Partial Deafness Treatment

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Continued improvement in the treatment of profound hearing impairment, particularly in very young children, encourages us to engage in new challenges and to implement new technological solutions. Our efforts have yielded a new generation of implants designed through the cooperation of scientists, bioengineers and implant manufacturers. Many specialists from different countries in Europe, Australia and the USA, are continually searching for novel improvements in implant efficacy in order to provide patients with the possibility of unconstrained communication with the environment (Lenarz et al., 2009; Baumgartner et al., 2007). We have witnessed extraordinary changes and rapid technological progress in the field of cochlear implantation such as improvements in speech coding strategies, surgical implantation procedures, implant fitting, novel electrode types and software enabling remote fitting of implants (Wąsowski et al., 2009).

The concept of preservation of the residual hearing

The development of implant technologies and growing expertise in surgery and rehabilitation have changed cochlear implant candidacy criteria. Increasingly younger children are undergoing implantation. Not only are individuals with a bilateral profound hearing loss receiving cochlear implants, but also those with considerable residual hearing. The implementation of a cochlear implantation program that preserves residual hearing is an ambitious challenge.

The combination of electrical stimulation through a cochlear implant with contra-lateral acoustic amplification of residual hearing provided by a hearing aid was initially described as ‘bimodal hearing’ (Dooley et al., 1993). Undoubtedly, one of the most important achievements was combination of electrical stimulation through a cochlear implant with the ipsilateral acoustic amplification by a hearing aid known either as ‘electro-acoustic stimulation’ (von Ilberg et al., 1999) or ‘hybrid stimulation’ (Gantz et al., 2004). Electro-acoustic stimulation was not only theoretically valid, but its benefits have been proven in numerous clinical studies.

Treatment of the partial deafness – results

The clinical team of the Institute of Physiology and Pathology of Hearing presented their results of significant residual hearing preservation and combining electrical stimulation in one ear with the acoustic stimulation in the other for the first time at the European Symposium on Paediatric Cochlear Implantation in Antwerp in 2000 (Skarzynski et al., 2000). In a study of 62 patients, who underwent cochlear implantation through cochleostomy, successful preservation of residual hearing was achieved in 77.3% of individuals . Continued study showed that in the group of 26 children and postlingual adults implanted through cochleostomy, loss of residual hearing after implantation occurred in 19% cases (Skarzynski et al., 2002). Those results suggested that the essential condition of successful preservation of residual hearing in cochlear implantation was to adopt the least invasive surgical technique possible, the round window approach.

Continually growing clinical material, including children with preserved residual hearing, has been presented by our team at international forums such as the ESPCI conferences in Spain, Switzerland and Italy, American conferences and the annual Hearing Preservation Workshops.

Steady development of our round window approach program permitted identification of a new group of 'partial deafness' (PD) patients, with normal low frequency hearing, but no hearing in the high frequencies. These individuals have a large population of spiral ganglion cells in the apex of the cochlea, representing normal tonotopy, and are successfully managed by cochlear implantation in a procedure known as the Partial Deafness Cochlear Implantation (PDCI), performed for the first time in 2002 in an adult patient with the partial deafness (Skarzynski et al., 2003).

Satisfactory preservation of residual hearing in 90% of adults implanted using the PDCI procedure provided justification for extending that method of treatment to children (Skarzynski et al. 2006). In 2004, the first cochlear implantation of a child with partial deafness was performed in our Institute (Skarzynski et al. 2006, 2007).

Our cochlear implant research group (Skarzynski, Lorens and Piotrowska, 2002, 2003a, 2003b; Skarzynski et al., 2006, 2007a, 2007b, 2009; Lorens et al. 2008) has been researching the benefits of hearing preservation, focusing initially on patients with steeply-sloping audiograms, whose hearing impairment is characterized by normal or slightly elevated

thresholds in the low-frequency band with nearly total deafness in higher frequencies. We have demonstrated that these patients achieved substantial improvement in speech discrimination and communication skills using a cochlear implant in the same ear with natural hearing without amplification. Other authors (von Ilberg et al., 1999; Kiefer et al., 2004; Gstoettner et al., 2006; James et al., 2006; Vermeire et al., 2008) reported on patients who, in most cases, had less residual hearing before the implantation and achieved benefit from Electric-Acoustic Stimulation (EAS): electrical stimulation of high frequency range with a cochlear implant (CI), combined with additional acoustic amplification in the form of a hearing aid (HA) for the preserved low-frequency range.

When we consider differences in pre-operative monosyllabic word test scores: 37% (Skarzynski et al., 2006) and 40% (Skarzynski et al., 2009) versus 7% (Kiefer et al., 2005) and 13.1 % (Gstoettner et al., 2006) it becomes clear that two different populations were implanted. In our population, the subjects had significantly better speech scores to begin with, reflecting better low frequency hearing before implantation. Moreover, if the modalities used by the patients in the implanted ear are taken into consideration (electric plus acoustic non amplified vs. electric plus acoustic amplified vs. electric) an additional small subgroup of patients may be identified. It includes individuals who lost residual hearing after implantation or whose remaining residual hearing in not sufficient to be amplified (termed 'non functional preservation'). Those patients rely solely on the CI and do not use amplification in the implanted ear. The term 'non-functional preservation' signifies postoperative thresholds for 125 Hz, 250 Hz and 500 Hz > 80 dB HL.

In sum, PDCI can lead to three approaches for three distinct groups of patients:

A. Electrical Complement (EC) in patients with normal or slightly elevated thresholds at low frequencies and with almost total deafness at higher frequencies. Non amplified low frequency hearing is complemented by electric stimulation with a cochlear implant.

B. Electric Acoustic Stimulation (EAS) in patients with mild to severe hearing loss in low frequencies and profound hearing loss in high frequencies. In the EAS group low frequency hearing is amplified and combined with electric stimulation in the same ear.

C. Electric Stimulation (ES) is used solely in the implanted ear in cases of loss of the low frequency hearing after implantation or non-functional hearing preservation.

The audiometric indication criteria for these three groups are shown in Figure 1

We have analyzed the audiometric and speech reception data of 95 subjects, 63 adults and 32 children, who were diagnosed with partial deafness and received either a MedEl Combi 40+ or a Pulsar cochlear implant. In all cases the 'round window' technique for hearing preservation was used, with the partial insertion of a 30 mm long standard (n=52) or Flex electrode (n=12), or full insertion of 20 mm M electrode (n=31). Patients selected for the analysis had at least one year of experience of using a cochlear implant.

The mean age at implantation was 32.58 years (ranging from 4.1 to 71.32 years). The patients were divided into the two subgroups, based on the preoperative audiograms: the EC-PRE group (59 individuals) and the EAS-PRE group (36 individuals), with the preposition that, if the full preservation of residual hearing was achieved after implantation, either the EAS or EC approach would be undertaken. Groups were assigned based on the audiometric criteria shown in Figure 1.

Pure tone audiometry data collected at 3 months before and after the surgery revealed that hearing preservation was achieved in 92 out of 95 (97%) subjects. The average hearing thresholds, measured before surgery and 3 months afterwards in 92 patients with preserved hearing are shown in Figure 2. Overall, for all audiometric frequencies, the hearing loss was not statistically significant ($p>0.05$). The differences in mean pre- and mean postoperative thresholds are presented in Table 1. They are consistent with what these authors had shown in the previous study, reporting the first ten cases (Skarzynski et al., 2007a), which validates our conclusion that the results of the 'round window' hearing preservation technique are repeatable.

Figure 3 shows the average hearing thresholds, measured before and after surgery in the two groups: EC-PRE and EAS-PRE. There are no significant differences in the hearing threshold changes between those groups.

The speech reception results, obtained preoperatively and 12 months after surgery were also analysed. The patients were examined using the Pruszewicz test - a consonant-vowel-consonant monosyllabic Polish word test (20 words per list, 20 lists). Pre-recorded words were presented in sound field at 60 dB SPL in quiet and in competition with speech-shaped noise at a speech-to-noise ratio (SNR) of +10 dB. Both words and noise were presented from the front. The subjects were tested using their natural bilateral acoustic hearing and

electrically stimulated hearing with the cochlear implant in one ear, or using the DUET Hearing System (Med-El Corporation, Innsbruck, Austria) and contralateral acoustic hearing. The DUET system includes a Tempo+ speech processor with precise Hilbert Transform envelope detection and a two-channel haring aid (HA) in one unit.

11 children out of 32 included in the current study could not be assessed using the standard monosyllabic test, because it was too difficult for them. For this reason, these 11 subjects were excluded from the speech reception evaluation, leaving 84 subjects with at least one year of experience of using the cochlear implant: 21 children and 63 adults. There is a significant increase in scores over time from pre-operative to 12 months after surgery. Monosyllabic word recognition increased from 34% to 73% under quiet conditions and from 7% to 54% under noisy conditions. These results are comparable to those achieved previously in the first group of ten adults (from 37% to 83% in quiet and from 10% to 60,5% in noise) (Skarzynski et al., 2006) and in the first group of nine children (from 30% to 69% in quiet and from 5% to 62% in noise) after PDCI (Skarzynski et al., 2007b). The data support our conclusion that the results of PDCI are highly reproducible.

The benefit of preserving residual hearing was demonstrated by Kiefer et al. (2005) and Gstoettner et al. (2004). The long-term evaluation of residual hearing has shown preservation and stability in about 75% of subjects (Gstoettner et al., 2006). That latter paper reported a monosyllabic word recognition score of 75% in the group of patients with complete hearing preservation. This is almost the same as the scores reported in our current study, although we included patients with partial preservation and with loss of hearing. Similar results were accomplished with another approach to acoustic-plus-electric speech processing using the application of a 10 mm Hybrid electrode (Gantz et al., 2006). In the group of Hybrid users hearing preservation was accomplished in 96%, compared to the 97% reported in this paper. Hybrid users who had more than 1 year of experience achieved an average score of 75 % correct monosyllabic words.

The mean scores and standard deviations of speech reception tests, performed preoperatively and postoperatively for the four groups of patients: EC-POST, EAS-POST, ES-EC and ES-EAS, are shown in Figure 4.

In the EC-POST group thresholds, measured after implantation, were ≤ 65 dB HL for 125 Hz, 250 Hz and 500 Hz (Figure 1a). In this group non-amplified low frequency hearing was

complemented by electric stimulation with a cochlear implant.

In the EAS-POST group thresholds, measured after implantation, were > 65 dB HL for 125 Hz, 250 Hz and 500 Hz . The patients in this group demonstrate functional hearing preservation with thresholds of ≤ 80 dB in the low frequencies (up to 500 Hz) (Figure 1b).

The ES-EC and ES-EAS groups consist of those patients from the EC-PRE and EAS-PRE groups who had non-functional preservation or lost hearing after surgery (Figure 1c). The average hearing thresholds, measured before surgery and 3 months afterwards in 19 patients with non-functional hearing preservation are shown in Figure 5. In the ES-EC and ES-EAS groups electric stimulation (ES) alone is used in the implanted ear.

In all four groups we observed a significant increase in scores between pre-operative and 12 months after surgery both under quiet and noisy conditions: EC-POST from 47% to 84% in quiet (p=0,000) and from 15% to 68% in noise (p=0,000); EAS-POST from 30% to 70% in quiet (p=0,000) and from 3% to 50% in noise (p=0,000); ES-EC from 39% to 75% in quiet (p=0,013) and from 6% to 52% in noise (p=0,000), ES-EAS from 14% to 68% in quiet (p=000) and from 2% to 50% in noise (p=0,001). Independent samples t-test with the Banferroni correction method revealed that both under quiet and noise condition the preoperative results were better in the EC-POST group than in the EAS-POST group by 17% (p=0,001) in quiet and by 12% (p=0,002) in noise, and the postoperative results were better by 15% (p=0,02) in quiet and 18% (p=0,002) in noise. Our data document that those individuals who have significantly better speech scores to begin with achieve significantly better scores after implantation.

No significant differences were observed between the EC-POST versus the ES-EC and the EAS-POST versus the ES-EAS groups. This finding reveals that there were no significant differences in post-operative scores between patients with functional preservation (group EC-POST and group EAS-POST) and patients with non functional preservation or with total loss of hearing (group ES-EC and ES-EAS).

Our results indicate that individuals with non-functional preservation and those who lost hearing have been able to obtain a significant advantage by using electric stimulation (ES) alone in one ear, and relying on low frequency hearing in the other ear (bimodal condition). This finding is consistent with results reported by Dorman et al. (2009), who did not find any significant differences in speech perception performance between EAS condition compared

with bimodal condition in patients implanted with the 10-mm Nucleus Hybrid electrode. Comparing speech perception scores before and after revision surgery in EAS cases, Helbig et al.(2009) also concluded that 20 mm insertion provides sufficient speech understanding, even in cases of loss of hearing or non-functional preservation.

These results indicate, on one hand, that, in this group of patients, there is no need for revision surgery to increase the electrode depth to 30 mm, but on the other, they cast doubt on the benefits of hearing preservation. However, we must treat this observation with caution, because in our tests both speech and noise were presented from the front, which limited the value of binaural cues, that would work to the advantage of patients with preserved low frequency hearing. Gifford et al. (2010) confirmed the value of hearing preservation in the study where sentence recognition in noise was assessed in a listening environment in which target and masker were spatially separated.

New approach to the treatment of patients with partial deafness

The first Polish cochlear implant program began in 1992. Based on the experience of over 2,600 adult and paediatric patients it was possible to consider hearing preservation from a new perspective.

The senior author found that present understanding of the term ‘partial deafness’ (PD) is different from the original definition, and the criteria for application of acoustic and electric amplification provided by the range of hearing aids, middle ear implants and cochlear implants may change and complement one another.

This new approach could reveal innovative possibilities for patients who obtain no benefit from hearing aids but do not qualify for cochlear implantation. Using the algorithm shown in Figure 6 we can realistically discuss the application of latest technologies in the patient with ‘partial deafness’.

Summary

The experience of the clinical team at the Institute of Physiology and Pathology of Hearing has lead to a turning point in the treatment of the various types of PD:

a) Ten years of management of PD adults and children, with varying levels of preservation of residual hearing, using combined stimulation (EAS)

b) Seven years of follow-up of PD adults who retained 93.2% of good low frequency hearing after implantation, complemented electrically (EC).

c) Nearly five years of follow-up of PD children, who retained 100% of good low frequency hearing after implantation, complemented electrically (EC).

d) Six years' experience with acoustic stimulation (AS) using the Vibrant Soundbridge middle ear implant, with temporary use of conventional hearing aids before the decision to implant was made.

e) Seventeen years of experience using the round window (RW) approach, gained during the initial stages of the Warsaw cochlear implant program. In the past ten years, the RW approach has given new meaning to the term 'residual hearing preservation' and set the grounds for successful treatment of the partial deafness (PDT).

f) Ten years experience demonstrating the feasibility of complete or partial residual hearing preservation with 20mm insertion of electrodes such as the Med-El Standard, Medium and Flex or the Nucleus SRA.

Conclusions

Treatment of partial deafness has allowed new directions to be set in the development of middle ear and cochlear implant programs for children and adults. Implementation of the partial deafness treatment (PDT) was connected with the development and implementation of novel diagnostic methods, hearing screening programs, batteries of audiological tests and psychoacoustic methods. Diagnostic imaging is very important in the determination of the type of hearing impairment in order to substantiate these extended indications for cochlear implantation.

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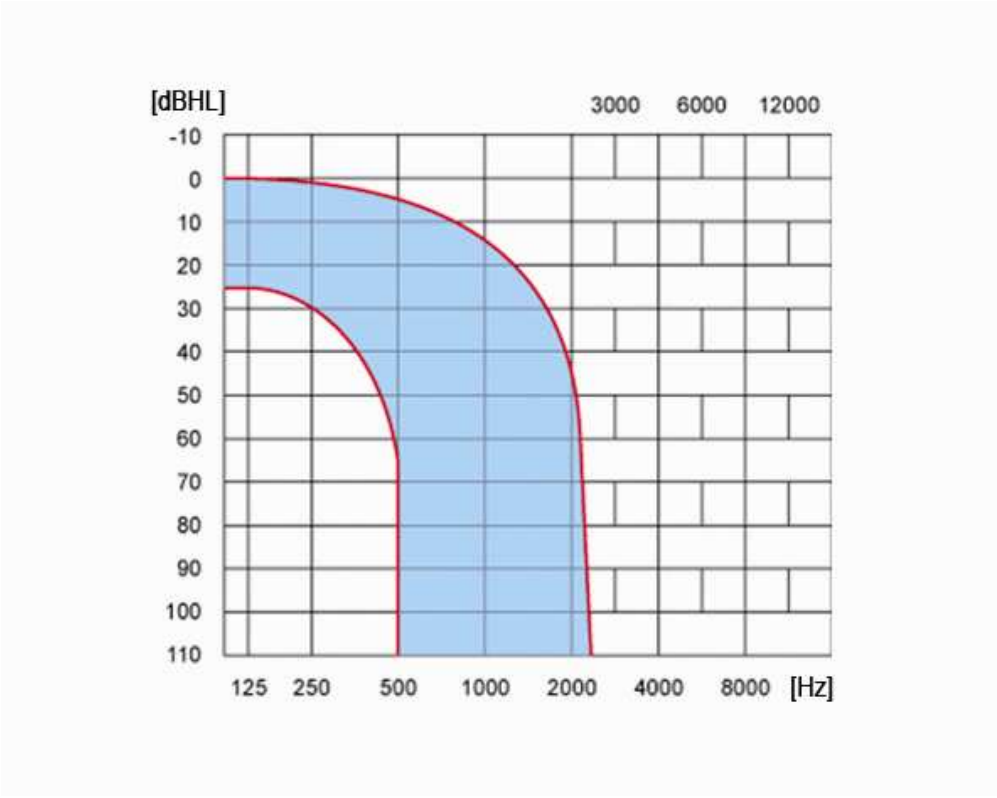
Table. 1 The differences between thresholds preoperatively and 12 months postoperatively in the previous study and in the current study

	125Hz	250Hz	500Hz	1000Hz	2000Hz	4000Hz
Preop-postop threshold differences (Skarzynski 2007) [dB]	7,8	16,3	26,1	11,6	0,3	5,7
Preop-postop threshold differences in the present group [dB]	13,9	19,5	21,4	12,5	2,9	3,2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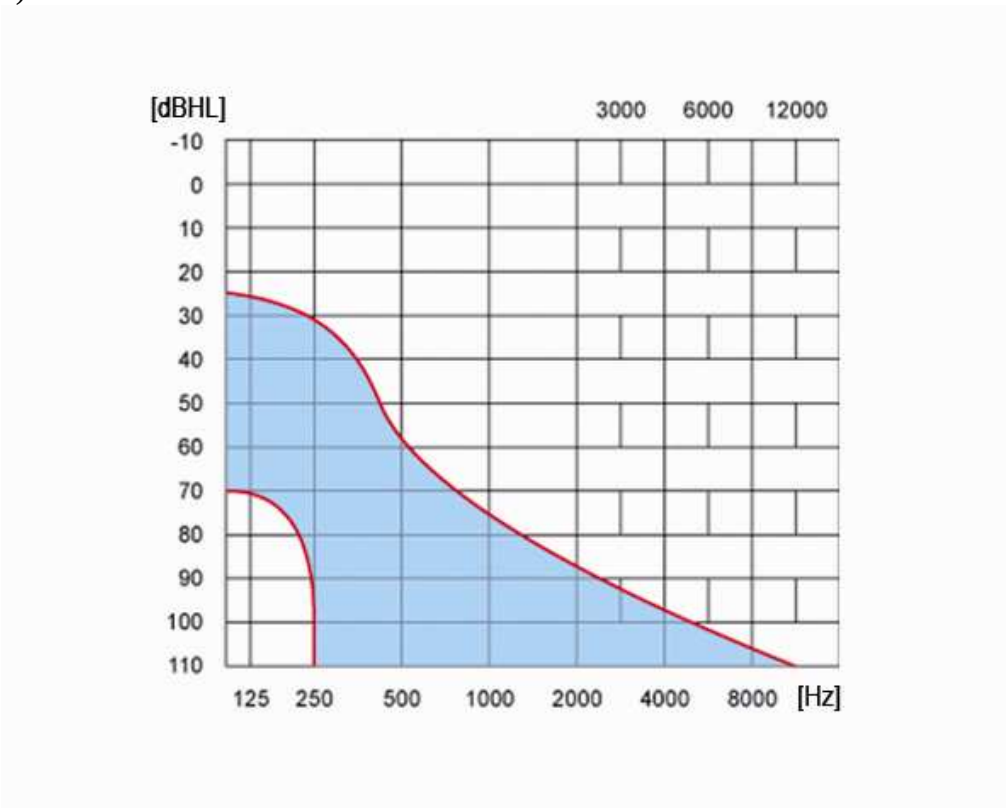
a)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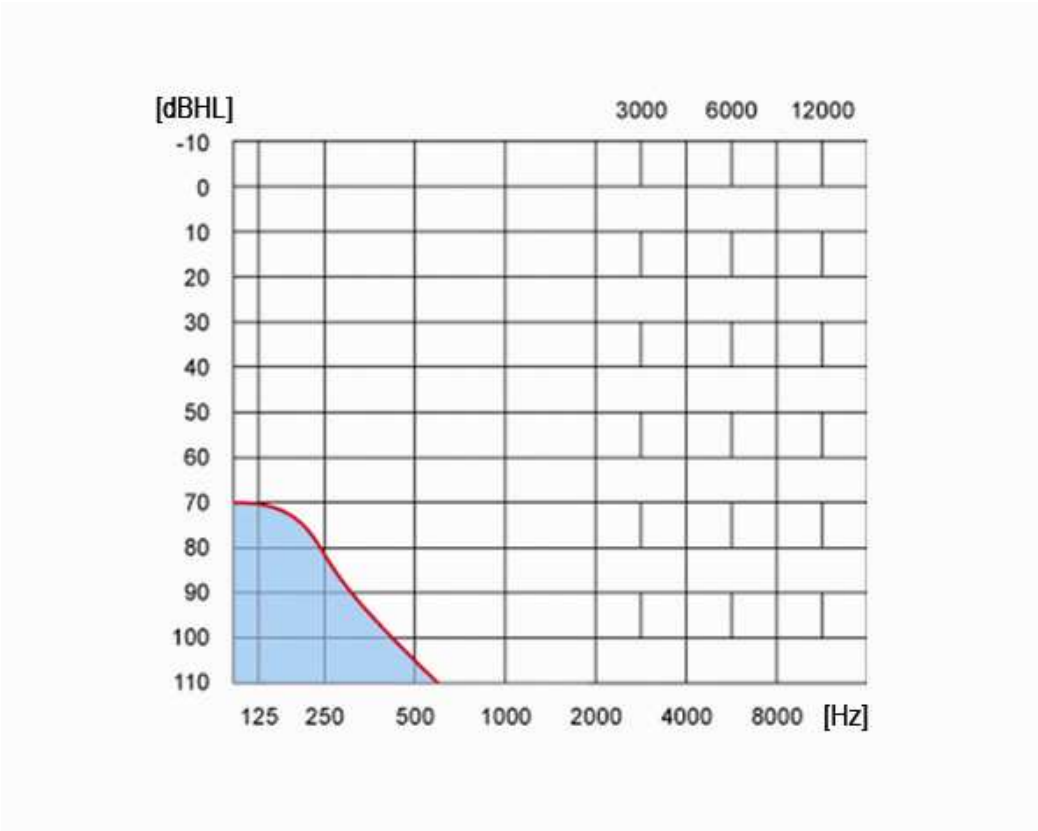
b)



c)

Fig. 1

Indication areas for: EC- a), EAS -b) and ES-c)

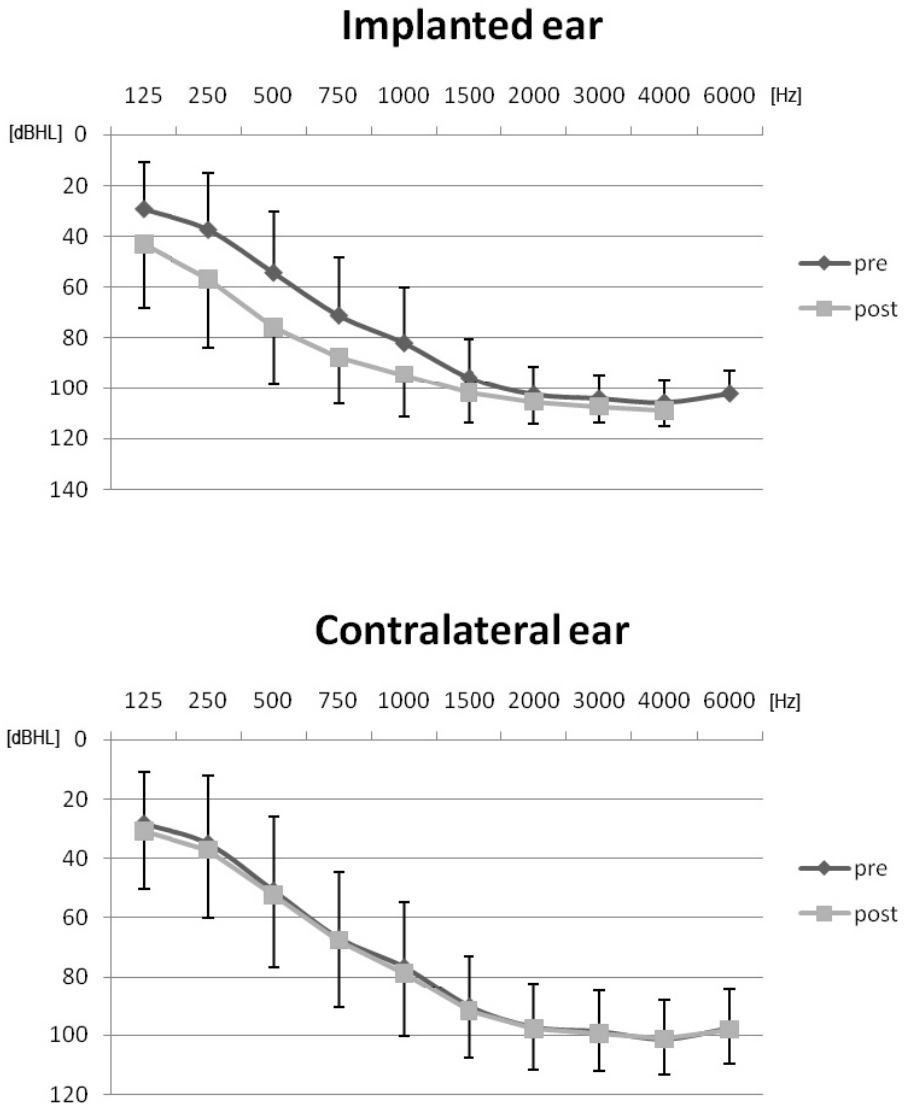
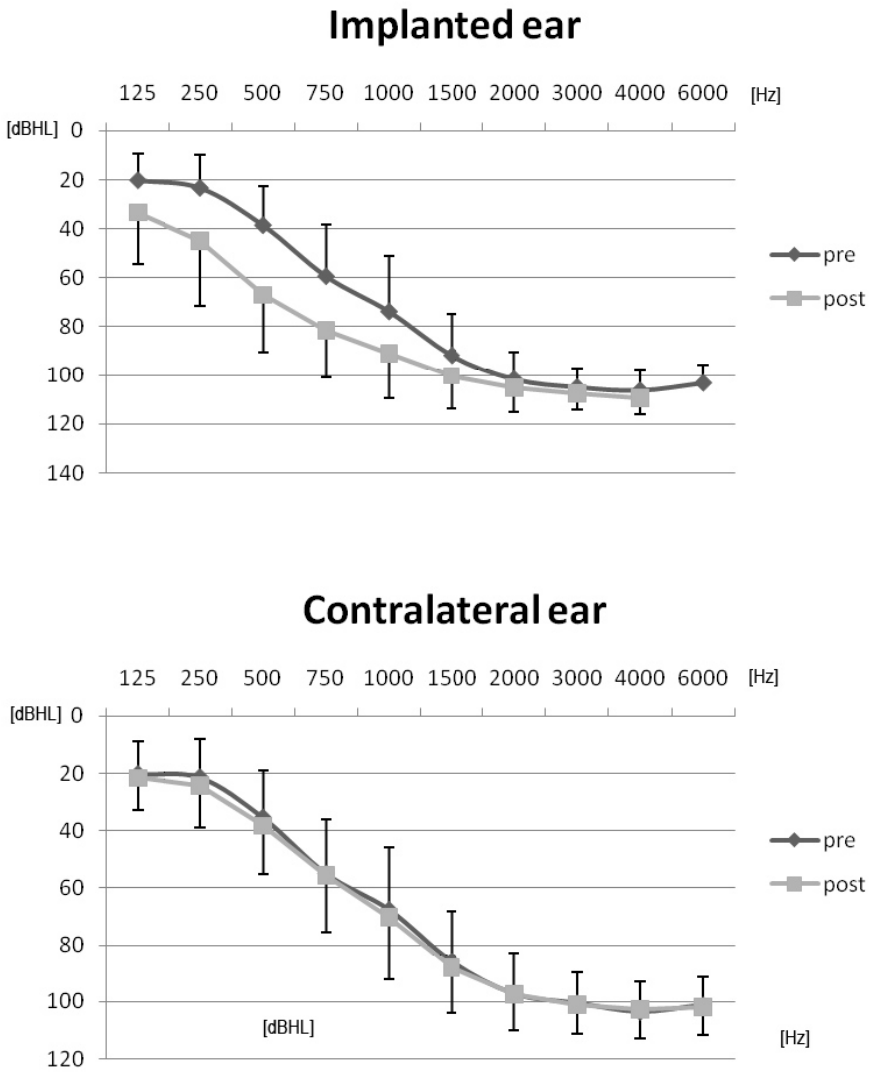


Fig. 2 Pre-operative and post-operative audiograms showing the mean and standard deviation for each frequency in 92 patients with preserved hearing


Implanted ear
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Contralateral ear
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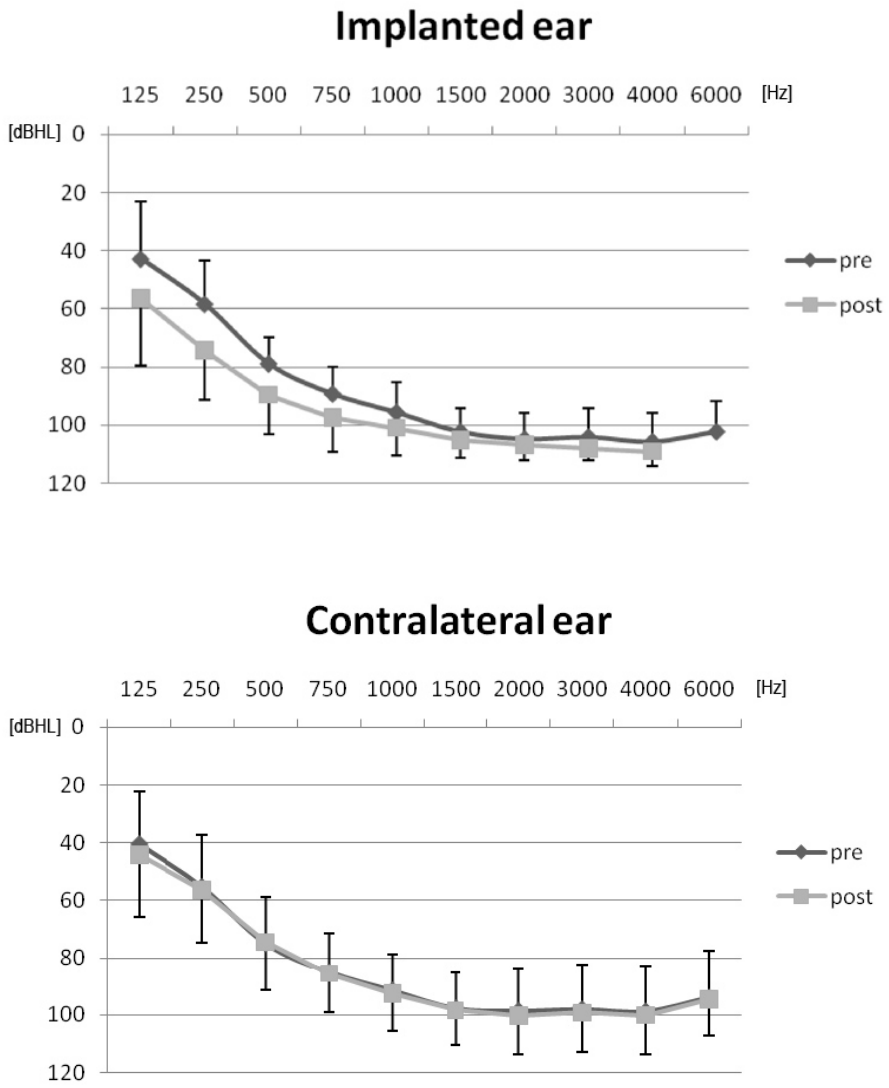
a)



b)

Fig. 3 Pre-operative and post-operative audiograms showing the mean and standard deviation for each frequency for two groups of patients: EC-PRE-a) and EAS-PRE-b)

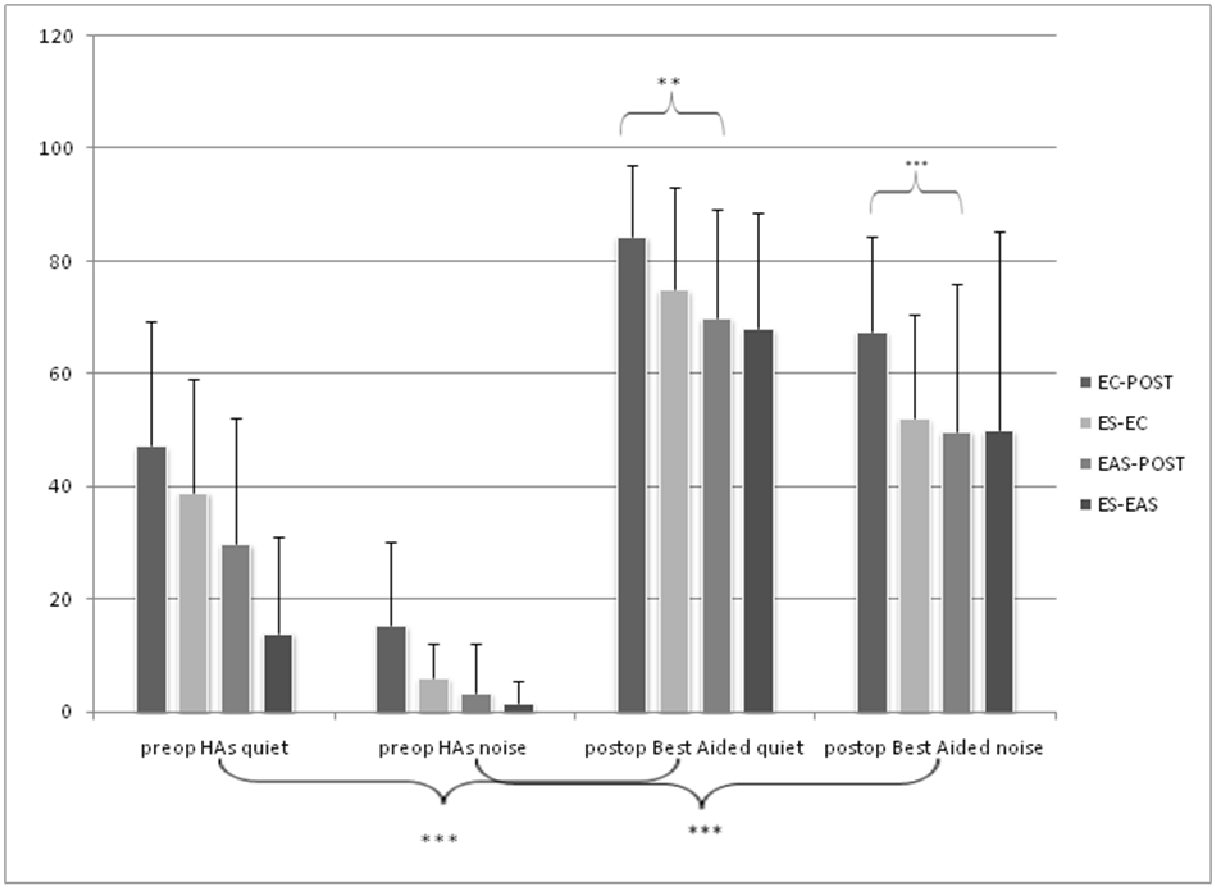


Fig 4 Preoperative and postoperative monosyllable scores in quiet and noise in the EC-POST, EAS-POST, ES-EC and ES-EAS groups

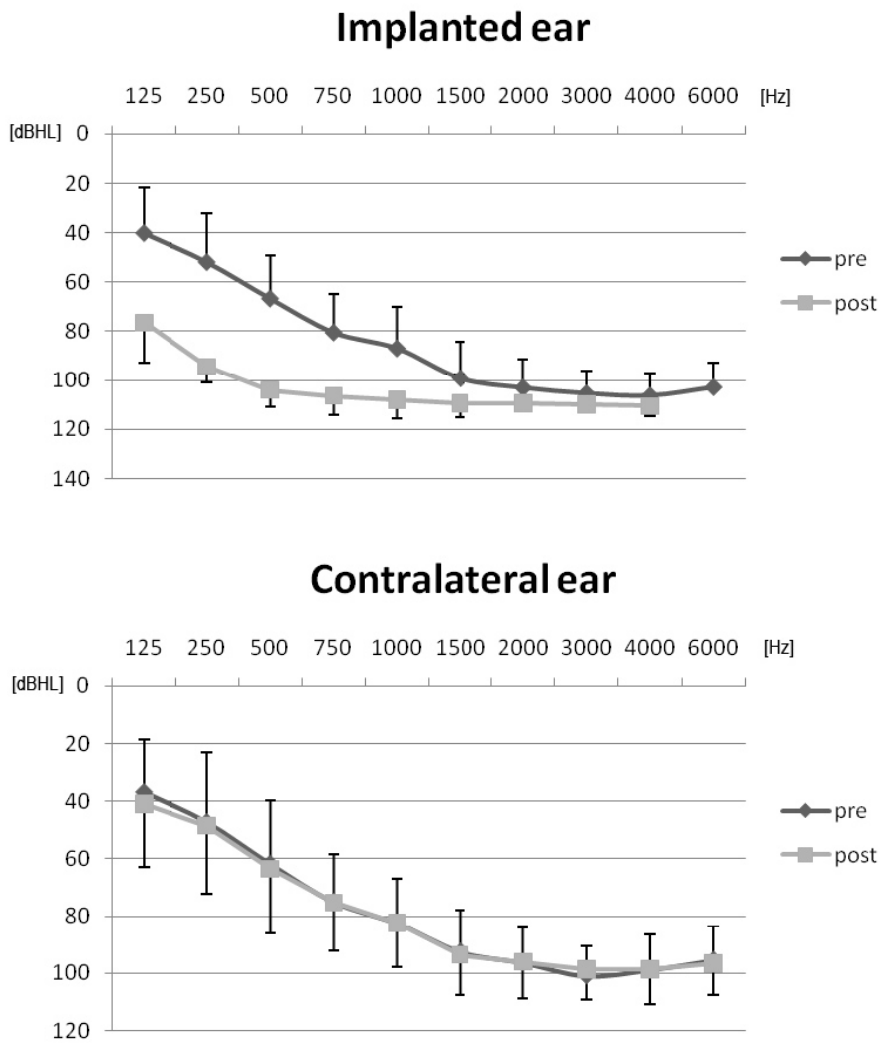


Fig. 5 Pre-operative and post-operative audiograms showing the mean and standard deviation for each frequency in 1 9 patients with non functional hearing preservation

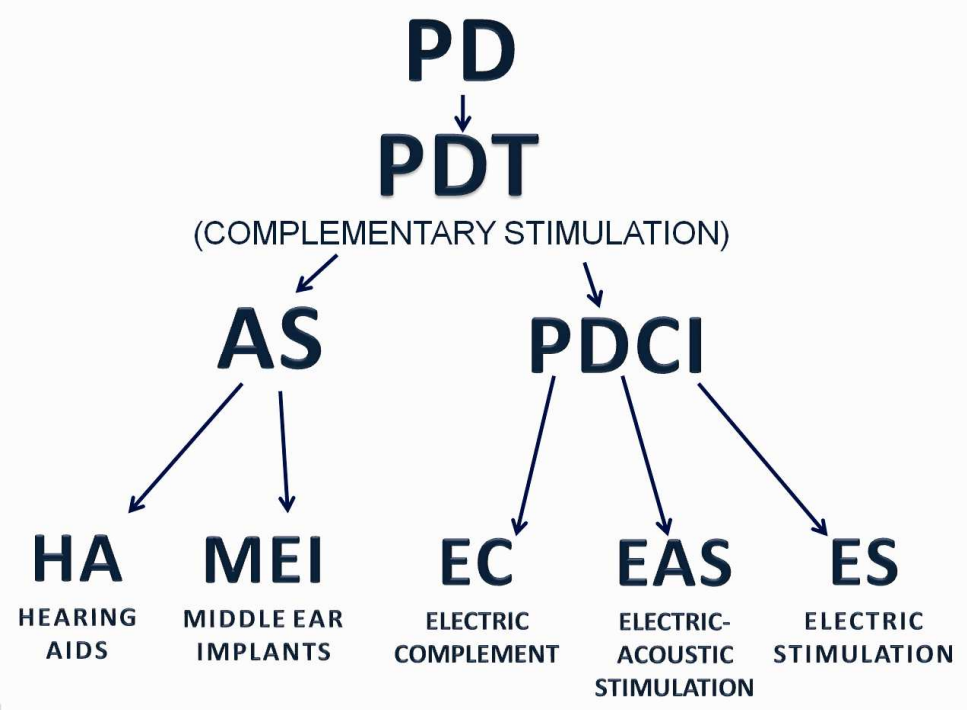


Fig.6 Algorithm of treatment of patients with the partial deafness:
AS – Acoustic stimulation – in borderline patients
PDCI – Partial Deafness Cochlear Implantation with electric stimulation
EC – Electric complement – to complement existing low-frequency hearing
EAS – Electro-acoustic stimulation – electric stimulation coupled with acoustic amplification
ES – Electric stimulation – purely electric stimulation in case of loss of the residual hearing, without changing the implant