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Assessment of the Outcomes Following Cochlear Implantation in Young Children

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Introduction

Since the first cochlear implants (CI) were fitted in children with congenital and acquired deafness at the ENT Department of the Medical University of Hannover, with follow-up rehabilitation at the Cochlear Implant Centre Hannover (CIC), we have gained over ten years of experience in this field. Today the operation is a standard procedure. To date CI surgery has been performed in Hannover on some 2000 people, around 950 of them children. The *Wilhelm Hirte* CIC currently (as of 18.08.2000) has 645 children in its charge, aged between seven months and 16 years; since 1994, however, the majority of children aged five and upwards have undergone rehabilitation at a different centre.

The implants used have an excellent safety record, as shown by the very low failure rates over the last 12 years (Bertram et al., 2000).

The launch of the children's CI programme necessitated the creation of facilities for intensive basic therapy in a purpose-built children's centre (Lehnhardt, Bertram, 1991; Bertram, 1996). The therapy lasts a total of twelve weeks (spread over a period of two and a half years) and includes the adjustment of the speech processor, interactive hearing and speech therapy, motopaedic and rhythmic/musical training and activities to encourage the children's creativity. Each week 21 children are admitted to the centre with their mother, father or guardian. The CIC comprises three accommodation units for children, each with seven single rooms, and a rehabilitation building within grounds covering an area of 7000 m^2. The centre's team of staff comprises three teachers of the deaf, three speech and language pathologists, one speech therapist, one specialist who conducts breathing exercises and vocal and speech training with the children, one childcare worker (for rhythmic/musical training), one motologist (for movement training) and three engineers responsible for the tuning of the speech processors. The medical care is provided by the ENT department. The CIC consults teachers and therapists in the child's place of residence before surgery is carried out. Reports are requested from teachers of the deaf and school psychologists on the child's previous development and the degree of success achieved with conventional hearing aids. This contact is maintained after the implant has been fitted, since the specialists from the establishments for the hearing-impaired are encouraged to visit the CIC and observe the period of basic therapy. Over the course of the two and a half years there is a continual two-way exchange of information, by means of reports and telephone calls, to provide updates on the rehabilitation process.

Staff advise parents and teachers in detail before and after the implantation on the potential and limitations of the cochlear implant. The centre is very much involved in the decision as to whether or not to go ahead with implantation. Although the ENT department at the Medical University of Hannover and the *Wilhelm Hirte* Cochlear Implant Centre are separate and autonomous institutions, the excellent teamwork has been and remains of crucial importance in terms of candidate selection, rehabilitation, long-term medical care and pedagogical support and the evaluation of success in the years following basic therapy in the CIC.

A pre-requisite for CI candidates is their suitability from the point of view of otolaryngological health (including such factors as profound sensorineural hearing loss in both ears, little or no benefit from hearing aid use, no medical contraindication) and with respect to pedagogical-psychological considerations (Lehnhardt, Hirschhorn, 1986; Lehnhardt, 1990; Bertram, 1991; Lenarz, 1994).

Why do we measure the development of auditory, speech and language skills following cochlear implantation?

Evaluating the development of auditory, speech and language skills and the personality of the implanted child is of interest for the parents, the teachers, the therapists and the health insurance companies which finance both the operation and the subsequent rehabilitation programme. Against the background of increasingly scarce financial resources it is understandable that the health insurance companies should raise the issue of the cost/benefit ratio. Examinations and tests carried out in the CIC and in the ENT department during and after the completion of basic therapy as well as annually during the long-term follow-up period should confirm an improvement over time in the above-mentioned abilities. The data obtained clearly demonstrates how much more effective a CI is compared to a conventional hearing aid in promoting hearing-based speech acquisition in children with a profound to total hearing loss. The test results obtained over a long period of time and with a large number of subjects can also be used as predictors with respect to the development of speech perception and speech competence in implanted children.

However, it is also pertinent to ask the related questions

- "What is success?" and

- "How do we define success and which parameters should be used to measure it?"

Within this context it is important to reflect on which parameters are of greater significance:

- The objectively obtained 'hard' data on auditory and speech abilities (poor or excellent outcome) or

- the mental well-being of the children and their subjective degree of satisfaction with the implant.

Considerable differences exist between individual children (in terms of maturity, age, onset and duration of deafness, auditory experience, residual hearing etc). Also subject to marked variation are the type and intensity of post-operative support at home and at school. These factors influence the long-term success, making comparisons between children more difficult.

What should be tested in order to measure success, and which procedures should be used?

Cochlear implantation of children with profound hearing loss is linked to the expectation that these children should demonstrably improve their speech perception skills over the years and show increasing speech competence (in terms of grammar and vocabulary) compared to their situation prior to implantation and rehabilitation. It is also to be expected that the development of their cognitive skills will be stimulated and that there will be improvements in their social behaviour.

There is a consensus of opinion that progress in the above-mentioned areas takes several years. This is in part due to the fact that the children's ability to use their sense of hearing for speech acquisition and for general development is delayed. In neurophysiological terms this corresponds to a delay in the supply of adequate sensual stimulation to the central auditory system. Only when this condition is met can the nerve cells responsible for the analysis of speech signals perform this function, and neuronal networks will only form, spread and functionally mature when the external stimuli are provided regularly (Klinke, 1998).

Various test procedures are in use which document the development of speech-perceptive performance, to record the length of time for which the speech processor is worn daily, to assess the spontaneous reaction of the children to environmental noise and to evaluate speech over the course of the day.

Auditory perception with the aid of the cochlear implant is objectively tested by CIC by means of the **HHPR test (Hannover-Hörpruefreihen)** (Bertram, 1996).

In order to assess the articulation and phoneme inventory we use the **Ravensburger Lautprüfbogen** (40 words). We obtain data both preoperatively and postoperatively with the aid of a comprehensive form for recording aspects of general development and the development of auditory and speech skills (Bertram et al., 1996).

For the test procedure the ENT Department at the Medical University of Hannover uses the **TAPS** (Test of Auditory Perception of Speech, Cochlear AG, Basel, 1992).

The **Mainzer Kindersprachtest,** which is a monosyllabic test, the **GASP** (Glendonald Auditory Screening Procedure; after Erber, 1982) and the **Mr Potatohead Task** (Robbins, 1993) are also employed, and the discrimination of **minimal pairs** (Schäfer test of vocal agnosia) tested.

The spontaneous reaction of implanted children to environmental noise is tested by means of the **MAIS** (Meaningful Auditory Integration Scale, Robbins et al., 1991) and the **MUSS**

(Meaningful Use of Speech Scale, Robbins, Osberger, 1990). MAIS and MUSS are so-called 'parent report scales'.

GASP, Mr. Potatohead Task, Monosyllabic Test, Minimal Pairs 7 and MAIS and MUSS are elements of the United States Food and Drug Administration (FDA) test.

Psychological testing of the children is performed using the age-dependent **SON-R 2½-7** (Snijders-Oomen non-verbal intelligence test, 1996) or the **SON-R 5½-17** (Snijders-Oomen non-verbal intelligence test, 1997) test.

Children with especially poor test scores are subject to additional preoperative and postoperative diagnostic procedures.

A two-year study on the acquisition of grammatical and lexical structures was carried out by G. Szagun (1998; 2000) of Oldenburg University using children implanted at CIC Hanover.

Difficulties in obtaining data on speech perception performance and speech competence in children with CI

The testing of speech-perceptive abilities in young children with cochlear implants is a considerable challenge. Difficulties are encountered owing to the fact that the vocabulary of very young congenitally deaf children is generally non-existent, or at best very minimal. They are unable to make use of auditory speech memory, since this is absent in most cases. Their cognitive immaturity must also be taken into account (Donoghue et al., 1998; Lenarz et al., 1999; Rizer, Burkey, 1999; Nikolopoulos et al., 1999; Lehnhardt ,1978; Tyler et al., 2000).

The ability to cooperate with the tester over an extended period is also limited, since young children tire more easily, with a corresponding decline in attentiveness and concentration. The ability to understand the tasks is to a large extent dependent on the child's age, speech competence and cognitive abilities. In particular, the associative skills of the children also influence their ability to recognise noises or speech as information and store and recall lexical material.

In order to overcome these difficulties as far as possible, it is necessary for the tester to deal sensitively with the child and use appropriate testing procedures, i.e. techniques which take into account the linguistic restrictions of these children. In order to be able to evaluate their speech perception and speaking skills a variety of tests need to be employed, with continuous monitoring over a long period of time.

As the age at which it is possible to diagnose children suffering from profound hearing loss is reduced by suitable screening methods (OAE and other procedures such as ABR), it will be increasingly possible for children under the age of two to be fitted with a cochlear implant. This places increased demands on the designers of testing materials to ensure their suitability for this age-group.

Materials and methods

Examples of these test procedures are presented below:

The **HHPR test** (Hannover Hörpruefreihen; Bertram, 1996) attempts to measure the different levels of auditory perception. It consists of **11 subtests**, which test

1. the discovery of noise and speech
2. the discrimination of fixed syllable patterns
3. the identification of monosyllables and
4. the identification of disyllables.

Tests on

5. phonemic discrimination of monosyllables and disyllables and
6. the identification of sentences

are also conducted.

Subtests 2-6 are **closed sets** with appropriate visual material. A random sequence of purely auditory stimuli (CD recordings of a woman's voice) is presented.

Subtests 7, 8 and **9** investigate **open speech perception** by means of words and sentences which vary in their degree of difficulty. Visual material is no longer used. These tests also involve the use of CD stimuli.

The aim of **subtests 10 and 11** is to assess speech perception and speech production with the aid of **22 different toys** and instructions for games or demonstrations. Phonemically balanced lexical material was not used, in order that words known to 3-4 year old children might be supplied. The children were tested before the operation, again one year after implantation and once a year thereafter.

The Test of Auditory Perception of Speech (TAPS), developed by Cochlear AG, Basel (1992) was also developed for cochlear implanted children aged three and above. The test is available in four languages, with German, English, French and Spanish versions.

The TAPS (German version) includes five test categories:

1. the detection of speech sounds
2. the perception of prosodic features
3. the identification of auditory and visual information
4. the integration of auditory and visual information and
5. the testing of open speech perception

The different test categories were presented live voice.

The Meaningful Auditory Integration Scale (MAIS) was designed by Robbins et al. (1991) as a parent report scale in interview format. The questioning technique avoids the common pitfall of provoking the parents into giving the ‘desired’ answer. It facilitates a dialogue between tester and interviewee and the style of questioning enables more information to be obtained. The parents are asked to report in as much detail as possible the reaction and behaviour of their child in response to noise and state for how long they are willing to have the speech processor switched on each day. A maximum of 40 points is obtainable under the scoring system. Each of the 10 questions is scored on a five-rung scale from 0 (lowest) to 4 (highest).

The Meaningful Use of Speech Scale (MUSS) is, as with the MAIS, a parent report scale. It also consists of 10 questions, the aim of which is to assess voice control, the use of speech without gestures or gesticulation and the employment of communication strategies in everyday life. The procedure and scoring system used are the same as those for the MAIS.

The MAIS and MUSS tests are particularly suitable for evaluating the auditory performance of very young children, multihandicapped children and foreign children, with whom it is not possible to objectively measure speech perception by means of standard speech perception tests (von der Haar-Heise et al., 2000).

Results

At present the CIC *Wilhelm Hirte*, Hannover, is caring for 645 CI-implanted children (as of August, 18, 2000). 405 of these children have since 1990 completed the initial rehabilitation stage of 2½ - 3 years. Once a year the speech processor programs are checked and the actual speech perception capabilities of the children monitored in the CIC and in the clinic.
240 children are currently within the initial rehabilitation stage. The overall Figure 1 does not include children (mostly older than 6) which were also implanted in the Otolaryngological University Clinic, Hannover, and are completing their rehabilitation at the CIC Werscherberg (about 150 children).

Since 1988 399 children were implanted with Nucleus systems. Since 1993 244 were implanted with the Clarion system. One child (reimplanted in Hannover, received initial treatment in another clinic) was implanted with a C40 + (MED EL). Another child was implanted with the same system (surgery in another clinic) (Figure 2).

467 of the children were born deaf, while 133 lost their hearing within two years of their birth. 45 lost it later (Figure 3). The majority of the implantation children is in the age group of 2 – 4 year old (in the center currently mainly children aged 0;7 to 6;0 are in the rehabilitation programs. The number of older children is lower) (Figure 4).

For evaluating the auditory speech perception capabilities, the three demanding words and sentences 1 and 2 test sets (all open sets) of the Hannover-Hörprüfreihen were used. The mean result achieved by each respondent in all test sets were converted into percentage values, from which the equally-weighted arithmetic mean was calculated.
All mean values of a group were totalled and divided by the respective number of respondents. Each group included usually a minimum of 20 to sometimes over 100 children (exception: "Group of children who lost hearing aged 2+" with at least 10 respondents per group). All respondent groups tested over the years showed improving speech perception skills, in terms of the age of deafness (Figure 5), of its cause of deafness (Figure 6) or the CI implantation side (Figure 7). No significant variations between the groups were found in the monitoring intervals (12, 24, etc.months). It must be noted, though, that within the different groups of respondents considerable variations of individual results were found. Multiple-Handicapped children with mental disorders were not monitored.
If despite intensive audio-lingual training the expected progress cannot be achieved, this might be caused by partial functional disorders which inhibit the development of perceptive and expressive oral communication skills (e.g. dyspraxia, legasthenia, concept formation disorders etc.). This should be particularly taken into account for parent counseling in the preliminary examination stage, as well as for the postoperative rehabilitation.

Of the 435 children who had been born deaf, prior to surgery 142 attended institutions for the deaf, 192 attended educational institutions for the hearing impaired, 17 attended regular schools, 23 attended integrated educational institutions and 48 attended other institutions or were cared for by therapists. 13 children had no educational training (Figure 8). The actual situation for the 192 children who prior to surgery attended institutions for the hearing impaired, is as follow: 26 are attending regular schools, 10 are attending integrated educational institutions, 3 are attending institutions for the deaf and 7 attending other educational institutions. 146 children remained in institutions for the hearing impaired (Figure 9). Of the children in institutions for the deaf, today 14 are attending regular, 51 are attending educational institutions for the hearing impaired, 12 are attending integrated educational institutions and 7 are attending other educational institutions such as kindergarten for children with speech, learning or physical handicaps. 58 children are still attending institutions for the deaf (Figure 10).
It remains to be seen whether and to what extend changes will happen within these groups over the following years, or whether no changes happen at all.

Illg et al. (1999) were able to show that congenitally deaf children aged between one and 15 years are able to benefit from cochlear implantation. Although older children had more experience of hearing aids and auditory training, younger children show more rapid and consistent improvement over a period of years, both in speech-perceptive abilities and speech competence (the latter, however, somewhat later). The performance of older children seems to level off between the 12th and 18th month following implantation.

Congenitally deaf children should be fitted with a CI as early as possible, since they achieve better results are better in terms of speech perception skills and level of speech intelligibility attained. Age should not, however, be the sole criterion for selection (Nikolopoulos et al., a.a.O.). Other studies published reach similar conclusions (Tyler et al., a.a.O.; Rizer, Burkey, a.a.O.; Lenarz et al., a.a.O.).

Discussion

Our own investigations and international comparative studies have shown that speech perception skills and speech acquisition are superior in those children implanted at an early age than in older children, assuming that no additional disorders are present. The performance of older children levels off, whereas younger children (aged 1-4) show steady improvement.

However, objective testing of auditory and perceptive skills in very young children proves difficult. In addition, children have very different backgrounds and requirements, with parental homes and educational environments varying considerably in respect of the support systems at their disposal.

There is also a noticeable trend towards increasing rates of cochlear implantation in younger children with severe hearing loss (Osberger, 1997; Lenarz et al., 1997). This makes it necessary that the medical and pedagogic-psychological selection criteria for implantation are subject to constant scrutiny. There is a need to find broadly reliable predictors which enable the likelihood of success to be anticipated, and suitable testing materials need to be developed for the objective assessment of the hearing and speech development of very young children following cochlear implantation.

Summary

The cochlear implant has proved its ability to enable hearing-assisted speech acquisition in young children with both congenital and acquired deafness, albeit to differing degrees. Evaluation of the results is interesting not only for parents, doctors and teachers but also for the financial backers, the health insurance companies. Such an appraisal can demonstrate that the intended benefits are indeed being realised and can provide useful guidance for selection criteria and predictions of success. It has been demonstrated that very young children benefit particularly from CI. It is therefore necessary to develop appropriate testing procedures for this age-group.

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Appendix

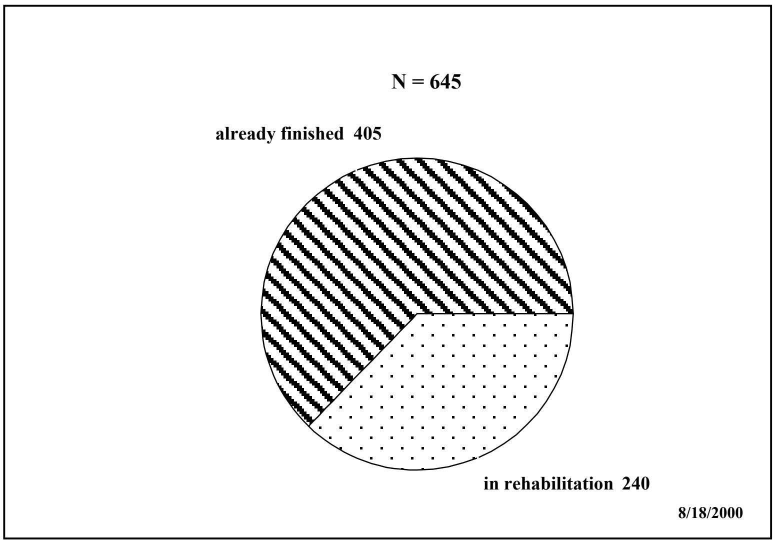

Figure 1: Number of CI-supplied

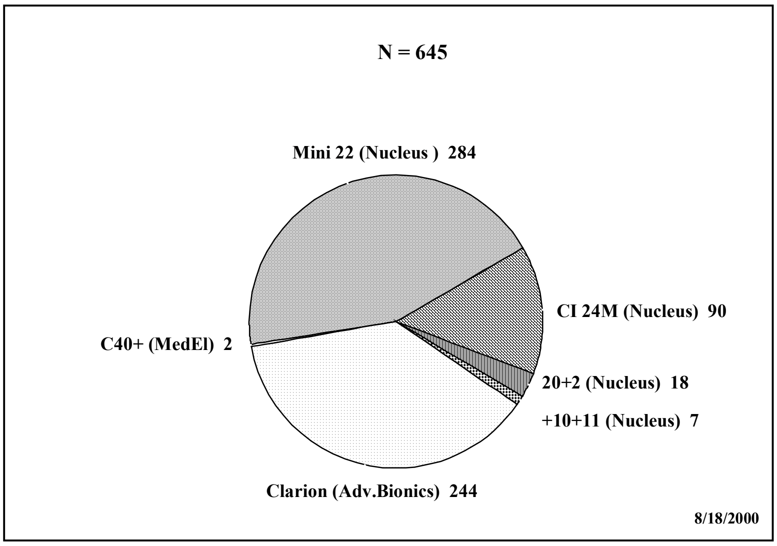

Figure 2: Distribution of the CI-systems

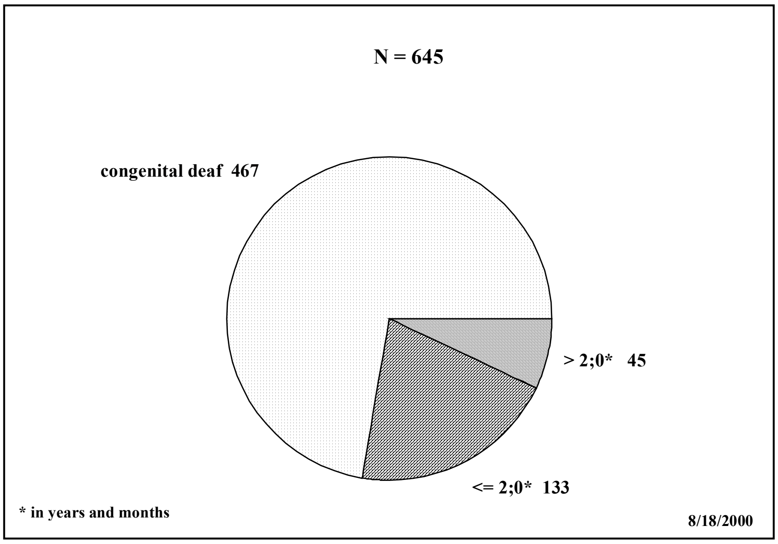

Figure 3: Beginning of deafness

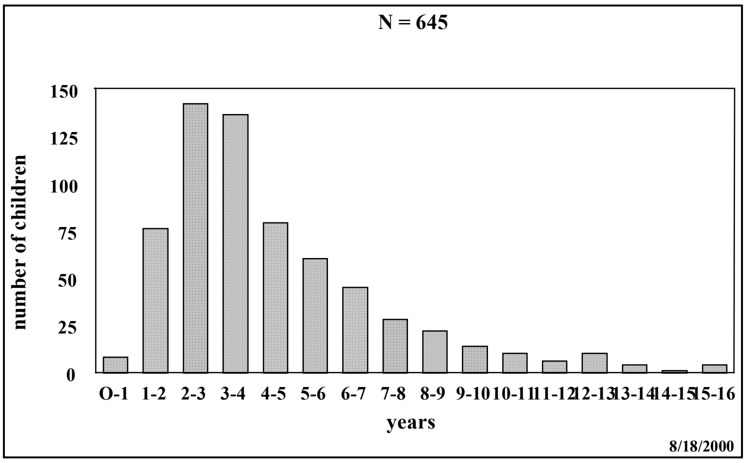

Figure 4: Age at the time of CI-supply

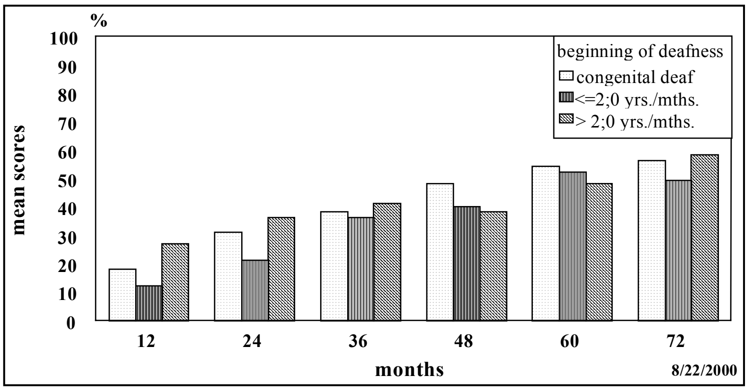

Figure 5: Development of auditive speech-perception regarding the age of deafness [words / sentences 1 / sentences 2 (open set)]

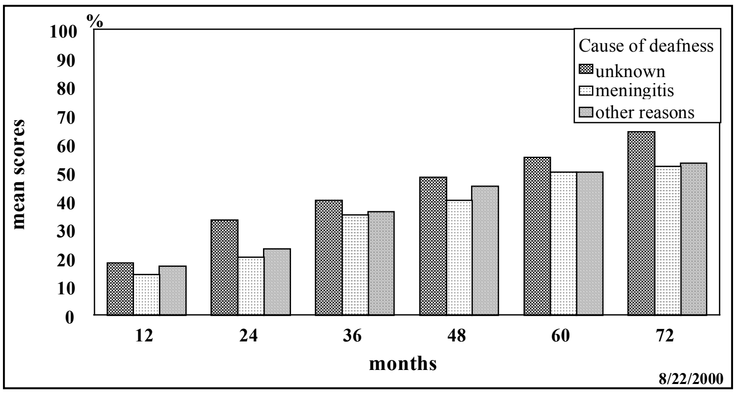

Figure 6: Development of auditive speech-perception regarding the reason for deafness [Words / sentences 1 / sentences 2 (open set)]

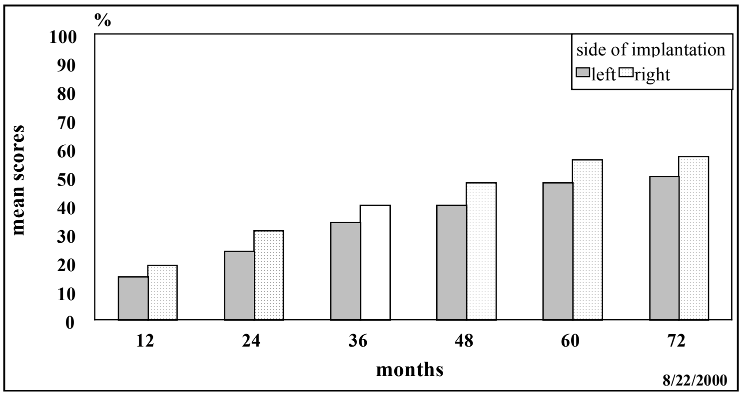

Figure 7: Development of auditive speech-perception regarding the side of implantation [Words / sentences 1 / sentences 2 (open set)]

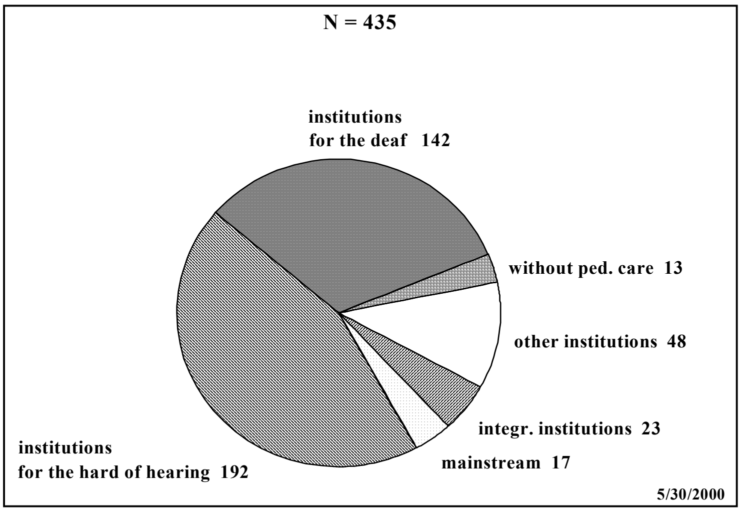

Figure 8: Pedagogic care of congenital deaf children before CI-supply

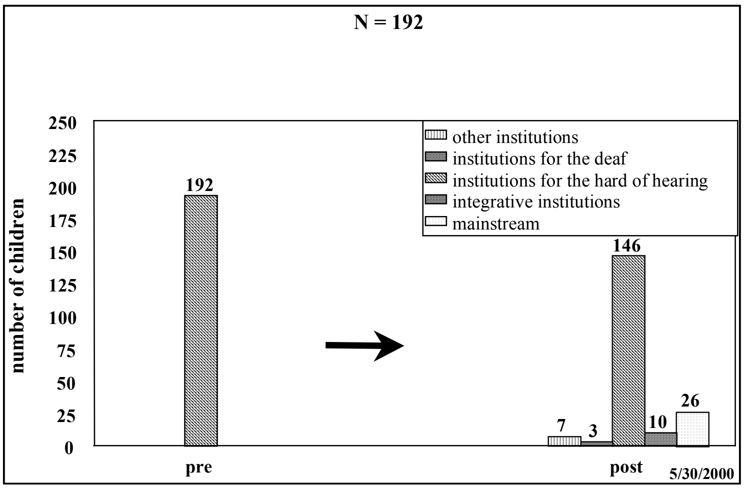

Figure 9: Pedagogic care of congenital deaf children from the institutions for hard of hearing after CI-supply

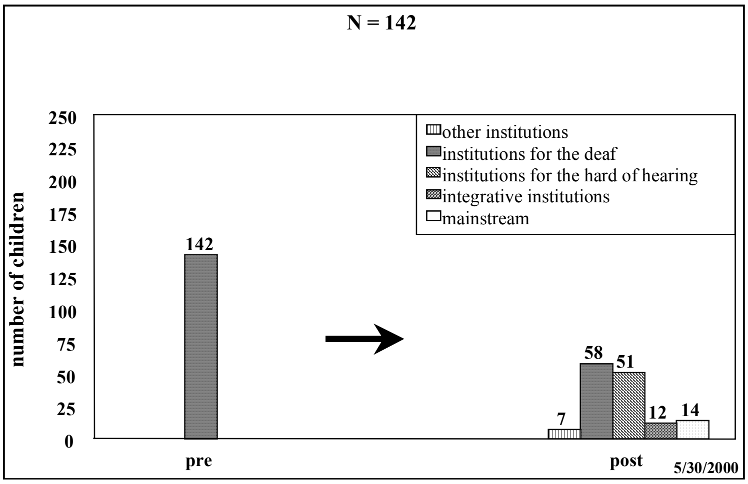

Figure 10: Pedagogic care of congenital deaf children from the institutions for the deaf after CI-supply