Special Communication

COCHLEAR IMPLANTATION IN DEAF CHILDREN

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A cochlear implant (CI) is a hearing device introduced in the 1980s for profoundly deaf subjects who gained little or no benefit from powerful hearing aids. This device comprises an electrode array inserted in the cochlea, connected to an internal receiver, and an externally worn speech processor. The CI transforms acoustic signals into electrical currents which directly stimulate the auditory nerve. Since the early 1990s, cochlear implantation in children has been developing rapidly. Although it is still difficult to predict how a child will perform with a cochlear implant, the success of cochlear implantation can no longer be denied. In this paper, some recent papers and reports, and the results of the various Nijmegen cochlear implant studies, are reviewed. Issues about selection, examinations, surgery and the outcome are discussed. Overall, our results were comparable with those of other authors. It can be concluded that cochlear implantation is an effective treatment for postlingually deaf as well as prelingually (congenital or acquired) deaf children with profound bilateral sensorineural deafness. *Ann Saudi Med 1997;17(5):533-539.*

Cochlear implantation is widely accepted as a routine clinical procedure for selected deaf children. It restores deaf children's perception of sound through the use of a special electronic device. The CI system comprises an electrode (single or multi-channel) placed in the cochlea, connected to an implanted receiver, and an externally worn microphone, signal processor and transmitter. The speech processor analyzes the sound signal from the microphone and transmits it transcutaneously to the internal receiver. The electrical stimulation by the CI bypasses the non-functional parts of the cochlea and delivers signals directly to the auditory nerve. Because of the direct stimulation of the nerve, most CI users perceive hearing sensations that cannot be obtained with even the most powerful conventional hearing aid. Owing to technological evolution, different types of CI devices have been introduced. The difference between the various types is in the electrode designs and/or speech processing strategies. However, they all consist of similar basic elements.

In recent years, remarkable progress has been made in the application of pediatric cochlear implantation, from the research stage to regular clinical application. At least 3400 subjects under 18 years have received CI worldwide. Of them, 400 were under the age of three years when implanted and a further 1250 were between three and six years.[1] The main goal of CI application is to restore hearing in children with profound hearing loss, thus enhancing their ability to participate in aural-oral communication.

Studies have revealed that the majority of children using a CI with a prelingual (before three years of age) or postlingual (after three years of age) onset of deafness obtain significant benefit from this prosthesis. However, speech perception abilities vary widely, ranging from the simple identification of sounds to the recognition of normal open speech.[2-6] Most users benefit more from their CI than from conventional hearing aids.[1,3,7,8]

The problem of post-implant, variable speech perception abilities continues to challenge research teams, and efforts are being made to find a means of predicting the result prior to cochlear implantation. So far, no preoperative factor has been found that can predict the outcome of cochlear implantation. However, it is known that some biographical factors, such as age at the onset of deafness and the duration of deafness, play a role.[1,9]

To obtain a good result, it is generally reported that careful candidate selection is necessary, and that a rehabilitation program should follow cochlear implantation. Success in this field can no longer be denied, in spite of initial skepticism in the scientific world and the deaf community.[10,11]

This paper presents an overview of the current concepts of cochlear implantation and reviews the results of CI studies at the University Hospital Nijmegen. In Nijmegen, the pediatric cochlear implantation program was initiated in 1989 in close cooperation with the Institute for the Deaf in St. Michielsgestel. Initially, the one-channel Med. El device was used, but later on, the 22-channel Nucleus

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device was introduced. By the end of 1996, 44 profoundly deaf children had been implanted (Table 1).

Selection Criteria and Preoperative Tests

Preoperative Assessments

Pediatric cochlear implantation requires medical, audiological and psychological evaluation. A routine ENT examination forms the initial part of the evaluation. Radical mastoidectomy or tympanoplasty, without any long-term problems, are not considered as contraindications.[12,13] In general, preoperative audiological assessment is considered as the major factor to determine the suitability of a child for cochlear implantation. The audiological test batteries consist of play audiometry or visual reinforcement audiometry, tympanometry and speech perception tests. Audiological assessment should confirm profound, bilateral sensorineural hearing loss, without useful residual hearing. To determine the potential of residual hearing, the use of powerful hearing aids with an appropriate auditory rehabilitation period is essential. Generally, speech perception tests quantify a child's ability to use his/her residual hearing effectively. Such data obtained preoperatively are also valuable as a reference for comparison with postimplant scores. To confirm the results of behavioral hearing tests, objective electrophysiological tests, such as auditory brain stem response (ABR) and/or electrocochleography measurements, are often used.[11]

The radiologic evaluation includes high-resolution computed tomography (HRCT) scanning, which is a prerequisite to determine possible ossification of the cochlea and congenital anomalies as well as anatomical landmarks.[14-17] Generally, ossification of the cochlea is not considered as a surgical contraindication for cochlear implantation.[7,13,17,18] However, in such cases full insertion of the electrodes is not always possible and the results of implantation might be poor.

Children undergo psychological testing as part of the preoperative assessment, to rule out any severe problems. The expectations and motivation of the child and the parents have to be realistic.[7,19]

Evolution of the Selection Criteria

Over the years, the selection criteria have changed as greater insight has been gained into the effect of several biographical factors upon CI performance. Various studies have revealed that postlingually deaf adults perform better with their CI than prelingually deaf adults.[1] The difference in performance between pre- and postlingually deaf children is far less pronounced.[2,20] Prelingually deaf subjects who received an implant during childhood achieved a higher level of performance than those who received CI during adulthood.[4]

TABLE 1. *Biographical data on the children (<14 yrs) who received a cochlear implant in Nijmegen.*

	Prelingual	Postlingual
Number of children	38	6
Male	13	5
Female	25	1
Age at onset of deafness		
Range, yrs	0.0-2.9	3.1-6.9
Mean, yrs	1.2	4.2
Duration of deafness		
Range, yrs	1.9-13.4	1.3-7.9
Mean, yrs	5.4	4.4
Age at cochlear implantation		
Range, yrs	2.9-13.4	4.3-12.3
Mean, yrs	6.6	8.6
Duration of CSI use		
Range, yrs	0.5-6.9	0.6-4.2
Mean, yrs	2.7	3.4

Dowell et al.[2] reviewed the speech perception results of all the children and adolescents (up to 19 years of age) implanted in Melbourne and Sydney. In agreement with other authors, they observed that the range of speech perception performance was wide. Their results indicate that the age at onset of hearing loss and the age at the time of cochlear implantation do not have any significant effect on speech perception. However, the duration of deafness and the duration of implant use had a significant effect.

In the recent literature, the youngest children implanted were under two years. Implantation at such a young age is only feasible if profound bilateral sensorineural hearing loss can be diagnosed with complete certainty, and if the child has not benefited from conventional hearing aids. Cochlear implantation at a young age may minimize the negative effect of auditory deprivation and in the case of meningitis, it might help to prevent labyrinthitis ossification which would impede later implantation. Cohen and Waltzman[10] reported that eight children under two years received CI at their institute and all showed significant benefit. The Hannover group has also implanted such young children, with encouraging results (personal communication). Nevertheless, more data are required to show the benefits of early implantation and help to guide future policy.

Surgery

Surgical Technique

CI surgery can be performed successfully in children, in spite of some difficulties, particularly with an ossified cochlea.[18] Access to the cochlea is obtained by a mastoid and facial recess approach, as is used in surgery

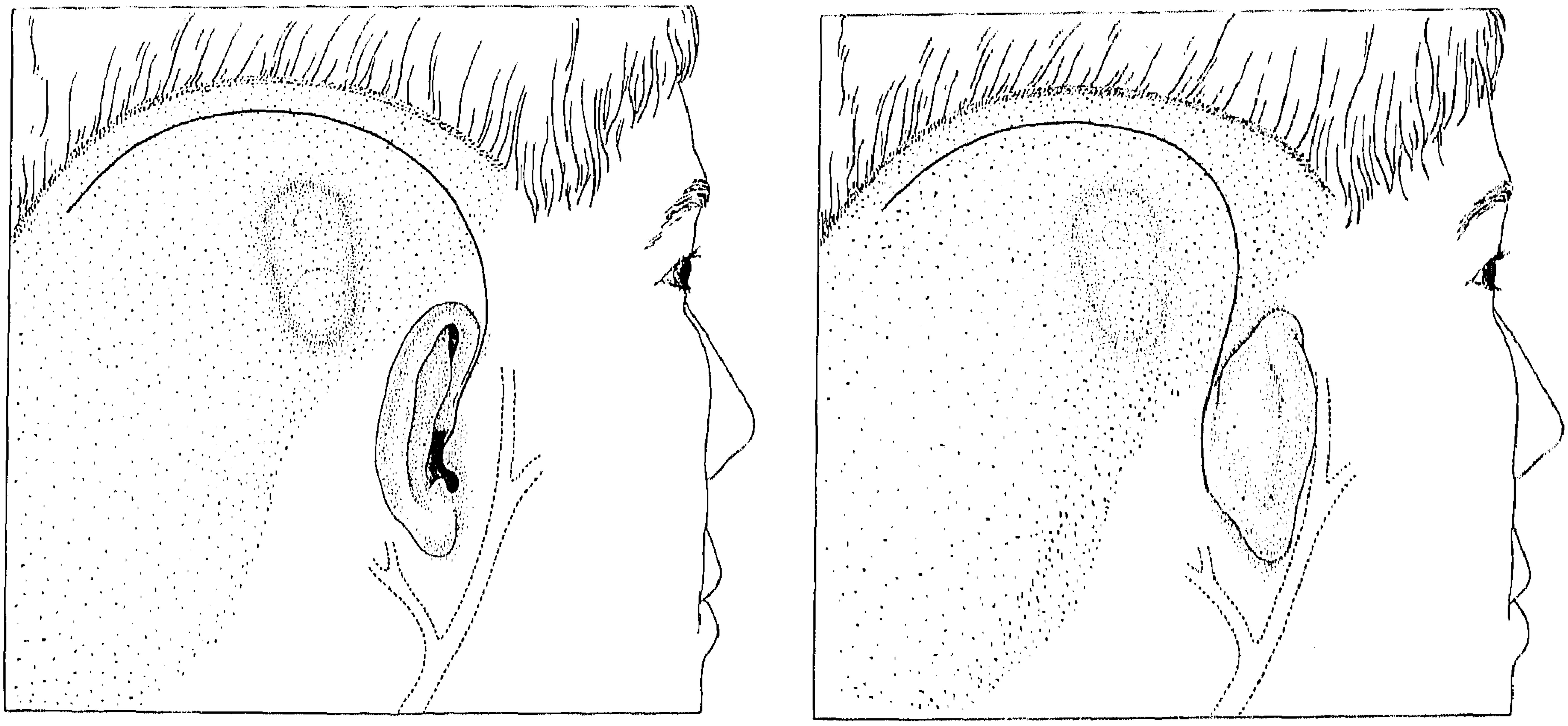

FIGURE 1A. Diagram of an endaural incision. 1B. A retro-auricular incision employed for cochlear implantation. Condensed dots indicate the site of placement of the receiver.

for chronic otitis media. The receiver-stimulator is positioned just above and behind the pinna (Figures 1A and 1B). The incision should be made at least 1 cm away from the planned site for the internal receiver. Several types of skin flap design have been advocated, and the basis for the designs is to maintain a good vascular supply to the flap.

In Nijmegen, two types of incision are used: an endaural incision (Figure 1A) or a retro-auricular incision (Figure 1B). Both curve upwards and backwards, high over the parietal region. Due to the dimension of a child's skull, the thinness of the skin and the later growth of the skull, the incision is made right down to the bone. The temporal muscle is lifted from the parietal portion of the temporal bone, with the subcutaneous tissue and skin as a single layer flap. This surgical modification minimizes problems with wound healing and possible electrode extrusion.

After elevation of the skin flap, the dura mater is sometimes exposed when drilling the well for placing the receiver-stimulator. It is usually necessary to gently push the dura mater down with a thin piece of bone to accommodate the receiver coil. Following mastoidectomy, a facial recess approach is used to gain access to the middle ear and round window niche.[21] The facial recess is opened, the facial nerve is skeletonized, avoiding exposure of the nerve sheath. Cochleostomy can be performed in two ways: through the promontory anterior to the round window membrane, or through the round window membrane itself.[11,21] The electrode array should be inserted gently to prevent damage to the delicate cochlear structures as much as possible.

Ossification of the cochlea, as is often found in post-meningitis cases, needs drilling to open the scala tympani for insertion of the electrode array. In some cases with severe ossification, extensive drilling of 6 to 8 mm is necessary. If no fluid-filled lumen is found, this may result in partial insertion.[15] Hartrampf et al.[18] reported that in cases with cochlear ossification, at least seven electrodes of the Nucleus 22-channel system can be inserted. After insertion, the cochleostomy is sealed with bone dust or soft tissue and glue. In general, the electrode lead is placed in a groove created in the superior part of the mastoidectomy fossa and fixed in the fossa incudis. This is because the distance from there to the round window does not change after birth. The receiver-stimulator should be tied down securely.

Complications

The surgical complication rate of the implant procedure is low in children. Largely, the complications are comparable with those of middle ear surgery. In addition to surgical complications, device migration or failure may occur. No major complications occurred in any of the 44 children who received a CI in Nijmegen. However, postoperative complications were found. A minor wound infection occurred in one child, while another child had a surgical hematoma. In five children, only partial insertion of the electrode array was possible due to severe ossification of the cochlea.

Revision surgery can be performed either to upgrade the CI system or to replace a failing device. It is possible to explant and reimplant without damage to the cochlea or the auditory nerve.[21]

Preoperative Imaging and Surgical Results

HRCT scanning has proven to be a valuable tool for the preoperative assessment of cochlear patency. However, minor or major cochlear ossification encountered during surgery is not always visible on preimplant radiological studies.[13,17,22] If the HRCT scan seems to be normal in children with a history of meningitis, the surgeon should suspect obliteration of the round window and part of the basilar turn.

A Dutch study on the predictive value of HRCT scanning carried out in 88 subjects (children and adults) with a CI showed a relatively large number of false-negatives, mainly in children, when compared to the intraoperative findings. The data are presented in Table 2. This means that in spite of its value, the accuracy of preoperative HRCT scanning is not optimal.

Electrophysiological Measurements

To achieve the best results with a CI, it is important to adjust the processor output to the user's dynamic range. This may be a problem in young children.[23] To tackle this problem, several investigators performed measurements to assess threshold and comfortable levels directly after placement of the CI, while the child was still under general anesthesia. For this purpose, electrically evoked ABR measurements (EABR) and/or electrically evoked stapedius muscle reflex (ESR) measurements were performed.[23,24]

A technical restriction of the EABR measurement is that it is more susceptible to noise and electrical artifacts than the ESR measurement. A specific problem with intraoperative ESR measurement is that anesthetic agents influence the outcome.[24,25] To illustrate this, Figure 2 shows an example of ESR thresholds recorded intraoperatively. During the measurement, the concentration of an anesthetic agent (Halothane) was increased and later readjusted to the original level. A significant effect of the Halothane concentration was seen. Figure 2 also shows the postoperative value obtained six months after device fitting. In general, postoperative ESR thresholds were lower than those measured during surgery.[24] Owing to the technical restrictions and the poor relationship with the behavioral results, intraoperative EABR and ESR data should be used with caution for device programming.

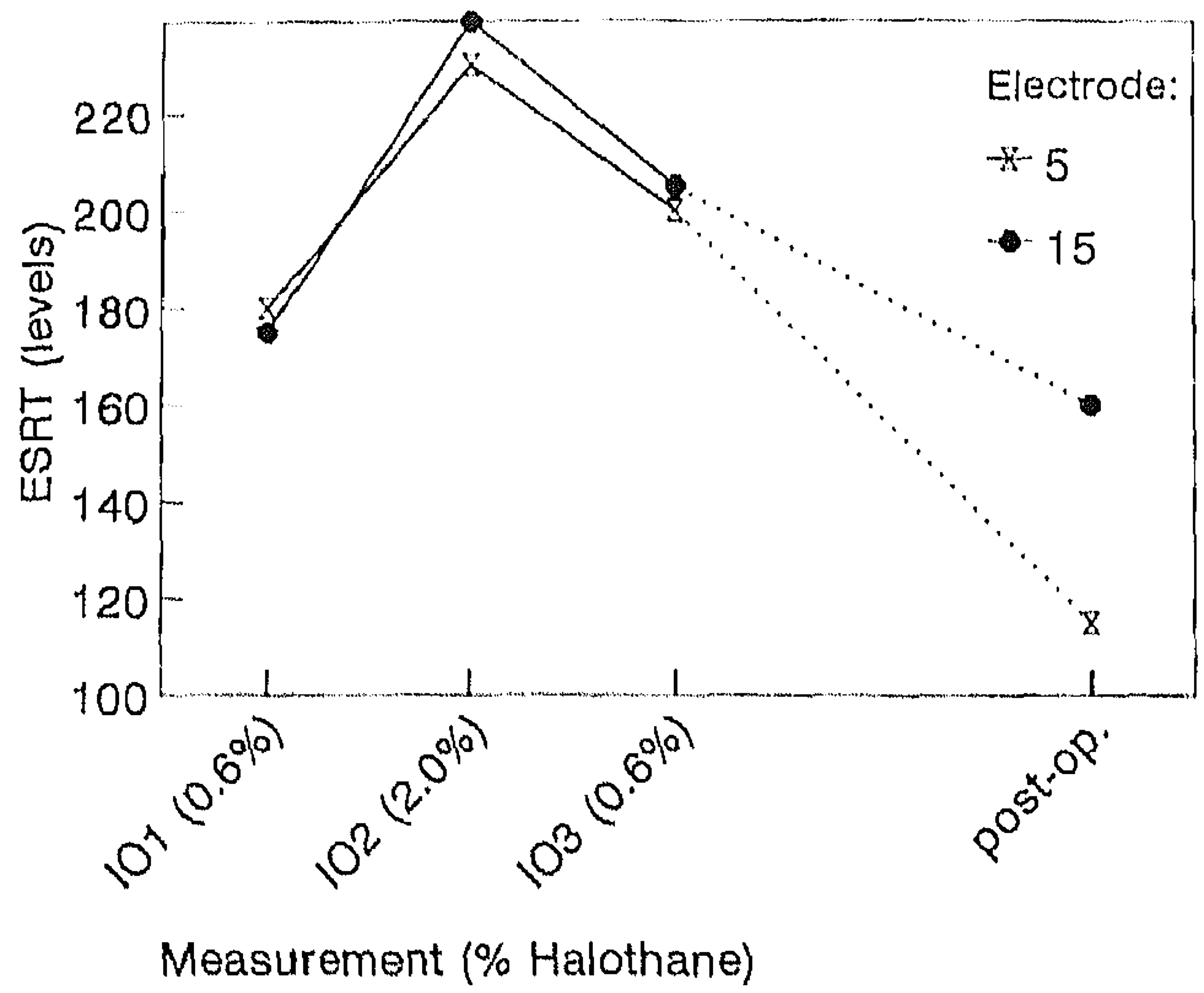

FIGURE 2. A typical example of electrical-evoked stapedius reflex thresholds (ESRT) recorded intraoperatively from electrodes 3 and 20, while the concentration of the anesthetic agent (Halothane) was varied from 0.6% (IO1) to 2.0% (IO2) and back again to 0.6% (IO3). Postoperative results obtained six months after device fitting are also indicated.

Rehabilitation

The aim of the rehabilitation program is to achieve optimal use of a CI. New auditory abilities should be utilized to develop new auditory and communication skills. Generally, speech perception skills improve after cochlear implantation. After the initial rehabilitation period, normal learning in daily life contributes to the optimal use of a CI.[19]

Collaboration of the CI team with tutors in a setting for the deaf is essential, especially for children. If a child does not receive rehabilitation and encouragement for spoken language, the outcome of cochlear implantation is likely to be disappointing. In cooperation with the Institute for the Deaf in St. Michielsgestel, the initial rehabilitation period in Nijmegen takes two weeks. After this period, implanted children return once every month for tutoring for at least one year. After rehabilitation, they should be able to continue learning at home and at school, at their own speed and in their own manner.

Outcome of Cochlear Implantation

Several studies have focused on speech perception skills in children with a CI. Gantz et al.[20] studied the benefit of the Nucleus multichannel CI in 54 children. They found that the speech perception skills of postlingually deaf children improved significantly during the first year after implantation. The prelingually deaf children progressed at a slower rate than their postlingual counterparts. However, some of the prelingually deaf children attained comparable, and in some instances better,

TABLE 2. *Preoperative high-resolution computed tomography findings in 88 candidates for cochlear implantation compared to the surgical findings.*

	Adults HRCT scan	Children HRCT scan	Total no. of subjects
True-positive finding	7 (11.3%)	4 (15.4%)	11 (12.5%)
False-positive finding	5 (8.1%)	1 (3.8%)	6 (6.8%)
True-negative finding	39 (62.9%)	12 (46.2%)	51 (58%)
False-negative finding	11 (17.7%)	9 (34.6%)	20 (22.7%)
Total	62	26	88

speech understanding than some of the postlingually deaf children. These authors also observed that speech perception and speech production continued to improve over the five-year follow-up period. The majority of the children showed open-set speech recognition. Waltzman et al.[6] analyzed the postoperative speech performance of 14 congenitally and prelingually hearing-impaired children whose age at the time of cochlear implantation was under three years. All the children developed good auditory skills. In addition, they concluded that congenitally and prelingually deaf children should receive a CI at an early age, because it will be more beneficial for the development of speech perception and because there was no difference in performance between the congenital and prelingual groups. It has become firmly established that the perceptual abilities of children with a CI continue to improve significantly over time. This is in contrast with profoundly deaf children who use conventional hearing aids, as they show plateau scores.[3,8]

Miyamoto et al.[26] compared the results of matched groups of CI users and hearing aid users. They showed that the mean score on speech perception tests 2.5 years after implantation was obviously better than the average score of children with conventional hearing aids with a hearing loss between 90 and 100 dB HL.

With regard to speech production, the earlier that cochlear implantation is performed in children, the better the intelligibility of their speech. Osberger et al.[27] reported that children with early onset of deafness who were implanted before the age of 10 years produced good intelligible speech, whereas a similar group of children who received a cochlear implant after the age of 10 years had the poorest speech intelligibility.

The auditory skills of 21 children with acquired profound deafness caused by meningitis, implanted in Nijmegen with a Nucleus multichannel system, were evaluated. These children were divided into four groups. The first group comprised children who became deaf between 0.3 and 2 years (n=5), the second group comprised children who became deaf between 2.1 and 3 years (n=6), and the third group comprised children who were older than 3 years at the onset of deafness (n=5). All these children had the electrode inserted over its full length. The fourth group comprised children who became deaf between 0.6 and 2.7 years of age (n=5) and who had only partial electrode insertion due to severe cochlear ossification. Preoperatively, the hearing thresholds were above 120 dB HL for all the children. The preoperative speech perception testing was carried out using high-gain postauricular hearing aids that the children had been wearing daily for at least one year. No significant speech perception using auditory presentation only (no lipreading) was found in any of the children.

After cochlear implantation, multiple speech performance measurements were conducted to determine the long-term benefits of rehabilitation with the CI. Two of the speech perception tests used were a picture-word identification test, which was a closed-set test of 12 monosyllables, and an open-set word recognition test, which consisted of 30 monosyllables. All test words were presented in auditory mode only, at normal conversational level of 70 dB.

Remarkable improvements in speech recognition performance were observed in the three groups of children with full insertion. Figure 3 shows the mean scores of these three groups on the picture-word identification test and Figure 4 shows the mean scores of the open-set word recognition test as a function of follow-up. There was constant improvement during the whole evaluation period. The most pronounced improvements were observed in the group of children who became deaf after the age of three years. Figure 4 shows that after three years of follow-up, the mean open-set speech recognition score lies between 55% to 75%. This is of great importance, because most children with such speech recognition abilities are able to develop normal oral-aural communication.[1,6,8] Several of these children can communicate with their relatives by telephone. These open-set speech scores are typically found in hearing impaired subjects with a hearing loss of 65 to 80 dB HL, using well-fitted conventional hearing aids.[8] So, the CI users perform with their CI after three years as well as well-fitted hearing aid users with a hearing loss between 65 and 80 dB HL. The age at onset of the deafness seems to affect the progress of the child (Figures 3 and 4). The figures suggest that the children who became deaf relatively earlier in life (group with onset of deafness between 0.3 and 2 years of age) and who had therefore little previous auditory experience, only showed delayed scores compared to children who acquired speech before they became deaf (group with onset of deafness after three years of age).

The result of the fourth group, with partial electrode insertion, was much poorer. The children in this group had scores below 10% on the open-set and closed word-identification tests, even after three years of follow-up.

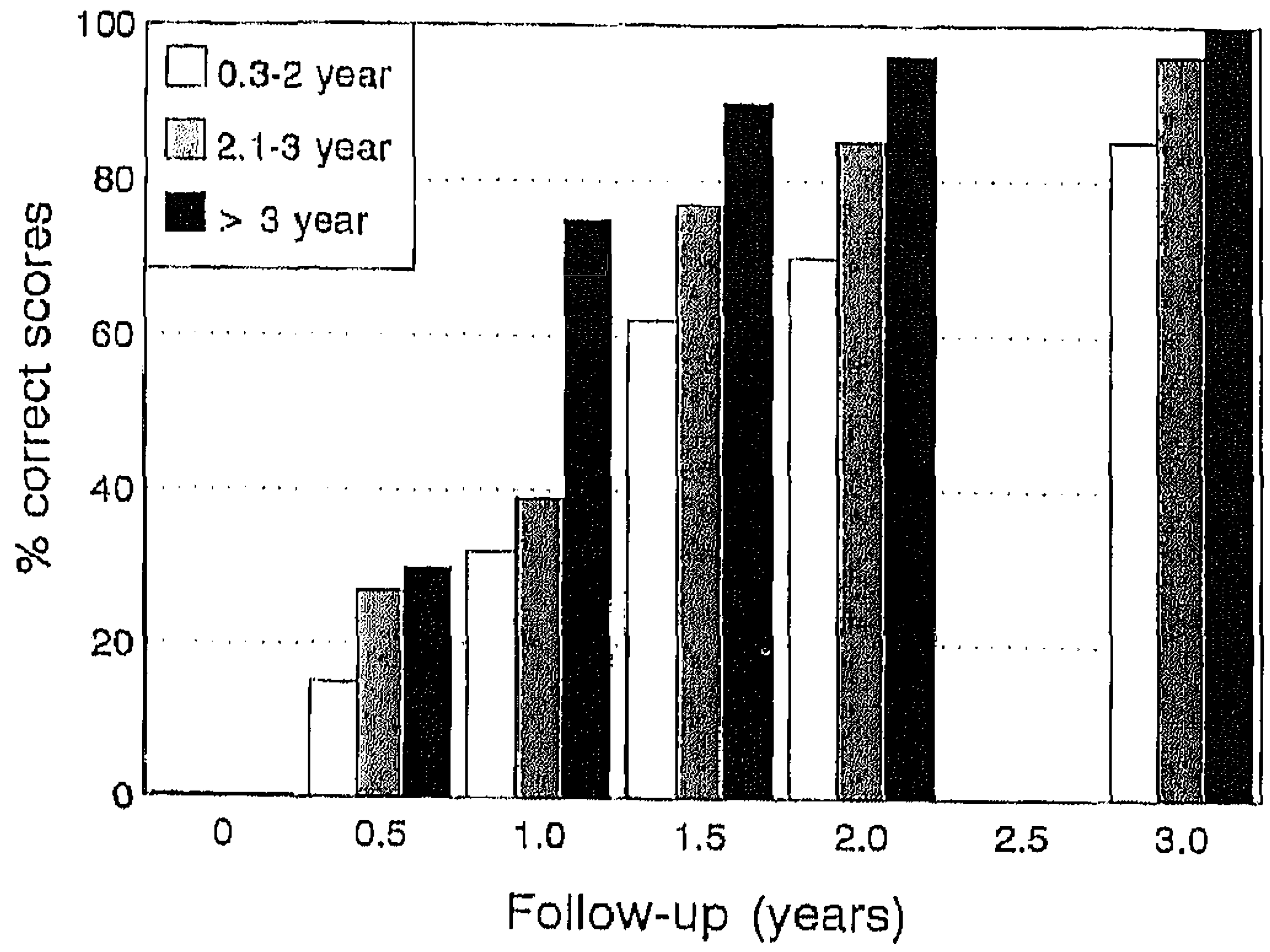

FIGURE 3. The mean score on the picture-word identification test as a function of follow-up of the three groups of children with full electrode insertion subdivided according to their age at the time of onset of deafness. The white bars indicate the mean scores of the children with acquired deafness between 0.3 and 2 years of age (n=5). The gray bars indicate the mean scores of the children with acquired deafness between 2.1 and 3 years of age (n=6) and the black bars indicate the mean scores of the children whose age at the onset of deafness was above 3 years (n=5). The values at a follow-up of 0 are the mean scores obtained before surgery with the children's own previous conventional hearing aids.

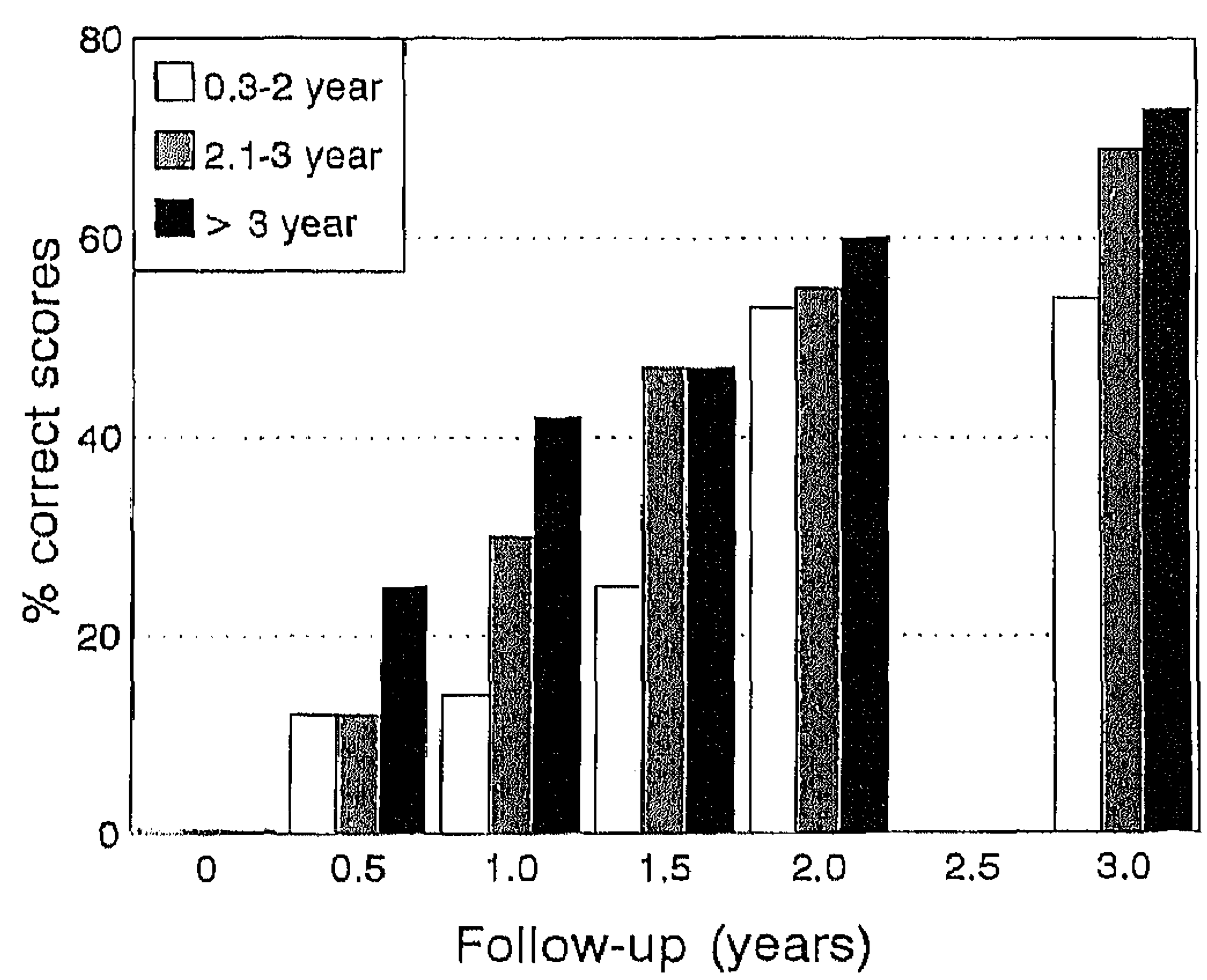

FIGURE 4. The mean score of the open-set speech perception test as a function of follow-up of the three groups of children with full electrode insertion subdivided according to their age at the time of deafness. The white bars indicate the mean scores of the children with acquired deafness between 0.3 and 2 years of age (n=5). The gray bars indicate the mean scores of the children with acquired deafness between 2.1 and 3 years of age (n=6) and the black bars indicate the mean scores of the children whose age at the onset of deafness was above 3 years (n=5). The values at a follow-up of 0 are the mean scores obtained before surgery with the children's own previous conventional hearing aids.

Therefore, results of a more basal speech perception test will be presented. The test used was a supra-segmental test in which the children only had to identify the number of syllables per word. Figure 5 shows the range (vertical lines) and the mean scores of the partial insertion group (white bars) and the average of the other three groups (gray bars). The values at the beginning of the follow-up are the mean scores obtained before surgery from the children with their own previous conventional hearing aids. This figure illustrates that the children with partial insertion are relatively poor performers, even after a three-year period of daily use. Therefore, to achieve the best result, the electrode array should be inserted into the cochlea over its full length. However, this cannot always be achieved in the case of severe ossification of the cochlea. Nevertheless, Kemink et al.[28] reported that the performance of children with partial insertion was comparable to that of children with full insertion. Our findings and those of other groups[11,29] showed that partial insertion leads to inferior long-term results.

In summary, our results and those of other studies[1,3,6] showed that significant (but variable) cochlear implantation outcomes can be achieved in children. The age at the onset of deafness and the method of insertion of the electrode array, either fully or partially inserted, play a role.

Conclusion

Nowadays, cochlear implantation is generally considered to be of significant value for pre- and postlingually profoundly deaf children. Adequate rehabilitation is most crucial for the children to maximize the benefits of cochlear implantation. Many studies reported that auditory performance with a CI varies among children. Until now, there has been no completely satisfactory explanation for this observation. However, performance seems to be best in children with a short duration of deafness, who acquired speech and language before their deafness occurred. Present results suggest that such children may acquire good speech perception and as a result, may develop normal aural-oral communication.

The outcome of partial insertion of multichannel electrode arrays is generally poorer than that of full insertion. This will depend on the position and number of active electrodes. It is highly questionable whether cochlear implantation in a severely obliterated cochlea is worthwhile.

Over the years, inclusion and exclusion criteria have gradually changed with growing knowledge. Generally, etiology and age at implantation do not seem to affect the post-implantation auditory performance. However, the earlier the implantation, the better the result, especially in prelingually deaf individuals. Nowadays, most CI teams

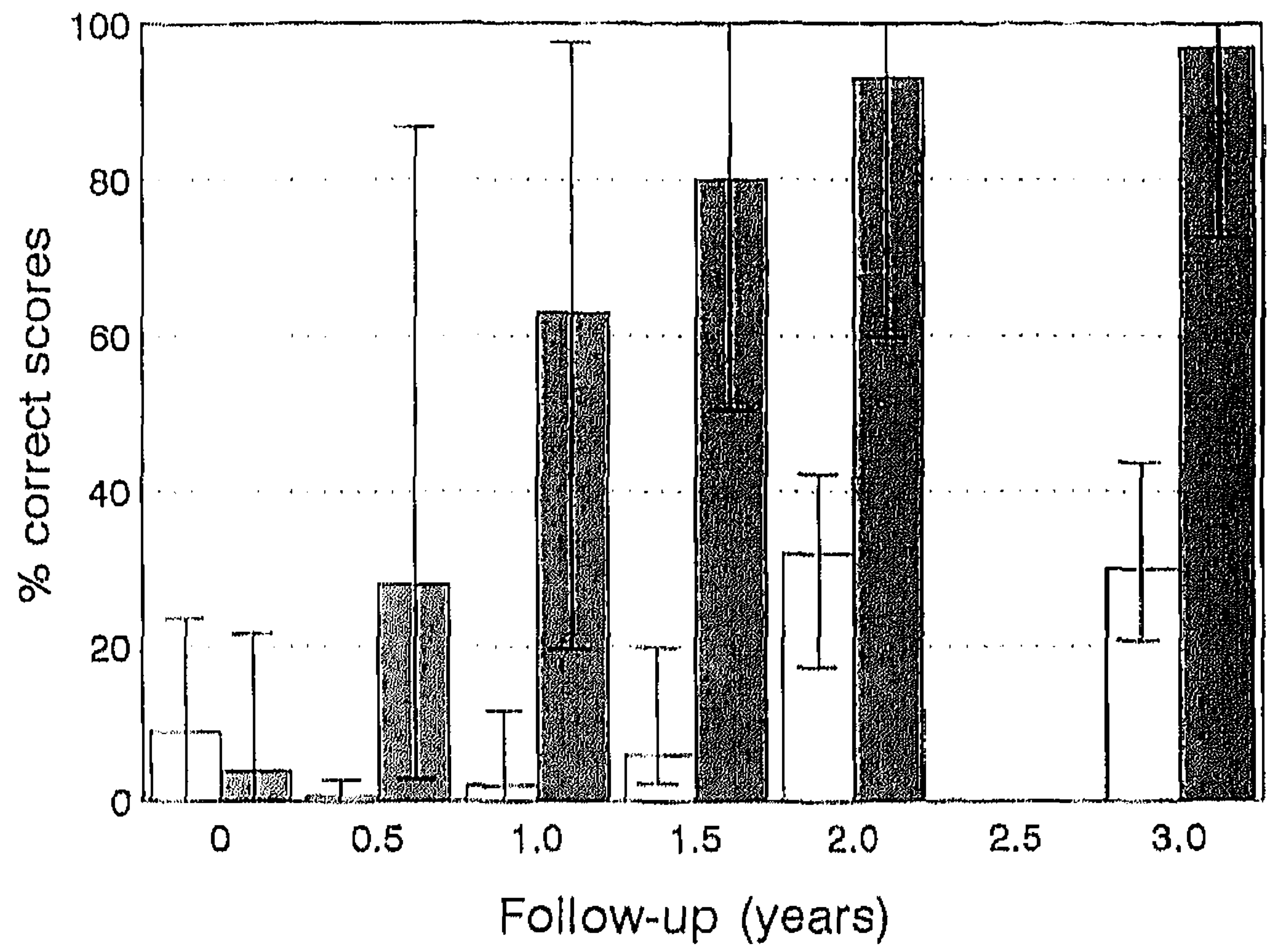

FIGURE 5. The mean score on the supra-segmental speech identification test and its range for the children with partial electrode insertion (white bar) and the averaged mean scores of the three groups of children with full insertion (gray bar) during a three-year follow-up period after cochlear implantation. The vertical lines indicate the range in the obtained scores. The values at follow-up of 0 are the mean scores obtained before surgery with the children's own conventional hearing aids.

only use a limited number of exclusion criteria. The most important exclusion criterion is the ability to utilize any residual hearing with well-fitted conventional hearing aids.

Owing to technological evolution and an increase in experience, the era of cochlear implantation is advancing rapidly. New techniques may enable wider groups of pre- and postlingually hearing-impaired children to benefit from cochlear implantation.

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