CMV CONFERENCE, SALT LAKE CITY
OCTOBER 2023

Hearing and Vestibular Monitoring Protocol for Infants and Children with Congenital Cytomegalovirus

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Children's Hospital Colorado
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Affiliated with
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About Us

Karen Hendrick, AuD

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- AuD from University of Washington in 2015

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- Vestibular team member
- AuD from University of Texas at Austin in 2016

Financial Disclosures: Employed by Children's Hospital Colorado.

CHCO Vestibular Program

Number of Vestibular Evaluations

- 2017 = 7
- 2018 = 44
- 2019 = 53
- 2020 = 83
- 2021 = 186 (Rotary Chair installed in January)
- 2022 = 170
- 2023 = 179 (year to date)

TOTAL = 722

Vestibular Appointment Types

Comprehensive Evaluation
- Children ≥ 7 years
- VEMP, vHIT, VNG, Rotational Chair, Caloric

Limited Evaluation
- Infants and children 12 months – 6 years
- VEMP, vHIT, Rotational Chair

Balance Clinic
- Otolaryngology, Audiology, Physical Therapy, Neurology

Vestibular Screen in Colorado Springs
- VEMP, vHIT, Bedside screens
- Combined with a PT evaluation

Vestibular Evoked Myogenic Potential (VEMP) Testing
- Pre-op Cochlear Implant surgery
- cCMV 12-month vestibular screening

Learning Objectives

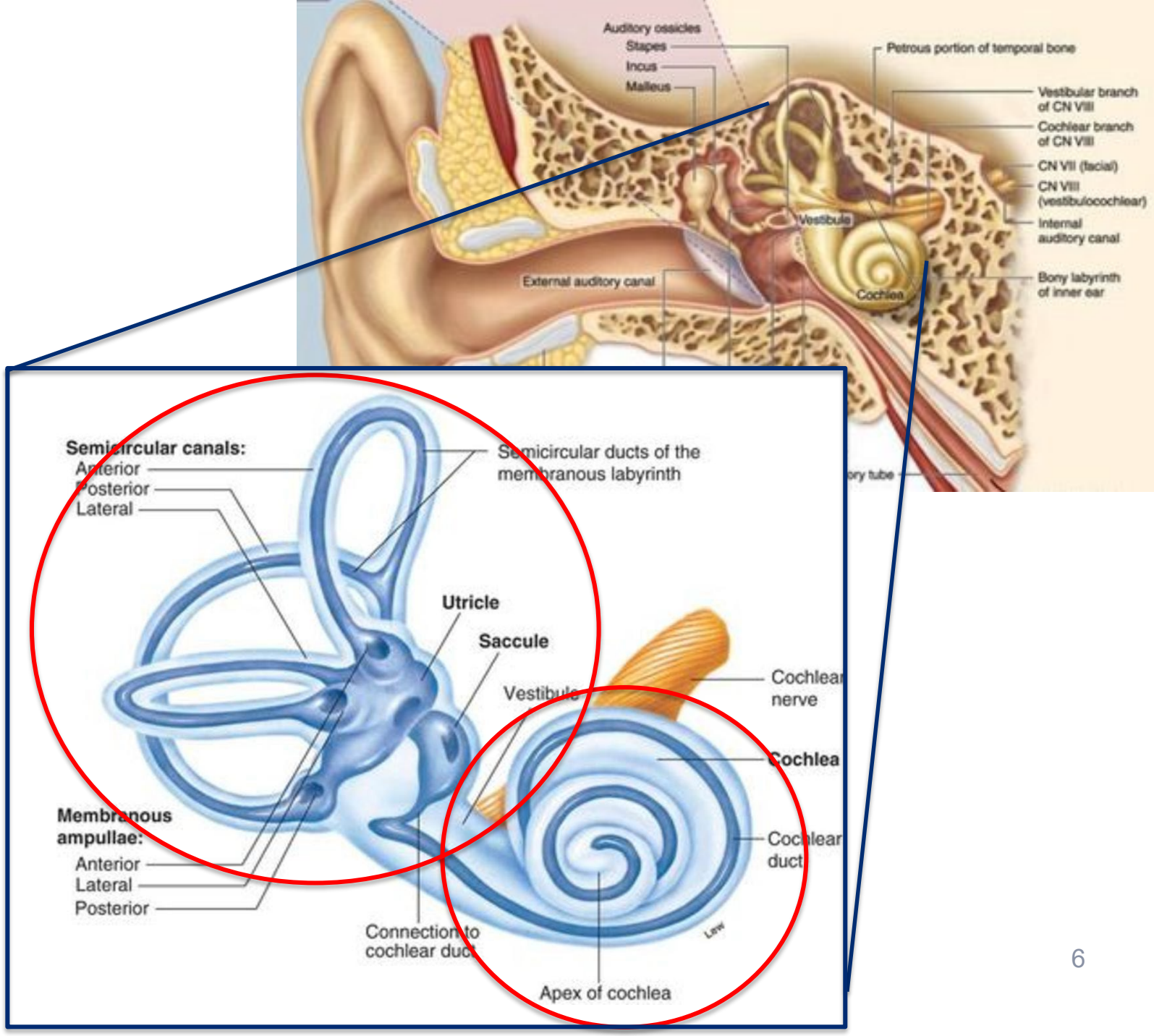
1. Hearing loss and vestibular dysfunction risks with cCMV
2. CHCO hearing and vestibular monitoring guidelines
3. Vestibular test modifications for all ages and developmental levels
4. Vestibular screens at different ages

Anatomy

Hearing:
- Cochlea

Vestibular:
- Utricle
- Saccule
- 3 Semicircular canals

All inner ear structures are connected through a continuous labyrinth



Hearing Loss and Congenital Cytomegalovirus (cCMV)

- It is well established that cCMV can cause sensorineural hearing loss.
  (Dollard et al., 2007; Goderis et al., 2014; Cannon et al., 2014)

- There are national recommendations to closely monitor hearing in children with cCMV.
  - Joint Committee on Infant Hearing (JCIH) Position Statement, 2019: recommends diagnostic follow-up at 3 months of age and annually until age 3
  - American Academy of Audiology (AAA) Position Statement, 2023: recommends diagnostic evaluations every 3-6 months for the first year of life, every 6 months until 3 years of age, annually until 6 years of age

CHCO Hearing Monitoring Guideline for cCMV

- Diagnostic auditory evoked potential (AEP) evaluation at birth/diagnosis of cCMV
- Monitor hearing:
  - every 3 months until 1 year of age
  - every 6 months until 3 years of age
  - annually until 6 years of age

❖ More frequent evaluations may be recommended if results are abnormal or incomplete, or per audiologist recommendation.

Vestibular Function and cCMV

Shears et al., 2022 published a systematic literature review of vestibular function in children with cCMV

- 12 studies performed vestibular tests on children with cCMV.
- Found 10/12 studies showed at least 40% or more of children with cCMV had vestibular loss.
    - Included was Bernard et al., 2015, who found 92% had vestibular loss.
- Vestibular dysfunction was more common in children with symptomatic cCMV, although still occurs in children with asymptomatic cCMV
- 2 studies showed a progression of vestibular dysfunction over time through serial testing

Considerations when developing our protocol:

- Vestibular dysfunction is common in children with cCMV.
- Vestibular dysfunction can occur with cCMV regardless of hearing status.
- Vestibular dysfunction can be progressive in children with cCMV.

CHCO Vestibular Monitoring Guideline for cCMV

- 12 months of age: Cervical vestibular evoked myogenic potential (cVEMP)
- 3 years of age: Limited vestibular evaluation
  - VEMP, vHIT, Rotary 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.

Why is Monitoring the Vestibular System Important?

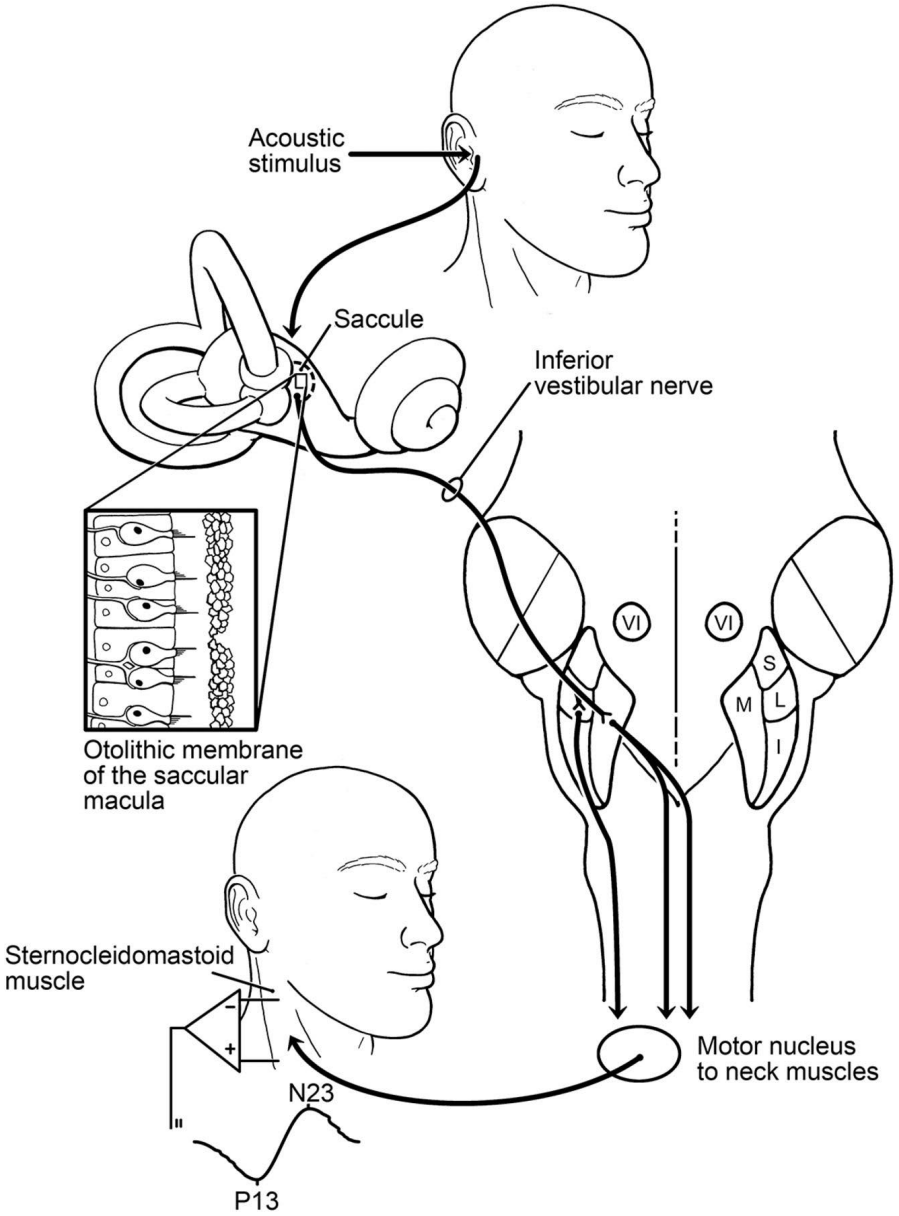
Bilateral vestibular hypofunction:

- Gross motor delays and imbalance
- Increases risk of cochlear implant internal device failure by 8 times (Wolter et al., 2015)
- Associated with deficits in memory, executive function, behavior, and school performance (Bigelow & Agrawal, 2015; Franco, 2008)

How can we reduce these problems?

- Early identification of hypofunction and participation in vestibular rehabilitation improves balance outcomes (Rine, 2018)

Cervical Vestibular Evoked Myogenic Potential



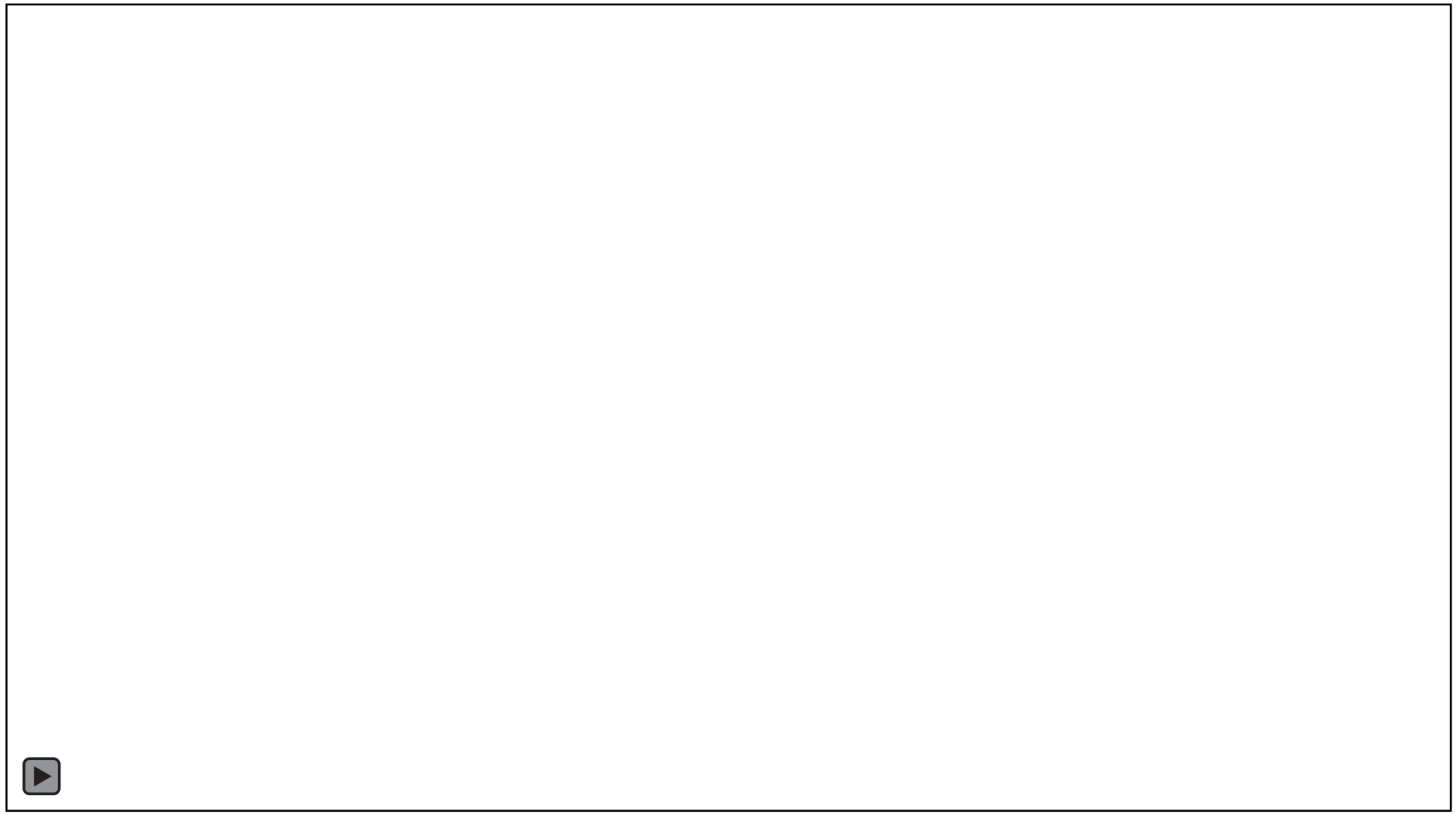
cVEMP: Saccule and Inferior Vestibular Nerve

- Saccule: senses vertical movement
- Utricle: senses horizontal movement (oVEMP)

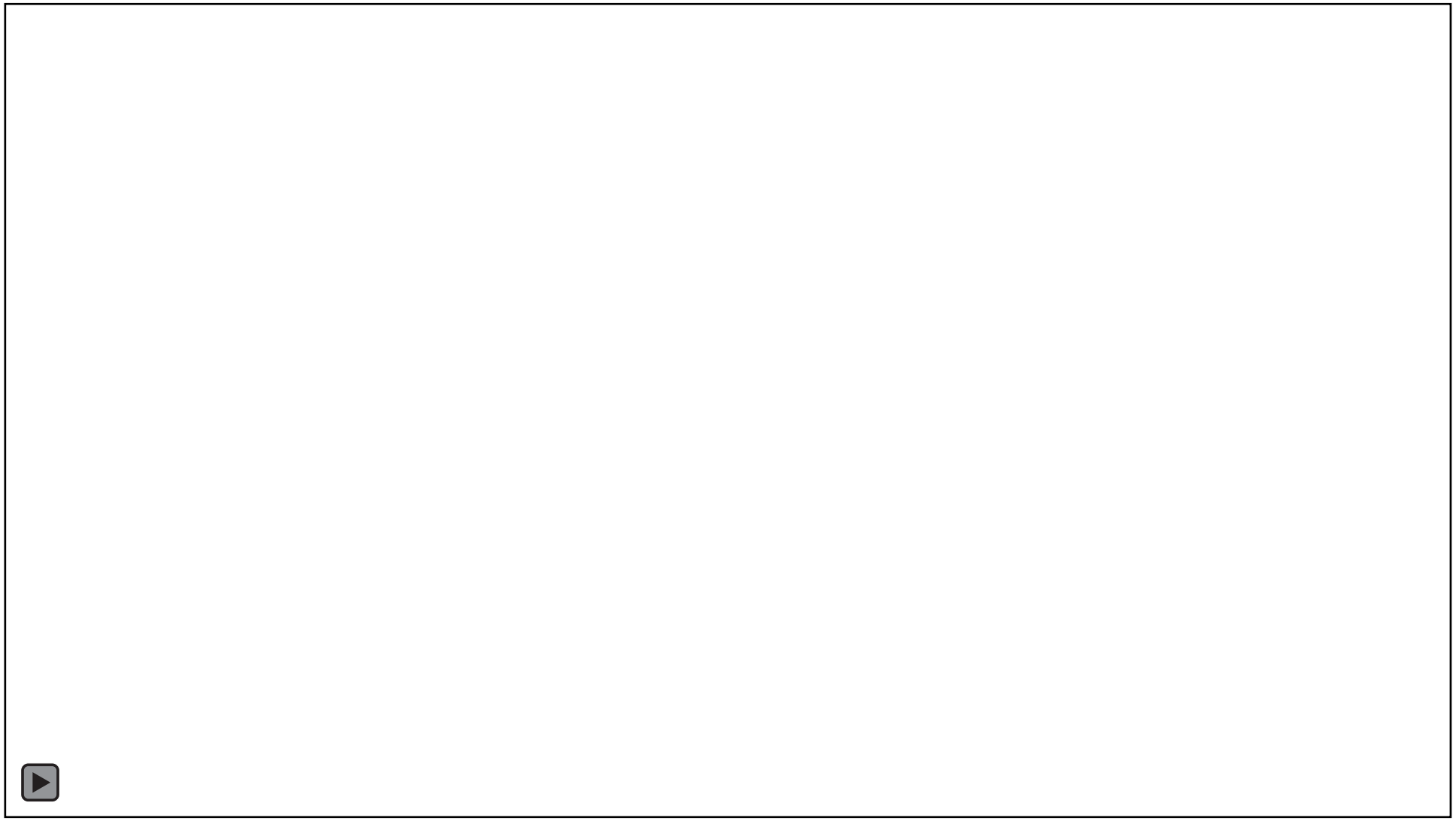
- Can be completed on infants
- Short and non-invasive
- Ear-specific
- Not affected by sensorineural hearing loss
- Air or bone conduction stimulus
- Contraction of the Sternocleidomastoid (SCM)
- Electrodes measure the response sent from the saccule along the vestibulospinal tract

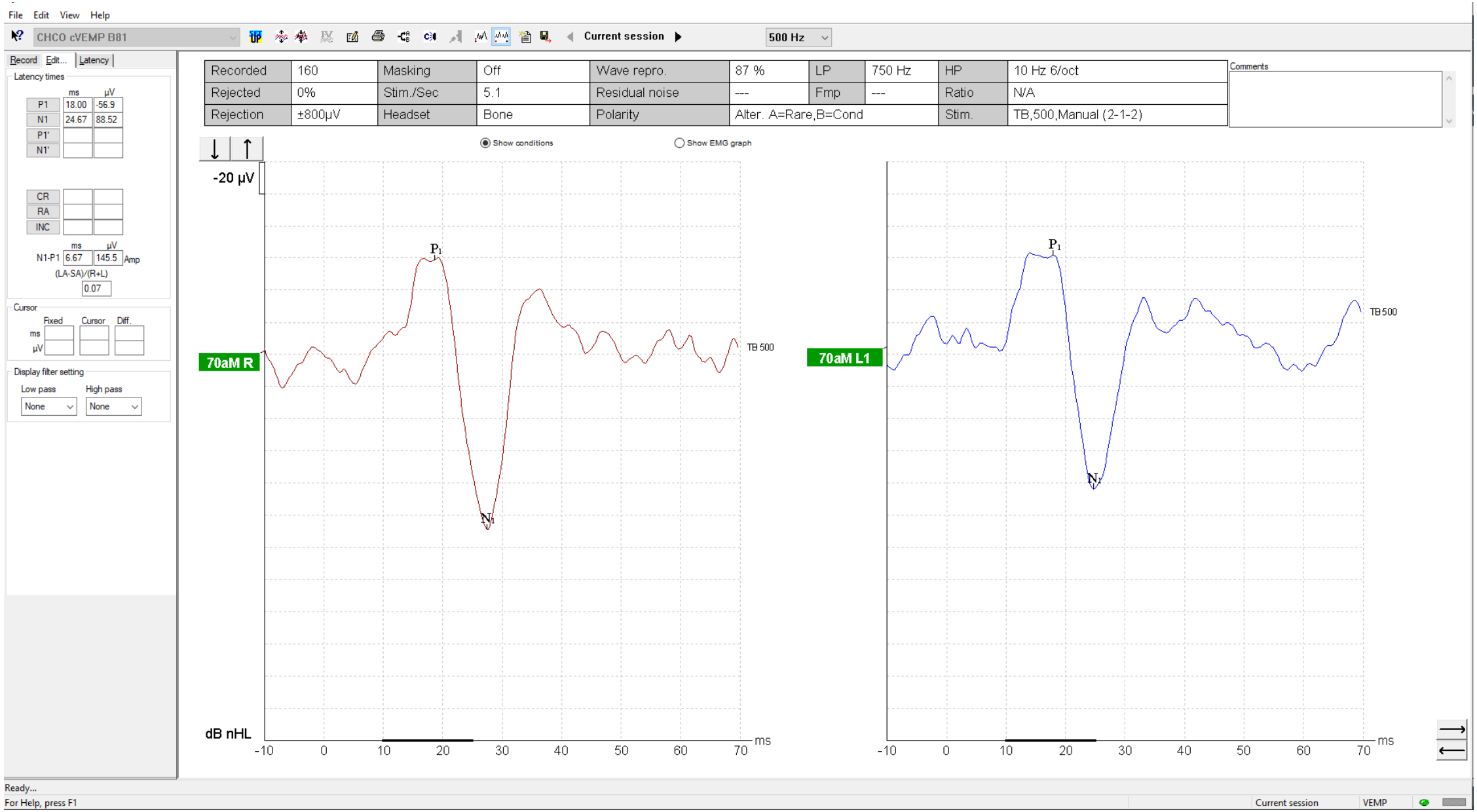
Photos 2 &3:
Fife, T.D. et al., 2017

cVEMP Testing: 12-month-old



cVEMP: Alternative Testing Position




File Edit View Help
CHCO cVEMP B81
Current session
500 Hz
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P1 18.00 -56.9
N1 24.67 88.52
P1'
N1'
CR
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INC
N1-P1 6.67 145.5 Amp
(LA-SA)/(R+L)
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Cursor
Fixed Cursor Diff.
ms
µV
Display filter setting
Low pass High pass
None None
Recorded 160 Masking Off Wave repro. 87 % LP 750 Hz HP 10 Hz 6/oct
Rejected 0% Stim./Sec 5.1 Residual noise --- Fmp --- Ratio N/A
Rejection ±800µV Headset Bone Polarity Alter. A=Rare,B=Cond Stim. TB,500,Manual (2-1-2)
Comments
Show conditions
Show EMG graph
-20 µV
70aM R
TB 500
P1
N1
dB nHL
-10 0 10 20 30 40 50 60 70 ms
70aM L1
TB 500
P1
N1
-10 0 10 20 30 40 50 60 70 ms
Ready...
For Help, press F1
Current session
VEMP

Abnormal cVEMP Follow Up

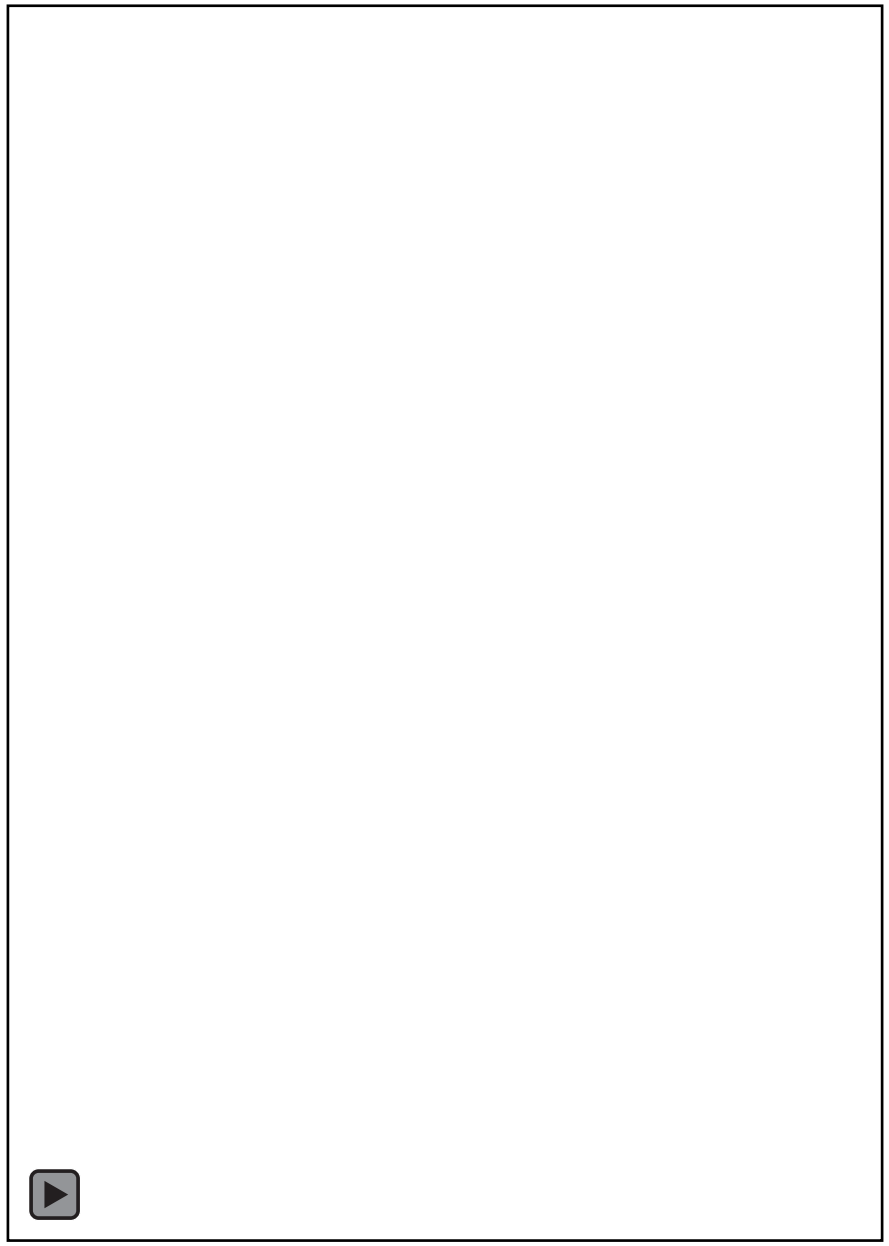
- Asymmetric cVEMPs at 12 months: Repeat in 3 months.
- Absent cVEMP bilaterally at 12 months: Rotational chair testing to assess for bilateral hypofunction.
- Vestibular physical therapy evaluation if there is bilateral hypofunction.

Rotational Chair Testing

Sinusoidal Harmonic Acceleration (SHA) Test

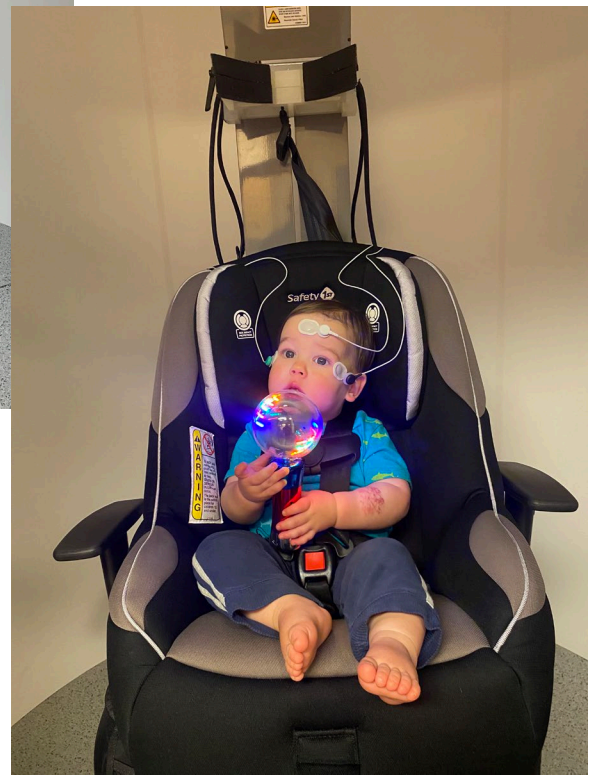
- Shows how the vestibular system senses different speeds of chair rotation.
- Continuous rotation of the chair to the right and left at multiple test frequencies in a blacked-out enclosure.
- Measure nystagmus – eye movement that occurs when there is a functioning vestibular system in response to the chair movement.
    - If there is bilateral hypofunction, there will be no nystagmus.

Nystagmus is measured by video goggles, electrodes, or infrared observation camera.



Pediatric Setup for Rotary Chair

Electrode setup in parent's lap or car seat

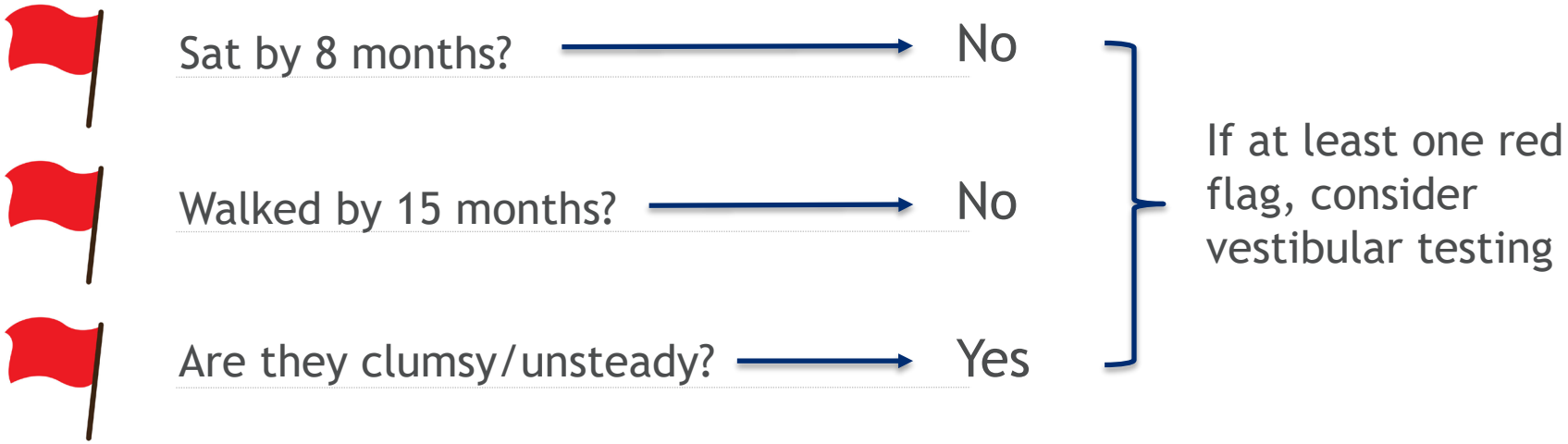


Pediatric Observation Camera

Screening for Possible Vestibular Dysfunction

- Pediatric vestibular centers are limited, and some states do not have this testing for younger ages at all.
- Due to the high prevalence of vestibular dysfunction in children with cCMV and/or sensorineural hearing loss, gross motor delays may be an indicator of vestibular involvement.
- If your patient/child has gross motor delays, vestibular rehabilitation physical therapy should be added and if possible, vestibular testing should be pursued.

Predictive Factors for Vestibular Dysfunction in Children with Permanent Hearing Loss



Children with sensorineural hearing loss, especially moderately-severe or greater, and who have at least one "red flag," should consider a vestibular evaluation.

Janky, K., et al., 2018

Reflex & Gross Motor Screening

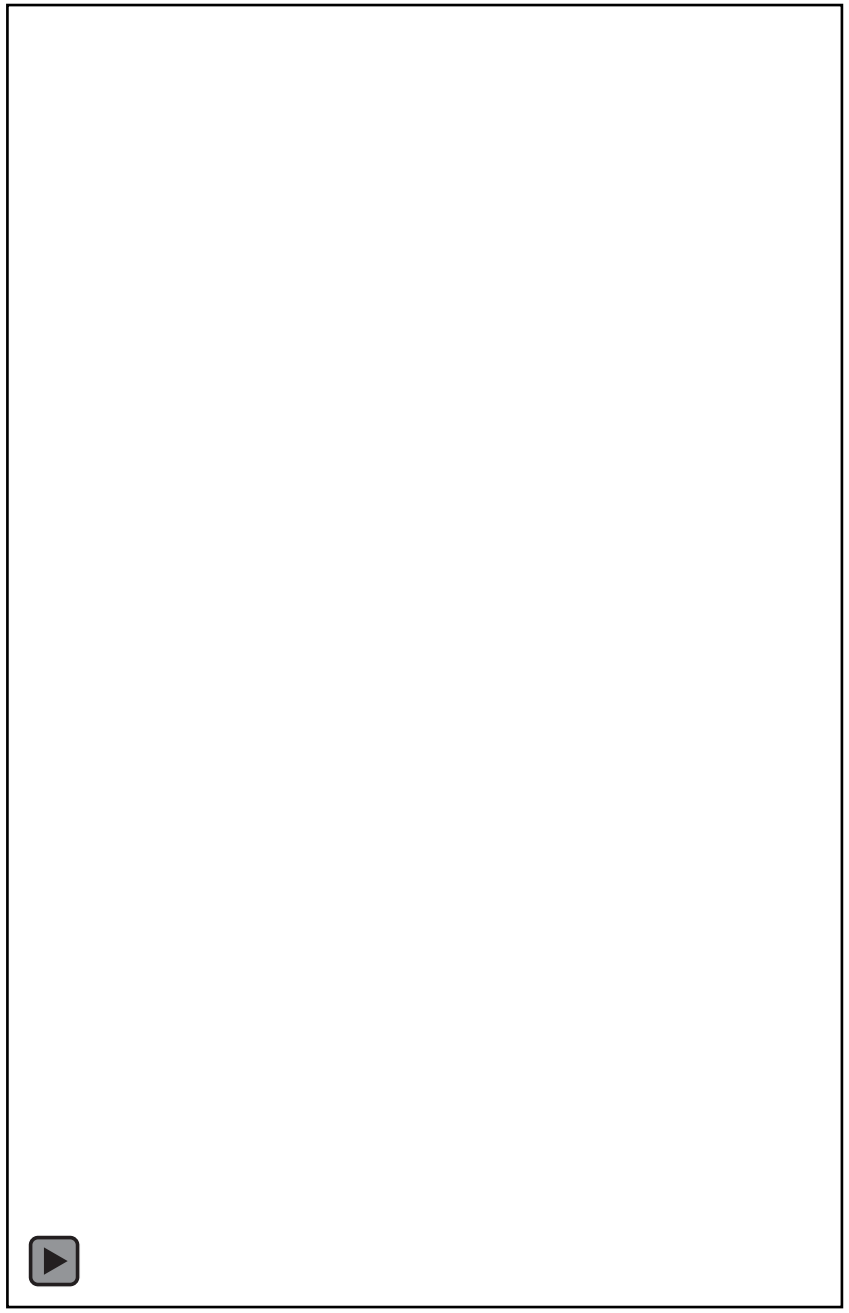
Patient Age	Screening
9+ months	Parachute Reflex
2y – 2y 11m	Stand on **two feet** with eyes closed for 5 seconds
3y – 3y 11m	Stand **tandem feet** with eyes closed for 5 seconds
4y – 6y 11m	Stand on **one foot** with eyes closed for 8 seconds
7+ years	Modified Clinical Test of Sensory Interaction on Balance (mCTSIB)

Parachute Reflex

- Begins to develop around 6 months of age and matures by 12 months of age. Most infants demonstrate by 9 months. (Romeo, D.M. et al., 2009)

- Infant should put their arms outward if they are suddenly moved towards the ground.

- Lack of arm extension may suggest a delay in reflexes/gross motor development.



Gross Motor Screening

2y – 2y 11m
Two feet for 5 seconds
With eyes closed

3y –3y 11m
Tandem feet for 5 seconds
With eyes closed

4y – 6y 11m
One foot for 8 seconds
With eyes closed

Modified Clinical Test of Sensory Integration on Balance (mCTSIB)

Floor
Eyes open

Floor
Eyes closed

Pad
Eyes open

Pad
Eyes closed

Grading: No sway, Some sway, Fall

Vestibular Rehabilitation

Physical Therapy Treatment Strategies: patient specific treatment programs that are goal oriented and based on dysfunction, activity and participation restrictions.

- **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)
- **Compensation/substitution:** alternative strategies for lost or ineffective system
- **Canal re-positional technique (CRT): e.g.,** Epley maneuver
- **Balance Training**
- **Oculomotor Exercises**

Final Takeaways

- Infants/children with cCMV are at high risk for progressive hearing AND vestibular loss.
- Bilateral vestibular hypofunction can lead to problems with balance, cognition, academics and social skills.
- Vestibular testing can be done on infants, young children or children with developmental delays.
- Vestibular Physical Therapy helps reduce the negative consequences of vestibular hypofunction and helps the child meet academic and recreational goals.
- Developmental and gross motor screens can help identify infants/children with cCMV who are likely to have vestibular problems, and they can begin PT without formal vestibular testing.

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Questions?

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