

*Cerebello-Pontine angle tumors
Otolaryngologist's perspective*



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Introduction:

Cerebellopontine angle tumors are encountered rather more commonly than anticipated by otolaryngologist. This group of tumors account for nearly 10% of all intracranial tumors. These tumors are rather fatal when not treated. Studies reveal that acoustic neuromas (vestibular schwannomas) constitute nearly 80% of all cerebellopontine angle tumors. Other tumors that could involve this area include:

1. Meningiomas
2. Dermoid tumors
3. Arachnoid cysts
4. Lipoma
5. Metastatic tumors
6. Vascular tumors

The entire science of neuro-otology developed because of tumors involving this area.

History:

Sir Charles Balance first documented successful removal of C-P angle tumor in London (1894). He also documented the presence of facial paralysis and facial anesthesia following surgery in the same patient.

Cushing coined the term “C-P angle syndrome” to describe the features of tumors involving this critical area. According to Cushing the components of this syndrome include:

1. Ipsilateral hearing loss
2. Ipsilateral facial hypesthesia
3. Hydrocephalus
4. Ultimately respiratory arrest due to brain stem compression (fatal)

Cushing suggested decompression of these tumors by partial resection via bilateral suboccipital craniectomy procedure.

Walter Dandy espoused more complete resection of tumor citing the risk of recurrence in partial decompression procedures. He advocated debulking followed by careful removal of capsule from the brainstem via suboccipital approach.

The modern era of C-P angle surgery was kickstarted by William House. He popularized the widely used translabyrinthine approach to the cerebellopontine angle. With the use of microdrills and microscope he was able to identify and preserve the facial nerve. Five years later he began to popularize the middle cranial fossa approach.

Anatomy of cerebello-pontine angle:

Location - This is an irregularly shaped potential space located at the posterior cranial fossa.

Boundaries -

Anterior – Posterior surface of temporal bone

Posterior – Anterior surface of cerebellum

Medial – Inferior olive

Superior – inferior border of pons and cerebellar peduncle

The cerebellar peduncle forms an important relationship inferiorly.

The 7th and 8th cranial nerves course superiorly and laterally towards the internal auditory canal in this space. These nerves carry along with them a thin sheet of arachnoid tissue. The 5th cranial nerve can be visualised superior to this space.

9th, 10th and 11th cranial nerves are located inferiorly.

Important contents of this space include:

Flocculus of cerebellum

Foramen Lushka of 4th ventricle

Anterior inferior cerebellar artery

The 7th and 8th cranial nerves are covered with glial tissue throughout their intracranial course. At the level of internal acoustic meatus the glial tissue is replaced by schwann cells. The junction between the glial tissue and schwann cells is known as Obersteiner Redlich zone. This zone is somewhat histologically unstable. Tumors are more prone to arise from this area.

Vestibular schwannomas:

These are the most common of CP angle tumors. It constitutes about 75-80% of all CP angle tumors. The overall incidence rate for vestibular schwannomas is rather difficult to ascertain due to the fact that majority of these tumors remain silent as they have a very slow growth rate. Routine autopsy studies have put the estimates to be about 2.5%. With the advent of sensitive imaging modalities more and more of these cases are being picked up at early stages.

These tumors arise from the vestibular segment of the 8th cranial nerve. Studies reveal that its incidence is approximately equal between superior and inferior vestibular nerves. These tumors have been thought to develop from the schwann cell, close to the glial – schwann junction. Current knowledge of course refutes this idea. These tumors most commonly arise lateral to this junction close to the internal acoustic meatus. It arises close to the sharp's ganglion. This ganglion contains the largest number of schwann cells. Hence the more appropriate term to describe this disorder is "Vestibular schwannoma". Rarely these tumors may also develop from the cochlear division of the 8th nerve.

Types of schwannomas:

Sporadic variety – This constitutes 95% of all vestibular schwannomas.

Type II neurofibromatosis – This may occur bilaterally. These lesions are known to occur in

patients of younger age group. Type II neurofibromatosis is associated with intracranial meningiomas and spinal cord tumors. There are two subtypes included under type II neurofibromatosis.

Wishart type is more severe whereas the Gartner subtype is less severe. Molecular genetic studies have revealed that defects involving the long arm of chromosome 22 to be the cause for Type II neurofibromatosis. More precisely this condition is inherited as an autosomal dominant condition. Mutation of tumor suppressor gene is known to be the cause. Tumor suppressor genes are important in reducing the incidence of tumors. Absence of these genes have been attributed to be the cause of this problem.

Molecular studies reveal that all cases of sporadic variety of schwannomas are caused due to inactivation of NF2 gene. This gene is actually one form of tumor suppressor gene. NF2 genes are known to control secretion of Merlin / Schwannomin. These are protein molecules which controls the contact based cell growth. They prevent cell proliferation when the cell gets into contact with the other. Lack of these protein molecules leads to loss of contact based cell proliferation inhibition. This causes increased proliferation of schwann cells leading on to schwannoma formation.

Biochemical factors contributing to the growth of schwannomas include:

1. Neuregulin – This protein is secreted by schwannomal cells. It is known to control the growth, proliferation and survival of schwann cells. This protein gets attached to erbB2 and erbB3 surface receptors present in the proliferating schwann cells.
2. Fibroblast growth factor – This has a vital role in the survival and proliferation of schwannomal cells
3. Platelet derived growth factor
4. Vascular endothelial growth factor

Some investigators conclude that growth of schwannomas accelerated during pregnancy. This according to them is due to the presence of hormonal receptors over their surface.

Clinical features of vestibular schwannomas:

Diagnosis of vestibular schwannomas needs high degree of astuteness and alertness on the part of the physician. Since this is a very slow growing tumor, there is ample time for brain accommodation to kick in.

Symptoms are most commonly vestibular / auditory and is unilateral. The rate of progression of symptoms pertaining to the vestibular schwannoma depends on the rate of growth of the tumor.

Intracanalicular tumors: These tumors are characterized by hearing loss (sensori neural), tinnitus and vestibular dysfunction (giddiness).

Tumor growth extending into the cerebellopontine angle: In this stage deafness worsens, dysequilibrium becomes more evident. As the tumor grows into the cerebellopontine angle it causes compression in this area causing involvement of trigeminal nerve. These patients present with midfacial hypesthesia. When the tumor expands further it could cause hydrocephalus and vision loss.

According to Cushing the diagnosis of acoustic schwannoma could be made with reasonable degree

of certainty only when auditory manifestations definitely precede other symptoms of CP angle tumor. These patients classically manifest with hearing loss which could be described by the patient as distortion in hearing. This could be evident while the patient attempts to converse through the phone. This distortion could be due to reduced speech discrimination. Some patients may have sudden hearing loss. Eventhough this is rare the presence of sudden hearing loss does not completely exclude the diagnosis of acoustic schwannoma. Studies reveal that sudden hearing loss in these patients could be caused by acute compression of the auditory nerve.

Ocular symptoms in these patients include:

1. Absent corneal reflex. This usually precedes hypesthesia of one side of the face due to involvement of the trigeminal nerve
2. Nystagmus
3. Diplopia – due to the rare involvement of 6th cranial nerve
4. Visual blurring – is very rare. When present papilloedema should be suspected. If untreated it could lead to optic atrophy resulting in tunnel vision. This papilloedema is usually due to increased intracranial tension.

Facial weakness is very rare in patients with vestibular schwannomas as the facial nerve tolerates even extensive stretching. Presence of facial weakness should prompt the suspicion of some other tumor in the CP angle rather than schwannoma.

Symptoms caused due to involvement of lower cranial nerves are rather rare and could cause symptoms like dysphagia, hoarseness and aspiration.

Symptoms due to cerebellar involvement occur rather late in this disorder. These symptoms include:

1. In co-ordination / wide based gait.
2. Tendency to fall forward

Presence of intense head ache and vomiting should arouse suspicion of hydrocephalus.

Any patient presenting with unilateral auditory / vestibular symptoms should be thoroughly investigated to rule out acoustic schwannoma.

Clinical examination of a patient with acoustic schwannoma:

Patients who are suspected to be suffering from unilateral auditory / vestibular symptoms should be thoroughly examined. During routine otoscopy sensation over the posterior bony meatal wall should be looked for. Decreased sensation in this area is known as “Hitselberger sign” a clear pointer towards the diagnosis of schwannoma. This is due to the involvement of sensory branches of 7th cranial nerve which is more sensitive to compression by shwannomas than its motor counterpart. This sign could even be present in patients with small tumors in the CP angle area.

Eyes should be examined for the presence of nystagmus. Extraocular movements are also accessed.

Tests for facial sensation:

Facial sensation could be blunted in these patients due to the involvement of 5th cranial nerve. Sensation should be tested for both pain and touch. This is usually performed using pin prick / wisp of cotton. It is imperative to test for both these sensations.

Evaluation of corneal sensation:

Corneal sensation can be evaluated using a wisp of cotton and looking for blink reflex. The cornea should be stimulated with a wisp of cotton while the patient is looking up, this is to ensure that the patient is not aware of the wisp of cotton coming.

Checking the integrity of masseter and temporalis muscle:

This is done to rule out the involvement of the motor component of the 5th cranial nerve. The patient is asked to clench the teeth while the masseter and temporalis muscle are palpated. Rigidity of these muscles indicate that the motor component of the 5th cranial nerve is intact.

Ophthalmoscopic examination is indicated in these patients when there are any visual disturbances.

Muscles of facial expression should be examined to rule out 7th nerve involvement. These patients with facial paralysis may not be able to close their eyes, wrinkle their forehead due to paralysis of facial muscles.

During examination of the oral cavity and oropharynx gag reflex should be looked for. This reflex is lost when the lower cranial nerves are involved.

Tongue should be examined for atrophic changes / deviation to one side during protrusion.

Sternomastoid / Trapezius muscles should be examined while the patient is shrugging the shoulders to rule out involvement of the accessory nerve.

Tests for cerebellar functions like finger to nose test, dysdiadokokinesis test should be performed to ensure that cerebellum is not involved.

Audiometric tests:

These tests should help to identify retrocochlear deafness which is commonly seen in these patients. Previously the following tests were commonly performed to rule out retrocochlear deafness.

1. SISI – Short increment sensitivity index
2. ABLB – Alternate loudness balance test
3. Tone decay test

These tests have been replaced due to poor sensitivity and specificity issues.

Currently used battery of audiological tests include:

1. Pure tone audiometry – These patients manifest with high frequency sensori neural hearing loss
2. Speech discrimination scores – These scores are classically out of proportion to that of pure tone audiogram scores. A patient with mild sensori neural hearing loss will be able to understand only 50% of the monosyllable words presented. This is very low when compared to that of a patient with cochlear hearing loss which could be as high as 80% for

the similar degree of PTA reading. Classically these scores become still worse in patients with acoustic schwannoma when the intensity of the stimulus sound is increased. The scores could get worse even by a factor between 20 – 30%. This phenomenon is called “Roll over” and is commonly seen in retrocochlear type of deafness. This phenomenon could also be seen rarely in patients with cochlear deafness.

3. Acoustic reflex thresholds – are evaluated by impedance testing. Reflex thresholds are obtained for 500, 1000, 2000 and 4000 Hz. The reflex threshold values may be elevated or totally absent in patients with vestibular schwannoma. If reflex is present then reflex decay should be measured by presenting the signal 10dB above the threshold levels for 10 seconds. The reflex decay test is considered to be positive if the power of stapedial contraction cannot be maintained by at least half its strength for 10 seconds.
4. Acoustic reflex decay – This test is also performed using an impedance audiometer. Reflex decay is tested at 500 and 1000 Hz.
5. Auditory brain stem evoked response – This is an electrophysiological test. It is the most sensitive and specific of all the tests for detection of vestibular schwannoma. Surface electrodes are placed in various locations of head and ear and these electrodes measure the electrical potentials generated in response to broad band clicks of short duration. The duration of these click stimuli could be 100 msec. Typically 1000 – 2000 clicks are presented at rates of 7-20/sec. The measured electrical potentials are averaged using a computer which displays the results in wave form. Five different types of wave forms have been identified representing the neural activity that takes place in progressively higher centres. These centers start with auditory nerve and ends at the inferior colliculus which lies at the level of brain stem. Abnormalities seen in auditory brain stem evoked response in patients with acoustic schwannoma include:
 - a. Presence of all 5 waves with an inter aural difference in the latency of wave V with a delay of more than 0.2 msec in the involved ear
 - b. No identifiable wave form is generated. This is a significant finding if the patient does not have hearing loss involving higher frequencies (more than 60 dB).
 - c. Presence of only wave I with absence of other waves involving higher centers
 - d. ABR could be normal in 10% of these patients

It has been suggested that ABR can be used to predict grossly the size of the tumor.

Vestibular schwannomas which are large enough to cause compression of brain stem would also cause lengthening of the interval between waves III and V on the contralateral side.

Vestibular testing in patients with acoustic schwannoma:

These tests include:

Electronystagmography

Computerized dynamic posturography – Response is somewhat below normal

Rotatory chair testing – Not so reliable

Studies reveal that close to 90% of vestibular schwannomas arise from the superior vestibular nerve. These lesions classically show reduced caloric response. Caloric tests usually assesses the function of the lateral semicircular canal and hence only lesions involving superior vestibular nerve can be picked up. Lesions involving the inferior vestibular nerve are thus missed in caloric tests. Hence caloric tests are not reliable investigation in the diagnosis of vestibular schwannomas.

Spontaneous nystagmus:

These are commonly observed only in large tumors. Smaller tumors are rarely symptomatic. If nystagmus is present in small tumors it is mostly of peripheral type and fatiguing. Large tumors cause central type of nystagmus which is non-fatiguing and lateral in nature. As a rule nystagmus beat away from the ear involved with acoustic schwannoma.

Role of imaging in the diagnosis of acoustic neuroma:

Most commonly vestibular schwannoma is identified by routine imaging of temporal bone, particularly its petrous part. During yesteryears plain radiographs and polytomographs were the mainstay in the diagnosis of acoustic neuromas. It was stressed that the presence of bony erosion of the internal auditory canal in these pictures is a must for a diagnosis of acoustic neuroma. Posterior cranial fossa myelography improved the accuracy to some extent. The diagnostic accuracy of conventional radiographs was caused by the presence of arachnoid cyst / loop of anterior inferior cerebellar artery creating an illusion of acoustic neuroma. That is the reason why the presence of erosion of the internal acoustic meatus was considered to be a must before a diagnosis of acoustic schwannoma could be safely made.

CT Scan:

Introduction of computerized tomography during 1980's increased the accuracy of diagnosis rather dramatically. Since CT imaging of temporal bone can be performed in an axial plane these lesions can be clearly made out due to excellent bone / soft tissue differentiation. If iodinated contrast agent is injected before performing the CT scan 90% of vestibular schwannomas became enhanced. Thus contrast CT increased the diagnostic yield and could really pick up small tumors above 5mm in size. Intracanalicular tumors less than 5 mm could be missed in the CT / Contrast CT imaging. Air contrast cisternography has increased the accuracy of CT imaging. Air contrast cisternography is performed by injecting 4ml of oxygen into the subarachnoid space. This clearly reveals the convex bulge of the mass at the level of porus of the internal auditory canal. The only problem with this investigation is the common presence of head ache for atleast a couple of days after investigation. When compared to MRI the soft tissue resolution of CT scan is lesser there by reducing the diagnostic yield.

MRI:

This is actually the gold standard investigation in the diagnosis of vestibular nerve schwannomas. Introduction of gadolinium scanning has infact increased the accuracy of MRI in the diagnosis of acoustic schwannomas as it is preferentially taken up by schwannomas. Now it is a must that gadolinium be used in all patients before scanning to accurately diagnose the presence of schwannomas in the Cerebello pontine angle. In gadolinium enhanced images the schwannomas enhance brightly in both T1 and T2 weighted MRI images. T1 weighted images are brighter on fat density and T2 weighted images which are brighter in fluid density.

Signs and symptoms of CP angle tumors:

1. Unilateral hearing loss
2. Unilateral tinnitus
3. Aural fullness
4. Lightheadedness

5. Facial hypesthesia
6. Decreased corneal reflex

Audiometric / vestibular tests:

1. PTA
2. Speech discrimination scores
3. Impedance audiometry
4. BERA
5. Electronystagmography

Imaging:

1. Plain radiographs – (Unreliable)
2. CT imaging
3. Air cysternography
4. MRI imaging (most accurate)

Presence of mass in the CP angle calls for differentiation between schwannoma and meningioma. Vestibular schwannoma is usually globular in shape and is centered at the internal auditory canal. Usually an acute angle is formed between the posterior face of temporal bone and the tumor surface. Vestibular schwannoma classically enters the internal acoustic meatus creating an appearance of an icecream cone. The internal acoustic meatus could be seen eroded by the enlarging schwannoma. Vestibular schwannomas may occasionally contain areas of cystic degeneration. Meningiomas on the other hand are more sessile, extends along the petrous ridge. It forms an obtuse angle with the petrous bone. These lesions also demonstrate dural tail of enhancement along its periphery. Meningiomas have a propensity to infiltrate the temporal bone.

Goals of treatment of vestibular schwannomas:

1. Primary objective is to preserve life
2. Preservation of facial function
3. If serviceable hearing is present preoperatively preservation of it
4. Complete tumor removal if feasible

Treatment algorithm of CP angle tumors:

1. Observation
2. Surgery – Trans-lab approach, middle cranial fossa approach, retrosigmoid suboccipital approach
3. Stereotactic gamma radiation

Role of observation in vestibular schwannoma:

The tumor doubling time of vestibular schwannoma is very long. There are instances where patients have been found to be asymptomatic even after being diagnosed with vestibular schwannoma for a period of 10 years. Studies reveal growth rate of these tumors to be about 1-2mm per year. The rule of the thumb being that if the patient is young then surgery is indicated because of the long life expectancy. It is ideal for all the patients under observation to keep repeating MRI scans every 6 months to assess tumor size. The advent of stereotactic gamma radiation has reduced the morbidity and mortality in most of these patients. It can also be safely performed even in elderly patients suffering from vestibular schwannomas.

Surgical management of CP angle lesions:

This is the most specific of the treatment modalities available in managing these patients. This is more so for vestibular schwannomas.

1. Surgery is a team effort needing a neurosurgeon, neurotologist who are familiar with microscopic anatomy of temporal bone with a good anesthetic support.
2. Be it any approach the most critical element is the availability of a good operating microscope.
3. It is ideally performed in specialized high volume centers where the experience in managing these patients are considerably high
4. The hospital staying time is also lesser when the surgery is performed in specialized centers
5. Facial nerve monitoring facilities are a must for all these procedures.

Translab approach:

This approach offers obvious advantages for patients with vestibular schwannomas.

1. This is the most direct route to cerebellopontine angle
2. It requires only minimal cerebellar retraction for adequate exposure
3. Facial nerve can be identified in the temporal bone always in the area free from disease
4. The fundus of the internal acoustic canal is widely exposed
5. Any damage to the facial nerve can be attended to by performing nerve grafting

Disadvantage of this procedure is the sacrifice of residual hearing in the operated side. If the patient has good residual hearing then it is worthwhile preserving it by performing other types of surgical procedures which preserves residual hearing.

Procedure:

Ideally performed under general anesthesia.

Incision is made 2 cms behind the post aural sulcus (William wild's incision)



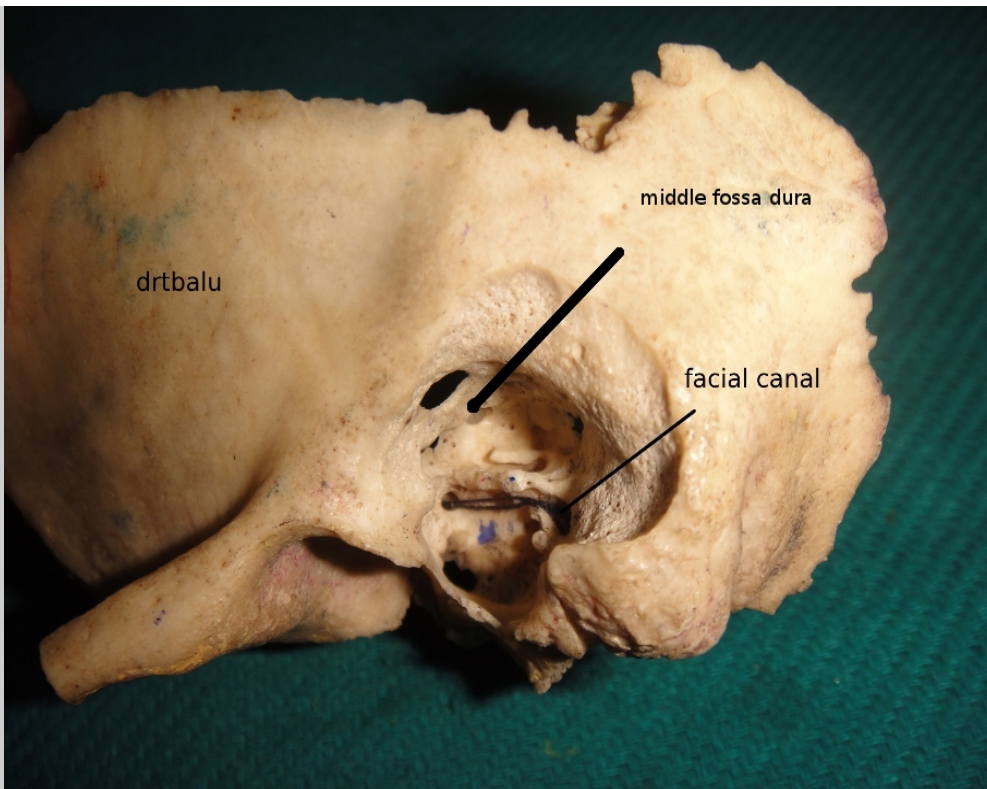
Figure showing post aural incision

Complete mastoidectomy is done.

Middle cranial fossa dura is identified.

Sigmoid sinus is identified

Lateral canal, fossa incudis and facial nerve are identified



Dissected temporal bone showing the structures seen after mastoidectomy

The sigmoid sinus is decompressed with the help of diamond burr. An island of bone is preserved over the dome of the sinus. This prevents the sigmoid sinus from bulging into the operating field. This island of bone is known as the Bill's island.

Bill's island method of decompressing lateral sinus:

This is performed by drilling an island of bone over the most prominent part of sigmoid sinus. The sinus plate is skeletonized using a large diamond burr till just 0.5 mm of bone remains over the sigmoid sinus. Then a groove is drilled circumferentially over the most prominent part of the bony covering exposing a strip of sinus all around it. The island of bone is fractured using a suction tip and is held away from the field while the anterior margin of the bony prominence is drilled. The superior and inferior margins of the bony prominence is drilled and the whole bony prominence which has been thinned out is pushed inwards along with the sinus.

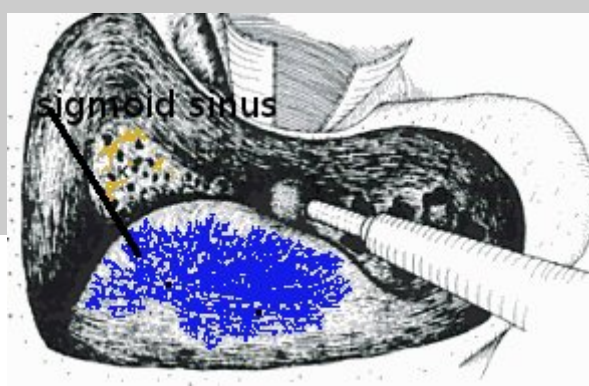


Figure showing the sigmoid sinus being skeletonized using Bill island method

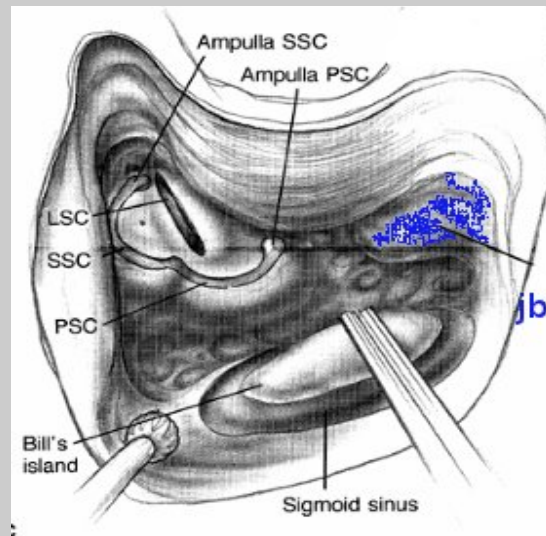


Figure showing sigmoid sinus being decompressed

Labyrinthectomy is begun by removal of bone in the sinodural angle along the horizontal semicircular canal. All the three semicircular canals are opened and followed into the vestibule. Enough care should be taken to identify the ampullae of the three semicircular canals and the subarcuate artery. These are vital landmarks in the identification of the internal auditory meatus. Bone is gently drilled out in the posterior cranial fossa dura area medial to the sigmoid sinus. The endolymphatic duct and sac comes into view. The inferior aspect of internal auditory canal is identified now. The variably placed jugular bulb is at risk at this stage. The jugular bulb can be identified by identifying the ampulla of the posterior semicircular canal. The jugular bulb is almost always located inferior to the ampulla of the posterior semicircular canal. Dissection inferior to the ampulla of posterior semicircular canal is carefully proceeded by using diamond burr till the jugular bulb is identified. The jugular bulb indicates the inferior level of dissection. After identification of the jugular bulb bone is removed along the inferior aspect of the internal acoustic meatus till the cochlear aqueduct is reached. Identification of cochlear aqueduct is really vital in order to avoid damage to the 9th, 10th and 11th cranial nerves. These nerves lie anterior and inferior to the cochlear aqueduct. The inferior edge of the internal acoustic meatus is clearly delineated and now the posterior aspect of the canal is skeletonized until the superior edge of the internal acoustic meatus is identified. Bone is then gently drilled between middle cranial fossa dura and the internal auditory canal. Once the medial portion of the internal auditory meatus is exposed the remaining piece of bone over the porus portion of the internal auditory meatus may be removed. The transverse crest is identified at the level of fundus of the internal acoustic meatus. Now is the time to identify the Bills

bar superiorly. This is a vital landmark for the labyrinthine portion of the facial nerve. When the dissection is over you will find the bone covering the middle and posterior cranial fossa is removed completely along with the bony covering of the entire posterior 2/3 of the internal acoustic meatus. The posterior cranial fossa dura is opened inferior to and parallel to the superior petrosal sinus over the midportion of the internal acoustic meatus. Using the Bills bar as a guide and with the help of a hook the superior vestibular nerve is separated from the facial nerve. If the tumor is smaller than 2 cm then it need not be debulked before separating it from the facial nerve. If the tumor is large then its posterior aspect should be inspected to find an avascular area for dissection and to ensure that the facial nerve is located in its customary anterior position. The capsule of the tumor is incised in the avascular plane and is removed using House-Urban dissector or by using ultrasonic aspirator. Continuous monitoring for facial nerve function during this stage will go a long way in preventing damage to the nerve. After tumor removal meticulous hemostasis is secured. The tensor tympani tendon is cut and the eustachean tube opening is occluded using surgicel or abdominal fat. The middle ear is obliterated by temporalis muscle flap and the dural defect is closed with dural silk sutures. A titanium plate is screwed into position in order to obliterate the defect in the temporal bone. Wound is closed in layers and mastoid dressing is applied.

The main limitation for this procedure is not the tumor size by the inferior portion of the cerebellopontine angle associated with an anteriorly placed sigmoid sinus and high jugular bulb. In patients in whom it is difficult to separate the mass fully from the facial nerve it is prudent to leave a sleeve of tumor tissue around the facial nerve in order to preserve it. Anatomical preservation of facial nerve is quite possible in more than 90% of cases when translab approach is used to remove the tumor.

Complications of translab surgical approach to excise vestibular schwannoma:

1. Facial nerve paralysis
2. CSF rhinorrhoea
3. Lateral rectus palsy
4. Meningitis due to persistent csf leak
5. Hearing is lost in a majority of these patients. If the semicircular canals are closed with the help of bone wax then some amount of hearing preservation is possible

Steps that should be taken to prevent csf otorrhoea:

1. By paying attention to proper dural closure
2. By obliterating the eustachean tube orifice completely
3. By using abdominal fat to obliterate the cavity and to layer the wound in a tight fashion
4. In patients with persistent csf leak then blind sac closure of the external auditory canal combined with a through closure of the eustachean tube orifice should be performed

Management of meningitis:

This is one of the most important complication of translab approach to acoustic schwannoma. The following features are pointers for the diagnosis of meningitis:

1. Mental status changes
2. Meningismus
3. Increasing intensity of post op headache
4. Changes in vital signs
5. Pyrexia

In patients with suspected meningitis early administration of culture specific antibiotics parenterally will prevent neurological sequelae. For purposes of culture CSF samples should be obtained from these patients as early as possible by performing lumbar puncture.

Middle cranial fossa approach to internal acoustic meatus:

This approach has the unique potential of hearing preservation while allowing exposure of lateral end of internal acoustic meatus. This approach can safely be used to remove only tumors that measure 2cms or less in their largest dimension, this include the intracanalicular portion.

Advantages of middle cranial fossa approach to internal auditory meatus:

1. The dissection is completely extradural and hence devoid of complications
2. The facial nerve is identified at the lateral end of internal acoustic meatus. This enables easy establishment of a plane between the facial nerve and the tumor mass
3. Hearing is preserved to a large extent

In order to remove large tumors extending up to the CP angle an extended middle middle cranial fossa approach has been devised.

Disadvantages:

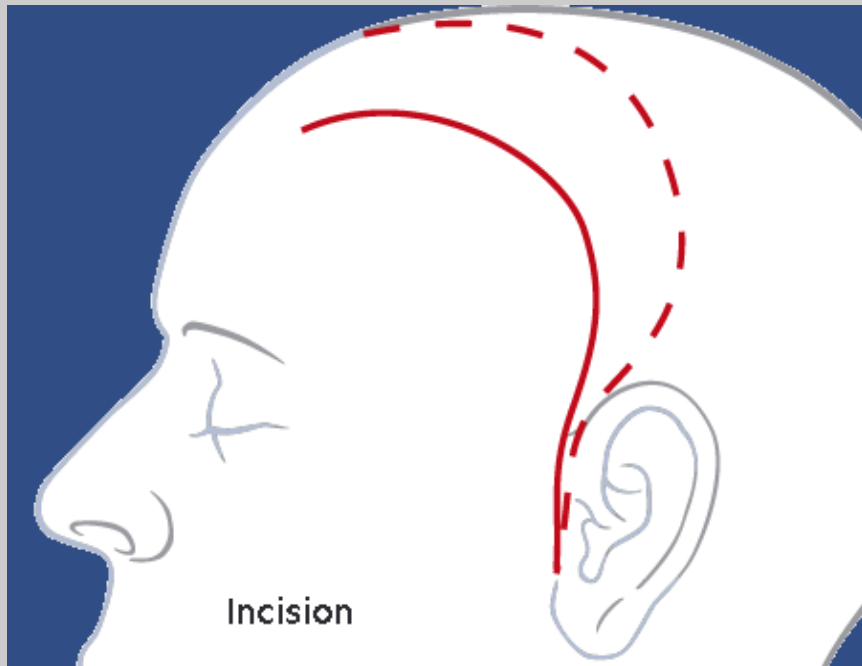
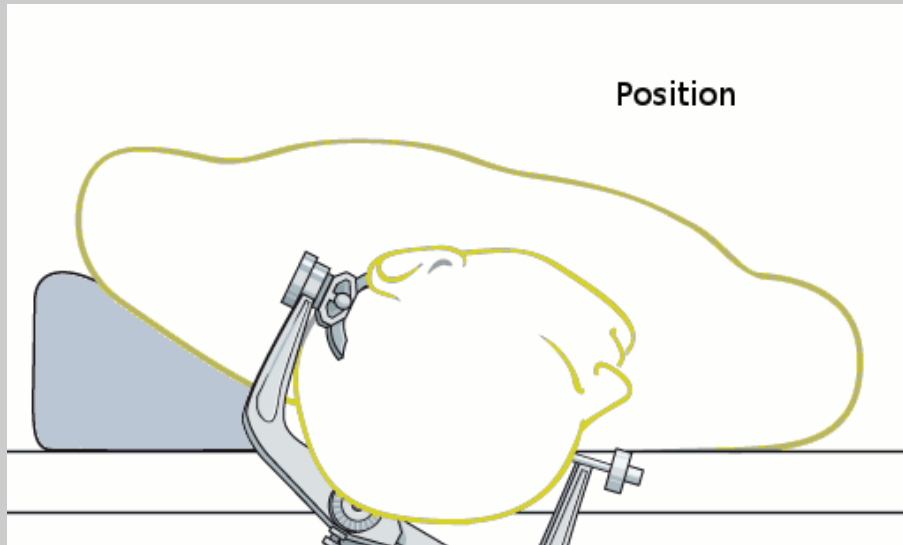
1. More challenging approach than trans lab
2. The surgeon should work around the facial nerve inorder to reach the tumor because the facial nerve is located in the superior portion of internal auditory canal
3. Landmarks in the floor of the middle cranial fossa may sometimes be absent adding to the woes.

Surgical technique:

The facial nerve and the auditory brainstem response is monitored throughout the procedure. The surgeon is seated at the head end of the patient. The head of the patient is turned away from the affected ear and the malar eminence should be at its highest point. A shoulder roll can be placed in order to prevent excessive traction to the cervical spine. The head is fixed with a head holder. In this position the frontal lobe of the brain falls away from the floor of the anterior cranial fossa thereby obviating the necessity for frontal lobe traction.

Incision:

The incision is begun from the pretragal area. The incision then extends superiorly and then curves posteriorly just above the auricle and then makes a semicircular curve back towards the eyebrow. The completed incision resembles a question mark.



Incision is deepened. Temporalis muscle is exposed. A T shaped incision is made cutting the temporal fascia along with the temporalis muscle. This creates a posterior musculofacial flap which can be used for reconstruction.

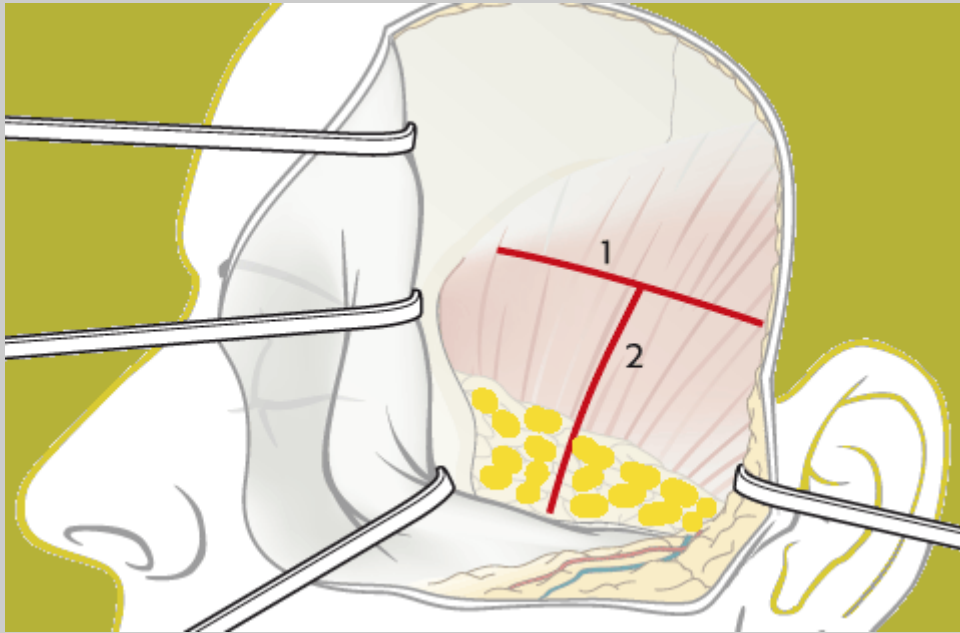


Figure showing T shaped incision over Temporalis fascia

It is always better to incise the temporalis muscle near its insertion close to zygomatic arch. The temporalis muscle is elevated in the subperiosteal plane and retracted anteriorly. A temporal craniotomy is performed using a high speed drill under continuous saline irrigation. A bone flap measuring 5 x 5 cms is raised. The temporal craniotomy is positioned in such a way that 2/3 of it is positioned anterior to the external auditory canal. More anterior the better it is.

Dural dissection:

The dura is dissected from the floor of middle cranial fossa till the arcuate eminence is reached. The greater superficial petrosal nerve and the petrous ridge should be identified at this juncture. The middle meningeal artery is identified at the level of foramen spinosum and it serves as the anterior limit of dissection. Posteriorly the dura is elevated up to the level of petrous ridge. House urban retractor is introduced in such a way that the blade of the retractor is placed over the posterior petrous ridge. The retractor should be positioned in such a way that it is in line with the external auditory canal and bisects the angle formed by the greater superficial petrosal nerve and arcuate eminence.

Using the greater superficial petrosal nerve and arcuate eminence as guides bone removal is begun and the internal acoustic meatus is identified. Drilling is continued till the contents of the internal acoustic meatus are exposed for 270 degrees. The internal auditory canal is followed laterally towards the fundus. Progressively smaller burr bits should be used as one gets closer to the fundus area to avoid damaging the basal turn of the cochlea and the ampula of the superior semicircular canal. The internal auditory canal is exposed only 90 degrees at the level of fundus. The proximal 3mm of the labyrinthine portion of the facial nerve canal should be drilled and exposed in order to

allow for swelling that could occur in the facial nerve during the process of dissection. The dura overlying the facial nerve can now be incised under high magnification. Fine hooks can be used to separate the superior vestibular nerve from the facial nerve.

The tumor is dissected from medial to lateral off the cochlear nerve. This step is rather crucial for preservation of residual hearing. At this juncture the nerve of origin of the tumor is cut while the cochlear division and the other vestibular division are preserved. After removal of tumor a cotton pledget soaked in papaverine is placed over the cochlear nerve at the internal acoustic meatus. This is known to increase the blood flow to the nerve there by facilitating preservation of hearing. The dural defect is packed with abdominal fat and the craniotomy bone plate is replaced and anchored with titanium plate and screws.

Suboccipital approach:

This is the traditional neurosurgical approach to the skull base. This is rather useful in patients with excellent residual hearing which needs to be preserved after surgery. It is ideal for patients in whom the tumor is medially located within the internal auditory meatus and does not protrude more than 2 cms into the cerebello pontine angle.

Advantages of this approach:

1. Excellent preservation of hearing
2. Good visualization of brain stem and lower cranial nerves

Disadvantages of this approach:

1. Lateral tumors cannot be removed without breaching the labyrinth
2. There is high risk of air embolism as the patients head is higher than that of the chest
3. Injury to endolymphatic duct could cause long term complications
4. Cerebellum needs to be retracted for better visualization with its attendant complications
5. There is a high incidence of post op headache

Procedure:

The patient is placed in supine position. The head is shaved. Facial nerve monitoring and ABR monitoring should be performed on a continuous basis during the entire surgical procedure.

Incision:

A curvilinear incision is given 3cms behind the post aural sulcus. This incision is curved slightly posteriorly in its inferior portion. A limited mastoidectomy is performed to decompress the sigmoid sinus. Cauterization of the mastoid emissary vein may be necessary at this juncture. The sigmoid sinus is exposed up to the level of the jugular bulb. A 4 cm craniotomy is performed over the suboccipital region. The underlying dura is stripped off the bone and the bone flap is removed. The dura is incised in a semilunar fashion with its base positioned medially. The dura is held apart by stay sutures. The sigmoid sinus is reflected anteriorly and the cerebellum posteriorly. The dissection is carried medially till the lateral cistern is reached. CSF starts to leak at this juncture and the cerebellum falls posteriorly. The dura over the posterior portion of the internal acoustic meatus is stripped down to the level of bone. A diamond burr is used to drill the internal auditory canal beginning from its posterior lip. The whole internal auditory canal is skeletonized up to 180 degrees of its circumference. The endolymphatic duct and sac are useful landmarks because the

duct lies superficial to the posterior semicircular canal. Tumor is dissected using House Urban dissector or Ultrasound aspirator. Care should be taken not to damage the facial nerve and cochlear division of 8th nerve. The bone edges are sealed with wax and the dural incision is sutured. The bone flap is replaced.

Stereotactic radiosurgery:

This is not true surgery but is a type of focussed irradiation. This technique was first introduced by Leskell in 1969. The term surgery is used to indicate that only a selective area of tissue is addressed as in a surgical procedure. Tissue destruction is achieved by delivering by multiple ionizing beams of radiation to a specific intracranial target. This is usually delivered in a single treatment session. This one session treatment has such a dramatic effect in the target zone the changes are considered “surgical”. With the use of three dimensional computer aided planning and immobilizing the patient the damage to adjacent normal tissue can be greatly minimized. This procedure is very useful in treating tumors involving brain tissue. Acoustic neuroma is amenable to stereotactic radiosurgery. It may hence be a preferred treatment modality in patients with tumors involving inaccessible areas of brain. This procedure can also be considered in treating radio recurrent tumors also.

Types of stereotactic radiosurgery:

1. Particle beam (proton)
2. Cobalt – 60 based (photon)
3. Linear accelerator based (Linac)

Among these machines available the Cobalt 60 based machines also known as gamma knife is extremely accurate in targetting and precisely treating patients with brain tumors.

Fractionated radiotherapy:

The source of radiation is often linear accelerator. The total dose is calculated using computer model of the lesion. This dose is delivered in a fractionated manner (in small doses). This minimizes damage to normal tissue while targetting the malignant ones.

Indications:

1. Vestibular schwannomas less than 2 cms in their greatest dimension
2. Vestibular schwannomas in elderly who are poor surgical risk

Patients who undergo stereotactic radiosurgery for vestibular schwannoma have prolonged sense of imbalance when compared to those who undergo micro surgical removal of the tumor. This has been attributed to the fact that in micro surgical procedures the vestibular system is totally damaged and central compensation is thereby complete. Where as in patients undergoing stereotactic radiourgery imbalance is prolonged because the labyrinthine function is diminished and nor totally destroyed leading to incomplete central compensation.

Radiotherapy also has the unique risk of causing malignant transformation of normal tissue.

Complications of CP angle tumors:

1. Sensorineural hearing loss
2. Facial nerve paralysis
3. CSF leak
4. Meningitis
5. Intracranial hemorrhage
6. Air embolism
7. Cerebellar ataxia
8. Head aches

Meningiomas involving CP angle:

This is the second most frequent tumor involving this area. Meningiomas arise from cap cells that collect in clusters around arachnoid villi tips. These cap cells are more prevalent around the dural venous sinuses and in areas where cranial nerves exit the brain tissue.

Meningiomas are of two types:

1. Globular
2. Flat

These tumors don't metastasize but commonly recur because of their propensity for bone invasion.

Meningiomas arising from the internal acoustic meatus cause symptoms similar to that of vestibular schwannoma. Since most meningiomas arise from the posterior surface of the petrous bone they don't enter the internal auditory meatus early. They are usually large when they cause auditory / vestibular symptoms. Meningiomas arising inferiorly close to sigmoid sinus cause:

1. Hoarseness of voice
2. Dysphagia
3. Tongue atrophy

Signs of meningioma in this area are commonly related to the eye:

1. Spontaneous nystagmus
2. Facial hypesthesia
3. Gait disturbances

Patients with meningiomas have better hearing than those with similar sized vestibular schwannomas.

Radiological differences between schwannomas and meningiomas:

1. Meningiomas are more homogenous in CT than schwannomas
2. They take up contrast material more uniformly than schwannomas
3. Schwannomas are heterodense because of areas of necrosis which are likely to be present
4. Angiomatous meningiomas are contrast enhancing and more invasive

Removal of meningiomas surgically is fraught with complications. Since they are radiosensitive stereotactic radiosurgery is preferred to conventional surgical procedure.

Epidermoids:

Epidermoid tumors in CP angle area are considered to be the third common lesion seen in this area. These patients classically have early facial paralysis and hemifacial spasm. In fact hemifacial spasm is more diagnostic of epidermoid in the CP angle area than any other symptom. Behaviorally these tumors resemble cholesteatoma of the ear.

Epidermoids need to be removed surgically. These lesions are adherent to adjacent structures which include the cranial nerves making their removal that much difficult.

Schwannomas involving other cranial nerves:

Schwannomas can arise from any cranial nerve in the posterior cranial fossa. The symptoms are determined by the nerve of origin and its location.

1. Schwannoma originating from the 5th cranial nerve causes hemifacial hyperesthesia along with enlargement of meckel's cave in the CT scan.
2. Schwannomas involving the 12th cranial nerve causes hemiatrophy of tongue.
3. When 9th, 10th and 11th cranial nerves are involved with schwannomas then they cause jugular syndrome which include dysphagia, hoarseness and shoulder weakness. Enlargement of hypoglossal canal can be seen in CT scan images of these patients.

Vascular tumors involving the CP angle:

1. Glomus jugulare
2. Hemangioma
3. Hemangioblastomas

These patients manifest with pulsatile tinnitus.

Miscellaneous tumors involving CP angle:

1. Arachnoid cysts: These are thin walled sacs containing CSF. They develop from adhesions within the arachnoid of the internal auditory canal.
2. Cholesterol granuloma – In this area spreads posteriorly
3. Dermoids
4. Teratomas
5. Chordomas

All these miscellaneous tumors in this area are amenable to surgical management.