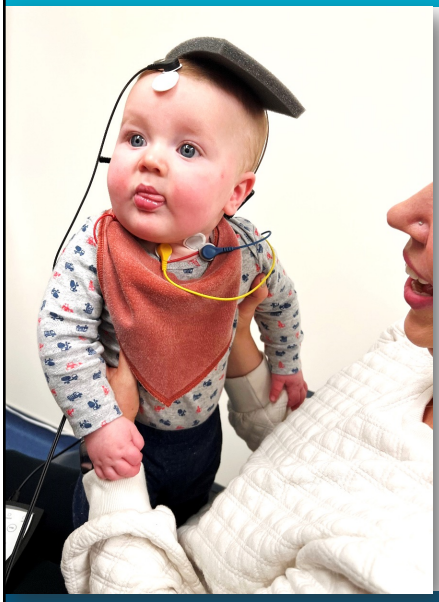

Newborn Vestibular Screening and the Consequences of Pediatric Vestibular Loss

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1

Objectives

After this course, participants will be able to:

- Describe the process of newborn vestibular screening
- List consequences of vestibular loss in children
- List areas of future research needed

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Three Years of Vestibular Infant Screening in Infants With Sensorineural Hearing Loss

Sarie Martens, MSc,[a] Ingeborg Dhooge, MD, PhD,[b,c] Cleo Dhondt, MSc,[b] Saartje Vanaudenaerde, MSc,[c] Marieke Sucaet, MSc,[a] Helen Van Hoecke, MD, PhD,[b,c] Els De Leenheer, MD, PhD,[b,c] Lotte Rombaut, MSc,[c] An Boudewyns, MD, PhD,[d] Christian Desloovere, MD, PhD,[e] Anne-Sophie Vinck, MD,[f] Sebastien Janssens de Varebeke, MD, PhD,[g] Dominique Verschueren, MD,[h] Margriet Verstreken, MD,[i] Ina Foulon, MD, PhD,[j] Charlotte Staelens, BSc,[k] Claudia De Valck, MD, PhD,[l] Robbe Calcoen, BSc,[m] Nele Lemkens, MD,[n] Okan Öz, BSc,[o] Mieke De Bock, MSc,[p] Lisa Haverbeke, MSc,[q] Christoph Verhoye, MD,[r] Frank Declau, MD, PhD,[s] Benoit Devroede, MD,[t] Glen Forton, MD, PhD,[u] Naima Deggouj, MD, PhD,[v] Leen Maes, PhD[a,c]

To cite: Martens S, Dhooge I, Dhondt C, et al. Three Years of Vestibular Infant Screening in Infants With Sensorineural Hearing Loss. *Pediatrics*. 2022;150(1):e2021055340

Methodological aspects of testing vestibular evoked myogenic potentials in infants at universal hearing screening program

Luca Verrecchia[1,2*], Niki Karpeta[1,2], Magnus Westin[1], Ann Johansson[1], Sonny Aldenklint[1], Krister Brantberg[1,3] & Maoli Duan[1,2]

SCIENTIFIC REPORTS
nature research

Comment > Pediatrics. 2022 Jul 1;150(1):e2022056986. doi: 10.1542/peds.2022-056986.

The Feasibility of Performing Vestibular Newborn Screening

Kristen L Janky [1], Christine Yoshinaga-Itano [2]

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3

Case 1: 9-month-old female

- Profound sensorineural hearing loss in the right ear
- Positive CMV testing by buccal swab; did a trial with valganciclovir
- Concerns with gross motor delay
  - She began sitting at age 9 months
- Interested in CI; therefore, imaging scheduled

Should vestibular testing be done?
What will it add to the clinical care of this patient?

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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 ~ 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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Vestibular Testing can be completed in infants and young children

Age birth to 3

- cVEMP
- Rotary Chair
- vHIT with remote camera

Age 3+

- Ocular VEMP
- Traditional vHIT
- Caloric testing




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< 12 mos, cVEMP most successful, > 12 months, remote vHIT most successful

TABLE 2. Success rate of the vestibular test protocol

	Success Rate (Number of Subjects With Successful Results) (%)						
	vHIT		VEMP		Rotatory Test		
Age Category (mo)	Hor SCC	Vert SCC	cVEMP	oVEMP	0.16 Hz	0.04 Hz	0.01 Hz
[5–12]	78.9%		89.5%		68.4%	76.3%	44.7%
n = 38	(30)		(34)		(26)	(29)	(17)
[12–24]	91.9%		81.1%		62.2%	56.8%	35.1%
n = 37	(34)		(30)		(23)	(21)	(13)
[24–36]	97.0%		84.8%		78.8%	72.7%	63.6%
n = 33	(32)		(28)		(26)	(24)	(21)
[36–48]	100.0%	60.0%	96.0%	92.0%	92.0%	96.0%	92.0%
n = 25	(25)	(15)	(24)	(23)	(23)	(24)	(23)
[5–48]	91.0%		87.2%		73.7%	73.7%	55.6%
n = 133	(121)		(116)		(98)	(98)	(74)

%, percentage of subjects with successful results; cVEMP, cervical Vestibular Evoked Myogenic Potentials; Hor, horizontal; n, number of subjects; oVEMP, ocular Vestibular Evoked Myogenic Potentials.; SCC, semicircular canals; VEMP, Vestibular Evoked Myogenic Potentials; Vert, vertical; vHIT, video Head Impulse Test.

Pediatric Vestibular Assessment: Clinical Framework

Sarie Martens,[1] Ingeborg Dhooge,[2,3] Cleo Dhondt,[2] Saartje Vanaudenaerde,[3] Marieke Sucaet,[1] Lotte Rombaut,[3] and Leen Maes[1,3]

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cVEMP deemed most feasible method for newborn vestibular screening due to overlapping use of ABR equipment

- ABR devices
  - are often used for NBHS fails
  - can be used to complete VEMP testing
  - cVEMP can be measured as early as days after birth
  - Thus, this represents a practical means for screening for vestibular loss (high feasibility).

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Children who did not pass their NBHS received a "vestibular screen"

- cVEMP using a **B-71**; At least 2 trials were recorded on each side to check waveform reproducibility.
  - Around the age of 6 months (Martens et al., 2022)
  - 2.3 ± 1.9 months (Verrechia et al., 2019)
  - Bone conduction used due to the high rate of otitis media in this age range of children

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Abnormal VEMP results were found in 13.8% (35 of 254) of the infants.

- Abnormal results were found significantly more often in:
  - unilateral or bilateral **severe to profound hearing loss** .
  - **early-onset hearing loss** (i.e., meningitis and cCMV)
  - **cCMV positive**
- **Etiology**:
  - Meningitis, Syndromic Hearing Loss, Hearing Loss caused by TORCHES infections, Cochleovestibular anomalies
- No significant effect of perinatal factors
  - prematurity, low birth weight, hospitalization > 5 days at NICU

Three Years of Vestibular Infant Screening in Infants With Sensorineural Hearing Loss

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What is our response at Boys Town to these findings?

11

Are any children missed by using cVEMP?

- YES
  - 4.6% of children are missed using cVEMP alone
  - 1.5% of children are missed using vHIT alone
  - Especially for children with CMV and cochleovestibular anomalies

Vestibular Infant Screening–Flanders: What is the Most Appropriate Vestibular Screening Tool in Hearing-Impaired Children?

Sarie Martens,[1,4] Leen Maes,[1,2,4] Cleo Dhondt,[3] Saartje Vanaudenaerde,[2] Marieke Sucaet,[1] Els De Leenheer,[2,3] Helen Van Hoecke,[2,3] Ruth Van Hecke,[1] Lotte Rombaut,[2] and Ingeborg Dhooge[2,3]

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Case Study 2 – 12 months

- **Primary Complaint**: mild hearing loss in the left ear and mild hearing loss of low- and mid-frequencies, rising to borderline normal hearing in the right ear. Hearing loss was confirmed via diagnostic auditory brainstem response (ABR) testing
- Mom expressed **concern for delay in gross motor milestones**; progress has slowed since fitting of a helmet for plagiocephaly.
  - Sits independently for at least 20 seconds before falling over.
  - Locomotes primarily by rolling.
- Other Health History:
  - Torticollis, Plagiocephaly, Congenital Brachycephaly, Cornelia de Lange syndrome, and 20 day stay in the NICU due to intra uterine growth restriction
- Is receiving weekly physical therapy services for trunk strengthening.

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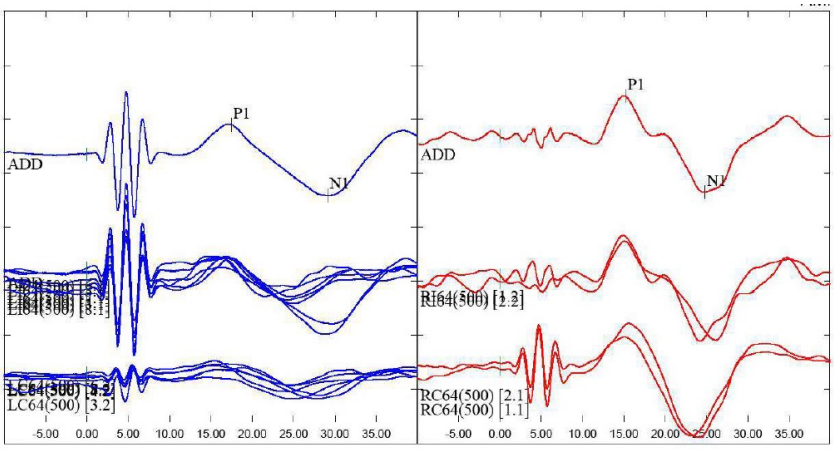
Case 2 – cervical VEMP



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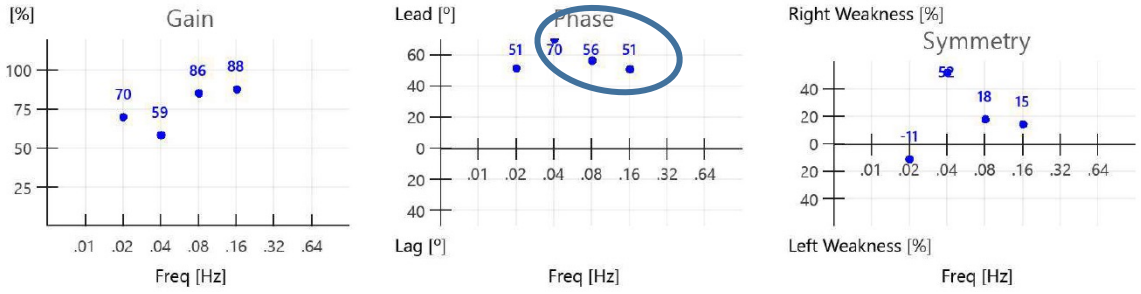
Case 2 – age 20 months

- At 20 months he is beginning to stand independently and is currently walking with assistance. He is receiving some physical therapy services



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Individuals with Cornelia de Lange can have a dysplastic vestibule

- CT abnormalities (n = 20) were correlated with audiometric outcomes; however, vestibular findings were not reported.
- "a bizarre-shaped vestibule with a focal protrusion at the posteroinferior aspect like a "tag" (75%)"

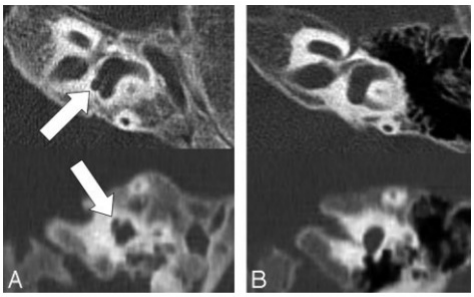

Fig 3. Axial section of a vestibule from a 2-year-old girl with Cornelia de Lange syndrome. Note the abnormal focal protrusion at the posteroinferior aspect of the vestibule (*arrows*) in the patient with Cornelia de Lange syndrome (*A*) as compared with a 2-year-old control with normal hearing (*B*).

J. Kim
E.Y. Kim
J.S. Lee
W.S. Lee
H.N. Kim

Temporal Bone CT Findings in Cornelia de Lange Syndrome AJNR Am J Neuroradiol 29:569–73 |

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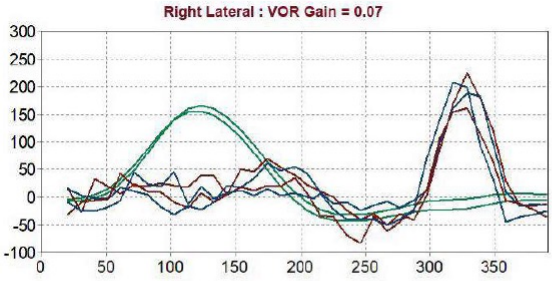
Case 2 – age 20 months



Impulses			VOR		
Canal		n	Mean gain	σ	Asymmetry
Ant.	R	0			
	L	0			
Lat.	R	2	0.07	0.12	64 %
	L	3	0.31	0.02	
Post.	R	0			
	L	0			

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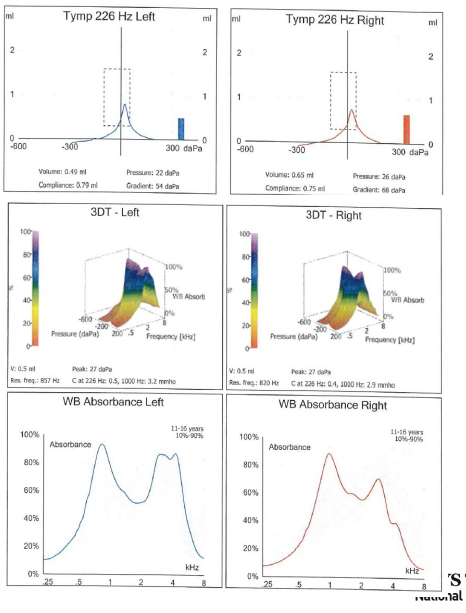
Case 3 – age 16 years

- Female with Down syndrome
- Mild hearing loss, bilaterally



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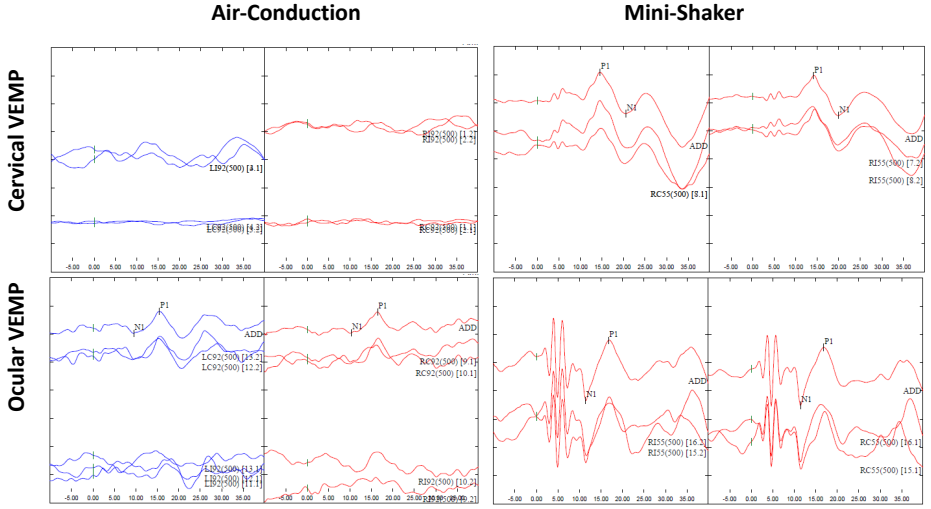
Case 3 – Absent air-conducted cervical VEMP



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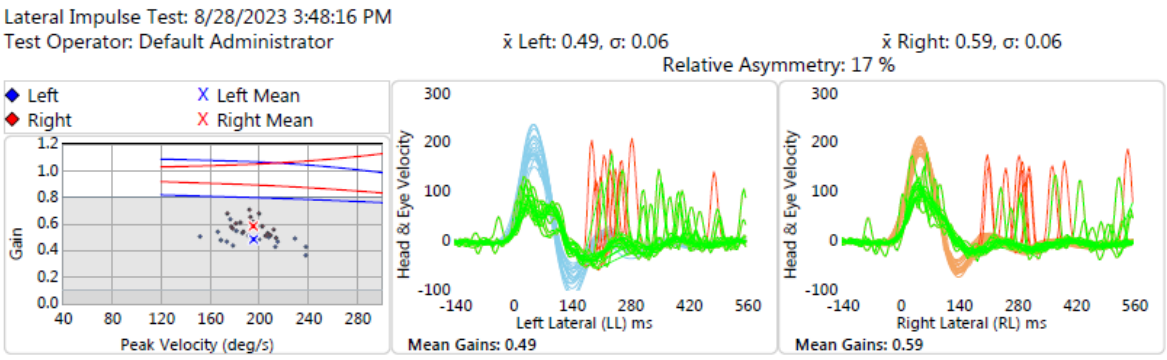
Case 3 – Bilateral Vestibular Loss



- Also verified on Ulmers

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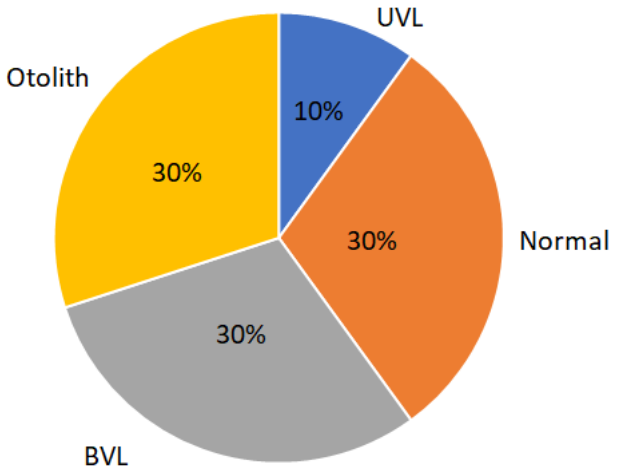
70% of individuals with Down syndrome demonstrated some degree of vestibular loss



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There was a higher percentage of VEMP responses using BCV compared to ACS

	cVEMP		oVEMP	
	ACS	BCV	ACS	BCV
Control	100%	100%	100%	100%
DS	22%	85%	28%	55%

This is attributed to the presence of middle ear dysfunction. Further analyses are planned using WAI to explain the differences in VEMP responses using ACS vs BCV.

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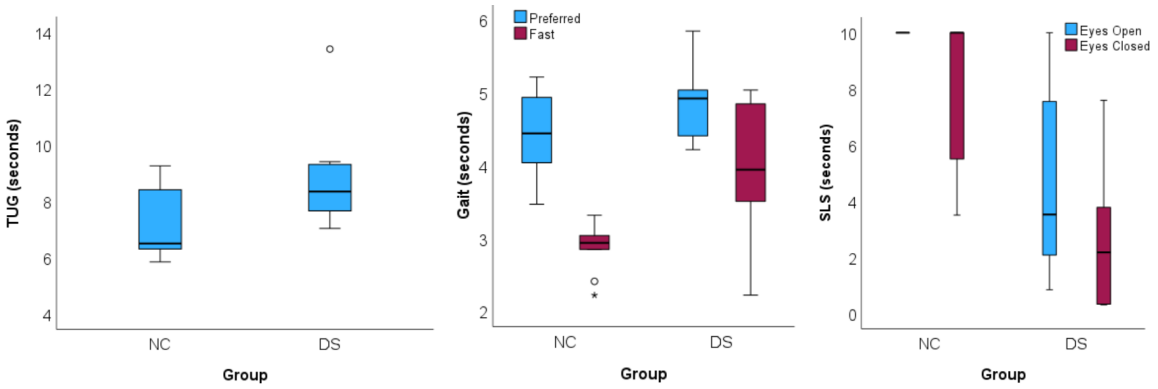
The normal control group performed significantly better on all balance measures, except preferred gait.



Balance ability significantly correlated with degree of vestibular loss.

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Are any children missed by using cVEMP?

- YES
- These data suggest that
  - The tests are complimentary
  - If 1 is normal, and additional concerns are present, keeping going
  - Some degree of follow-up is needed
- If I must pick one test, depends on symmetry of hearing
  - If asymmetrical: cVEMP
  - If symmetrical + gross motor delay: rotary chair
  - Best if I can get both

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Is the presence of gross motor delay enough?

- NO
  - Gross motor delay can differentiate children with bilateral vestibular loss from normal but does not differentiate those with unilateral vestibular loss.
  - However, it is a great marker for those whose hearing loss does not follow the typical "rules"

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Do all children with SNHL need to be screened?

- It seems reasonable that vestibular testing be recommended in the following groups of children:
  1. Children with **severe to profound sensorineural hearing loss**, which justifies testing all children prior to cochlear implantation;
  2. Children who pass their newborn hearing screening and **acquire hearing loss or have progressive hearing loss**; and
  3. Children with any degree of hearing loss and **concomitant gross motor delay** due to the known association between vestibular loss and gross motor function.

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Added inclusion of children with ANSD

- Vestibular involvement in ANSD occurs in 42 – 91% of patients; with a higher likelihood of vestibular loss if ANSD occurs post-lingually (Nash, 2015; El-Badry 2018; Laurent 2022; Hu 2020)
- Vestibular abnormalities have been related to vestibular or brainstem radiological alterations (Laurent et al., 2022)
  - Vestibular dysplasia, vestibular anomalies and absence of SSCs
  - Children with canal areflexia all demonstrated delayed gross motor development and benefited from PT
  - Bilateral vestibular involvement with unilateral ANSD cochlear nerve deficit

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Pediatric Vestibular Loss, Clinical recommendation:

- Screen all kids with **unilateral/bilateral severe-to-profound hearing loss** and **acquired hearing** loss secondary to meningitis, CMV, any cochleovestibular anomalies, or unilateral ANSD.
- Screening will occur with cVEMP and/or rotary chair.
  - Prioritize VEMP because you can localize side involved
  - Eventually transition to remote-vHIT
- Parents given information sheet
- For any child that fails, at minimum:
  - Yearly PT evaluation
  - Follow-up assessment at age 3 – 4 for additional testing (cVEMP, oVEMP, vHIT/RC, gross motor screen) and additional PT evaluation.

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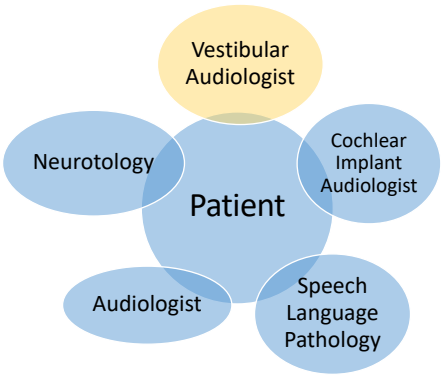
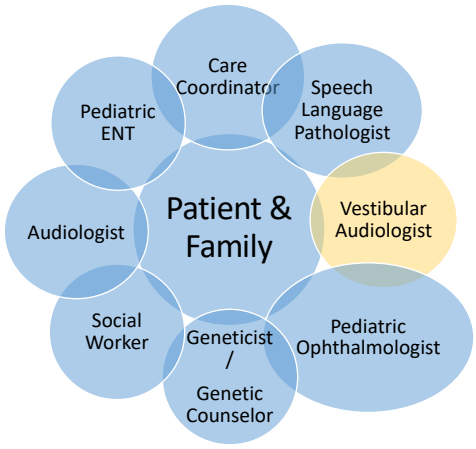
Pediatric Vestibular Testing

1. Cochlear Implant Candidacy



2. Pediatric Hearing Clinic



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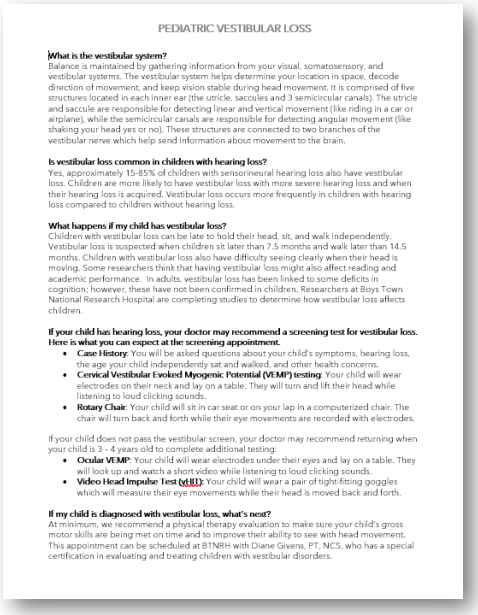

PEDIATRIC VESTIBULAR LOSS

What is the vestibular system?
Balance is maintained by gathering information from your visual, somatosensory, and vestibular systems. The vestibular system helps determine your location in space, decode direction of movement, and keep vision stable during head movement. It is comprised of five structures located in each inner ear (the utricle, saccules and 3 semicircular canals). The utricle and saccule are responsible for detecting linear and vertical movement (like riding in a car or airplane), while the semicircular canals are responsible for detecting angular movement (like shaking your head yes or no). These structures are connected to two branches of the vestibular nerve which help send information about movement to the brain.

Is vestibular loss common in children with hearing loss?
Yes, approximately 15-85% of children with sensorineural hearing loss also have vestibular loss. Children are more likely to have vestibular loss with more severe hearing loss and when their hearing loss is acquired. Vestibular loss occurs more frequently in children with hearing loss compared to children without hearing loss.

What happens if my child has vestibular loss?
Children with vestibular loss can be late to hold their head, sit, and walk independently. Vestibular loss is suspected when children sit later than 7.5 months and walk later than 14.5 months. Children with vestibular loss also have difficulty seeing clearly when their head is moving. Some researchers think that having vestibular loss might also affect reading and academic performance. In adults, vestibular loss has been linked to some deficits in cognition; however, these have not been confirmed in children. Researchers at Boys Town National Research Hospital are completing studies to determine how vestibular loss affects children.

If your child has hearing loss, your doctor may recommend a screening test for vestibular loss. Here is what you can expect at the screening appointment.

- **Case History**: You will be asked questions about your child's symptoms, hearing loss, the age your child independently sat and walked, and other health concerns.
- **Cervical Vestibular Evoked Myogenic Potential (VEMP) testing**: Your child will wear electrodes on their neck and lay on a table. They will turn and lift their head while listening to loud clicking sounds.
- **Rotary Chair**: Your child will sit in car seat or on your lap in a computerized chair. The chair will turn back and forth while their eye movements are recorded with electrodes.

If your child does not pass the vestibular screen, your doctor may recommend returning when your child is 3 – 4 years old to complete additional testing:

- **Ocular VEMP**: Your child will wear electrodes under their eyes and lay on a table. They will look up and watch a short video while listening to loud clicking sounds.
- **Video Head Impulse Test (vHIT)**: Your child will wear a pair of tight-fitting goggles which will measure their eye movements while their head is moved back and forth.

If my child is diagnosed with vestibular loss, what's next?
At minimum, we recommend a physical therapy evaluation to make sure your child's gross motor skills are being met on time and to improve their ability to see with head movement. This appointment can be scheduled at BTNRH with Diane Givens, PT, NCS, who has a special certification in evaluating and treating children with vestibular disorders.

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How to Complete VEMP Testing in Children less than 5 years old

Pediatric Vestibular Assessment: Clinical Framework

Sarie Martens,[1] Ingeborg Dhooge,[2,3] Cleo Dhondt,[2] Saartje Vanaudenaerde,[3] Marieke Sucaet,[1] Lotte Rombaut,[3] and Leen Maes[1,3]

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Important Considerations

- Use of cVEMP to screen for vestibular loss is feasible
- A play partner is essential!
- Use of EMG monitoring needs to be integrated into the same electrode due to small anatomy
- Would be helpful to know the minimum number of trials needed to obtain a reliable response

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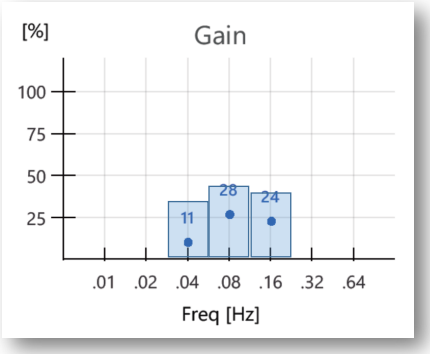
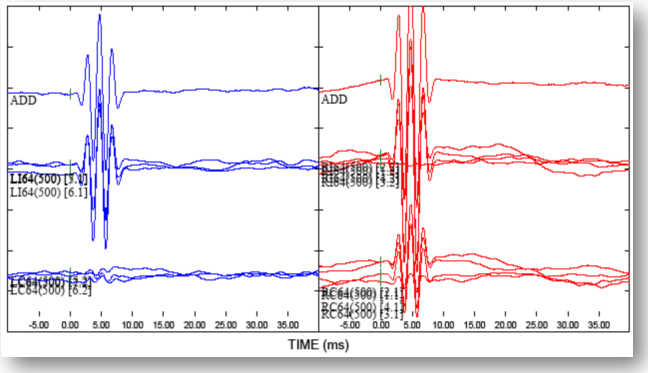
Case 1: 9-month-old female
Profound sensorineural hearing loss in the right ear





Should vestibular testing be done?
What will it add to the clinical care of this patient?

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- Most reported effects of vestibular loss involve gross motor and dynamic visual acuity deficits
- **What does this mean for children and families?**

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Action Items

1. Increase the number of qualified practitioners who assess and treat pediatric vestibular loss.
   - At minimum, have at least one qualified center in every state or geographic region that can complete pediatric vestibular testing and rehabilitation
2. Characterize the full spectrum effects of pediatric vestibular loss.
   - Impact of varying degrees of vestibular loss (unilateral, otolith only, bilateral) (Van Hecke et al., 2021)
3. Investigating the effectiveness of vestibular rehabilitation in remediating these functional effects in children

Defining the consequences of vestibular loss and the effectiveness of treatment provides further justification for the importance of early diagnosis and rehabilitation

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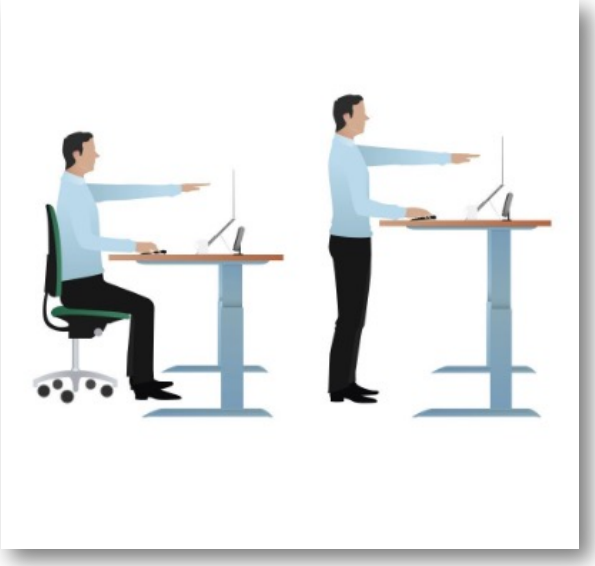
BREAK

Five Times
Sit to Stand



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Long-term Goals

- Define the long-term effects of pediatric vestibular loss across the spectrum of severity

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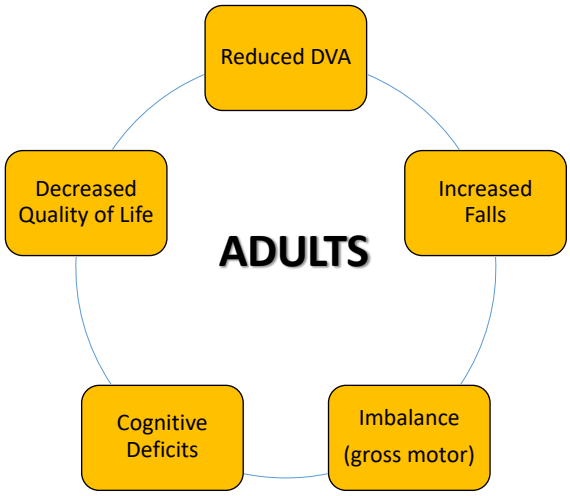
ADULTS are significantly affected by vestibular loss



Do children suffer from the same effects of vestibular loss as adults?

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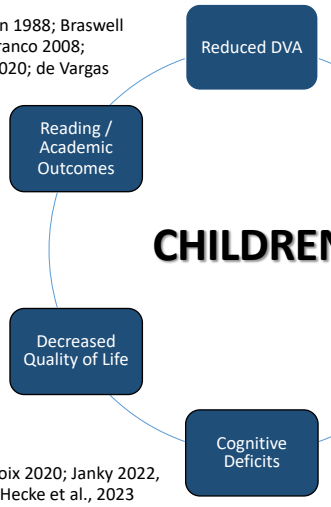
The effects of vestibular loss in CHILDREN are not well established

VESTIBULAR AND BALANCE LAB
BOYS TOWN
NATIONAL RESEARCH HOSPITAL
EST. 2011

Snashall 1983; Levinson 1988; Braswell 2006a; Franco 2007; Franco 2008; Tomaz 2014; Caldani 2020; de Vargas Romero 2020



Rine 2003; Braswell 2006; Martin 2012, Christy 2014, Janky 2015, Janky *in press, etc*

Wolter 2015

Janky *in press*

Kaga 1999, Rine 2000, Inoue 2013, Maes 2014, Christy 2014, Janky 2018, Sokolov 2019, Janky *in press,* etc.

Lacroix 2020; Janky 2022, Van Hecke et al., 2023

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- **It remains to be proven that early detection of vestibular deficits results in better functional outcomes**

Identify → Characterize the Effects of VL → Investigate the Effects of VBRT

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Reliable?

Test Set Up Accurate?

Vestibular Loss?

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Vestibular Evoked Myogenic Potential (VEMP) Test-Retest Reliability in Children

*†Elizabeth Fuemmeler, *‡Amanda I. Rodriguez, *Megan Thomas, *Tom Creutz, *Denis Fitzpatrick, and *Kristen L. Janky

*Boys Town National Research Hospital, Omaha; †Purdue University, West Lafayette, Indiana; and ‡University of Nebraska - Lincoln, Lincoln, Nebraska

Reliable? YES

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

Gabrielle R. Merchant,[1] Kyli M. Schulz,[1,2] Jessie N. Patterson,[3] Denis Fitzpatrick,[1] and Kristen L. Janky[3]

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]

BCV reliable? YES

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

	Cervical 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	4.41	22.51	7.42
p	0.001	0.036	<0.001	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.

VEMP response rates increase when using BCV for individuals with CI

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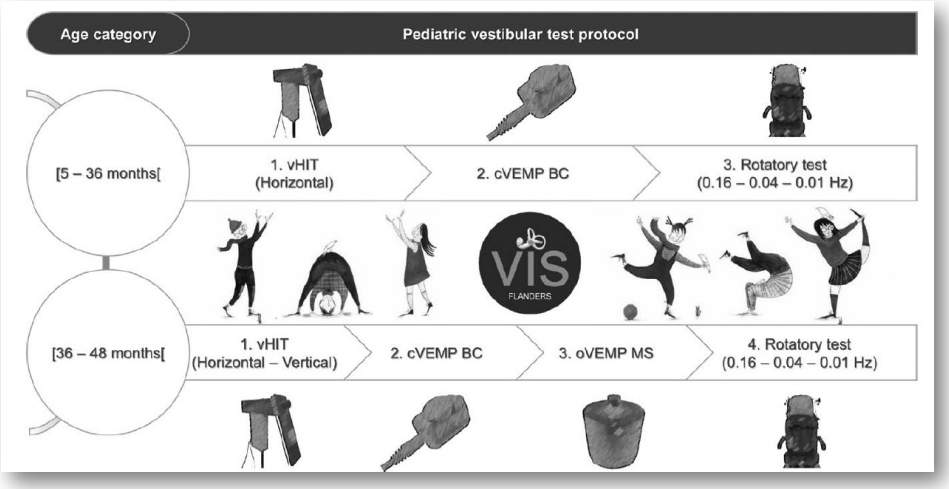
41

Pediatric Vestibular Assessment: Clinical Framework

Sarie Martens,[1] Ingeborg Dhooge,[2,3] Cleo Dhondt,[2] Saartje Vanaudenaerde,[3] Marieke Sucaet,[1] Lotte Rombaut,[3] and Leen Maes[1,3]



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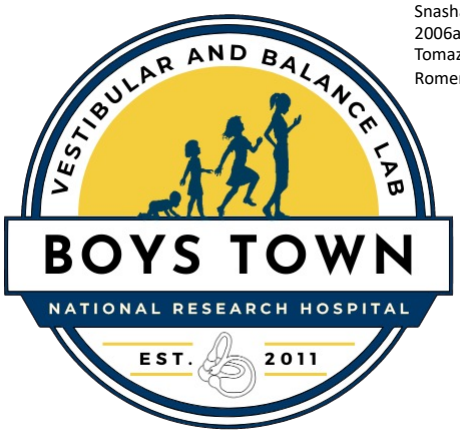
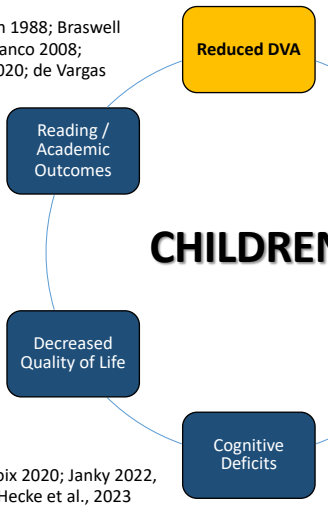
The effects of vestibular loss in CHILDREN are not well established



Snashall 1983; Levinson 1988; Braswell 2006a; Franco 2007; Franco 2008; Tomaz 2014; Caldani 2020; de Vargas Romero 2020



Rine 2003; Braswell 2006; Martin 2012, Christy 2014, Janky 2015, Janky 2021

Wolter 2015

Janky *in press*

Kaga 1999, Rine 2000, Inoue 2013, Maes 2014, Christy 2014, Janky 2018, Sokolov 2019, Janky 2021, etc.

Lacroix 2020; Janky 2022, Van Hecke et al., 2023

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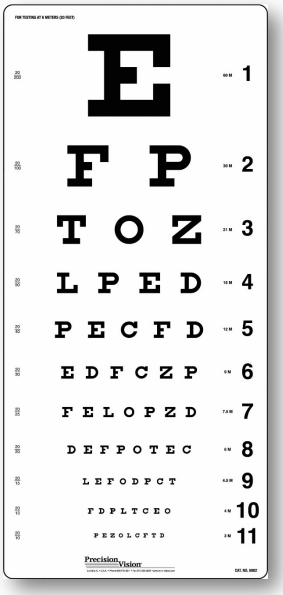
43

Children with vestibular loss have reduced dynamic visual acuity

- Rine 2003; Braswell 2006; Martin 2012, Christy 2014, Janky 2015, Janky 2021*, etc*



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A clinical test of dynamic visual acuity for children

International Journal of Pediatric Otorhinolaryngology

Rose Marie Rine*, Jennifer Braswell[1]

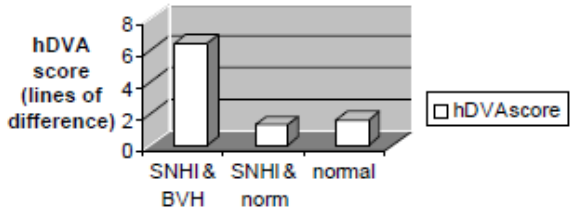

Fig. 1 Comparison of hDVA scores between groups of children. The DVA scores attained by children with concurrent sensorineural hearing loss (SNHI) and bilateral vestibular hypofunction (BVH) were significantly higher than peers with normal vestibular function with (SNHI & norm) or without (normal) SNHI.

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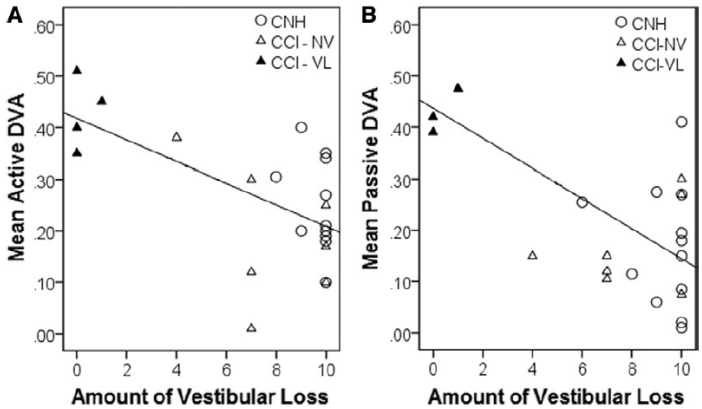
Vestibular, Visual Acuity, and Balance Outcomes in Children With Cochlear Implants: A Preliminary Report

Kristen L. Janky[1] and Diane Givens[2]



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What are the long-term effects (if any) of decreased visual acuity?

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Evidence that vestibular hypofunction affects reading acuity in children

International Journal of Pediatric Otorhinolaryngology

Jennifer Braswell [a,*], Rose Marie Rine [b,1]

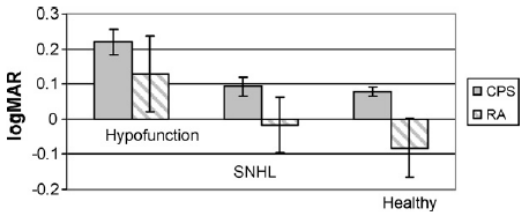

Fig. 2 Critical print size (CPS) and reading acuity (RA) scores were significantly higher in children with SNHI and VH, as compared to peers without VH regardless of hearing status. Hypofunction = vestibular hypofunction ($n = 5$), SNHL NVF = sensorineural hearing loss and normal vestibular function ($n = 9$), healthy = normal hearing and vestibular function ($n = 43$).

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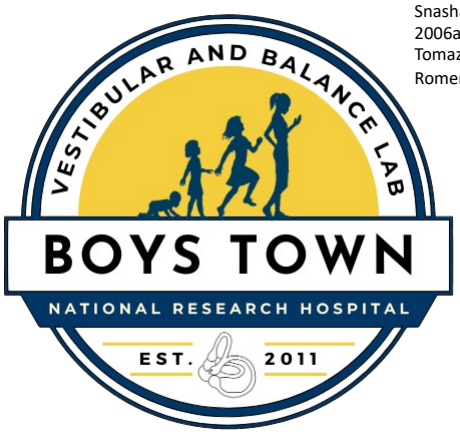
48

The effects of vestibular loss in CHILDREN are not well established



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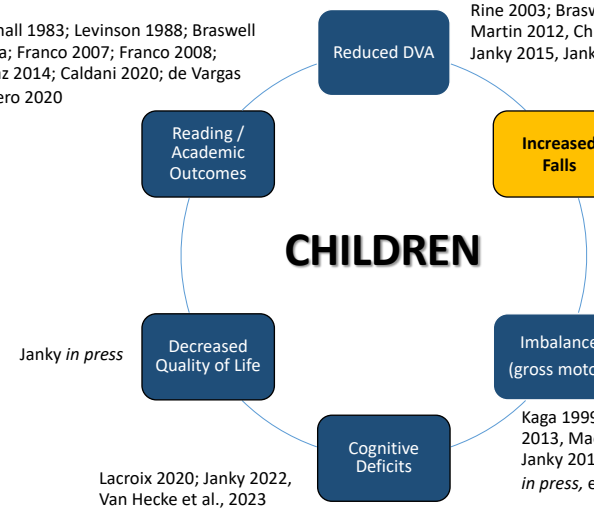
Rine 2003; Braswell 2006; Martin 2012, Christy 2014, Janky 2015, Janky *in press, etc*

Wolter 2015

Janky *in press*

Kaga 1999, Rine 2000, Inoue 2013, Maes 2014, Christy 2014, Janky 2018, Sokolov 2019, Janky *in press,* etc.

Lacroix 2020; Janky 2022, Van Hecke et al., 2023



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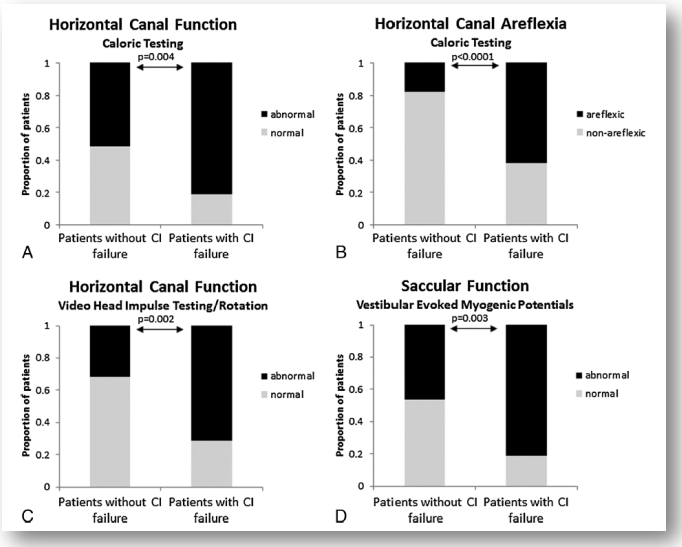
Children with Vestibular Loss have Imbalance

- Kaga 1999, Rine 2000, Inoue 2013, Maes 2014, Christy 2014, Janky 2018, Sokolov 2019, Janky 2021, etc.

Which can lead to increased falls and CI Failure

- Wolter 2015



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Children with Vestibular Loss have Imbalance

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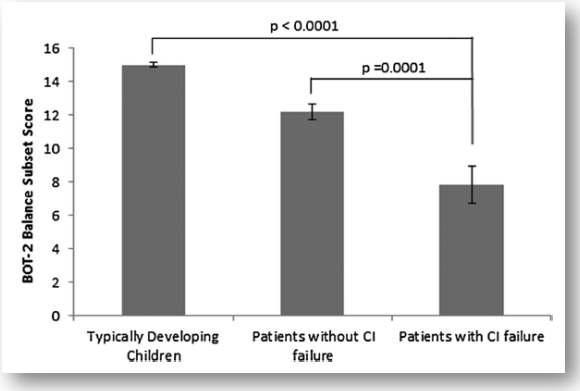

Which can lead to increased falls and CI Failure

- Wolter 2015

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Additional work is needed characterizing the prevalence of falls in children.

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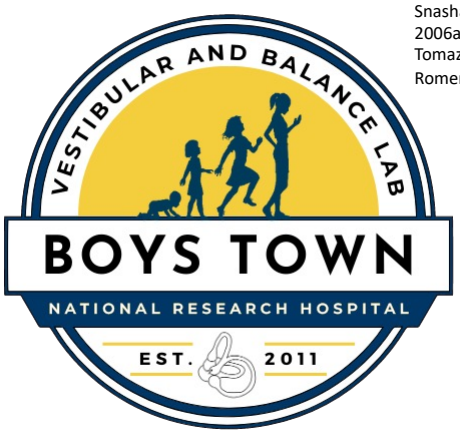
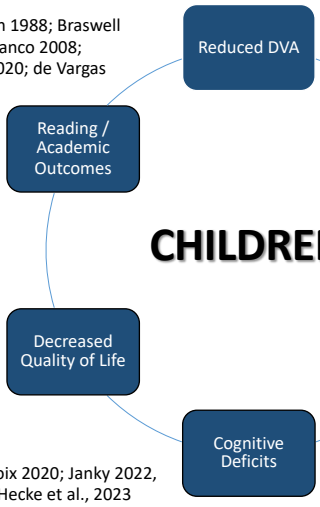
The effects of vestibular loss in CHILDREN are not well established



Snashall 1983; Levinson 1988; Braswell 2006a; Franco 2007; Franco 2008; Tomaz 2014; Caldani 2020; de Vargas Romero 2020



Rine 2003; Braswell 2006; Martin 2012, Christy 2014, Janky 2015, Janky *in press, etc*

Wolter 2015

Janky *in press*

Kaga 1999, Rine 2000, Inoue 2013, Maes 2014, Christy 2014, Janky 2018, Sokolov 2019, Janky *in press,* etc.

Lacroix 2020; Janky 2022, Van Hecke et al., 2023

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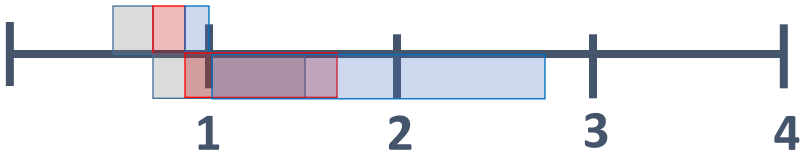
Vestibular Loss Results in Gross Motor Delay

- Children with significant vestibular loss are delayed in gaining head control (Inoue 2013) and in sitting, standing, and walking.
  - Typically developing children sit, stand, and walk at 6 – 8 m, 10 – 11 m, and 10 – 12 m, respectively
  - Children with vestibular loss sit, stand and walk at 8 – 18 m, 9 – 20 m and 12 – 33 m, respectively (Kaga1999).



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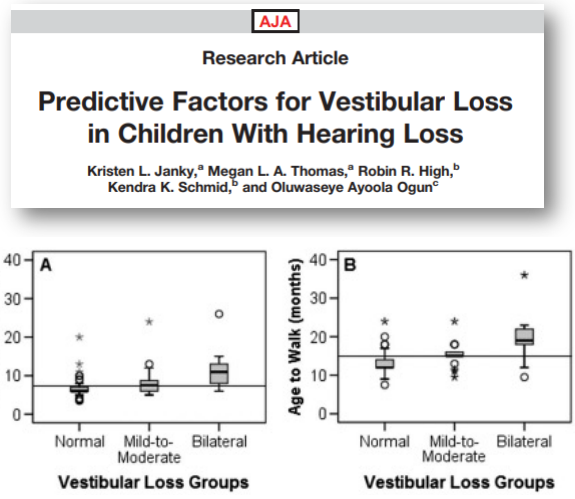
Children are more likely to have vestibular loss if:


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]

- Sat later than 7.2 months or

- Walked later than 14.5 months

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These values **do** overlap normal expectations:

- Sitting without support (3.8 to 9.2 months)
  - Age to sit > 7.25 months
- Standing with assistance (4.8 to 11.4)
- Hands and knees crawling (5.2 to13.5)
- Walking with assistance (5.9 to 13.7)
- Standing alone (6.9 to 16.9)
- Walking alone (8.2 to 17.6)
  - Age to walk > 14.5 months

WHO Multicentre Growth Reference Study

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Gross Motor Delay Extends into Later Childhood

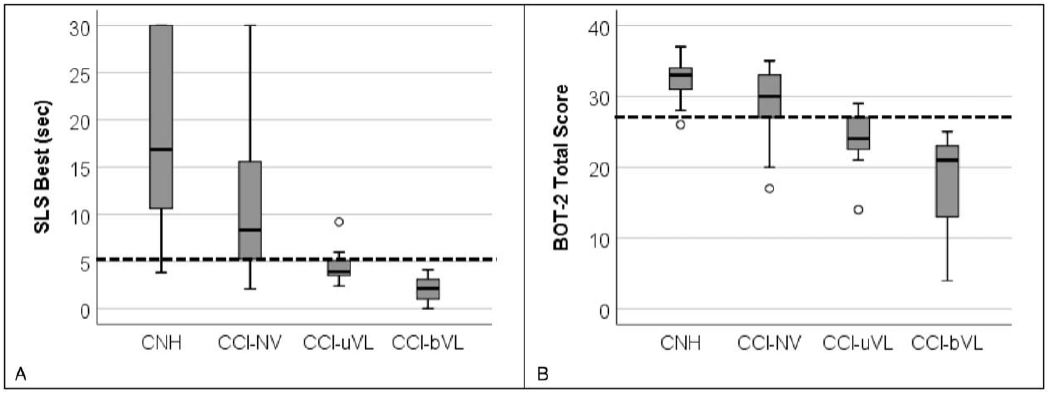

FIG. 1. *A*, Single Leg Stance (SLS) best performance across groups. Dotted line denotes 5-second cut-off criterion; *B*, BOT-2 Balance subtest, total score across groups. Dotted line denotes 27.5 cut off criterion.

Using Functional Outcomes to Predict Vestibular Loss in Children

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

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57

Perceptual and Motor Skills, 2000, 90, 1101-1112. © Perceptual and Motor Skills 2000

EVIDENCE OF PROGRESSIVE DELAY OF MOTOR DEVELOPMENT IN CHILDREN WITH SENSORINEURAL HEARING LOSS AND CONCURRENT VESTIBULAR DYSFUNCTION [1, 2]

- "Both gross motor and balance development scores were lower on repeated testing"

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What are the long-term effects (if any) of decreased gross motor skills?

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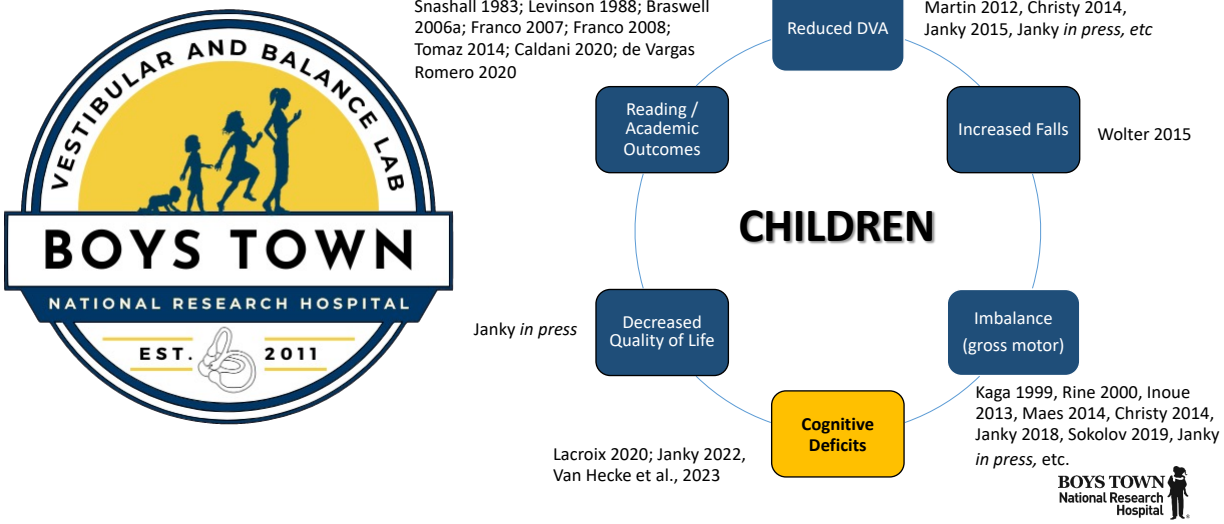
The effects of vestibular loss in CHILDREN are not well established



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Cognitive Deficits and Vestibular Loss

- In adults, vestibular loss has been linked to deficits in spatial navigation, spatial memory, visuospatial, attention, executive function, and cognitive processing (e.g., Xie 2017; Kremmyda 2016; Smith 2019; Popp 2017; Bigelow 2015).
- However, the extent to which cognitive deficits exist in children with hearing and comorbid vestibular loss is unclear.

61

Cognitive Deficits and Vestibular Loss in Children

- Reduced visuospatial working memory, mental rotation and space orientation was recently reported in a small cohort (n = 13) of children with vestibular loss (Lacroix 2020); however, these children varied in their degree of hearing loss.
- Hearing loss alone results in reduced working memory, executive function, and attention (Beer 2014; Nittrouer 2013; Nittrouer 2017; Davidson 2018, Pisoni 2016),
- Both language and auditory experience significantly impact working memory and attention (McCreery *under review*),
- Cognitive deficits could be misattributed to vestibular loss without the inclusion of a control group of children with normal vestibular loss, matched on hearing loss severity. suggesting that in order to isolate the contribution of vestibular loss, careful control of language and auditory experience is critical.

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Does vestibular loss result in cognitive deficits in children with cochlear implants?

- 38 CNH (mean age: 12.2 years, range 7 - 18)
- 37 CCI (mean age: 13.4, range 7 – 20)
  - 26 normal vestibular function
  - 11 vestibular loss (5 UVL, 6 BVL)
- All children had normal non-verbal intelligence
- As expected, children with hearing loss had lower language (PPVT-4) and speech perception (AZ Bio) compared to children with normal hearing, but there were no differences among the children with CI.

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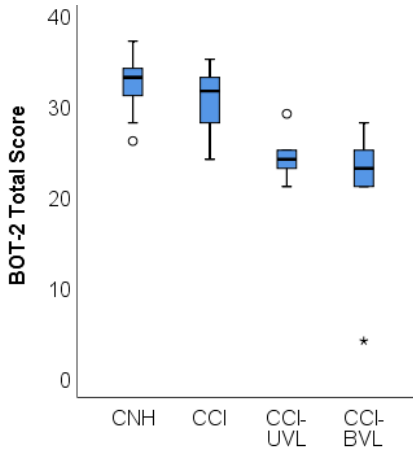
63

Children with Vestibular Loss Have Reduced Balance



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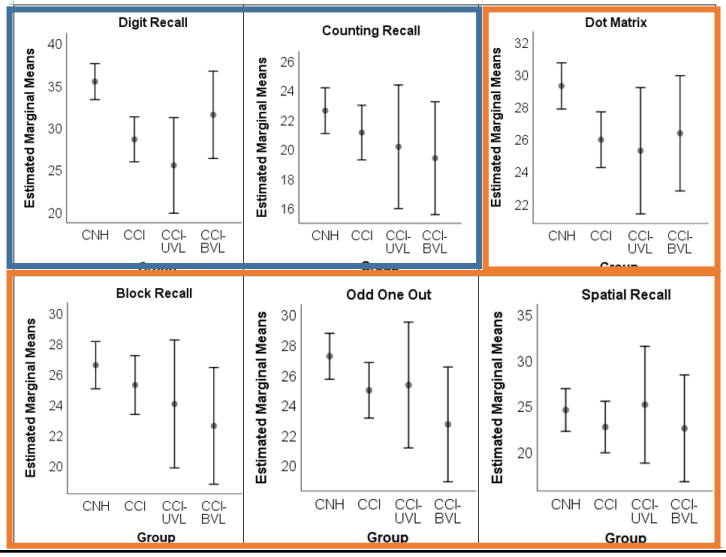
Children with vestibular loss did not have *significantly* worse working memory performance

VERBAL

VISUOSPATIAL



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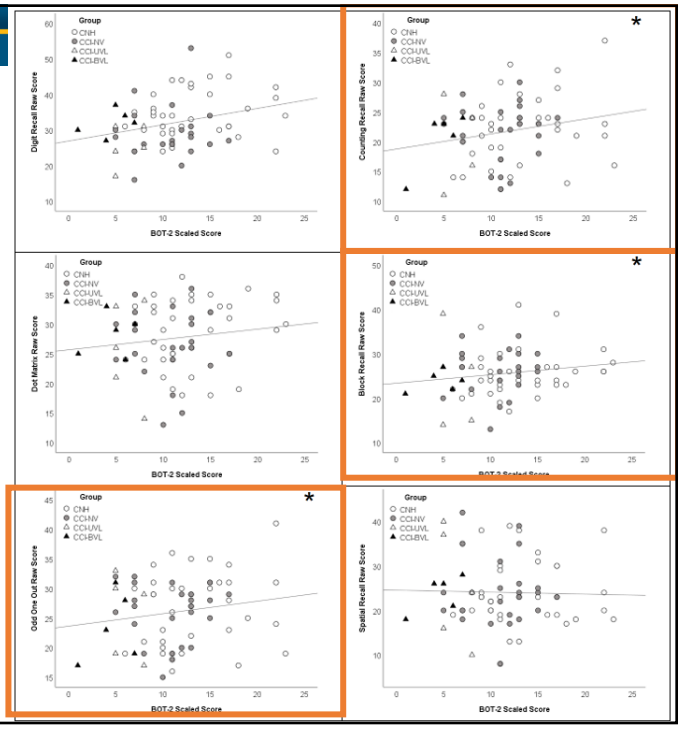
- Regression analysis
  - vHIT, age, and language
  - Balance, age and language
- Interestingly balance, not vHIT, was a significant predictor for counting recall, block recall, and odd one out performance,
- Suggests that the degree of central pathway integration may be a factor in the relationship between cognition and vestibular loss; those with poorer central pathway integration have poorer cognition.



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Does vestibular loss result in cognitive deficits in children with cochlear implants?

- Need more subjects
- Control group matched on hearing loss is crucial
- Balance – in addition to vestibular status – may also be crucial

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67

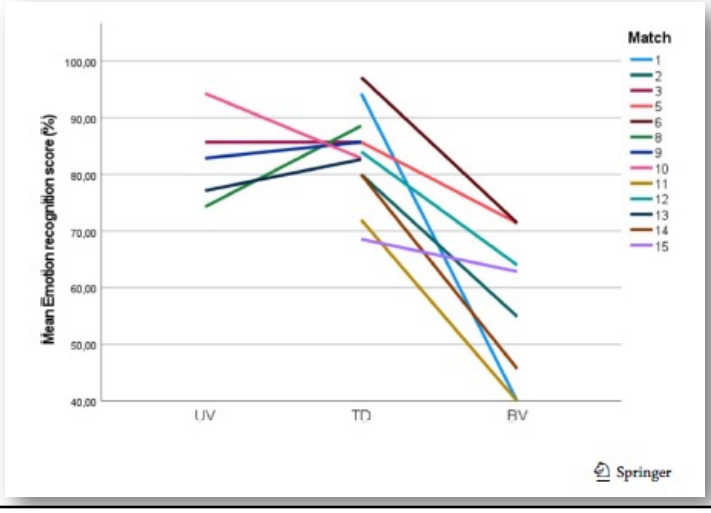
Journal of Neurology
https://doi.org/10.1007/s00415-023-11774-3

ORIGINAL COMMUNICATION

A cross-sectional study on the neurocognitive outcomes in vestibular impaired school-aged children: are they at higher risk for cognitive deficits?

Ruth Van Hecke[1] · Maya Danneels[1] · Frederik J. A. Deconinck[2] · Ingeborg Dhooge[3,4] · Laura Leyssens[1] · Emmely Van Acker[1] · Hilde Van Waelvelde[1] · Jan R. Wiersema[5] · Leen Maes[1,2]

Children with bilateral vestibular dysfunction exhibited more difficulties with emotion recognition compared to their typically developing peers



68

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ORIGINAL COMMUNICATION

A cross-sectional study on the neurocognitive outcomes in vestibular impaired school-aged children: are they at higher risk for cognitive deficits?

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- Executive function
  - Inhibition task (NEPSY II)
  - BRIEF questionnaire
- Lower emotion recognition and inhibition for the children with bilateral vestibular loss.

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Additional work is needed characterizing the effect of vestibular loss on cognition in children.

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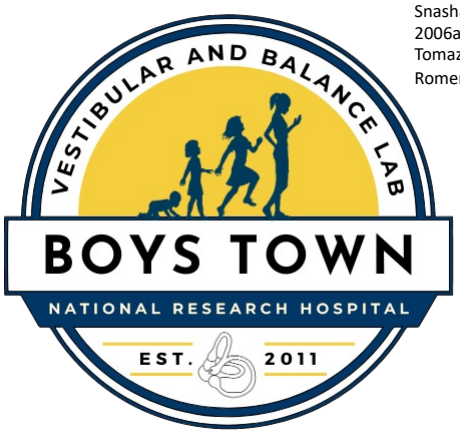
The effects of vestibular loss in CHILDREN are not well established



Snashall 1983; Levinson 1988; Braswell 2006a; Franco 2007; Franco 2008; Tomaz 2014; Caldani 2020; de Vargas Romero 2020

Rine 2003; Braswell 2006; Martin 2012, Christy 2014, Janky 2015, Janky *in press, etc*

Reduced DVA

Increased Falls

Wolter 2015

Imbalance (gross motor)

Kaga 1999, Rine 2000, Inoue 2013, Maes 2014, Christy 2014, Janky 2018, Sokolov 2019, Janky *in press,* etc.

Cognitive Deficits

Lacroix 2020; Janky 2022, Van Hecke et al., 2023

Decreased Quality of Life

Janky *in press*

Reading / Academic Outcomes

CHILDREN

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71

The effects of vestibular dysfunction on balance and self-concept in children with cochlear implants☆

Kristen L. Janky[a,*], Jessie Patterson[a], Megan Thomas[a], Sarah Al-Salim[b], Sara Robinson[b]

[a] *Boys Town National Research Hospital, Department of Audiology, Omaha, NE 68131, United States*
[b] *Boys Town National Research Hospital, Center for Childhood Deafness, Language and Learning, Omaha, NE 68131, United States*

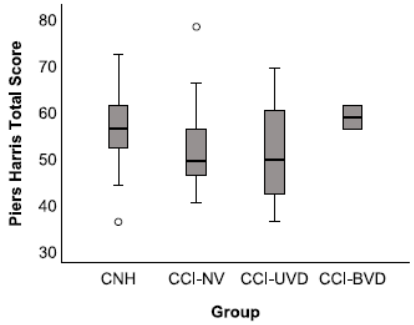

Table 3
PCA predictors and component Structure.

Predictors	Component	
	1	2
	"Vestibular"	"Auditory-Language"
BOT-2 – Total Raw Score	.856	
Mean HC vHIT gain	.860	
Passive DVA	−.729	
AZBio		.730
PPVT-4		.801

BOT-2 = Bruininks-Oseretsky Test of Motor Proficiency; HC = horizontal canal; vHIT = video head impulse test; DVA = dynamic visual acuity; PPVT-4 = Peabody Picture Vocabulary Test – 4.

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Additional work is needed characterizing the effect of vestibular loss on quality of life in children.

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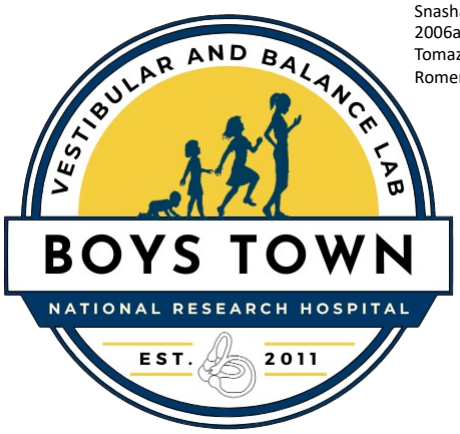
The effects of vestibular loss in CHILDREN are not well established



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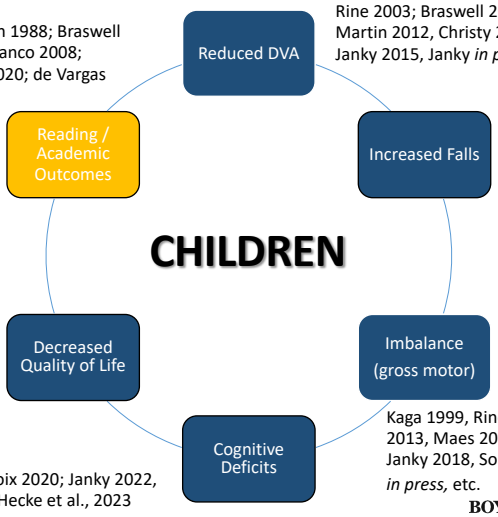

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Wolter 2015

Kaga 1999, Rine 2000, Inoue 2013, Maes 2014, Christy 2014, Janky 2018, Sokolov 2019, Janky *in press,* etc.

Lacroix 2020; Janky 2022, Van Hecke et al., 2023

Janky *in press*

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Hearing Loss is Associated with Poorer Reading

- Reading is a fundamental part of academia and is an essential component to learning.
- Compared to typically developing peers, children with hearing loss have poorer reading outcomes (Lederberg 2013; Qi 2012)
- Reading performance in children with hearing loss has been related to reductions in:
  - Language and phonological processing (Geers 2011, Harris 2017; Lederberg 2013; Lederberg 2019; Werfel 2017),
  - Decoding and/or language comprehension (Gough 1986), and
  - Auditory access (i.e., severity of hearing loss, hearing aid use, auditory environment, duration of hearing loss, and properly fit auditory devices (Geers 2011; Tomblin 2015).

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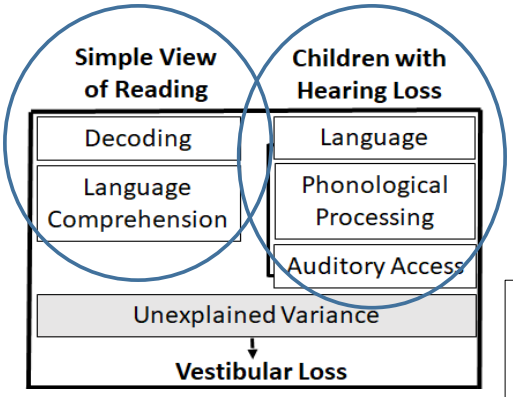

Children with moderately severe hearing loss are more likely to demonstrate reductions in reading comprehension (Tomblin 2020)

Language, phonological processing, Auditory Access

Vestibular Loss

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Vestibular Loss is Associated with Poorer Reading

- It has been suggested that the gross motor and visual acuity deficits present in children with vestibular loss have cascading effects on reading and academic outcomes (Levinson 1988; Snashall 1983; Braswell 2006a)
- Children with reading difficulties have vestibular deficits (Snashall 1983; Franco 2007; Franco 2008; Tomaz 2014; Caldani 2020; de Vargas Romero 2020).
- It has also been suggested that reading difficulties could improve with vestibular rehab (Braswell 2006b; Caldani 2020).

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The Problem

- While vestibular loss has been speculated to influence reading outcomes in children (Braswell 2006a; Snashall 1983; Franco 2007; Franco 2008; Tomaz 2014; Caldani 2020)
  - **The association between reading difficulties and vestibular loss remains unexplained.**
  - **These studies have not controlled for variables known to predict reading performance.**

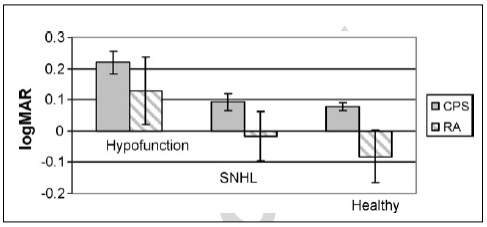
78

Evidence that vestibular hypofunction affects reading acuity in children

Jennifer Braswell[a,*], Rose Marie Rine[b,1]

International Journal of Pediatric Otorhinolaryngology

1. Reading acuity* scores were significantly worse in children with vestibular loss
2. Reading acuity scores correlated with dynamic visual acuity.



* Reading acuity = smallest print size that can be read

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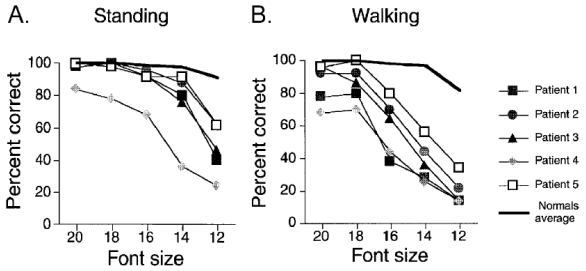
79

Dynamic visual acuity while walking in normals and labyrinthine-deficient patients

Journal of Vestibular Research 9 (1999) 49–57

Edward J. Hillman[a], Jacob J. Bloomberg[b,*], P. Vernon McDonald[c] and Helen S. Cohen[a]

- Adults with bilateral vestibular loss performed significantly worse at visual acuity while standing and walking.



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Why would vestibular loss cause breakdowns in ***static*** acuity?

Neither Hillman et al (1999) nor Braswell and Rine (2004) provide a rationale for **why** static visual acuity may be affected in adults and children with vestibular loss.

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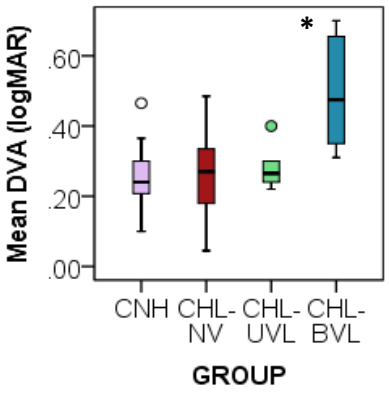
Children with Vestibular Loss Have Reduced DVA

- Subjects:
  - Control children with normal hearing (n = 22, mean age: 12.3 years, range 6 - 18)
  - Children with hearing loss (n = 37, mean age: 12.3 years, range 6 - 18), grouped by vestibular status:
    - Normal (n = 22)
    - Unilateral loss (n = 6)
    - Bilateral loss (n = 9).



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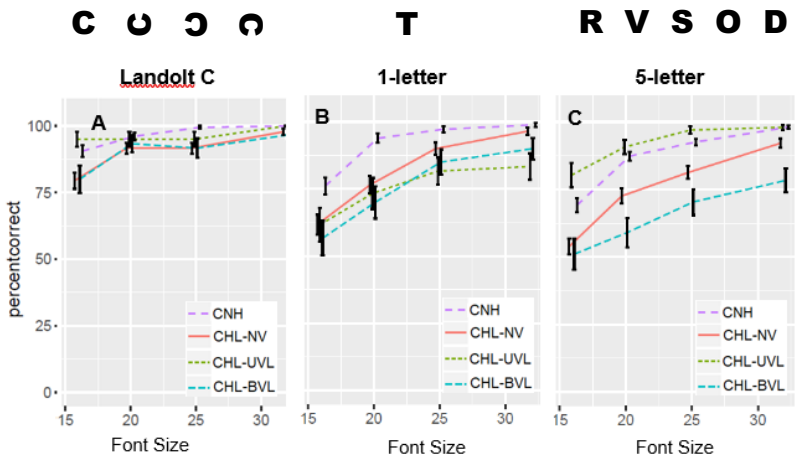
Children with Vestibular Loss Have Reduced Static Acuity



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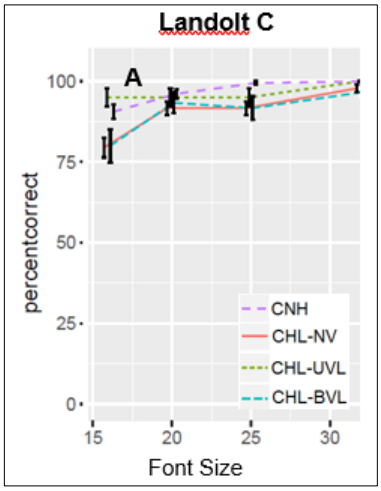
Equal Performance for Landolt C

- Landolt C has the lowest linguistic complexity suggesting visual acuity is not a factor driving group differences



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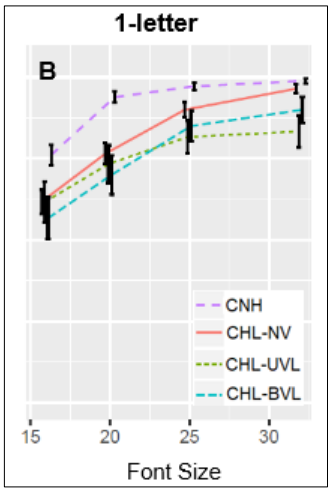
Performance declines for ALL children with hearing loss in 1-letter condition

- Children with hearing loss do not have difficulties with alphabet knowledge (Werfel 2014); however, they do have lower language (Tomblin 2015; Werfel 2017).
- Thus, the reduced static visual acuity from the Landolt C to the 1-letter condition is *hypothesized* to be related to language deficits secondary to hearing loss.



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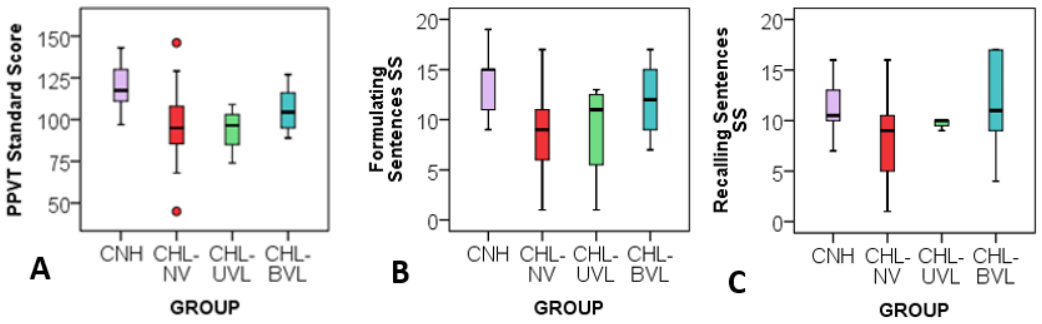
Children with Hearing Loss have lower language scores



A: Peabody Picture Vocabulary Test (PPVT),
B: CELF Formulating Sentences,
C: CELF Recalling Sentences

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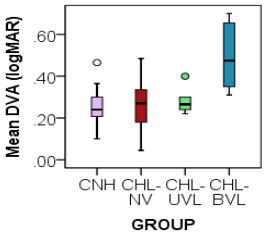
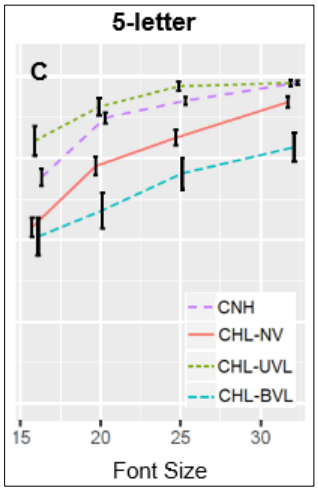
Performance significantly declined for children with bilateral vestibular loss

- The breakdown in performance with increased visual target complexity is *hypothesized* to be due to abnormal vestibulo-ocular reflex (like dynamic visual acuity) and/or working memory, attention, or cognitive processing.





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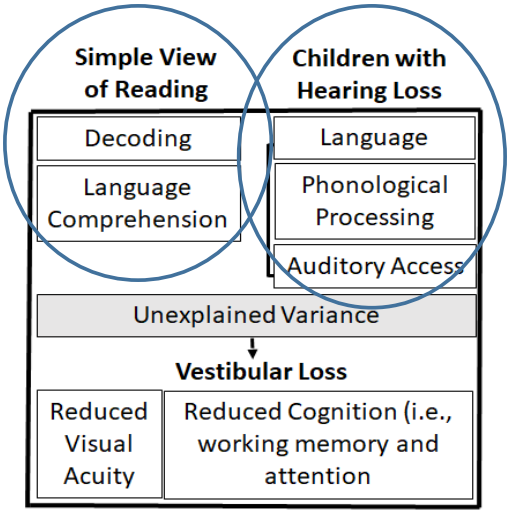

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Reduced Visual Acuity and Reading

- Better reading performance has been associated with children who have better developed vestibulo-ocular reflex latencies (de Vargas Romero 2020).
- Children with learning disabilities require an increased number of saccades for accurate visual acuity, which has been attributed to an insufficient vestibulo-ocular reflex (Petri 1980).
- The co-occurrence of saccade deficits in children with vestibular loss while reading has not been established.
- Our preliminary data demonstrated there was no main effect of head position (fixed vs free). While this suggests the vestibulo-ocular reflex does not significantly affect *static* visual acuity, we did not evaluate eye movement patterns *during* static visual acuity testing.

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Children with Vestibular Loss have longer reaction times

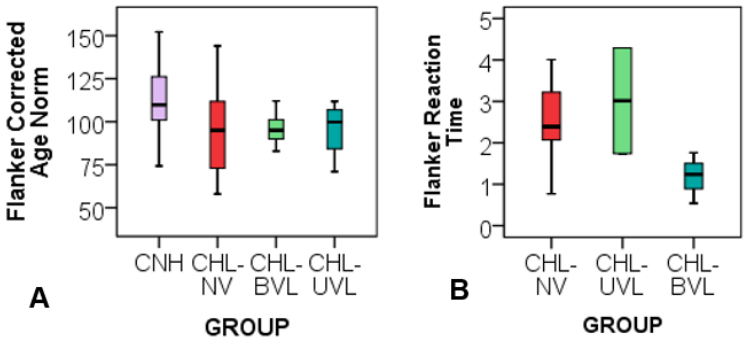

Fig. 8: Flanker A: There was no significant difference in performance between groups. B: Children with bilateral vestibular loss had slower reaction time; lower is worse due to log transformation. CNH = children with normal hearing; CHL= children with hearing loss, NV= normal vestibular, UVL = unilateral vestibular loss; BVL = bilateral vestibular loss.

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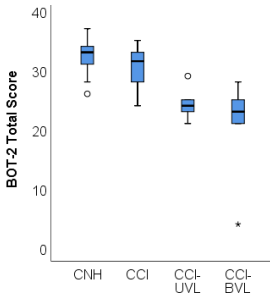
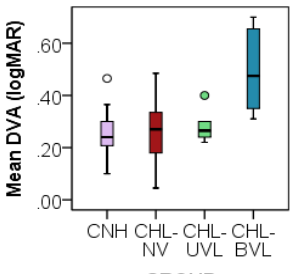
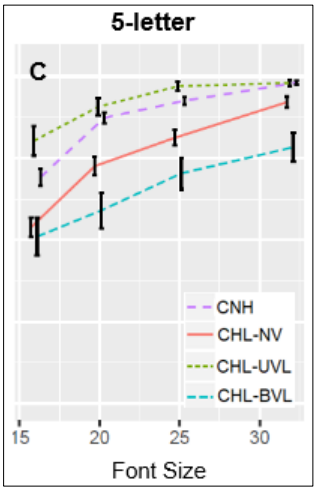
Preliminary data suggest that children with bilateral vestibular loss have deficits in:

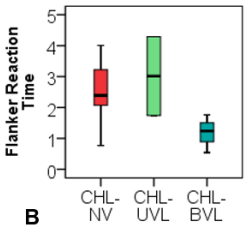


Balance

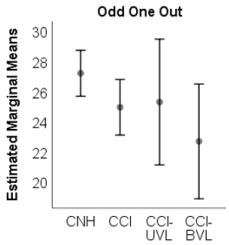


Dynamic Visual Acuity





Reaction Time



91

K-Award Goals:

1. Disambiguate the variations in visual target complexity and the factors that affect performance

2. Determine if vestibular loss affects reading performance

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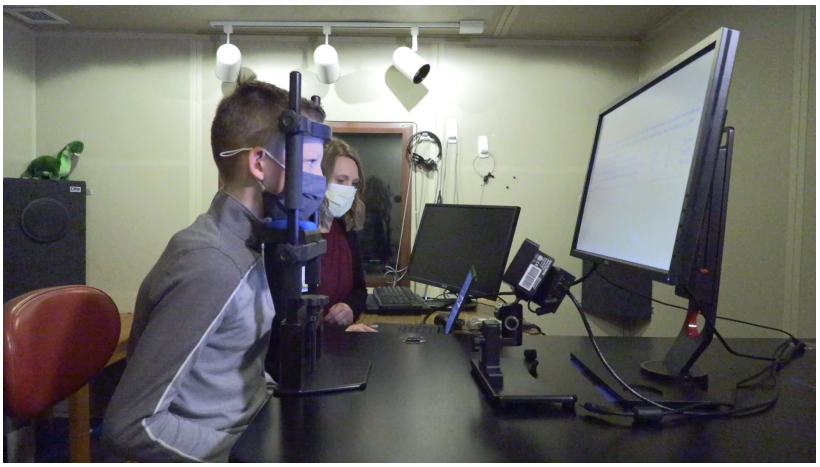
Reading (& Cognition & QOL) Outcomes in Children with Vestibular Loss



Contact me if you have children with CI (age 8 – 14) AND vestibular loss that might participate!

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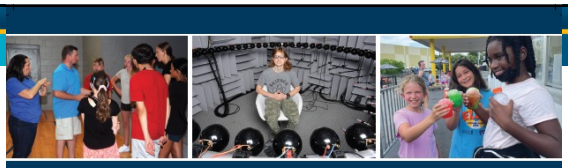

Participate in Cochlear Implant Research

Who?
6- to 17-year-olds with one or two cochlear implants

What?
3- to 4-day research camp that may include the following activities:

- Hearing, speech, language and balance (vestibular) assessments
- Reading words or paragraphs
- Listening to words and repeating them back
- Judging the emotion of talkers
- Listening to sounds and detecting where they are from
- Playing thinking games on a computer or tablet

Where?
Boys Town National Research Hospital
555 North 30th Street, Omaha, NE 68131

What will the research camp daily schedule look like?
You'll receive a family-centered schedule with activities based on your child's age and needs. Typically, a full-day camp will last 5-7 hours, including lunch. Participating children will receive prizes and $20/hour for time spent in research.

What will your family receive for travel accommodation?
The camp provides a hotel stay in Omaha and a daily food allowance for each participating child with a cochlear implant(s) and one parent. Roundtrip mileage will be reimbursed, or air travel will be arranged.

Research participation is also about FUN!
Your family will receive FREE admission to visit local attractions, including the world-renowned Henry Doorly Zoo!

Who are we?
We are a group of researchers passionate about improving outcomes for children with cochlear implants.

Interested in signing up?
Please fill out this short survey. Our research coordinator will reach out shortly to discuss participation eligibility.

https://redcap.link/boystown-ci-research

For general questions:
Please email ci.research@boystown.org.

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94

Functional Hearing Laboratory

- **Members:** Ellen Peng (Director), Jeff Simmons (Research Audiologist) & Victoria Sweeney (Core Research Audiologist)
- **Goal**: To personalize cochlear implant fitting for children with bilateral cochlear implants to maximize functional hearing in real-world (noisy & reverberant) listening environments
- **Tools**
  - Psychoacoustics, speech perception, neuroimaging (fNIRS), direct-streaming (acoustic) virtual reality
- **Example Studies**
  - Individual Profiling of Spatial Release from Masking (SRM) in Reverberation
  - Role of Balance & Vestibular Function in Functional Spatial Hearing (i.e., localization, acuity & SRM)
  - Effect of CI Processing Strategy on Functional Spatial Hearing (e.g., MED-El FS4)
  - Top-down Speech Processing in Complex Listening Environments
- **Recruitment & Typical Study Session**
  - Pediatric bilateral CI users of 8-17 years old of all three CI brands
  - 2-3 hours per study/protocol with CI participants with breaks
- **Current support**
  - NIH R21DC020532, Hearing Health Foundation

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Auditory Prostheses and Perception Laboratory

- **Members:** Monita Chatterjee (Director), Aditya Kulkarni (Research Lab Project Manager)
- **Goal**: To investigate the relation between cue weighting for emotions and identification of emotional prosody, in addition to perception-production link for emotional prosody by cochlear implant recipients
- **Tools**
  - Speech perception, psychophysics, developmental and cognitive measures
- **Example Studies**
  - Emotion recognition and perception in a sound booth using 2-AFC or 5-AFC procedures
  - Naturalistic and scripted emotion production
  - Speech processing in complex listening environments
- **Recruitment & Typical Study Session**
  - Pediatric CI users (7-18 years old) and adults (19 and up) of all three CI brands and modalities
  - 4-6 hours per study/protocol with breaks
- **Current support**
  - NIH R01 DC019943-01A1

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What I hope you learned today…..

- Vestibular Newborn Screening is feasible!!
- Bone Conducted cVEMP, Rotary chair, or remote based vHIT are good tools.
- Children to be tested:
  - Severe-to-profound hearing loss
  - Acquired / progressive hearing loss
  - ANSD
  - Gross motor delay
- Degree of vestibular loss matters!
- More research is needed to define the long-term effects of vestibular loss in children

97

Specific Research Questions

- Testing:
  - The minimum number of trials needed to obtain a reliable response.
  - Validity of vHIT findings
- Functional outcomes:
  - What's the next step?
    - How does reduced DVA affect children?
    - How does reduced gross motor function affect children?
    - Additional work is needed characterizing the prevalence of falls, the extent of cognitive dysfunction, and in children with vestibular loss

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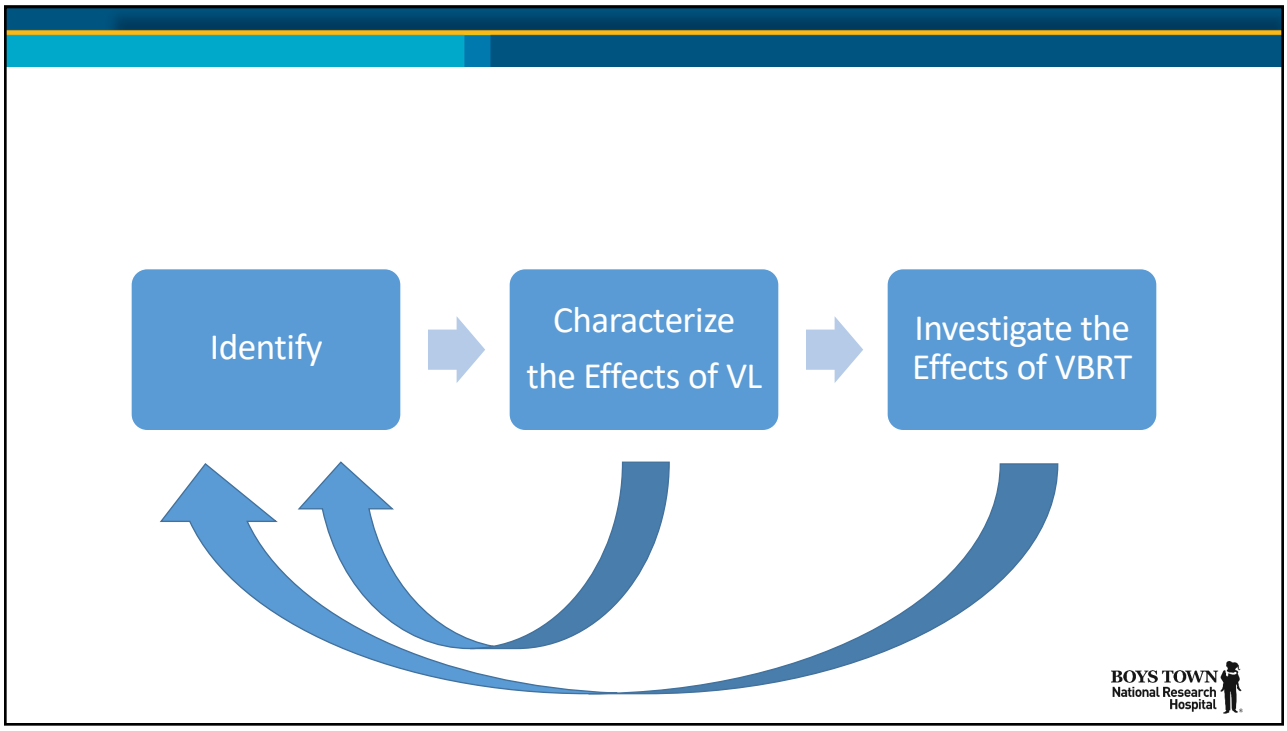
98



99

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- Liz Marler
- Nicole Greenwalt
- Kyli Schulz
- Gabby Merchant
- Katie Gordon
- Kayla Samuelson
- Sarah Al Salim
- Sophie Ambrose
- Casey Vandervelde
- Manuel Vicente

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100

Questions?

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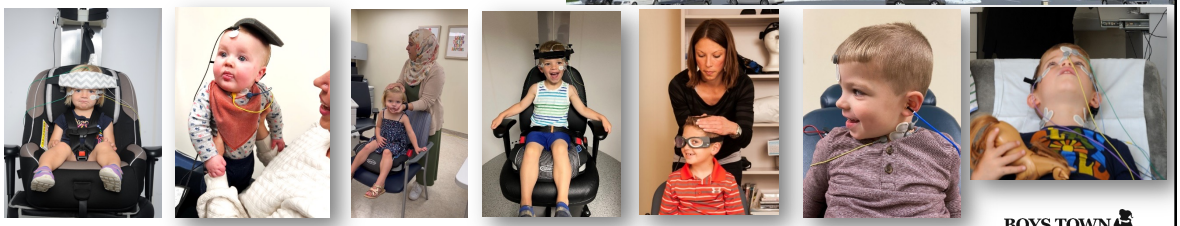

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101