Guidelines and Protocols for Testing Children with High Risk Factors

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High Risk Factors

- High risk factors for hearing loss were first discussed and published in the 1972 JCIH statement.
- Between 1982 and 1994, other risk factors were added to the statement.
- With each new statement, other recommendations are added.

JCIH Guidelines 2000

“All infants who pass newborn hearing screening but who have risk indicators for other auditory disorders and/or speech and language delay receive ongoing audiologic and medical surveillance and monitoring for communication development. Infants with indicators associated with late-onset, progressive, or fluctuating hearing loss as well as auditory neural conduction disorders and/or brainstem auditory pathway dysfunction should be monitored.”

Current 2007 JCIH Guidelines

“The timing and number of hearing reevaluations for children with risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss. Infants who pass the neonatal screening but have a risk factor should have at least 1 diagnostic audiology assessment by 24 to 30 months of age. Early and more frequent assessment may be indicated for children with cytomegalovirus (CMV) infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, or culture-positive postnatal infections associated with sensorineural hearing loss; for children who have received extracorporeal membrane oxygenation (ECMO) or chemotherapy; and when there is caregiver concern or a family history of hearing loss.”

“Surveillance and screening in the medical home”

“For all infants, regular surveillance of developmental milestones, auditory skills, parental concerns, and middle-ear status should be performed in the medical home, consistent with the American Academy of Pediatrics (AAP) pediatric periodicity schedule. All infants should have an objective standardized screening of global development with a validated assessment tool at 9, 18, and 24 to 30 months of age or at any time if the health care professional or family has concern. “

“Infants who do not pass the speech-language portion of a medical home global screening or for whom there is a concern regarding hearing or language should be referred for speech-language evaluation and audiology assessment. “

Appendix 2: Risk Indicators Associated with Hearing Loss in Childhood

- Caregiver concerns regarding hearing, speech or developmental delay
- Family history of permanent childhood hearing loss
- NICU care of more than 5 days or any other following regardless length of stay: ECMO, assisted ventilation, ototoxic meds, hyperbilirubenemia requiring blood transfusion
- Low birth weight < 1500 grams
- In utero infections: CMV, herpes, rubella
- Physical findings that are associated with a syndrome known to include SNHL or permanent CHL
- Craniofacial anomalies
- Syndromes associated with hearing loss
- Neurodegenerative disorders
- Culture positive postnatal infections associated with SNHL: meningitis
- Head trauma
- Chemotherapy

3 Urban Boston Hospitals with different protocols…

Hospital A

- ABR at 3 months
  - Hyperbilirubenemia at > 20 mg/dl
  - In utero infection of CMV
- ABR at 6 months
  - Syndromes or Neurodegenerative Disorders associated with hearing loss
  - Family history of hearing loss
  - In utero infection such as syphilis, herpes, rubella
- ABR within first 9 months
  - ECMO
- Behavioral evaluation by 1 year of age
  - Low birth weight <1500 grams

Hospital B

- ABR at 3 months
  - Family history of hearing loss, CMV, meningitis
- ABR at 6 months
  - Craniofacial anomalies and other risk factors
- Behavioral testing at 9-12 months
  - NICU stay and prematurity

Hospital C

- ABR at 3 months
  - CMV, family history of hearing loss, and meningitis
- Behavioral testing at 6 months
  - All other risk factors

High Risk Cases

Case A

- 32 week NICU graduate
- History of:
  - Respiratory distress syndrome
  - Hypotension
  - Hypoglycemia
  - Hyperbilirubenemia
  - Grade 4 IV hemorrhage
  - Intraparenchymal hydrocephalus
  - Prosencephaly
- Ventilator for a month and then transferred to O2 (at home/Rehab facility as well)

- Hearing screen (OAE and ABR screen)
  - Refer - both ears
- 3 week follow-up (bilateral)
  - ABR response down to 15 dB eHL via click stimuli
  - Robust OAEs (Diagnostic)
  - Normal tympanometry and present reflexes

- 6 month follow-up
  - Normal tympanometry and absent reflexes
  - Absent OAEs (Diagnostic)
  - Response to 75 dB HL at 1000 Hz (no startle)
  - No response to speech at 85 dB HL
  - Threshold ABR
    - Right – DNT
    - Left – Poor morphology and no repeatable response at 80 dB nHL

- Continued testing…
  - Tympanometry: Normal
  - Acoustic reflexes: Absent
  - OAEs: Absent
  - ABRs
    - Responses not seen at 90 dB nHL via click
    - Rarefaction and condensation stimuli ruled out auditory dys-sychrony

- Recommendations:
  - Fit with binaural Oticon Vigo Power BTE
  - Early Intervention (already receiving due to developmental delays)
  - Referred to hospital in Boston for Cochlear Implant Evaluation

Case B

- Normal full term in well baby nursery
- Family history of hearing loss - sibling has mild to moderate SNHL with binaural hearing aids
- Passed ABR screen

- Follow-up at 2 1/2 months
  - Mild to moderate SNHL – bilateral

- Recommend binaural hearing aids
  - Further information is unknown at this time. Lost to follow-up.

Case C

- 33 week NICU Graduate
- Hyperbilirubenemia
  - Required multiple full body transfusions
- ABR screen - passed both ears

- Follow-up Testing at 6 months
  - Diagnostic OAE - absent
  - Threshold ABR
    - Right - moderately severe SNHL
    - Left - moderate SNHL
- Recommendations
  - Binaural hearing aids

Case D

- 24 week NICU graduate
- Diagnosed with Cytomegalovirus
- Required ventilation - tracheotomy
- ABR screen - passed both ears

- Initial Testing - 2.5 months old
  - Diagnostic ABR
    - Right - Moderately severe SNHL
    - Left - Severe SNHL
- Treatment
  - Ganciclovir

- Most recent follow-up testing after Treatment - 4 months old
  - Diagnostic ABR
    - Right - Grossly normal hearing
    - Left - Mild to moderate SNHL

Case E

- Normal delivery
- ABR screen - pass both ears
- Parental Concern
  - No response to sounds
- No familial history of hearing loss

- Testing - 3.5 months old
  - Diagnostic ABR
    - 500 - 8000 Hz
      - Severe to profound SNHL
    - Auditory dys-sychrony ruled out
- Recommendations
  - Repeat testing
  - Consider binaural hearing aids

Conclusion and Thoughts

- The guidelines are correct in the fact that follow up protocols 'should be customized and individualized' and that 'early and more frequent assessment may be indicated' for children with various risk factors.
- The responsibility for determining any given time schedule for follow up falls on the medical home AND the audiologist. The resulting protocols may be, as we have seen, highly variable.
- Availability of diagnostic pediatric audiology services may be limited in many areas, so more specific guidelines for primary care physicians might be helpful.

- There is a need for Audiology to develop more standard guidelines within the structure of the JCIH Guidelines to assist screening and diagnostic programs in making decisions about follow-up.
- Guidelines will need to be evidence-based, so more research regarding level of risk for delayed onset and progressive hearing loss for various factors may be needed (example 32 weeks vs 35 weeks prematurity)
- Parent education materials geared to risk factors may be helpful for screening programs.
- Where audiologists are in short supply, are there other acceptable screening tools that can be used as interim measures, such as OAEs and tympanometry?

Final Thought and Future Discussion….

- Once the infant has been seen for initial follow-up testing, should there be guidelines and protocols regarding the continued follow-up and what should they be?

References

- Joint http://www.jcih.org/posstatemts.htm
- PEDIATRICS Vol. 120 No. 4 October 2007, pp. 898-921 (doi:10.1542/peds.2007-2333)

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Thank you and have a great day!