Third Window Abnormalities a spectrum of disorders

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Introduction

Defects in the integrity of the bony structure of inner ear

First described by Minor et al in 1998

Potential third window sites include:
- Dehiscence of semicircular canals
- Enlargement of vestibular aqueduct
- Dehiscence of scala vestibuli side of cochlea

Mechanism of hearing loss

1. Merchant & Rosowski proposed a universal theory for the underlying mechanism of hearing loss in these patients
2. Normal sound conduction is transmitted through the oval and round windows which serve as fluid interfaces between the middle ear and perilymphatic spaces of inner ear. Presence of dehiscence / third window in the labyrinthine bone will cause disruption of this sound conduction mechanism leading on to deafness
The fluid spaces of inner ear are completely surrounded by the bone of otic capsule. There are several openings or windows that connect the inner ear fluid spaces to the cranial cavity or the air filled middle ear cavity.

What is it?

Oval window is the primary window

Round window is the secondary window
Fluid spaces of the normal inner ear are nearly completely surrounded by the bone of otic capsule. There are several openings (windows) that connect the inner ear fluid spaces to the cranial cavity or to the air filled middle ear cavity.

Oval window is the primary window

Round window is the secondary window

Role of primary & secondary windows

These windows are large in area and short in length. This minimizes the impedance of fluid flow thereby facilitating sound transmission from middle to inner ear.
Normal Third Window

Presence of third windows enabled normal bone conducted hearing even when both oval & round windows are closed.

Under normal conditions they are functionally closed.

These are longer and are of smaller caliber. They have high impedance.

Third window

1. Vestibular aqueduct
2. Cochlear aqueduct
3. Foramina for blood vessels

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Pathologic Third windows

Anatomic

Semicircular canal
1. Superior canal dehiscence
2. Posterior canal dehiscence
3. Lateral canal dehiscence

Vestibule
1. Large vestibular aqueduct
2. Inner ear malformations causing dehiscence between internal auditory canal and vestibule

Cochlea
1. Dehiscence between carotid canal & Scala vestibuli
2. Inner ear malformation causing dehiscence between internal auditory canal and scala vestibuli. E.g. DFN-3 (X linked deafness with stapes gusher)

Diffuse / distributed third window

Paget’s disease of temporal bone
Incidence

- 2-10% in superior canal (CT studies)
- 0.3 – 4.5% posterior canal
- Perilabyrinthine fistula involving lateral canal 15%
- Enlarged vestibular aqueduct seen in 1% of dissected temporal bones
Superior semicircular canal Dehiscence

SSD is characterized by loss of bone covering the superior canal.

There is a potential communication between the canal and cranial cavity.

Minor et al. described this condition in 1998.

There is lack of middle ear pathology associated with this lesion.

This is the best documented and most investigated 3rd window lesion.
SSC Dehiscence syndrome Signs & Symptoms

Some patients develop involuntary head movements in response to loud sounds. Vestibule contributes nerve to nerve supply to neck muscles.

Triggers symptoms of migraine.

The question whether this lesion is progressive is still unanswered.

**Symptoms**
1. Bone conduction hyperacusis
2. Autophony
3. Pulsatile tinnitus
4. Sound / Pressure induced vertigo
5. Chronic disequilibrium
6. Aural fullness
7. Deafness (conductive)

**Signs**
1. Oscillopsia
2. Tullio phenomenon – giddiness induced due to exposure of loud noise
3. Henebert sign – Giddiness induced on pressure being applied to external canal
4. Autophony – patient hears his / her voice loudly due to bone conduction hyperacusis
SSCD Clinical Examination

Nystagmus in the plane of superior canal

Nystagmus is observed when audiometer is used with a range of different frequency tones. Eye movements can be observed by using Frenzel glasses.

Tuning fork tests reveal conductive deafness with increased sensitivity to bone conducted sounds.

Weber test with 512 tuning fork will be heard more loudly in the ear with greater bone conduction. Hyperacusis.

Nystagmus can be observed on compressing the tragus of external canal.

Ear drum appears normal with no evidence of middle ear pathology.

In patients with large dehiscence >5mm superior semicircular canal rather than being stimulated will be impaired due to autoplugging effect of temporal bone dura which prolapses.

Sometimes tuning fork can be heard in the affected ear even when placed over the medial malleolus.
SSCD Diagnosis Imaging

CT Imaging planes

1. In the planes of semicircular canal (Poschl view)
2. In a plane orthogonal (perpendicular) to the semicircular canal (Stenver’s view)

Many patients with CT evidence of dehiscence may be asymptomatic due to the protective role of inelastic dura which prevents pressure transmission through dehiscences.

CT Temporal bone would be a pointer but not sufficient. Artifacts due to partial averaging can resemble dehiscence.

HRCT 1 mm cuts improves diagnostic accuracy.

MR Imaging

1. Alternative to CT imaging
2. Advised if surgery is being planned
3. Soft tissues are better visualized
4. T2 weighted images are preferred. In these images semicircular canal fluid signal is bright. Loss of this signal can be useful for assessing adequacy of prior surgical plugging.
Pure tone Audiometry

- Mistaken for otosclerosis. Acoustic reflexes are normal in SSCDS.
- Large air bone gap at lower frequencies (250, 500 & 1000 Hz).
- Increased dehiscence length correlates with large air bone gaps.
- Bone conduction threshold in low frequencies is negative or better than normal.
This test in a non-dehiscent ear is thought to reflect function of the saccule (cervical VEMP) or utricle (ocular VEMP).

Cervical VEMP involves inhibitory neural reflex pathway from the saccule to the ipsilateral sternomastoid muscle.

Ocular VEMP involves an excitatory pathway from the utricle to the contralateral inferior oblique muscle.

Ocular VEMP amplitudes have been found to be highly sensitive and specific for diagnosis of SSCD.

Patients with SSCD syndrome have abnormal VEMP findings. They have lower than normal thresholds for cervical VEMP responses to an audible click / tone burst and elevations in the ocular VEMP amplitude responses.
ECoG was used to diagnose endolymphatic hydrops. Patients with SSCD syndrome have elevations in the summing potential to action potential ratio. Elevated SP:AP ratio gets corrected on successful plugging of the dehiscence.
Diagnostic Criteria for SSCD Syndrome

High resolution CT images at least 1mm slice reformatted in the plane of SCC demonstrating dehiscence

At least one of the following symptoms

1. Bone conduction hyperacusis in the form of autophony, audible eye movements, audible foot steps etc.
2. Sound induced vertigo
3. Pressure induced vertigo (pressure induced via nasal / glottic valsalva, pressure applied to external canal
4. Pulsatile tinnitus

At least one of the following diagnostic tests indicating a third mobile window

1. Negative bone conduction thresholds on pure tone audiometry
2. Enhanced VEMP responses (low cervical VEMP thresholds or high ocular VEMP amplitudes)
3. Elevated AP:SP ratio in an ECocG in the absence of sensorineural hearing loss
SSCD Management

Medical

Labyrinthine sedatives. Useful during acute conditions. Vestibular exercises

Surgical

1. Canal plugging and resurfacing
2. Round window procedures
Canal plugging & Resurfacing

**Middle cranial fossa approach**

1. This approach was first described by Minor et al.
2. 4x4 craniotomy is drilled
3. Temporal lobe of brain is retracted
4. Arcuate eminence is identified
5. Point of dehiscence identified
6. SSC is opened using diamond drill bit
7. It is plugged
8. It is resurfaced with bone pate, bone wax, hydroxyapatite cement or soft tissue

**Transmastoid approach**

1. Classic mastoidectomy
2. Following structures skeletonized (sigmoid sinus, posterior fossa dura, presigmoid area
3. Three semicircular canal identified and skeletonized using a diamond burr
4. Area of dehiscence identified, middle cranial fossa dura elevated carefully and the dehiscence is closed
5. In patients with dehiscence of superior petrosal sinus, it can be exposed at the sinodural angle

Advantages of this approach include direct access to arcuate eminence without need for removal of labyrinthine bone. Concomittant resurfacing of tegmen mastoideum and tympani is performed.

Indicated only in patients with debilitating symptoms

Resurfacing of dehiscent canal prevents chronic stimulation from pulsating temporal lobe

Tragal perichondrium should be placed in the space between the dura and dehiscence
Round window Reinforcement

This is a low risk procedure and should be the first one to be offered to the patients.

Can be done under LA / GA

TM flap is elevated

Round window niche and promontory are denuded of mucosa and the round window is reinforced with temporalis fascia / tragal cartilage / fat / connective tissue
Posterior canal Dehiscence

Dehiscence can be between posterior canal and cranial cavity or between posterior canal and jugular bulb.

Findings are similar to SSCD syndrome.

First described radiographically in 1986 by Bony.

Nystagmus is vertical and beats in opposite directions.

High jugular bulb has been attributed to be one of the causes.

VEMP is usually diagnostic.
PSCCD Syndrome Management

Management

Surgical management should be offered to the patient only if the symptoms are debilitating.

Majority of these patients have high jugular bulb

Temporalis fascia graft is applied into the area of dehiscence and then supported by placement of a layer of bone pate. Cortical bone graft is placed beneath the bone pate to complete the repair.

Majority of these lesions can be managed conservatively without resorting to surgery

Jugular bulb should be decompressed and then reduced inferiorly using bipolar cautery and the space is packed with surgicel and bone wax.

Transmastoid approach is preferred
Lateral canal Dehiscence

Fistula sign positive. Giddiness and nystagmus can be elicited by applying alternating pressure by pressing tragus.

Caused by middle ear disease like cholesteatoma / chronic otitis media with granulation.

Commonly associated with disruption of middle ear conduction mechanism.

If associated with middle ear pathology then deafness is mixed.

Difficult to categorically say the amount of conductive deafness caused by this third window.

Worst case scenario – Dead labyrinth.

Management

Cholesteatoma clearance by performing mastoidectomy.

Fistula of lateral canal is exposed and is closed with temporalis fascia graft.
Enlarged Vestibular aqueduct Syndrome

Clinical features

1. Clinical presentation mimics those of middle and inner ear disorders such as otosclerosis and endolymphatic hydrops
2. Mixed / S/N hearing loss is seen in majority of these patients
3. Aural fullness and tinnitus is observed
4. Hearing loss could be fluctuating ranging from mild to profound
5. Episodic vertigo has been observed
6. Unsteadiness / in co-ordination

In adults normal vestibular duct has a diameter of 0.4-1mm with a mean value of 0.62 mm

EVA syndrome is the most common congenital inner ear malformation

EVA syndrome is characterized by vestibular aqueduct with an AP diameter of 1.5 mm or more. Ideally it is measured halfway between the common crus and the operculum
EVA Syndrome Diagnosis

**MR Imaging**
- T2 weighted images allow visualization of membranous labyrinth
- This is the only imaging technique that visualizes the extraosseous portion of membranous labyrinth

**Management**
- No treatment has been successful in halting the progression of the disease.
- Cochlear implantation can be resorted to in the event of profound sensorineural hearing loss.
- Intratympanic corticosteroid injections have been proposed. This has an added advantage of producing local therapeutic concentrations of the drug without significant side effects. The anti-inflammatory effects of steroid has been postulated to play a role in alleviating the symptoms.
DFN-3 (X-Linked Deafness with stapes gusher)

**Clinical Features**
- Conductive deafness is caused by fixation of foot plate seen in these patients
  - 1. Mixed hearing loss
  - 2. Occurrence of perilymph gush while foot plate is fenestered for stapedectomy
  - 3. Stapedeal reflex is preserved
  - 4. Air-bone gap is greater in lower frequencies
  - 5. Some patients have congenital fixation of foot plate of stapes

**Radiology**
- 1. Radiology demonstrates dilatation of the internal auditory canal
- 2. There is deficiency of bone between internal auditory canal and cochlea
- 3. There may be deficiency of bone between internal auditory canal and the vestibule

**Management**
- 1. Conservative management directed at alleviating the troubling symptoms
- 2. If stapes is fixed then stapedectomy may be resorted to. There is risk of perilymph gush
- 3. Hearing aids may be prescribed to improve hearing

In patients with mobile stapes conductive deafness is caused by abnormal communication between internal auditory canal and the inner ear (either scala vestibuli of cochlea or the vestibule)
Dehiscence between the Cochlea and Carotid Canal

History

Kim & Wilson described a patient with air-bone gap that persisted even after successful stapedectomy. On further examination a communication between cochlea and carotid canal was identified. This communication dissipated acoustic energy away from cochlear partition.

Symptoms & signs are more or less similar to other lesions producing third window in the otic capsule

VEMPS are normal and preserved.

Imaging

CT images reveal a dehiscence on the Scala vestibuli side of basilar membrane. The presence of a large dehiscence in scala vestibuli can decrease cochlear input impedance and reduce sound pressure within scala vestibuli produced by air conducted sound.

Cochlear implant may be resorted to in the presence of severe to profound deafness.
Karlberg et al. described a patient with predominantly low frequency conductive hearing loss characterized by supranormal bone conduction thresholds, presence of acoustic reflexes and presence of air conducted VEMPs despite the presence of conductive hearing loss.

Radiology showed Mondini – like deformity of the cochlea. Modiolus was deficient and a communication was present between the basal turn of cochlea and the internal auditory canal. This abnormal communication caused conductive hearing loss.

Cochlear implant may be resorted to in the presence of severe to profound sensorineural hearing loss.

Apert’s syndrome:

Patients with this syndrome showed mixed hearing loss with an air bone gap of 20-60 dB. Exploratory tympanotomy showed no evidence of middle ear pathology.

CT images demonstrated a dilated vestibule, slight dilatation of the internal auditory canal and enlargement of lateral semicircular canal. Conductive deafness in these patients have been attributed to the presence of third window.
Paget’s Disease

Paget’s disease of otic capsule classically presents with mixed hearing loss with an air-bone gap in the lower frequencies. Multiple microfractures within the otic capsule on the Scala vestibuli side of cochlear partition is responsible for the formation of third window.

These microfractures present in the otic capsule act as a distributed third window dissipating energy transmitted through the stapes foot plate away from the cochlea.

Management:

This is usually conservative. In the presence of severe to profound deafness cochlear implant could be considered.

For mild and moderate deafness hearing aid can be prescribed.
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<thead>
<tr>
<th>Lesion</th>
<th>Middle Ear</th>
<th>Third Window</th>
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<tbody>
<tr>
<td>Air Bone Gap</td>
<td>0-60 dB involving all frequencies</td>
<td>0-60 dB involving lower frequencies</td>
</tr>
<tr>
<td>Bone conduction Tresholds</td>
<td>Rarely &lt; 0 dB</td>
<td>May be negative</td>
</tr>
<tr>
<td>Acoustic reflex</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>VEMP</td>
<td>Absent</td>
<td>Present thresholds lower than normal</td>
</tr>
<tr>
<td>OAE</td>
<td>Absent</td>
<td>Present</td>
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<tr>
<td>Umbo velocity on laser doppler vibrometry</td>
<td>Variable</td>
<td>High Normal</td>
</tr>
<tr>
<td>Sound / pressure induced vertigo</td>
<td>Absent</td>
<td>May be present</td>
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<tr>
<td>CT / MRI</td>
<td>Shows ME abnormality</td>
<td>Shows inner ear abnormalities</td>
</tr>
<tr>
<td>Exploratory Tympanotomy</td>
<td>May show ME abnormality</td>
<td>No ME abnormality will be evident</td>
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