UNILATERAL HEARING LOSS IN CHILDREN: MANAGEMENT AND OUTCOME MEASURES

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MANAGEMENT CHALLENGES IN PEDIATRIC UHL

Will intervention delay have irreversible consequences to a child with UHL?

Is there a way to know *a priori* that a child with UHL will experience academic failure or binaural deprivation in a way that affects daily living activities?

Does the implementation of a holistic UHL evaluation protocol help in determining appropriate outcomes for children with unilateral hearing loss?
PREVALENCE OF PEDIATRIC UHL

• Prevalence of UHL is of the order of 1 in 1000 in neonates, but increases to as high as 1–5% in school-aged children, though reports vary widely (Boyd 2015; Gordon et al., 2015).

• Data on severity of hearing losses are generally lacking, but the proportion of severe-profound cases is probably around 30–50%.

• The majority of cases have a genetic origin or are from perinatal complications, but include some acquired losses (largely from meningitis, head trauma, and viral infections) in older children.

• A significant proportion of children with severe/profound UHL have sensorineural deafness due to structural abnormalities of the cochlea and IAC (reportedly between 29 and 67%), and a smaller number have outer and middle ear malformations with severe conductive hearing loss.

• Newborn hearing screening programs have decreased the age at diagnosis of hearing loss. When hearing loss occurs in only 1 ear, the screening result may be overlooked or dismissed as unimportant, particularly when hearing in the opposite ear is normal (Gordon et al., 2015).
WILL INTERVENTION DELAY HAVE IRREVERSIBLE CONSEQUENCES?

MANAGEMENT CHALLENGE #1
The primary consequence of UHL is the development of an “aural preference syndrome” caused by a central overrepresentation of the better or “good” ear. (Gordon et al., 2015).

- Affected by degree of hearing loss
- Affected by the length of deprivation

The primary behavioral consequence of UHL is the disruption in spatial hearing caused by absent or conflicting binaural acoustic cues.

- Degraded sound localization
- Near complete dependence on head shadow

The consequences on “functional hearing” can vary widely.

- Persistent evidence of educational, psycho-social, linguistic, and cognitive co-occurring challenges for 30 – 60% of children with UHL.
- Subset of children with UHL who have abnormal/insufficient central processing.
BRAIN DEVELOPMENT AND NEUROPLASTICITY

Hebb’s theory

Let us assume that the persistence or repetition of a reverberatory activity (or “trace”) tends to induce lasting cellular changes that add to its stability. . . . When an axon of cell A is near enough to excite a cell B and repeatedly or persistently takes part in firing it, some growth process or metabolic change takes place in one or both cells such that A’s efficiency, as one of the cells firing B, is increased.—D. O. Hebb (1949)

Neurons that fire together...wire together
Initial state

First stimulus primes neighboring neuron

With repeated firing, links between neurons are strengthened, and they begin to fire in near synchrony, establishing a distinctive firing pattern – a memory

Neurons

Activated

Primed

Consolidation

Short-term memory

Long-term memory
USE IT OR LOSE IT?

Neural activity drives neural plasticity during development.
Developing synapses that work together flourish.

The Normal Ear

Auditory neural pathways form but do not mature.
When these “silent” immature pathways are eventually stimulated, will they respond and develop normally?

The Impaired Ear
**REVIEW OF PATHWAY**

**AUDITORY SYSTEM**

- **Peripheral System**
  - Outer Ear
  - Middle Ear
  - Cochlea
  - 8th Cranial Nerve

- **Central System**
  - Caudal & Rostral Brainstem
  - Thalamus
  - Cerebral Cortex

- Conductive loss
- Sensory or Sensorineural loss
- Neural loss
Larger than normal variability in the laterization of cortical dipoles in children receiving bilateral cochlear implants sequentially with long delay.

UNCI caused strong ipsilateral laterization.

Unilateral driven strengthening of projections to the contralateral left auditory cortex was not reversed by the addition of a second CI in long delay group.

Children with long delay sequential BICI experienced a strengthening of pathways from their hearing ear to both the ipsilateral corex and contralateral cortex.
“Aural preference syndrome” characterized by:
1. asymmetric hearing during early childhood development;
2. asymmetric speech understanding in each ear that is resistant to treatment (i.e., persisting after compensation of the initial asymmetry); and
3. deficits in binaural hearing, including sound localization, resistant to therapy of the weaker ear.

Gordon et al., 2015
SUMMARY – AURAL PREFERENCE SYNDROME

Electrophysiological and imaging studies demonstrate reorganization of the auditory pathways in UHL as a result of neural plasticity, particularly in young children with congenital hearing loss, and that such changes are associated with abnormal central auditory activation.

Monaural auditory deprivation that lasts longer than 2 years results in poorer speech understanding in the affected ear over time, both by acoustic and electrical (via a CI) stimulation, and electrophysiological studies demonstrate a link between the reduced performance and neural plasticity, also more marked in young children.

Severe-to profound UHL requires effective restoration of hearing to the impaired ear within 2 years of onset to thwart development of “aural preference syndrome.”
Can “aural preference syndrome” occur for specific frequency bands? If so, what constitutes a “long delay” in treatment?
Can “aural preference syndrome” occur for specific frequency bands? If so, what constitutes a “long delay” in treatment?
CONSEQUENCES OF PEDIATRIC UHL

• The primary consequence of UHL is the development of an “aural preference syndrome” caused by a central overrepresentation of the better or “good” ear. (Gordon et al., 2015).
  • Affected by degree of hearing loss
  • Affected by the length of deprivation

• The primary behavioral consequence of UHL is the disruption in spatial hearing caused by absent or conflicting binaural acoustic cues.
  • Degraded sound localization
  • Near complete dependence on head shadow

• The consequences on “functional hearing” can vary widely.
  • Persistent evidence of educational, psycho-social, linguistic, and cognitive co-occurring challenges for 30 – 60% of children with UHL.
  • Subset of children with UHL who have abnormal/insufficient central processing.
BINAURAL HEARING

Binaural hearing allows for:

1) the identification/localization of sound sources in space;
2) increased perception of loudness via binaural summation;
3) improved hearing in quiet and in noisy environments through head shadow and squelch effects;
4) overall communication is less tiring.

Evidence exists for a short sensitive period for bilateral input in human auditory development.

Outcomes linked to underlying changes to the developing auditory system before and after unilateral and bilateral stimulation.
HOW CAN SPATIAL HEARING BE ASSESSED?

COCKTAIL PARTY EFFECT

SOUND LOCALIZATION

THE POORER EAR MUST BE TESTED AS PART OF A BINAURAL SYSTEM
Frequencies below 1000 Hz – Interaural Time Differences (ITD)
Frequencies above 3000 Hz – Interaural Level Differences (ILD)

Known as the Duplex Theory
UHL CAUSES CONFLICTING ACOUSTIC CUES

Conflicting interaural cues degrade localization accuracy.
Stimuli = Spondee word “Baseball” recorded with male voice.
Average Level = 60 dB SPL, Level randomly roved ± 8 dB
Total Number of Trials = 150, Each loudspeaker was randomly selected 10 times.
Reinforcement = Picture puzzle piece on every trial; stickers and prizes every 20 to 25 trials
The mammalian auditory system requires much practice in early years to learn to localize to sound accurately (Mrsic-Flogel et al., 2003; Campbell et al., 2008).

Adult Localization error = 2 – 9 degrees azimuth (For review see: Middlebrooks & Green, 1991)

Children with normal hearing reach adult localization accuracy sometime after six years of age (Van Deun, et al., 2009).

Horizontal sound localization is a second or third order approximation dependent entirely upon central auditory computation of interaural differences in sound level, arrival time, and spectrum.
SOUND LOCALIZATION ACCURACY

Age-Matched Control (NH)

Perceived Target Location

Actual Target Location

-60 -40 -20 0 20 40 60

-60 -40 -20 0 20 40 60

TENNESSEE HEALTH SCIENCE CENTER
NORMAL HEARING EXAMPLES

CAAR, 13y

CAAP, 6y

Ave RMS Error: 1.2 Deg

Ave RMS Error: 6.4 Deg
UHL CAUSES CONFLICTING ACOUSTIC CUES

Older

UAAE

Audiogram

Frequency (Hz)

Threshold (dB HL)

100 1000 10000

0 20 40 60 80 100

Unaided

Target Location

RMS Error = 6.85

Response Location

-60 -40 -20 0 20 40 60

Younger

UAFF

Audiogram

Frequency (Hz)

Threshold (dB HL)

100 1000 10000

0 20 40 60 80 100

Unaided

Target Location (Deg AZ)

RMS Error = 21.17 Deg

Response Location (Deg AZ)

-60 -40 -20 0 20 40 60

Younger

UAAG

Audiogram

Frequency (Hz)

Threshold (dB HL)

100 1000 10000

0 20 40 60 80 100

Unaided

Target Location (Degs Azimuth)

RMS Error

L Hemifield

7.86 degs

R Hemifield

5.95 degs
Can performance be affected by a hearing aid?
HEARING LOSS AND HEARING AIDS AFFECT LOCALIZATION ACCURACY

**ADULTS**

UHL increases localization error (Hausler et al., 1983)

Localization is better without hearing aids than with them (Van den Bogaert et al., 2006)

Aided localization error linked to:
- Occluding ear molds (Noble et al., 1998)
- Fast-acting WDRC (Keidser et al., 2006; Wiggins & Seeber, 2011)
- Microphone directionality (Keidser et al., 2006)

**CHILDREN**

UHL increases localization error.

Younger children show greater localization error than older children (Van Deun et al., 2009; Johnstone et al., 2010)

Age at intervention (HA use) is correlated to localization performance, earlier intervention was associated with greater benefit in pediatric UHL. (Johnstone et al., 2010)
Bilateral Benefit in Localization Acuity (Deg. Azimuth) Unaided - Aided

Age (yrs) Child First Fit with a Hearing Aid

Johnstone et al., 2010

$r = -0.671, p < 0.05$
SUMMARY OF UHL AND LOCALIZATION

The degree of UHL, the age of the child with UHL, and the age at intervention affect localization accuracy and long term outcomes with personal hearing aid.

Evidence of an “aural preference syndrome” exists for children who received intervention (first hearing aid) after age 5 yrs.

It may be that partial UHL or mild-moderate UHL may affect central development more slowly than severe-profound losses.
“COCKTAIL PARTY” PROBLEM
PROBLEM

Children with UHL hearing loss are affected more than their normal hearing peers by the negative consequences of background noise and distance on speech understanding.
Binaural Redundancy or Summation: target and noise from front location (~6 dB)

Binaural Squelch: target and noise spatially separated (~3 - 4 dB)

Head Shadow Effect: monaural cue, target and noise spatially separated (~8 dB)

This figure was adapted from: Litovsky, Johnstone, and Godar (2006). “Benefits of bilateral cochlear implants and/or hearing aids in children.” IJA, 45(Supplement 1): S78-S91, page S84.

SHOULD BE TESTED AIDED AND UNAIDED IN RANDOM ORDER AND RESULTS COMPARED
TEST PROCEDURE

4 ALTERNATIVE FORCED CHOICE

ADAPTIVE TRACKING PROCEDURE

Litovsky, 2005; Johnstone & Litovsky, 2006; Johnstone et al., 2013
<table>
<thead>
<tr>
<th>Condition</th>
<th>Comparison</th>
<th>Significance</th>
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<tbody>
<tr>
<td>Listening</td>
<td>Main Effect</td>
<td>F(3,12)=77.523, p&lt;.0005</td>
</tr>
<tr>
<td>Quiet</td>
<td>POST HOC</td>
<td>Front, NIE, and NNE</td>
</tr>
<tr>
<td>Front</td>
<td>POST HOC</td>
<td>NNE</td>
</tr>
<tr>
<td>NIE</td>
<td>POST HOC</td>
<td>Front</td>
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<td>NNE</td>
<td>POST HOC</td>
<td>NIE</td>
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<tr>
<td>Age</td>
<td>Main Effect</td>
<td>Younger vs. Older</td>
</tr>
<tr>
<td>Amplification</td>
<td>Main Effect</td>
<td>Unaided vs. Aided</td>
</tr>
</tbody>
</table>
Most advantage with the Head Shadow Effect when the normal ear is in head shadow, regardless of amplification.

Binaural Redundancy Noise:
\[ t(13) = -0.469, \ p = 0.646 \]

Binaural Squelch:
\[ t(13) = -0.748, \ p = 0.468 \]

Head Shadow:
Imp Ear in Shadow
\[ t(13) = 0.333, \ p = 0.744 \]
INDIVIDUAL DATA BILATERAL BENEFIT

Example: Unaided SRT minus Aided SRT
35dB Unaided SRT - 31dB Aided SRT = 4dB

Audiogram
Frequency (Hz)
100 1000 10000
Threshold (dB HL)
0 20 40 60 80 100

Younger Group Bilateral Benefit
Younger (6-9yrs)

Older Group Bilateral Benefit
Older (10-14yrs)
Unaided

Aided, Skeleton Mold

Aided, Open Fit

RMS Error = 21.17 Deg

RMS Error = 11.62 Deg

RMS Error = 6.32 Deg

Phonak Eleva 411; WDRC, Omni
Example: Unaided SRT minus Aided SRT
35dB Unaided SRT - 31dB Aided SRT = 4dB

Audiogram

Younger Group Bilateral Benefit

Older Group Bilateral Benefit
UHL THAT ARE BEST FIT WITH OPEN-FIT DOMES

UAAE  Audiogram

UAAF  Audiogram

UAAG  Audiogram

THRESHOLD (dBA HL)
CONCLUSIONS

Group data: overall, using a hearing aid in the impaired ear does not facilitate binaural hearing when listening to speech in spatially distributed interfering speech. Is this evidence of an aural preference syndrome in partial UHL?

Group data: children with UHL primarily rely on monaural listening strategies to function in a “cocktail party environment.” SRT’s were lowest (best) when interfering speech was located near the impaired ear putting the ear with normal hearing in head shadow. Is this evidence of an aural preference syndrome in partial UHL?

Group data: children with normal hearing may utilize monaural hearing to function in “cocktail party environments.”

Individual data: aided test results in quiet do not appear to predict performance in spatially distributed interfering speech.

Individual data: could be useful in documenting efficacy of hearing aid interventions (e.g. use of OFDs).
CAN WE PREDICT ACADEMIC FAILURE?

MANAGEMENT CHALLENGE #2
CONSEQUENCES OF PEDIATRIC UHL

• The primary consequence of UHL is the development of an “aural preference syndrome” caused by a central overrepresentation of the better or “good” ear. (Gordon et al., 2015).
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WHAT IS FUNCTIONAL HEARING?

“Functional Hearing refers to the way in which an individual uses whatever hearing he or she has [in the real world].” National Consortium on Deaf-Blindness

http://nationaldb.org/
WHY IS FUNCTIONAL HEARING IMPORTANT?

People rely on functional hearing to achieve and maintain personal autonomy and power.

People rely on functional hearing to obtain and maintain intimate personal relationships.

People rely on functional hearing to navigate and communicate in complex acoustic environments.
UHL AND FUNCTIONAL HEARING

CHILDHOOD

In USA, from 30% – 50% of children with UHL in schools need special education services. (English & Church, 1999)

Greater likelihood of psychosocial and communication problems. (Stein, 1983)

Delayed development of spatial hearing. (Johnstone et al., 2010)

ADULTHOOD

Compared to peers with normal hearing, adults with UHL are no different in educational level, employment type, social problems, or substance abuse. (Colletti et al., 1988)

Continue to show impaired spatial hearing measures. (Viehweg & Campbell, 1960)
Evidence that ABRs evoked with complex sounds (like speech) may predict auditory-based communication skills (for review see: Skoe & Kraus, 2010)

cABRs are unique from click-evoked ABRs because they may predict communication skills such as reading and speech-in-noise perception.

There are published or established normative data for 3- to 4-yr olds, 5- to 12-yr olds, and young adults.

Developed the BioMARK (Biological Marker of Auditory Processing; Natus Medical http://www.natus.com/; see also http://www.brainvolts.northwestern.edu under “Clinical Technologies”), a clinical measure of speech-sound encoding.

Although cABRs can be used to assess a possible disorder, they do not provide the specificity needed to pinpoint the site of the disorder. This is because an abnormal outcome on a single measure may reflect more than one underlying cause or disorder.
• Time domain representation of a 40 msec stimulus /da/ (gray) and response (black)
• This stimulus evokes seven characteristic response peaks V, A, C, D, E, F, and O. These peaks relate to major acoustic landmarks in the stimulus.

Could you collect cABR from the normal ear in a child with UHL and predict academic problems?
Children with UHL with normal cABR values

2/5 had an IEP

Children with UHL with abnormal cABR values

4/6 had an IEP

All children were also given the Woodcock Reading Mastery Test – Revised (WRMT-R) and the Phonemic Synthesis Test

An age-matched control group with normal hearing and some with IEP was also tested
## WRMT SUB-TESTS ALIGNED WITH COMMON CORE

<table>
<thead>
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<th>WRMT</th>
<th>Common Core domain K-5</th>
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<tbody>
<tr>
<td>Letter Identification</td>
<td>Print Concepts</td>
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<tr>
<td>Phonological Awareness</td>
<td>Phonological Awareness</td>
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<tr>
<td>Word Identification; Word Attack; Word Comprehension</td>
<td>Phonics and Word Recognition</td>
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<tr>
<td>Oral Reading Fluency; Passage Comprehension</td>
<td>Fluency</td>
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</tbody>
</table>

**Reading readiness score**
**Basic skills score**
**Reading comprehension score**
**Total score**
Significant difference between UHL group and NH group for wave A amplitude.

Significant positive correlations were found for the UHL group between:
1) wave amplitudes for V, A, and O and the WRMT-R Reading Readiness score;
2) wave amplitudes for V, A, D, O and the WRMT-R Reading Comprehension score;
3) wave amplitudes for V and A and the WRMT-R Total Reading score.

Significantly lower scores for the UHL group compared to the NH group on the WRMT-R Basic Skills (Word and Passage Comprehension) and Comprehension subtests. (Listening Comprehension and Oral Reading Fluency)
Significant correlations between the WRMT-R Reading Readiness cluster percentiles and waves V, A, and O amplitudes. The triangles = children with an IEP in place; The single star = child who is home schooled; symbols filled black = children with UHL; symbols filled grey = children with normal hearing.
RESULTS SUMMARY

Significantly smaller amplitude for wave A for the UHL group as compared to the NH group.

Significantly lower scores for the UHL group compared to the NH group on the WRMT-R Basic Skills (Word and Passage Comprehension) and Comprehension subtests. (Listening Comprehension and Oral Reading Fluency)

Significant positive correlations were found for the UHL group between: wave amplitudes for V, A, and O and the WRMT-R Reading Readiness score; and between wave amplitudes for V, A, D, O and the WRMT-R Reading Comprehension score.; and between the WRMT-R Total Reading score and wave amplitudes for V and A.

No such correlations were found between wave amplitude and Woodcock scores for the NH group.

Significant correlation between the PST score and wave C amplitude.
Significant correlations between the WRMT-R Reading Readiness cluster percentiles and waves V, A, and O amplitudes. The triangles = children with an IEP in place; The single star = child who is home schooled; symbols filled black = children with UHL; symbols filled grey = children with normal hearing.
SUMMARY

Currently we cannot predict academic performance from the audiogram, localization accuracy, speech-in-noise performance.

Electrophysiological studies that exploit complex signals like speech sounds may help identify children earlier, though no evidence for sufficient specificity exists.

It seems likely however, that some children with UHL have abnormal speech processing along the central pathways of the normal ear.

A child with an “aural preference syndrome” for an ear with abnormal processing would certainly be at risk for deficits associated with impaired functional hearing.
DOES A HOLISTIC APPROACH IMPROVE OUTCOMES?

MANAGEMENT CHALLENGE #3
OUTCOMES FOR CHILDREN WITH UHL

1. Will following a formalized holistic protocol reveal areas of deficit not evidenced by the audiogram alone?

2. Will following a holistic protocol help guide the clinician in choosing the most appropriate outcome for children with unilateral hearing loss?

- Hearing Aid
- CROS
- Osseo Integrated
- FM System

- Return to Clinic
- Speech Language Therapy
- Psycho-Educational Referral
CONSIDERATIONS FOR CROS AND OSSEO-INTEGRATED DEVICES

There are psychosocial considerations that argue against fitting UHL young (pre-school) children with amplification devices including conventional CROS aids and BAHA. Older children may potentially benefit from such aids but outcomes from ipsilateral amplification (of milder losses) may be less positive in congenital cases due to the effects of monaural auditory deprivation.

Reported outcomes in adults and children using CROS aids and BAHA are mixed, with no significant improvement of localization and only modest improvements of speech understanding in noise for specific listening situations. Subjective and QoL outcomes, however, suggest greater benefit than that indicated by objective measures, particularly for BAHA.

Mounting evidence shows that more than 50% of people who receive a BAHA reject it within 5 years. Highest rejection rates for those who received a BAHA by age 10 yrs. Most common reason for rejecting a BAHA is that it reduces the ability to hear in background noise.

Current published guidelines do not recommend use of these devices in young children as they are often unable to utilize appropriate listening tactics to maximize benefit.
AUDIOLOGIC DIAGNOSTIC BATTERY

• Case History
• Otoscopy
• Tympanometry
• Otoacoustic Emissions
• Pure tone air & bone
• Speech Testing
• Speech-In-Noise Testing
LOCALIZATION PROTOCOL

• Adapted from pediatric localization studies (Martin, Johnstone, Hedrick, 2015; Johnstone, et al., 2010)
  • Number of total trials reduced to 35 (5 at 7 loudspeaker locations) as compared to 150 (10 at 15 loudspeaker locations).
  • Include one visual localization test of 35 trials for 3 and 4 yr old children

• 5 conditions: unaided sound; unaided light, aided with HA, aided with CROS, and aided with BAHA
SCREENING INSTRUMENT FOR TARGETING EDUCATIONAL RISK (SIFTER)

5 Content Areas:
1. Academics
2. Attention
3. Communication
4. Class Participation
5. Social Behavior

• Parent circles the number best representing the behavior of his/her child.
• Score Range: 3-15
  Higher the score = Better Result
Unilateral Hearing Loss Worksheet

Areas Of Testing

Parent Support | S/N | Localization | S/L eval | SIFTER Com | SIFTER Att. | SIFTER Academics | SIFTER class participation | SIFTER Social/Behavior

- Pass
- Exc
- Good
- Poor
- Visual
- Auditory
- WNL
- Abnormal
- At Risk
- At Risk
- Pass
- Pass
- Pass
- At Risk
- At Risk
- At Risk
- At Risk

NO CONCERNS

AUDIOLOGICAL

RTC
- 6 months
- 1 year
- PRN
- Other

CROS
- HA
- FM
- BAHA
- Accommodations
- Modifications
- OTHER

SPEECH/LANGUAGE

Therapy Recommended:
- CHS
- UTHSC
- School
- Private
- Other

PSYCHO-EDUCATIONAL

Referral to Psychologist
Name:

Patient name: ____________ Date: ____________ Audiologist: ____________

Developed by: Erin Plyler, AuD
DATA ANALYSIS TECHNIQUE

• Pattern-Matching Logic Technique (Yin, 2003; Trochim, 1998)
  • Compares an empirically based pattern to one or more predicted pattern(s)
  • If the patterns match, it strengthens the internal validity of the case studies
PATTERN MATCHING METHODOLOGY

1. Create predicted pattern(s)/proposition(s)
2. Collect data from different testing areas
3. Analyze to find empirical based pattern(s)
4. Compare empirical based pattern(s) to the predicted pattern(s)

* Specific outcomes correlated to specific patterns
PATIENT DATA

- Five (5) existing patients
- Age Range: 8 years to 17 years of age
- All patients had a unilateral hearing loss
- Each patient was assigned a code and symbol
<table>
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<th>Audio</th>
<th>SIFTER</th>
<th>Localization</th>
<th>SIN</th>
<th>Our Recmd</th>
<th>Dcts Recmd</th>
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Audiograms

SIFTER Results

Sound Localization

Speech In Noise Conditions
SUMMARY

Will a holistic protocol reveal areas of deficits not shown on the audiogram?

-YES-

The protocol clearly shows that a deficit/deficits are present that would not be revealed by the audiogram.
SUMMARY

Will the formalized protocol help guide the clinician in choosing the most appropriate outcome for children with unilateral hearing loss?

-YES-

Many different complexities involved.
CLINICAL IMPLICATIONS

• The decision of amplification should not be based on just the audiogram, but a variety of different tests
• Help make thorough, evidence based decisions in regards to appropriate outcomes for children and adults with UHL
• Stresses the importance for low frequency testing, specifically 125 Hz
REFERENCES

Will be provided upon request.