Single-Sided Deafness (SSD) and Cochlear Implantation:

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Introduction and Rationale

Definition: **Single-sided deafness (SSD) has been defined by an international consensus statement as a 'severe-to-profound' hearing loss in one ear (4 frequency pure tone average >70dB HL) and normal or near-normal hearing in the contralateral ear (4 frequency pure tone average ≤35 dB HL) and has a prevalence of about 1% of the general population (Vincent, Arndt & Firszt, 2015; Lucas, Katiri & Kitterick 2018; Davis, 1995).**

Cochlear implantation for single-sided deafness (SSD) is a groundbreaking but emerging field in cochlear implantation. Cochlear implants (CIs) were developed for individuals with profound hearing loss in both ears, but recent clinical advances have extended the benefits of CIs to those affected by SSD. **For patients with SSD, a cochlear implant is the only device that can provide auditory stimulation to the hearing nerve and give access to binaural hearing cues, improving sound localization, speech perception in noisy environments, and overall listening quality.** However, the field remains new and complex, with ongoing research exploring optimal candidate selection, assessment techniques, and post-implantation outcomes.

One influential researcher in this area, Lisa Park, has made significant contributions to understanding SSD CI outcomes. Her studies emphasize that SSD patients present unique auditory challenges and responses, unlike traditional bilateral CI users. For example, her work has highlighted the variability in outcomes, showing that some patients report substantial improvements in spatial hearing and speech perception, while others experience more limited benefits. Park's research also stresses the need for more tailored post-operative auditory rehabilitation, noting that SSD CI users may benefit from specific training focused on integrating the implant with their natural hearing in the unaffected ear.

Additionally, Park's findings underscore the variability in how quickly SSD CI recipients adapt to binaural hearing, pointing to the necessity of individual rehabilitation programs. Her research, along with others in the field, suggests that audiological testing for SSD CI candidates and post-implantation evaluations should be adapted to measure localization and speech-in-noise perception specifically, as these areas are particularly challenging for SSD patients. Although access to one well-functioning ear allows individuals with SSD to appear mostly unimpaired in a quiet listening environment, receiving sound to one side only has a negative impact on the development of binaural listening skills which help with localization and separating relevant from irrelevant signals in noise (Oosthuizen et al., 2021). SSD results in significant cortical reorganization (Ullah et al., 2023), and early intervention during a period of high neuroplasticity promotes optimal outcomes and may prevent or reduce the "aural preference syndrome" (Arras et al., 2024) or " Single ear syndrome**"** that can lead to complex neural and functional challenges that require targeted intervention.

This document aims to address the issues surrounding SSD and recommend assessment and intervention protocols compliant with international standards and will ensure best practice within the South African context.

> The lack of access to two well-functioning ears can lead to a clinically significant degree of disability in everyday life due to the contributing factors listed below (Choissoine-Kerdel et al., 2000; Dwyer, Firszt & Reeder, 2014; Iwasaki et al., 2914; Newman et al., 1997; Lucas, Katiri & Kitterick 2018).

Functional consequences of SSD:

- Severe disruption in the spatial aspects of hearing leads to an inability to detect the source of a sound (Douglas et al., 2007; Arndt et al., 2017; Lucas, Katiri & Kitterick 2018). This is a safety concern particularly for children who might not be consciously aware of this deficit (for example they can't detect the speed or direction of oncoming traffic based on their hearing) (Gifford, 2017).
- Impaired ability to recognize and understand speech in the presence of background noise (Hawley, Litovsky & Cullin, 2004; Welsh, Welso & Rosen, 2004; Lucas, Katiri & Kitterick 2018).
- Reduced speech discrimination and poor localization abilities result in increased cognitive efforts required to process auditory information (Santopietro et al., 2024) which leads to high levels of fatigue, particularly in situations when the individual is unable to move to a more favourable listening position (Lucas, Katiri & Kitterick 2018; Arndt et al., 2017).
- Understanding speech that is directed to the ear with SSD while there is noise on the side of the normal hearing ear is particularly difficult and is typical of classroom situations such as group work (Oosthuizen et al., 2021). Turning their head to ensure that the sound source is on the non-impaired side is tiring and uncomfortable (Lucas, Katiri & Kitterick 2018).
- Unable to hear warning/emergency sounds such as alarms, telephones ringing, or babies crying while sleeping with the good ear on the pillow (Lucas, Katiri & Kitterick 2018).
- Tinnitus can be a consequence of SSD that can range from mild to catastrophic.

Psychological consequences of SSD:

- Increased stress levels related to their need to seek out optimal positions within social situations to hear and participate (Wie, Pripp & Tvete, 2010; Lucas, Katiri & Kitterick 2018).
- Heightened anxiety and concern about losing the hearing in their contralateral ear (Lucas, Katiri & Kitterick 2018).
- Experience feelings of self-stigma (negative perception of oneself due to hearing loss) and low self-efficacy (belief in one's ability to participate) (Lucas, Katiri & Kitterick 2018; Arndt et al., 2017).

Social consequences of SSD:

- Feel excluded from social situations (Wie, Pripp & Tvete, 2010).
- Experience problems with social interactions in their work environment and personal lives (Lucas, Katiri & Kitterick 2018).
- Perceive their social life to be restricted by their hearing loss (Subramanium, Eikelboom & Eager, 2005).
- Develop negative coping strategies such as withdrawing from or within everyday listening situations (Lucas, Katiri & Kitterick 2018).

Developmental and academic consequences of SSD for children:

- Increased academic risk of repeating a grade or falling behind in academic work compared to normal hearing peers (Bovo et al., 1988; Bess et al., 1998; Tharpe, 2008; Gifford, 2017).
- Poorer speech and language scores compared to normal hearing peers and siblings (Lieu et al., 2010; Lieu et al., 2013).
- At risk for delays in cognition development (compared to siblings)

that can affect academic outcomes (Lieu et al., 2013).

- At risk-of-behavioral problems (compared to siblings) that can affect academic outcomes (Lieu et al., 2013).

In essence, individuals with SSD experience high levels of hearing handicap regardless of their age, aetiology, or duration of monaural auditory deprivation (Dwyer, Firszt & Reeder, 2014; Iwasaki et al., 2013; Lucas, Katiri & Kitterick 2018). This proves that even though hearing loss is confined to one ear only, it has a large effect on the individual's health and well-being (Wie, Pripp & Tvete, 2010; Lucas, Katiri & Kitterick 2018). It also affects their brain as Sharma and colleagues (2016) found that abnormal auditory and cross-modal plasticity occurs in response to acquired unilateral deafness.

Conventional high-powered acoustic hearing aids cannot restore access to sound in the impaired ear due to the sensorineural nature and degree of hearing loss in these individuals (Valente et al., 2015; Lucas, Katiri & Kitterick 2018: Arndt et al., 2017). Potential rehabilitation options are the contralateral routing of signals (CROS) aid which is a device that reroutes sound from the side of the impaired ear to the hearing ear for the benefit of speech understanding in noise or a similar effect can be achieved using a bone- conduction hearing device (Kitterick et al., 2014; Armdt et al., 2011; Arndt, Laszig & Aschendorff, 2017; Busk, Linnebjerg & Wetke, 2014; Hol et al., 2010; Arndt et al., 2017). The bone conduction hearing device has the potential for even better benefits to speech perception and sound quality compared to CROS devices (Kitterick, Smith & Lucas, 2016). Furthermore, some studies have suggested that there can be an increased aversion to loud sounds with the use of CROS devices (Lin et al., 2006).

The auditory deprivation associated with SSD causes irreversible changes in the auditory cortex which rerouting devices would not be expected to prevent as they do not provide hearing to the affected side (Park et al., 2022). Only cochlear implantation can allow the additional benefit of restoring access to binaural cues that underpin speech perception in noise and sound localization that is sustained over the long term (>10 years) (Arndt et al., 2011; Arndt, Laszig & Aschendorff, 2017; Finke, Bönitz & Lyxell, 2017b; Hassepass et al., 2016; Jacob et al., 2011; Mertens et al., 2015; Távora-Vieira et al., 2015; Vermeire & Van de Heyning, 2009). (Lewis, et al., 2015) (Lucas, et al., 2018) (Mertens, et al., 2015) (Mertens, et al., 2017) (Newman, et al., 1997)

Benefits of cochlear implantation for SSD:

- Restoring access to binaural cues that underpin speech perception in spatially separated noise (Arndt et al., 2011; Arndt, Laszig & Aschendorff, 2017; Finke, Bönitz & Lyxell, 2017b; Hassepass et al., 2016; Jacob et al., 2011; Mertens et al., 2015; Távora-Vieira et al., 2015; Vermeire & Van de Heyning, 2009; Van de Heyning et al., 2017; Arndt et al., 2017).
- Restoring access to binaural cues that underpin sound localization which reduces difficulty with identifying the location of sound sources (Arndt et al., 2011; Arndt, Laszig & Aschendorff, 2017; Finke, Bönitz & Lyxell, 2017b; Hassepass et al., 2016; Jacob et al., 2011; Mertens et al., 2015; Távora-Vieira et al., 2015; Vermeire & Van de Heyning, 2009; Arndt et al., 2017).
- Improved hearing-specific quality of life (Arndt et al., 2011; Arndt et al., 2017).
- Broader benefits and improvements on health-related quality of life as measured by the Health Utilities Index Mark 3 (Arndt et al., 2011; Arndt et al., 2017).
- Reduced difficulty in navigating everyday environments (Arndt et al., 2011; Arndt, Laszig & Aschendorff, 2017; Fine et al., 2017a; Härkönen et al., 2015; Mertens et al., 2015; Ramos et al., 2015; Távora-Vieira et al., 2015).

- Tinnitus relief (Van de Heyning et al., 2008; Van de Heyning et al., 2017).
- Safe and effective treatment for SSD (Van de Heyning et al., 2017).
- The abnormal auditory and maladaptive cross-modal reorganization of the central auditory system, caused by acquired unilateral deafness can be reversed following cochlear implantation (Sharma et al., 2016; Wedekind, 2018). However, evidence confirms that better outcomes are generally achieved with a shorter duration of deafness (Arndt et al., 2017).
- Evidence of daily use of the device also proves the functional success of cochlear implant treatment for individuals with SSD (Arndt et al., 2017; Polonenko et al., 2017).
- The goal of cochlear implantation in children with SSD is to provide bilateral input to encourage the development of binaural hearing. (Park, et.,2022) Optimal outcomes are achieved when children with congenital SSD are implanted by the age of 3 years (Arndt et al., 2023).

Medical Considerations in SSD

Imaging studies are an essential component in the evaluation of adults and children presenting with SSD. The identification of an anatomical cause is beneficial since it provides the patients or parents with a diagnosis, natural history, and expected prognosis (Lipschitz, et al., 2019; Parks et al, 2022). The performance of Magnetic Resonance Imaging (MRI) with contrast and high-resolution Computed Tomography (CT) or a cone beam CT scan(CBCT) is strongly supported in literature reviews. MRI with contrast is the imaging modality of choice to provide the best accuracy in the diagnosis of any pathology of the brain, cerebellopontine angle, internal acoustic meatus, labyrinth and hypoplasia or aplasia of the eighth nerve. The CT temporal bone imaging demonstrates normal bony landmarks including the diameter of the internal auditory canal (IAC) and cochlear aperture (Parks et al., 2022).

Lipschitz, et al. (2019) identified the aetiology in half of the paediatric cases with SSD using imaging studies. These cases had congenital causes for hearing loss, classified as inner ear anomaly, syndromic or non-syndromic genetic aetiology and congenital cytomegalovirus (CMV) infection. The most common finding in the SSD cohort was cochlear nerve deficiency, followed by cochlear dysplasia & enlarged vestibular aqueduct. Cases of semi-circular dysplasia, temporal bone fracture, skull base legions, and labyrinthitis ossificans were also observed to a lesser amount (Lipschitz et al. 2019). The imaging studies also identified cases with intracranial and brain abnormalities, such as white matter changes (associated with CMV), intracranial lesions, trauma-associated intracranial hematomas, Chiari 1 malformation, and ventricular enlargement.

Numerous studies have shown that the most common causes for SSD in adults were sudden or progressive idiopathic hearing loss and inflammatory aetiologies, e.g. otitis media, labyrinthitis, meningitis, cholesteatoma or mumps (Dillon et al., 2022 & Kurz, et al., 2019). Other common causes are retrocochlear tumours (most commonly vestibular schwannoma), Meniere's disease and trauma (Dillon et al., 2022).

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As for all other CI patients, a good history and clinical examination will determine which further referrals and special examinations (e.g. a battery of blood tests and sonars, etc.) are important.

Individuals with sudden and/or rapid progression of SSD should undergo standard medical workup and monitoring to determine if the hearing spontaneously improves or is recoverable with treatment, such as oral or intratympanic steroids or hyperbaric oxygen therapy. The presence of ossification should be explored in patients with a history of meningitis, otosclerosis, labyrinthitis, temporal bone fracture and prior vestibular schwannoma microsurgery. (Dillon et al., 2022).

Diagnosis of the cause of deafness is valuable in the prediction of estimated outcomes with cochlear implantation. Kurz, et al., (2019) found a significant correlation between inflammatory disease and the duration of deafness of longer than 10 years leading to poorer speech perception outcomes.

Medical contra-indications and special considerations for CI in candidates with SSD:

Significant hypoplasia or aplasia of the eighth nerve, often referred to as cochlear nerve deficiency (CND), is a contraindication to cochlear implantation for children and adults with SSD. The electrical signal presented to the ear with CND would be significantly degraded, the prognosis is poor and the likelihood of non-use is high (Parks et al., 2022; Dillon et al., 2022).

The cochlear anatomy of children with SSD should allow for full or adequate insertion of the electrode array. Partial insertions may result in poor performance and increase the likelihood of non-use (Parks et al., 2022). Advanced cochlear ossification may prohibit adequate insertion of the electrode array and is therefore a possible contra-indication for cochlear implantation (Dillon et al., 2022).

Children and adults with SSD at risk of progressive hearing loss in the better hearing ear merit special consideration. It is advantageous to implant the SSD ear rather than wait for the contralateral ear to decline. This approach reduces the period of auditory deprivation that will improve performance and enable binaural hearing. Two common causes of progressive hearing loss in the initially better hearing ear of children and adults with SSD are congenital cytomegalic-virus infection (cCMV) and cochlear malformation such as enlarged vestibular aqueduct. CMV is one of the most common causes of acquired SNHL in children but often undiagnosed in asymptomatic children. Individuals with enlarged vestibular aqueduct are twice as likely to have bilateral cochlear malformation and therefore at risk for progressive hearing loss in both ears (Parks et al, 2022 & Dillon et al., 2022). Aetiologies such as Meniere's disease, auto-immune inner ear disease, and neurofibromatosis type 2 (NF2) also warrant special consideration (Dillon et al., 2022).

SSD due to bacterial meningitis requires timely intervention. If profound SNHL occurs, rapid ossification may obliterate the cochlea and preclude successful implantation (Parks et al., 2022 & Dillon et al, 2022). Other conditions with a risk of rapid progressive ossification are otic capsule fractures and vestibular schwannoma microsurgery (Dillon et al., 2022).

Individuals with sudden and/or rapid progression of SSD should undergo standard medical workup and monitoring to determine if the hearing spontaneously improves or is recoverable with treatment, such as oral or intratympanic steroids or hyperbaric oxygen therapy. In most cases it is recommended that cochlear implantation should not occur 3 to 6 months after sudden hearing loss to allow time for the recovery of hearing (Dillon et al., 2022).

Candidacy and Exclusion from Cochlear Implantation

Inclusion Criteria:

Ear to be implanted:

Severe-to-profound sensorineural hearing loss with a PTA of ≥70dBHL

Aided word recognition (one 50- monosyllabic word list) of ≤40% as measured with standardised monosyllabic words.

Sudden loss – wait 3-6 months before considering implant unless related to conditions associated with high risk of progressive ossification

Contralateral ear:

Normal to mild hearing (PTA ≥ 35dBHL) levels

Aided word recognition of 80% or more as measured with standardised Monosyllabic words

Special consideration for people with the risk of a progressive loss in the better ear

Lack of or limited perceived benefit from conventional treatment options for SSD, including hearing aid, bone-conduction device or CROS technology.

Completed Audiological trial period of different treatment options as indicated in this document.

A recent systematic review suggests improvement in tinnitus outcome compared to other devices like CROS and BC devices for candidates that meet audiological and medical criteria.

Counselling about possible tinnitus suppression should be included and patients should be aware of the possibility in a very small percentage of patients, of their tinnitus symptoms worsening after implantation.REF

Realistic expectations

In children, the following factors are important for candidacy (Gordon, et al., 2018):

Adequate speech and language development

Normal overall development

Typical social & learning abilities

Good family support and structure

Availability and commitment to rehabilitation

Availability of appropriate educational support services

Exclusion Criteria:

- Adults and children with congenital SSD loss older than 5 years, are not CI candidates.
- Any medical condition that is considered a contra-indication to undergoing cochlear implantation.
- Radiological contra-indications like a hypoplastic nerve.
- Children with global developmental delay and/or multiple handicaps must be carefully selected on a case-by-case basis.
- Evidence of active middle-ear pathology based on otologic examination and/or immittance testing.
- Medical or psychological conditions that contra-indicate undergoing surgery.
- Ossification or any other cochlear anomaly that might prevent complete insertion of the electrode array.
- Hearing loss of neural or central origin, including auditory neuropathy.
- Unrealistic expectations on the part of the subject regarding the

possible benefits, risks, and limitations inherent to the surgical procedure and prosthetic device.

- Poor compliance during the assessment of alternative technology e.g. poor datalogging on trial devices and questionnaires not returned.

Cochlear Implant Testing Protocol for Adults and Children with Single-Sided Deafness (SSD)

This protocol is designed to evaluate candidacy for cochlear implantation in adults and children with single-sided deafness (SSD), incorporating the American Cochlear Implant Alliance Task Force guidelines, 2024 Clinical Recommendations for the Treatment of Unilateral Hearing Loss/Single-Sided Deafness with Cochlear Implantation and current evidence-based practices and the Minimum Speech Test Battery (MSTB-3) Dan et al. 2024. The evaluation includes audiological testing, speech-in-noise measures, and questionnaires assessing quality of life, tinnitus, and spatial hearing.

Adult Testing Protocol for SSD (children older than 5 years with acquired SSD included)

Adults and children with congenital SSD loss older than 5 years, are not CI candidates.
The onset and duration of the hearing loss significantly impact the outcomes of cochlear implantation.

Medical and audiological evaluation

Questionnaires:
The following questionnaires must be completed before the patient's audiological evaluation. The audiologist can use the information from the questionnaires to discuss treatment options and guidance in treatment decision-making.

- **Speech, Spatial, and Qualities of Hearing Scale 12 (SSQ 12)**:

The **SSQ** is used to assess the subjective experience of hearing in real-world situations. It evaluates speech perception, spatial awareness, and the overall quality of hearing.

- **Cochlear Implant Quality of Life (CIQOL) Survey**:

This assesses the impact of a cochlear implant on daily life, social interactions, and mental well-being.

- **Tinnitus Handicap Inventory (THI)**:

Used to assess the impact of tinnitus on daily activities, emotions, and hearing.

- **Revised Hearing Handicap Inventory for Adults (RHHIA)**:

Assesses the social and emotional impact of hearing loss on daily life.

- **Vanderbilt Fatigue Scale (VFS-A)10 item version:**

Designed for individuals aged 18 and older, the VFS-A evaluates fatigue across various domains, including physical and cognitive aspects related to listening. *10-Item Version:* Suitable for both clinical and research applications.

Treatment options:

- No intervention
- CROS or Bone Conduction device
- Assistive devices e.g. FM system
- Cochlear implantation

Counselling to guide the patient about treatment options. This is one of the most important aspects of the evaluation process and time should be taken to explain the implications of all the treatment options. Patients should understand the physiological mechanisms of restoring hearing with a cochlear implant and that other treatment options like CROS and BC devices provide "pseudo-hearing".

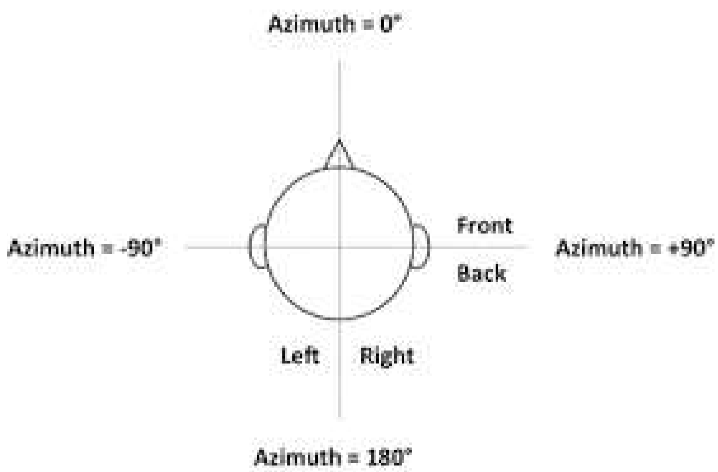
Indications:

Affected ear: Severe to Profound sensorineural hearing loss (≥70 dB HL).
Contralateral ear: Normal hearing (≤35 dB HL) or mild hearing loss.

Audiological evaluation: Unaided testing:

- Standard diagnostic baseline measurements using objective and subjective measures to confirm that the patient falls within the criteria of SSD.
- **Unaided speech-in-noise** measurements to evaluate the patient's speech-in-noise abilities using words and sentences.

- **Test material for speech in noise testing:**
  Sentences: CID sentences in noise, CID sinne in geraas, other material like SASIN/SASIG/HINT/QuickSin can also be used. This is a baseline test for comparison pre- and post-implantation.
- The normal ear should be plugged and/or masked. To ensure that the normal hearing ear is not responding, masking should be 40-45 dB HL minimum.



See appendix for testing conditions (MSTB-3 PRE-OPERATIVE PROTOCOL).

If a 1-speaker set-up is available: present speech and noise using a 0 SNR (signal 65dBA: noise 65dBA) at 0 degrees.
If a 2-speaker set-up is available: present speech and noise using a 0 SNR (signal 65dBA: noise 65dBA), with signal presented from the front and noise presented to the better ear.
The presentation level should be 45dB HL (65 dB SPL *refer to calibration figures for audiometer to determine dB HL relative to dB SPL).

Audiological evaluation: Aided testing

Treatment options trial

It is recommended that adults and older children try **Contralateral Routing of Signal (CROS)** hearing aids or **bone conduction devices (BC)** for at least one week, ideally two weeks. No aided testing with the Cros and BC devices is recommended as this does not contribute to decision-making for intervention.

The BBSS - Bern Benefit in Single-Sided Deafness Questionnaire, SSQ 12 and Vanderbilt Fatigue Scale (VFS-A)10 should be administered and the patient should fill out the questionnaire for both treatment options e.g. CROS and BC device on a softband. If these devices do not provide significant improvement, CI may be considered.

Table 1: Potential auditory benefits from different treatment options (taken from Cochlear White Paper Nov. 2024):

	No treatment	**CROS**	**BC Device**	**Cochlear Implant**
Overcome Head-Shadow	X	✓	✓	✓
Sound Lateralisation	X	✓	✓	✓
Improved localisation	X	X	X	✓
Binaural summation	X	X	X	✓
Squelch	X	X	X	✓

Children Testing Protocol for SSD (Children younger than 5 years with congenital SSD)

Audiological Evaluation for Children with SSD:

Congenital SSD:

Children with congenital SSD are evaluated differently because their brains have developed without binaural input from birth. Key elements of the evaluation include:

Congenital SSD is typically diagnosed early through:

- **Newborn hearing screening** programs.
- **Objective Tests**: **Otoacoustic emissions (OAE)** and **auditory brainstem response (ABR)** testing are essential for confirming profound hearing loss in the affected ear. These tests are typically done shortly after birth or following a failed newborn hearing screen.
- **Visual Response Audiometry and Conditioned Play Audiometry** can be used in older children to assist/confirm objective test results.
- **Developmental Monitoring**: Regular tracking of speech and language milestones by a speech-language therapist is critical. Children with congenital SSD may show **delays in speech and language development**, particularly in complex listening environments like classrooms.
- **Speech Perception Tests**: Speech perception tests do not have to be performed on young children, and the SI index will indicate the deficit.

Congenital SSD candidates benefit from earlier cochlear implantation because the brain's plasticity allows for more efficient adaptation to auditory input if the cochlear implant is provided early in life. The goal is to prevent or mitigate developmental delays in speech and language acquisition.

Summary of Findings from the Literature:

Before Age 3–4: The majority of studies, including Arndt et al., Zeitler et al., and Eisenberg et al., recommend implantation **before age 3 or 4** for the most effective outcomes. These children show significant improvements in speech perception, sound localization, and binaural auditory processing.

After Age 5: Delayed implantation, particularly after age 5, is associated with poorer outcomes. Studies by Kuhn et al. and Buss et al. emphasize that the brain's auditory plasticity declines after this age, leading to more limited gains in speech and spatial hearing abilities.

Ideal Window: Most experts agree that implantation should ideally occur **before age 3** to take advantage of the critical period for auditory development.

These findings support the consensus that **early cochlear implantation** for SSD—ideally within the first few years of life—maximizes auditory outcomes by capitalizing on the brain's neuroplasticity during the critical period for auditory learning.

Audiological Evaluation

Pure-Tone Audiometry:

- **Affected ear**: Severe to Profound sensorineural hearing loss (≥70 dB HL).
- **Contralateral ear**: Normal hearing or mild hearing loss (≤35 dB HL).
- Objective test measures ie. Acoustic reflexes, OAE, ABR if the child/adolescent is unreliable during testing.

Imaging

CT and MRI: Imaging is required to evaluate the cochlear anatomy, ensuring there are no malformations that could hinder implantation success.

Questionnaires to Assess Benefit

Speech, Spatial, and Qualities of Hearing Scale for Children (SSQ-C):

This child-friendly version of the SSQ evaluates how children perceive speech, spatial sound, and the quality of sound in daily environments.

Auditory Behaviour in Everyday Life (ABEL):

The ABEL questionnaire evaluates how hearing impacts a child's everyday life, including in social settings and at school.

Parents' Evaluation of Aural/Oral Performance of Children (PEACH):

PEACH is a parent-report questionnaire that assesses how the child functions in everyday environments, including speech understanding and sound localization.

Children's Home Inventory for Listening Difficulties (CHILD):

CHILD evaluates listening difficulties from the child's perspective in various environments, providing insights into challenges in speech perception and hearing.

Vanderbilt Fatigue Scale VFS for Pediatrics:

Intended for children aged 6 to 17, this suite includes:

- *VFS-Child (VFS-C):* A self-report version for children to complete.
- *VFS-Parent (VFS-P):* A proxy-report version for parents or guardians.
- *VFS-Teacher (VFS-T):* A proxy-report version for teachers.

Medical Evaluation:

For both congenital and acquired SSD:

Imaging (CT and MRI): This is crucial to assess the condition of the cochlear and auditory pathways and rule out any anomalies that might hinder implantation (e.g., cochlear nerve deficiency).

Medical History: In cases of acquired SSD, the medical team will investigate possible causes, such as viral infections (e.g., mumps) or trauma, which may have broader implications for auditory rehabilitation post-implant.

Treatment Options Trial:

It is not recommended that young children with congenital SSD trial a bone conduction device and a CROS system (Park et al. 2022). Parents should be carefully counselled about binaural auditory brain development.

Speech and Language Development:

Congenital SSD: Early intervention is critical to prevent delays in speech and language development, which are common in congenital SSD if not addressed. Evaluations focus on the use of **auditory verbal therapy** to ensure the child can integrate the auditory input provided by the implant.

Acquired SSD: These children typically show less language developmental delay, the focus should be on the **re-integration of auditory stimuli** and sound localization after implantation.

Psychological and Family Considerations:

The decision to implant in cases of congenital SSD requires an understanding of the family's commitment to long-term **auditory rehabilitation**, especially since the child has no prior binaural hearing experience.

Committed to full-time use of their cochlear implant speech processor, 10 Hours or more a day.

For acquired SSD, psychological evaluations may include assessing the child's **adjustment to hearing loss** and their motivation levels to undergo surgery and auditory rehabilitation to regain function in their affected ear.

Key Differences Between Congenital and Acquired SSD:

Timing and Brain Plasticity: Congenital SSD patients benefit more from early implantation due to the brain's heightened plasticity during the critical window for speech and language development. In contrast, acquired SSD patients may be more focused on restoring previously held binaural hearing abilities.

Speech and Language Concerns: Congenital SSD presents a higher risk of **speech and language delays**, while acquired SSD tends to cause difficulties in sound localization and speech perception, especially in noise (eg. Classroom) with a lower risk of language delays if intervention is timely.

Considerations for Auditory Re/Habilitation (AR) in SSD

Important considerations in AR with SSD:

The elimination of sound entering the contralateral ear (with normal hearing or some degree of residual hearing) during AR exercises: This can be achieved through occlusion with silicone earplugs/moulds, noise-cancelling headphones, or a combination of both. The amount of time and the number of exercises where the better-hearing ear is occluded should be determined by the therapist according to the auditory skill level and needs of the CI recipient.

The use of assistive listening devices (ALDs): A device where direct streaming options to the implanted ear only ("*direct audio input*") allows the therapist and patient to practice auditory exercises over a distance, where the device sends the signal directly to the CI and thus without assistance from the contralateral, better- hearing ear.

Device use: Research by Park et al. indicates that patients with 10+ hour use a day has better outcomes with their cochlear implants.

Time required to reach optimal outcomes: Patients with SSD should receive up to 12 months of audiological rehabilitation before optimal outcomes can be measured/evaluated. This aspect is important to cover in counselling of adult patients with SSD and the parents of children with SSD.

Benefits of AR for adults with CI: Audiological rehabilitation can be holistically defined as the reduction of hearing-loss-induced deficits in functionality, activity, participation, and quality of life through sensory management, instruction, perceptual training and counselling (Boothroyd, 2010). A growing body of research is currently displaying the benefits or improved outcomes of audiological rehabilitation for adults post-implantation (Erber, 1988; Hogan, 2001; Plant, 2006). This includes benefits such as improvements in speech perception in quiet, speech perception in noise, speech perception over distances, sound localization, awareness of environmental sounds, music perception, and overall quality of life (Hogan, 2001; Plant, 2006; Pedley & Hogan, 2005). Further evidence suggests that even moderate training on targeted phonemes can improve speech perception by as much as 15 – 20% (Fu, 2008). In general, AR training programs have been developed to optimize hearing and quality of life outcomes for adult CI users with SSD (Távora-Vieira, et al., 2015).

Benefits of AR for children with CI: For children, the benefits of (re)habilitation have been thoroughly documented and researched. Children who are identified with hearing loss by 3 months and enrolled in family-centred intervention programs by 6 months, can develop similar speech and language skills as their typical hearing peers (Fulcher et al., 2012). They also develop better reading skills, educational outcomes (Yoshinaga-Itano, 2003) and better social-emotional growth (Langereis & Vermeulen, 2015).

AR for children and adults with SSD and CI: Research with regards to the benefits of audiological rehabilitation for adult CI recipients with single-sided deafness is currently limited and mostly focused on adults with SSD who are fitted with hearing aids, rather than CI's. Existing evidence does, however, point to improvements in speech discrimination in noise, improvement in sound localization, and tinnitus reduction (Zhang et al., 2012; Nawaz et al., 2014). For children with SSD, post-implant (re)habiltation demonstrates similar benefits: improved speech understanding in noise and quiet and improved sound localization (Hassepass et al.,2013).

AR components for adult SSD CI recipients: The components of aural rehabilitation for adult CI recipients with single-sided deafness are the same as those addressed with bilateral adult CI recipients. The components are as follows:

Informational counselling: Providing information regarding hearing loss, cochlear implants, and the rehabilitation process to optimize the recipient's use of the cochlear implant, assistive devices, online programs, resources, and tools.

Psychosocial counselling: Providing support with psychological or social issues in everyday life as a result of hearing loss to foster motivation, a positive attitude and realistic expectations of a cochlear implant device.

Analytic auditory training: For improvement of the discrimination of specific speech features, phonemes, words, and short phrases without contextual background information.

Synthetic auditory training: To improve understanding of longer phrases, sentences, paragraphs, and conversations using contextual, syntactic, and semantic cues.

Communication strategies training: To improve the CI recipient's ability to follow typical daily conversations, especially in more challenging listening environments.

Frequent communication partner training: To enable training or programs to be continued at home or on a regular basis. Also to foster empathy and understanding from the communication partner regarding the CI AR process.

Telephone training: If deemed necessary but not crucial for patients with SSD. It might be useful, especially during emergency situations.

Music therapy: To relearn appreciation or enjoyment of music; to improve the perception of acoustic elements of rhythm, pitch, and tone colour (timbre); to further improve the perception of speech in noise; to improve the overall quality of life.

It is important to note that the content and amount of time spent on each component is patient-specific and should be tailored to each recipient's individual needs, in both bilateral and single-sided CI recipients.

AR components for child SSD CI recipients:

Estabrooks et al. (2016) provide an outline for rehabilitationists, particularly auditory-verbal (AV) practitioners, on how to provide therapy sessions to children with SSD:

Beginning the session with both ears: A conversation or activity during which the child wears the CI device and the typical hearing ear is unoccluded.

Listening with the hearing device only: A variety of activities focusing on auditory skill development are presented with the hearing device on and with the typical hearing ear occluded with an earmould or earplug.

Listening with both ears again: A variety of tasks are presented to facilitate the refinement of binaural interaction skills (e.g. localization skills, understanding of speech- in-noise, etc.), speech sound discrimination skills, temporal processing skills, perception of music and/or dichotic listening skills.

Parent guidance: The practitioner and parents discuss the session outcomes and exchange ideas of ways to incorporate the short-term objectives into the child's daily life.

Child guidance: The child needs to become an "active listener", so he/she is encouraged to repair communication breakdowns and to use self-advocacy strategies to control the listening environment for successful communication.

Conclusion

Cochlear implantation for single-sided deafness (SSD) is becoming more common, particularly in children, evaluation and intervention must be tailored to the nature of the hearing loss—whether congenital or acquired. Early identification and implantation in congenital SSD maximize developmental outcomes, while acquired SSD requires careful assessment of prior hearing ability and the individual's adjustment to hearing loss. Both groups require thorough audiological, medical, and psychosocial evaluations to ensure optimal outcomes.

This guideline provides a standardized protocol for the audiological management of SSD, enabling clinicians to guide their patients with SSD to choose the best treatment option for them taking in account their individual variables. It offers a consistent and comprehensive approach that can be implemented within the resources available to most clinics.

In recognition of the growing body of evidence supporting cochlear implantation for SSD, the U.S. Food and Drug Administration (FDA, 2019) has expanded CI indications to include patients aged five years and older with SSD and asymmetric hearing loss. This decision is based on extensive research demonstrating the significant benefits of cochlear implants over conventional treatments such as contralateral routing of signals (CROS) and bone-conduction devices.

By following the recommendations outlined in this guideline, healthcare professionals can ensure that SSD patients receive evidence-based care, ultimately improving their auditory function, quality of life, and long-term developmental outcomes.

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