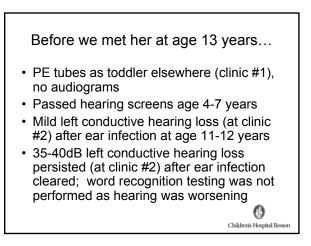


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Before we met her at age 13 years...

- Because of language-based learning disability, she presented to clinic #3 for a central auditory processing evaluation at age 12 years
- CAP eval was deferred because unilateral severe hearing loss with poor word recognition was found
- MRI at clinic #3 was initially interpreted as unremarkable (later reread as showing an area of enhancement in the basal turn)
- Neuroepithelial dysfunction within the left cochlea was concluded
- · Unilateral hearing loss management

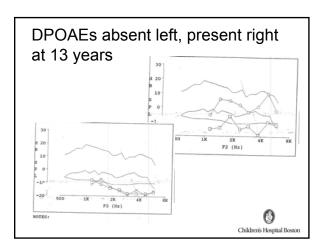
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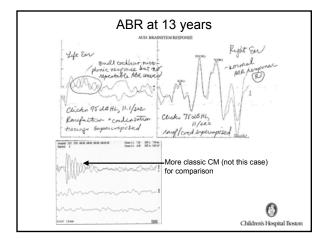
- absent DPOAEs
- small cochlear microphonic with absent ABR waves

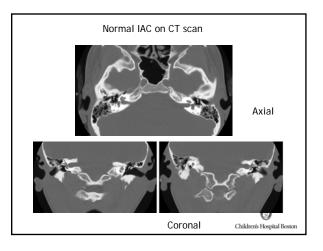
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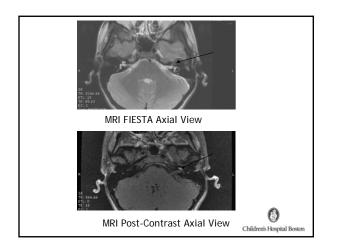
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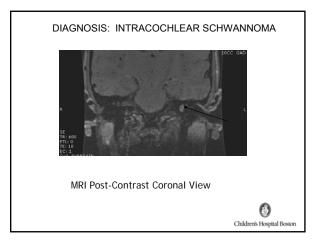
FREQUENCY IN HERTZ (Hz) 125 250 500 1000 2000 4000 8000 KEY -10 R L (HL) IN DECIBELS (dB) 0 0 0 0 0 0 0 AC (AIR) \odot Ψ JNMASKED ASKED 4由 ٦ BC (BONE) Γ °< INMAS ASKED TYMPANOMETRY: NORMAL AU IPSI REFLEXES: PRESENT AD, ABSENT AS NU-6 AD 100%, **NU-6 AS 0%** ୷ HEARING LEVEL 70 П 80 90 100 AT 100 DBHL AGE 13 YEARS 110 0 120 is Hospital Bosto

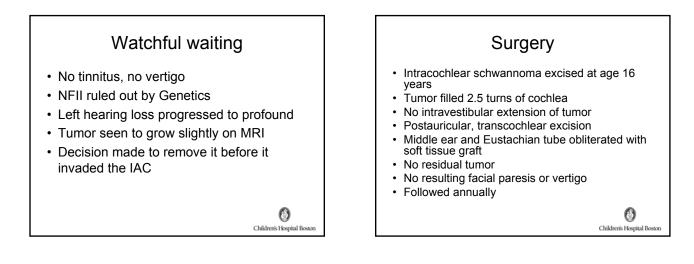


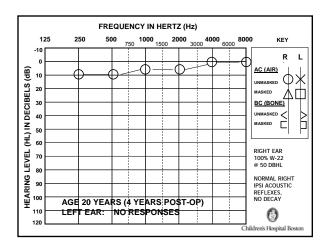


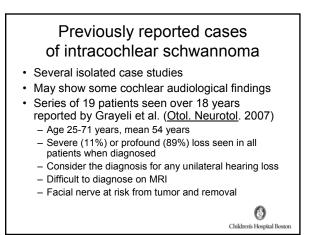








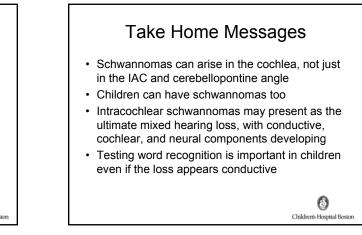


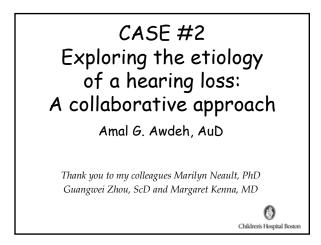


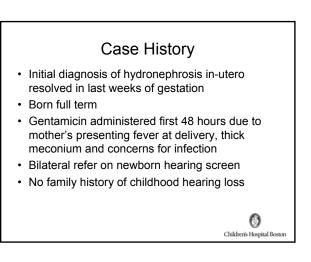
Pediatric acoustic neuromas

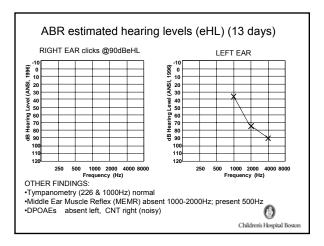
- · Rare outside neurofibromatosis type II
- Chen et al. (Am. J. Otol. 1992) reviewed 16
 cases age 1-14 years; none were described as
 intracochlear
- Mazzoni et al. (Int. J. Ped.ORL 2007) described 10 non-NFII pediatric cases, none intracochlear
- Laury et al. (Int. J. Ped. ORL, 2009) reported a 13 year old with vestibular schwannoma in a pediatric series with unilateral neural loss

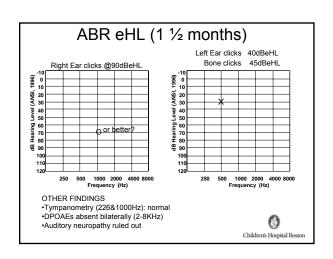
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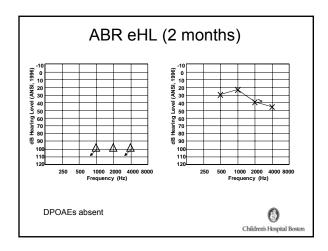


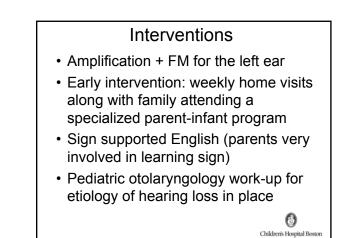




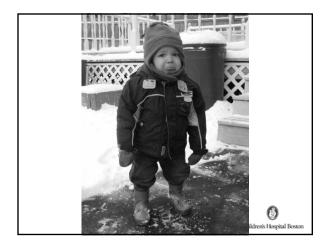


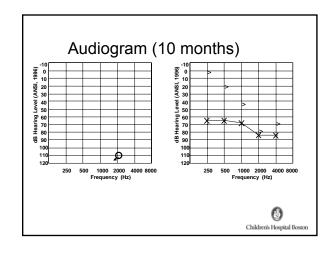






ABR eHL (4 1/2 months) Work-up to determine etiology · CT-scan of the temporal bones unremarkable (966) · Genetic studies: (ISN 20 30 (ISNE) 30 DN - negative for mitochondrial mutations 40 50 19 40 50 evel-- negative for connexin 30 test dB Hearing L 06 02 02 09 09 60 70 80 - negative for Pendred syndrome - connexin 26 test showed 1 pathogenic 100 110 120 110 mutation of 35delG (suggesting he is a carrier -250 500 1000 2000 4000 8000 Frequency (Hz) 250 not the likely cause of the hearing loss) • Negative CMV test at 2 weeks of age Tympanometry (226&1000Hz) normal Normal ophthalmology evaluation 0 Children's Hospital Bost





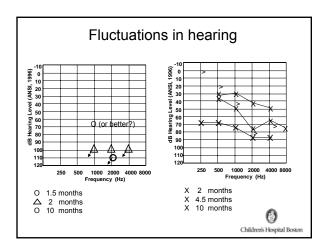
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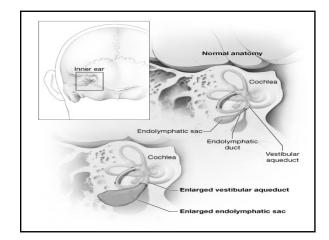
1000 2000 Frequency (Hz)

4000 8

0

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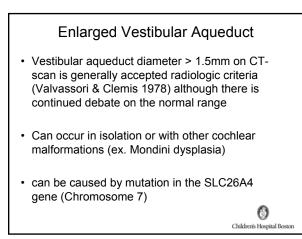




Enlarged Vestibular Aqueduct (EVA)

- · Hearing loss congenital or develops later on
- Progressive/fluctuating
- May or may not be accompanied by vestibular symptoms;
- Unilateral or bilateral (Mori et al J Otolaryngol Head Neck Surg. 2008 in their systematic literature review found bilateral EVA 6 times more common than unilateral EVA)
- Previously reported in the literature as sensorineural hearing loss; however Zhou et al (Laryngoscope 11/08) suggests 80% of the 54 children with EVA in their retrospective study had either conductive/mixed hearing loss

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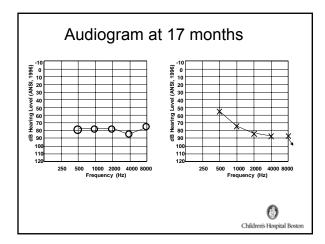


Recent hypothesis on air-bone gap in EVA EVA acts a "third mobile window" (Merchant et al. Ann Otol Rhinol Laryngol. 2007) shunting of air-conducted sound away from the cochlea (through the enlarged vestibular aqueduct) elevates *air conducted* thresholds "third mobile window" increases the difference in impedance between the scala vestibuli side and the scala tympani side of the cochlear partition, improving *bone-conducted* thresholds.

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Testing for EVA Tympanometry, MEMR, DPOAEs Pure-tone audiometry (supra-normal bone thresholds including 250Hz) CT-scan or MRI imaging studies for clinical diagnosis VEMP - abnormally low threshold response an audiological sign in children in presence of non-middle ear related mixed/conductive hearing loss (Zhou et al, Layngoscope 11/08)

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Changes in management plan

- · Continue re-examining right ear
- Added amplification on the right side cautiously in light of improved thresholds (auditory nerve stimulation opportunity)
- Education on avoidance of head trauma/sudden barometric pressure changes
- · Family awareness of possibility of progression
- Informational CI consult
- Early EVA diagnosis may prevent unnecessary surgical/exploratory procedure to correct lowfrequency conductive loss
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Take Home Messages

- Important to try to determine site of origin of airbone gap using tympanometry, MEMR, DPOAEs; VEMP
- EVA one of the contenders to consider when there is a conductive/mixed loss unexplained by healthy middle ears
- EVA a case of conductive hearing loss of inner ear origin

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CASE #3 AND #4 Conductive Hearing Loss in Children: Expect the Unexpected

Guangwei Zhou, Sc.D.

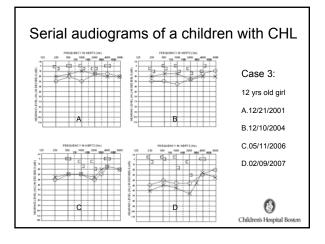
In collaborating with Dennis Poe, M.D., Quinton Gopen, M.D., Manali Amin, M.D., Laurie Ohlms, M.D., Dwight Jones, M.D., Jane Liberman, Au.D.

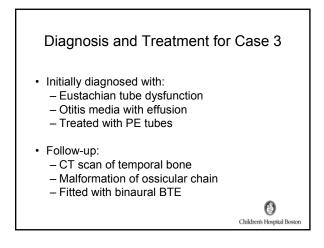
guangwei.zhou@childrens.harvard.edu

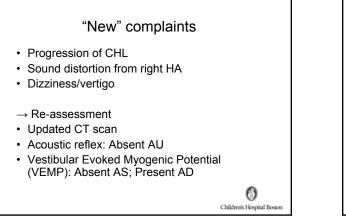
Common Etiologies of CHL in Children External ear Microtia and Atresia

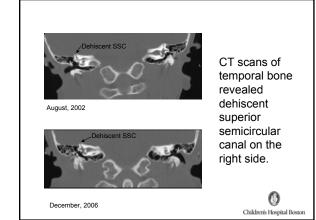
- Impacted cerumen
- Middle ear
 - Tympanic membrane perforation
 - Eustachian tube dysfunction
 - Otitis media with effusion
 - Irregularities of ossicular chain
 - Cholesteatoma

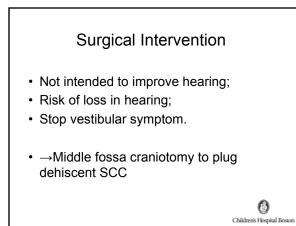
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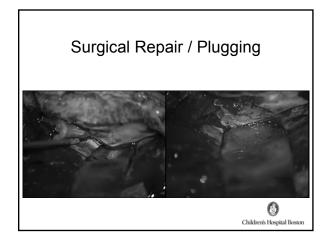








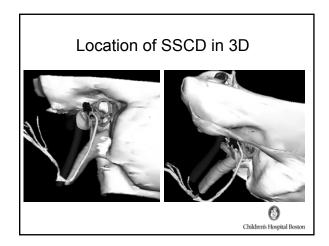


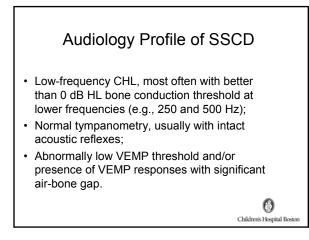


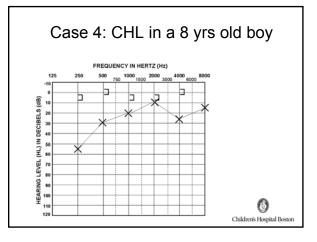
Superior Semicircular Canal Dehiscence (SSCD)

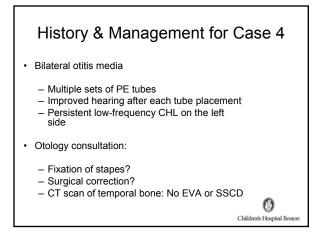
- Sound and/or pressure-induced vertigo due to dehiscence of the superior semicircular canal, Verticaltorsional eye movements.
 - Minor et al. 1998
- Patients can present with vestibular, auditory, or symptoms of both.
 - "Minor Syndrome"

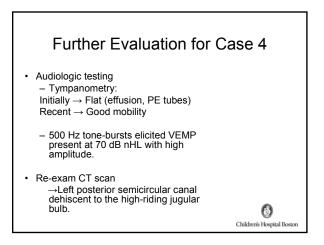
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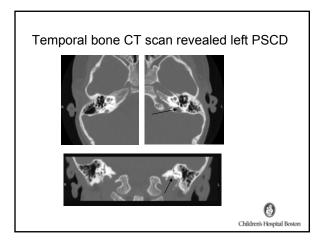


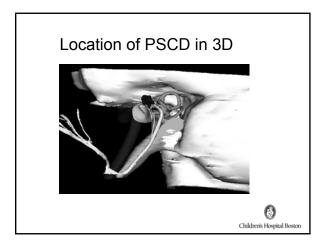


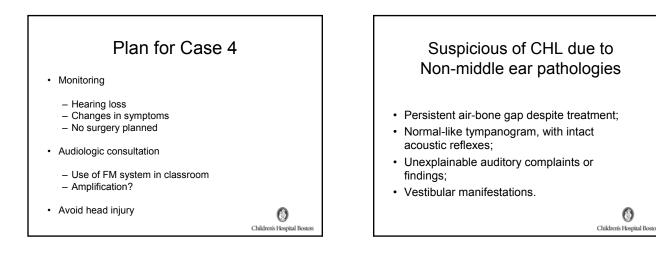












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CHL attributable to Inner ear abnormalities

- Superior semicircular canal dehiscence (SSCD)
- Posterior semicircular canal dehiscence (PSCD)
- Enlarged vestibular aqueduct (EVA)
- · Enlarged cochlear aqueduct
- · Malformed cochlea and/or dilated vestibules
- Others

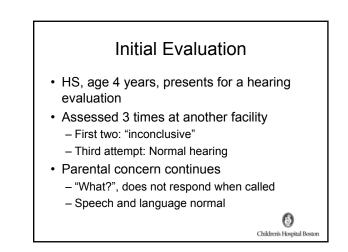
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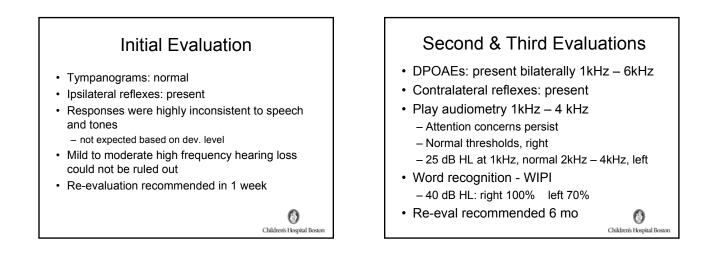
CASE #5 When Poor Reliability is Reliable

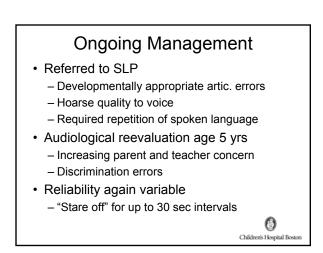
Cheryl Edwards, M.S. cheryl.edwards@childrens.harvard.edu

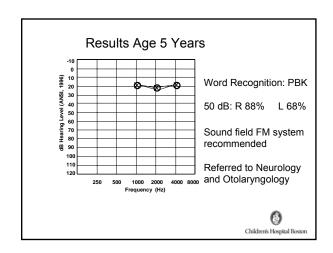
With special thanks to Katie West, M.A.

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Medical Evaluations

- Neurology
 - EEG unremarkable
- Otolaryngology
 - Brain MRI was normal
 - CT revealed bilateral Mondini malformation

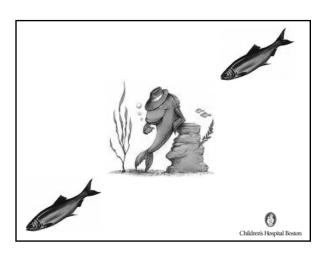
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Subsequent Evaluations

- · Mild/minimal low freq. fluctuating CHL
- Word recognition
 poorer than expected based on pure tones
- ABR: normal
- · Good days/bad days observed
- · Discrimination errors persist with FM
- Presence of Mondini the explanation?

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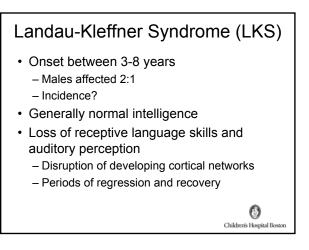


6 Years of Age HS began "acting deaf" could only respond with visual cues "Staring off" behavior noted again 24 hour EEG Abnormal bilateral spike and wave discharges activated by sleep, temporal lobe Diagnosis: Landau-Kleffner Syndrome

Landau-Kleffner Syndrome (LKS)

- · Described 1957
- · Acquired aphasia due to seizure activity
 - Spikes or sharp waves over temporal and/or parietal lobes on EEG
 - Activated by sleep, not behaviorally obvious
- · Inability to recognize sounds
 - May appear HOH or deaf
 - Environmental sounds cannot be identified
- Normal pure tone audiogram

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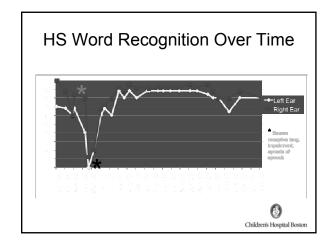
Landau-Kleffner Syndrome (LKS)

- · Can read and write if skills already in place
- Some children recover completely

 Earlier onset associated with poorer outcomes
 - Most have no seizure activity by adulthood
- About 50% are left with residual deficits
 - Functionally inappropriate connections during critical period?
 - Seizures controlled with medication

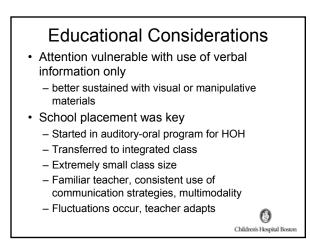
· Multiple subpial transection

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Age		9 years	10 years	11 years	11 years	12 years
Filtered Words	R			48		72
	L			44		36
Dichotic Digits	R	95	98	90		85
	L	18	30	40		63
Comp Sent	R	100	100	100	100	100
	L	0	0	0	0	0
		Binaural				
Pitch Patterns		0%		20 %		

Treatment - HS Variety of medications for seizures Understanding decreased if seizures were not well controlled Completed FastForWord Some subjective improvement Has difficulty with learning musical instrument





Follow-up

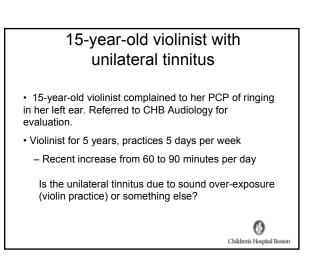
- Now 15 years old
- · Slightly more resistant to FM use
- Seen annually, sooner if concerns – Mother extremely good observer
- Scheduled for 24 hour EEG in June
 Monitor for seizure activity

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Closing Thoughts • Parent concern drove this diagnosis • Collaboration • If the answers don't add up, keep looking! • If the answers don't add up, keep looking!

CASE #6 Prescriptive Fitting of Custom Hearing Protection for a Teenage Violinist *or...* Why a pediatric audiologist still needs to know hearing science Brian J. Fligor, Sc.D.

> (i) Children's Hospital Bost



Evaluation <u>Primary Questions:</u> 1. Does this patient have a noise-induced hearing loss (NIHL)? 2. Is this patient's violin practice the source of a hazardous sound exposure, accounting for her tinnitus, and sufficient to place her at risk for NIHL? (otologic and noise history otherwise unremarkable) 3. If so, what is the best approach for reducing her NIHL risk?

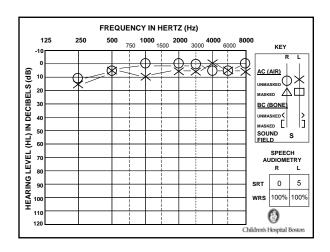
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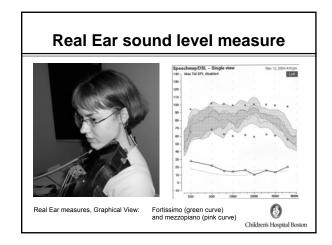
Elements of a Hearing Loss Prevention Program (HLPP)

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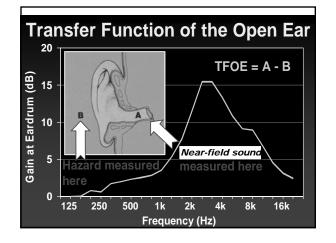
- Noise Survey (assessment)
- Engineering Controls
- Audiometric Monitoring
- Education and Motivation
- Hearing Protection Devices (HPD)
 - Apply in a pediatric setting?

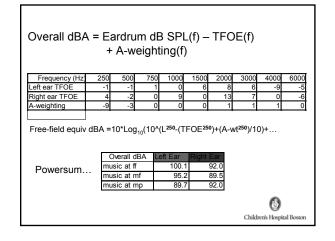




eft Ear	Eardrum of								
Frequency (Hz) 250 57	500 83	750 97	1000 87	1500 97	2000 100	3000 89	4000 83	6000 76
ortissimo nezzoforte	5/	83 50	97 52	87 92	97 49	99	89	83	64
nezzotorte nezzopiano	41	50	52	92	49	99	73	63	55
lezzopial lo	#3 Eardrum (/1	02	01	92	13	63	55
Frequency (Hz		500	750	1000	1500	2000	3000	4000	6000
ortissimo	46	78	58	85	89	2000	92	77	57
nezzoforte	80	85	80	91	81	92	87	74	49
nezzopiano	52	60	82	90	90	89	87	80	57
Powersum a Convert dB					Level	(OAL):			

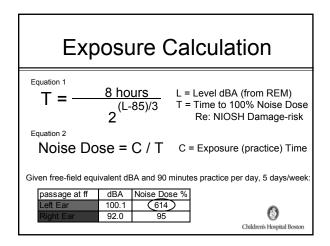
.eft Ear	Eardrum o	IB SPL								
Frequency (Hz) 250	500	750	1000	1500	2000	3000	4000	6000	OAL SPL
ortissimo	57	83	97	87	97	100	89	83	76	103.4
nezzoforte	41	50	52	92	49	99	81	71	64	99.9
nezzopiano	43	88	71	82	81	92	73	63	55	94.0
light Ear	Eardrum o	B SPL								
Frequency (Hz) 250	500	750	1000	1500	2000	3000	4000	6000	OAL SPL
ortissimo	46	78	58	85	89	96	92	77	57	98.3
rezzoforte	80	85	80	91	81	92	87	74	49	96.0
nezzopiano	52	60	82	90	90	89	87	80	57	95.5
Powersum across frequencies for Overall Level (OAL): Convert dB SPL to intensity = 10 ^(dB/10) OAL dB SPL = 10*Log ₁₀ (10 ^(L250/10) +10 ^(L500/10) ++10 ^(L6000/10)) (at the eardrum) But "Hazard" is measured in A-weighted decibels in the free-field										

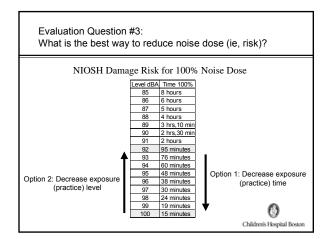


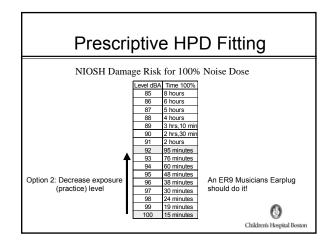


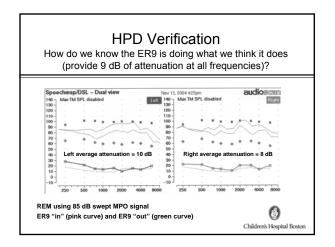
Evaluation Question #2: Is her violin practice sufficient to explain left-sided tinnitus?									
i.e., given her practice duration and regularity, is her noise exposure potentially hazardous?									
Overall dBA	Left Ear	Right Ear							
music at ff	100.1	92.0							
music at mf	95.2	89.5							
music at mp	89.7	92.0							
			0						

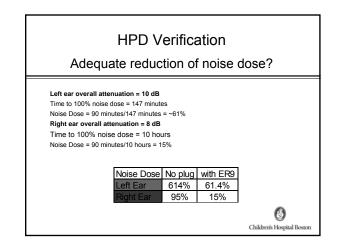
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Elements of a Hearing Loss Prevention Program (HLPP)

- Noise Survey (assessment)
- Engineering Controls
- Audiometric Monitoring
- Education and Motivation
- Hearing Protection Devices (HPD)
- The finances:
 - You are more obviously "selling" a service
 - 92596 "Ear Protector Evaluation"

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