Syndrome of the Third Mobile Windows

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Third mobile window

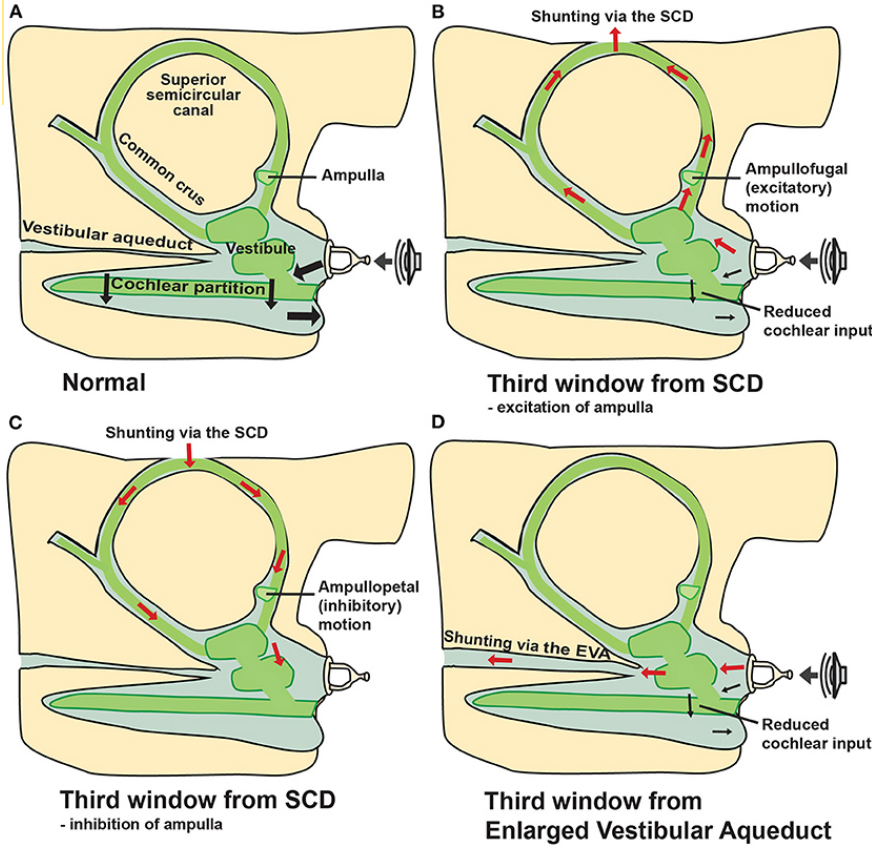
- The leading symptom of these rare diseases is recurrent attacks of vertigo which may be associated with oscillopsia and which are induced by changes in **pressure or sounds**.
- Many patients can hear the sounds of their own body louder in the affected ear (**autophony**) or external sounds louder.
- These symptoms are caused by a pathological transduction of pressure to the inner ear due to a bony defect, which leads to a "**third mobile window**."
- The most frequent subtypes are canal dehiscence syndromes such as the **superior semicircular canal dehiscence syndrome (SCDS)**
- The posterior and the horizontal canal can also be affected; very few patients have bilateral SCDS.
- Similar symptoms can also occur due to an **enlarged vestibular duct**. A very rare entity is "**perilymphatic fistula**"

Superior Semicircular Canal Dehiscence Syndrome

01

Pathophysiology and Etiology

- Symptoms and signs are caused by a pathological transduction of pressure via a third window (in addition to the round and oval windows) .
- There is a transduction of pressure from the oval window to the inner ear and a conduction via the round window. The third window changes the biomechanics of the inner ear considerably.
- SCD **facilitates the flow of sound volume velocity** through the **cochlear partition at low frequencies**, resulting in bone hyperacusis which can also explain autophonia as well as the **increased amplitudes of the oVEMP** and **reduced cVEMP thresholds** (Ward et al. 2017).
- The precise etiology is still unclear. It is assumed that there is a **primary congenital delay in bone growth above the superior canal** (Carey et al. 2000) **destructing osteoclastic processes** (Kamakura and Nadol Jr. 2017).
- Trauma, barotrauma, or changes of pressure after lifting heavy weights, coughing, or pressing, which may lead to a rupture of the bone due to increased intracranial pressure.



"Third window" mechanism due to SCD and enlarged vestibular aqueduct.

(A) Normal anatomy allows volume velocity across the cochlear partition from the oval to the round window (two windows).

(B) Air conducted sound stimulation results in volume velocity from the stapes to be shunted toward the SCD (third window) and away from the cochlea, resulting **in increased air-conduction thresholds at low frequencies** and/or sound-induced vertigo **(Tullio's phenomenon)**. Positive static pressure in the middle-ear cavity may result in ampullofugal fluid motion exciting the ampulla, resulting in nystagmus **(Hennebert sign)** and oscillopsia/vertigo

(C) Elevated intracranial pressure from **Valsalva** against closed glottis (e.g., straining, heaving lifting) may result in ampullopetal endolymphatic fluid motion, inhibition of the ampulla, also leading to nystagmus (Hennebert sign) and oscillopsia/vertigo

(D) Enlarged vestibular aqueduct (EVA) can also act as a third window, shunting volume velocity away from the cochlear partition and toward the widened vestibular aqueduct .

Eberhard, Kristine Elisabeth, et al. "Current trends, controversies, and future directions in the evaluation and management of superior canal dehiscence syndrome." Frontiers in Neurology 12 (2021): 638574

Box 12.1 Diagnostic Criteria for Superior Canal Dehiscence Syndrome (SCDS)

The diagnosis of superior semicircular canal dehiscence syndrome requires all of the following criteria:

1. At least one of the following symptoms consistent with a third window lesion of the inner ear:
    1. Bone conduction hyperacusis
    2. Sound-induced vertigo and/or oscillopsia time-locked to the stimulus
    3. Pressure-induced vertigo and/or oscillopsia time-locked to the stimulus
    4. Pulsatile tinnitus
2. At least 1 of the following signs or diagnostic tests indicating a "third mobile window" of the inner ear:
    1. Nystagmus characteristic of excitation or inhibition of the affected superior canal evoked by sound, or by changes in middle ear pressure or intracranial pressure
    2. Low-frequency negative bone conduction thresholds on pure tone audiometry
    3. Enhanced VEMP responses (low cervical VEMP thresholds or high ocular VEMP amplitudes)
3. High-resolution temporal bone CT imaging with multiplanar reconstruction demonstrating dehiscence of the superior canal
4. Not better accounted for by another vestibular disease or disorder

Adapted from (Ward et al. 2021)

Diagnosis - Patient History

- Attacks, lasting **seconds to minutes**, of spinning or non-spinning vertigo induced by changes in pressure (sneezing, coughing, pressing, lifting heavy weights), and/or sounds most often of a low frequency or fremitus
- These attacks can be associated with **oscillopsia, postural imbalance, pulsatile tinnitus, pulsatile oscillopsia, movement-induced postural imbalance and gait disorder and recurrent positional vertigo**
- **Autophony** (pulse, swallowing, speaking, eye movements, blinking, and even bowel movements) can be heard in the affected ear.
- Some patients can hear **external sounds also louder**, namely in the low frequency range which can lead to a severe impairment.
- Other potential inducing factors are: significant changes in altitude, changes in pressure when entering or leaving a tunnel, changes in body position, or pressing a finger on the external auditory canal of the affected ear.
- Traumas Hx (barotrauma, head trauma, ear trauma, operations of the ear, Valsalva maneuvers due to lifting of heavy weights)

Diagnosis - Physical Examination

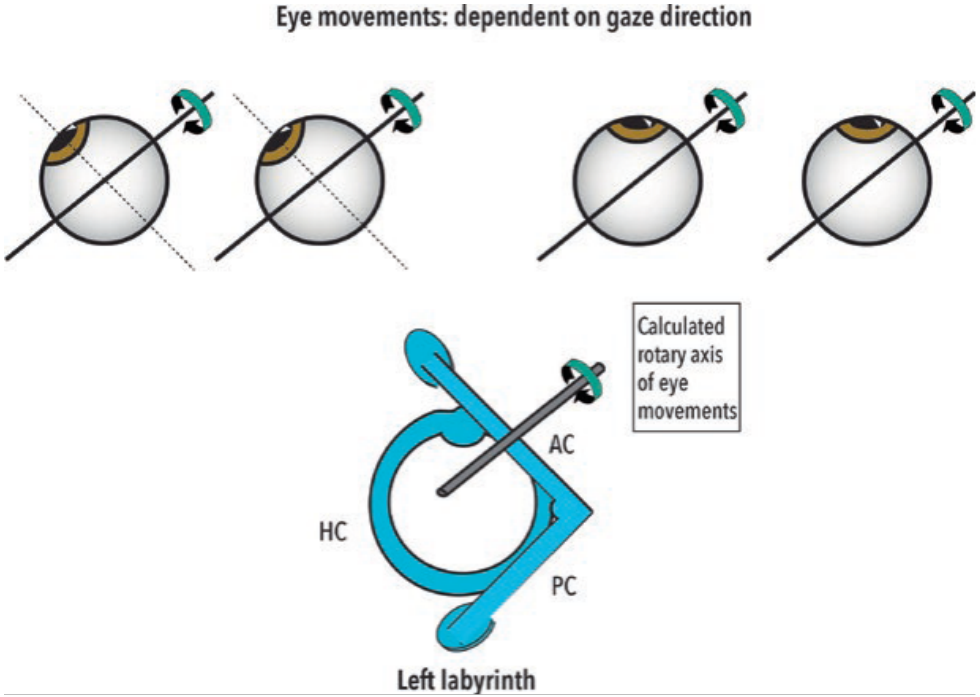
- ***Changes in pressure* via *the external auditory canal***
  1. By the Politzer balloon or pressure on the tragus (Hennebert's sign); the increase in pressure should persist for **at least 30 seconds**. Positive pressure from the outside leads – via the oval window – to an excitation of the affected superior canal due to an **ampullofugal** movement of the cupula.
  2. Nystagmus can occur during the increase and/or the subsequent decrease in pressure.
  3. a **vertical torsional nystagmus** in the plane of the superior canal
- **Valsalva maneuver**
  1. Increase of intracranial pressure by abdominal pressing against the closed vocal cord. Here also one can observe a **vertical torsional nystagmus**
  2. This maneuver leads to increased pressure in the direction of the oval window with an inhibition of the superior semicircular canal (**ampullopetal** deviation of the cupula). Pressing against the closed nostril leads to a combined increase of intracranial and middle ear pressure, which can also induce nystagmus.

Diagnosis - Clinical Testing of Hearing and ENT Examination

1. Weber test (512 Hz), the sound is typically louder on the affected side, which correlates with autophony.
2. The Rinne test can provide evidence of impaired bone conduction on the affected side in the low-frequency range.
3. When vibratory sensation is tested with a tuning fork on the extremities or the sternum, patients can hear the vibration in the affected ear, which allows easy identification of the affected side
4. Tympanoscopy can provide evidence of a hypermobile stapes.

Diagnosis - Vestibular Laboratory Testing

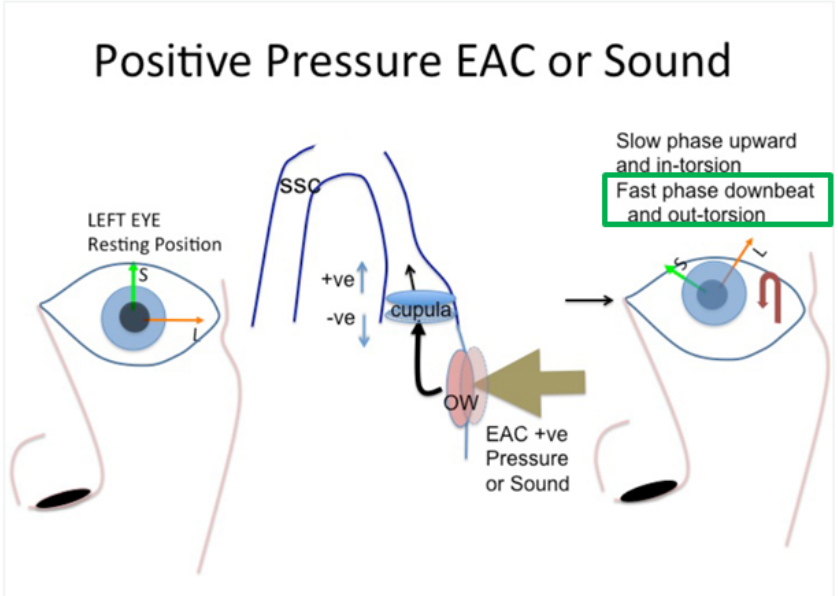
- **Videooculography :** In patients with SCDS, it is helpful for the patient to look in the direction of the plane of the superior canal (e.g., **45°** to the left if one expects left-sided SCDS) during such a recording, because a **pure vertical nystagmus** will be induced, which supports the diagnosis
- **Testing for Tullio phenomenon**: using an audiometer Sounds of certain frequencies can induce vertigo and nystagmus in the plane of the affected canal, which should ideally be recorded by videooculography.

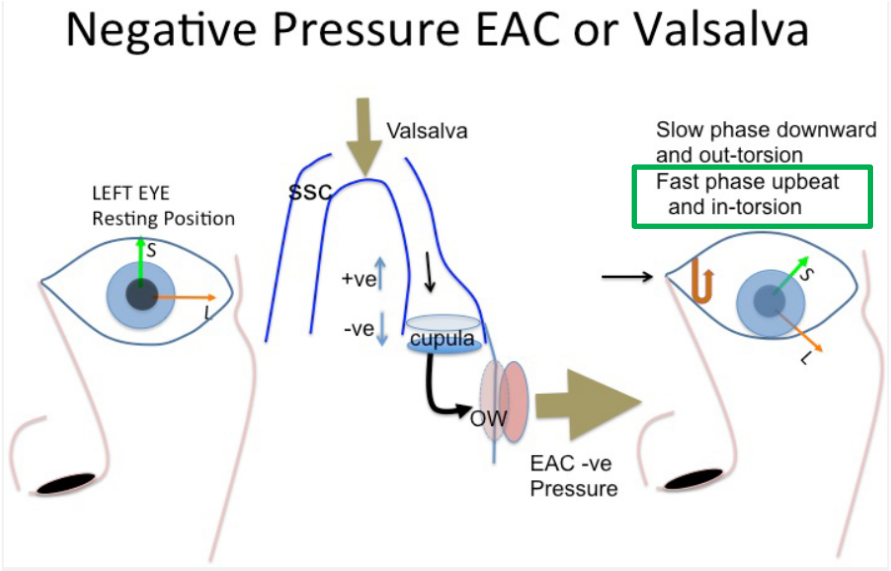


Vestibular-evoked

Dependence of the direction of the nystagmus on the gaze direction in a patient with left-sided SCDS. If the patient looks 45° to the left, there will be a pure vertical nystagmus (top left). If the patient looks straight ahead (top right), there will be a vertical torsional nystagmus

Left SCD nystagmus

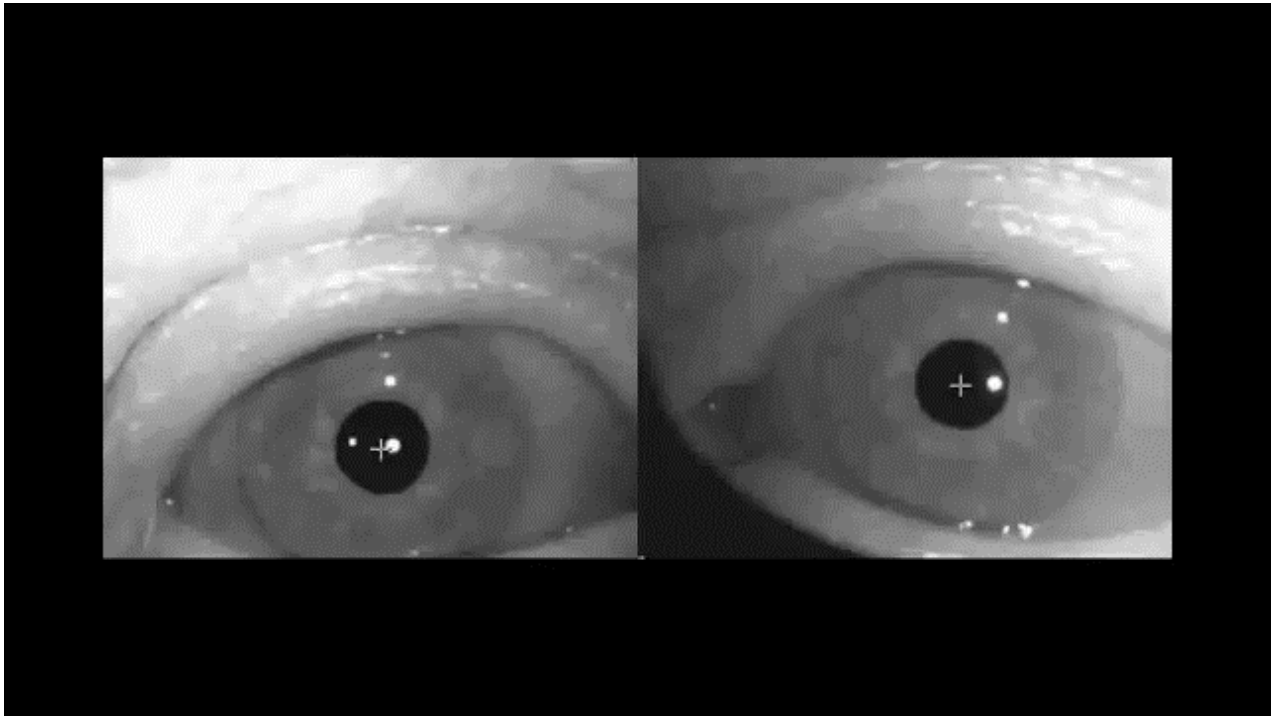




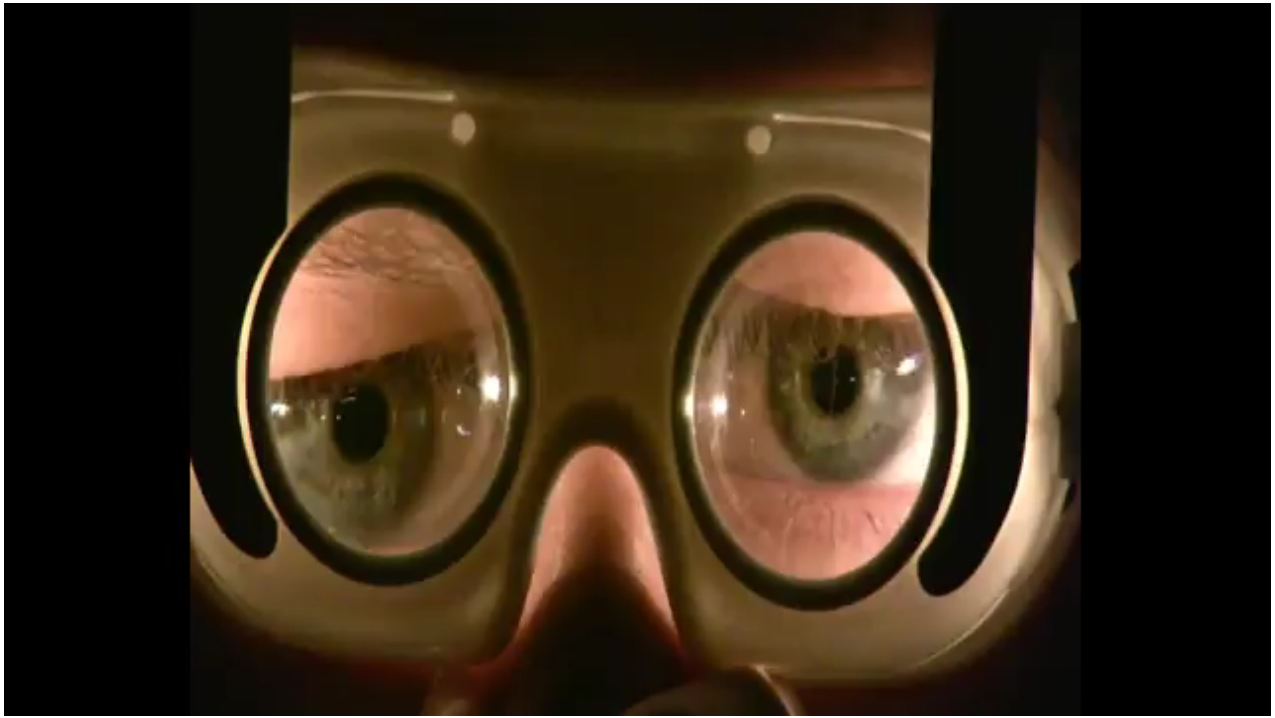
This stimulates the superior semicircular canal, and so would cause an in-torsion and upward slow phase eye movement from Ewald's first law which states that the **direction of the elicited eye movement is the plane of the stimulated canal.**

https://canadianaudiologist.ca/superior-semicircular-canal-dehiscence/

Pressure induced nystagmus (Hennebert's sign) in left sided SCDS



Vertical torsional nystagmus in a patient with Lt SCD syndrome during the Valsalva maneuver



Diagnosis - Vestibular Laboratory Testing

Vestibular-evoked potentials (VEMP): VEMP can considerably contribute to the diagnosis of SCDS because the amplitudes are typically increased on the affected side.

- **ocular VEMP (oVEMP):** the optimal stimulation frequency is **4 kHz** (Tran et al. 2020).

1. Another study showed that if the **amplitude of the oVEMP is higher than 16.7 µV**, the sensitivity is 100% and the specificity is 89% (Verrecchia et al. 2019).
2. oVEMP can also be elevated in patients without SCDS, namely in Menière's disease which underlines that for the diagnosis several consistent criteria have to be fullfilled.

- **cervical VEMP (cVEMP)** : the best stimulation frequency is 2 kHz; under these conditions a specificity of almost 100% and a sensitivity of 92% was reported.

Since **oVEMP** are easier to perform and **do not depend on threshold measurements**, they are recommended for clinical routine. In addition, a review of published papers showed that they are evidently more sensitive and more specific than cVEMP (Fife et al. 2017).

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Vestibular Evoked Myogenic Potential (VEMP) Testing: Normative Threshold Response Curves and Effects of Age

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Table 2

Normative Mean Values (across all ages for right and left ears combined). All parameters are reported at threshold with the exception of 500 Hz toneburst maximum presentation level (123 dBSPL).

	Click	250 Hz	500 Hz	750 Hz	1000 Hz	500 Hz MAX
			THRESHOLD			
OVERALL	122.17 (4.09)	116.87 (6.45)	114.16 (6.45)	115.75 (5.53)	116.65 (5.2)	NA
20–29	123 (2.58)	116.58 (5.01)	113.25 (3.35)	113.75 (3.58)	115.5 (3.2)	NA
30–39	121.25 (4.43)	114.58 (7.22)	110.31 (6.45)	112.5 (4.47)	114.69 (4.64)	NA
40–49	119.17 (5.85)	114.69 (6.94)	111.84 (6.08)	116 (7.36)	116.05 (6.36)	NA
50–59	125	119.38 (5.63)	117.81 (5.15)	117.81 (4.46)	119.06 (5.83)	NA
60+	125	120.83 (5.97)	117.78 (7.52)	119.33 (4.17)	118.57 (4.57)	NA
			AMPLITUDE			
OVERALL	27.17 (9.13)	27.86 (11.61)	27.65 (11.13)	29.96 (13.36)	29.83 (13.37)	57.34 (34.32)
20–29	27.88 (9.87)	29.59 (9.56)	32.76 (10.92)	35.46 (16.35)	33.57 (15.22)	66.12 (31.49)
30–39	26.71 (8.33)	29.9 (12.63)	25.75 (12)	26.13 (11.8)	24.39 (11.18)	74.54 (40.96)
40–49	30.57 (11.20)	28.15 (11.87)	27.64 (11.32)	27.14 (9.81)	27.02 (8.39)	56.13 (30.21)
50–59	22.1 (7.47)	23.10 (9.25)	20.63 (6.76)	27.42 (15.58)	31.93 (16.77)	42.37 (35.16)
60+	28.69	25.89 (14.92)	29.92 (10.94)	33.19 (10.22)	32.16 (13.05)	50.61 (29.3)
			P13 LATENCY			

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PMID: 30677753

Toward Optimizing cVEMP: 2,000-Hz Tone Bursts Improve the Detection of Superior Canal Dehiscence

Table 1. Sample criterion values and the corresponding sensitivities and specificities for the ROC curves in Figures 4 and 5

	Threshold			TWI			VEMPn (123 dB peSPL)			VEMPid (123 dB peSPL)		
	criterion value, dB peSPL	sensitivity, %	specificity, %	criterion value, dB	sensitivity, %	specificity, %	criterion value (dimensionless)	sensitivity, %	specificity, %	criterion value %	sensitivity, %	specificity, %
500 Hz	98	52.0	100.0	103	88.0	100.0	1.44	68.0	100.0	20.9	68.0	100.0
	103	80.0	97.8	108	96.0	84.8	1.00	84.0	91.3	15.0	84.0	93.5
	108	88.0	93.5	113	96.0	60.9	0.75	92.0	78.3	10.0	92.0	82.6
	118	100.0	43.5	123	100.0	21.7	0.49	100.0	56.5	7.2	100.0	69.6
750 Hz	103	76.0	100.0	93	76.0	100.0	1.31	76.0	100.0	19.0	80.0	100.0
	113	88.0	69.6	103	92.0	91.3	1.00	92.0	93.5	15.0	84.0	95.7
	123	96.0	19.6	113	92.0	58.7	0.75	92.0	78.3	10.0	92.0	84.8
	128	100.0	6.5	133	100.0	2.2	0.51	100.0	63.0	6.8	100.0	63.0
1,000 Hz	103	68.0	100.0	103	92.0	100.0	1.73	60.0	100.0	27.5	64.0	100.0
	108	92.0	97.8	113	96.0	76.1	1.00	92.0	95.7	20.0	68.0	97.8
	118	96.0	58.7	123	96.0	71.7	0.75	96.0	87.0	15.0	84.0	97.8
	123	100.0	39.1	133	100.0	4.3	0.54	100.0	65.2	4.4	100.0	50.0
2,000 Hz	118	*92.0*	*100.0*	108	*92.0*	*100.0*	0.67	*96.0*	*100.0*	9.2	*96.0*	*100.0*
	123	96.0	91.3	118	92.0	97.8	0.50	96.0	95.7	7.0	96.0	97.8
	128	96.0	67.4	128	96.0	63.0	0.40	96.0	82.6	5.0	96.0	93.5
	138	100.0	41.3	138	100.0	26.1	0.28	100.0	82.6	1.9	100.0	60.9

If all cases with a threshold or third window indicator (TWI) below the criterion value or a VEMPn or VEMPid above the criterion value are assumed to indicate the presence of a dehiscence, then the data show that this judgment will have the sensitivity and specificity listed. For example, for cVEMP threshold at 2,000 Hz (lower left in the Table), a 118-dB criterion cutoff value corresponds to a 92% sensitivity and a 100% specificity (italicized in the Table). This means that none of the healthy ears had a 2,000-Hz threshold below 118 dB peSPL (specificity = 100%), while 23 out of 25 ears with SCD met this criterion (sensitivity = 92%).

Figure 3

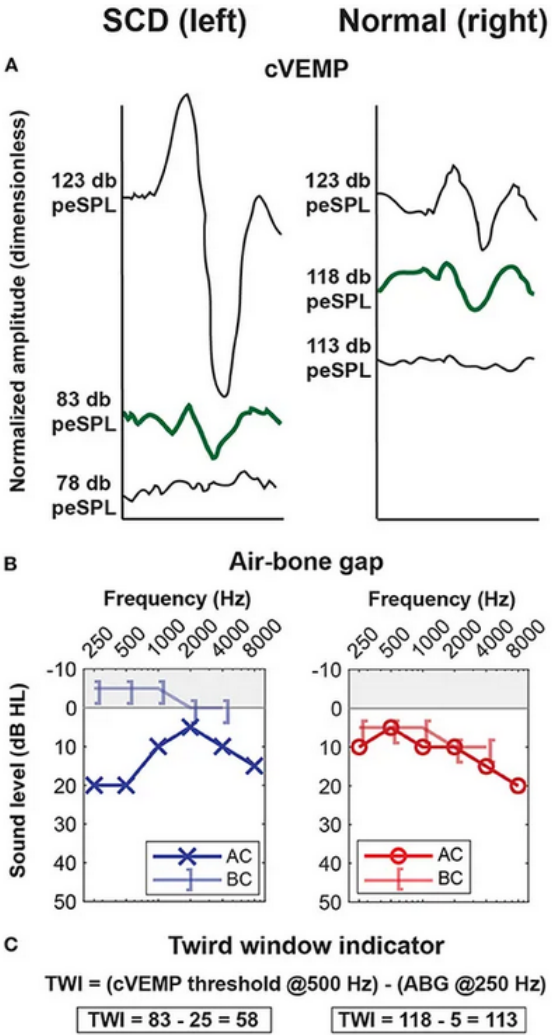
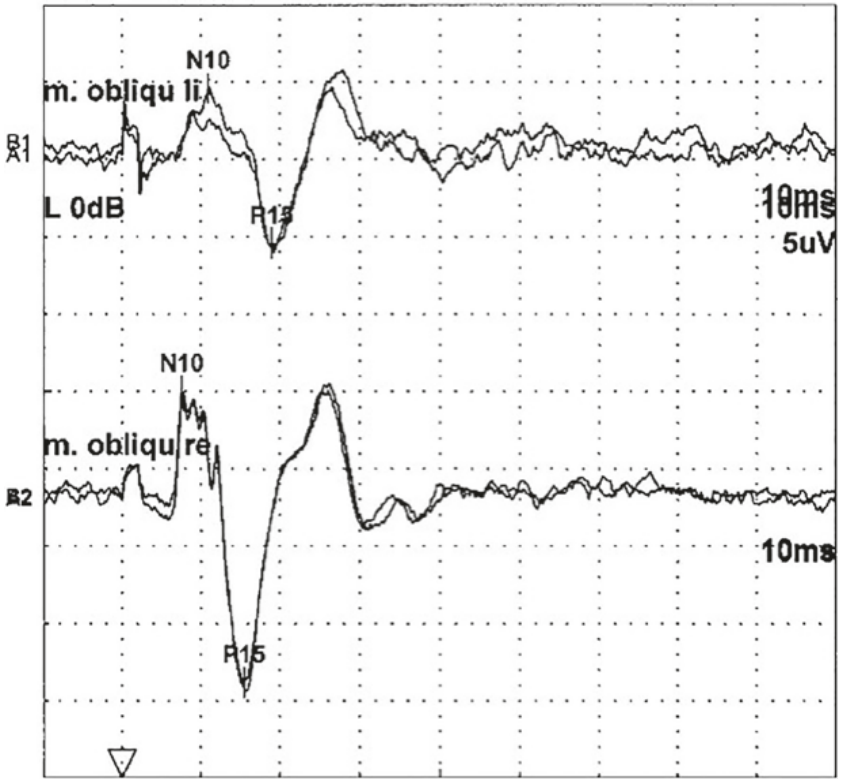


Figure 3. Third Window Indicator (TWI) improves ability to differentiate SCD ears from non-third window ears. Example of an adult patient with symptoms of left-sided SCDS. **(A)** Low threshold cVEMP of 83 dB peSPL [peak sound pressure level, 123 dB peSPL is equivalent to 90 dB HL (65)] at 500 Hz in the left SCDS ear and 118 peSPL in the unaffected right ear. **(B)** Air-bone gap of 25 dB HL at 250 Hz in the left SCDS ear, compared to 5 dB HL in the unaffected right ear. **(C)** The TWI is the difference between the cVEMP threshold at 500 Hz and the air-bone gap at 250 Hz (65). In this case, the TWI is **58** dB for the left SCDS ear, and **113** dB for the unaffected right ear. *Modified from Noij et al. (65).

TMUH

Eberhard, Kristine Elisabeth, et al. "Current trends, controversies, and future directions in the evaluation and management of superior canal dehiscence syndrome." Frontiers in Neurology 12 (2021): 638574.



a Ocular vestibular-evoked myogenic potentials (oVEMP) in a patient with left-sided SCDS. When recording from the right inferior oblique muscle (lower trace) during the stimulation of the left labyrinth, the amplitude of the N10-P15 is 19.8 μV compared to 10.1 μV on the non-affected side (upper trace).

Diagnosis - Audiological Examination

- Patients often have **negative bone conduction in the low-frequency range (≤500 Hz)**, which corresponds to autophony and supports the diagnosis.

Audiogram

Typically, SCD causes a low frequency conductive loss. Most often, the conductive loss is mild, and in the low frequencies only. Figure 6 shows a typical audiogram in SCD. Whether this can ever be large enough to be confused with a true otosclerotic type hearing loss is controversial. In temporal bone experiments, even when the SCD was open to air, losses were only below 1 kHz, and when covered with dura, the losses were minimal.[15]

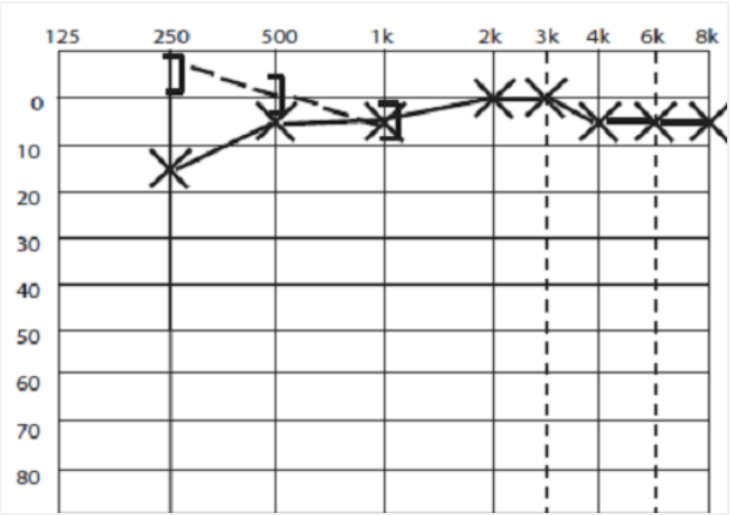
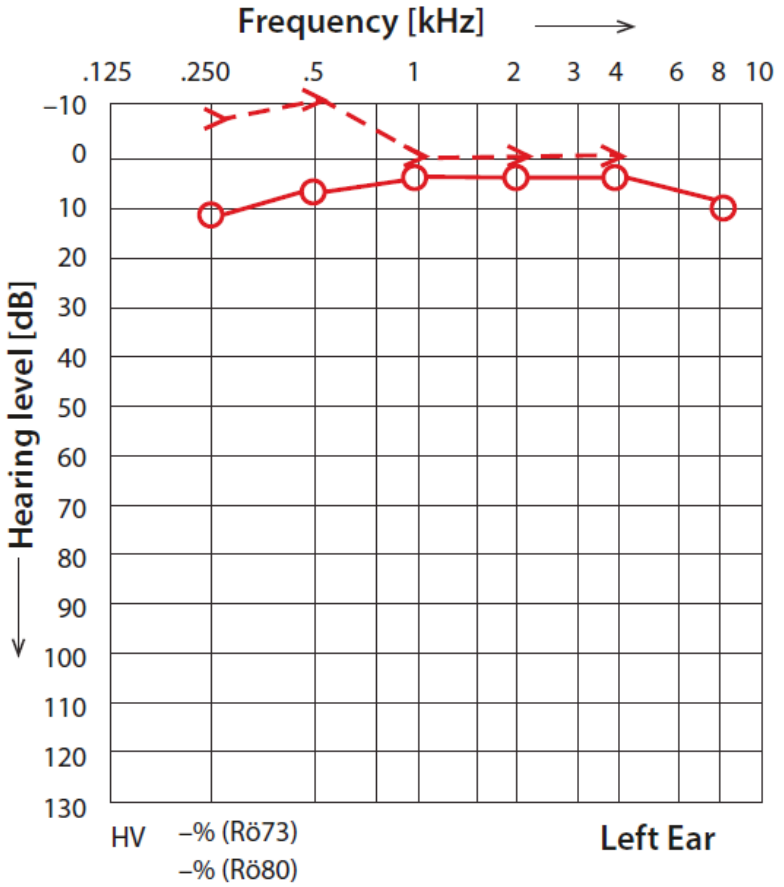


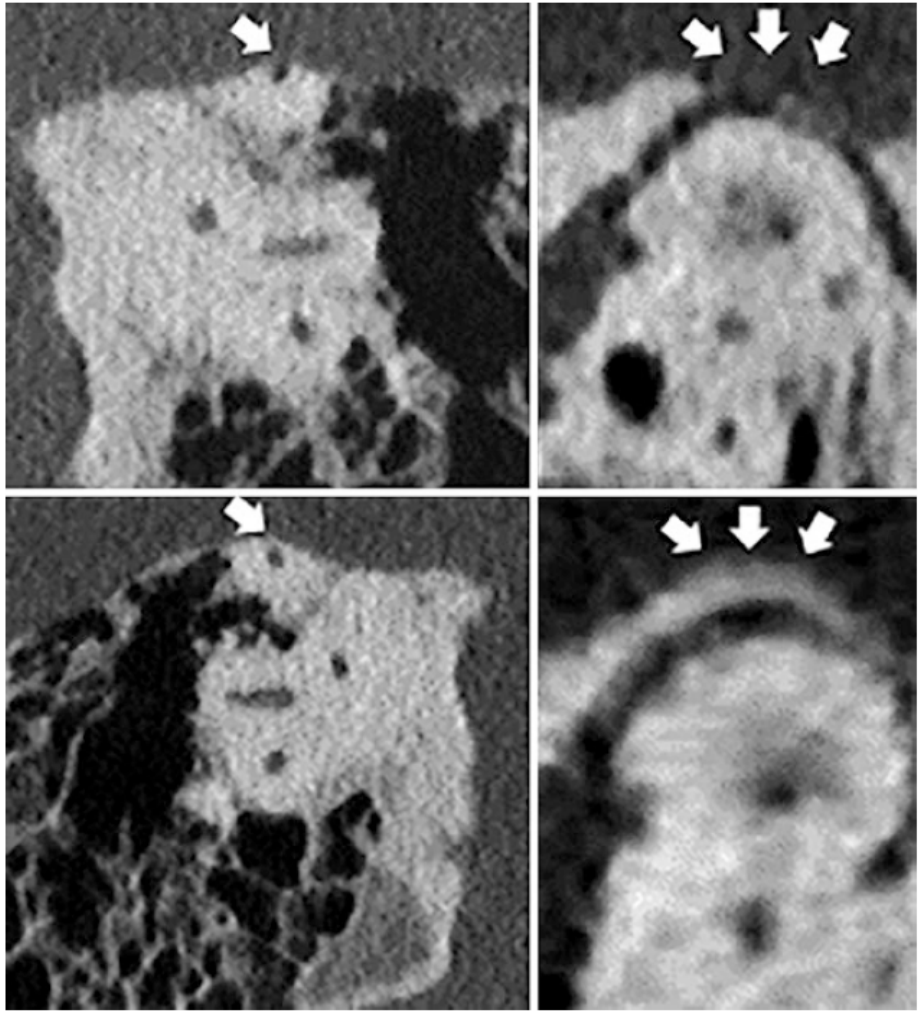
Figure 6, typical audiogram in SCD, showing low frequency conductive hearing loss, and supranormal bone conduction thresholds for bone conduction in the lowest frequencies.

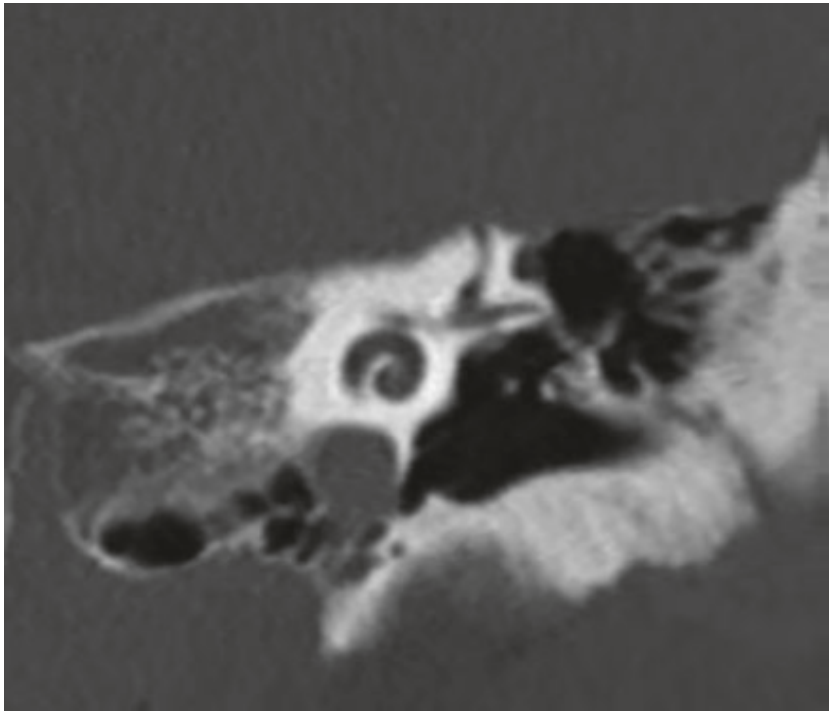


https://canadianaudiologist.ca/superior-semicircular-canal-dehiscence/

Diagnosis - Imaging

- The bony defect on the superior canal can be visualized by a high-resolution temporal bone CT. The resolution must be ≤0.6 mm with 3D reconstruction perpendicular and parallel to the affected semicircular canal
- Healthy subjects can also have a bony defect on a temporal bone CT of the superior canal. [In a study on 500 healthy subjects, this was found on one side in 2% and on both sides in 0.6%; in 110 patients who had audiological and/or vestibular symptoms, 14% had a unilateral and 1.8% a bilateral bony defect without symptoms of SCDS (Berning et al. 2019).]
- Even small punctiform bony defects, which can hardly be visualized by CT, can lead to SCDS. MRI should be used to exclude other pathologies





High-resolution CT of the temporal bone shows the bony defect on top of the superior canal

Syndrome of Different Third Mobile Windows, Differential Diagnoses and Clinical Problems

- **Horizontal canal dehiscence syndrome or posterior semicircular canal dehiscence syndrome :**The symptoms are similar to SCDS. However, analysis of the direction of the induced nystagmus and imaging allow a differentiation between the three subtypes.
- **Enlarged vestibular duct :** can lead to similar symptoms (Stahl and Otteson, 2022), namely autophony and a negative bone conduction. Therefore, this entity must always be considered as a subtype of the syndrome of the third mobile windows which can be seen on CT.
- **Bilateral SCDS** Up to 50% of patients with SCDS anatomically have a very thin roof of the superior canal, without necessarily having bilateral symptoms. Therefore, to make a correct diagnosis of symptomatic bilateral SCDS, not only the imaging, but also the other above-mentioned criteria should be fulfilled for both sides.
- **Secondary syndrome of the third mobile windows** The above-mentioned symptoms and signs can also occur in other inner ear diseases that impair the integrity of the temporal bone, in particular cholesteatomas, meningiomas, or meningoceles and after head trauma. Patients with Menière's disease can also develop a secondary syndrome of third mobile window.
- **Near dehiscence syndrome (N-SCDS)** (i.e., roof of the superior semicircular canal thin, but not missing). It was found that patients with N-SCDS had less frequent pressure- or sound-induced nystagmus, the cVEMP thresholds were higher and the oVEMP amplitudes were lower. In both cohorts, the results of the surgery were similar.
- **Perilymphatic fistula**

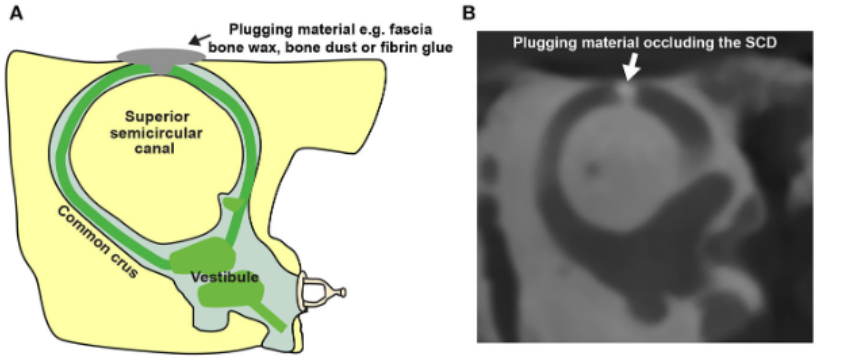
Box 12.2 Alphabetical List of the Important Differential Diagnoses of Syndrome of the Third Mobile Windows

- Bilateral syndrome of the third mobile windows
- Bilateral vestibulopathy
- BPPV
- Central positional vertigo
- Functional dizziness
- Menière's disease
- Panic attacks
- Vertebral artery compression/occlusion syndrome
- Vestibular paroxysmia
- Vestibular migraine

Pragmatic Therapy

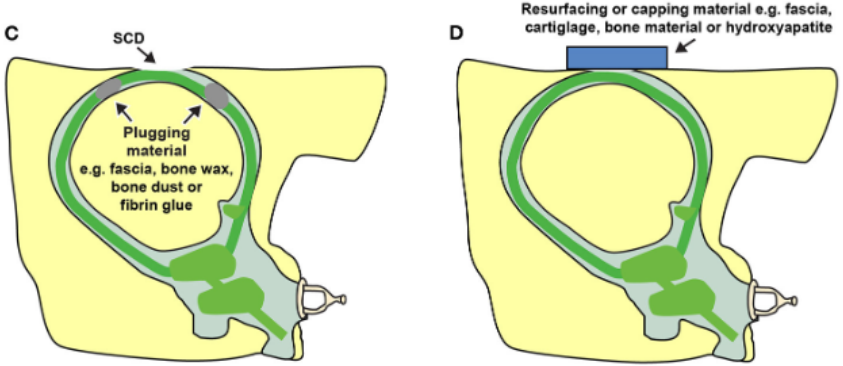
Explanation of the mechanism and the cause of the disease.

- In mild cases with tolerable symptoms, a conservative approach is justified, i.e., recommendation to avoid changes in pressure as much as possible.
- In cases with severe signs and symptoms and impairment of quality of life, surgery is possible **(no consensus about the best surgical methods and approach)**
  1. Canal plugging
  2. Resurfacing, i.e., restoring the surface of the superior semicircular canal
  3. Capping, i.e., simply covering the bony defect.



Occlusion of SCD via the middle fossa approach

CT of occlusion of SCD via the middle fossa approach



Isolation of SCD via the transmastoid approach

Resurfacing or Capping

FIGURE 9 | Illustrations depicting surgical repairs of SCD. **(A)** Plugging or occlusion of an arcuate eminence defect via middle fossa craniotomy approach. **(B)** CT image in the Pöschl view following repair. Occlusion was performed in a cadaveric temporal bone model of SCD using contrast-infused surgical bone wax. **(C)** Transmastoid approach for repair of SCD. A labyrinthotomy is created in the ascending and descending limbs of the superior semicircular canal and plugged to isolate the SCD. **(D)** Resurfacing or capping an arcuate eminence defect. This approach attempts to create a seal without occluding the superior semicircular canal lumen. *Modified from Cheng et al. (16).

Case of SCDS with different treatment options

- Possible surgical approaches are via the **middle fossa** or through the **mastoid**; large case series have been published (e.g., Romiyo et al. 2019).
- In a retrospective study in ten patients, it was demonstrated that **transmastoid resurfacing of the SCDS with cartilage** is associated with many complications (33%), in particular CSF leaks, and the long-term results were not promising: 33% of the patients required a second operation (Al et al. 2019).
- In a study in 118 patients, it was shown that **canal plugging can lead to significantly more vestibular symptoms** due to the defect in the superior canal function, but with a lower incidence of tinnitus (Wung et al. 2019).
- A **fixation of the stapes** or a stabilization of the oval window can also be performed (Gona and Phillips 2020) which reduces the resonance oscillations.

Perilymphatic Fistula/Transient Perilymphatic Leakage

- The leading symptom of “perilymphatic fistula” (PLF), which is **a very rare subtype of syndrome of the third mobile windows** and which can also be called transient perilymph leakage is recurrent attacks of vertigo induced by changes in pressure or certain sounds, which are thus also caused by a third mobile window.
- Most patients who were previously misdiagnosed with a PLF and had surgery most likely had SCDS, in particular since the fixation of the stapes can also result in an improvement of symptoms of SCDS.
- In addition to the above-mentioned tests for SCDS and the imaging studies of the temporal bone, measurement of **Cochlin-tomoprotein** has proved to be helpful to support the diagnosis of PLF.

Audiol Neurootol. 2009;14(5):338-44. doi: 10.1159/000212113. Epub 2009 Apr 15.

Cochlin-tomoprotein: a novel perilymph-specific protein and a potential marker for the diagnosis of perilymphatic fistula

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Box 12.3 Suggested Japanese Classification of the Subtypes and Possible Etiologies of Perilymphatic Fistula (2017)

- Category 1 In the history, trauma, diseases of the middle or inner ear, operations:
  - (1a) Direct labyrinthine trauma (luxation of the stapes, fracture of the capsula otocius, etc.)
  - (1b) Other trauma (brain trauma, polytrauma, etc.)
  - (2a) Diseases of the middle or inner ear (cholesteatoma, other tumors, malformations, dehiscence, etc.)
  - (2b) Iatrogenic (surgery or other medical treatment, etc.)
- Category 2 External barotrauma: history of massive pressure changes (e.g., scuba diving or flying)
- Category 3 Internal barotrauma: history of internal pressure changes (e.g., physical exertion, sneezing, coughing, pressing)
- Category 4 No evident cause in the history

Terminology

- A fistula (Latin *fistula*, i.e., a tube or pipe) is, by definition, a pathological tube-like connection between two hollow organs or between a hollow organ and the surface of the body. If one transfers this terminology to the perilymphatic space, one can assume that there is a leak of the perilymph, which leads to a impairment of function or a persisting loss of function of the inner ear. Therefore, one can also call this "**transient perilymph leakage**" (TPL).

Pathophysiology

- The perilymphatic and the endolymphatic space lie within the bony labyrinth. The border of tissue between the bony labyrinth and the middle ear forms the annular ligament of the base of the stapes and the membrane of the round window.
- The TPL can be localized at the **round or the oval window** or may be due to **bony defects**, such as fractures, erosion of the bone, or microfissures of the labyrinth.
- A TPL is thus caused by a pathological connection between the perilymphatic space of the inner ear and the middle ear, the mastoid, or even in an intracranial direction.

Etiology

- Possible causes are head trauma, barotrauma, chronic inflammation with erosion of the bone, tumors, in particular cholesteatomas, or surgical procedures such as stapedectomy or a cochlear implantation, simple traumas (sneezing, coughing, or pressing or lifting heavy weights)

- Specific causes can be:

1. A bony defect in the direction of the epidural space, as in SCDS
2. A pathological hypermobility of the stapes or the membrane of the oval or round window or the ossicular chain with a hypermotility of the stapes footplate.
3. A congenitally pathological bulging/prominence of the oval window causing a trans-stapedial CSF fistula, which leads to a perilymphatic hydrops and sensorineural hearing loss in children.
4. Fractures or microfissures, which extend, for instance, from the ampulla of the posterior canal to the round window.
5. Bony defects in the region of the lateral wall of the labyrinth (toward the middle ear) together with a partial collapse of the perilymphatic space (“floating labyrinth” (Nomura et al. 1992)) or
6. Chronic otitis media, which can also lead to a dehiscence of the horizontal canal (Chien et al. 2011).

- All of these can lead to a pathological pressure transduction from the middle ear to the labyrinth, which leads to symptoms during the Valsalva maneuver. The therapeutic principles derive from the pathophysiology: conservative (spontaneous) or operative closure of the leakage.

Treatment

- The therapy of the first choice for TPL is conservative, as most fistulas close spontaneously.

Conservative Therapy

- Conservative therapy consists of **1–3 weeks** of bed rest with moderate elevation of the head, if necessary a mild sedative, the administration of laxatives, and several weeks of limited physical activity that avoids all heavy lifting, abdominal pressing, strong coughing, or sneezing, even after improvement.
- The benefit of the frequently used tympanoscopy in patients with sudden hearing loss and suspected TPL: only 11–14% were identified with a leakage. It was concluded that there was **no significant improvement after exploratory tympanostomy and sealing of the membranes** for patients with a TPL (Heilen et al. 2020).

Surgical Therapy

- If conservative therapy fails and disturbing vestibular symptoms persist, exploratory tympanoscopy has been recommended to examine the oval and round windows.
- Surgical closure of the leakage successfully relieves vertigo in only up to 70% of patients (overview in Deveze et al. 2018); the pre-existing hearing loss generally does not improve at all.
- The operative procedure involves the removal of the mucous membrane in the region of the fistula and its substitution with autologous material
- Leakage of the oval window adjacent to the stapes footplate require a stapedectomy and prosthesis.
- Even if the operation is successful, the postoperative sensitivity of the patients to extreme physical strain (abdominal pressing, barotrauma) is greater than that of healthy subjects.

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EVA audiogram

- **Type and Configuration of Hearing Loss**
- Conductive, mixed, and sensorineural losses have all been reported in the literature. In the case of EVA, conductive hearing loss or mixed components are most likely to occur in the low frequencies, such as 250 Hz, 500 Hz and sometimes up to 1000 Hz. Oftentimes, bone conduction can be very good and present at the lowest end of normal, such as -10 dB, which may be missed if you are not testing below 0 dB HL. It is likely to see those conductive or mixed components in the lows, but we often see sensorineural components in the highs. As hearing declines, whether it be in a stepwise fashion, a gradual change, or more of a sudden change in hearing, we tend to see the hearing loss shift from conductive or mixed to sensorineural.
- The most common reported configurations of hearing loss for patients with EVA are downsloping, flat, and reverse cookie bite where the low frequencies have a hearing loss and hearing comes up and is better in the mid frequencies, and then becomes poorer again in the high frequencies.
- Degree of hearing loss can be all over the audiogram. There is no rule here. Recall that many patients with EVA will have additional inner ear abnormalities that may impact the progression or the degree of hearing loss. The degree of hearing loss can range from mild to profound. It is reported to potentially fluctuate, rapidly change, or gradually change over time with no specific incident or head injury. Hearing loss can also range from deafness in childhood to stable hearing loss into adult life.
- A review completed in 2011 by Dr. Gopen and colleagues revealed that 30% to 40% of ears had a stable hearing loss from a seven to nine year period, meaning that roughly over nine years, about 30-40% of ears had stable hearing. While that is great, we have to consider the other 60% of patients who have a gradual progression or sudden changes in hearing.
- Hand in hand with pure-tone thresholds, we likely will test word recognition ability. Oftentimes, word recognition will decline with the progression of hearing loss, just as we would see with typical progressive hearing losses, although word recognition may be poorer than expected when compared to other individuals who have conductive or mixed components of middle ear origin. Often in those cases, as soon as we bring the intensity up to a louder level, patients do very well on word recognition. This is not always the case in EVA, despite the presence of the mixed or conductive components.
- Additional measurements we include in our clinical workup will become very important. If we have a patient in front of us with a conductive and mixed component, our goal with tympanometry is to determine if the eardrum is moving well or if there is negative pressure in the ear. Even in the presence of a conductive or mixed component for patients with EVA, tympanometry is expected to be within normal limits, given that there is no fluid there.
- Acoustic reflexes performed with either tonal stimuli or broadband noise can typically be present with conductive or mixed components. If there is a straight sensorineural hearing loss in that ear, the general rules apply where we would expect the reflex to present, elevated, or absent. If you have a normal tymp, a conductive or mixed component and you are obtaining reflexes, that is an indication that the ossicles are moving as they should; that pattern is helpful in ruling out middle ear pathology. In EVA we are not looking to the middle ear as the source of the issue, so oftentimes reflexes are present.

- https://www.audiologyonline.com/articles/advanced-management-complex-cases-enlarged-15571

Left SCD nystagmus