UNILATERAL HEARING LOSS IN CHILDREN: NEW DIRECTIONS IN MANAGEMENT

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PHONAK PEDIATRIC CONFERENCE - MUNICH
MAY 17, 2019
I. Background
Definitions of Unilateral Hearing Loss

- Loss of any degree in one ear (Bess et al., 1986)
- Non-functional hearing in one ear: Single-Sided Deafness or Limited Usable Hearing Unilaterally (LUHU)
Limited Usable Hearing Unilaterally

- Unaidable hearing
  - Profound SNHL
  - Very poor word recognition
  - Marked intolerance for amplified sounds

(Valente et al., 2002)
Prevalence of UHL

- Infants: 0.6-0.7 per 1000
  - CDC, 2014
- School-age: 3-6 per 100
  - Bess et al., 1998
- Adolescence: 14 per 100
  - Shargorodsky, et al. 2010
# Causes of or Associated Factors with UHL

<table>
<thead>
<tr>
<th>Association</th>
<th>Prevalence</th>
<th>Reference</th>
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</thead>
<tbody>
<tr>
<td>Unknown/No risk factors</td>
<td>31-54%</td>
<td>Declau et al., 2008; Ghogomu et al., 2014</td>
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<tr>
<td>Cochlear nerve deficiency</td>
<td>26-50%</td>
<td>Clemmens et al., 2013; Nakano et al., 2013</td>
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<tr>
<td>Enlarged vestibular aquaduct</td>
<td>23%</td>
<td>Clemmens et al., 2013</td>
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<td>Premature birth</td>
<td>20%</td>
<td>Haffey et al., 2013</td>
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<tr>
<td>Prolonged NICU stay</td>
<td>14-20%</td>
<td>Friedman et al., 2013; Haffey et al., 2013</td>
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</table>
Unilateral Enlarged Vestibular Aqueduct

Of children with severe-to-profound UHL who were imaged:
• 41% had abnormal temporal bone anatomy
• Most commonly EVA (14%)
• EVA most common bilaterally
• Those with unilateral EVA commonly progress to bilateral HL and >50% initially had UHL in ear without EVA

Greinwald et al., 2013
II. Psychoeducational Outcomes
“...audiologists and otolaryngologists are not usually concerned over such deafness, other than to identify its etiology and assure the parents that there will be no handicap.”

Northern & Downs, 1978
Binaural Advantages

Head Shadow = 6 – 12 dB
Binaural Advantages

Binaural Squelch

Monaural

Binaural
Today, 43% of children with UHL identified prior to 6 months of age

(Bess & Tharpe, 1986; Ghogomu et al., 2014)
Academic Outcomes Yesterday & Today

- Additional 13% have IEPs
- 50% have academic difficulty
- 35% require grade repetition
Effect of UHL on Infant and Toddler S/L Development


- Average age for first words = 12.7 months (WNL)
- Average age for first 2-word utterances = 23.5 months (significant delay)

Kishon-Rabin et al. (2015)

- 41% of infants showed delays in preverbal vocalizations
UHL and Speech-Language Scores
(Lieu, Tye-Murray, & Piccirillo, 2010)

- Sibling-controlled study of 6-12 y.o. with UHL
- n = 148
- Oral & Written Language Scales (OWLS)

Results:
- Children with UHL had poorer language comprehension, oral expression, and oral composite scores
Impact of Unilateral Conductive HL on Academic Performance
(Kesser, Krook, Gray, 2013)

- Case control survey
- School children with aural atresia
- None repeated a grade but **65% required resource help**
- 47% had IEPs
- 45% received speech therapy
But we still didn’t know WHY these children were having trouble or WHAT to do about it.

- Presented data on identification, assessment, & intervention
- Break-out groups for discussion
- Publication of proceedings

www.cdc.gov/ncbddd/ehdi/
Listening Effort & Fatigue
Growing evidence of listening effort and fatigue in children with hearing loss
Discussion paper

Listening effort and fatigue: What exactly are we measuring? A British Society of Audiology Cognition in Hearing Special Interest Group ‘white paper’

Ronan McGarrigle*, Kevin J. Munro*,†, Piers Dawes*, Andrew J. Stewart*, David R. Moore‡,*
Johanna G. Barry§ & Sygal Amitay§
Listening Effort –
Attentional requirement
necessary to understand speech
Assuming a limited effort capacity, performance on a secondary task will decrease when the primary listening task is made more difficult.

Bourland-Hicks & Tharpe, JSHLR, 2002
What does this mean for children in the real world? Depends…

- attention
- Classroom acoustics
- cognition
Fatigue

No commonly accepted definition – Can be physical or mental, subjective or objective (Hornsby, Naylor, & Bess, 2016)
Consequences of ongoing, severe, fatigue

- Inattention
- Poor concentration
- Distractibility
- Poor school achievement
- High absenteeism
Degree of HL in adults and self-reported fatigue

- No association between degree of HL and fatigue
- Strong relationship between high levels of hearing handicap (HHIE/A) and subjective fatigue

(Hornsby & Kipp, 2016)
What is the effect of hearing loss on subjective reports of fatigue in school-age children?
What they did...

- 10 children with hearing loss (CHL) and 10 age-matched peers with normal hearing (CNH)
- Subjective ratings of fatigue using the PedsQL Multidimensional Fatigue Scale
- All had normal non-verbal intelligence
- CHL had poorer language abilities than CNH
What they found...

PedSQL-MFS: Pediatric Quality of Life-Multidimensional Fatigue Scale (Varni et al., 2002)

PedsQL Score vs More Fatigue

17-30 point differences!

CHL vs CNH
Why is this important?

- The fatigue scores reported herein indicated more fatigue experienced by CHL than children with cancer, rheumatoid arthritis, diabetes, and obesity (Varni et. Al, 2002; 2004; 2009; 2010)
No association between fatigue ratings and degree of HL
Cognitive fatigue ratings are associated with language ability (CELF scores)

![Cognitive Fatigue Graph](image)

- CHL Rating vs. CELF Score
- More Fatigue on the x-axis, Better Language Ability on the y-axis
- Spearman's Rho (r) = 0.304, P-value = 0.020
Listening-Related Fatigue in Children with UHL (Bess et al., in review)
How do you feel RIGHT NOW?

1. I feel tired.

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<tr>
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<th>Some</th>
<th>Quite a bit</th>
<th>A lot</th>
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<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
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Please circle one for each statement.

2. It is easy for me to do these things.

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3. My head hurts.

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4. It’s hard for me to pay attention.

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5. I have trouble thinking.

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Bess et al., in preparation
III. Management
Hearing Technology Options for UHL

- Traditional hearing aids
- Contralateral Routing of Signal (CROS) hearing aids
- Remote microphone systems (FM and DM)
- Cochlear implants
Degree of Unilateral Hearing Loss

Severe to Profound

- **FM trial first line**
  - intervention

- Educate about hearing aid use as second line

Mild to moderately severe

- **HA trial first line**
  - intervention

- And/or FM system (classroom vs coupled with HA)
Management Drivers

- Developmental status (S/L, academics, behavior, fatigue)?
- Aidable?
- OME?
- Daycare? School? Home?
- Readiness, motivation, acceptance
- Child Factors
- Family Factors
- Environment
- Audiolologic / otologic History
- Degree of Loss

Case-by-Case Reasoning
Mothers of children with mild HL reported difficulty accepting the need for amplification because their infants responded to many sounds with or without amplification.

Moeller, Hoover, Peterson, & Stelmachowicz (2009)
IV. Moving Forward
Consensus Practice Parameter: Audiological Assessment and Management of Unilateral Hearing Loss in Children

- Identification and Audiologic Assessment and Monitoring
- Medical Management
- Technology Management
- Information Counseling for Families
- Priorities for Research
Known etiology?

- Vestibular concerns?
  - Perform vestibular assessment and vestibular rehabilitation, as needed

- Hearing, learning, or speech/language concerns?
  - Functional hearing problems?
    - Perform screenings
      - speech in noise
      - functional auditory screening (e.g., Early Listening Function)
      - localization
  - Academic or pre-academic concerns?
    - Perform academic screening (e.g., SIFTER)
    - Consider referral for educational or developmental evaluation
  - Speech or language deficits?
    - Screen for speech-language deficits (e.g., Early Language Development Scale) and, if indicated, consider speech or language therapy

Advantages/disadvantages of available hearing technologies

Speech or language concerns?

Academic or pre-academic concerns?
Hearing technologies

- Cochlear nerve deficiency?
- Cochlear implant
- Hearing aid
- CROS
- RMS

Questions:
- Aidable or seeking sound detection?
- Child able to handle?
- What are the child’s listening environments?
What we Know

- A significant portion of children with permanent MHL have been found to demonstrate difficulties observed
  - In academic settings
  - Under laboratory conditions
  - By parents and teachers
  - By the children themselves
What distinguishes

These children from these children?
What are the contributing stressors?

- Listening conditions?
- Listening effort and/or fatigue?
- Lack of early or aggressive intervention?
- Lack of effective amplification?
- Concomitant recurrent otitis media?
- Connectivity between brain networks involved in executive function, cognition, & language comprehension?
- Etiology?
How do we find those drivers?
Priorities for Research

- Separate reporting of children with minimal bilateral hearing loss and children with UHL
- Improve descriptions of study samples
  - Gender
  - Ear with UHL
  - Age at diagnosis
  - Etiology
  - Degree of hearing loss
  - Age at and type of intervention
We still have much to discover!
II. Endowment History

Seven year growth of 147%

>$7m in documented bequests
Vanderbilt Bill Wilkerson Center - Green Hills

- Audiology and Hearing Aid Clinic
- Opened July 9, 2018