Integrating diagnostic information to optimize management of hearing loss in infants
- *Putting diagnostic audiology information together to plan habilitation*

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Overview

1. Joint Committee on Infant Hearing (JCIH), 2007
   - Goals for infant diagnostic testing in the context of universal newborn hearing screening (UNHS)
   - Recommendations for audiologists

2. Tools available to audiologists in the diagnostic test battery

3. Prioritizing tests & techniques to maximize efficiency in diagnosing different types of hearing loss (case studies)

4. Key messages
JCIH Goals for infant diagnostic testing in the context of universal newborn hearing screening
Joint Committee on Infant Hearing (JCIH), 2007 - UNHS timeframe goals

Universal Newborn Hearing Screening (UNHS)

- Screen by 1 mth
- Diagnose by 3 mths
- Early Intervention by 6 mths

Joint Committee on Infant Hearing (JCIH)
Year 2007 Position Statement:
Principles and Guidelines for Early Hearing Detection & Intervention Programs

Pediatrics 2007;120;898-921
Joint Committee on Infant Hearing (JCIH) 2007
- Diagnostic Audiology

• **Experience:** Comprehensive audiological evaluation of newborn and young infants who fail newborn hearing screening should be performed by audiologists experienced in pediatric hearing assessment

• **Degree, Type & Configuration:** .........to assess the integrity of the auditory system in each ear, to estimate hearing sensitivity across the speech frequency range, to determine the type of hearing loss, to establish a baseline for further monitoring, and to provide information needed to initiate amplification-device fitting.
Joint Committee on Infant Hearing (JCIH), 2007 - Diagnostic Audiology test battery

The audiological assessment should include:

1. **History**: Child and family history (risk factors & parent observations)

2. **ABR – frequency specific**: A frequency-specific assessment of the ABR using air-conducted .....and bone-conducted tone bursts when indicated.

3. **ABR – ANSD check**: Click-evoked ABR testing using both condensation and rarefaction single-polarity stimulus, if there are risk indicators for neural hearing loss (auditory neuropathy/auditory dyssynchrony) such as hyperbilirubinemia or anoxia, to determine if a cochlear microphonic is present. .....any infant who demonstrates “no response” on ABR elicited by tone-burst stimuli must be evaluated by a click-evoked ABR

4. **OAEs**: Distortion product or transient evoked OAEs.

5. **Tympanometry**: using a 1000-Hz probe tone.

6. **Behavioral Observation**: Clinician observation of the infant’s auditory behavior as a cross-check in conjunction with electrophysiologic measures.
What are we trying to measure & manage?

To accurately determine:
» Degree
» Type
» Configuration

Medical:
• Advice & treatment for conductive hearing loss
• Investigation of possible neurological conditions
• Investigate possible structural abnormalities (scans)

Amplification:
• Optimal amplification as early as possible
• Identify infants needing early referral for cochlear implant evaluations

Habilitation:
• Discuss whether the baby should learn language using auditory alone or may need visual communication
Tools available to us in the diagnostic test battery & what are we looking for?
What tools do we have in our audiological test battery?

**Behavioural**

1. Parent Observations
2. Behavioural Observation Audiometry (BOA)
3. Visual Reinforcement Observation Audiometry (VROA)

**Objective**

1. Tympanometry
2. Otoacoustic Emissions (OAEs)
3. Auditory Brainstem Response (ABR)
4. Auditory Steady State Response (ASSR)
5. Cortical Auditory Evoked Potentials (CAEPs)
What are we looking for?

- Normal hearing
- Auditory Neuropathy Spectrum Disorder (ANSD)
- Sensorineural hearing loss
  (moderate & greater)
- Mild sensorineural hearing loss
- Conductive hearing loss
- Mixed hearing loss

<table>
<thead>
<tr>
<th>Normal</th>
<th>Mild</th>
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<tbody>
<tr>
<td>ANSD</td>
<td>CHL</td>
</tr>
<tr>
<td>SNHL</td>
<td>Mixed</td>
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Maximizing test efficiency for diagnosis of different types of hearing loss (case studies)
What is the most important thing to test next?

Always assume this is the last piece of information you will get

There is more than one way you can approach this
Here are some suggestions
The first 10 minutes!
- Tympanometry & OAEs

**Scenario 1**
- Tympanometry Normal
- OAEs Present

**Scenario 2**
- Tympanometry Normal
- OAEs Absent

**Scenario 3**
- Tympanometry Abnormal
- OAEs Absent

Tympanometry & OAEs help to narrow down the “Type” of hearing loss & what intensity we should start with for ABR testing

ANSD could be ‘hidden’ under the middle-ear problems
Scenario 1
- Case 1A

What frequency & intensity will you start with for ABR?

4 kHz

Test session is over!
- Parents have to come back (increases stress)
- Your waiting time for appointments increases
Scenario 1  
- Case 1A

What frequency & intensity will you start with for ABR?

4 kHz

Test session is over
- But, you have been able to complete testing for one frequency
Toneburst ABR
- Minimum of 2 frequencies
Scenario 2
- Case 2

What intensity will you start with for 1 kHz ABR?

Tympanometry
Normal

OAEs Absent/
partially present

Mixed
Normal
Mild
ANSD
SNHL
CHL
ANSD
Mild
Normal

4 kHz

1 kHz

Time (ms)

4 kHz

1 kHz

4 kHz

1 kHz

Time (ms)
Toneburst ABR
- Bone conduction ABR

Be Aware
Conductive!

Superior Semicircular Canal (SSC) Dehiscence

Expecting SNHL

Tympanometry
Normal

OAEs Absent/
partially present

Should do Bone-conduction ABR even if you are expecting a SNHL
Bone conductor
Positioned Superior & Posterior to the ear

Bone conductor
Can be placed closer to mastoid but more likely to record artifact

Guiding Management

- Bone conduction thresholds elevated
  - SNHL confirmed

- Bone conduction thresholds normal
  - CHL confirmed

Hearing Aid prescription for SNHL according to degree & configuration of loss &
High-resolution imaging of the temporal bones to check for SSC dehiscence

Hearing Aid prescription for CHL according to degree & configuration of loss &
Scenario 2
- Case 3

Tympanometry
Normal

OAEs Absent

4 kHz

‘click’

Cochlear Microphonic (CM)

Absence of ABR
Profound loss?

Cochlear Implant?

4 ms

Time (ms)

Rarefaction click
(95 dBnHL)

Condensation click
(95 dBnHL)

Alternating click
(95 dBnHL)

Auditory Neuropathy Spectrum Disorder (ANSD)

Absence of ABR
Profound loss?
Scenario 2
- Case 3, Auditory Neuropathy Spectrum Disorder (ANSD)

Parents observed the baby waking up when there was a noise in the next room.

Behavioural Observation Audiometry (BOA) showed a ‘startle’ response with a loud rattle.

Cortical Auditory Evoked Potential (CAEP) testing arranged.
CAEPs & ANSD: Just because there isn’t synchrony to rapid stimuli at brainstem level doesn’t mean sound isn’t getting through at all

Hair cells, synapse or nerve fibers may struggle to keep up with the stimulus

CAEP is measured with a slow rate stimulus

ABR is not clearly identified unless the hair cells & nerves are firing rapidly

Stimulus rate for ABR is fast
**Scenario 2**
- **Case 3, Auditory Neuropathy Spectrum Disorder (ANSD)**

![Graph showing hearing levels and frequency]

- **CAEPs present at 65 dB SPL**
  (conversational levels)

- Later **behavioral testing showed a mild to moderate hearing loss**

**Visual Reinforcement Observation Audiometry (VROA)**
Key messages for the diagnosis & management of infants with hearing loss
Summary: Key messages

1. Infant diagnostic audiologists play a very important role in linking newborn hearing screening to timely & effective early intervention.

2. Audiologists should choose the order of each test carefully to get the maximum amount of information at each appointment.

3. A full battery of tests needs to be used to determine the degree, type, and configuration of the hearing loss. This includes ABR (air & bone conduction), OAEs, Tympanometry, behavioral observation, and CAEPs (if possible).
Thank-you for listening

谢谢你听