Third windows in children

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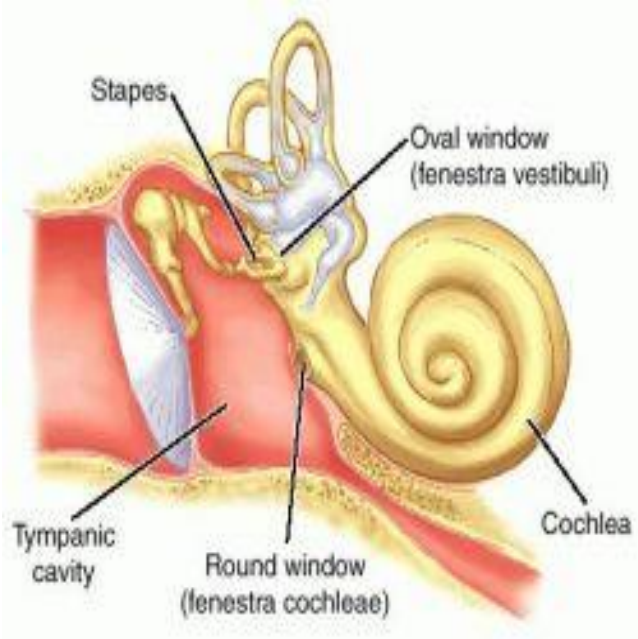
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Pathophysiology

Physiology

What are the 2 natural windows ?



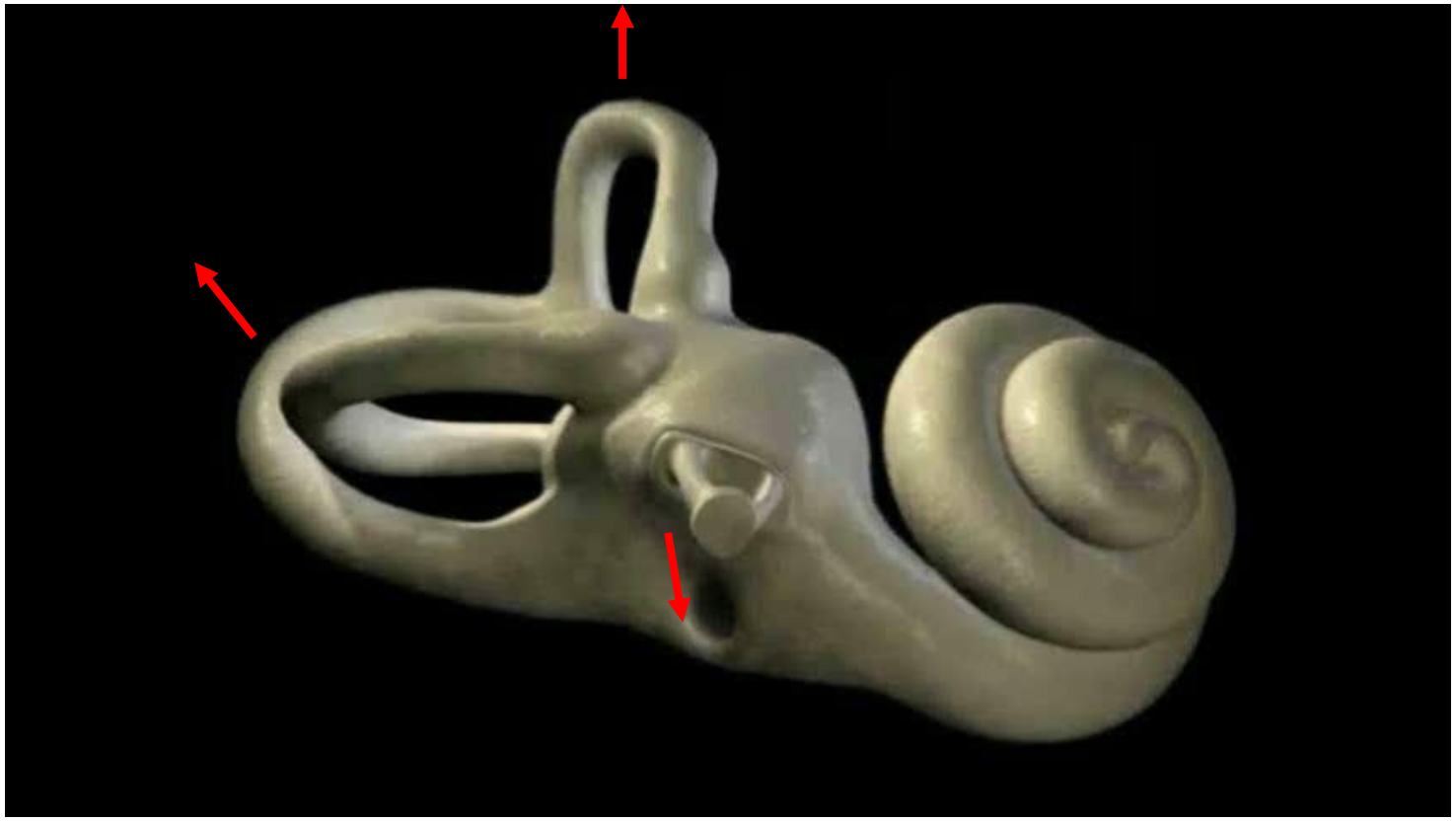
OW – preferential distribution
RW – Tympanic baffle

The cochlear travelling wave propagates from a zone of high SPL (OW) to a zone of low SPL (RW) thereby stimulating the basement membrane and processes responsible for hearing

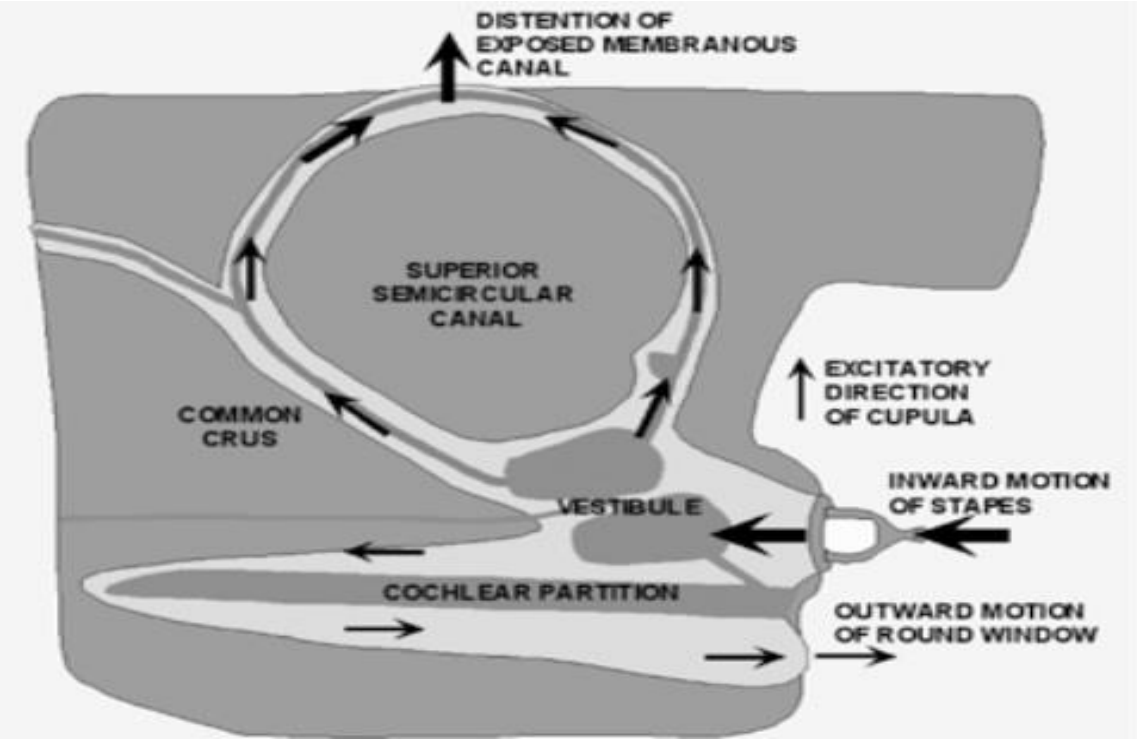
Pathology

The third window disorders are a group of conditions that are characterised by a direct or a virtual connection between either the middle and the inner ear or the inner ear and the cranial cavity which lead to pathological effects in the inner ear as the cochlear travelling wave follows a path of least resistance between the oval and the third window.

First identified by Lloyd Minor (1998) with his first case series of superior semicircular dehiscences.



The preferential distribution of stapes-oval window acoustic transference is overridden and shunted by the pathological third window – therefore you get a conductive hearing loss in PTA



- The cochlear travelling wave now travels from the OW to the third window where the SPL is lower than the natural RW
- Therefore, *BC improves due to lessening of resistance of the travelling wave*
- Stimulation of OHC by third window due to bony inertia may also improve BC
- Net result is an air – bone gap
- In later stages, may be direct cochlear involvement due to aberrant endolymphatic movement

Epidemiology

Incidence of Third windows in children

- Excluding endolymphatic hydrops, 0.1% of the population in adults (Hain 2016)
- Non syndromic semicircular canal dehiscences in children 1.7% SSCCD and 1.2% PSCCD out of 649 temporal bones (Saxby 2014)
- 6% of hearing impaired children have DVA (Madden 2014) that includes DES; DVA commonest paediatric third window
- About 90 children reported with X linked perilymph gusher (Dasgupta 2023)
- Others are extremely rare (Wackym 2020, Dasgupta 2022)

BAAP Mid Year 2023

Examples

1. **Superior, posterior and lateral semicircular canal dehiscence/near dehiscence**
2. Cochlear-facial nerve dehiscence
3. Cochlea-internal carotid artery dehiscence
4. Cochlea-internal auditory canal dehiscence
5. **X linked gusher syndrome**
6. **Perilymph fistula**
7. **Facial nerve canal dehiscence**
8. **Wide vestibular aqueduct in children**
9. **Dilated internal auditory meatus**
10. Hypermobile stapes footplate
11. Otosclerosis with internal auditory canal involvement
12. **Bone dyscrasias**
13. **Endolymphatic hydrops**

Causes and natural history

- May be congenital or follow other anatomical abnormalities for example high riding jugular bulbs for PSCD; venous sinus abnormalities for SSCD
- Syndromic – as part of Pendred, CHARGE or BOR, even Down's
- Or may be idiopathic
- The conditions may be late onset and evolving as the bony labyrinth undergoes dynamic remodelling and any exposed dura may be osteogenic, later becoming osteopenic (Saxby et al 2014)
- In bony dehiscences, dural plate deficiency leads to dural herniation into the third window that exacerbates symptoms (Ward 2017)
- Traumatic (post head injury or intracranial radiation)

Clinical features and diagnosis in children

Important points to consider

- Third windows are bony abnormalities subjected to accelerated osteogenesis and bony remodelling in a growing skull
- This means that the behaviour of such abnormalities will be different from those in adults
- This leads to a different phenotype of third windows in children both in terms of clinical features and in laboratory diagnosis and as a result often not diagnosed
- Classical features become apparent in mid to late teens
- May exist as a part of a syndrome
- TWS is a diagnosis based on anamnesis that is difficult to obtain from children
- The angular and gravitational motion sensors may be fundamentally weak that can be quantified

Symptoms

- Hearing loss
- *Vestibular symptoms*
- *Conductive dysacusis - misophonia*
- Gaze evoked tinnitus
- *Autophony*
- Positive pressure disorientation
- Pulsatile tinnitus/oscillopsia
- Tullio’s phenomenon
- *Aural fullness*
- *Motor developmental delay*
- *Disproportionate functional hearing*

Signs

- Hennebert’s sign
- Vestibular signs in VOR and VST
- Mastoid vibration provoking nystagmus
- The video head impulse test in third windows show variability and in canal dehiscences may localise to canals that do not show dehiscence (Dasgupta 2020 and 2023,Tikka 2021, Castellucci 2024) due to modified endolymphatic fluid mechanics
- VEMPS may not show the classical hyper response in amplitude but show lower thresholds or absent/reduced responses (Dasgupta 2020, 2022, 2023)
- PTA with variable loss and normal middle ear – CHL/SNHL/mixed

- Commonest narrated symptom if possible is conductive dysacusis, misophonia, pulsatile tinnitus, dysequilibrium and autophony – Hennebert's, Tullio's are uncommon - hearing loss ranges from CHL, mixed and SNHL – vestibular assessment shows VEMP and vHIT abnormalities

- History is often surrogate

- Final diagnosis by HRCT – thinning rather than a frank dehiscence can generate a similar phenotype

- Symptoms may generate a significant emotional reaction due to their oddity and not infrequently children sent to mental health services due to a lack of awareness

Diagnosis in children

The art of rational thinking and inductive reasoning



Inspector Gregory: Is there any point to which you wish to draw my attention?

Holmes: To the curious incident of the dog in the night-time.

Gregory: The dog did nothing in the night-time.

Holmes: That was the curious incident.

High index of suspicion when:

- A conductive hearing loss regardless of tympanometry which cannot be explained by OME for example > 50dBHL
- Mixed loss of any intensity
- Negative BC threshold
- Normal Impedance studies including SRT and OAE
- Balance symptoms and abnormal vestibular function tests especially vHIT and VEMPs

- In case of sensorineural hearing loss, TWS is always a possibility
- Progressive but fluctuating hearing loss
- ***Rarely third window features in children unless older***
- Paradoxical cochleovestibular involvement
- Disproportionate functional hearing as compared to measured hearing or disproportionate result with HA

- Vestibular investigation is crucial as the majority develops a vestibular problem in the vertical canals; may not localise to symptomatic side
- HRCT is the investigation of choice as by definition third window affects the bony labyrinth; MRI picks up only the dilated endolymphatic sac and duct system
- All patients with refractory middle ear disorder need to be investigated and followed up
- VEMPS indicated in all cases

Every individual paediatric vestibular laboratory must have its own vHIT and VEMP norms and extrapolation of adult norms should be avoided

The Alder Hey criteria for requesting HRCT to confirm/exclude a third window (two or more of the following):

- Mixed or conductive hearing loss with AB gap exceeding 60 dBHL
- Normal SRT and OAE
- Symptoms of disequilibrium and overt TWS
- Abnormal cVEMP – high amplitude/low threshold; absent response; pathological asymmetry between 2 sides
- Abnormal vHIT with pathological saccades
- Abnormal VNG suggestive of TW
- Sensorineural hearing loss with normal MRI

The Alder Hey series

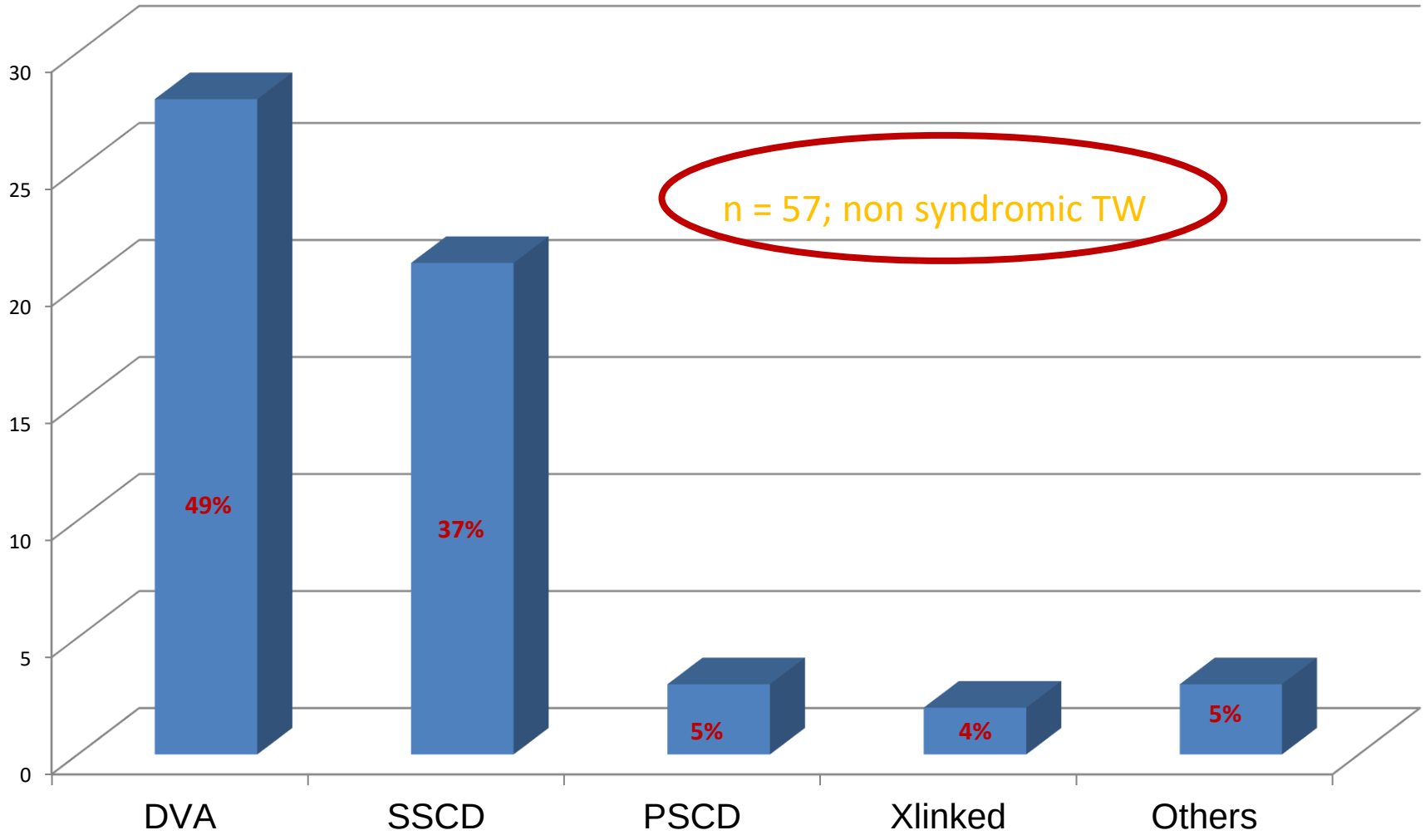



Audio Vestibular Medicine
Vestibular Laboratory



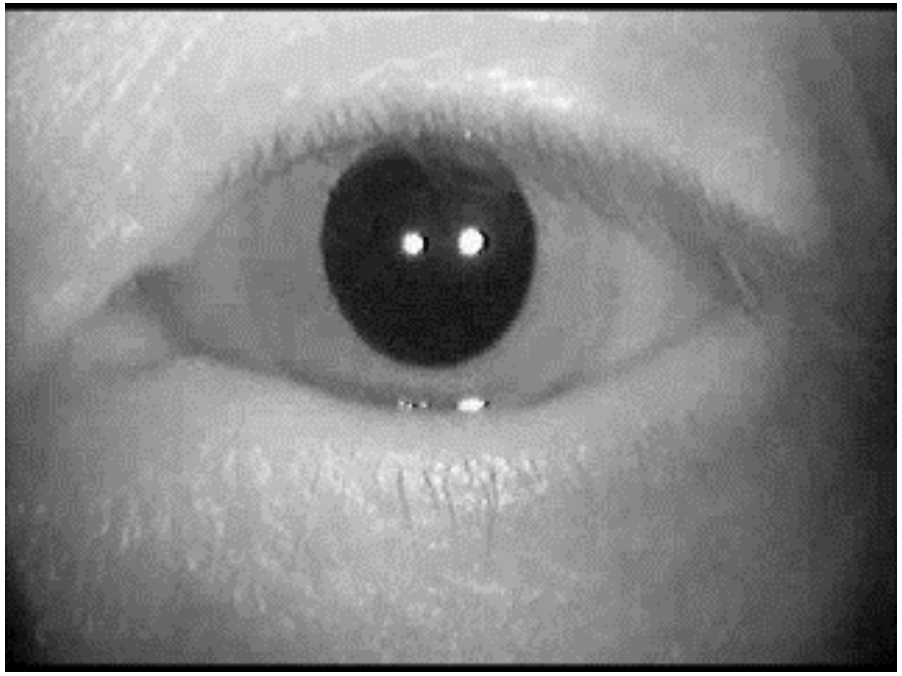
920 children referred to a tertiary vestibular unit from general paediatrics, community paediatrics, audiology, neurology, endocrinology, rheumatology, cardiology, gastroenterology, oncology, neurosurgery, ENT and *CAMHS* between February 2016 and September 2019

- Full audiological quantification along with PTA, OAE and SRT
- Full vestibular quantification with VNG especially Hennebert's sign, vHIT and VEMP
- vHIT may localise to the same canal or involve a different canal (Dasgupta 2020 and Tikka 2021)
- VEMPs must be used with caution unless you have your own norms in children due to reported variability in children; also, if the vestibular function is deranged, VEMPS might be absent or diminished (Dasgupta et al 2020)
- HRCT of temporal bones reformatted at less than 0.5mm in coronal, axial, Stenver and Poschl's views (to specify this in request)
- MRI in some instances as an initial screen (DES, dilated IAM)

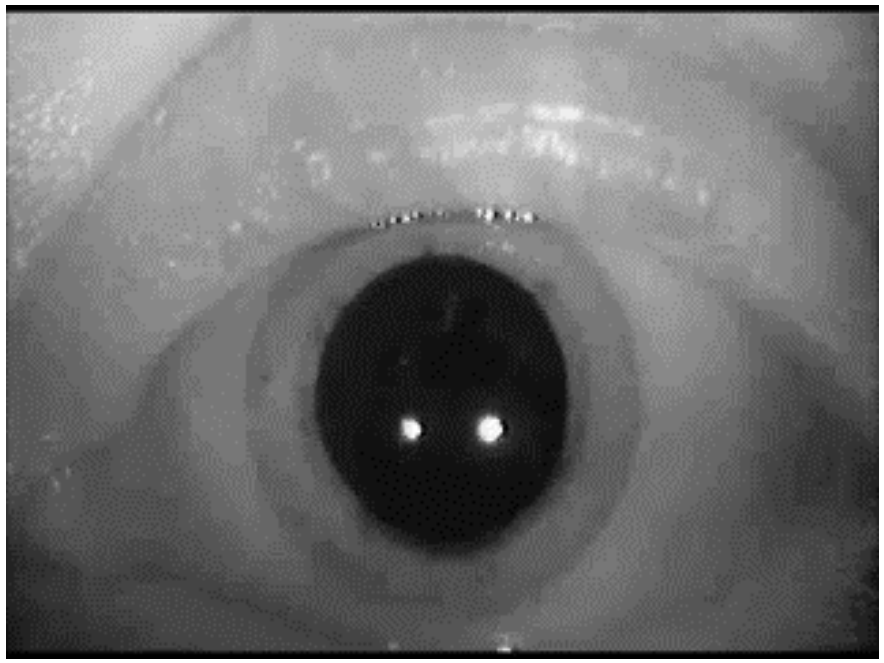


Third window in whole cohort: 6.19%

Hennebert’s sign

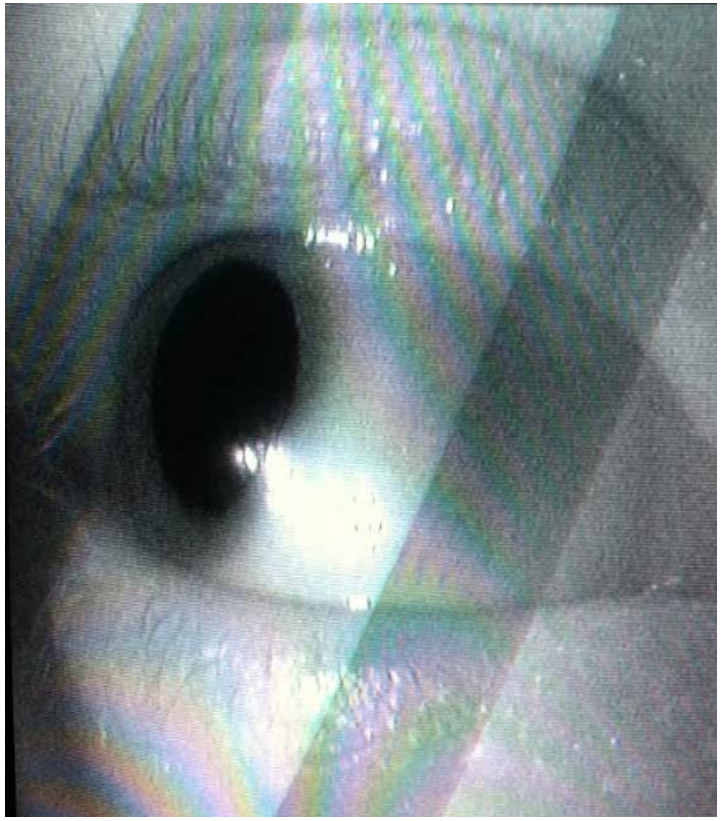


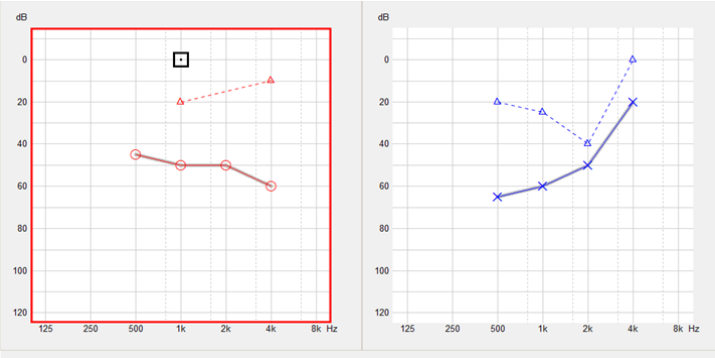
SSCDS



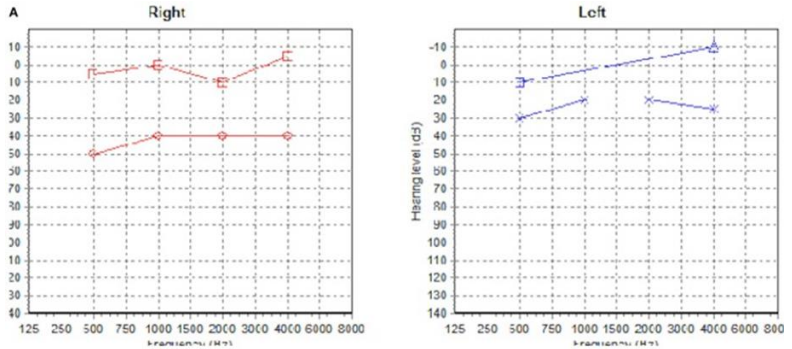
PSCDS

Pulsatile downbeat nystagmus

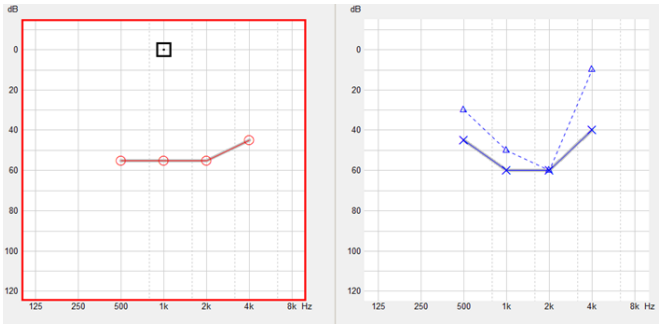




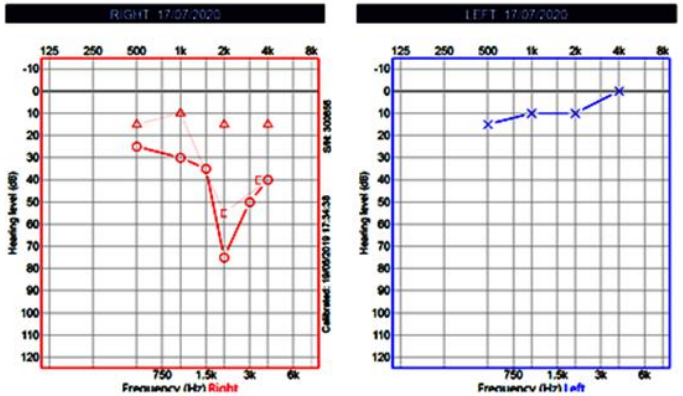
DVA



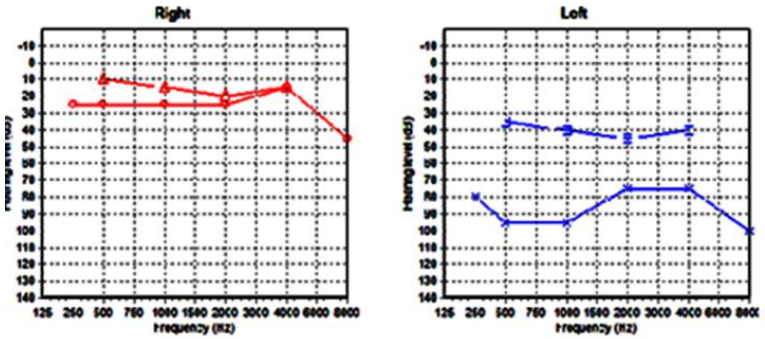
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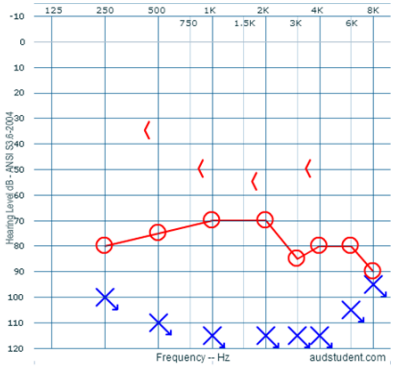
PSCD



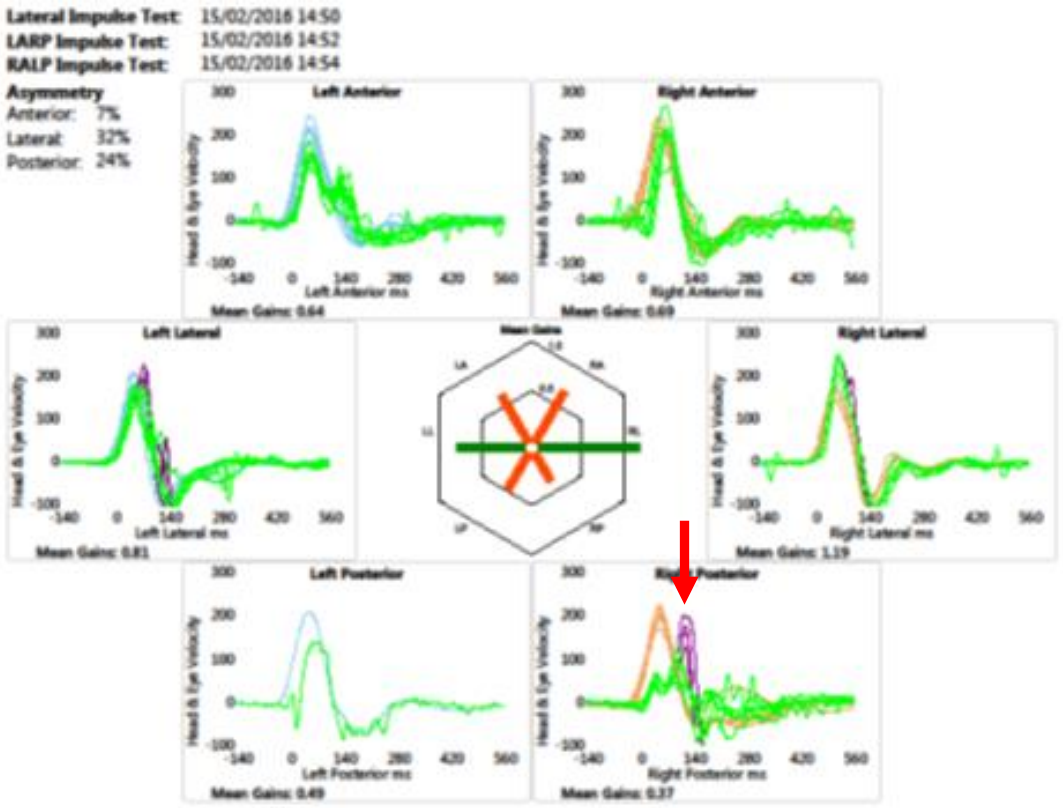
PSCT



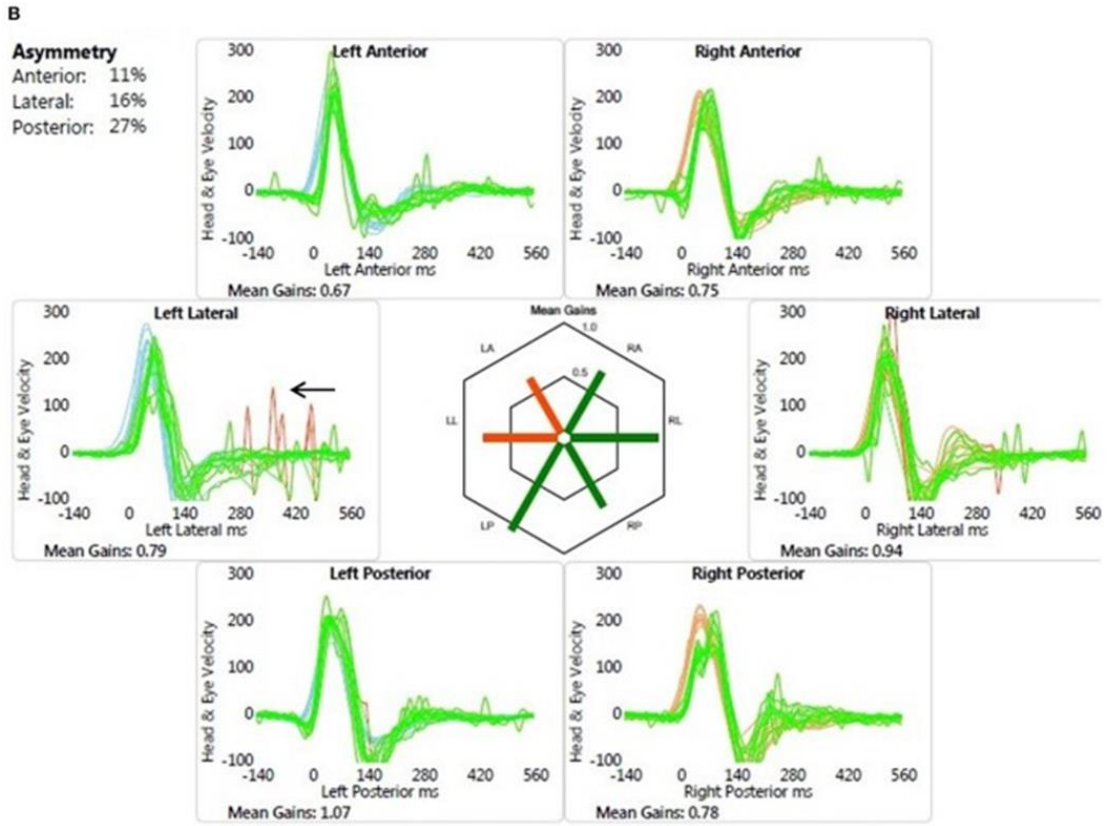
Dilated IAM/FCD



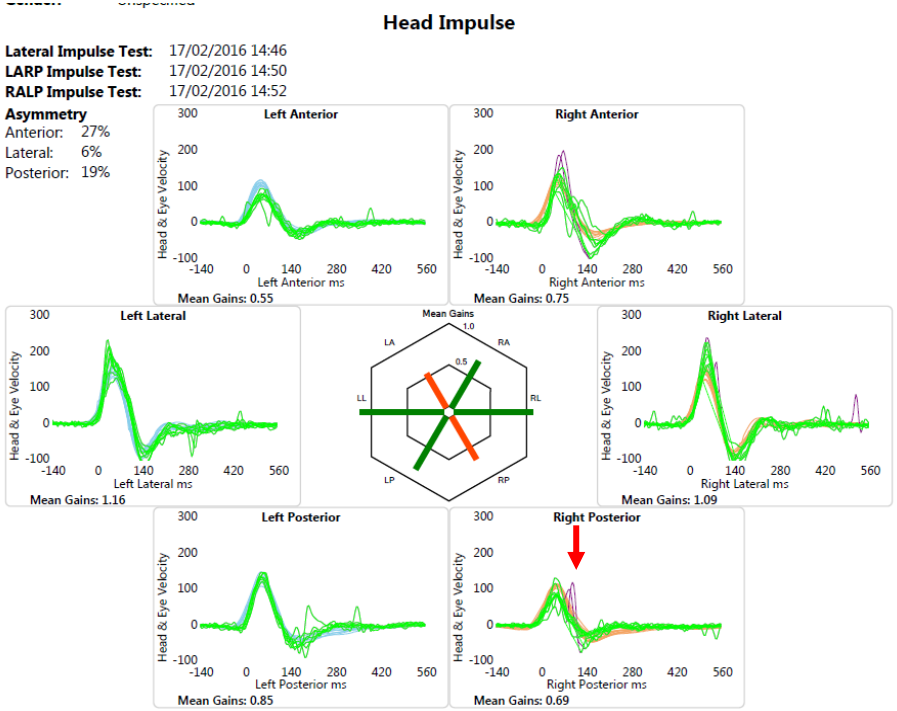
X linked gusher


Lateral Impulse Test: 15/02/2016 14:50
LARP Impulse Test: 15/02/2016 14:52
RALP Impulse Test: 15/02/2016 14:54
Asymmetry
Anterior: 7%
Lateral: 32%
Posterior: 24%
Left Anterior
Right Anterior
Left Lateral
Right Lateral
Left Posterior
Right Posterior
Mean Gains: 0.64
Mean Gains: 0.69
Mean Gains: 0.81
Mean Gains: 1.19
Mean Gains: 0.49
Mean Gains: 0.37

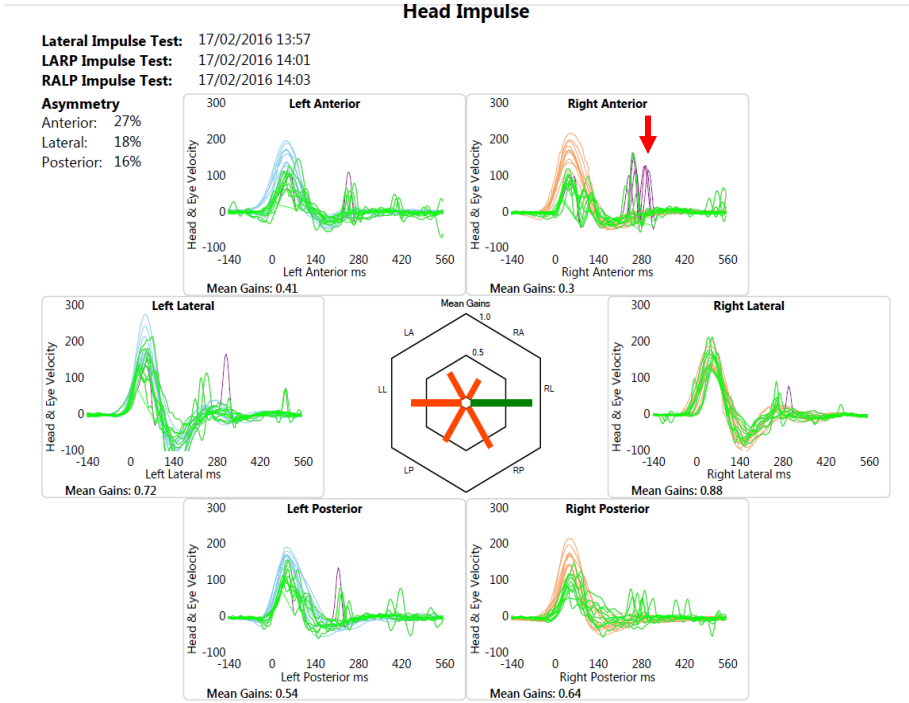
DVA


B
Asymmetry
Anterior: 11%
Lateral: 16%
Posterior: 27%
Left Anterior
Right Anterior
Left Lateral
Right Lateral
Left Posterior
Right Posterior
Mean Gains: 0.67
Mean Gains: 0.75
Mean Gains: 0.79
Mean Gains: 0.94
Mean Gains: 1.07
Mean Gains: 0.78

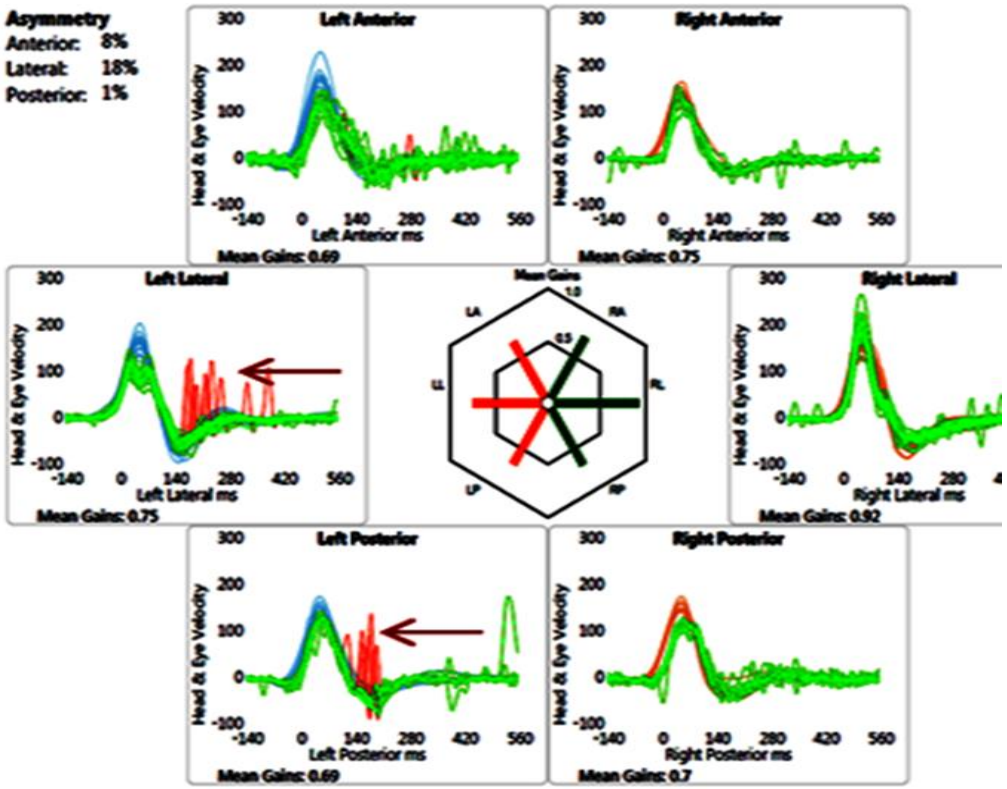
SSCD



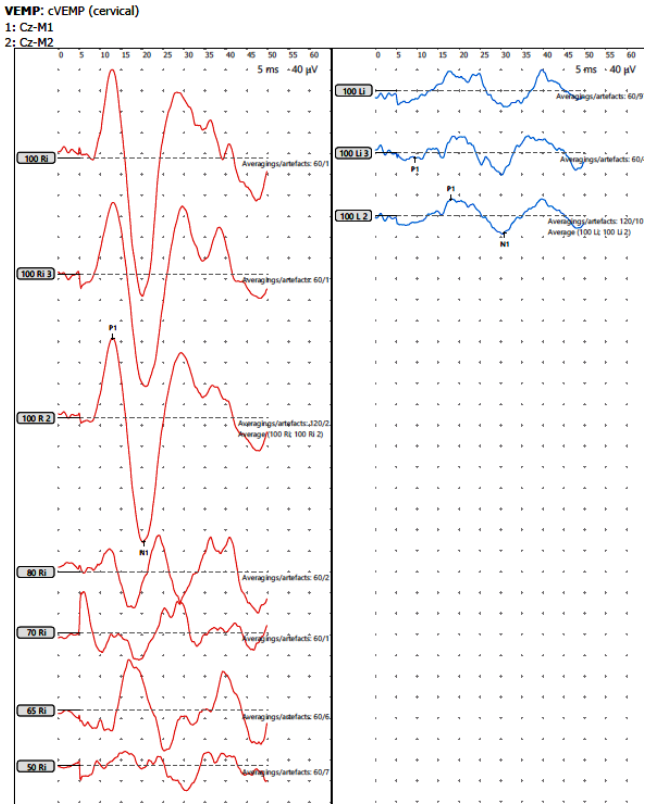
PSCD



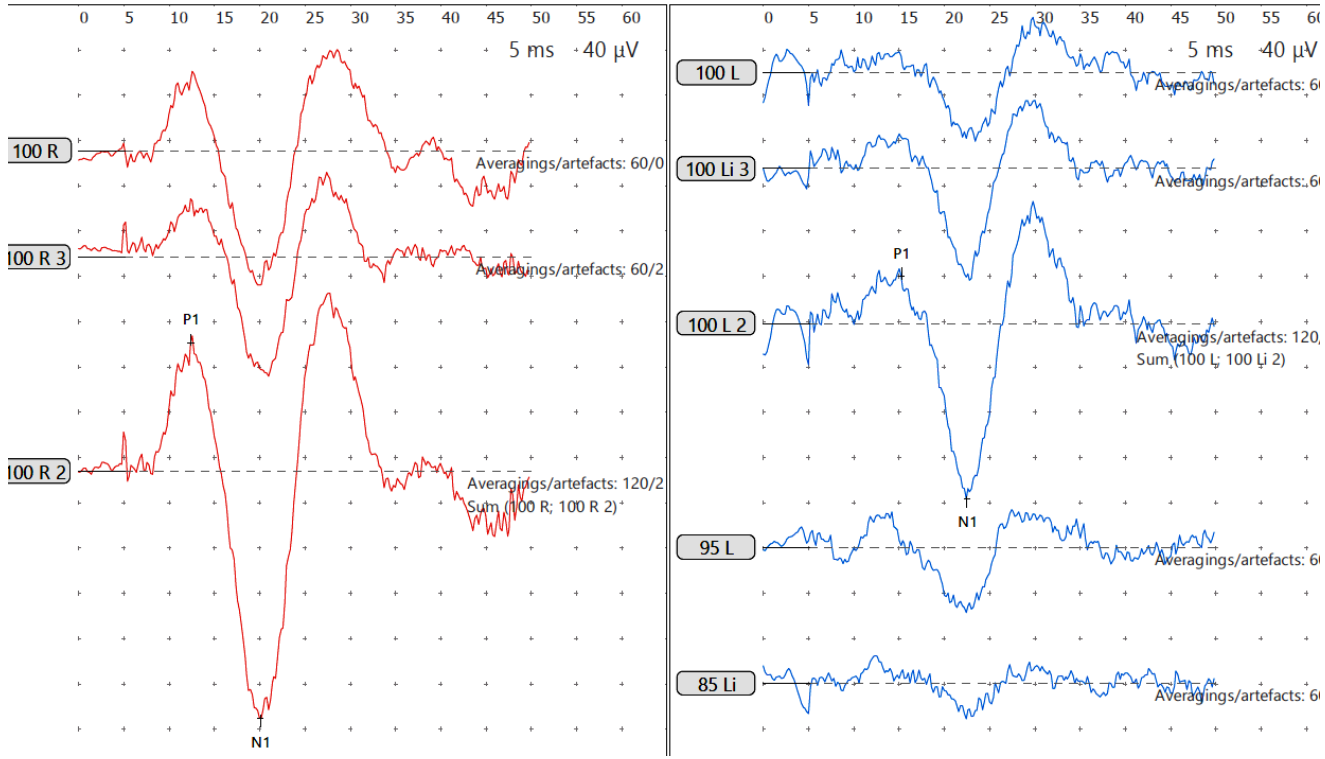
X linked gusher



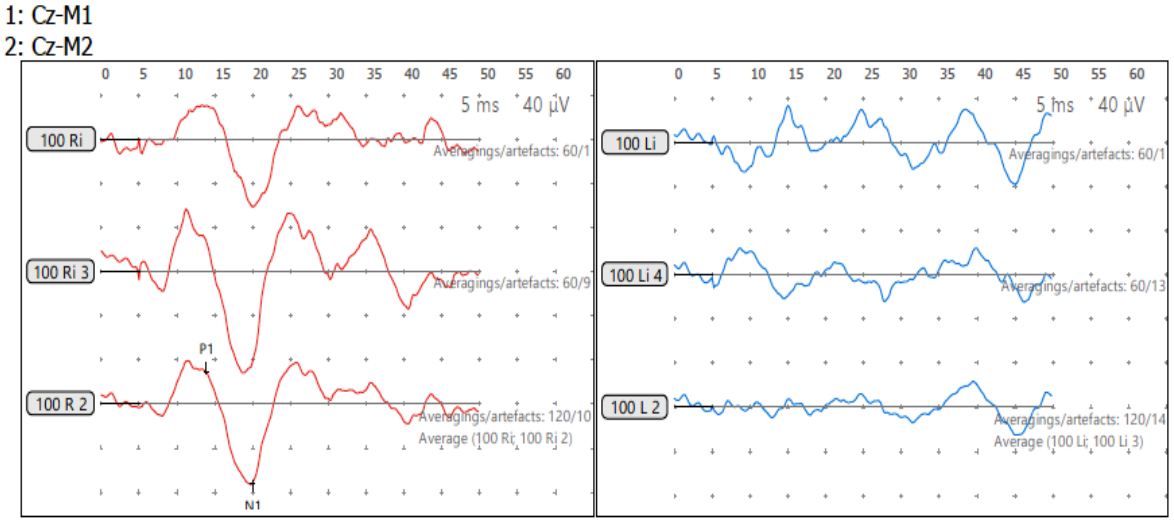
Facial canal dehiscence and dilated IAM


VEMP: cVEMP (cervical)
1: Cz-M1
2: Cz-M2
5 ms 40 µV
100 Ri
100 Ri 3
100 R 2
80 Ri
70 Ri
65 Ri
50 Ri
100 Li
100 Li 3
100 L 2
P1
N1

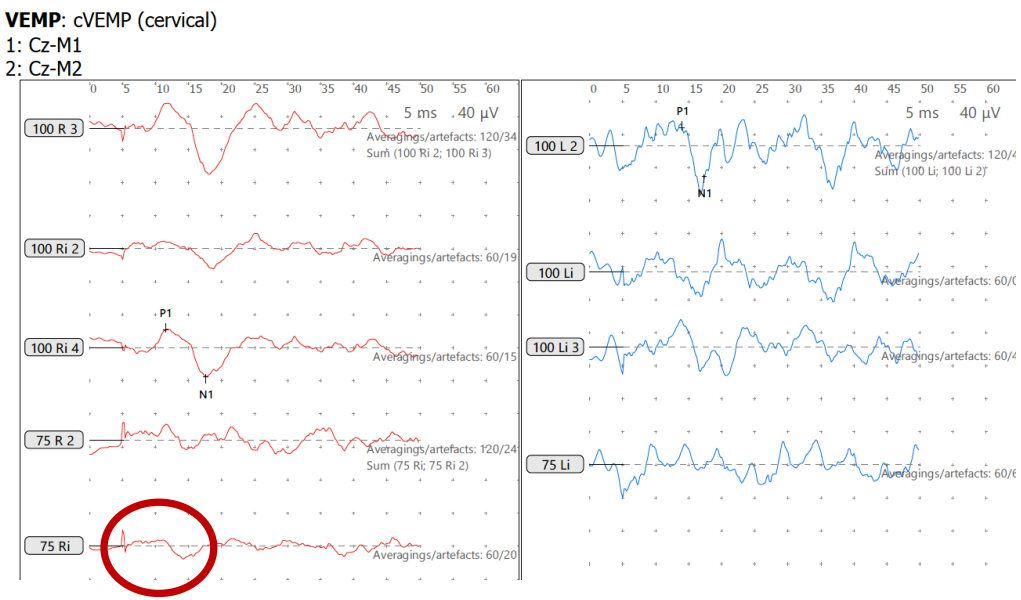
DVA


5 ms 40 µV
100 R
100 R 3
100 R 2
Averagings/artefacts: 60/0
Averagings/artefacts: 60/2
Averagings/artefacts: 120/2
Sum (100 R; 100 R 2)
P1
N1
100 L
100 Li 3
100 L 2
95 L
85 Li
Sum (100 L; 100 Li 2)

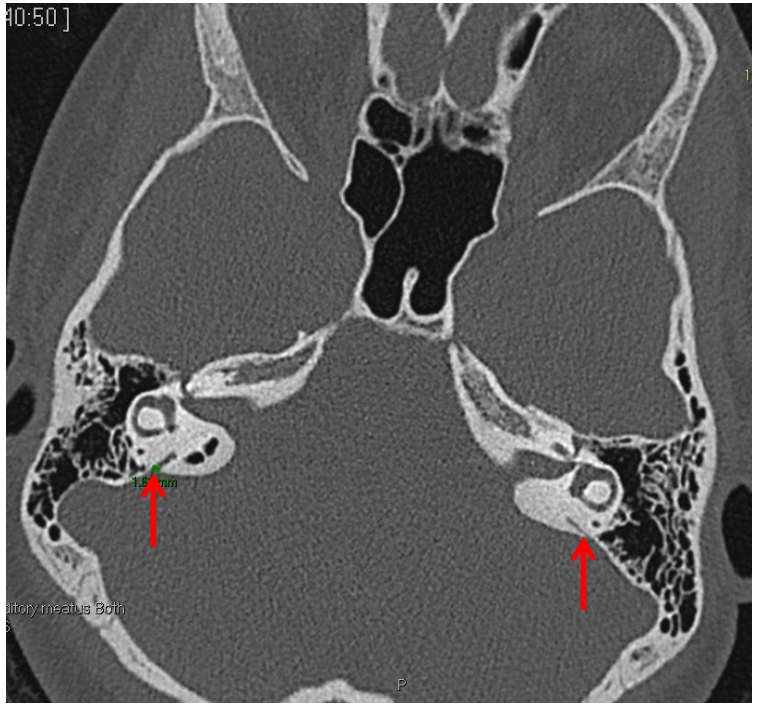
SCDS



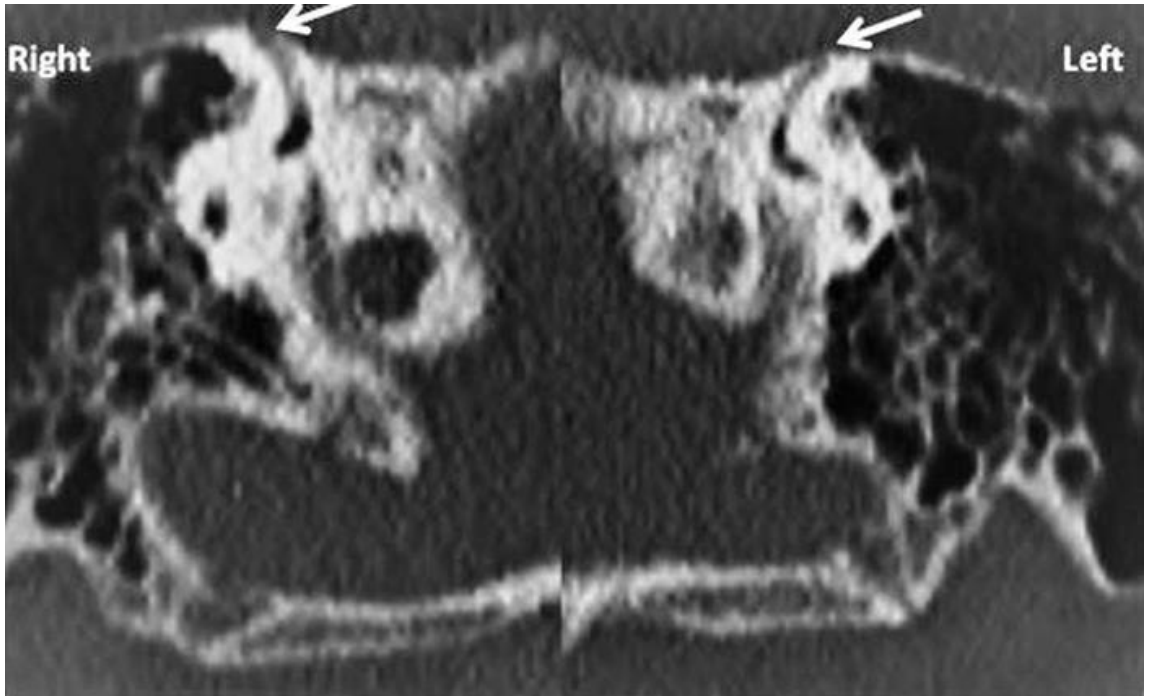
Facial canal dehiscence



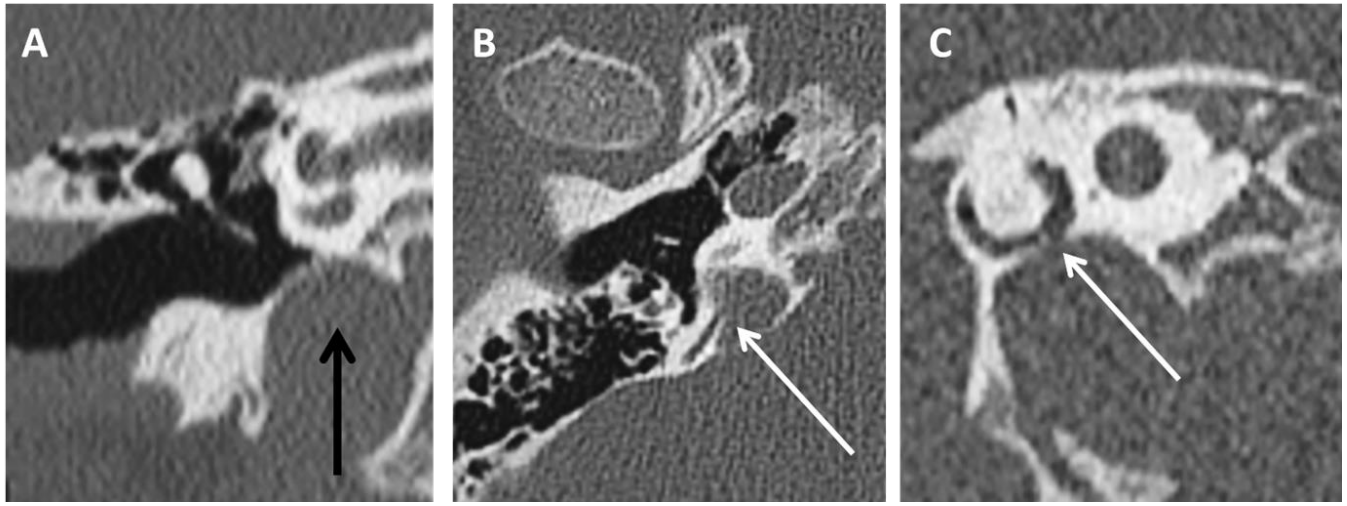
X linked gusher

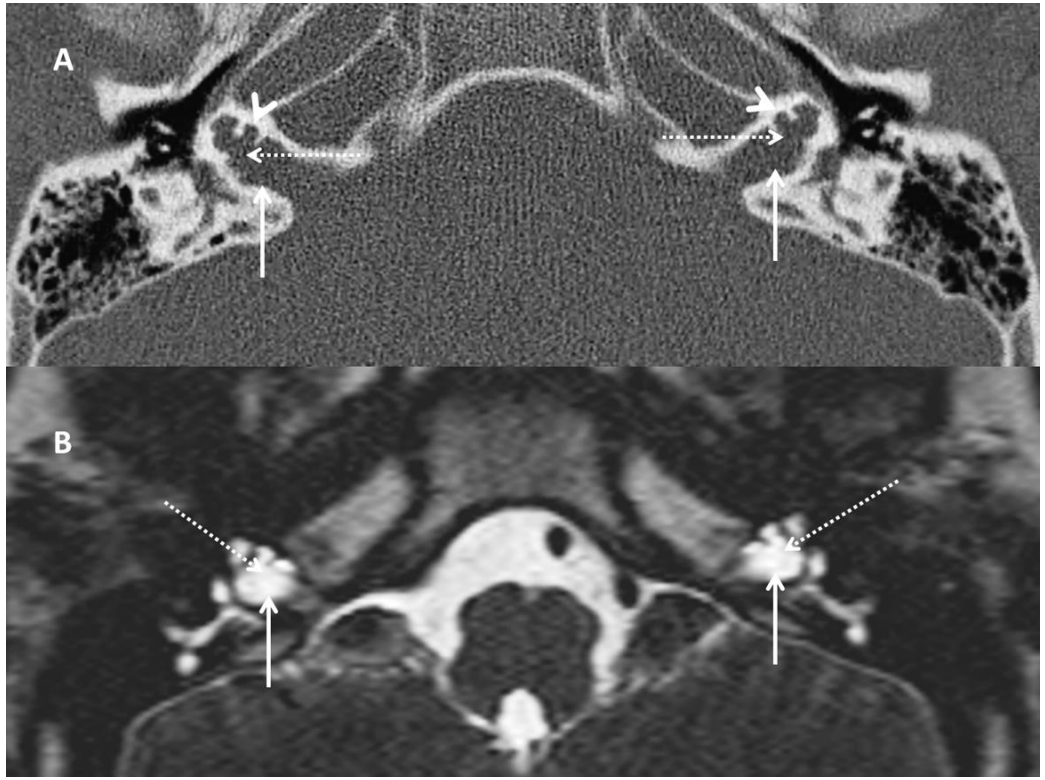


DVA



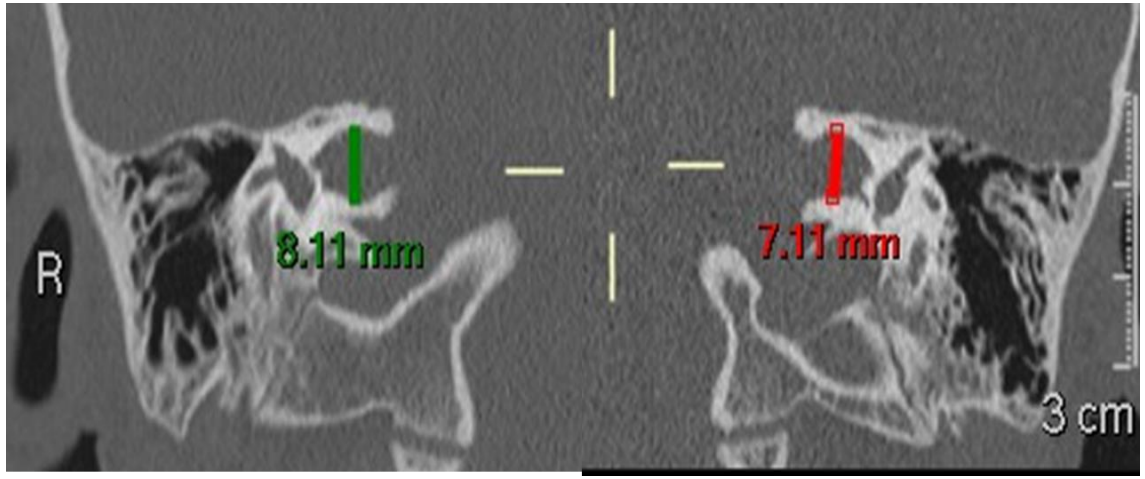
SSCD



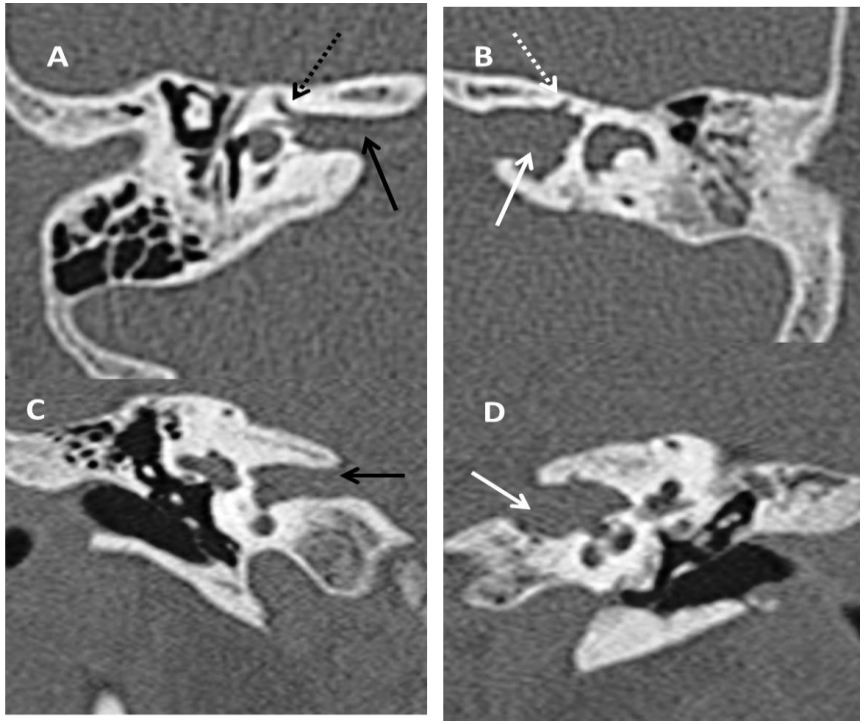


PSCD

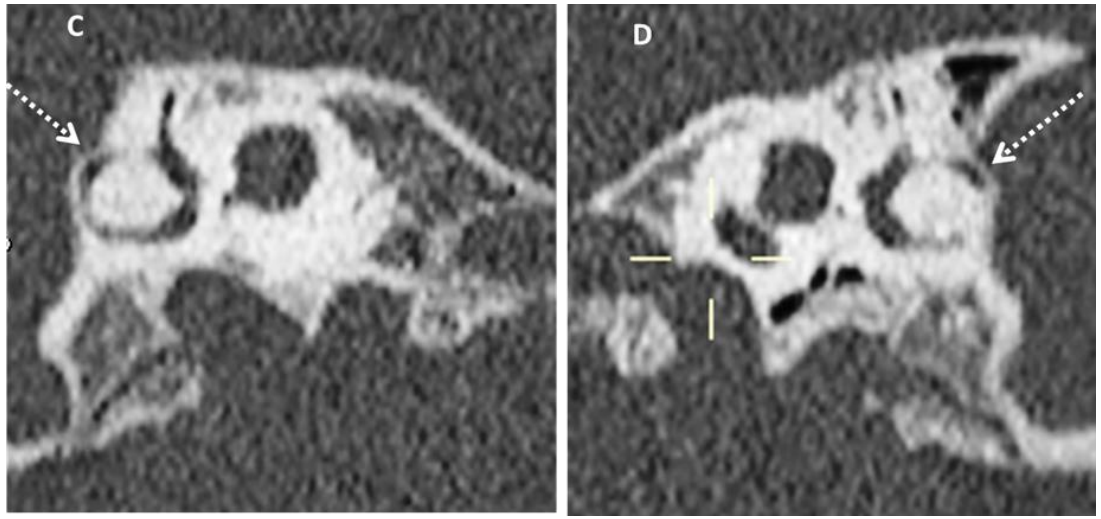
X linked gusher



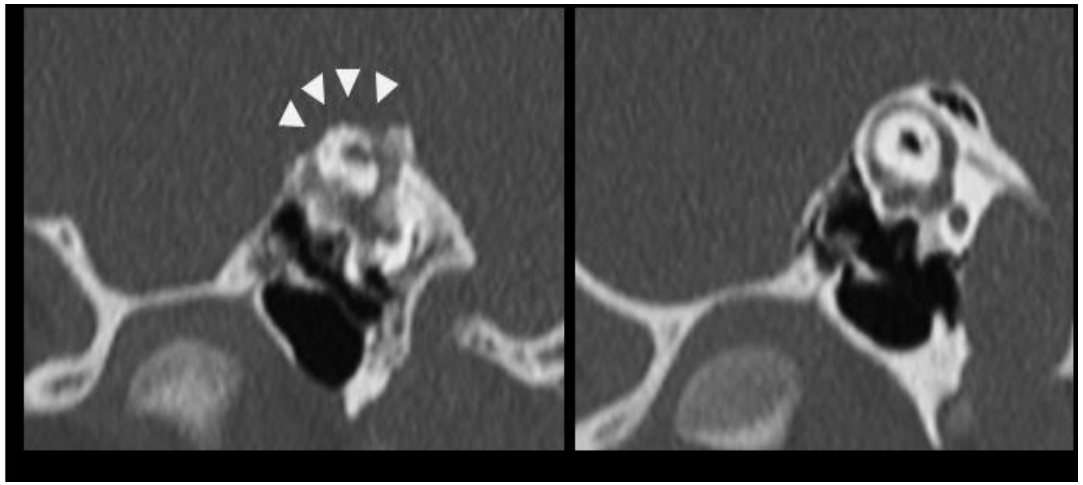
Dilated IAM



Facial canal dehiscence and dilated IAM



PSCT



Post radiation SSCD

Management

- Accurate diagnosis information leads to adjustment and self awareness
- Hearing rehabilitation with variable outcome
- Vestibular rehabilitation with variable outcome
- For distressing conductive dysacusis surgery involving capping and plugging of canal if dehiscence is the cause or even plugging round window

- Cognitive well being achieved by diagnosis (Wackym 2017)
- To keep under follow up with serial audiovestibulometry to see evolution of condition
- Situational counselling regarding body movements, posture, sounds and positive pressure

Conclusions

- Third window disorders are rare in children and are great mimickers (Carey 2013).

- A strong index of suspicion suggested by clinical features often result in confirmation of diagnosis and significantly positively affect cognitive skills in children to deal with their illness much better regardless of treatment options.

- Children may not present with classical third window symptoms and may show variable vestibular function test results including VEMPS.

Therefore, it must be kept in mind that a third window phenotype and the management algorithm in children may be different from adults.

frontiers in Neurology

METHODS
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Functional and Objective Audiovestibular Evaluation of Children With Apparent Semicircular Canal Dehiscence–A Case Series in a Pediatric Vestibular Center

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frontiers in Neurology

ORIGINAL RESEARCH
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Audiovestibular Quantification in Rare Third Window Disorders in Children

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X-Linked Gusher Disease DFNX2 in Children, a Rare Inner Ear Dysplasia with Mixed Hearing and Vestibular Loss

Soumit Dasgupta, Conceptualization, Methodology, Software, Validation, Formal analysis, Investigation, Resources, Data curation, Writing – original draft, Writing – review & editing, Visualization, Supervision, Project administration, Funding acquisition,[1,2,*] James Hong, Methodology, Formal analysis, Investigation, Data curation,[2] Rhyanna Morris, Conceptualization, Formal analysis, Investigation, Data curation, Writing – review & editing,[2] Javed Iqbal, Software, Validation, Formal analysis, Investigation, Data curation, Writing – review & editing,[3] Amy Lennox-Bowley, Software, Validation, Formal analysis, Investigation, Data curation, Writing – review & editing,[4] and Jeyasakthy Saniasiaya, Methodology, Validation, Formal analysis, Investigation, Data curation, Writing – review & editing[5]



When treating a child with conductive or mixed loss, never forget the elusive third window!

Thank you – soumit.dasgupta@alderhey.nhs.uk

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