumours in all positions • no brain retraction • Can be extended to translab/trans cochlear for clival extension
Disadvantages	• hearing destroyed • fat graft required – no primary dural closure
Contraindications	only-hearing ear
Follow up	MRI at 1 and 5 yrs post op to screen for recurrence

Retrosigmoid	
General	Hearing preservation achievable for AN up to 2cm or <1.5cm medial to the porous acousticus
Indications	Reserved for AN that do not extend far laterally in IAC • <90% of the length of IAC • >3mm of space from the fundus of the IAC
Advantages	• Hearing preservation rate 22-58% o Risk of delayed hearing loss • good inferior access for large CPA tumours • Facial nerve preservation slightly better than other techniques for AN >2cm
Disadvantages	• Incomplete tumour removal: o VN in the lateral portion of IAC o Lateral dissection is limited by posterior SCC, common crus, or endolymphatic duct • *Headaches*: high incidence of headaches post-op (bone dust in CSF) • difficult access for intracanalicular tumours • cerebellar retraction (oedema, infarct, haematoma) o Increased risk of *gait instability* o *Ipsilateral cerebellar atrophy* seen on follow up MRI • higher CSF leak rate
Contraindications	Relative: intracanalicular extension
Follow up	MRI at 1 and 5 yrs post op to screen for recurrence

Middle Cranial Fossa approach	
Indications:	good hearing < 1.5cm intracanalicular (lateral) tumour

<table>
<tr><td></td><td>(< 0.5cm extension into CPA) in young patient</td></tr>
<tr><td>Advantages</td><td>Hearing preservation:
60- 73% of patients
Lower when tumour is lateral in the IAC
Lower risk of delayed hearing loss as no risk of entry into the endolymphatic duct.
<table><tr><td>Tumour size</td><td>Hearing preservation rate</td></tr><tr><td>< 1.0cm</td><td>76%</td></tr><tr><td>1.0 to 1.5cm</td><td>39%</td></tr><tr><td>1.5 to 2.5 cm</td><td>33%</td></tr></table></td></tr>
<tr><td>Disadvantages</td><td>limited medial access
Postoperative seizures: temporal lobe retraction
Transient dysphasia / hemipareisis / memory impairment

worse facial nerve outcome
No statistically significant difference for size matched tumours
Trend toward better outcomes in TL approach
Higher rate of FNP where extension to CPA / inferior nerve origon of tumour
Tumours <1.0cm – 94% HB I, 100% HB grade I or II
CSF leak rate the same as for TL (1 – 5%)</td></tr>
<tr><td>Contraindications</td><td>Elderly
medically unwell
large CPA component (> 1cm)</td></tr>
<tr><td>Follow up</td><td>MRI at 1 and 5 yrs post op to screen for recurrence</td></tr>
</table>

Summary of treatments:

	Advantages	**Disadvantages**	**Indication**
Translab	Best facial n results	Dead Ear	Large tumours > 2cm
	No size / position limitations		Small tumours + no serviceable hearing
	Limited cerebellar retraction		Good contralateral hearing
MCF	Hearing preservation	Temporal lobe retraction	<1cm in IAC
		Medial access poor	Good Hearing

		Worst VII outcome	
Retrosig	Hearing preservation Good inferior access large CPA tumours	Cerebellar retraction Intradural drilling – severe headache Lateral access poor	<1.5cm Not in IAC Good Hearing

Outcomes with surgery

Mortality	0.5%
Recurrence	0.5 – 2%
Facial nerve	Temporary • 10 – 30% delayed palsy • Majority make complete recovery Permanent • Related to the size of the tumour • Portion of patients with HB I – II outcome by size: **Size of tumour** / **Portion with HB I/II** <1.5cm / > 90% 1.5 – 3cm / 85-90% >3cm / 50- 75%
Hearing	17 – 65% in AAO-HNS grade A or B with hearing preservation approaches For translab – dead ear No difference between hearing preserveing surgery and gamma knife
Dizziness	
QOL:	54% have worse QOL poorer with larger tumour size Younger patients more affected by facial dysfunction Most difficult aspects: Hearing Loss, FNP, Eye problems, Headache

AAO-HNS CLASSIFICATION

CLASS	PURE-TONE AVERAGE (0.5, 1, 2, 3 kHz)	SPEECH DISCRIMINATION (%)
A	≤ 30 dB	≥ 70
B	≤ 30 dB	50 - 69
OR	30 – 50 dB	≥ 50
C	≥ 50 dB	≥ 50

D	any	≤ 50

Differential Diagnosis

- Acoustic Neuromas 80%
- Meningiomas 10 %
- Cholesteatomas / Epidermoids 5 %
- Lipomas
- Arachnoid Cysts
- Aneurysms
- Metastases

Clinical manifestations of NF2: