He was the first to notice. Or, more likely, the first to say it. “There’s something up with her ears.”….Within minutes, Nxxx [husband] 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.dpuf




Children with Microtia/Atresia

An Introduction

CLARKE
SCHOOLS FOR HEARING & SPEECH

Participants will…

1	2	3
….discuss unique experiences of parents of children with microtia/atresia and the effect of those experiences in meeting their child's needs.	….will analyze research on supports and psychosocial needs of children with microtia/atresia and their families.	….identify strategies for supporting families of children with microtia atresia that align with the goals of JCIH and EHDI.

Annual Births

3.6 million babies born in the US annually

11,000-12,000 identified as deaf/hh

< 500 with microtia/atresia

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- Clarke School/New York
- PhD candidate
  - Teachers College, Columbia University
- Parent of 2
  - 1 with microtia/atresia

*disclosure-uses cochlear bahas

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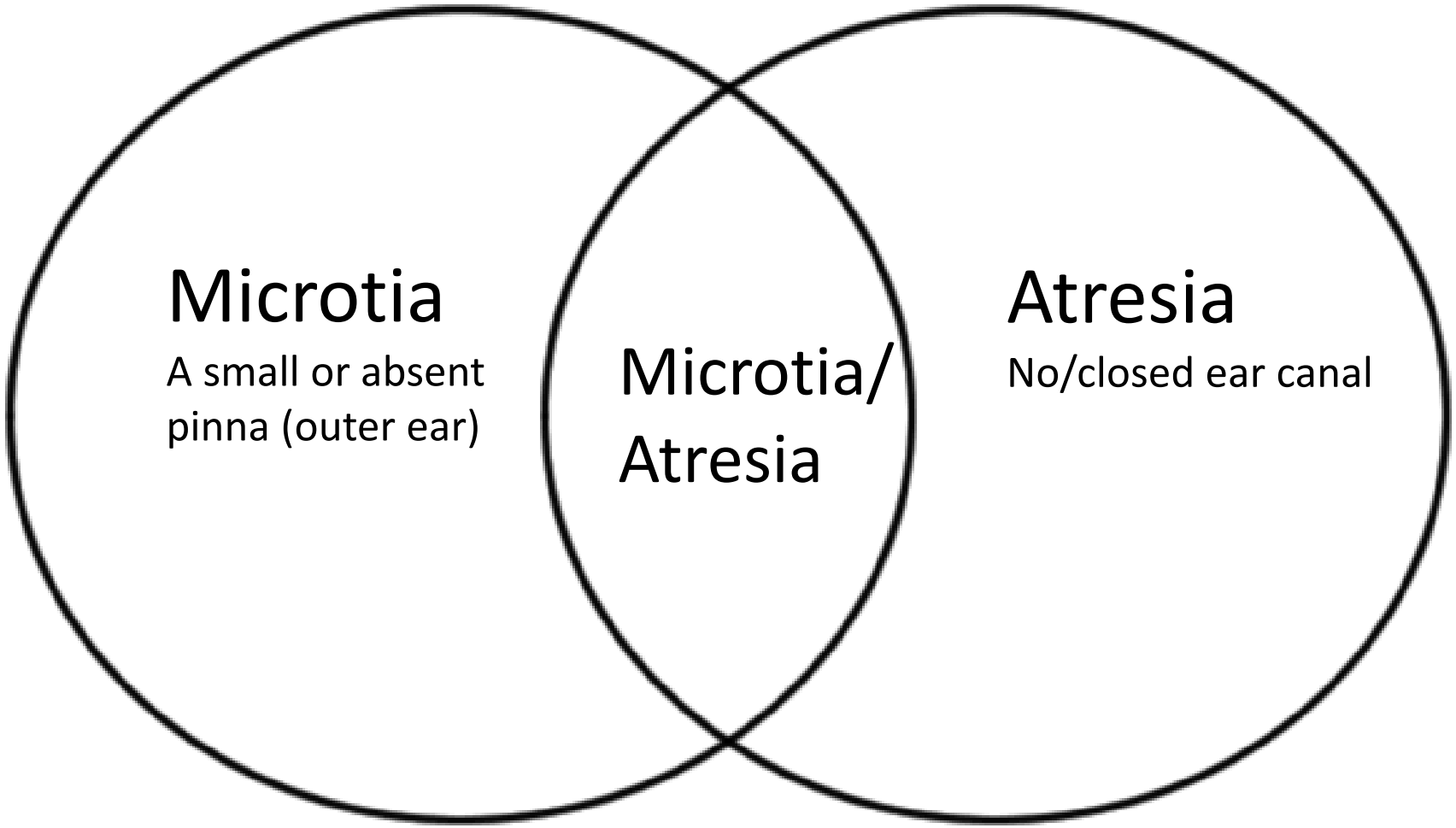
SCHOOLS FOR HEARING & SPEECH

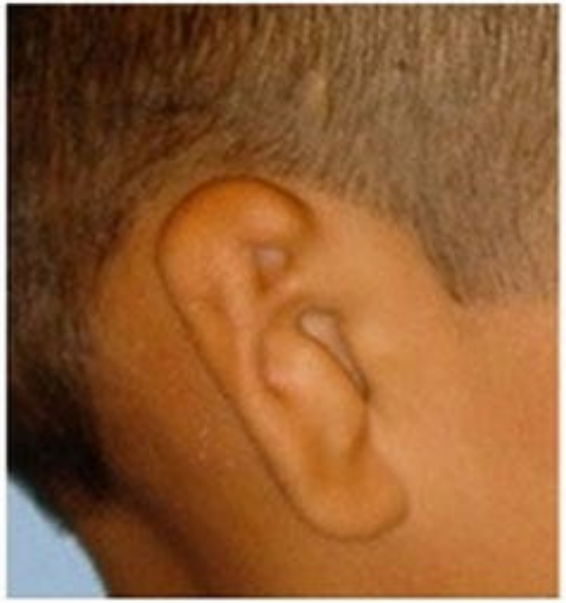
The basics

WHAT IS MICROTIA/ATRESIA?



Types

Grade I Microtia



Grade II Microtia

Grade III Microtia

Grade IV Microtia

What's in a name?

Microtia/Atresia

Aural Atresia

Unilateral Atresia

Unilateral Conductive Hearing Loss

Permanent Unilateral CHL

Atresia

Anotia

Single Sided Deaf (SSD)

Microtia only

Stenotic Canal

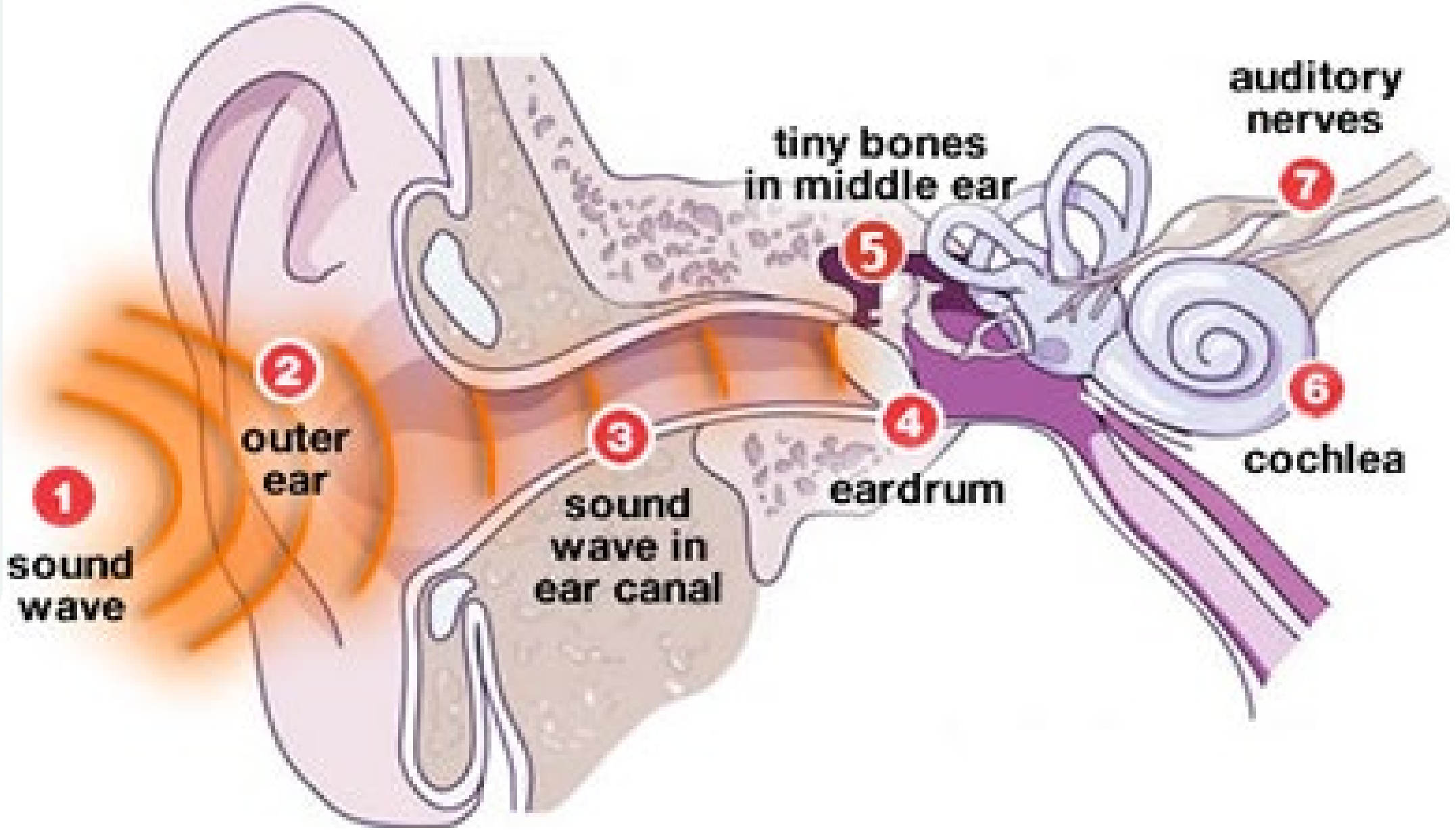
Transient Conductive loss

Quick Facts

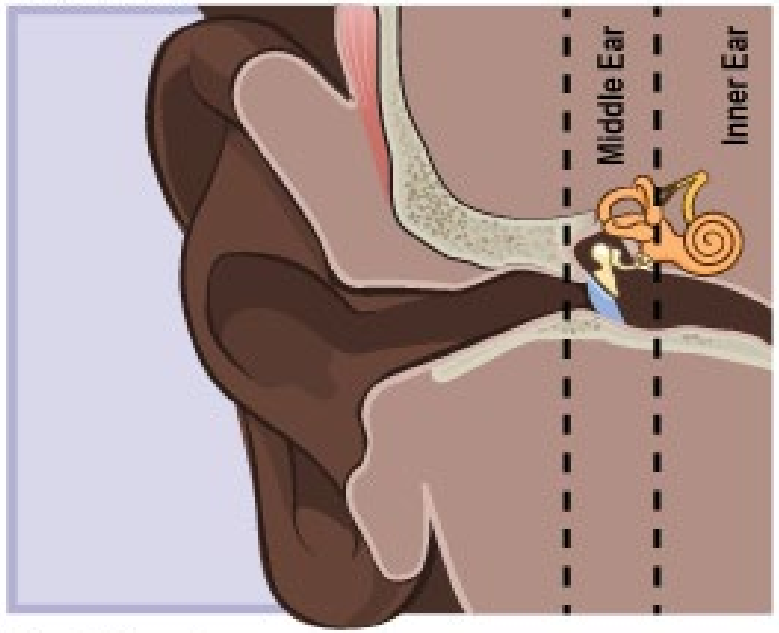
Estimates of 1/10,000 - 5/10,000
(Center for Disease Control and Prevention, 2018)

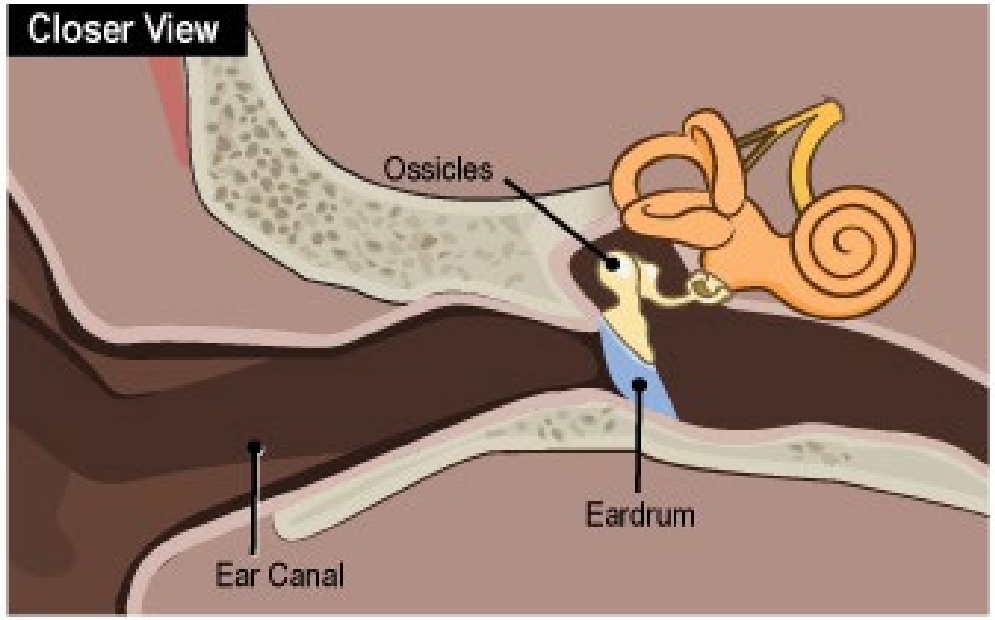
- Hispanic/Latino-7x higher
  - Ecuador 17/10,000 (Noroña et al., 2024)
- Asians-3x higher
- African/descent .86/10,000 (Shaw et al., 2004)

- More common in males
- unilateral (>90%), right ear (60%)
- Syndrome related -<20%
  - Oculo-Auriculo-Vertebral-Spectrum (OAVS), sometimes referred to as **hemifacial microsomia**
  - Craniofacial Microsomia and Goldenhar

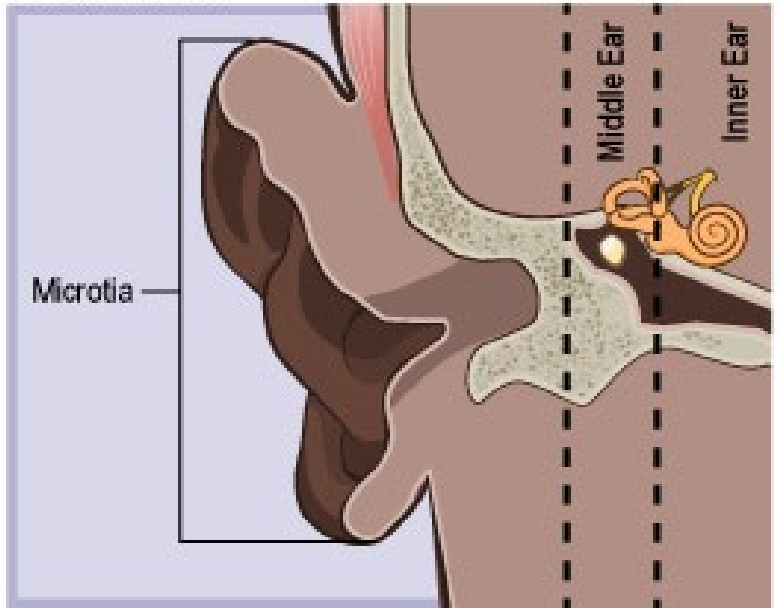

auditory nerves
tiny bones in middle ear
7
5
2
outer ear
3
sound wave in ear canal
4
eardrum
6
cochlea
1
sound wave

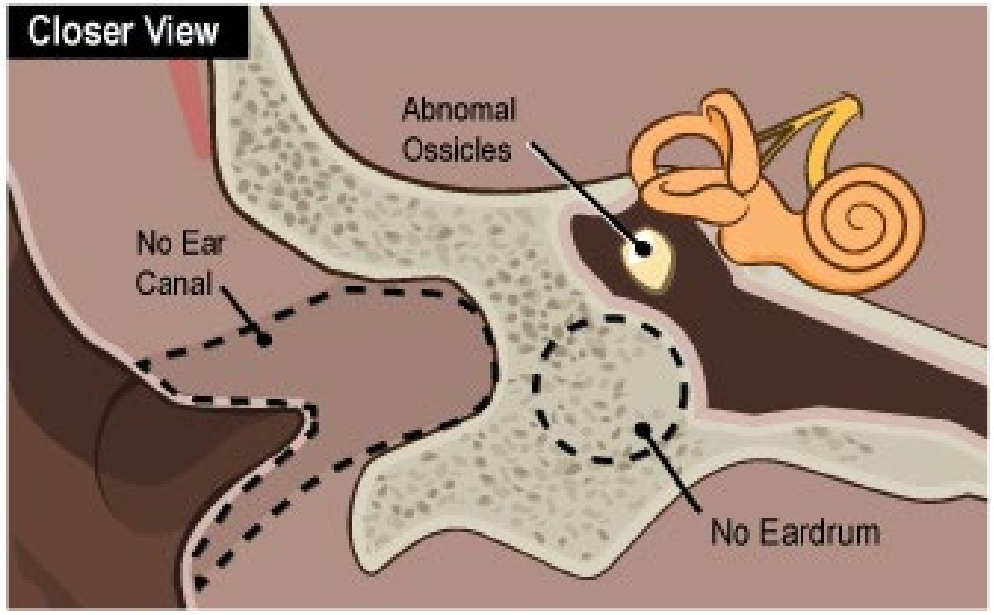
Normal Ear





Aural Atresia



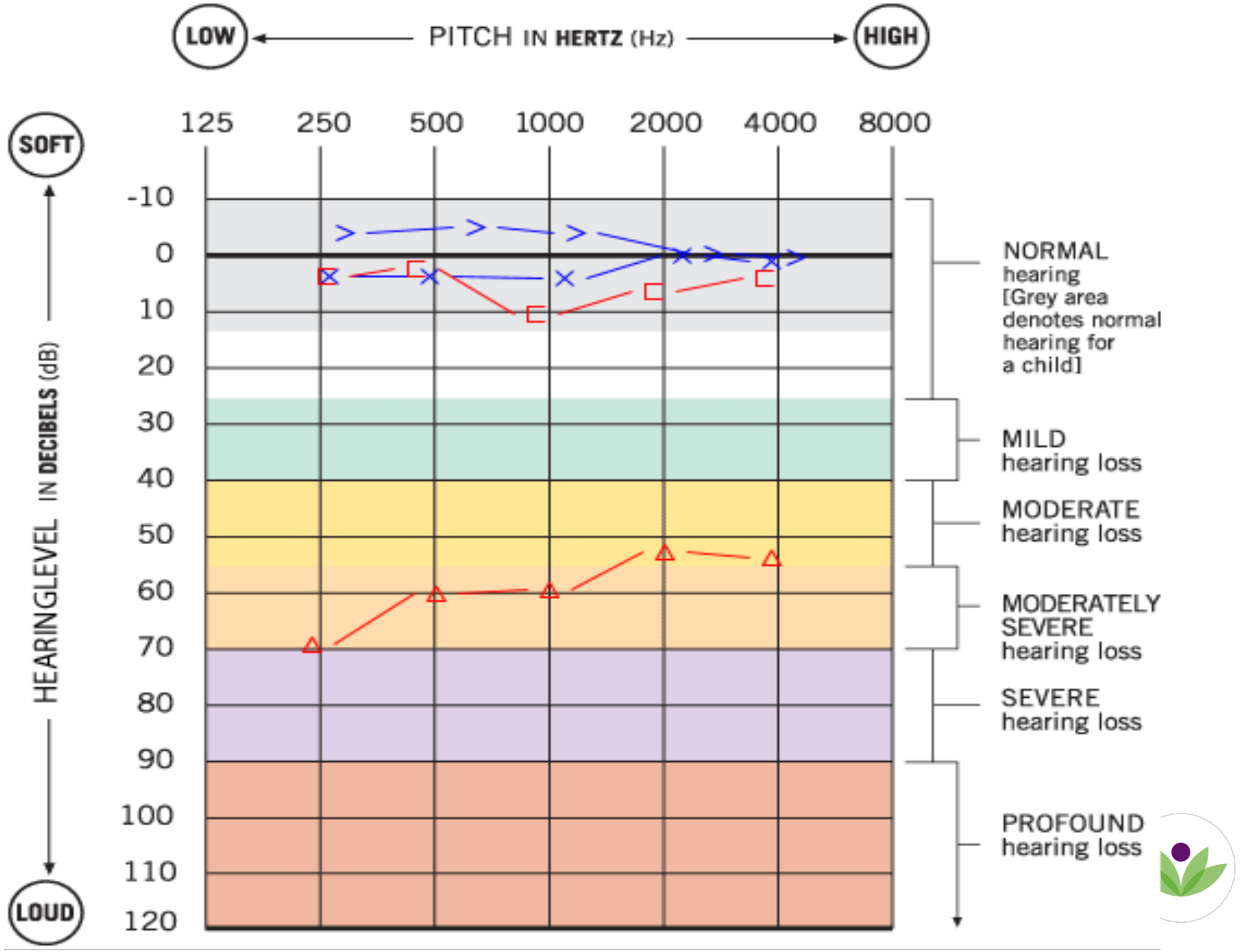


Maximum Conductive Loss- Air/bone gap= cochlear function

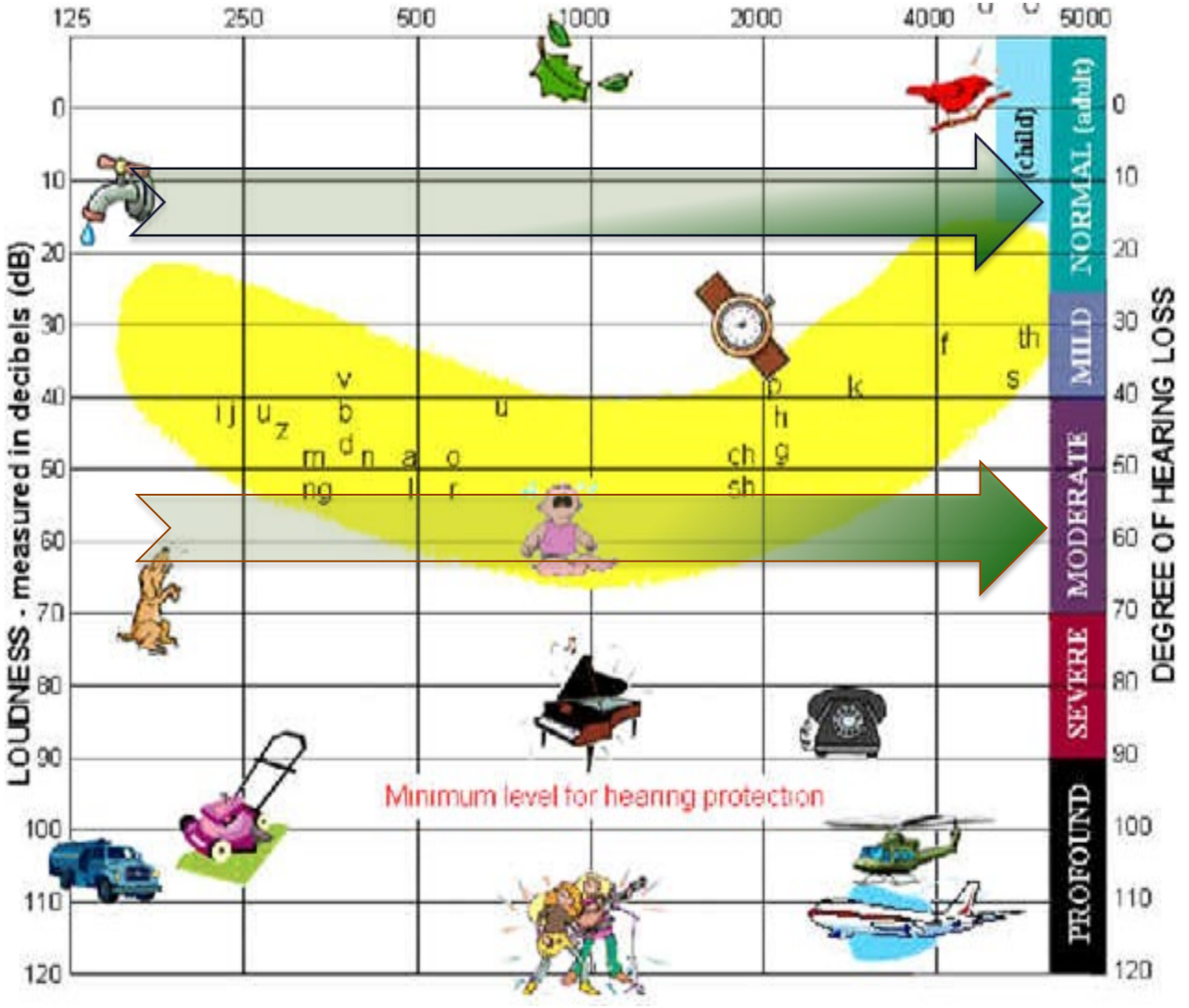
"Normal cochlea function"

≠

"normal hearing"



Lost in the Gap



Bone Responses

Air Responses

Microtia/Atresia 2014-2018

(National Birth Defects Prevention Network , 2022)

	White, Non-Hispanic	Black, Non-Hispanic	Hispanic	Asian or Pacific Islander Non-Hispanic	American Indian Alaska Native, Non-Hispanic	Total*
FL	58 1.2/10,000	28 1.2/10,000	48 1.5/10,000	<5	0	142 1.3/10,000
MA	46 2.2/10,000	7 2/10,000	28 4.2/10,000	9 2.8/10,000	0	93 2.6/10,000
NY	95 1.6/10,000	22 1.3/10,000	109 4/10,000	37 2.8/10,000	0	264 2.2/10,000
PA	Birth Defects Surveillance Program (PA-BDSP) doesn't collect data on anotia/microtia.					

Joint Committee on Infant Hearing (JCIH) re: microtia/atresia

1994	UNHS and 1:3:6 model endorsed
2000	UNHS target “permanent bilateral or unilateral, sensory or conductive hearing loss….”
	…any degree of bilateral or unilateral permanent hearing loss should be considered eligible for early intervention services.
2019	EI referral made by the birth hospital
	Diagnostic audiologic evaluation immediately upon discharge or can be done in NICU/hospital before discharge

EI services represent the purpose & goal of the entire EHDI process. Screening and confirmation that a child is D/HH are largely **meaningless** without appropriate, individualized, targeted & high-quality **intervention.**

(Joint Committee on Infant Hearing, 2013)



Options. (Reinisch and Lewin, 2009),

Age	Audiological	Ear Canal	Reconstruction	Therapy and support
Birth-3	ABR/Testing Amplification: Non-surgical (Baha, Ponto, Adhear			Early Intervention Language Development Parent Supports
>3 yrs			Medpor/Supor Reconstruction Atresiaplasty	
>5 yrs	Surgical bone conduction devices (BCD)	Atresiaplasty Canalplasty		
≳ 7 yrs			Rib Cartilage/Graft (3-4 surgeries per ear)	

EI Regulations

Part C of IDEA

Each state's definition of an infant or toddler with a disability **must** include an infant or toddler with….A diagnosed physical or mental condition with a high probability of resulting in developmental delay. (34 CFR § 303.21)

Florida

- Auditory Neuropathy
- **Aural Atresia (bilateral or unilateral)**
- Sensorineural hearing loss >25 dB HL

Massachusetts

- Hearing loss - permanent
- Bilateral, left ear, or right ear
- Neural hearing loss/auditory neuropathy

New York

- a diagnosed hearing loss that cannot be corrected with treatment or surgery
- *Clinical Guidelines supplement*
- *Cost of HAs/BCDs/HAT covered*

Pennsylvania

- State policy does not go beyond federal regulatory language.

Counseling Topics

- Effects of unilateral hearing loss
- Air/Bone Gap- what does it really mean
- Critical period of time for auditory input and language development
- Keep Up vs Catch Up
- Surgical Timeline
- Canalplasty success/complications/age when surgery can occur
- Need for amplification post-surgery
- Delay to Amplification Usage
- Higher incidence of middle ear issues in non-M/A ear. (Billings, 2015)

Considerations

Medical Surveillance

- Genetics
- Kidney
- Heart
- Vertebral anomalies
- Soft Palate
- Genital anomalies
- Syndromes

Other Needs

- Information access and accuracy
  - Craniofacial team
- Isolation
- Bullying
- Psychosocial Needs
- Surgical questions
- Parenting challenges
  - Hats, glasses, lost devices

Soft Palate and VPI

Soft Palate Dysfunction in Children With Microtia.

(van Hövell tot Westerflier et al., 2019)

Prospective study- 67 children

Unilateral (n=40) results

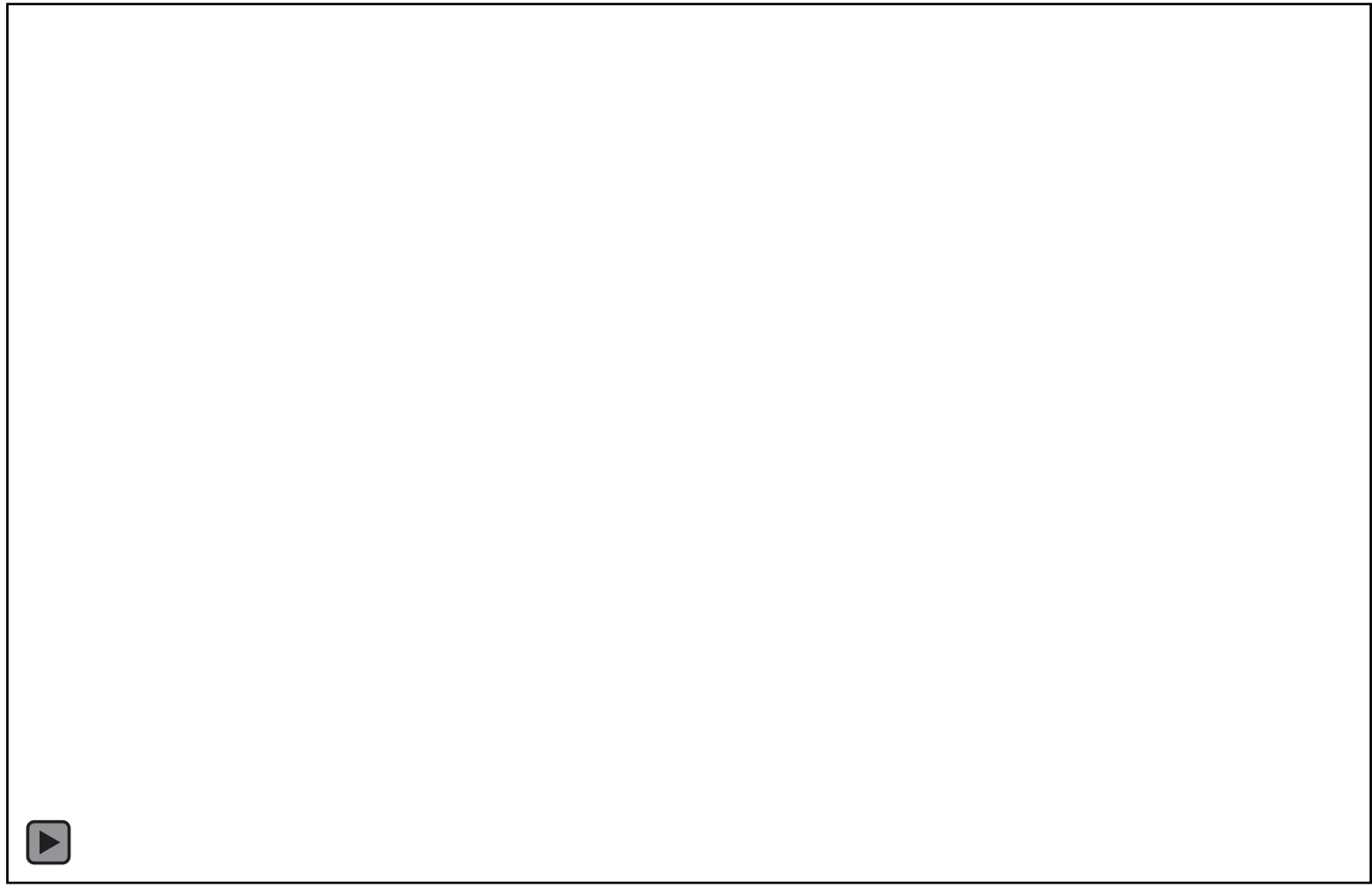
- VPI-60%
- Uvular Deviation- 95%

Bilateral (n=27) results

- VPI- 85%
- Uvular Deviation-
- 59%- almost no movement
- 40%-deviation

Recommendation:

Children with isolated and non-isolated microtia should have in depth evaluation of soft palate movement and VPI



Clinical consensus document for fitting non-surgical transcutaneous bone conduction hearing devices to children. (Bagatto et al., 2021)

Purpose: Address the assessment, selection, and fitting for non-surgical bone conduction devices for children under age 5

- Goal is to fit within 1 month of diagnosis
- Children as young as 2 months
- Bilateral m/a=bilateral BCDs (sequential, based on age)

Psychosocial Needs

infants > toddlers > children> tweens> teens> adults

What helps?

- ✧Connect with other parents
- ✧Therapists with M/A experience
- ✧Acknowledge contradictory advice
- ✧Family and strangers' questions, comments, and stares
- ✧Awareness of unique situations

Car seats, highchairs, strollers

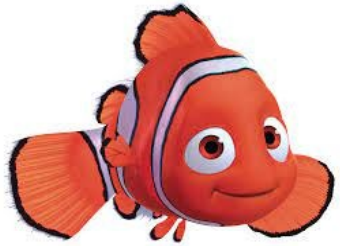
Hats and helmets

Thermometers

Masks

Banz (on amazon)

Paradigm Shift



Resources

The Ear Community Facebook, website, Ear Community picnics

Facebook groups Microtia Parents & others specific to countries/regions

myFace myFace.org

Little Baby Face Foundation https://www.littlebabyface.org/

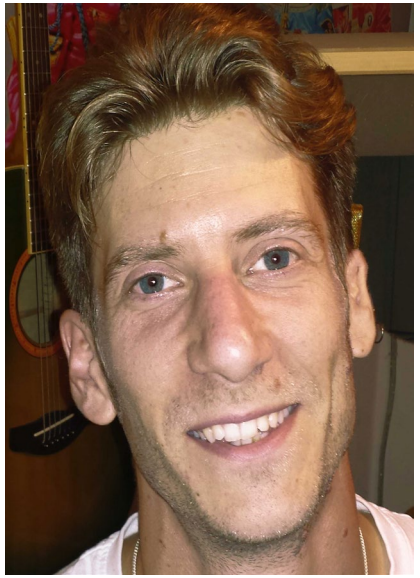
Hearing First Sophie's Story https://www.hearingfirst.org/celebrate-lsl/lsl-life/sophie

Give an Ear Foundation (South Africa based/Africa focused)

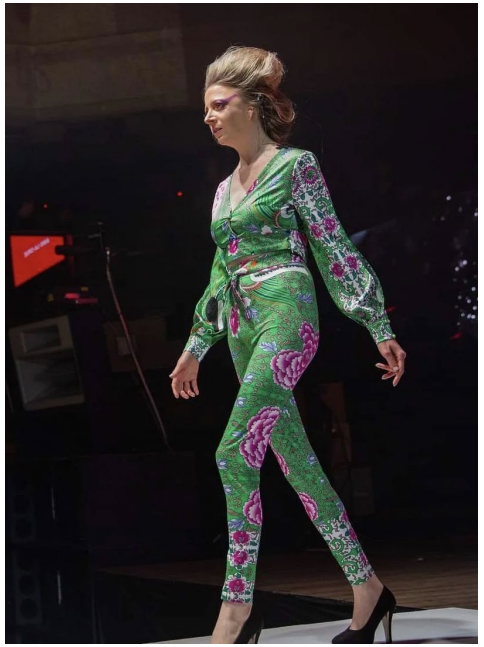
https://giveanearfoundation.org

Microtia UK http://microtiauk.org/

Microtia Atresia Australia https://microtiaatresia.com.au/



Jono Lancaster	Advocate *love me, love my face*
Camilla Gilbert	Motivational speaker
Gideon Glick	Actor
Nick John Wilson	Singer/song writer
Cynthia Murphy	Model/Advocate
Paul Stanley	KISS drummer
Salia Baligh	2023 Rose Parade Princess



Books

Jojo’s Tiny Ears

Stefania Munzi-Logus

Hi, I’m Me

Hi, I’m Me in Kindergarten

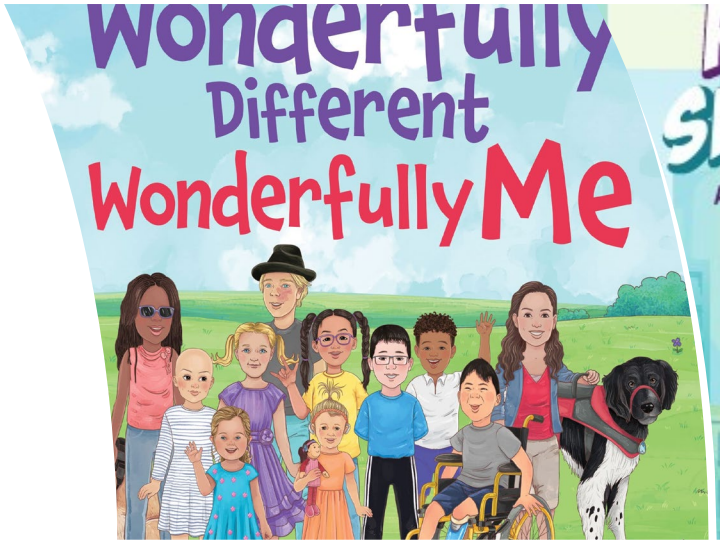
Kelly Vurinaris

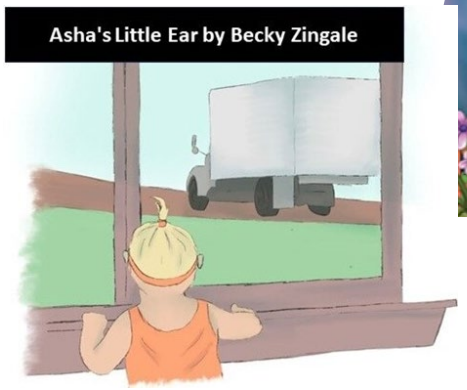
Ricky the Rock That Couldn’t Roll Mr. Jay

Rebekah's Superpower Emma Bilyk

Wonderfully Different, Wonderfully Me

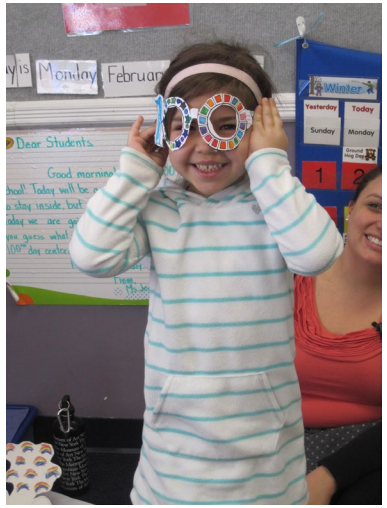
Becky Zingale





Priscila Soares

- BCD user (not m/a)
- Artist/illustrator
- Myluckyears (Instagram)
- Myluckyears.com


Dear Students
Winter
Yesterday
Today
Sunday
Monday

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