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Single Sided Deafness/ Asymmetric Hearing Loss in the Pediatric Population

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Prevalence of Single-Sided Deafness (SSD) in the United States

- **The prevalence of SSD is estimated to impact up to 345,000 individuals in the United States.** SSD was defined as normal hearing (pure-tone average [PTA] of ≤25 dB) in one ear and severe or worse hearing (PTA > 70 dB) in the other ear.

- Although the prevalence of SSD is overall low at 0.14%, it should be noted that an additional 0.13% of participants in this cohort had unilateral moderately severe hearing loss. In total, this study identifies over **650,000 individuals who could potentially benefit from hearing rehabilitation**, including a cochlear implant, for SSD or for unilateral moderately severe hearing loss.

- Prevalence ranges from estimates like the above, 350K to 650k. **Age and range in definitions between UHL and SSD can significantly impact this number.**

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Etiology of SSD/UHL in Pediatrics

In **pediatrics** 35-60% of cases: unknown etiology

- Progression to other side, 10-15%
- Most common causes:
  - Inner ear anomalies
  - Cochlear nerve deficiency, 26-50%
  - Enlarged vestibular aqueduct (EVA), 23-41%
  - TORCH, CMV infections, Meningitis
  - Genetic causes

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Why is Losing Hearing From One Ear a Problem?

The brain relies on binaural cues to best determine location of sound, as well as, to provide optimal signal-to-noise ratio (SNR) to decipher speech in a background of noise.

1. Timing cues (low frequency)
2. Amplitude cues (high frequency)
3. Spectral cues

Patients cannot always control their orientation to noise in their environment.

1. Buss E, Dillon MT, Rooth MAet al. Effects of Cochlear Implantation on Binaural Hearing in Adults With Unilateral Hearing Loss. Trends Hear 2018; 22:2331216518771173.

2. Firszt JB, Reeder RM, Holden LK, Dwyer NY, Asymmetric Hearing Study T. Results in Adult Cochlear Implant Recipients With Varied Asymmetric Hearing: A Prospective Longitudinal Study of Speech Recognition, Localization, and Participant Report. Ear Hear 2018; 39:845-862.

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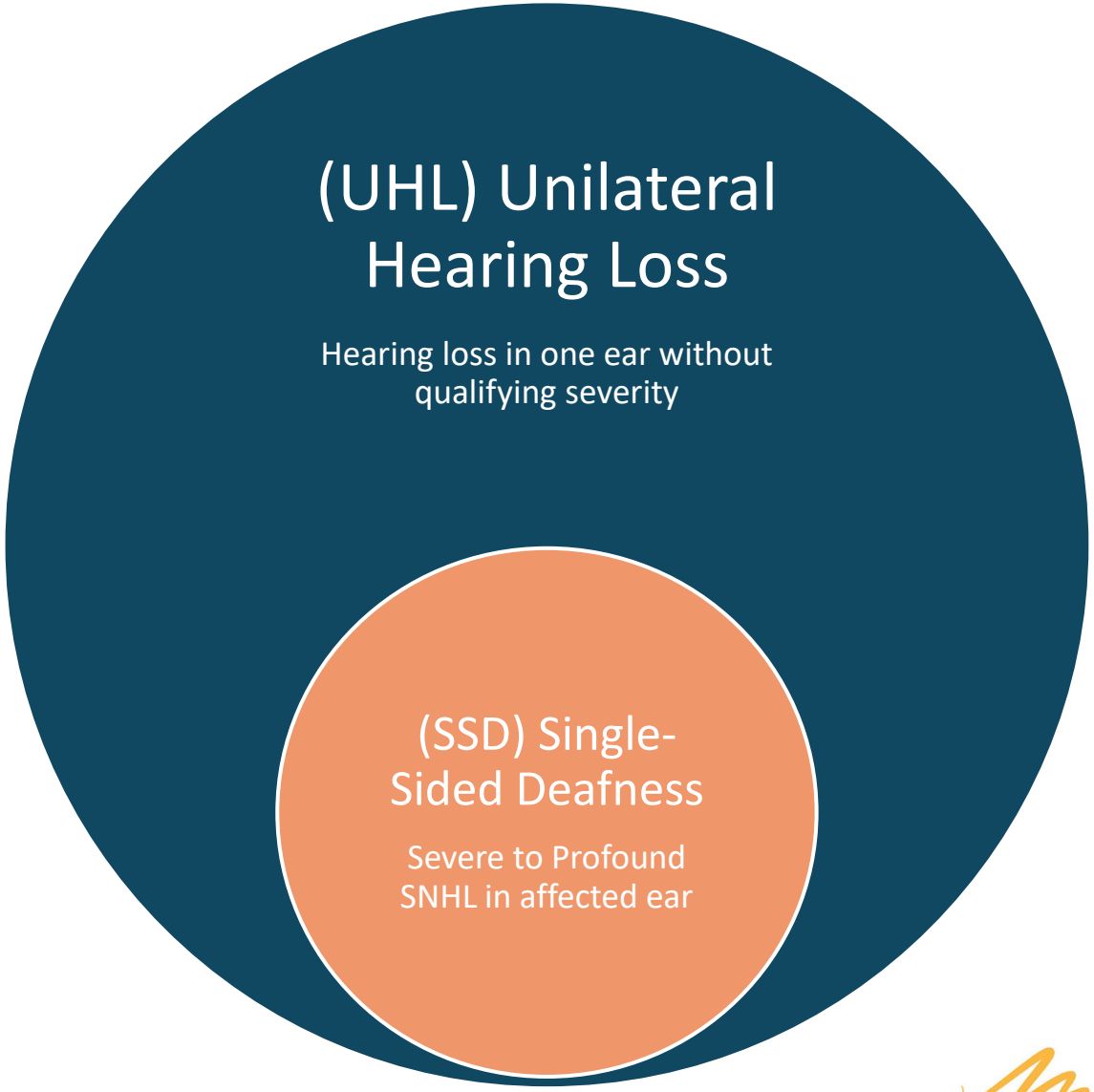
Terminology – UHL & SSD

- Both UHL and SSD are terms used when describing hearing loss in one ear.
- UHL refers to any type of hearing loss in one ear without describing severity, with normal to near-normal hearing in the opposite ear.
- SSD refers specifically to severe to profound SNHL in one ear and normal to near-normal hearing in the other.
- Research may be collected & reported on patients with UHL or only SSD depending on scope.
- Professionals will use UHL or SSD interchangeably while patients may be more familiar with SSD.



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Potential Impacts of UHL/SSD

Auditory

- Difficulty **hearing in background noise**[1]
- Decrease in **sound localization** leads to **both communication** and **safety concerns**[1]
- Difficulty understanding **soft speech**, especially from the side of the poor hearing ear or at a distance[2]
- Loss of **balanced hearing** ("stereo-effect")

Psychosocial

- Increased **auditory effort** (cognitive load) leading to increased **fatigue**, speech confusion, decreased **short term memory retention**[3]
- Poorer **quality of life**, reduced **social interactions**[4]
- Often accompanied by severe **tinnitus**[5]

Specific to Children

- Higher risk of **delayed speech-language development**[6,7]
- **Social, emotional, cognitive and behavioral challenges**[8]
- Requires **greater auditory effort** in the classroom, leads to **fatigue**, negatively affects **learning**, more likely to **fail a grade**[9]

1. Arndt S, Aschendorff A, Laszig R, Beck R, Schild C, Kroeger S, Ihorst G, Wesarg T: Comparison of pseudo binaural hearing to real binaural hearing rehabilitation after cochlear implantation in patients with unilateral deafness and tinnitus. Otol Neurotol 2011a;32: 39–47.
2. Firszt JB, Reeder RM, Holden LK, Dwyer NY, Asymmetric Hearing Study T. Results in adult cochlear implant recipients with varied asymmetric hearing: a prospective longitudinal study of speech recognition, localization, and participant report. Ear Hear 2018;39:845–862
3. Alhanbali, S., Dawes, P., Lloyd, S., & Munro, K. J. (2017). Self-Reported Listening-Related Effort and Fatigue in Hearing-Impaired Adults. *Ear and Hearing*, *38*(1), e39–e48. https://doi.org/10.1097/AUD.0000000000000361
4. Wie, O. B., Pripp, A. H., & Tvete, O. (2010). Unilateral deafness in adults: Effects on communication and social interaction. *Annals of Otology, Rhinology and Laryngology*, *119*(11), 772–781.
5. Vermeire K, Van de Heyning P. Binaural hearing after cochlear implantation in subjects with unilateral sensorineural deafness and tinnitus. Audiol Neurootol 2009;14:163–171.
6. Lieu JE, Tye-Murray N, Karzon RK, Piccirillo JF. Unilateral hearing loss is associated with worse speech-language scores in children. Pediatrics 2010;125:e1348–e1355.
7. Sangen, A., Dierckx, A., Boudewyns, A., Dhooge, I., Offeciers, E., Wouters, J., Desloovere, C., & van Wieringen, A. (2019). Longitudinal linguistic outcomes of toddlers with congenital single-sided deafness—Six with and twelve without cochlear implant and nineteen normal hearing peers. *Clinical Otolaryngology*, *44*(4), 671–676. https://doi.org/10.1111/coa.13347
8. Lieu, J. E. C. (2018). Permanent Unilateral Hearing Loss (UHL) and Childhood Development. *Current Otorhinolaryngology Reports*, *6*(1), 74–81. https://doi.org/10.1007/s40136-018-0185-5
9. Hornsby BW, Werfel K, Camarata S, Bess FH. Subjective fatigue in children with hearing loss: some preliminary findings. Am J Audiol 2014;23: 129–134

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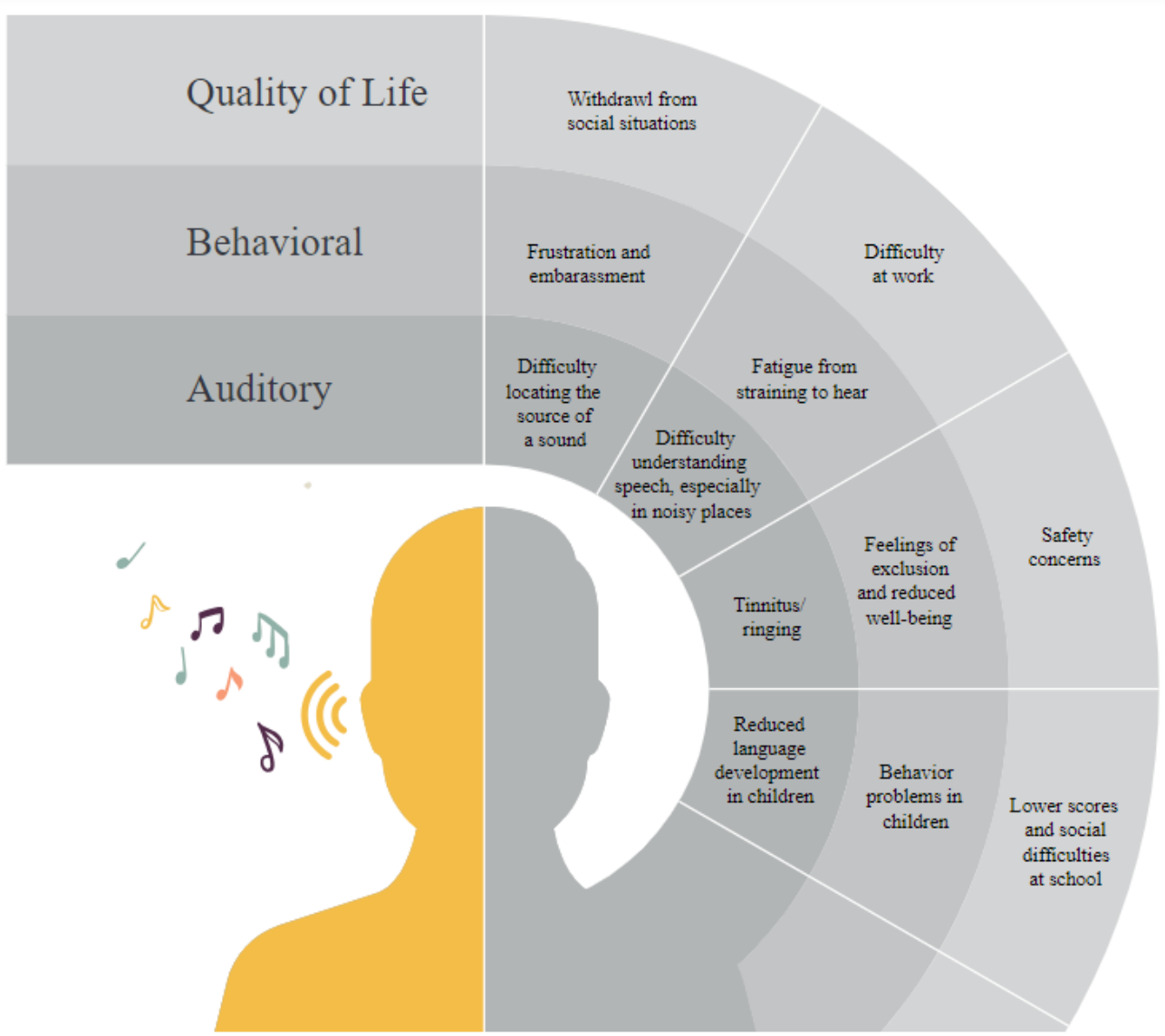
Potential Impacts of UHL/SSD

Some impacts are more immediately noticed.

Others may become more of a concern over time.



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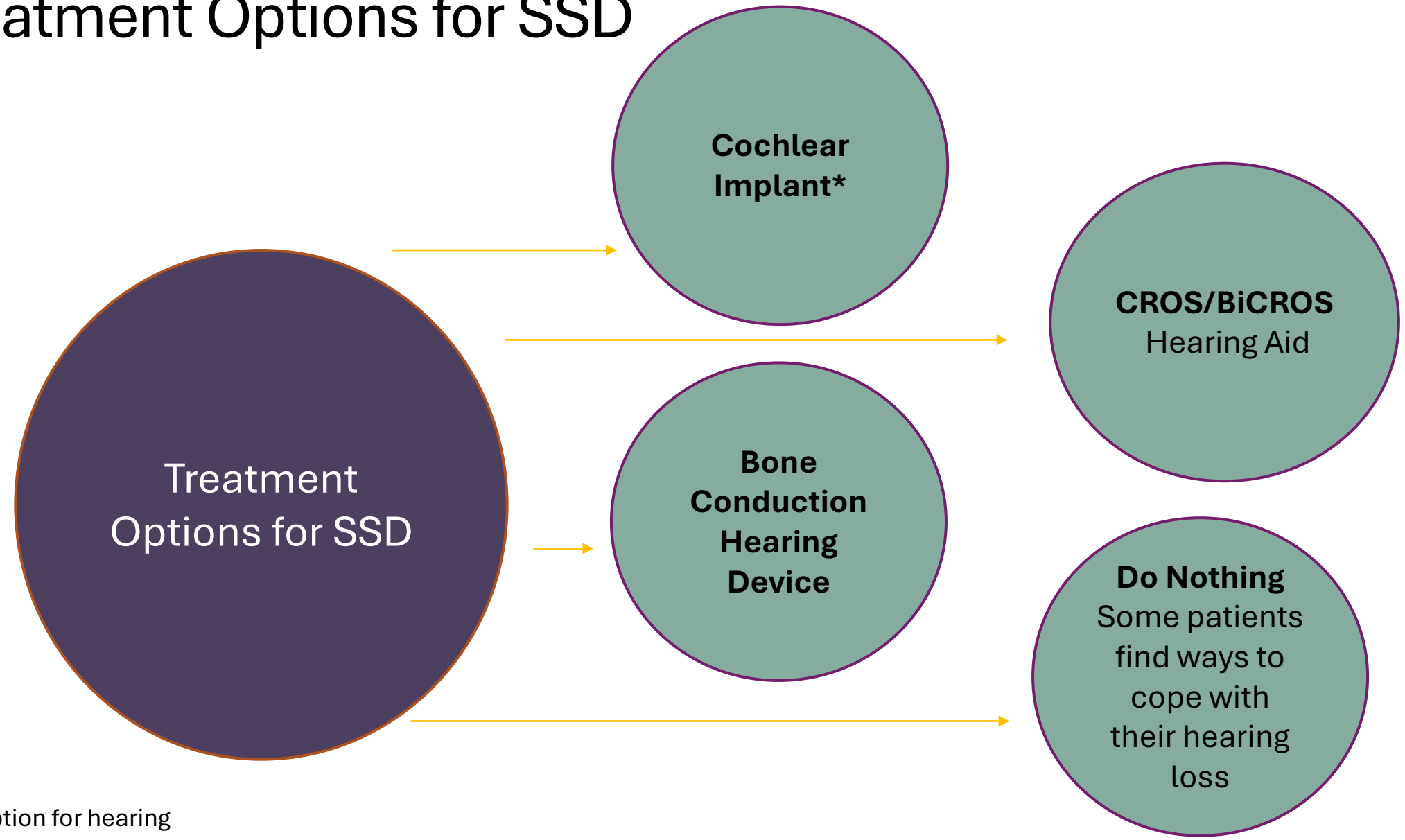
Treatment Options for SSD



*Only option for hearing from that ear

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		TREATMENT OPTION			
		No Treatment/ Observation	CROS/BiCROS	Baha®/Osia® System	Nucleus® Cochlear Implant
POTENTIAL AUDITORY BENEFIT	Overcome Head Shadow	✗	✓	✓	✓
	Sound Lateralization	✗	✓	✓	✓
	Improved Localization	✗	✗	✗	✓
	Binaural Summation	✗	✗	✗	✓
	Squelch	✗	✗	✗	✓

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Cochlear Implant Candidacy

- Cochlear implant candidacy was expanded by the FDA in January 2022 to include patients with SSD aged 5 years or older.
- Ear to be implanted severe to profound SNHL – Pure tone average at 500, 1, 2, 4, >80 dB HL.
- Aided CNC word score (or developmentally appropriate) <5%
- Off label considerations are at centers discretion
  - Implanted SSD children as young as 12 months old
  - Implanted SSD children with less than 80 dB HL PTA with poor word discrimination and little to no benefit from a hearing aid

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Cochlear Implants

- The number one recommendation (if surgery is an option).
  - Reasons may not be an option:
    - Hypoplastic auditory nerve
    - Malformed cochlea
    - Not a surgical candidate
    - Longer than 8-10 years of profound hearing loss
- Provides hearing and most likely open set identification in that ear
- Helps with localization of sound
- Binaural Summation

Cochlear Implant Recommendations

- Wear sound processor all waking hours (10+ hours)
- Direct Streaming 30-60 minutes per day, 3-5 days a week
  - Ideas for young kids
    - Favorite show/song
    - Favorite nursery rhyme
    - Favorite books
  - Ideas for older kids
    - Favorite movie/show with subtitles
    - Audio books, reading along with book
    - Apps/Websites

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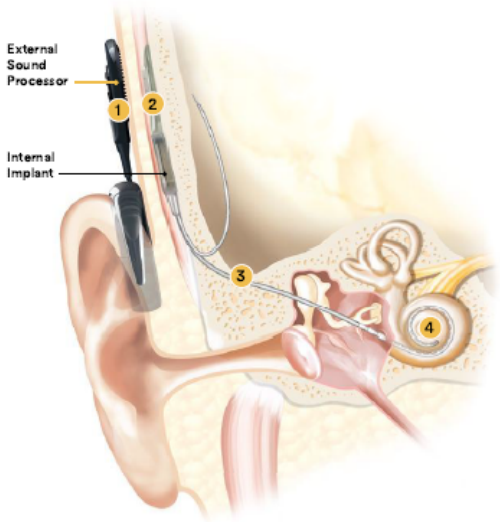
Treatment option for single-sided deafness

Cochlear implant with the Cochlear™ Nucleus® System

A cochlear implant helps restore hearing to your deaf ear

How the Nucleus System works

1. Microphones on the sound processor pick up sounds and the processor converts them into a digital signal.
2. This signal is transferred to the implant just under the skin.
3. The implant sends the digital sound signal to the electrode array in the cochlea.
4. The hearing nerve picks up the signal and sends it to the brain, which is understood as sound.



Potential benefits[9–17]

- Improved speech understanding in noise
- Improved speech understanding in quiet
- Improved quality of life
- Reduced listening effort
- Improved localization

Nucleus Sound Processors

The Nucleus System provides small, simple and smart solutions designed to maximize hearing performance.

Cochlear Kanso® 2 Sound Processor

Cochlear Nucleus 8 Sound Processor

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Bone Conduction Options for SSD

- Profound SNHL
  - Contralateral Ear- Within normal limits 500, 1, 2, 4K <=20 dB HL
  - Osia and Baha surgical options 5 years and older
  - Non-surgical Baha options – Softband, Sound Arc, ADHEAR.

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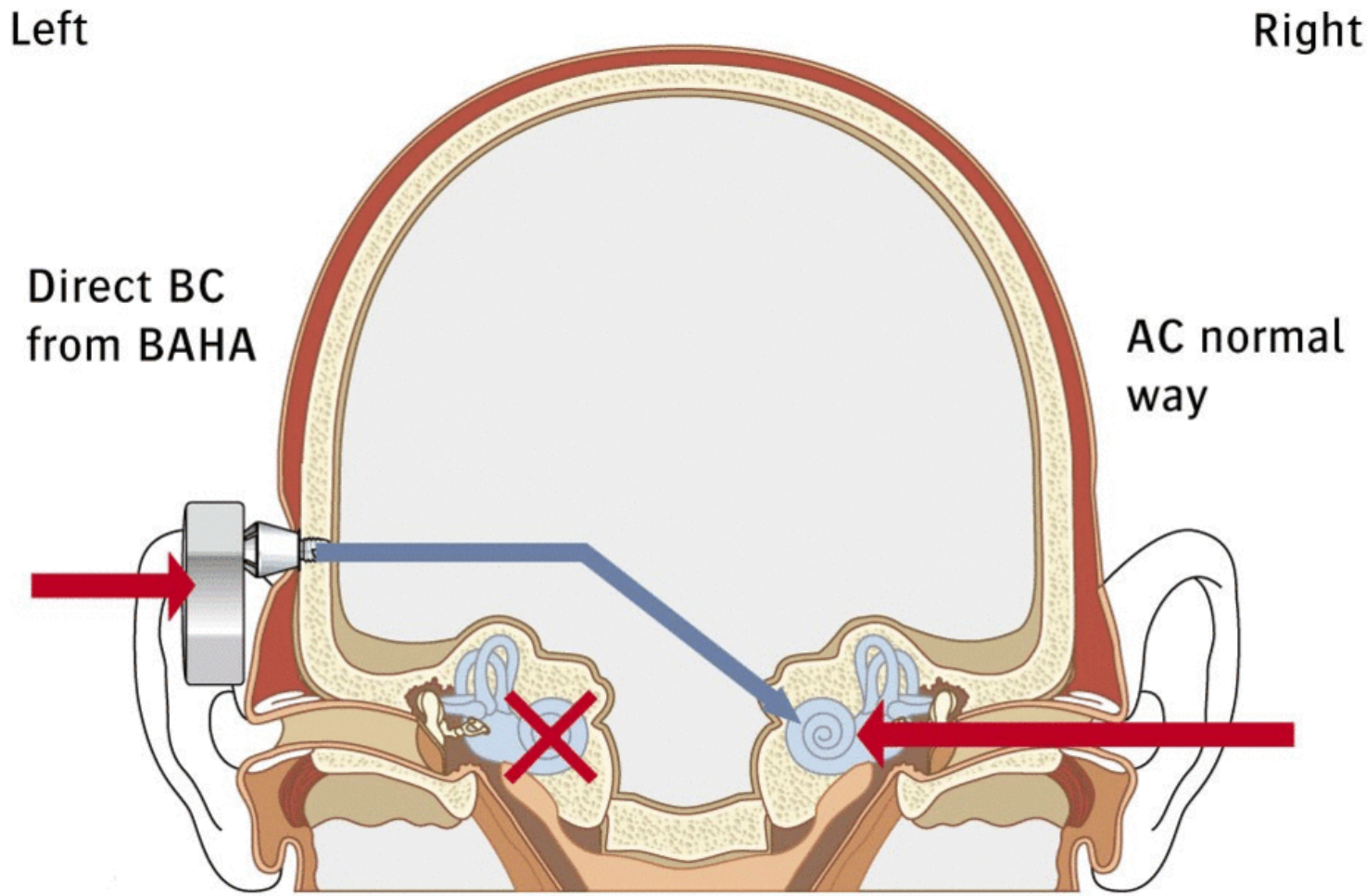

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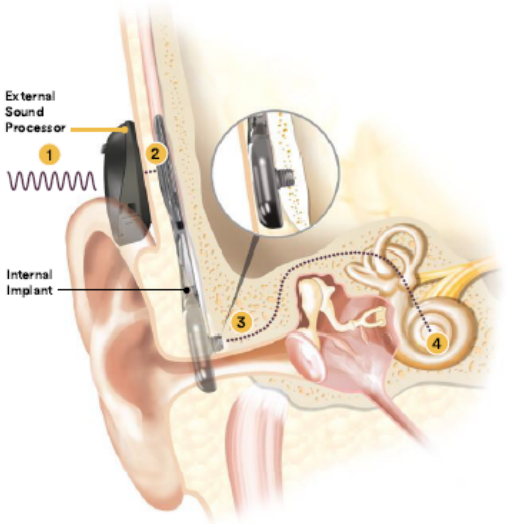
Treatment option for single-sided deafness

Bone conduction implant with the Cochlear™ Osia® System

The Osia® System sends sounds to your normal hearing ear to help enhance hearing

How the Osia System works for single-sided deafness

1. Microphones on the sound processor pick up sounds and the processor converts them into a digital signal.
2. This signal is transferred to the implant just under the skin.
3. The Piezo Power™ transducer vibrates, sending vibrations through the implant to the bone.
4. The vibrations travel around the head to the hearing inner ear where they are converted into electrical impulses and sent to the brain to be interpreted as sound.



Potential benefits

- Improved speech understanding in noise[1,2]
- Improved speech understanding in quiet[1–4]
- Improved quality of life[5]
- Better transmission of high frequency sounds compared with other bone conduction implant systems[6–7]
- Proven transmission of natural sound[8]

Osia Sound Processor

The slim off-the-ear Osia Sound Processor is discreet, light and comfortable to wear.

Cochlear Osia Sound Processor

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Osseointegrated Surgical and Non-Surgical options

- Osia connection around 6 weeks post surgery
- Baha Connect connection around 12 weeks post surgery
- Baha Start- Softband or Sound Arc immediately
- ADHEAR – immediately

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ADHEAR

- Bone conduction option using adhesive
- Same idea as Baha on soft band or sound arc

Baha on Soft Band vs. Surgical Options

- Abutment system can provide more high frequency stimulation to the cochlea. There can be a loss of energy transferring through the soft band compared to abutment.
- Baha uses electromagnetic transducer – best at lower frequencies.
- Osia uses piezoelectric transducer – best at high frequencies
  - Osia can provide more high frequency output which can give better performance in quiet and noise

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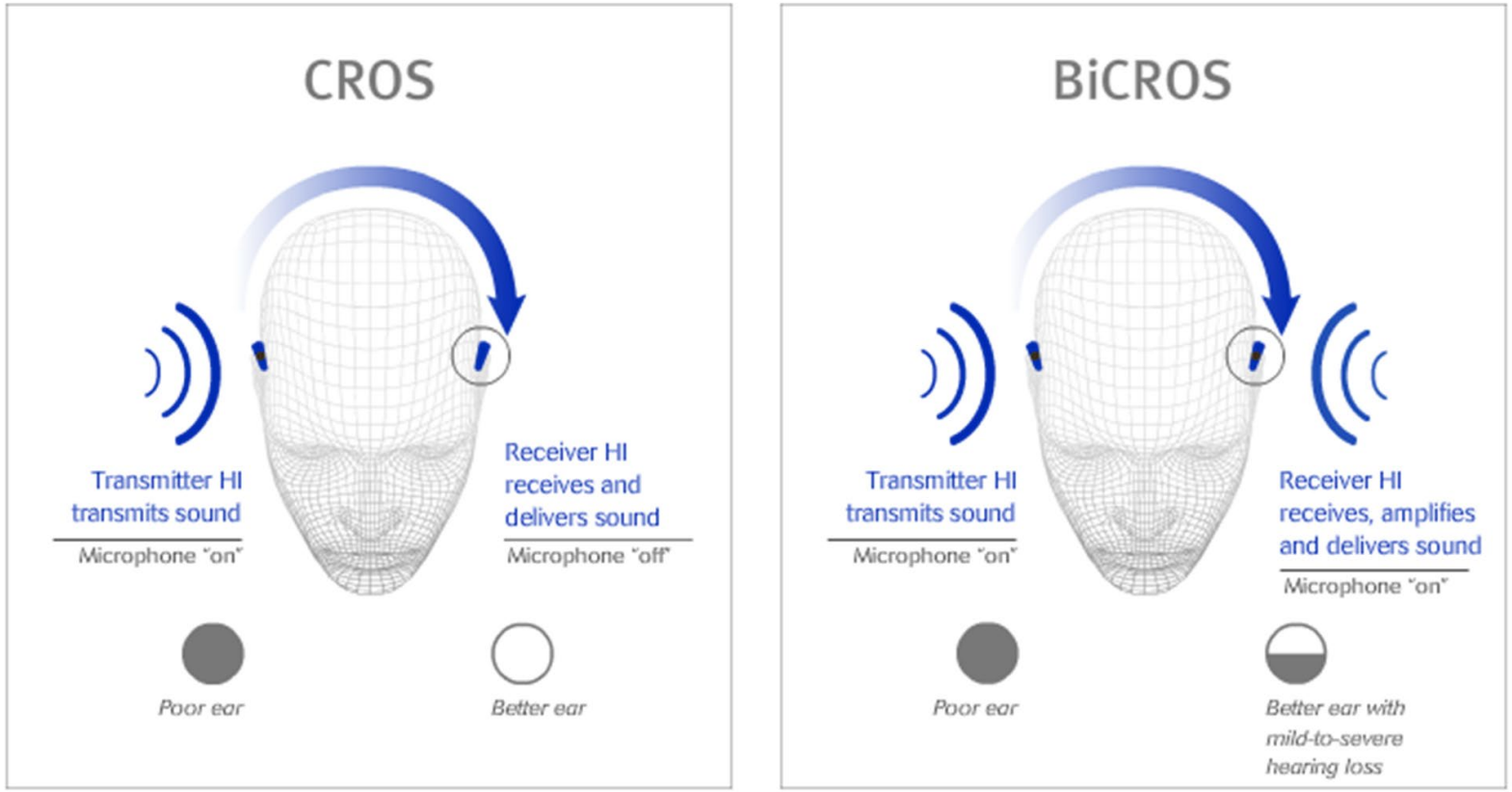

CROS/BiCROS System

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CROS/BiCROS System in Pediatrics

A limitation of a CROS hearing aid is that the earmold can occlude the “good” ear if not sufficiently open. Therefore, CROS systems are typically not recommended for small children.

A good option for older kids and teens

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Educational Recommendations

- Strategic Seating
  - Position student in the classroom so their better ear faces the teacher and other primary speakers and away from noise sources (heater, hallway, etc.)
- Direct Streaming Options
  - Roger Focus II – allows kids with normal hearing in one ear and hearing loss in the other to have the teacher's voice direct to their normal hearing ear.

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Questions?

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