Advanced Tools for Early Newborn Diagnosis

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I. Summary and Review of Newborn Screening
II. Middle Ear Problems in Infants
III. 1000 Hz and Wideband Tympanometry
IV. Frequency Specific Toneburst Air and Bone ABR
V. Cases
Universal Newborn Hearing Screening Status

• With the growth of Universal Newborn Hearing Screening, 95% of all infants are evaluated at birth, and the average age of identification is 3 months in the U.S. (JCIH, 2007).
• The refer rate for NHS averages 8% at Stage I screening but improves to less than 2% at re-screening with either a second OAE test or ABR screening (Clemens et al., 2000; Clemens & Davis, 2001; Hall et al., 2004).

Effectiveness and Efficiency of UNHS

• The positive predictive value of screening for SNHL is only about 10% since the incidence of congenital SNHL is approximately 2/1000 newborns.
• The over-referral rate for SNHL may cause unnecessary anxiety for some parents (Vohr et al., 2001) and increased cost to the medical system (Clemens et al., 2000).

What is the underlying cause of NHS false positive results?

• Transient outer or middle-ear conditions may result in a “referred” test result regardless of the presence of normal cochlear function.
• ~80% of hearing screening referrals are associated with middle-ear dysfunction as detected by wideband immittance (Sanford & Keefe, 2009; Hunter, Feeney et al. 2010)
Middle Ear Problems in Infants

• In newborns, the middle-ear space may contain residual amniotic fluid, mesenchyme or meconium (Northrup et al., 1986; Jaisinghani et al. 1999; Miura et al. 2008).

• The outer ear canal of infants is flaccid, easily collapsible and the bony portion ossifies over the first year after birth, causing large developmental changes (Holte et al., 1991).

Should middle ear conditions be considered “false-positive screening”?  

• What is the importance of early middle ear dysfunction?
• Are there reasons to follow children who have early conductive hearing loss?
• What can we learn about this large proportion of screening referrals?
• What should be the clinical protocol to follow-up these cases?

Conductive Hearing Loss – “So What?”

• Infants failing NHS due to conductive hearing loss are at high risk for persistent or fluctuant hearing loss.
• Average hearing for infants with CHL approximately 30 dB with range of 15-45 dB. (Maxon et al., 1993).
• Conductive hearing loss may sometimes be structural in origin.
Failed Newborn OAEs Resulting From Otitis Media
Boone, Bower and Martin, 2005

• Retrospective review of 76 patients referred from failed TEOAE at birth.
• Otitis media was found in 64.5%.
• Mean age at OM diagnosis was 3 months.

• Tube insertion was required in 35% to allow for complete hearing assessment.
• After resolution of OME, 11% had SNHL.
• In infants without OME, 79% had SNHL.
• Thus, the risk of SNHL in infants without OME who refer is nearly 8 times greater.

• Newborn is referred for repeated OAE or ABR failure:
  – Is hearing loss present?
  – If so, sensorineural or conductive?
• Possible sources of information:
  – Pneumatic Otoscopy
  – Toneburst ABR
  – 1000 Hz Tympanometry
  – Wideband Reflectance
Challenges in Examining Newborn Ear

- Narrow, collapsing ear canal
- Flaccid ear canal on pneumatic pressure
- Horizontal orientation of TM
- Unclear demarcation of TM and ear canal walls
- "Normal" TM appearance opaque and thickened
- Requires otolaryngologist experienced in newborn ear examination and microscopic exam

226 versus 1000 Hz tympanometry in newborns

- 226-Hz tympanogram usually normal or notched in confirmed MEE under 6 months of age
- Paradise et al., (1976) first observed this problem and attributed it to simple ear canal wall movement

Difficulty of Identifying MEE with Conventional Tympanometry

- Marchant et al. (1986) first reported that higher frequency tympanograms (660 Hz) were effective in identifying MEE
- Later report (Rhodes et al., 1999) found that 1000 Hz was more sensitive than 660 Hz to OAE screening failures with normal ABRs
Tympanometry in infants with middle ear effusion having been identified using spiral computerized tomography.


In the right ear without fluid, tympanograms were single peaked at both probe-tone frequencies.

In left ear with fluid, 226 Hz tympanogram was single peaked, 1000 Hz tympanogram was flat.
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Tympanometry in infants with middle ear effusion having been identified using spiral computerized tomography.

Percent Abnormal 226 Hz Tympanometry Compared to CT Results

- 226 Hz not specific
- 226 Hz not sensitive

Percent Abnormal 1000 Hz Tympanometry Compared to CT Results

- 1000 Hz is specific
- 1000 Hz is sensitive

Wideband Power Reflectance Principle

\[
\text{Power Reflectance} = \frac{\text{Reflected Power}}{\text{Incident Power}}
\]
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Wideband Reflectance (WBR)
Keefe et al., 2008

Normal ears have lower (better) reflectance than ears with confirmed Middle ear Effusion (MEE).

AROC = .87 for absorbance versus .75 for 1-kHz tympanometry
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Wideband Reflectance is Poorer in Newborns with Abnormal OAE

Hunter et al., 2010.
Pass = 352
Refer = 141

Wideband reflectance improves during first 3 days after birth

Hunter et al., 2010.
Pass = 352
Refer = 141

Wideband reflectance improves with OAE improvement

Hunter et al., 2010.
Pass = 352
Refer = 141
Case 120R

- 2 day-old baby boy
- 37 weeks GA
- 2685 gms
- No risk factors
- Passed DPOAE
- Could not obtain seal for tympanometry
Case 120R Discussion

- All frequency regions demonstrated normal reflectance, correlating with normal OAE signal levels.
- This normal pass agrees with reflectance.
- Tympanometry could not be obtained.
Case 97R

- 1 day-old baby girl
- 38 weeks GA
- 3100 gms
- No risk factors
- DPOAE Referred

**1000 Hz Tympanometry and Acoustic Reflex**

<table>
<thead>
<tr>
<th>Frequency (kHz)</th>
<th>Amplitude (dB SPL)</th>
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<tbody>
<tr>
<td>2</td>
<td>-20</td>
</tr>
<tr>
<td>3</td>
<td>-13</td>
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<tr>
<td>4</td>
<td>-10</td>
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<tr>
<td>8</td>
<td>-20</td>
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</tbody>
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- Static admittance = 0.0 mmho
- Tympanometric Width = >500 daPa
- Equivalent volume = .34 cc
- BBN acoustic reflex: Present below 90 dB

*Profile of Tympanometry and Acoustic Reflex for 1000 Hz:*

- **TestID 911:**
  - **Reflex:**
    - Present
    - Antidromic
    - PTA
  - **Referred:**
    - DPOAE
Toneburst ABR Protocol to Detect Conductive Hearing Loss

- If tympanograms or reectance abnormal, important to perform bone ABR rather than waiting.
- Risk of loss to follow-up is higher when testing is incomplete or inconclusive.
- Testing bone conduction helps to rule out sensorineural hearing loss earlier.

Toneburst ABR Protocol

- System: Vivosonic Integrity V50C
- Stimulus rate: 37.1(stimuli/s)
- Windowing: Blackman 2-0-2
- Averaging: Kalman weighted
- High pass filter cutoff frequency: 30 Hz
- Low pass filter cutoff frequency: 3000 Hz
- Tone-burst frequencies: 0.5, 1, 2, 4 kHz.
- Cond/Rare with correlation ≥ 0.7 for wave V threshold.

Recording Montage

- High forehead to ipsilateral mastoid recording montage.
- ER-3A insert earphones.
- Hand-held B-71 vibrator at temporal bone, contralateral masking.
Typical Tone-burst ABR Recording

Mean AC wave V TB Thresholds
Elsayed et al., Audiology NOW! 2012

Mean BC wave V TB Thresholds
Elsayed et al., Audiology NOW! 2012

*Average and upper cut off values are frequency-specific.
Latency-Intensity Functions for AC
Elsayed et al., Audiology NOW! 2012

- Latency and TB frequency are inversely related due to cochlear travel time.
- Latency generally decreases with increased intensity.

Latency-Intensity Functions for BC
Elsayed et al., Audiology NOW! 2012

- Trends for latency related to frequency and intensity are similar to AC TB.

Case 3: Baby Girl 2003

- Enrolled from NICU, 38 week gestational age, >3000 gms
- Treated with Gentamicin
- Screening in NICU:
- Referred on ABR and OAE Left Ear
- Passed ABR and OAE Right Ear
• DPOAE: High noise level, both ears, no significant S/N Ratio at any Frequency
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Recorded May 2, 2012

Left Air Conduction ABR
- 500 Hz = 50 dB, 1000 Hz = 30 dB, 2000 Hz = 50 dB

Left Bone Conduction ABR
- 500 Hz ABG = 40 dB, 1000 Hz ABG = 10 dB

Recommended UNHS Follow-Up
- Failed (x2) Hearing Screen
- Diagnostic Air and Bone ABR
  - Normal
  - Abnormal
- ENT - Otoscopy
  - Normal
  - Fail
- 1000 Hz Tympanometry or Wideband Reflectance
  - Normal
  - Abnormal
- Monitor for progressive if needed
- Antibiotics M&T if unresolved by 4 months
Take Home Messages

• At least 80% of newborn OAE screening refers are associated with abnormal wideband absorbance at birth.
• 1 kHz tympanometry has much better sensitivity and specificity than 226 Hz tympanometry for infants less than 6 months old.
• Wideband reflectance has higher sensitivity and specificity than 1 kHz tymps in newborns and adds information to interpret OAE screening results.
• Use of WBR or 1 kHz tympanometry in Newborn Screening can help in follow-up decisions.

Take Home Messages

• Normative thresholds for air and bone conduction are available across frequencies from 500-4000 Hz.
• Hand-held BC is more feasible than elastic headbands for obtaining reliable threshold values.
• Handheld bone conduction requires practice, but adequate force can easily be maintained for low thresholds.
• It is feasible to obtain clicks at high intensity, AC and BC thresholds for both ears at two or more toneburst frequencies within 1 hour in most cases during natural sleep at 2-12 weeks of age.