# Vestibula schwannoma

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**Introduction:**

Also known as Acoustic neuroma is the most common tumor involving the cerebellopontine angle. It constitutes nearly 80% of CP angle tumors. During the past century the management goal of vestibular schwannoma has shifted from that of total resection to functional preservation.

These are benign slow growing lesions that could present as both intracranial / extracranial mass. Symptoms may be minimal as long as the mass is confined to the internal acoustic meatus, when it extends out of the IAM to occupy the cerebellopontine angle then compression symptoms begin to manifest.

**Location:**

Vestibular schwannoma (Acoustic neuroma) commonly originates from the vestibular division of the $8^{th}$ nerve sheath. It commonly arises from the area of transition between the central and peripheral myelin which happens to be near the vestibular ganglion at the fundus area of internal auditory meatus.

These tumors are asymptomatic during early phases. Even if diagnosed in early stages, it is purely an incidental finding reflected in imaging studies performed for other problems.

**Epidemiology:**

Incidence of these tumors as per published records is about 10-15 / million a year. This is the most common tumor involving the cerebellopontine angle. Nearly 80% of CP angle tumors are vestibular schwannomas.

Mean age at the diagnosis of this tumor ranges between 46-58. Occurrence in younger patients are often associated with neurofibromatosis type 2. Bilateral acoustic schwannomas are associated with neurofibromatosis type 2.

**Etiology:**

Majority of patients diagnosed with an acoustic neuroma have no risk factors. Exposure to high dose ionizing radiation is the only definitive environmental risk factor associated with the development of acoustic schwannoma. Multiple studies have determined that cell phone usage is not associated with increased incidence of acoustic schwannoma.

Neurofibromatosis type II occurs in individuals who have defective tumor suppressor gene located on chromosome 22q12.2. The defective protein produced by this gene is known as Merlin or schwannomin. Bilateral acoustic schwannomas are a principal clinical feature of neurofibromatosis type II along with other features that include: peripheral neurofibromata, meningioma, glioma, and juvenile posterior subcapsular lenticular opacities. Peripheral neurofibromata and café au lait spots are less frequent when compared to neurofibromatosis type I. Many patients with neurofibromatosis type II present in late adolescence or early adulthood. Rarely it may be present in the $5^{th}$ – $6^{th}$ decades.

**Pathophysiology:**

Majority of acoustic neuromas develop from the Schwann cell covering of the vestibular portion of the vestibulocochlear nerve. Only less than 5% arise from the cochlear nerve. The superior and inferior vestibular nerves appear to be the nerves of origin and they are involved in equal frequency.

Growth patterns:

Three types of growth patterns have been observed in acoustic tumors. They include:

1. No growth or very slow growth
2. Slow growth (0.2 cm / year) on imaging studies
3. Fast growth (>= 1.0 cm / year) on imaging studies

Majority of acoustic schwannomas grow rather slowly. Some may grow quite quickly and can double in volume within a span of 6 months to 1 year.

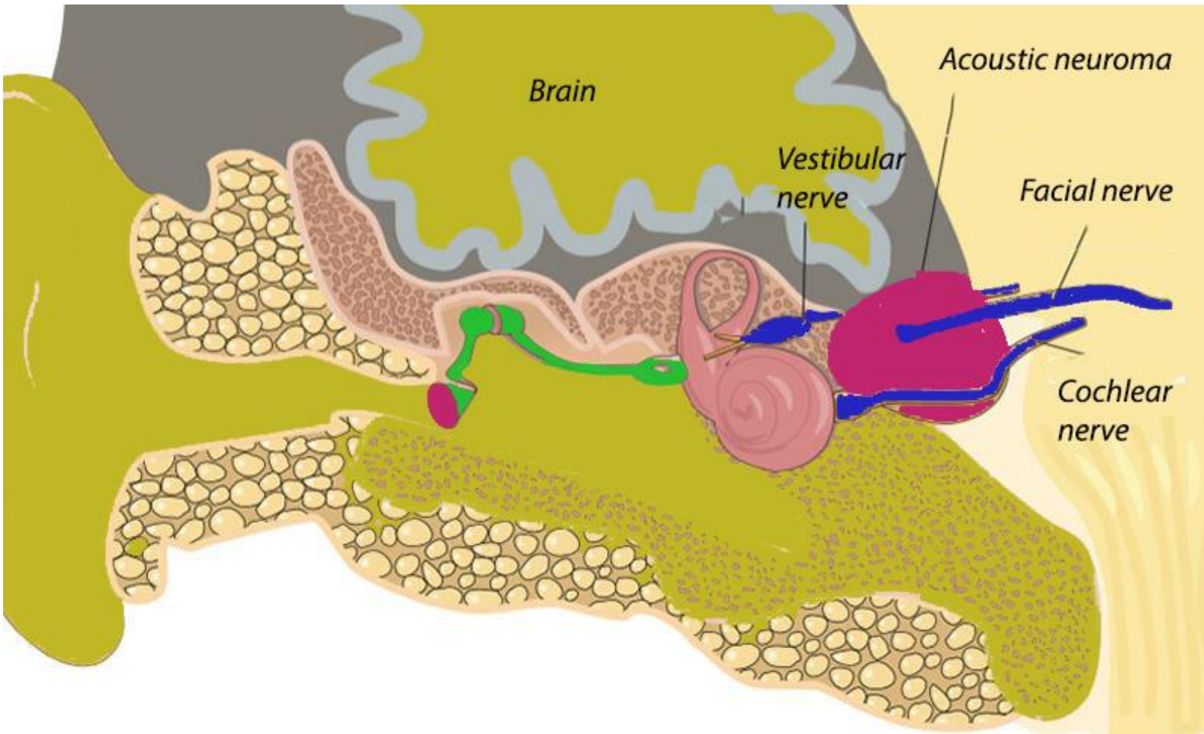
Some tumors adhere to one or the other of the above-mentioned growth patterns, others may alternate between periods of no or slow growth and rapid growth. Tumors that have undergone cystic degeneration (because they have outgrown their blood supply) are sometimes capable of relatively rapid expansion because of selective enlargement of their cystic component.

Acoustic tumors arise from the investing Schwann cell layer. Tumor growth will generally compress the vestibular fibers present on the surface of the nerve. Since vestibular nerve fibers are phylogenetically old, they withstand forces of destruction. So, nerve fiber destruction due to compression by schwannoma is rather slow. This is the reason why many of the patients with vestibular schwannoma experience very little symptoms like giddiness, imbalance etc.

Once the tumor has grown sufficiently large to fill the internal acoustic meatus, its growth continues either by expansion of bone of the internal acoustic meatus or by extending into the cerebello-pontine angle. Growth within the CP angle is generally spherical.

Acoustic schwannoma produces symptoms by the following mechanism:

1. Compression / distortion of the spinal fluid spaces
2. Displacement of the brain stem
3. Compression of blood vessels producing ischemia / infarction
4. Compression / attenuation of nerves




Diagrammatic representation of acoustic neuroma

The cerebello-pontine angle is relatively empty; hence tumors can continue to grow without causing symptoms till they reach a critical size (3-4 cm). Symptoms arise only when the mass gets into contact with critical structures. Tumors arising from the internal acoustic meatus usually produce early symptoms (hearing loss, vestibular imbalance) due to compression of the cochlear nerve, vestibular nerve or labyrinthine artery against the bony walls of the internal auditory canal.

As the tumor approaches 2 cm diameter, it begins to compress the lateral surface of the brain stem. Further growth is possible only by compressing / displacing the brain stem towards the contralateral side. Tumors greater than 4 cms often extend anteriorly to compress the trigeminal nerve and produce facial hypesthesia. As the tumor continues to grow beyond 4 cm then progressive effacement of cerebral aqueduct and $4^{th}$ ventricle occurs leading on to the development of hydrocephalus.

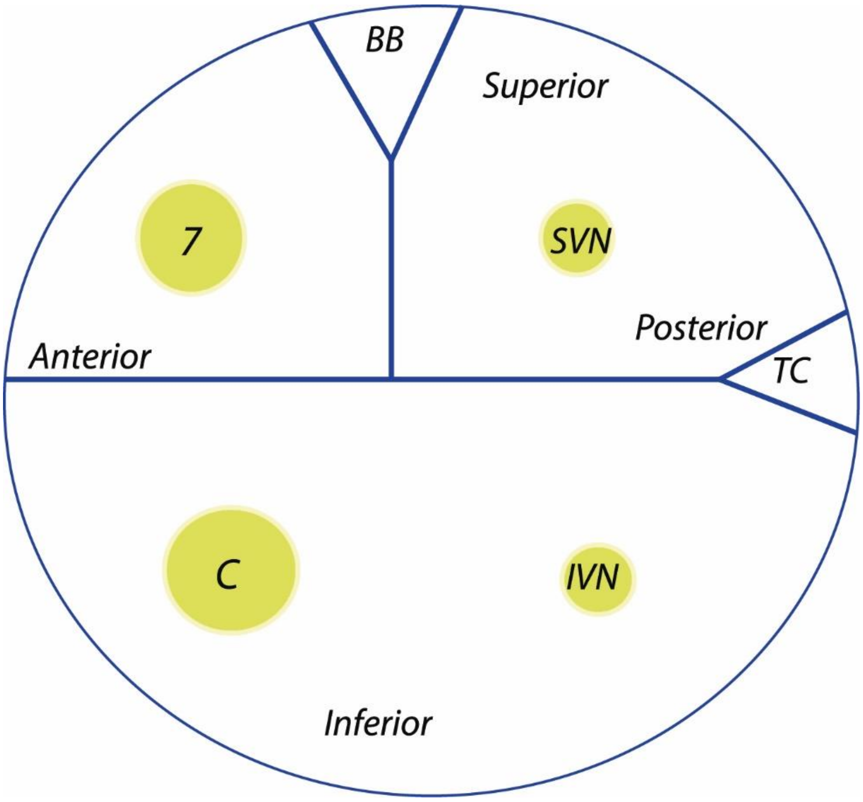



Image showing the cut section of internal acoustic meatus

7 – Facial nerve

BB – Bills's bar

SVN – Superior vestibular nerve

C – Cochlear nerve

IVN – Inferior vestibular nerve

TC – Transverse crest

**Clinical Presentation:**

Unilateral hearing loss is the most common presenting symptom at the time of diagnosis. Any case of unilateral sensorineural hearing loss should be evaluated to rule out acoustic schwannoma. Hearing loss can be caused by two mechanism i.e. direct injury to the cochlear nerve, or disruption of blood supply to the cochlear nerve. Progressive sensorineural hearing loss is the common manifestation of this disorder. If these patients manifest with fluctuating hearing loss then disruption of blood supply to cochlea should be seriously considered.

A significant number of patients with acoustic schwannoma have speech discrimination scores which is reduced out of proportion to that of pure tone average.  This feature is typical of retro-cochlear lesions.  This feature can be identified through audiological testing due to a phenomenon known as “roll-over”.  Roll-over phenomenon indicate that speech discrimination scores decrease as the volume of speech stimulus increases.  A normal speech discrimination score should not be used to rule out acoustic schwannoma.  In fact, patients with acoustic schwannoma may have normal to near normal hearing and speech discrimination scores.

Tinnitus is the next frequent symptom along with sensorineural hearing loss in these patients.  Majority of these patients have asymmetrical hearing and there is an association between the severity of deafness and lower speech discrimination scores.

Hearing loss in patients with acoustic schwannoma could be sudden or fluctuating in nature in 15% of these patients.  Hearing in these patients improve spontaneously or in response to steroid therapy.  Gadolinium enhanced MRI should be performed in patients with sudden or fluctuating hearing loss even if hearing returns to normal.

It has been observed that tumor size has very poor correlation with hearing status as patients with large tumors may have normal hearing and some patients with small tumors may be profoundly deaf.

Unilateral tinnitus should be one of the reasons for further evaluation to rule out acoustic schwannoma.  Nearly 15 – 20% of these patients may present with tinnitus alone at the time of diagnosis.  They may not even complain of hearing loss.

Vertigo and disequilibrium are uncommon presenting symptom in these patients.  Vestibular nerve is highly resistant to compressive forces.  Rotational vertigo may occasionally be seen in these patients even in the presence of small tumors.  Disequilibrium is more common in patients with large tumors.  Since the destruction of vestibular fibers if fairly slow there is adequate time for compensation.

Headaches are present in nearly 50% of these patients.  It is more common during presentation even than that of facial weakness.  Headache is more common in patients with larger lesions.  Larger lesions are known to produce obstructive hydrocephalus which could be one the reason for headache.

Facial numbness occurs in nearly a third of these patients.  It is relatively more common than facial weakness at the time of diagnosis.  Objectively demonstrable hypoesthesia involving teeth, buccal mucosa, or skin over the face is associated with large tumors.  Subjective blunting of facial sensation that cannot be documented on objective examination occurs commonly in medium / small sized tumors.  Majority of the patients with large tumors have facial hypoesthesia which is objectively demonstrable but the patient is often unaware of it.

Blunting of corneal reflex occurs earlier and more commonly than facial hypoesthesia. This occurs earlier and more commonly in majority of patients with acoustic schwannoma.

Motor fibers of facial nerve can withstand substantial amount of stretching as long as it occurs rather slowly when compared to the sensory fibers of trigeminal nerve. Facial weakness is hence uncommon in these patients with an incidence of less than 1%. When facial weakness is associated with small / medium sized tumor then it should raise suspicion that it is not an acoustic schwannoma. Other diagnosis like facial neuroma, hemangioma, meningioma, granuloma or AV malformations should be considered.

Tumors greater than 4 cms can obstruct the flow of CSF through the ventricular system by distorting or obstructing the $4^{th}$ ventricle. In early decades 75% of these patients presented with hydrocephalus.

**Management:**

Treatment depends on the following factors:

1. Age
2. Medical status of the patient
3. Size of the tumor
4. Location of the tumor
5. Status of hearing
6. Preference of the patient

In old patients with small tumors careful examination of the patient which include serial MRI's is the option. In older patient with a tumor that is growing then stereotactic radiosurgery may be the best option. Young patients with large tumors 2.5 – 3 cms and patients with small tumors with intact hearing may choose surgery as an option.

Acoustic schwannomas can be managed by the following ways:

1. Surgical excision of the mass
2. Arresting the growth using stereotactic radio surgery
3. Careful serial observation

**Observation:**

Observation without therapeutic intervention is advised in the following patients:

Elderly

Patients with small tumors with good hearing

Patients who refuse treatment

Patients with a tumor on the side of an only hearing ear or only seeing eye

Studies reveal that 40% of individuals who are being observed ultimately require therapeutic intervention.

During observation period nearly 70% of these patients who were eligible for hearing conservation surgery initially lost their eligibility.

Telian's criteria which should be followed during observation:

1. Preoperative hearing in both ears
2. Risk of immediate hearing loss as a consequence of surgery
3. The risk of facial nerve paralysis
4. Risk of surgical complications and their seriousness
5. Life expectancy of the patient
6. Size of the tumor
7. Tumor growth rate
8. Patients with neurofibromatosis type 2 or bilateral tumors

**Stereotactic radiosurgery:**

This has emerged as an alternative to microsurgery in select cases with acoustic schwannoma. This procedure makes use of radiation source which is administered using a variety of machines (Gamma Knife, Cyber knife, BrainLab etc.) This procedure uses the concept of delivery radiation to a precise point / series of points to maximize the amount of radiation that is delivered to target tissues while at the same time minimizing the exposure to adjacent normal tissues. Radiation can be delivered as a single dose or as multiple fractioned doses.

The precise delivery of radiation reduces the amount of radiation needed for tissue effect. It obliterates tumor growth by causing obliterative endarteritis of the blood supply of the tumor. It can also affect tumor cells undergoing mitosis by causing double stranded DNA breaks.

The classic study of Hensen et al. demonstrated that acoustic neuroma cells are radioresistant at the current low dose radiation used in radiosurgery. Boari et al in his study revealed that acoustic schwannomas with mean tumor volume of 1.94 cm was amenable to radiotherapy administered via gamma knife. He demonstrated a tumor volume reduction of 80% in these patients.

**Advantages of stereotactic radiosurgery:**

1. Decreased length of hospital stays
2. Decreased cost of therapy
3. Rapid return to full employment
4. There is negligible post treatment morbidity and mortality

**Disadvantages of stereotactic radiosurgery:**

1. There is a need for long term monitoring and follow up of these patients with periodical MRI imaging. The cost of follow up would be much more than the actual cost of stereotactic radiosurgery
2. Does not eliminate the tumor completely. It may fail to control tumor growth requiring subsequent salvage surgery
3. There is a higher incidence of trigeminal nerve injury
4. Possibility of secondary malignancies developing
5. Does not address disequilibrium and hence could lead to long term balance dysfunction
6. It carries the risk of patient developing hydrocephalus, hence these patients should be closely monitored following the procedure

**Dose of radiation used:**

Hearing preservation depends on the irradiation dosage. Ideal dose being 12-13 Gy is ideal for hearing preservation. Hearing preservation is dependent on the radiation dose to the cochlea, cochlear nerve, and cochlear nucleus. Following irradiation there is a transient tumor volume expansion which can lead to hearing loss.

**Surgical therapy:**

This is the treatment of choice for tumor eradication. Various approaches can be used to remove acoustic tumors. Person under the age of 65 and with medium to moderately large tumors prefer surgery.

Other groups of patients in whom surgery is preferred are those with rapidly growing tumors, large grade tumors, significant hearing loss following acoustic schwannoma, and severe headache.

Surgical approaches:

Three different surgical approaches are used in the management of acoustic schwannomas. They include:

Retro sigmoid approach

Translabyrinthine approach

Middle cranial fossa approach

Advantages of retrosigmoid approach:

1. This approach can be used for all acoustic tumors. It can be used for operations that sacrifice hearing and operations that attempt to conserve hearing. Its only limitation is its inapplicability for small tumors that occupy far lateral positions in the internal auditory canal.
2. This approach provides the best wide field visualization of the posterior cranial fossa. The inferior portion of the CP angle and the posterior surface of the temporal bone anterior to the porus acusticus are clearly observed via the translabyrinthine approach. Panoramic view is helpful when displacement of nerves is not predictable. This commonly occurs in meningiomas.
3. Hearing conservation surgery can be attempted even for relatively large tumors via the retrosigmoid approach. In this approach destruction of labyrinth is not needed.

Disadvantages of retrosigmoid approach:

1. This approach may require cerebellar retraction / resection. Manipulation of the cerebellum provides opportunities for post op oedema, hematoma, infarction and bleeding.
2. There is always increased risk of CSF leak
3. There is a greater likelihood of severe protracted post op headache
4.

Advantages of Translab approach:

1. This provides the best view of the lateral brainstem facing the acoustic tumor
2. Cerebellar retraction is not needed
3. 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. This reduces the risk of facial palsy post operatively.
4. If the facial nerve needs to be sectioned then this approach allows restoration of the facial nerve continuity by rerouting the nerve and performing a primary anastomosis.

Disadvantages of Translab approach:

1. Hearing is totally sacrificed
2. The inferior portions of the CP angle and cranial nerves are not clearly visualized as seen in retrosigmoid approach. The temporal bone anterior to the porus acusticus is also less well visualized.
3. A fat graft is needed. Harvesting abdominal fat can cause donor site complications like hematoma, bleeding and infection.
4. The sigmoid sinus is vulnerable to injury. Bleeding from the sigmoid sinus can be difficult to control adding to significant blood loss. If a dominant sigmoid sinus is occluded during the surgery, then post-operative intracranial pressure elevation can occur.
5. A high jugular bulb or anteriorly placed sigmoid sinus can substantially compromise the space available for tumor removal.

Advantages of middle cranial fossa approach:

1. This is the only procedure that fully exposes the lateral third of the internal acoustic meatus without sacrificing hearing
2. It is totally extradural

Disadvantages of middle cranial fossa approach:

1. The facial nerve usually courses across the antero superior portion of the tumor. It comes in the way of tumor removal and is vulnerable to injury during the process of tumor removal. Temporary facial paresis is more common than full blown facial palsy.

2. In elderly there is increased risk of dural laceration and avulsion. It should be noted that the dura in elderly is very friable. This complication is more common during the $6^{th}$ and $7^{th}$ decades of life.
3. This approach provides very little exposure of posterior cranial fossa
4. This approach is technically difficult and more demanding
5. Some of the patients may develop trismus due to manipulation / injury to temporalis muscle
6. Temporal lobe must be retracted, risking injury. Hematoma formation in this area is more common. Seizure disorder has been reported in some patients due to temporal lobe injury.

Selection of ideal surgical approach:

The following factors should be considered while deciding on the optimal approach to remove these tumors.

Pre-operative hearing level:

If the patient has no useful hearing then Translab approach or retrosigmoid approach is preferred. The exact choice depends on the experience and training of the surgeon. In majority of centers performing surgery for acoustic schwannoma a trans lab approach is preferred.

What level of hearing constitutes useful salvageable hearing?

The rule of thumb being a Puretone average greater than 50dB and speech discrimination of less than 50% is not considered to be salvageable hearing level.

A normal Preoperative ABR favor hearing conservation. Marked abnormalities of ABR wave morphology / increased wave I-III and I-V latencies make hearing conservation less feasible.

An abnormal caloric test on ENG increases the likelihood of successful conservation surgery. The ENG is considered to test the lateral semicircular canal which is innervated by the superior vestibular nerve and this nerve should be considered to be normal in the presence of positive caloric test. In this scenario it can be safely assumed that the tumor could have originated from the inferior vestibular nerve which is adjacent to the cochlear nerve. Surgical removal in this scenario is likely to injure the cochlear nerve or could interfere with blood supply to the cochlear nerve. VEMP is abnormal when the inferior vestibular nerve is affected. An abnormal VEMP coupled with normal caloric testing on ENG strongly suggest an inferior vestibular nerve tumor and hearing conservation is difficult in these patients.

Tumor size:

Hearing conservation opportunities diminish as the tumors become large. Hearing is difficult to conserve when tumors are 1.5-2.0 cm in diameter. Hearing can be conserved when tumors are small and intracanalicular. A school of surgeons believe that hearing conservation surgeries are indicated for removal of small tumors and trans lab approach is preferred to remove larger ones. Trans lab approach maximizes the chance of facial nerve conservation when large tumors are to be removed.

Position of the tumor:

If hearing conservation is to be attempted that the tumor lies within the lateral portion of the internal auditory canal, most surgeons prefer a middle cranial fossa approach. The middle cranial fossa approach permits direct exposure of the lateral end of the internal auditory meatus without sacrificing hearing. This approach is the frequently used one to remove tumors lying completely within the internal acoustic meatus. Tumors limited to the medial portion of internal acoustic meatus can be managed via retrosigmoid approach. An extended middle cranial fossa approach can be resorted to remove tumors 0.5-1 cm within the CP angle. Division of superior petrosal sinus is required to gain sufficient access to the posterior cranial fossa with larger tumors.

Tumors that have a large volume medial to the plane of porus acousticus can be extirpated using retrosigmoid approach if hearing is to be conserved. If hearing conservation is not an issue again retrosigmoid approach is preferred for tumors with significant inferior extension since lower cranial nerves are better visualized via this approach. Rarely retrosigmoid approach can be combined with trans lab approach to remove such tumors.

Anatomical compulsions:

The following anatomic variations can make trans lab approach more difficult and at times nearly impossible to perform.

1. High jugular bulb. In some individuals this could rise up to the level of internal acoustic meatus
2. Anteriorly placed sigmoid sinus. In this scenario the distance between the sigmoid sinus and external canal could be just a few millimeters. This causes a huge limitation as far as space is concerned. This scenario places a huge risk on injury to itself as well as injury to the facial nerve.
3. Contracted sclerotic mastoid. These small cavities provide very little room for tumor removal. They are also commonly associated with suppurative otitis media which itself is a contraindication for a Translab approach.
4. Reduced / absent flow in the contralateral venous sinus. This can very well happen in patients who had undergone previous surgery in the area, trauma, congenital anomalous development, previous disease can cause a markedly reduced / absent venous flow through contralateral sinus. Retrosigmoid approach will injure the only draining sinus causing venous infarction
5. Surgeon’s preference
6. Patient’s choice

Follow up:

Follow up MRI should be obtained within 6 months to 1 year after surgery in order to document the completeness of tumor removal. Even after complete removal follow up MRI need to be performed at 5th year and at 10th year following surgery. If post op MRI is indicated then fat suppression technique should be used because fat would have been used to close CSF leak.

**Complications:**

Injury to AICA (anterior inferior cerebellar artery) and rarely to PICA (posterior inferior cerebellar artery). AICA could be loosely attached to the tumor capsule, hence separating it is rather easy. Sacrificing AICA can have varying effects on the patient depending on the anatomy. It can also lead to devastating neurologic injury or rarely even death.

Branches of AICA that are most vulnerable to injury are the labyrinthine artery and branches supplying the facial nerve. The perplexing post-operative facial palsy could be accounted for due to interruption of facial nerve vascular supply due to coagulation of small branches of AICA.

Failure to conserve hearing could be the result of injury to cochlear blood supply. Conservation of labyrinthine artery becomes rather difficult as the size of the tumor increases, this accounts for the relatively less success in hearing preservation while operating on large sized tumors.

Neurologic injury / cerebral oedema secondary to venous injury usually occurs as a result of injury to sigmoid sinus itself, the petrosal vein of Dandy, or to the vein of Labbe.

Occlusion of sigmoid sinus will have varying effects depending largely on patient's unique vascular anatomy. If contralateral venous outflow tract is patent then communication is possible through the torcula herophili which is rather adequate. Usually the size of the sigmoid sinus on both sides are not symmetrical, with greater volume of blood flowing through the right sided sinus. Depending on the volume of drainage by the dominant sinus occlusion of it can cause complications like raised intracranial tension, venous infarction and sometimes even death. Since a number of potential collaterals are present between the torcula herophili and the jugular bulb, occlusion of sigmoid sinus close to the torcula herophili is more likely to have significant adverse effect than occlusion close to the jugular bulb.

Petrosal vein of Dandy is a single large outflow tract in some patients, in some it would comprise of a series of large veins. Occlusion can result in oedema and infarction of either the temporal lobe or the brain stem. Attempt should be made to preserve this vein.

Occlusion of veins of Labbe result in severe oedema of temporal lobe and temporal infarct. The oedema can be so severe as to cause brain herniation and death in some cases. This vein generally enters the superior petrosal sinus or transverse sinus between the torcula herophili and the point at which the superior petrosal sinus joins the transverse sinus. It thus does not come directly into the surgical field. Rarely injury to superior petrosal sinus may result in its obliteration. The presence of this vein should always be kept in mind during surgery.

Bleeding into the posterior cranial fossa in the immediate PO period can produce brain stem compression and death rather rapidly. Sudden rapid deterioration in the immediate post op period should raise suspicion of posterior fossa hemorrhage and mandates immediate decisive action.

Cerebellar injuries:

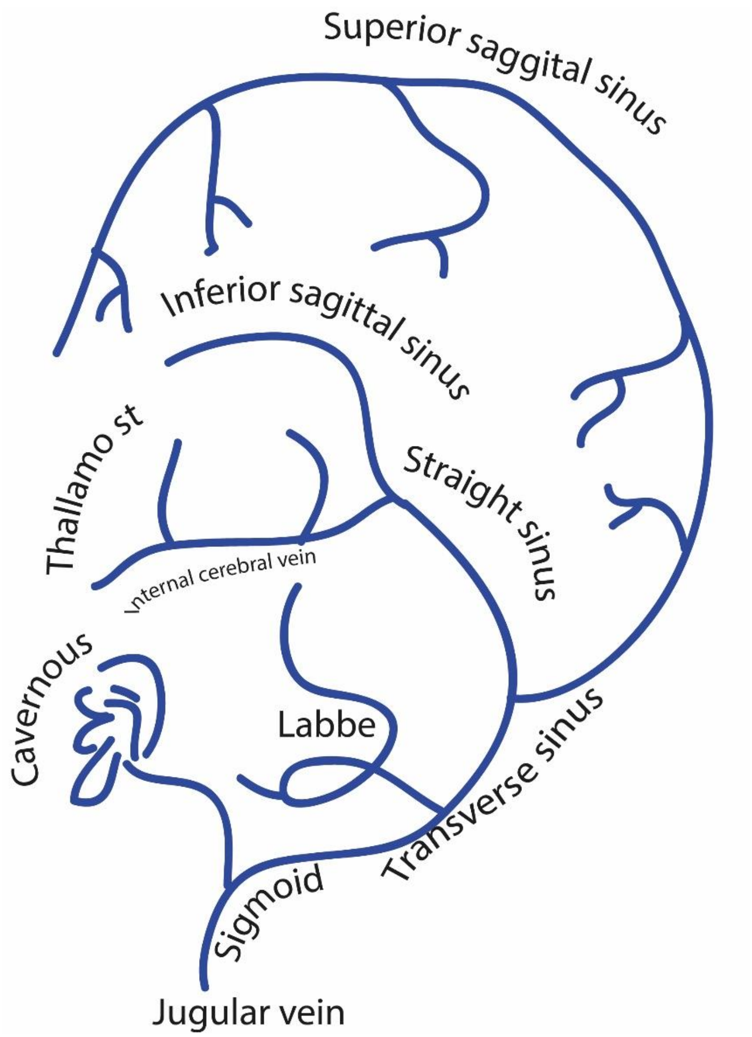



Figure showing the anatomy of dural venous sinuses

Cerebellar injuries:

This was rather common in early days of this century. With the advancement of surgical techniques and tools its incidence has dramatically fallen in recent times. Even now cerebellar injuries do occur but are generally not so troublesome. Common culprit happens to be injury due to rotating burrs. Shaft of the burr happens to be the culprit as it causes damage to the cerebellum while the surgeon is focusing on the rotating burr head during the phase of bone removal. Small areas of injury have very minimal sequelae.

Bleeding from injury to cerebellum can be controlled using oxidized cellulose, cautery or gelatin sponges. Direct injury to cerebellum due to retraction, intracerebral bleed, infarction due to altered blood supply due to surgery can produce severe edema of cerebellum. This can lead to brain stem compression / intracranial herniation and eventually death. Obstruction to the fourth ventricle and cerebral aqueduct can produce significant hydrocephalus.

This crisis can be managed by aggressive use of osmotic diuretics, hyperventilation and steroids. If medical management fails then the part of involved cerebellar hemisphere need to be resected.

Facial paralysis:

Post op facial paralysis is unavoidable in some cases. The tumor tissue may be attached to the facial nerve and attempts to free the mass away from the nerve can cause facial paralysis. Tumor can sometimes totally envelop the facial nerve. Removal of the tumor is possible only if that portion of facial nerve is resected along with the tumor. In this situation facial nerve repair using sural nerve graft should be attempted.

In the event of facial nerve paralysis then eye care should be the most important measure that must be considered till such time the facial nerve function resumes. Inability of the patient to close the eye will cause corneal ulcers and exposure keratitis. Patient should be advised to wear glasses with side protector always to prevent dust from getting into the eye. Dry eye can be managed by liberal use of artificial tears. During night time ocular lubricants should be applied. Even after aggressive use of artificial tears during day time and lubricants during nights if exposure keratitis occurs then use of eye patch, or placement of gold weight to keep the eyelid closed at least partially can be resorted to. Coexistent injury to $5^{th}$ cranial nerve could cause corneal anesthesia which obviously compounds the problem. Tarsorraphy will have to be resorted to if $5^{th}$ nerve is damaged along with facial nerve.

CSF complications:

Transient abnormalities involving CSF resorption may lead to temporary and mild post-operative hydrocephalus. Shunting is almost very rarely or never required. Even if hydrocephalus is present post operatively it generally resolves without difficulty in the first few weeks following surgery.

Post-operative meningitis:

This complication usually occurs in two forms. Bacterial meningitis is potentially life threatening and occurs very rarely i.e. in less than 1% of patients. It can occur within first couple of days post operatively. Sometimes it could be delayed by a couple of weeks. Once started it has a relentless progression and individuals can progress to unconscious state rather rapidly. In the even of suspicion of meningitis, lumbar puncture should be resorted to for diagnosis and for culture. It should be performed only after CT scan reveals that there is no threat of brain herniation.

CSF should be subjected to the following investigations:

1. Grams stain
2. CSF glucose levels
3. CSF protein levels

If gram stain is positive, CSF glucose level is less than 40 m/dL and WBC count in CSF is higher than 2500 cells/$mm^3$ antibiotics should be started immediately without waiting for culture results. If CSF values do not reflect any of the above said criteria then the patient should be closely monitored and repeat Lumber puncture can be performed in the event of deterioration of patient's condition. Aseptic meningitis has been reported in nearly 1/3 of these patients. It more or less symptomatically resembles bacterial meningitis i.e. increasing headache, fever, neck stiffness and elevated CSF pressure. Spinal fluid profile in patients with aseptic meningitis would show marked elevation of White cell count, and increased CSF protein levels. But CSF glucose levels remains within the normal reference range. Culture results are usually negative. Steroids are very useful in managing aseptic meningitis and prompt administration results in decrease in head ache, neck stiffness within few hours.

CSF leak either through the wound or Eustachian tube and middle ear occurs in 2% of these patients. It is common after Translab or retrosigmoid approaches. It is less common after middle cranial fossa approach. When CSF leak occurs following retrosigmoid approach then it traverses the path through pneumatized air cell tracts.

Pathophysiology of CSF leak following surgery for acoustic neuroma removal:

CSF is produced within the ventricular system at a rate of 0.3 ml / min and it makes up to 500 ml /day. It enters the subarachnoid space in the posterior fossa via the midline and lateral foramen of the 4th ventricle. Contamination of CSF by blood, bone dust and necrotic debris during surgery impairs CSF absorption directly by mechanical interference in the arachnoid villi or indirectly by inciting inflammatory response within the subarachnoid space. The symptom complex could vary from brief asymptomatic elevation of CSF pressure to clinical evident aseptic meningitis. CSF escaping through the wound could be initially managed by wound resuturing. If CSF leak persists for more than 24 hours after initiation of conservative management which include pressure dressing, consistent elevation of head then CSF pressure reduction steps should be considered. CSF pressure reduction techniques include:

1. Multiple lumbar punctures
2. Continuous / intermittent drainage of CSF via lumbar catheter
3. Permanent diversion of CSF by means of indwelling shunt

If the process of diversion is selected then the most common method is to place an indwelling subarachnoid catheter placed into the lumbar subarachnoid space. The drain is opened episodically so as to remove 200-400 ml of spinal fluid in any given 24-hour period. This drain should be performed for 2-5 days. If the drain has been in place for more than 5 days then it should be replaced to avoid infection.

Post op headache:

This is common with retrosigmoid procedures. This complication has reduced a lot after introduction of these steps:

1. Care should be taken to avoid contamination of CSF with blood and bone dust.
2. Bone flap is replaced and bony defect if any is eliminated by using methyl methacrylate of hydroxyapatite. This site prevents direct attachment of posterior cervical musculature to the dura.

When post op headaches occur, then it should be managed with relatively high dose non-steroid anti-inflammatory agents and aggressive regimen of manipulative physiotherapy.

**Treatment outcome and prognosis:**

Tinnitus worsens in about 20% of patients after tumor removal. In a great majority of patients' tinnitus either improves or remain unchanged. Roughly 30% of patients who did not have tinnitus on presentation may develop it post surgically. It is often not troublesome.

Studies conducted by Bell etal showed that patients who undergo acoustic schwannoma resection had poor resolution of tinnitus if they are young and whose per operative hearing is considered to be serviceable. Tinnitus is also severe in patients who have residual tumor left behind post operatively.

Recurrence / residual tumor:

Recurrence is uncommon if the tumor has been removed completely. Overall recurrence rate has been estimated to be about 5%. Majority of recurrences are seen in patients who have undergone retro sigmoidal removal of the mass. This could possibly due to a small amount of tumor tissue left behind in the lateral end of the internal auditory meatus where intraoperative visualization is rather difficult when this approach is used. Tumor recurrence should be suspected if there is recurrent headache, altered sensation over face, dysarthria and dysphasia if lower cranial nerves get involved.

Tumor bed inflammation could be seen persisting for months or even years after tumor removal. This could consequently cause areas of contrast enhancement when post-operative gadolinium MRI scan is performed. This makes it difficult to differentiate recurrence from contrast enhancement caused by inflammation of tumor bed. Tumor recurrences appear globular in shape in contrast MRI while inflammation appears to be linear in contrast MRI. Fat suppression techniques should be used during post op surveillance to distinguish from recurrent from fat packing. Surveillance for post op tumor recurrence should be performed for at least 10 years following surgery.

Facial function:

Preservation of facial function is really improving with widespread use of facial nerve monitoring during surgery. Facial nerve outcomes vary and it depends on tumor size. If tumors are small less than 1.5 cm then good facial function following surgery is to be expected. Pre-op electrophysiologic testing can help in predicting post op outcome of facial nerve function. It has been proved that in the presence of significant electrophysiologic abnormalities on nerve conduction studies would increase the incidence of loss of facial nerve function following surgery. Blink reflex testing helps in this prediction.

Facial nerve paralysis could be delayed. It can develop within a few hours to a week or more following surgical removal of acoustic schwannoma. Incidence of delayed facial palsy ranges between 10-30%. The mechanism of action is rather unclear. Ischemia secondary to vasospasm, vascular injury, traction, nerve oedema due to stretching have been proposed. Viral reactivation following trauma to nerve is also possible. Nearly 90% of these patients with delayed facial nerve paralysis make complete recovery of the nerve function.

Hearing outcome:

Hearing preservation has increased substantially during the last decade. Hearing can be preserved in nearly 80% of these patients. It has also been proved that gamma knife surgery does not appear to have significantly higher rate of hearing preservation when compared to that of conventional surgery.