

Pediatric Vestibular Loss: The Need for Multi-Disciplinary Assessment

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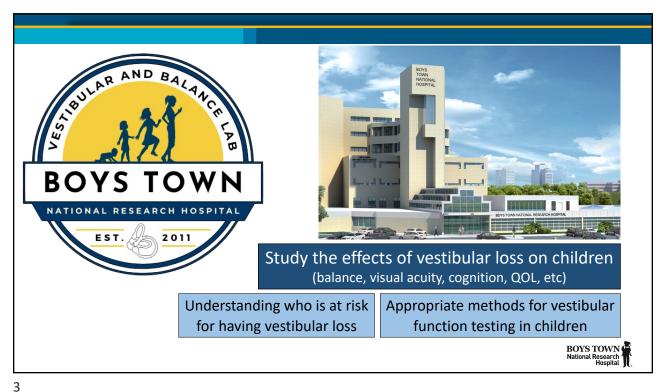
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Objectives

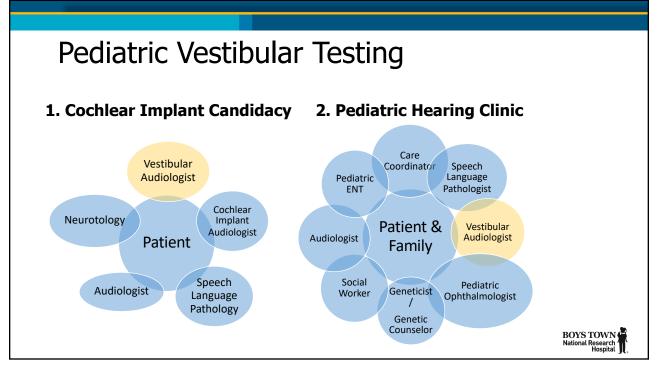
After this course, participants will be able to:

- Identify children who are candidates for vestibular testing
- 2. List causes of pediatric vestibular loss and dizziness
- 3. List practitioner types needed for multidisciplinary assessment of pediatric vestibular loss





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Pediatric Vestibular Testing

3. Vestibular Clinic

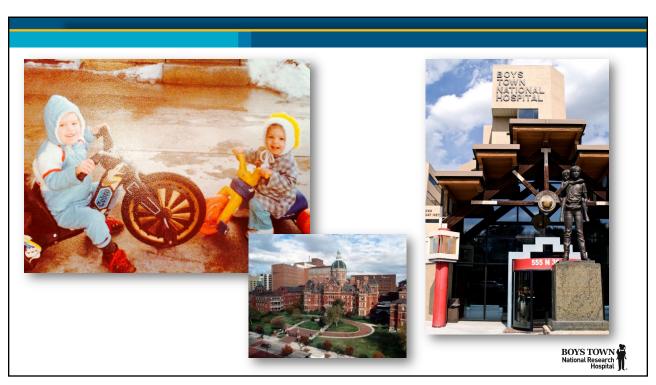


Administrative

- Lead Audiologist, BTNRH Vestibular Program
- Director, T35 Short-term Research Training Program for AuD Students
- Externship Selection Committee



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Prevalence of Vestibular and Balance Disorders in Children

Otology & Neurotology 31:1441–1444 © 2010, Otology & Neurotology, Inc.

*Robert C. O'Reilly, †Thierry Morlet, *Brian D. Nicholas, ‡Gary Josephson, ‡Drew Horlbeck, §Larry Lundy, and ||Arnel Mercado

- Reviewed 561,151 patient encounters
 - 2,546 (0.45%) were diagnosed with a balance disorder
 - 159 (6.2%) peripheral vestibular disorders
 - · Odds ratio: 43 times more likely to have SNHL
 - 109 (4.1%) central involvement was diagnosed
 - Odds ratio: 16 times more likely to have headache
 - 2,283 (90%) unspecified dizziness



Epidemiology of Dizziness and Balance Problems in Children in the United States: A Population-Based Study

Chuan-Ming Li, MD, PhD¹, Howard J. Hoffman, MA¹, Bryan K. Ward, MD², Helen S. Cohen, EdD, OTR³, and Rose Marie Rine. PT. PhD^{4,5}

(J Pediatr 2016;171:240-7).

- Parents were asked if during the past year their child was bothered by symptoms of dizziness and balance problems (10, 954)
 - Prevalence 5.3% (female: 5.7%, males 5.0%)
 - · Prevalence increased with age
- · Associations:
 - Hearing loss increased the risk of dizziness and balance by 4.8
 - Difficulty seeing increased the risk of dizziness and balance by 3



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Epidemiological studies tell us that there are at least 2 categories of kids at risk for vestibular loss:

Kids who complain of dizziness

 Children can develop similar etiologies of vestibular loss and thus similar symptoms as adults, including dizziness, imbalance, falls, and hearing Loss

Kids with Hearing Loss

 Are all children with hearing loss at equal risk for vestibular loss?



SNHL is a risk factor for vestibular loss

(Verbecque et al 2017)

- Children are more likely to have vestibular loss with:
 - Severe-to-profound sensorineural hearing loss (Janky et al., 2018; Martens et al., 2022)
 - · And with specific etiologies of hearing loss:
 - Cytomegalovirus (CMV) infection (30 60%)
 - Auditory Neuropathy (show decline in function over time)
 - Waardenburg Syndrome (up to 70/80%)
 - Rubella (30 40%)
 - Pendred Syndrome
 - Malformations (depends on severity of malformation)
 - Meningitis (~100%, depends on rate sensor)
 - Ototoxicity
 - Usher Syndrome (~100%, depends on type)

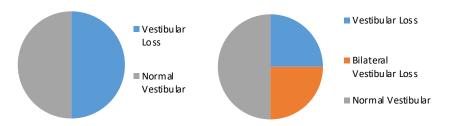
Hearing loss etiology is unknown in $\sim 35-42\%$ of cases (Sokolov 2019, Cushing 2019) and not all etiologies result in complete vestibular loss (Cushing 2019), suggesting etiology alone is not a sufficient predictor of vestibular loss.



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Not all children with HL have vestibular loss

- · Specific to children with CI
 - 50% have some degree of vestibular loss
 - 20 30% have bilateral loss
 - Jacot et al. (2009), Jin et al. (2006), Cushing et al (2013), Janky et al. (2015)





Vestibular End-Organ Dysfunction in Children With Otology & Neurotology Sensorineural Hearing Loss and Cochlear Implants: An Expanded Cohort and Etiologic Assessment [*†Sharon L. Cushing, *†Karen A. Gordon, †‡John A. Rutka, *†Adrian L. James, and *†Blake C. Papsin HORIZONTAL CANAL FUNCTION SACCULAR FUNCTION HOHIL-CALORIC **ROTATIONAL** VEMP □Normal □ Unilateral ■ Bi MENINGITIS 50% of children pre-CI **ABNORMAL** have some degree of **COCHLEA** vestibular loss - 30 % have bilateral loss **CONNEXIN 26** (homozygous)

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OTHER

UNKNOWN

Not all children with hearing loss have vestibular loss

AJA

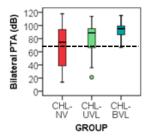
Research Article

Predictive Factors for Vestibular Loss in Children With Hearing Loss

Kristen L. Janky,^a Megan L. A. Thomas,^a Robin R. High,^b Kendra K. Schmid,^b and Oluwaseye Ayoola Ogun^c

Severe to profound sensorineural hearing loss and specifically in those with meningitis, syndromes, congenital cytomegalovirus, and cochleovestibular anomalies (Martens, 2022)

 Children are more likely to have vestibular loss if their bilateral PTA is > 66 dB





Gross Motor Delay is a Common Consequence of Vestibular Loss in Children

- Gross motor function can be used as a screening tool for vestibular loss (Oyewumi, 2016; Janky 2018; Janky 2022)
- Children are more likely to have vestibular loss if:
 - Parents report concern for gross motor delay
 - Infants sit later than 7.5 months or walk later than 14.5 months
 - Older children cannot stand on one leg with eyes closed for 4 - 5 seconds

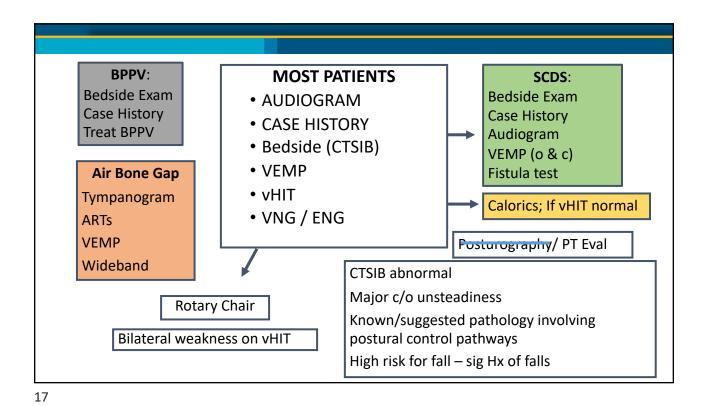


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What protocol are we using with children?





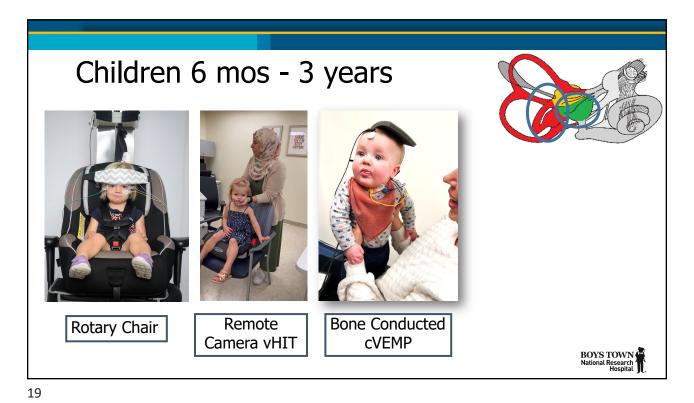
AUDIOGRAM

- CASE HISTORY
- Bedside (SLS, CTSIB, ocular motor, HIT)
- Dix Hallpike
- Assessment of Canal function
- Assessment of Otolith Function

- Chief complaint of gross motor delay

- PT Eval

- BOYS TOWN Research Research



Children 3 - 7 years

Air/bone-Conducted ovemp

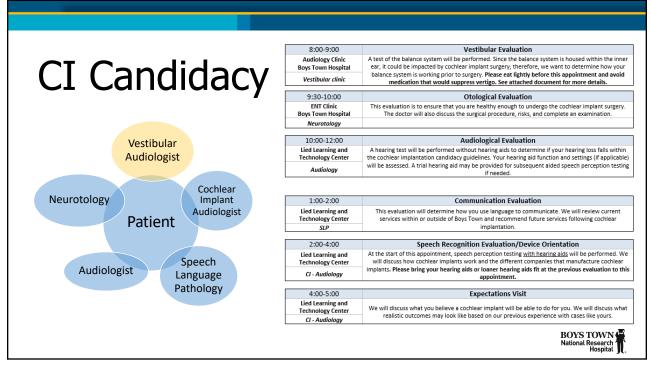
Rotary Chair

Goggle vHIT

Air/bone-Conducted cVEMP

Rotary Chair





Case: 33-month-old male

- · Normal hearing in the right ear
- Profound sensorineural hearing loss in the left ear
- Gross motor skill development:
 - Sat independently at 6 months
 - · Walked independently at 18 months
 - Parents expressed concern for delayed gross motor development,
 - No signs of dizziness or imbalance



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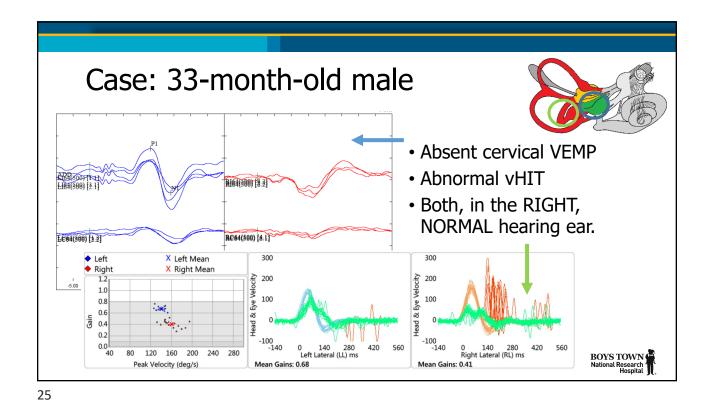
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Is this child at risk for vestibular loss?

YES

- o Degree of hearing loss
- Presence of gross motor delay
- o Parent concern



Case: 33-month-old male

- Counseling points:
 - The presence of vestibular loss can help account for delays in gross motor development (reassurance)
 - Consultation with vestibular rehabilitation
 - Vestibular loss not lateralizing with the hearing loss, is suggestive of congenital CMV
 - · Considerations for CI in the left ear:
 - risk of vestibular loss from implantation, which would result in bilateral vestibular loss.



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cCMV: Laterality of vestibular loss is **not** always with the hearing loss

TABLE 6 Correlation Between Laterality of Hearing Impairment and Laterality of Vestibular Disorders

Hearing Deficits	Vestibular Deficits					
	Bilateral Symmetric	Right	Left	Normal	Total	
Bilateral symmetric	13	9	4	2	28	
Right	3	3	(4)	0	10	
Left	8	0	2	0	10	
Normal		0	\bigcirc	2	4	
Total	25	12	11	4	52	

Data are expressed as number of children. There is a significant relationship between the laterality of the disorders (P = .03), but these are not correlated $(\kappa = 0.05)$.

(Bernard et al., 2015)

Only 7.7% of children (4/52) had normal vestibular function.

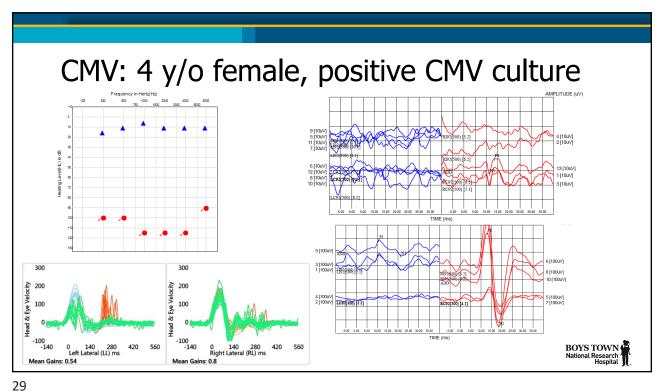


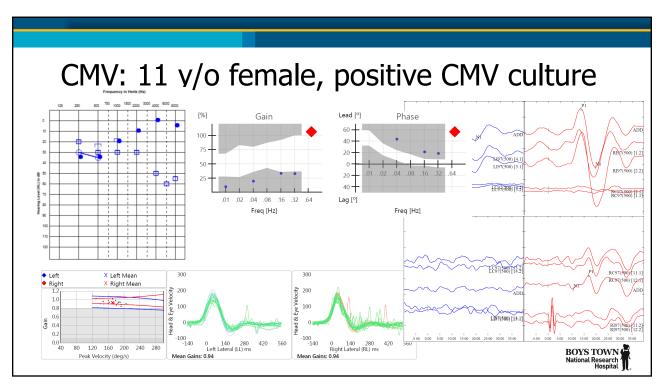
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A quick word about Cytomegalovirus (CMV)

- Laterality is **not** always with the hearing loss.
- More likely to have inner ear dysfunction if they have imaging lesions and if the infection occurs in the first trimester (Chebib 2022)
- More likely to have vestibular loss if hearing loss is present (Dhondt 2020)







Changes in Vestibular Function Following Pediatric Cochlear Implantation: a Prospective Study

Cleo Dhondt,¹ Leen Maes,^{2,3} Saartje Vanaudenaerde,³ Sarie Martens,² Lotte Rombaut,³ Ruth Van Hecke,² Romain Valette,⁴ Freya Swinnen,³ and Ingeborg Dhooge^{1,3}

Complete ipsilateral loss of function after implantation was seen in 5% (3/54)*

*however, all these children had CMV, suggesting that etiology of hearing loss and not the CI could have resulted in the vestibular loss



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The most common abnormalities following CI are reduced or absent vestibular evoked myogenic potential (VEMP) and caloric responses.

• HOWEVER:

- VEMP:
 - most investigations have used air-conduction stimuli.
 - air-bone gaps can occur following Cl^{4,5} causing reduced air-conducted VEMP response^{6,7}, thus inflating the estimations of vestibular loss following Cl.
 - Bone-conducted stimuli is preferred over air-conducted stimuli for VEMP testing to overcome the presence of post-CI air-bone gaps^{6,7}



Increased VEMP response rates using BCV

TABLE 3. Response rates for cervical and ocular VEMP to ACS and BCV stimuli

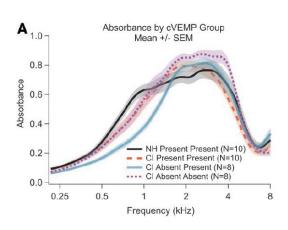
	Cervica	I VEMP	Ocular	VEMP
Group	ACS	BCV	ACS	BCV
Ears with NH (n = 10)	100%	100%	100%	100%
Ears with CI (n = 27)	41%	67%	15%	52%*
χ^2	10.41 0.001	4.41 0.036	22.51 <0.001	7.42 0.006

Significantly higher response rates compared with ACS (p = 0.004).

ACS, air-conducted stimuli; BCV, bone-conducted vibration; CI, cochlear implant; NH, normal hearing; VEMP, vestibular evoked myogenic potential.

Effect of Cochlear Implantation on Vestibular Evoked Myogenic Potentials and Wideband Acoustic Immittance

Gabrielle R. Merchant, Kyli M. Schulz, Bessie N. Patterson, Denis Fitzpatrick, and Kristen L. Janky



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The most common abnormalities following CI are reduced or absent vestibular evoked myogenic potential (VEMP) and caloric responses.

- HOWEVER:
 - Caloric:
 - Caloric testing is not recommended due to changes in temporal bone anatomy which can confound pre- and post-CI caloric test comparisons



Age Group	Semicircular Canal	Otolith	Other
6 m to 3 yrs	Rotary Chair	Bone-Conducted* (B-71) cervical VEMP	Bedside HIT, spontaneous nystagmus, post head shake
3 to 19 yrs	Rotary Chair or vHIT	Bone-Conducted* (B-71 or Mini- shaker) cervical and ocular VEMP	nystagmus, vibration-induced nystagmus
Adults	Rotary Chair or vHIT	Bone-Conducted* (<u>Mini-shaker</u>) cervical and ocular VEMP	

^{*}If bone conduction responses are absent, complete air conduction Tympanometry to be completed on all patients prior to testing

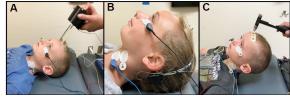


Bone Conduction Vibration Vestibular Evoked Myogenic Potential (VEMP) Testing: Reliability in Children, Adolescents, and Young Adults



Nicole, L. Greenwalt, ^{1,2} Jessie N. Patterson, ² Amanda I. Rodriguez, ^{2,3} Denis Fitzpatrick, ² Katherine R. Gordon, ² and Kristen L. Janky ²

• Objective: determine which BCV VEMP method (Mini-shaker, B-71, or impulse hammer) yields the highest response rates and reliability in a group of adults, adolescents, and children.

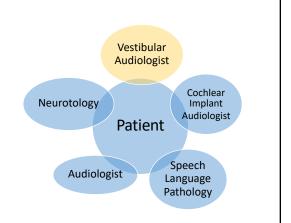


 All BCV methods resulted in consistent cervical and ocular VEMP responses with at least moderate reliability for all groups; EXCEPT for the B-71 ocular VEMP in adults.

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CI Candidacy

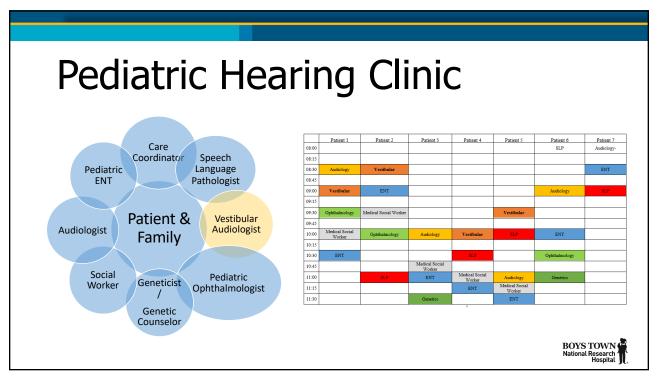
- Vestibular testing is NOT used for candidacy
- In RARE instances, can be part of discussions
- Awareness of vestibular loss
 - The presence of vestibular loss can help account for delays in gross motor development (reassurance)
 - · Consultation with vestibular rehabilitation
- Pre/post CI evaluations:
 - Consider vHIT, rotary chair and VEMP using BCV!

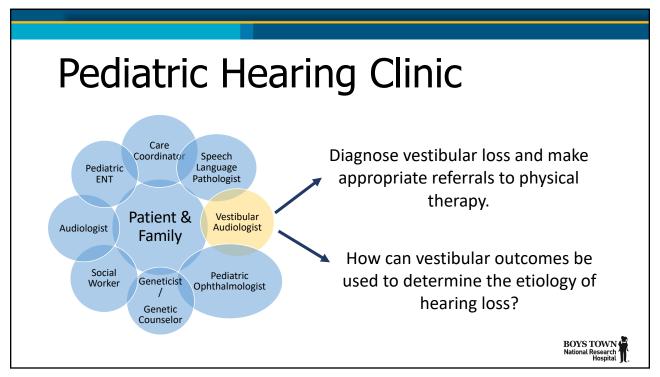


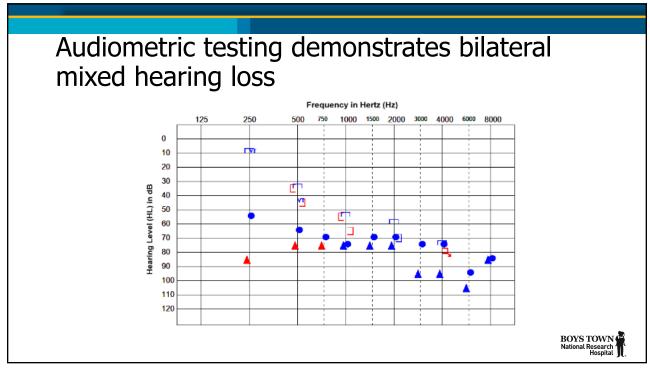


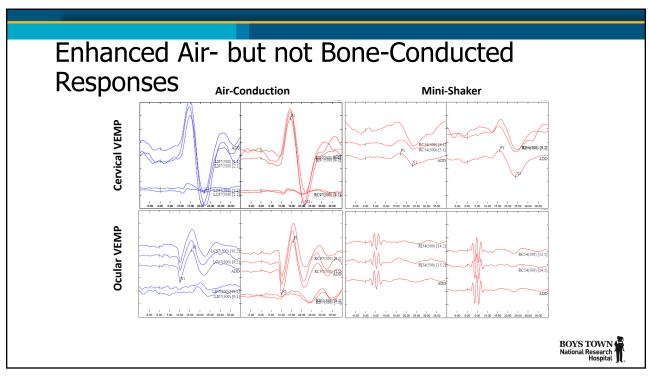
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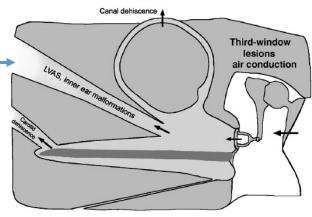




Large Vestibular Aqueduct Syndrome (LVAS) (represents one type of 3rd Window Disorder)

Vestibular aqueduct (VA) is enlarged if >0.9 mm at the midpoint or >1.9 mm at the operculum

LVAS is the most common inner ear malformation, affecting up to 15% of children with early-onset hearing loss

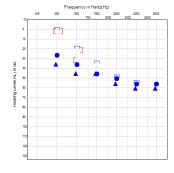


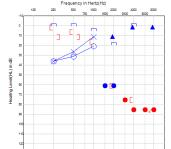
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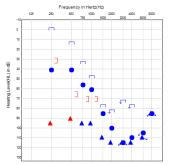
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Hearing loss with LVAS is progressive, so early diagnosis is critical

• Hearing loss type (i.e., conductive, sensorineural, or mixed⁴), configuration, and rate of progression is variable with LVAS



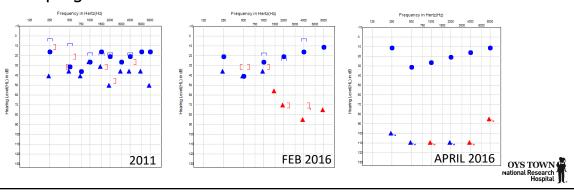




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Hearing loss with LVAS is progressive, so early diagnosis is critical

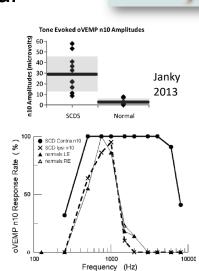
• Because children are unable to reliably report changes in auditory function, LVAS is not suspected until significant hearing loss progression has occurred.



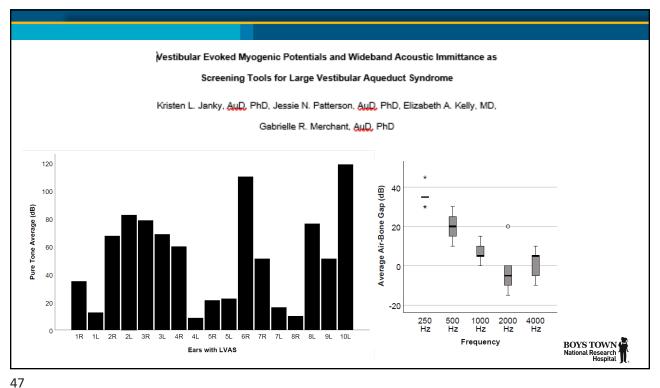
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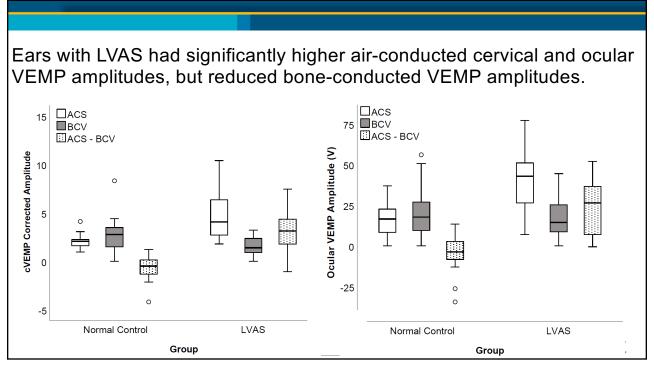
Primarily what we know about 3rd Window Disorders comes from superior canal dehiscence syndrome (SCDS)

- Low cervical VEMP thresholds
- High ocular VEMP amplitudes
- Broad frequency tuning (i.e., present VEMP responses at high frequencies)
- Making VEMP an excellent screening tool for SCDS⁸⁻¹¹.



Manzari 2013





Subtracting air-conducted amplitudes from boneconducted amplitudes was the best individual factor for differentiating control from LVAS ears

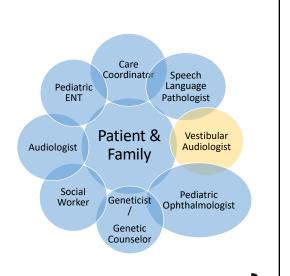
- 82% sensitivity and 80% specificity for cervical VEMP
- 88% sensitivity and 80% specificity for ocular VEMP.



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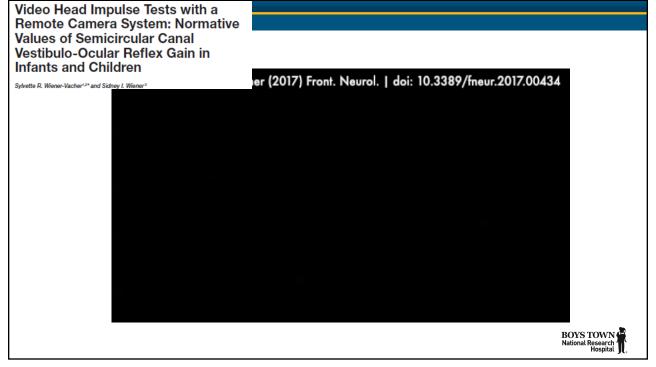
Pediatric Hearing Clinic

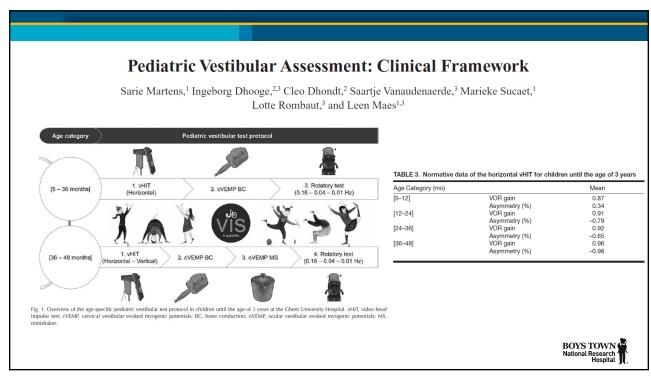
- Awareness of vestibular loss
 - The presence of vestibular loss can help account for delays in gross motor development (reassurance)
 - Consultation with vestibular rehabilitation
- Help in determination of hearing loss etiology
 - CMV
 - LVAS
 - Usher Syndrome



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How can this be used as a clinical tool?



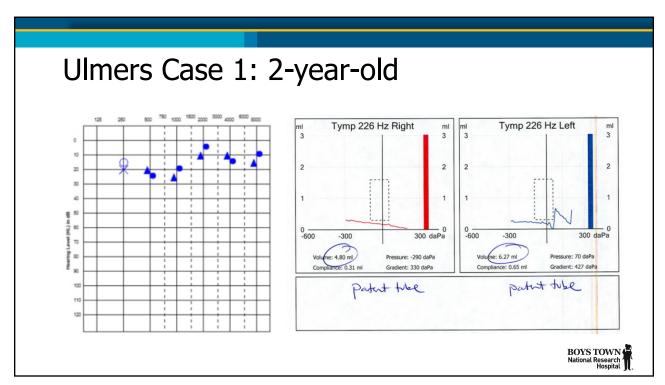
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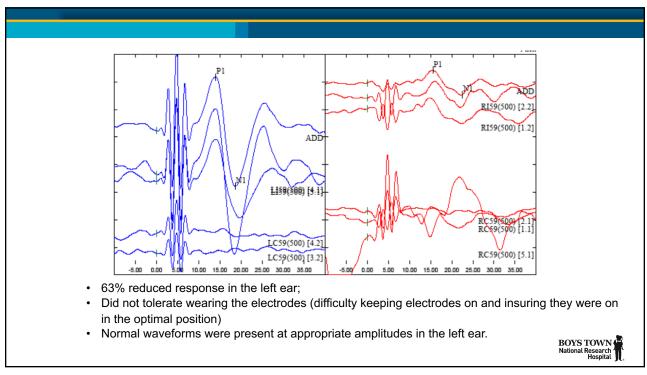
Ulmers Case 1: 2-year-old

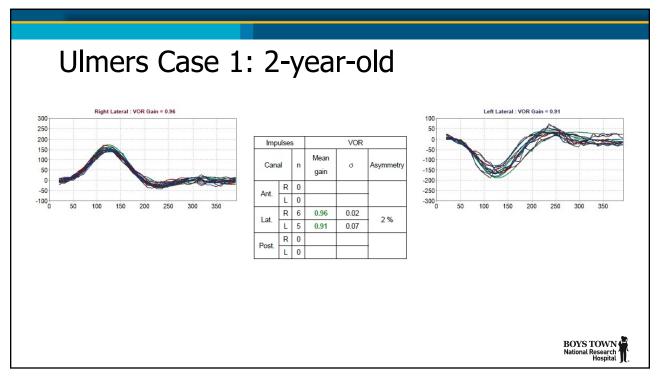
- · Chief Complaint: Imbalance, runs into walls, falls frequently
- She has been receiving PT services twice per week.
- Mild hearing loss in the left ear; she has been utilizing amplification since she was 3 months.
- Balance is better when she is wearing her hearing aid; however, audiometric testing today revealed
 that hearing in the left ear has improved within the normal range, and the hearing aid is no longer
 necessary.
- Previous vestibular testing noted normal rotary chair and asymmetric VEMP responses, with a 74% reduced response in the left ear.
- She is being reassessed today due to continued issues with imbalance and to determine if there has been any change in vestibular function.
- · Mom reports that she has a difficult time seeing things that are not in her central vision.
- · PT had also suggested possible ocular motor deficit.



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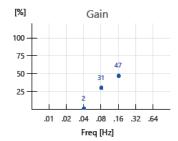
CASE 2 – 8-month-old female

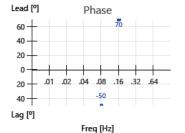
- Chief Complaint: Patient is currently being followed for bilateral profound sensorineural hearing loss, secondary to bilateral cochlear nerve deficiency.
 - · born full-term
 - birth history is significant for meningomyelocele.
 - She underwent went spinal cord untethering at age 5 months.
 - There were perinatal concerns for neural tube defect.
 - · She did not pass her newborn hearing screening
 - Follow-up ABRs at age 2 and 5 months suggest bilateral profound sensorineural hearing loss.

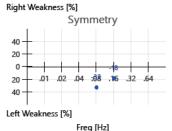


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CASE – 8-month female

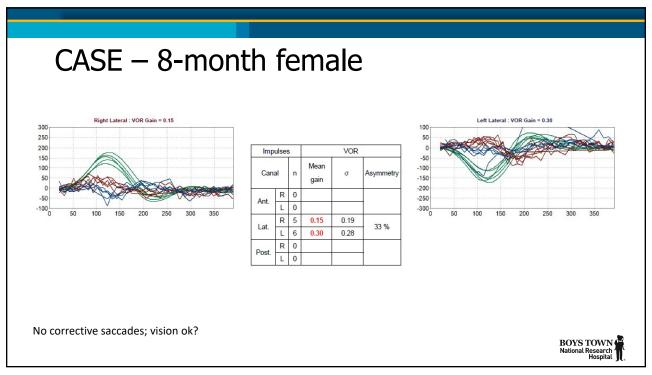


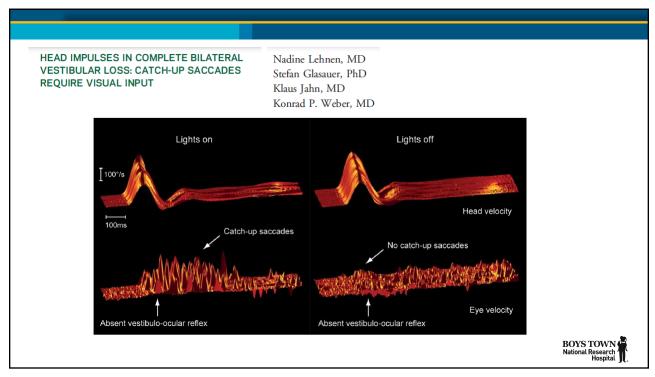




Family from 4 hours away; wanted to confirm for moving forward







How can this be used as a clinical tool?

- Helpful when:
 - Patient Intolerance of cVEMP and Rotary Chair
 - Confirmatory
 - Confirmatory with conflicting test findings
- If/when FDA approved, first tier assessment



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Cleo Dhondt, MSc ©; 1	Ingeborg Dhooge, PhD,	MD 😊; Leen Maes, PhD	D				
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		Across the Different Co	Age Categorienducted		Success Ra	ate (%)	
Age Category	No. of Subjects	Across the Different Co	Age Categorienducted	cVEMP	Success Ra	ate (%) Rotatory Test	
Age Category 5 months–1 year	No. of Subjects	Across the Different Co Median Age (mo) 7.0	Age Categorie nducted vHIT*	cVEMP	Success Ra	Rotatory Test	
Age Category 5 months–1 year 1 year–2 years	No. of Subjects	Across the Different Co Median Age (mo) 7.0 17.0	Age Categorie nducted vHIT* 72.2 100.0†	cVEMP 100.0 100.0	Success Ra	Rotatory Test 88.9 62.5	
Age Category 5 months–1 year 1 year–2 years 2 years–3 years	No. of Subjects 18 8 8	Across the Different a Co Median Age (mo) 7.0 17.0 29.0	Age Categorie nducted vHIT* 72.2 100.0† 85.7†	cVEMP 100.0 100.0 75.0	Success Ra	Rotatory Test 88.9 62.5 50.0	
Age Category 5 months–1 year 1 year–2 years 2 years–3 years Total group < 3 years	No. of Subjects 18 8 8 34	Across the Different a Co Median Age (mo) 7.0 17.0 29.0 10.5	Age Categorie nducted VHIT* 72.2 100.0 [†] 85.7 [†] 81.3	cVEMP 100.0 100.0 75.0 94.1	Success Ri	Rotatory Test 88.9 62.5 50.0 73.5	Caloric Test
Age Category 5 months–1 year 1 year–2 years 2 years–3 years Total group < 3 years 3 years–4 years	No. of Subjects 18 8 8 34 8	Across the Different a Co Median Age (mo) 7.0 17.0 29.0 10.5 44.0	Age Categorie nducted VHIT* 72.2 100.0 [†] 85.7 [†] 81.3 100.0	cVEMP 100.0 100.0 75.0 94.1 100.0	Success Ri oVEMP	Rotatory Test 88.9 62.5 50.0 73.5	Caloric Test

cVEMP or vHIT?

Using Functional Outcomes to Predict Vestibular Loss in Children

Kristen L. Janky, Megan LA. Thomas, Jessie Patterson, and Diane Givens

Boys Town National Research Hospital, Department of Audiology, Omaha, Nebraska

- Average horizontal canal vHIT gain was a significant predictor of all functional outcomes [standing balance test; passive DVA; BOT-2] while neither corrected cVEMP amplitude nor oVEMP amplitude predicted performance.
- Average horizontal canal vHIT best predicts functional performance; if using a tiered approach, horizontal canal vHIT should be completed first.



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Vestibular Clinic





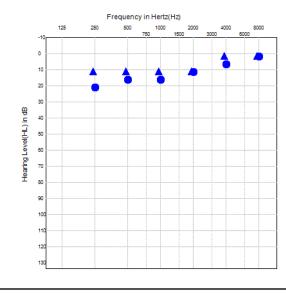
Case Study: Dizzy 9-year-old female

- Chief complaint:
 - spontaneous onset room spinning dizziness.
 - Symptoms began when the family was flying home from vacation, during descent. Patient also reports headache when she does not have her head down to the right. Dizziness is constant.
 - Aggravating factors include sitting up and standing.
 - Relieving factors include lying on stomach or with head down to the right.
 - Associating symptoms include emesis and lightheadedness.
 - Parents report she is clumsy when walking and veers right.



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Case Study: Dizzy 9-year-old female

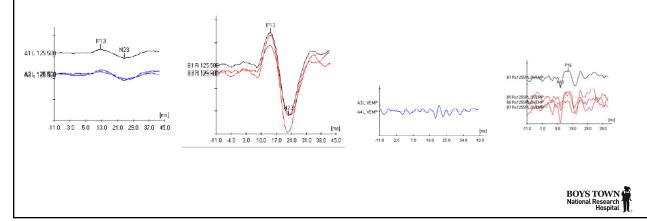


- Patient needed to lay on the floor during testing.
- Normal tympanograms
 - ECVs were 0.9
 - 125 dB SPL could be used for VEMPs



Case Study: Dizzy 9-year-old female

- Reduced cVEMP left, present right
- Absent oVEMP left, present right



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Case Study: Dizzy 9-year-old female

- MRI
 - Normal
- VNG
 - Normal Ocular Motor Battery:
 - saccades, smooth pursuit and gaze with fixation
 - Gaze without fixation
 - Center: RBN 19 d/s
 - Right: RBN 12 d/s
 - Left: RBN 9 d/s
 - Refused caloric testing; vHIT is not present at our satellite location
- INTERPRETATION
 - Left Involvement: absent left VEMPs and RBN



Vestibular Neuritis?

- Inflammation of the vestibular nerve; most often superior branch; however, can affect either/both
- Acute onset room spinning dizziness
- No associated hearing loss
- Symptoms are worse at their onset and improve over the course of days to weeks
- Represents 1 5% of pediatric vertigo
- Children recover more quickly; children < 15 years were found to recover more quickly (Brodsky 2016)



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CASE – MALE 31 months

- Chief Complaint: delayed gross motor development.
 - born full-term with no birth complications
 - passed his newborn hearing screening.
 - bilateral myringotomy with tube placement at 8 months and again 5 months ago.
 - rolled at 8-9 months of age, sat independently at 11 months, and began walking at about 1.5 years of age after starting physical therapy.
 - Currently enrolled in physical therapy.
 - Continues to struggle with balance and occasionally falls, especially on uneven terrain; has a medical history significant for a tibial fracture that occurred when he scooted off a bed at 13 months.

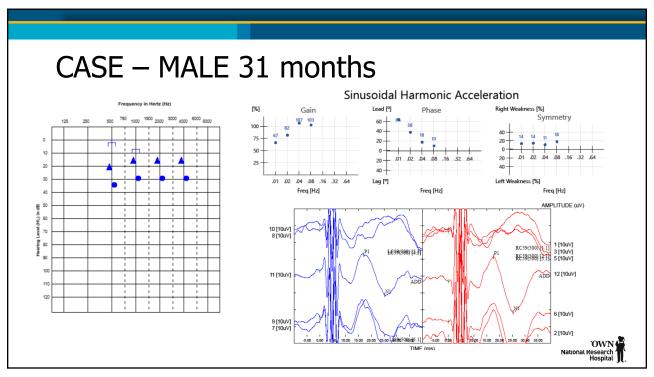


CASE - MALE 31 months

- Previous Testing: age 20 months
 - Patent tube left; negative pressure right.
 - Phase lead on Rotary Chair and normal cVEMP testing. Thus, unilateral peripheral vestibular system involvement could not be ruled out at that time.
 - Mom was counseled that recurring otitis media and the presence of a broken leg can be contributing factors to delayed gross motor development and at minimum a unilateral vestibular loss is present; however, the likelihood of this is low, particularly if hearing is normal.



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There are several theories suggesting that middle ear dysfunction directly affects the vestibular system.

- 1. an ionic transfer via the round window, which causes a change in endolymph composition (Pazdro-Zastawny et al., 2018). This transfer results in an indirect change to the kinocilia and stereocilia in the inner ear, which could result in clumsiness and/or imbalance.
- 2. Abnormal vestibular function could be the result of an infection in the middle ear space which causes inflammation of the labyrinth; the infection could travel via the round window to the inner ear resulting in inflammation (Goycoolea et al., 1987).
- 3. The presence of significant negative pressure in the middle ear space which causes involuntary movement of inner ear fluids (Kobrak, et al., 1935).



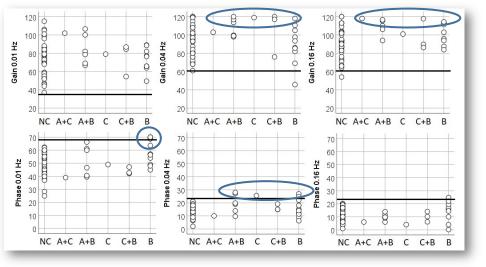
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Effects of middle ear status on rotary chair outcomes in children

- The purpose of this study is to evaluate how middle ear status affects rotary chair outcomes in children 6 to 77 months.
- 21 children with middle ear dysfunction (mean: 30 months, 11 males) and 36 control children with normal middle ear function (mean: 33 months, 19 males) participated. All participants completed tympanometry, wideband acoustic immittance, and rotary chair testing at 0.01, 0.04, and 0.16 Hz.



Effects of middle ear status on rotary chair outcomes in children



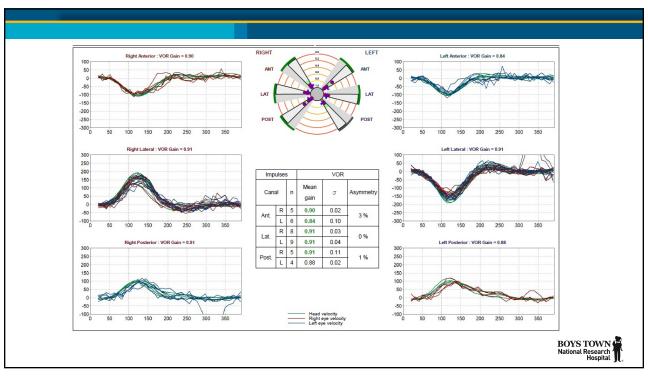
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CASE MALE 31 months

- Rotary chair in the presence of middle ear dysfunction is valid; however, isolated phase abnormalities can be present that are not related to middle ear dysfunction severity.
- Repeat rotary chair and VEMP?
- vHIT?





CASE MALE 31 months

- 1. Continue with ESU physical therapy services to address balance concerns.
- 2. Normal results today suggest the vestibular system is functioning appropriately. As such, repeat vestibular testing is no longer indicated. Repeat if concerns arise in the future.
- 3. Mom was counseled that balance is achieved with a combination of vestibular, vision and somatosensation. If balance issues persist, vision assessment would be recommended.



Vestibular Clinic

- · Vestibular Migraine
- Vestibular Neuritis
- OME
- SNHL
 - Cochleovestibular anomaly
 - CMV
- Concussion
- Psychogenic
- 3PD
- BPPV
- · Meniere's disease
- Orthostasis
- CNS [seizure, CPA tumor, autoimmune, syncope]
- SSCD

*Manuel of Pediatric Vestibular Disorders

Neurotology

Pharmacy

Vestibular

Audiologist

Patient



Physical

Therapist

Audiologist

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Objectives, revisited

After this course, participants will be able to:

- Identify children who are candidates for vestibular testing
- List causes of pediatric vestibular loss and dizziness
- List practitioner types needed for multidisciplinary assessment of pediatric vestibular loss



1. Identify children who are candidates for vestibular testing

- · Children with:
 - Dizziness
 - Hearing loss (> 66 dB)
 - Gross motor delay/imbalance





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2. List causes of pediatric vestibular loss and dizziness

Hearing Loss (severe-to-profound)

- Cytomegalovirus (CMV) infection
- Auditory Neuropathy (show decline in function over time)
- Waardenburg Syndrome
- Rubella
- Pendred Syndrome
- Malformations
- Meningitis
- Ototoxicity
- Usher Syndrome

Dizziness

- Vestibular Migraine
- Vestibular Neuritis
- OME
- Concussion
- Psychogenic
- 3PD
- BPPV
- · Meniere's disease
- Orthostasis
- CNS SSCD



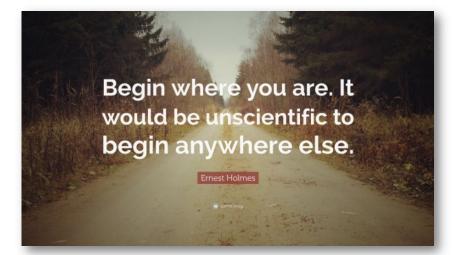
3. List practitioner types needed for multi-disciplinary assessment of pediatric vestibular loss

- · Depend on the focus of the clinic
 - Cochlear Implant
 - · Pediatric Hearing Clinic
 - · Dizzy Clinic
 - Neurotology
 - · Audiology
 - · Physical Therapy
 - Ophthalmology
 - Social work
 - SLP
 - Genetics
 - Neurology
 - Psychiatry



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Thinking about starting a pediatric vestibular program?



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