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New Methods of Deafness and Partial Deafness Treatment

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Implantable hearing prostheses are a preferred medium for (re)habilitation in individuals with sensorineural deafness and profound hearing loss. Cochlear implants are used in hearing impairments of the cochlear origin, while in cases of the retrocochlear origin, auditory brainstem implants are applied. The obtained results indicate that open-set speech understanding has now become achievable for the majority of cohlear and brainstem implant users. Moreover, the new method of partial deafness treatment has been offered for patients remaining beyond the scope of effective treatment with application of cochlear implant or hearing aids.

Keywords: cochlear implants, auditory brainstem implant, partial deafness cochlear implantation

1. Introduction

Hearing is one of the most important of human senses. Access to information and benefiting from modern technologies can be seriously limited or rendered impossible by hearing impairment. Hearing disorders in people of various age have a particularly unfavourable influence on development of the speech and language communication in young children. Usually, etiology of hearing impairments is very complex and includes congenital and acquired defects. The most frequent among acquired diseases are hearing impairments due to upper airways infections and noise, which can also be a cause of acoustic traumas. Development of modern technologies and medical techniques enables us to provide surgical treatment in almost all cases of damages of the middle ear. It also creates possibilities of effective help in damages of the inner ear in patients with total deafness or profound hearing loss, who don't benefit from conventional hearing aids. Application of implantable de-

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vices has had a crucial impact on the development of otosurgery, audiology, biomedical engineering and psychoacoustics. Currently, implants of various types are used as prostheses implanted in the external ear, temporal bone, middle ear, inner ear and brainstem. Appropriate acoustic or electric stimulation allows to effectively improve auditory abilities and introduce born deaf patient or bring back postlingually deafened patient to the world of sounds. However, there is a large group of patients who remain beyond the scope of effective treatment by hearing aids or implantable prostheses. Their hearing impairment is characterized by normal or slightly elevated threshold in the low-frequency band, with nearly total deafness in higher frequencies. It is proposed to describe this type of hearing impairment as partial deafness.

The aim of the paper is to present new and more effective methods for restoring audition in deafened and partially deafened patients, which have been developed and implementd in the Institute of Physiology and Pathology of Hearing.

2. Deafness and Sensorineural Profound Hearing Loss

The hearing part of the inner ear — cochlea contains cells, tissues, fluids, and nerves necessary for conversion of the sound induced fluid pressure waves within the cochlea into neural impulses of the auditory nerve. The hair cells in conjunction with the basilar membrane are responsible for this conversion. If the hair cells are damaged, the auditory system has no way of transforming acoustic pressure waves (sound) to neural impulses, and this, in turn, leads to profound hearing loss or deafness. In this situation, the sound travels through the outer ear, the middle ear, and the inner ear but never makes all the way to the brain because of the broken link — the damaged hair cells. The hair cells can be damaged by certain diseases (e.g., meningitis, Meniere's disease), congenital disorders, by certain drug treatments, by noise or due to many other causes. In sensorineural deafness and profound hearing loss of the cochlear origin, the current treatment is based on the cochlear implant placement [1].

The cochlear implant bypasses the normal hearing mechanism (outer, middle, and part of the inner ear including the hair cells) and electrically directly stimulates the remaining auditory neurons [2].

The system of cochlear implant is the electronic hearing prosthetic device, whose action is based on electric stimulation of the acoustic nerve. The cochlear implant system consists of the external and internal part (Fig. 1).

The internal part is surgically placed in the mastoid portion of the temporal bone and contains a receiver and an electric stimulator in the same housing with the electrode array. The electrode array extending from the implant housing is inserted into the cochlea. The external part is the speech processor that transforms sound into electric stimuli (Fig. 2).

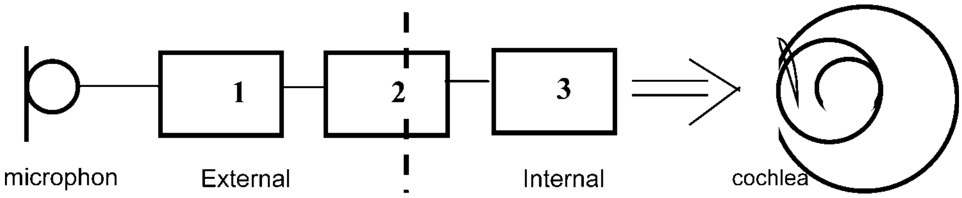

Fig. 1. Components of cochlear implant systems: 1— speech processor, 2 — transcutaneous transmission link, 3 — implanted receiver, decoder and stimulator in the same housing

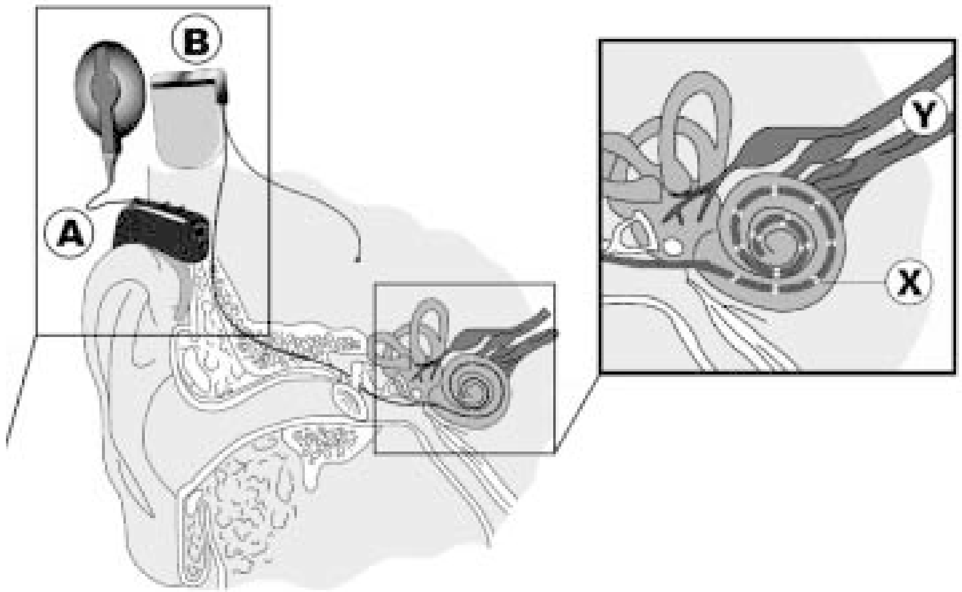

Fig. 2. Scheme drawing of the cochlear implant system: A — speech processor; B — implant; X — active electrodes; Y — auditory nerve

In patients without the functional auditory nerve, as it often in case of Neurofibromatosis type 2 is, the cochlear implant system is not applicable. The auditory brainstem implant was developed for this patient population. The auditory brainstem implant system is similar in design and function to the cochlear implant, but the electrode is placed on the cochlear nucleus complex in the brainstem rather than in the cochlea [3].

3. Cochlear Implant (CI) Program

In Poland, the Cochlear Implants Program was launched at the Institute of Physiology and Pathology of Hearing in Warsaw in July 1992. For 12 years of the Program it has evolved into a complex organizational professional structure which is included in the system of health care for patients with hearing impairment. To maximize the benefit, a detailed protocol for the pre-, intra- and postoperative procedures was implemented [2]. To determine the cochlear implant candidacy, a lot of factors should be taken into account because successful use of the cochlear implant depends on a combination of variables associated with development, cognition and psychosocial functioning as well as auditory experience [1].

According to the current candidacy criteria, children with severe-to-profound bilateral sensorineural hearing loss and as young as 12 months or older and adults with moderate hearing loss in the low frequencies and profound hearing loss in the middle to high speech frequencies bilaterally are consider to receive a cochlear implant [4]. However, guidelines for the implant candidacy are not categorical because of the variety of communication choices available to hearing-impaired populations and because of different responses and benefits from hearing aids, in different patients, for a given level of sensorineural hearing loss. The decision on eligibility can be also determined by patient's motivation and expectations. Thus the current candidacy criteria present a variety of options, and preoperative evaluation represents a complex and multidisciplinary assessment and consists of audiological, medical and psychological evaluation.

The surgical procedure is performed under general anesthesia. A postaurical incision is made and a well is created in the skull, to accommodate the internal part of the device [5]. Then posterior tympanotomy is performed to visualize the niche of the oval window. Cochleostomy is made into the basal turn of the scala tympani and the electrode array is inserted [6]. The implant is then placed into the well behind the mastoid. The incision is closed and a pressure dressing is placed.

The postoperative procedure consists of speech processor fitting, audiological assessment of hearing benefits and the hearing and language rehabilitation.

From the beginning of the program, over 700 patients have been provided with cochlear implants.

4. Results of CI Program

In order to evaluate the hearing and language improvement of cochlear implanted patients diferent tests of speech comprehension were performed in the quiet and the noise [7, 8]. One of them was the monosyllabic Pruszewicz monosyllabic word test in the Polish language (20 words per list, 20 lists). Lists of each test were randomized among test conditions. The speech items were presented via a loudspeaker situated in front of the subject (0° azimuth), at a level of 70 dB SPL. The subject was seated 1m away from the speaker. A speech-weighted noise was presented from the same speaker at a speech-to-noise ratio of +10dB. An unselected sample of 65 deaf adults who had undergone a common protocol of implantation and rehabilitation with Nucleus and Combi 40 + cochlear implant systems were assessed.

Age of the patients at implantation ranged from 18y to 55y. The onset of deafness ranged from 14 to 52, and the duration of deafness ranged from 2 to 11 years. The mean word recognition scores obtained postoperatively are shown in Fig. 3.

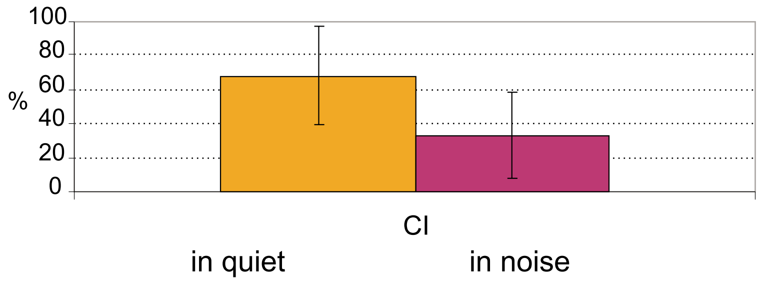

Fig. 3. The postoperative mean score in monosyllabic word test in quiet and in noise

5. Partial Deafness

There is a large group of patients with hearing impairment characterized by normal or slightly elevated thresholds in the low-frequency band, with nearly total deafness in the higher frequencies. We define this type of hearing impairment as partial deafness [9]. Patients in this group remain beyond the scope of effective treatment only by hearing aids. Such patients have not been considered before for cochlear implantation, for fear that this intervention could damage the functioning part of the cochlea. Placement of intracochlear electrode was feared to lead to a complete damage of the remaining hear cells which, in turn, could cause a loss of the low frequencies hearing.

However, considerable improvement in the cochlear implant technology, on one side, and development of new surgical technique called "soft surgery" on the other side, have resulted in preservation of residual hearing after cochlear implant placement [10]. The residual hearing is usually defined as a remaining hearing sensitivity measured in terms of a threshold below 90 dB for low frequencies up to 1000 Hz. Moreover, new concept called electro-acoustic stimulation (EAS) was proposed by von Ilberg et al. [11]. They suggest that the use of a hearing aid and a cochlear implant in the same ear can result in hearing and speech perception that is better than with either device alone. Although the preservation studies and EAS preliminary studies were conducted in patients with much lower thresholds in the low frequencies than partially deafened patients, they were inspiration for consideration of cochlear implants in partial deafness [12].

In partial deafness, a basal part of the cochlea is damaged and it's apical part works according to the tonotopical organization of cochlea (Fig. 4).

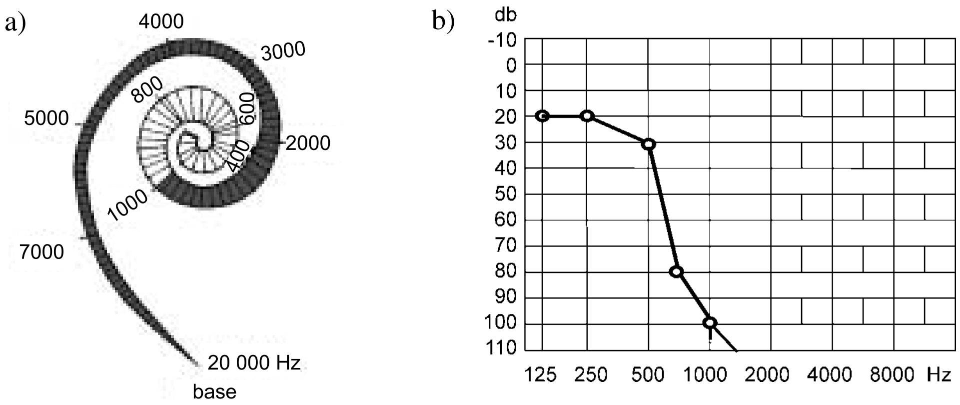

Fig. 4. Partial deafness. a) schematic drawing of the tonotopical organization of cochlea; b) an example of a partial deafness audiogram

Because the total deafness has successfully been treated by means of cochlear implant for quite a long time a decision was made to implant patients with partial deafenes in the way of inserting part of a standard electrode in the region of damaged cochlea. And than to stimulate only the damaged part of the cochlea, preserving the low-frequency acoustic hearing in the implanted ear. So, in the result, electric stimulation on one side would be combined with acoustic stimulation on both sides.

6. Partial Deafness Cochlear Implantation (PDCI) Program

The first Partial Deafness Cochlear Implantation (PDCI) operation was performed in July 2002. Up to now, 14 PDCI surgeries have been performed. All cases were implanted with a Med-El Combi 40+ system, using the standard electrode array. One of the most important and difficult part of the proposed method is preservation of the low frequency acoustic hearing (natural for patient) after the electrode insertion. Therefore, to avoid a loss of the low-frequency hearing, partial electrode insertion is performed, with an approach to the scala tympani directly through the round-window membrane. This surgical approach is different from that used in the standard cochlear implantation. The most commonly used technique is cochleostomy. However, drilling of the cochleostomy hole may lead to a damage of the remaining hear cells and, in turn, may cause loss of the low frequency hearing. The approximated depth of insertion is 20 mm. The electrode array is fixed in its final position using fibrin glue at the round-window niche. The round-window membrane is left partially uncovered, to preserve its mobility. The device is fixed in a well made in the temporal bone during the surgery.

7. Results of PDCI Program

To evaluate preservation of the acoustically-stimulated hearing after the cochlear implantation, audiometric measurements were performed in the implanted ears under the conditions like preoperatively. Pure-tone testing was performed using a Siemens SD5 audiometer calibrated according to standards established by the American National Standards Institute (ANSI). The maximum output of the audiometer was 130dB HL, and a standard clinical procedure was followed in the threshold determination. Testing was performed in IAC soundproof booth under Sennheiser HDA 200 headphones. Hearing was fully preserved (within 0–10dB) in 7 patients, and partially (within 11–20 dB) in 6 patients. In one patient, the maximum threshold difference was 40 dB and in another one hearing was completely lost.

In order to evaluate the hearing and language improvement of partially deafened cochlear implanted patients, tests of speech comprehension were performed in the quiet and the noise using the monosyllabic word test 6 months after the implantation. The mean word recognition scores obtained preoperatively and postoperatively with the cochlear implant plus acoustically-stimulated hearing (without amplification), are shown in Fig. 5.

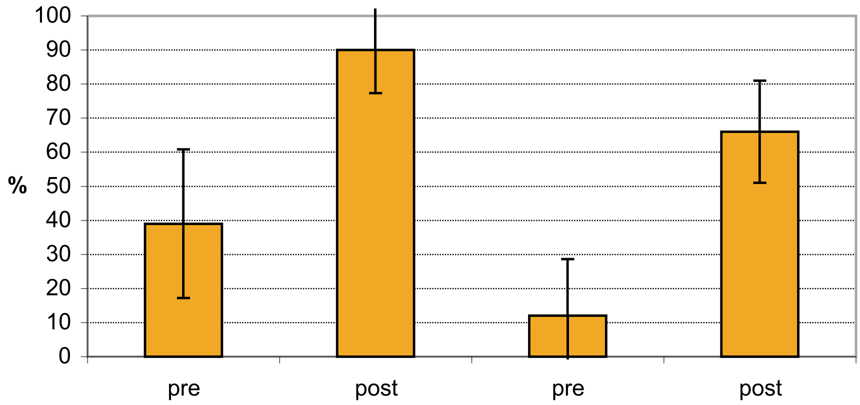

Fig. 5. The pre- and postoperative mean score in monosyllabic word test

8. Auditory Brainstem Implant (ABI) Program

Three patients were provided with auditory brainstem implants at the Institute of Physiology and Pathology of Hearing in 1998–2001 [13]. Auditory Brainstem Implants are a modern method of treatment of total bilateral deafness in cases of the retrotracochlear origin. Development of this highly specialized program was possible thanks to a close collaboration of many specialists in anatomy, audiology, otosurgery, neurosurgery, biomedical engineering, psychology and speech therapy. Bilateral deafness cased by the tumors of cerebello-pontine angle, in vast majority, occurs in cases of neurofibromatosis type 2. Treatment of this pathology for many years was limited to the surgical tumor removal or radiation ("gamma knife"). After successful removal of the tumor of the VIII nerve, cochlear nuclei were localized and multichannel Combi 40+ system was implanted. After activation of the implant system, the patients took part in a two-year rehabilitation program, which is obligatory for adult patients provided within the Brainstem Implant Program.

9. Results of ABI Program

Word recognition scores obtained twelve months after activation of the brainstem implant system were 45% for the first patient and 58% for the second patient. The third patient was not able to discriminate words in the open set test due to poor general conditions (multiple cranial nerves damages, ataxia and other disabilities associated with central and periferal tumors). However, the ability to detect sounds, detection of the beginning and the end of sounds, recognition of some perceptual features of sounds and identification of speech in close sets were proved [14].

10. Conclusions

The analysis of the benefits experienced by cochlear implant users indicates the undeniable success of cochlear implants in many deaf people, although noticeable individual differences in the outcome and benefits were observed. The results of the new method of partial deafness treatment indicate that the combination of the electric plus acoustic stimulation provided a more complete representation of speech frequencies than would be possible with either modality alone. Our results demonstrate a significant benefit of the new method in respect to standard implantation, especially in the noise, an often reported difficult listening condition for cochlear implant users.

Finally, simultaneous removal of the tumors and ABI implantation allows for treatment of life-threatening pathologies. The results obtained in two patients prove that the auditory brainstem implant is the kind of prosthesis that can effectively restore hearing sensations and can be a valuable aid in patient's everyday communication. The case of the third patient indicates a possibility of improvement in the life quality even in the presence of severe neurological handicaps.

The study demonstrates that deafness, partial deafness and profound hearing loss of sensorineural origin is successfully treated using implantable prosthetic devices — either cochlear implants or brainstem implants.

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