

# **Risk Factors for Infant Hearing Loss: Practical Information and Clinical Implications**

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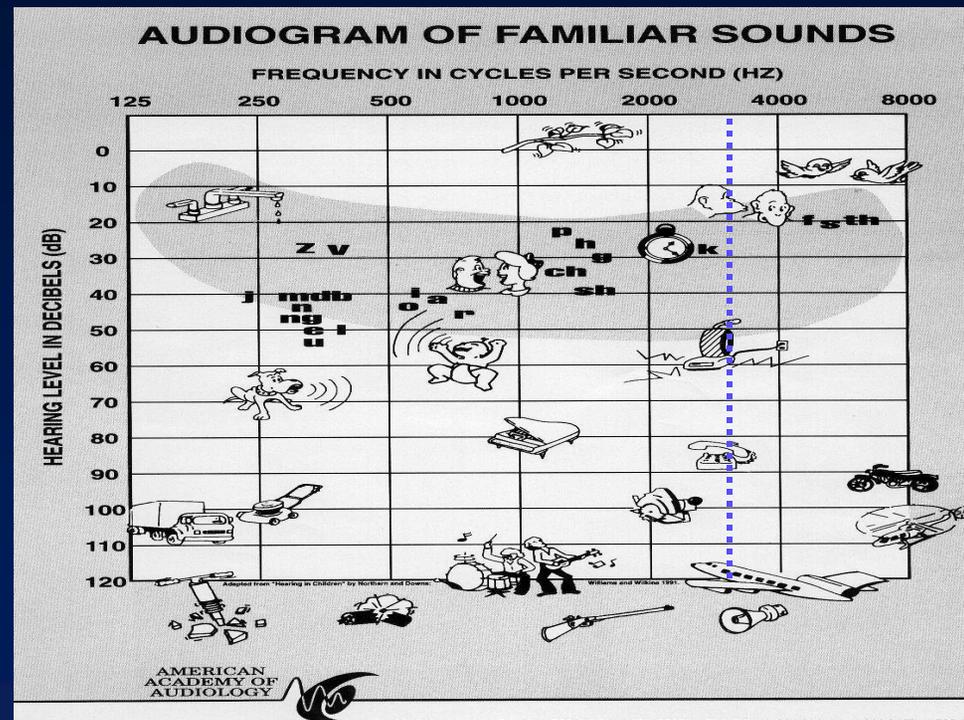
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## Risk Factors for Infant Hearing Loss: Important in Early Hearing Loss Diagnosis and Intervention (EHLI)



## **Joint Committee on Infant Hearing: Defining Standard of Care for Early Detection and Identification of Hearing loss**

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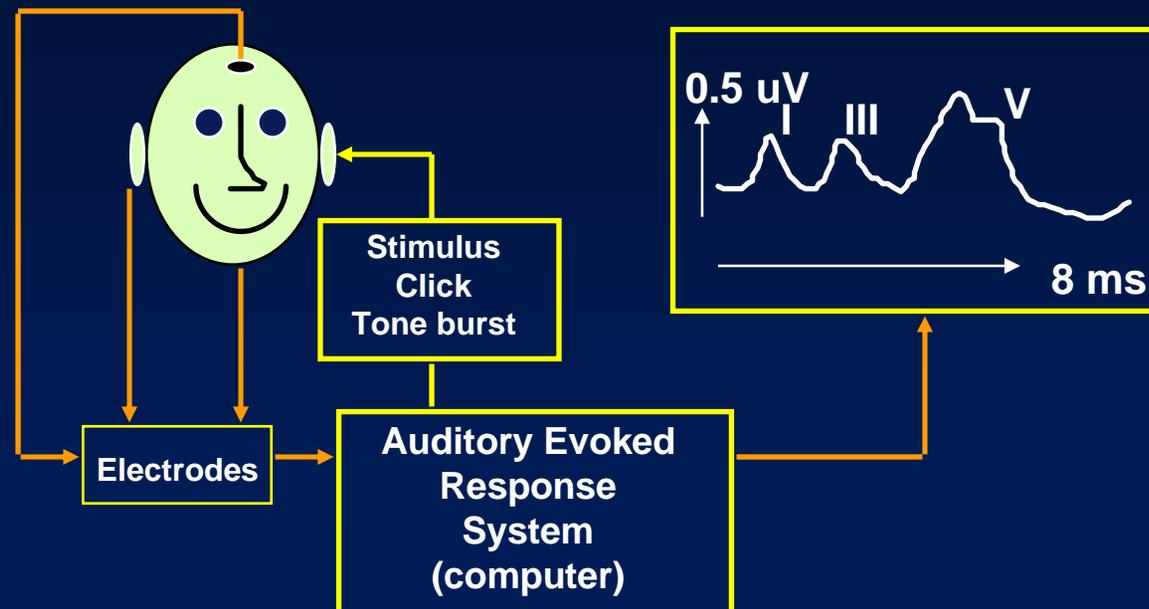
### **□ Member organizations:**

- Alexander Graham Bell Association for the Deaf and Hard of Hearing**
- American Academy of Audiology**
- American Academy of Otolaryngology-Head & Neck Surgery**
- American Academy of Pediatrics**
- ASHA**
- Council on Education of the Deaf**
- Directors of Speech and Hearing Programs in State and Welfare Agencies**

## Diagnosis of Hearing Loss: Protocol for Confirmation of Hearing Loss in Infants and Toddlers (0 to 6 months) *Year 2007 JCIH Position Statement*

- ❑ Child and family history
- ❑ Otoacoustic emissions
- ❑ ABR during initial evaluation to confirm type, degree & configuration of hearing loss
- ❑ Acoustic immittance measures (including acoustic reflexes)
- ❑ Supplemental procedures (insufficient evidence to use of procedures as “sole measure of auditory status in newborn and infant populations”)
  - Auditory steady state response (ASSR)
  - Acoustic middle ear reflexes for infants < 4 months
  - Broad band reflectance
- ❑ Behavioral response audiometry (*if feasible*)
  - ✓ Visual reinforcement audiometry *or*
  - ✓ Conditioned play audiometry
  - ✓ Speech detection and recognition
- ❑ Parental report of auditory & visual behaviors
- ❑ Screening of infant’s communication milestones

## PEDIATRIC AUDIOLOGY: Auditory brainstem response (ABR)



*ABR mature by 18 months*

## Infant Hearing Screening and Diagnostic Assessment: Auditory Brainstem Response



## Infant Hearing Screening and Diagnostic Assessment: Otoacoustic Emissions



## **Year 2007 JCIH Position Statement: Risk Indicators Associated with Permanent Congenital, Delayed-Onset, or Progressive Hearing Loss in Childhood (1)**

- ❑ Caregiver concern regarding hearing, speech, language, or developmental delay.
- ❑ Family history of permanent childhood hearing loss
- ❑ NICU stay of > 5 days or
  - ECMO
  - Assisted ventilation
  - Exposure to ototoxic medicines
  - Hyperbilirubinemia requiring exchange transfusion
- ❑ In utero infections, e.g.,
  - CMV
  - Herpes
  - Rubella
  - Syphilis
  - Toxoplasmosis
- ❑ Craniofacial anomalies, including involvement of the
  - Pinna
  - Ear canals
  - Ear tags and pits
  - Temporal bone anomalies

## **Year 2007 JCIH Position Statement: Risk Indicators Associated with Permanent Congenital, Delayed-Onset, or Progressive Hearing Loss in Childhood (2)**

- ❑ Physical findings associated with a syndrome, e.g., white forelock
- ❑ Syndromes associated with hearing loss, e.g.,
  - Neurofibromatosis
  - Osteopetrosis
  - Usher syndrome
  - Waardenburg
  - Alport
  - Pendred
  - Jervell
  - Lange-Nielson
- ❑ Neuro-degenerative disorders, e.g.,
  - Hunter syndrome
  - Sensory motor neuropathies
    - ✓ Friedreich ataxia
    - ✓ Charcot-Marie-Tooth syndrome
- ❑ Culture positive post-natal infections associated with sensorineural hearing loss, e.g.,  
Confirmed bacterial and viral meningitis
- ❑ Head trauma requiring hospitalization
- ❑ Chemotherapy

## Year 2007 JCIH Position Statement: New Definition of Hearing Loss and New Screening Protocols

- “The definition [of targeted hearing loss] has been expanded from congenital permanent bilateral, unilateral, or permanent conductive hearing loss to include neural hearing loss (e.g., “**auditory neuropathy/dyssynchrony**”) in infants admitted to the NICU”
- Separate protocols are recommended for NICU and well-infant nurseries.
  - NICU infants admitted for more than 5 days should be screened with ABR to detect neural hearing loss.
  - Infants who do not pass the ABR screening in the NICU should be referred directly to an audiologist for re-screening and, as indicated, comprehensive diagnostic evaluation (including ABR).
  - Re-screening should be completed for both ears, even if 1 ear failed initially (there is considerable risk for progressive hearing loss in ear that initially passed hearing screening).

**Year 2007 Position Statement: Principles and Guidelines for  
Early Hearing Detection and Intervention Programs  
Joint Committee on Infant Hearing (JCIH). *Pediatrics* 120: 898-921**

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- ❑ “Physiologic measures must be used to screen newborns and infants for hearing loss. Such measures include OAE and automated ABR testing.” (p. 903)
- ❑ “Both OAE and automated ABR techniques provide noninvasive recordings of physiologic activity underlying normal auditory function.” (p. 903)
- ❑ “**Neural conduction disorders or auditory neuropathy/dys-synchrony** without concomitant sensory dysfunction will not be detected by OAE testing.”
- ❑ “The JCIH recommends ABR technology as the only appropriate screening technique for use in the NICU.” (p/ 904)
- ❑ “Some programs use a combination of screening techniques (OAE and ABR) to decrease the fail rate at discharge.” (p. 904)

Guidelines for  
Identification and Management  
of Infants and Young Children  
with Auditory Neuropathy Spectrum Disorder



**The Children's Hospital**  
• Bill Daniels Center for Children's Hearing  
Guidelines Development Conference  
at NHS 2008, Como, Italy

**Year 2007 JCIH Position Statement:  
Risk Indicators Associated with Permanent Congenital,  
Delayed-Onset, or Progressive Hearing Loss in Childhood (1)**

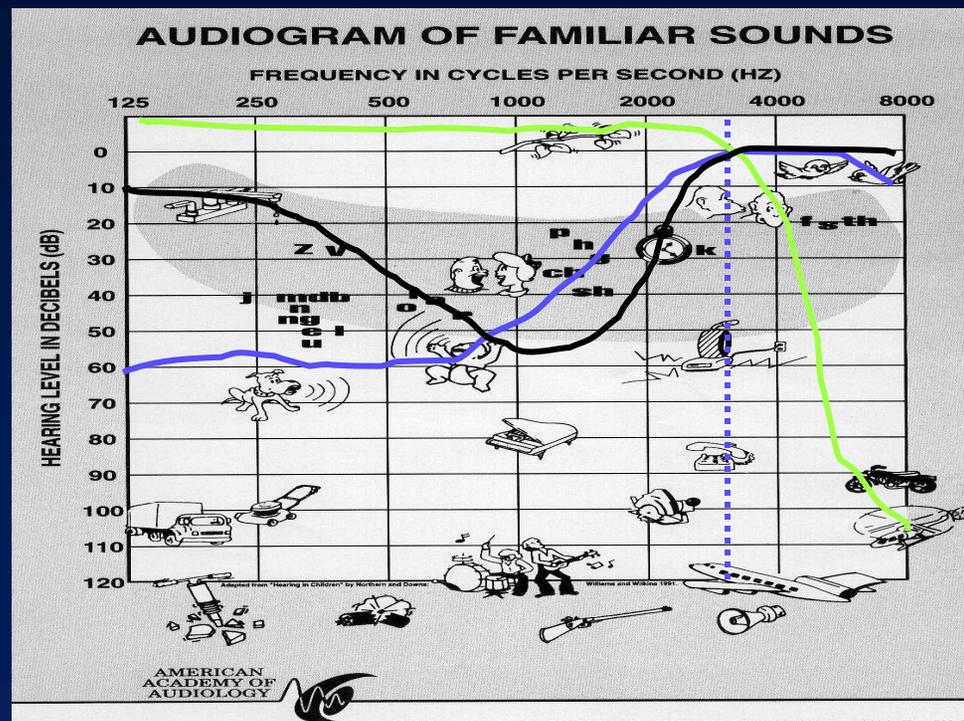
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- ❑ **Delayed onset, late onset, or “acquired” hearing loss**
  - **Normal auditory function (hearing) at birth with the onset of auditory dysfunction (hearing loss) in infancy or early childhood**
- ❑ **Progressive hearing loss**
  - **Normal auditory function (hearing) at birth with the onset of auditory dysfunction (hearing loss) in infancy or early childhood**

## Audiologic Factors Influencing the Distinction between Progressive versus Delayed/Late Onset Sensorineural Hearing Loss

- Hearing screening method and protocol, e.g.,
  - ABR
    - ✓ Dependent mostly on hearing sensitivity in 2000 to 4000 Hz region
    - ✓ When elicited with 35 dB nHL click stimulation, not sensitive to low frequency or mild high frequency cochlear auditory dysfunction
  - OAE
    - ✓ Screening protocol usually includes limited frequency region of 2000 to 5000 Hz
    - ✓ Dependent only on outer hair cell status. Insensitive to inner hair cell auditory dysfunction
- Configurations of hearing loss mostly likely contributing to “false negative” screening outcome
  - High and very high frequency hearing loss > 5000 Hz
  - Mid-to-low frequency hearing loss < 2000 Hz (often genetic etiology)
  - Mid-region “cookie bite” hearing loss (often genetic etiology)
- Hearing loss may initially be unilateral
  - Hearing screening must be completed for both ears
  - Children with unilateral hearing loss at birth are at risk for later bilateral hearing loss
  - Follow up screening or diagnostic assessment of both ears is indicated for children with unilateral screening failures

# Congenital or Progressive Hearing Loss Masquerading as Delayed/Late Onset Hearing Loss



## **Year 2007 JCIH Position Statement Recommendations for Timing and Frequency of Audiologic Follow up for Infants with Risk Indicators Associated with Permanent Congenital, Delayed-Onset, or Progressive Hearing Loss**

- ❑ Prior (2000 JCIH) recommendations for follow up at 6-month intervals of all NICU graduates (approximately 400,000 babies annually) placed an excessive burden on audiologists
- ❑ 2007 JCIH shifts responsibility for surveillance of all infants to the primary care provider who will refer to audiologists as needed, e.g.,:
  - Concerns or findings consistent with hearing loss
  - Risk factors for delayed/late onset or progressive hearing loss
- ❑ 2007 JCIH recommends at least one audiologic referral for low risk infants by age 24 to 30 months
- ❑ Early and more frequent referral (every 6 months) to audiologists for risk factors associated with delayed onset and progressive hearing loss, e.g.,:
  - Family history
  - CMV
  - ECMO therapy
  - Potentially ototoxic chemotherapy (e.g., cisplatin)
  - Neurodegenerative disorders

## Infants with Risk Indicators Associated with Permanent Congenital, Delayed-Onset, or Progressive Hearing Loss

- Fortnum, Summerfield, Marshall, Davis & Bamford. (2001). *BMJ*, 323, 536-554
  - Prevalence within 17,160 children increased from 1.07% at age 3 years to 2.05% at age 9 to 16 years
  - “Prevalence of confirmed permanent childhood hearing impairment increases until the age of 9 years to a level higher than previously estimated. Relative to current yields of universal neonatal hearing screening in the United Kingdom, which are close to 1/1000 live births, 50-90% more children are diagnosed with permanent childhood hearing impairment by the age of 9 years.

## Infants with Risk Indicators Associated with Permanent Congenital, Delayed-Onset, or Progressive Hearing Loss

- ❑ At least 50% of all congenital hearing loss is hereditary (2007 JCIH)
  - Almost 600 syndromes with hearing loss
  - 125 genes associated with hearing loss
- ❑ Approximately 30 to 40% of children with hearing loss have associated disabilities
- ❑ Common genetic causes of delayed onset or progressive hearing loss
  - Connexin 26
    - ✓ Accounts for >50% of non-syndromic and some causes of syndromic hearing loss
  - Connexin 30
    - ✓ Severe to profound hearing loss usually occurs with no other medical problems
  - Cytomegalovirus (CMV)
    - ✓ Progressive, sometimes into school age
    - ✓ Hearing loss may fluctuate
    - ✓ Hearing loss may be unilateral or bilateral
    - ✓ Accounts for 21 to 25% of congenital hearing loss
  - Pendred syndrome
    - ✓ Recessive syndromic hearing loss
    - ✓ May develop later in infancy or early childhood
    - ✓ Usually progressive
    - ✓ Accounts for up to 8% of all congenital hearing loss

## **Year 2007 JCIH Position Statement Guidelines About Surveillance for Delayed-Onset, or Progressive Hearing Loss**

- ❑ 2007 JCIH recommends inclusive strategy of surveillance within medical home based on the pediatric periodicity model
- ❑ At each visit infants should be monitored for
  - Auditory status
  - Middle ear status
  - Developmental milestones
  - Concerns should be validated with administration of validated global screening tool at 9, 18, 24, and 30 month
- ❑ An audiologic evaluation is recommended if a child doesn't pass speech-language portion of screening tool in medical home
- ❑ Siblings of infants with diagnosed hearing loss should also be referred for audiologic assessment
- ❑ All infants with risk indicators, regardless of surveillance findings, should be referred for audiological evaluation by at least 24 to 30 months of age

# Objective Assessment of Hearing



James W. Hall, III  
De Wet Swanepoel



A volume in the Core Clinical Concepts in Audiology Series.