

COCHLEAR IMPLANT: INDICATIONS, CONTRAINDICATIONS AND COMPLICATIONS

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ABSTRACT

Cochlear implant (CI) is a surgically implanted electronic device that provides a sense of sound to a person who is profoundly deaf. CI represents the current treatment for patients affected by profound sensorineural hearing loss (SNHL). At first times, only deaf adults were considered to be candidates for a CI; however, with the development of technology and gained experience by physicians, indications for cochlear implantation have been expanded. Today, CIs are implanted also in children and broader indications are followed. There are, however, a number of patients who are potential candidates for CI but do not completely fulfill the current indications. Residual hearing and duration of deafness represent prognostic indicators for CI performance; however, prelingually deafened adults and children with residual hearing are still point at issue. Anatomical variations such as cochlear malformation, ossification and chronic otitis media still represent a contraindication to CI for some surgeons. Although the technology for cochlear implant surgery is advancing everyday, there may be several complications related to the device or operation and some of them can be healed only with medical therapy but some are serious enough to process to reimplantation. In this article the indications and contraindications for cochlear implant candidates and complications that may occur during or after the operation were reviewed under the light of the literature.

Keywords: Cochlear implant, indication, contraindication, complication, reimplantation

INTRODUCTION

Hearing loss is one of the most common health problems. Approximately 1-3 out of 1000 babies is born with a hearing loss (1). Lack of acoustic stimuli to the auditory system arising from sensorineural hearing loss has been usually coped with the inception of cochlear implantation in the 1980s (2). Cochlear implants (CI) can improve speech and sound

perception, promote development of verbal language and auditory skills and educational development and also suppress tinnitus in some patients (2). Depending on the grade of hearing loss, affected individuals can be fitted successfully with hearing aids. For patients with hearing loss that is not relieved with hearing aids, a cochlear implant may present an opportunity for hearing.

The CI is a device which is surgically placed into the cochlea and converts sound to an electrical signal. This signal is conducted via electrodes to the spiral ganglion cells in the cochlea. As of 2012, an estimated 324,000 patients worldwide have CIs implanted (3). But, this number represents only a small number of individuals potentially benefitted from implantation.

Although individual reactions to CIs depend on a number of psychosocial and physical factors, the new trend for improving performance with more

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sophisticated hardware i.e. electrodes and programming strategies has loosened the indications for cochlear implantation in recent years (3). Plenty of devices are approved recently for use in patients with unilateral hearing loss rather than a bilateral profound loss that was required previously (2). Additionally, some studies have begun using shorter electrode arrays for shallow insertion in patients with substantial low-frequency residual hearing. This technique will allow the recipient to continue to use amplified hearing aid for the low frequencies simultaneously with a CI for the high frequencies.

Complications are unavoidable with cochlear implantation as with all surgical procedures. They may be easy to treat like temporary vertigo or terrible enough like death due to meningitis. It is advisable that surgeons without prior experience should not perform an operation themselves; they should work with a more experienced surgeon when performing their first implant.

In this article, indications and related etiologic factors, contraindications and complications of cochlear implantation were discussed according to the literature.

INDICATIONS OF COCHLEAR IMPLANTATION

From the first introduction of this device, FDA which has great influence on cochlear implantation candidacy has pursued several clinical trials on this issue. Many supplements about the technological improvements of the devices have been presented to the FDA. Therefore, implant candidacy is generally based on the best knowledge and the decision of the surgeon following the patient (2). Patients, child or adult, with severe-to-profound bilateral sensorineural hearing loss and some pathologies limited to the cochlea are potential applicants for cochlear implantation. There are some classical criteria for candidacy but changes have firstly included implanting patients with increasing degree of preoperative open-set speech skills, patients with increasing degree of residual hearing, patients, especially children at younger ages and with abnormal cochlea as much as possible (2).

The most widely used criteria for candidacy include bilateral severe-to-profound sensorineural hearing loss (SNHL), the optimal age should be between 12 months and 5 years but younger ages can

also be the candidate, lack of improvement in hearing and speech benefits from hearing aids after a 3 months usage, the family of the patient will promote the surgery and postoperative rehabilitation, the family and the patients have convenient hope for the outcome of the surgery and are available for rehabilitation and education and also there should be no contraindications for surgery (4). If the audimetric candidacy is described distinctly, it ranges from severe to profound SNHL for ages from 2 to 17 years and profound SNHL for ages from 12 to 24 months, that is for children aged 12-24 months, the pure-tone average (PTA) should equal or exceed 90 dB (3). For patients older than 24 months, the PTA should equal or exceed 70 dB. If the patient can detect speech with best-fit hearing aids, a speech-recognition test in a sound field of 55 dB HL sound pressure level (SPL) is performed (3). A number of speech recognition tests are currently in use. Speech recognition criteria include limited benefit from binaural amplification trial with word recognition tests like MLNT (Multisyllabic Lexical Neighborhood Test) and LNT (Lexical Neighborhood Test) scores $\leq 30\%$ for 2-17 years and limited benefit from binaural amplification trial for 12-24 months (2,3) (Table 1).

If the prelingual patient is at younger age when the implantation is performed, the outcome will be better. As a result, the patient will make full usage of the brain, auditory deprivation can be prevented, and speech and language abilities can be developed easily because the brain cannot lose its plasticity. Before implantation, children bigger than 6 years of age and adolescents should wear fitted hearing aids and have some auditory practice. Also they should be given language training while they are young (4).

For postlingual patients, some additional criteria can be as postlingual deafness of all ages, no or little benefit from hearing aid amplification with preoperative scores for speech recognition test is lower than 50% in ear to be implanted and 60% bilaterally in best-aided condition (2,4). Also, emotional and psychological stability with good motivation is important for candidacy. The language environment of the home and workplace after surgery affect the outcome. Another affecting factor for the outcome is the onset age and duration of hearing loss. Generally, an early onset and a long duration of hearing loss have poorer outcomes for postlingual patients (4,5).

Table 1. Indications for patients related with cochlear implant candidacy (SNHL – sensorineural hearing loss, PTA – pure-tone average)

Indications of the Cochlear Implantation		
Audiometric candidacy		
12-24 months	2-17 years	>18 years
Profound SNHL - PTA \geq 90 dB	Severe to profound SNHL- PTA \geq 70 dB	Severe to profound SNHL- PTA \geq 70 dB
Speech recognition tests		
12-24 months	2-17 years	>18 years
Limited benefit from binaural amplification	Limited benefit from binaural amplification with scores \leq 30%	Limited benefit from binaural amplification with scores lower than 50% in ear to be implanted and 60% bilaterally
Prelingual SNHL Perlingual SNHL Postlingual SNHL New concepts in indications		
Younger age, Hearing preservation, Unilateral deafness, Tinnitus, Bilateral implantation		

In addition to these criterias, the indications have been loosened for candidacy because of some reasons mentioned above e.g. technological developments or influences by some health agencies. New concepts in the candidacy are dealing with the following topics; CI and hearing preservation, cochlear implantation at younger ages, CI and relation with genetic hearing loss, CI for unilateral deafness and tinnitus, CI and neural plasticity and bilateral cochlear implantation (2,5). In the ongoing part of the article, some etiologic factors resulting in hearing loss related with the candidacy of cochlear implantation will be discussed.

ETIOLOGIES OF HEARING LOSS AND CANDIDATES FOR COCHLEAR IMPLANTATION:

A-Prelingual Causes

Genetic

Hereditary hearing loss accounts for almost 50% of all congenital sensorineural hearing loss, and it is caused by genetic mutations. It can be the result of a mutation in a single gene, or combination of mutations of different genes (6). Hereditary hearing loss can either be non-syndromic, which is restricted to the inner ear, or syndromic and a part of multiple anomalies affecting the body. Non-syndromic HL can further be categorised by its mode of in-

heritance (6). Approximately 20% of non-syndromic sensorineural hearing loss (NSSHL) is inherited as autosomal dominant, which is also referred to as DFNA; this type of hearing loss is usually delayed onset. Eighty percent of inherited HL is autosomal recessive (DFNB), in which hearing loss is generally congenital, but some forms may emerge later in life. The inheritance of the remaining types of HL is either mitochondrial or X-linked (DFN) (less than 1 percent). To date, 125 deafness loci have been reported in the literature; 58 DFNA loci, 63 DFNB loci and 4 X-linked loci (7).

The GJB2 gene encodes for the protein Connexin 26 (Cx26) and the most common cause of nonsyndromic autosomal recessive deafness is mutation in Cx26, which is responsible for approximately 20% of hereditary hearing loss in children (8). Connexins are transmembrane proteins forming gap-junction channels which allow cell-to-cell communication and ion transport. Some examples of syndromic hearing loss are Waardenburg, Stickler, Brachio-oto-renal, Treacher Collins, Neurofibromatosis, Usher, Jervell Lange-Neilsen, Alport, and Pendred syndromes (8).

Infectious

About the nonmeningitis causes, the most common environmental cause of hearing impairment in children is congenital Cytomegalovirus (CMV) (9). Twenty five to thirty three percent of childhood deaf-

ness can be generated by non-genetic or environmental causes. Asymptomatic congenital CMV infections can be a cause of mild to profound sensorineural hearing loss in 10-15% of patients (9). Rubella, toxoplasmosis, syphilis, measles and mumps are some of the other potential infectious agents causing hearing loss.

Auditory neuropathy/dyssynchrony

Auditory neuropathy is a kind of hearing loss in which the outer hair cells are present and functional, but sound can not be transmitted truly to the auditory nerve and brain. It is also known as auditory neuropathy/auditory dyssynchrony or auditory neuropathy spectrum disorder and implicates the vestibulocochlear nerve as the site of pathology (10). Otoacoustic emissions, the cochlear microphonic of electrocochleography or pure-tone audiometry can be used to assess outer hair cell function. A defect in spiral ganglion cells, inner hair cells or the synapse between the two can be the cause of this disorder (10).

Auditory neuropathy is seen in both children and adults. Normal to profound hearing loss can be obtained on pure-tone audiometry, but especially speech discrimination testing with poor performance is the common criteria in patients with auditory neuropathy (10). The most potent risk factors are anoxia, hyperbilirubinemia, and prematurity for auditory neuropathy spectrum. In several studies it was reported that patients had some forms of hereditary nonsyndromic autosomal dominant auditory neuropathy progressing to outer hair cell malfunction causing decline in pure-tone audiometer results. Some of the patients in these series benefitted from hearing aids or FM devices but in some a CI is the only way to communicate (10-12).

Hyperbilirubinemia

In the neonatal jaundice, bilirubin may pass the blood-brain barrier and be deposited in the ventral cochlear nucleus and can cause sensorineural hearing loss. On the ABR test, transient loss of wave IV and V is seen in 1 to 3 of newborns with bilirubin levels of 15-25 mg/dL. Higher concentrations of bilirubin in the blood are also a risk factor for auditory neuropathy (13).

B-Perlingual and/or Postlingual Causes

Infectious

One of the infectious causes for hearing loss is meningitis. The most common organisms causing

meningitis include *Haemophilus influenzae*, *Streptococcus pneumoniae* and *Neisseria meningitidis*. Approximately 9% of childhood deafness can be caused by meningitis (14). The highest incidence of organisms that cause hearing loss has *S. pneumoniae* with 31% (14). If *S. pneumoniae* is the causative organism for hearing loss, patients are developing obstruction of the cochlear lumen ie labyrinthitis ossificans and it makes CI surgery difficult (14,15).

Ototoxicity

Plenty of medications can cause hearing loss and the most well known ototoxic medication are aminoglycoside antibiotics (16). Other commonly known ototoxic medications include salicylates, loop diuretics, erythromycin, vancomycin, quinine and cisplatin. Because of severe to profound sensorineural hearing loss after ototoxicity, CI is a good treatment option.

Trauma

After temporal bone trauma, conductive hearing loss most commonly happens because of tympanic membrane perforation, ossicular displacement or hemotympanum. The otic capsule is more resistant to trauma but sometimes breaks may involve the labyrinth or/and cochlea. If a fracture passes from the otic capsule, it almost always results in severe-to-profound sensorineural hearing loss. It is very uncommon to see bilateral otic capsule fractures but it would be an indication for cochlear implantation (17). After the fracture of the otic capsule, there would be intraluminal ossification or fibrosis which can make electrode insertion much more difficult. There should be preoperative imaging before surgery for providing additional information about fibrosis or ossification.

Noise-induced hearing loss

According to the World Health Organization (WHO), the number of people affected worldwide by hearing loss increased from 120 million to 250 million worldwide from 1995 to 2004 (18). Much of this impairment (16%) may be caused by exposure to noise on the job (18). Noise induced hearing loss can be temporary or permanent. Acoustic trauma occurs after exposure to sounds louder than 140 dB, such as sounds of gunshots or explosions. Outer hair cells are the most susceptible to noise exposure. After exposure to noise, the outer hair cells swell typically but if

the noise exposure ceases, they may normalize. Most commonly a notch at 4000 Hz on pure-tone audiometry can be seen in the setting of hearing loss secondary to noise exposure, but hearing loss may advance to a degree and amplification can not provide good benefit then cochlear implantation may be considered (3).

Tinnitus

Tinnitus refers to abnormal perception of sound although there is actually no sound and it currently affects nearly 10% of the population (19). When tinnitus starts to reduce the quality of life, patients want to be cured and look for appropriate treatment. For patients experiencing severe tinnitus, tinnitus retraining therapy (TRT) is one of the proven treatment options. Because of its expense, limited number of programs, and duration of therapy, a significant number of patients with severe tinnitus leave TRT and remain without treatment (19).

In some studies it was reported that some patients with preoperative tinnitus which were CI recipients had informed either abolition or reduction of their tinnitus when they wore their CI (20, 21). In a recent study, patients with single-sided deafness were implanted for severe tinnitus and in 20 out of 21, tinnitus was reduced or ceased significantly. Three of the patients in this study even displayed constant residual inhibition while not wearing their processor. Up to date for severe tinnitus, cochlear implantation may be one of the most successful treatment options (20).

Ménière disease

Meniere disease is characterized by room spinning vertigo, aural fullness, tinnitus and fluctuating hearing loss. The diagnosis of this disease is made with a whole history and complete examination of the head and neck region, as well as imaging modalities to rule out retrocochlear pathology. Exact diagnosis can be made by examination of histology of temporal bones in subjects with Meniere disease and it reveals the dilation of the endolymphatic compartment. It is not clear whether endolymphatic hydrops is the final result of some other pathologic processes causing symptoms of Meniere or is the main reason for the symptoms.

House et al reported that in their study sample, in 11% of patients, Meniere's disease was bilateral at presentation and meanwhile an additional 12% (14%

of unilaterals) became bilateral during the follow-up period (22). It is difficult to predict progression of hearing loss in Meniere but severe to profound sensorineural hearing loss may be a result. Recent studies on patients with Ménière disease underwent CI had overall better performance (23,24).

Presbycusis

With aging, hearing loss initially begins with high frequencies, than progresses to lower frequencies. Thirty to thirty five percent of 65 year olds have some hearing loss; this percentage rises to 40-50% for people over 70 years (25). Men are more commonly affected from presbycusis. Conventional hearing aids often supply benefit for patients, but only 20% wear this device. Patients with profound hearing loss who are unable to take advantage of traditional amplification may utilize from a CI (26). Sometimes some patients have residual low frequency hearing, and they may use hearing aid/CI combination providing both electrical and acoustic stimulation (27).

CONTRAINDICATIONS

Contraindications to cochlear implantation may include deafness because of lesions of eight cranial nerve or brain stem. They can be analyzed in two ways; relative and absolute. The absence of cochlear development demonstrated on CT and MRI as in Micheal deformity and cochlear atresia and lack of auditory nerve are some of the absolute contraindications. Several other absolute contraindications may be severe mental disease, severe mental retardation to cooperate with speech training and acute or chronic otitis media and mastoiditis without eradication of the disease (2-4,6). Certain medical conditions like pulmonary, cardiac and hematologic conditions, uncontrolled epilepsy and not being available for rehabilitation remain relative contraindications. But secretory otitis media is not a surgical contraindication for cochlear implantation (Table 2).

Single-stage or more than one-staged surgery can be applied in chronic otitis media with tympanic membrane perforation (4). In single-stage surgery, any disease within the middle ear and mastoid cavity is eliminated by performing a myringoplasty and then the cochlear implantation is performed in the same procedure. More than one-staged surgery means firstly eliminating any disease, performing a

Table 2. Contraindications for the cochlear implant

Contraendications for the Cochlear Implantation	
Absolute	Relative
Agenesis of Cochlea	Inappropriate medical conditions (e.g. pulmonary, cardiac, hematologic)
Agenesis of Cochlear Nerve	Uncontrolled epilepsy
Severe mental disease	Not being available for postoperative rehabilitation
Acute/chronic otitis media and mastoiditis without eradication of the disease	
Mental retardation to cooperate with speech training	

myringoplasty or tympanoplasty and then performing the cochlear implantation 3–6 months later (4,5).

COMPLICATIONS OF THE SURGERY

Complications of cochlear implantation are in a wide spectrum from temporary vertigo to death due to meningitis and related with medical, surgery or device problems (28). In literature, several forms of complication classification have been reported.

Kempf et al. classified the complications as early and late (29). They named the complications emerging in one week to 3 months – early complications and after 3 months – late (29). The complications of cochlear implantation were classified as major and minor by Summerfield and Marschall (Table 3) and they argued that minor complications can be healed with medical therapy without hospitalization but major complications may need hospitalization, revision surgery or reimplantation (30,31).

Infections are the most common complications of the cochlear implantation. The frequency is between 7-16.6% (32). The most common causative organism in simple skin and wound infections is *Staphylococcus aureus*. It can be cured with oral antibiotic treatment without any need for hospitalization. Perioperative antibiotic prophylaxis may decrease the infection rates significantly.

Skin flap infections course is more serious and may need reimplantation. Hematoma may be a predisposing factor (33). Little incisions rather than bigger ones have decreased the incidence of all type of infections.

Pseudomonas infections can cause resistant infections around the implant and the possible reason for resistance is biofilm layer which decreased

the penetrance of antibiotics and makes a barrier to immune mechanisms. Despite prolonged antibiotic therapy, reimplantation may be required (34).

Cholesteatoma is one of the late complications of cochlear implantation. During surgery, attention should be paid to protect the external auditory canal and especially bony tympanic annulus. Patients with persistent otorrhea after surgery should arouse suspicion for cholesteatoma (35).

Perilymph gusher can be seen during surgery as one of the intraoperative complications. It shows itself with leakage of cerebrospinal fluid to surgical field during cochleostomy. It is commonly seen in children with cochlear anomaly.

Facial nerve damage is rare. Intraoperative monitorization helps to protect the nerve. If not treated carefully, burr shaft can damage the nerve and temporary facial paresis occurs. Also chorda tympani can be damaged during surgery and deterioration may be observed in the sense of taste. Therefore, surgery must be performed with the rich irrigation (36).

Magnet or receiver may be dislodged secondary to minor head traumas. But also allergic reactions, new bone formation or infections may be the causes of displacement. According to some authors, placing the implant into the bone socket properly and suturation to the neighboring tissues can prevent the dislodging (37).

Because of some complications, reimplantation surgery should be performed (32). These are:

- ❖ Serious skin infection
- ❖ Device failure
- ❖ Serious device failure

Table 3. Major and minor complications after cochlear implant surgery

Major and Minor Complications of Cochlear Implantation	
Major	Minor
Cerebrospinal fluid fistula	Pain
Device Failure	Simple skin infection
Displacement of the electrode	Dizziness
Misplacement of the electrode	Imbalance
Facial nerve paralysis or damage	Temporary facial paresis
Facial stimulation	Otitis media
Flap infection or hematoma	Hematoma on scalp
Perilymph gusher	Tinnitus
Rejection of the implant	Vertigo
Displacement of the implant	Chorda tympani damage
Mastoiditis	Damage to the dura
Meningitis	
Retraction pockets or cholesteatoma	
Subdural hematoma	

❖ Mild device failure

And some complications may need revision surgery but no need for reimplantation (32);

- ❖ Skin infection (for drainage or debridement)
- ❖ Cholesteatoma
- ❖ Displacement of the magnet
- ❖ Displacement of the receiver
- ❖ Displacement of the electrode or early recognition of wrong placement.

CONCLUSION

The purpose of this review article was to provide an overview on the etiologies of hearing loss circumvented with cochlear implantation and to provide a discussion on indications, contraindications and complications of CIs. A great number of changes occur and indications are loosened for candidacy for cochlear implantation. Generally, accompanying and satisfactory changes in surgical techniques have also been accompanied by. Together, this has improved the benefit and safety of cochlear implantation. Indications for, and the advances in design of

cochlear implantation have been paired by improvements in surgical practices and decrease in complications. The improvements in safety and influences have further encouraged the use of these devices. To reduce the surgery-related complications, the patient should be carefully assessed preoperatively and surgeons without prior experience should not perform an operation themselves; they should work with a more experienced surgeon when performing their first implantation.

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