J Am Acad Audiol 19:602–611 (2008)

Speech Perception Outcome in Multiply Disabled Children following Cochlear Implantation: Investigating a Predictive Score

DOI: 10.3766/jaaa.19.8.4

Keith Trimble*
Laura C. Rosella†
Evan Propst*
Karen A. Gordon‡
Vicky Papaioannou‡
Blake C. Papsin*

Abstract

Background: Children with multiple disabilities account for a small percentage of implantees in a cochlear implant program, but they remain the most challenging group for which to predict benefit from the implant and for cooperation with habilitation postoperatively.

Purpose: To assess the relationship of pre-implant functional disabilities with postoperative speech perception scores and determine the feasibility of predicting outcome with a cochlear implant in a multiply disabled pediatric population.

Research Design: Retrospective cohort study.

Study Sample: Sixty-six children with a cochlear implant and at least one additional disability.

Data Collection and Analysis: We retrospectively examined the relationship between pre-implant Graded Profile Analysis (GPA) scores and postimplant speech perception scores. A pre-implant functional disability score (based on the Battelle developmental screen) was applied to the same cohort of patients and its association with postimplant speech perception scores was examined.

Results: The functional disability score significantly predicted high ($k > 24$) and low ($k < 7$) speech perception scores ($p < 0.001$ and $p < .0001$) and had excellent discrimination ability (c statistic = 0.88 and 0.93 respectively). The GPA score was not significantly associated with speech perception scores ($p = 0.519$ and $p = 0.146$) and demonstrated no ability to discriminate postimplant speech perception scores in this implant population (c statistic = 0.49 and c = 0.57).

Conclusions: Prediction of outcomes following cochlear implantation in multiply disabled children can be facilitated using this newly developed functional disability score as an adjunct to traditional candidacy assessments.

Key Words: Cochlear implantation, disabled children, hearing impaired persons, logistic regression, outcome measures

Abbreviations: GPA = Graded Profile Analysis; PROSPER = Pediatric Ranked Order Speech Perception score; MAIS = Meaningful Auditory Integration Scale; ESP = Early Speech Perception; WIPI = Word Intelligibility by Picture Identification; GASP = Glendonald Auditory Screening Procedure; MLNT = Multisyllabic Lexical Neighbourhood Test; BKB = Bamford-Kowal-Bench; PBK = Phonetically Balanced Kindergarten

*Department of Otolaryngology—Head and Neck Surgery, The Hospital for Sick Children, 555 University Avenue, Toronto, M5G 1X8; †Department of Public Health Sciences, University of Toronto, Health Sciences Building, 6th Floor, M5T 1P8, Toronto; ‡Cochlear Implant Laboratory, The Hospital for Sick Children, 555 University Avenue, Toronto, M5G 1X8

Keith Trimble, MB FRCS, Department of Otolaryngology—Head and Neck Surgery, The Hospital for Sick Children, 555 University Avenue, Toronto, M5G 1X8, Canada; Phone: +1 416-813-4938; Fax: +1 416-813-5036; E-mail: keith.trimble@utoronto.ca

Sumario

Antecedentes: Los niños con discapacidades múltiples constituyen un pequeño porcentaje de los implantados en un programa de implantes cocleares, pero sigue siendo el grupo más desafiante para predecir el beneficio del implante y para la cooperación en una habilitación post-operatoria.

Propósito: Evaluar la relación de las discapacidades antes del implante con los puntajes post-operatorios de percepción del lenguaje y determinar la factibilidad de predecir el resultado del uso de un implante coclear en una población pediátrica con discapacidad múltiple.

Muestra del Estudio: Sesenta y seis niños con un implante coclear y al menos una discapacidad adicional.

Recolección y Análisis de los Datos: Examinamos retrospectivamente la relación entre los puntajes del Análisis de Perfil en Grados (GPA) pre-implante y los puntajes de percepción del lenguaje post-implante. Se aplicó un puntaje de discapacidad funcional pre-implante (con base en el tamiz de desarrollo de Battelle) en la misma cohorte de pacientes y se examinó su asociación con los puntajes de percepción del lenguaje post-implante.

Resultados: El puntaje de discapacidad funcional predijo significativamente los puntajes altos de percepción del lenguaje ($k > 24$) y los bajos ($k > 7$) ($p < 0.001$ y $p < 0.0001$) y mostró una excelente habilidad de discriminación (estadística $c = 0.88$ y 0.93, respectivamente). El puntaje GPA no se asoció significativamente con los puntajes de percepción del lenguaje ($p = 0.519$ y $p = 0.146$) y no demostró capacidad para discriminar los puntajes de percepción del lenguaje post-implante (estadística $c = 0.49$ y $c = 0.57$).

Conclusiones: La predicción de los resultados después de una implantación coclear en niños con discapacidades múltiples puede facilitarse utilizando el puntaje de discapacidad funcional recientemente desarrollado como un adicional a las evaluaciones tradicionales para la candidatura al implante.

Palabras Clave: implantación coclear, niños discapacitados, persona hipoacúsica, regresión logística, medidas de resultados.

Abreviaturas: GPA = Análisis de Perfil en Grados; PROSPER = Puntaje Pediátrico de Percepción del Lenguaje por Orden de Rango; MAIS = Escala de Integración Auditiva Significativa; ESP = Percepción Temprana del Lenguaje; WIPI = Inteligibilidad del Lenguaje por Identificación de Dibujos; GASP = Procedimiento de Tamizaje Auditivo de Glendonald; MLNT = Prueba Léxica Multisilábica de Barrio; BKB = Bamford-Kowal-Bench; PBK = Palabras Fonéticamente Balanceadas de Jardín de Infantes

Prediction of postimplant performance in children with severe to profound hearing loss is often facilitated through preoperative evaluation of factors such as age at onset and duration of deafness, age at implantation, language level, etiology of deafness, communication strategy before surgery, mode of education after surgery, parental support, and presence of delayed cognitive and motor milestones (Clark, 2003). However, candidacy criteria for cochlear implantation have evolved and expanded over the years to include younger children, children with cochlear abnormalities, and those with multiple disabilities (McConkey Robbins et al, 2004; Papsin, 2005; Waltzman et al, 2000), making predictions of outcome more complex. The aim of the present study was to investigate a user-friendly multi-domain psychometric test and its ability to predict speech perception scores of children with multiple disabilities after implantation.

Our institution has previously reported on a structured approach to evaluating potential implant candidates that provides a means of predicting postimplant speech perception benefits (Daya et al, 1999; MacDonald et al, 2004). This approach was entitled "Graded Profile Analysis" (GPA) and consisted of 14 categories, each of which was assessed by a team member with expertise in that field. Each category was evaluated with respect to the degree of concern (severe, mild/moderate, or none) present. The GPA score was related to the decision to provide an implant with three separate groups identified: children who generally did not receive implants, children who did receive implants; and a group in which decisions were made on a case-by-case basis. Generally speaking, the GPA score correlated well with open set speech perception tests (the child repeats the word heard), but not closed set tests (the child hears a word and chooses a representation from a set of pictures or objects); likely because many children achieved good scores on the latter measures. Either the GPA instrument was not sensitive to the disabilities of these children or the full extent of developmental delay or language disorders were not yet apparent at the time of pre-implant assessment. Consequently it was felt that in children with multiple disabilities and low open set speech discrimination, the GPA score was limited in its ability to predict outcomes.

Table 1. The Eight Domains and Four Ratings Categories of the Modified Battelle Developmental Inventory Screening Test Used to Determine the Pre-implant Functional Disability Score

DOMAIN	1: Severe	2: Moderate	3: Mild	4: Age appropriate
Personal-Social	no social interaction	minimal interaction	meaningful interaction	age appropriate
Gross Motor	virtually no motor function	disability affecting half of body	disability affecting part/whole limb	age appropriate
Fine Motor	no fine motor skill	moderate disability	mild disability	age appropriate
Receptive Vision	no vision	one eye or half of each eye	wears corrective lenses	normal vision
Receptive Language	no understanding	understands 50%	understands 75%	age appropriate
Expressive Language	not speaking	50% intelligible	75% intelligible	age appropriate
Articulation	inarticulate	articulates 50% of words	articulates 75% of words	age appropriate
Cognition	no conditioned response	inconsistent conditioned response	age-inappropriate conditioned response	age appropriate

Ratings scales are commonly used to assess young children, and in general, the developmental level of the child should be considered, rather than simply applying an age-appropriate tool. In the context of pain assessment in preverbal children, for example, assessment of behaviors is the primary tool used. This is particularly difficult in cognitively impaired children, and there is not yet a consensus on a standard assessment procedure (Breau et al, 2002). These same difficulties are anticipated when rating children who are multiply disabled and deaf because of the large variability in cognitive and physical traits that can impact on speech perception scores.

Postimplant speech perception scores in children with disabilities have previously been reported to be variable (Edwards et al, 2006; Waltzman et al, 2000). Some patients with CHARGE syndrome, Down syndrome, and autism have been poor implant users, as measured by speech perception scores, and compliance for mapping and testing postimplant can also prove difficult (Bauer et al, 2002; Daneshi and Hassanzadeh, 2006). The situation is more complicated when implanting very young children because they may not yet have displayed significant developmental delay or neurologic deficit (Waltzman et al, 2000).

Although children with multiple disabilities account for a small percentage of implantees in our program (12%, 66 out of 550), they remain the most challenging group for which to predict benefit from the implant and for cooperation with habilitation postoperatively. Even though these children may not progress as well as their peers who do not have additional disabilities, they may nevertheless realize benefit from the additional auditory stimulation offered by a cochlear implant, representing a sufficiently successful outcome to justify the procedure. Another concern is that benefit following implantation can be difficult to measure objectively in these children. In some cases, the only outcome data available relates to the hours of cochlear implant use or anecdotal reports of awareness of environmental sounds.

This study investigates the ability of a multidomain psychometric test (which could be easily administered by the cochlear implant team) to predict speech perception scores of children with multiple disabilities after cochlear implantation. We hypothesized that there would be a significant correlation between a child's global developmental disability and postimplant speech perception scores, and a weaker correlation between the GPA and postimplant speech perception scores. If true, this tool might be useful to counsel parents regarding potential outcomes of implantation.

MATERIALS AND METHODS

Inclusion/Exclusion Criteria

Patients were included if they underwent cochlear implantation at the Hospital for Sick Children between July 1988 and March 2005, had a disability in addition to sensorineural hearing loss, were younger than 18 years old with at least six months of cochlear implant use, and had speech outcome data. Disabilities were known at the time of candidacy assessment. Cleft lip and palate patients were excluded as their functional impairment was usually corrected and was not a persistent disability. Procedures followed for the study were in accordance with the ethical standards of the Hospital for Sick Children's Research Ethics Board.

Pre-implant Functional Disability Score

The Battelle Developmental Inventory Screening Test has seven domains and is validated for use in children from six months to eight years (Glascoe and Byrne, 1993). The adaptive domain was removed from this test and vision and articulation domains added (see Table 1) to constitute the functional disability score. Each patient was scored retrospectively for each of the eight domains by their managing implant audiologist, based on information from comprehensive pre-implant assessments by a speech therapist, auditory verbal therapist, and implant audiologist. The audiologists were asked to be as objective as possible using information from the pre-implant assessments in

Table 2. The Graded Profile Analysis Score: A Structured Assessment Allowing Potential Cochlear Implant Candidates to Be "Scored" in Each Category, Giving a Potential Range of −14 to +14

Team Impressions of Factors Important in Suitability for Cochlear Implant	No Concern (+1)	Mild-Moderate Concern (0)	Great Concern (−1)
Chronological Age	Under 3.5 years	3.5 to 5 years	Over 5 years
Duration of Deafness	Under 3.5 years	3.5 to 5 years	Over 5 years
Developmental Milestones	On target	Mild delay	Significant delay
Medical/Radiological	Implantable	Potential for nonstimulation/partial insertion	Not implantable
Multiple Handicap	No additional developmental delay	Mild developmental delay	Moderate-severe developmental delay
Functional Hearing	No usable aided hearing	Questionable aided hearing	Usable aided hearing
Speech and Language	Under 3.5 years delay	3.5 to 5 years delay	Over 5 years delay
Family Structure and Support	Supportive	Questionable support	Unreliable support
Expectations (Patient)	Appropriate	Questionable	Inappropriate
Expectations (Parent)	Appropriate	Questionable	Inappropriate
Interpersonal Social Skills	Age appropriate	Questionable	Very delayed
Educational Environment	Mainstream	Hearing impaired class with some sign support	Signing program
Support Services	Auditory verbal therapy	Limited availability of therapy	No availability of therapy
Cognitive/Learning Style	Age appropriate	Questionable	Delayed

the patient's chart to complete the functional disability score retrospectively. For example, tests used during the communication assessment part of candidacy assessment, such as the Goldman Fristoe test of articulation skills and the Preschool Language Scale, were translated into their respective domain scores in the pre-implant functional disability score. Other difficult-to-assess domains such as personal-social, cognitive, or motor skills were assessed by appropriateness of responses during the audiological assessment. Categories in these domains are fairly self-explanatory (see Table 1), except for cognition, which is a particularly difficult skill to assess in children with other physical limitations and hearing loss. A formal psychology cognitive assessment was not routinely performed. The pediatric audiologist and auditory verbal therapist have a wealth of experience with deaf infants and young children and thus can judge the relative abilities of an individual child to perform age-appropriate tasks in audiological and speech-language testing. These tasks typically involve both cognition and motoric capabilities. Audiologists judged cognitive abilities based on the child's ability to learn a conditioned response to either auditory or vibrotactile stimulation during audiological testing. The scale ranged from an inability to learn to perform any conditioned response over repeated test sessions to consistent responses in an age-appropriate task. All domains were scored from one to four (severe disability to age appropriate, see Table 1), and a total score from all eight domains was assigned (8 = most severe, 32 = most age appropriate) with the audiologist assigning a score for each category. In order to assess intra-rater reliability, each child's pre-implant function was rescored in all eight domains by the same audiologist twelve weeks later. Correlation analysis (Pearson's r) suggests moderate intra-rater reliability ($r = 0.69$) regarding the pre-implant functional disability score.

Pre-implant GPA Score

Pre-implant GPA scoring was completed for each child prospectively by a multidisciplinary team, including a speech therapist, auditory verbal therapist, and implant audiologist. This tool has been reported and shown to be a valid predictor of postimplant speech perception scores (Daya et al, 1999; MacDonald et al, 2004). It is used during the candidacy assessment process and helps to steer decisions on implantation. The categories are listed in Table 2, and each is scored according to the level of concern relating to suitability for a cochlear implant. Among our otherwise nondisabled children, those receiving a score of greater than or equal to eight typically receive cochlear implants.

Speech Perception Outcomes

In the early postoperative period following implantation, many different tests were used to assess speech perception; many of the tests were too difficult for this population, resulting in a large amount of missing data. To remedy this, the Pediatric Ranked Order Speech Perception (PROSPER) score was created to integrate all available results into one score that could be followed over time (see Table 3). This was performed by hierarchizing the individual speech perception tests, ranging from simple detection through pattern perception and closed set and open set phoneme, word, and sentence recognition, as described by Geers and Moog (Geers and Moog, 1987). The

Table 3. Pediatric Ranked Order Speech Perception (PROSPER) Score: Hierarchized Individual Speech Perception Tests, Ranging from Simple Detection through Pattern Perception, Closed-Set and Open-Set Phoneme to Word and Sentence Recognition

Pediatric Ranked Order Speech Perception Score	
Blank	Did not test
0	Could not test
1	MAIS <50%
2	MAIS ≥50%
3	ESP low verbal pattern perception <50%
4	ESP low verbal pattern perception ≥50%
5	ESP low verbal spondee <50%
6	ESP low verbal spondee ≥50%
7	ESP low verbal monosyllable <50%
8	ESP low verbal monosyllable ≥50%
9	ESP standard pattern perception <50%
10	ESP standard pattern perception ≥50%
11	ESP standard spondee <50%
12	ESP standard spondee ≥50%
13	ESP standard monosyllable <50%
14	ESP standard monosyllable ≥50%
15	WIPI <50%
16	WIPI ≥50%
17	GASP sentences <50%
18	GASP sentences ≥50%
19	GASP word <50%
20	GASP word ≥50%
21	MLNT phoneme <50%
22	MLNT phoneme ≥50%
23	MLNT word <50%
24	MLNT word ≥50%
25	BKB word <50%
26	BKB word ≥50%
27	LNT phoneme <50%
28	LNT phoneme ≥50%
29	LNT word <50%
30	LNT word ≥50%
31	PBK phoneme <50%
32	PBK phoneme ≥50%
33	PBK word <50%
34	PBK word ≥50%

speech perception test battery included one parental questionnaire (Meaningful Auditory Integration Scale [MAIS]), two closed set tests (Early Speech Perception [ESP] test and the Word Intelligibility by Picture Identification [WIPI] test) and five open set tests (the Glendonald Auditory Screening Procedure [GASP] word and sentence tests; the Multisyllabic Lexical Neighbourhood Test [MLNT]; the Lexical Neighbourhood Test [LNT]; Bamford-Kowal-Bench [BKB] words; and the Phonetically Balanced Kindergarten [PBK] test). The hierarchy of these tests as described by Geers and Moog suggests that, for any one child, test scores (%) would be higher on the MAIS questionnaire (which asks questions about the child's general auditory behavior) than on closed or open set tests of speech perception. Closed set tests, in which the child hears a word or sentence and chooses the most representative picture or object from a set of pictures (two or more), would yield higher scores than an open set test, in which the child must repeat what was heard or answer a question without the option for guessing. Consideration of the test material is also important. As described by Erber (Erber, 1982), children with hearing loss learn to discriminate between multisyllabic words before discrimination of spondee words, and discrimination between monosyllabic words is most difficult. In addition, the test becomes more challenging as the level of the vocabulary increases. An ordinal scale was developed based on both the type of test and the test content. The low-verbal ESP, using vocabulary typical for toddlers, was therefore rated as a simpler test than the high-verbal ESP, which uses a higher level vocabulary set. Each of the three subtests of the closed set ESP was ranked according to the type of words (multisyllabic, spondee or monosyllables). The high-verbal ESP monosyllable test was considered to be simpler than the WIPI (also monosyllables) based on the increased level of vocabulary of the latter. Although open set tests were generally ranked as more complex tests, the GASP test was placed between the ESP and WIPI tests due to the very simple vocabulary that it uses. Open set tests were then ordered by word type from multisyllabic (used in the MLNT) to monosyllables (LNT), with the BKB word score somewhere between. The PBK was considered to be the most complex test based on the higher level of vocabulary used.

Phoneme scores were considered to be lower in rank than word scores as the phoneme scores are more likely to be slightly better, allowing differentiation between fair performance on the test (0–50%) and better results (50–100%), and resulting in 34 levels. The rank ordering was quantified from 0 (least complex) to 34 (most complex). In order to interpret the scores as they relate to high and low performance following implantation, two cutoffs in the distribution of postimplant PROSPER outcome scores were evaluated: one at 24 to focus on the high performers (those achieving open set speech perception skills) and one at 7 to focus on the low performers (those achieving perception of speech patterns only).

Habilitation

Habilitation methods were recorded for each child at the time of implantation. Method of therapy was auditory verbal (53 children) or auditory oral (5). Method of communication was oral (46 children), manual (3), or total communication (9).

Statistical Analysis

The data indicated that PROSPER scores tended to cluster at high and low ends of the outcome scale (see

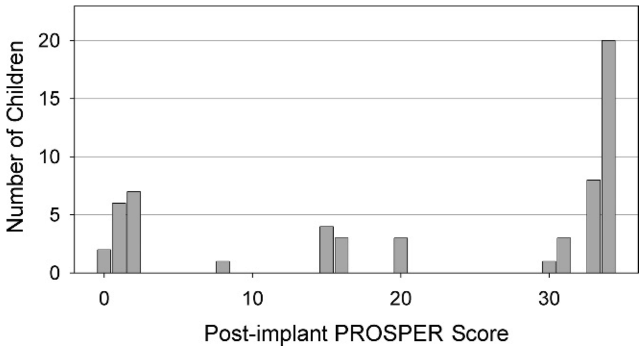

Figure 1. Distribution of postimplant PROSPER scores are plotted for the 58 children.

Figure 1). We therefore used logistic regression to analyze the relationship between the pre-implant functional disability score (continuous variable) or pre-implant GPA score (continuous variable) and the dichotomized postimplant PROSPER outcome score. By dichotomizing our outcome variable, we minimized false inferences made by assuming a continuous distribution. High (≥24) or low (≤7) dichotomous PROSPER outcome cutoffs were chosen primarily as they reflect meaningful clinical outcomes but also because the sample size was not powered for ordinal or nominal outcomes. Effects of patient characteristics (i.e., age, duration of implant use) and pre-implant scores (GPA and functional disability score) on the dichotomized postimplant PROSPER score were examined by t-tests. The GPA variable was analyzed in two ways: first by its continuous form (using the t-test) and by a dichotomized form (>8) to reflect the range of scores for children typically receiving a cochlear implant in our center (Daya et al, 1999; MacDonald et al, 2004). The latter was tested using a chi-square test; details are provided in Table 4.

Probability plots were produced based on the probability of scoring high or low from the logistic regression equation (remembering this is only the unadjusted relationship—it is the only relationship that can be modeled in two dimensions). Both the pre-implant GPA score and the pre-implant functional disability score were modeled to contrast the performance of these scores. Odds ratios and 95% outcomes for the two scores were calculated. In addition to the two pre-implant scores, additional variables were assessed for a relationship with the PROSPER score. A further adjustment of age was included to demonstrate the predictive significance of the PROSPER score, independent of the age at activation, chronological age, or duration of implant use. The discrimination ability of the scores for predicting high and low outcomes was examined. Discrimination is the ability to correctly classify those who will and will not experience the event, in this case a high or a low PROSPER score following cochlear implantation based on the pre-implant prognostic score. It is a key feature of prognostic models used to assess patient performance and is assessed by the c-index, which is identical to the area under the receiver operating characteristic curve (Hanley and McNeil, 1982). A c statistic of 0.5 indicates no discriminative ability of the model whereas c statistics of 0.8 to 0.9 indicate excellent discrimination (Hosmer and Lemeshow, 2000). All statistical analysis was performed using SAS software (Version 9.1; SAS Institute, Cary, North Carolina).

RESULTS

Sixty-six patients with profound hearing loss and other disabilities were identified from the cochlear implant database. Eight patients were excluded: five did not have postoperative speech perception scores available; one child moved to another province; and two were explanted for reasons unrelated to speech perception scores (one developed postoperative wound infection and chose to use American Sign Language, another had Goldenhar syndrome and facial nerve stimulation at stimulation levels and parameters that

Table 4. Age at Activation, Sex, and Prognostic Scores by Postimplant PROSPER Outcome Score

	High PROSPER Outcome Score*			Low PROSPER Outcome Score*		
Factor	≥24 (n = 32)	<24 (n = 26)	*P-value*	≤7 (n = 15)	>7 (n = 43)	*P-value*
Mean (SD) Age at Activation	5.2 (0.6)	2.4 (0.3)	<0.001	2.3 (0.3)	4.6 (0.5)	<0.001
Male (%)	46.9	38.5	0.52	33.3	46.5	0.38
Graded Profile Analysis Score						
Mean (SD)	10.0 (0.6)	9.5 (0.8)	0.60‡	8.5 (1.3)	10.2 (0.5)	0.24‡
% above score of 8	64.0	73.1	0.49†	66.7	69.4	0.85†
Pre-implant Functional Disability Score						
Mean (SD)	25.4 (0.7)	17.6 (1.1)	<0.001‡	15.2 (1.1)	24.4 (0.7)	<0.001‡

*High PROSPER outcome score (>24) is equivalent to open set word recognition; Low PROSPER outcome score (<7) is equivalent to pattern perception only.
†P-value is from the χ^2 test.
‡P-value is from the t-test.

Table 5. Selected Characteristics of Pediatric Cochlear Implant Users Identified with a Disability Other Than Deafness

Characteristic	*n = 58*
Male (%)	56.9
Right ear implanted (%)	82.8
Oral communication (%)	86.0
Auditory verbal mode of therapy (%)	91.4
Disability (%)	
Developmental delay	31.0
Syndrome*	15.5
Cerebral palsy	15.5
Language disorder	13.8
Deaf/blind	12.1
Learning disability	8.6
Kernicterus	3.4
Median duration of implant use in years (range)	3.5 (0.5 to 12)
Age at activation	
Median in years (range)	3.2 (0.7 to 14.8)
Mean in years (SD)	4.0 (3.1)
Pre-implant Graded Profile Analysis Score (−14 to 14)	
Median (range)	10 (−4 to 14)
Mean (SD)	9.7 (3.6)
% above score of 8	68.6
Pre-implant Functional Disability Score (8 to 32)	
Median (range)	23.5 (9 to 32)
Mean (SD)	21.8 (6.0)
Post-implant PROSPER Score (0 to 34)	
Median (range)	31 (0 to 34)
Mean (SD)	21.8 (14.0)
% above score of 24	55.2
% below score of 7	25.9

*CHARGE x3, split hand/foot malformation, Noonan's, Leopard, Klippel-Fiel, Waardenburg.

severely compromised any auditory detection). Fifty-eight patients were included whose demographic details and descriptive statistics are presented in Table 5.

A wide spread of pre-implant functional disability scores was noted, ranging from both extremes (9 to 32) with a median score of 23.5 and a mean of 21.8 (±6.0). Figure 1 displays the distribution of postimplant PROSPER scores. The median score was 31 with minimum and maximum scores (0 to 34). The scores clustered at each end, with the uppermost score representing open set speech and the lowest score corresponding to awareness of environmental sound.

Table 4 presents demographic characteristics (age and sex) by the high and low PROSPER scores. The mean pre-implant GPA score did not significantly differ with the high and low PROSPER scores ($p = 0.60$ and $p = 0.24$); however, the mean pre-implant functional disability score was significantly associated with the low and high PROSPER outcome scores ($p < 0.001$ and < 0.001).

Table 6 shows the crude and adjusted odds of "High" and "Low" PROSPER score in relation to various patient characteristics, modeling the GPA score and pre-implant functional disability score as predictors in the logistic regression analysis. The age at activation-adjusted odds ratio for the high and low PROSPER score (for each unit increase in the pre-implant functional disability score) was 1.58, 95% CI (1.22, 2.04) and 0.66, 95% CI (0.53, 0.83), respectively. The discrimination of the unadjusted pre-implant functional disability score was excellent using both high and low cutoffs ($c = 0.89$ and $c = 0.93$). In contrast, the pre-implant GPA score demonstrated no ability to discriminate outcomes in these 58 implant patients ($c = 0.49$ and $c = 0.57$). The logistic regression analysis that modeled the relationship between GPA score and pre-implant functional disability score to the probability of a high ($k > 24$) and low ($k < 7$) PROSPER score demonstrated that the GPA score was not significantly associated with the PROSPER scores ($p = 0.60$ and $p = 0.15$) (see Table 6).

In order to apply the pre-implant functional disability score as a predictor tool in other children with multiple disabilities undergoing candidacy assessment, we used the logistic regression equation detailed in Figures 2 and 3 to produce a "p versus x plot" of the predicted probability of achieving a high postimplant PROSPER score. This plot, shown in Figure 2, indicates that the probability of achieving a high postimplant PROSPER score increases with increasing values of the pre-implant functional disability score. Based on the predictive curve shown in Figure 2, a patient scoring 20 on the pre-implant functional disability score has a 50:50 chance of a "High" PROSPER score following implantation. For a patient scoring 29, there is a 90% probability of achieving a high PROSPER score. By adjusting the cutoff points for speech outcome, we developed probability plots for low PROSPER scores (<7, ESP low verbal spondees only) as shown in Figure 3. Thus, a child with a pre-implant functional disability score of 15 has a 70% probability of achieving a low PROSPER score and has only a 15% chance of achieving a high PROSPER score postimplant.

DISCUSSION

In this study, a modified version of the Battelle Developmental Inventory Screening Test was used to evaluate children with multiple disabilities prior to cochlear implantation. A PROSPER score was created to compare postimplantation speech perception scores of subjects with different ages at implantation (0.7 to 14.8 years) and duration of implant use (0.5 to 12 years). Fifty-five percent of children with multiple disabilities achieved open set speech perception scores of greater than 50%; however, the PROSPER scores

Table 6. Crude and Adjusted Odds of "High" and "Low" PROSPER Score in Relation to Patient Characteristics, Using Graded Profile Analysis Score and Pre-implant Functional Disability Score

Factor	Crude odds ratio	95% CI	P-value†	C statistic	Adjusted odds ratio*	95% CI	P-value†	C statistic
PROSPER >24								
Graded Profile Analysis score	1.04	(0.89, 1.22)	0.60	0.49				
Pre-implant functional disability score	1.39	(1.17, 1.64)	<0.001	0.89	1.58‡	(1.22, 2.04)	0.001	0.95
					1.67§	(1.20, 2.33)	0.003	0.98
					1.37**	(1.14, 1.65)	0.001	0.92
Age at activation	2.12	(1.32, 3.41)	0.002		3.18	(1.34, 7.56)	0.009	
Chronological age	1.76	(1.33, 2.34)	<0.001		2.52	(1.31, 4.86)	0.006	
Duration implant use	1.62	(1.20, 2.20)	0.002		1.91	(1.15, 3.19)	0.013	
PROSPER <7								
Graded Profile Analysis score	0.88	(0.74, 1.05)	0.15	0.57				
Pre-implant functional disability score	0.70	(0.58, 0.83)	<0.001	0.93	0.66‡	(0.53, 0.83)	0.001	0.95
					0.58§	(0.40, 0.85)	0.005	0.97
					0.52**	(0.31, 0.88)	0.014	0.92
Age at activation	0.54	(0.32, 0.90)	0.017		0.47	(0.22, 0.99)	0.048	
Chronological age	0.61	(0.45, 0.82)	0.001		0.49	(0.28, 0.86)	0.013	
Duration implant use	0.56	(0.38, 0.84)	0.005		0.23	(0.06, 0.92)	0.038	

*Adjusted for age at activation, chronological age, and duration of implant use in separate models; note Graded Profile Analysis score has an age component and cannot be adjusted for age.
†P-value is from the χ^2 test.
‡Estimate adjusted for age at activation only.
§Estimate adjusted for chronological age only.
**Estimate adjusted for duration of implant use only.

tended to cluster at either end of the outcome scale as shown in Figure 1. This discontinuous distribution may indicate that the continuous form of the PROSPER score is not effectively capturing the range of outcomes in this population or that the raters (audiologists) assessing the patient tend to cluster their evaluations at certain levels.

A logistic regression model best fit this data and indicated a significant relationship between pre-implant measures of functional disability and postimplant speech perception scores, which persisted even after adjustments for chronological age, age at activation, or the duration of implant use (though these variables did have an important prognostic significance). The pre-implant GPA score (Daya et al, 1999), which we routinely use in decision making and performance prediction, was unable to reliably predict PROSPER scores in this group. This was reflected in the lack of statistical significance of the GPA with the

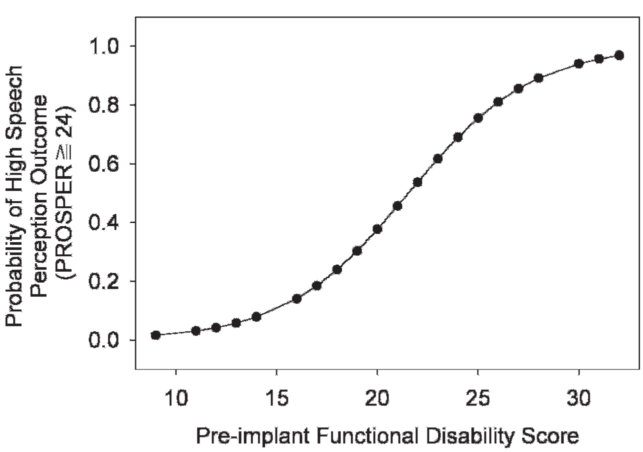

Figure 2. Predicted probability of scoring high ($k > 24$) postimplant PROSPER score as a function of pre-implant functional disability score (plot generated by the logistic regression model: P [k ≥ 24]/P [k < 24] = exp [−7.0272 + 0.3262 {pre-score}]).

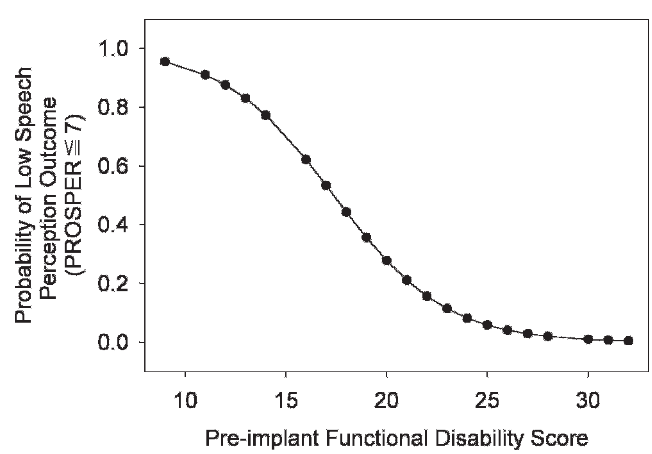

Figure 3. Predicted probability of scoring a low ($k < 7$) postimplant PROSPER score as a function of pre-implant functional disability score (plot generated by the logistic regression equation: [k ≤ 7]/P [k > 7] = exp [6.3082 + −0.3631 {pre-score}]).

outcome variable seen in all the t-tests, chi-square tests, and logistic regression analyses. Furthermore, the GPA was unable to discriminate high or low PROSPER scores in patients as measured by the c-statistic. This result is likely due to the complexity of developmental skills in children with multiple disabilities, which are more fully considered in the domains of the pre-implant functional disability score. Probability plots in Figures 2 and 3 demonstrate how this functional disability score can be used clinically to predict high or low speech perception scores.

Even though children with multiple disabilities may benefit from cochlear implantation, some neither develop closed set nor open set speech discrimination. This was shown by the 15 children who could not perform speech perception tests and had only MAIS scores available (PROSPER ≤2, see Figure 1). The lack of functional speech perception scores in this group can be partially explained by the younger age of 9 of these 15 children. In the remainder, however, the limited improvement in speech perception scores after cochlear implantation is likely related to their additional disability. It is possible that these children do experience improved hearing but cannot use it to achieve functional benefits. Previous reports used oral communication mode and programming ability as acceptable endpoints for evaluating success with a cochlear implant in this subpopulation (Hamzavi et al, 2000; Waltzman et al, 2000; Wiley et al, 2005; Winter et al, 2004). However, without measurable functional outcomes, the benefits and the disadvantages of cochlear implantation cannot be easily assessed.

It is true that the speech perception tests themselves might not completely capture the hearing abilities of the child. On the other hand, the battery of speech perception tests contained in the PROSPER score represent reasonable and well-documented outcome measures for assessment—allowing comparison between patients and tracking changes over time. Overall, the data presented suggest that a measure of global disability such as pre-implant functional disability score can aid prediction of good versus poor speech perception scores after cochlear implantation. This information is useful both from a financial perspective, given the fiscal limitations imposed by the envelope-funded health-care system in Canada, and to counsel parents regarding potential outcomes of implantation. We believe that it is sometimes appropriate to recommend against cochlear implantation. There may be children for whom the additional sensory input provided by the cochlear implant is not only ineffectual in promoting speech and language development but may also be a disruptive influence on other areas of development. Moreover, the follow-up required after cochlear implantation might take the child away from time and energy spent on other therapies and support. Unfortunately, multiple involvements can take a toll on families and caregivers both from a social and economic perspective (Barton et al, 2006).

Certain limitations must be considered when interpreting these study findings. There was variation in age, duration of implant use, and degree of disability across the subjects. Duration of implant use was limited to six months in some patients, and it is possible that outcomes may have improved with longer duration of implant experience. The PROSPER score developed for use in this study is based on a hierarchy of listening skills (Geers and Moog, 1987) and standard speech perception scores and consequently has the same limitations as the tests themselves. Skills that cannot be captured by these tests are not recognized in the predictive method described in this study. The GPA was conducted prior to cochlear implantation, but there is potential for recall bias in the functional disability score, as this was recorded after implantation. The developmental assessment of the deaf child by psychologist or developmental pediatrician is a logical addition to improve the accuracy of the functional disability score. In our experience, these assessments can be difficult to administer and interpret in the deaf child. Moreover, we have long waiting lists in our institution to be seen by these teams. In contrast, the pediatric audiologist and pediatric auditory verbal therapist have a wealth of experience with deaf infants and young children and thus can judge the relative abilities of an individual child to perform age-appropriate tasks in audiological and speech-language testing. These tasks typically involve both cognition and motoric capabilities. The gap between the actual and age-appropriate performance of the functional disability score may increase with age, resulting in relatively better scores for disabled younger children whose performance may appear age appropriate. The functional disability score is designed as a predictive tool to aid in counseling of parents regarding implanting their multiply disabled and deafened child, with the primary intention of being easily and quickly administered by a team of regular cochlear implant professionals.

The functional disability score used in the present study has the potential to aid decisions regarding cochlear implantation in children with multiple disabilities. Given the limitations discussed, the use of more appropriate assessors (occupational therapy, clinical psychology, and developmental pediatrics, for example) completing contemporaneous assessments should be considered in future work to further validate the tool. This initial study should be validated in another cohort of children with additional disabilities, including a factor analysis to confirm which items of the multidomain score are most heavily contributing to the prediction of postimplant speech perception scores. Future studies should address additional outcome measures other than just speech perception scores, as it may be argued that any evidence of auditory awareness in a child with multiple disabilities may represent a sufficiently successful outcome to justify the procedure.

REFERENCES

Barton GR, Fortnum HM, Stacey PC, Summerfield AQ. (2006) Hearing-impaired children in the United Kingdom, III: cochlear implantation and the economic costs incurred by families. *Ear Hear* 27:563–574.

Bauer PW, Wippold FJ, 2nd, Goldin J, Lusk RP. (2002) Cochlear implantation in children with CHARGE association. *Arch Otolaryngol Head Neck Surg* 128:1013–1017.

Breau LM, McGrath PJ, Camfield CS, Finley GA. (2002) Psychometric properties of the non-communicating children's pain checklist-revised. *Pain* 99:349–357.

Clark G. (2003) *Cochlear Implants, Fundamentals and Applications*. New York: Springer.

Daneshi A, Hassanzadeh S. (2006) Cochlear implantation in prelingually deaf persons with additional disability. *J Laryngol Otol* 121:635–638.

Daya H, Figueirido JC, Gordon KA, Twitchell K, Gysin C, Papsin BC. (1999) The role of a graded profile analysis in determining candidacy and outcome for cochlear implantation in children. *Int J Pediatr Otorhinolaryngol* 49:135–142.

Edwards LC, Frost R, Witham F. (2006) Developmental delay and outcomes in paediatric cochlear implantation: implications for candidacy. *Int J Pediatr Otorhinolaryngol* 70:1593–1600.

Erber N. (1982). *Auditory Training*. Washington, DC: Alexander Graham Bell Association for the Deaf.

Geers AE, Moog JS. (1987) Predicting spoken language acquisition of profoundly hearing-impaired children. *J Speech Hear Disord* 52:84–94.

Glascoe FP, Byrne KE. (1993) The usefulness of the Battelle Developmental Inventory Screening Test. *Clin Pediatr (Phila)* 32: 273–280.

Hamzavi J, Baumgartner WD, Egelierler B, Franz P, Schenk B, Gstoettner W. (2000) Follow up of cochlear implanted handicapped children. *Int J Pediatr Otorhinolaryngol* 56:169–174.

Hanley JA, McNeil BJ. (1982) The meaning and use of the area under a receiver operating characteristic (ROC) curve. *Radiology* 143:29–36.

Hosmer DW, Lemeshow S. (2000) *Applied Logistic Regression, 2nd Ed*. New York: John Wiley and Sons.

MacDonald L, Sohn G, Papsin BC, Gordon KA. (2004) Use of a Graded Profile Analysis to assess cochlear implant candidacy: recent findings. *Int Congress Ser* 1273:215–218.

McConkey Robbins A, Koch DB, Osberger MJ, Zimmerman-Phillips S, Kishon-Rabin L. (2004) Effect of age at cochlear implantation on auditory skill development in infants and toddlers. *Arch Otolaryngol Head Neck Surg* 130:570–574.

Papsin BC. (2005) Cochlear implantation in children with anomalous cochleovestibular anatomy. *Laryngoscope* 115:1–26.

Waltzman SB, Scalchunes V, Cohen NL. (2000) Performance of multiply handicapped children using cochlear implants. *Am J Otol* 21:329–335.

Wiley S, Jahnke M, Meinzen-Derr J, Choo D. (2005) Perceived qualitative benefits of cochlear implants in children with multi-handicaps. *Int J Pediatr Otorhinolaryngol* 69:791–798.

Winter M, Johnson KC, Vranesic A. (2004) Performance of implanted children with developmental delays and/or behavioural disorders: retrospective analysis. *Int Congress Ser* 1273:277–280.