

Review Article

Current criteria for selecting cochlear implant in deaf patients: a review

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Received: 26 October 2021

Accepted: 24 November 2021

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ABSTRACT

Cochlear implantation is indicated in patients with severe to profound hearing loss that cannot be adequately treated by other auditory rehabilitation measures. The definitive indication of cochlear implantation is made on the basis of an extensive interdisciplinary clinical, audiological, radiological, and psychological diagnostic work-up. There are numerous changes are happening in cochlear implant candidacy. These have been associated with concomitant changes in surgical techniques, which enhanced the utility and safety of cochlear implantation. Currently, cochlear implants are approved for individuals with severe to profound unilateral hearing loss rather than previously needed for bilateral profound hearing loss. Studies have begun using the short electrode arrays for shallow insertion in patients with low-frequency residual hearing loss. The advancement in designs of the cochlear implant along with improvements in surgical techniques reduce the complications and result in the safety and efficacy of the cochlear implant which further encourages the use of these devices. This review article aims to discuss the new concepts in the candidacy of the cochlear implant, cochlear implant in younger children and hearing preservation, a cochlear implant for unilateral deafness, bilateral cochlear implant, and cochlear implant with neural plasticity and selection of patients for the cochlear implant.

Keywords: Cochlear implant, Deaf patients, Profound hearing loss, Indications for cochlear implantation

INTRODUCTION

In children, adequate hearing is vital for age-appropriate social and lingual development.¹ Hearing has a key role in verbal communication and significantly contributes to social well-being.² Hearing impairment hamper cognitive performance and promotes social isolation, as well as the development of depressive and dementia-related disorders.^{3,4} The cochlear implantation is done for hearing rehabilitation of children and adults with severe to profound sensorineural hearing loss and/or poor speech discrimination with limited benefit from conventional hearing aids.⁵ The candidacy requirements for children and adult cochlear implantation have gradually loosened. The obvious aim is to never a single patient

performs more poorly with their cochlear implant than previously used hearing aids alone.⁶ Now the indications for cochlear implantation have been expanded for achieving open-set word recognition. Bilateral cochlear implantations are highly helpful for providing access to sound information from both ears, allowing improved speech perception in noise and quiet places as well as sound localization.⁷ There is not much literature concerning criteria for the cochlear implant. Changes in candidacy have primarily included implanting individuals with increasing amounts of residual hearing, implanting candidates with increasing amounts of pre-implant open-set speech skills, implanting at younger ages, and implanting in individuals with the abnormal cochlea. This review article will discuss history, evaluation of

candidacy, audiological evaluation, cochlear implant in children, cochlear implant in unilateral deafness, bilateral cochlear implant, cochlear implant in auditory neuropathy, and the cochlear implant in genetic deafness.

Methods for literature search

Multiple systematic methods were used to find current research publications on current criteria for selecting cochlear implant patients. We started by searching the Scopus, PubMed, Medline, and Google Scholar databases online. A search strategy using PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analysis) guidelines was developed. This search strategy recognized the abstracts of published articles, while other research articles were discovered manually from the citations. Randomized controlled studies, observational studies, comparative studies, case series, and case reports were evaluated for eligibility. There were total numbers of articles 96 (32 case reports; 34 cases series; 30 original articles) evaluated. This paper focuses on the history, evaluation of candidacy, audiological evaluation, cochlear implant in children, cochlear implant in unilateral deafness, bilateral cochlear implant, cochlear implant in auditory neuropathy, and cochlear implant in genetic deafness. This analysis provides a foundation for future prospective trials in current criteria for the cochlear implant. It will also serve as a catalyst for additional study into the cochlear implant and its relation to criteria.

History

Dr. William House in the 1960s, along with Jack Urban, an engineer made the first implantable device that could stimulate the auditory nerve, making the cochlear implants a reality in clinical practice.⁸ The first cochlear implant program was established at the House Ear Institute in 1980. A 9 year old boy was implanted in 1980 and by 1982, 12 children with age ranges from 3.5 to 17

years had been implanted in their program.⁹ The House/3M device was approved by food and drug administration (FDA) for implantation in adults in 1984 and children in 1986. Nucleus-22 channel implant received FDA approval in June 1990 for children aged 2 years and older. Then momentum gained speedily and by the mid-1990, a greater number of children were being implanted than adults.⁹

Evaluation of candidacy

The basic evaluation of the cochlear implant candidates includes medical, audiological, and radiographic evaluation.¹⁰ A thorough evaluation should be done to determine the etiology of hearing loss. Pre-lingual versus post-lingual hearing impairment as well as the duration of deafness is important history to predict the outcome after cochlear implantation. The majority of a pediatric cochlear implant is pre-lingual deaf children who are born with sensorineural hearing loss due to genetic mutation (e.g. Connexin 26), perinatal environmental exposures, or idiopathic etiologies.¹¹ These pediatric patients usually get good speech outcomes after cochlear implantation with best outcomes, occurring when implantation is done within 1 to 2 years of age.¹² However, pre-lingual adolescents are not good candidates for cochlear implantation and have a high chance of being non-users of the device. Because of the long duration of hearing loss, reorganization takes place at the auditory cortex, leading to take over of the auditory cortex by other somatosensory inputs like vision.¹³ The FDA criteria for cochlear implantation are not consistent across the devices or manufacturing companies. To complicate the candidacy, insurance companies also have varied criteria/guidelines, but are similar to FDA guidelines (Table 1) provides a broad overview of the candidacy guidelines for conventional cochlear implantation.

Table 1: Candidacy guidelines for cochlear implant.

| Parameters | Children (12-24 months) | Children (2-17 years) | Adult |
|-----------------------------|---|---|---|
| Hearing threshold | Profound SNHL (>90 dB) | Severe to profound SNHL (>70dB) | Moderate to profound SNHL in both ears(>40dB) |
| Recognition of words | Limited benefit from binaural amplification trial based on MAIS | Limited benefit from binaural amplification defined by ≤20 to 30% word recognition scores | Limited benefit from binaural amplification defined by ≤50% sentences recognition in the ear to be implanted (or ≤40% by CMS criteria) and ≤60% in contralateral ear or binaurally. |

CMS: Centers for medicare and Medicaid services; dB: Decibel; SNHL: Sensorineural hearing loss; MAIS: Meaningful auditory integration scale.

Audiological assessment before cochlear implantation

In children with congenital prelingual profound hearing loss and perilingual deafness, the indication of cochlear implantation is primarily based on the audiological threshold, which is objectively determined by using

frequency-specific brainstem evoked response audiometry (BERA).¹⁴ The limit of cochlear implant indication presently is considered to be an average audiological threshold of >70 dB, as it is usually no longer possible to achieve sufficient speech intelligibility with conventional hearing aids above this threshold.¹⁵

Addition to audiological assessment, one should consider the level of language development, communication skills, general level development and socio-family aspects when considering the indications as part of the interdisciplinary pre-diagnostic work-up.¹⁶ Pure tone audiometry and otoacoustic emissions (OAEs) are two important audiological tests are done other than BERA before cochlear implantation.

Cochlear implant in children

Presently universal newborn hearing screening is helpful to detect infants with hearing loss. The early identification of hearing loss provides greater opportunities for early intervention. Early cochlear implantation in children is highly helpful for normal hearing and speech development.¹⁷ Speech development in children occurs from birth and is usually complete by the age of six years.¹⁸ The speech quality, language skill, expressive and receptive vocabulary are improved by exposure to aural language from as early as possible. One study showed that children implanted between the age of 12 to 36 months outperformed those implanted between the ages of 37 and 60 months.¹⁹ For several years, the lower limit for age at cochlear implantations was two years. The first clinical trial to select children less than two years of age was with the Clarian device. The Clarian device can be selected for children of 18 months if the treatment showed ossification. Cochlear implant devices can be safely used for children 12 months and older. A decrease in age at the time of implantations is presently limited by the nature of audiological tests in children. Use of modern techniques, a confident evaluation can be made at younger ages than adults. If hereditary deafness or meningitis, a confident assessment can be done at younger ages than the adult. Evaluation of children with a cochlear implant in less than one year of age should include behavioral audiometry, bilateral OAES, ear-specific and frequency-specific auditory brainstem response (ABR), auditory steady-state response (ASSR), bilateral tympanometry, and acoustic reflexes.

Cochlear implant in unilateral deafness

There are different treatment modalities of unilateral deafness to dates such as no treatment, conventional contralateral routing of signals (CROS), or bone-anchored hearing aid (BAHA) hearing aid. Cochlear implantation is a new treatment option available for patients with unilateral deafness. One study was done on eleven adult patients with unilateral deafness of different causes.²⁰ that study aimed to evaluate the use of one-sided electrical stimulation with normal hearing on the contralateral side and after six months in comparison to pre-operative unaided condition, conventional CROS or BAHA inteno mounted on the soft band/tension clamp or with a CROS hearing aids. After test periods with both devices, the patients received a cochlear implant. Here, the authors revealed that cochlear implantation enhanced the hearing abilities in a person with single-sided

deafness and is superior to alternative treatment modalities. The use of a cochlear implant did not hamper the speech understanding of the normal hearing ear. Their study suggested that the bilateral integration of electric and acoustic stimulation is possible even with one-sided normal hearing. Detail indications for bilateral and unilateral cochlear implantations are given in (Table 2).

Table 2: Indications for cochlear implantation.

| Hearing impairment | Cochlear implantation |
|---|---|
| Bilateral profound hearing loss | Bilateral cochlear implant |
| Unilateral hearing loss with normal hearing in contralateral ear | Unilateral cochlear implantation |
| Unilateral hearing loss with impaired hearing on contralateral ear | Bimodal treatment (Cochlear implantation and hearing aid in contralateral ear) |
| Congenital bilateral hearing loss | Bilateral cochlear implantation in first year of life |
| Acquired hearing loss | Bilateral or unilateral cochlear implantation with no age limit |
| Hearing loss following meningitis/labyrinthitis/trauma | Bilateral or unilateral cochlear implantation preferably within 4 to 6 weeks |
| Hearing loss with usable residual hearing in low frequency range | Electric acoustic stimulation (EAS) cochlear implantation with hearing aid on the affected side |

Auditory neuropathy and cochlear implant

Auditory neuropathy is a type of hearing impairment with progressive or transient moderate to profound sensorineural hearing loss where the function of outer hair cells of the cochlea is preserved but the afferent neural activity of the auditory nerve and central auditory pathways is disordered.²¹ The incidence of auditory neuropathy is approximately 10% to 14% in children diagnosed with severe to profound sensorineural hearing loss.²¹ Audiological assessment shows normal pre-neural cochlear activity in otoacoustic emission responses but evoked response from the auditory pathway is commonly absent. Cochlear microphonic responses (generated by polarization and depolarization of cochlear outer hair cells) are also present. Hearing aids are rarely helpful in these cases. However, cochlear implants are debated if the lesion is in the cochlea, so bypass the inner hair cells with direct stimulation to the VIII cranial nerve and give good results. If the lesion is in the auditory nerve, the electrical stimulations may give limitations, so many clinicians are very conservative for cochlear implantation as an option in auditory neuropathy. In a large study

conducted at Sydney cochlear implant centre, Australia, many of the children with auditory neuropathy received successful cochlear implantation with a very smaller number of failures to get maximum benefit.²²

Cochlear implant in genetic deafness

The hereditary etiology is responsible for more than 60% of all prelingual hearing loss whereas iatrogenic or environmental causes are responsible for the remaining 40%.²³ Some of these patients show good results with amplification, but the majority of them need cochlear implantation. The scientific literature is not clear about the success of cochlear implantation in genetic hearing impairment. Connexin 26 (Cx26) mutation is responsible for approximately 50% of non-syndromic autosomal recessive hearing loss, which is the most common etiology for non-syndromic hereditary hearing loss. Dominant Cx26 mutations are associated with syndromic diseases with skin manifestations such as keratitis ichthyosis deafness syndrome and palmoplantar keratoderma with hearing loss and these are rare.²⁴ A temporal bone study of a heterozygous Cx26 mutation showed intact spiral ganglion cells, no neural degeneration, lack of hair cells in the organ of Corti, and agenesis of the stria vascularis, which may explain good results occur after cochlear implants in these patients, as they have higher neural integrity of the peripheral and central and auditory system.²⁵ Usher syndrome is a common cause for deaf-blindness in humans and it is an autosomal recessive syndromic deafness characterized by sensory impairment of ears and eyes which leads to congenital sensorineural hearing loss and retinitis pigmentosa and posteriorly, retina degeneration, loss of night vision after ten years of age, restriction of visual field and sometimes blindness in adolescence temporal bone study of Usher syndrome patients, severe degeneration of basal turn of the organ of Corti, atrophy of stria vascularis and reduction in spiral ganglion cells were seen. The cochlear neurons were decreased with approximately 68% of neuronal loss compared with age-matched controls.²⁶ Patients with Usher syndrome have low-frequency residual hearing and little help from amplification, so cochlear implantation is done as early as possible because vision impairment makes sign language a temporary solution to the patient. One study showed that early implantation is critical for developing effective oral auditory skills before vision loss, so the best output is seen in children implanted before age of 3 years.²⁷ The best prognostic factor following cochlear implants is thought to be the age of implantation and not genotypic mutations. Other benefits of cochlear implantations are quality of life and independent living.²⁸ Waardenburg syndrome is an autosomal syndrome characterized by sensorineural hearing loss, dystopia cantorum, hyperplasia of the eyebrows, heterochromia iridis, and a white forelock which affects 1 in 40,000 live births and accounts for 2 to 5% of all congenitally deafened children.²⁹ Study on temporal bone Waardenburg syndrome shows atrophy of the organ of Corti and stria

vascularis.²⁹ Treatment includes amplification and cochlear implantation for profound sensorineural hearing loss. There is also an increased incidence of auditory neuropathy in this patient, which may undermine the efficacy of a cochlear implant.³⁰

Bilateral cochlear implant

The environmental sound sources are usually multiple sound origins which are a challenge for the auditory system. The binaural system helps provide cues that segregate target signals from competing sounds and it identifies sound sources in normal-hearing listeners. Binaural hearing is the result of integration between inputs from two ears and auditory pathways. The result of the binaural hearing is better speech understanding when competing sounds are present. The head shadow effects occur during listening when speech and noise are spatially separated. For example, background noise coming from the right side would interfere with the right ear but the head would obstruct (create an acoustic shadow) some of the interfering noise from reaching the left ear. So, the head shadow effect results in a better signal-to-noise ratio in the protected left ear. A listener is selectively attending to the ear with a better signal-to-noise ratio to improve intelligibility. The head shadow decreases high-frequency sounds by approximately 20 decibels but low frequency by only 3 to 6 decibels, and this effect does not need central auditory processing.³¹ The head shadow effect has a larger impact on hearing with binaural cochlear implantation. The brainstem auditory nuclei process differences in timing, amplitude in improved separation of noise and speech. Evidence of benefit of binaural squelch effect is limited, not found in all users, and is not as large as seen with head shadow effect.³² Best moments to perform a second cochlear implant should be informed to the patient if sequential or simultaneous implantation is planned. Surgeons should inform the patient about the same risks from the first operation and the additional benefits with the second one may not be so great (it only increases approximately 20%).³³ In this way, simultaneous than sequential binaural cochlear implantation is advisable because of avoidance of a second hospitalization and general anesthesia and it will be possible to use a similar process for both implants with a reduction of the cost. In the case, children using binaural cochlear implants, the development of normal patterns of cortical activity occurs when inter-implant delays are reduced.³⁴ In contrast to bilateral cochlear implantation, bimodal stimulation refers to a condition where a unilateral cochlear implant is placed in one ear and a conventional hearing aid is kept in the non-implanted ear. This dynamic is done in patients who have residual acoustic hearing in the nonimplanted ear and it offers selected benefits over the monaural condition in terms of speech understanding in quiet and in noise as well as sound localization.³⁵ The addition of acoustic stimulation to conventional cochlear implant technology can broaden auditory inputs to the brain, so improve the hearing abilities.^{36,37} The

disadvantages of bimodal hearing are the subjects fuse the electrical and acoustic signals centrally.^{38,39}

CONCLUSION

In current years, the criteria for cochlear implantations are changing. To get optimal benefits, early cochlear implantations in prelingually deaf children are required. Additional disabilities are no longer contraindicated for performing cochlear implantation. Nowadays the criteria for cochlear implantation include deafness but also residual hearing. The combination of electric and acoustic stimulation is an established technique for the treatment of hearing loss still functioning in the low frequencies. Because of the benefits of bilateral hearing, bilateral cochlear implantation has become the standard option in the last decade. Although rare, recent experiences show the benefits of a cochlear implant in unilateral deafness and with severe tinnitus. Clinicians often expect rehabilitation of language communication skills with cochlear implantation.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Swain SK. Current criteria for selecting cochlear implant in deaf patients: a review *Int J Adv Med* 2022;9:50-5.