Partnership between Parents, Academic Medicine, and EHDI Improves the Care of Children At Risk of Becoming Deaf or Hard of Hearing

Presented by:

Dr. Liza Creel, University of Louisville School of Public Health and Information Sciences

Cathy Lester, KY EHDI program, Office for Children with Special Health Care Needs

Dr. Judy Ann Theriot, KY AAP Chapter Champion

Roles and Responsibilities of EHDI Program

- Monitoring and surveillance for all refers and risk factors
- Monitoring of NBHS programs- screening procedures and data
- Tracking of diagnostic evaluations to monitor compliance with “3” of “1-3-6”
- Follow infants with PCHL until enrolled in early intervention*

History of KY EHDI

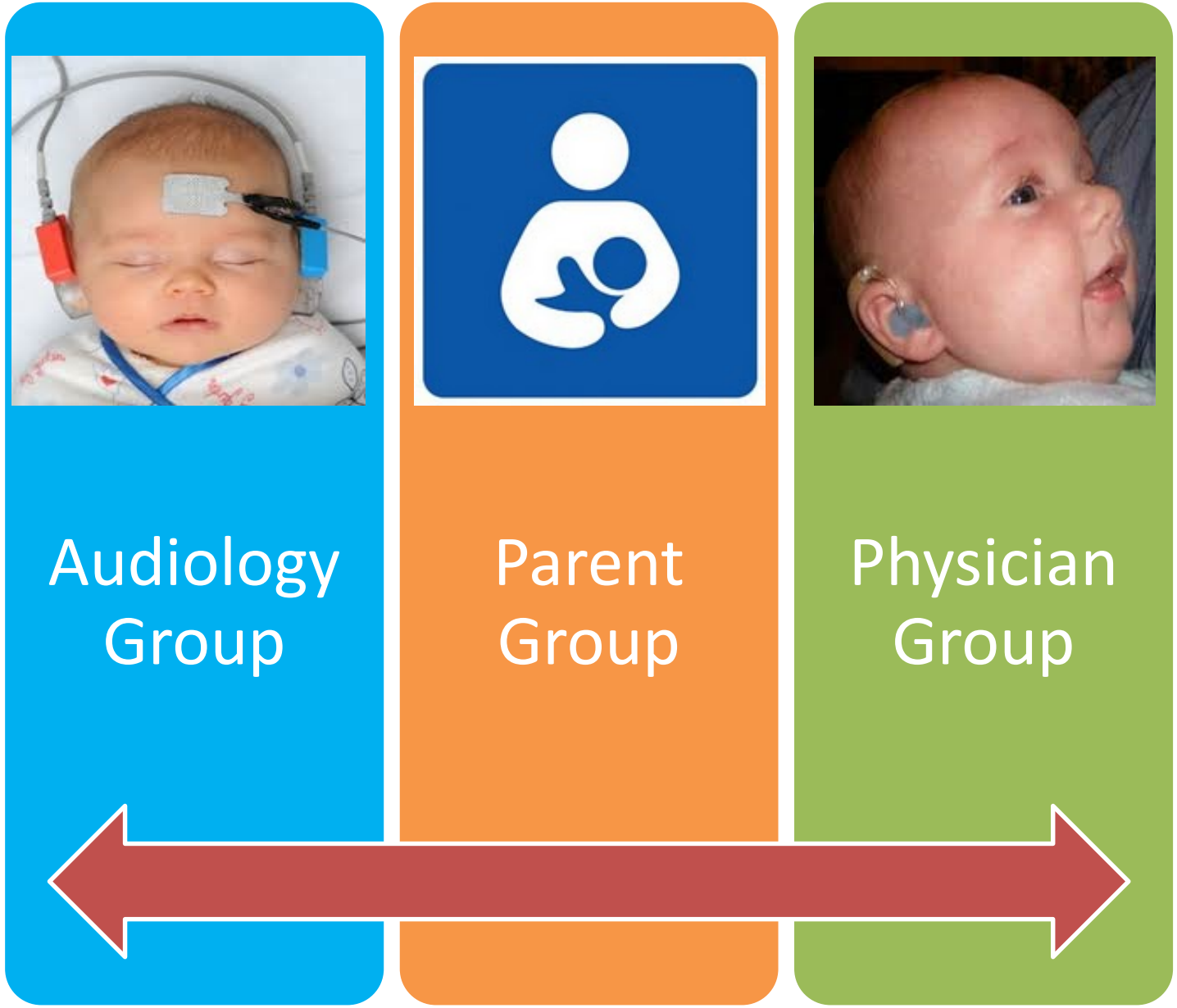
- January 2001: Implementation of Universal NBHS
  - 52% screened prior to this … 92% screened now!
- February 2001: EHDI sending letter to families when follow-up testing is needed
- 2009: Audiologists submitting evaluation results to EHDI electronically through KY-CHILD
- 2015: Physicians receive letters from EHDI for all NBHS referrals and all infants identified with PCHL
- 2018: Physicians receive letters from EHDI on all babies that pass UNHS but have risk factors for late onset/progressive hearing loss

KY EHDI Advisory Board

- Parents of children who are DHH
- Audiologists and SLP's
- Pediatricians, neonatologists, ENT's
- Epidemiologist/Public Health
- Hospital screening program representatives
- State stakeholder organizations
  - Hands & Voices, KCDHH, DBHID, First steps/Part C
- KY EHDI staff from OCSHN (advisory role only)

Working Meetings

- Meeting agenda
  - Initial discussion/reports
  - Work group breakout (Parents, audiology, medical)
  - Return & report out
- Groups were working on similar projects so they combined into a provider group and a parent group

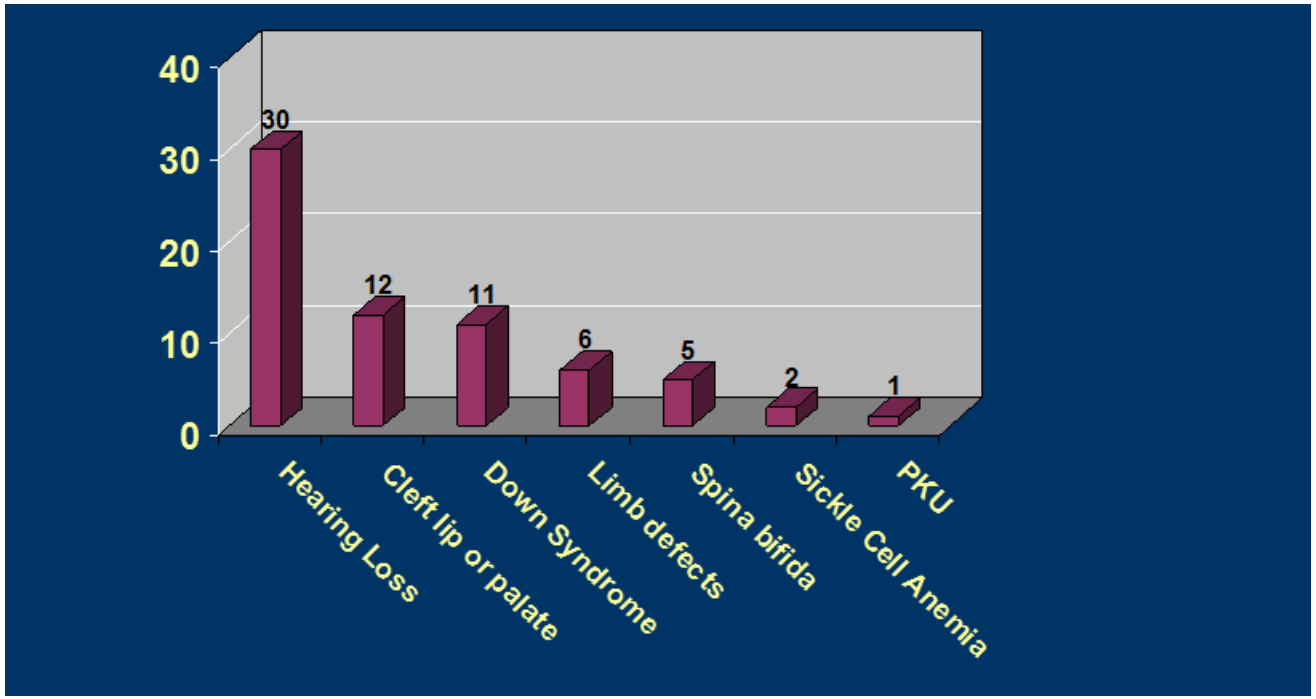



Education and Outreach
Data Collection and Analysis
Community Engagement and Linkages
Quality Improvement
Early Intervention
EHDI Advisory Board

KY EHDI Advisory Board Activities

- Quality Improvement
  - Learning Communities
  - Risk factor protocol
- Community Engagement
  - Hands & Voices
  - Guide by your Side
- Early Intervention
  - Unilateral hearing loss as a qualifier
- Education & Outreach
  - Parent groups
  - Physicians
  - students
- Data Collection
  - Surveys
  - Research studies

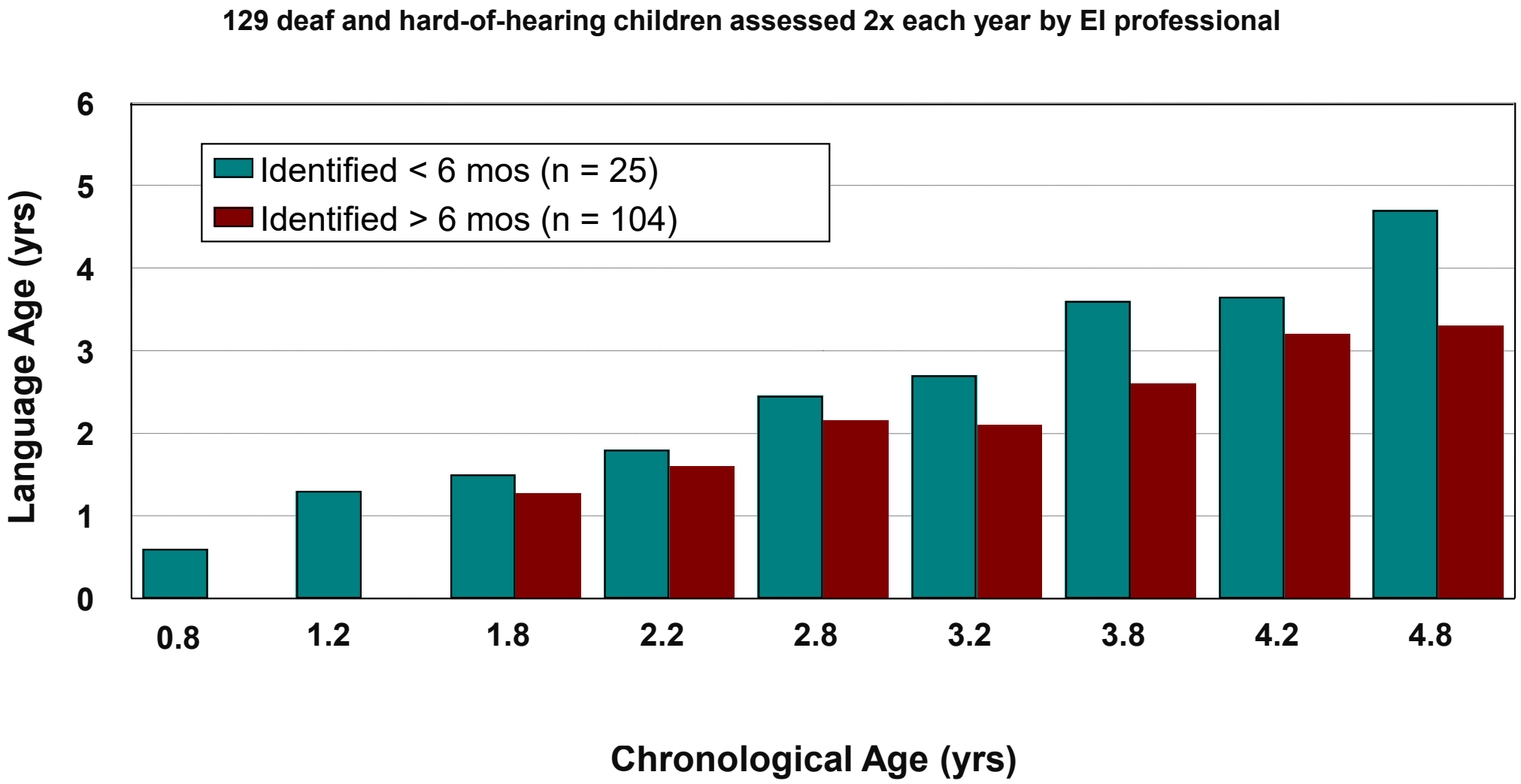
PCHL: The Most Common Congenital Disability/Birth Defect



Why is Early Identification of PCHL So Important?

- Congenital hearing loss can interfere with the most basic human need – COMMUNICATION
  - Narrow window of brain development for spoken language
  - Non-signing parents must learn sign quickly to serve as language models if this is the chosen communication modality
- Undetected hearing loss has life-long negative consequences
- Newborns identified with PCHL can receive intervention services immediately if desired!
- Intervention by 6 months boosts language development to levels comparable to typically hearing peers
- Cost of education is reduced; lifetime achievement and earning potential are increased

Boys Town National Research Hospital Study: Earlier vs. Later ID



Long Term Implications of PCHL Without Intervention

- **Any** degree of hearing loss can have an affect on a child's speech and language development, social/emotional development, self-image, and academic success
- Children with mild to moderate hearing loss miss up to 50% of classroom discussions
- 37% of children with minimal hearing loss fail at least one grade
- Children with unilateral hearing loss are 10 times more likely to repeat a grade than their typically hearing peers
- Cost to society for child with HL: Over $1M, with $433K due to loss of productivity

Risk Factors for Permanent Childhood Hearing Loss (PCHL)

- Certain conditions can cause increased risk for progressive and/or delayed onset hearing loss
  - JCIH specifies risk factors
- KY EHDI program captures risk factors for individual infants in order to provide resources to these families, in the event that there is concern about late onset/progressive hearing loss after passing NBHS

Risk Factor Collection System

- KY CHILD is the electronic system whereby hearing screening results and risk factors are collected by hospitals
- KY CHILD is “Kentucky Certificate of Birth, Hearing, Immunizations and Lab Data”
- Hospital staff enter all relevant data, and this information is transferred electronically each night to the EHDI program

Risk Factors Captured in KY-CHILD

INFANT HAD BILIRUBIN LEVEL EQUAL TO OR GREATER THAN 18 MG *(List highest level)* _____	INFANT DIAGNOSIS OF PERSISTENT PULMONARY HYPERTENSION
CRANIOFACIAL ANOMALY/SYNDROME *(Specify)* _____	INFANT DIAGNOSIS OF CYTOMEGALOVIRUS
INFANT DIAGNOSIS OF SEPSIS	MOTHER PRE/PERINATAL DIAGNOSIS OF CYTOMEGALOVIRUS
INFANT DIAGNOSIS OF SEIZURES	MOTHER PRE/PERINATAL DIAGNOSIS OF SYPHILIS
INFANT DIAGNOSIS OF MENINGITIS	MOTHER PRE/PERINATAL EXPOSURE TO RUBELLA
OTOTOXIC MEDICATIONS (INCLUDING BUT NOT LIMITED TO AMINOGLYCOSIDES) USED FOR FIVE DAYS OR LONGER; AND/OR LOOP DIURETICS USED IN COMBINATION WITH AMINOGLYCOSIDES.	FAMILY HISTORY OF PERMANENT CHILDHOOD HEARING LOSS (Excludes acquired hearing losses) *(Specify)* _____

Also captured: low birth weight (<2500 grams); anoxia; 5 minute Apgar < 3

Advisory Board Workgroups

- Providers in KY were aware of the importance of Newborn Hearing screening but….
  - They were unaware of risk factors for PCHL
  - They were not following children with risk factors with ongoing testing

Liza’s Data

- Liza, add as many slides as you need, and feel free to move to whatever location you think this section needs to be.

EHDI Advisory Board Develops SOC Document for Risk Factor Based Follow-Up

- Reviewed JCIH 2007 listed risk factors
- Reviewed KY-specific data regarding the incidence of PCHL with specific risk factors
- Consulted peer-reviewed references to determine timing and frequency of follow-up hearing evaluations
- This took nearly a year!

KY EHDI Advisory Board Recommended Follow-Up for Children with Risk Factors for Hearing Loss (Birth to 5 Years) June 2018

Risk Factor	Special Considerations	Initial Evaluation	Follow-Up
Parental Concern	Any parent concern regarding hearing status	Full evaluation upon request	If pass, PRN if concerns persist or new concerns present
Speech or Developmental Delays	Any concern regarding speech-language or hearing status	As soon as possible upon request	If pass, PRN if concerns persist, or annually with speech services
Family History	1st or 2nd degree relative with permanent hearing loss in childhood (under age 18).	If pass NHS, full evaluation by 3 mos of age	If pass, annually until age 5
Cytomegalovirus (CMV) or Zika (Congenital)	Confirmed Diagnosis of baby	If pass NHS, full evaluation by 3 mos of age	Every 3 months until age 3, every 6 months until age 6, annually thereafter
Cleft Lip/Palate		If pass NHS, full evaluation by 3 mos of age	Based on audiological evaluation & physical findings
Atresia/Microtia	Child should not be screened in affected ear	Schedule full evaluation for as soon after discharge as possible	Based on audiological evaluation & physical findings
Extracorporeal Membrane Oxygenation (ECMO)		Full evaluation upon discharge	If full evaluation at 3 months WNL: every 3 months until 3, every 6 months until 6, annually thereafter
Syndromes associated w/ Hearing Loss (Confirmed or Suspected)	Confirmed Diagnosis https://www.american-hearing.org/disorders/congenital-deafness/#syndronic	If pass NHS, full evaluation by 3 mos of age	If full evaluation at 3 months WNL: every 3 months until 3, every 6 months until 6, annually thereafter
Meningitis	Confirmed or Suspected (Viral and/or Bacterial)	Full evaluation upon discharge	If pass re-evaluate every 3 months until age 1, annually until age 5
Refer Newborn Hearing Screening (NHS)		Full evaluation including Auditory Brainstem Response (ABR) by 3 months of age	Every 3-6 months if confirmed hearing loss
Chemo therapy	https://www.childrensoncologygroup.org/index.php/sensory/hearingproblems	Per oncology protocol	Per oncology protocol
Head Trauma	Penetrating or blunt force trauma to the head or ear, or barotrauma to the ear	Full evaluation upon discharge	If pass, PRN
Risk Factor	**Special Considerations**	**Initial Evaluation**	**Follow-Up**
Ototoxic Medication	Loop diuretics (furosemide), gentamycin, vancomycin, streptomycin, cisplatin, carboplatin	If pass NHS, full evaluation by 1 year of age	PRN for concerns; start w/ tympanograms and OAE's if previous tests were normal
Elevated Bilirubin	>/= 18 mg/dl or exchange transfusion	If pass NHS by AABR, full evaluation by 1 year of age	If pass, PRN
Craniofacial Anomalies	Congenital microcephaly, ear canal, temporal bone, (excluding isolated ear pits or tags)	If pass NHS, full evaluation by 3 mos of age	If pass, annually until age 5
Mechanical Ventilation	Intubation, not C-PAP or Bi-PAP	If pass NHS, full evaluation by 1 year of age	If pass, PRN
In Utero Infection	Confirmed Diagnosis of baby after birth including Herpes, Rubella, Syphilis, Toxoplasmosis	If pass NHS, full evaluation by 1 year of age	If pass, full evaluation annually
NICU Stay	Greater than 5 days for any reason	If no other risk factors, full evaluation by 1 year	If pass, PRN
Other Severe Medical Conditions	Including Persistent Newborn Pulmonary Hypertension (PPHN), Hydrocephalus, Perinatal Asphyxia	If pass NHS, full evaluation by 1 year	If pass, PRN

Abbreviations: ABR—Auditory Brainstem Response, AABR—Automated Auditory Brainstem Response (screen), NHS—Newborn Hearing Screening, OAEs—Otoacoustic Emissions, PRN— "As Needed", WNL— Within Normal Limits

"RED" Risk Factors: Urgent

- Parental Concern: If pass, PRN
- Speech or Language Delay: If pass, PRN; or annually w/ speech
- Family Hx: Annually until 5
- CMV or Zika: If WNL every 3 months until age 3; every 6 months until age 6; annually
- Cleft Lip/Palate: Based on Eval & Findings
- Atresia/Microtia: Based on Eval & Findings
- ECMO: If WNL 3 until 3, every 6 until, annually
- Meningitis: If WNL, every 3 months until age 1, annually until 5**
- Refer NBHS: If HL confirmed, every 3-6 months
- Chemotherapy: Per oncology Protocol
- Head Trauma: If WNL, PRN

A Note About Family History of PCHL

ASK:

Do you or the baby's father know of anyone in either family (siblings, cousins, aunts, uncles, parents) who has had a hearing loss ***since childhood?***

DO NOT ASK:

Does anyone in either the mother's or the father's family have a hearing loss?

"BLACK" Risk Factors:
Slightly Less Urgent than Red Risk Factors

- Ototoxic Meds: PRN for concerns
- Elevated Bilirubin (≥ 18 mg/dl or exchange transfusion needed): If WNL PRN
- Craniofacial Anomalies: If WNL, annually until 5
- Mechanical Ventilation: If pass, PRN
- In Utero Infection excluding CMV/Zika: If WNL, annually
- NICU Stay (>5 days): If WNL, PRN
- Other Medical Conditions: If WNL, PRN

Next Steps

- EHDI Advisory Council plans to develop a SOC document for intervention services in 2019
  - Will consider best practices, evidence-based protocols, parent choice of intervention modality(ies)

- Does your center follow these SOC guidelines for Diagnosis of PCHL and recommended follow-up based on risk factors for late onset/progressive PCHL?

  - Contact KY EHDI for support, training, guidance!

THANK YOU!

Questions?

Contact Us:

Liza Creel – liza.creel@Louisville.edu
Cathy Lester – cathy.lester@ky.gov
Judy Theriot – judy.Theriot@ky.gov