Unilateral Hearing Loss in Children: What’s a Clinician To Do?

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What do you do about the child who comes in to your office with unilateral hearing loss?

If there is a subset of children with UHL that are at risk:

- Identify the risk factors (as a specialty and within each child)
- Identify the at-risk child
- Put in place resources to support that child’s language development, academic and psychosocial success

So what’s the big deal?

Children with UHL experienced barriers due to their hearing loss but learned to adapt.

Quantitatively, statistically significant differences between groups (UHL, BHL, NoHL) were not observed on the 3 main HRQOL scales (Total, Psychosocial, and Physical).


Disclosures

I receive royalties for an ear simulator I and my UVA colleagues developed and patented to teach otoscopy and tympanostomy tube placement.

What do you do about the child who comes in to your office with unilateral hearing loss?

It depends…

- Age
- CHL vs. SNHL vs. Mixed
- Coexisting (risk) factors
- Psychosocial support
- Academic progress

The Big Deal

- Prevalence/incidence of UHL
- Disabilities associated with UHL
Prevalence

- 391,000 school-aged children in the US
- 6-12 per 1,000 with USNHL
- 0-5 per 1,000 with mod.-profound USNHL


Prevalence

- The 2001-2006 NHANES survey shows an overall prevalence rate of bilateral and unilateral hearing loss (≥ 25 dB) in adolescents (ages 12-19) of 2.3% (range, 1.5-3.1%)


What’s the binaural advantage?

Hearing in noise (cocktail party effect)

Binaural advantage in noise

Summation (1.5-3 dB)


Binaural advantage in noise

Head shadow (4-7 dB)

Binaural advantage in noise

Binaural squelch (~3 dB)

What’s the binaural advantage?

Physiology of Sound Localization

Interaural time difference (ITD)

Applicable for sounds < 1500 Hz

Interaural intensity difference (IID/ILD)

Applicable for sounds > 1500 Hz

Physiology of Binaural Hearing

Central auditory pathways

Does the physiology have any practical application?

Do two ears make a difference?

The more adverse the listening condition, the greater the discrepancy between listeners with normal hearing and those with unilateral impairment.

Bess FH and Tharpe AM. Pediatrics 1984;74:206-16
USNHL in children

<table>
<thead>
<tr>
<th>Study</th>
<th>Failed</th>
<th>Resource Help</th>
<th>Combined</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bess and Tharpe, 1986</td>
<td>35%</td>
<td>13%</td>
<td>48%</td>
</tr>
<tr>
<td>Oyler et al., 1988</td>
<td>27%</td>
<td>41%</td>
<td>68%</td>
</tr>
<tr>
<td>Bovo et al., 1988</td>
<td>22%</td>
<td>12%</td>
<td>34%</td>
</tr>
<tr>
<td>Jensen et al., 1989</td>
<td>18%</td>
<td>60%</td>
<td>78%</td>
</tr>
<tr>
<td>Lieu, JEC., 2004</td>
<td>22-35%</td>
<td>12-41%</td>
<td>34-76%</td>
</tr>
</tbody>
</table>

Adapted from Tharpe, AM. Trends in Amplification. 2008;12:7-15

Options for Hearing Habilitation

- Preferential seating in class
- Individualized Education Program [IEP; 504(c)]
- FM System
- Conventional amplification
- Bone conducting technology
- Surgery

Courtesy, Cochlear Corp.

Unilateral Conductive Hearing Loss

- Should I put a tympanostomy tube in a child with unilateral OME?
- Should I perform tympanoplasty in a 7 yo with a unilateral TM perforation?
- Child with cholesteatoma?
- Child with unilateral congenital aural atresia (CAA)?
PET in unilateral OME

Lack of evidence for the benefits of surgical intervention for children with unilateral effusion and hearing loss, even if persistent.

Robb and Williamson, Paediatrics and Child Health, 2015

PET in unilateral OME

AAO-HNSF Updated Guidelines on Management of OME

Clinicians should manage the child with OME who is not at risk with watchful waiting for 3 months from the date of effusion onset (if known) or 3 months from the date of diagnosis (if onset is unknown).

Rosenfeld et al. Otolaryngol Head Neck Surg. 2015

PET in unilateral OME

Identify the at risk child

• Structural abnormalities of the TM
• Retraction of the TM
• Medial rotation of the malleus
• Attic retraction pocket
• ADHD
• Parental concern
• Behavioral problems
• Change in academic performance

Unilateral TM perforation

Who is at risk?

• Overall disability – swimming, etc
• Preferential seating in school
• Hearing aid?
• Surgery
  – Presence of cholesteatoma
  – Age: >7-8
  – Status of contralateral ear
  – Craniofacial abnormalities

Unilateral Cholesteatoma

• Surgery to eradicate disease
• Two stage or one stage - ossiculoplasty

Unilateral Congenital Aural Atresia

Options for hearing habilitation

• Observation (if unilateral)
  – Preferential seating in class
  – IEP/504(c)
  – FM system
  – Conventional hearing aid
  – Bone conducting hearing device
  – Osseointegrated bone conduction device

• Bone anchored hearing device
  – FDA approval ≥5 years old
  – Canalplasty/Atresia repair if candidate

What if ossiculoplasty fails?

• Preferential seating in class
• IEP/504(c)
• FM system
• Conventional hearing aid
• Bone conducting hearing device
• Osseointegrated bone conduction device

Study question

Does unilateral conductive hearing loss (secondary to congenital aural atresia) cause the same academic disabilities on school age children as unilateral sensorineural hearing loss?

Unilateral Congenital Aural Atresia

Should we be placing bone conducting hearing devices on all children with unilateral aural atresia?

(A MUST in children with bilateral CAA!)

Unilateral Congenital Aural Atresia
Bone conducting hearing devices on all children with uCAA?

**PRO**
- Stimulates the atretic ear/cochlear and central auditory pathways
- Supports speech and language development
- Head shadow

**CON**
- Cost
- Comfort/Compliance
- Cosmesis
- Sound localization?
- Benefit in noise?
- Perceived benefit overall?

Do BCHDs provide SL and HIN?

- Patients with UCHL showed no difference in sound localization in the aided vs. unaided conditions
- In children with UCHL, a hearing aid helped somewhat with hearing in noise but did not improve sound localization
- No improvement in objective measures but some subjective improvement

Hypothesis

Children with unilateral aural atresia have similar grade retention rates and need for resources as children with unilateral sensorineural hearing loss.

Materials and Methods

- 132 families of children with aural atresia and NO prior surgery surveyed (Bess and Tharpe survey) June – Nov. 2011
  - UVA IRB #15369
  - 91 returned (69%)
  - 23 excluded for prior atresia surgery
  - 26 excluded child < 5 yrs
  - 2 excluded for bilateral atresia
  - N=40

Survey – USNHL Control

- 48 families of children with USNHL surveyed June – Nov. 2011
  - 12 returned (25%)
  - 1 excluded for child < 5 yrs
  - N=11

Demographics

<table>
<thead>
<tr>
<th>Atresia Group (n=40)</th>
<th>SNHL Group (n=11)</th>
<th>Bess and Tharpe (n=60)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male-Female</td>
<td>23:17 (56% M)</td>
<td>7:4 (64% M)</td>
<td>0.36</td>
</tr>
<tr>
<td>Mean age (range)</td>
<td>8.9 (5-31)</td>
<td>12.6 (7-19)</td>
<td>0.034</td>
</tr>
<tr>
<td>Right/Left</td>
<td>29:11</td>
<td>4:7</td>
<td>NW</td>
</tr>
<tr>
<td>Race (C:A:H:AA)</td>
<td>30:4:0:6:0</td>
<td>9:0:1:3:0</td>
<td>0.14</td>
</tr>
<tr>
<td>Syndromic: HFM/Goldenhar</td>
<td>14</td>
<td>0</td>
<td>NW</td>
</tr>
</tbody>
</table>

Kesser et al, 2013
Audiometric data

<table>
<thead>
<tr>
<th>Atresia Group</th>
<th>SNHL Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atretic ear</td>
<td>Normal ear</td>
</tr>
<tr>
<td>AC PTA 67.2 (46-91)</td>
<td>8.1 (0-23)</td>
</tr>
<tr>
<td>BC PTA 9.9 (0-38)</td>
<td>NR</td>
</tr>
<tr>
<td>SRT 63.9 (45-75)</td>
<td>NR</td>
</tr>
</tbody>
</table>

Bess and Tharpe: PTA ≥ 45 dB HL = poorer ear
≤ 15 dB HL = better ear

Academic Progress

<table>
<thead>
<tr>
<th>Resource</th>
<th>Atresia Group (n=40)</th>
<th>SNHL Group (n=11)</th>
<th>Bess and Tharpe (n=60)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Repeated a grade</td>
<td>0 (0%)</td>
<td>2 (18.2%)</td>
<td>21 (35%)</td>
</tr>
<tr>
<td>Any resource</td>
<td>26 (65%)</td>
<td>7 (63.6%)</td>
<td>8 (13.3%)</td>
</tr>
<tr>
<td>Behavior problem</td>
<td>5 (12.5%)</td>
<td>3 (27.3%)</td>
<td>12 (20%)</td>
</tr>
</tbody>
</table>

Kesser et al, 2013

National Data


Resource assistance

<table>
<thead>
<tr>
<th>Resource</th>
<th>Atresia Group (n=40)</th>
<th>SNHL Group (n=11)</th>
<th>Bess and Tharpe (n=60)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Any resource</td>
<td>26 (65%)</td>
<td>7 (63.6%)</td>
<td>8 (13.3%)</td>
</tr>
<tr>
<td>Amplification</td>
<td>5 (12.5%)</td>
<td>3 (27.2%)</td>
<td></td>
</tr>
<tr>
<td>Speech Rx</td>
<td>18 (45%)</td>
<td>4 (36.4%)</td>
<td></td>
</tr>
<tr>
<td>FM System</td>
<td>13 (32.5%)</td>
<td>3 (27.2%)</td>
<td></td>
</tr>
<tr>
<td>IEP</td>
<td>19 (47.5%)</td>
<td>5 (45.5%)</td>
<td></td>
</tr>
<tr>
<td>Special Ed.</td>
<td>8 (20%)</td>
<td>3 (27.2%)</td>
<td></td>
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</table>

Analysis – Grade retention

<table>
<thead>
<tr>
<th>Study Comparison</th>
<th>p Value</th>
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<tbody>
<tr>
<td>Atresia (0%) vs. Bess and Tharpe (35%)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Atresia (0%) vs. SNHL (18%)</td>
<td>0.04</td>
</tr>
<tr>
<td>SNHL (18%) vs. Bess and Tharpe (35%)</td>
<td>0.32</td>
</tr>
</tbody>
</table>

Fisher's exact test

Kesser et al, 2013
**Analysis – Need for resource**

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<tr>
<td>Atresia (65%) vs. Bess and Tharpe (13%)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Atresia (65%) vs. SNHL (64%)</td>
<td>1</td>
</tr>
<tr>
<td>SNHL (64%) vs. Bess and Tharpe (13%)</td>
<td>0.001</td>
</tr>
<tr>
<td>Pooled SNHL+Atresia vs. Bess and Tharpe</td>
<td>&lt;0.0001</td>
</tr>
</tbody>
</table>

Fisher’s exact test

Kesser et al, 2013

**Discussion**

Children with aural atresia are less likely to repeat a grade but use resources to a far greater degree.

Why?

**Bess and Tharpe**

- A wakeup call...?
- Children getting the resources to succeed (64% vs. 13%)

**Grade Retention**

| SNHL (18%) vs. Bess and Tharpe (35%) | 0.32 |

**Aural Atresia is seen...**

**Classrooms are much different now...**

- Children in small groups at tables
- Projects
- So why aren’t grade retention rates better for today’s USNHL children than those in 1986?

**CHL is not as significant a disability as SNHL**

Cochlea => Otocyst

Middle ear/ear canal => Branchial apparatus

Most children with aural atresia have normal cochlear function!
CHL is not as significant a disability as SNHL

Stimulation of both central auditory pathways by the **good** ear

Adapted from Pickles, 1988, p.180.

CHL is not as significant a disability as SNHL

Stimulation of both central auditory pathways by the **atretic** ear

…test the bad ear!

Adapted from Gadland, 2004, p.72.

CHL is not as significant a disability as SNHL

Crossed acoustic reflex data (n=11)

Kesser, Gray, Hildebrand, 2011

Unilateral Congenital Aural Atresia

Options for hearing habilitation

- Observation (if unilateral)
  - Preferential seating in school
  - FM system
  - IEP
- Bone conducting hearing aid
  - A MUST in bilateral patients
  - Benefits unclear in unilateral patients
- Bone anchored hearing device
  - FDA approval ≥5 years old
- Canalplasty/Atresia repair if candidate

Should we be placing bone conducting hearing devices on all children with unilateral aural atresia?

**PRO**
- Stimulates the atretic ear/cochlear and central auditory pathways
- Supports speech and language development
- Head shadow

**CON**
- Cost
- Comfort/Compliance
- Cosmesis
- No sound localization
- Maybe some benefit in noise
- Perceived benefit overall?

A Rational Approach

- Discuss but do not discourage; support a family’s desire to trial. Benefits unproven:
  - No sound localization
  - Maybe some benefit in noise
  - Head shadow
- Monitor the hearing
- Support the child’s speech and language development and academic success
  - IEP
  - FM system
  - Sleep
Sensorineural Hearing Loss

Who is at risk?
- Degree of hearing loss
- Laterality (right ear advantage?)
- Academic progress
- Behavioral problems
- Psychosocial environment

Options for Hearing Habilitation – Sensorineural Hearing Loss
(Look familiar?)
- Preferential seating/FM system/IEP
- Conventional hearing aid
- CROS hearing aid
- Bone conducting hearing device for SSD
- Osseointegrated bone conducting device (OBCD for SSD)
- Cochlear implantation

Sensorineural Hearing Loss
Who is at risk - Mild-moderate SNHL
- Reduce background noise
- Face person talking
- Hearing aid trial

OBCD (BAHA) in Children with SSD
- Improvement in head shadow
- No sound localization
- Distraction in background noise?

CI in Children with SSD
- ...up to 3 years post-implantation, congenitally deaf children (n=3) who received a cochlear implant after 4 years of age do not demonstrate binaural hearing benefits. Tavora-Vieira D; Rajan GP. Eur Ann Otorhinolaryngol Head Neck Dis. 133 Suppl 1:S12-4, 2016 Jun
- NYU group (n=4): “varying degrees of open set SD” 1 user; 1 intermittent user; 1 non-user
**CI in Children with SSD**

- N=13; Age 1.8–15.7 years
- Improvements in objective measures of hearing in background noise and sound localization
- Results varied
- Better with acquired vs. congenital hearing loss
- Better with shorter duration of deafness

*Arndt et al., Audiol Neurotol 2015;20(Suppl 1):21-30*

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**CI in Children with SSD**

- N=21; Age 10 mos.–11.3 yrs
- Head shadow improved by 2.1 dB
- Squelch improved by 0.95 dB
- Summation improved by 0.98 dB
- No difference between children <6 vs. those >6
- High overall satisfaction, BUT
- Of 5 patients followed > 3 yrs, 3 were limited or non-users

*Thomas et al. Otol Neurotol 2017;38:496-503*

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**CI in Children with SSD**

- Small case series
- Widely heterogenous findings
- Findings often not measures of true binaural hearing

*Peters JP et al. Laryngoscope 2016;126(3):713-21*

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**Future directions**

- Identify the at risk child
- Which resource is best? For each individual child?
- Longitudinal studies to track academic progress
- *Outcome measures*??
- fMRI studies on language
- Cochlear implantation

*Fatigue*

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**Fatigue**

Children with UHL (age 5–18) and their parents completed the PedsQL Multidimensional Fatigue Scale (MFS).

- Response rates:
  - CAA 54% (43/80)
  - SNHL 29% (12/41)
  - NH 4% (2/41)
- Average age: 10 (5–18)
- Sex: 25 male, 32 female
- Amplification: 12/43 (28%) CAA
  - 5/12 (42%) SNHL
- Surgery: 29/43 CAA
**Fatigue**

*Child report of Multi-dimensional fatigue survey*

- Normative vs. uCHL
- Normative vs. uSNHL
- uCHL vs. uSNHL

**Fatigue**

*Parent report of Multi-dimensional fatigue survey*

- Normative vs. uCHL
- Normative vs. uSNHL
- uCHL vs. uSNHL

**Fatigue**

- Children with BHL show more fatigue than those with UHL and normative controls
- Children with UHL show more fatigue than normative controls
- High variance in responses - fatigue itself is multidimensional...
- Subject to recall bias

**Conclusions**

- Is the child at risk?
- Every child and every family is different - optimize resources: preferential seating, FM system, Speech Rx, IEP, Amplification
- Role of surgery?
  - Osseointegrated BC
  - Abreisa surgery
  - Cochlear implantation

**Conclusions – UHL in Children**

- Early detection of hearing loss is important
- Trial of early intervention
- Intervention may help the child meet his/her full potential
- Monitor hearing and academic progress – set the child up for success and make sure the child RESTS!!