Superior semicircular canal dehiscence syndrome and its management

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Introduction:

This condition was first described by Minor et al in 1998. This is a troublesome disorder characterized by vertigo and oscillopsia induced by loud sounds or changes in the pressure of external auditory canal or middle ear cavity. These patients may present with the following features:

1. Autophony
2. Hyperacusis
3. Pulsatile tinnitus
4. Hearing loss

When these symptoms are mild, they can be managed conservatively. Surgical intervention is very rarely needed if they are debilitating.

Pathophysiology:

This syndrome is characterized by the formation of a “third opening” i.e. “third window” between the middle cranial fossa and the superior semicircular canal. This is formed because of a bony defect in the canal. This abnormal communication results in vertigo and oscillopsia which is commonly induced by loud sounds. This particular phenomenon is known as Tullio phenomenon. Giddiness is caused when valsalva maneuver is performed or even when pressure changes occur in the external auditory canal.

In the presence of dehiscence of superior semicircular canal, the hydro-acoustic waves flowing through the cochlea are inadvertently transmitted through out the labyrinthine system causing activation of vestibular system which manifests as giddiness.

Hydraulically this dehiscence could serve as a pressure release mechanism thereby causing less pressure to be distributed in the inner ear. Invariably there is an outward bulging caused through the dehiscence in the middle cranial fossa. This causes ampullofugal deflection of cupula of the superior semicircular canal which is perceived by the brain as body movement. This results in vestibular symptoms. Similarly, intracranial pressure fluctuations or negative pressure in the external / middle ear results in inward bulging of the membranous canal leading on to ampullopetal deflection of cupula. These two scenarios support noise / pressure induced vertigo which is observed in these patients.

Increased compliance of inner ear is known to contribute to conductive hearing loss and perception of pulsatile tinnitus observed in these patients. The so called third window could weaken / dampen the energy transmission produced by movement of foot plate of stapes resulting in reduction of sound transmission to the cochlea and hence the hearing loss. It has also been observed that the presence of third window alters the normal impedance inequality between the oval and round windows causing a certain amount of hyper conduction of sound which is perceived by the patient as autophony / hyperacusis.

Clinical features:

Giddiness

Oscillopsia

Tullio phenomenon – Giddiness induced due to exposure to loud noise.

Henebert's sign – Giddiness is induced by pressure changes in the external auditory canal. This can be clinical demonstrated by alternate pressing and releasing tragus of the ear. This leads to pressure changes in the external auditory canal causing giddiness.

Autophony – This is another manifestation of this condition where the patient hears his / her own voice loudly. This is due to hypersensitivity to bone conducted sounds. These patients can hear a vibrating 512 tuning fork placed at the ankle in the affected ear.

Diagnosis is suspected based on the above stated clinical features. Nystagmus evoked on exposure to loud noise / increased pressure in the external auditory canal is usually oriented in the same plane as the dehiscent semicircular canal.

Audiometric evaluation:

Findings typically include supernormal bone thresholds and low frequency conductive hearing loss. Tympanometry and acoustic reflexes are near normal.

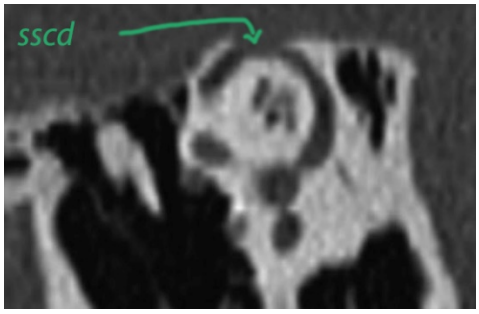
Cervical vestibular evoked myogenic potential (cVEMP) in these patients will elicit responses at lower thresholds (less than 70dB) to tone burst testing than in persons without the syndrome. Studies conducted by Zuniga et al showed that cVEMP threshold results showed a sensitivity and specificity ranging from 80% - 100% for diagnosis of superior semicircular canal dehiscent syndrome. Ocular amplitude VEMP had a sensitivity had a sensitivity and specificity greater than 90%.

Imaging:

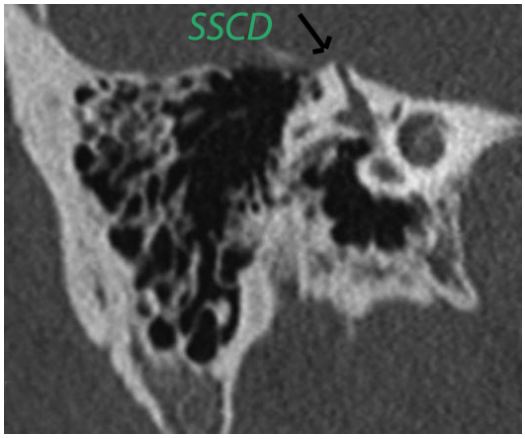
Temporal bone CT imaging should be performed for all patients with suspicion of SSC dehiscence. CT temporal bone confirms the diagnosis. HRCT with 0.5 mm cuts of temporal bone is ideal. Projections in the plane of superior canal (Poschl views) and in a plane perpendicular to the SSC (Stenver's view) is taken.

Based on CT imaging two types of SSC dehiscent syndromes have been described. They include:

1. Dehiscence of the arcuate eminence
2. Dehiscence in the region of superior petrosal sinus



Superior semicircular canal dehiscence as seen in Poschl view



Superior semicircular canal dehiscence as seen in Stenver's view

MR imaging is very useful as it accurately diagnoses this condition nearly 100%.

Etiology:

Unknown. There is a high degree of suspicion that congenital anomaly could play a part. Thinning of bone overlying superior semicircular canal occurs bilaterally in nearly a third of these patients. Some patients even have been reported to be symptomatic during their childhood. In contrast to adults SSCD in children usually presents with auditory symptoms first and conservative management should be started as soon as possible.

Low intensity cranial trauma:

Repeated low intensity cranial trauma in patients who are into combative sports or diving. The defect in the floor of the middle cranial fossa at the level of SSC could occur secondary to increments in the pressure of CSF.

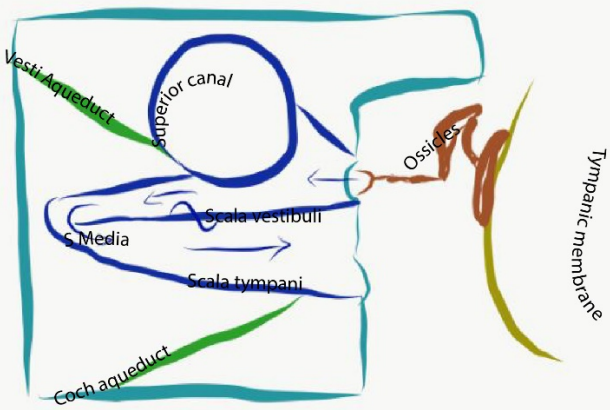


Diagram showing anatomy of Inner ear and sound conducting mechanism

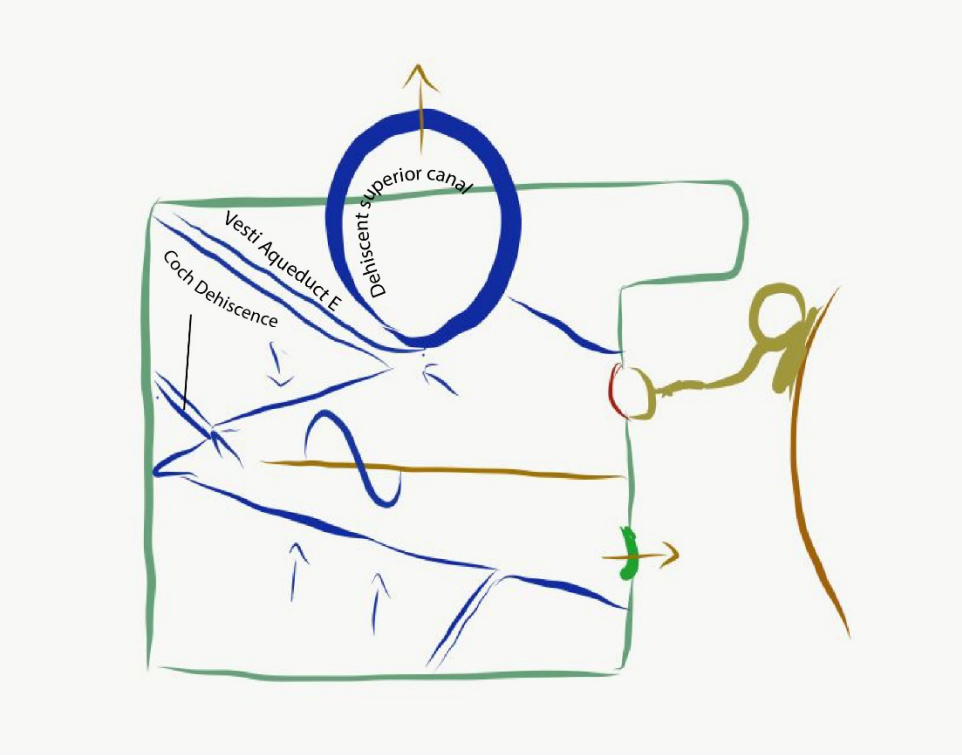


Image showing cochlear dehiscence in addition to dehiscent superior canal.
Bone conducted sounds gets accentuated as shown in the diagram because of the existance of multiple windows

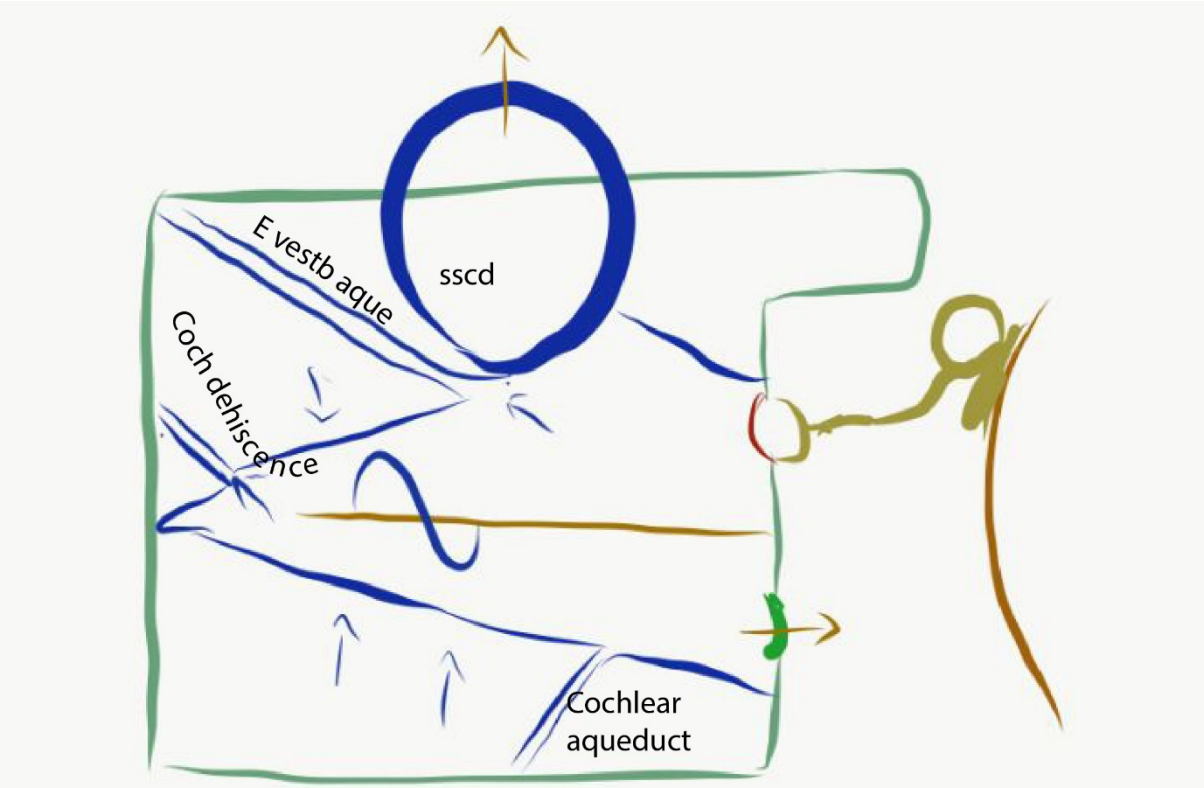


Figure showing bone conduction of sound in the presence of cochlear dehiscence

Management:

Patients with mild symptoms should be managed conservatively. Surgery is reserved only for patients with debilitating symptoms. When surgery becomes necessary the bony dehiscence can be resurfaced, plugged or capped using different surgical approaches.

Middle cranial fossa approach:

This approach to treat SSCD syndrome was first described by Minor et al. A 4x4 cm craniotomy is drilled. The temporal lobe is retracted, arcuate eminence is identified. Invariably at this point the dehiscence could easily be identified. The superior semicircular canal is opened using a diamond drill and then it is plugged. In addition, the canal may also be capped or resurfaced with bone pate, bone wax, hydroxyapatite cement or soft tissue.

The advantage of this approach includes the direct access to arcuate eminence defect without the need for labyrinthine bone removal and the exposure of surrounding cranial base if the repair is needed for other associated tegmen defects. Concomitant resurfacing of the tegmen mastoideum and tympani is performed since the floor of the middle cranial fossa is usually thin. Resurfacing of the dehiscent canal also prevents chronic stimulation from the pulsating temporal lobe of the brain.

Transmastoid approach:

This procedure begins with a classic post aural incision as done for mastoidectomy. Mastoidectomy is completed using a cutting burr and suction irrigation. The following structures are skeletonized:

Sigmoid sinus

Posterior fossa dura

Middle fossa dura

Presigmoid area

The horizontal, posterior and superior semicircular canals are identified and skeletonized with a diamond burr. The area of dehiscence is identified, the middle cranial fossa dura is carefully elevated from the dehiscent superior canal. In patients with dehiscence at the superior petrosal sinus, the sinus can be exposed at the sinodural angle, which lies posterior to the solid angle and can be followed to the superior canal.

Two points of the bony labyrinth are fenestrated with a 1 mm diamond burr, and the endosteum is opened just inferior to the fenestrated apex of the superior canal, on the ampullated and non-ampullated portions of the canal. Care is taken to avoid suction or manipulation of the membranous labyrinth to prevent damage to these vital structures. Bone dust, bone wax, bone pate, fascia and even bone chips can be used to fill the lumen of the superior canal at the points of fenestration. If bone wax is used then it is recommended that application of two wax spheres, 2 mm in size would be sufficient to occlude the canal without risk of damaging the neuroepithelium. Conchal cartilage could be harvested and placed in an intracranial extradural position, repairing the middle cranial fossa floor defect. Finally, the wound is closed in layers.

If Transmastoid approach is selected tragal perichondrium is placed in the space between the dura and the dehiscence by folding it over the top of an annulus elevator and inserting it over the superior canal. This procedure is performed after dissection of the middle cranial fossa dura from the dehiscent canal is completed. The dura and overlying temporal lobe / superior petrosal sinus will stabilize the position of the graft, with no tendency to extrude.

This approach avoids the risk of performing craniotomy in the middle fossa area. This results in lower morbidity and shorter hospital stay. One disadvantage with this approach is that the dehiscence is visualized with more difficulty compared to that of middle cranial fossa approach. Another huge limitation is when the

tegmen hangs too low, precludes the safe exposure of dura just lateral to the superior canal. This is the reason why pre op CT image is very important in order to anticipate this difficulty in addition to the fact that the image identifies the precise size, and location of the dehiscence.

Endoscope assisted middle cranial fossa repair:

This is more advantageous than the binocular microscopy, since it provides high definition view of the middle cranial fossa. In this technique, following the incision, a temporalis fascia graft is harvested. A periosteal flap is made and a minicraniotomy (3x2cm) is preferred. Under microscopic vision, the dura is gently lifted off the tegmen mastoideum and tegmen tympani until the arcuate eminence is visualized. Then, a 30-degree endoscope is introduced in order to visualize the defect. The House Urban middle cranial fossa retractor is used to maintain the retraction of the temporal lobe while working with the endoscope. The dura is dissected from the medial aspect of the defect under endoscopic view. The ampullated and non-ampullated limbs of the defect are gently occluded using bone wax.

Round window reinforcement:

This procedure is a low risk one and hence should be offered as the first procedure in patients with mild symptoms. Tympanomeatal flap is elevated under local / general anesthesia. Canal plasty may be performed if better exposure is needed. The round window niche and promontory are denuded of mucosa and the round window is reinforced with temporalis fascia, tragal cartilage, perichondrium, fat or connective tissue. The tympanomeatal flap is repositioned and the external canal is packed.

Complete round window occlusion is another technique that has been used in the management of SSCD syndrome. Authors caution against this procedure fearing that symptoms might worsen during late post op period.

Studies reveal that a combination of plugging and resurfacing achieves better long-term control of symptoms than resurfacing alone. When resurfacing alone is performed, a complete of the defect is not guaranteed and this area may remain sensitive to pressure changes.