Cochlear Implantation in Prelingually Deafened Adolescents

Daniel M. Zeitler, MD; Abbas Anwar, BS; Janet E. Green, AuD; James S. Babb, PhD; David R. Friedmann, MD; J. Thomas Roland Jr, MD; Susan B. Waltzman, PhD

Objectives: To determine the efficacy of cochlear implantation (CI) in prelingually deafened adolescent children and to evaluate predictive variables for successful outcomes.

Design: Retrospective medical record review.

Participants: Children aged 10 to 17 years with prelingual hearing loss (mean length of deafness, 11.5 years) who received a unilateral CI (mean age at CI, 12.9 years).

Intervention: Unilateral CI.

Main Outcome Measures: Standard speech perception testing (Consonant-Nucleus-Consonant [CNC] monosyllabic word test and Hearing in Noise [HINT] sentence test) was performed preoperatively, 1 year postoperatively (year 1), and at the last follow-up/end of the study (EOS).

Results: There was a highly significant improvement in speech perception scores for both HINT sentence and CNC word testing from the preoperative testing to year 1 (mean change score, 51.10% and 32.23%, respectively; $P < .001$) and from the preoperative testing to EOS (mean change score, 60.02% and 38.73%, respectively; $P < .001$), with a significantly greater increase during the first year ($P < .001$). In addition, there was a highly significant correlation between improvements in performance scores on the CNC word and HINT sentence speech perception tests and both age at CI and length of deafness at the year 1 testing ($P \leq .009$) but not from the year 1 testing to EOS testing. Adolescents with progressive deafness and those using oral communication before CI performed significantly better than age-matched peers.

Conclusions: Adolescents with prelingual deafness undergoing unilateral CI show significant improvement in objective hearing outcome measures. Patients with shorter lengths of deafness and earlier age at CI tend to outperform their peers. In addition, patients with progressive deafness and those using oral communication have significantly better objective outcomes than their peers.

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SENSORINEURAL HEARING LOSS (SNHL) affects 1 to 3 of every 1000 children born in the United States and other developed countries, with the rates likely higher in less developed nations.[1-4] Approximately 4000 infants are born each year in the United States with bilateral severe to profound hearing loss,[5,6] defined as hearing thresholds above 60 dB in each ear, and 4 infants per 10 000 births are born profoundly deaf in both ears (thresholds >80 dB).[7,8]

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Cochlear implantation (CI) has become an accepted treatment paradigm for individuals 12 months or older who have bilateral severe to profound SNHL. The efficacy of CI in the rehabilitation of hearing relies on the conversion of acoustic sound to a series of electrical impulses that stimulate the auditory nerve via a surgically implanted electrode in the cochlea. Outcomes in young, prelingually deafened patients undergoing CI have been uniformly excellent.[9-11] With the implementation of universal newborn hearing screening in some countries, early diagnosis of bilateral severe to profound SNHL has markedly improved, allowing for early intervention in many of these infants. However, many infants with congenital bilateral severe to profound SNHL are not identified early in life owing to the unavailability of newborn screening, false-positive screening results, or a lack of access to health care. Similarly, a large subset of patients have progressive, bilateral severe to profound SNHL beginning after infancy owing to a variety of causes, such as otosclerosis, enlarged vestibular aqueduct syndrome, labyrinthitis ossificans, ototoxic medication administration, Meniere disease, idiopathic sudden SNHL, and genetic causes, including pro-

Author Affiliations: Denver Ear Associates, Englewood, Colorado (Dr Zeitler); and Department of Otolaryngology–Head and Neck Surgery, New York University Langone Medical Center (Drs Green, Friedmann, Roland, and Waltzman), and Department of Radiology (Dr Babb), New York University School of Medicine, New York, New York. Mr Anwar is a medical student at the New York University School of Medicine.

gressive nonsyndromic hearing loss. These patients, therefore, become candidates for CI as adolescents.

For adolescents deafened at an early age who do not undergo CI early in life, speech perception scores after implantation almost universally improve but are worse when compared with age-matched control peers undergoing implantation at an earlier age,[12-15]; this duration of deafness before CI has been shown to be negatively correlated with the ability to understand and use spoken language.[16-18] Initial hesitation among the parents of many deaf adolescents to subject their child to CI in the interest of waiting for long-term data to emerge, "saving the ear" for new technological advances, or preventing sweeping changes in the social, academic, and personal lives of their children has given way to acceptance and understanding of the CI procedure and the profound effect it can have on the lives of adolescents.

Adolescents represent a unique subgroup of candidates for CI that has not been extensively examined. Although early reports[19-21] have shown that adolescents benefit from unilateral CI, few studies have examined the effects of factors, such as age at CI, duration of deafness, cause of SNHL, and mode of communication, on speech perception outcomes in this population. The purposes of this study were to determine the efficacy of CI in prelingually deafened children and adolescents aged 10 to 17 years and to evaluate possible predictive variables.

METHODS

SUBJECTS

We performed an institutional review board–approved retrospective review of all children and adolescent patients aged 10 to 17 years with bilateral severe to profound SNHL who derived minimal to no benefit from conventional amplification and underwent unilateral CI from 1986 through 2009. The study population included subjects with congenital deafness and those with postnatal, prelingual progressive or sudden deafness, defined as children with severe to profound hearing loss before age 3 years regardless of hearing status at birth or cause of deafness. Patients with postlingual deafness were excluded from the study. We identified 67 patients who met inclusion criteria. Thirty-three subjects (49%) had congenital deafness of unknown etiology. Other causes of deafness included progressive hearing loss (n=11), meningitis (n=6), enlarged vestibular aqueduct syndrome and/or Mondini malformation (n=5), ototoxic effects (n=4), Waardenberg syndrome (n=3), cytomegalovirus infection (n=3), and high fever (n=2). These diagnoses were used to divide the cohort into 5 main groups according to the cause of hearing loss: (1) congenital (including syndromic) (41 patients [61%]); (2) idiopathic progressive (11 [16%]); (3) hearing loss due to ototoxic medication (4 [6%]); (4) meningitis (6 [9%]), and (5) other, including cytomegalovirus infection and fever (5 [7%]). The mean age at CI was 12.9 years (range, 10-17) and the mean length of deafness was 11.5 years (range, 0.25-17). All patients were English speaking, and patients with a diagnosis of mental retardation or other associated disabilities were excluded from the study. Collected data included length of deafness, age at the time of CI, ear undergoing CI, device, and mode of communication. Forty-seven of 67 patients (70%) attended mainstream schools, and an additional 18 of 67 (27%) were schooled within self-contained classroom settings with special education resources. Only 2 patients (3%) were homeschooled. Fifty of the adolescents (75%) used oral communication before CI, 7 (10%) used manual communication, and 10 (15%) used total or cued communication. Fifty-four devices were Cochlear Corporation (Sydney, Australia), 12 were Advanced Bionics (Valencia, California), and 1 was Med-El (Innsbruck, Austria). All had full insertion of their devices, and there were no postoperative complications. Mean follow-up was 60 months (range, 12-168). Eight patients were unavailable for follow-up after CI (moved, changed centers, international patients, etc) and were excluded from analysis. Five patients were nonusers of their device and were excluded from analysis. Fifty-four patients with at least 1 year of follow-up after CI were included in the data analysis.

METHODS

Pure-tone and speech audiometry and open-set speech perception testing designed to assess word and sentence recognition were performed preoperatively with the subjects wearing their conventional amplification devices and at 3, 6, and 12 months and then yearly after initial CI device stimulation. Postoperative testing was performed using the same materials so that each subject could be self-compared, allowing for a single-subject repeated-measures design. Scores reported included preoperative, year 1 (after 1 year of device use), and end of the study (EOS; requiring ≥2 years of device use). The score at the final evaluation was treated as missing for subjects who did not undergo evaluation at least 2 years after CI. Changes for these subjects are completely characterized by the preoperative to year 1 change score. The changes in word and sentence scores were computed for each subject as the score at the later time minus the score at the earlier time; a positive change reflects an increase in score.

The Consonant-Nucleus-Consonant (CNC) monosyllabic word test and the Hearing in Noise (HINT) sentence test in quiet and noise were performed preoperatively and postoperatively on all subjects. The CNC test consists of ten 50–monosyllabic word lists that are scored as percentages of phonemes and words correct. Each list represents the phonemic balance of the English language, and the distribution is matched across lists. The HINT test is made up of 250 sentences divided into 25 phonetically balanced lists of 10 sentences each and can be administered in quiet or with competing noise at a signal to noise ratio of +10 dB. All tests were administered in a standard soundproof suite using recorded material presented at 60-dB sound pressure level.

Study end points (CNC word score and HINT sentence score) were summarized in terms of the mean (SD) and the median (interquartile range); the latter is provided because it is generally deemed appropriate when nonparametric analyses are used. A Wilcoxon matched-pairs signed rank test was used to assess whether there was a pre-CI to post-CI change in each study end point. Spearman rank correlations were used to characterize the association of length of deafness, length of CI use, and age at CI with the change in each end point. We used Mann-Whitney and Kruskal-Wallis tests to evaluate covariance among nonparametric populations (only significant independent subject-level factors were retained in each final model). For each test, the error variance was allowed to differ across comparison groups to eliminate the unnecessary assumption of variance homogeneity. All reported P values are 2 sided, and $P < .05$ was considered statistically significant.

RESULTS

Table 1 shows the mean and median percentage of correct preoperative, year 1, and EOS scores for the 2 test end points, namely, the HINT sentence test and the CNC

Table 1. Preoperative to EOS Change in the CNC Word and HINT Sentence Scores

	HINT Sentence Change Score			CNC Word Change Score		
Period	Mean (SD)	Median (IQR)	*P* Value[a]	Mean (SD)	Median (IQR)	*P* Value[a]
Preoperative to year 1	51.10 (37.85)	54 (8-86)	<.001	32.23 (28.19)	30 (4-53)	<.001
Preoperative to EOS	60.02 (37.11)	71 (22-93)	<.001	38.73 (27.97)	38 (18-63)	<.001
Year 1 to EOS	6.77 (16.05)	2 (0-12)	.008	9.30 (15.86)	6 (0-24)	<.001

Abbreviations: CNC, Consonant-Nucleus-Consonant; EOS, end of study; HINT, Hearing in Noise; IQR, interquartile range.
[a] Calculated from an exact paired-sample Wilcoxon signed rank test of whether the score increased.

monosyllabic word test. There was a highly significant increase in mean and median scores for both tests during each period studied. There was also a significantly greater increase in the CNC and HINT scores during year 1 than from year 1 to EOS, implying that improvement in objective measures shows a significant tendency to slow down over time (**Table 2**). After Spearman rank correlation testing, there were highly significant and negative correlations between CNC word and HINT sentence scores with the age at implantation and length of deafness for both the preoperative to year 1 scores and preoperative to EOS scores (data not shown). When scores were adjusted for the length of follow-up, these correlations remained statistically significant (**Table 3**). For both the HINT sentence and CNC word scores, the correlation of the year 1 improvement with length of deafness was significant when adjusted for age ($P<.001$ and $P=.01$, respectively), whereas the correlation of scores with age at implantation was not significant when adjusted for deafness duration ($P=.19$) (data not shown), implying that the greater improvement among younger subjects is mainly explained by the fact that these subjects tended to have been deaf for a shorter time.

Figure 1 shows the mean performance change score on the HINT sentence and CNC word tests stratified by the cause of deafness. These results are also summarized in **Table 4**. Kruskal-Wallis analysis showed that the cause of deafness had no significant effect on the improvement in HINT sentence score during any period measured ($P>.17$). Cause of deafness did show a significant effect on the improvement in CNC word score during year 1 ($P=.007$) and across the study period as a whole (preoperative to EOS) ($P=.01$); subjects with progressive deafness performed significantly better than did patients with postmeningitic deafness or congenital deafness (**Table 5**). There were no significant differences between any of the other etiological variables. The cause of deafness had no significant effect on improvement in CNC word score after the first year; therefore, the significant effect of cause on CNC word score improvement can be explained by the fact that overall improvement occurred mainly in the first year. In analysis of covariance testing using rank change in the CNC word score as the dependent variable, the effect of cause of deafness on year 1 improvement in CNC word score failed to retain significance (P .70) when adjusted for length of deafness. This implies that the effect of etiology was mainly due to differences among patients with various causes in terms of length of deafness before CI.

Table 2. Difference Between the Changes During Year 1 and From Year 1 to EOS

	Change Score		
Period	Mean (SD)	Median (IQR)	*P* Value[a]
Preoperative to year 1	46.95 (45.97)	65 (0-92)	<.001
Year 1 to EOS	19.80 (35.44)	10 (0-41)	.002

Abbreviations: EOS, end of study; IQR, interquartile range.
[a] Calculated from an exact paired-sample Wilcoxon signed rank test to assess whether the outcome showed a greater increase during year 1 than during the remainder of follow-up.

Figure 2 shows the mean performance change score on the HINT sentence and CNC word tests stratified by pre-CI mode of communication (ie, manual, oral, or total). These results are also summarized in **Table 6**. Kruskal-Wallis analysis showed that the primary mode of communication before CI had a significant effect on mean improvement during the first year of CI device use (preoperative to year 1) for oral communicators compared with manual communicators (CNC word score, $P<.001$; HINT sentence score, $P<.001$) and total communicators (CNC word score, $P=.03$; HINT sentence score, $P=.03$). There was a significant improvement in the CNC word and HINT sentence change scores during the entire study period (preoperative to EOS; $P=.03$). There was no significant improvement in either objective outcome measure after the first year (year 1 to EOS, $P=.29$ [sentence] and $P=.80$ [word]). There was no significant difference in scores for either test between total communicators and manual communicators ($P=.09$ [sentence] and $P=.11$ [word]). Analysis of covariance testing using rank change in CNC word score as the dependent variable demonstrated that the effect of mode of communication on year 1 CNC word and HINT sentence score improvements retained statistical significance ($P<.001$) when adjusted for length of deafness (data not shown).

COMMENT

The results of this study are consistent with previously reported data[12,19-22] showing significant improvement in speech perception skills in prelingually deafened adolescents after unilateral CI. However, the adolescents with shorter length of deafness and earlier age at CI (within the adolescent age range) showed significantly greater im-

Table 3. Association of the Improvement in the CNC Word and HINT Sentence Scores With Patient Variables on the Relevant Test at Each Time Point Adjusted for Length of Follow-up[a]

	HINT Sentence Score		CNC Word Score	
Period by Patient Variable	**Spearman Correlation, *r***	***P* Value**	**Spearman Correlation, *r***	***P* Value**
Year 1				
Age at CI	**−0.36**	**.009**	**−0.51**	<**.001**
Length of deafness	**−0.51**	<**.001**	**−0.65**	<**.001**
Length of CI device use	−0.16	.27	−0.06	.67
Preoperative score	−0.10	.50	0.21	.14
Preoperative to EOS				
Age at CI	**−0.33**	**.04**	**−0.46**	**.003**
Length of deafness	**−0.45**	**.004**	**−0.63**	<**.001**
Length of CI device use	−0.24	.13	−0.23	.15
Preoperative score	−0.18	.27	−0.02	.89
Year 1 to EOS				
Age at CI	−0.10	.53	−0.04	.79
Length of deafness	−0.05	.74	0.06	.71
Length of CI device use	−0.19	.24	−0.15	.35
Preoperative score	0.13	.44	−0.12	.46

Abbreviations: CI, cochlear implantation; CNC, Consonant-Nucleus-Consonant; EOS, end of study; HINT, Hearing in Noise.
[a] Significant values are in boldface type.

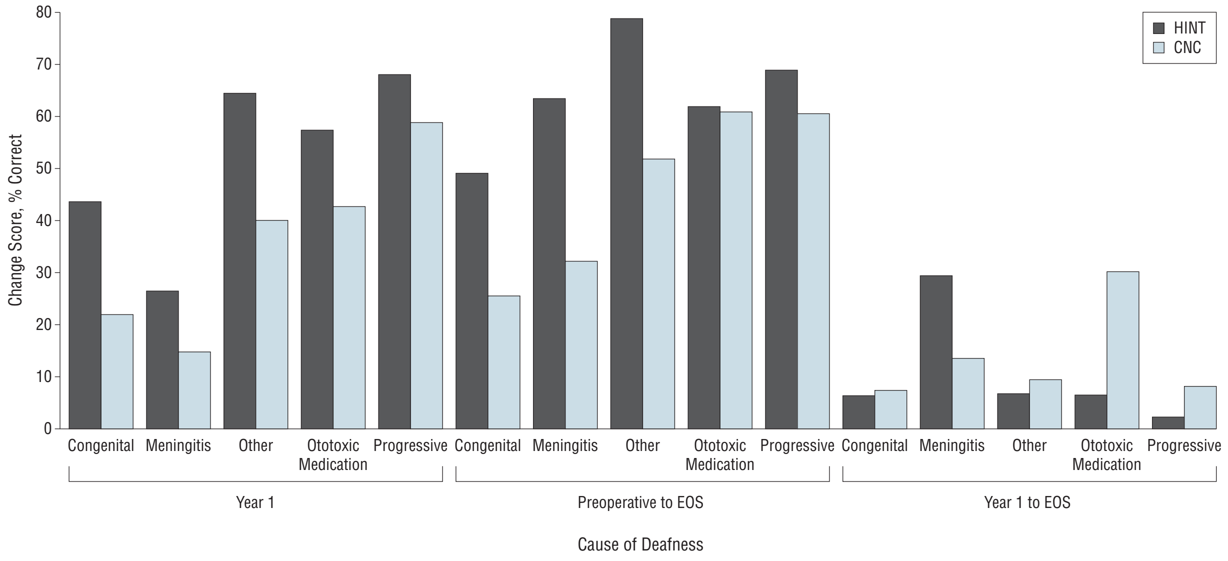

Figure 1. The mean change score for the Hearing in Noise (HINT) sentence test and Consonant-Nucleus-Consonant (CNC) monosyllabic word test in the 3 study periods analyzed stratified by cause of deafness. EOS indicates end of study.

provement in word and sentence testing than did those undergoing CI in late adolescence. Because there was no correlation of age at CI and length of deafness with improvement in speech understanding after year 1, it seems that most of the gains occurred in the first year after CI.

Neurophysiological data[23,24] have suggested that there may be a sensitive period for central auditory development in humans that ends at 7 years of age, with children who undergo CI after 7 years of age showing abnormal brain responses to auditory input and poorer language skills, even after several years of experience with a CI device. In addition, others have explained that this observed phenomenon may be related to cortical plasticity, whereby disorganized cooperation between bottom-up and top-down processes leads to colonization of the auditory cortex by other sensory modalities during critical periods of central nervous system development.[25] Although cortical reorganization may in fact occur in children deprived of early auditory stimuli, our results clearly demonstrate significant objective benefit in children undergoing CI after this so-called sensitive period.

Cause of deafness had no significant effects on the improvement in HINT sentence scores during any period. However, cause of deafness had a significant effect on the improvement in CNC word scores during year 1 of CI device use only, with no significant effects between the

Table 4. Change in CNC Word and HINT Sentence Scores Stratified by Cause of Deafness

Period by Cause of Deafness	HINT Sentence Change Score		CNC Word Change Score	
	Mean (SD)	Median (IQR)	Mean (SD)	Median (IQR)
Year 1				
Congenital	43.12 (39.39)	50 (0 to 85)	21.04 (22.58)	20 (0 to 42)
Meningitis	25.75 (21.39)	26 (5 to 46)	14.00 (11.43)	14 (4 to 24)
Other	64.10 (40.15)	84 (25 to 96)	39.45 (34.68)	26 (14 to 62)
Ototoxic medication	56.75 (39.42)	72 (15 to 84)	42.00 (30.20)	44 (12 to 70)
Progressive	67.56 (29.30)	79 (46 to 88)	58.22 (17.45)	52 (43 to 75)
Preoperative to EOS				
Congenital	48.70 (38.66)	58 (3 to 85)	24.80 (24.67)	21 (1 to 40)
Meningitis	63.00 (15.62)	71 (45 to 73)	31.33 (10.26)	34 (20 to 40)
Other	78.22 (34.22)	98 (61 to 100)	50.89 (31.15)	44 (27 to 79)
Ototoxic medication	61.33 (53.12)	92 (0 to 92)	60.00 (9.17)	62 (50 to 68)
Progressive	68.33 (32.23)	80 (46 to 91)	60.00 (20.67)	62 (38 to 74)
Year 1 to EOS				
Congenital	5.37 (18.37)	0 (0 to 11)	6.53 (15.74)	6 (0 to 19)
Meningitis	28.67 (10.97)	25 (20 to 41)	12.67 (11.55)	6 (6 to 26)
Other	6.13 (7.85)	4 (1 to 10)	8.67 (11.36)	4 (0 to 18)
Ototoxic medication	5.67 (4.93)	8 (0 to 9)	29.33 (15.28)	26 (16 to 46)
Progressive	1.67 (16.50)	1 (−6 to 9)	7.33 (20.89)	2 (−6 to 26)

Abbreviations: CNC, Consonant-Nucleus-Consonant; EOS, end of study; HINT, Hearing in Noise; IQR, interquartile range.

Table 5. Mann-Whitney Test Comparison of Patients With Different Causes of Deafness in Terms of Year 1 Improvement in CNC Word Score

Causes Compared		*P* Value
Meningitis	Other	.09
Meningitis	Congenital	.52
Meningitis	Ototoxic medication	.10
Meningitis	Progressive	.004
Other	Congenital	.07
Other	Ototoxic medication	.77
Other	Progressive	.08
Congenital	Ototoxic medication	.13
Congenital	Progressive	<.001
Ototoxic medication	Progressive	.30

Abbreviation: CNC, Consonant-Nucleus-Consonant.

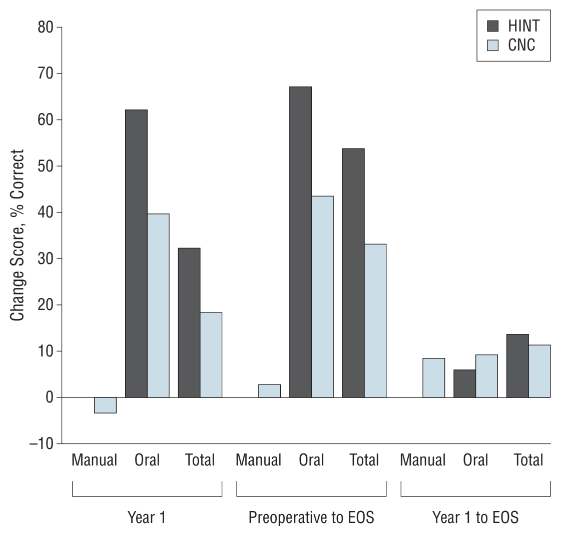

Figure 2. The mean change score for the Hearing in Noise (HINT) sentence test and Consonant-Nucleus-Consonant (CNC) monosyllabic word test in the 3 study periods analyzed stratified by primary mode of communication before cochlear implantation. The HINT sentence scores for the manual communication category in each of the 3 periods are not visible because the mean change scores were 0. EOS indicates end of study.

first year of use and the last follow-up period. In addition, when scores were adjusted for length of deafness, the significant effect of cause of deafness on performance was lost. It is important to realize that the number of subjects in each group was small, and perhaps subtle differences in performance outcomes between different causes of deafness were not observed owing to a lack of statistical power. Conversely, the length of deafness before CI rather than the cause of deafness may be the stronger determinant of success.

Although some studies suggest better performance in patients with *GJB2* mutations,[26] other studies report no relationship when subjects are matched for age at CI and length of device use.[27-29] These equivocal data perhaps suggest that the cause of deafness is not a reliable predictor of successful hearing outcomes. That patients with progressive deafness had the highest mean change scores for both HINT sentence and CNC word scores at year 1 in our study is likely because these patients tended to have shorter lengths of deafness before CI, a factor known to improve post-CI performance. In addition, although our study did not specifically evaluate genetic causes of deafness, we assume that a portion of the patients in the progressive hearing loss group have *GJB2* mutations and therefore improved post-CI performance. Conversely, patients with meningitis are known to perform more poorly than matched control subjects, perhaps accounting for the poorer performance observed in the group of patients with postmeningitic deafness in our study.[30]

The effect of pre-CI mode of communication also had an effect on the speech perception outcomes. Before CI,

Table 6. Change in CNC Word and HINT Sentence Scores Stratified by Mode of Communication

Period by Communication Mode	HINT Sentence Change Score Mean (SD)	HINT Sentence Change Score Median (IQR)	CNC Word Change Score Mean (SD)	CNC Word Change Score Median (IQR)
Year 1				
Manual	0 (0.00)	0 (0 to 0)	−3.40 (7.60)	0 (−8.5 to 0)
Oral	62.26 (32.16)	77 (38 to 88)	39.48 (26.75)	40 (1 to 56)
Total	32.33 (41.59)	10 (0 to 79.5)	18.25 (21.27)	12 (0 to 35)
Preoperative to EOS				
Manual	0 (0.00)	0 (0 to 0)	2.67 (4.62)	0 (0 to 8)
Oral	67.23 (32.18)	75 (47 to 96)	43.48 (27.05)	38 (22 to 68)
Total	53.86 (43.46)	69 (0 to 92)	33.14 (27.32)	42 (0 to 60)
Year 1 to EOS				
Manual	0 (0.00)	0 (0 to 0)	8.33 (14.43)	0 (0 to 25)
Oral	5.79 (16.21)	2 (0 to 15)	9.03 (17.12)	6 (0 to 24)
Total	13.71 (17.96)	9 (0 to 29)	11.17 (10.78)	11 (0 to 21)

Abbreviations: CNC, Consonant-Nucleus-Consonant; EOS, end of study; HINT, Hearing in Noise; IQR, interquartile range.

75% of subjects in this study used oral speech and language as their primary if not sole means of communication, although 10% used sign language and 15% total communication. Our data show that adolescent patients who used oral communication performed significantly better on word and sentence testing measures than did their peers who used total or manual communication. Only 2 of the 7 adolescents using manual communication achieved measurable benefit from their CI device, but many of these patients reported subjective benefits, including sound awareness and improved self-confidence. Not surprisingly, 2 of the 7 patients using manual communication became nonusers of their device because of the inability to achieve objective benefits. These results are similar to those published previously[20,31] and support the notion that prelingually deafened adolescents using only manual communication will rarely obtain any significant auditory benefit after CI and, barring exceptional cases, will not perform as well as their peers who used oral communication before CI. However, these patients should undergo evaluation on a case-by-case basis to assess candidacy based on the needs and expectations of each individual.

The most recent population studies estimate that approximately 33 000 children in the United States aged 6 to 19 years have profound bilateral hearing loss.[32] Despite the advances in CI technology and successful post-CI hearing outcomes in adolescents, multidisciplinary CI teams continue to struggle with counseling patients and their families regarding prognostic factors and predictors of successful CI device use in the prelingually deafened adolescent cohort. The present study evaluates data from CI in prelingually deafened adolescents, many of whom have prolonged lengths of deafness. The rationale for evaluating these patients was 2-fold. We demonstrated that there is significant improvement in objective hearing outcome measures in this group of patients. Furthermore, we identified factors that predict improved performance after CI, that is, patients with shorter length of deafness and earlier age at CI. In addition, patients with progressive deafness of unknown causes outperform their peers with congenital or postmeningitic deafness. Finally, patients who used only oral communication before CI have significantly better objective outcomes than do their peers who used manual or total communication.

Some of the variables studied appear to predict performance during the first year after CI but lose their significance after year 1. Perhaps this is primarily caused by the ceiling effect; many adolescents using CI devices are able to reach high levels of speech understanding within the first year after CI. However, other variables are clearly confounded by the length of deafness before CI, highlighting the importance of early evaluation and aggressive treatment for prelingually deafened adolescents. Some adolescents find minimal to no benefit from their CI, as evidenced by the 5 patients who were nonusers of their devices. Although studies are currently under way to further examine contributing factors to outcomes in the adolescent population, CIs should be considered a viable option for hearing rehabilitation for this group so long as patient and family expectations are realistic and appropriate.

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Correspondence: Susan B. Waltzman, PhD, Department of Otolaryngology–Head and Neck Surgery, New York University Langone Medical Center, 660 First Ave, Seventh Floor, New York, NY 10016 (susan.waltzman@nyumc.org).

Author Contributions: *Study concept and design:* Zeitler and Waltzman. *Acquisition of data:* Anwar, Green, Friedmann, Roland, and Waltzman. *Analysis and interpretation of data:* Zeitler, Babb, and Waltzman. *Drafting of the manuscript:* Zeitler, Green, Babb, Friedmann, and Waltzman. *Critical revision of the manuscript for important intellectual content:* Zeitler, Anwar, Babb, Roland, and Waltzman. *Statistical analysis:* Zeitler and Babb. *Obtained funding:* Waltzman. *Administrative, technical, and material support:* Anwar, Green, Friedmann, Roland, and Waltzman. *Study supervision:* Zeitler and Waltzman.

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