Received: 2002.12.05 Accepted: 2003.02.10 Published: 2003.04.23	A new method of partial deafness treatment
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	Summary
Background:	There is a significant group of patients whose hearing impairment is characterized by normal or slightly elevated thresholds in the low frequency band with nearly total deafness in high frequency range. These patients remain beyond the scope of effective treatment by hearing aids. We name this kind of hearing loss 'partial deafness'.
Case Report:	A new method of partial deafness treatment was applied in the case of a young woman. A par- tially-inserted cochlear implant was used to restore hearing at high frequencies, while preserv- ing low-frequency acoustic hearing in the implanted ear.
Conclusion:	The results demonstrate a substantial improvement in speech discrimination and communica- tion skills when electric stimulation on one side was combined with acoustic stimulation on both sides.
key words:	severe hearing loss • cochlear implants • hearing aids • electroacoustic stimulation
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Background

Hearing is one of the most important of the human senses. Access to information and modern technology can be seriously limited or rendered impossible by a hearing impairment. Damaged audition can adversely impact the individual and society. A recent epidemiological study conducted in Poland found that one Pole in three has hearing problems [1]. Hearing impairment is viewed as a serious social problem. These observations emphasize the need for research to develop new and more effective methods for restoring audition.

At present, hearing loss is treated using prosthetic devices: either a hearing aid or a cochlear implant. Hearing aids are chiefly used for mild to severe hearing loss. In profound hearing loss and deafness, current treatment is based on cochlear implants [2].

Considerable improvement in cochlear implant technology has resulted in the broadening of selection criteria [3]. As more positive results of implantation are demonstrated, there is considerable emphasis on implanting individuals who are not only totally deaf, but also those with residual hearing in the low frequencies [4,5]. Moreover, recent studies have shown that residual hearing can be preserved after cochlear implant placement [6,7]. Further extension of selection criteria has been proposed by von Ilberg et al. [2], who 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. This concept is called electro-acoustic stimulation (EAS) [2].

There is another large group of patients, however, whose hearing impairment is characterized by normal or slightly elevated thresholds in the low-frequency band, with nearly total deafness in higher frequencies. We propose to describe this type of hearing impairment as partial deafness. The patients in this group remain beyond the scope of effective treatment by hearing aids only. Such patients have not been considered before for cochlear implantation, because it was feared that this intervention would damage the functioning part of the cochlea.

The main purpose of this report is to present a new method of partial deafness treatment applied in the case of a young woman.

CASE REPORT

The patient is a 25-year-old woman with prelingual onset of deafness of unknown etiology. Profound highfrequency hearing loss was diagnosed when she was 4. Multiple fittings of hearing aids did not improve her speech understanding. This resulted in numerous problems related to social communication, and made social and occupational activity challenging if not impossible. Problems that were not so crucial in childhood became significant during school and university education. Due to her normal age-related speech production abilities, she was not considered a hearing impaired person, but

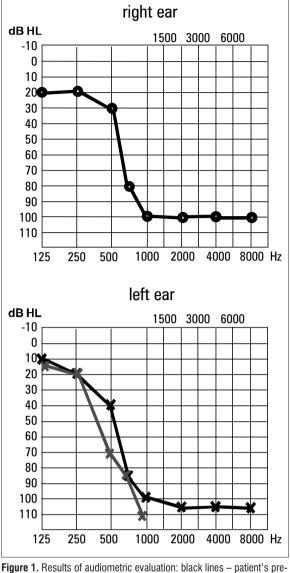


Figure 1. Results of audiometric evaluation: black lines – patient's preoperative audiogram; red line – patient's postoperative left ear audiogram

on the other hand her hearing was too limited to assure normal communication in everyday life.

Since the beginning of 2001 she has been a patient at the Institute of Physiology and Pathology of Hearing in Warsaw. Audiometric and auditory brainstem response (ABR) measurements were used to assess her hearing sensitivity. 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 130 dB HL, and standard clinical procedure was used for threshold determination [8]. Testing was performed in an IAC soundproofed booth under Sennheiser HDA 200 headphones.

The patient's preoperative audiogram is shown in Fig. 1. The mean Pure Tone Average (PTA) for 500, 1000 and

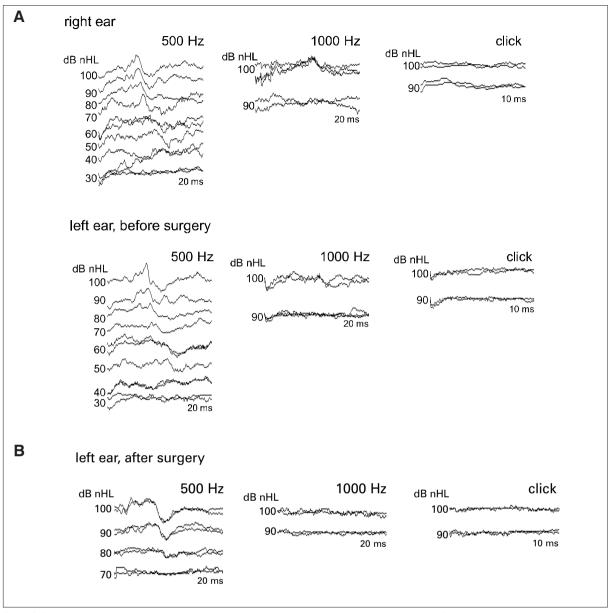


Figure 2. Results of auditory brainstem response evaluation: A) preoperative responses in the left and right ear; B) postoperative responses in the left ear.

2000 Hz was 77 dB for the right ear and 82 dB for the left. According to the classification of hearing impairment proposed by Goodman and Clark, each of these PTAs indicates severe hearing loss [9,10]. In terms of audiometric classification, the audiogram is precipitously sloping. This type of audiogram is also called a 'corner audiogram' or a 'steeply sloping' audiogram. Unfortunately, this is a common pattern of hearing loss.

Tests of speech comprehension were performed using the Pruszewicz monosyllabic word test in the Polish language (20 words per list, 20 lists). The lists for each test were randomized among various test conditions. The results given are the mean values for 3 lists. The word recognition scores obtained in the right and left ear separately were 20% and 25% respectively at 70 dB HL. In bilateral conditions, the score was 23% in quiet surroundings, and 0% in noise without hearing aid, vs. 30% in quiet surroundings and 0% in noise with optimal fitting of hearing aids on both sides. The patient did not tolerate the use of hearing aids in her daily life due to severe discomfort.

ABRs were recorded using electrodes at the forehead and the ipsilateral mastoid. The recording bandwidth was 200–2000 Hz (6 dB/octave slope). A computer-controlled system (Eptest) was used to record the ABRs. The time of response analysis was 20 ms. The ABRs were recorded for various amplitudes of 100 μ s clicks, presented at 37/s and in alternating polarity. Madsen insert earphones were used. The stimuli also included tone bursts with a Gaussian envelope. The frequencies of the bursts were 500 Hz and 1000 Hz. The ABRs obtained before surgery are shown in Fig. 2a.

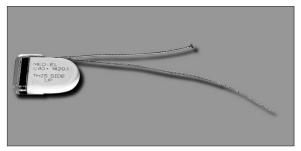


Figure 3. Med-El Combi 40+ implant with standard electrode.

In June 2002 the patient was qualified for cochlear implant surgery. The operation was performed on July 12. The patient's left ear was implanted with a Med-El Combi 40+ system, using the standard electrode array (Fig. 3). In order to avoid loss of low-frequency hearing, a partial electrode insertion was performed, with an approach to the scala tympani directly through the round-window membrane. The approximate depth of insertion was 20 mm. Eight of the 12 electric contacts were inserted. The electrode array was fixed in its final position using fibrin glue at the round-window niche. The round-window membrane was left partially uncovered to preserve its mobility. The device was fixed in a well made in the temporal bone during surgery.

In order to evaluate the preservation of acousticallystimulated hearing after cochlear implantation, audiometric and ABR measurements were performed in the implanted left ear using the same conditions as preoperatively. The patient's postoperative left ear audiogram is shown in Fig. 1. The audiometric thresholds measured at 125 Hz and 250 Hz were the same as those measured preoperatively. However, decrements in sensitivity were observed at 500 Hz and at 1000 Hz. In all, a 15 dB deterioration of the PTA was observed postoperatively. The ABR recordings obtained after the surgery are shown in Fig. 2b. Speech comprehension in quiet and in noise was tested at one week, one month, and three months after activation of the cochlear implant system. The speech items were presented via a loudspeaker from in front of the subject (0° azimuth), at a level of 70 dB SPL. The subject was seated 1m away from the speaker. Speech-weighted noise was presented from the same speaker at a speech-to-noise ratio of +10dB. Word recognition scores obtained with the cochlear implant plus acoustically-stimulated hearing (without amplification) are shown in Fig. 5.

Word recognition in quiet was also measured for the condition of implant activation only, at a 3-month interval. Direct input to the implant speech processor was used instead of the sound-field presentations described above, in order to eliminate any possibility of acoustic stimulation. The score for the implant-alone condition was 23% correct.

DISCUSSION

The patient presented here had a pattern of hearing loss that did not quite meet standard criteria for either a

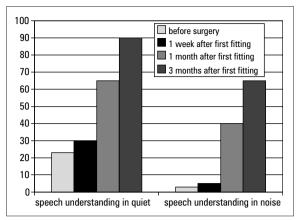


Figure 4. Results of monosyllabic speech understanding after cochlear implantation in quiet surroundings and in noise.

cochlear implant or combined EAS, as usually practiced. Her hearing at low frequencies was too good, and there was a concern that insertion of a cochlear implant would damage that low-frequency hearing [11].

However, the patient's low-frequency hearing was grossly insufficient for communication in everyday life. Repeated attempts with different fittings of hearing aids did not offer any significant help. She obtained a score of only 30% correct in the monosyllabic word test, using optimally-fitted aids. Amplification at high frequencies was only marginally useful.

Such results are consistent with those of many other studies. In particular, amplification at frequencies above the region of substantial residual hearing provides little or no benefit for people with steeply-sloping audiograms [12–15]. Moreover, attempts to transpose high frequencies into the region of residual low-frequency hearing have not been successful [16].

In contrast, cochlear implants can restore useful perception of high-frequency information [17]. This information is critical to speech reception. Furthermore, von Ilberg and coworkers [2] proved that the central auditory system is able to integrate inputs from electrical stimulation with a cochlear implant with those from acoustically-stimulated hearing.

Based on the encouraging results reported by von Ilberg et al, the decision was made to apply a cochlear implant in the present case of partial deafness. We assumed that the restoration of high-frequency perception with electrical stimulation would improve speech comprehension if the residual, low-frequency hearing could be preserved. The combination of electric plus acoustic stimulation would then provide a more complete representation of speech frequencies than would be possible with either modality alone.

The results obtained fully support this assumption. The low frequency hearing was preserved to the large extent, as proved by audiometric and ABR evaluation. Changes in audiometric thresholds, particularly above

1000 Hz, could be caused by the presence of the electrode in the cochlear. The sound field audiogram with the cochlear implant is shown in Fig. 4, indicating restoration of high frequency perception. After a short period following activation of the cochlear implant, a quite large and highly significant improvement in the recognition of monosyllabic words was observed. As shown in Fig. 5, scores for recognition of the words in quiet surroundings increased from 23% to 90%, and scores for recognition in noise increased from 0% to 65%, after 3 months of experience with the implant. This increase was the result of the combination of electric and acoustic stimulation, due to the fact that the score for the implant-alone condition was only 23%. Such high scores with this combination are most encouraging, as the monosyllabic word test is the most difficult of those given in standard audiological practice. Moreover, the scores are consistent with an almost complete restoration of communication abilities in everyday life. The patient now understands with ease most everything said to her, using her hearing alone. This is a remarkable outcome.

Results from the presented case support further applications of cochlear implants for people with steeplysloping audiograms and substantial low-frequency hearing. The low-frequency hearing can be preserved, and the combination of electric and acoustic stimulation can provide high levels of speech recognition.

CONCLUSIONS

The results obtained in the reported case can be considered to be the first step towards the application of a new method for the treatment of partial deafness. To implement the method, a 3-step procedure is proposed, with pre-, intra- and post-operative parts. The preoperative procedure includes clinical and audiological assessment to confirm fulfillment of qualification criteria, i.e, thresholds of 40 dB HL at 125, 250 and 500 Hz, and thresholds of 70 dB HL or higher at all higher audiometric frequencies. The subject should obtain minimal benefit only from the most-optimally fitted hearing amplification, with monosyllable scores in quiet of 55% correct or lower in both ears in the best-aided condition, at 70 dB SPL.

The proposed surgical procedure includes the following steps:

- antrotomy;
- posterior tympanotomy to allow visualization of the round window niche;
- tympanopunction in the inferior part of the round window membrane;
- approach to the scala tympani directly through the round window membrane;

- partial insertion of the electrode array;
- electrode fixation in the round window niche with fibrin glue (the membrane must be partially uncovered to preserve its mobility);
- fixation of the device in a well made in the temporal bone.

The postoperative part includes audiological assessment of preserved hearing sensitivity at low frequencies and cochlear implant system fitting, with the latter focused on selection of appropriate parameters of electrical stimulation.

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