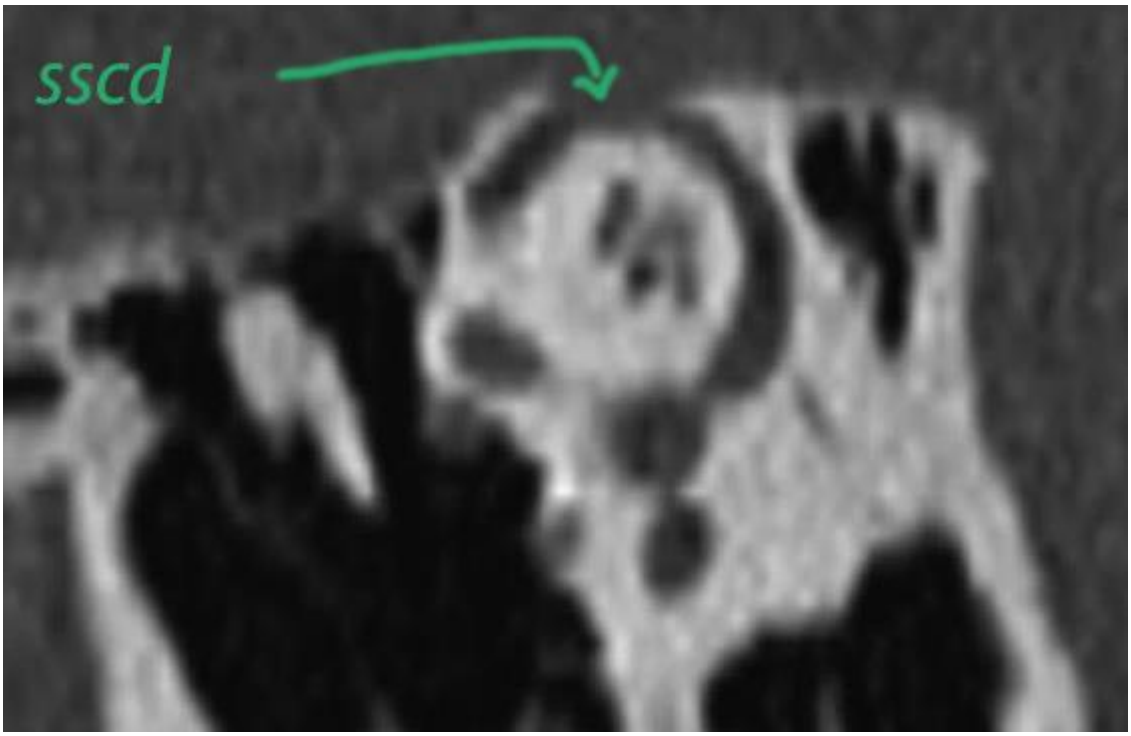
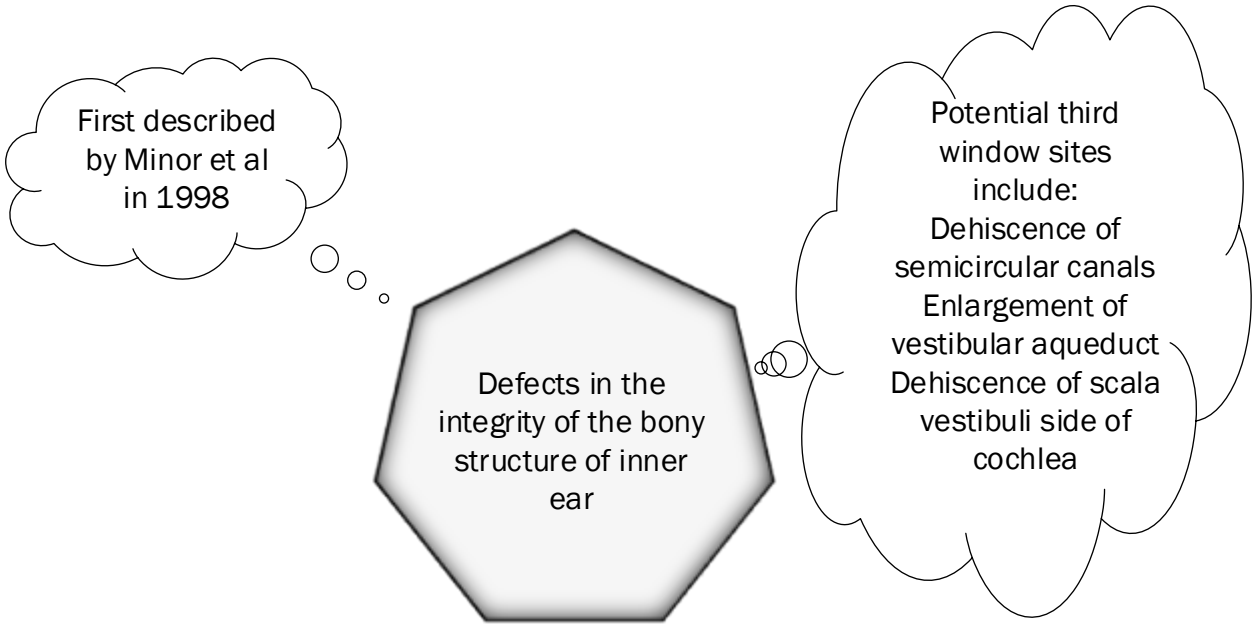
Third Window Abnormalities a spectrum of disorders

Prof Dr Balasubramanian Thiagarajan (drtbalu)



Introduction



Mechanism of hearing loss

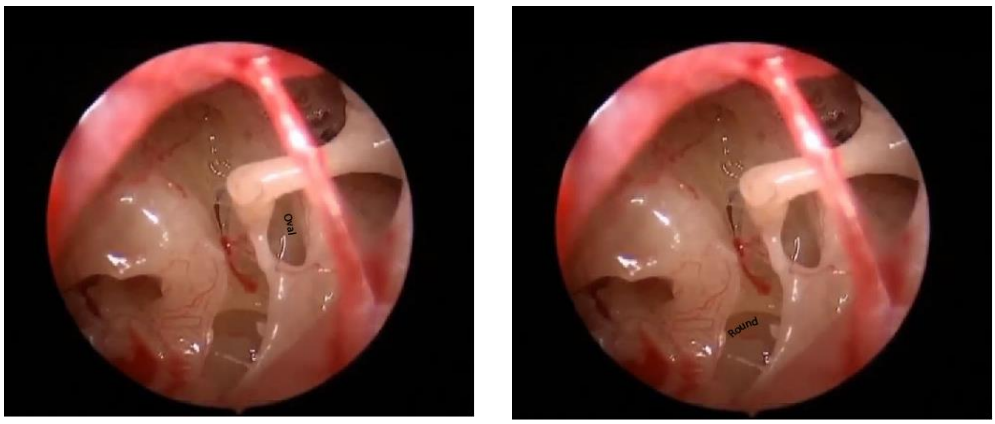
1. Merchant & Rosowski proposed a universal theory for the underlying mechanism of hearing loss in these patients
2. Normal sound conduction is transmitted through the oval and round windows which serve as fluid interfaces between the middle ear and perilymphatic spaces of inner ear. Presence of dehiscence / third window in the labyrinthine bone will cause disruption of this sound conduction mechanism leading on to deafness

Third Window

What is it ?

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.

Oval window is the primary window

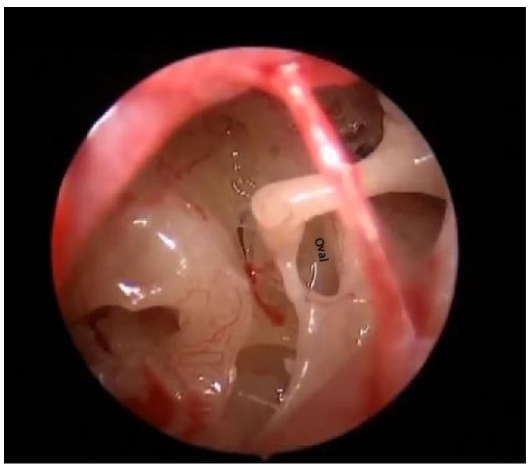


Round window is the secondary window

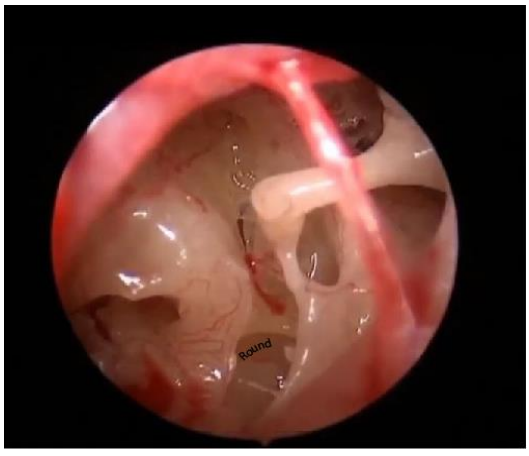
Physiology

Physiology

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

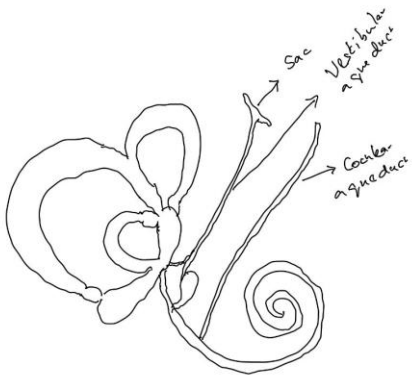
Presence of third windows enabled normal bone conducted hearing even when both oval & round windows are closed

Under normal conditions they are functionally closed

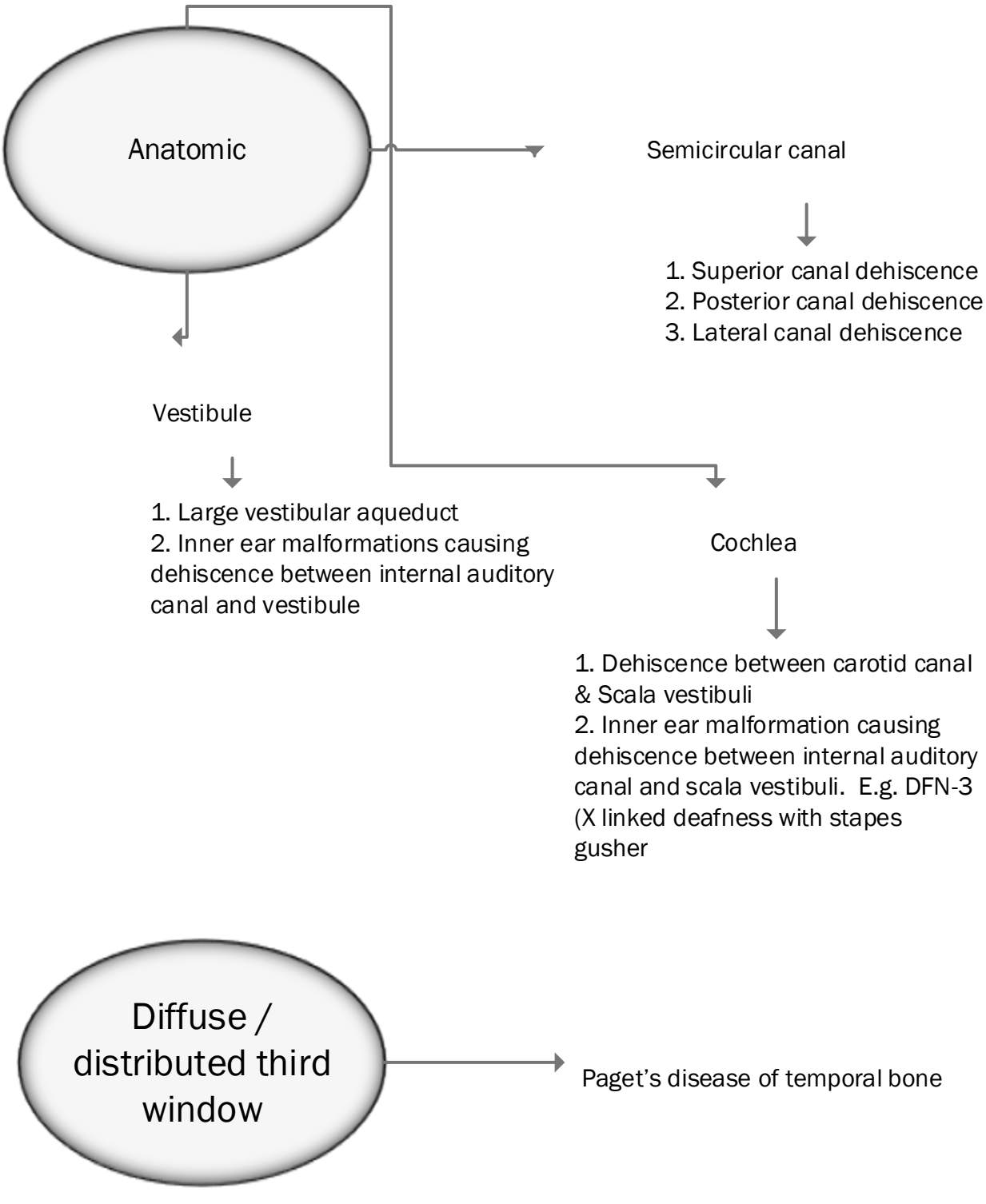
Third window

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

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



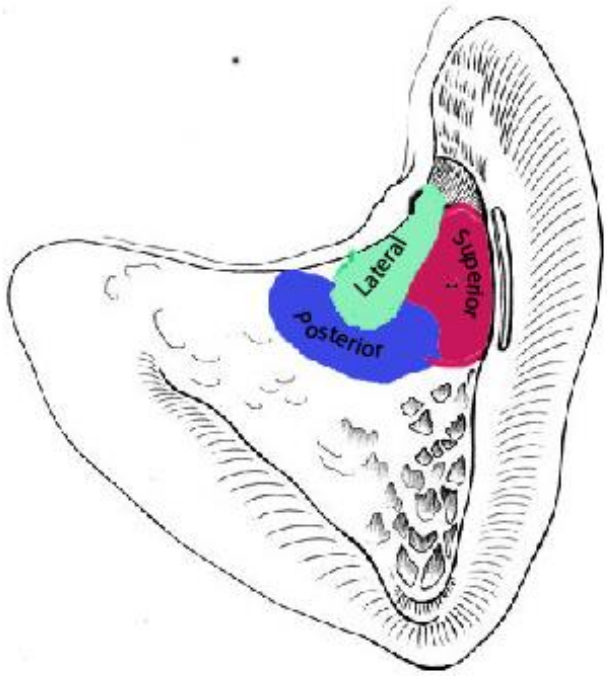
Pathologic Third windows



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

There is a potential communication between the canal and cranial cavity

SSD is characterized by loss of bone covering the superior canal

There is lack of middle ear pathology associated with this lesion

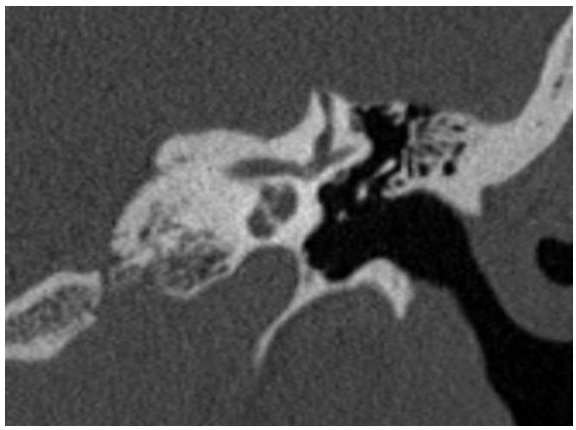
This is the best documented and most investigated 3rd window lesion

Minor etal. Described this condition in 1998

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

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

Nystagmus in the plane of superior canal

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

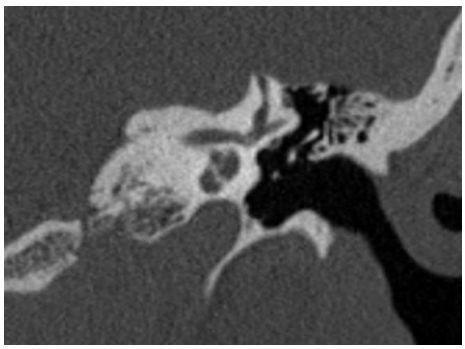
Ear drum appears normal with no evidence of middle ear pathology

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

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

HRCT 1 mm cuts improves diagnostic accuracy

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

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

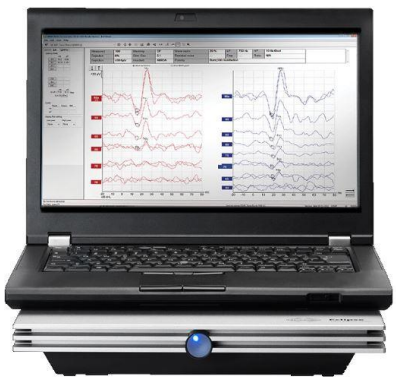
Bone conduction threshold in low frequencies is negative or better than normal

VEMP

Vestibular evoked myogenic potentials

This test in a non-dehiscent ear is thought to reflect function of the saccule (cervical VEMP) or utricle (ocular VEMP).

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



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

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

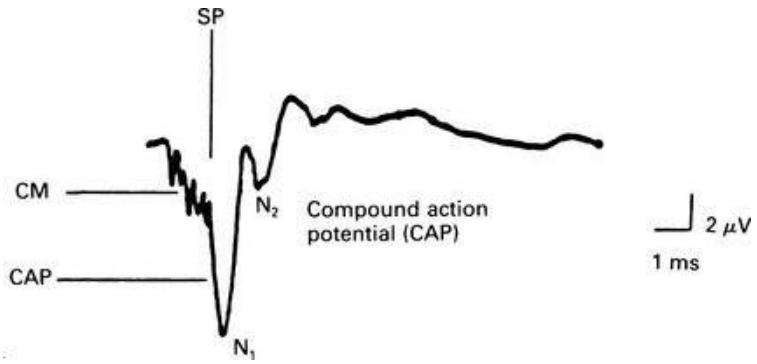
SSCD Syndrome

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

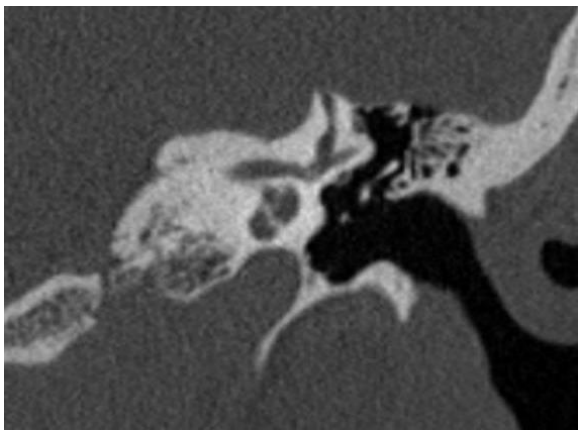
Patients with SSCD syndrome have elevations in the summating potential to action potential ratio

Was used to diagnose Endolymphatic hydrops



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

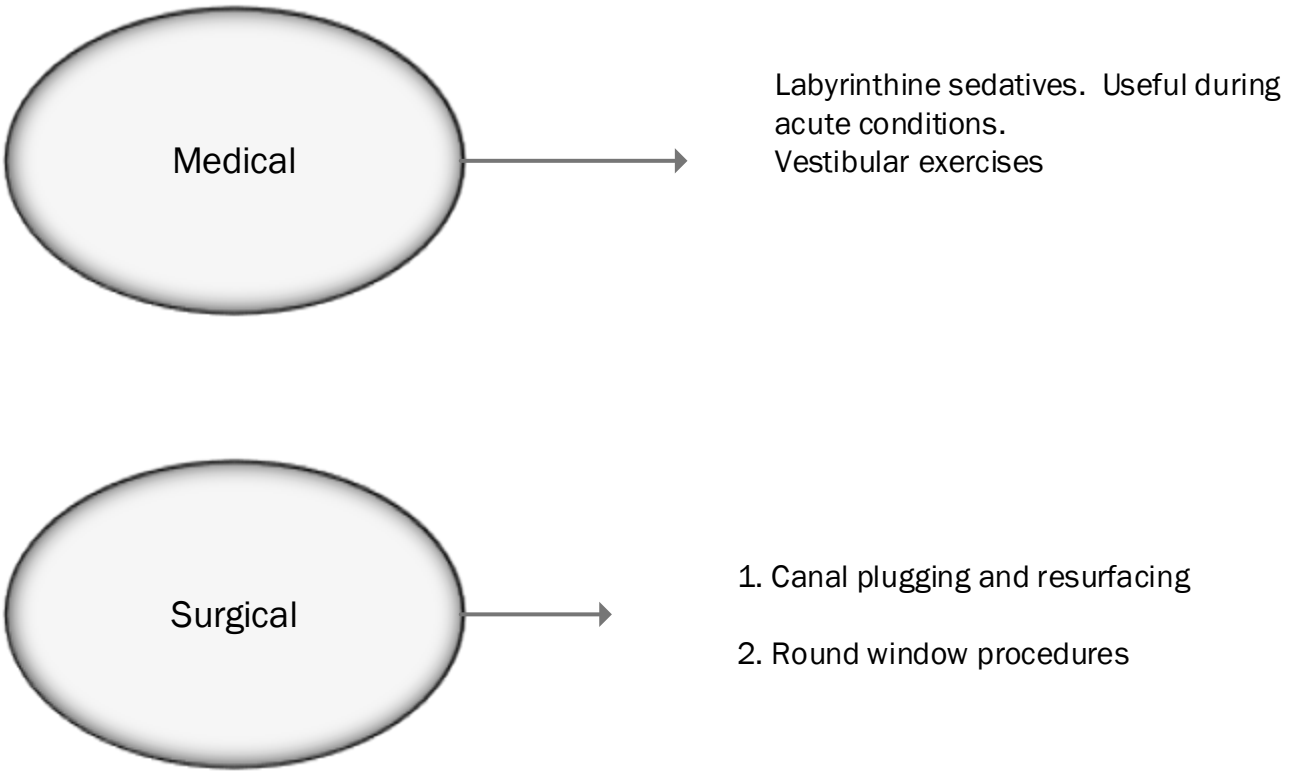
At least one of the following symptoms

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



Canal plugging & Resurfacing

Indicated only in patients with debilitating symptoms

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

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

Resurfacing of dehiscent canal prevents chronic stimulation from pulsating temporal lobe

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

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

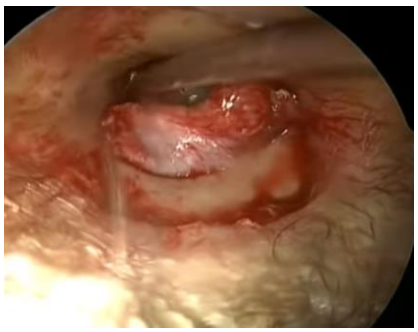
Round window Reinforcement

Round window Reinforcement

TM flap is elevated

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

Can be done under LA / GA



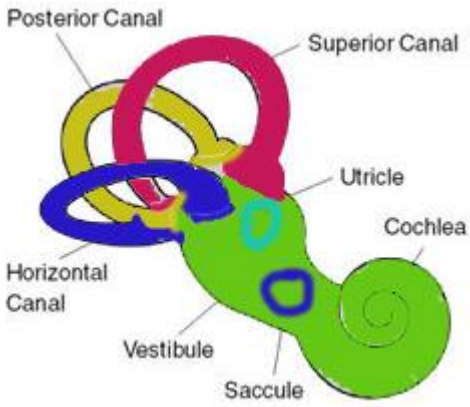
Round window niche and promontory are denuded of mucosa and the round window is reinforced with temporalis fascia / tragal cartilage/ fat / connective tissue

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 radiographicall y in 1986 by Bony

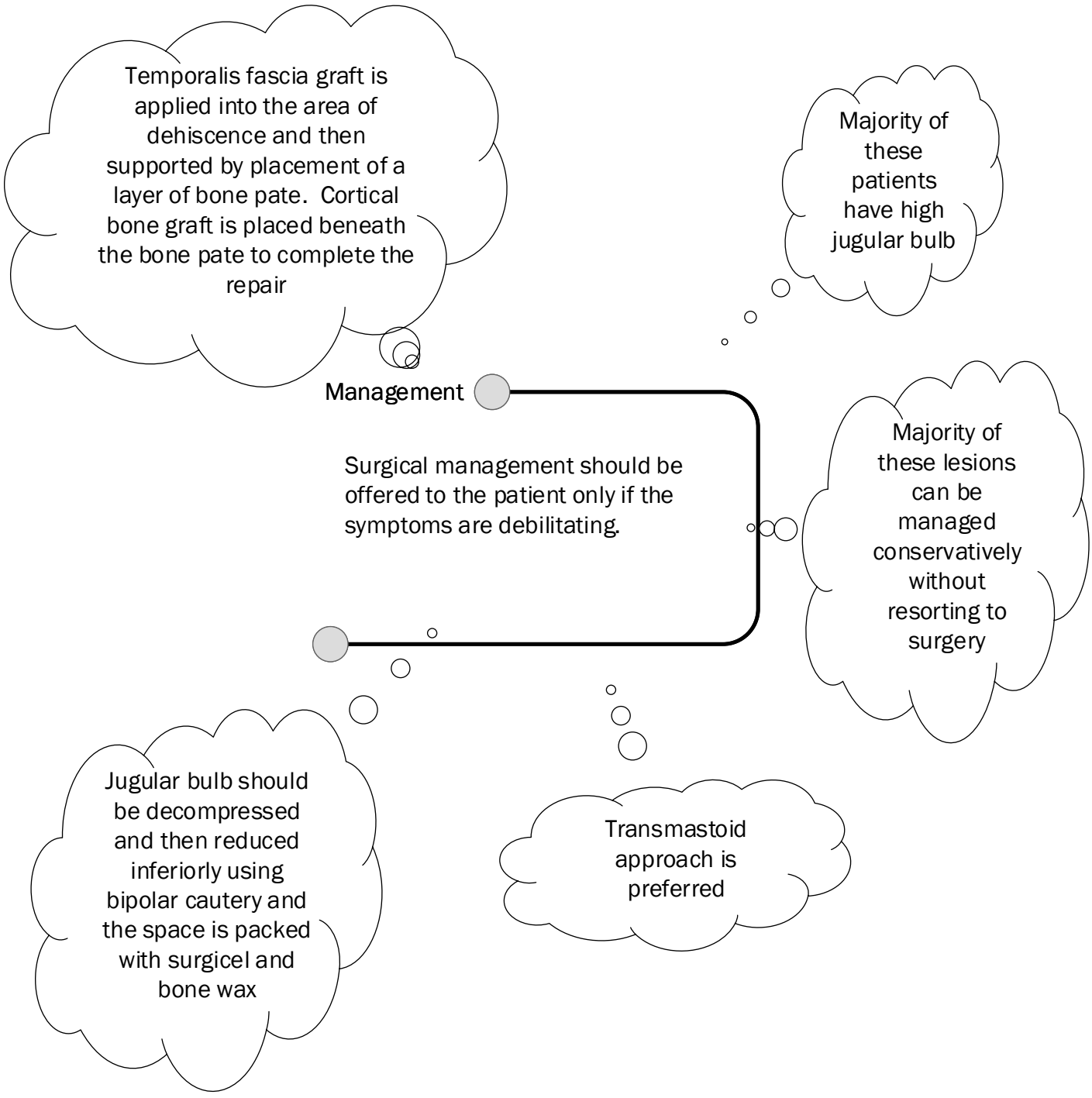


VEMP is usually diagnostic

Nystagmus is vertical and beats in opposite directions

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

PSCCD Syndrome Management

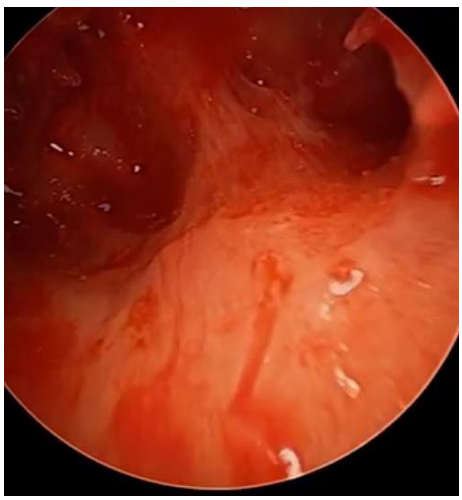


Lateral canal Dehiscence

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

Commonly associated with disruption of middle ear conduction mechanism

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



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

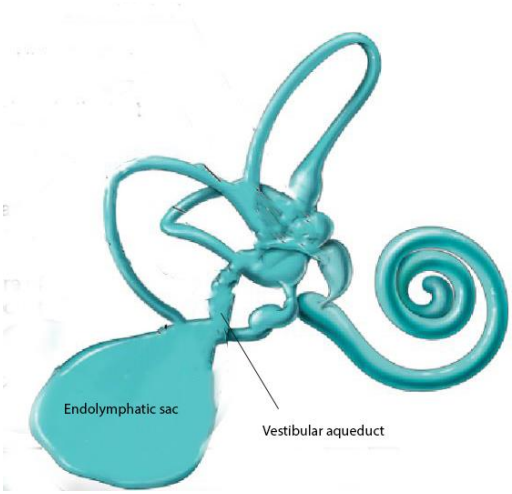
Cholesteatoma clearance by performing mastoidectomy.

Fistula of lateral canal is exposed and is closed with temporalis fascia graft

Enlarged Vestibular aqueduct Syndrome

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

EVA syndrome is the mot 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

Clinical features

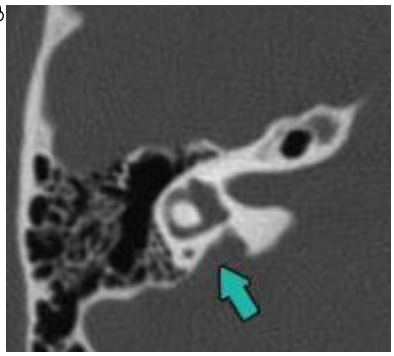
1. Clinical presentation mimics those of middle and inner ear disorders such as otosclerosis and endolymphatic hydrops
2. Mixed / S/N hearing loss is seen in majority of these patients
3. Aural fullness and tinnitus is observed
4. Hearing loss could be fluctuating ranging from mild to profound
5. Episodic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

EVA Syndrome Diagnosis

Axial CT temporal bone 1.5 mm cuts show bony labyrinth anatomy.

Vestibular aqueduct is seen over the posterior surface of petrous bone in imaging

Diagnosis is radiological



MR Imaging

1. T2 weighted images allows visualization of membranous labyrinth
2. 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

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

Conductive deafness is caused by fixation of foot plate seen in these patients

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

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

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.

Inner Ear Malformations

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.

Differentiating Middle ear and Third window lesions		
Lesion	Middle Ear	Third Window
Air Bone Gap	0-60 dB involving all frequencies	0-60 dB involving lower frequences
Bone conduction Tresholds	Rarely < 0 dB	May be negative
Acoustic reflex	Absent	Present
VEMP	Absent	Present thresholds lower than normal
OAE	Absent	Present
Umbo velocity on laser doppler vibrometry	Variable	High Normal
Sound /pressure induced vertigo	Absent	May be present
CT / MRI	Shows ME abnormality	Shows inner ear abrnomalities
Exploratory Tympanotomy	May show ME abnormality	No ME abrnormality will be evident