Single-Sided Deafness (SSD) and Cochlear Implantation: SACIG Position Statement & Clinical Guidelines

AUTHORS:

Dr Elnèmarie Burden (ENT-specialist)

Mrs Nicolize Cass (Audiologist)

Miss Estienne Havenga (Speech-, Language Therapist)

Dr Maurice Hockman (ENT-specialist)

Ms Mirisa Meyer (Audiologist)

Dr Kurt Schlemmer (ENT-specialist)

Mrs Tamara van Zyl (Audiologist)

Ms Jenny Perold (Audiologist)

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1) Introduction & Rationale

Single sided deafness (SSD) refers to a highly asymmetric unilateral hearing loss and is a unique type of hearing loss as one ear has completely normal hearing (Lucas, Katiri & Kitterick 2018; Gifford 2017).

SSD has been defined by an international consensus statement as a 'severe-to-profound' hearing loss in one ear (pure tone average >60dB HL) and normal or near-normal hearing in the contralateral ear (pure tone average <30 dB HL) and has a prevalence of about 1% of the general population (Vincent, Arndt & Firszt, 2015; Lucas, Katiri & Kitterick 2018; Davis, 1995). This does not however mean that all individuals with SSD are candidates for cochlear implantation as they still need to fulfill all cochlear implant assessment criteria in accordance with South African Cochlear Implant Group guidelines.

Although access to one well-functioning ear allows individuals with SSD to appear mostly unimpaired in a quiet listening environment, their 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 leading 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).
- Increase in listening effort lead to high levels of fatigue, particularly in situations when they are unable to move to a more favourable listening position (Lucas, Katiri & Kitterick 2018; Arndt et al., 2017).

- 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).

Psychological consequences of SSD:

- Increased stress levels related to their need to seek out optimal positions within social situations in order 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 ones' 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 behavioural 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 the 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 has an effect on 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 the 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).

However, 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

a;, 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).

2) Audiological Considerations in SSD

Although hearing aids for a contralateral routing of signals (CROS-HA) and bone conduction devices have been the traditional treatment options for single-sided deafness (SSD) and asymmetric hearing loss (AHL), in recent years, cochlear implants (CIs) have gradually become a viable treatment option, particularly in countries where regulatory approval and reimbursement schemes are in place (Vlastarakos, et al., 2014). This shift occurred since the CI is the only device capable of restoring bilateral input to the auditory system - therefore of possibly restoring binaural hearing and it may also result in the suppression of tinnitus (Van Zon, et al., 2015). Even though a number of studies have independently shown that the CI is a safe and effective treatment option for SSD and AHL, clinical outcome measures in those studies and across CI centres differ significantly (Van Zon, et al., 2015) (Van den Heyning, et al., 2017). Only with the consistent implementation of defined and agreed-upon outcome measures across implant centres, can reliable evidence be generated to measure the safety and efficacy of CIs and alternative treatment options in recipients with SSD and AHL (Van den Heyning, et al., 2017).

Protocol:

The following clinical treatment options for single-sided deafness (SSD) or asymmetric hearing loss (AHL), have already received regulatory approval:
(1) a contralateral routing of signal hearing aid (CROS-HA);
(2) a bone conduction device (BCD),
(3) The latest research proposes adding cochlear implantation as a third possible treatment option. Cochlear implantation as a treatment option was approved by CE mark in 2013 and 2019 by FDA.

However, despite the broader availability of at least some of these options in most countries, SSD remain untreated in the vast majority of patients.

The efficacy of each of the treatment options, including no treatment, is generally measured in terms of:

1. Speech understanding in quiet
2. Speech understanding in noise
3. Sound localization
4. Quality of life (QoL)
5. Tinnitus reduction (when applicable)

The following protocol was developed, discussed and agreed upon by expert panels that convened at the 2015 APSCI conference in Beijing, China, and at the CI 2016 conference in Toronto, Canada (Van den Heyning, et al., 2017). The protocol is based on a set of minimum outcome measures and aims at coordinating assessment methods across centres and thus at generating a growing body of high-level evidence for these treatment options (Van den Heyning, et al., 2017).

The protocol compares longitudinal outcome measures, comparing the CROS-HA, BCD and CI treatments. The recommended outcome measures include:

1) **Speech in noise testing**, using the same set of 3 spatial configurations to compare binaural benefits such as summation, squelch, and head shadow across devices;
2) **Questionnaires** to collect **quality of life measures** and the frequency of device use;
3) **Questionnaires** for assessing the **impact of tinnitus before and after treatment**, if applicable.

It is proposed that these outcome measures be performed after a trial period of 2 weeks was conducted with a (Bi) CROS HA and BCD headband. This protocol would allow results to be compared and the advantages and disadvantages of various treatment options to be more clearly seen (Meyer & Van Zyl, 2017).

Outcome measures should be collected at the following intervals:

1) At baseline
2) After each of the initial 2 weeks (Bi) CROS HA and BCD headband trials

Should the patient opt to proceed with the cochlear implant, continue with the outcome measures after device activation at:

3) 3 months
4) 6 months
5) 12 months
6) Annually thereafter according to the protocol outlined in Appendix B.

Should patients opt for non-treatment, it is advised that the patient should be seen for follow-up outcome measures as indicated for post-cochlear implantation.

Outcome measures explained:

- Pure Tone Audiogram:

At the baseline assessment, the audiological assessment should include the measurement of air conduction hearing thresholds (PTA averaged over frequencies of 0.5, 1, 2, and 4 kHz) in both ears. Conduct the assessments using a standard audiological setup in a sound-treated room. Present stimuli via headphones / insert earphones with the better ear masked.

Therefore, in order to demonstrate the benefit of either intervention, it is necessary to track the aided and unaided (CROS, BCD, or CI deactivated) performance measures at all intervals.

- Masked speech perception:

Masked speech perception should be assessed with a standard audiometric and validated recorded sentence testing, using a free-field setup in a sound-treated room. Present the masker at a fixed level of 50dBHL (Meyer & Van Zyl, 2017). Adapt the level of the target signal to measure the speech reception threshold at which 50% of the sentences are understood correctly (Van den Heyning, et al., 2017). Multi-speaker babble should be used as

the masking signal, since it is a (1) a more effective speech masker than other noise signals and (2) more sensitive in showing benefits of a CI or a second good ear over a first good ear (Bernstein, et al., 2016)

Different testing configurations can possibly demonstrate the binaural benefit in terms of the head shadow, summation, binaural squelch, and spatial release from masking (Gartrell, et al., 2014). To calculate the binaural effect with each of the 3 treatment options for SSD is demonstrated in the table below (Test time adds up to 30–45 min for all spatial configurations and listening conditions, assuming 5–7 min per test list.).

The spatial configuration S_{ssD} N_{AH} measures the effect of the head shadow on speech recognition by presenting the speech signal to the side of the poorer ear and the masker to the side of the better ear and by comparing speech reception thresholds for the aided condition with those of the unaided listening condition (Van Zon, et al., 2015). The negative head shadow effect is overcome by adding a CROS-HA, BCD headband, or CI device on the SSD side (thus, the side with the better signal-to-noise ratio). According to Firszte et al., 2012, this spatial configuration is the most sensitive to evaluate pre- and post- treatment improvements.

Using the S_0N_{SSD} configuration, the signal is presented from the front and the masker from the side of the poorer ear. In the aided condition, the CROS-HA, BCD, or CI device receives the more adverse signal-to-noise- ratio (SNR). While the binaural squelch still produces a benefit of approximately 3 dB in normal-hearing listeners (Bronkhorst & Plomp, 1988), CROS-HA and BCD devices consistently show diminished speech recognition in this configuration (Peters, et al., 2015) since they basically route the masker to the better ear and consequently reduce the effective SNR ratio on that side. On the other hand, CI users with SSD (Peters, et al., 2015) or AHL, have consistently showed that they perform equally or better with the CI activated in this environment, suggesting that a CI is able to reinstate binaural hearing (Van Zon, et al., 2015)

With the spatial configuration S_0N_0, the effect of binaural summation on speech recognition is evaluated by presenting both the signal and the masker from the front and by comparing speech reception thresholds for the aided condition with those of the unaided listening condition (Van Zon, et al., 2015).

Binaural effect measure:

Spatial Configuration	Listening conditions	Binaural Effect Measure
S_{SSD} N_{AH}	Aided, Unaided	**Head shadow (dB)** = SRT S_{SSD} N_{AH} unaided – SRT S_{SSD} N_{AH} aided
S_0N_{SSD}	Aided, Unaided	**Squelch (dB)** = SRT S_0N_{SSD} unaided – SRT S_0N_{SSD} aided
S_0N_0	Aided, Unaided	**Summation (dB)** = SRT S_0N_0unaided - SRT S_0N_0 aided **SRM[1] (dB)** = SRT S_0N_0 unaided – SRT S_0NSSD aided

The measures commonly used to quantify binaural effects and the way those measures are derived from the proposed testing configurations are shown in the column on the right. For all measures, a positive effect size in decibels indicates a binaural benefit (Van den Heyning, et al., 2017).

- Quality of life (QoL) and tinnitus assessments:

Impact of hearing loss:

The Speech, Spatial, and Qualities of hearing (SSQ) questionnaire (Noble, et al., 2013) is a sensitive and specific measure to assess the impact of hearing loss on speech perception, sound localization, and QoL (Horsman, et al., 2003). The majority of studies exploring the benefits of the different treatment options for SSD or AHL have used the SSQ as the primary assessment tool for assessing subjective outcomes in the subdomains addressed by the questionnaire (Van den Heyning, et al., 2017).

[1] SRM: Spatial Release from Masking

Generic QoL Questionnaire:

The Health Utilities Index Mark 3 (HUI III) (Horsman, et al., 2003) is a generic measure of general health status, and should be used to determine a single index value of the health status. The HUI III is the standard assessment tool for health technology assessments evaluating the cost-effectiveness of a treatment.

Tinnitus Questionnaire:

For those patients with tinnitus, falling within the CI indication criteria, the Tinnitus Handicap Inventory (THI) questionnaire (Meikle, et al., 2012) should be used to identify, quantify, and evaluate the handicap from tinnitus at the pre-operative interval and at all follow-up intervals.

Additional Questionnaires:

The Bern Benefit in Single-Sided Deafness (BBSS) questionnaire should be administered after the trail of both the CROS- HA and the BCD headband. The questionnaire consists of 10 visual analogue scales rating the subjectively perceived benefit of the Baha or any other CROS device in different situations (Meyer & Van Zyl, 2017).

A summary of the treatment process for adults is available in Appendix A & B and for children in Appendix C (Meyer & Van Zyl, 2017).

3) Medical Considerations in SSD

Imaging studies are an essential component in the evaluation of adults and especially children presenting with SSD. In children, the identification of an anatomical cause is beneficial since it provides the parents or caregivers with a diagnosis, natural history and expected prognosis (Lipschitz, et al., 2019). Performance of Magnetic Resonance Imaging (MRI) with contrast and high resolution Computed Tomography (CT) (or a CBCT: cone beam CT) scan is strongly supported in literature reviews. MRI with contrast is the imaging modality of choice to provide

the best accuracy in diagnosis of any pathology of the brain, cerebello-pontine angle, internal acoustic meatus and labyrinth.

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 aquaduct. Cases of semi-circular dysplasia, temporal bone fracture, skull base legions and labyrinthitis ossification 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.

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.

Numerous studies have shown that the most common causes for SSD in adults were sudden onset hearing losses and inflammatory aetiologies, e.g. otitis media, labyrinthitis, meningitis, cholesteatoma or mumps (Kurz, et al., 2019).

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 for inflammatory disease and duration of deafness of longer than 10 years leading to poorer speech perception outcomes.

Considerations for Auditory Re/Habilitation (AR) in 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 counseling (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 has 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 has been thoroughly documented and researched. Children who are identified with hearing loss by 3 months and enrolled in family-centered 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:

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

2. **Psychosocial counseling**: 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.

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

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

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

6. **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.

7. **Telephone training**: To enable some degree of speech discrimination or the following of basic conversations over the telephone where the signal is often distorted, especially during emergency situations.

8. **Music therapy**: To relearn appreciation or enjoyment of music; to improve perception of acoustic elements of rhythm, pitch and tone colour (timbre); to further improve perception of speech in noise; to improve 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) provides an outline for rehabilitationists, particularly auditory-verbal (AV) practitioners, on how to provide therapy sessions to children with SSD:

1. **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.

2. **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.

3. **Listening with both ears again:** A variety of tasks are presented to facilitate 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 skils.

4. **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.

5. **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.

Important considerations in AR with SSD:

a. **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.
b. **The use of assistive listening devices (ALDs):** A device where direct streaming options to the implanted ear only ("*direct audio input")* gives the therapist and patient the opportunity 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.
c. **Device use:** Research indicates that children and adults with SSD may require the use of their devices only in school, work or social settings, unlike those with bilateral hearing loss **(Lucas et al., 2018).** Device use with SSD depends on the patient's communicative or listening needs within his/her daily life.
d. **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 counseling of adult patients with SSD and the parents of children with SSD.

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4) Candidacy and Exclusion from Cochlear Implantation

Inclusion Criteria:

- Ear to be implanted:
    - Moderate-to-profound sensorineural hearing loss with a PTA of ≥ 70dBHL
    - Aided word recognition of ≤ 60% as measured with standardised monosyllabic words.

- Contralateral ear:
    - Normal to mild hearing (PTA ≥ 35dBHL and ≤55dBHL) levels
    - Aided word recognition of 80% or more as measured with standardised Monosyllabic words
- 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.
- Realistic expectations
- In children, the following factors are important for candidacy (Gordon, et al., 2018):
    - Age-appropriate speech and language development
    - Normal overall development
    - Social & learning abilities
    - Good family support and structure
    - Availability of rehabilitation services
    - Availability of educational support services

Exclusion Criteria:

- Any medical condition considered a contra-indication to undergoing cochlear implantation.
- Alleviation of tinnitus as the stated primary or sole motivation for seeking implantation by the subject and/or investigator.
- 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.

5) Conclusion

This protocol for the audiological management of patients with SSD, will allow results to be compared and the advantages and disadvantages of various treatments to be more clearly seen. The protocol proposed herein is consistent, comprehensive, and may be completed by clinics within the range of their normally available resources.

The FDA (FDA, 26/07/2019) has endorsed expanding cochlear implantation indications to include patients 5 years and above with single sided deafness (SSD) and asymmetric hearing loss (AHL) who have profound sensorineural hearing loss in one ear and normal hearing or mild sensorineural hearing loss in the other ear (Racey, 2019). The expansion of the cochlear implant indications have been based on vast amounts of literature that support the benefits of this treatment option over other treatment options for SSD (Park, et al., 2019).

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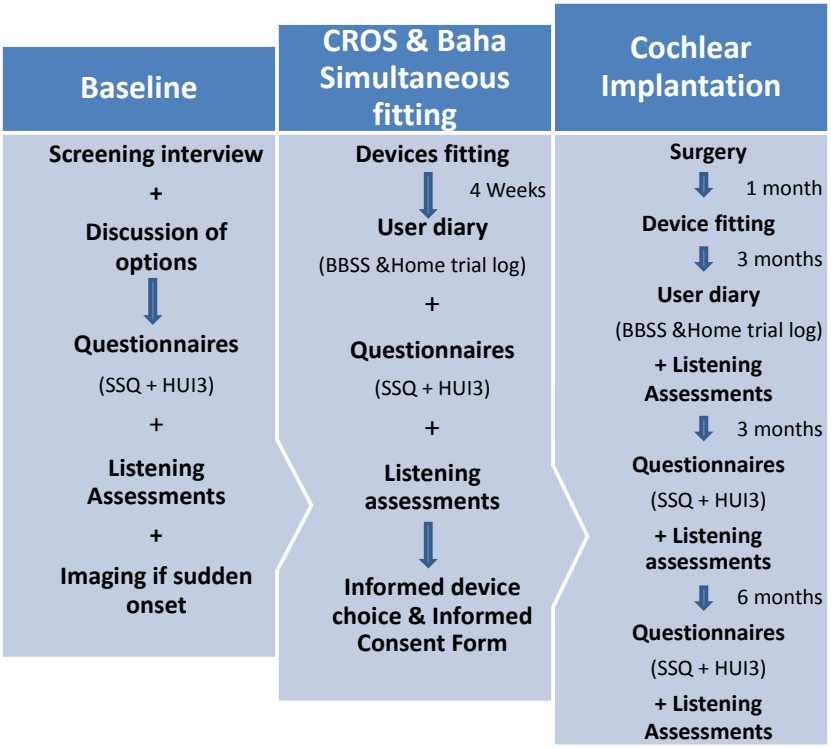
Appendix A

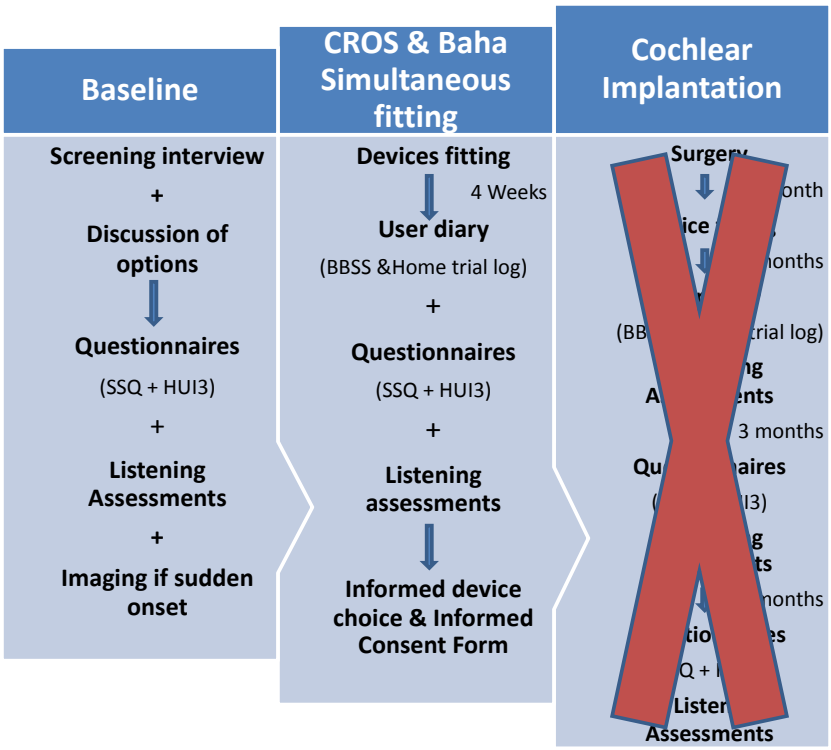
UHL/SSD Adult Protocol <15 year duration of deafness



Definitions:
UHL is often termed single-sided deafness (SSD) however as the term 'deafness' implies a profound or total hearing loss, then SSD is not really the ideal term for cases where there is some functional hearing.
Some aetiologies may result in markedly asymmetric hearing loss (AHL) where both ears have a significant hearing loss however only one ear falls within standard CI criteria.
Definitions for hearing in the better ear:
SSD: better ear ≤30 dB HL to 4 kHz inclusively,
AHL: better ear ≤60 dB HL to 4 kHz inclusively, and >30 dB HL at one or more frequencies to 4 kHz inclusively and with an inter-aural asymmetry of >30 dB (four-frequency average).

Appendix B

UHL/SSD Adult Protocol >15 year duration of deafness*



Definitions:
UHL is often termed single-sided deafness (SSD) however as the term 'deafness' implies a profound or total hearing loss, then SSD is not really the ideal term for cases where there is some functional hearing.
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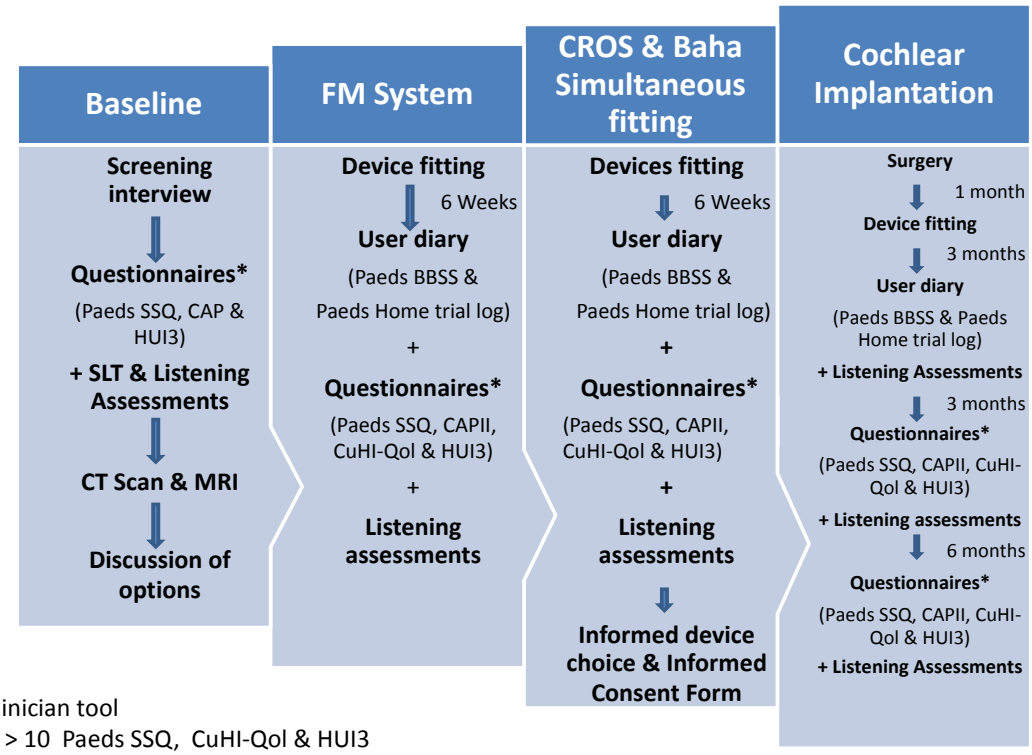
AHL: better ear ≤60 dB HL to 4 kHz inclusively, and >30 dB HL at one or more frequencies to 4 kHz inclusively and with an inter-aural asymmetry of >30 dB (four-frequency average).

Appendix C

UHL/SSD Paediatric Protocol



CAPII= clinician tool
Children > 10 Paeds SSQ, CuHI-Qol & HUI3
Children < 10 Paeds SSQ Parents version & CuHI-Qol

Appendix D – Home Trial log instructions: Afrikaans & English

Instruksies vir gebruik van die verskillende versterkings opsies vir die behandeling van my unilaterale gehoorverlies:

U het vandag 1 of meer verskillende opsies ontvang, om uit te probeer, om u te help met die unilaterale gehoorverlies.

Lys die verskillende opsies wat aan u geleen word om te proef:

1.
2.
3.

Identifiseer, met die hulp van ‘n oudioloog, 3 omgewings waar u probleme ondervind as gevolg van ‘n unilaterale gehoorverlies:

1.
2.
3.

In stilte, watter opsie is vir u beter?

In geraas, watter opsie is vir u beter?

Watter opsie stel u in staat om meer bewus te wees van u ruimtelike omgewing (indien enige):

In ‘n groepsgesprek, watter opsie is vir u beter?

U keuse, indien enige:

Opmerkings:

Instructions for the use of different amplification options for the treatment of your unilateral hearing loss:

You received more than one treatment options today, to try out to see whether you benefit from any of them:

List the different options that was given to you, to try out:

1.
2.
3.

Identify, with the assistance of your audiologist, 3 environments that you struggle to cope because of your unilateral hearing loss:

1.
2.
3.

In quiet, which option is better?

In noise, which option is better?

Which option makes you feel more spatially aware of where sound is coming from (if any):

In a group conversation, which option is better?

Your choice of device, if any:

Comments:

Appendix E – Ticksheet for Treatment options for SSD – English & Afrikaans

TICKSHEET FOR TREATMENT OPTIONS FOR SSD

The different options to treat a unilateral hearing loss at present are the following:

1. FM system ☐
2. Bone conduction device ☐
3. CROS hearing aids ☐
4. Cochlear Implant ☐
5. No treatment ☐

Treatment choice: _____

Date: _____ Signature: _____

LYS VAN BEHANDELINGSOPSIES VIR UNILATERALE GEHOORVERLIES

Die volgende behandelings opsies is tans beskikbaar vir ‘n unilaterale gehoorverlies:

1. FM sisteem ☐
2. Beengeleidingsapparaat ☐
3. CROS gehoorapparate ☐
4. Kogleêre inplanting ☐
5. Geen behandeling ☐

Behandelingskeuse: _____

Datum: _____ Handtekening: _____