

EI services represent the purpose and goal of the entire EHDI process. Screening and confirmation that a child is D/HH are largely meaningless without appropriate, individualized, targeted and high-quality intervention. (Joint Committee on Infant Hearing, 2013)



MAKING THE CASE:

*FOLLOWING 1:3:6 FOR CHILDREN WITH
MICROTIA/ATRESIA*

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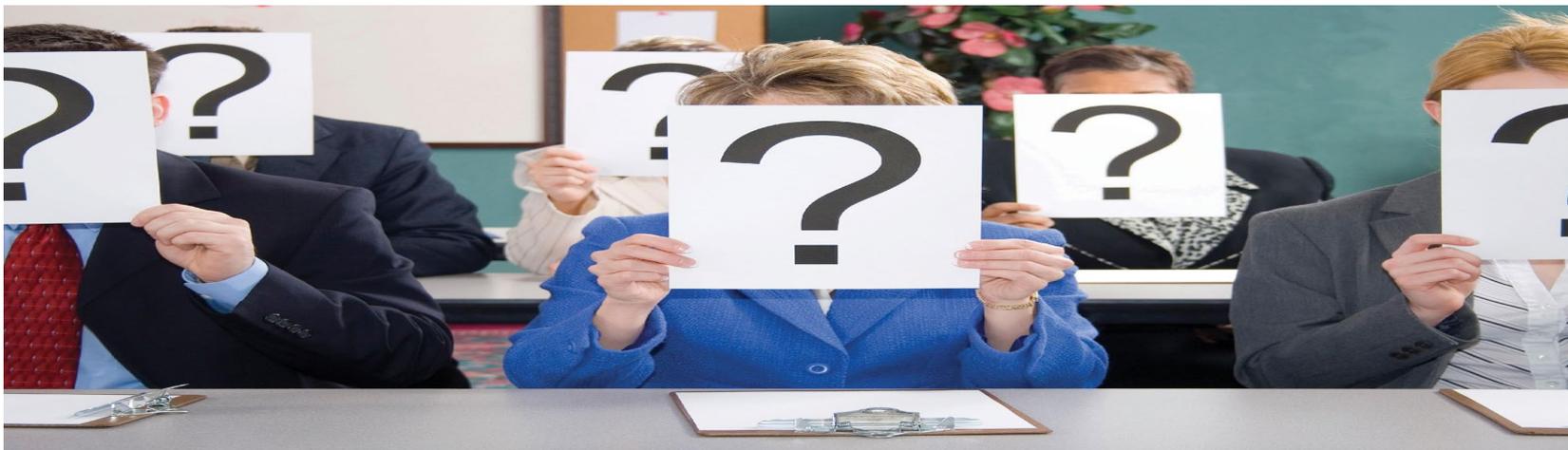
Learning Outcomes

Participants will...

...review and identify how the JCIH position statement and recommendations apply to children with microtia/atresia.

...identify strategies for supporting families of children with microtia/atresia.

...identify treatment options and school-based implications in the microtia/atresia population.



WHO ARE YOU?

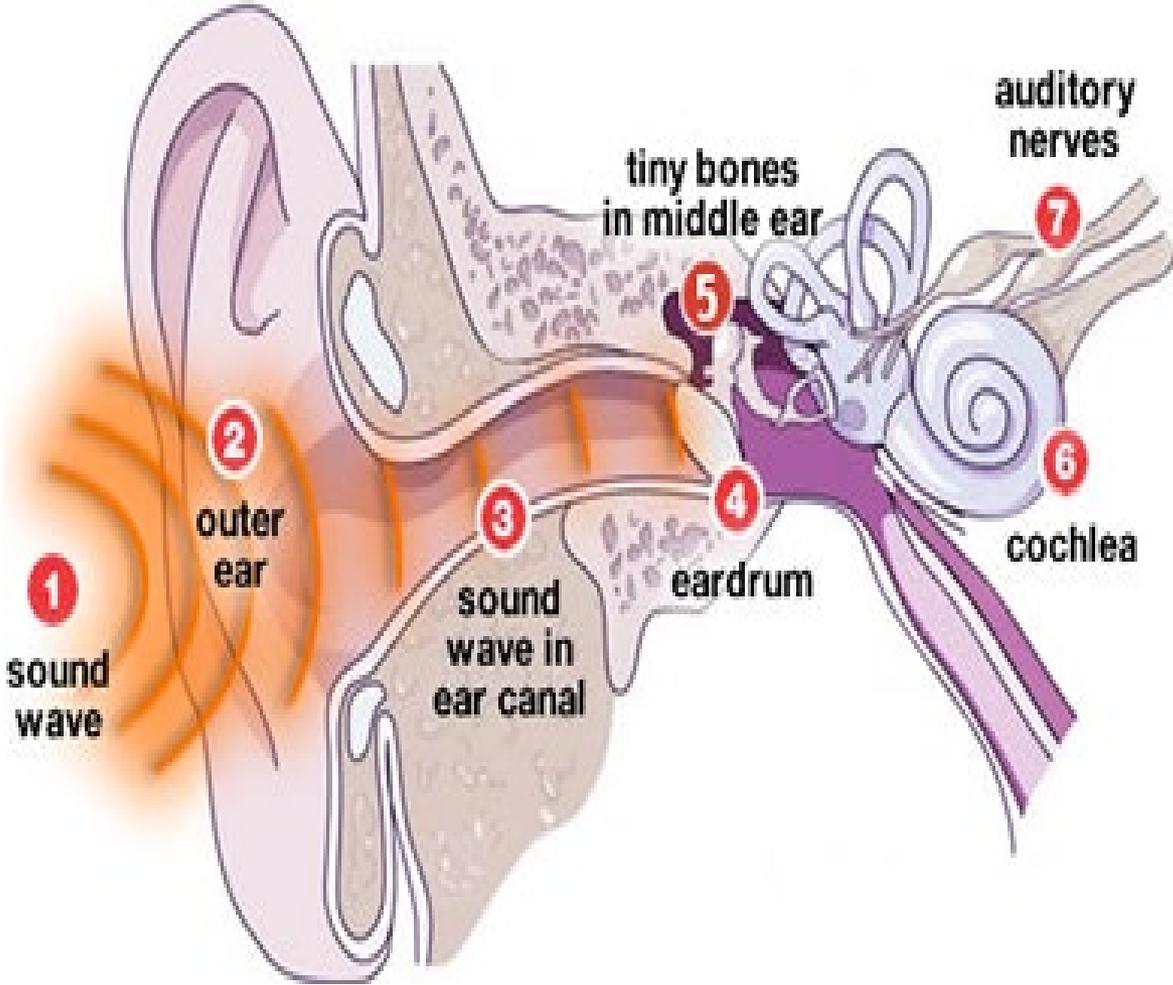
*WHY ARE YOU
HERE?*

What is microtia / atresia ?



- AKA aural atresia
- Small ➤ No outer ear , closed/absent ear canal
- Conductive hearing loss
- 1 in 10,000 - 5 in 10,000 (Center for Disease Control and Prevention, 2018)
- Asians -3X Higher, Hispanics-7x (Shaw et al., 2004)
- Most common: male, unilateral (>90%), right ear (60%)
- Oculo-Auriculo-Vertebral-Spectrum (OAVS), sometimes referred to as hemifacial microsomia spectrum is the most common “other” (includes Hemifacial Microsomia and Goldenhar)

Effects on hearing



*What does
JCIH say
about
children with
CHL?
JCIH 2000*

Defines the targeted hearing loss for UNHS programs as “permanent **bilateral or unilateral**, sensory or **conductive** hearing loss.....”

Infants in whom permanent hearing loss is diagnosed should be fitted with an amplification device within 1 month of diagnosis.

All families of infants with any degree of bilateral or unilateral permanent hearing loss should be considered eligible for early intervention services.

In the beginning...

He was the first to notice. Or, more likely, the first to say it. “There’s something up with her ears.”Within minutes N was in the corner on his iPhone, diagnosing our daughter before anyone else had the chance to. Microtia, he said. Atresia. It meant nothing to me. But then, I *looked*. There wasn’t just something wrong with her ears—she barely *had* ears. More alarmingly, there were no ear canals. Just two little peanuts on either side of a perfect little head, so perfect on its face that it seemed to mock the malformations on its sides.

<http://ivyleagueinsecurities.com/2013/05/my-sisters-birth-story/#sthash.s3P3j1AE.pdf>



Options. (Reinisch and Lewin, 2009),

Age	Audiological	Ear Canal	Reconstruction	Therapy and support
Birth -3 	ABR/Testing Amplification: Non-surgical <ul style="list-style-type: none"> • Baha 5 • Ponto • Adhear 			Early Intervention Language Development Parent Supports
>3 yrs			Medpor/Supor Reconstruction	
>5 yrs	Surgical implantation of baha	Atresiaplasty		
≥ 7 yrs			Rib Cartilage/Graft (3-4 surgeries per ear)	

*Identify what we
are talking
about in EHDI
when we talk
about children
with
microtia/atresia*

Microtia repair

Atresia opening/repair

Hearing ✓

Amplification ✓

Early Intervention ✓

Medical Needs



Early Intervention-Topics

- Critical period of time for auditory input/language development/brain development
- Keep Up vs Catch Up
- Connect with other parents
- Therapists with M/A experience
- Acknowledgement of contradictory advice





- Family and strangers questions, comments, and stares
- Awareness of unique situations:
 - Car seats
 - highchairs & strollers
 - Hats and helmets
 - Thermometers
- Self-identity and psychosocial health

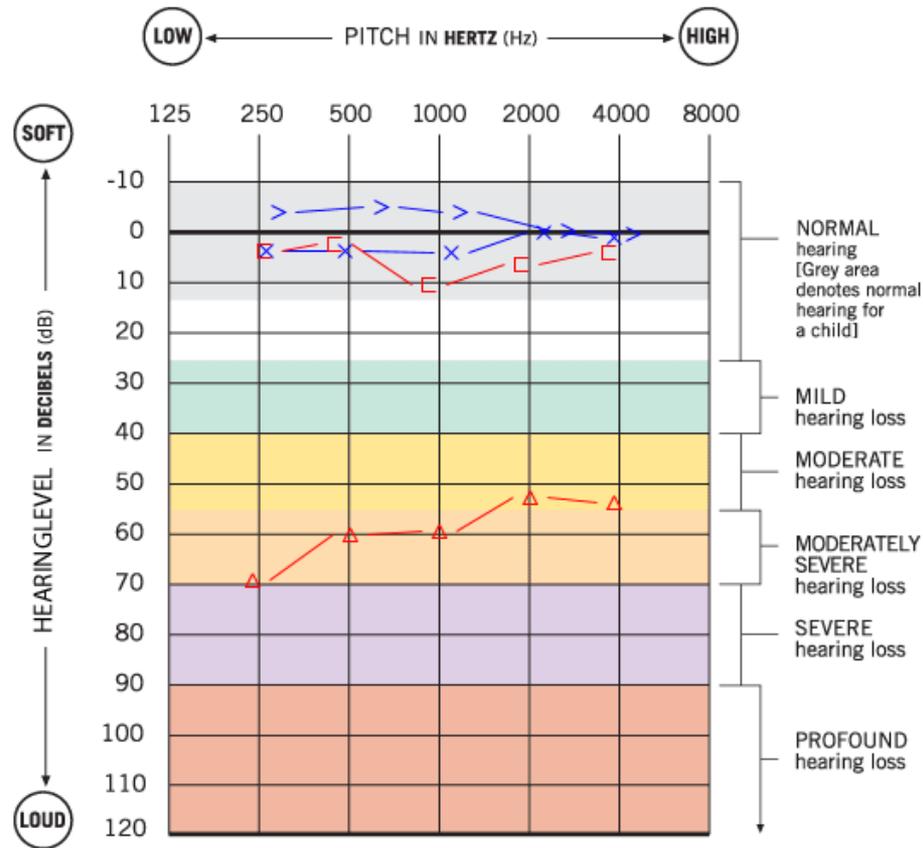


Audiology/Medical Topics

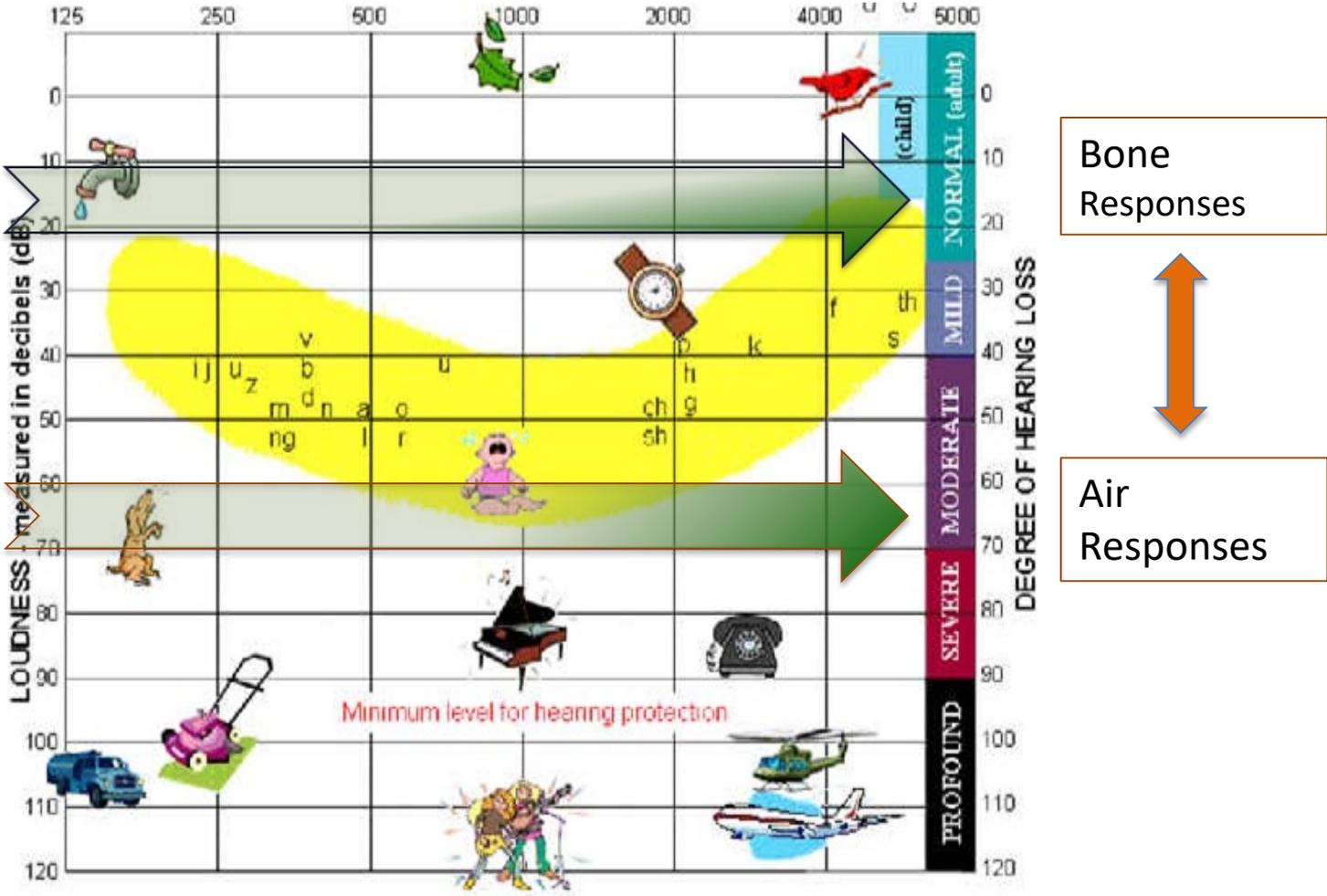
- Impact of unilateral hearing loss
- Air/Bone Gap- what does it really mean
- Surgical Timeline
- Canalplasty success/complications/age when surgery can occur
- Need for amplification post-surgery
- Higher likelihood of tube placement in the non-atretic ear than the general population (Billings et al, 2015)
- Incidence of velar palsy/velopharyngeal insufficiency in children with OAVS (Oculo-Auriculo-Vertebral-Spectrum) and children with isolated microtia. (Kolodzynski et al, 2018)

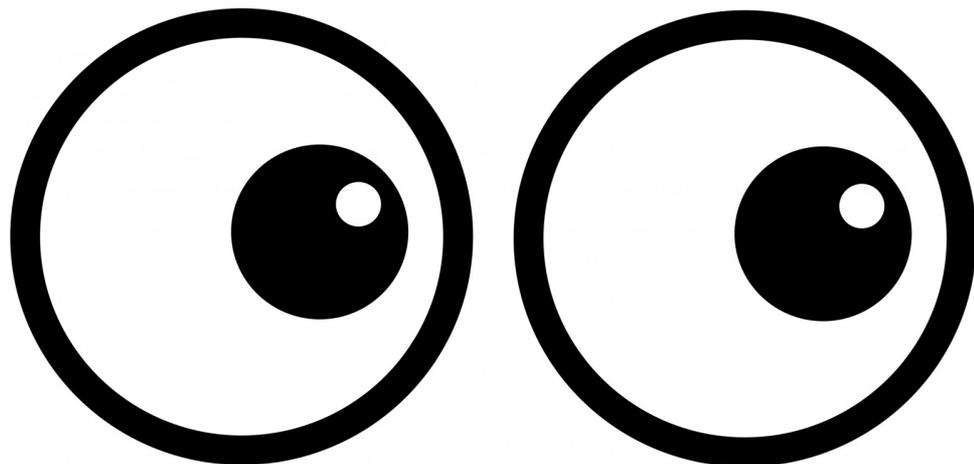
Air/bone gap= cochlear function

Parents hear “normal hearing”



Lost in the Gap





Research

Readability and Quality Assessment of Websites Related to Microtia and Aural Atresia (Alamoudi & Hong, 2015)

Objective: How readable are websites for parents?

- 30 sites analyzed based on google search
- Content:
 - Microtia sites (good)
 - Atresia Sites (fair)
- Readability
 - 10th grade reading level needed overall.
 - 1 microtia and 1 atresia site-“reasonable”



Parental Preferences for the First Consultation for Microtia van Hövell tot Westerflie et al (2018).

Purpose: Identify what information parents were presented with and what information they wished they had been provided

- 87 parents responded to the survey
 - ¼ report getting no information after their child was born
 - ¾ received diagnosis from pediatrician or otologist
 - Few had an audiologist as part of the initial team
 - Most parents describe the information/experience as “terrible”, or “bad”.
 - Parents want accurate and patient/family centered information and care



Effect Of Amplification On Speech And Language In Children With Aural Atresia.

Attaway, J., Stone, C. L., Sendor, C., & Rosario, E. R. (2015)

16 children

- 10 unilateral- 5 boys and 5 girls
- mean age 4.4 years (3-6 year span)
- 87% were Hispanic

Amplification

- Average age of first amplification- 26.5 mo (range 5.6 mo-6.11 years)
- Children who were fit earlier were more compliant users

Speech/Language

- Children with right side atresia and speech/language WNL were fit one year earlier with amplification than children with speech and language delays



Impact of Unilateral Conductive Hearing Loss Due to Aural Atresia on Academic Performance in Children

Bradley W. Kesser, MD; Kaelyn Krook, BS; Lincoln C. Gray, PhD *The Laryngoscope* VC
2013

Of the 40 atresia patients, none repeated a grade, but **65%** needed some resources:

- 12.5% currently use a hearing aid
- 32.5% use(d) a frequency-modulated system in school
- 47.5% had an Individualized Education Plan
- 45% utilized speech therapy

Qualitative study, survey, parents attending a conference



Effects of Aural Atresia on Speech Development & Learning

Retrospective Analysis From a Multidisciplinary Craniofacial Clinic

Jensen DR1, Grames LM, Lieu JE. JAMA Otolaryngol Head Neck Surg. 2013;139(8):797-802

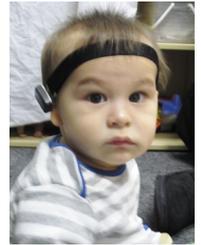
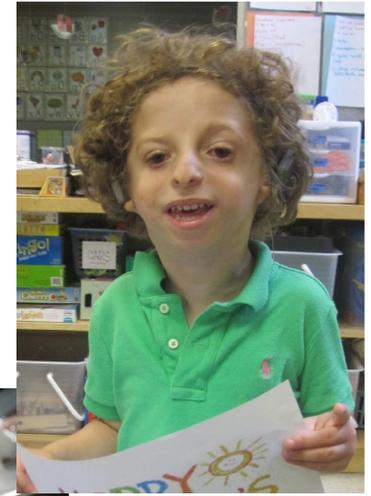
- N=74 patients PTA of 60dB in the affected ear across all groups
 - 48 right sided AA
 - 19 left sided AA
 - 7 bilateral AA All used amplification **WHAT KIND?**
- } **3/67 amplified. 1 FM & 2 SB**
- Speech Therapy: 86% among bilateral, 43% among unilateral
- Educational interventions : 33% right, 21%left, 43% bilateral
- School problems:
 - right (31%) left (11%) bilateral(0%)



Next Steps

1. Diversify the N's
2. Data collection
3. Better education and tools for professionals
4. Funding for amplification/therapy
5. Research! Research! Research!





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