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[Review Article]

Cochlear implants and electrical brainstem stimulation in sensorineural hearing loss

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Abbreviations ABI: auditory brainstem implant; MRI: magnetic resonance imaging

Abstract

Cochlear implants and multichannel auditory brainstem implants enable patients with bilateral total or profound hearing loss to receive at least acoustic information. Both types of prosthesis are based on electrical stimulation of the auditory pathway. Different speech coding strategies and the number of electrodes used may influence the postoperative results. The preoperative evaluation of patients is of utmost importance. The cochlear implant is suitable for patients with hearing loss due to inner ear disorders, but who have functioning hearing nerve. Patients with a defect of the hearing nerve can be provided with an auditory brainstem implant. Curr Opin Neurol 12:41-44.

Introduction

Cochlear implants and auditory brainstem implants (ABIs) are electronic audioprostheses. Implantation of both is indicated in functional impairment of peripheral parts of the auditory pathway with complete deafness or severe hearing loss [1-3]. Different coding strategies of cochlear implant systems are making use of the tonotopic organization of the cochlea and cochlear nucleus and the temporal processing of acoustic signals along the auditory pathway. In cochlear implants, an electrode array with up to 22 single electrodes is placed into the scala tympani. In ABIs, disk electrodes are placed on the cochlear nucleus complex in the foramen of Luschka. Implanted patients need postoperative rehabilitation. Postlingually deafened adults are able to achieve an open set speech understanding in 80%, whereas prelingually deafened or children born deaf may attend mainstream schools in at least 60%. Patients provided with an ABI will reach some open set speech understanding in 7-8%. At present, the specific advantage of ABIs is a clear benefit in lipreading capabilities and control of the voice. The number of cochlear implant users worldwide is about 25000 and the number of ABI users 140.

Patient selection

Selection criteria in electrical stimulation of the auditory pathway follow standardized protocols. Our experiences in the 1980s showed that prelingually deafened adults and adolescents had no measurable benefit from cochlear implants. Out of 15 patients implanted from 1985 to 1986, only one was still using the device in 1998. This patient has no open set speech understanding, but some limited acoustic orientation. Congenitally deaf or very early deafened patients should not receive a cochlear implant as adults or adolescents [4]. The aim should be to perform implantation as early as possible, even before the age of 2 years [5-10]. The surgical technique allows an implantation at this age. Bilateral complete deafness or at least severe hearing loss are indications for cochlear implant surgery. Severe hearing loss is accepted as an indication for a cochlear implant if speech understanding is up to 30% for monosyllabic words at 70dB speech sound level with optimally fitted hearing aids. Speech testing, however, is not possible in children under the age of 4 years. In these cases, electrocochleography and measurement of electrically and acoustically evoked potentials is necessary. Detection of thresholds is of greater importance than latency of evoked potentials. Optimal hearing aid fitting is self-evident and allows assessment of the development of children in audioverbal education. Maturation and development of neural networks may be detected with the help of positron emission tomography [11].

Early implantation should follow the diagnosis of deafness in all cases, regardless of the cochlear implant system used. Indication for cochlear implant surgery in the case of inner ear malformation or obliteration due to bacterial meningitis should be discussed carefully [12]. Malformation with aplasia of the eighth nerve is a contraindication for cochlear implant surgery [13]. Neural structures of the internal auditory canal may be differentiated with the help of magnetic resonance imaging (MRI) [14••]. Inner ear malformations with a missing separation between cochlea and internal auditory canal will result in a cerebrospinal fluid gusher, and will thus have an increased risk of developing meningitis postoperatively. Development of hearing and speech cannot be predicted in these cases because information regarding the number of functioning neurones is not available.

Computed tomography and MRI may reveal a large vestibular aqueduct syndrome. Patients with this kind of malformation often suffer from a progressive hearing loss and/or sudden deafness after minor head trauma [15]. Results after cochlear implant surgery in these patients can be predicted to be good to excellent. This type of malformation may also be associated with Pendred,s syndrome.

Deafness may occur as a complication of bacterial meningitis. Especially when caused by *Streptococcus pneumoniae* and *Haemophilus influenzae* infections, deafness is associated with the development of tissue or bony obliteration of the inner ear. Obliteration may occur within several weeks, and our experience showed that visible obliteration seems to start in the region of the semicircular canals, as confirmed by one author [16•]. Early implantation in these cases can result in good rehabilitation. In patients with bilateral obliteration, bilateral implantation has to be considered carefully [17].

Surgical treatment

The surgical procedure follows standardized protocols and varies because of specific features of different devices. Mastoidectomy, posterior tympanotomy and creating a well to house the receiver-stimulator are accepted aspects of the surgical procedure. The receiver-stimulator should be fixed with sutures to the skull, especially in children. Electrode insertion is performed into the scala tympani using a cochleostomy or a round window approach. In cases of obliterated cochleae, several approaches for insertion of the electrode array have been reported [18•,19].

Careful insertion of a suitable electrode array allows preservation of residual hearing [20•,21]. Up to now there have been no reports regarding whether conventional hearing aids have been used in an implanted ear. Our experience in approximately 700 cochlear implant users showed that conventional hearing aids were not used; these patients, however, had overall good to excellent results. Some reports of animal experiments revealed degeneration of the neural elements after trauma of the cochlea. Whether this is relevant to speech, understanding cannot be assessed. Even the latest results have not revealed how many neurones of the spiral ganglions have to survive in order to provide speech understanding with electrical stimulation [22•,23••].

The new generation of cochlear implant systems permit telemetric measurements which allow the functioning of the implant to be assessed during the operation; they also permit electrically evoked potentials to be registered. Registration of these potentials facilitates the finding of hearing thresholds and discomfort thresholds postoperatively and helps to avoid overstimulation. Neural response telemetry might also be helpful in obtaining information regarding the quantity of functioning neural populations, and may allow speech coding strategies to be mixed during use. Thus, an objective measurement can be developed that may allow the stimulation rate for certain areas of the basilar membrane to be individually determined, or that may put more emphasis on the tonotopic coding of signals. Intraoperative measurement of electrically evoked stapedius reflexes has proved to be a useful tool to determine discomfort thresholds [24,25,26•].

Chronic electrical stimulation, also at high rates, has shown no negative effect on hearing capacity so far [27-29]. It seems advisable to place the electrode array within the cochlea as close to the modiolus as possible. This appears to allow better stimulation of the ganglion cells within the spiral ganglion and also allows a sharper differentiation of the canals. Speech understanding should improve and power consumption be reduced. This will influence the development of small speech processors that can be worn as behind-the-ear devices. This will result in better comfort with body-worn parts of the cochlear implant and also in better results in speech understanding.

Results

Results after cochlear implantation are good to very good. About 80-85% of postlingually deafened adults will achieve open set speech understanding. Similar results are obtained in children, even those who are congenitally deaf. Results decline when the number of electrodes is reduced, however. It has also been shown that the duration of deafness has a significant influence on the results of implantation. The longer the duration of deafness (whether congenital or acquired), the more limited the results that can be expected. At least for a certain group of patients, speech understanding can be improved by higher stimulation rates with a reduced number of stimulating electrodes. Another important factor influencing the results appears to be the age of children at the time of implantation. As yet results have not been statistically proved, but the tendency is clear. Younger children are better performers than older ones [30].

Future developments

Future developments of cochlear implant systems will have to deal with increasing the number of electrodes while minimizing the potential risks of electrical stimulation [31•]. Higher stimulation rates and overlapping stimulation of single electrodes may improve speech understanding [32]. The possibility of an implantation close to the modiolus seems to be significant, but it also has to be taken into account that explantation and reimplantation should be possible without further lesions [33•]. Different investigations have been made regarding the compatability of MRI. Furthermore, it is important to decide about the optimal time for implantation. Regarding this, different examinations have to be performed to determine the maturity of the auditory pathway. Completely implantable systems are being considered, which are similar to hearing aids. At present, cochlear implants exist with speech processors that have been reduced in size so that they can be worn like behind-the-ear hearing aids.

Auditory brainstem implants

The ABI is indicated in cases of functional defects of information transfer from cochlea to brainstem (i.e. from spiral ganglion to cochlear nucleus) [34]. Neurofibromatosis type 2 has been found in nearly all patients implanted so far [35••]. Tumours located in the cerebellopontine angle or the internal auditory canal have to be resected when clinical symptoms are present. Because preservation of function of the cochlear nerve is not possible in cases of radical tumour removal, an electrical stimulation can be performed by implantation of an electrode array on the cochlear nucleus, which leads to the perception of acoustic signals [36••,37]. Multiple-channel electrodes have been used since 1992. Thirty-seven patients have been implanted in Europe so far.

We prefer a translabyrinthine/transmastoidal approach for anatomical reasons. The suboccipital (retrosigmoidal) approach will only be chosen in cases of prior surgery via a translabyrinthine approach or in cases of tumour recurrence. The facial and glossopharyngeal nerves will be monitored intraoperatively. Electrical stimulation may help identification of early potentials of the lower auditory pathway. The electrode array with up to 21 single disk electrodes is placed in the foramen of Luschka and fixed with tissue after identification of the cochlear nucleus complex. Receiver-stimulator and body-worn parts of the ABI are similar to those of cochlear implant systems. The electrode array consists of 21 disk electrodes with a diameter of 0.7mm. The number of electrodes should be as high as possible to allow flexibility of stimulation because of somatosensory side effects. An average of 10 (two to 21) electrodes are used. Nearly 96% of ABI patients implanted in nine European clinics as part of a European study can detect acoustic information with electrical stimulation. Nearly 8% of patients have some open set speech understanding and 83% of implanted patients report a benefit regarding their quality of life.

Conclusion

At present, cochlear implants and ABIs help deaf patients to gain access to the acoustic environment. This is of special importance to ABI users. Cochlear implant users, however, reach an open set speech understanding allowing communication in more than 80% of cases. Early treatment of congenitally deaf children results in integration of these children into mainstream schools and in social integration into the hearing world. Development of hardware as well as of new speech coding strategies will help to improve rehabilitation results further. Moreover, understanding of the function of the auditory pathway will be better. Cochlear implant technology may serve as a basis for the development of other sensory prostheses.

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