Monitoring for Hearing Loss in Children: Risk Indicator Practices

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Introduction

• Joint Committee on Infant Hearing (JCIH)
  • Established in 1969, guidelines since 1971
  • Recommend protocol for early identification of children with hearing loss
  • Latest statement released in 2007, with 11 risk indicators
  • 2018 statement will be published later this year
Purpose

- Review the current literature on each individual risk indicator and discover the prevalence of postnatal hearing loss in children with one single risk indicator or multiple risk indicators.
• **Surveillance**: all children should receive monitoring of developmental milestones in hearing and language development at well-child visits

• **At Risk**: if a child passes newborn hearing screening, but is identified as having a risk indicator for hearing loss, JCIH guidelines recommend at least **one hearing evaluation before 30 months of age**.
Risk Indicators
1. Caregiver concern
2. Family history
3. NICU stay
4. In-utero infections
5. Craniofacial anomalies
6. Physical findings associated with a syndrome
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss
8. Neurodegenerative disorders
9. Postnatal viral or bacterial meningitis
10. Head trauma or temporal bone fracture
11. Chemotherapy
Eleven question survey sent to all EHDI program coordinators in the U.S. and Canada.

42 total responses
- 37 from US
- 5 from Canada

Question 1
- “Does your EHDI program monitor risk indicators for delayed-onset and/or progressive hearing loss?”

Question 3
- “If yes, which risk indicators does your EHDI program monitor?”
Results

- Craniofacial anomalies
- Family history of permanent childhood hearing loss
- In utero infections (CMV, Herpes, Rubella, Syphilis, Toxoplasmosis)
- Hyperbilirubinemia (aka jaundice) that requires exchange transfusion
- Extended NICU stay/greater than 5 days
- Syndromes associated with hearing loss or progressive or late-onset hearing loss
- Assisted ventilation
- Exposure to ototoxic medications
- Physical findings that are associated with a syndrome
- Low birth weight
- Culture positive postnatal infections associated with sensorineural hearing loss
- ECMO (extracorporeal membrane oxygenation)
- Exposure to loop diuretics
- Neurodegenerative disorders
- Caregiver concerns regarding hearing, speech, language or developmental delay
- Head trauma
- Chemotherapy
- Apgar score
- Jaundice not requiring an exchange transfusion
- Other
• Question 3 responses not listed as JCIH Risk Indicator:
  • Congenital diaphragmatic hernia
  • Hypoxic ischemic encephalopathy-Sarnat II or III
  • Intraventricular hemorrhage-grade 3 or 4
  • Periventricular leukomalacia
  • Medevac to larger center
  • Kidney dysfunction
  • Persistent pulmonary hypertension of the newborn
  • Genetic hearing loss: Connexin 26
Conclusion

- Across EHDI programs, different risk indicators are monitored
- Not all JCIH risk indicators are monitored by EHDI programs
• Cleft Lip and/or Palate:
  • Hearing loss
    • High risk of otitis media with effusion and conductive hearing loss
    • Without identification as a risk indicator, children’s hearing loss may not be detected due to lack of monitoring
  • Recommendation:
    • Recommend that cleft lip and/or cleft palate be added to the risk indicator list either under its own risk indicator or clearly stated under craniofacial anomalies
Recommendations

- Revised list of risk indicators for identifying children at risk for hearing loss
- More specific guidelines for a timeline for monitoring
**My Recommended Guidelines 2018**

1. Caregiver concern or family history of childhood hearing loss
2. Neonatal intensive care of more than 5 days with the following occurring: ECMO treatment of longer than 5 days, assisted ventilation, exposure to multiple or high doses of ototoxic medications such as aminoglycosides or loop diuretics, or hyperbilirubinemia that requires exchange transfusion
3. Cytomegalovirus (CMV)
4. Craniofacial anomalies involving the ear canal or temporal bone anomalies
5. Cleft lip and/or cleft palate
6. Physical findings that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss such as a white forelock or malformations of the ears
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss such as neurofibromatosis, osteopetrosis, and Usher syndrome: other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson
8. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome
9. Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis
10. Head trauma, especially basal skull/temporal bone fracture that requires hospitalization
11. Chemotherapy

**JCIH Guidelines 2007**

1. Caregiver concern regarding hearing, speech, language, or developmental delay
2. Family history of permanent childhood hearing loss
3. NICU of more than 5 days or any of the following regardless of length of stay: ECMO, assisted ventilation, exposure to ototoxic medications or loop diuretics, and hyperbilirubinemia that requires exchange transfusion
4. In utero infections, such as CMV, herpes, rubella, syphilis, and toxoplasmosis
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher syndrome: other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson
8. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome
9. Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis
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Questions?