

# Cochlear Implantation in Adults with Prelingual Deafness. Part I. Clinical Results

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**Objectives/Hypothesis:** To examine the hypothesis that the newer generations of cochlear implants could provide considerable speech understanding to late-implanted, prelingually deaf adult patients. **Study Design:** Retrospective review. **Methods:** Speech perception scores of 103 patients with long-term prelingual deafness obtained from the recent clinical trials were compared with other previously published results. **Results:** Unlike patients receiving implants in early childhood, the audiologic performance of most patients with long-term prelingual deafness rapidly approached asymptotic levels within 1 year after implantation. Beginning at 3 months postimplantation, statistically significant improvement was noted in their speech perception scores compared with their preimplant levels. However, the average performance plateau achieved by this group of patients was significantly below the levels published for postlingual patients. No statistically significant differences were observed between the test scores at any testing interval among patients with different devices. **Conclusions:** Most patients with long-term prelingual deafness achieve their performance plateau within 1 year after implantation. The results suggest that patient characteristics, rather than device properties, are likely to be the major contributing factor responsible for the observed audiologic outcomes. **Key Words:** Cochlear implants, critical period, congenital deafness, long-term deafness.

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## INTRODUCTION

Over the last 2 decades, cochlear implantation has gradually been accepted as an effective treatment for many patients with profound sensorineural hearing loss (HL). Since the approval by the Food and Drug Adminis-

tration in 1984, the communicative benefits provided by cochlear implants (CIs) to postlingually deafened adults have been well documented.<sup>1</sup> It is now expected that most of these patients will score above 80% correct on high-context sentences, even without visual cues.

Despite objections from the deaf community, thousands of prelingually deaf children have also received CIs, and many have shown excellent outcome on a wide range of measures of hearing, speech, and language.<sup>2–4</sup> Using auditory inputs from their CIs, some prelingually deafened pediatric CI users have been able to acquire spoken language at a pace that is similar to normal-hearing children.<sup>5</sup> Unfortunately, the same level of benefits has not been observed in all children, and some pediatric patients derive only minimal benefits from their CIs.<sup>3</sup>

Several key findings have been consistently observed in the outcome measures of prelingually deafened CI users. Chief among them, age at implantation and duration of deafness were found to have the most significant impact on the postimplant outcome measures. Children who received an implant at an early age consistently performed better on all clinical tests than children who received implants at an older age, and adolescents and adults with long-term prelingual deafness derived the poorest benefits from their implants. A few early studies showed that these latter groups of patients could achieve only limited postimplant improvement in closed-set auditory perception, with some awareness of environmental sounds, but no open-set speech recognition ability.<sup>6,7</sup> Consequently, few patients with long-term prelingual deafness were considered good CI candidates.

As recent advances in CI technology continue to push the performance levels of most CI users to higher levels, interest in cochlear implantation for prelingually deafened adolescents and adults has been rekindled. Several recent studies have suggested that the latest implant technology could indeed provide some open-set speech perceptual abilities to these patients.<sup>8–11</sup> These conclusions, however, are based on analyses of results obtained with only a very small number of patients, and the data often showed enormous variability among individuals, making the true assessment of their effectiveness an exceedingly difficult task.

Prelingually deafened adults consist of a very heterogeneous group of patients. A substantial number of individual factors, such as etiology of deafness, communication mode, residual hearing, and educational experience,

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could all affect the postimplantation outcomes. Consequently, a valid assessment of the effectiveness of CIs would require a study with a large number of patients or a very well-controlled group of subjects. However, with no evidence proving their clinical efficacy, it is difficult to justify the cochlear implantation of a large number of such patients. In this article, we review all available published evidence in the CI literature on late-implanted prelingually deafened adults, and report new speech perception data obtained from an additional 103 patients from the recent CI clinical trials. The speech recognition scores of these patients were examined longitudinally over the 12-month clinical trial period to evaluate the effectiveness of cochlear implantation in providing auditory perceptual benefits.

## MATERIALS AND METHODS

Published evidence in the CI literature on late-implanted prelingually deafened adults was reviewed. In addition, speech perception data from the recent clinical trials were requested from three major CI companies (Advanced Bionics, Cochlear, and MedEL). The inclusion criteria included prelingual profound deafness (onset of deafness at < 3 years old) and implantation at older than 13 years of age, thus ensuring that these patients had at least 10 years of auditory deprivation. These criteria netted a total of 103 patients. The Nucleus devices were equipped with the SPEAK strategy, whereas the Clarion and MedEL devices made use of the CIS strategy.

## DATA AND RESULTS

Although a great deal of audiologic outcome data are available for postlingually deafened adults and prelingually deafened young children who have received CIs, few studies have focused on implantation outcomes of prelingually deafened adults. The earliest studies by Clark et al.,<sup>12</sup> Skinner et al.,<sup>7</sup> and Waltzman et al.<sup>6</sup> were mainly case reports of a few selected patients. The subjects in these studies all showed uniformly poor open-set speech perceptual skills, even though some improvements were noted on closed-set speech recognition tests over time after implantation. However, it should be noted that all of these earlier studies were conducted with speech processing strategies that are now deemed obsolete, and thus the results may not apply to the users of the newer generation of CIs.

Fryauf-Bertschy et al.<sup>3</sup> published the first large series of longitudinal data that included CI patients with prolonged deafness. However, their results were obtained mainly from children. Only 4 of the 34 prelingually deafened patients in their study received implants after the age of 12. Nonetheless, some inferences can be drawn from the clear trends observed in their data. A summary of the most representative results in that article using PB-K word scores is replotted in Figure 1. The PB-K test is an open-set word recognition test that is routinely used to measure speech perception skills without visual enhancements because of lipreading. The patients in that study were divided into four groups on the basis of their age at implantation. The results showed a strong influence of duration of deafness before implantation on the patterns of long-term improvement observed in these patients. Significant differences were observed in performance between children receiving implants before age 5 and those receiving implants after age 5 at the 3- and 4-year test intervals. Qualitatively, the results showed that the older the patient was at the time of implantation, the slower the

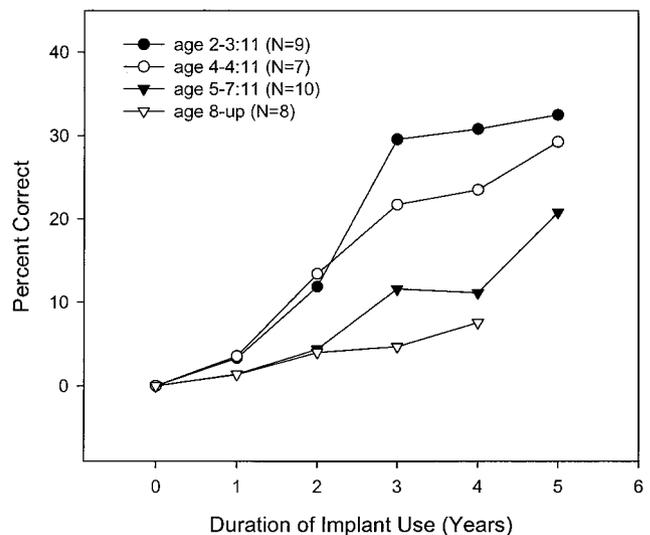


Fig. 1. A summary of the PB-K word test results published by Fryauf-Bertschy et al.<sup>3</sup> Average test scores of the prelingually deafened cochlear implant patients grouped by their age at implantation.

rate of improvement and the lower the average scores—even after 4 or 5 years of implant use. For the group receiving implants after 8 years of age ( $n = 8$ ), only minimal open-set word recognition ability was demonstrated. Although the data set did not specifically include adult patients, the general trend suggests that prelingually deafened adults would receive minimal to no auditory benefits from a CI. It should be noted that all the patients in the Fryauf-Bertschy et al.<sup>3</sup> study used the older F0F1F2 or the MPEAK strategy in their Nucleus processors.

A similar trend was also present in the study reported by Manrique et al.,<sup>4</sup> which assessed patients who used the newer SPEAK strategy or the MPEAK strategy. In their study, 98 prelingually deafened patients were followed over a period from 1 to 6 years after implantations, with another 58 postlingually deafened patients serving as a comparative group. Twenty-three of the 98 prelingually deaf patients had greater than 14 years of profound deafness before implantation, and another 17 patients received implants between 11 and 14 years of age. Figure 2 shows a summary of their longitudinal follow-up data. The average percent correct scores of the patients are plotted as a function of the duration of their implant use. For the closed-set recognition test of daily words delivered under auditory presentation (Fig. 2A), the prelingually deafened patients in all five age groups appeared to achieve a level close to their asymptotic level after only 1 to 2 years postimplantation.

For the early implanted children and for postlingual adults, ceiling effects may have masked any possible improvement over time in their auditory skills. With the more difficult open-set recognition test of bisyllabic words (Fig. 2B), the ceiling effects were reduced, and the results showed continuing improvement in their auditory development 3 to 5 years after implantation. For the older patients, however, the ceiling effects did not apply, and yet no significant changes were observed after the first year of implant use in either the open- or the closed-set tests. In general, the results of Manrique et al.<sup>4</sup> support the earlier

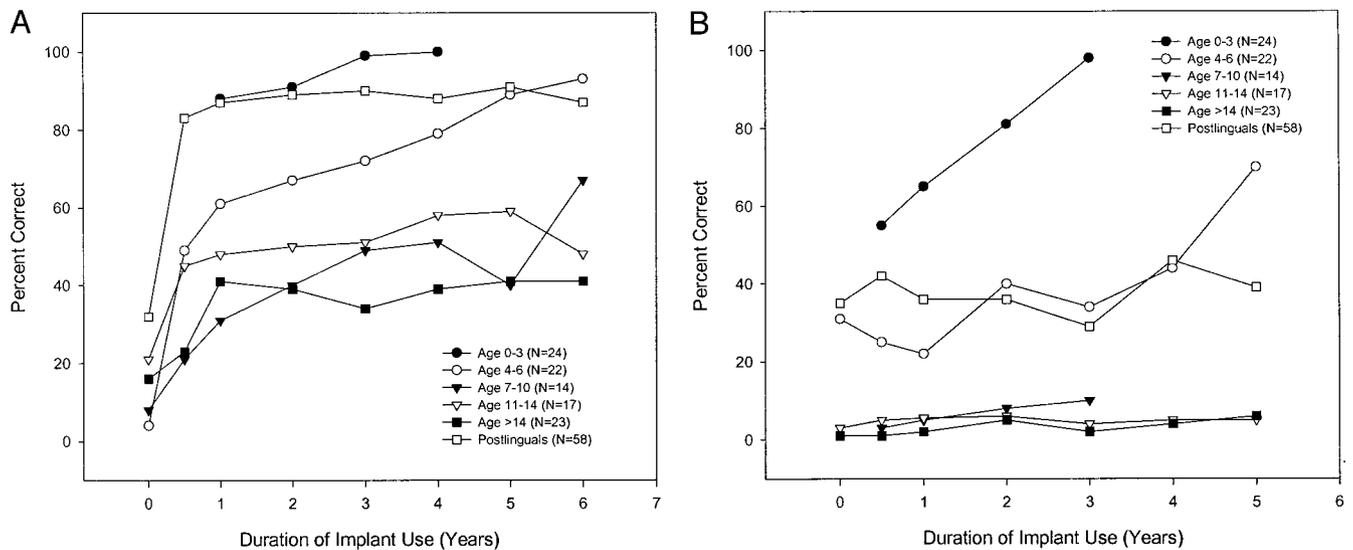


Fig. 2. A summary of the longitudinal follow-up data reported by Manrique et al.<sup>4</sup> Average scores of prelingually deaf patients grouped by their age at implantation. Results of 58 postlingually deafened adults are also included for comparison purposes. The tests are (A) daily words test (a closed-set test) and (B) bisyllabic words test (an open-set test).

findings that postimplant performance is inversely related to the duration of deafness before implantation. Prelingually deaf patients implanted at 11 to 14 years of age, and after 14 years of age, showed virtually no open-set speech-perception understanding even after 5 years of implant use.

In another study, Snik et al.<sup>13</sup> attempted to address the issue of the upper age limit for cochlear implantation in congenitally deaf subjects by assessing the performance of 12 patients longitudinally. The patients in their study received implants from age 4 to 33 years. The authors combined the test scores from a battery of speech perception tests, which included word discrimination, vowel discrimination, supra-segmental speech identification, word identification, and open-set speech recognition tests, into one single measure called the “equivalent HL.”<sup>14</sup> The combined measure was obtained by matching the CI patient’s overall performance on the test battery with an age-matched reference group of severely and profoundly hearing-impaired children with conventional hearing aids. Their results are summarized in Figure 3, which plots the average equivalent HL levels of patients in three different age groups over the 3-year follow-up period. The results showed that adolescents (implanted at 11–13 years old) and adults all reached a plateau in performance within their evaluation period (2–3 years), with most progress leveled off between 6 to 12 months after implantation. In contrast, the 4 to 8 year-old group continued to display improvement throughout their follow-up period, without reaching an asymptotic level. On the basis of these results, the authors concluded that implantation of congenitally deaf patients during or after puberty offers only limited benefit.

More recently, several articles have reported more promising results with the use of newer generations of implant technology.<sup>9–11,15</sup> These studies found varying degrees of improved open-set speech understanding in children and adults with long-term congenital deafness. Osberger et al.<sup>15</sup> reported some measurable open-set speech understanding in

prelingually deaf children using the Clarion CIs. However, their study focused on children older than 5 years of age, and thus the duration of deafness in these patients was relatively short compared with the previous studies of prelingually deafened adults. In 1999, Waltzman and Cohen<sup>9</sup> reported case studies of five patients (age at implantation ranged from 10.5–20 years old) with moderate improvement in speech understanding as measured by a variety of clinical tests. More patients were included in the subsequent studies by Schramm et al.<sup>11</sup> (9 adults and 6 children) and Waltzman et al.<sup>10</sup> (14 adults and 35 children). Both articles reported substantial individual variability in performance, but some open-set speech perception was demonstrated by several

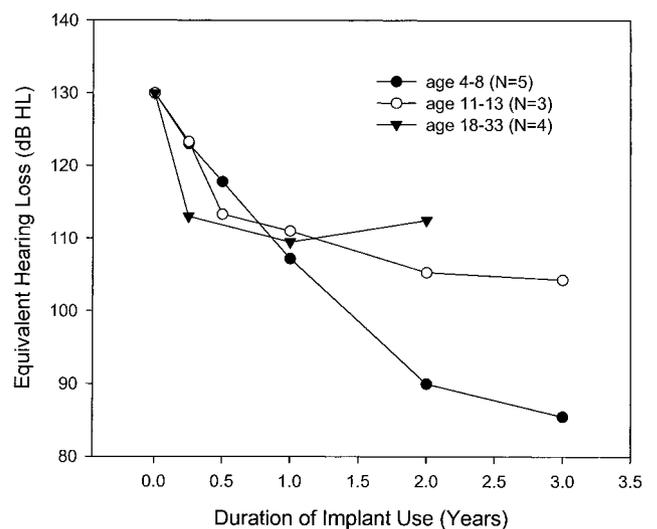


Fig. 3. Postimplant performance results adapted from data reported by Snik et al.<sup>13</sup> Each plot represents the average “equivalent hearing loss” (see text for details) of prelingually deaf patients grouped according to their age at implant.

patients, as measured by their postimplant improvement in open-set word and sentence recognition scores. The patients in both of these studies received either the Clarion or Nucleus implant systems, using the latest CIS, SPEAK, or ACE coding strategies. The authors suggested that the improved technology was the major factor contributing to the improved implantation outcomes. Unfortunately, none of the articles reported any longitudinal data on their patients, making a comprehensive evaluation of their outcome measures difficult.

To assess the hypothesis that the latest CI technology was the primary factor responsible for the improved audiologic outcomes of these prelingually deafened patients, speech perception data from the recent clinical trials were requested from three major CI companies (Advanced Bionics, Cochlear, and MedEL). The data include speech perceptual scores of 103 patients over the 12-month clinical trial period.

Figure 4 shows the average percent correct scores and standard errors for three different open-set speech perception tests over the 12-month clinical trial period. For comparison purposes, Figure 4 also includes previously unpublished data from a group of postlingually deafened adult MedEL Combi-40+ users tested at the Indiana University School of Medicine. The open-set perceptual tests included the CUNY sentences in quiet (Nucleus and MedEL patients), the HINT sentences in quiet (MedEL and Clarion patients), and the CNC monosyllabic words (Nucleus and MedEL patients) tests. The CUNY and HINT are open-set tests of sentence recognition. The CNC is a more difficult open-set word recognition test.

Figure 5 shows the individual HINT scores obtained from the Clarion and MedEL prelingually deaf patients. The scores shown in this figure illustrate the enormous variability in test scores typically observed among individual patients. Although the average scores at 6 and 12 months postimplant were approximately 20%, several individual patients were able to achieve much higher scores (40–100%).

## DISCUSSION

Several important conclusions can be drawn from an examination of the clinical trial data shown in Figure 4. First, the results demonstrate that the overall performance

of the three different implants are remarkably similar. No statistically significant differences were observed between the test scores at any testing interval for the prelingually deafened patients ( $P \geq .2$  on all paired-samples *t* tests). Such similarity among vastly different processing devices suggests that patient characteristics, rather than device properties per se, are likely to be the major contributing factor responsible for the observed outcome measures. Second, beginning at 3 months after implantation, statistically significant improvement ( $P \leq .001$  on paired-samples *t* tests) was noted in the scores of all three speech perceptual tests compared with their preimplant levels. However, the average performance plateau achieved by these patients was still significantly below the findings published for postlingual adult patients or prelingually deaf children with short-term deafness.<sup>1,2,5–12</sup> This finding was observed despite the use of the latest speech processors and coding strategies. Finally, most prelingually deafened patients rapidly approached their asymptotic level of performance within 12 months after implantation. This finding is consistent with the data reported in the earlier studies by Manrique et al.,<sup>4</sup> Fryauf-Bertschy et al.,<sup>3</sup> and Snik et al.,<sup>13</sup> but differs from previous findings, suggesting that prelingually deaf patients may require a long period of time to achieve their maximal audiological benefit.<sup>10</sup>

The tremendous variability observed in the clinical trial data may be attributable to the wide-ranging characteristics of the patient populations. Because clinical trials are routinely conducted at several medical centers, it was unavoidable that the patient population would be quite heterogeneous in nature (e.g., disease etiologies, communication modes, and educational experiences). Indeed, Waltzman et al.<sup>10</sup> attributed the improved patient performance in their study in part to the significant number of oral communicators enrolled in that study. Similar conclusions were reached by Osberger et al.<sup>15</sup> The precise implications of oral communication versus total communication (i.e., using sign and speech) on the performance of CI patients will be explored in greater detail in Part II of this series. In addition, several patients with some residual hearing may have had some auditory experience through their hearing aids before cochlear implantation.

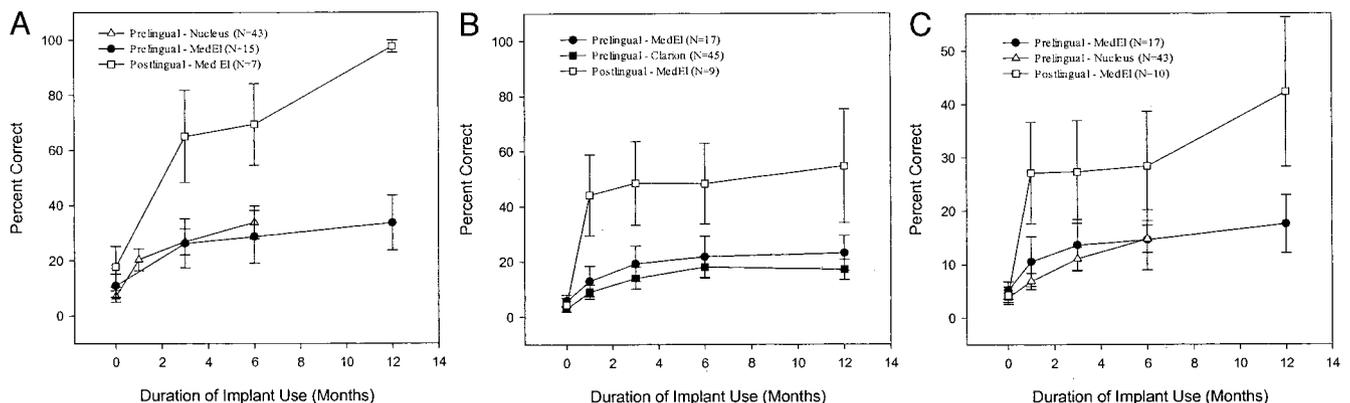


Fig. 4. Average test scores and standard errors of prelingually deaf adult cochlear implant (CI) patients obtained from clinical trials of the three major CI companies. For comparison purposes, the figure also includes scores obtained from postlingually deafened MedEL Combi-40+ adult CI users. The tests are (A) CUNY sentences in quiet, (B) HINT sentences in quiet, and (C) CNC mono words test.

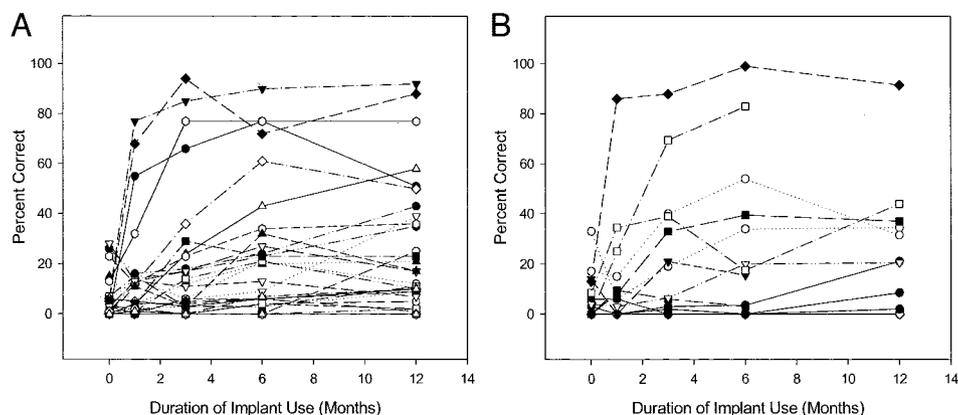


Fig. 5. Individual HINT scores of (A) Clarion and (B) MedEl patients. Data from 46 Clarion and 17 MedEl users. The HINT test was performed in quiet.

These factors could explain the results of Schramm et al.,<sup>11</sup> in which many of their subjects had significant residual hearing before implantation.

## CONCLUSIONS

In this article, the postimplant performance data of prelingually deaf patients with long-term deafness were obtained from recent clinical trials and other previously published studies. Several general trends were observed after an examination of the outcome data. First, the longitudinal audiologic outcome measures clearly indicate a relationship between the duration of auditory deprivation before implantation and the patient's eventual audiologic performance plateau. Even with the latest CI technology, a gradual lowering of the performance plateau was observed as the age at implantation was delayed. If receiving implants after age 12, most profoundly deaf patients achieved only very limited closed-set speech perception and minimal to no open-set speech understanding, indicating the presence of a sensitive period for cochlear implantation. Second, prelingually deaf patients with long-term deafness reached their performance plateaus significantly earlier than patients receiving implants during their early childhood. Adult prelingual patients typically reach their performance plateau within 6 months to 1 year. Third, substantial performance variability was observed among individuals. A few CI users were able to score significantly above chance even in open-set speech perception tests. However, the number of such patients was very small, and they represent the exception rather than the rule. And finally, there were no significant differences noted in efficacy among the Nucleus, Clarion, and MedEl CIs at any of the postimplant intervals. Taken together, the pattern of results suggests that patient characteristics, rather than implant device properties, are likely to be the major contributing factors that are responsible for the observed outcome measures. In an effort to better understand the features that define the sensitive period of cochlear implantation, we will discuss in Part II of this series the possible anatomic and physiologic correlates of the observed performance limitations and outcome variabilities associated with long-term prelingual deafness.

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