International Journal of Pediatric Otorhinolaryngology (2007) **71**, 1407—1413

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International Journal of Pediatric Otorhinolaryngology

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Partial deafness cochlear implantation in children

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Received 2 February 2007; received in revised form 15 May 2007; accepted 15 May 2007

KEYWORDS
Partial deafness cochlear implantation;
Electric-acoustic stimulation;
Children;
Paediatric;
Outcomes;
Hearing preservation;
DUET

Summary

Objective: Partial deafness cochlear implantation and electric-acoustic stimulation have proven to be a useful method of treating adults with a ski-slope type hearing loss. Good hearing preservation and speech perception outcomes have been reported. This study aims to assess partial deafness cochlear implantation in children.
Method: Nine children, ranging in age from 4.2 to 12 years, received a cochlear implant following the round window surgical technique for partial deafness cochlear implantation. Hearing preservation was assessed by pure-tone audiometry and speech perception outcomes were measured using monosyllable word tests in quiet and noise. Data are available for most children up to a period of 1 year.
Results: Hearing could be preserved partially in all cases, however, one child does not have sufficient preservation to make use of electric-acoustic stimulation. The eight children with sufficiently preserved hearing either use the natural low frequency hearing in combination with a cochlear implant to hear or use the DUET combined hearing system. Speech perception tests showed improvement in quiet and noise over time.
Conclusion: Results suggest that partial deafness cochlear implantation is a viable treatment method in children. However, surgery should only be conducted by an experienced surgeon and parents need to be carefully counselled about the risks and benefits of partial deafness cochlear implantation.

1. Introduction

Preservation of hearing is considered a "hot" topic in the field of cochlear implantation. Hearing preservation allows the potential to use remaining hearing for either natural or acoustic amplification of the low frequency sounds. This would provide for more natural sound perception and reception of the consonants in speech. Hearing preservation may also allow for better music perception. Hearing preservation is also promising for future development of electrodes and drug delivery systems.

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doi:10.1016/j.ijporl.2007.05.014

Studies have shown that it is possible, in some cases, to preserve residual hearing after cochlear implantation with a deep insertion electrode array (31 mm, MED-EL COMBI 40+), in cases who have a corner audiogram [1,2]. Success has also been reported in subjects with some residual hearing who received a shorter electrode array (17 mm, Nucleus 24 Contour Advance) [3].

Following on from this, careful consideration has been paid to preservation of hearing in individuals with significant low frequency hearing, i.e. mild-to-moderate hearing loss in the low frequencies and severe-to-profound hearing loss in the high frequencies. The concept of electric-acoustic stimulation was first presented in 1999 [4], where a subject was implanted with a limited insertion depth to preserve low frequency hearing. This would allow for electrical stimulation of the high frequencies via the cochlear implant and acoustic stimulation of the low frequencies via a hearing aid. Since then, hearing preservation rates have been reported in about 80% of cases, as well as significant improvements in speech perception scores, particularly in noise [5—9].

The International Centre of Hearing and Speech published data on their first case of Partial Deafness Cochlear Implantation (PDCI) in 2003 [10]. In the case of PDCI, there is essentially normal hearing in the low frequencies, and often this is not amplified with a hearing aid, but the EAS principle uses natural low frequency hearing. Positive hearing preservation and speech results have been reported for PDCI [11,12].

The question arises, what would the possibilities of PDCI in children be? In a previous study [1] on preservation of residual hearing in traditional candidates, 7 of 26 subjects were children. Corner audiograms were able to be preserved in 16 (62%) cases. A further review of residual hearing preservation in 30 children showed preservation for 22 cases (73.3%) (unpublished data). Given the success of preserving hearing in traditional CI users and the outstanding success of PDCI in adults, our centre implanted its first child for PDCI in 2004.

This paper will demonstrate outcomes for nine children who have undergone the PDCI procedure. Fitting of the DUET™, a behind-the-ear processor combining a hearing aid and a cochlear implant in one device, will be discussed.

2. Method

2.1. Subjects

Nine children with partial deafness were implanted using the round window technique to increase the chance of preserving hearing [11]. Four girls and five boys have been implanted using the PDCI technique. The mean age at implantation was 9.07 years (range 4.2—12.1 years). The reported aetiologies were as follows: unknown (6), hypoxia (2) and ototoxicity over a period of 3 years (1). An attempt was made to ensure that any subjects with a progressive hearing loss were excluded from PDCI. Progressive hearing loss is defined as a 10 dB shift at two consecutive frequencies or a 15 dB shift at one frequency over a period of 1 year. The cases of unknown aetiology are probably congenital — this would reflect the aetiology in our adult population — most of whom believe that their ski-slope loss was present from birth. All subjects are in mainstream education, except the youngest child, who is in kindergarten. Four subjects were fitted with the DUET™ combined speech processor during the assessment period as they had some degree of hearing loss in the low frequencies that would benefit from acoustic amplification. One subject lost most residual hearing and he uses a cochlear implant only; and four subjects use a cochlear implant with limited frequency range

Table 1 Description of each subject including age at implantation, cochlear implant type (C40+ = COMBI 40+ Standard Electrode array, C40+ M = COMBI 40+ Medium Electrode array, PULSAR M = $\text{PULSAR}_{\text{CI}}^{100}$ Medium Electrode array), side implanted, device fitted (TEMPO+ and normal low frequency hearing, cochlear implant only, DUET speech processor), and aetiology

Subject	Age at CI	Cochlear implant	Side	Device fitted	Aetiology
EG	9.2	C40+	R	DUET at 10 years 9 months	Unknown
MK	10.11	C40+ M	L	DUET at 11 years 1 month	Unknown
SP	11.9	C40+ M	L	CI-only	Unknown
WM	6.1	PULSAR M	R	DUET at 6 years 5 months	Hypoxia
AB	4.2	PULSAR M	R	TEMPO+ and normal low frequency hearing	Unknown
KC	8.7	PULSAR M	R	TEMPO+ and normal low frequency hearing	Unknown
AA	10.2	PULSAR M	R	DUET at 11 years 1 month	Ototoxicity
DA	9.2	PULSAR M	R	TEMPO+ and normal low frequency hearing	Unknown
PS	12.1	PULSAR M	L	TEMPO+ and normal low frequency hearing	Hypoxia

and unaided low frequency hearing in the ipsilateral ear. The subjects' families were aware of the benefits and risks posed by PDCI prior to implantation. Table 1 details subject demographics.

2.2. Surgical considerations for children

Special care must be taken to achieve good access to the round window area. The electrode should be inserted using a posterior tympanotomy and entry to the round window should be at a right angle to the surface of the round window membrane. This approach is designed to avoid insertion into the scala tympani wall. The electrode itself seals the insertion puncture. It is often difficult to insert the electrode in children because of the short distance between the facial nerve canal and the annulus fibrocartilagineus of the tympanic membrane. For this reason, an anterior tympanotomy is performed more frequently in children than in adults, to improve visualization of the round window niche. Additionally, it is necessary to close the mastoid with spongostan, fibrin glue and a piece of bone taken during mastoidectomy. This prevents air flow from the middle ear to the implant area under the skin.

2.3. Devices used

One subject was implanted with a MED-EL COMBI 40+ standard electrode array which was partially inserted to a depth of 20 mm. Insertion depth was based on the subject's pre-operative audiogram and was estimated according to tonotopical organization of the cochlea. Two subjects were implanted with the MED-EL COMBI 40+ Medium electrode array and six subjects were implanted with the PULSAR$_{CI}{}^{100}$ Medium electrode array. All subjects were implanted in the worse ear, as determined both by audiogram and subjective reports.

In some cases, children were fitted with the DUET^{TM} Hearing system. This device was released in November 2005 and allows for electric stimulation via a speech processor system and amplification of low frequency hearing via a hearing aid component, all within the same device. This is more appropriate than fitting with a cochlear implant and in-the-ear hearing aid in the ipsilateral ear, in cases were there is low frequency hearing that would benefit from amplification.

2.4. Programming

The cochlear implant is programmed in such a way that the there is no overlap with acoustic perception, so as to not interfere with this perception. This means that the low frequency cut-off point is somewhere between 500 and 1000 Hz. This frequency cut-off point is determined by the audiogram. The low frequencies are then heard using the preserved natural low frequency hearing. In cases where the DUET^{TM} was fitted, the fitting of the cochlear implant component was conducted as described above. The hearing aid component was fitted according to the required calculated gain, using the half-gain rule (where the threshold of a given frequency is divided in half to obtain the predicted gain).

2.5. Audiological testing

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. Testing was performed in IAC soundproofed booth under Sennheiser HDA 200 headphones. A standard clinical procedure was used for threshold determination [13].

2.6. Speech perception testing

Subjects were tested using their natural bilateral acoustic hearing and electrically stimulated hearing via the cochlear implant in one ear or with a DUET speech processor, if they had been fitted with this. Tests of speech comprehension were performed using the Pruszewicz monosyllabic Polish word test (20 words per list, 20 lists). The test lists were randomised across test condition, and the results shown are a mean value of three test lists. Tests were also conducted in noise at a signal-to-noise ratio of +10 dB. Speech tests were conducted pre-operatively, then at first fitting, and then at 1, 3, 6 and 12 months after the first fitting.

2.7. Statistical analysis

Demographic data were analysed using descriptive statistics. Quantitative data are expressed as mean, median, standard deviation, quartiles and range (minimum and maximum); qualitative data are presented as absolute and relative frequencies. Analyses of variance for repeated measurements (general linear model) with time as a factor were performed for each test condition. If data were not nearly normally distributed, a Friedman test was used. With Kolmogorov—Smirnov-tests, data were checked for their distribution. Statistical significance was defined as $p < 0.05$. To detect differences between the test intervals (difference from the pre-operative to the first fitting assessment, first fitting to 3 months after first fitting assessment difference, 3—6 months difference, and 6—12 months difference), parametric paired Student's *t*-tests or nonparametric Wilcoxon signed ranks tests were used, depending on the data distribution. After adjustments for multiple comparisons with the use of Bonferroni's procedure *p*-values of less than 0017 were considered to indicate statistical significance. SPSS for Windows 12.0 software (Chicago, IL) was used for all analyses.

Missing data were not imputed for children with missing data. Statistical testing was performed with complete data only. If patients have had single missing values, "single mean imputation" was used for missing values at the pre-operative assessment and "last observation carried forward" was used for missing values at the other intervals.

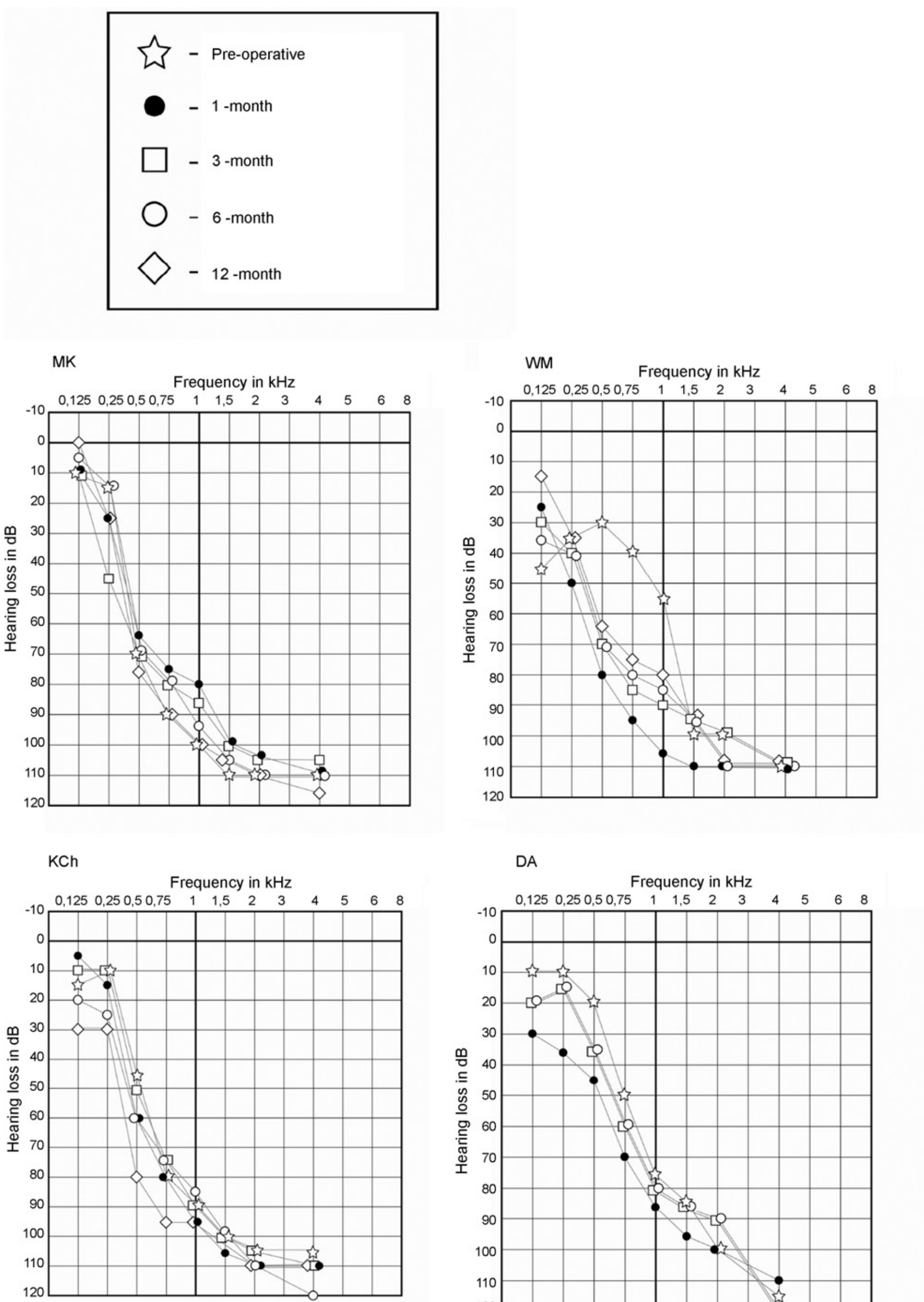

Fig. 1 Individual audiograms, showing pre-operative audiogram and post-operative audiograms for the following test intervals: 1, 3, 6 and 12 months.

3. Results

3.1. Hearing preservation

All surgeries went without incident and no post-operative problems were noted. Individual audiograms showing results over time can be seen in Fig. 1. Four subjects show full preservation of hearing. Five subjects show partial preservation, however one subject (SP) could be considered non-functional partial preservation, as the low frequency hearing cannot be amplified sufficiently with the hearing aid component of the DUET in the implanted ear to provide a useful result. However, he could combine the electrical stimulation with available acoustic hearing in the contralateral ear. This indicates that 8/9 subjects can be classified as PDCI users. Overall, hearing loss across all subjects was not statistically significant over time for 125, 250 and 500 Hz ($p > 0.05$).

Subject EG preserved hearing over 12 months and subject KC has preserved hearing over 6 months. Subject AA lost some hearing initially, and then remained stable up to 12 months, a similar pattern was seen for subject DA, and then a stable audiogram has been recorded up to 6 months. MK showed a drop at 250 Hz at 3 months, which resolved at 6 months and may indicate a middle-ear component. Similarly, WM had a large drop in hearing at 250 Hz which resolved at 3 months. Hearing thresholds at 500, 250 and 1000 Hz increased at 1 month for this subject as well, but have remained stable since. Subject SP appears to have a gradually progressive loss over time, which is not seen in the contralateral ear, which has remained stable over time. Finally, subject AB has had a drop in hearing at 3 months and again at 6 months. Interestingly, this drop was also noted in the contralateral ear at 6 months as well (an average drop of 50 dB), and has remained the same for both ears at 12 months. This indicates that some other element is involved in this case, beyond the introduction of a device into the cochlea.

3.2. Speech perception

Three subjects could not be assessed on the standard monosyllable test. Subject AB was too young to participate in formal testing. However, on the MTP test (a closed-set test) of the EARS test battery, designed for young children he scored 20/24 at 12 months. His parents report that he understands most things and that his speech production and language development have shown significant improvement since the implant and over time. Subject WM could not perform the tests due to her age; she has poor pronunciation and could not be scored accurately on the tests. Her parents report a large change in her behaviour, it is easier for her to understand people; she is more sociable and requires less listening effort. She also has better understanding when watching television. Finally, KC did not participate in speech testing as she has severe depression and other psychiatric issues. All three of these children are full-time users of PDCI.

Individual data for monosyllable testing in quiet can be seen in Fig. 2. Children performed better with EAS compared to the pre-operative acoustic hearing aid condition over time. There were no significant differences between individual test intervals. Individual data for monosyllable testing in noise can be seen in Fig. 3. Four subjects were tested for five intervals. These subjects included EG, AA, SP and MK. Their scores were evaluated by the separate repeated measures ANOVA. This ANOVA was highly significant (d.f. = 4, $F = 7.033$; $p = 0.004$) and post hoc comparison using the Holm—Sidak method showed that the 1-year scores were significantly higher than AS pre and first fitting scores, and that the 6-month scores were significantly higher than AS pre scores as well. The results are shown in Fig. 2b.

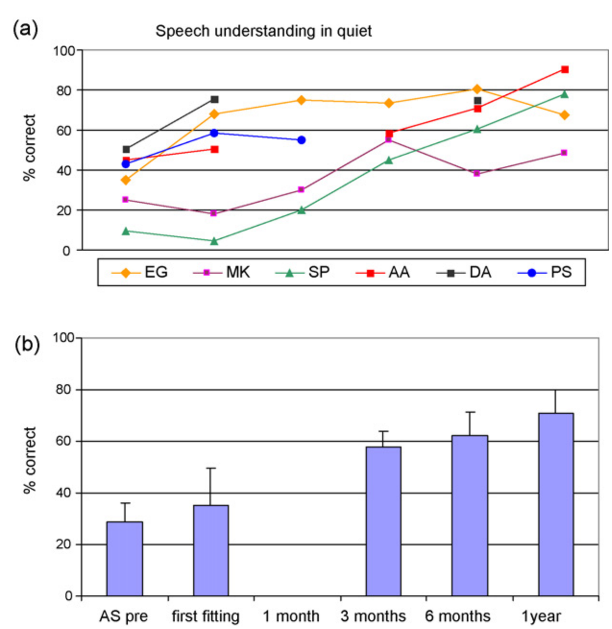

Fig. 2 Monosyllable scores in quiet showing individual data for each child, at each interval tested. Fig. 2a shows the raw data for each subject, demonstrating progress over time for each, up to their most recent test interval. Fig. 2b shows the mean data, with standard deviation for the four children (subjects EG, AA, SP and MK) who have reached the 1-year test interval to demonstrate the benefit of PDCI over time.

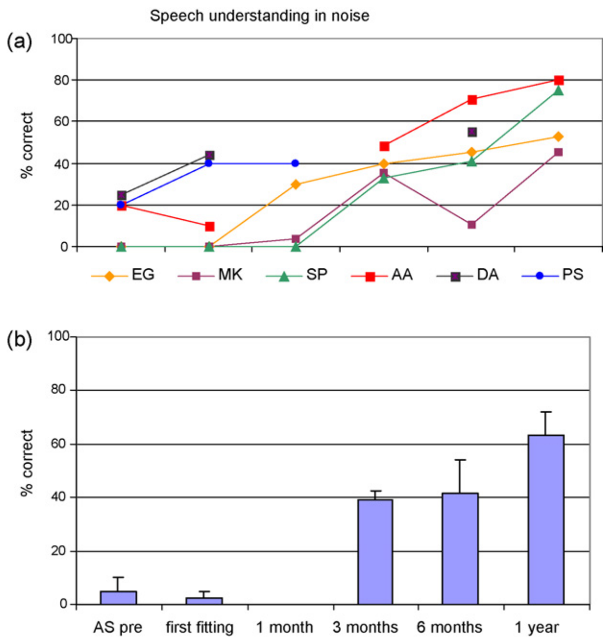

Fig. 3 Monosyllable scores in noise (+10 dB SNR) showing individual data for each child, at each interval tested. Fig. 3a shows the raw data for each subject, demonstrating progress over time for each, up to their most recent test interval. Fig. 3b shows the mean data, with standard deviation for the four children (subjects EG, AA, SP and MK) who have reached the 1-year test interval to demsontarte the benefit of PDCI over time.

3.3. Fitting the DUET™

Four children have been fit with the DUET™ Hearing System. Two children were fitted after 1 year of device experience, one child was fit at 1 month and one child was fit at first fitting. The recommended fitting procedures were followed and no special considerations had to be made for fitting the children. The children readily accepted the combination of electric and acoustic information. These results suggest that there are no special issues when fitting children with the DUET™.

4. Discussion

The results demonstrate that eight of nine subjects are able to utilise electric-acoustic hearing either via their natural low frequency hearing in their implanted ear or via the hearing aid component of their DUET™ hearing system. The results also demonstrate that hearing can be preserved or partially preserved in all cases. All six children tested showed improvement over time for speech perception skills, particularly in noise.

We consider our hearing preservation results to be particularly good. We have total preservation of 44.5% and partial preservation of hearing of 55.5%. However, although it is important to be able to demonstrate hearing preservation, one needs to consider whether this preservation is functional or not. This means, can the individual be fit both electrically and acoustically in the same ear, and use their preserved natural low frequency hearing as a complement to their electrical acoustic stimulation. In our case, we can demonstrate that 8/9 children had functional preservation—a rate of 88%. As there are no studies showing preservation of low frequency hearing in EAS or PDCI children, we need to compare our data to those studies of EAS and PDCI in adults.

Our results compare rather favourably to the reported results for adults. In studies using a shorter electrode array (6 and 10 mm) 6 subjects showed preserved hearing [8]; in a larger sample size from the same group 11 of 11 preserved hearing within 10—15 dB over the short term [9]. A follow-up of this sample [14] demonstrated an average loss of 9 dB for 125—1000 Hz, with one subject losing hearing at 3 months. It is important to note that these all had a significantly shorter depth of insertion when compared to our study. Also of interest is that there is some later hearing loss, which we also see in our study. This suggests some other element affecting stability of hearing, beyond the electrode and the surgical procedure.

Variable preservation also was reported in a study using a different electrode and different surgical technique, but a similar insertion depth. This study [3] reported preservation of 33, 26 and 19% for 125, 250 and 500 Hz in 27 subjects. When the surgeons followed the ''soft surgery technique'' ($n = 12$) preservation improved to 50, 50 and 33% within 20 dB at 125, 500 and 1000 Hz, respectively. Studies using the same electrode as we have used also demonstrate better hearing preservation results of 85.7% [7]. Partial preservation was maintained for over 3 months (12/18 subjects, 86%) and six subjects with 1-year data showed stable audiograms [5]. In a further elaboration of this study [6], 8/15 subjects had preserved hearing within 0—10 dB, 3/13 within 11—20 dB, and 2 subjects lost hearing immediately. These results suggest complete preservation in 53% of the subjects and partial preservation in 23%—a total preservation of 84%. This is similar to the preservation rate of 90% in a publication on adult PDCI patients [11]. Essentially, hearing preservation in children is comparable to that of adults, suggesting PDCI is viable in children. However, it should be noted that the surgeon (first author) in our clinic had vast experience with PDCI surgeries in adults (currently 20 adults) before attempting the procedure in children. He also had considerable experience in preserving corner audiograms in conventional cochlear implant candidates with some residual hearing [1]. The authors strongly suggest that such surgery should not be attempted in children without significant successful experience in adults.

As with adults, the speech outcomes for our children also show improvement over time, both in quiet and in noise. An average of 69% for monosyllables at 6 months and a 9 dB improvement of speech in noise have been reported for adults [14]. This synergistic effect in noise was also reported by others [5,7]. The results for children also mirror the improvement over time seen in our adult PDCI population [12]. In another study at another centre in 2005 [6], varying use of EAS was noted: seven subjects performed better with EAS compared to CI-only, four scored the same, and two could not use EAS. This is in contrast to our data, where all subjects except one use either EAS via the DUET™ or via natural acoustic low-frequency hearing. Interestingly, in a study using a similar insertion depth to ours [3], only 10 of 27 subjects had sufficiently preserved hearing to use EAS. This is quite a low functional hearing preservation rate compared to the data we present. This might be reflected in the fact that the ''soft surgery technique'' was followed in only 12 cases. This again highlights the special attention that must be paid to the surgical technique in order to preserve hearing for EAS.

5. Conclusion

There is a certain group of children, with a ski-slope type hearing loss, who obtain benefit from electric-acoustic stimulation. Following surgery they show improvements in speech perception abilities, both in quiet and in noise. Parents of these children report a change in listening behaviour and ease of listening. It is important to note that this group of children do not demonstrate any benefit with traditional acoustic amplification and are often reported to be ''loners'' or socially excluded from their peer group. They are neither deaf nor hearing, and such hover in a world ''in-between''. Provision of PDCI opens up the hearing world to these children.

Acknowledgments

The authors would like to thank Miss Martina Deibl for the statistical analysis in this paper and Miss Anke Weninger for drawing the audiograms.

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