Unilateral Hearing Loss in Children:
Treatment Options and Guidelines for Clinical Management

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Objectives

- Unilateral Hearing Loss:
  - » Definitions
  - » Prevalence
  - » Etiology
  - » Evidence
  - » Challenges in Screening and Diagnosis
  - » Available Treatment Options
  - » Management Considerations

A Definition

Unilateral Hearing Loss:

"A permanent unilateral hearing loss exists when the diagnosis indicates there is a calculated or predicted average pure tone air conduction threshold at 0.5, 1, 2 kHz of any level greater than or equal to 20 dB HL or pure tone air conduction thresholds greater than 25 dB HL at two or more frequencies above 2 kHz in the affected ear with an average pure tone air conduction threshold in the good ear less than or equal to 15 dB"

(National Workshop on Mild and Unilateral Hearing Loss 2005)

Prevalence

- » Approximately 1/3 of infants with congenital HL identified from NHS programs have unilateral hearing loss
- » Estimates vary due to variations in method of ascertainment:
  - In some cases only SNHL are counted, omitting permanent CHL
  - Poor follow up
  - Definition of UHL

Lieu 2010 Seminars in Hearing

Prevalence

- UHL at birth:
  - » 0.83/1000:
    - 0.41/1000 well baby
    - 3.2/1000 NICU

    (2013-2014 CDC EHDI database)
- UHL at school age::
  - » 2.5-3%

  (Bess, Dodd-Murphy and Parker 1998, Shargrodsky et al 2010)

Possible reasons for increased prevalence form birth to school-age

- Cases of progressive or late onset HL
- Differing definitions/inclusion criteria
- Low follow up rates in newborn screening programs underestimating true prevalence

1. Adapted from Chapter 10 Children with Unilateral Hearing Loss in Comprehensive Handbook of Pediatric Audiology, Seewald and Tharpe ed. 2011

Progression from Unilateral to Bilateral Hearing Loss (Neault, 2005)

- Two groups of newborns move from unilateral refer to bilateral hearing loss status:
  1. Those who actually had bilateral HL at time of screening.
     - Mild HL in ear that passed screening because stimulus level may have exceeded intended SPL in infant's ear
     - Hearing loss mild enough to pass the intended screening intensity in the better ear.
  2. Those who had UHL at time of screening but develop bilateral HL later
     - Unilateral permanent loss at time of screening with development of bilateral permanent loss because of underlying etiology.

Progression of UHL

According to Lieu (2018):

- For children with mild to severe UHL at presentation, the risk of progression in the worse hearing ear may be as high as 40%
- Risk of progression to bilateral HL approaches 20%
- Risk for progression various by etiology with greater risk of progression for:
  - » Children with enlarged vestibular aqueduct (EVA),
  - » Cochlear canal bony stenosis (or hypoplastic cochlear nerve)
  - » CMV

Etiology of UHL[1]

- Inner Ear Malformations
  - » Enlarged Vestibular Aqueduct Syndrome (EVAS)
  - » Cochlear Nerve Deficiency (aplasia, hypoplasia)
- Congenital Cytomegalovirus (CMV)
  - » Most common cause of nongenetic HL and leading cause of UHL in children (Nance, 2007)
- Congenital Aural Atresia and Microtia
  - » Occurs in 1/10,000 births; 70% unilateral (Schuknecht, 1989)

1. Adapted from Chapter 10 Children with Unilateral Hearing Loss in Comprehensive Handbook of Pediatric Audiology, Seewald and Tharpe ed. 2011

Etiology of UHL[1]

- Sudden SNHL
  - » Prevalence lower in children than adults. Approximately 3.5% of sudden UHL occurs in children under 14 years (Nakashima & Yanagita, 1993)
  - » Approximately 85% of cases in children are unilateral (Roman et al., 2001)
- Bacterial Meningitis
  - » Approximately 30% of cases of HL from bacterial meningitis are unilateral (Kutz et al. 2006)
- Viral/Bacterial Mumps
  - » Approximately 80-90% of cases of HL caused by mumps are unilateral
  - » Leading cause of UHL <1967 when vaccine introduced
- Prematurity

1. Adapted from Chapter 10 Children with Unilateral Hearing Loss in Comprehensive Handbook of Pediatric Audiology, Seewald and Tharpe ed. 2011

Diagnostic Challenges in UHL

- Ability to perform speech recognition testing in poorer ear difficult prior to two years of age
- Masked testing is difficult before 2 1/2-3 years of age using behavioral audiometric procedures (VRA).

Advantages of Binaural Hearing

- Improvements in:
  - Threshold
  - Localization
  - Speech understanding in adverse listening conditions
- Facilitated by:
  - Binaural summation
    - Sound presented to two ears perceived louder than if same sound presented monaurally (3dB at threshold, 6dB at 30dB SL)
    - 18% improvement in monosyllabic word recognition and 30% improvement in sentence scores
  - Head shadow effect
    - Provides reduction of sound intensity between ears of 6-12dB for complex signals
  - Binaural release from masking
  - Precedence effect

Frequently Cited Concerns with UHL

- Greater difficulty hearing in noise
  - » Children with UHL lose binaural advantage and are likely to have more difficulty understanding speech in noisy or reverberant environments
  - » Children require a higher SNR than adults for speech recognition (Corbin et al 2016) and are constantly learning in a noisy environment.
- Difficulty with localization
  - » Lack of interaural time and intensity cues
    - Interaural Time Difference (ΔIT) Below 800Hz
    - Interaural Level Difference (ΔIL) Above 1600Hz
- Concerns regarding auditory deprivation:
  - » Reorganization of auditory system following reduced stimulation
  - » Poorer word recognition in ears without amplification

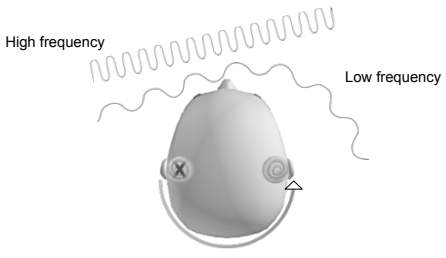
(Silverman et al., 2006)

Head Shadow Effect

- With the loss of auditory function in a unilateral configuration, sounds originating from the same side as the impaired ear will be present in the "shadow" of the head.
- Long wavelength (low frequency sounds) readily "bend" around the head and are largely unaffected by the baffle of the head.
- Short wavelength (high frequency sounds) are "reflected" or "baffled" by the head and are then attenuated by this process, creating difficulty.
- When one considers that consonant sounds contain much of the meaning of speech, the Head Shadow Effect is the primary or root cause of the communication problem.

Slide Courtesy of Cochlear Corporation

Head Shadow Effect



-3dB Head Shadow Effect

Slide Courtesy of Cochlear Corporation

Studies on Children with UHL

- Quigley & Thomure, 1969
  - » 116 children with UHL not receiving services
  - » Majority of children (82%) had HL <26dBHL
  - » As a group, identified children scored below expected levels on Sanford Achievement Test
  - » Children were one grade behind expected levels based on age
- Bess et al (1984,1986)
  - » 35% of children with UHL failed at least one grade compared to 3.5% with normal hearing
  - » 13% in need of resource room help
  - » 20% described by teachers as having behavioral problems

Studies on Children with UHL

- Majority of studies since that time have reported variety of difficulties in speech, language, academics and behavior in children with UHL:
  - » Oyler, 1988; Bess, Dodd-Murphy & Parker 1998;Culbertson & Gilbert 1986, Jenson, Johansen & Borre, 1989, yet…
- A few studies have reported no difficulties:
  - » Hallmo, Moller Lind & Tonning, 1986; Stein,1983; Tieri, Masi, Ducci & Marsell, 1988.

The Unilateral Hearing Loss in Children Study(UHLCS) Lieu, 2013

- Funded by NIDCD (Recruitment period 2005-2010)
- Case controlled study of children 6-12 years of age
- Compared children with UHL to siblings with normal hearing
- Standardized measures of achievement and language controlling for cognitive ability
- PTA of >30dB for 3 consecutive test frequencies
- Children with any medical condition associated with cognitive impairment and fluctuating or temporary HL excluded
- One objective was to determine whether school aged children with UHL demonstrate significantly poorer language skills than their siblings with normal hearing

The Unilateral Hearing Loss in Children Study (UHLCS) Lieu, 2013

- Primary Outcome Measure:
  - » Oral and Written Language Scales (OWLS)
- Analysis of 74 sibling pairs showed children with UHL had significantly poorer scores than normal siblings in:
  - » Language comprehension (91 vs 98 for controls; p=0.003)
  - » Oral expression (94 vs 101 for controls; p=0.007
  - » Oral composite (90 vs 99 for controls; p <0.001)
- No differences found between right or left UHL
- No differences found for varying degrees of HL
- Using sibling pairs allowed control over many factors such as: family, genetic, socioeconomic, parental educational level

Studies on Children with UHL

- Children with limited useable hearing unilaterally (LUHU) (those with poor word recognition or greater than severe unilateral SNHL are at greater risk (Culbertson & Gilbert, 1986; Lieu et al., 2013)
- Large variability in outcomes
  - » Although on average, children with UHL are 10 times more likely to repeat a grade or require academic assistance in school, nearly 70% of children with UHL are considered academically successful, not requiring educational support (Bess & Tharpe, 1984; Lieu, 2004;Oyler, Oyler & Matkin, 1988)

Picou et al., 2020

Treatment Options

Device Options for Children with UHL

- Conventional air conduction hearing aid
- Remote microphone systems (FM/DM)
  - Sound Field systems
  - Personal systems
- Bone Conduction Devices:
  - Conventional bone conduction hearing aid
  - BAHA Softband
  - Surgical BAHA (>5 years of age and depended on skull thickness)
  - BAHA Attract (>5 years of age and depended on skull thickness)
  - MedEl AdHear (non-surgical, adhesive)
- Middle ear Implants (e.g. MedEl Bonebridge)
- Cochlear Implants

Management
Guidelines and Recommendations

National Workshop on Mild and Unilateral Hearing Loss (2005)

- In 2005, The Early Hearing and Detection Program at the Center for Disease Control and Prevention (CDC) in collaboration with Marion Downs Hearing Center:
  - » Recognized that children with moderate to profound HL were successfully being identified through NBHS
  - » Acknowledged that a “substantial body of evidence” exists to suggest that mild and UHL has detrimental effect on children’s development
  - » Convened to discuss issues related to identification, assessment and intervention for infants and children with mild or UHL

National Workshop on Mild and UHL Technology Considerations

- Once degree, configuration and type of HL can be definitively determined those with slight and mild degrees of long-standing HL in the speech frequency range may be considered for hearing aids
- For those with UHL, CROS aids might not be recommended until the child is able to control his environment
- Other technologies such as bone anchored hearing aids (BAHA or transcranial hearing aids may be considered for children 5 years or older.
  - » However, it was noted that there are no available data to support either of these technologies

Cincinnati Children's Hospital Medical Center **Best Evidence Statement (BESt)** Audic

Date published/posted: August 20, 2009

Recommendations

1. In ***all*** children with unilateral SNHL:

- It is recommended managing providers discuss the potential impact of unilateral hearing loss (UHL) with the child and family to help them understand potential gains, realistic goals, costs, and physical requirements of amplification so they can make an educated decision regarding interventions
- Be cognizant of cost, which can be an issue in providing a HA or FM system. Most insurance companies do not cover HAs or other amplification devices, nor do they pay for FM systems as covered benefits and many schools do not uniformly provide FM systems for children with UHL

Cincinnati Children's Hospital Medical Center **Best Evidence Statement (BESt)** Audio

Date published/posted: August 20, 2009

2. It is recommended, whether or not amplification is provided, that the child and care team (family, health care professionals, clinicians and school personnel) consider monitoring the impact on functional, educational, and behavioral performance as well as academic performance and behavior (family selected outcomes) in the classroom to guide care decisions

(Lieu 2004, McKay 2008, Local Consensus, McKay 2005)

Cincinnati Children's Hospital Medical Center **Best Evidence Statement (BESt)** Audiol

Date published/posted: August 20, 2009

In children with severe to profound unilateral SNHL:

3. It is recommended that school-aged children with severe to profound unilateral sensorinerual hearing loss (USNHL) be fit with an FM system as the first line of amplification technology (Kenworthy 1990 [3b], Updike 1994 [4b]). Select an FM system with the most open fit to decrease occlusion in the good ear (Kopun 1992 [4b]).

4. It is recommended that provision of a HA in children with severe-profound UHL be on a case-by-case basis (Kiese-Himmel 2002 [4b], McKay 2002 [4b]).

Audiol

Best Evidence Statement (BESt)

Date published/posted: August 20, 2009

In children with mild to moderate unilateral SNHL:

5. It is recommended that children with mild to moderate sensorineural UHL be fit with a hearing aid (FM ready) as the first line intervention (Kenworthy 1990 [3b], McKay 2002 [4b], Shapiro 1977 [4b], McKay 2005 [5b]).
6. It is recommended provision of an FM system with or without a hearing aid be discussed with the family (McKay 2002 [4b], Local Consensus [5]).

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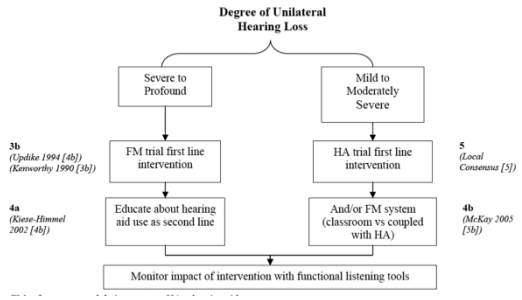
Best Evidence Statement (BESt)

Date published/posted: August 20, 2009



American Academy of Audiology
Pediatric Amplification Guidelines (2013)

The purpose of providing amplification for children is to minimize the negative impacts of hearing loss on communication development and academic achievement.

Amplification systems should, therefore be considered for any type or degree of HL that could possibly interfere with normal developmental processes, including minimal/mild or unilateral hearing loss or Auditory Neuropathy Spectrum Disorder.

Summary of Evidence for Audiologic Candidacy
AAA Pediatric Amplification Guidelines 2013

- *Children with aidable unilateral hearing loss should be considered candidates for amplification in the impaired ear due to evidence for potential developmental and academic delays.*
- *Children with unilateral hearing loss are at greater risk than children with normal hearing for speech and language delays and academic difficulties.*

Quick Practice Guideline

May 2018

Tools and considerations for assessing and managing unilateral hearing loss in children

Authors:
Bagatto, M., DesGeorges, J., King, A., Kitterick, P., Laurnagaray, D., Lewis, D., Roush, P., Sladen, D., Tharpe, A.

Retrieved on 1.20.2020 from:
https://www.phonakpro.com/content/dam/phonakpro/gc_hq/en/resources/evidence/white_paper/documents/technical_paper/quick_practice_guideline_btb_uhl.pdf

Quick Practice Guideline

The following table briefly summarizes these available technologies and provides consideration of their relative advantages and limitations.

Device	Benefit	Disadvantage	Considerations
Conventional	Improved detection of sound on the affected side	Might not provide benefit for profound SNHL	Supports fitting on affected side if degree is mild to severe Environmental sound awareness might be the goal for more severe degrees of loss
Bone conduction	Suitable for unilateral microtial atresia. Might be considered in lieu of a conventional CROS	No fitting protocols Sound quality for profound UHL is inferior to routed signal in CROS	Surgical option not available for children under the age of 5 years in many areas
Contralateral Routing of Signal (CROS)	Improved detection of speech on the side with UHL in quiet Improved speech understanding in noise when speech is the dominant signal on the side with hearing loss	Reduced speech understanding when noise is the dominant signal on the side with hearing loss Unlikely to help localization	Ability to manage device and listening environment – especially for young children Need to avoid occluding normal hearing ear – use large vent or open fitting No data available regarding outcomes of non-implanted bone conduction devices for children with profound UHL. Transcranial CROS requires custom earmold that sits in bony portion of canal
Cochlear implant (CI)	Improved speech recognition on affected side Improved overall speech recognition in noise Improved localization (adults and children)	Surgical intervention Might prevent candidacy from future advancements in hearing restoration	Must have compelling audiologic data showing that the ear to be implanted will not benefit from other non-surgical forms of technology Subject to medical and regulatory candidacy criteria
Personal Remote Microphone System (RMS)	Improved access to primary auditory signal Addresses effects of noise, and reverberation	For each transmitter/microphone, system is beneficial for hearing a single talker only. Multiple microphones are needed for multiple talkers Requires talker's cooperation Use of a personal receiver might affect compliance for some users	Deciding which ear to fit depends on multiple factors, including degree of hearing loss in poorer ear and ability to ensure an open fitting in the better ear

Quick Practice Guideline

Device	Benefit	Disadvantage	Considerations
Classroom Audio Distribution System	Improved access to primary auditory signal Delivers primary talker's voice evenly throughout the learning space through one or more loudspeakers Benefits all listeners in the learning space	Each transmitter/microphone system is beneficial for hearing a single talker only. Multiple microphones are needed for multiple talkers Requires the talker's cooperation Limited portability and flexibility for use across a range of environments Might not provide the same degree of benefit as personal or desktop RMS	Improved signal-to-noise ratio, but dependent upon classroom acoustics Systems are more effective when classroom acoustics have been optimized
Desktop RMS	Improved access to primary auditory signal Addresses effects of noise, distance, and reverberation on speech understanding	Each transmitter/microphone system is beneficial for hearing a single talker only. Multiple microphones needed for multiple talkers Requires the talker's cooperation Limited flexibility for use across a range of environments Use of a desktop receiver might affect compliance for some users	

International Journal of Audiology

ISSN: 1499-2027 (Print) 1708-8186 (Online) Journal homepage: https://www.tandfonline.com/loi/iija20

Consensus practice parameter: audiological assessment and management of unilateral hearing loss in children

Marlene Bagatto, Janet DesGeorges, Alison King, Padraig Kitterick, Diana Laurnagaray, Dawna Lewis, Patricia Roush, Douglas P. Sladen & Anne Marie Tharpe

International Journal of Audiology, 58:12, 805-815, DOI:10.1080/14992027.2019.1654620

Table 2. Hearing technologies for children with unilateral hearing loss.

Device	Benefit	Disadvantage	Considerations
Conventional	Improved detection of sound on the affected side for mild to severe UHL	Might not provide benefit for profound SNHL. Not clear whether prescriptive targets for binaural paediatric fittings require adjustments for UHL fittings	Environmental sound awareness might be the goal. Speech perception scores alone should not be used to determine whether a hearing aid recommendation should be made
Bone-Conduction	Suitable for unilateral microtia/atresia. For profound UHL, the device can be considered in lieu of conventional CROS device	No fitting protocols for these devices. Sound quality of bone-conducted signal for profound UHL is inferior to routed signal in a CROS system	Surgical BCD is not available for children under the age of 5 years in many jurisdictions
CROS	Improved detection of speech on side with UHL in quiet. Improved speech understanding in noise when speech is the dominant signal on side with hearing loss	Reduced speech understanding when noise is the dominant signal on the side with hearing loss. Unlikely to help localisation	Requires ability to manage device and listening environment – especially for young children Need to avoid occluding normally-hearing ear when possible –large vent or open fitting required Remote microphone system might still be required to improve hearing ability in noise, especially in educational settings No data available regarding outcomes of non-implanted bone conduction devices for children with profound UHL No evidence of the benefits of transcranial CROS fittings to children Transcranial CROS requires a custom earmold that sits in bony portion of canal, which can affect feasibility of fittings for some children
CI	Improved speech recognition on affected side, improved overall speech recognition in noise, improved localisation (adults and children) and improved health related quality of life (adults)	Might prevent candidacy from future advancements in hearing restoration Performance might depend on amount of aural rehabilitation	Length of auditory deprivation can have a negative impact on performance Must have compelling audiologic data showing that the ear to be implanted will not benefit from other non-surgical forms of technology Cost is prohibitive and reimbursement is an obstacle in non-socialized health care models All children considered for CI must have appropriate imaging to establish normal temporal bone anatomy
Personal RMS	Addresses effects of noise, distance and reverberation on speech understanding	Multiple microphones are needed for multiple talkers. Requires the talker's cooperation Use of a personal receiver could affect compliance for some users	Decisting which ear to fit depends on multiple factors, including degree of hearing loss in the poorer ear and the ability to ensure open fitting in better ear
Classroom Audio Distribution System	Improved access to primary auditory signal. Delivers primary talker's voice evenly throughout the learning space, through one or more loudspeakers. Benefits all listeners in learning space	Multiple microphones are needed for multiple talkers. Requires the talker's cooperation Limited portability and flexibility for use across a range of environments	Systems are most effective when classroom acoustics have been optimised
Desktop RMS	Improved access to the primary auditory signal. Addresses effects of noise, distance, and reverberation on speech understanding	Multiple microphones are needed for multiple talkers. Requires the talker's cooperation. Flexibility for use across a range of environments is limited	

BCD: bone-conduction device; RMS: remote microphone system; SNR: signal-to-noise ratio; CI: cochlear implant; CROS: contralateral routeing of signal; UHL: unilateral hearing loss.

LSHSS

Prologue

Evidence-Based Practices and Outcomes for Children with Mild and Unilateral Hearing Loss

Elizabeth A. Walker[a]

Language, Speech and Hearing Services in Schools Vol.51 Jan.2020

- 10 articles
- Overview of current research and clinical practice for children with mild bilateral
- and unilateral hearing loss

Language, Speech and Hearing Services in Schools Vol.51 Jan.2020

"*Growing body of evidence that children with so-called 'minimal hearing loss' are at risk for academic difficulties, behavioral problems and language and listening deficits.*"

Despite this evidence, these children continue to present challenges for several reasons:

1. NBHS not intended to identify degrees of HL <35-40dBHL complicating early diagnosis
2. Lack of consensus among researchers and clinicians on optimal intervention strategies for mild and UHL because:
   a. Not all children with experience delays and
   b. There is insufficient evidence that hearing assistive technology or early intervention will lead to better outcomes if there are delays (Walker, 2020)

Evidence re Amplification for UHL

Evidence regarding amplification

Davis et al, 2001:

- Survey of 150 families with MBHL or UHL
- Return rate 40%
- Of 27 fitted with unilateral amplification
  - » 26% reported wearing it all of the time
  - » 4% wore only at school
  - » 50% reported never wearing it
- Of 36 children with mild bilateral hearing loss
  - » 44% reported wearing it all the time
  - » 3% wore only at school
  - » 25% reported never wearing it

Evidence regarding amplification

McKay et al 2002:

- Children's Hospital of Philadelphia
- 28 children (Ages 2-17) with UHL fit with HA on poorer ear
- Criteria for amplification was hearing loss of 25-65dBHL in poorer ear and usable speech recognition in that ear.
- Loaner hearing aids used
- 20/28 returned survey
- Majority of children liked the HA
- Parents reported they were happy with decision to get a hearing aid and majority wished they had done it sooner

Support for recommending air conduction HAs to promote development of bilateral auditory pathway in infants with mild to severe USHL (Bagatto, 2020)

Johnstone, Nabelek and Robertson (2010):

- Improved sound localization abilities in children with USHL if they received HA <5 years of age
- Children who were aided at 9 years of age or older had impaired localization on the affected side

Gordon, Wong and Papsin (2013):

- Reorganization of bilateral auditory pathways could be avoided if use of unilateral CI in early childhood did not occur for more than 1.5 years prior to getting a second CI

Management Considerations for UHL by Type and Degree

- Is hearing loss conductive or sensorineural?
- What is the degree of HL on affected side?
- Is hearing loss on affected side "aidable"
  - » Is there sufficient hearing to provide audibility
  - » What is speech recognition on affected side?
    - (Difficult to determine in very young children)

Remote Microphone Systems: Consider alone or in combination with HA or CI

Touchscreen Mic

Roger Pen

Roger Select

Images from www.phonak.com

Personal Remote Microphone Systems (FM/DM) Consider for severe/profound UHL (or other "unaidable" UHL)

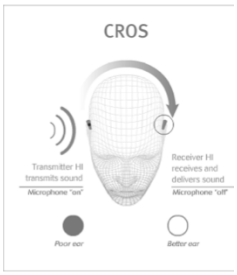
- If receiver in normal hearing ear, important that device be non-occluding
- Difficult in past due to size of receiver and tubing
- Newer device options with slim tube make open fit possible
- Verify by testing speech recognition with and without device in ear

Classroom Audio Distribution Systems

Important to consider audibility of teacher's voice but those of peers as well

Contralateral Routing of Signal (CROS)

- HA microphone on the impaired side picks up sound and delivers to the good side
- Cumbersome in past with wired components, newer wireless devices available
- Useful only for older children who can control their environments (by positioning or turning off mic on side of noise)
- May be good option for children who have cochlear nerve hypoplasia or aplasia and not candidates for CI



LSHSS

Clinical Focus

Considerations for Choosing Microphone Technologies for Students With Limited Useable Hearing Unilaterally

Erin M. Picou,[a] Hilary Davis,[a] and Anne Marie Tharpe[a]

Language, Speech and Hearing Services in Schools Vol. 51, 74-83 Jan.2020

Conclusions (Picou et al 2020):

- Results of review demonstrate discrepancy between CROS benefits measured via survey and lab settings
  - » Scientific rigor, age of studies (outdated equipment designs) or differences between lab and classroom listening situations
- It is likely that CROS systems provide more benefit than prior lab studies suggested. However complicated by several factors:
  - » Location of the student's seat
  - » Classroom configuration
  - » Whether peers, the teacher or everyone are talkers of interest

Conclusions (Picou et al 2020):

- There is no microphone system recommendation that will work for all school-aged children with LUHU
- A combination of RM and CROS could accommodate most students in most classrooms.
- More work is necessary to confirm the hypothesized effects of CROS and RM in classrooms.

Bone Conduction Hearing Devices

- Many more options to consider:
  - » Baha on softband
  - » Surgical Baha
  - » MED EL Bonebridge
  - » Cochlear BAHA Attract
  - » MED EL AdHear
  - » Cochlear OSIA
- Bone conduction devices generally accepted in cases of permanent, unilateral CHL
  - » Benefit in cases of profound UHL in children less clear
    - Reports of improved quality of life
    - Sound quality may be inferior to CROS system
    - As with CROS, child must be old enough to control listening environment

Baha® on Softband (Non-surgical Option)

Retrieved 1/21/20 from: https://www.cochlear.com/us/en/home/products-and-accessories/baha-system/baha-softband-and-soundarc

https://thebahablog.com/2018/06/14/guest-blog-baha-system-hazel-infant-hearing-loss/hazel_baha2/#main

The Baha® Connect System

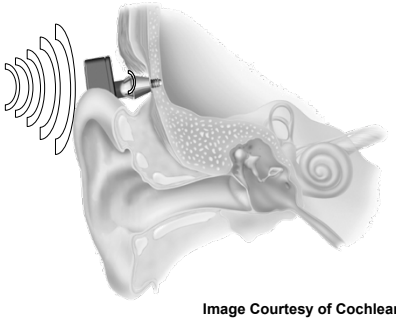
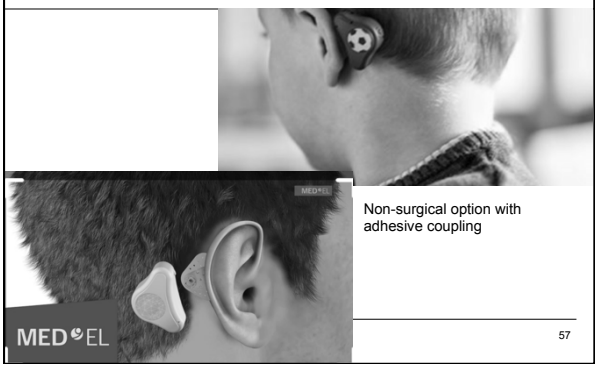


Image Courtesy of Cochlear Corporation

Baha Attract System

- Transcutaneous bone conduction

Med EL AdHear System



Non-surgical option with adhesive coupling

57

What about cochlear implants for unilateral hearing loss?

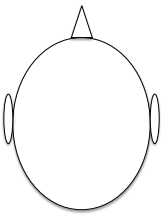
Cochlear Implantation in Unilateral Hearing Loss: Pediatric Studies

- Consistent device use (Hassepass et al 2013, Polonenko et al 2017, Greaver et al 2016)
- Improvements in localization (Hassepass et al 2013, Sharma et al 2016, Rahne et al 2016)
- Improvements in QoL (Sladen et al 2017, Thomas et al 2017)
- Better hearing in noise (Hassepass et al 2013, Sladen et al 2017, Thomas et al 2017, Rahne et al 2016)
- Improved speech perception in the implanted ear (Sharma et al 2016, Sladen et al 2017, Greaver et al 2016, Rahne et al 2016)
- Cortical reorganization to age appropriate levels (Sharma et al 2016)

Not FDA approved. Investigational

Rationale for Consideration of CI in SSD

- Localization
- Speech in noise
- Auditory effort



Cochlear Implantation In Pediatric Cases of Unilateral Hearing Loss

PUHL Clinical Trial

Aim: To investigate the effectiveness of cochlear implantation in children with moderate-to-profound unilateral hearing loss.

Kevin Brown, MD PI

Lisa Park, AuD, Co-Investigator and Study Coordinator

PUHL Study

- Children age 3.5-6.5 years
- Typically developing
- PTA of ≥ 70 dB HL in one ear and normal hearing in the contralateral ear
- Aided CNC word score of ≤ 30% in the ear to be implanted
- No evidence of cochlear nerve deficiency (CND)
- No evidence of ossification
- No significant malformations
- English is the primary language

Not FDA approved. Investigational Device Exemption

Device

- MED-EL SYNCHRONY Flex28 or Flex24
  - Array choice at surgeon's discretion
- SONNET Speech Processor
  - All programmed in FS4
  - Omni-directional mode with wind noise reduction disabled
  - No use of RONDO processors

Participant Characteristics

ID	Age at CI	Years Profound	Etiology	ID	Age at CI	Years Profound	Etiology
01	6.50	1.87	Infection	11	6.17	3.55	Unknown
02	6.42	1.37	Unknown	12	3.97	1.79	cCMV
03	4.59	1.41	Trauma	13	4.90	1.26	Unknown
04	6.13	6.18	Mondini	14	5.46	4.99	Unknown
05	4.74	4.79	Waardenburg	15	5.58	0.79	Unknown
06	12.85*	2.78	Unknown	16	3.79	3.83	Unknown
07	4.00	4.04	Mondini	17	5.45	5.49	Unknown
08	6.50	4.62	Unknown	18	3.58	3.63	Unknown
09	6.50	6.62	Unknown	19	3.93	3.96	Unknown
10	7.09*	2.38	cCMV	20	3.61	3.66	cCMV

DA Approval of compassionate use;

Not FDA approved. Investigational Device Exemption

Test Battery

- Speech Perception
  - Early Speech Perception (ESP-S) and CNC
    - Aided with the contralateral ear masked
    - Normal Hearing Ear alone
    - CI only via direct connect
- Localization
- Parent and Child questionnaires

Not FDA approved. Investigational Device Exemption

Test Battery

- Quality of Life Scales:
  - Peds-QL (parent and child)
    - Measures general, cognitive, and sleep fatigue
  - SSQ for Children with Impaired Hearing
    - Measures perception of speech understanding, spatial hearing, and quality of sound
  - Bern Benefit in Single Sided Deafness Questionnaire
    - Compares perception of hearing with and without a hearing device
    - Modified for children with the permission of Dr. Kompis

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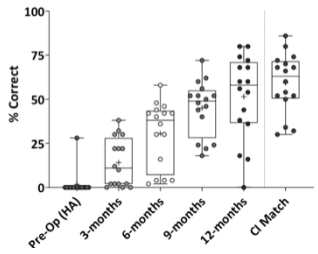
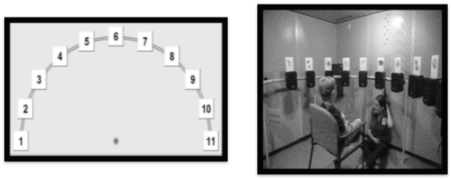
CNC Word Scores

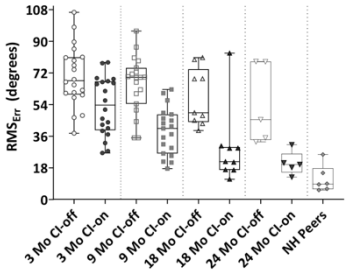


Localization

Localization



Localization



Conclusions

Hearing in Noise

- Without the CI: Noise on the implanted side presents a significant struggle.
- With the CI: Benefits of head shadow and binaural summation may be emerging by 6 months of CI use.
- Speech perception CI alone does not seem to predict head shadow benefit.

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Conclusions

Speech Perception

- Pre-operatively with a hearing aid: Open set speech perception is non-existent and closed set skills are limited.
- After 3 months of CI use: Open set skills are emerging in the implanted ear.

Localization

- Without a device: Poor for the majority of subjects
- With a device: Already improved by 3 months of listening experience

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Conclusions

QoL

- Children report greater difficulty with fatigue than their parents
- With a hearing aid: Subjects report little to no benefit
- With a CI: Benefit is reported by 3 months of use
  - Quality of life scores are showing improvement in speech understanding, spatial hearing, and quality of hearing

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Conclusions

- There is not a single management approach in cases of UHL. Decisions often need to be made on case by case basis depending on the child's needs and family's preference.
- There is little evidenced-based research to guide the clinician on management of UHL
- Important to remember that infants who fail screening in one ear are at risk for bilateral hearing loss
- Families should be informed of the risk factors associated with UHL and the options available for management and intervention
- The family's values and wishes must be given priority when working with infants and young children, regardless of degree or configuration. (Bagatto, 2020)

Unanswered Questions…

- If amplification is recommended for children with mild to moderate UHL, what age should it begin?
- Will speech, language and academic delays found in children with UHL be ameliorated by early hearing aid fitting and consistent use for those with 'aidable' hearing in poorer ear?
- Will speech, language and academic delays found in children with profound UHL be ameliorated by use of CI in children with severe/profound UHL?
- Is there a time sensitive window in which hearing interventions must take place in order for children to be able to receive benefits such as improved localization ability or listening in noise?

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THANK YOU!

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References

- American Academy of Audiology
  Pediatric Amplification Protocol (2013)
- National Workshop on Mild and Unilateral Hearing Loss (2005)
  - » http://www.cdc.gov/ncbddd/hearingloss/conference.html
- Cincinnati Children's Hospital Medical Center
  - » Audiologic management for children with permanent unilateral sensorineural hearing loss. Best Evidence Statement (BESt) 2009
- Bagatto, M., DesGeorges, J., King, A., Kitterick, Laurnagaray, D., Lewis, D., Roush, P., Sladen, D. and Tharpe, A. Consensus practice parameter: audiological assessment and management of unilateral hearing loss in children. *International Journal of Audiology*, 58:12, 805-815, DOI: 10.1080/14992027.2019.1654620
- Bagatto, M. Audiological Considerations for Managing Mild Bilateral or Unilateral Hearing Loss in Infants and Young Children. *LSHSS,* 2020 51, 68-73

References

- Management Guidelines for Children with UHL
  The Center for Childhood Communication at CHOP, McKay, S. and Iyer, A. ASHA Leader 2005
- Picou, E., Davis, H. and Tharpe, AM, Considerations for Choosing Microphone Technologies for Students with Limited Useable Hearing Unilaterally. *LSHSS,* 2020;51, 74-83
- Porter, H. and Bess, F. Children with Unilateral Hearing Loss. In R. Seewald and AM Tharpe (eds.) Comprehensive Handbook of Pediatric Audiology (pp 175--191), San Diego, California. Plural Publishing 2010
- Lieu, J. Children with Unilateral Hearing Loss, Seminars in Hearing 2010
- Lieu, JE, Tye-Murray, N., Karzon, RK, Piccirillo, JF, Unilateral Hearing Loss is Associated with Worse Speech and Language Scores in Children. Pediatrics 2010;125 (6)