



Transforming Lives:
Management of Hearing Loss
Associated with Cleft Lip/Palate
and Other Craniofacial Conditions

Generous funding of this program provided by the Milbank Foundation

Thursday, October 13, 2022

1

Welcome and Introductions



Stephanie Paul
Executive Director
myFace



2



Presentation



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Pediatric Audiologist
Seattle Children's Hospital
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Pediatric Otolaryngology – Head and Neck Surgery
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University of Washington School of Medicine
Seattle, WA



3

Disclosures

- **Aijiana Zanders, AuD, CCC-A**
Dr. Aijiana Zanders is an employee of Seattle Children's Hospital with no other relevant financial disclosures.
- **Randall A. Bly, MD**
Dr. Randall A. Bly is co-founder and holds a financial interest of ownership equity with Wavely Diagnostics, Inc. and EigenHealth, Inc. He is a co-inventor and consultant, Spiway, LLC.



4

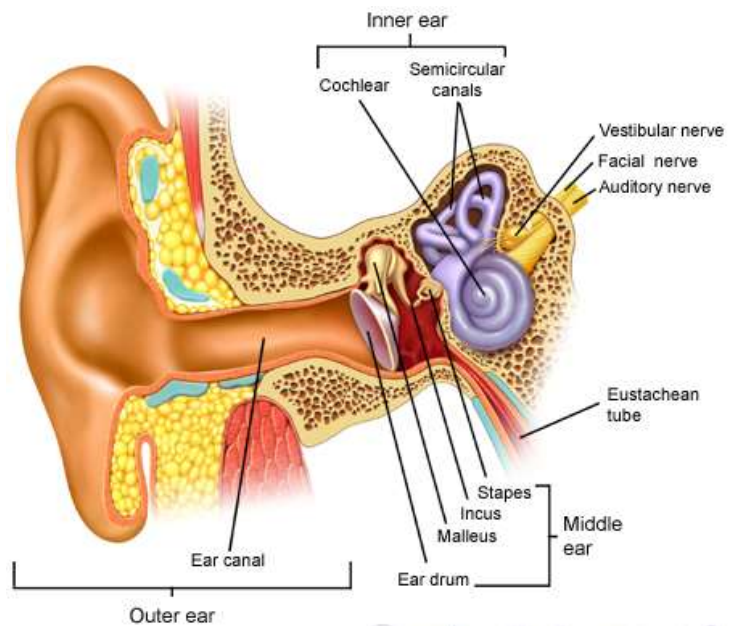


Presentation Outline: Dr. Aijiana Zanders

- a. Ear Anatomy
- b. Types of hearing loss
- c. Ways to test hearing
- d. Potential hearing device recommendations (surgical and non-surgical)
- e. Recommendations for kids with hearing loss

5

Ear Anatomy

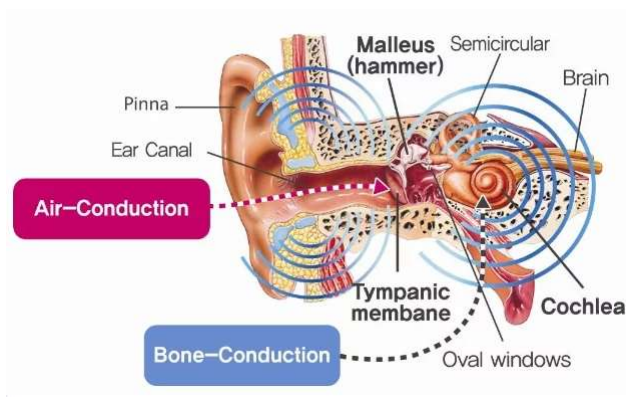


6



Transmission of Sound

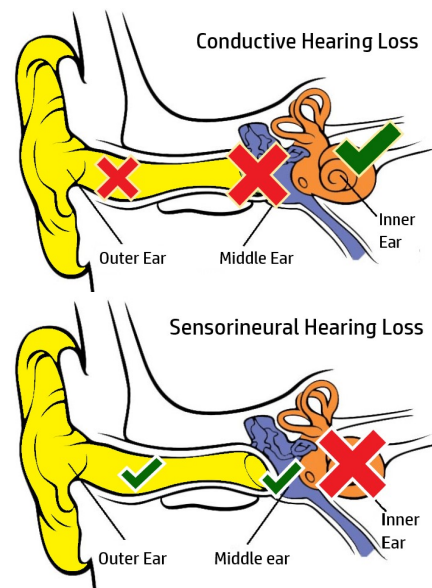
- **Air Conduction**
Sound passes through the outer and middle ear before reaching the cochlea
- **Bone Conduction**
Sound passes through bones of the skull to the inner ear directly, bypassing (skips) the outer and middle ear



7

Types of Hearing Loss

- Conductive Hearing Loss (CHL)
 - Outer/middle ear problem
 - Often temporary, but can be permanent
- Sensorineural Hearing Loss (SNHL)
 - Inner ear/auditory nerve problem
 - Permanent
- Mixed Hearing Loss
 - Mixture of CHL and SNHL



8



Different Ways to Test Hearing

9

NORMAL HEARING →

HEARING LOSS →

The Audiogram

10



Hearing Devices – Traditional Hearing Aids



11

Osseointegrated Bone Conduction Sound Processor

- For conductive hearing loss (as well as mixed hearing loss)
- FDA approved at 5 years old (*12 years old for OSIA)



Cochlear Implant

- For severe to profound sensorineural hearing loss; limited to no benefit from traditional amplification
- FDA approved at 9 months of age



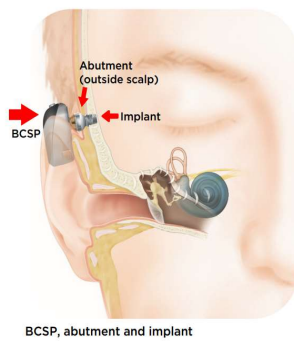
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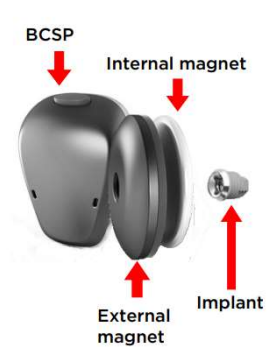
Bone Conduction Sound Processor (Surgical)



OSIA



ABUTMENT



ATTRACT



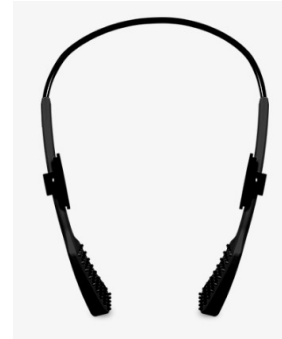
13

Bone Conduction Sound Processor (Non-Surgical)

SOFTBAND



SOUNDARC



14



Recommendations For Kids with Hearing Loss

- If recommended, establish full time hearing device use
- Establish care with Early Intervention
 - Special programs designed to support learning and development
 - Purpose is to achieve optimal child and family outcomes
 - The earlier the better (JCIH, 2019)
- Monitor hearing regularly
- Consistent follow-up with Otolaryngology



15

Recommendations For Kids with Hearing Loss (continued)

- Using good communication strategies:
 - Gain attention before speaking
 - Decrease background noise
 - Decrease distance
 - Speak using a reasonable volume
 - Enunciate and pronounce words clearly
 - Ensure your child can see your face and mouth when speaking to them



16



Presentation Outline: Dr. Randall Bly

- a. Common hearing loss presentations for craniofacial conditions
- b. The cause of the hearing loss
 - Focus on conductive hearing loss (problem with sound getting in)
 - Sensorineural hearing loss can also be present, sometimes both – mixed hearing loss
- c. Management options
 - Observation
 - Access to sound through a variety of device types
 - Surgery

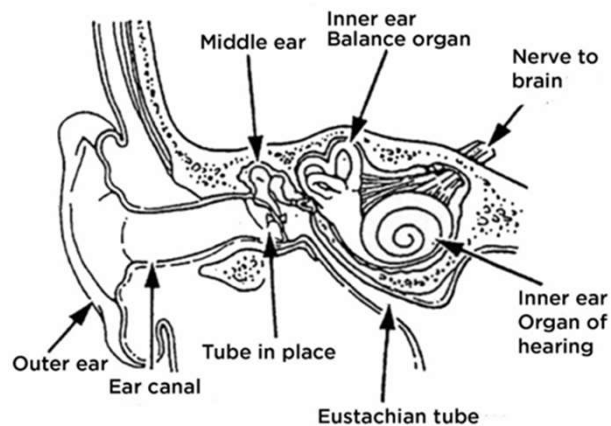


17

Mild conductive hearing loss (as seen with fluid behind ear drum)

Any condition that impacts eustachian tube function:

- i. Cleft palate +/- cleft lip
- ii. Craniofacial microsomia, microtia, aural atresia
- iii. Apert, Crouzon, Pfeiffer
- iv. Robin sequence
- v. 22q deletion



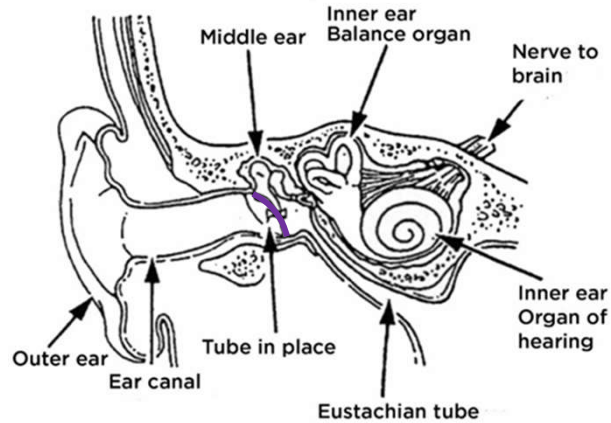
18



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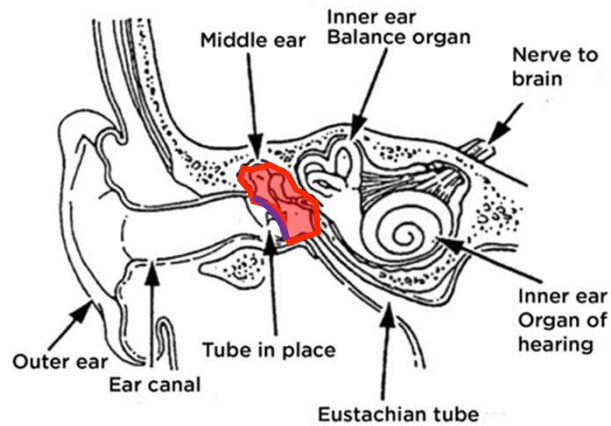


19

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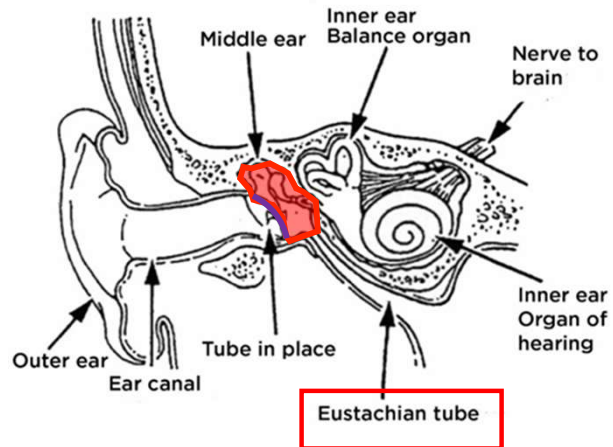
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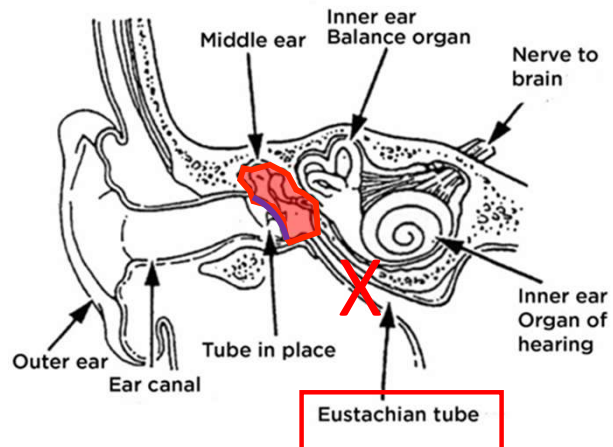


21

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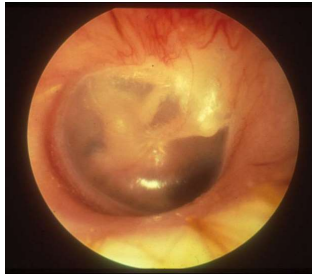


22



Mild conductive hearing loss, 8 months of age

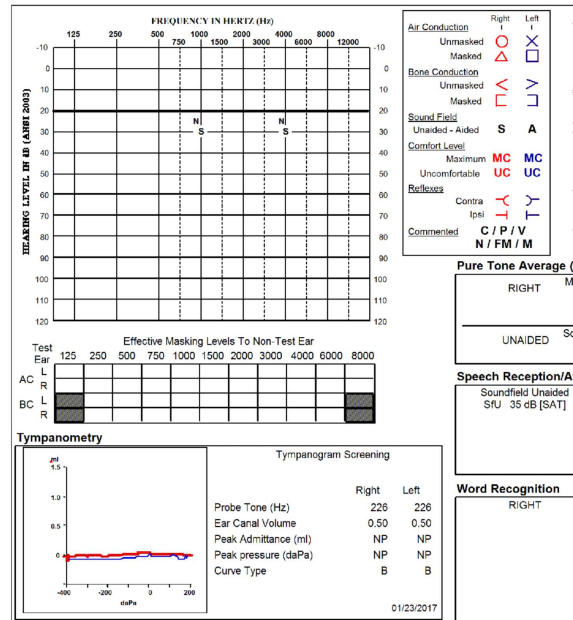
a. Patient with cleft palate example



Normal right ear drum



Ear drum with fluid behind it



23

Ear tube placement, typically at same time as palate repair

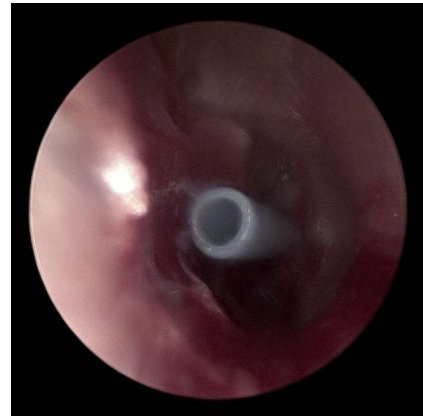
- 5-minute procedure with microscope
- Small incision away from ear bones
- Remove fluid, place ear tube
- OK to swim, especially surface-level swimming
- Ear tube typically comes out spontaneously 1–2 years

Benefits

- Improved hearing by removing fluid
- Possibly improved management of ear infections (ear drops)

Risks

- 1–3% risk for ear drum perforation afterwards
- May need to repeat procedure after ear tube comes out, if fluid returns
- Chronic ear drainage (rare after palate repair)

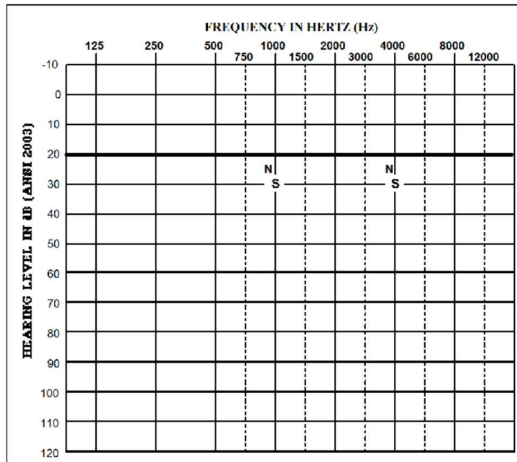


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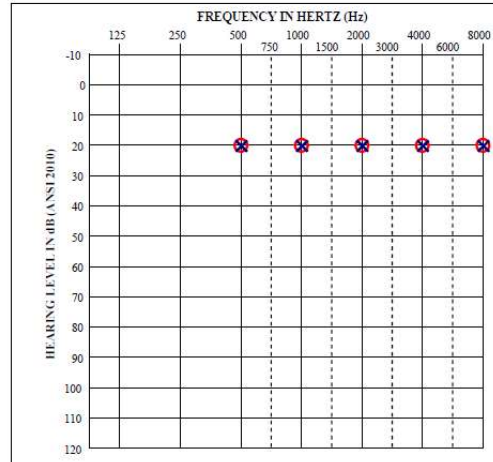


Normal hearing after ear tube placement

Before ear tube placement



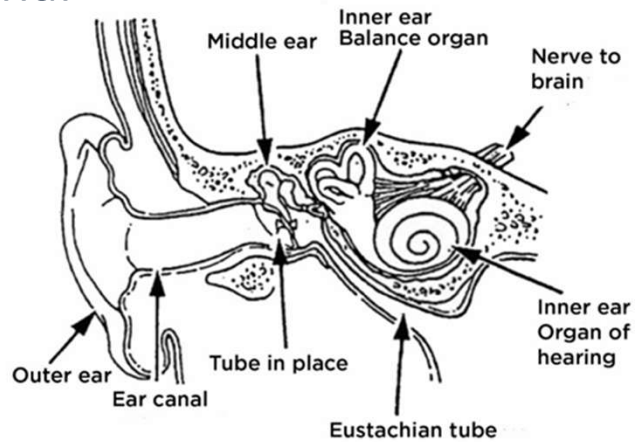
After ear tube placement



25

Conductive hearing loss because of abnormal ear bones or ear canal

- Aural atresia
- Abnormal ear bones (ossicles) preventing movement
- Usually a severe hearing loss (compared to mild for fluid)

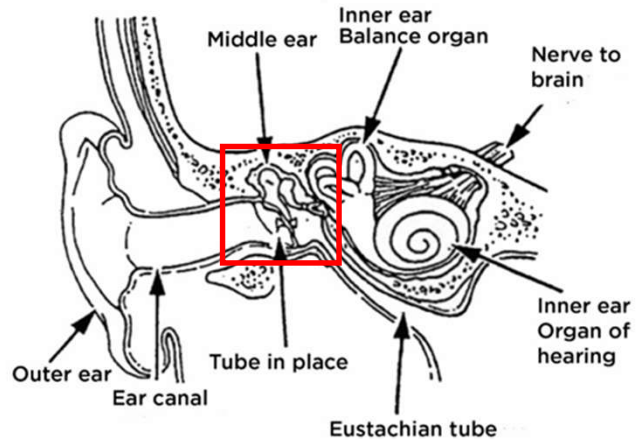


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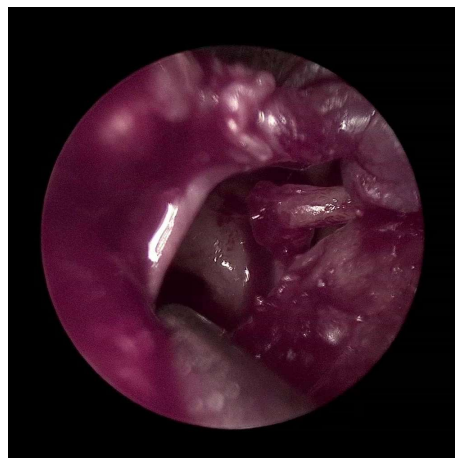
Conductive hearing loss because of abnormal ear bones or ear canal

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27

Ear bones conduct sound energy into cochlea

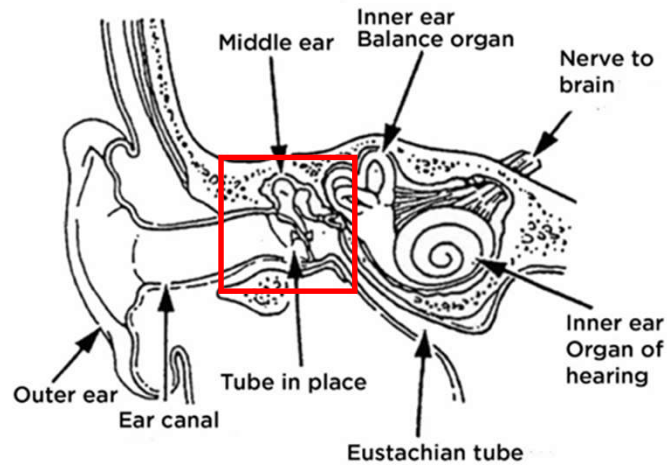


[Video01_093022]

28



Aural atresia (ear canal did not form fully)



29

Aural atresia (ear canal did not form fully)



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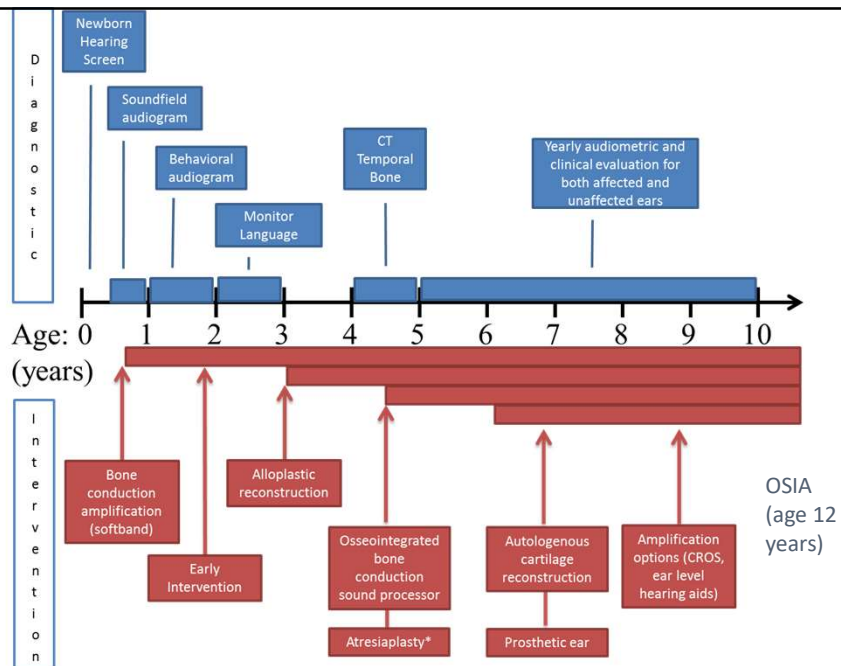


Aural atresia (ear canal did not form fully)



31

Aural atresia and microtia – management overview

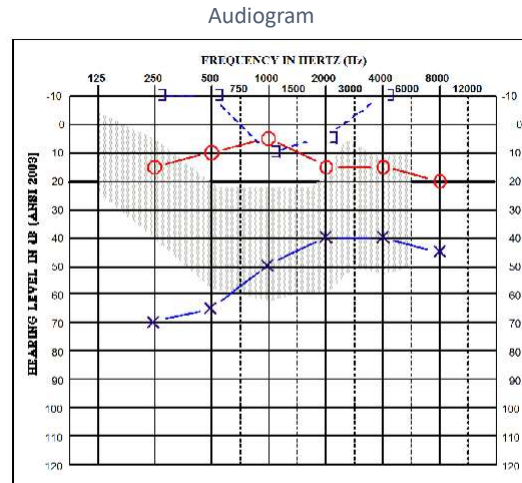


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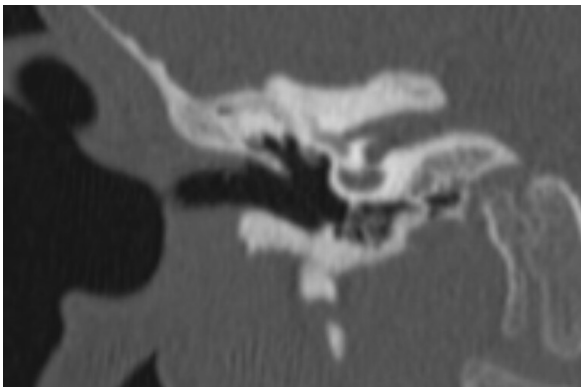
Very narrow ear canal with tympanic membrane that did not fully form

Example of narrow ear canal and conductive hearing loss

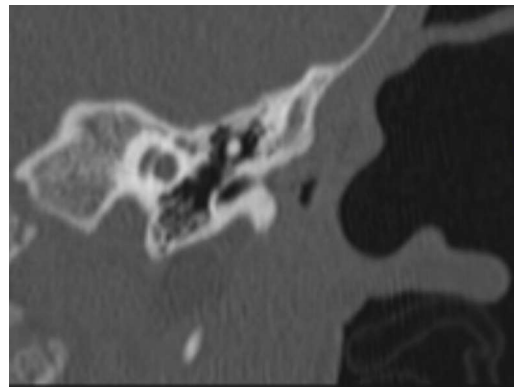


33

Normal ear canal with normal thin ear drum (without bone)



Very narrow ear canal with bony plate where ear drum should be



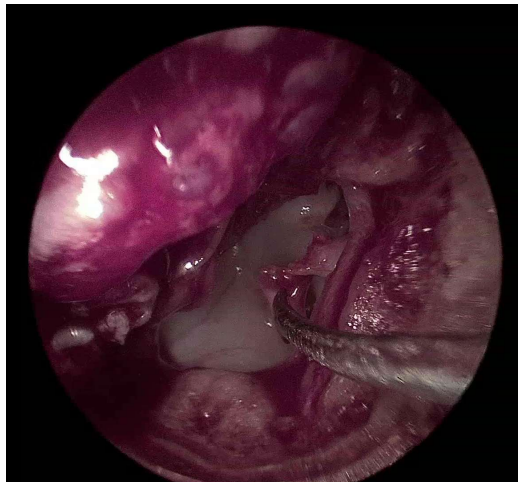
34



Ossiculoplasty (rebuilding ear bones)



35



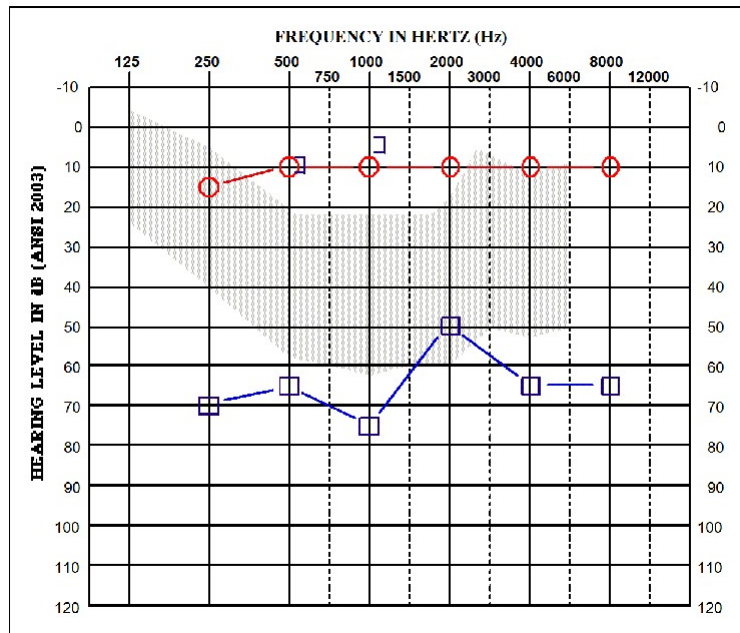
[SlideVideo]

36

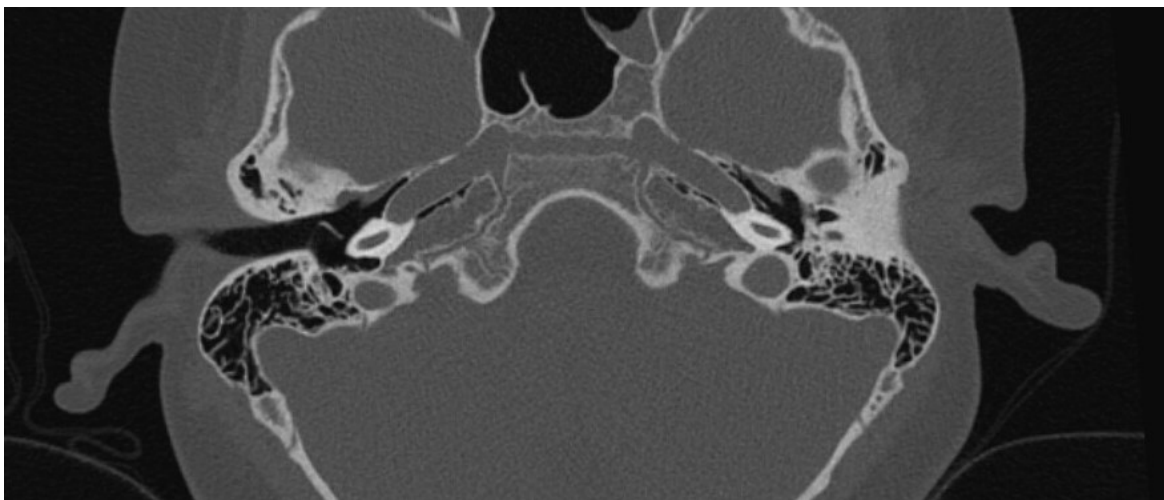


Complete aural atresia, patient example

Audiogram with normal hearing on right, maximum conductive hearing loss on left



37

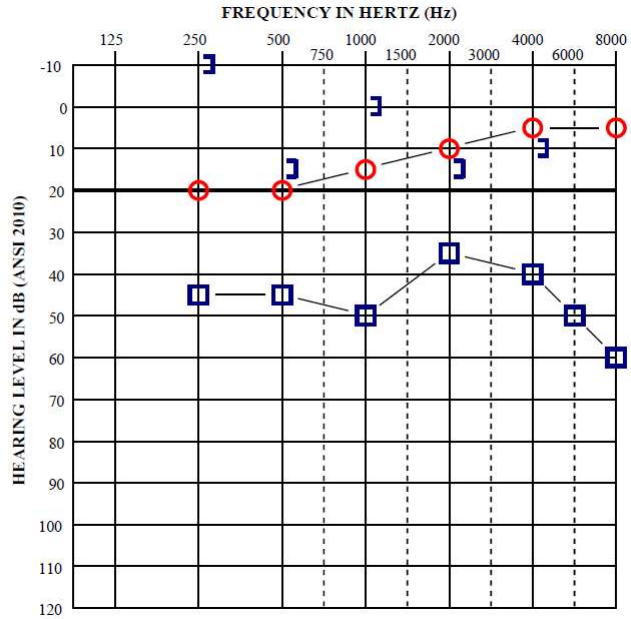


Left aural atresia

38

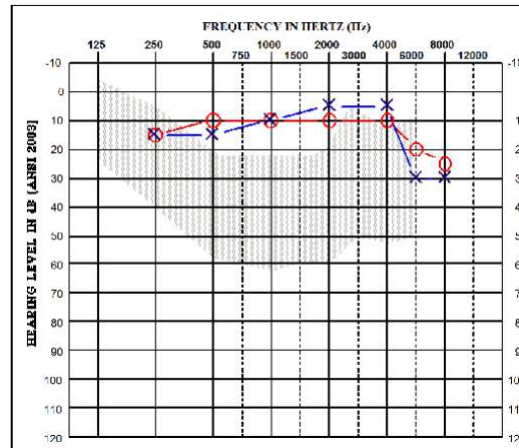
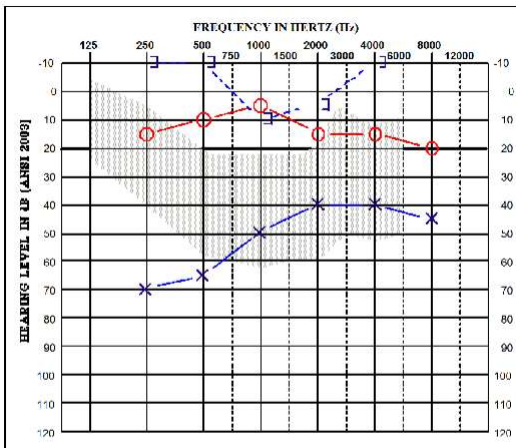


Post operative audiogram.
Pure tone average: 63 to 40 dbHL



39

Pre and post audiogram after ossiculoplasty



40



Conclusion

- Important to understand severity and type of hearing loss
- Surgery is one of many options available to patients
- Access to sound is really important
 - Can be done through a variety of techniques



41

Meet the Cole Family



42



Austin's Early Years



43

Before surgery (ear exams
in clinic were tough!)



44



After surgery



45

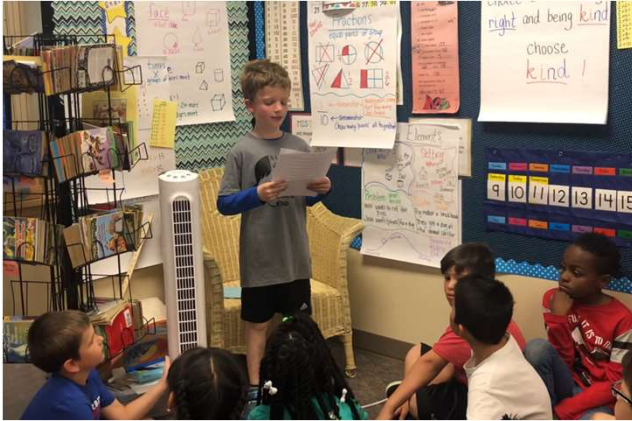
Austin's Experience with ear exams and surgery



46



Austin presenting in 2nd grade! Ear canal needs ongoing maintenance though



47

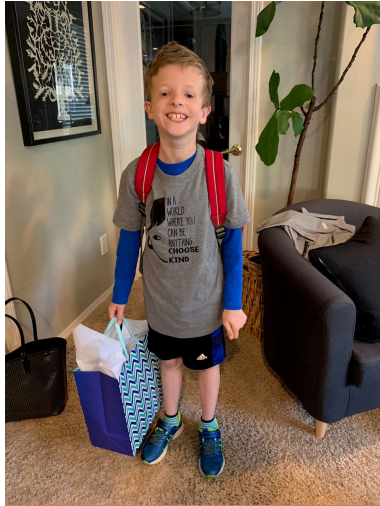
And more surgery



48



Austin Today – and Plans For the Future



49

Resources

- <https://www.asha.org/public/hearing/Hearing-Loss-in-Children/>
- <https://www.cdc.gov/ncbddd/hearingloss/facts.html>
- <https://www.cdc.gov/ncbddd/hearingloss/ehdi-programs.html>
- <https://www.seattlechildrens.org/pdf/PE3312.pdf>
- <https://www.seattlechildrens.org/pdf/PE1259.pdf>

50



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51

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52



Questions & Answers



53

Closing Remarks



Stephanie Paul
Executive Director
myFace



54



Please Ask Congress to Support Ally's Act

- Ally's Act is a bill that would require insurance companies to provide coverage for osseointegrated hearing devices such as Bone Anchored Hearing Aids (BAHAs) and Cochlear Implants
- Ally's Act is currently under consideration in the U.S. House of Representatives and in the Senate
- Please consider sending a pre-written email to your Representatives asking them to support Ally's Act at: myface.org/allysact

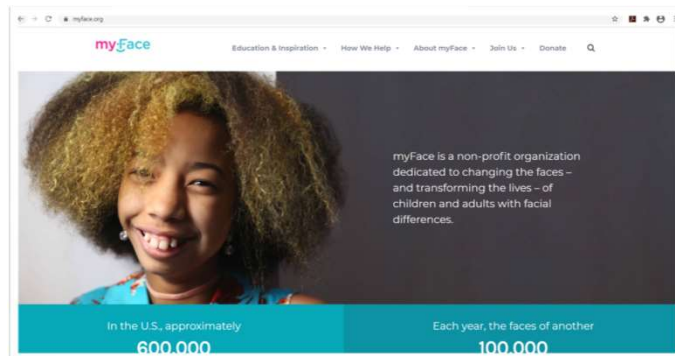


Ally Tumblin



55

For additional information and resources
visit [myFace.org](https://myface.org)



Or email us at
info@myface.org



56



Upcoming Events at myFace

Join us on
Wednesday, Oct. 26th at 12 PM ET
for a discussion on Exploring Face Equality
with Phyllida Swift, CEO of Face Equality
International (FEI)!

Register at:
www.myface.org/mystory/



57

Upcoming Events at myFace

Join us in person in New York City for our annual **Holiday Party**

Thursday, December 8th from 4:30 – 7:30 PM

Stay tuned for further information about this event



58



Upcoming Events at myFace

Join us for the next webinar in our **Transforming Lives Webinar Series:**
Diagnosis and Management of Craniosynostosis – Coming in Early 2023



Mark Urata, MD
Division Chief
Division of Plastic and Maxillofacial Surgery
Children's Hospital Los Angeles (CHLA)



Alexis Johns, PhD, ABPP
Pediatric Psychologist
Children's Hospital Los Angeles (CHLA)

Further information about this webinar will be
available on the myFace website soon

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59

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Thank You

Please complete the evaluation at: myface.org/hearingloss

60