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Semicircular Canal Dehiscence: Lessons for Collaboration, in partnership with the American Academy of Audiology

Rebekah F. Cunningham, PhD

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Rebekah F. Cunningham, PhD



Rebekah F. Cunningham, PhD, is a clinical applications specialist for Natus Medical – Sensory, where she provides education and training on newborn hearing screening as well as hearing and balance assessment. She is an adjunct Associate professor at A.T. Still University, teaching both online and on campus. Her professional experience includes academia, hospital/medical settings, primary and secondary schools, and neurotology offices. Her passion has always been pediatric-focused, teaching and mentoring audiology students, providing services, and programs responsible for ensuring timely and appropriate evaluations and follow-up for children who are deaf and hard of hearing. Dr. Cunningham has created and taught courses on Auditory Evoked Potentials, Otoacoustic Emissions, Pediatric Audiology (including Early Hearing Detection and Intervention and Educational Audiology), Cochlear Implants, and Auditory Processing Disorders. She has served for more than 25 years on committees and task forces for local and national audiology organizations and is an Associate Editor of Audiology Today (AT) and sits on the Editorial Board for The Journal of Early Hearing Detection and Intervention (JEHDI). Dr. Cunningham has presented and published on the topics of assessment and management of children who are deaf and hard of hearing or experience auditory processing difficulties.



Devin L. McCaslin, PhD

Devin L. McCaslin, PhD currently serves as the Clinic and Academic Program Director of Audiology at Michigan Medicine in Ann Arbor and holds the rank of Professor in the Department of Otolaryngology-Head and Neck Surgery. Dr. McCaslin's major academic, clinical and research interests relate to clinical electrophysiology, vestibular assessment, healthcare economics, and the application of artificial intelligence to manage and treat dizzy patients. He also serves as the Deputy Editor-in-Chief of the Journal of the American Academy of Audiology and is a past member of the Board of Directors for the American Auditory Society and American Academy of Audiology. He is also a Past President of the American Balance Society.



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- 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:

- Discuss the incidence of superior semicircular canal dehiscence in children.
- Describe subtle symptoms of SSCD in children and how they vary from those seen in adults.
- Explain the need for inter professional collaboration in their clinical or school-based patient population.



Agenda

- What is SSCD
 - Symptoms, demographics
- Case study
- Treatment
- Reflections

All about you!



Poll

Which best describes you?

- See children (0-18) regularly (3-5 days/week)
- I do not see children clinically
- See children (0-18) sometimes (2 or less days/week)
- Other

Poll

Do you do vestibular testing for children?

- Yes, at least once a week
- Yes, less than once a week
- No
- Other

Superior Semicircular Canal Dehiscence Syndrome

For the purposes of this presentation, we will refer to this syndrome as either SSCD or SCDS



Lloyd D. Minor, M.D.

History

- 1998: Minor describes SCDS
 - 8 patients with sound +/- pressure induced vertigo
 - Eye movements align with superior canal
 - Common history of fistula repair or stapedectomy
 - Mobile “third window” theory

Case Reports > Arch Otolaryngol Head Neck Surg. 1998 Mar;124(3):249-58.

doi: 10.1001/archotol.124.3.249.

Sound- and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal

L B Minor ¹, D Solomon, J S Zinreich, D S Zee



Definition

- Rare inner ear disorder (but is it?)
 - Thinning or dehiscence of the bone in the otic capsule over the semicircular canal is more prevalent in children younger than 12 months and with age, the semicircular canals may develop more bony covering
 - The dehiscence may be part of normal development under the age of 5 years
- Abnormal thinning of the bone over the superior semicircular canal
- Results in mobile third window
 - Auditory symptoms
 - Vestibular symptoms



Symptoms

Hearing

- Autophony
- Hearing loss
- Ear pressure/fullness
- Sensitivity to loud sounds

Vestibular

- Sound-induced
- Pressure-induced
- Dizziness
- Imbalance
- Oscillopsia



Pathophysiology

Obesity



Increased
intrathoracic
pressure



Decrease
venous return



Increased
intracranial
pressure



Pathophysiology

- When the bony capsule overlying a semicircular canal is disrupted, one of the major protective mechanisms that prevent sound energy from stimulating the canal is lost.
- Tullio (1929) demonstrated this in his seminal experiments with sound as a stimulus for the labyrinth in pigeons after he fenestrated their semicircular canals.

Incidence of SSCD

- 0.5% incidence complete dehiscence
- 1.4% incidence bony thickness <0.1 mm
- Average thickness over canal .96 mm
- Median age ≈ 40
- Women = Men
- Right $>$ Left



Poll

Are you familiar with the signs/symptoms of SSCD?

- Yes
- No
- Yes, but maybe not in children (?)



SSCD Symptoms

- When I cough, I get dizzy
- I get dizzy when I sneeze
- Loud sounds make me dizzy
- I get dizzy when I have a bowel movement
- Loud sounds make my vision blurry
- I get dizzy when I strain to lift something
- I become dizzy when I am in large crowds

What are the symptoms?

- Vestibular signs: 95%
 - Sound-evoked eye movement: 82%
 - Valsalva induced eye movements: 75%
 - Tragal pressure induced eye movement: 45%
 - Sound induced head movement: 20%



SSCD Protocol

CANAL DEHISCENCE REPORT SHEET

DATE:

MR:

CT completed:

Condition	How long stimulate	How long record	Result:
Valsalva holding nose	2-3s	10-15s	+ / -
250Hz pulsed @ 100dB	10s	10-15s	+ / -
500Hz pulsed @ 100dB	10s	10-15s	+ / -
1000Hz pulsed @ 100dB	10s	10-15s	+ / -
Positive pressure in canal	Second half of tympanometry	Duration of tympanometry	+ / -

*all conditions listed above should show torsional nystagmus with fast phase TOWARD affected ear.

Condition	How long stimulate	How long record	Result:
Negative pressure in canal	First half of tympanometry	Duration of tympanometry	+ / -
Valsalva "bearing down"	2-3s	10-15s	+ / -
Hyperventilation	3-5s	10-15s	+ / -

*all conditions listed above should show torsional nystagmus with fast phase AWAY FROM affected ear.

Condition	VEMP latency (ms)	VEMP amplitude (uV)	cVEMP threshold level or oVEMP present @ 4000Hz
Left cVEMP			
Right cVEMP			
Left oVEMP			
Right oVEMP			



SSCD in Children

Pediatric Semicircular Canal Dehiscence: Radiographic and Histologic Prevalence, With Clinical Correlation

*Duncan A. Meiklejohn, †C. Eduardo Corrales, *Brian M. Boldt,
‡Jeffrey D. Sharon, *§Kristen W. Yeom, ‡John P. Carey, and *Nikolas H. Blevins

Semicircular canal dehiscence in the pediatric population[☆]

Eunice Y. Chen^a, Angelisa Paladin^b, Grace Phillips^b, Molly Raske^b,
Liah Vega^c, Do Peterson^d, Kathleen C.Y. Sie^{a,*}

Incidence of SSCD in children

- Rare in the pediatric population
- Limited research
- Disequilibrium symptoms and typical third window symptoms were absent or difficult to elicit in children
- Only three (0.5%) fulfilled the adult criteria of a superior semicircular canal dehiscence syndrome

Incidence of SSCD in children

- The use of VEMPS in semicircular canal dehiscence in children has limited evidence.
- Meiklejohn et al. ([13](#)) who in their series including 19 children from the age of 2 months until 7 years observed a 0% prevalence of the condition at the age beyond 3 years, which is the starting age range of the study. The majority of this case series presented with hearing loss and included variable comorbidities and cochleo-vestibular dysplasia as well.
- Only 30% in our series described classical third window symptoms that are crucial for a diagnosis in an adult but seldom available from children. These include gaze evoked tinnitus or audible eye movements, conductive dysacusis, autophony, Hennebert's phenomenon or Tullio's phenomenon and pulsatile tinnitus.



Incidence of SSCD in children

- Rare in the pediatric population
- Limited research
- VNG examination with and without optic fixation, rotatory chair tests and vestibulo-spinal tests as well as mastoid vibration tests were normal in the majority of children in the current case series (76.9%) suggesting that gravitational sensor and static, low and mid frequency angular motion sensor vestibular function tend to be preserved in semicircular canal dehiscence in children.
- It appears that saccades may be important to assess high frequency vestibular function in semicircular canal dehiscence in children.

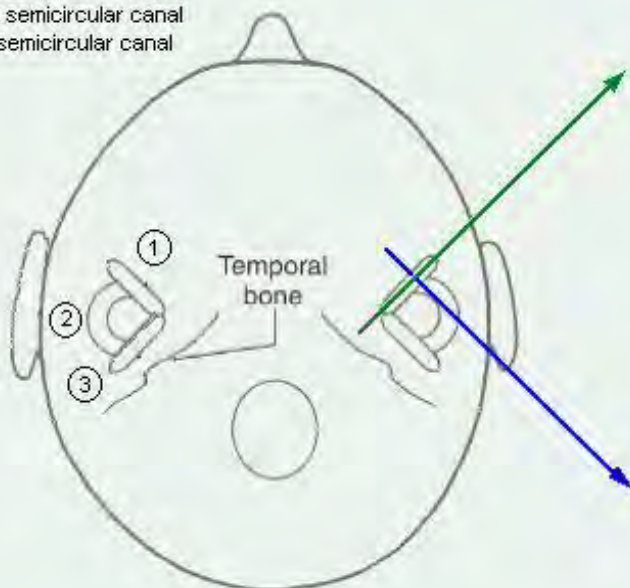
What are the symptoms in children?

Child	HS	BS	TWS	Tymp	SRT	OAE	VNG/VFT	VOR LLSC	VOR RLSC	VOR LASC	VOR RASC	VOR LPSC	VOR RPSC	AC/BC R	AC/BC L	Type HL	Saccade	Diagnosis
1	Yes	Yes	Nil	Norm	Norm	Abn	Norm	1.16	1.09	0.55	0.75	0.85	0.69	55/46	58/46	Mixed	Yes	R PSCD
2	Nil	Yes	GET	Norm	Norm	Norm	Norm	0.67	0.48	0.62			1.46	7/-5	4/3	Nil	No	L SSCD R PSCD
3	Yes	Nil	Nil	Norm	Norm	Norm	Norm	0.96	0.96	0.41	0.85	0.82	0.51	27/7	10/na	CHL	Yes	R SSCD
4	Yes	Nil	Nil	Flat	Reduced	Abn	Norm	1.1	0.88					100/nt	17/4	SNHL	No	R SSCD
5	Yes	Yes	Nil	Norm	Norm	Norm	Abn	1.14	1.17	1.13	1.08	0.90	1.1	45/9	45/10	CHL	Yes	RSSCD
6	Nil	Nil	Nil	Norm	Norm	Norm	Norm	0.79	0.94	0.67	0.75	1.07	0.78	42/2.5	23/10	CHL	Yes	Bilateral SSCD
7	Nil	Nil	CD	Norm	Norm	Norm	Norm	0.94	0.94	0.79	0.92	0.99	0.83	9/6	7/6	Nil	Yes	Bilateral SSCD
8	Yes	Nil	Nil	Norm	Norm	Norm	Norm	0.86	0.83	0.50	0.89	0.82	0.39	25/14	45/20	CHL	Yes	R SSCD
9	Yes	Yes	Nil	Norm	Norm	Norm	Abn	0.86	0.90	0.73	0.48	0.54	0.47	36/30	41/26	Mixed	Yes	R SSCD R PSCD
10	Yes	Nil	Nil	Norm	Norm	Norm	Abn	0.82	0.90	0.73	0.88	0.75	0.73	17/15	21/15	Mixed	Yes	L SSCD
11	Nil	Yes	Auto/ tinnitus	Norm	Norm	Norm	Norm	0.95	0.90	0.41	0.37	0.66	0.59	12/7	17/9	Nil	Yes	R SSCD
12	Yes	Yes	CD	Norm	Norm	Norm	Norm	1.07	1.01	0.07	0.07	1.06	1.03	44/35	39/36	Mixed	Yes	Bilateral SSCD
13	Nil	Nil	Nil	Norm	Norm	Norm	Norm	1.18	0.99	0.65			0.98	4/10	55/51	SNHL	No	R SSCD

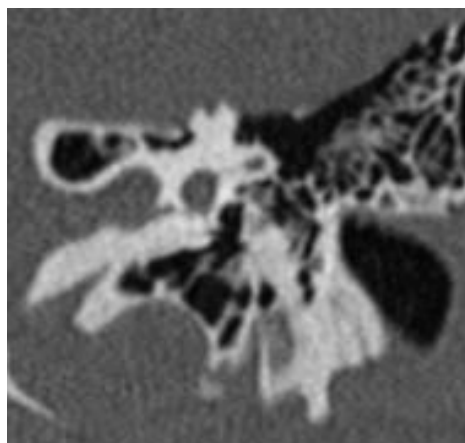
Children in case group; Legends: HS, hearing symptoms; BS, balance symptoms; TWS, Third window symptoms; Tymp, tympanometry in dehiscent ear/s; SRT, stapedial reflex test in dehiscent ear/s; OAE, transient otoacoustic emission in dehiscent ear/s; VNG/VFT, videonystagmography/other vestibular function tests; VOR, vestibulo-ocular reflex gain; LL/RL/LA/RA/LP/RP SC, left lateral, right lateral, left anterior, right anterior, left posterior, right posterior semi-circular canals; PTA AC R and L, pure tone audiometry air conduction thresholds averaged 500 Hz–4 kHz right and left; BC R and L, pure tone audiometry bone conduction thresholds averaged 500 Hz–4 kHz right and left; na, not performed; nt, no detectable threshold; HL, hearing loss; CHL, conductive hearing loss; SNHL, sensorineural hearing loss; GET, gaze evoked tinnitus; CD, conductive dysacusis; Auto, autophony; Tinn, tinnitus; Norm, normal; Abn, abnormal.

Imaging

- 1) Superior semicircular canal
- 2) Horizontal semicircular canal
- 3) Posterior semicircular canal



Pöschl - parallel



Stenver - perpendicular



This is Piper

- Now 11 (almost 12!)
- Funny, smart, kind!
- Plays flute
- Strong academically
- Anxious, takes med
- Celiac disease



Identification

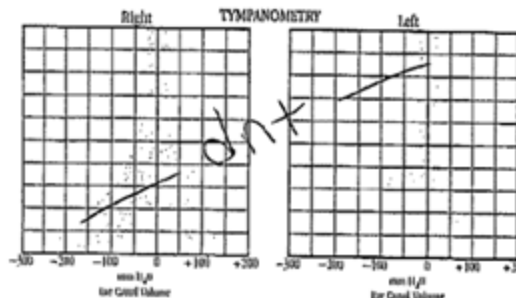
- Passed UNHS, AU
- Identified at age 6 (09/25/2018)
- Placed on steroids
 - 1 week after, hearing worse
 - 3 weeks after, back to baseline
- Recs: recheck 6-12 months, HAF outside facility



	Unaided		Masked		Reliability	Transducer
	Air	Bone	Air	Bone		
R	☐	☐	☐	☐	☒ Good	☐ Headphones
L	☒	☐	☐	☐	☐ Fair	☒ Insert phones
	☐	☐	☐	☐	☐ Poor	☐ Sound field

SPEECH AUDIOMETRY					
	Monosyllabic	Phonetic	Word	Picture	Visual
R	35	70	100	40 (m)	%
L	40	100	100	100	%
SD	dB	dB	dB	dB	dB

CONTRA			IPSI		
Hz	dB	dB	dB	dB	dB
250					
500					
1000					
2000					
4000					



Chief complaint: flw @ hearing loss

Impression: R: moderate rising to slight
Schwannoma hearing loss
L: normal hearing

- Recommendations:
1. Flw w/ ENT
 2. Recheck routinely to monitor hearing loss (in 6-12 mo)
 3. HAF w/ outside facility
 4. Visual accommodation (i.e., preferential seating, FM system)



Poll

After the identification of hearing loss in an infant or young child, which of the following would you like to see done?

- Referral to Pediatric ENT
- Imaging
- Bloodwork
- Genetics
- All of these
- Other



Immediately after ID 2018

- Mother reported an MRI was ordered to rule out a tumor
 - Came back clear
- No bloodwork
- Mother asked about genetics but was told no, it would not change what they do
- Referred to outside facility to be fit with HA



HAF

- Phonak Sky
- Roger TS mic in class/specials/therapy
 - Piper is increasingly uncomfortable using the TS Mic – peers
- https://youtu.be/7SOtz9FETY4?si=DR6zgasJTjPk_SEr:
1:56-2:59



Hearing History: Audiograms

- Slight decrease overall (5/7/2019) (compared to baseline 8 months prior)
- No significant changes (7/16/2020)
- APD Evaluation – AP skills WNL for age but deficits in auditory memory (2/22/2021)
 - Thoughts on APD with children with SNHL?
- Slightly improved hearing (6/17/2021)
- Stable 10/13/2022
- Hearing loss has always been sensory

Multidisciplinary evaluation

07/13/2021

- She utilizes spoken English at the conversational level that is sometimes difficult to understand due to low vocal volume, cessation of voice at the end of sentences, and/or vocal fry at the end of sentences.
- She presents with language skills within expectations but demonstrates below average understanding of multiple meanings and use of complex grammar structures within her narratives.
- Shared concerns regarding Piper's increased anxiety across environments is apparent in the current assessment; rating scales completed by Piper and her mother suggest elevated ratings for anxiety and panic symptoms, including feelings of being tense or restless



Multidisciplinary evaluation

07/13/2021

- Elevated scores indicate more concerns than are typical for learning problems and inattention when compared to her same age peers
- Less developed executive functioning skills; Piper exhibits initiation difficulties and may struggle with some timed tasks; she will benefit from extra cues to begin tasks as well as maintain attention to task
- Study strategies are inadequately developed for her age as she has difficulties using a variety of resources when a concept is not understood and using strategies for memory and encoding

Multidisciplinary evaluation

07/13/2021

Areas of need:

- Discussion of mental health supports related to her feelings of panic and anxiety
- Increased understanding of her fluctuating hearing loss and self-advocacy skills



IEP

- First created for 2019-2020 school year – still in effect
- Consult with CDHHE's team for DHH recommendations and accommodations
- **DHH, OHI, SI**



Elementary contracts with ...

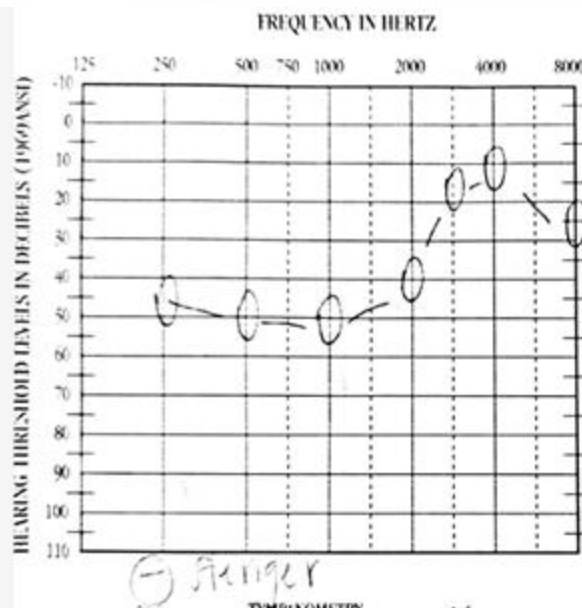
- Start seeing Piper Fall '22
- First complaint from her regarding dizzy with loud sounds in late February 2023 – I recorded it in our database 3/3/23
- She tried turning the HA off but still felt dizzy
- Tried earplug(s)
- Could not eat in cafeteria any longer
- Told nurse and teacher
- <https://youtu.be/JVAqaCO1x1w?si=HYOVxHLqiQ85-7B0>: 00:00-1:28



After reporting dizziness with loud sound

- I reached out to parent
- Piper was taken to her ENT office
- Audio revealed a significant decrease
- Mother requested
 - Genetics
 - Referral for balance testing
- How long?
- <https://www.youtube.com/watch?v=7SOtz9FETY4>: 1:13-1:58

03/08/2023
and
03/14/2023



	Unmasked		Masked		Reliability	Transducer
	Air	Bone	Air	Bone		
R	○	<	△	□	☒ Good ☐ Fair ☐ Poor	☐ Headphone ☒ Insert phones ☐ Sound field
L	×	>	□	]		

SPEECH AUDIOMETRY

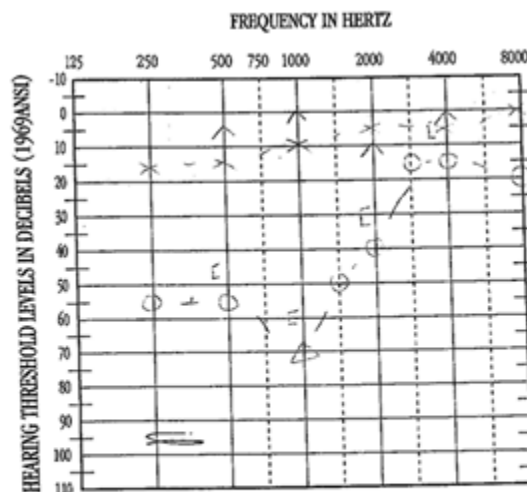
	SRT	Discrimination					MCL dB	UCL dB
		dB	%	%	%	%		
R	30	40	100	100	100	100		
L	40	100	100	100	100	100		
SF	40	100	100	100	100	100		

Chief complaint: updated audio

- ☒ Hearing difficulty
☐ Tinnitus
☐ Pressure fullness
☐ Ear infections
☐ Surgery follow up
☐ Head trauma
☐ Ear pain
☒ Vertigo

Impressions: immediate to NNL

INFL



	Unmasked		Masked		Reliability	Transducer
	Air	Bone	Air	Bone		
R	○	<	△	□	☒ Good ☐ Fair ☐ Poor	☐ Headphone ☒ Insert phones ☐ Sound field
L	×	>	□	]		

SPEECH AUDIOMETRY

	SRT	Discrimination					MCL dB	UCL dB
		dB	%	%	%	%		
R	55	75	100	100	100	100		
L	10	80	100	100	100	100		
SF	40	100	100	100	100	100		

Chief complaint: my hearing is getting worse

- ☒ Hearing difficulty
☐ Tinnitus
☐ Pressure fullness
☐ Ear infections
☐ Surgery follow up
☐ Head trauma
☐ Ear pain
☐ Vertigo

Impressions: immediate to NNL

INFL

INFL - SNHL

Structural SNHL 10/13/22



Balance consult/testing

3/23-24/2023

Reason for Appointment

1. Dizziness

History of Present Illness

Dizziness:

Piper reports she has dizziness in the cafeteria with loud noise "like spinning really fast". It resolves quickly after leaving the cafeteria and she has nausea only when the loud noise is present. No problems with balance. She is wanting to play the flute in band. She has no history of headaches. No autophony, no bodily noises are heard in her right ear.

Assessments

1. Dizziness - R42 (Primary)
2. Asymmetrical hearing loss of right ear - H91.8X1

She has lost hearing as of 5 years ago with some fluctuation in the low frequency range and has been told she may have Meniere's, in spite of her age. She has some sensitivity to loud noise causing vertigo and nausea, so consider superior canal dehiscence syndrome. I will have her return for vestibular function testing and to further evaluate her symptoms, then see her back to review findings - we will be especially interested in the result of

the VEMP and regardless consider sending her for a CT IACs with Stenver and Poschl views to look for any signs of SCDS.

Problems - 1 chronic problem with worsening/not at goal addressed today - dizziness (Moderate)

Data - 1 external consult note reviewed; 1 audiogram reviewed and independently interpreted

Risk -.

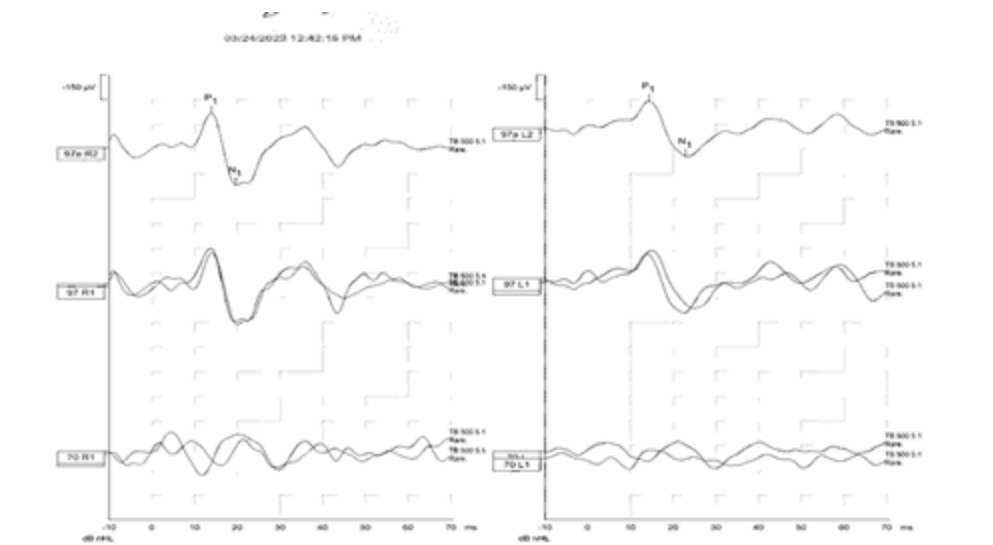
Treatment

1. Dizziness

PROCEDURE: Vestibular Function Testing

cVEMP

cVEMP was Normal. Repeatable cVEMP waveforms showed comparable and expected latencies and amplitudes, bilaterally when evaluated using a 500 Hz tone burst stimulus presented at 97dB nHL via insert earphones. No asymmetry was present after EMG scaling was employed. VEMP asymmetry ratio was 13%. No reduced threshold was seen using a 70dBnHL stimulation level, bilaterally.



Curve	Latencies (ms)					
	P1	N1	P1'	N1'	N1-P1 Lat	N1-P1 Amp
97 R						
97 R1						
97a R2	14.00	19.67			5.67	439.6
97 L						
97 L1						

(LA-SA)/(R+L)



VNG

VNG test results were normal. The VNG showed no gaze, spontaneous or positional nystagmus. The Dix-Hallpike test for benign paroxysmal positioning vertigo (BPPV) was negative for both sides. Cool caloric irrigations produced robust and symmetrical nystagmus. Caloric fixation suppression was normal. Saccadic eye velocity and accuracy were normal. Visual pursuit and optokinetic tests were normal and symmetrical.

VNG test results were normal. The VNG showed no gaze, spontaneous or positional nystagmus. The Dix-Hallpike test for benign paroxysmal positioning vertigo (BPPV) was negative for both sides. Bithermal caloric irrigations produced robust and symmetrical nystagmus. Caloric fixation suppression was normal. Saccadic eye velocity and accuracy were normal. Visual pursuit and optokinetic tests were normal and symmetrical.

VOR suppression resulted in a normal percentage of gain reduction at all frequencies tested.

F/U Visit 3/27/2023

Subjective:**Chief Complaints:**

1. Dizziness.

HPI:Dizziness:

Piper returns for follow-up of noise-induced dizziness, noting right ear hearing loss years ago. Vestibular testing was completely normal. VEMP, in particular, was normal, revealing no evidence for superior canal dehiscence syndrome. Audiogram 3/8/2023: L ear normal hearing, R low frequency moderately severe hearing loss rising to normal hearing in higher frequencies.

TODAY, she reports continued dizziness with loud noises in the cafeteria. Her parents continues to be concerned about fluctuating hearing loss.

Assessment:**Assessment:**

1. Dizziness - R42
2. Asymmetrical hearing loss of right ear - H91.8X1

She has lost hearing as of 5 years ago with some fluctuation in the low frequency range and has been told she may have Meniere's, in spite of her age. She has some sensitivity to loud noise causing vertigo and nausea, so consider superior canal dehiscence syndrome. But her vestibular evaluation was normal, including VEMP, no subthreshold response, thus no evidence for SCDS. Despite this I discussed the option of getting a CT IAC to look for SCDS, but given the negative test result and the fact that the only symptoms she has to go along with possible SCDS would be the cafeteria noise-induced dizziness, such a CT scan would likely be normal. Parents would prefer to hold on CT at this time, as the likelihood of finding any abnormality is quite small. I discussed seeing how she does playing the flute and if this causes dizziness, look into ear molds (plugs). I will refer her to Riley Audiology as they would like second opinion regarding fluctuating hearing loss.

Problems - 1 chronic problem with worsening/not at goal addressed today - dizziness (Moderate)

Data -

Risk - low



After normal balance assessment

- Mother returns to ENT
- Requests bloodwork
- Requests imaging again

Imaging 3/31/24

TECHNIQUE:

Low dose, multichannel computerized tomography was performed on the temporal bones without IV contrast. Multiplanar reformats were reviewed.

COMPARISON:

None.

FINDINGS:**RIGHT:**

EAC/ Tympanic membrane: Normal.

Middle Ear Cavity: Normal.

Ossicular chain: Normal.

Tegmen Tympani: Normal.

Mastoid Air Cells: Normal.

Vestibular and Cochlear Apparatus: There appears to be dehiscence and/or thinning of the superior right semi circular canal.

Inner Ear Structures: Normal.

IAC/CP angle: Normal.

Miscellaneous Findings: None.

LEFT:

EAC/ tympanic membrane: Normal.

Middle Ear Cavity: Normal.

Ossicular chain: Normal.

Tegmen Tympani: Normal.

Bloodwork

Connexin 26 68 KD Antigen

Results/Notes:

Copy for: [REDACTED] (Role: ORDERING)

PT: RALICH, PIPER L

MRN: [REDACTED]

D

ACCT: [REDACTED]

Accession: [REDACTED]

Location:

Collected: 03/22/2023 10:27

Received: 03/22/2023 10:29

Ordered by: [REDACTED]

*** CONNEXIN 26 (GJB2) RNA SEQUENCING TH

Procedure	Value	Flag	Units
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INTERPRETATION

This test identified one pathogenic variant in the GJB2 gene, therefore this individual is at least a carrier for nonsyndromic sensorineural hearing loss.

TECHNICAL RESULTS

Interpretive Result Table

INTERPRETIVE RESULT: Carrier

GENE/TEST: GJB2

TECHNICAL RESULT: c.35del (p.Gly12Valfs*2)

VARIANT TYPE: Heterozygous Frame Shift

INHERITANCE: Autosomal Recessive

CLINICAL RELEVANCE: Pathogenic

PUB MED ID: 34440441 29501291

FootNote

No other abnormal variants were detected.

COMMENTS

Comments: This result confirms the carrier status. In an individual with symptoms consistent with a recessive disease, possible explanations for symptomatic carrier status include, but are not limited to: (1) the existence of a second pathogenic variant that cannot be detected using this methodology or (2) the clinical symptoms, while consistent with the disease, are actually caused by other factors.



Connexin 26

- The GJB2 variants tend to be associated with **bilateral** hearing loss
 - Denoyelle et al (1999) 89% of children studied, severity between ears did not differ between ears
 - Orzan et al (2015) more than 90% of the 46 children with GJB2 variants demonstrated a symmetrical hearing loss (inter-ear difference of <15 dB at two frequencies or 10 dB at four frequencies).
 - However, "population-based data on all individuals with any type of hearing loss are needed to clarify the issue" (Kenneson et al, 2002)



Consult with CCHMC

- Mother shares with me bloodwork and imaging results
- Requests help in finding another ENT (neurotology) to see Piper

Pre-op

CSN: 572200266

Date of birth: 03/19/2012

Date of visit: 05/26/2023

Dear Dr. Burke,

I had the pleasure of evaluating Piper. This 11-year-old comes in with a diagnosis of hearing loss that is fluctuating as well as some vertigo that is triggered by loud noises. She has had an extensive workup in the past and there is also concern from previous CT scan in that she has a right-sided superior canal dehiscence syndrome. We reviewed all of her past medical records in addition to her imaging and it is indeed suspicious for a possible right superior canal dehiscence syndrome. However, I mentioned to her family that it is exceedingly unusual to see this in pediatric populations and even when we do see this, it is rare that it causes symptoms. However, Piper's symptoms align really nicely with this and on her exam, there is really nothing else that would explain any of hearing fluctuations or her dizziness symptoms. The ears are perfectly clear on both sides. There are no signs of fluid or infection and her nose and throat exam was equally benign. We talked through the different intervention spectrum of options which ranges from doing nothing and seeing if this spontaneously resolves over time which is a clear possibility although it is exceedingly unsatisfying for the family. They do find that she has her day to day life perturbed by this and even daily activities such as the cafeteria where the noise level gets louder, triggers dizziness on a fairly consistent basis. We talked about superior canal remodeling and resurfacing which would be done through a middle cranial fossa approach and while not a particularly risky operation it does certainly have intuitively more concerns given the need for a temporal craniotomy. As one alternative, we also talked about doing a round window patch in order to try and address the third window effect that we see from patients who have a superior canal dehiscence syndrome. This has some small risk of causing a hearing loss if there is too robust a round window obliteration. However, I do think that this is one of the more conservative options before considering a craniotomy. Her family is very much in agreement, will consider this over the weekend and then they will let me know if they would like to proceed or not.

Sincerely,



https://youtu.be/7SOtz9FETY4?si=DR6zgasJTjPk_SEr: 0:00-1:13



Why does she have a HL?

- SS CD can cause hearing problems such as: (Cleveland Clinic, 2023)
 - Autophony: Hearing your voice or self-generated sounds, like breathing or blinking, louder than normal.
 - Hyperacusis: Heightened sensitivity to sound (meaning sounds may feel unusually loud, uncomfortable or even painful).
 - Pulsatile tinnitus: Hearing noises in your head that occur in time with your pulse, or heartbeat.
 - Hearing loss (especially at low frequencies).
- Connexin 26 can cause hearing loss:
 - While more commonly bilateral, can be unilateral and this has not been well-studied
- Question: Why do you think she has hearing loss?

Question

Why does Piper have a unilateral hearing loss?

- Type your responses in the Q/A

How do you think Piper is post-op?

- Better!
- Same
- Worse (dizziness and/or hearing)

Interdisciplinary Collaboration

- Need for collaboration with other professionals!
- Educational audiologist, ENT, physicians, vestibular audiologist, surgeons



Symptoms different in children

- She did have the Tulio effect
- Her balance testing was normal
- If SSCD was the cause of her hearing loss, or if Connexin 26 is, why were neither of these suggested to family?
- Need for education
 - ENT
 - Pediatric ENT
 - Other physicians



Poll

Do you believe SSC dehiscence repair surgery is a good option for children with SS CD? (or adults)

- No
- Yes
- Thoughts



Questions?

Rebekah F. Cunningham, PhD

Devin L. McCaslin, PhD



Thank you for attending!

- Please remember to complete the exam after the event is over.
- The exam will be available 10-15 minutes after the conclusion of this event.
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 - Choose take exam
 - For Live credit complete the exam within 7 days