SOUTH AFRICAN COCHLEAR IMPLANT GROUP

SUID - AFRIKAANSE KOGLEERE INPLANTINGSGROEP

APPENDIX B:

Guidelines for Referral and Candidacy for Cochlear Implantation, including guidelines for unilateral and bilateral cochlear implantation

The following guidelines have been developed to represent the most common practice through consideration of evidence-based criteria for best practice and clinical experience. It is acknowledged that guidelines/criteria for implantation are under active research internationally and, it is therefore expected that these guidelines will develop further over time and will need to be updated no less than every 18-24 months.

Cochlear implantation should be considered only after a rigorous, valid and individualised evaluation process is completed and consensus is reached by the multidisciplinary cochlear implant team (who are recommended to all be members of SACIG). This core team should include an ENT surgeon, audiologist, radiologist and speech-language therapist. For more complex cases other specialist inputs may be required (e.g., a psychologist / social worker / medical specialist, Allied Health specialist(s)).

Please refer to the main Quality Standards document regarding the recommended qualifications of the core cochlear implant team (ENT surgeon, audiologist, speech-language therapist).

It should be noted that delays in implantation have detrimental and life-long effects on auditory brain development, negatively impacting spoken language development with consequent negative effects on learning, educational and ultimately vocational outcomes.

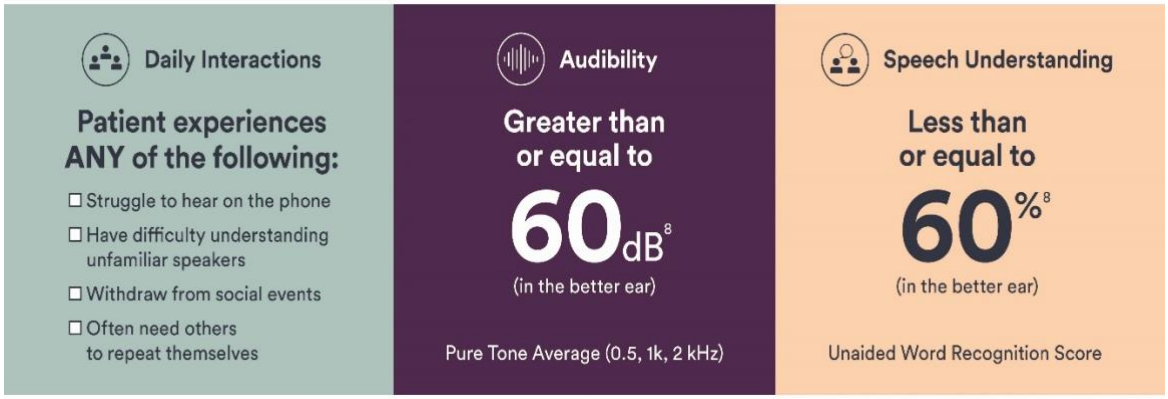
Children who receive early, bilateral cochlear implantation have substantial speech perception improvements, speech intelligibility, receptive-expressive language ability, reading, social, reading, and cognitive skills (Sanju et al. 2022), often exceeding that of their normal-hearing peers.

Evidence supports bilateral implantation under the age of 1 year in cases of bilateral severe-profound congenital hearing loss as this leads to the most positive outcomes (McKinney, 2017).

Referral criteria for adults and older children:

The revised 60/60 guidelines are recommended for referral to a cochlear implant programme for in-depth evaluation by a cochlear implant team for possible candidacy. If used as a screening measure, the 60/60 measure results in a 96% detection rate and a 34% false-positive rate for identifying adults who would meet traditional indications for a cochlear implant.

Zwolan, T A., Schartz-Leyvartz., K C., Pleasant, T. (2020) Development of a 60/60 Guideline for Referring Adults for a Traditional Cochlear Implant Candidacy Evaluation. Otology Neurotology. 41(7): 895-900.



Referral criteria for younger children:

A 50/70+ guideline for referral of children is recommended: that is, clinicians should refer paediatric patients for evaluation if they meet any of the following criteria: appropriately selected word recognition scores <50% and unaided pure tone thresholds >70dBHL (Davidson 2006; Fitzpatrick et al. 2009; Leigh et al. 2011, 2016; Bittencourt et al. 2012; Leal et al. 2016); or poor functional performance, limited progress in language or auditory development, or poor quality of life (Mondain et al. 2002; Lazaridis et al. 2010; Vickers et al. 2016). (Warner-Czyz et al, 2022)

AUDIOLOGICAL CRITERIA FOR CANDIDACY FOR COCHLEAR IMPLANTATION

Cochlear implants are recommended for:

CHILDREN (6 months and older)

Candidacy determination involves specification of audiological and medical criteria for cochlear implantation in children. Recommendations for evaluation for cochlear implantation should maintain flexibility and consider a child's skill progression (month-by-month progress in speech, language and auditory development) and quality of life with appropriately fitted hearing aids. Paediatric cochlear implant candidacy is a rapidly changing and evolving process

due to new research and technological advancements. These evidence-based guidelines for current clinical protocols encourage a team-based approach focused on the whole child and the family system (Warner-Czyz et al, 2022).

1. Bilateral moderate to severe sensorineural hearing loss in the low frequencies and severe to profound hearing loss in the high frequencies.

2. In the case of asymmetrical hearing loss (mild to moderate sensorineural hearing loss in the better ear and moderate to profound sensorineural hearing loss in the poorer ear), special consideration should be given to speech, language and educational progress with best fitted hearing aids in determination of candidacy for the poorer ear.

3. Single-sided deafness (SSD): Congenital onset - implantation after careful consideration and evaluation. It is not always possible to determine whether the hearing loss is congenital or acquired. If the hearing loss is congenital, implantation is ideal before the age of five years. Merit for implantation should be assessed on a case-by-case basis. **(Appendix C).**

4. Who obtain minimal or no benefit from appropriately fitted binaural hearing aids and show limited progress in spoken language development while under management of an appropriately trained speech-language therapist (**Appendix A**)

5. ±6 months to ±36 months of age (this is the optimal period of auditory plasticity therefore early implantation results in better outcomes). Later implantation can be considered in special circumstances if the child has developed some spoken language. Later implantation is contra-indicated if there is no evidence of any development of spoken language unless in special cases (e.g. diagnosis of progressive loss of vision).

6. In cases of sudden onset or progressive hearing loss (as opposed to congenital onset of bilateral severe-profound sensorineural hearing loss), speech perception scores should reflect minimal hearing aid benefit which would negatively impact on further speech / language / academic progress. Cochlear implantation should not be delayed due to the critical period of neuroplasticity to avoid the negative effects of poor or insufficient auditory access to sound.

7. The degree of hearing loss in children with confirmed Auditory Neuropathy Spectrum Disorder (ANSD) does not correlate with their speech perception ability and progress in speech and language development. Children should be evaluated as potential CI candidates based on their speech perception scores, parent questionnaires and therapist reports even if audiometric thresholds do not meet typical candidacy guidelines (Rance & Barker, 2008). ANSD often presents with disproportionately poor

speech recognition abilities – particularly in noise – relative to degree of hearing loss (Rance & Barker, 2008; Berlin et al. 2010). Radiological evidence should support cochlear implantation (i.e. present auditory nerve).

8. Children with multiple handicaps: cochlear implantation should be considered within the context of improvement of quality of life and be within the financial reach of the family for life-long management considering the cumulative medical and rehabilitation expenses required for these children. Outcomes may not necessarily include the development of spoken language but can provide improved life quality and communication.

9. In cases of deafness resulting from meningitis or an inflammatory condition of the cochlea where there is radiological evidence of ossification cochlear implantation should be considered an emergency and (bilateral) implantation should be recommended. This may need to be prior to the age of 6 months.

ADULTS (18 years and older)

1. Bilateral, postlingual moderate to severe sensorineural hearing loss;

2. Asymmetrical hearing loss where the better ear has mild to moderate sensorineural hearing loss and severe to profound sensorineural hearing loss in the poorer ear;

3. Achieve limited benefit from appropriately fitted binaural hearing aids after a minimum trial period of ±3 months. Hearing aid limited benefit is confirmed by aided word recognition scores (of poorer than) ≤ 40% in the ear to be implanted (see **Appendix F**)

4. The degree of hearing loss in adults with confirmed Auditory Neuropathy Spectrum Disorder (ANSD) does not correlate with their speech perception ability and they should be considered for cochlear implantation.

5. Single-Sided deafness – Adults with acquired, progressive, or sudden single-sided deafness can be considered for cochlear implantation. Duration of deafness should be considered when selecting candidates. **Refer to Appendix C.**

PRELINGUALLY OR PERILINGUALLY DEAFENED OLDER CHILDREN AND ADULTS

These patients with bilateral severe to profound sensorineural hearing loss are considered for cochlear implantation if they use spoken language as their primary mode of communication, have a degree of speech intelligibility and have developed oral communication and speech production (Lahlou et al, 2022) and obtain inadequate benefit from hearing aids.

CANDIDATES FOR ELECTROACOUSTIC STIMULATION (EAS)

Hearing thresholds in the low frequencies (250Hz and 500Hz) could be at normal to mild levels in low frequencies and in the middle to high frequencies (>1000Hz) at severe to profound levels. Word recognition scores are more representative of the impact of the hearing loss on speech perception, communication ability and quality of life with appropriately fitted hearing aids.

Hearing preservation surgery is an important consideration in these cases (surgical method and choice of electrode).

Consideration should be given to the device selected which offers access to electroacoustic stimulation.

MEDICAL CRITERIA

1. The auditory nerve should be present.
2. The cochlea should be sufficiently patent for electrode insertion
3. The surgical procedure can be performed with minimal risk to the person
4. No active middle ear inflammatory condition

REHABILITATIVE CRITERIA

Prospective implant recipients and their families should:

1. be well motivated
2. have demonstrated commitment to the rehabilitative process, and
3. have appropriate expectations of the potential benefits of an implant.

FINANCIAL RESOURCES

Socioeconomic factors should ensure a realistic probability that the family will be able to support and maintain the device as well as rehabilitation over the period of their lives.

BILATERAL COCHLEAR IMPLANTATION:

A. BILATERAL COCHLEAR IMPLANTATION IN CHILDREN

Guidelines for simultaneous and sequential implantation are listed below.

Care of the child who is a cochlear implant (CI) candidate should include a strategy for the treatment of hearing loss in both ears.

There is no restriction on age of implantation for children with progressive hearing loss (assuming they have had auditory input via hearing aids for children with a longer duration of progressive hearing loss).

A.1 SIMULTANEOUS IMPLANTATION

Unimplanted children are considered for a simultaneous implantation when they meet the following criteria:

- Bilateral severe to profound sensorineural hearing loss
- ±6 to ±36 months of age (this is the optimal period of auditory plasticity therefore early bilateral implantation results in better outcomes). Later implantation can be considered in special circumstances if the child has developed some spoken language. There is no restriction on age of implantation for children with progressive hearing loss assuming they have had auditory input via hearing aids and have developed some spoken language.
- Labyrinthine (cochlear and vestibular), Internal Auditory Canal, mastoid, middle ear, and external ear canal anatomy that does not preclude appropriate electrode insertion.
- Recent history of meningitis with otologic involvement. Unilateral cochlear implantation should be avoided to minimise the risk of future exclusion of potential implantation of the other ear due to ossification of the cochlear structures.
- No active inflammatory middle ear or mastoid disease.
- No medical conditions that significantly increase surgical risk.

- No co-existing significant neurological condition that may negatively influence CI benefit.

A.2 SEQUENTIAL IMPLANTATION

A.2.1. Criteria for children already unilaterally implanted

Implantation of the second ear is appropriate when the following criteria are met:

- In addition to the second ear meeting CI candidacy criteria, there should be minimal measurable binaural advantage on age-appropriate speech perception measures in the bimodal condition (CI + contralateral HA) compared to the CI alone. Tests should be selected to avoid ceiling and floor effects.
- Evidence of good compliance during therapeutic intervention with the first implantation.
- If there is a history of vestibular problems a comprehensive vestibular assessment should be completed prior to implantation of the second ear.

A delay in implantation of the second ear should be avoided when both ears meet criteria to avoid the development of Aural Preference Syndrome.

Bilateral hearing in early development protects auditory cortices from reorganizing to prefer the better ear (Polonenko, 2017). Unilateral hearing in childhood restricts input along the bilateral auditory pathways, causing permanent reorganization (Gordon, 2013). Sequential implantation should ideally take place within 1.5 years.

A.2.2 Criteria for un-implanted children

Un-implanted children may be considered for a sequential approach when they do not meet criteria for simultaneous implantation because any of the following conditions exist:

- Question about the usefulness of residual hearing in either ear (implant worst hearing ear first).
- History of vestibular disturbance that raises concern about the effect of CI on vestibular function and/or symptoms. Prior to implantation a vestibular assessment should be completed to assist with selection of the ear.
- Presence of abnormal labyrinthine or altered mastoid/middle ear/ear canal anatomy in either ear requiring special surgical techniques for implantation (implant best anatomically developed ear first if equal hearing in each ear).
- Concern about the influence of co-existing medical/developmental conditions (e.g. neurological conditions and sensory processing disorders) on CI benefit or that increase the risk of surgery.
- History of poor compliance during therapeutic intervention with the first implantation.

B. BILATERAL COCHLEAR IMPLANT CANDIDACY IN ADULTS

Care of the patient who is a cochlear implant (CI) candidate should include a strategy for the treatment of hearing loss in both ears. Guidelines for simultaneous and sequential implantation are listed below.

B.1 SIMULTANEOUS IMPLANTATION

It is generally recommended that sequential implantation is preferable due to the risk of vestibular involvement post-implantation. An exception would be where there are radiological signs of potential ossification which could preclude future implantation in that ear. A vestibular assessment should be considered for adults and older children (refer to Appendix R).

Un-implanted adult patients may be considered for simultaneous implantation when they meet the following criteria:

- Bilateral severe-profound sensorineural hearing loss
- Postlingual or progressive onset of hearing loss in both ears.
- Duration of profound hearing loss <30 years in both ears with a history of good bilateral aid use.
- History of recent meningitis with otologic involvement (should be considered as emergency for implantation)
- Normal labyrinthine (cochlear and vestibular) anatomy.
- No active inflammatory middle ear or mastoid disease or history of canal wall mastoid surgery in either ear.
- No history of significant vestibular disorders, however the patient needs to be counselled regarding potential complication of vestibular sequelae. If an existing history of vestibular/disequilibrium disorders exists, the patient should undergo a comprehensive vestibular assessment.
- No medical conditions that significantly increase surgical risk or co-existing conditions that may influence CI benefit (such as neurologic disorders).

B.2 SEQUENTIAL IMPLANTATION

B.2.1. CRITERIA FOR UNIMPLANTED PATIENTS

Un-implanted adult patients may be best considered for a sequential approach when they do not meet criteria for simultaneous implantation because any of the following conditions exist:

- Prelingual or perilingual onset of hearing loss or long term (>30 years) profound deafness in either ear (implant ear with best hearing history first.)
- History of vestibular disturbance that raises concern about the effect of CI on vestibular function and/or symptoms. A vestibular assessment should be conducted prior to implantation, the ear with the weaker vestibular system should be implanted first.
- Presence of abnormal labyrinthine or altered mastoid/middle ear anatomy in either ear requiring special surgical techniques for implantation.
- Concern about the effect of coexisting medical conditions on CI benefit or that increase the risk of surgery.
- No history of significant vestibular disorders, however the patient needs to be counselled regarding potential complication of vestibular sequelae.
- Concern re insufficient evidence of previous hearing aid use.

B.2.2. CRITERIA FOR PATIENTS ALREADY UNILATERALLY IMPLANTED

Implantation of the second ear is appropriate when the following criteria are met:

- In addition to the second ear meeting CI candidacy criteria, there should be minimal measurable binaural advantage demonstrated in the bimodal condition (CI + contralateral HA) compared to the CI alone condition. Tests should be selected to avoid ceiling and floor effects.
- Although good function of the first CI is preferred, implantation of the second ear can be considered in the event of less than expected first CI performance if there is hope of “capturing” a better performing ear.
- No history of significant vestibular disorders, however the patient needs to be counselled regarding potential complication of vestibular sequelae. A vestibular assessment should be conducted prior to implantation of the second ear (Appendix R).

REFERENCES

The following references are recommended to use as a guide for current SACIG selection criteria:

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