

Navigating Pediatric Audiology with 1-3-6

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Kids deserve the best.

Outline

- Learning Objectives
- Program Overview
- Current Program Goals
- Case Studies
- Research



Key Terms

- Auditory Evoked Potential (AEP)
- Automated Auditory Brainstem Response (AABR) screening
- Auditory Brainstem Response (ABR) screening/evaluation
- Children's Hospital of Wisconsin (CHW)
- Distortion Product Otoacoustic Emissions (DPOAEs)

Key Terms

- Early Hearing Detection and Intervention (EHDI)
- Ear Nose and Throat (ENT)
- Joint Committee on Infant Hearing (JCIH)
- Universal Newborn Hearing Screening (UNHS)
- Wisconsin EHDI-Tracking Referral and Coordination (WE-TRAC)

Learning Objectives

- Understand the JCIH 1-3-6 recommendations and how they are applied at Children's Hospital of Wisconsin through specific case examples.
- Evaluate a hospital based EHDI program and identify barriers to meeting the JCIH 1-3-6 goals.
- Develop strategies to implement a system of management and more effective programs for Early Hearing Detection and Intervention.

Why we exist

- Nothing matters more than our children. At Children's Hospital of Wisconsin, we believe kids deserve the best. Our belief drives everything we do. It defines our vision for the future and our mission for today.



Our Vision at CHW

- Wisconsin kids will be the healthiest in the nation.



UNHS Program at CHW Mission

Our Mission

- Our mission (HEAR) is the work we do to achieve our vision:
- **H**elping children hear their best
Offering the nation's best Audiologic and Early Intervention care
- **E**ducation
Maintaining and advancing as a leader in the field of Audiology and Early Intervention by making education and interdisciplinary collaboration of our professionals a priority
- **A**dvocacy
Working with and for families to ensure that all options are given and resources and guidance is provided in order for families and patients to be their best advocates
- **R**esearch
Striving to answer questions and finding ways to improve systems within our department. Collaborating with community partners to better the overall goal of providing children with timely and effective care

EHDI and UNHS at CHW Goals:

- 1) 100% of newborns are screened for a hearing difference before discharge at CHW
- 2) 100% of newborns who do not pass the hospital screening are re-screened by **1 month of age**
- 3) 100% of newborns who refer from the re-screen to an Audiologist receive a diagnostic exam **by 3 months of age**
- 4) 100% of newborns diagnosed with a hearing difference enter an early intervention program (i.e. Birth to Three) by **6 months of age** and receive appropriate amplification, if applicable

UNHS Program Overview



Current State of UNHS at CHW

Total Number of Babies on CHW UNHS WE-TRAC Queues



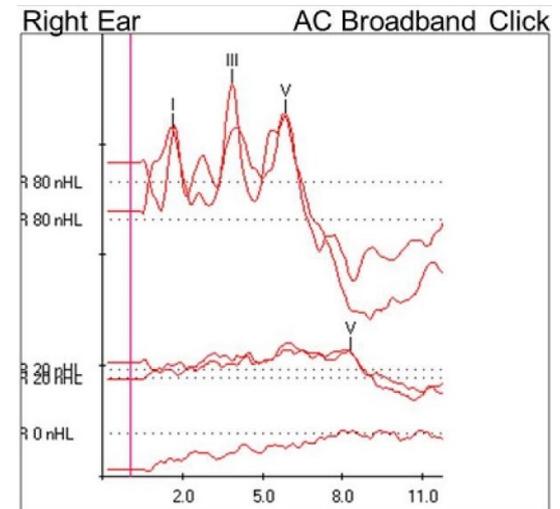
What is a “passed” UNHS at CHW?

Passed AABR in both ears via Algo3i:



What is a “passed” UNHS at CHW?

ABR Click screening at 35 dB HL in each ear via Interacoustics Eclipse system:



UNHS Outpatient Queue

Check EPIC (Medical Records), UNHS data base & Outpatient Queue

Scheduled?
No

Scheduled ?
Yes

Connect with family?
No

Contact Family 3 times

Connect with Family?
Yes

Refer to Wisconsin Sound Beginnings for LTFU prevention

Willing to schedule
Yes

Willing to schedule?
No

ABR if needed

3 way call to Central Scheduling

- Provide Patient & Family Education
- Identify Barriers Connect to Resources

Arrives?
No

Arrives?
Yes

Diagnosed as Deaf/Hard of Hearing

Normal hearing, bilaterally

Birth to Three referral placed and FU appointments made

Documented into WE-TRAC, UNHS Database & EPIC

UNHS Inpatient Queue

Check EPIC (Medical Records), UNHS data base & Outpatient Queue

Hearing screen complete?
No

Hearing screen complete?
Yes

Request NICU nurse to complete ALGO screening

Refer result for one or both ears?

Pass result for both ears?

Documented into WE-TRAC, UNHS Database & EPIC
Case temporarily closed

Audiology consult placed

Screening at 35dBHL using Interacoustics Eclipse equipment

Pass result for both ears?

Refer result for one or both ears?

Diagnostic ABR

Normal hearing, bilaterally

Diagnosed as Deaf/Hard of Hearing

Birth to Three referral placed and FU appointments made

- Provide Patient & Family Education
- Identify Barriers Connect to Resources

Audiologic Framework



Risk Factors for Hearing Loss

- Congenital heart defects/cardiac surgery
- Family history of childhood hearing loss
- Syndromes associated with hearing loss (Usher, Waardenburg, Pendred, Neurofibromatosis, etc.)
- Craniofacial abnormalities of the head, face, ears, or neck (cleft lip/palate, ear pits/tags, atresia, microtia, etc.)
- Maternal infections during pregnancy or delivery: Toxoplasmosis, Syphilis, HIV, Hepatitis B, Rubella, CMV, Herpes simplex (TORCH), Zika virus
- Postnatal infections such as viral or bacterial meningitis

Risk Factors for Hearing Loss

- Born premature (less than 37 weeks gestation)
- Neonatal Intensive Care Unit (NICU) stay greater than 5 days
- Elevated hyperbilirubinemia
- Blood transfusion
- Required mechanical ventilation of 5 days or longer: Extracorporeal Membrane Oxygenation (ECMO), Continuous Positive Airway Pressure (CPAP), and/or High-frequency Oscillatory (HFO) Therapy
- Traumatic Brain Injury (TBI)
- Ototoxic medications

Test Battery

- Cursory Otoscopy
- 1000 Hz/226 Hz Tympanometry
- Acoustic Reflexes
- Behavioral Audiometry
- Distortion Product Otoacoustic Emissions (DPOAEs)*
- Auditory Brainstem Response (ABR)



Children's Hospital of Wisconsin: Case Studies

Universal Newborn Hearing
Screening in a Hospital Setting

Case 1:

- Born at 31 weeks gestation
- 7 month NICU stay
- Congenital syphilis
- Ventilator dependence
- Followed by ENT for:
 - Vocal cord granuloma
 - Tracheostomy dependent

Case 1: Audiologic History

- 8/3/2017: Audiology consult ordered while inpatient over weekend. Patient discharged before audiology team able to see the patient in the NICU.
 - Outpatient follow up recommended, however did not follow up.
- 8/11/17: Mom contacted regarding need for diagnostic hearing test, sedated ABR intake completed.
- No showed audiology appointments:
 - 7/17/18
 - 12/21/19
 - 2/11/19
- Case sent to social work at CHW and to the Wisconsin Sound Beginnings Program for follow up efforts



Case: 1

<i>Early congenital syphilis</i>	<i>Late congenital syphilis</i>
<ul style="list-style-type: none">• Presentation before 2 years of age• Prematurity and intrauterine growth retardation• Hepatosplenomegaly• Nasal chondritis ("snuffles")• Skin rash• Osteochondritis• Neurologic symptoms and signs, including hydrocephalus and cranial nerve palsies	<ul style="list-style-type: none">• Presentation after 2 years of age• Craniofacial malformation• Dental abnormalities• Interstitial keratitis• Deafness• Neurosyphilis• Paroxysmal cold hemoglobinuria

Case 1:

- Barriers to care:
 - UNHS not completed prior to discharge due to transfer of hospitals
 - Inpatient communication with Audiology team
 - No showed multiple appointments
- Victories:
 - Continuing to follow up with mom
 - Enlisted other assistance (social work, Wisconsin Sound Beginnings)

Case 2:

- Born at 37 weeks gestation
- NICU stay of 4 months
- CHARGE syndrome
- Bilateral microtia Type 2, atresia
- Bilateral conductive hearing loss
- Bicuspid aortic valve
- Chronic lung disease
- Dysphagia G
- Global developmental delay
- Gastrostomy tube dependent
- Foster care

CHARGE Syndrome Diagnostic Criteria

Table 1: CHARGE syndrome diagnostic criteria⁵³

Major Criterion	Minor Criterion
+Coloboma	+ Heart defect
+Choanal atresia	– Orofacial cleft
+Characteristic ear anomalies	+ Genital hypoplasia
+Cranial nerve dysfunction (facial palsy, vestibular dysfunction, swallowing difficulties)	+ Growth deficiency
	+ Developmental delay
	– Tracheo-esophageal fistula
	+ Distinct facial appearance

Note:—CHARGE indicates Coloboma, Heart defects, choanal Atresia, mental Retardation, Genitourinary, and Ear anomalies; +, pertinent positive finding; –, pertinent negative finding. A CHARGE diagnosis is indicated by 4 major criteria or 3 major and 3 minor criteria. Exclude other conditions such as velocardiofacial syndrome and DiGeorge sequence using FISH test to exclude 22q11 deletion.

Case 2: Audiologic History

- Never received UNHS due to bilateral microtia and atresia
 - Followed through outpatient Audiology clinic
- 11/9/18: (19 months old) Sedated ABR
- 12/14/18: (20 months old) Fit with unilateral amplification with loaner device
- Fit with bilateral amplification – SOON!

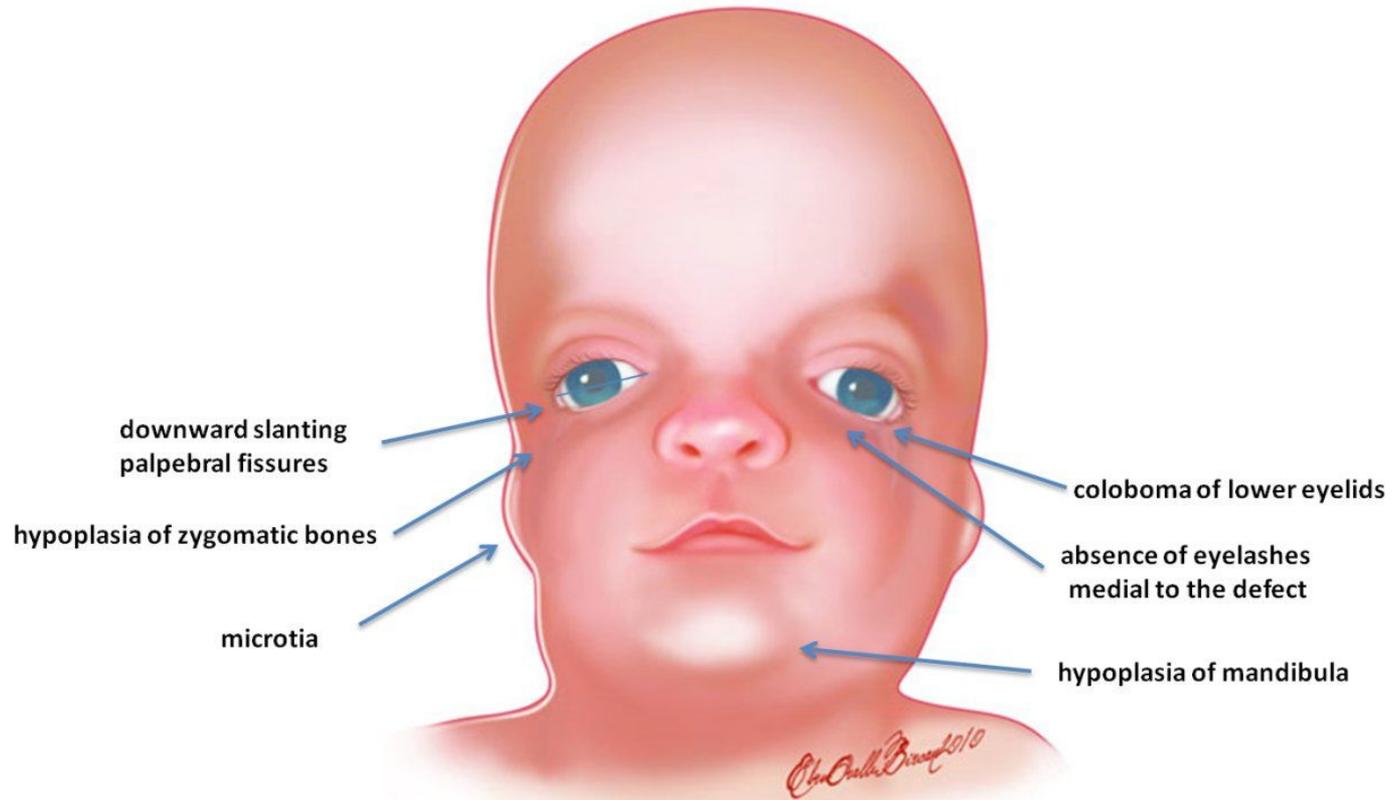
Case 2:

- Barrier to care:
 - Child in foster care, consent from mom was difficult to obtain.
- Victories:
 - Utilizing Sound of Hope loaner BAHA hearing aid
 - Sound of Hope: Loaner devices are for patients that need temporary assistance with hearing aids while at CHW.

Case 3:

- Born at 38 weeks gestation
- NICU stay of 2 months
- Treacher Collins syndrome
- Congenital micrognathia
- Bilateral microtia grade III/atresia
- Tracheostomy dependent
- Choanal stenosis

Case: 3



Case: 3

Spectrum of Microtia Severity

Least Severe  Most Severe



The ear is smaller but still looks like an ear because most normal features are present



Some normal features are present but the upper ear is severely deficient. The canal may be present or absent.



A small piece of cartilage is present just above the ear lobe which is displaced upward and forward. The canal is almost always absent.



Anotia is when there is a complete absence of the ear and canal.

Case 3: Audiologic History

- 9/14/18: (21 days old) Inpatient Natural Sleep ABR:
 - Maximum conductive hearing loss, bilaterally with normal hearing sensitivity from 500-4000Hz at the level of the cochlea in at least the better ear.
- 9/28/18: (35 days old) Inpatient unilateral BAHA fitting via Sound of Hope BAHA 5 device
- 10/10/18 & 10/12/18: Inpatient Hearing Aid Follow Up
- 11/30/2018: (3 months old) Hearing Aid Consultation
- Bilateral BAHA Fitting to be scheduled **SOON!**

Case 3:

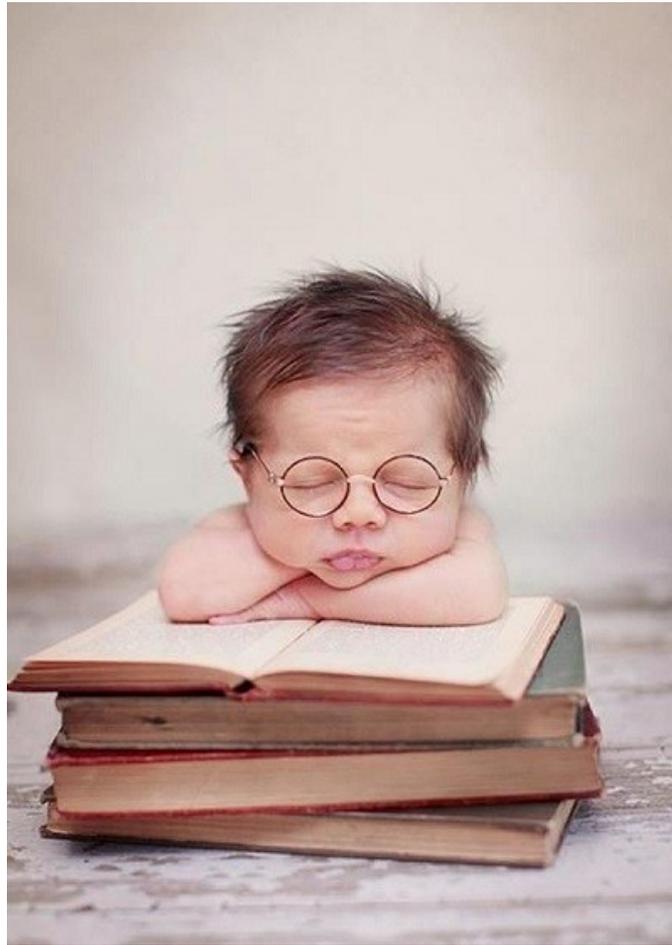


- Barrier to care:
 - Would not be able to achieve JCIH 1-3-6 guidelines with out inpatient diagnostic, fitting and follow up appointments.
 - Education of inpatient providers
- Victories:
 - First inpatient hearing aid fitting and follow up appointments.
 - Exciting step for CHW!

Current UNHS Inpatient Program Process

- Educating medical residents on UNHS
- Interdepartmental presentations
- Training NICU staff on AABR and appropriate referral
- Fit 3 patients with BAHA devices while inpatient
 - Many more to come!

Research Directions

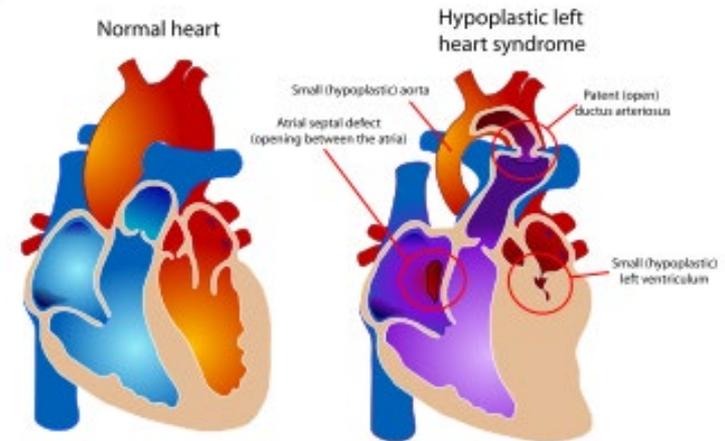


Research Directions

- Identifying barriers to care in achieving the 1-3-6 guidelines of WE-TRAC at CHW:
 - One of which is implementing a successful inpatient Audiology program to:
 - Screen, diagnose, and fit amplification in accordance to the JCIH 1-3-6 guidelines, regardless of length of hospital stay.



Research Directions



- Partnering with the Herma Heart Institute (HHI) to identify the following:
 - What is the prevalence of children with cardiac anomalies at CHW that do not pass their UNHS?
 - What is the prevalence of these children that are diagnosed with a hearing loss?
 - When were they diagnosed?

Research Directions

- What is the ototoxic medication exposure in children with cardiac anomalies?

OTOTOXICITY



There are more than 200 medications (prescription and over-the-counter) on the market today that are known to be ototoxic – which, quite literally, means **“poisonous to the ears”**.

Questions?



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THANK YOU

**COMPANIONS FOR
YOUR ATTENTION**

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Let's Connect!

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