EHDI and the Alabama Newborn Screening Program

Satellite Conference and Live Webcast
Friday, June 12, 2016
9:00 – 11:00 a.m. Central Time

Produced by the Alabama Department of Public Health
Video Communications and Distance Learning Division

Faculty

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Newborn Screening Program

- Currently screen for 30 primary conditions
  - Sickle Cell Disease
  - Cystic Fibrosis
  - Endocrine Disorders
  - Metabolic Disorders
  - Hearing Loss
  - Critical Congenital Heart Disease

Hearing Loss Facts

- **Hearing loss is more prevalent than all other newborn screening disorders**
- **One to three out of 1000 infants have hearing loss**

Hearing Loss Facts

- **Prior to universal newborn hearing screening, the average age of identification of permanent hearing loss in the United States was 2 years of age**
- **Infants with hearing loss who receive intervention by 6 months of age have the best opportunity to develop speech and language skills closest to their hearing peers**

Hearing Loss Facts

- **Hearing loss may occur at a later age**
- **All infants should be monitored for developmental delays that may indicate late onset hearing loss**
- **Any infant with risk factors should have at least one diagnostic audiology assessment by 24-30 months of age**

What is EHDI?

- Originally enacted by Congress in 2000, the Early Hearing Detection and Intervention (EHDI) Act provides funding for early hearing and detection programs nationwide

Early Hearing Detection and Intervention

- The goal of EHDI is to "maximize linguistic competence and literacy development for children who are deaf or hard of hearing"
- Can be accomplished by meeting the goals outlined in the 1-3-6 plan

What is 1-3-6?

- A hearing screen should be completed by 1 month of age
- A comprehensive audiological evaluation (diagnostic) should be completed by 3 months of age
- Appropriate intervention should be initiated by 6 months of age

Hearing Screen

- Every infant should receive a hearing screen prior to hospital discharge
- An infant should receive no more than two inpatient hearing screens
- An infant who does not pass the hospital hearing screen should be referred for another screen before 1 month of age
- Both ears should be tested each time

Care Coordinator Role

- The Care Coordinator (CC) may follow up with the hospital to verify final hearing screen result
- CC should ensure *appropriate* testing completed

Audiological Evaluation

- Screener or physician should refer an infant with a non-passing repeat screen for evaluation before 3 months of age
  - Medical evaluation and treatment for ear infection
  - Referral to ENT for evaluation of middle ear effusion
  - Appointment with audiologist for diagnostic evaluation

Care Coordinator Role

- CC should educate physician and family, providing needed assistance, to ensure appropriate follow up care completed
  - Diagnosis – Normal hearing or Hearing Loss?
  - Report confirmed diagnosis (type and severity of hearing loss)

Early Intervention

- An infant with probable or confirmed hearing loss, should be referred to an early intervention program by 6 months of age
- Intervention options will vary from infant to infant

Care Coordinator Role

- CC should ensure referral to local early intervention (EI) program and provide support until enrollment verified

What is Hearing Screening?

- A hearing screen is a noninvasive, automated test to detect potential hearing loss
- A hearing screen cannot tell you how much hearing loss is present or the underlying cause of hearing loss

Determining Appropriate Screening Method

- An OAE can be performed on any well baby with no known risk factors
- An OAE should not be used as the follow up screen for an infant that initially did not pass an AABR
- An infant who is in NICU greater than 5 days, or who has other risk factors, should always receive an AABR

Risk Factors for Hearing Loss

- Family history of permanent childhood hearing loss
- NICU stay of more than 5 days
- Exposure to ototoxic medications or loop diuretics
- Hyperbilirubinemia that requires exchange transfusions
- In utero infections, such at cytomegalovirus (CMV)
- Craniofacial anomalies

Risk Factors for Hearing Loss

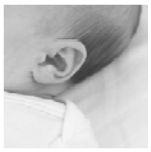
- A syndrome, or physical findings associated with a syndrome, which is known to include permanent hearing loss
- Neurodegenerative disorders
- Postnatal infections associated with sensorineural hearing loss, both bacterial and viral
- Head trauma
- Chemotherapy

Hearing Loss Diagnosis

- Types of hearing loss
  - Sensorineural Hearing Loss
  - Conductive Hearing Loss
  - Auditory Neuropathy Spectrum Disorder

Hearing Loss Diagnosis

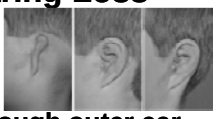
- Severity of hearing loss
  - Mild
  - Moderate
  - Severe
  - Profound



Sensorineural Hearing Loss (SNHL)

- Caused by damage to inner ear (cochlea) or auditory nerve pathways
- Reduces ability to hear faint sounds
- Usually cannot be medically or surgically corrected
- Most common type of permanent hearing loss

Conductive Hearing Loss (CHL)



- Caused by inefficient sound conduction through outer ear canal to the eardrum and the tiny bones (ossicles) in the middle ear
- Results in reduction of sound level or ability to hear faint sounds
- Can often be medically or surgically corrected

Auditory Neuropathy Spectrum Disorder (ANSD)

- Caused by inconsistent electrical signals from the 8th nerve to the brain
- Suspect ANSD
  - Infant has abnormal ABR/AABR with normal OAEs present
  - Diagnosed infant/child has residual hearing with good high-quality hearing aids, but still does not progress in speech and language

For More Information

Contact the Alabama Newborn Screening Program at (334) 206-5556 or toll free at (866) 928-6755.

Order brochures for parents online at:
https://www.adph.org/Extranet/Forms/Form.asp?ss=s&formID=5561