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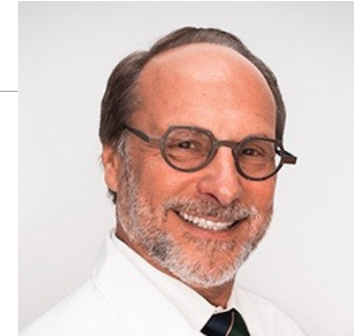
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Pediatric Balance Assessment: It's as Simple as A-B-C, in partnership with the American Academy of Audiology

Richard E. Gans, PhD, CCC-A, DFAA



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Dr. Gans received his PhD from The Ohio State University. Over his 40-year career he has presented or published 150 presentations, research articles and textbook chapters and presented in 38 countries. He served as an American Academy of Audiology Board of Directors for 6 years serving as PAC Chair, Treasurer and President (2004-05). He was awarded the Academy's Distinguished Achievement Award (2016) and is a Legacy Distinguished Fellow (DFAAA). He is a Life Member of ASHA and a Research Scientist Member of the American Academy of Otolaryngology-HNS and the American Academy of Neurology.



Disclosures

- **Presenter Disclosure:** Financial Disclosure: Dr. Gans is the founder and CEO of the American Institute of Balance. Non-financial Disclosure: Dr. Gans has no relevant non-financial disclosures.
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- As healthcare is rapidly evolving, new findings may change the validity of the information presented in this course. Providers should always consult the latest research and guidelines from professional associations.
- The interventions discussed in this course may be limited in generalizability, requiring practitioners to consider specific individual and population factors.
- The course may not cover all possible risk factors, diagnostic methods, or treatment options, and complex cases may require additional considerations.
- Healthcare providers should apply cultural competence and sensitivity to ensure effective and respectful care based on their patients' diverse needs.
- Providers should always evaluate the limitations of diagnostic and screening tools, considering their potential for false results as well as any ethical implications.
- It is the responsibility of course participants to critically evaluate the evidence from multiple sources to guide professional practice.



Learning Outcomes

After this course, participants will be able to:

- List the 3 most common congenital and/or acquired conditions.
- Describe 3 behavioral and electrophysiological assessment protocols.
- Explain the best techniques for how to administer C-EMP to neonates.



Course Information

Pediatric Balance Evaluation: It's as Simple as A-B-C



Development

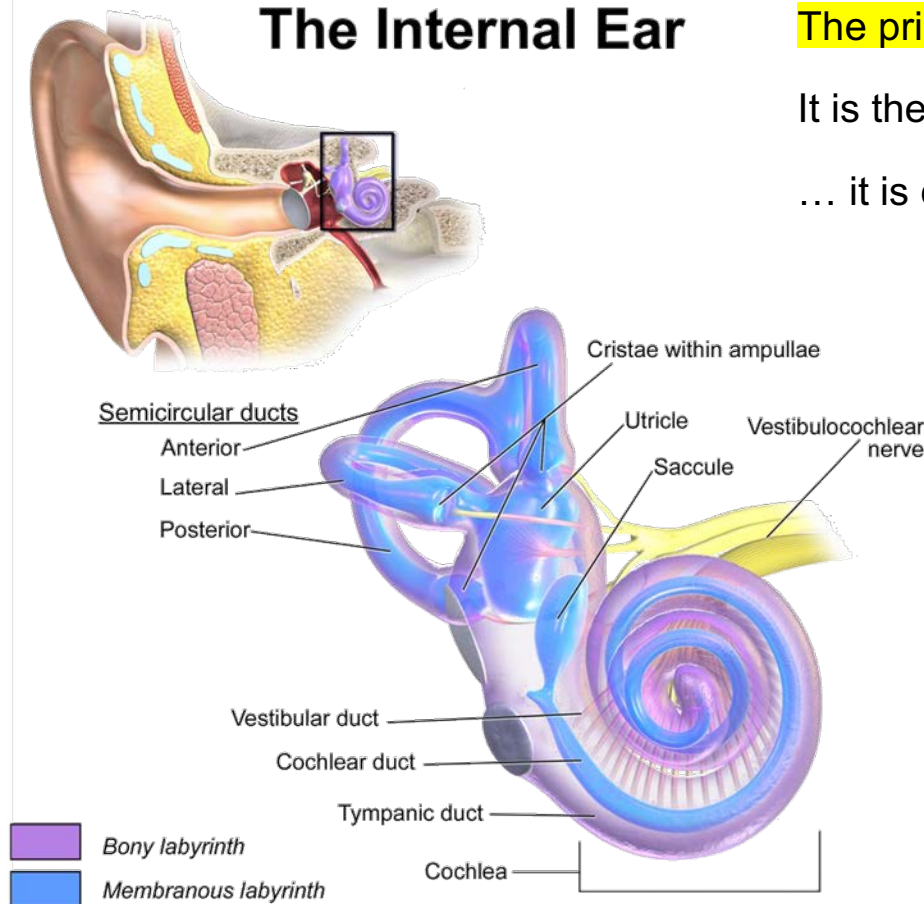
✓ **Embryological Development**

The Internal Ear

The primary function of the inner ear is equilibrium.

It is the gravity and velocity sensor and response system

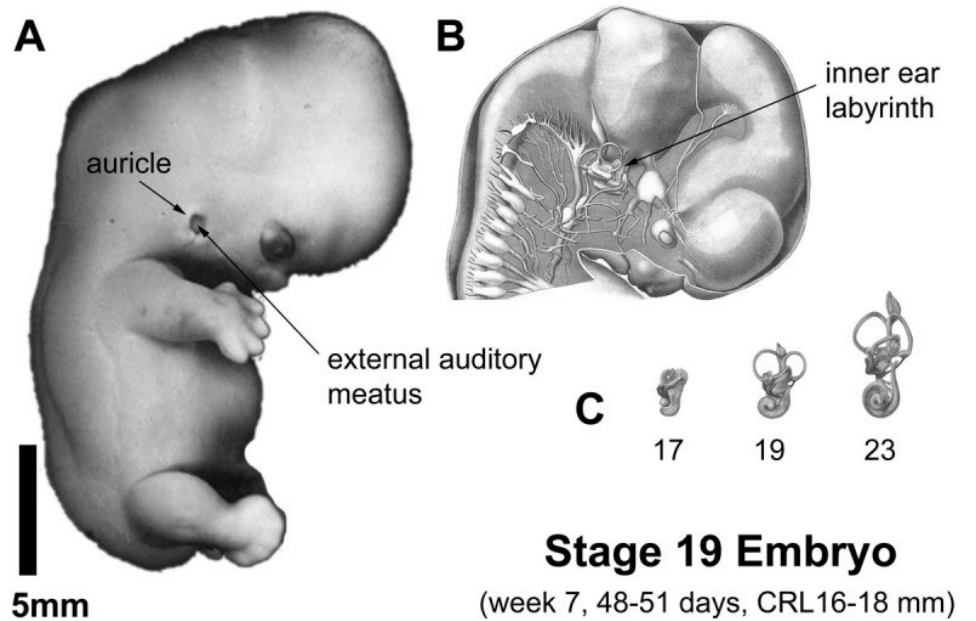
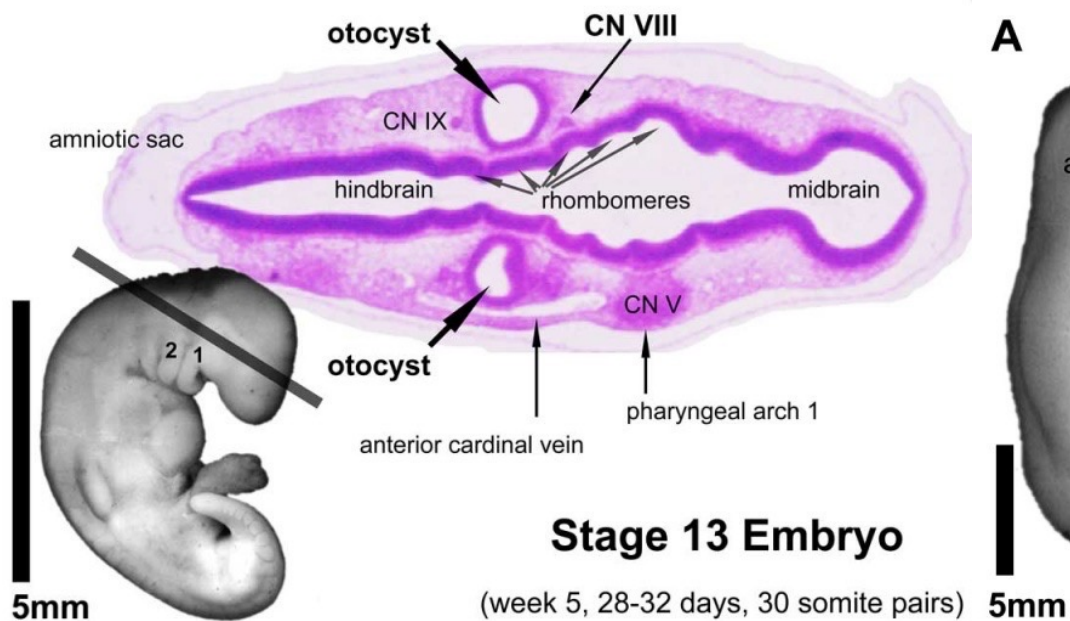
... it is critical to survival



- The vestibular system precedes the cochlea which develops from the saccule.
- Conditions which cause a defective cochlea there may first have first caused a labyrinthine malformation or dysfunction.

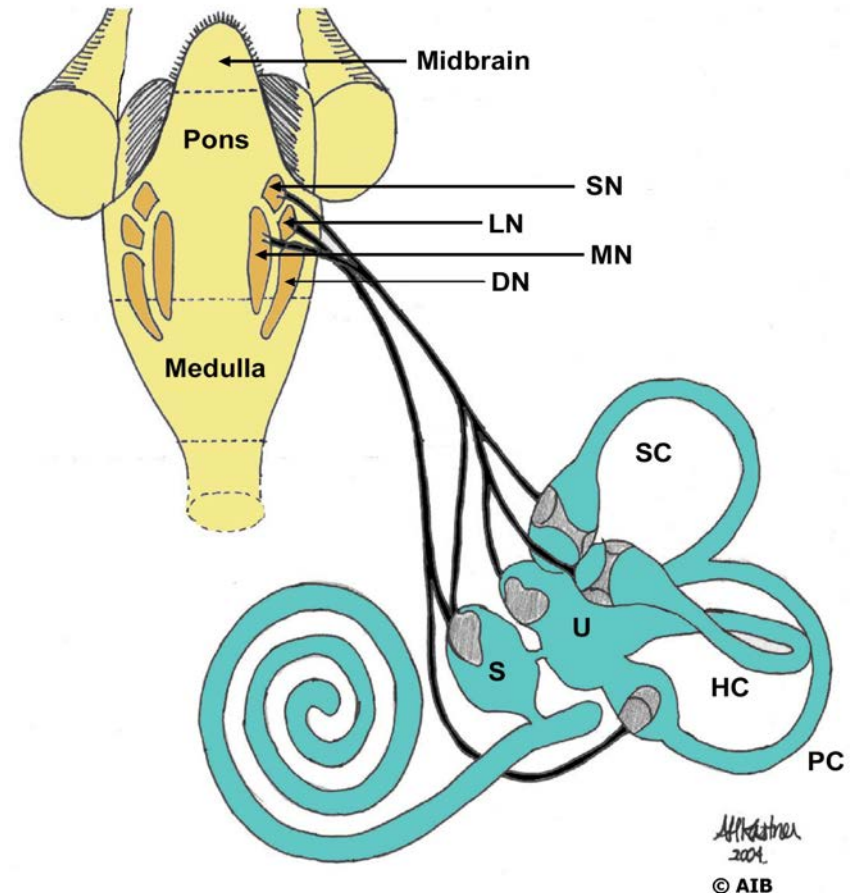
✓ Embryological Development

- Phylogeny- vestibular is the 1st sensory system to develop.
- Vestibular end-organ complete by 49th day of gestation.



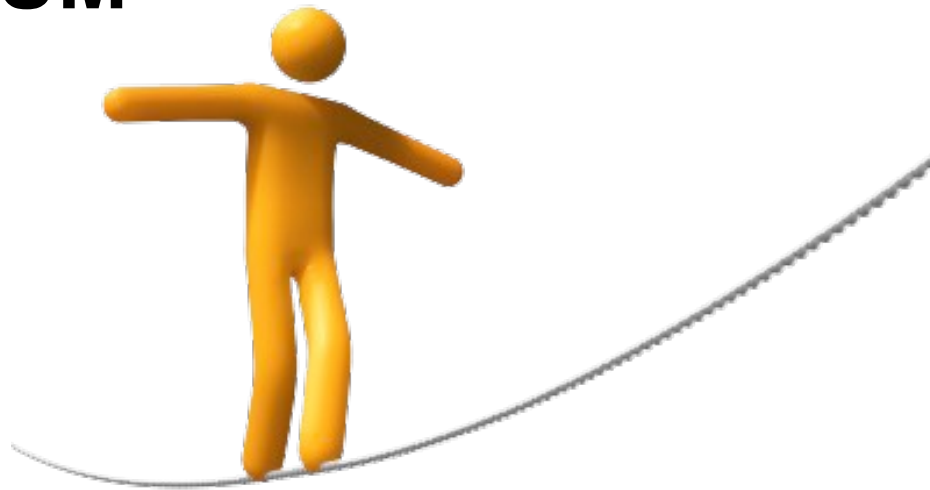
✓ Embryological Development

- Neural connections complete between 12-24th weeks.
- Myelination complete by 8-9th month of gestation.
- VOR will be adult functioning at 6 months
- It will continue to develop through the first 6 years of life



EQUILIBRIUM

Internal
Vestibular



External
Visual
Somatosensory

Central Nervous System (CNS)

Sensory motor integration and appropriate motor response

✓ Motor Milestones

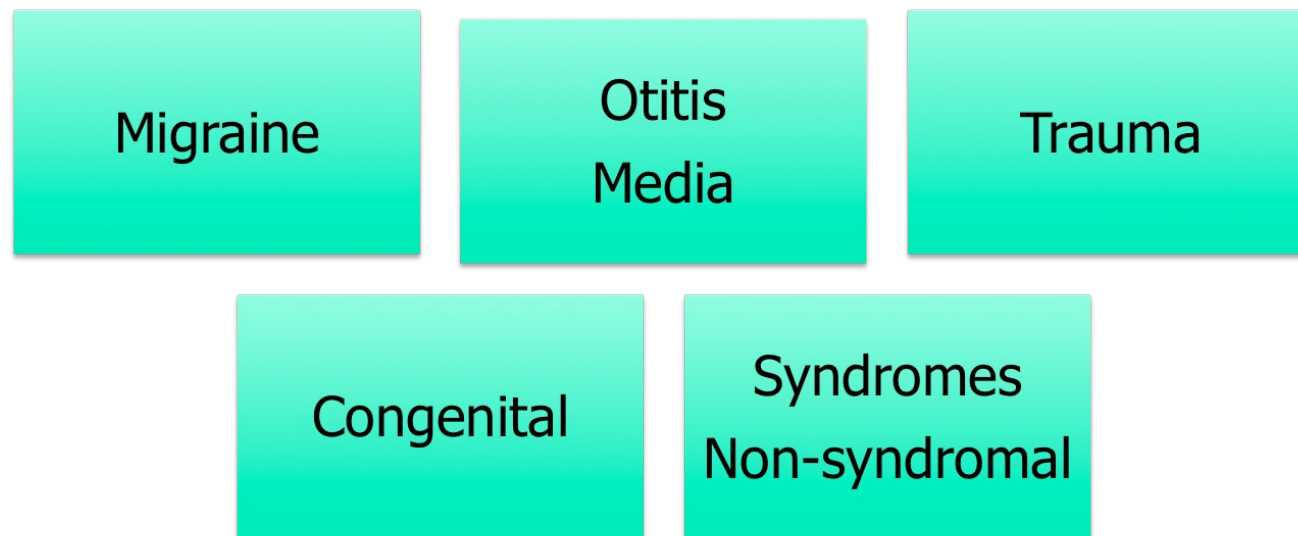
Fully developed at birth, maturation continues through early childhood.
This will allow further development of ...



- Balance
- Control
- Movement
- Coordination

Causes

Most common causes of vestibular dysfunction in children



Who are at Risk ?

- Infants and children identified with SNHL
- Identified with any syndromes or other acquired complicating medical factors e.g. meningitis, onset of imbalance with otitis media etc.
- Paroxysmal torticollis of childhood, periodic vomiting, with or without headache
- Infants and children with delayed motor milestones including children who may be considered “uncoordinated or “clumsy”
- Post head trauma w/wo Dx of concussion followed by dizziness or imbalance

Genetics

- Over 500 nDNA syndromes known to affect the audiovestibular (AV) system.
- 30-40% of children with SNHL also have vestibular symptoms. New study reported 90%+.
- AV disturbances the most commonly found defect across all mitochondrial diseases.
- Approximately 70 different nonsyndromal loci for hereditary AV impairment.
- Vestibular dysfunction without hearing loss may go undiagnosed.

✓ **Common Congenital and Acquired Conditions**

Syndromes-Descriptions

Usher	Type I - Congenital-bilateral profound SNHL, Retinitis Pigmentosa. Type II- Mild-severe progressive high frequency SNHL.	Waardenburg	Congenital SNHL, pigmentary disturbances of iris, hair, skin. Vestibular disturbances without hearing loss.
Branchiootorenal	Preauricular pits or tags, branchial cysts, hearing loss and/or abnormal development of the kidneys.	Von Hippel-Lindau	Hemangioblastomas of brain, spinal cord and retina. Renal cysts and renal cell carcinoma (40%). Dizziness/imbalance and hearing loss may be initial symptoms, may mimic Meniere's.
Pendred	Congenital, severe-profound SNHL, abnormality of bony labyrinth. Abnormal thyroid development with goiter in early puberty or adulthood.	CHARGE	Coloboma-heart-atresia-retarded-genital-ear. Vestibular symptoms prevalent.
Neurofibromatosis Type 2 (NF2)	Bilateral vestibular schwannomas, tinnitus, hearing loss and balance dysfunction. Schwannomas of other peripheral nerves, Meningiomas and juvenile cataract.	Marshall	Saddle nose, myopia, early-onset cataracts and short stature. Vestibular symptoms prevalent.
		Spinocerebellar Ataxia	Complex and progressive. 23 distinct genetic disorders. May also include hearing loss.

Earliest form of Migraine: BPV of Childhood

- Most frequent cause of pediatric dizziness (35%).
- Paroxysmal torticollis of childhood, periodic vomiting, and basilar migraine in the migraine family.
- Patients presents different clinical patterns of vestibular symptoms, with a common pathophysiology. Vasoconstriction of vestibular nuclear vessels followed by vasodilation.

BPV of Childhood

- Onset 1-4 years old.
- Migraine variant.
- Attacks are brief and may occur weekly-monthly.
- Symptoms may include: vomiting, dizziness - vertigo, imbalance – ataxia, nystagmus, torticollis.
- 30 – 50% BPV Childhood cases can evolve to classic migraines by puberty.
- May also evolve to Meniere's later in life.
- Usually will recover spontaneously.
- Torticollis is managed with physical therapy.

Vestibular Migraine: Puberty Onset



Aura: An abnormal sensory perception

- Vertigo: experienced as an aura in 25-30% of migraineurs
- Hearing Loss: 5% fluctuant or permanent
- Tinnitus
- Phonophobia: hypersensitivity to sound (misophonia)

Trauma: Concussion

Signs and Symptoms vary, and may include one or many of the following:

- Emotions out of proportion
- Slurred or incoherent speech
- Gross observable incoordination
- Disorientation (time, date, location)
- Any period of LOC
- Headaches and Irritability
- Sleep Disturbances
- Depression may develop
- Unequal pupil size
- Vacant Stare
- Tinnitus (ringing in the ears)
- Nausea & Vomiting
- Delayed verbal responses
- Delayed motor responses
- Confusion & inability to focus
- Memory deficits

Labyrinthine Concussion

- Soft tissue damage to the membranous labyrinth post-trauma
 - May be from amusement park ride, car accident, head trauma, etc. Does not require head impact
- Immediate vertigo post-trauma
 - Possible tinnitus and hearing loss in affected ear
 - Duration of symptoms may last 1 hour to several weeks
- Patient may need Vestibular Rehabilitation Therapy after vertigo subsides



Assessment: Behavioral

Pediatric Assessment: What and How to Test...?



3 systems to consider...

- Vestibular Ocular (VOR)
 - Vestibular Spinal (VSR)
 - Vestibular Collic (VCR)
-
- Age appropriate...maturational development.
 - Child-directed
 - Behavioral/Clinical and Electrophysiologic evaluation

Evaluation Recommendations

Child directed- age appropriate

- **Vestibulo Spinal Reflex (VSR)**
 - Norms and motor development
- **Vestibulo Ocular Reflex (VOR)**
 - Spontaneous nystagmus, conjugate eye movement, ocular-motility, rotation testing, optokinetic reflex
- **Vestibulo Collic Reflex (VCR)**
 - Vestibular Evoked Myogenic Potential (VEMP): 3+ months

✓ Behavioral Assessment

Maturational Motor Milestones

	3 months	7 months	9 months	12 months	24 months
	Raises head and chest when lying on stomach	Sits with and then without support of hands	Crawling on hands and knees	Sits without assistance	Walks alone by 18 months
	Starts to use eyes and hands in coordination	Supports weight on legs	Walking with assistance	Crawls forward on belly by pulling with arms and pushing with legs	Begins to run
	Begins to support head	Ability to track moving objects improves	Upper body- turns from sitting to crawling position	Creeps on hands and knees and supports trunk	Can push a wheeled toy
	Pushes down with legs when feet placed on floor	Rolls over		Pulls self up to standing position	
	Moves eyes in all directions	Supports head when sitting		Walks holding on to furniture	
				Stands momentarily without support	

Age-based Protocols by Month

	3	6	12	24	36
VOR	Eye movement OPK		vHIT		
VSR	Motor norms				Clinical Test Sensory Integration of Balance (CTSIB)
VCR	VEMP				

Righting Reflexes-Equilibrium Responses

- Neck Righting- birth- 6 months
- Body Righting - 6 months to 4-5 years
- Lifting child in space or changing position (Landau Reflex) 6 months to 2-3 years
- Age guidelines-maturational milestones
 - head righting
 - postural control
 - tonic neck reflexes-posture

What to look for

1. Loss of postural tone
2. Inability to produce normal muscle tension on ipsilateral side
3. Lesion within inner ear or vestibular nuclei- will produce less activity in the extensor muscles- greater strength on opposite side

✓ Behavioral Assessment

4 months



Physio-Ball destabilizes infant
forces neck and body righting reflexes

Righting Reflex - neonate/infant



✓ Behavioral Assessment



Physio-Ball - good activity for young children as well, as it encourages a “play” activity which destabilizes- forces neck and body righting reflexes

✓ Behavioral Assessment



Dynamic surfaces (CTSIB)
- good activity for young children as well, as it encourages a “play” activity- while looking at vestibular contribution to VSR and postural stability

30 month - BPV-Childhood

Recovering torticollis - note the right lateral head tilt

Physical evaluation includes:

1. CTSIB with extension cervico-vestibular
 - Limits of Stability (LOS)
2. Physio-ball- trunk stabilization
3. Physio-ball- righting reflex



14 month - BPVC

- Recovering right torticollis: note right lateral tilt
- Physical exam- watch her reflexively touch and push down with legs when in contact with ground



Landau Reflex

- Suspended in prone- supported under the chest
- Watch the righting reflex when head is positioned downward toward ground as she is “flown”, challenging reference to gravity



✓ Behavioral Assessment

- Watch the righting reflex when head is positioned downward toward ground
- When rolled forward she is able to hold her head upright



14 month – **CHARGE Syndrome**

- Note physical examination-
observation of motor milestones-
- Infant cannot hold herself up or sit
upright unassisted



14 month – **CHARGE Syndrome**

- Note physical examination
- Poor head and trunk control on Physio-ball



✓ Behavioral Assessment



Assessment: Electrophysiological

✓ **Electrophysiological
Assessment**

Summary of Electrophysiological Tests

VNG

4-5 years old- may not be compliant

Rotary Chair

Good for infants- facility may not own RC system-cost prohibitive

cVEMP

Testing as early as 3 months of age

VORTEQ

Good for young children and older

vHIT

May be used with infants as young as 10 months of age

All tests may be limited by child's compliance and size limitations e.g. size of video goggles etc.

Pediatric VEMP Studies

Table 17.1 Review of pediatric VEMP studies

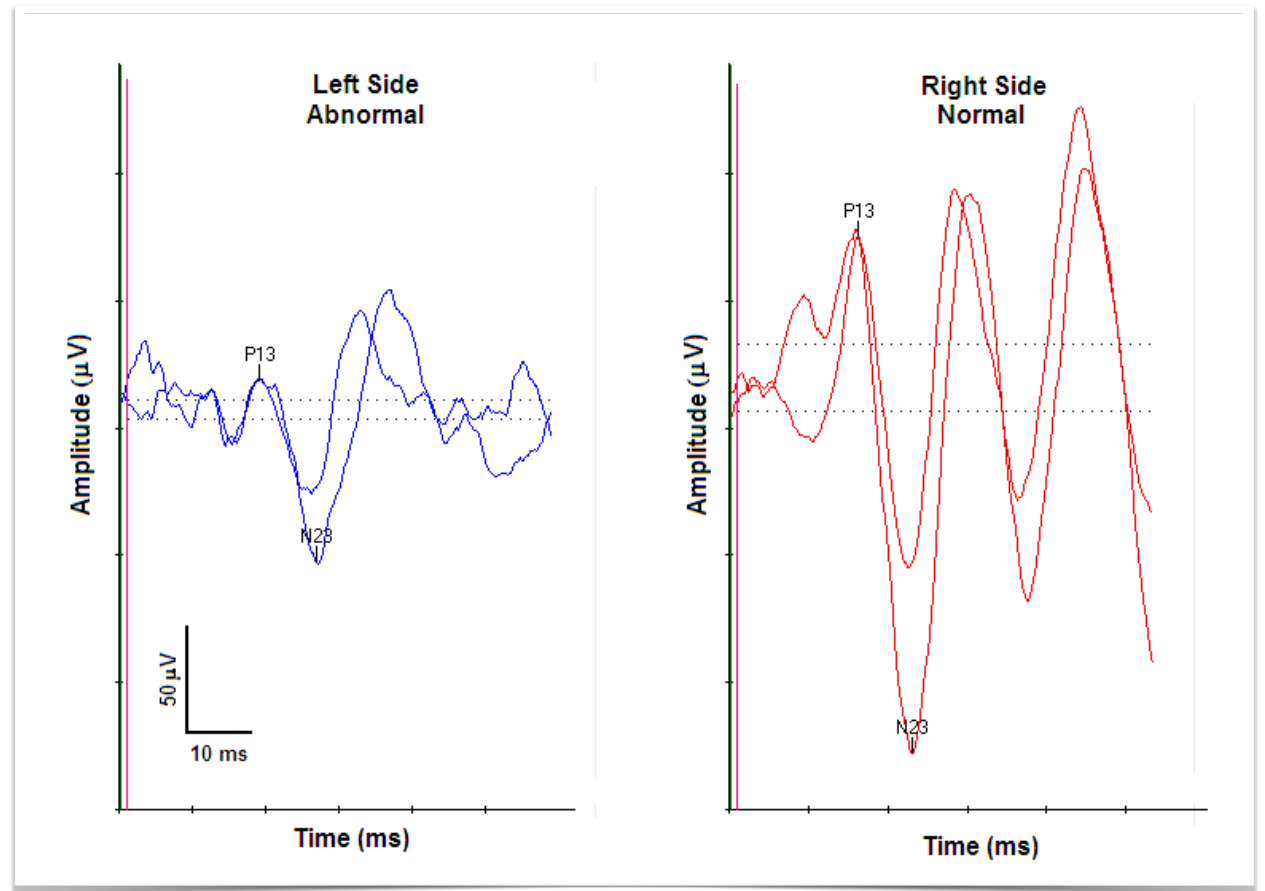
Investigators	Study
Pereira et al 2015 ¹⁶	Normative data on children ages 8–13 years
Maes et al 2014 ¹⁷	Compared cVEMP on normal and hearing impaired children ages 3 to 13 years
Ecevit et al 2012 ¹⁸	Compared cVEMP in late preterm and term births
Zhou et al 2009 ¹⁵	21/23 (91%) SNHL had abnormal amplitudes
Picciotti et al 2007 ¹³	Ages 3–15 years
Kelsch et al 2006 ¹²	Ages 3–11 years
Sheykholeslami et al 2005 ¹⁴	Neonates

Abbreviations: cVEMP, cervical vestibular evoked myogenic potentials; SNHL, sensorineural hearing loss.

VEMP Responses and Pathology

<i>Pathology</i>	<i>VEMP Responses</i>			
	<i>Absent</i>	<i>Reduced</i>	<i>Enhanced</i>	<i>Delayed</i>
<u>Otologic</u>				
Meniere's Disease	X	X	X	
Superior Canal Dehiscence Syndrome			X	
Neurolabyrinthitis	X	X		
Vestibular Neuritis	X	X		
<u>Neurologic</u>				
Migraine	X	X		X
Spinocerebellar Degeneration	X			X
Multiple Sclerosis	X			X
Brainstem Stroke	X			X

Vestibular Evoked Myogenic Potentials (VEMPs)



Abnormal left ear VEMP shows 50% reduction in amplitude.

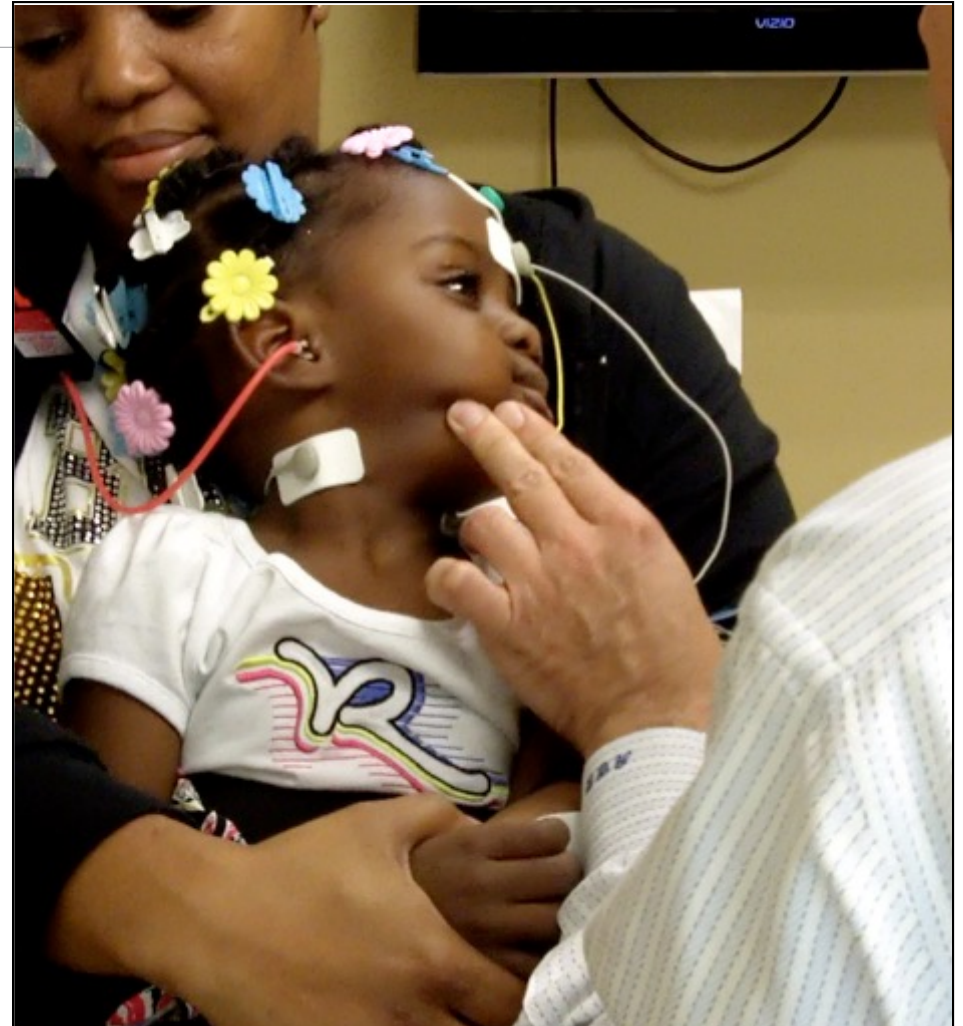
✓ Electrophysiological
Assessment

cVEMP neonate



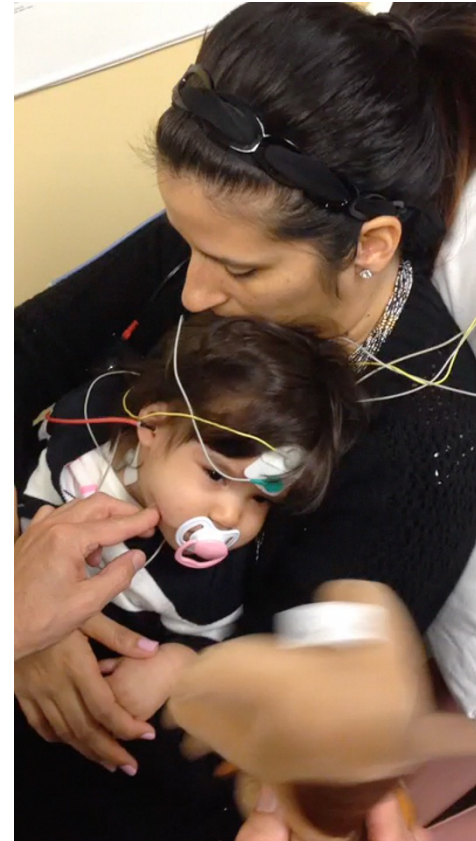
✓ Electrophysiological
Assessment

c-VEMP
BPV- Childhood- 4 yr. 6 month old
Right side torticollis- resolving



Cervical Vestibular Evoked Myogenic Potential

- (c-VEMP)
- Using some play techniques along with the mother's participation



Children: video Head Impulse Test (VHIT) has great test sensitivity

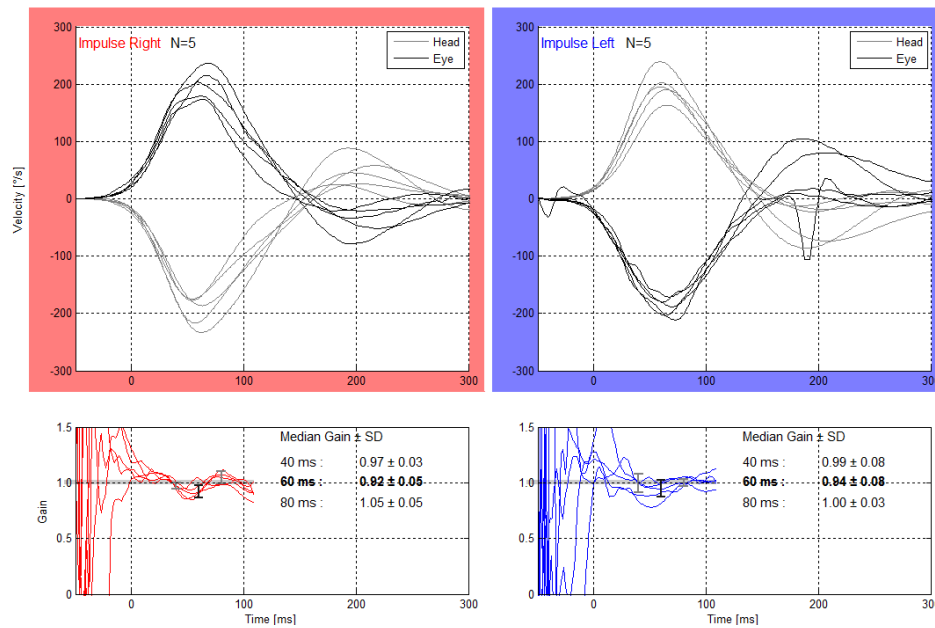
Because the human eye cannot see everything.

There are two types of 'catch up' saccade

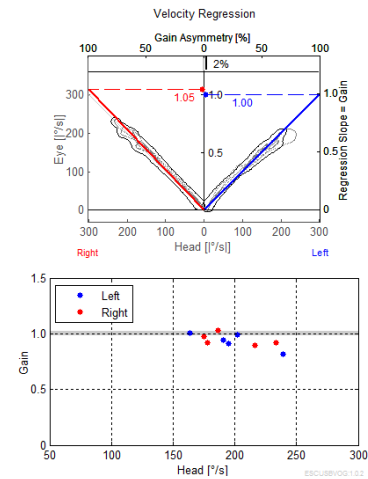
1. Saccades that happen **after the head had been moved**. We can see these eye movements, these are known as **overt saccades**
2. Saccades that happen **during the head movement**. These are known as **covert saccades**. Even the most experienced clinician cannot see these as they happen so quickly.



10 months old



Name:
Age:
Pers.ID:
Examination: Head Impulse
Condition: Lateral
Date: 2013-10-02 / 12:22:35.
Examiner:

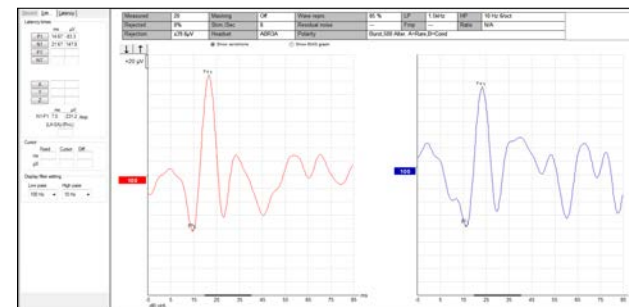
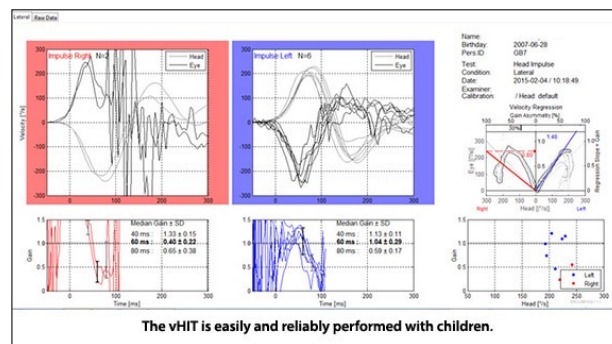
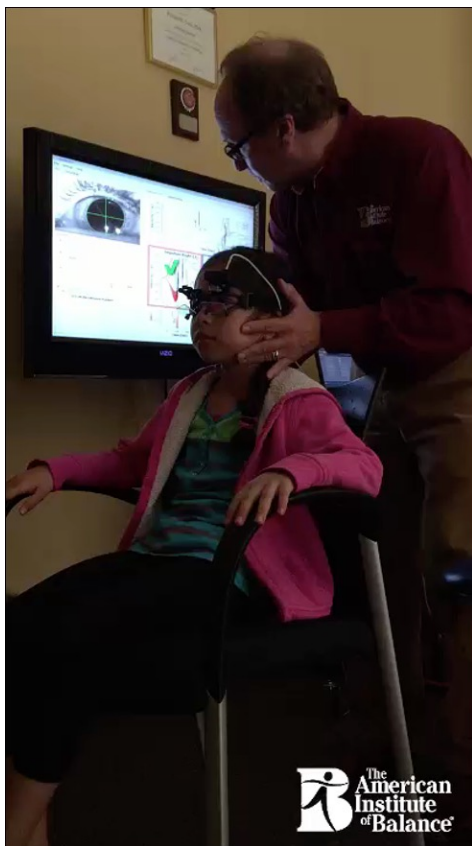


✓ Electrophysiological
Assessment

Vestibular Migraine

vHIT

cVEMP



✓ Case Studies

Case Studies



✓ Case Studies

HC-BPPV

- A 16-year-old male with a severe form of Spina bifida (S.B.) (lower body paralysis, AV shunt, and thoracic cavity) was referred to AIB by a pediatric otolaryngologist for evaluation of positional vertigo.



Osteogenesis Imperfecta

- 14-year-old female with Osteogenesis Imperfecta (OI), who is pre-pubescent, has a radiographically confirmed basilar invagination for many years. In addition, she has experienced over 50 fractures related to her OI condition.
- According to NIH, OI is typically a group of genetic disorders (autosomal dominant) associated with imperfect bone formation. Recently over the past 3-4 weeks she has been troubled by headaches and the past week with an acute onset positional vertigo

✓ Case Studies

HC-BPPV



Paraplegic w/limited upper body mobility



13-year-old –3 years post MVA.
Onset of acute positional vertigo
5 months



✓ Case Studies

Special needs populations, children or adults requires patient-centered management strategies



Summary: Pediatric PEARLS

- Children have audio - vestibular systems. Testing may begin as young as 3 months of age.
- Normal or abnormal hearing thresholds (unilateral or bilateral sensorineural hearing loss (SNHL)) cannot be used in isolation to indicate status of vestibular function.
- Behavioral screening protocols may provide valuable findings which can be validated by clinical or electrophysiological vestibular function tests.
- Undiagnosed and untreated vestibular deficits in children pose potential quality of life and safety issues.

Thank You!

Richard E. Gans, PhD, CCC-A, DFAA





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