MAYO CLINIC

Progression of Balance Disorders

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Sled dog racing interest growing in area

Story and photos
by Steve Krikava

Sled dog racing came out of the Alaskan gold rush days, explains Daryl McCaslin, a modern sled dog racing enthusiast from Amasa, Mich.

He says that during the gold rush, dog sleds were the only practical means of shipping freight; so freighting companies sponsored races in order to find the fastest dogs. The Alaskan Husky isn't an American Kennel Club (AKC) registered breed, Daryl McCaslin emphasizes. AKC registered sled dogs are the Alaskan Malamute, the Samoyed and the Siberian Husky, he says.

Those dogs are bred "primarily for conformation and show purposes . . . whereas the Alaskan Husky is bred for speed and endurance," he says.

Daryl McCaslin and his wife Laurie are natives of lower Michigan who have been raising sled dogs for 11 years. They moved from Tennessee to Amasa six years ago. "We probably had the only sled dog team in the state of Tennessee," Daryl McCaslin says.

The McCaslins currently have 15 adult dogs and nine pups. Daryl McCaslin says he'd like to keep all the pups since they have a good blood line, but in order to keep his kennel from getting too large, he'd have to sell some of his older, ex-

is 6 months old, he says. By the time the dog is a year and a half, the owner should be able to tell if he has a good racer.

Training also involves obedience to verbal commands, Daryl McCaslin says. "They're very strictly disciplined, a sled team is. When a dog is in harness, it stands there and waits for orders."

All dogs in the team should know "stay," "whoa" or "stop," and "hike," "go" or some other command for start, he says.

A lead dog is trained further to "set a good pace," to stay out in front of the team so the rest will follow him, Daryl McCaslin explains. "It makes your team a lot more manageable if you've got one (lead dog) who can take commands (for turning)," he says.

"Rowdy's what you call a perfect command," says Laurie McCaslin. "That means you've got perfect control from way back," Daryl McCaslin explains.

Races, especially sprints, can be won by 1/100 of a second, he continues, and racers don't want to lose time trying to get the team around a corner.

Sprint races generally are two-day events involving relatively short races for four divisions of teams: D — three dogs, C — three to five dogs, B — five to seven dogs. A — open, seven and up. "Usually you never race more than 16 dogs," Daryl McCaslin says. "More than that gets kind of unruly, hard to handle."

"The Midwest is starting to have a real good (sprint racing) circuit," he con-

Sled dogs . . .

(continued from page 1)

However, there was no electric service to their new property and the electric company said it would cost $10,000 to run a line to their home, so the McCaslins learned to live without it.

They have gas heat, gas lights and a gas refrigerator. A gas powered generator provides enough electricty for pumping water.

Laurie McCaslin sews with an antique, pedal-powdered Singer sewing machine and plays an 1897 reed organ bought from a church in Norway, Mich.

The McCaslins gave their color television console to a friend who had helped them get settled in Amasa, and the McCaslin children now read a lot and listen to wind-up phonographs and other antique music machines.

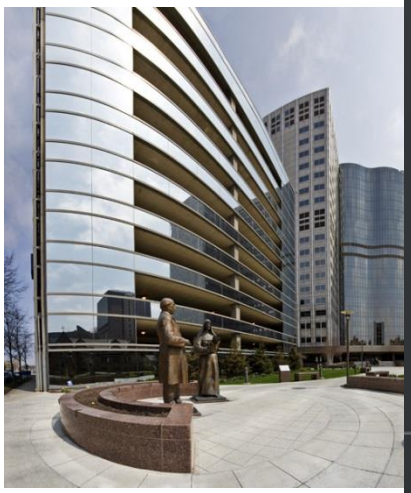
Now the power company has said they can bring in power for a reasonable cost, Daryl McCaslin says, but he and his family are so used to living without it that they've decided not to have electricity brought in yet.

Daryl McCaslin says he's quite happy now, living in a rural setting, getting involved with local politics, working with his dogs in the wilderness. He says there's so much publicly owned land in the Upper Peninsula that "I can sled 20 or 30 miles without ever crossing private land."

Disclaimer

- Grant support
  - U.S. National Institutes of Health (Grant # 2R44DC014611-02)
  - Mayo Clinic Internal Funding

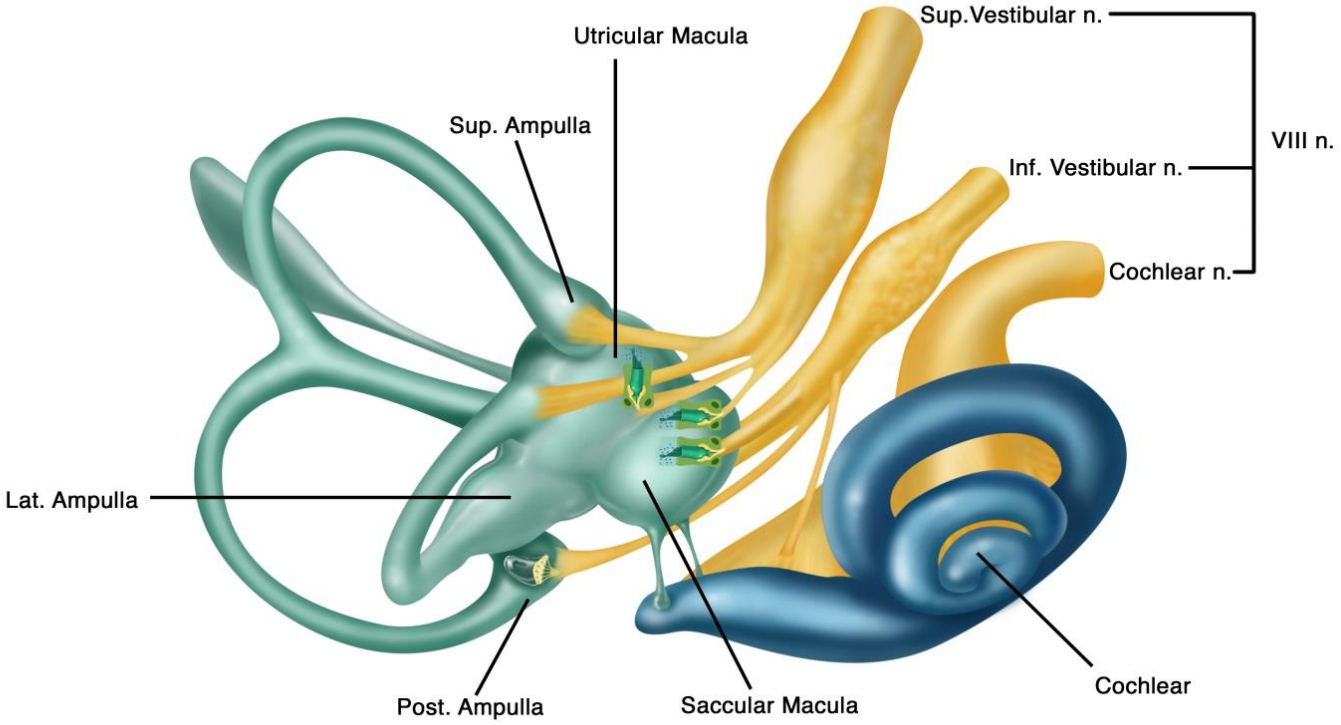
Mayo Cl
Locations



Outline

- Background
- Case
- Epidemiology
- Critical Period for the Vestibular System
- Migraine
- Cytomegalovirus
- Genetics
- Enlarged Vestibular Aqueduct
- Summary

Peripheral Vestibular System



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Background-History

- A 9-year-old girl presents with report of longstanding dizziness. She has unilateral hearing loss and a history of multiple head injuries.
- According to the family, from age 7, the patient began to experience episodes of nausea and dizziness lasting for minutes to hours at a time.
- The case history revealed multiple falls with significant injuries warranting a visit to the emergency room, including:

Background-History

- Age 2: the patient fell off a couch at home and had a bruise on her neck.

- Age 3: the patient fell and hit her head (occiput) on a rock with no imaging follow-up at the time.

- Age 4: the patient fell off a swing onto the back of her head and had bleeding from her injury. There was no loss of consciousness.

Background-History

● Age 7: the patient ran into a parked car and fell to the ground, hitting her head and yielding abrasions on her forehead and
around her eye.

Outside Otolaryngology Report
(5/9/2011)

History Of Present Illness

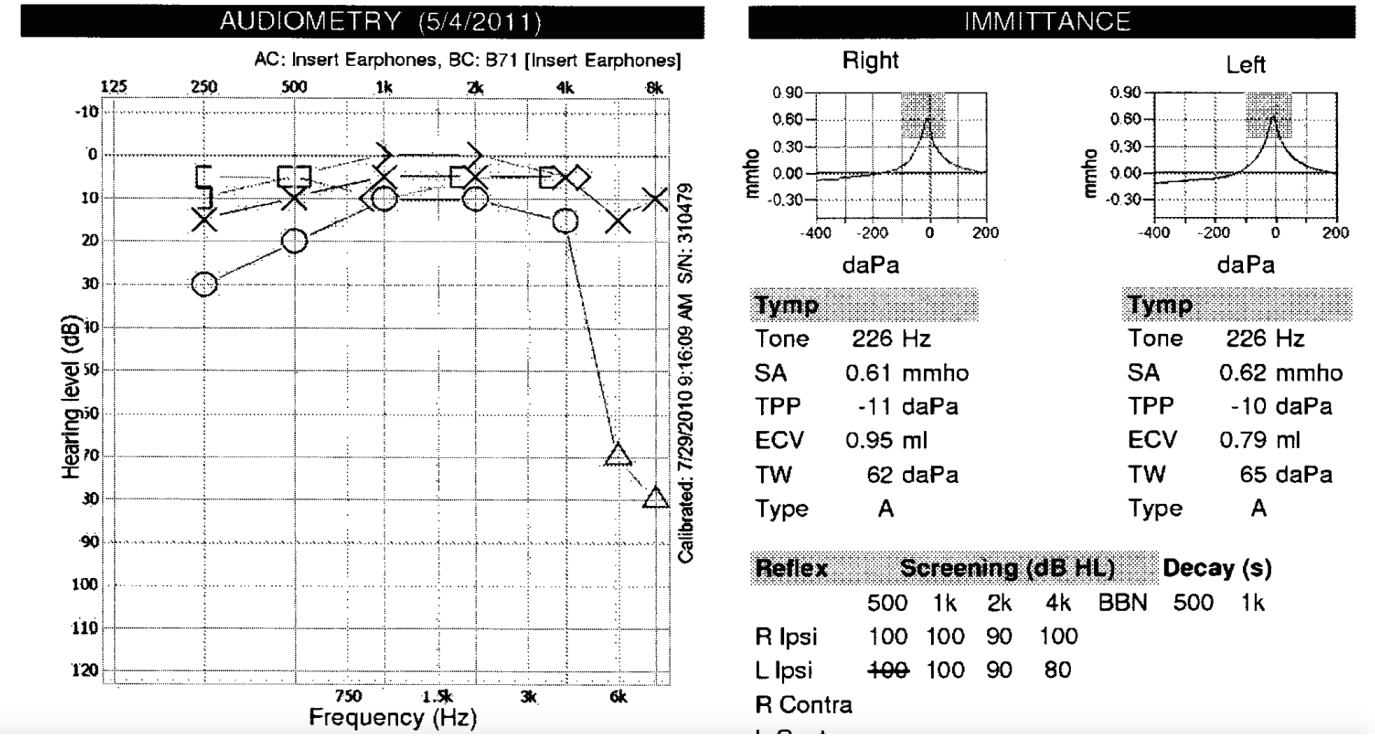
The patient is a 9 year old White female , who is brought to the office by her parents and who presents for evaluation of The episodes last hours at a time. The onset is described as gradual. vertigo. The motion sensation is described as movement in multiple directions. The vertigo began 5 years ago and has since been intermittent , occurring frequently with episodes lasting almost daily. The vertigo has not been associated with any precipitating events. The patient's past medical history is noncontributory.

The symptom onset is not thought to be associated with a particular event. The symptoms are not worsened by any known factor. No alleviating factors have been identified.

The patient states that she has also experienced nausea, vomiting, and headache. The patient reports moderately severe temporal throbbing headaches. Complaints denied: ear pain, decreased hearing, fluctuating hearing loss, tinnitus, ear drainage, ear fullness, and ear pressure.

A few ear infections as a infant. No family hx of inner ear. Possitive fm hx of hearing loss. Will get vertigo without a headache. Mom has migraines. Triaged by

Outside Audiogram



Audiogram showing a mild low frequency conductive hearing loss with severe high frequency hearing loss at 6K-8K on the right

Initial CT examination

PROCEDURE: CT TEMPORAL BONE WITHOUT CONTRAST

TECHNIQUE: Computerized tomography of the temporal bones with coronal and axial sections was performed. CPT 70480

HISTORY: peripheral vertigo, right sensorineural hearing loss

COMPARISONS: None .

FINDINGS:

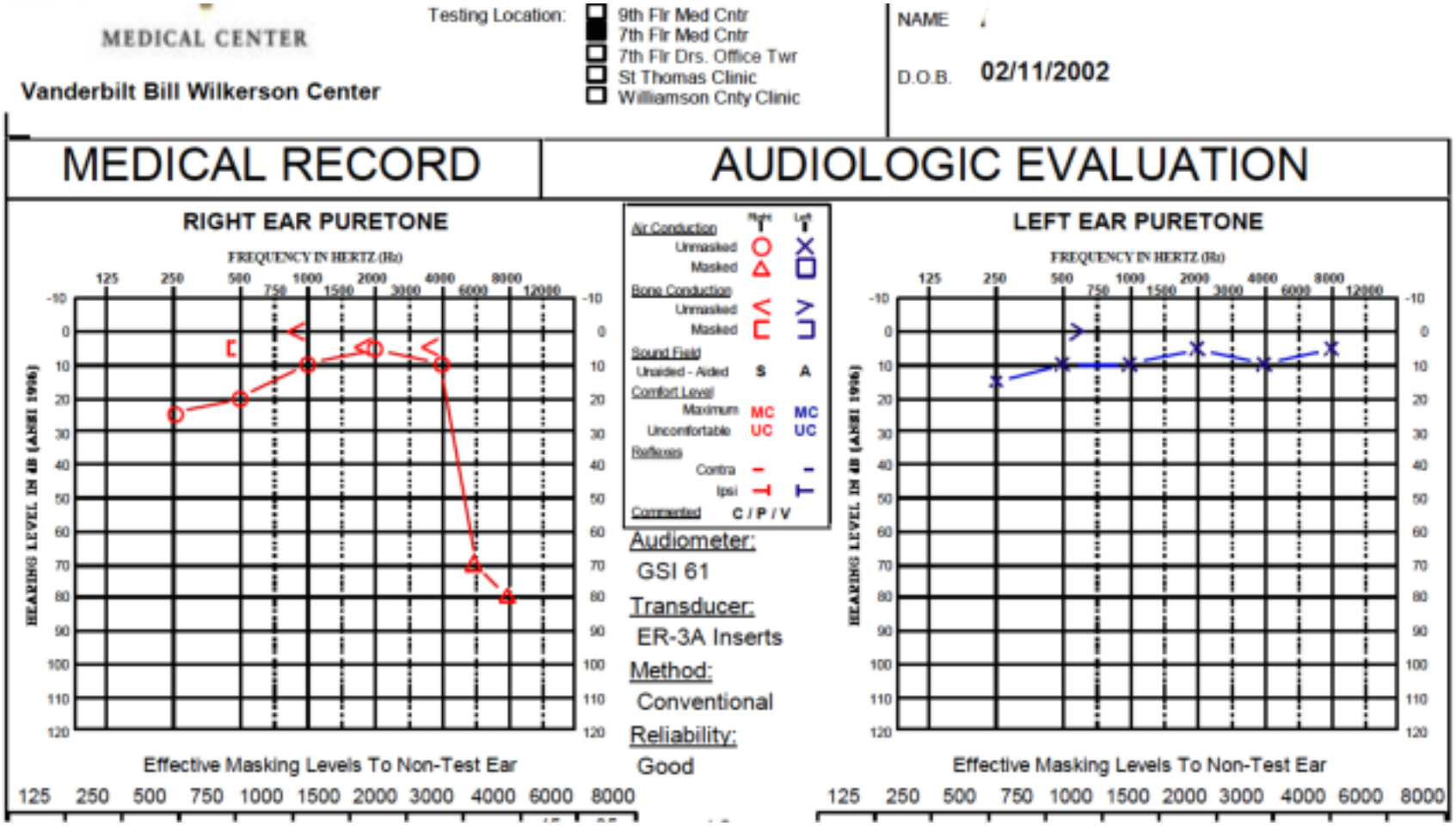
RIGHT temporal bone: External auditory canal appears normal. Tympanic membrane appears normal without evidence for thickening or retraction. Scutum is normal. The ossicles appear normal in morphology. No abnormal soft tissue is appreciated within the middle ear cavity. Cochlea, vestibule, and semicircular canals appear normal. Facial nerve is well delineated within the temporal bone and appears normal. The internal auditory canal appears normal. Mastoid air cells appear well-aerated. Jugular bulb and carotid canal appear normal.

LEFT temporal bone: External auditory canal appears normal. Tympanic membrane appears normal without evidence for thickening or retraction. Scutum is normal. The ossicles appear normal in morphology. No abnormal soft tissue is appreciated within the middle ear cavity. Cochlea, vestibule, and semicircular canals appear normal. Facial nerve is well delineated within the temporal bone and appears normal. The internal auditory canal appears normal. Mastoid air cells appear well-aerated. Jugular bulb and carotid canal appear normal.

Intracranial contents appear within normal limits. Globes and orbits are normal. The paranasal sinuses are clear.

IMPRESSION:
1. Normal appearance to the bilateral temporal bones. Specifically, no morphologic abnormalities to account for the patient's vertigo and sensorineural hearing loss.
\.br.\

10/3/11 (First internal Audiogram)



Neurotology Visit #1

- Dix-Hallpike maneuver demonstrated upward beating nystagmus in the right head-hanging position. An Epley maneuver was performed.
- Radiology: CT Temporal bone from May 2011 showed no underlying anatomical abnormality.
- Assessment: right-sided unilateral hearing loss, right-sided vestibular weakness and BPPV. She has had some improvement with Epley maneuvers in the past. CT Temporal bone is unremarkable.

Pediatric Neurology Report

- 9 year old female with longstanding symptoms of vertigo that may or may not be related to the fall at 3 years of age.
  - “Further review of the outside head CT done demonstrated a previous right occipital skull fracture”
  - “Continue repositioning exercises that have helped w/ the symptoms and frequency”.
  - “The child should avoid activities that induce the symptoms of vertigo and extreme heights as she could fall if Sx occur”.

2nd Neurotology appointment

- Visit scheduled because of frequent vertigo
  - Daily dizzy attacks is the main complaint. MRI ordered.
  - Patient complains that she feels like she is spinning for 7-8 hours. She also gets a headache at those times with light sensitivity and sound sensitivity.
  - “Patient is having trouble in school with them notifying her mom”.
  - “I have suggested that she go on a migraine diet”.

MRI w/o Contrast (temporal bone)

- Ordered for comparison of CT scan done on may 9, 2011
- Findings:
  - There is enlargement of the right endolymphatic sac. No appreciable cochlear dysplasia.
  - Left endolymphatic sac, internal auditory canals and their contents, remaining membranous labyrinths, and cerebellopontine angle structures are unremarkable.

MRI

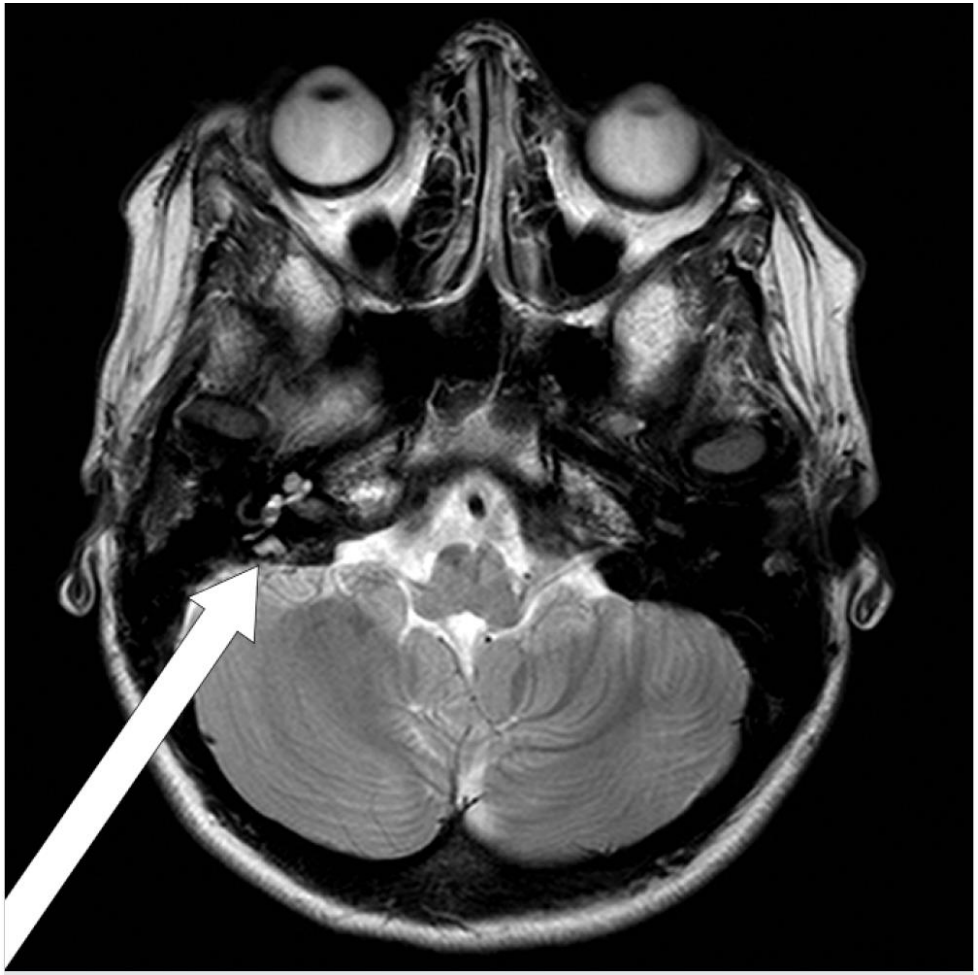
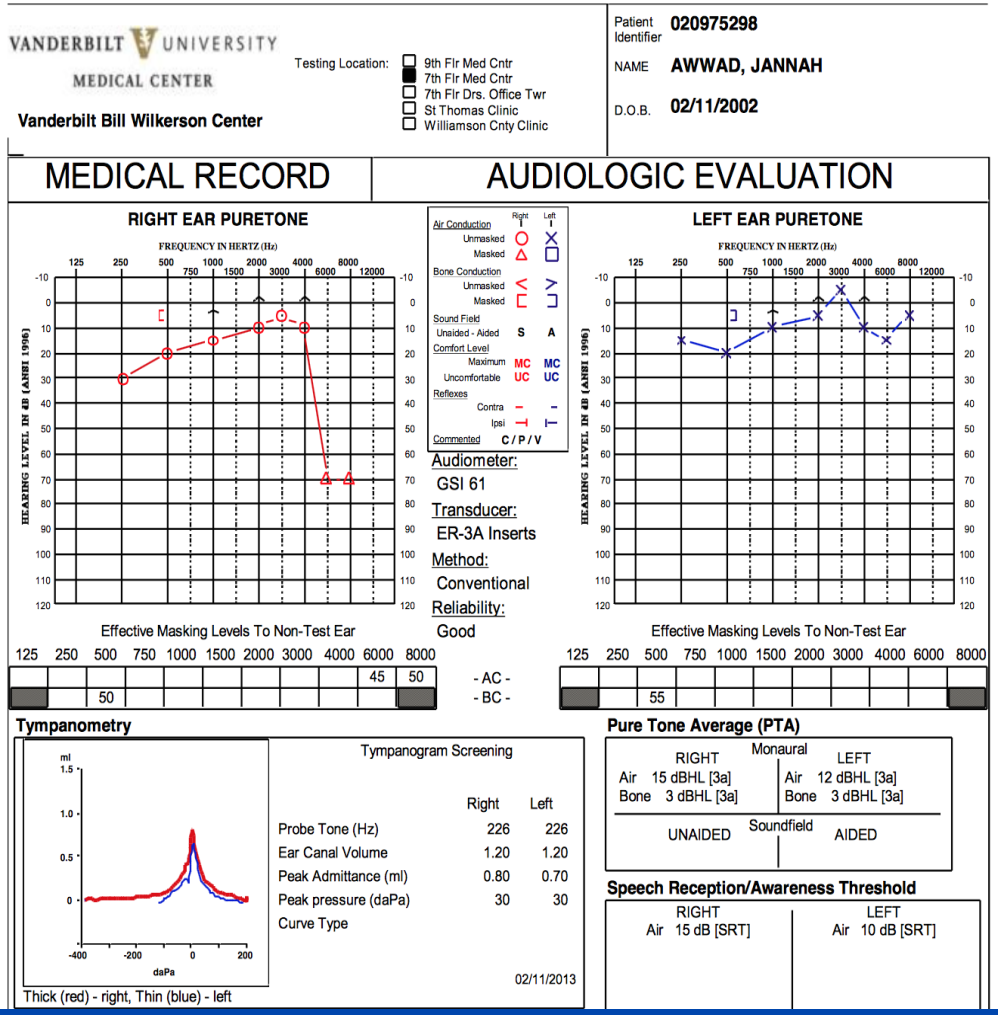


Fig. 17.3 Computed tomographic scan. An axial cut viewed from below the patient. The white arrow indicates the location of the vestibular aqueduct.

Audiogram
2/11/13



VANDERBILT UNIVERSITY MEDICAL CENTER

Vanderbilt Bill Wilkerson Center

Testing Location: ☐ 9th Flr Med Cntr ■ 7th Flr Med Cntr ☐ 7th Flr Drs. Office Twr ☐ St Thomas Clinic ☐ Williamson Cnty Clinic

Patient Identifier 020975298

NAME AWWAD, JANNAH

D.O.B. 02/11/2002

MEDICAL RECORD | AUDIOLOGIC EVALUATION

RIGHT EAR PURETONE

FREQUENCY IN HERTZ (Hz)

125 250 500 750 1000 1500 2000 3000 4000 6000 8000 12000

HEARING LEVEL IN dB (ANSI 1996)

-10 0 10 20 30 40 50 60 70 80 90 100 110 120

Air Conduction Unmasked Masked

Bone Conduction Unmasked Masked

Sound Field Unaided - Aided S A

Comfort Level Maximum MC MC Uncomfortable UC UC

Reflexes Contra – – Ipsi ⊣ ⊢

Commented C / P / V

Audiometer: GSI 61

Transducer: ER-3A Inserts

Method: Conventional

Reliability: Good

LEFT EAR PURETONE

FREQUENCY IN HERTZ (Hz)

125 250 500 750 1000 1500 2000 3000 4000 6000 8000 12000

HEARING LEVEL IN dB (ANSI 1996)

-10 0 10 20 30 40 50 60 70 80 90 100 110 120

Effective Masking Levels To Non-Test Ear

125 250 500 750 1000 1500 2000 3000 4000 6000 8000

45 50 - AC -

50 - BC -

Effective Masking Levels To Non-Test Ear

125 250 500 750 1000 1500 2000 3000 4000 6000 8000

55

Tympanometry

ml 1.5 1.0 0.5 0

-400 -200 0 200 daPa

Thick (red) - right, Thin (blue) - left

Tympanogram Screening

	Right	Left
Probe Tone (Hz)	226	226
Ear Canal Volume	1.20	1.20
Peak Admittance (ml)	0.80	0.70
Peak pressure (daPa)	30	30
Curve Type		

02/11/2013

Pure Tone Average (PTA)

Monaural

RIGHT: Air 15 dBHL [3a], Bone 3 dBHL [3a]

LEFT: Air 12 dBHL [3a], Bone 3 dBHL [3a]

Soundfield

UNAIDED

AIDED

Speech Reception/Awareness Threshold

RIGHT: Air 15 dB [SRT]

LEFT: Air 10 dB [SRT]

Neurotology Visit #3
(1/1/2013)

- The patient has recently had an MRI which does not show any evidence of retrocochlear pathology.
- The patient does have an enlarged vestibular aqueduct on the right.
- The patient has seen remarkable improvements in her overall dizziness and vertigo.
- Recommend continuing with migraine diet

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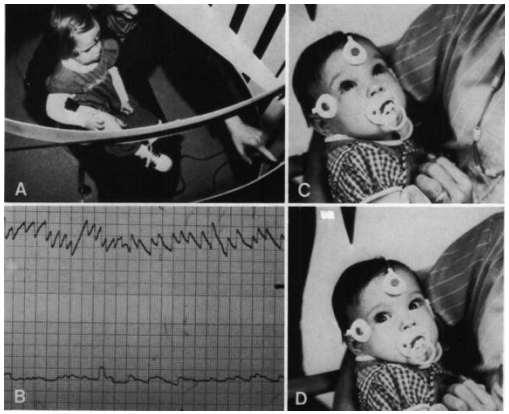
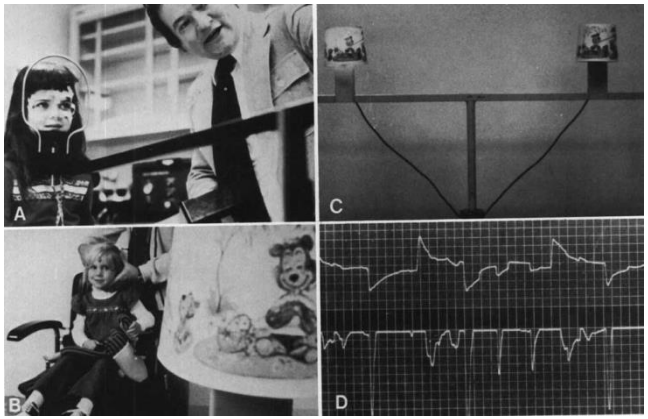
VESTIBULAR TESTING IN CHILDREN

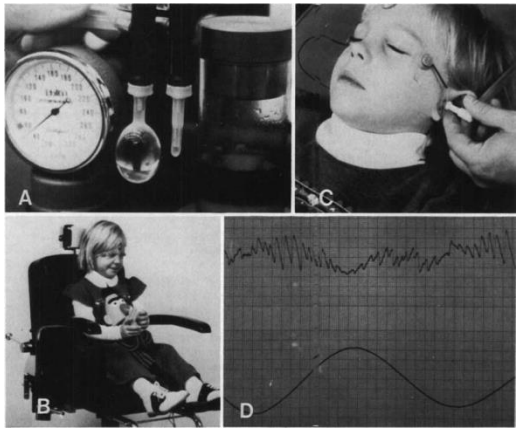
DAVID G. CYR, MA

OMAHA, NEBRASKA

Vestibular evaluation in the pediatric population has in the past taken several forms. For the most part, the pediatric vestibular evaluation has been more subjective than objective. One of the primary reasons for this has been the obvious difficulties encountered in trying to conduct a standard, adult electronystagmography (ENG) procedure on a pediatric patient population not capable of performing in a manner conducive to a good ENG recording. The purpose of this paper is to suggest certain modifications of the standard adult ENG battery for use with young children and infants. Discussion consists of modifications in the areas of various ocular movement tests including sinusoidal pursuit, calibration, optokinetics and gaze testing. In addition, procedures relative to peripheral vestibular output from perrotational and caloric stimulation are discussed. Topics also include the use of a closed-loop caloric irrigator and simultaneous caloric irrigation as viable alternatives to standard, alternate water irrigation when testing the vestibular output of a young child or infant.







Epidemi

- Until re … een conduc … e underta … d an otolaryn …
- How do … vestibular impairm …

Vestibular Infant Screening-Flanders (VIS-Flanders)

The start of an exciting project

Maes, L.[1,2]; Martens, S.[1]; Dhondt, C.[3]; Sucaet, M.[1]; Vanaudenaerde, S.[2]; Rombaut, L.[2]; Dhooge, I.[2,3]

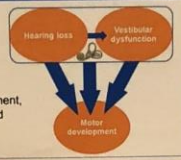
[1]Ghent University, Faculty of Medicine, Department of Speech, Language and Hearing Sciences
[2]Ghent University Hospital, Department of Otorhinolaryngology
[3]Ghent University, Faculty of Medicine, Department of Ear Nose Throat

Background

1. Importance

➢ Children with a hearing loss have a higher risk of vestibular problems, due to the close anatomical relationship between the auditory and vestibular organs.

➢ Given that the vestibular function is important for the (motor) development, the early detection of vestibular problems is essential in hearing-impaired children.



2. State-of-the-art

➢ An extensive vestibular and motor test protocol in hearing-impaired children is standard of care in the Ghent University Hospital since 2016.

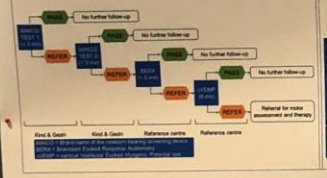
➢ This is currently not a standard component in the diagnostic follow-up in other care centers in Flanders because these tests are very challenging in the paediatric population. Consequently, a lot of vestibular problems go unnoticed!

Methods: Clinical implementation

1. Vestibular Infant Screening – Flanders

➢ **Subjects**: All Flemish infants with a permanent congenital hearing loss at the age of 6 months. For this purpose, all diagnostic reference centers of Child and Family (Kind en Gezin) in Flanders will participate.

➢ **Protocol**: If a hearing loss is confirmed by the existing auditory screening programme (MAICO and BERA test), the vestibular screening (cVEMP) will be performed.



2. Extensive follow-up protocol

➢ **Subjects**: All children with a confirmed hearing loss at the Ghent University Hospital, will be subjected to a more extensive test protocol, which will be repeated up to the age of 3 years.

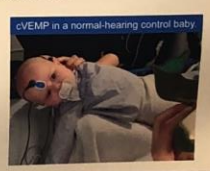
➢ **Protocol**: This age-dependent protocol consists of a vestibular test battery, motor assessment and 2 questionnaires.

	Vestibular assessment	Ocular motor and nystagmus assessment	Motor assessment	Questionnaires
6mo	vHIT	Subjectively (without ENG registration)	AIMS	Bayley II-questionnaire
1yr	Rotatory test		PDMS-2	
2yr	cVEMP	Objectively (ENG registration)	PDMS-2	Vestibular screening-questionnaire
3yr	+ Caloric test + oVEMP		GDBT	

3. Determination of normative data

➢ **Subjects**: Typically developing normal-hearing children between 6 months and 3 years of age will be examined once.

➢ **Protocol**: The extensive vestibular (including the cVEMP screening) and motor test protocol will be used to obtain normative data.



Objectives

1. Vestibular Infant Screening – Flanders

The implementation of a standard vestibular screening will lead to:

➢ Early identification of vestibular deficits.
➢ Early referral for motor assessment and therapy.
➢ More insight into the incidence of vestibular deficits in congenitally hearing-impaired children in Flanders.
➢ An increasing awareness for vestibular disorders and associated symptoms in children.

2. Extensive follow-up protocol

Linking the results of the screening to those of an extensive test protocol will give insight in:

➢ The sensitivity of the cVEMP screening in detecting vestibular deficits.
➢ A further fine tuning of the screening protocol.
➢ The development of uniform guidelines for further referral.

3. Determination of normative data

Normative data will be collected to enable a correct interpretation of:

➢ The results of the extensive follow-up protocol at the Ghent University Hospital.
➢ The results of the vestibular screening in all diagnostic reference centers in Flanders.

Conclusion

➢ This project investigates the vestibular function in Flemish hearing-impaired children.

➢ The cervical Vestibular Evoked Myogenic Potential test (cVEMP) will be used as a vestibular screening tool at the age of six months.

➢ This vestibular screening ensures an early detection of possible vestibular problems and makes a timely referral for further motor investigation and appropriate therapy possible.

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Vestibular Infant Screening - Flanders

- Migraine
- BPVC
- Otitis Media
- Viral Infection
- Trauma

The Predominant Forms of Vertigo in Children and Their Associated Findings on Balance Function Testing

Devin L. McCaslin, PhD*, Gary P. Jacobson, PhD,
Jill M. Gruenwald, AuD

KEYWORDS

• Benign paroxysmal vertigo of childhood
• Migraine-associated vertigo • Electronystagmography
• Trauma • Vestibular neuritis • Otitis media
• Auditory neuropathy • Vestibular

CHAPTER 5

An Update on the Predominant Forms of Vertigo in Children and Their Associated Findings on Balance Function Testing

DEVIN L. MCCASLIN, PHD • JAMIE M. BOGLE, PHD • GARY P. JACOBSON, PHD

Vestibular Deficits in Children with Hearing Impairment

- Migraine
  - root cause has been suggested to be asymmetrical activation of brainstem vestibular nuclei or defective Ca2+ channels
  - Distinct criteria
- Otitis Media
  - Have been shown to have delayed development of motor skills (Cohen et al, 1997)
  - Treatment (PE tubes) have been shown to improve balance impairments

Vestibular Deficits in Children with Hearing Impairment

- GJB2 (Connexin 26)
  - Mutations result in DFNB1 and DFNA3
- Cytomegalovirus
  - Have a host of co-morbities and vestibular impairment can be one of them
  - Often associated with hearing loss
- Mondini Malformation
  - Inner ear malformation
  - Can be unilateral or bilateral

Vestibular Deficits in Children with Hearing Impairment (Con't)

- Waardenburg Syndrome
- Ototoxcity
- Pendred Syndrome

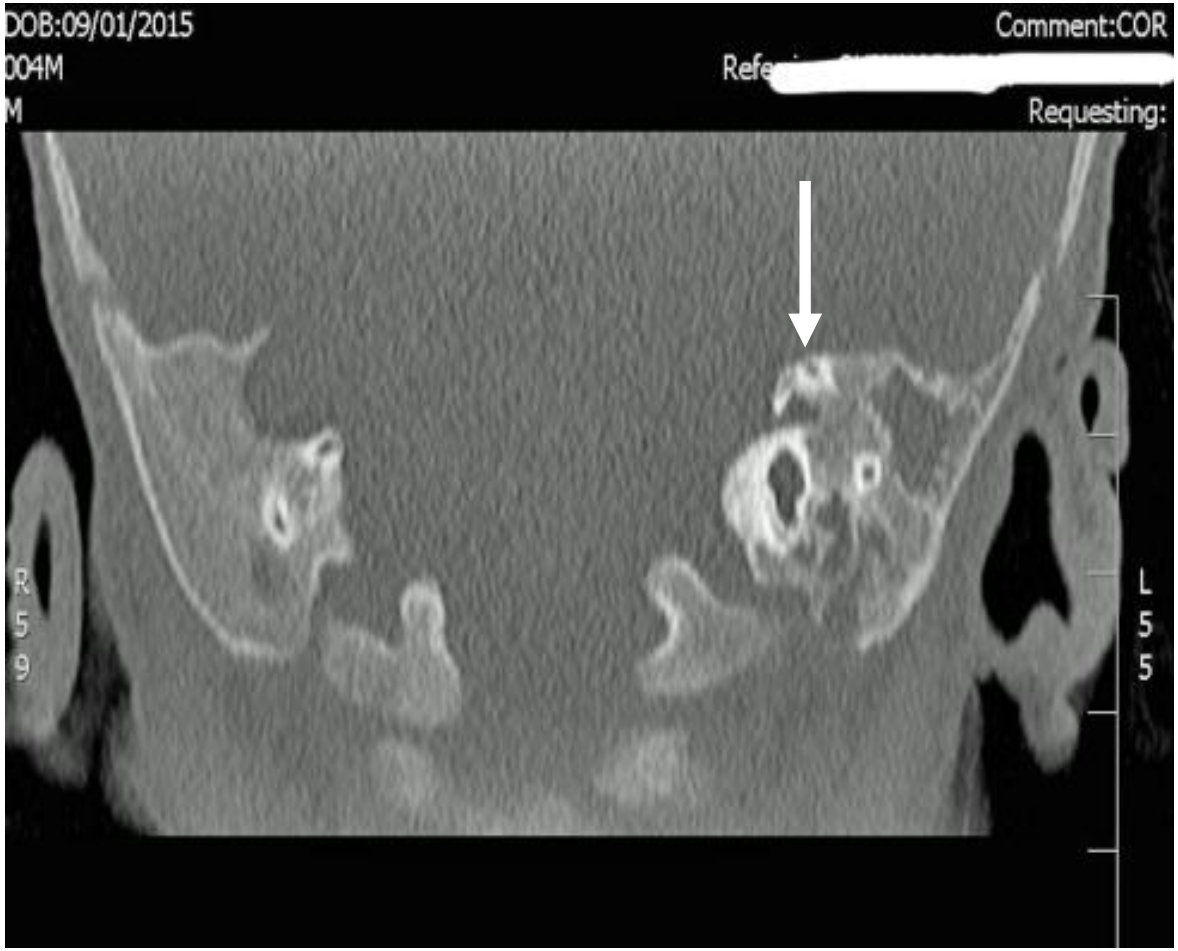
Vestibular Deficits in Children with Hearing Impairment (con't)

- Usher Syndrome (Chris Zalewski NIH)
  - Affects hearing and vision
  - Three types
    - Type I – profound hearing loss, vestibular impairments and vision loss that is progressive
    - Type II – typically mild sloping to profound sensorineural hearing loss. Often starts in teenage years
    - Type III – Progressive sensory loss

Vestibular Deficits in Children with Hearing Impairment (con't)

- Enlarged vestibular aqueduct
- Superior canal dehiscence
- Vestibular Neuritis
  - Inflammation of one or both of the branches of the vestibular nerve (superior or inferior)

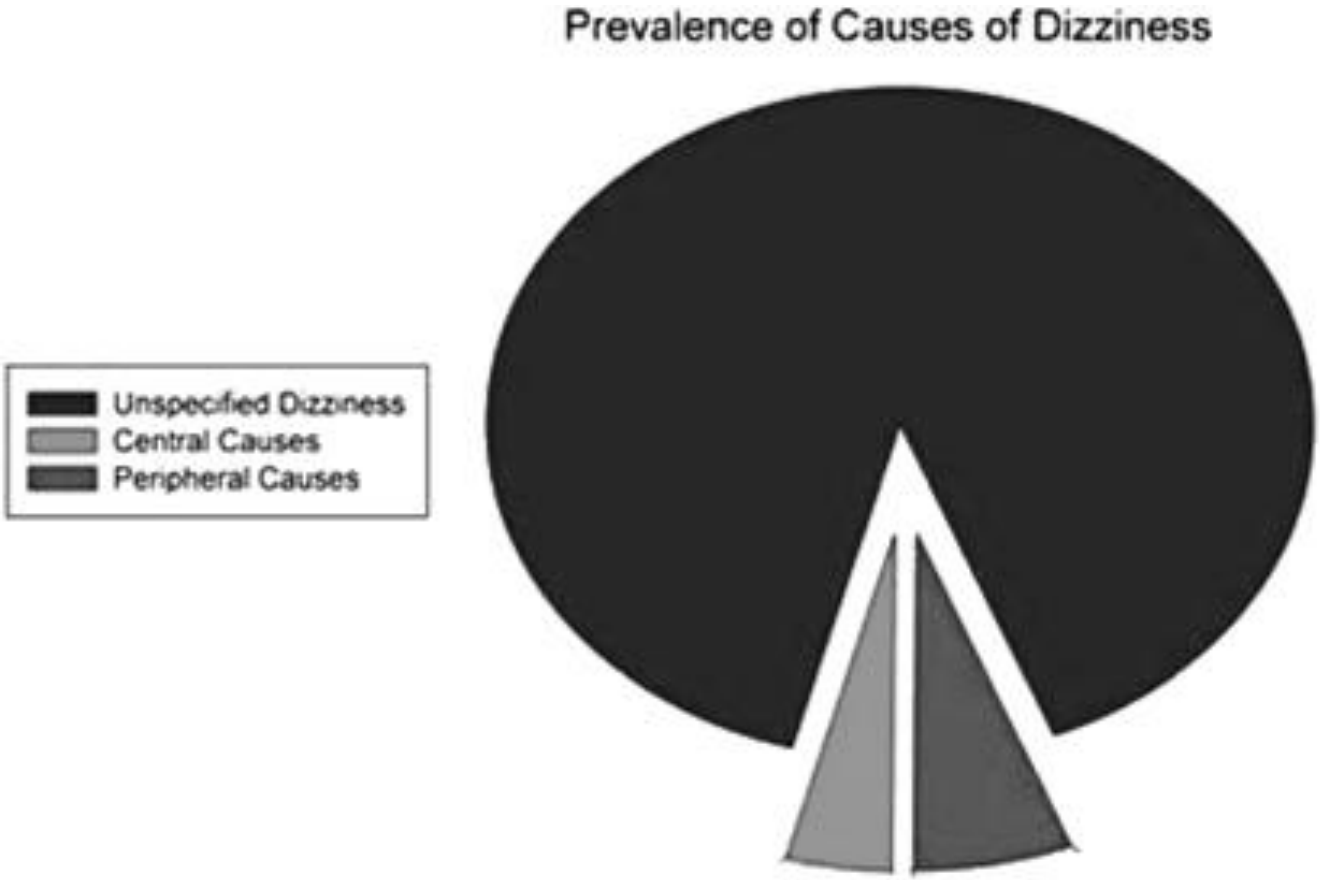
Pediatric Superior Canal Dehiscence



Prevalence

- 1.03% (5,793) of patients 0-18 yoa present with primary complaint related to balance
- 0.45% of patients 0-18yoa diagnosed with a balance disorder (O'Reilly et al 2010)
  - 4 year retrospective review of a pediatric health system records for ICD9 codes related to balance disorders: 561,151 inpatient and outpatient encounters were reviewed
  - 2,546 pediatric patients diagnosed with a balance disorder

What they found . . .



But…

- Vestibular impairment is the single most common associated feature of SNHL (Cushing, 2015 presentation)
- Studies have suggested that up to 85% of children with SNHL have some degree of vestibular impairment (Arnvig, 1955, Cushing et al., 2008, O'Reilly et al., 2011).
- So, 0.45% is likely an underestimation of prevalence in our clinical populations

Epidemiology of Dizziness and Balance Problems in Children in the United States: A Population-Based Study

Chuan-Ming Li, MD, PhD[1], Howard J. Hoffman, MA[1], Bryan K. Ward, MD[2], Helen S. Cohen, EdD, OTR[3], and Rose Marie Rine, PT, PhD[4,5]

- A multistage, nationally representative, probability sample of children (n = 10,954; aged 3-17 years) was examined based on the 2012 National Health Interview Survey Child Balance Supplement.
- Prevalence of dizziness and balance problems was 5.3% (3.3 million US children).
- 36.0% of children with dizziness and balance problems were seen by healthcare professionals during the past year and 29.9% received treatment.

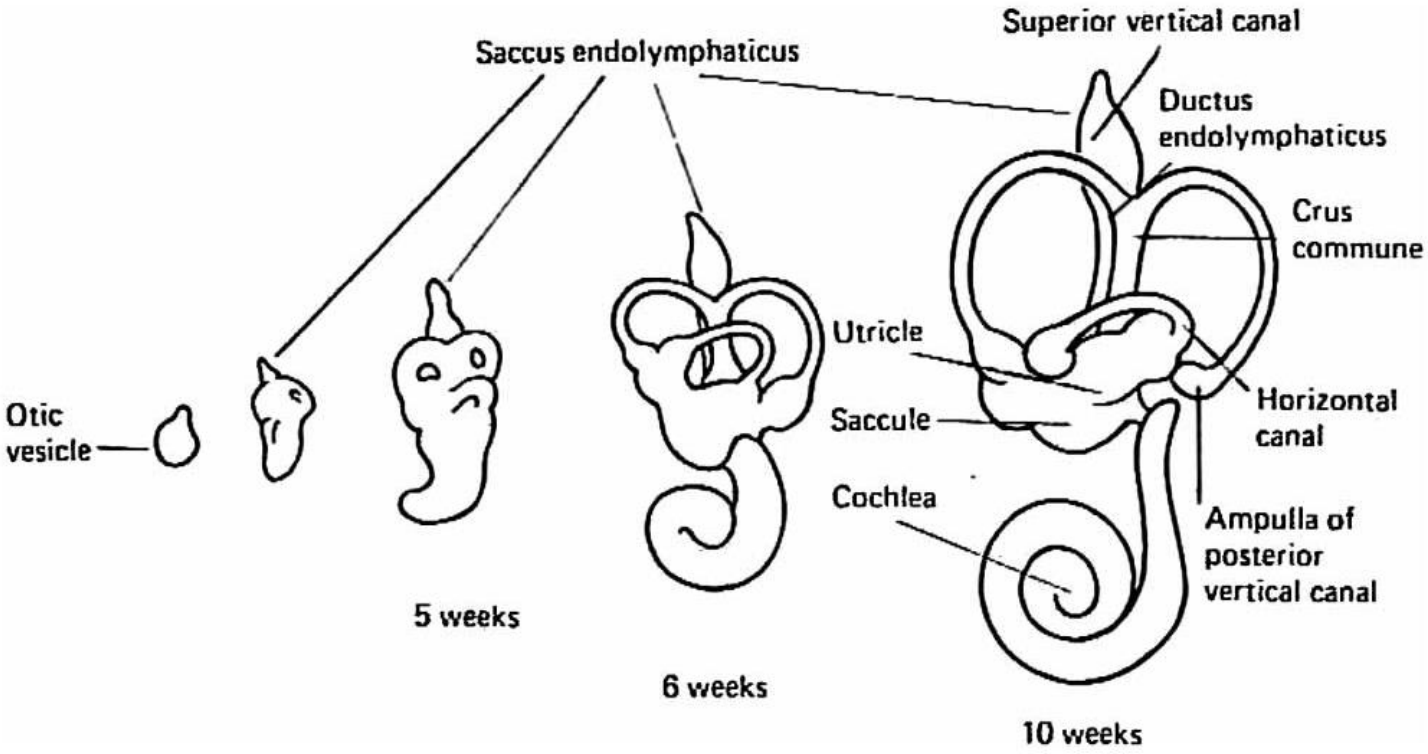
Outline

- Background
- Epidemiology
- Critical Period for the Vestibular System
- Migraine
- Genetics
- Cytomegalovirus
- Elarged Vestibular Aqueduct
- Summary

Development of the Vestibular System

- The vestibular system in humans is the first sensory system to develop.
- The vestibular nerve is the first cranial nerve to complete myelination and the system itself becomes functional in the 8th to 9th month of intrauterine life (Blayney, 1997).

Development of the Ear



Development of the Vestibular System

- Vestibular end-organ is fully developed at birth.
- Inhibitory influences and cerebellar control continue to develop until age 15.
- These findings make the case for developing and using normative data for tests of vestibular function.

(Casselbrant et al., 2010)

Development of the Vestibular System

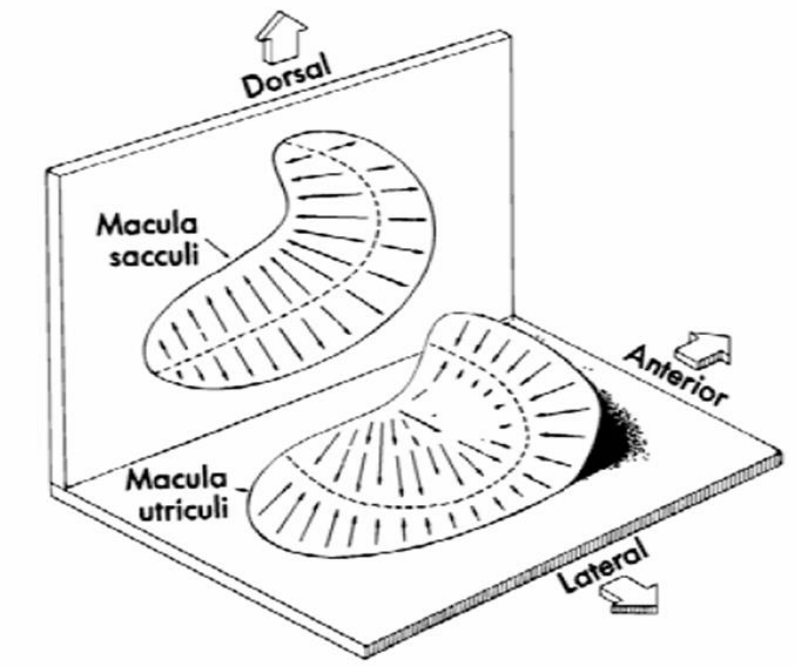
- Bony Labyrinth – 10 weeks gestation
- Membranous Labyrinth – 12 weeks gestation
- Myelination of neurons in the brain -20 weeks gestation
- Receptors in the vestibular system functional – 32 weeks gestation
- Vestibular nerve myelination – at birth

Is There a Critical Period for the Vestibular System?

- Gravity is a constant and all organisms on earth use it as a frame of reference for orientation in space.
- In most cases the critical periods were hypothesized when a definitive or at least a long lasting change was observed after the exposure to altered gravity during some developmental stages.

Otolith Organs

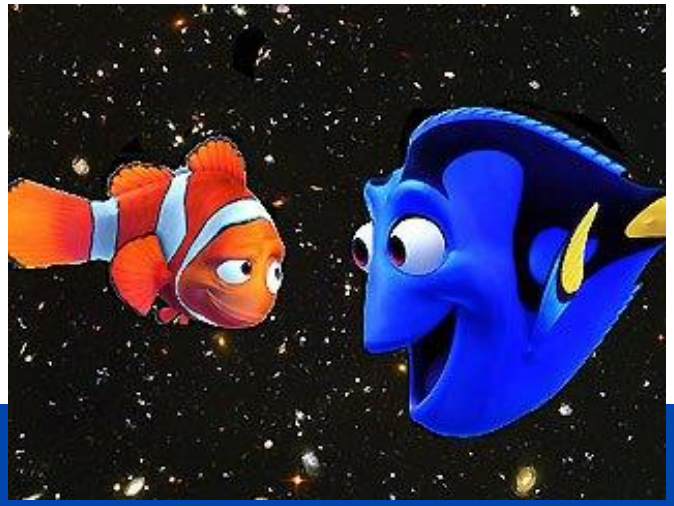
- Utricle has 2:1 surface area vs saccule
- Saccule is firmly attached to the temporal bone (less compliant)
- Utricle is attached only in the anterior region (more compliant)



Todd et al. 2009

Development

- Exposure to altered gravity during development of the vestibular system will alter the vestibular central structures (vestibular nuclei).
- In fish, the exposure to microgravity increased the number of synapses in some vestibular nuclei (Anken et al., 2002).



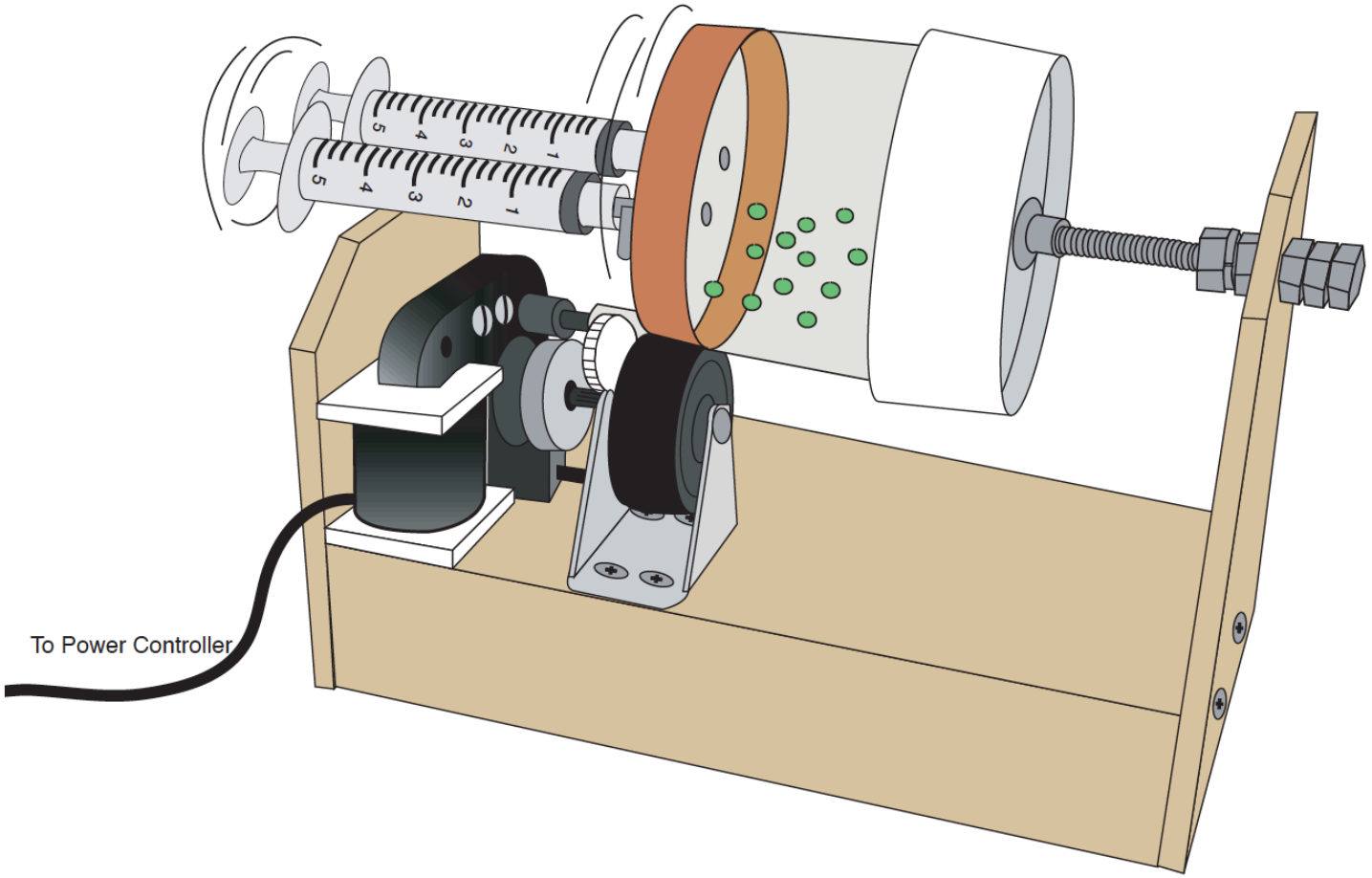
Is There a Critical Period for the Vestibular System?

- The central nervous system is believed to need environmental experience to calibrate the gravity information during critical periods of the development.
- This hypothesis has been explored many times and is one of the key issues in the developmental biology research of space and with regard to the colonization of different planets.

Zebrafish

- Tropical fresh-water fish that are part of the minnow family
- 70% of human genes are found in the Zebrafish
- Breed every 10 days and produce 50-300 eggs per day
- Knock-out/in

Is There a Critical Period for the Vestibular S

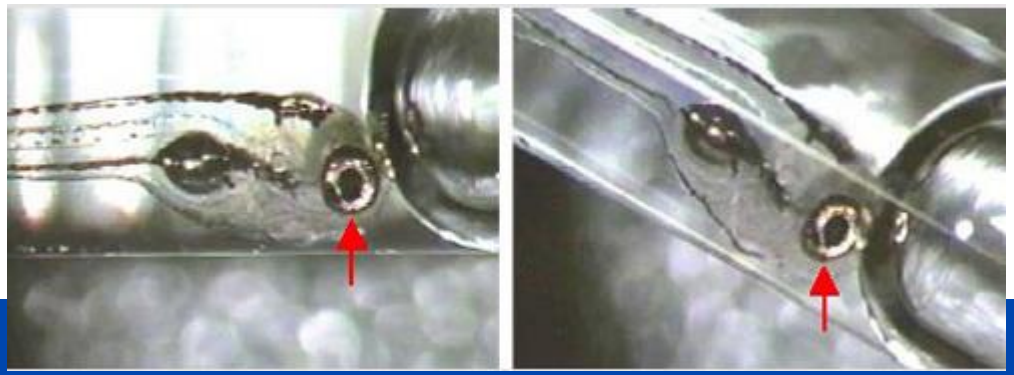


The Classroom Bioreactor

key element, a rotating plastic cylinder enclosing a tubular membrane that infuses growth media and oxygen and removes wastes.

Critical Period for the Vestibular System

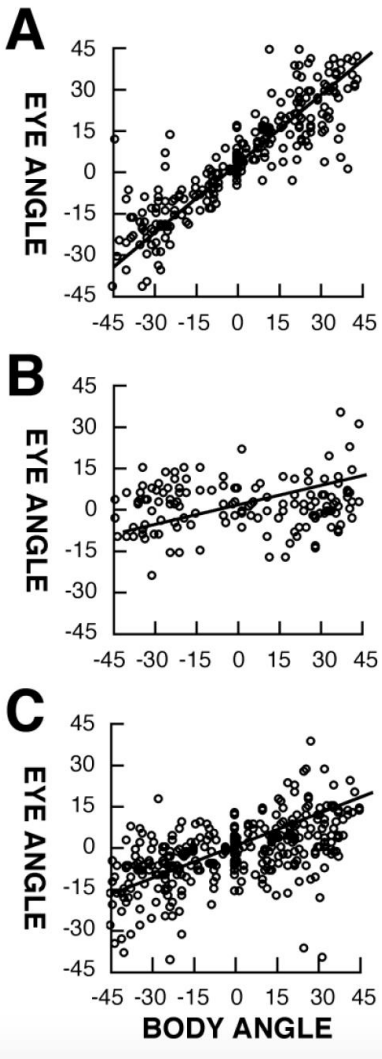
- Placed zebrafish embryos in the bioreactor at different time spans following fertilization.
- Once the fish matured they tested their vestibular reflexes
- Would perform the head tilt test – normal would keep their eyes in the plane relative to gravity



Head-tilt test performed on the same 96-hour old zebrafish hatchling to check for vestibular deficits.

Moorman

Moorman, 2002

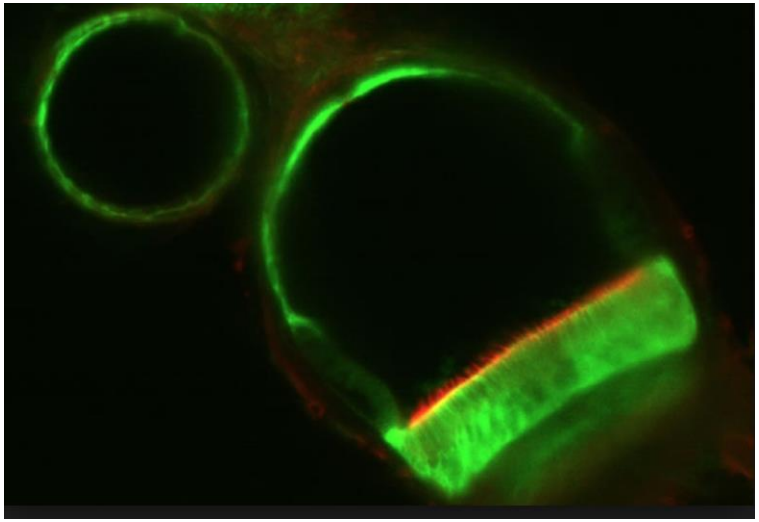


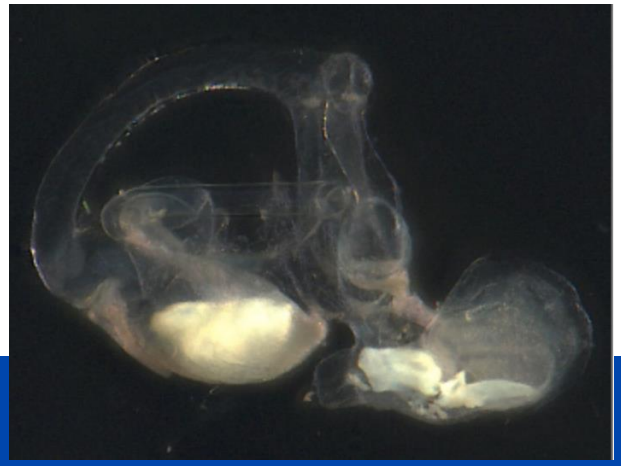
Normal controls

Experimental fish

Experimental fish – evaluated again

Mayo Clinic Zebra Fish Lab





Lisa Schimmenti, M.D., professor of pediatrics

Outline

- Development of the Vestibular/Balance System
- Balance Deficits in Children
- Research in Identification
- Research in Assessment
- Summary

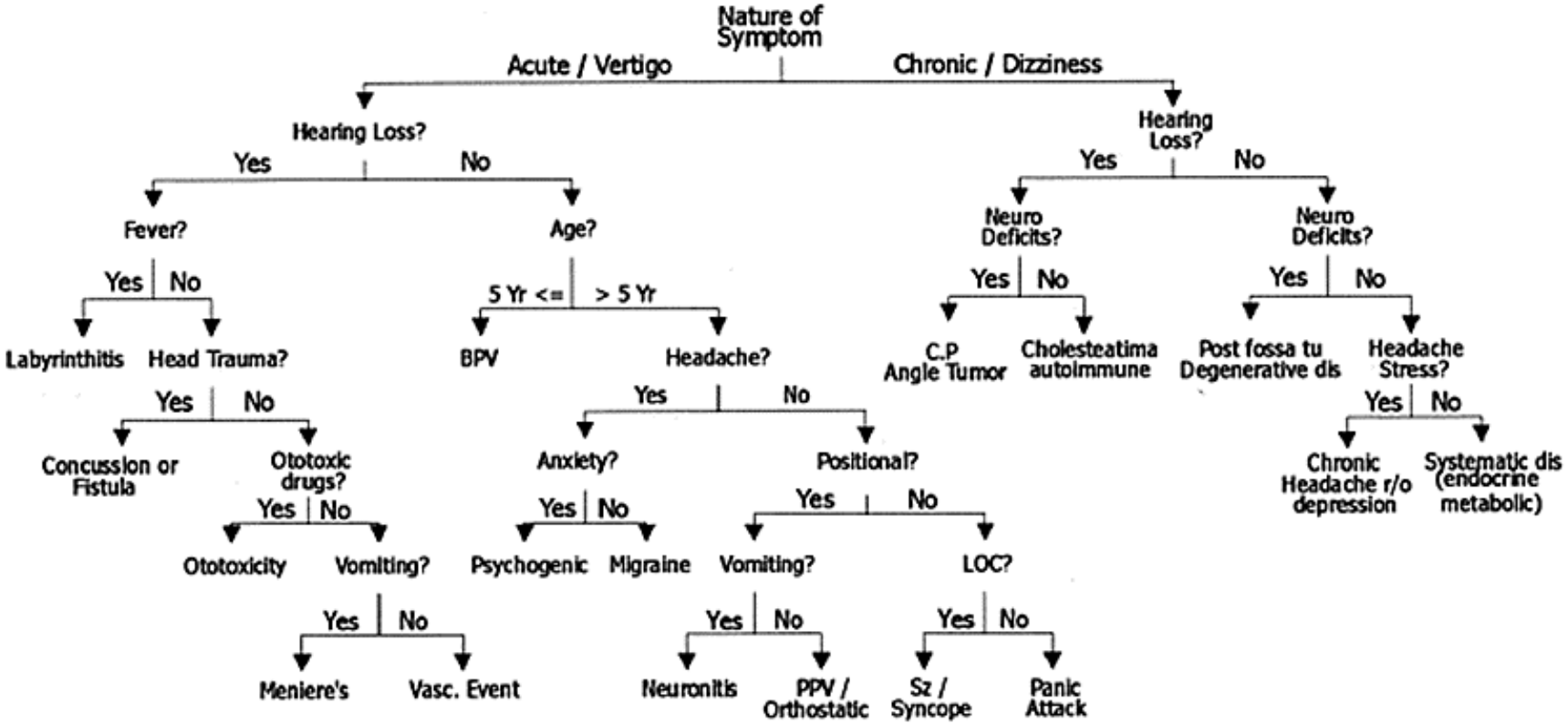
Early Identification-The problem

- Widespread national initiatives for early identification of permanent hearing loss in infants have brought an increased focus to the early auditory development of children.
- It is now well established that hearing loss can delay a child's receptive and expressive speech and language, result in reduced academic achievement, and have significant social consequences.

Early Identification

- The Joint Committee on Infant Hearing (JCIH) endorses early detection of and intervention (EDHI) for infants with hearing loss.
- The goal of EDHI is maximizing speech, language, and literacy development (JCIH position statement 2007), as children with hearing loss are known to be at risk for communication, cognition, reading, and social-emotional delays

How do we identify children with balance impairments?



Ravid et al., Pediatr Neurol 2003;29:318

How do we identify children with balance impairments?

Contents lists available at ScienceDirect

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The development of the vanderbilt pediatric dizziness handicap inventory for patient caregivers (DHI-PC)

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Purpose

- The purpose of the investigation was to develop a psychometrically sound dizziness disability/handicap outcome measure for use with a pediatric population between 5 and 12 years of age.
- Items comprising the alpha version of the DHI-P were created based on reports from parents, providers and patients.

DHI-P- (40 item)

NAME: DATE:

PEDIATRIC DIZZINESS HANDICAP INVENTORY (DHI)
(Age 5-12)

Instructions: The purpose of this questionnaire is to identify difficulties that your child may be experiencing because of your dizziness or unsteadiness. Please answer "yes", "no", or "sometimes" to each question. **Answer each question as it pertains to your child's dizziness problem only**.

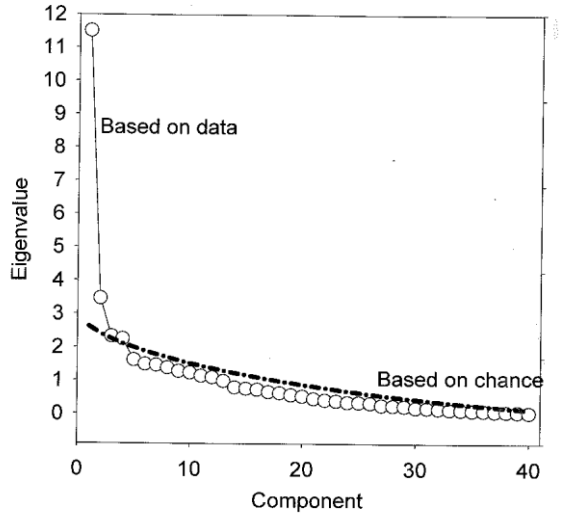
	Yes (4)	Sometimes (2)	No (0)
Does looking up increase your child's problem?			
Does performing more ambitious activities like sports or active play, (running, jumping, etc.) increase your child's problem?			
Do quick movements of your child's head increase his/her problem?			
Does turning over in bed increase your child's problem?			
Because of your child's problem, is it difficult for him/her to walk unassisted?			
Does bending over increase your child's problem?			
Do other people ask if there is something wrong with your child's balance?			
Is your child's balance unpredictable?			
Does your child use a great deal of effort to keep his/her balance?			
Is your child unable to run and move as he/she likes?			
Does your child's problem make him/her feel tired?			

Design

- Phase 1 – Initial item development
- Phase 2- Statistical analysis
- Phase 3 – Test-retest

Phase 1 Results

- A factor analysis showed there to be a single factor (eigenvalue of 11.51) that explained 29% of the total variance.



The Questionnaire for Dizziness, Eye, and Balance (Q-DEB) Function for Children and Adolescents

- Currently, no screening device exists to identify infants, children, and adolescents at risk for vestibular related impairments
- The purpose of this study is to develop a questionnaire to be used for screening vestibular related problems in children fror 1-21 years of age.

Development of the Q-DEB

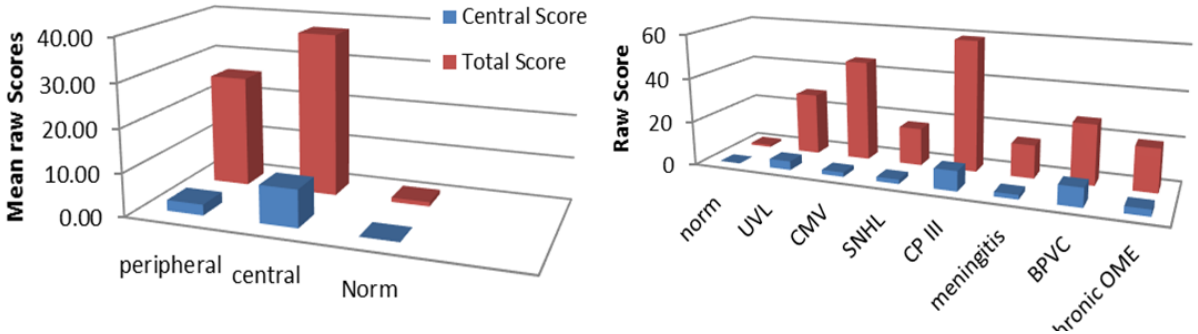
- Pilot and feasibility testing of the Q-DEB was completed using hypothetical clinical scenarios as well as participants from UAB, Children's of Alabama, and Vanderbilt University Medical Center.
- Participants were categorized as peripheral vestibular, central vestibular, or typically developing.

Q-DEB

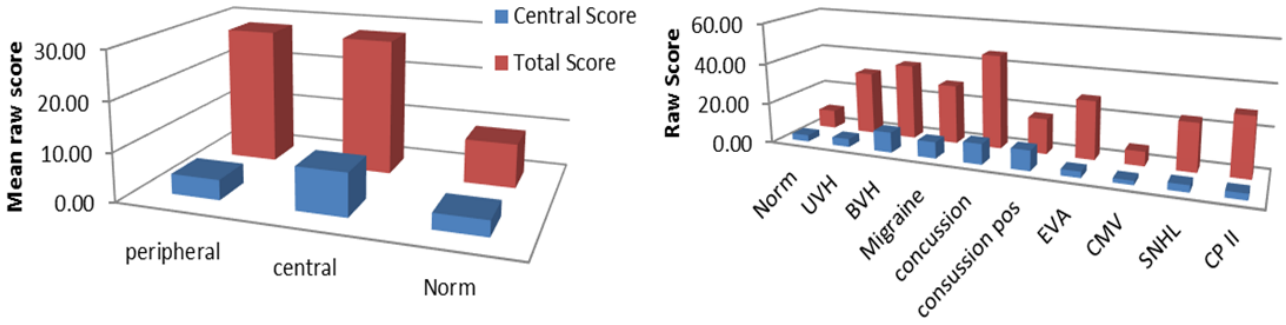
Catches a large playground ball thrown to him/her	Catches and holds with hands away from body	Traps against body	Is unable to do this
Catches a tennis ball thrown to him/her	Catches and hold with hands away from body	Traps against body	Is unable to do this
Throws a tennis ball	Overhand directly to a person or target	Overhand 3-5 feet in random direction	Is not able to throw small ball overhand
Pedaling a tricycle	Able to get on and pedal 5-10 feet	With help to get on it, can pedal 1-5 feet	Is unable to pedal the tricycle
Walking upstairs or a step	Steps up without rail or holding anything	Steps up but uses rail	Is unable to step up onto a step
Walking up multiple stairs	Alternating feet, 1 foot on each step	Steps up placing both feet on each step	Is unable to step up
walking down stairs or a step	Without a rail	Using a rail or someone's hand	Is not able to walk down stairs
walking down multiple stairs	Alternating feet, one on each step	Steps down bringing both feet onto each step	Cannot walk downstairs

Q-DEB
Results

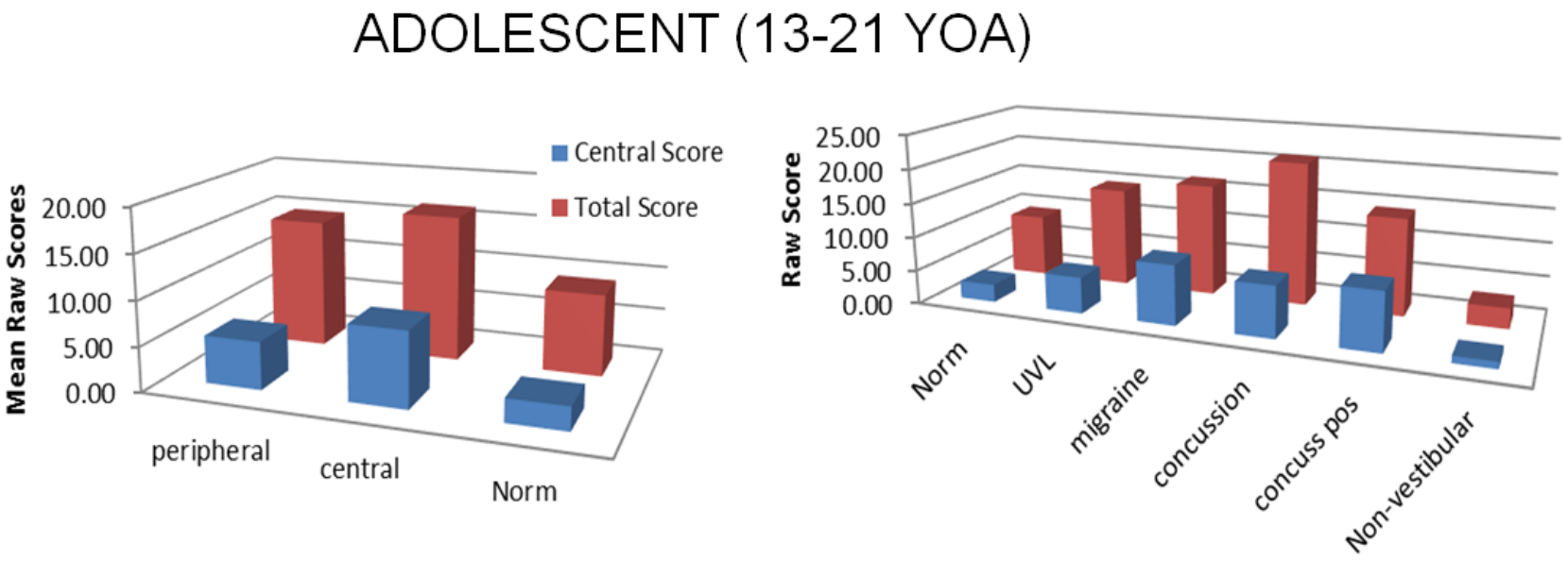
INFANT (1-5 YOA)



CHILD (6-12 YOA)



Q-DEB
Results



Additional Questionnaires

- The Pediatric Vestibular Symptom Questionnaire: A Validation Study (Pavlou et al., 2016).

Table I. The PVSQ

The following questions ask about how often you feel dizziness and unsteadiness. Please circle the best answer for you.
How often in the past month have you felt the following?

1. A feeling that things are spinning or moving around

3	2	3	4	?
Most of the time	Sometimes	Almost never	Never	Don't know

2. Unsteadiness so bad that you actually fall

3	2	3	4	?
Most of the time	Sometimes	Almost never	Never	Don't know

3. Feeling sick

3	2	3	4	?
Most of the time	Sometimes	Almost never	Never	Don't know

4. A light-headed or swimmy feeling in the head

3	2	3	4	?
Most of the time	Sometimes	Almost never	Never	Don't know

5. Feeling of pressure in the ear(s)

3	2	3	4	?
Most of the time	Sometimes	Almost never	Never	Don't know

6. Blurry vision, difficulty seeing things clearly, and/or spots before the eyes

3	2	3	4	?
Most of the time	Sometimes	Almost never	Never	Don't know

7. Headache or feeling of pressure in the head

3	2	3	4	?
Most of the time	Sometimes	Almost never	Never	Don't know

8. Unable to stand or walk without holding on to something or someone

3	2	3	4	?
Most of the time	Sometimes	Almost never	Never	Don't know

9. Feeling unsteady, about to lose balance

3	2	3	4	?
Most of the time	Sometimes	Almost never	Never	Don't know

10. A fuzzy or cotton wool feeling in the head

3	2	3	4	?
Most of the time	Sometimes	Almost never	Never	Don't know

11. Do any of these symptoms stop you doing what you want to do?
If yes, which ones? .....

AJA

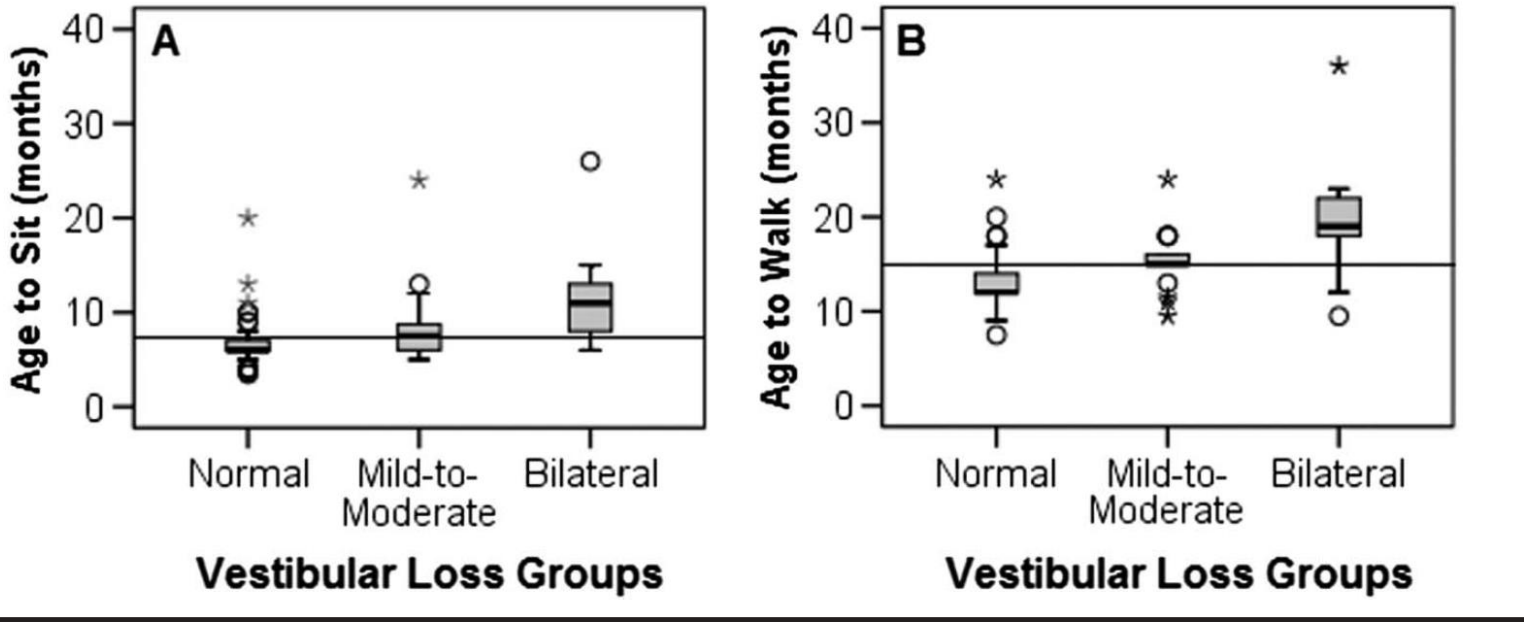
Research Article

Predictive Factors for Vestibular Loss in Children With Hearing Loss

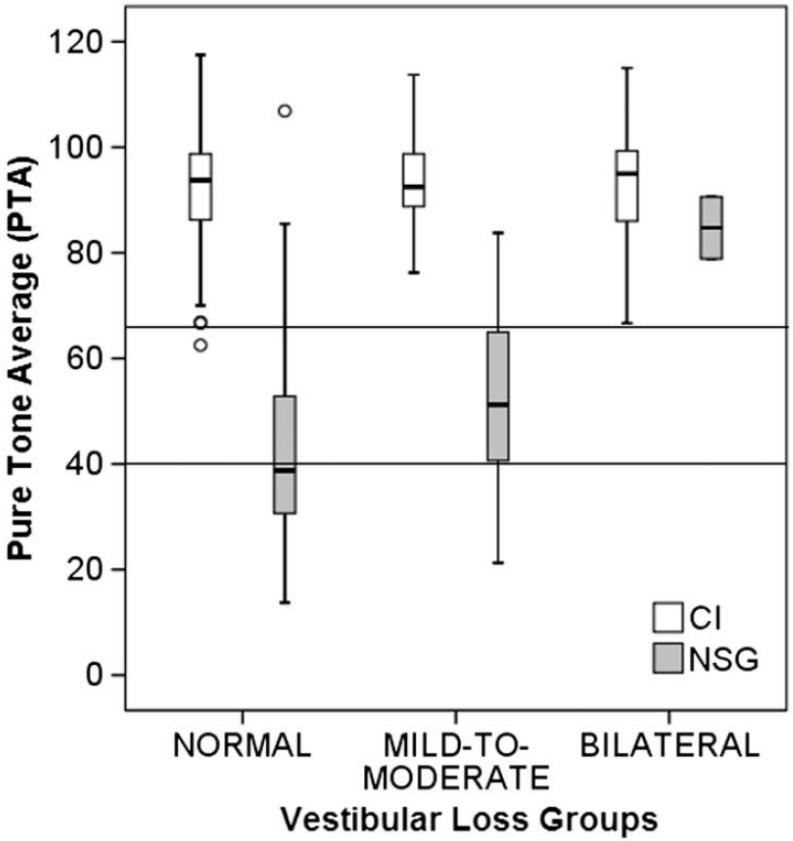
Kristen L. Janky,[a] Megan L. A. Thomas,[a] Robin R. High,[b]
Kendra K. Schmid,[b] and Oluwaseye Ayoola Ogun[c]

- 186 medical records were reviewed for:
  - Degree of hearing loss
  - Degree of vestibular impairment
  - Imaging
  - Parental concerns regaring gross motor skills
  - Developmental Profile (DP-3)
  - Any co-morbidities

Age to Sit and Walk (Significantly increased with degree of vestibular impairment



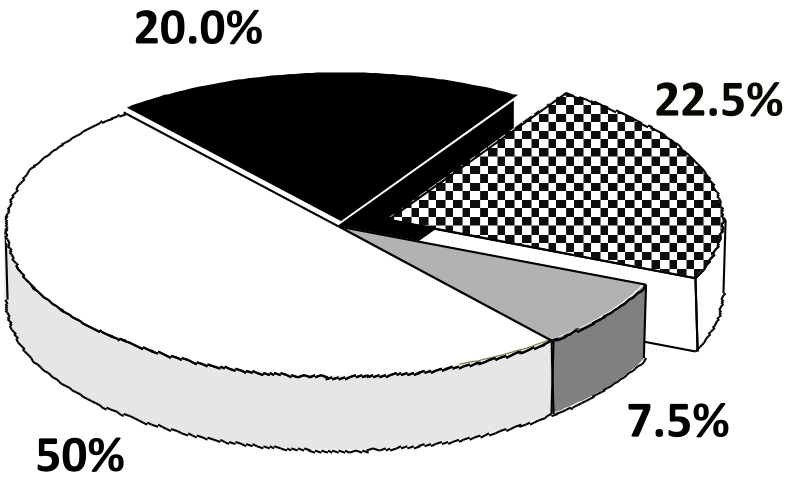
Degree of Hearing Loss



Take Home

- Degree of HL > 66 dB
- Parent concern for gross motor development
- Age to sit >7.25 months
- Age to walk >14.5 months

Vestibular Function and CI Candidates



Normal vestibular function

Asymetrical Vi

Symetrical Vi

Complete bilateral vestibular loss

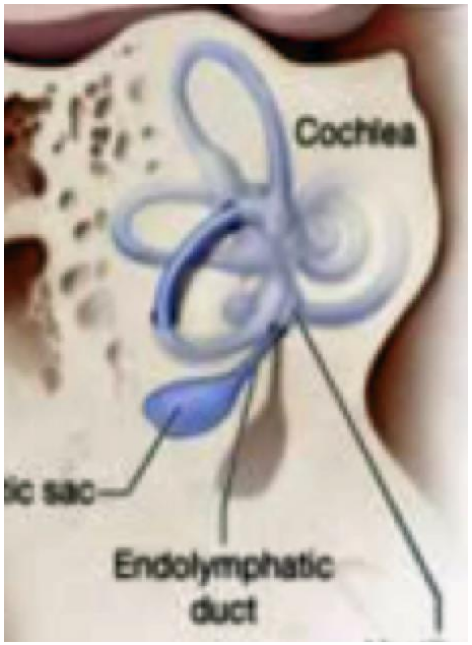
Wiener-Vacher & Quarez , 2018

Outline

- Background
- Epidemiology
- Critical Period for the Vestibular System
- Meniere's and Migraine
- Genetics
- Cytomegalovirus
- Elarged Vestibular Aqueduct
- Summary

Vestibular Deficits in Children with Hearing Impairment (con't)

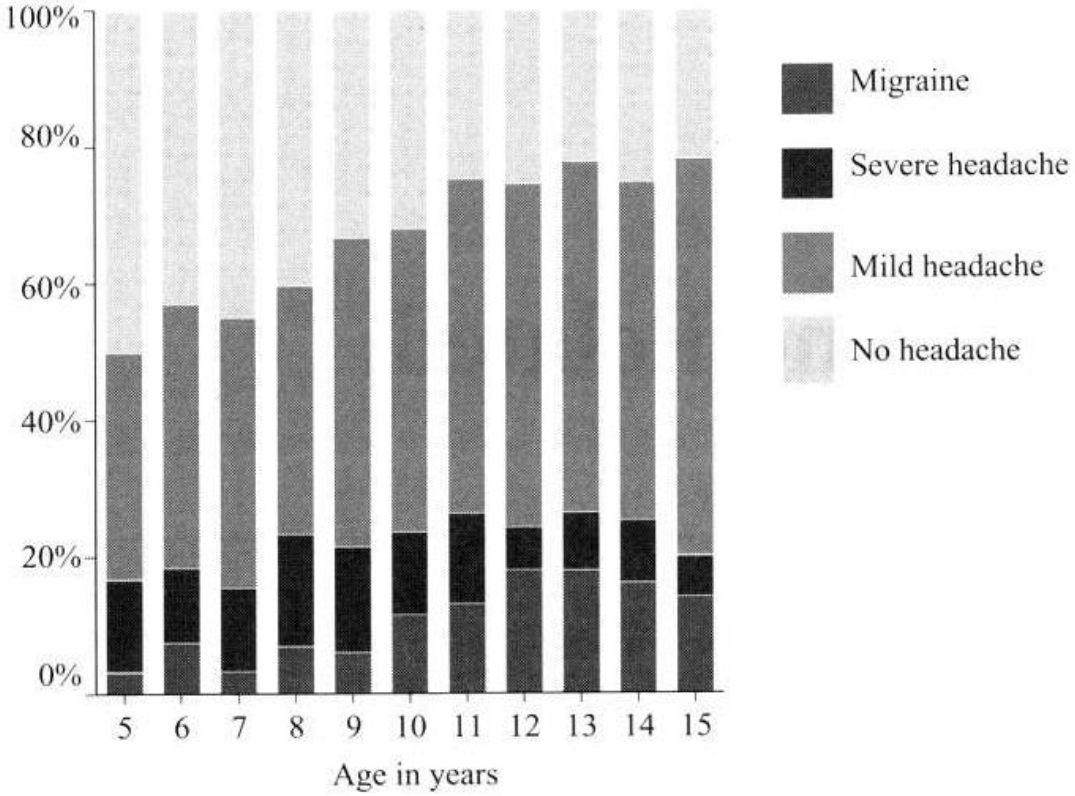
- Meniere's disease
  - Earliest report is 9 years of age by Mizukoshi (2001)-Interesting work emerging at Harvard – Steve Rauch, MD.
  - Menières-like syndrome (1.1%) Wiener-Vacher, 2018.



Epidemiology

- The prevalence of migraine increases with age, from 0-3% in preschool children to 20% in adolescents (Gunner and Smith, 2007).
- The peak age is 12-14 years
- Migraines are more common in boys early on but after age 12 it is more common in girls`

Headache in Children



Abu-Arefeh and Pearlman, 2006

Migraine Variants in Children

- Benign Paroxysmal Torticollis (BPT)
- Benign Paroxysmal Vertigo of Childhood (BPV)
- Benign Recurrent Vertigo of Adulthood (BRVA)

Basilar Migraine

- Recurrent headaches localized to occipital region
- Multiple neurologic symptoms localized to the posterior fossa including:
  - Tinnitus
  - Fluctuating hearing loss
- Can occur in both sexes at any age
- Neurological symptoms caused by ischemia in the basilar artery distribution (vasospasm).

Basilar Migraine

- Reports of fluctuating low frequency hearing loss, often bilateral and that is reversible between attacks
- Patients respond to migraine prophylactic medications including beta blockers and calcium channel blockers.

(Harker & Rassekh, 1987; Olsson, 1991)

Basilar Migraine

Table 1.—Most Common Symptoms in 49 Patients With Basilar Migraine*

	% of Cases
Headache (usually occipital)	96
Nausea	83
Vomiting	71
Vertigo	63
Gait ataxia	63
Paresthesias (usually bilateral)	61
Dysarthria	57
Weakness (usually bilateral)	55
Tinnitus	26
Impaired hearing	20
Double vision	16

From: Baloh, 1997

Vestibular Migraine

- Until recently, there has been a lack of a universally accepted definition of vestibular
- Accordingly, the Barany Society, which represents the international community mandated a classification group to develop diagnostic criteria for vestibular migraine.

Vestibular Migraine

Vestibular migraine: Diagnostic criteria

Consensus document of the Bárány Society and the International Headache Society

Thomas Lempert[a,*], Jes Olesen[b], Joseph Furman[c], John Waterston[d], Barry Seemungal[e], John Carey[f], Alexander Bisdorff[g], Maurizio Versino[h], Stefan Evers[i] and David Newman-Toker[j]

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Vestibular Migraine Symptoms

- Spontaneous vertigo including internal vertigo (false sensation of self motion)
- External vertigo (false sensation that the visual surround is turning)
- Positional vertigo
- Visually-induced vertigo (triggered by a complex or large moving visual stimulus)

Vestibular Migraine Symptoms

- Head motion-induced vertigo.
- Head motion-induced dizziness with nausea.
- Dizziness is characterized by a sensation of disturbed spatial orientation.

Vestibular Migraine

- At least 5 episodes with vestibular symptoms1 of moderate or severe intensity, lasting 5 min to 72 hours.
- Current or previous history of migraine with or without aura according to the ICHD.
- Not better accounted for by another vestibular or ICHD diagnosis.

MAYO CLINIC

Lempert et al., *Journal of Vestibular Research*, 2013

Vestibular Migraine

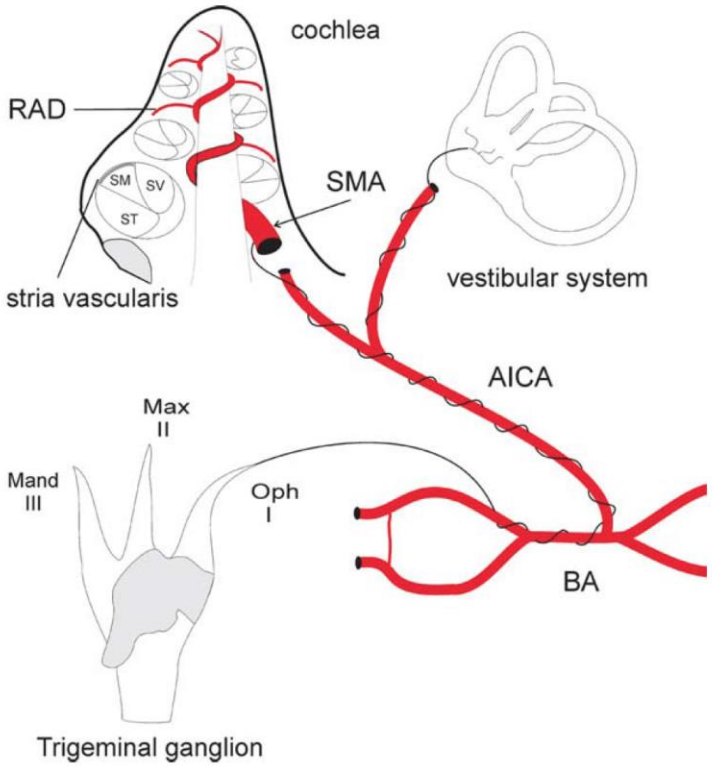
- One or more migraine features with at least 50% of the vestibular episodes:
  - headache with at least two of the following characteristics
  - one sided location, pulsating quality
  - moderate or severe pain intensity, aggravation by
  - routine physical activity
  - photophobia and phonophobia
  - visual aura

Auditory Symptoms

- Less common in migraine:
  - Phonophobia occurs in ~67% of migraine
  - Low frequency hearing loss (as seen in Meniere's Syndrome) occurring most often in young women during menstrual period
- Migraine has been identified as one cause of sudden hearing loss, often profound, that persists

Pathophysiology of VM

- Capsaicin stimulation of the cochlea and electric stimulation of the trigeminal ganglion mediate vascular permeability in cochlear and vertebro-basilar arteries



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- Cytomegalovirus
- Genetics
- Enlarged Vestibular Aqueduct
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Cytomegalovirus (CMV)

- The effects of congenital CMV on the brain and auditory system of newborns have been well documented
- Vestibular dysfunction due to CMV is less well understood but, nevertheless, exists.
- Both hearing and balance should be routinely assessed in individuals with congenital CMV.

Cytomegalovirus (CMV)

- Cytomegalorivus (CMV) is associated with a herpes simplex virus that can be passed from mothers to babies during pregnancy, affecting 1% of all newborns.
- The majority (90%) of babies infected with CMV in utero are asymptomatic at birth
- Approximately 20% will go on to show hearing or vision loss, seizures or other neurologic problems.

Cytomegalovirus (CMV)

- Symptoms of an active infection at birth include:
  - jaundice
  - Petechiae
  - hepatosplenomegaly (HSM)
- Asymptomatic and symptomatic infants can develop later health problems or disabilities.
- At the moment, there is no vaccine to prevent CMV. Preventative measures and antiretroviral treatment is important in infants manifesting symptoms.

Cytomegalovirus (CMV)

- Sequelae include
  - Sensorineural hearing loss (SNHL)
  - Retinitis
  - Intellectual disability
  - Microcephaly
  - Seizures
  - Cerebral Palsy
  - Balance impairments

Cytomegalovirus (CMV)

- Sensorineural hearing loss (SNHL)
  - Incidence: 10-16% will develop hearing loss (Goderis, 2014)
    - 1 in 3 with symptomatic CMV
    - 1 in 10 with asymptomatic CMV
  - Prevalence difficult to estimate due to late-onset hearing loss (33-50% estimated) as well as asymptomatic CMV
    - Suggested that 25% of children under 4yoa with hearing loss may be due to congenital CMV (Fowler, KB)

Audiovestibular sequelae of congenital cytomegalovirus infection in 3 children presumably representing 3 symptomatically different types of delayed endolymphatic hydrops

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Cytomegalovirus (CMV)

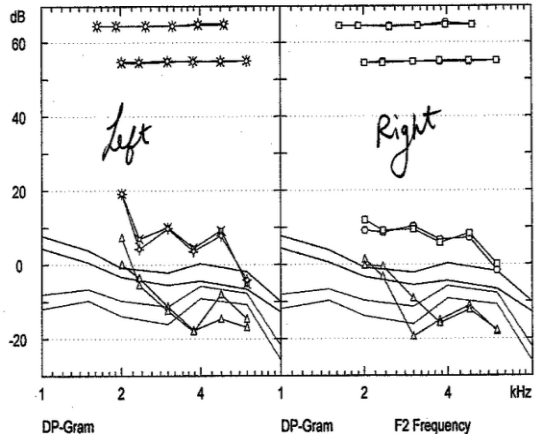
- Case 1 had no hearing in one ear and severe progressive hearing loss in the other ear; he showed vestibular symptoms at the age of 4.5 years.
- Case 2 had severe but stationary hearing loss in one ear and showed hearing impairment symptoms in the other ear at 9-13 years of age. May go on to develop delayed endolymphatic hydrops.

Cytomegalovirus (CMV)

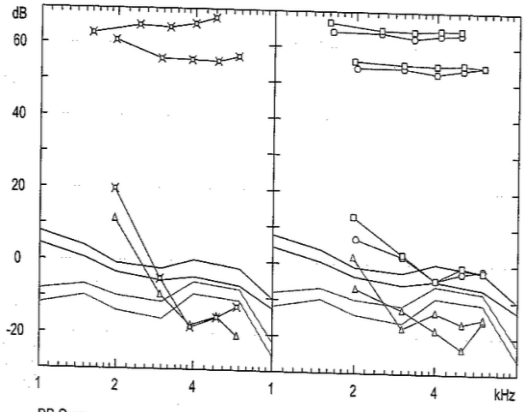
- Case 3 did not have hearing impairment symptoms, or vestibular symptoms, but was found to have severe progressive hearing loss from the age of 15 months onwards, which led to profound deafness at the age of 2 years and vestibular areflexia at or before the age of 4 years.

Cytomegalovirus (CMV)

- 6 year old
- Mother identified with CMV at 22 weeks
- Passed newborn hearing screening
- 4 months old
  - Present DPOAEs and reflexes

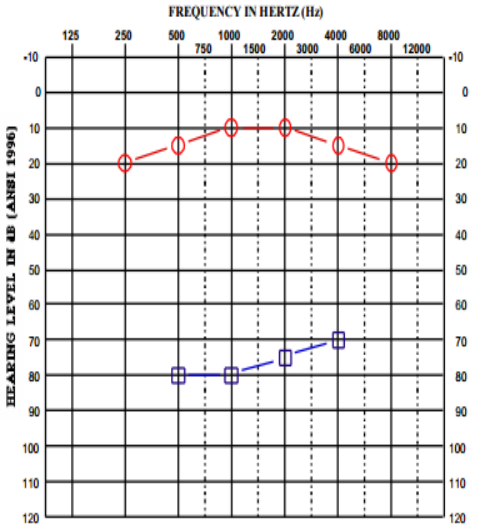


Cytomegalovirus (CMV)



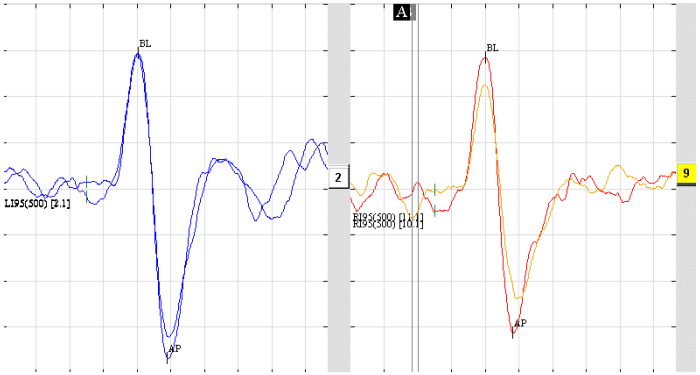
- 13 months of age

  DPOAEs absent 3000-6000Hz left ear

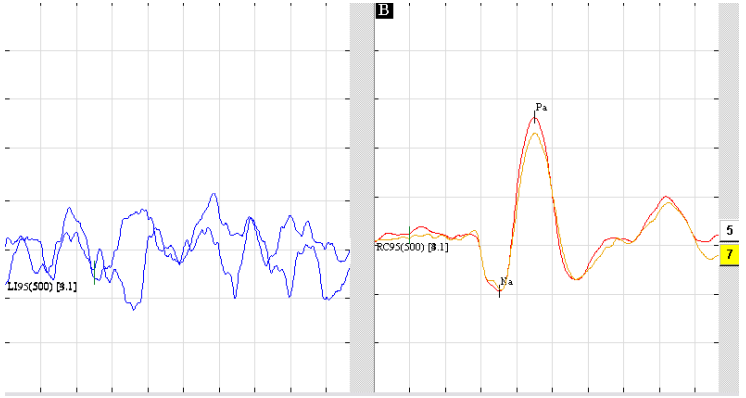


- Current

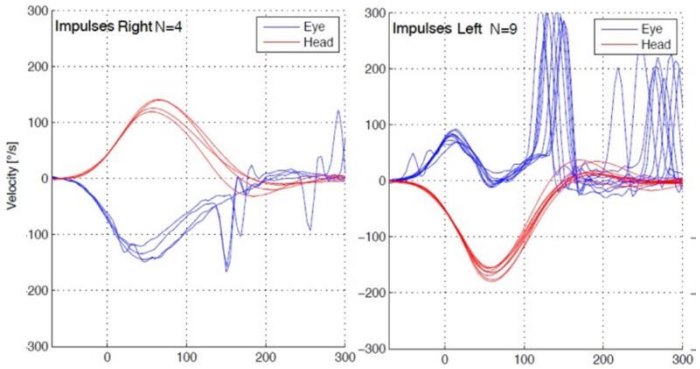
Vestibular Laboratory Testing (6 yo)



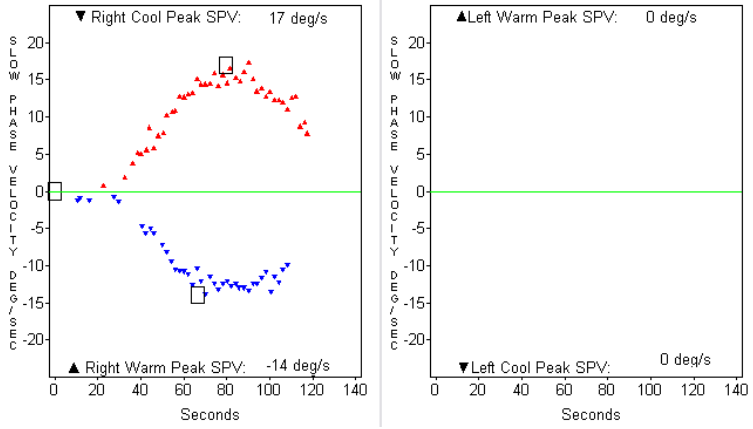
Cervical VEMP



Ocular VEMP



Video Head Impulse



Caloric

Outline

- Background
- Epidemiology
- Critical Period for the Vestibular System
- Migraine
- Cytomegalovirus
- Genetics
- Enlarged Vestibular Aqueduct
- Summary

Vestibulopathy due to Heredity

- This refers to an individual having a phenotypic trait of vestibular dysfunction due to a gene that may or may not be directly involved in the structure or function of the inner ear
- Genetics of the inner ear have been largely focused on hearing loss.
- Numerous opportunities exist for the study of genetics of the vestibular system.

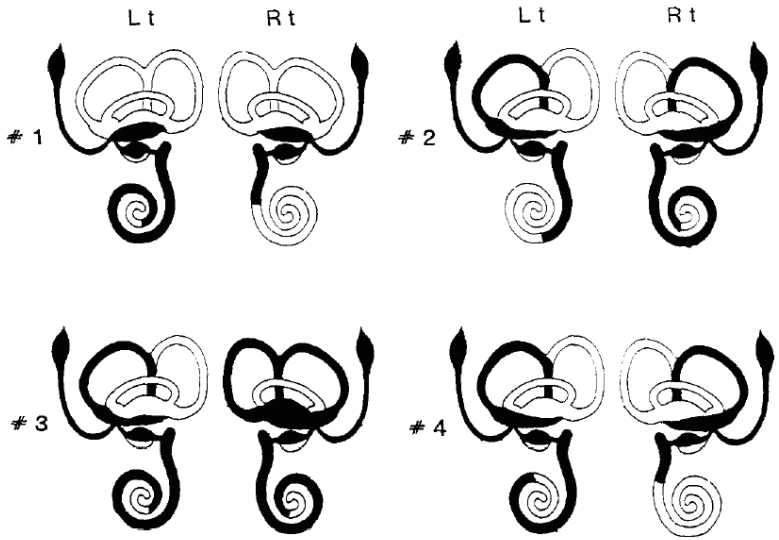
Genetics and Vestibulopathy

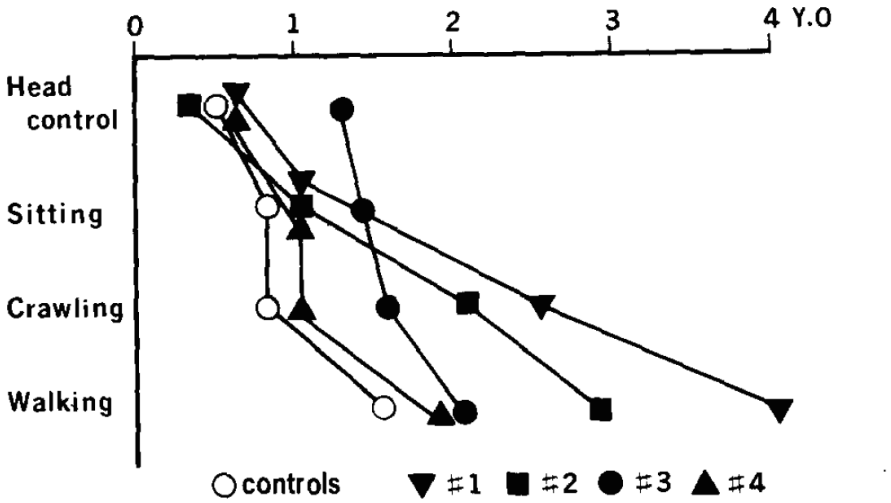
- Numerous advancement in understanding the the genetics of hearing loss has occurred over the last 10 years,
- There is very little is known about what role genes play in vestibular impairment.
- . Not all genetic mutations produce both cochlear and vestibular abnormalities

Inner Ear Malformations

- Mondini Malformation
  - Development is interrupted during the 6th-7th week gestation
  - Basal turn of the cochlea often develops to approximately 1.5 turns.
  - Variable hearing loss
  - Vestibular function is variable as well
- The most frequent etiology of vestibular impairments are inner ear malformations.
- They represent 13.5% (n=140/1037) of the children referred for balance disorders (Wiener-Vacher, 2018).

Milestones and Inner Ear Abnormalities





White represents absent portions system.

Syndromes Associated with Vestibular Deficits

- Usher syndrome
- Alport syndrome
- Pendred syndrome
- Waardenburg syndrome
- CHARGE syndrome

Usher Syndrome

- Usher syndrome type 1
- The Usher syndromes (USH) are a heterogeneous group of autosomal recessive disorders.
- Characterized
  - congenital hearing loss
  - retinitis pigmentosa
  - Vestibular impairment
- 10 Usher syndrome genes
- USH1 - USH3 associated with vestibular impairment

Vestibular dysfunction of patients with mutations of Connexin 26

Ingo Todt,[1] Hans Christian Hennies,[2] Dietmar Basta[1] and Arne Ernst[1,CA]

[1]Department of Otolaryngology at UKB, Hospital of the University of Berlin (Charité Medical School); [2]Department of Molecular Genetics and Gene Mapping, Max Delbrück Center for Molecular Medicine, Berlin, Germany

Connexin 26

- Connexin 26 is a beta type gap junction protein.

  GJB2 = gap junction beta 2

- Connexin 26 is expressed in the cochlea: stria vascularis, basilar membrane, spiral prominence and limbus.
- Connexins form gap junctions believed to be involved in K+ recycling in the cochlea.
- Vestibular system has a smaller potential (endocochlear-vestibular) 150mV vs 65 mV

Connexin 26 – vestibular findings

- Findings:
  - 5/7 Five had abnormal cervical VEMPs. Suggests a loss of saccular function.
  - Utricular and semicircular canal function was determined to be normal.
  - Authors felt that connexin 26 mutations could be associated with saccular impairment only.

Table I. Overview of genetic, audiologic (maximum hearing loss) and neurotological test results of all patients investigated.

Patient ID	Mutation in *GJB2*	Consequence	Hearing loss	VEMP	SHV	VOR	Subjective dizziness
I.B.	homo. c.35delG	Gly12fsX2	Deaf	None	Regular	Regular	None
R.S.	hetero. c.79G > A	Val27Ile	Low frequencies 40 dB	None	Regular	Regular	None
J.S.	hetero. c.457G > A	Val153Ile	Low frequencies 55 dB	None	Regular	Regular	None
I.L.	hetero. c.109G > A	Val37Ile	High frequencies 20 dB	Regular	Unilateral pathological	Regular	Yes
C.J.	hetero. c.101T > C	Met34Thr	Low frequencies 60 dB	None	Regular	Regular	None
C.D.	hetero. c.355G > A	Glu119Lys	High frequencies 50 dB	Regular	Regular	Regular	None
A.T.	comp. hetero. c.30delG c.313_326del	Gly12fsX2 Lys105fsX5	Pantonal 40 dB	None	Regular	Regular	None

"However, these defects are well compensated and do not decisively influence the quality of daily life of those patients."

Diagnosing Genetic Disorder in Children with Balance Disorders

- Typically there are a two red flags:
  - Multiple organ systems effected (as in a syndromic disorder)
  - Presence of more than one affected family member
- Detailed family history
- Referral to clinical geneticist should be pursued.

Why does it matter?

- Better understanding of biology of normal and abnormal hearing
- Facilitate development of animal models (useful for studying pathophysiology)
- Early molecular diagnosis
- Genetic counseling
- Possible prevention
- New strategies for therapeutic intervention (gene therapy?)

Outline

- Background
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- Critical Period for the Vestibular System
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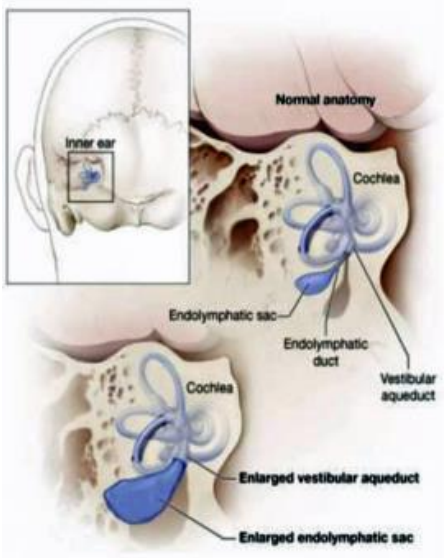
- Enlarged Vestibular Aqueduct
- Summary

Enlarged Vestibular Aqueduct (EVA)

- EVA describes a dilation of the vestibular aqueduct, which is the bony canal located in the petrous portion of the temporal bone that houses the endolymphatic duct and sac.
- First described by Valvassori and Clemis (1978)
- It is the most common inner ear malformation associated with early-onset sensorineural hearing loss, usually affecting children within the first few years of life.

Enlarged Vestibular Aqueduct (EVA)

- Although it can manifest itself in many different ways, Steinbach et al., (2006) reported that as many as 15% of pediatric patients with sensorineural hearing loss have EVA.



Vestibular Dysfunction in Patients with Enlarged Vestibular Aqueduct

Chris K. Zalewski, PhD[1,*], Wade W. Chien, MD[1,2,*], Kelly A. King, PhD[1], Julie A. Muskett, MS[1], Rachel E. Baron[1], John A. Butman, MD, PhD[3], Andrew J. Griffith, MD, PhD[1], and Carmen C. Brewer, PhD[1]

- 45% of patients with EVA had vestibular signs and symptoms
- 44% of tested patients with EVA had abnormal VNG test results.
- An increased number of vestibular signs and symptoms was correlated with the presence of bilateral EVA ($P = .008$) and a history of head injury ($P < .001$).
- Abnormal VNG results also correlated with a history of head injury ($P = .018$).

Prognostic Factors for Sudden Drops in Hearing Level After Minor Head Injury in Patients With an Enlarged Vestibular Aqueduct: A Meta-analysis

Bo Jan Noordman, Eveline van Beeck Calkoen, Birgit Witte, Theo Goverts, Erik Hensen, and Paul Merkus

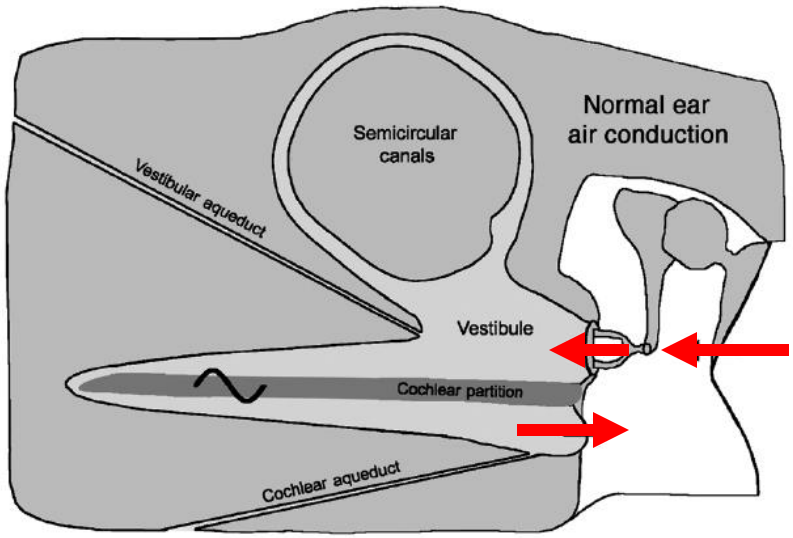
- Objective: To identify factors associated with sudden drops in hearing level after minor head trauma in patients with EVA.
- 1/3 of the patients with EVA experienced sudden drops in hearing level because of head trauma.
- A significant association was found between preexisting fluctuating HL and sudden drops in hearing level caused by trauma.

Progressive Hearing Loss and Head Trauma in Enlarged Vestibular Aqueduct: A Systematic Review and Meta-analysis

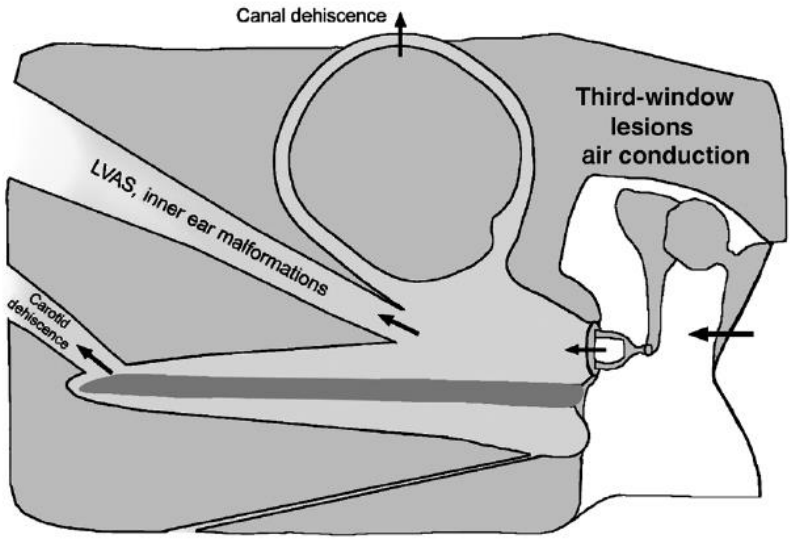
A. Sean Alemi, MD[1], and Dylan K. Chan, MD, PhD[1,2]

- Twenty-three studies (1115 ears with enlarged vestibular aqueduct) met inclusion criteria.
- Progressive SNHL hearing loss was found in 39.6% of ears, with trauma associated progression in 12%.

Pathophysiology



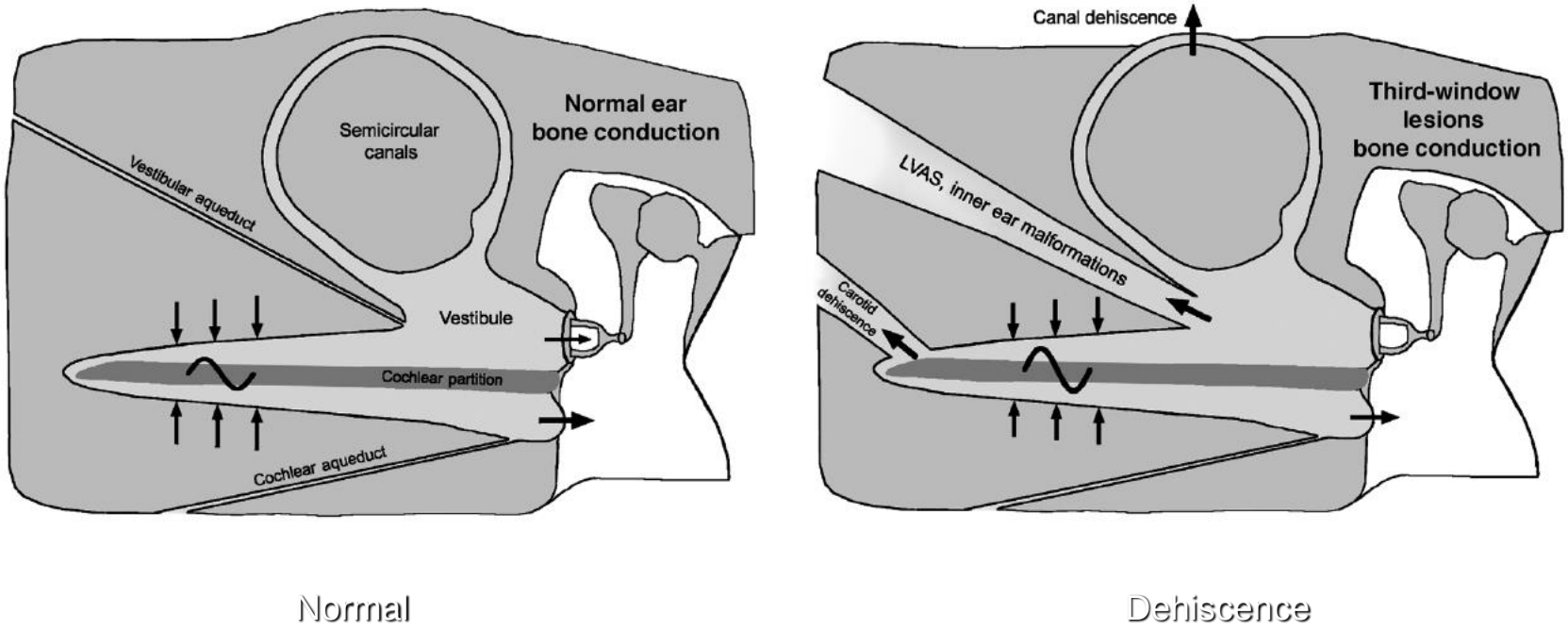
Normal



Dehiscence

Otology & Neurotology
29:282–289 © 2008, Otology & Neurotology, Inc.

Bone Conduction



Normal Dehiscence

Sets up an inequality in the impedance between the oval and round windows.

Otology & Neurotology
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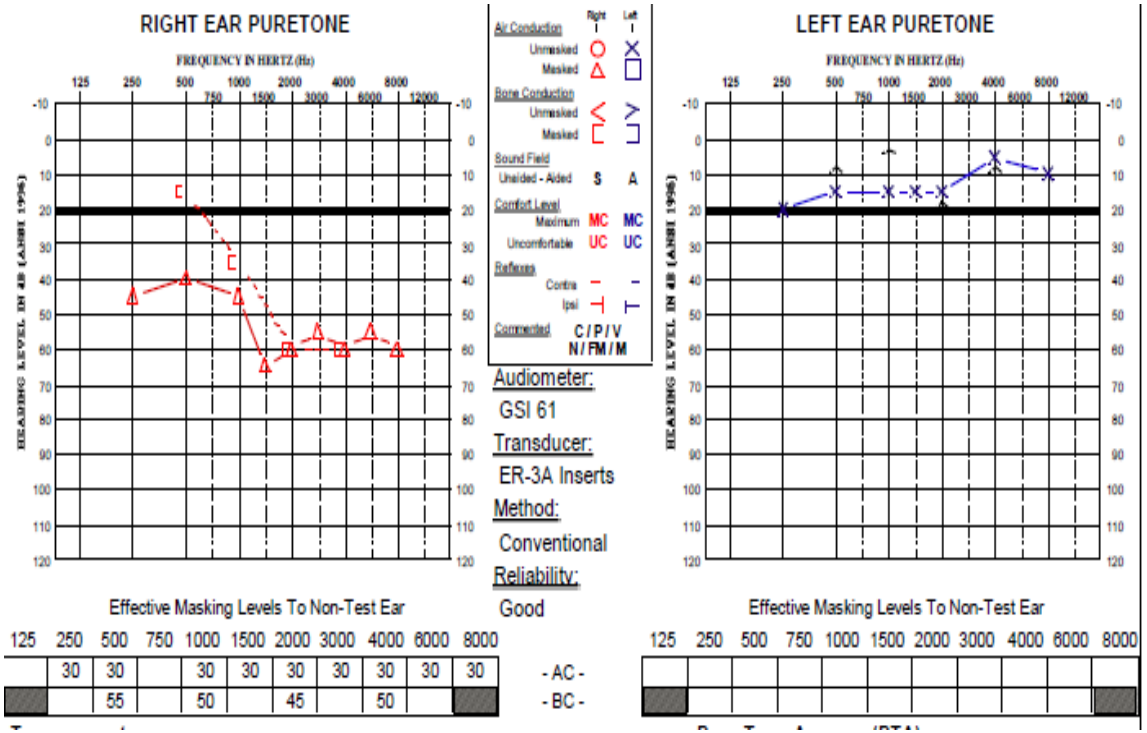
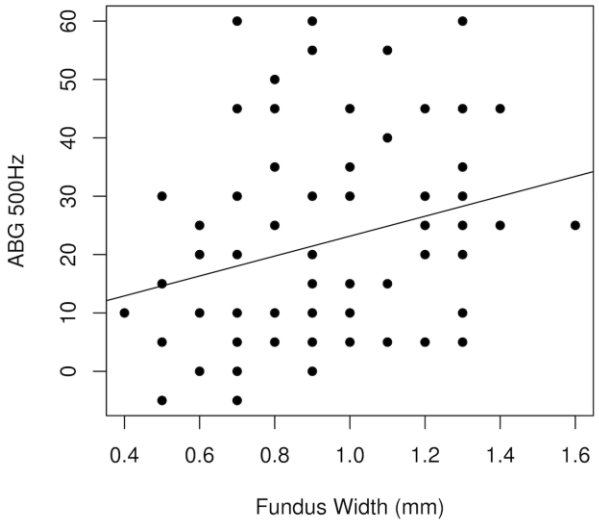
How can we screen for EVA?

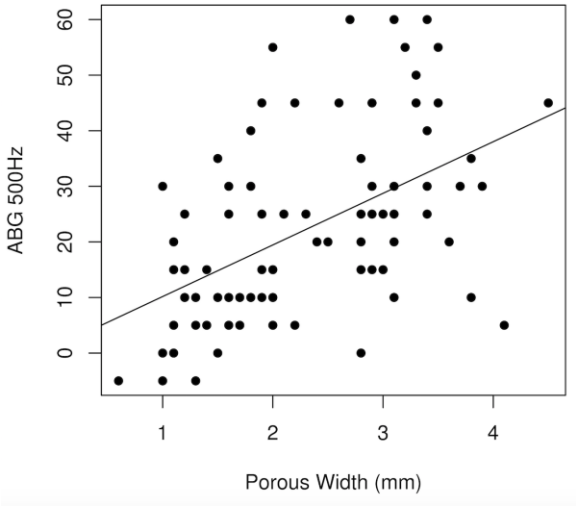
- Newborn Hearing Screening
  - Due to progressive nature, may pass and exhibit hearing loss later



- Audiometry
  - Degree of hearing loss quite variable (normal to profound PTAs)
  - Configuration: usually sloping with air-bone gaps in low frequencies
  - Requires an ABR or the patient to provide masked bone responses

Measurement of EVA





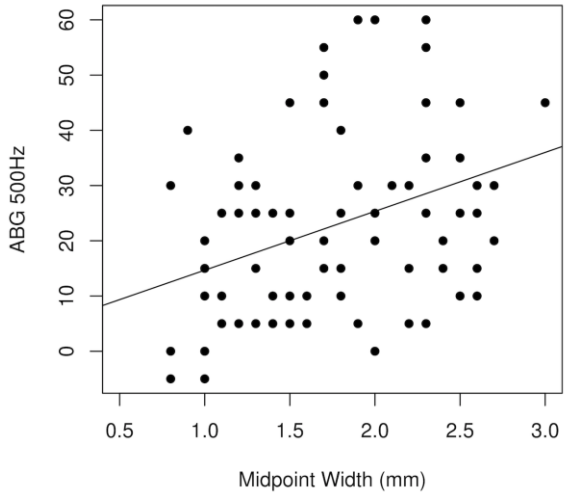


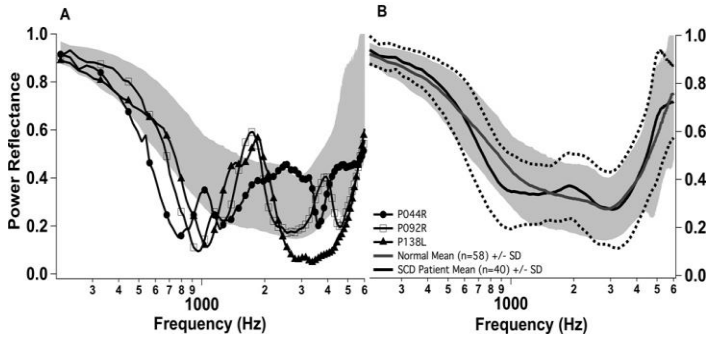
Table 3: Association of VA measurements with hearing outcomes, N=110

	Hearing Outcome		
	ABG 500 Hz	ABG 1000 Hz	Average ABG
VA Measurement	*Correlation Coefficient* *P-value*		
Fundus width	0.28 p=0.010	0.14 p=0.21	0.24 p=0.025
Midpoint width	0.38 p<0.001	0.27 p=0.017	0.33 p=0.002
Porous width	0.50 p<0.001	0.39 p<0.001	0.47 p<0.001

McCaslin et al., in Review

Why Wideband Acoustic Immittance?

- Sato et al., 2012 - Multifrequency Tympanometry
  - Type A tymps in patients with EVA despite air-bone gaps
  - Resonant frequency significantly lower compared to normative data
- Merchant et al., 2014 - Wideband Reflectance in patients with SCD
  - Screening tool for SCD using notch detection algorithm in WR
  - Sensitivities of 80% to 93% specificities of 69% to 72%



Merchant et al., 2014

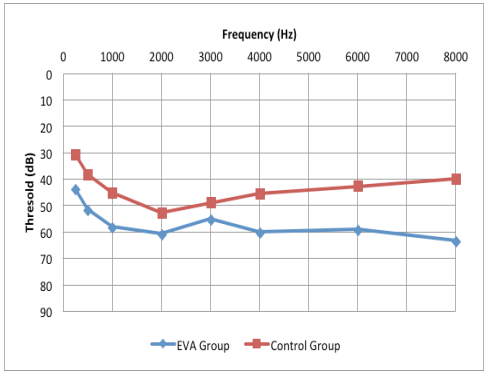
EVA and Dizziness

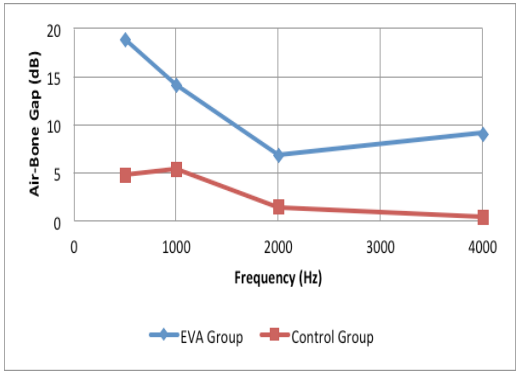
P	Age	Side of EVA	Size of EVA (mm)	DHI-P	Hearing	vHIT	oVEMP	cVEMP	Vibration	Caloric	Rotary Chair	Power Reflectance Peak
1	12	Right	R: 7.1	52	R: ✗ L: ✓	R: ✓ L: ✓	R: ✗ L: ✓	R: ✗ L: ✓	R: ✗ L: ✓	R: ✗ L: ✓ 72% UW	✓	R: ✗ L: ✓
2	8	Bilateral	R: 4.0 L: 3.2	34	R: ✗ L: ✗	R: ✓ L: ✓	R: ✓ L: ✓	R: ✓ L: ✓	R: ✓ L: ✓	R: ✓ L: ✓	✓	R: ✗ L: ✗
3	8	Left	L: 4.0	0	R: ✓ L: ✗	R: ✓ L: ✓	R: ✓ L: ✓	R: ✓ L: ✓	R: ✓ L: ✓	R: ✓ L: ✗ 30%UW	✓	R: ✓ L: ✗
4	6	Bilateral	N/A	0	R: ✗ L: ✗	R: ✗ L: ✗ Anterior canals only	R: ✗ L: ✓	R: ✓ L: ✓	R: ✓ L: ✓	R: ✓ L: ✓	✓	R: ✓ L: ✓
5	9	Left	N/A	6	R: ✓ L: ✗	R: ✓ L: ✓	R: ✗ L: ✗	R: ✓ L: ✓	N/A	R: ✓ L: ✓	✓	R: ✗ L: ✗

Stone & McCaslin, *ENT and Audiology News*, 2015

Subjects

- 14 subjects diagnosed with EVA
  - 23 EVA ears, 4 “normal” ears, 1 “possible EVA” ear
  - Average age: 10.1 (6-16)
- 12 SNHL controls
  - 22 ears
  - Average age: 8.5 (4-14)
  - Hearing loss not expected to be confounding factor





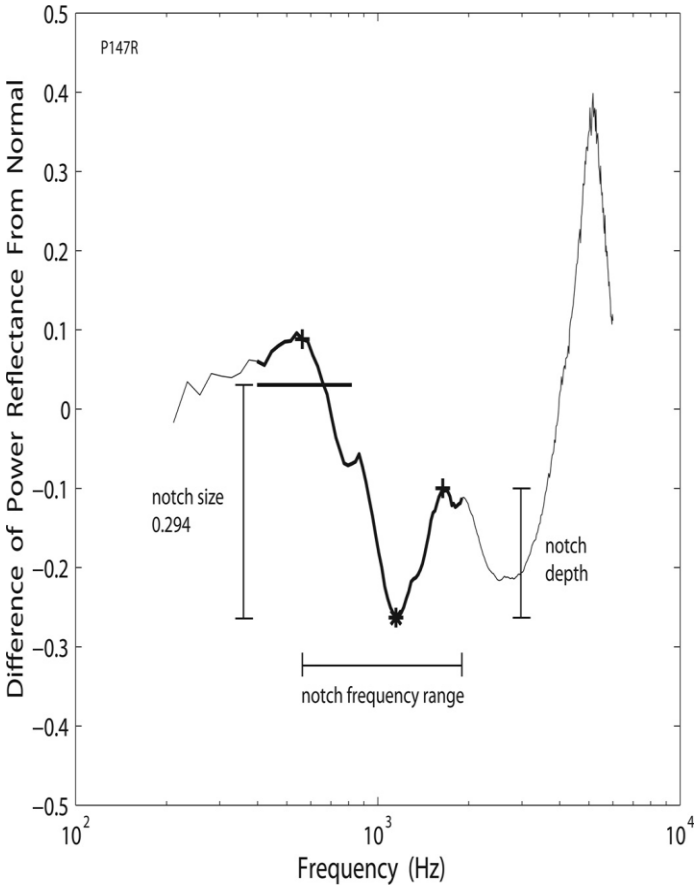
Notch Detection Algorithm

From Merchant et al., 2015

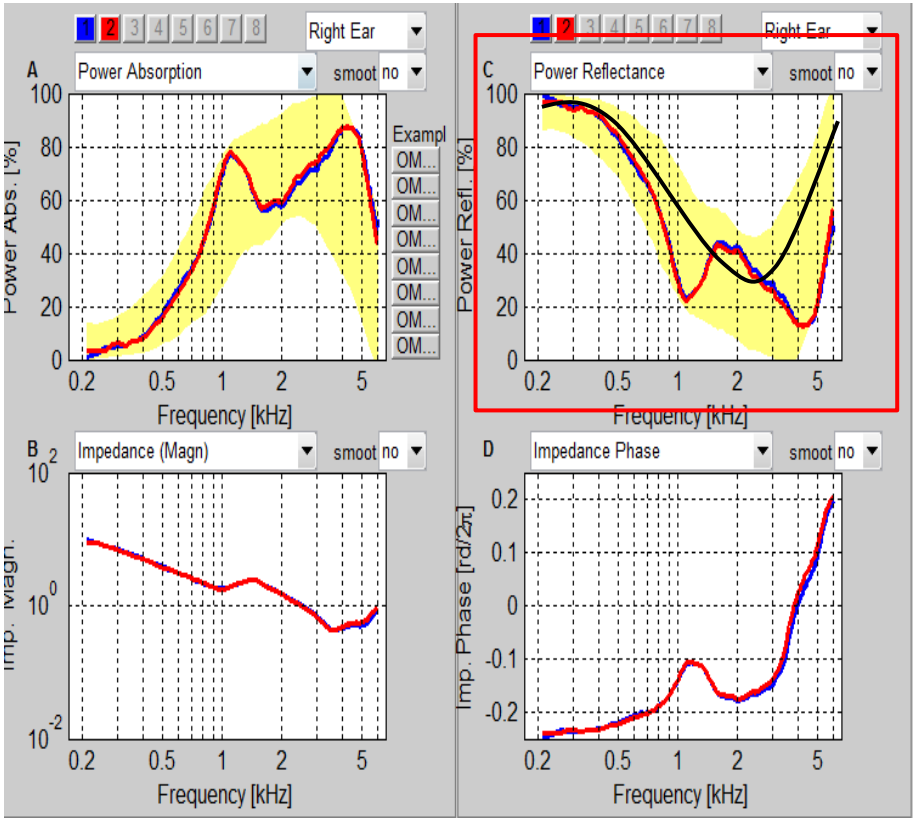
Parameters include:

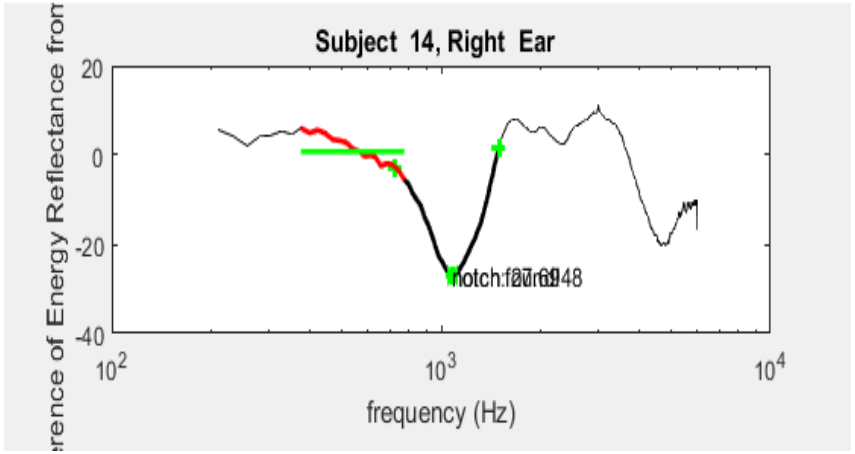
- Notch size
- Notch frequency range
- Notch depth
- Baseline frequency range

Normalized with WAI data from Rosowski et al., 2012



Notch Detection Algorithm





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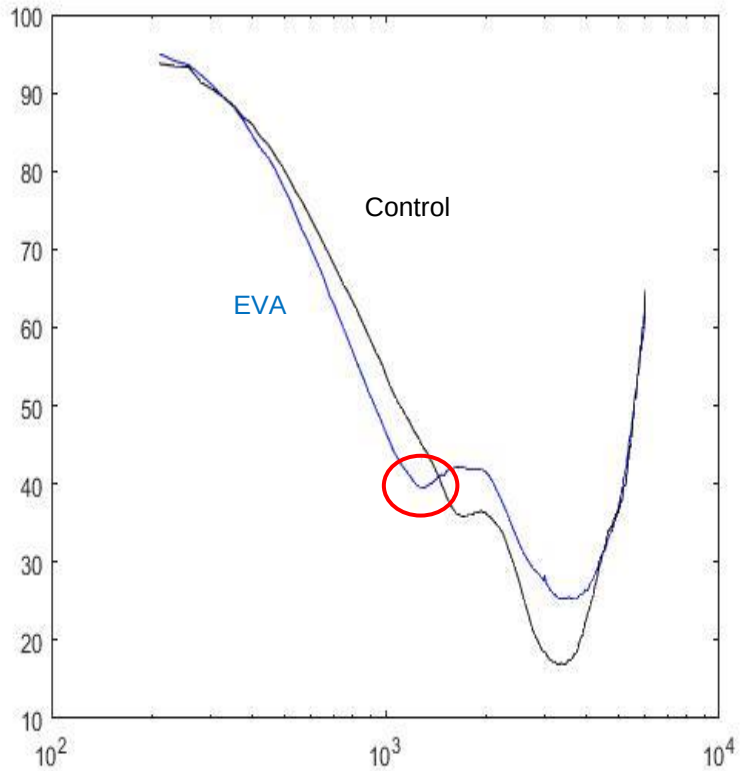
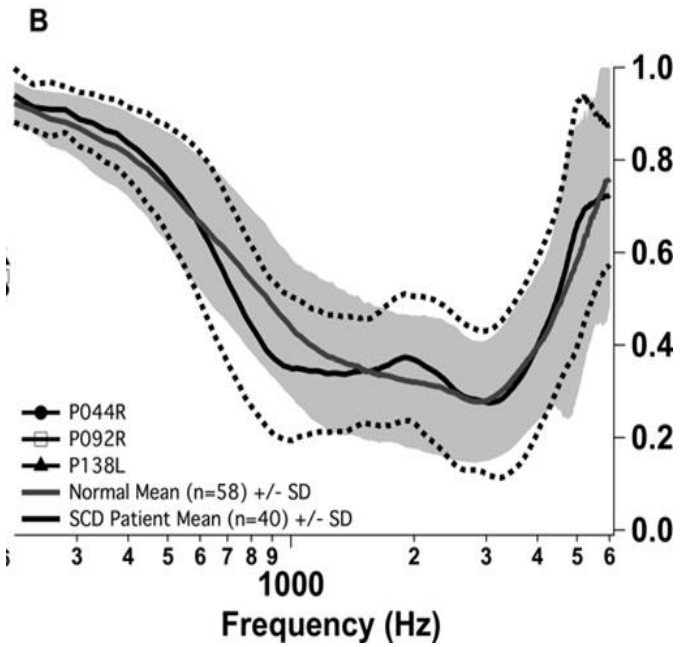
Results: Wideband Acoustic Immittance





Frequency range: **750-1500 Hz**

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	Parameters from Merchant	Optimized Parameters	Merchant et. al, 2014
Sensitivity	73%	**73%**	92.5%
Specificity	20%	**44%**	69%

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Results: Dizziness Handicap Inventory

	EVA	Control
Average Score	18.00 *	4.80
Mild Handicap	0/13	1/10
Moderate Handicap	2/13	0/10
Severe Handicap	2/13	0/10

* $p < .05$

Results: Dizziness Symptom Inventory

- 6/12 subjects surveyed: “My child has had at least one **head injury** that required a visit to the doctor.”
- 6/12 subjects surveyed: “My child has complained of **headaches** (head pain).”
- 6/12 subjects reported symptoms associated with dizziness
  - 5/12 subjects: “My child is clumsy all the time.”
  - 5/12 subjects: “My child’s dizziness come in **spells or attacks** (it is not constant).”
  - 5/12 subjects: “My child has had **repeated clear spells** of dizziness.”
  - 4/12 subjects: “My child is absolutely **fine after the dizzy spell.**”
  - 3/12 subjects: “My child’s dizziness attacks last **seconds to minutes.**”

Discussion

- EVA subjects reported symptoms of dizziness in higher incidences on average compared to control group
  - Guides how we counsel these patients
- Large group of subjects and parents chose to participate to learn more about EVA
  - Important to counsel patients, especially given progressive nature of hearing loss, possible presentation of vestibular symptoms, and possible associated hearing loss due to head trauma

Counseling is KEY!!!

A PARENT'S GUIDE TO **THE DIZZY CHILD**

SIGNS AND SYMPTOMS

These are some of the signs you might notice if your child has a balance disorder:

- headache or migraine
- dizziness or spinning sensation
- trouble seeing when moving his or her head
- motion sensitivity and/or sickness
- difficulty playing sports
- clumsiness

WHAT NEXT

Your first step is to talk with your child's primary care physician or pediatrician about your concerns. If a balance disorder is suspected, your child can be referred to an audiologist for assessment.

TREATMENT OPTIONS

Depending on the results of your tests, your doctor may treat your child medically or refer him or her for a special type of physical therapy targeted at treating balance disorders of the inner ear.

Medical Treatment
It is important that you follow up with a physician who specializes in treating balance disorders of the inner ear. He or she may be able to recommend treatments involving special medications to help with your child's inner ear problems.

Physical Therapy
Vestibular rehabilitation is a specialized area of physical therapy that uses specific exercises and balance training to improve symptoms of dizziness and imbalance caused by problems of the inner ear. The physical therapist can evaluate and provide individualized treatment interventions specific to the diagnosis, needs, and goals of you and your child.

CONTACT

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Back to the case…

Vestibular Assessments

Table 17.1 Caloric and rotational chair data

Patient age	Right warm	Left warm	Right cool	Left cool	Asymmetry (%)	Total SPV	SHA
9 y 3 mo	6	27	6	36	68	75	Normal
10 y 11 mo	4	27	4	31	76	66	Low gain, 0.01–0.04 Hz
12 y 4 mo	1	7	4	10	55	22	Phase leads with low gain, 0.01–0.04 Hz
12 y 9 mo	1	17	4	14	72	36	Normal

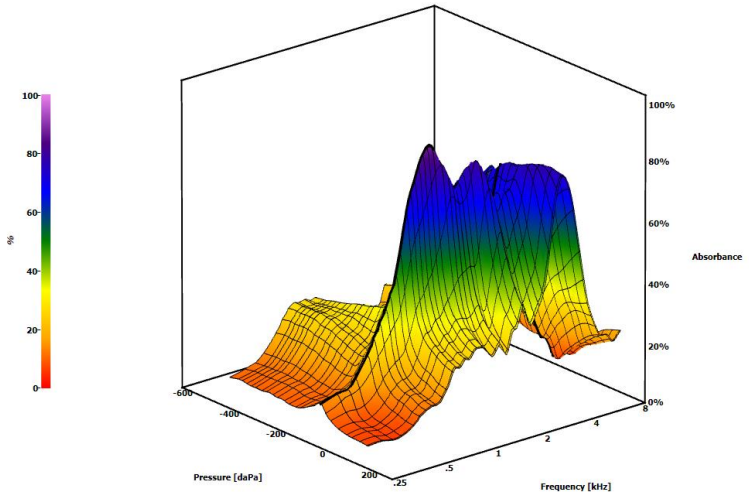
Abbreviation: SHA, sinusoidal harmonic acceleration; SPV, slow phase velocity.

Table 17.2 VEMP data

Patient age	cVEMP P1 latency		cVEMP amplitude		oVEMP N1 latency		oVEMP amplitude	
	Right	Left	Right	Left	Right	Left	Right	Left
9 y 3 mo	NR	13.58	NR	471.19	NR	11.19	NR	5.18
10 y 11 mo	NR	14.55	NR	63.97	NR	13.26	NR	2.94
12 y 4 mo	NR	15.72	NR	37.60	NR	12.18	NR	7.01
12 y 9 mo	NR	NR	NR	NR	NR	NR	NR	NR

Abbreviations: cVEMP, cervical vestibular-evoked myogenic potential; NR, no response; oVEMP, ocular vestibular-evoked myogenic potential.

Wideband and pDHI



PEDIATRIC DIZZINESS HANDICAP INVENTORY (DHI)
(Age 5-12)

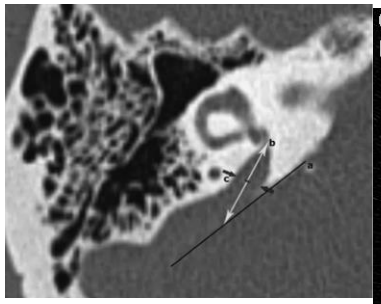
Instructions: The purpose of this questionnaire is to identify difficulties that your child may be experiencing because of his or her dizziness or unsteadiness. Please answer "yes", "no", or "sometimes" to each question.
Answer each question as it pertains to your child's dizziness problem only.

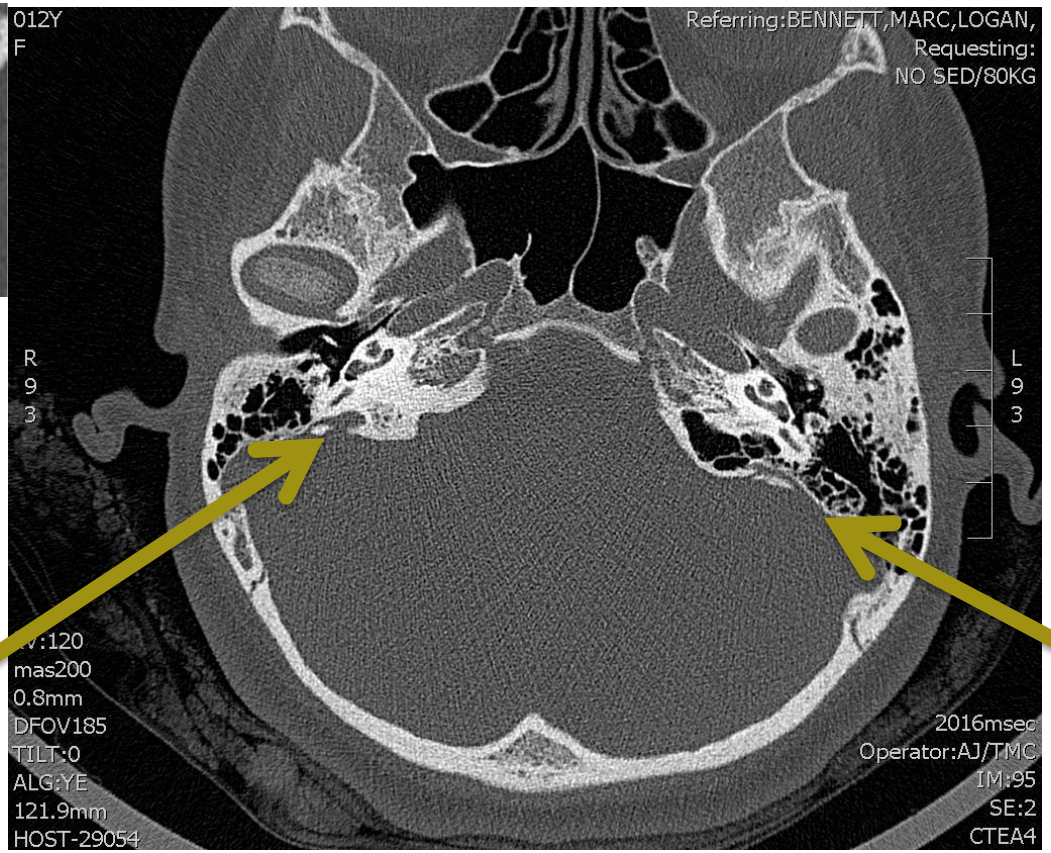
	Yes (4)	Sometimes (2)	No (0)
1. Because of your child's problem, is it difficult for him/her to walk unassisted?		X	
2. Does your child use a great deal of effort to keep his/her balance?		X	
3. Does your child's problem make him/her feel tired?			X
4. Is your child's life ruled by his/her problem?			X
5. Does your child's problem make it difficult for him/her to play?		X	
6. Because of his/her problem, does your child feel frustrated?		X	
7. Because of his/her problem, has your child been embarrassed in front of others?			X
8. Because of his/her problem, is it difficult for your child to concentrate?		X	
9. Because of his/her problem, is your child tense?		X	
10. Do other people seem irritated with your child's problem?			X
11. Do you find other people do not understand your child's problem?	X		
12. Is your child's balance unpredictable?	X		
13. Because of his/her problem, does your child worry?		X	
14. Because of his/her problem, does your child feel angry?			X
15. Because of his/her problem, does your child feel "down"?			X
16. Because of his/her problem, does your child feel unhappy?		X	
17. Because of his/her problem, does your child feel different from other children?			X
18. Does your child's problem significantly restrict his/her participation in social or educational activities, such as going to dinner, meeting with friends, field trips, or to parties?		X	
19. Because of your child's problem, is it difficult for him/her to walk around the house in the dark?		X	
20. Because of his/her problem, does your child have difficulty walking up stairs?			X
21. Because of his/her problem, does your child have difficulty walking one or two blocks?			X
22. Because of his/her problem, does your child have difficulty riding a bike or scooter?			X
23. Because of his/her problem, does your child have difficulty reading or doing schoolwork?		X	
24. Does your child's problem make it difficult to successfully do activities that others his/her age can do?		X	
25. Because of his/her problem, does your child have trouble concentrating at school?		X	
Version 2		TOTAL SCORE	

CT result again 6/12/14

- CT Results
  - The right vestibular aqueduct is enlarged, measuring 2.5 mm in diameter. Left side:
  - The left Vestibular aqueduct is upper limits of normal on the left. It measures 1 mm in maximal diameter.
- Impression: Enlarged right vestibular aqueduct (2.5 mm in diameter). Left vestibular aqueduct, within upper limits of normal.

CT





Enlarged Vestibular Aqueduct

Normal Vestibular Aqueduct

Vestibular Rehabilitation Eval

- Head impulse – abnormal right (UW)
- Patients gait speed was at above or expected norms.
- Function gait assessment – normal
- Posturography - deficit in the use of vestibular cues
- Motor Control test – normal
- Given home exercises
- Discharged 1/28/2015

Conclusions

- BPPV- Right Side (rare) confused with BPV?
- Unilateral vestibular loss involving the otolith organs and lateral SCC
- Hearing loss is stable (for now)
- WBT was abnormal (peak at 1000 Hz)
- Is this due to the head injury?
- Awareness and counseling is key!!

Thank You!

