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Vestibular Monitoring Protocol for Cochlear Implant and cCMV Patients

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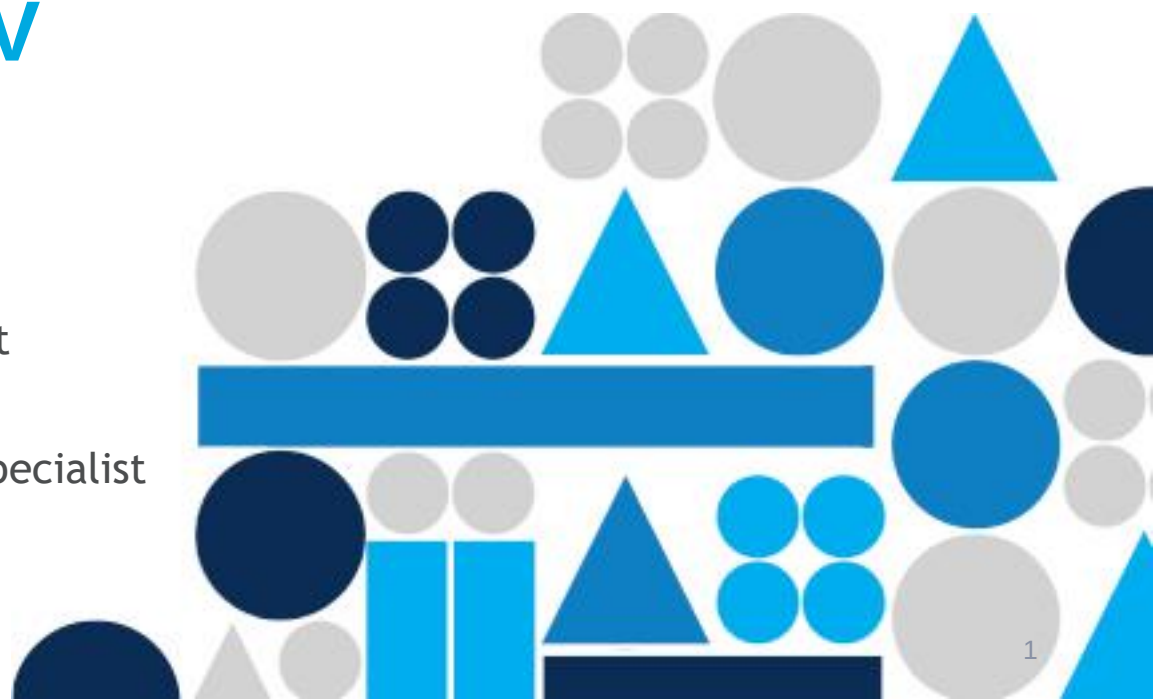

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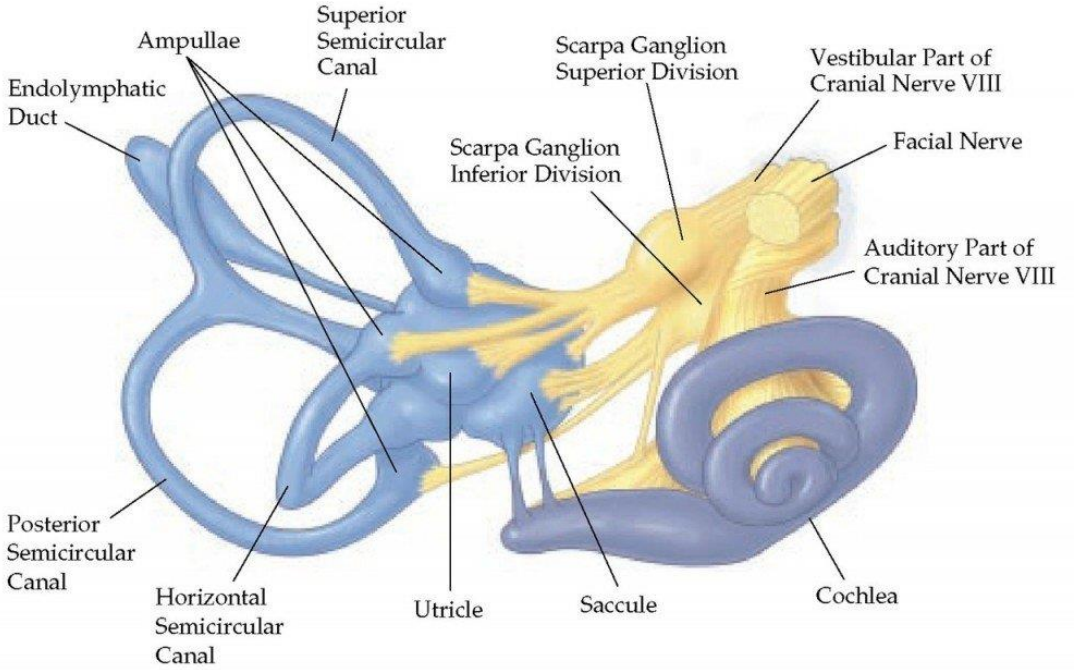
Learning Objectives

1. Vestibular dysfunction and SNHL
2. Vestibular dysfunction and cochlear implants
3. Vestibular dysfunction and cCMV

Why are balance concerns common in people with SNHL?



The Inner Ear

Peripheral vestibular organs & Cochlea

- Share endolymph fluid
- Share similar blood supply
- Comprise the 8th cranial nerve
- Congenital malformations

If something has affected one half of the inner ear, it can also affect the other half.

About 50% of children with severe-profound sensorineural hearing loss will have vestibular hypofunction. Half of them will have bilateral hypofunction.

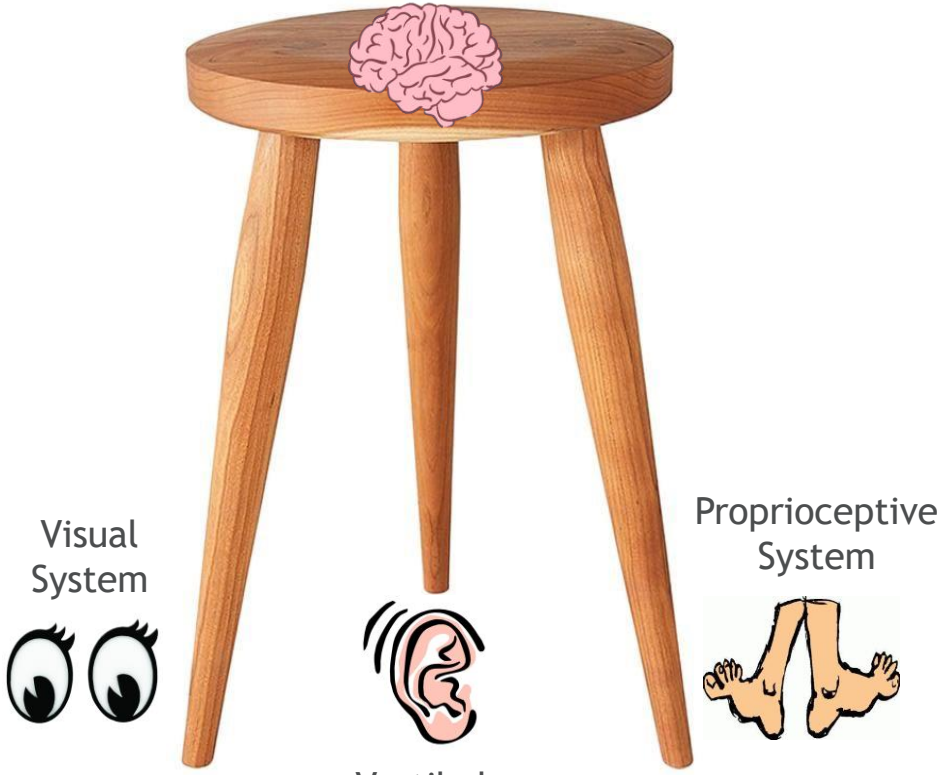
Cushing, et al., 2013

Prevalence of Vestibular Dysfunction by Hearing Loss Etiology

- Congenital Cytomegalovirus (cCMV)
  - Asymptomatic: **45%** (Pinninti, et al., 2021)
  - Symptomatic: **92%** (Bernard et al., 2015)
- Bacterial Meningitis: **80-100%** (Cushing et al., 2013)
- Usher's Syndrome: **50%** (Toms et al., 2020)
- Waardenburg: **77%** (Black et al., 2001)
- Alport: **45-75%** (Barozzi et al., 2020)
- Ototoxicity (aminoglycosides): **79%** (Handelsman et al., 2017)
- Enlarged Vestibular Aqueduct (EVA): **89%** (Yang et al., 2016)
- Connexin 26: **44%** (Cushing et al., 2013)
- Inner ear or 8th nerve malformations
- CHARGE

Vestibular dysfunction is common in children with SNHL

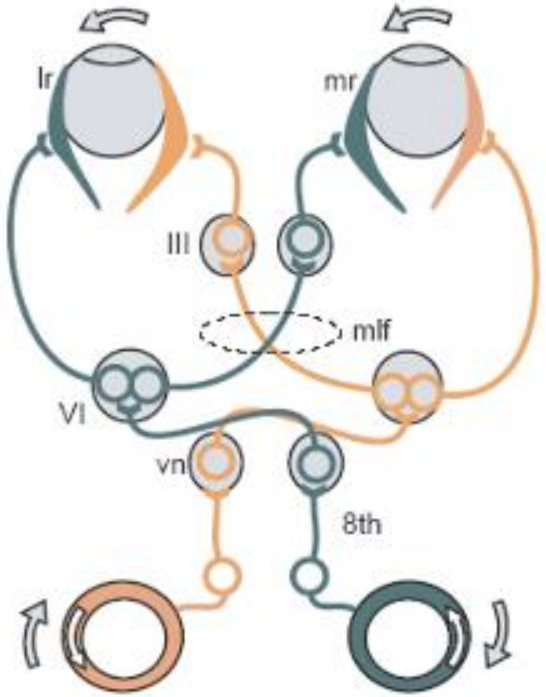
The Balance System



Bilateral Vestibular Hypofunction

Vestibular-Ocular Reflex (VOR)

- Our eyes move exactly opposite of our inner ear movement, stabilizing our visual field.
- Without a VOR, objects will appear to bounce/move in response to head movement (oscillopsia).
- People with bilateral semicircular canal areflexia do not have a VOR.
- Vestibular physical therapy can help the patient develop gaze stabilization strategies in the absence of a functioning VOR.



Over ¼ of children with severe to profound SNHL will have bilateral areflexia

Vestibular Rehabilitation

Physical Therapy Treatment Strategies: patient-specific, goal oriented, and based on dysfunction, activity, and participation restrictions.

Vestibular results help determine the best treatment strategy.

- **Habituation:** repeated exposure to dizzy provoking stimulus to help habituate the nervous system
- **Adaptation:** the vestibular system changes to adapt to the neural stimulus (head and/or body movement)
- **Substitution:** alternative strategies for lost or ineffective system
- **Canal re-positional technique (CRT):** Treatment for BPPV (e.g. Epley maneuver)

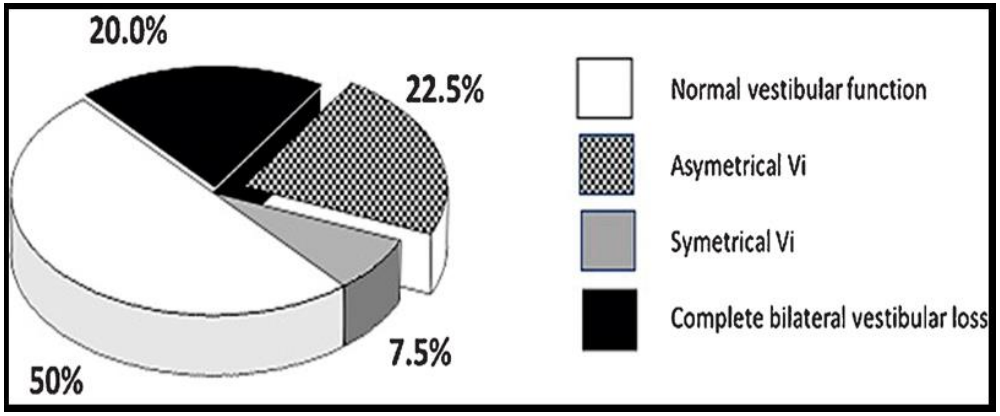
Call to Action:

Provide vestibular testing to our highest risk patients and connect them with vestibular rehabilitation therapy early.

Prevalence in Cochlear Implant Patients

The high prevalence of vestibular disorders in children with hearing loss has been well established.

Wiener-Vacher et al. (2018)



- Prevalence: 50%
- 1,491 children with profound bilateral SNHL underwent vestibular testing
  - 20% of children had complete bilateral vestibular areflexia
  - 30% had partial vestibular hypofunction
  - 50% had normal vestibular function

Prevalence in Cochlear Implant Patients

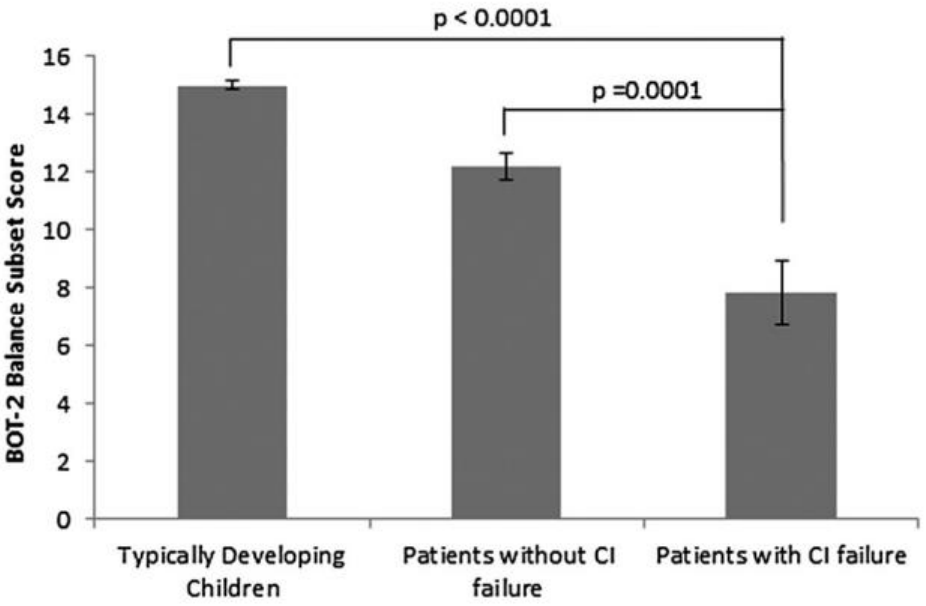
Cushing, 2013

- Prevalence: 50%
- 153 children with profound SNHL had VEMP, Caloric, and Rotational Chair testing
  - 13% had partial vestibular loss
  - 37% had complete bilateral areflexia

Jacot et al., 2009

- Prevalence: 50%
- 224 children with profound SNHL had vHIT, Caloric, and VEMP testing. 89 had both pre and post implant testing.
  - 20% had partial vestibular loss
  - 30% had complete bilateral areflexia
  - 71% demonstrated changes post-implantation compared to pre-implantation

Vestibular Hypofunction and Device Failure



Wolter et. al., 2015

Compared rates of CI failure between children with normal vestibular function and children who had bilateral vestibular hypofunction.

Children with vestibular hypofunction were 8 times more likely to experience an internal device failure secondary to injury from falls.

- Poorer BOT-2 scores in patients with CI failure = More at risk for falls/trauma
  - Single, powerful head impact
  - Repeated smaller impacts over time

Cochlear Implant Summary

- Children with cochlear implant(s) are at high risk for vestibular hypofunction.
- Children with cochlear implants and bilateral vestibular hypofunction are at increased risk for device failure, but the risk can be minimized by early identification and participation in vestibular rehabilitation.
- Untreated vestibular hypofunction can impact the child's ability to participate in daily activities and reading abilities.
- Treatment is effective once a hypofunction is identified.
- **All children receiving cochlear implant(s) at CHCO will undergo vestibular testing to rule out or identify underlying vestibular hypofunction.**

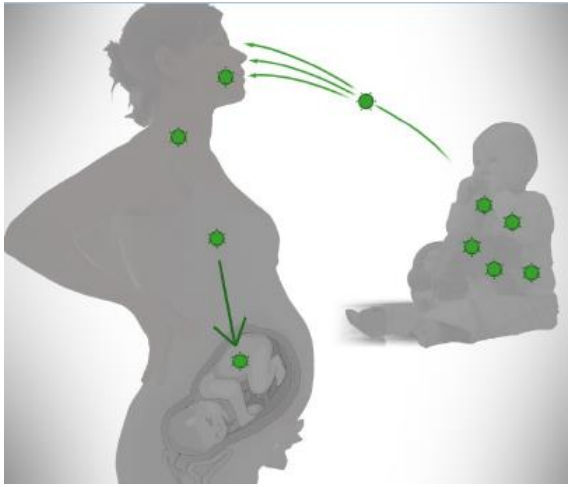
Vestibular Monitoring for Cochlear Implant Patients

- **VEMP and HIT prior to CI surgery (all ages)**
  - Results do not impact candidacy
- **Implanted before age 3 years: Follow up at age 3 years**
  - Vestibular Testing
  - Physical Therapy evaluation
  - If pre-operative testing was abnormal, follow up at 3-months post-activation instead
- **Implanted after age 3 years: Follow up at 3-month post-activation**
  - Vestibular Testing
  - Physical Therapy evaluation if results are abnormal


0:00 / 0:23

Congenital Cytomegalovirus (cCMV)

- CMV is a common virus that majority of adults have been exposed to through body fluids.
- Pregnant women can pass the virus to their unborn baby.
  - Occurs in 0.7 – 1% of all live births (Pontes, et al., 2024)
  - About 11% are symptomatic at birth and 89% are asymptomatic (Pontes, et al., 2024)



- Can cause hearing and vision loss, as well as brain, liver, spleen, lung abnormalities and growth restriction
- Leading cause of hearing loss and neurologic disabilities worldwide (Boppana, et al., 2013)
- Cause of 25% of congenital sensorineural hearing loss and can be progressive (Pontes, et al., 2024)

Vestibular Function and cCMV

Shears et al., 2022: Systematic literature review of vestibular function in children with cCMV

- 12 studies performed vestibular tests on children with cCMV.
- 10/12 studies showed at least 40% or more of children with cCMV had vestibular hypofunction.
  - 92% of symptomatic children (Bernard et al., 2015)
  - 45% of asymptomatic children (Pinninti et al., 2021)
- 2 studies showed a progression of vestibular hypofunction over time.
  - 10% showed progression over 10 months (Dhondt et al., 2021)
  - 50% showed progression over 26.3 months (Bernard et al., 2015)

Considerations when developing our protocols:

Vestibular hypofunction…

- is common in children with severe-profound SNHL and/or cCMV
- can occur with in children with asymptomatic cCMV who have normal hearing
- can be progressive in children with cCMV
- can cause problems with reading and academics
- leads to an increased risk of internal device failure secondary to falls/injury
- can be improved (not cured) with vestibular rehabilitation therapy

CHCO Vestibular Monitoring Guideline for cCMV

- **12 months of age:**
  - Cervical vestibular evoked myogenic potential (cVEMP)
  - Head Impulse Test (HIT)
    - If abnormal, add rotational chair testing
- **3 years of age:**
  - Limited vestibular evaluation
    - VEMP, vHIT, Rotational chair
- **7 years of age:**
  - Comprehensive vestibular evaluation
    - VEMP, vHIT, Rotary chair, VNG (oculomotor, positionals, calorics)

❖ Additional or repeat testing may be recommended if there are other abnormal findings.

Gross Motor Milestone Screening

Table 1
Normal developmental gross motor milestones[67,68]

Developmental Motor Milestone	Expected Age to Reach Milestone
Head control	4 mo
Sit without support	6–9 mo
Crawling	6–9 mo
Walking	12–18 mo
Standing on 1 foot	30 months—briefly 36 months—2 s 4 years old—5 s 5 years old—10 s

Data from Gerber RJ, Wilks T, Erdie-Lalena C. Developmental milestones: motor development. Pediatr Rev 2010; 31:267-276; quiz 277., Syed MI, Rutka JA, Sharma A, Cushing SL. The 'dizzy child': a 12-minute consultation. Clin Otolaryngol 2014; 39:228-234.

Consider vestibular testing for all children with SNHL who have gross motor delays.

Future Research

Comprehensive Vestibular Findings in Children with Congenital Cytomegalovirus

- IRB approved
  - Children's Hospital Colorado
  - Cincinnati Children's Hospital Medical Center
  - Children's Hospital Philadelphia
  - University of Michigan

Case Studies

Children's Hospital Colorado
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Case Study 1:

Congenital Cytomegalovirus

Asymmetric Sensorineural Hearing Loss

Cricket

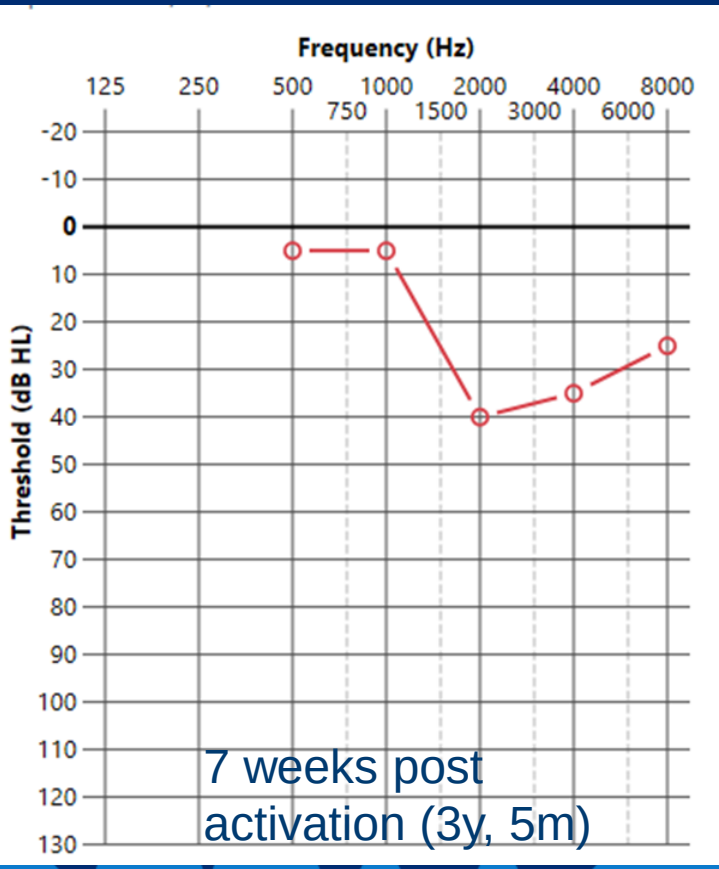
- Born at 34 weeks gestation due to pre-eclampsia
- Birth weight 3 lbs, 10 oz (IUGR)
- CCMV diagnosis at 2 days of age
  - Due to other complications and test results
- Valganciclovir started 6 days after birth - 6 months
- 25 days in NICU due to prematurity
- 1 month of age, seen by Infectious Disease for ongoing management
  - Recommended audiologic and vision evaluations

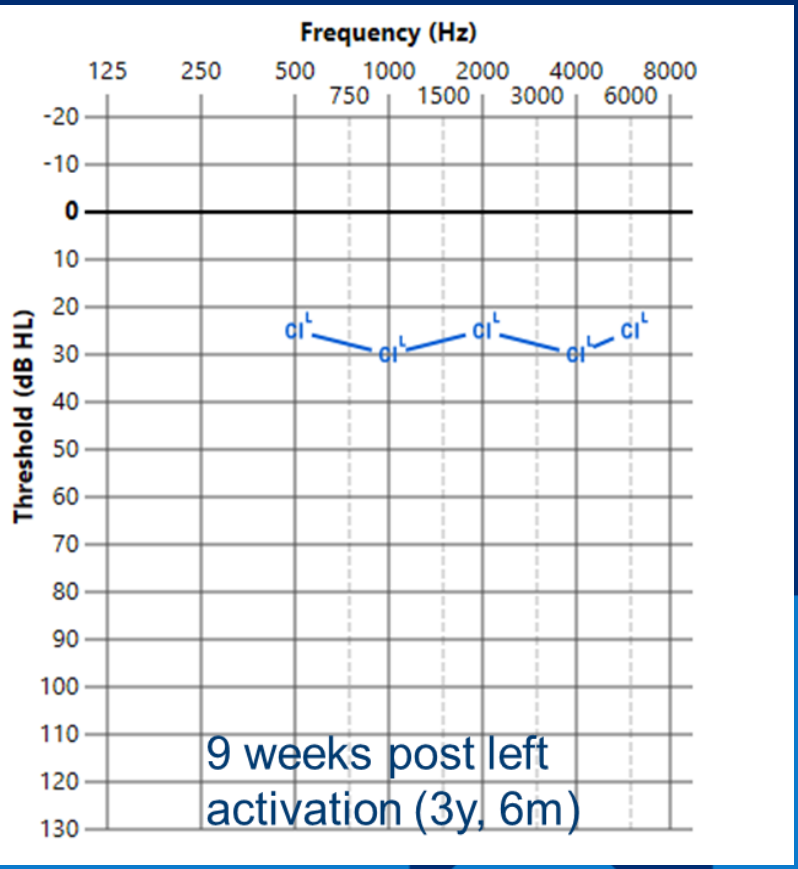
Cricket's Hearing Loss

- Passed newborn hearing screening
- 1st ABR at 54 days of age
  - Right ear: Normal hearing sensitivity
  - Left ear: Mild hearing loss rising to normal hearing sensitivity
- 2nd ABR at 82 days of age
  - Right ear: Normal hearing sensitivity
  - Left ear: Mild to moderate SNHL
- 3 months: Left hearing aid fitting
- 12 months: Suspected decrease in hearing sensitivity
- 14 months:
  - Right ear: Normal hearing 500-1000 HZ, mild SNHL 2000-4000 HZ
  - Left ear: Moderate to moderately-severe SNHL
- 15 months: Bilateral hearing aid fitting

Hearing Loss continued

- 18 months: Increased HA use, 30 spoken words and 5 signs
- 21 months: 70-word vocabulary and using 2-word utterances, suspected decrease in left ear
- 25 months: Decrease in right ear hearing
- 34 months: Unmasked thresholds in severe HL range, new HAF left ear, speech clarity decreasing
- 36 months: CI eval, struggling in background noise, want to continue progress rather than waiting for failure
    - Age appropriate receptive and expressive language skills
    - Decrease in localization reported/observed
    - Fine motor and speech/language milestones within normal limits
    - Gross motor delays


Frequency (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
Threshold (dB HL)
-20
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
7 weeks post activation (3y, 5m)


Frequency (Hz)
125
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Threshold (dB HL)
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-10
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30
40
50
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70
80
90
100
110
120
130
CI L
CI L
CI L
CI L
CI L
9 weeks post left activation (3y, 6m)

Gross Motor Milestones

1. Early intervention for hearing loss and physical therapy
2. **12 months:** Nearly able to sit up on own
3. **14 months:** Sitting un-supported ~2 minutes at a time
4. **15 months:** Scooting on her bottom
5. **28 months:** Walking

Physical Therapy and Motor Milestones

PT evaluation at age 3 years, 3 months prior to cochlear implant surgery

Poor muscle endurance and strength for age

Very unwilling to do any task that taxed her balance

Started PT after CI surgery

Continued PT until March 2020, when clinic closed

Home exercises and good progress noted

Physical Therapy

- Reinitiated at 3 years, 10 months
- Made small gains but plateaued after 1.5 years of therapy
- Enrolled in dance, karate, and gymnastics to help with balance
- Very small for age and challenging to increase strength due to size
- PT recommended an evaluation with a Vestibular Physical Therapist

Note: Normal vision evaluations at 1 and 3 years of age

Physical Therapy Evaluation: 5 years of age

Video Frenzel Goggles:

- Right gaze: right beating nystagmus
- Left gaze: left beating nystagmus
- Upward gaze: no nystagmus
- Downward gaze: no nystagmus

Vestibulo-Ocular Reflex (VOR)

- Horizontal: absent
- Vertical: absent

Head Impulse Test

- Saccades observed bilaterally

One leg balance:

	Left	Right
• Eyes open firm surface	3 seconds	0 seconds
• Eyes closed firm surface	Fall	Fall

Sharpened Rhomberg

- Abnormal

Modified Clinical Test of Sensory Integration of Balance (mCTSIB)

- Falls in conditions with eyes closed

Recommendations:

Vestibular Evaluation

Continue weekly vestibular rehab physical therapy

Vestibular Testing: 5 years of age

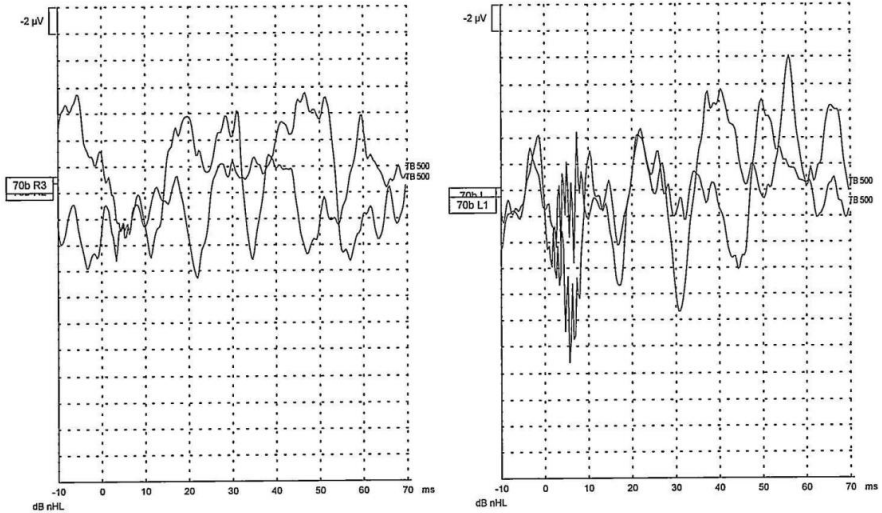
Limited Vestibular Evaluation due to patient age

- cVEMP
- oVEMP
- vHIT
- Rotational Chair

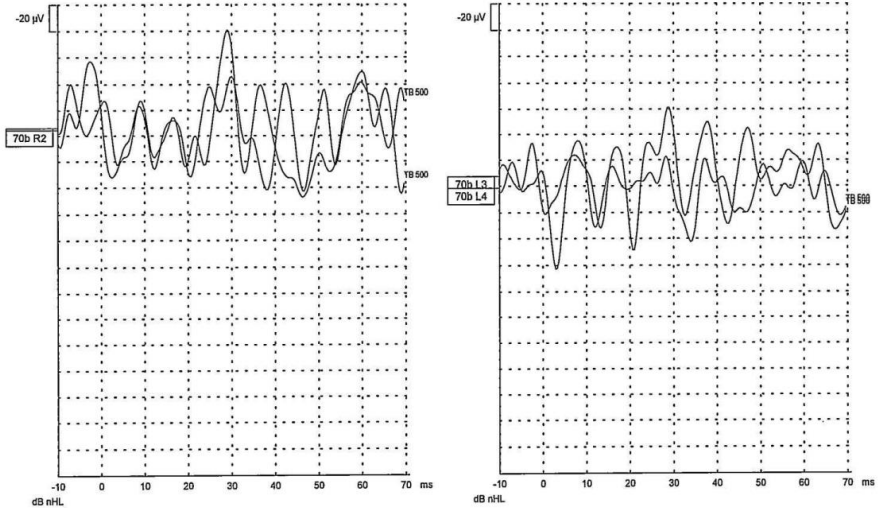
Not included: oculomotor testing, VNG positional testing, caloric testing

Vestibular Evoked Myogenic Potential (VEMP)

oVEMPs: Absent bilaterally

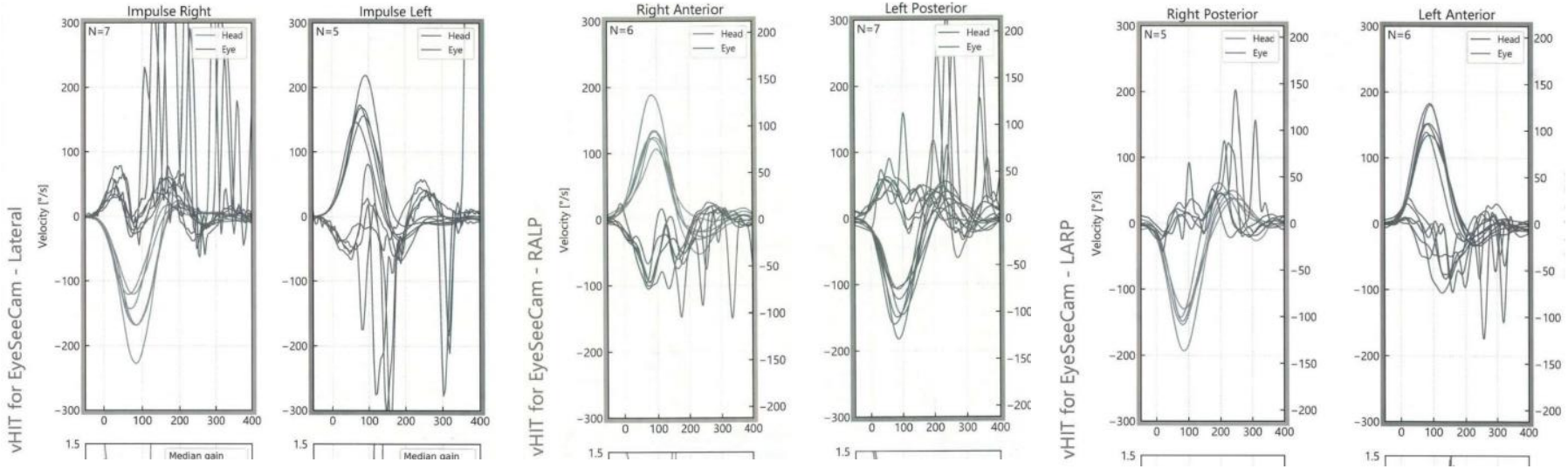


cVEMPs: Absent bilaterally



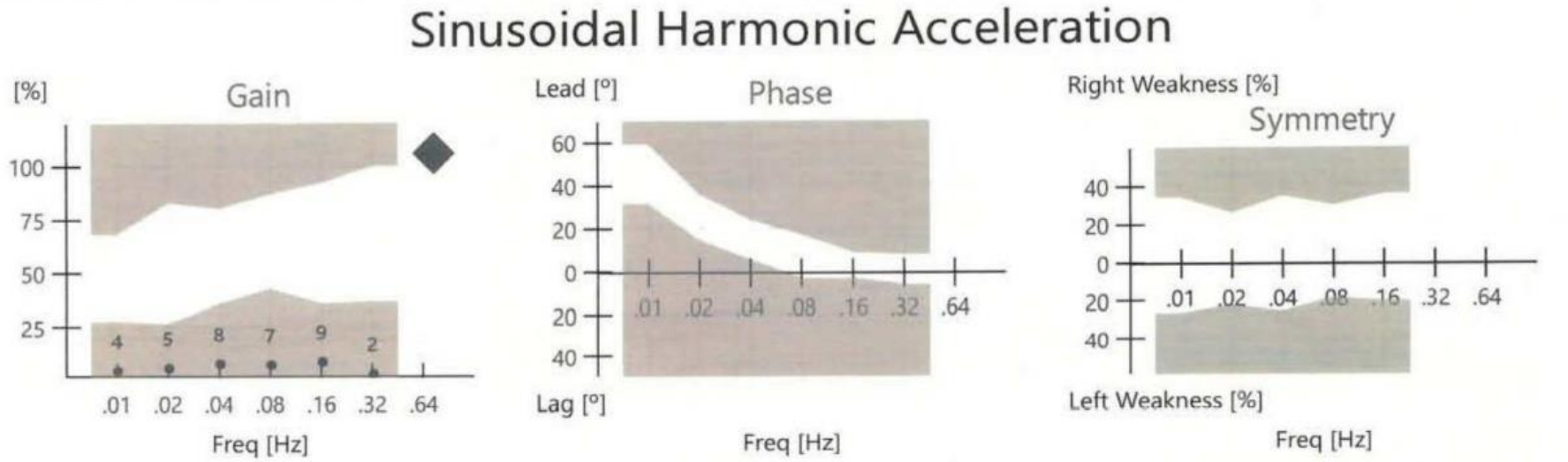
Video Head Impulse Test (vHIT)

vHIT: Low gain and saccades in all 6 semicircular canals

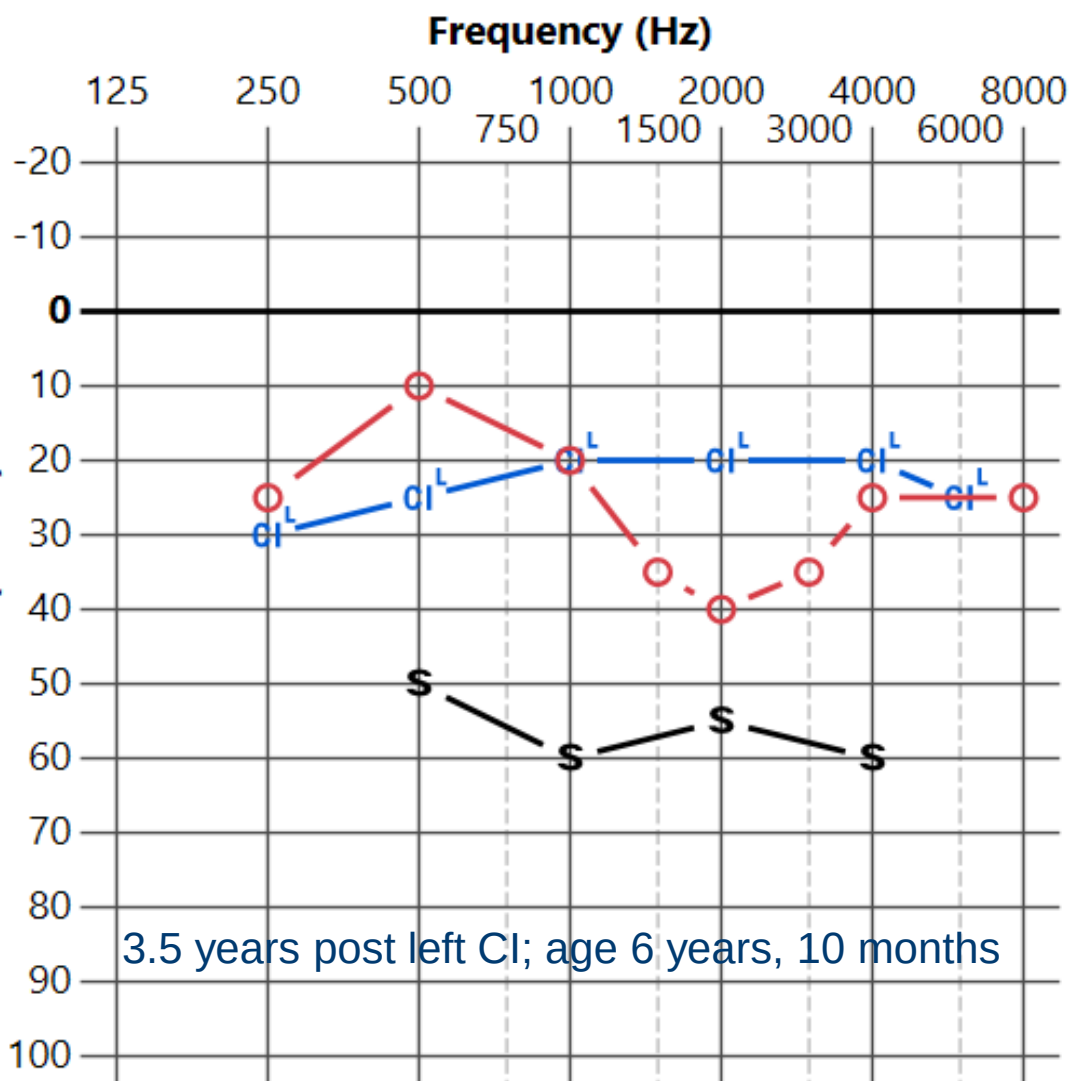


Sinusoidal Harmonic Acceleration (SHA) Rotational Chair Test

Low gain at all frequencies = Bilateral Vestibular Hypofunction



Overall indication: Bilateral vestibular areflexia



6 years, 10 months

- ~3.5 years post left cochlear implant
- Stable hearing in right ear
- Uses hearing aid and cochlear implant full time
- Loves streaming to both
- Thriving in school academically and socially
- Discharged from vestibular rehab
- Adhering to home program
- Has made excellent progress
- Still has an aversion to activities that are challenging or cause her to lose balance
- Incredible family support and follow through

Case Study 2:

Congenital Cytomegalovirus

Normal hearing at birth

Molly

Birth history:

- MOC 14 years of age
- Induced at 37 weeks due to severe intrauterine growth restriction (IUGR)
- NICU 1 week
- Passed NBHS
- Tested for cCMV due to IUGR: positive

Dx AEP at 22 days of age:

- Right ear: Normal hearing, absent DPOAEs, normal tympanogram
- Left ear: Normal hearing, present DPOAEs, normal tympanogram
- PCP referred to Infectious Disease for antiviral treatment—family did not follow up

Lost to follow up until 20 months of age:

- Right ear: Mild hearing loss, Absent DPOAEs
- Left ear: Normal hearing, partially absent DPOAEs

Molly

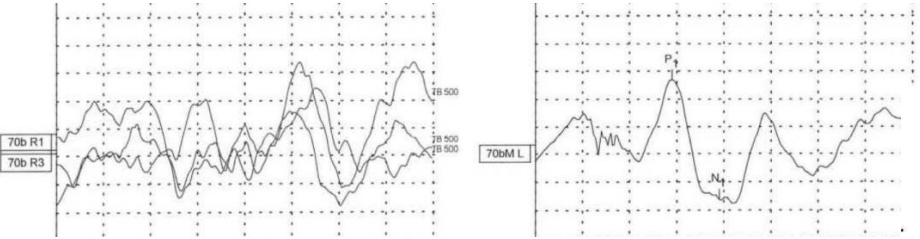
Gross motor delays and balance concerns discussed at hearing test

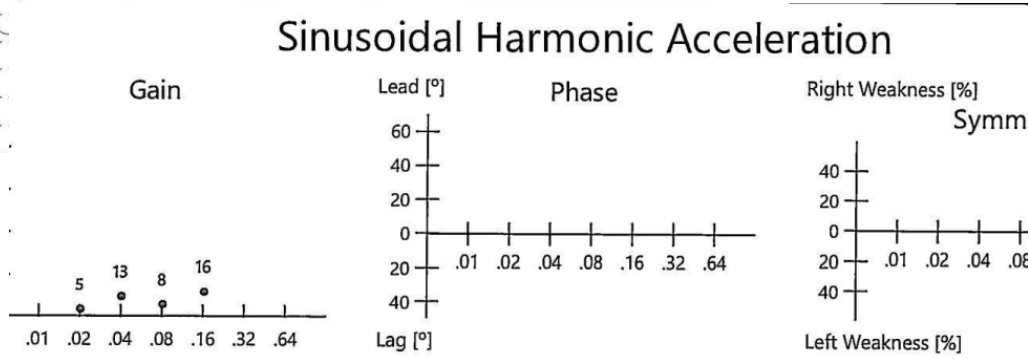
- Not yet walking at 20 months
- Referred for vestibular testing

Vestibular evaluation at 20 months of age

- cVEMP: absent right ear, present left ear
- HIT: Catch up saccades bilaterally
- Rotational Chair: Low gain at all frequencies

→ Bilateral vestibular hypofunction





Molly's Interventions

Interventions

- Recommended they discuss results with her early intervention PT, who recommend they pursue a vestibular PT evaluation at Children's.
  - Continuing PT and OT early interventions weekly

Vestibular Rehab evaluation at 22 months of age

- Peabody Developmental Motor Scales:
  - Stationary: 37 percentile
  - Locomotion: 1 percentile
  - Equivalent age: 13 months
- Interventions planned:
  - Weekly vestibular rehab for 12 weeks (will likely extend)
  - Shoe inserts recommended
  - Home exercise program with caregiver education
  - Balance training
  - Neuro-developmental treatment
  - Motor skills training
  - Gait training
  - Postural training

Molly's Follow Up Recommendations

Sedated Auditory Evoked Potentials evaluation is scheduled

- Hearing aid recommendations pending results of testing
- Monitor hearing every 6 months until age 3 years, then annually until 6 years

Repeat vestibular testing at 3 years of age

- cVEMP
- oVEMP
- vHIT
- Rotational chair

Repeat vestibular testing at 7 years of age

- VEMPs
- vHIT
- Oculomotor
- VOR Suppression
- Subjective Visual Vertical
- Positional testing
- High Frequency Headshake Test
- Caloric Testing


Questions?

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