VESTIBULAR SCHWANNOMA (Acoustic Neuroma)

- **DEFINITION**
  - Vestibular schwannoma (VS) is a **benign tumour** arising from abnormally proliferative Schwann cells, which envelope the lateral portion of the vestibular nerve in the internal acoustic meatus.

- **PATHOLOGY**
  - Site of Origin ➔ **OBSTIENER REDLICH ZONE** i.e. distal portion of 8th cranial nerve where neural elements cease and is a zone of cellular instability usually found in IAM or CP angle.
  - There are plenty of Schwann cells at this zone of instability.
  - Common nerve of origin is Superior vestibular nerve.

- **HISTOPATHOLOGICAL EXAMINATION**
  - **Antoni A** pattern (Fasciculated pattern)
    - In which there are closely packed cells with small spindle-shaped and densely stained nuclei. A whorled appearance of Antoni type A cells is called a **Verocay body**.
  - **Antoni B** pattern (Reticular)
    - In which there is a **looser cellular aggregation of vacuolated pleomorphic** cells. Cystic acoustic neuromas with predominant Antoni B cells have high propensity of unpredictable and sudden growth.
    - In any particular VS, one type of cellular pattern may predominate or both types can be completely admixed.
ETIOLOGY

- Unknown, Chr 22q may be responsible for the development of both the unilateral sporadic VS and the bilateral VS in neurofibromatosis type 2.
- Because the tumour develops in the nerve sheath, it compresses rather than invades the nerve on which it arose, thereby leaving a plane between the nerve fibres and the tumour.
- As the VS grows, it gradually fills all the internal acoustic meatus and eventually protrudes out of the porus.
- The degree of bone resorption of the internal acoustic meatus varies in VS.

GROWTH AND EXTENSION

- Slow growing → 0.02 cm/year
- Medium Growing → 0.2 cm/year
- Fast Growing → 1 cm per year
- Erosion through the bony meatus
- Extra – meatal expansion of the tumour into the relatively large and empty pontine cistern initially develops silently.
- Growth and extension in this direction causes some displacement and stretching of the seventh and eight cranial nerves on the anterior aspect of the tumour and of the anterior inferior cerebellar artery (AICA) on the inferior aspect.
- Blood supply from AICA and Middle meningeal artery.
- Angioneogenesis is visible at surgery, with new small vessels running from the porus to the extrameatal portion of the tumour.
- After further growth, the tumour expands sufficiently to touch and compress the cerebellum and trigeminal nerve.(around 2cm tumour size), Even 9,10,11 nerves displaced.
- During this process, the seventh and eight nerves are thinned or ribboned, become compressed and even more stretched.
- At the same time, the internal acoustic meatus continues to become more and more widened.
- The tumour may extend to the tentorium and can obstruct the cochlear aqueduct.
- Over time, the trigeminal and abducens nerves become stretched over the surface of the tumour and thinned in much the same way as the facial nerve.

SO → IAC → pontine cistern (7 & 8 nerves stretched anteriorly and AICA inferiorly) → Cerebellum & 5 nerve (7 & 8 ribboned by now) → IAC widened further by now → tentorial extension leading to obs. of Cochlear Aqueduct → 5 & 6 nerves thinned out by now.

CLASSIFICATION

- Is based on the CT based size of the largest extrameatal diameter.

<table>
<thead>
<tr>
<th>Jackler system</th>
<th>Classification of VS according to size</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intrameatal tumour</td>
<td>Extrameatal size</td>
</tr>
<tr>
<td>Grade 1</td>
<td>Small</td>
</tr>
<tr>
<td>Grade 2</td>
<td>Medium</td>
</tr>
<tr>
<td>Grade 3</td>
<td>Moderately large</td>
</tr>
<tr>
<td>Grade 4</td>
<td>Large</td>
</tr>
<tr>
<td>Grade 5</td>
<td>Giant</td>
</tr>
</tbody>
</table>
SYMPTOMS AND SIGNS:

- **CLINICALLY**:
  - Otological Stage
  - Stage of Vth nerve involvement
  - Stage of Brainstem and cerebellar compression
  - Stage of rising Intracranial tension
  - Terminal Stage

OTOLOGICAL STAGE ➔

- **DEAFNESS**
  - U/L SNHL, gradual 2-20yrs.
  - DUE TO = Direct compression of 8th nerve and also interruption of blood supply of auditory nerve and cochlea.

- **TINNITUS**
  - I/L, high pitched continuous
  - Same mech
  - 4000 Hz dip on PTA (2000 Hz dip is Otosclerosis, 4000Hz dip is Acoustic Neuroma, 3 to 6000Hz dip NIHL)

- **VESTIBULAR DYSFUNCTION**
  - Small tumours = Vertigo, Large tumours = Dysequilibrium

STAGE OF TRIGEMINAL NERVE INVOLVEMENT ➔

- Absent corneal reflex
- **Facial nerve involvement** leads to facial numbness and facial muscle weakness

STAGE OF BRAINSTEM AND CEREBELLUM INVOLVEMENT ➔

- Ataxia, dysmetria, past pointing, dysdiadokokineses, disturbance of gait, intention tremors, Spontaneous nystagmus, dysnergia. (Cerebellar signs Usually seen when tumour >4cm size)

STAGE OF INCREASED INTRACRANIAL PRESSURE ➔

- Headache, Vomiting, neck rigidity, raised ICT.

TERMINAL STAGE ➔

- Failures of vital centers in brainstem

SIGNS:

- **HOUSE AND HITSELBERGER'S SIGN** ➔ Loss of sensation of posterior superior meatal wall

DIAGNOSIS:

- CT scan. Nowadays, all patients with unilateral audio vestibular symptoms that cannot be readily explained should proceed to MR imaging with gadolinium enhancement.
**SPECIAL MANIFESTATIONS OF VS**

- An extraordinary large internal acoustic meatus with extensive bone resorption is found in up to 11 percent of small- and medium-sized tumours with an extrameatal diameter of 1-25 mm.
- Spontaneous haemorrhage and calcification within the tumour is seen occasionally.

**Medial vestibular schwannoma**

- The medial VS is defined as a tumour without lateral extension into the internal acoustic meatus. In this situation, the internal auditory canal is filled with yellow cerebrospinal fluid which has a high protein content.
- Typically, all four nerves are clearly visible in the empty fundus (Superior Vestibular, Inferior Vest., Acoustic nerve and facial nerve).
- The tumour is located mainly extrameatally and the internal acoustic meatus is not widened in the majority of cases. Why this should happen is not known.
- The smallest medial tumour had an extrameatal diameter of 15 mm and the size of medial tumours was significantly larger, and the involvement of the cerebellum and the brain stem significantly higher than in the nonmedial tumours.
- Most of the medial tumours with normal internal acoustic meatuses were giant and large tumours with cerebellar and trigeminal symptoms and relatively better hearing.

**Cystic vestibular schwannoma**

- Cyst formation within VS are seen regularly and are easily detected by MR. This has been thought to represent degenerative change or coalescence of micro cysts in Antoni A tissue.
- More recently, it has been shown that cystic tumours contain an increased amount of Antoni B tissue which is surrounded by a membrane-like structure composed of Antoni A type cells. SO MIXED
- Therefore, three criteria are required to be present before a tumour can be termed 'cystic'.
- First : There must be a hypo dense/hypo intense area on CT/MR.
- Second : Per operative identification of the cystic elements must be achieved.
- Third : there must be histological verifcation of S-100 positive membrane.
- Surgical outcome of cystic VS is less favourable than that of solid tumours of comparable size.
- The large size of these tumours at diagnosis, and their cysts which can increase in size dramatically, have been considered by some to represent a contraindication for treatment by either radiotherapy or 'wait and scan' policy.

**INCIDENCE OF VESTIBULAR SCHWANNOMA**

- Three categories.
  1. Asymptomatic or silent tumours
  2. Symptomatic undiagnosed tumours
  3. Diagnosed tumours

**SURGICAL APPROACHES**

- It is interesting to look at the history of the two main
• Surgical approaches now in common use, **the Retrosigmoid (suboccipital) approach and the translabyrinthine approach**.

• **SURGICAL APPROACHES TO THE CEREBELLOPONTINE ANGLE**

### TRANSLABYRINTHINE APPROACH

- **This is now the favoured approach for the removal of VS for the majority of neurotologists.**

- **Extent**: It can be extended **posteriorly** to allow further access to the **posterior fossa, jugular foramen, hypoglossal canal and foramen magnum**.
- It can be extended **anteriorly** to allow access to the petrous apex and clivus (transcochlear approach).
- It can be extended superiorly to allow access to the middle fossa.
- Inferiorly to allow access to the jugular foramen and upper neck.
- Morrison and King combined the translabyrinthine approach with the middle fossa approach as the **translabyrinthine transtentorial approach for large or giant tumours**.
- The technique gives excellent exposure of the tumour and of the brainstem, but there is an unacceptable incidence of postoperative epilepsy and occasional temporary dysphasia if the dominant temporal lobe is retracted.
- The **key stages in the operation** are:
  1. Skin and periosteal flaps.
  2. Extended cortical mastoidectomy.
  4. Skeletonization of the jugular bulb and vertical portion of the facial nerve.
  5. Skeletonization of the internal auditory meatus.
  6. Identification of the facial nerve at the lateral end of the internal meatus.
  7. Opening of the posterior fossa through the dura of the posterior surface of the petrous bone.
  9. Closure with obliteration of the middle ear and petrosectomy defect, usually with abdominal fat.
- **POSITION**
- The patient is placed on the operating table in the supine position with the head turned 30° away from the surgeon and supported on a soft head ring. Two-channel neuromonitoring for the facial
nerve is usually sufficient, but with very large tumours it may be necessary to monitor the lower cranial nerves as well.

- **SKIN INCISION**
  - For tumours up to about 2.5 cm intracranial diameter the incision can be about 3 cm behind the postauricular sulcus but for larger tumours the incision should be sited further back.
  - The anterior limb should extend to a line tangential to the anterior wall of the external auditory canal.

- **MUSCULOPERIOSTIAL FLAP**
  - Pedicled superiorly or anteriorly. The superiorly based flap, favoured by the authors, has the advantage that, if necessary, it can easily be extended upwards to allow access to the middle cranial fossa.

- **CORTICAL MASTOIDECTOMY**
  - Bone is removed up to the middle fossa dura, exposing it widely, both over the floor of the middle fossa but also some 3 or 4 cm up the squamous portion of the temporal bone.
  - In a similar manner, bone is removed from the sigmoid sinus and from the bone overlying the posterior fossa dura for 2 or 3 cm behind the sinus.
  - Care must be taken to avoid damage to the **superior petrosal sinus**, which runs in the angle between the middle and posterior fossa dura.
  - Bleeding from the superior petrosal sinus and, indeed, even from the sigmoid sinus is easily controlled with pressure and the application of Surgicel or similar haemostatic mesh.

- **BONY LABYRINTHECTOMY**
  - A standard total bony labyrinthectomy is performed.
  - Care must be taken in drilling out the ampulla of the posterior canal, which lies medial to the second genu of the facial nerve.
  - The ampulla of the superior semicircular canal should be retained, as it is a landmark for the superior vestibular nerve.
  - In drilling out the superior canal, the surgeon will encounter the subarcuate artery.
  - The bone over the posterior fossa dura between the labyrinth and the anterior margin of the sigmoid sinus should be removed.
  - Skeletonization of the jugular bulb and the vertical portion of the facial nerve.
  - The jugular bulb is the lower limit of bone removal and in nearly all cases bone should be removed down to its level.
  - The retrofacial air cells are exenterated and bone may be removed over the vertical portion of the facial nerve until the sheath is visible through the bone.

- **SKELETONIZATION OF THE INTERNAL MEATUS**
  - A U-shaped gutter is drilled above, behind and below the internal meatus.
**Superiorly**, bone is removed between the meatus and the dura of the middle cranial fossa, from which the bone should be removed to allow superior elevation of the temporal lobe and easier access.

One should constantly keep in mind the position of the facial nerve in the **anterosuperior** quadrant of the meatus.

**Inferiorly**, one frequently encounters the cochlear aqueduct in a position superomedial to the jugular bulb. It is heartening if a brisk flow of CSF results as this allows the intracranial contents to slacken off and ease the subsequent opening of the posterior fossa.

The cochlear aqueduct is a useful guide to the position of the lower cranial nerves, which are just anteroinferior to the duct. Subsequent bone removal round the meatus should be above the level of the aqueduct.

At the **lateral** end of the meatus the transverse crest and the canal for the SVN should be sought. The latter runs from the lateral end of the meatus towards the retained ampulla of the superior semicircular canal, and is a constant and reliable landmark, it runs from 12 o'clock to 6 0' clock in the surgical field.

**Opening the posterior cranial fossa**

This is done through a U-shaped dural flap, based laterally close to the lateral sinus. ped dural flap, based laterally close to the lateral sinus. The upper limb is close to the superior petrosal sinus and the lower limb close to the jugular bulb. The medial limb is at the level of the porus.

The dura of the internal meatus should be cut from lateral to medial at the level of the transverse crest.

**Identification of the facial nerve**

It runs along the anterosuperior quadrant of the meatus as far as the porus where it is displaced to a variable extent anteriorly and/or superiorly. Thus, in the translabyrinthine approach, the tumour is usually between the surgeon and the facial nerve.

At the lateral end of the meatus, the SVN has already been identified.

It is also useful to try to identify the facial nerve on the brainstem at the earliest opportunity.

With a small tumour it may be immediately obvious in front of the audio vestibular nerve and usually separated from it by a loop of the anterior inferior cerebellar artery (AICA).

Once both proximal and distal ends of the nerve have been identified the surgeon starts to form a mental image in his mind of the likely course of the nerve in relation to the tumour.

**Tumour removal**

With tumours confined to the internal meatus or with little intracranial extension, dissection can start at the fundus and proceed medially, keeping to the arachnoid plane.

With larger tumours debulking of the inside of the tumour is carried out so that the tumour is converted from a solid ball to a hollow ball.

As the tumour bulk reduces it becomes progressively easier to manipulate the tumour capsule and careful retrograde dissection of the capsule off the brainstem end of the facial nerve.

When dissecting at the lower pole one must be careful to protect the lower cranial nerves and AICA.

When dissecting the tumour off the facial nerve in the internal meatus, he is within the arachnoid plane. When dissecting in a retrograde manner from the stem, he is outside the arachnoid plane. Failure to appreciate this subtle difference in location carries the risk of dividing the nerve.
In the internal meatus and close to the brainstem the plane between the facial nerve and the tumour is usually quite easy, but at a point at or just medial to the porus it may be almost impossible to identify and sharp dissection may be necessary to get the tumour off the nerve. This is the point where the facial nerve is most likely to be lost.

- The surgeon may, in fact, decide to leave a small nubbin of tumour on the nerve at this point to reduce the risk to the facial nerve.
- There is good evidence that these small fragments become devitalized and either disappear on follow-up scanning, or at least, seem to remain biologically inert.
- Special attention should be drawn to the unique surgical problems with the so-called medial tumour that arises in the CPA without involving the internal meatus and is usually large at presentation.
- After tumour removal, haemostasis must be secured.

**Closure**
- CSF fistula remains one of the most common postoperative problems
- To minimize the risk, careful obliteration of the middle ear and the temporal bone defect is essential.
- The temporal bone defect is obliterated with abdominal fat either in strips or in one large piece.
- The periosteal flap is then sutured back over the fat and the skin closed in two layers.

**MIDDLE FOSSA APPROACH**
- The middle fossa approach is one of the possible routes of access for hearing preservation surgery.
- The internal meatus is approached extradurally from above through a small craniectomy.
- The maximum size of tumour that can comfortably be removed is approximately 1-1.5 cm in intracranial diameter which is in fact about the limit beyond which the possibility of hearing preservation recedes.
- Furthermore, there is a small but real risk of epilepsy following extradural retraction of the temporal lobe.
- The key stages in the middle fossa approach are:
  1. Skin and soft tissue incisions.
  2. Middle fossa craniectomy.
  3. Extradural approach to upper surface of temporal bone and to posterior fossa.
  4. Skeletonization of internal meatus.
  5. Identification of facial and vestibular nerves.
  7. Closure.

**DETAILS OF THE MIDDLE FOSSA APPROACH**

**Position**
- The patient is supine with the head on a head ring or in a neurosurgical clamp. The essential point is that the intermeatal line should be perpendicular to the floor.
- This ensures that the internal meatus is in the same plane as the external meatus.

**Incision**
- A 6-7 cm vertical or gently backward curving incision starts at the level of the zygomatic arch just in front of the pinna.

**Craniectomy**
A 5 x 5 cm square bone flap is cut with about two-thirds in front of the intermeatal line and one-third behind it.

The dura over the temporal lobe is exposed. The lower edge of the craniectomy should be at the level of the floor of the middle fossa.

**Exposure of upper surface of petrous bone**

The dura is elevated off the surface of the petrous pyramid.

This is facilitated considerably by the administration of intravenous mannitol at this stage and the dura can be elevated as far medially as the petrous ridge and the superior petrosal sinus.

A middle fossa retractor is introduced.

As the dura is elevated, the greater superficial petrosal nerve is identified running up from the region of the middle meningeal artery to the geniculate ganglion.

The arcuate eminence is the other important landmark to be identified. It corresponds approximately to the superior semicircular canal.

**Location of the internal auditory meatus**

Two favoured approaches to the internal meatus.

The geniculate ganglion is identified and the facial nerve is followed medially along its labyrinthine segment until the meatus is reached. This method involves drilling between the cochlea anteriorly and the superior semicircular canal posteriorly.

If either of these structures is breached, the hearing will be lost.

The other approach was suggested by Fisch and others. The angle between the line of the GSPN and the plane of the superior semicircular canal is bisected and that gives the line of the internal meatus.

The internal meatus is skeletonized anteriorly, superiorly and posteriorly. At the lateral end Bill's bar is identified and the ampulla of the superior canal blue-lined. Medially, the porus is skeletonized and the dura of the posterior fossa increasingly exposed.

**Identification of the facial, cochlear and vestibular nerves**

The facial nerve is identified in the anterosuperior quadrant

The cochlear nerve is concealed under the facial nerve and cannot be seen at this stage.

**Tumour removal**

The surgeon must recognize that the facial and cochlear nerves lie in the arachnoid plane. Debulking of the inside of the tumour can proceed safely both inside the meatus and in the posterior fossa.

One major difference from the translabyrinthine approach is that the facial nerve lies between the surgeon and the tumour and is thus more vulnerable to damage from instrumentation.

**Closure**

The internal meatus is closed with a free muscle plug.

The free bone flap is replaced and secured with nonabsorbable ties and the muscle and skin closed in layers.

**Retrosigmoid approach**

A very large portion of the occipital bone was removed, from the transverse sinus above to the foramen magnum.

Cerebellar retraction may be necessary. Postoperative headache > translabyrinthine operation

**DETAILS OF THE RETROSIGMOID APPROACH**
- **Positioning**
  Neurosurgeons will favour the lateral position or parkbench position with or without the use of a clamp.
  Neurotologists may find that the supine position with the head turned to the opposite side provides access just as easily.
- **Incision**
  A vertical or slightly curving incision is made about 3 cm behind the mastoid process, from above the level of the transverse sinus to the level of the tip of the mastoid.
- **Craniotomy and exposure of the tumour**
  A 5 x 5 cm craniotomy is made using the drill, taking the mastoid emissary vein as the starting point.
  The anterior and superior limits of bone removal are the sigmoid and the transverse sinuses, respectively.
  The dura is opened through a U-shaped flap based anteriorly on the posterior edge of the sigmoid sinus or simply opened in a cruciate fashion.
  The cerebellar hemisphere is exposed & retraction is needed.
- **Tumour removal**
  Intracapsular debulking is safe and allows one progressively to handle the capsule of the tumour. It is desirable to identify the facial and audiovestibular nerves on the brainstem as quickly as possible, and to note the position of the AICA.
  For complete tumour removal the IAM must be drilled out.
- **Closure**
  Dural repair, bone dust and tissue glue which sits on the dural repair and fills the bony defect.
  A separate periostial flap is then used to cover the pate repair and the skin and subcutaneous tissues closed in layers.

The auditory brainstem implant is increasingly used to rehabilitate patients with neurofibromatosis type 2 (NF2) who are totally deaf and have no cochlear nerves as a result of their bilateral VS or the surgery to remove them.

- **OUTCOMES AND COMPLICATIONS**
  - Death, stroke, haemorrhage, brain injury - around 1 percent or less in specialist centres.
  - The actual causes of death have not changed much over the years: vascular events and their effects on the brain, and general medical complications associated with major surgery such as myocardial infarction, pulmonary embolus and pneumonia.
  - **ARTERIAL BLEEDING**
    - Serious injury to the main trunk of the AICA requiring clamping, is of grave significance and carries a high risk of death.
  - **VENOUS BLEEDING**
    - Mastoid emissary vein (readily controlled by a combination of bipolar cautery and the use of bone wax), Jugular bulb (compression using bone wax and a neurosurgical cottonoid).
Excessive compression of the jugular bulb should be avoided as it may give rise to lower cranial neuropathies.

During tumour removal itself, the petrosal vein (Dandy’s vein) is invariably encountered.

**POSTOPERATIVE HAEMATOMA**

- The risk of a postoperative haematoma is greatest in the first 72 hours.
- Failure to regain consciousness after surgery is the classic sign of a posterior fossa haematoma and necessitates immediate re-exploration of the operative site.

**BRAIN INJURY**

- Cerebellum
- Brainstem
- Temporal lobe and seizures

**CRANIAL NEUROPATHIES**

- **Facial nerve**
  - <1.5 cm tumour size facial preservation can be near 100% otherwise don’t be fool to try excising every bit of tumour

- **Nervus Intermedius**
  - Difficulties with a dry eye (even with normal facial and trigeminal function), altered taste and dryness of the nose and mouth can be very troublesome for the patient.

- **Trigeminal Nerve**
  - Tumours less than about 2 cm intracranial diameter do not usually come into contact with the trigeminal nerve.

**ABDUCENS AND TROCHLEAR NERVES**

**LOWER CRANIAL NERVES**

- CSF leakage
- CSF otorrhoea
- CSF rhinorrhoea

**INTRACRANIAL INFECTIONS**:

- Meningitis, encephalitis, cerebritis.