Unilateral Cochlear Implants in Children Single Sided Deafness (SSD)

Current Research

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Learning Objectives

- Understanding the impact of single sided deafness in Children
- Discuss traditional and new treatment options for SSD in children.
- Determine the best candidates for unilateral CI in Children.

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Unilateral hearing loss in Children

- Incidence of hearing loss at birth is 1.86/1000 newborns in the US.
- 30-40% are unilateral.
- **Single sided deafness**
  - 1 in 3,700 newborns
- 2 to 5 of 1000 children and teenagers
  - Delayed onset on congenital hearing loss or acquired hearing loss such as infection, trauma, or ototoxic exposures (including noise exposure).

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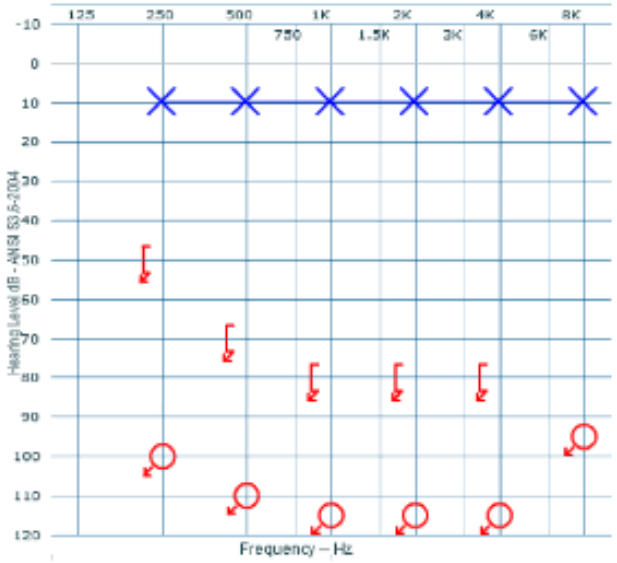
Single Sided Deafness (SSD)

- The most severe form of UHL.
- Defined as Severe to profound SNHL in the worse ear, with normal to no more than mild hearing loss in the better ear (PTA less than 30 dB)



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Consequences of Unilateral Hearing Loss in Children

- In the past… ☹
  - UHL was of little consequence b/c it was assumed that speech and language presumable developed appropriately with one normal hearing ear.
  - Prior to NBHS: UHL was usually not diagnosed until school aged when children would get school screening.

Consequences of SSD in Children

- Children with SSD often struggle with:
  - Speech perception in noise
  - Sound localization due to lack of binaural hearing effects such as the **head shadow effect** (intensity is decreased to opposite ear due to head), **squelch effect** (hearing in noise), and **binaural redundancy effect** (better hearing w/ two ears vs. one).
  - Delayed speech-language development
  - Increased risk for psychosocial difficulties
  - Cognition (lower IQ)
  - Increased behavioral problems
  - Inferior functioning in educational settings compared with normal hearing peers.

Academic effects of SSD

- 1980-1990s
  - Studies of UHL showed
    - 24-35% of children w/ UHL failed grades compared to 3% of their normal hearing peers.
    - 22-59% of children required additional educational assistance
    - 36% UHL had IEPs

Traditional Treatment for Children with SSD

- Until recently clinical practice consisted of observation alone
- Preferential seating in the classroom
- Contralateral routing of the signal (CROS hearing aids)
- Bone anchored hearing aids/Osseo-integrated hearing devices.

Limitations to traditional treatment options for SSD

- **Bone anchored hearing aids/Osseo-integrated hearing devices**
  - Can be uncomfortable
  - Inconvenient for early intervention
    - Babies being held
    - Feedback
  - Surgical intervention options are limited to age 5 and older
  - Decreased benefit in high frequencies
- **CROS hearing aids**
  - Require good patient feedback
  - Two devices instead of one
  - Compromising normal hearing in better ear.
  - Decreased benefit in high frequencies.
  - Not recommended until about age 8 years of age
- Can still bed difficult to localize
- Do not restore binaural hearing
- **DELAY IN INTERVENTION DUE TO LIMITAIOTNS! ☹**

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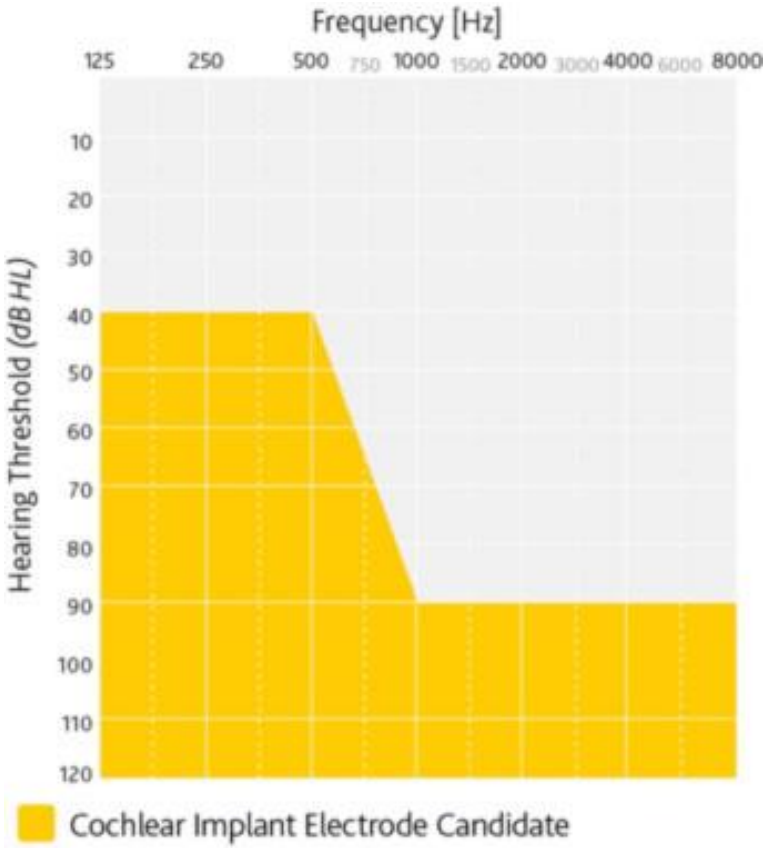
Traditional CI Candidacy

- **Adults**
- **Individuals 18 years of age or older**
  - Moderate to profound sensorineural hearing loss in both ears
  - Limited benefit from amplification defined by preoperative test scores of ≤ 50% sentence recognition in the ear to be implanted and ≤60% in the opposite ear or binaurally.
- **Children (2-17 Years)**
  - Severe to profound sensorineural hearing loss in both ears
  - Limited benefit from binaural amplification
  - Multisyllabic Lexical Neighborhood Test (MLNT) or Lexical Neighborhood Test (LNT) scores ≤ 30%
- **Children (9-24 Months)**
  - Profound sensorineural hearing loss in both ears
  - Limited benefit from binaural amplification



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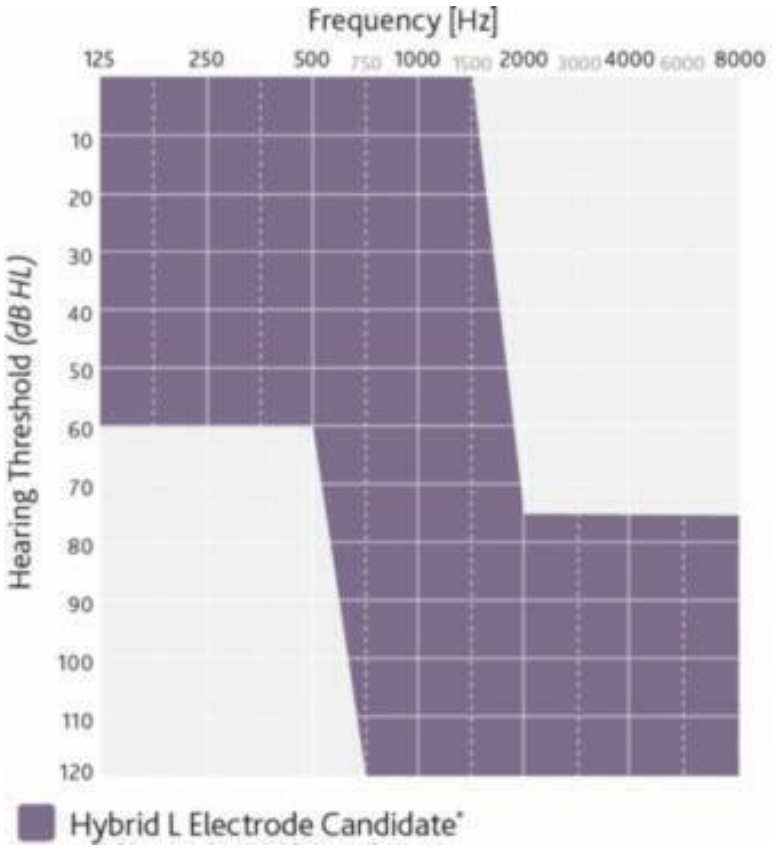
Unilateral CI options for Adults 18+

- Cochlear America‘s The Nucleus Hybrid System is indicated for unilateral use in patients aged **18 years and older** who have residual low frequency hearing sensitivity and severe to profound high-frequency sensorineural hearing loss
- limited benefit from appropriately fitted bilateral hearing aids.
- Precipitously sloping hearing losses
- Less than 60% word rec in ear to be implanted
- The contralateral ear’s no better than 80% word rec.
- Moderately severe to profound mid- to high frequency hearing loss in the contralateral ear.
- Acoustic component

- Still requires bilateral hearing loss.
- Not recommended for children! ☹



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FINALLY! FDA Approval for Unilateral CI in Children!!

- In 2019, the US FDA approved cochlear implantation for children w/ SSD!! ☺
- Med-El: Synchrony and Synchrony 2
  - 5 years and older
  - Profound in one ear
  - Normal to mild hearing loss in opposite ear.
- Cochlear Americas
  - January 2022
  - Cochlear Nucleus 24 implant systems.

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Criteria for Unilateral CI Candidacy

- When should you consider CI for Children?
  - **BEST** candidates:
    - Children born with SSD – off-label implantation that is considered medically necessary
      - CMV
    - Progressive hearing loss
    - Sudden hearing loss
    - High risk for losing hearing in contralateral ear
      - EVA
  - Less than 3 years of age is ideal – prime neural plasticity.
  - Short duration of deafness
    - Less than 3.5-4 years duration of deafness
    - Later implantation is possible but REALISTIC EXPECTATIONS need to be established.
  - **MOTIVATED & COMPLIANT PARENTS!!**

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Limited Research ☹

- Although Unilateral CI have been implanted all over the world off-label, there is limited research at this time to support the benefit of unilateral CI in SSD.
- This may create a barrier in counseling parents at this time.

Concerns: Integrating stimulus input to the brain

- **Concerns:** Integrating acoustic and electric stimulation
  - Can auditory processing centers integrate both an acoustic signal and an electric signal from a cochlear implant together?
  - Can the brain combine two signals in a beneficial manner to permit binaural hearing?
  - **Bimodal hearing**:
    - Well-demonstrated with bimodal hearing in which patients with a cochlear implant benefit from a hearing aid in the contralateral, better hearing ear.
  - **Residual hearing**:
    - Due to the tonotopic organization of the cochlea, patients hear both acoustically with their residual low-frequency hearing and electrically in the mid- and high-frequency ranges with their cochlear implant.
  - **Hybrid Cochlear Implants**

Acoustic/Electrical stimulation cont'd

- If the brain can combine information from acoustic hearing and a cochlear implant in the same ear, and from a CI and hearing aid on opposite ears, it seems reasonable to expect that information from a cochlear implant could be integrated with normal hearing from an opposite ear as well.

- **Adults:**
  - Tinnitus treatment:
    - Unilateral CI to treat tinnitus, also found to significantly increase ability to understand speech in noise & improved localization.
  - increase speech in quiet from an average of 4% to a mean of 55% with the cochlear implant alone at 12 months after implantation.
  - Increased quality of life
  - Better localization
  - Better hearing in noise

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Cochlear Implantation in Children With Single-Sided Deafness
A Systematic Review and Meta-analysis

Liliya Benchetrit, MD1; Evette A. Ronner, BA2; Samantha Anne, MS, MD3,4; et al

- Is cochlear implantation in children with SSD associated with improved audiological and patient-reported outcomes?

- Systematic review and meta-analysis of **12 studies that evaluated 119 children** with SSD.

- **Objective of the study:**
  - To evaluate the **audiological & patient-reported outcomes** in children who underwent cochlear implantation for SSD and to
  - Assess the association between time of implantation, subjective outcomes, and cochlear implant device use rates.

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Inclusion Criteria Cont'd

- Inclusion criteria
  - Younger than 18 years
  - Diagnosis of SSD for which they underwent a cochlear implantation
  - **Single-sided deafness** was defined as a 1-sided pure-tone average of 90 dB or higher or an ABR of 80 or higher, and WNL in the contralateral ear.
  - At least 1 outcome of interest measured numerically: speech perception, sound localization, device use, and patient-reported outcomes.
  - **Of the 526 articles reviewed, 12 (2.3%) met the selection criteria.**
  - This study analysis was conducted from January 4, 2020, to April 4, 2020.

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Research Measures

- **Speech perception in quiet**
  - Measured as a proportion of correct responses.
  - CI side only w/ masking in normal ear.
  - Age appropriate Monosyllabic and multisyllabic word lists were used.
- **Speech perception in noise**
  - Measured as decibel (dB) signal to noise ratio for speech reception threshold.
  - background noise was presented at 60 to 70 dB
  - measured the sound and noise from 0° azimuth testing configuration
- **Sound localization**
  - Measured in degree of localization error
  - Researchers used between 3 to 13 loudspeakers, with various stimuli presented at 55 to 70 dB.
  - preoperative vs postoperative (1-2.2 years) binaural performance and were combined for a meta-analysis

Research Measures Cont'd

- **Device use**
  - Measured by hours per day of device use.
- **Patient-reported outcomes**
  - Measured by the speech, spatial, and qualities of hearing scale (SSQ).
    - standardized questionnaire that has been shown to be sensitive in measuring bilateral and unilateral hearing abilities across 3 domains, thus yielding 3 subscale scores (score range: 1-10, with the highest score indicating the best result).
    - The SSQ has been validated for adults, children, and parent proxy for young children
    - The questionnaire items were scored from 1 to 10, completed before and after (1-3 years) cochlear implantation
    - Outcomes were divided into congenital vs. acquired SSD

Results

- **Speech in Noise:**
  - Perception in noise reported a clinically meaningful improvement with the implant among all patients.
  - Lack of improvement in the 4 children with congenital or perilingual SSD to long duration of deafness (>4 years in congenital SSD; >7 years in perilingual SSD)
  - For 5 children, lack of improvement due to a ceiling effect created by the excellent normal hearing ear
  - lack of improvement in 1 child attributed to long duration of deafness (>6.8 years), which included the critical period for binaural hearing development

Results cont'd

- **Speech in Quiet**
  - Overall, 81.0% experienced improvement from the cochlear implantation, and their mean scores ranged from 56% to 100%.
  - Children who received an implant after a shorter duration of deafness (3 years) had greater improvement than children who underwent an implantation later.
  - Long duration of deafness > 4-7 years, was the probable reason for lack of observed improvement

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Results Cont'd

- Localization
  - 88.7% children showed improvement in sound localization 1 to 2 years after cochlear implantation, with mean reduction of 24.78° in localization error.
  - All studies reported clinical improvement of sound localization at most angles.
  - Device use was associated improved sound localization

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Results Cont'd

- **SSQ**
  - The questionnaire items were scored from 1 to 10
  - completed before and after (1-3 years) cochlear implantation
  - outcomes were divided into congenital vs acquired SSD
  - Children with acquired SSD had statistically significantly greater improvements compared with children with congenital SSD in the speech measure.
  - implantation age of children with congenital SSD was statistically significantly younger than the implantation age of children with acquired SSD
    - 5.1 [3.2] years vs. 9.5 [3.0] years.
  - The median duration of deafness was significantly shorter in the acquired SSD group vs. the congenital SSD group
    - 1 [0.8-1.5] years vs. 4.1 [1.7-6.8] years

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Results Cont'd

- **Device Use**
  - Eleven studies reported on both the duration of deafness and the frequency of device use after a 1-2 years.
  - Of 101 children, 74.3% used the device regularly.
  - The remaining 20.8% reported limited device use
  - 4.9% became nonusers
    - Nonuse was explained by the lack of advantage
    - unpleasant electrical stimulation
    - lack of adequate family support
    - duration of deafness (9.0 [5.8-11.7] years vs 3.3 [1.2-5.4] years regular users

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Clinical Findings:

- Speech perception in noise and quiet as well as sound localization improved after cochlear implantation. Patient-reported audiological outcomes and cochlear implant use rates were higher among children with a **shorter duration of deafness**.

- Meaning Findings from this study can be used to inform future research efforts, refine cochlear implantation candidacy criteria, and aid in family counseling and shared decision-making.

Discussion

- Comprehensive clinical data that evaluate the outcomes are limited.
- Most children (79.6%) experienced improved speech perception in noise and in quiet after cochlear implantation
- Lack of improvement among children with congenital SSD who received an implant after age 4 years
- Children with shorter duration of deafness (<4-7 years) acquired a clinically significant speech recognition improvement after implantation.
- Most children (88.7%) showed improvement in sound localization 1 to 2 years after cochlear implantation.

Limitations & Future Research Opportunities

- Small sample sizes
- Unable to control for the heterogeneity of the pediatric SSD population, including:
  - Deafness cause
  - Onset and duration
  - Age at implantation,
  - Device manufacturers
  - Extent and availability of social and rehabilitative support systems

Binaural Vs. Monaural Hearing

Binaural Hearing

Monaural Hearing

Binaural Hearing and Single-Sided Deafness (SSD)

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References

- Benchetrit, L., Ronner, E., Anne, S., Cohen, M. Cochlear Implantation in Children With Single-Sided Deafness: A Systematic Review and Meta-analysis. *JAMA Otolarygology-Head & Neck Surgery.2021;147: 58-69.*
- Benchetrit, L., Ronner, E., Anne, S., Cohen, M. Cochlear Implantation in Children With Single-Sided Deafness: A Systematic Review and Meta-analysis. *JAMA Otolarygology-Head & Neck Surgery.2021;147: 58-69.*
- *Cochlear implant candidacy. Cochlear. (2022, June 22). Retrieved October 12, 2022, from https://www.cochlear.com/us/en/professionals/products-and-candidacy/candidacy/cochlear-implant*
- *Cochlear Implants for Single-Sided Deafness. Med-El. (2022, Oct 12). Retrieved October 12, 2022, from Cochlear Implants for Single-Sided Deafness*
- *Anne, S.,* Unilateral Hearing Loss in Children: Evaluation and Management. Audiology Online. (2021, Dec 13). Retreived October 10, 2022. Audiologyonline.com.

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