Pediatric Vestibular Dysfunction: More Than a Balancing Act

Kay Bachmann, PhD

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Introduction

1. The learner will be able to identify 3 causes of vestibular disorders in children.
2. The learner will be able to identify at least 3 common symptoms of vestibular dysfunction in children.
3. The learner will be able to administer a short screening to assess balance function in children.

3

Childhood Hearing Loss

- 1.4 in 1,000 newborns have hearing loss (CDC)
- 5 in 1,000 children have hearing loss ages 3-17 yrs (CDC)

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Childhood Vestibular Loss

- Balance disorders may make up .45% of chief complaints per chart review in a Pediatric ENT department. (O'Reilly et al 2010)
- Children with hearing impairment are twice as likely to have vestibular loss than healthy children
- Studies show vestibular loss in 30-79% of children with HL
- There is 10% increase in vestibular loss as a result of trauma from receiving a CI (Jacot et al, 2009)

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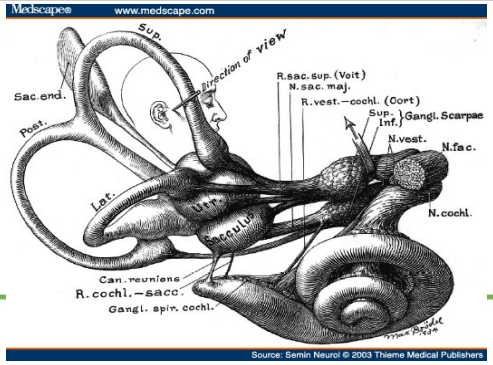

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Common Disorders with Hearing Loss and Vestibular Abnormalities

- Syndromes
  - Usher Syndrome Type 1
  - Pendred Syndrome (also non syndromic Enlarged vestibular Aqueduct Syndrome)
  - Branchio-oto-renal Syndrome
  - CHARGE association
- Cochlear Malformations
- VIII Nerve Defects
- Cytomegalovirus (CMV)
- Meningitis
- Cochlear implant patients

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What Does a Typically Functioning Vestibular System Do?

Two Main Reflexes:

Vestibular Ocular Reflex- Helps hold an object steady when the head/body are in motion.

Vestibular Spinal Reflex- Helps keep our posture and body steady when it senses movement.

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What Does an Abnormal Vestibular System Do?

VOR defect

VSR defect

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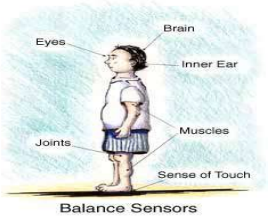
The Balance System

- Vision
- Proprioception
- Inner Ear
  (Vestibular End Organs)
- Central Pathways
  (Brain)



Balance Sensors

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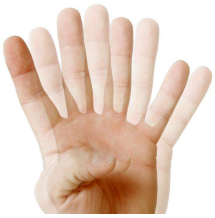
Visual Problems

- Blurriness
- Double vision
- Difficulty focusing
- Headaches
- Eye strain
- Difficulty with or avoidance of reading



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Visual Problems

- Symptoms with reading
  - Horizontal deficits
- Symptoms when looking up (at the board) and down (at the desk) or while walking up/down the stairs
  - Vertical deficits
- Symptoms while running and trying to focus on a target (i.e. ball, goal)
  - Gaze stabilization

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International Journal of Pediatric Otorhinolaryngology (2006) 70, 1957–1965

ELSEVIER

International Journal of Pediatric Otorhinolaryngology

www.elsevier.com/locate/ijporl

Evidence that vestibular hypofunction affects reading acuity in children

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

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Methods

- 72 children with normal hearing/normal vestibular system
- 5 children with vestibular loss and SNHL
- 9 children had normal vestibular tests and SNHL
- Each was asked to read a series of words at a comfortable pace.
- Each passage became sequentially smaller in print size.

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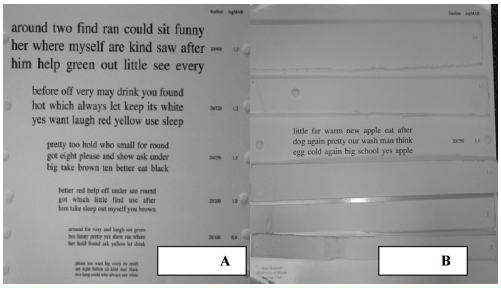

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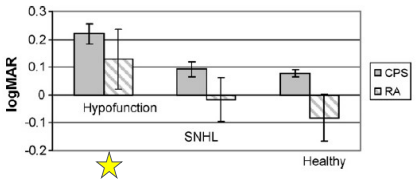
Results



- CPS: Critical print size
- RA: Reading Acuity

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Discussion

The normal hearing children and the children with SNHL (normal vestibular function) scored statistically the same.

What does this tell us about the effects of language impairment vs. vestibular impairment?

Suggestions for increasing font size for these children in school.

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Balance Problems

- Unsteady
- Falling
- Clumsy
- Not keeping up with peers
- Difficulty in areas of low light
- Difficulty on uneven or angulated surfaces

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Year : 2013 | Volume : 29 | Issue : 3 | Page : 199-201

Clinical balance tests for evaluation of balance dysfunction in children with sensorineural hearing loss

Eman A.F. Said

Audiology Unit, Department of Otolaryngology, Assiut College of Medicine, Assiut, Egypt

	NH children	HI children	*P* value
BOT-2			
Standing with feet apart on a line			
Eyes open			
Row	10±0.00	10±0.00	–
Point	4.0±0.00	4±0.6	–
Eyes closed			
Row	9.06±1.4	7±1.4	0.000**
Point	3.9±0.58	2.9±0.36	0.000**
Walking forward on a line			
Eyes open			
Row	6±0.0	5.6±0.6	0.372
Point	4.0±0.00	3.75±0.4	0.423
Standing on one leg on a line			
Eyes open			
Row	9.7±1.7	7±1.2	0.001**
Point	3.9±0.69	2.3±0.49	0.02*
Eyes closed			
Row	8.6±2.4	4.4±0.7	0.001**
Point	3.6±0.39	1.8±0.5	0.000**
Walking forward heel to toe on a line			
Eyes open			
Row	6±0.5	4.0±0.7	0.01*
Point	4±0.7	2.7±0.4	0.01*
Total point score	22.8±2.7	17.4±3.5	0.01*
mCTSIB			
Standing on a firm surface			
Eyes open	10±0.00	10±0.00	–
Eyes closed	9.00±1.4	6.1±1.3	0.02*
Standing on a foam cushion			
Eyes open	9.5±2.5	7.3±1.8	0.04*
Eyes closed	8.3±2.6	4.5±0.6	0.001**
Stand on one leg			
Eyes open	9.03±1.7	6.8±1.2	0.01*
Eyes closed	8.6±2.4	4.4±0.7	0.001**
Tandem stand			
Eyes open	9.9±2.04	8.2±1.8	0.372
Eyes closed	9.2±0.62	5.17±1.5	0.04*

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What We Know

- Children with vestibular loss were thought to compensate on their own due to neural plasticity
  - Latest research (Janky and Givens 2015):

    "Children with vestibular loss do not naturally recover to levels of their healthy peers, particularly with activities that utilize vestibular input and that attention should be given to vestibular loss in the pediatric population."

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International Journal of Pediatric Otorhinolaryngology (2004) 68, 1141–1148

International Journal of Pediatric Otorhinolaryngology

www.elsevier.com/locate/ijporl

Improvement of motor development and postural control following intervention in children with sensorineural hearing loss and vestibular impairment☆

Rose Marie Rine*, Jennifer Braswell, Donna Fisher, Kelly Joyce, Kristen Kalar, Margaret Shaffer

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Methods

25 children (3-8 yrs old) with SNHL

Tested through rotary chair, CDP, and Peabody Motor Development test

Separated by diagnosis: normal (4), and bilateral vestibular hypofunction (21)

Given treatment exercises or placebo exercises for 12 weeks (30-minute sessions 3x weekly)

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Exercises

- Designed to work on hand-eye coordination, balance, visual motor exercises, visual/somatosensory awareness
- Placebo group had language therapy

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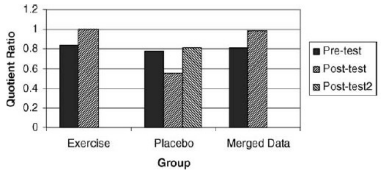
RESULTS



Various motor tasks including balance tasks

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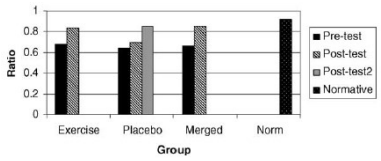
RESULTS



Somotosensory ratio results:

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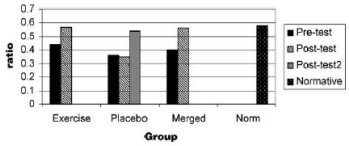
RESULTS



Vision ratio scores:

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Discussion

All the children showed improvement.

The author noted the importance of the young age of participants (3-8 years old).

Another study sited used older adolescents and teens and did not show the same improvement.

The author reported previously published data showing compounding and progressive motor skills.

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Symptoms of Vestibular Dysfunction in Children with HL

- ~~Complaints of vertigo~~
- ~~Difficulty driving~~
- ~~Unable to go to work~~
- ~~Roaring tinnitus~~
- Behind learning to walk
- Falling
- Difficulty with reading acuity
- Not keeping up physically with their peers
- Other issues: Cognitive? Academic? Mental health?

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Screening in the Literature

- Recommendations from:
- Boys Town National Research Hospital (2018)
- Hospital for Sick Children, Toronto CAN (2016)
- Thoughts from Cincinnati Children's Hospital

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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]

American Journal of Audiology • Vol. 27 • 137–146 • March 2018

- Retrospective chart review of 186 charts

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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]

Each child's medical chart was reviewed to obtain the following data:

- vestibular loss severity (classified as normal, bilateral, or mild to moderate)
- degree of hearing loss (bilateral pure-tone average [PTA])
- imaging abnormalities
- parental concerns for gross motor delay
- parent report of age when their child sat (months)
- walked independently (months)
- comorbidities
- score on the Developmental Profile-3.

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Findings

- Of these factors which had the greatest ability to differentiate children with vestibular loss from children with normal vestibular function:
  - age-to-sit (cut off 7.25 months)
  - age-to-walk (cut off 14.5 months)
  - PTA (cut off of 66 dB)
  - parental concerns for gross motor delay

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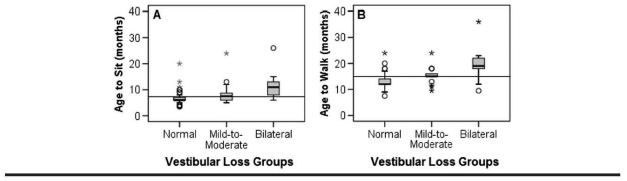
Figure 1. Both age-to-sit (A) and age-to-walk (B) increase as the severity of vestibular loss increases. The line in each panel represents the cutoff values from receiver operating characteristic analysis.



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Otology & Neurotology
37:926–932 © 2016, Otology & Neurotology, Inc.

Using Balance Function to Screen for Vestibular Impairment in Children With Sensorineural Hearing Loss and Cochlear Implants

*Modupe Oyewumi, *Nikolaus E. Wolter, †Elise Heon, ‡Karen A. Gordon, §Blake C. Papsin, and §Sharon L. Cushing

- 45 children with total vestibular loss and CI
- 20 peers with normal hearing
- Bruininks Oseretsky Test of Motor Proficiency II (BOT-2)

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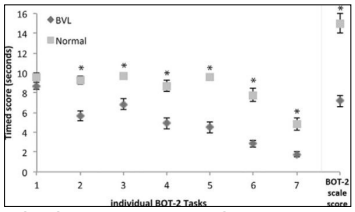

- FIG. 1. Comparison of performance on overall BOT-2 score and individual tasks of the BOT-2 balance subtest by group for children with and without bilateral vestibular loss (TBVL).

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Recommendations from Toronto Group

Children with hearing loss ages 4 yrs 8 months and older:

1) One-foot standing—eyes open (cutoff 8 s).
2) One foot standing—eyes closed (cutoff 4 s).
3) Tandem stance—eyes closed (cutoff 8 s).

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- What else could we do to catch the kids with unilateral loss or mild/moderate vestibular losses?

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Screening Protocol with Ohio Valley Voices School (protocol is evolving)

- Local oral/aural school now screens children in the fall
  - **SCREENING OF visual issues**
  - Head Thrust
  - Dynamic Visual Acuity assessment
  - **SCREENING OF balance issues**
  - Foam challenge
  - Standing on one foot
  - Ages and Stages

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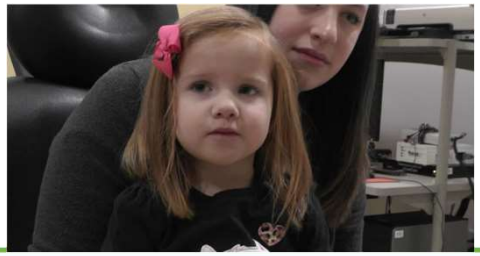
Head Thrust



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Head Thrust-Video

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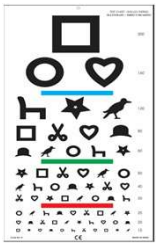
DVA Screening

- Have patient stand 10 feet from eye chart
- Obtain static vision acuity
- Obtain dynamic vision acuity with headshake
- Increase of greater than 2 lines is abnormal



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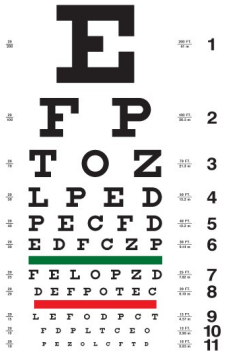
DVA Screening



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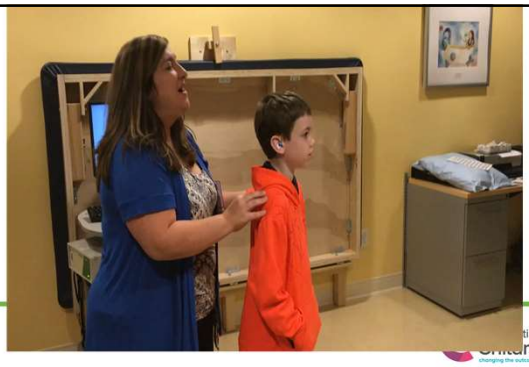

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Modified Clinical Test of Clinial Test of Sensory Interaction on Balance (m-CTSIB)

- 4 conditions
  - Stable surface, eyes open
  - Stable surface, eyes closed
  - Neurocom foam, eyes open
  - Neurocom foam, eyes closed

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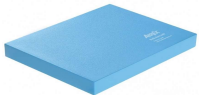
Other Options: Compliant Surface

- Airex balance pad



- Pillow

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Typical 5 year old (10 seconds)

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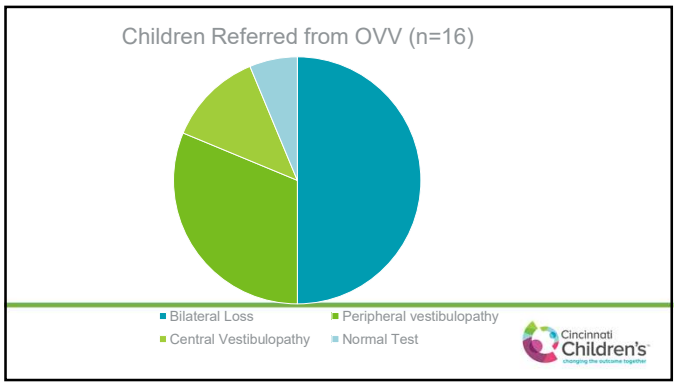

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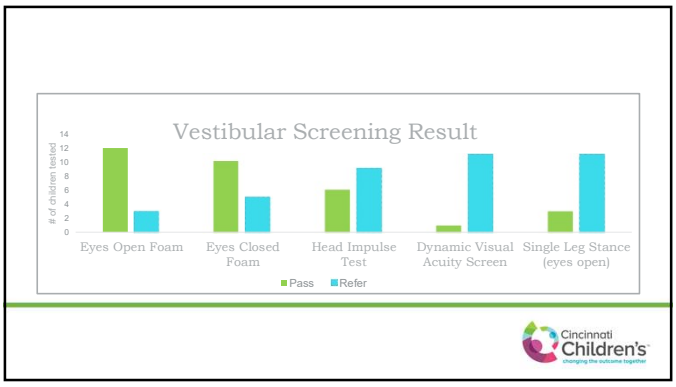

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Next Phases

- Run statistics on which test is most sensitive/specific
- Remember, all of these children have significant hearing loss (how do we translate to those with less hearing loss)?

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What can we do for these children once they are found to be at risk for a vestibular disorder?

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Cases

Cincinnati Children's

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Pendred Syndrome

- caused by changes/ mutations in the gene *SLC26A4* on chromosome 7.
- Recessive trait: needs to inherit two mutated *SLC26A4* genes—one from each parent—to have Pendred syndrome.
- Goiter or large thyroid may develop in adolescence or adulthood.
- Bilateral hearing loss, often progressive
- Enlarged Vestibular Aqueducts (all children with confirmed EVA get screenings)
- 5 to 10 percent of hereditary hearing loss
- protein that the *SLC26A4* gene makes, called pendrin, found in the inner ear, kidney, and thyroid gland.
- Researchers have identified more than 150 deafness-causing mutations or alterations of this gene.

Cincinnati Children's

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Pendred Syndrome

- 7 year old
- Did not pass balance screening at school
- Bi-modal (CI right-HA left)
- History of torticollis
- Walked at 18 months
- Duane's Syndrome (affecting left eye)
- Rides a bike with training wheels
- Difficulty standing on one foot (i.e. getting dressed or putting on shoes, getting out of tub)
- Greater difficulty standing on right leg per parent
- Holds on to railing going up and down stairs

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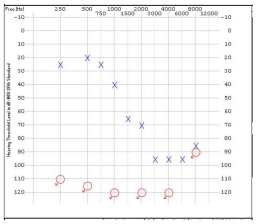
Pendred Syndrome



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Our little girl

- **SCREENING OF VOR**
- Head Thrust (catch up saccade turning both directions)
- Dynamic Visual Acuity assessment (failed x 4 lines)
- **SCREENING OF VSR**
- Foam challenge (could not stand foam eyes closed)
- Standing on one foot (could not stand at all on right leg, 2 seconds on left leg)
- Ages and Stages (showed at risk for not meeting milestones)

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Formal Vestibular Evaluation

- Posturography
- Rotary Chair
- Videonystagmography
- Vestibular Evoked Myogenic Potentials
- Video Head Impulse Test

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Conclusion for our case?

- Total right peripheral vestibulopathy
  - Uncompensated
  - Might be unstable- meaning it still flares up (she never has reported vertigo!)
  - Referred for vestibular rehabilitation
  - You need vision and proprioception to be working 100% to recover easily, she has an ocular motility issue

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Vestibular Rehab Eval

- Single leg stance: 1-2 seconds per trial
- Standing on Airex (unstable surface) 2-5 seconds per trial
- Wants to ride a 2 wheeler

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Vestibular Rehab Evaluation

Subtest scaled scores have a mean of 10 and a standard deviation of 3. Test results indicated:

Performance was **well-below average** in Balance . (Subtest Scaled Score 5 or less)

Subtest	Max Point Score	Pt's Total Score	Scaled Score	Low End	High End	Age Equivalence	Z-score
5 Balance	37	15	3	2	8	below 4	-2

Comments: *The scores that include single leg stance reflect* [redacted] *balance when using right (laterality of vestibular hypofunction) foot for balance.*
Standing with feet apart on a line - eyes open: 9.6 sec
Walking forward on a line: 6 steps
Standing on one leg on a line - eyes open: 1.5 sec
Standing with feet apart on a line - eyes closed: 1.8 sec
Walking forward heel-to-toe on a line: 2 steps
Standing on one leg on a line - eyes closed: 0.8 sec
Standing on one leg on a balance beam - eyes open: 2.8 sec
Standing heel-to-toe on a balance beam: 10.3 sec
Standing on one leg on a balance beam - eyes closed: 0.6 sec

Balance: standing
Single limb stance: 1.5 seconds on the right, 7.4 seconds on the left

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Pendred Syndrome – Vestibular Rehab Treatment

- Working on:
  - Eye-head movements
  - Balance exercises
    - Standing on one foot
      - » Modified initially
    - Eyes closed
    - Uneven or compliant surfaces
    - Obstacle courses

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Home Exercise Program

- Single leg stance, finger tips on counter top progress to arms out like superman eyes open/eyes closed
- Walking with head turns iPad “hidden pictures”
- standing with feet on pillow with eyes open/eyes closed,
- VOR x 1 with flashcards, seated

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Vestibular Rehab Update-3 months Later

- Goal: maintain single leg stance on RLE x 6 seconds
  - At session, 3-6 seconds per trial
- Goal: demonstrate improved vestibular balance control as evidenced by maintaining balance on Airex with eyes closed x 15 seconds
  - 5-15 sec per trial with one trial x 26 sec
- Working on riding that bike!

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Pendred Syndrome: At Discharge

- Every other week x 4 months
  - 8 total visits with focused HEP
- Single leg stance x 10 sec each leg
- Maintains balance on foam pad with eyes closed 20-30 sec each trial
- "She just got on her bicycle and started riding!"

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Thank you!
Questions???

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Cincinnati Children's
changing the outcome together

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