

Evaluating Benefits of Cochlear Implantation in Deaf Children With Additional Disabilities

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Objectives: Cochlear-implanted deaf children having additional disabilities may develop speech perception and language skills at a slower pace than their implanted peers without such disorders. Nevertheless, it has been shown that, even for these special cases, cochlear implantation (CI) provides benefits for a larger range of neuropsychological functions including social and relational skills. These benefits are frequently mentioned by parents, but rarely objectively measured by tests. This article presents a new evaluation tool aimed at assessing the global benefits offered by CI in these special cases.

Design: The new tool has been designed as a closed-format questionnaire, divided into five areas. It is based on observing the frequency of preselected behaviors in daily activities, which imply specific social, neuropsychological, and perceptual skills. The questionnaire has been presented to the parents of 50 deaf children with additional disabilities, before and at least 6 months after CI.

Results: The data show significant improvements in all investigated areas. However, not all skills improve in the same way, and only those skills related to language and communication correlate positively with time after implantation. The present article further discusses changes in skills for which parents have higher expectations, such as the preferred communication mode, speech intelligibility, and the ability to communicate on the telephone.

Conclusions: The questionnaire has a simple-to-use format, and it has been proven to be sufficiently sensitive for the detection of changes in each examined area. Because the questionnaire is based on observed behaviors, it can be used even when other existing tests involve tasks that are too complex for these children.

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INTRODUCTION

Cochlear implantation (CI) is the most suitable treatment for patients with profound deafness (Nikolopoulos et al. 2004). However, predicting the outcome of cochlear implants in children with multiple disabilities to select the candidacy for implantation is a complex task. This article focuses on this special group of children, which represents about 30 to 40% of children affected by hypoacusis (Lesinski et al. 1995; Holden-Pitt & Albertorio 1998; Hamzavi et al. 2000; Fortnum et al. 2002; Filipo et al. 2004; Wiley et al. 2005; Gallaudet Research Institute 2008; Chilosi et al. 2010).

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In the early days of CI, candidacy criteria strictly excluded children with multiple disabilities. With growing experience, improvements in technology, and good results, inclusion criteria are being widened and an increasing number of such children have now been implanted (Lesinski et al. 1995; Hamzavi et al. 2000; Waltzman et al. 2000; Edwards 2003, 2007; Donaldson et al. 2004; Filipo et al. 2004; Daneshi & Hassanzadeh 2007; Pundir et al. 2007; Berrettini et al. 2008; Nikolopoulos et al. 2008; Wiley et al. 2008; Bacciu et al. 2009; Gérard et al. 2010; Hans et al. 2010; Meinzen-Derr et al. 2010). Nevertheless, deciding on candidacy remains a challenging task. The reason for the complexity of this decision is because of the difficulty of assessing the true benefits that cochlear implants provide to this special group of patients (Lesinski et al. 1995; Holden-Pitt & Albertorio 1998; Hamzavi et al. 2000; Fortnum et al. 2002; Filipo et al. 2004; Gallaudet Research Institute 2008).

The methods commonly used for determining appropriateness of candidacy of deaf children without additional disabilities are standardized testing protocols and behavior observation tests, carried out both in everyday activities and in structured environments. These tests are aimed at drafting profiles about perceptual, linguistic, and neuropsychological skills, speech intelligibility, and social behavior. However, children who have additional disabilities are often unable to complete standardized testing. Therefore, a comprehensive general evaluation is usually made, based on objectively measured hearing levels and on qualitative behavioral testing. In the literature, many studies focus on evaluating the improvements, after cochlear implantation, of auditory and speech-perception skills (Hamzavi et al. 2000; Waltzman et al. 2000; Fukuda et al. 2003; Donaldson et al. 2004; Filipo et al. 2004; Vlahovic & Sindija 2004; Yang et al. 2004; Daneshi & Hassanzadeh 2007; Pundir et al. 2007; Berrettini et al. 2008; Wiley et al. 2008; Bacciu et al. 2009; Trimble et al. 2009; Hans et al. 2010), language development (Fukuda et al. 2003; Rajput et al. 2003; Donaldson et al. 2004; Wiley et al. 2005; Trimble et al. 2009; Gérard et al. 2010; Meinzen-Derr et al. 2010) and speech intelligibility (Rajput et al. 2003; Vlahovic & Sindija 2004; Nikolopoulos et al. 2008; Bacciu et al. 2009; Hans et al. 2010). Some studies further focus on the need to perform multifactorial developmental evaluations in other domains, such as non-verbal cognition, cognitive performance, precursors of language development, gross and fine motor skills, receptive vision, or social interactions (Wiley et al. 2008; Trimble et al. 2009; Meinzen-Derr et al. 2010).

Depending on age and communication skills of the pediatric patients, many studies turn to administering questionnaires to parents. Parental assessments have positively correlated with results of objective tests, for example, regarding auditory capacity, level of speech, and language and communication skills, which confirm that parents are valid reporters (Glascoe & Sandler 1995; Glascoe 2000; Percy-Smith 2010). Some of those questionnaires

measure speech perception and language abilities (Knors et al. 2003; Stacey et al. 2006; Lanson et al. 2007; Gérard et al. 2010). Others assess changes in behaviors, such as communication mode, production skills, reaction to sounds, eye contact, play with other children, family, and social interactions (Donaldson et al. 2004; Vlahovic & Sindija 2004; Berrettini et al. 2008). Others determine improvements in quality of life (Filipo et al. 2004; Wiley et al. 2005; Lin & Niparko 2006; Sach & Barton 2007; Schorr et al. 2009), or evaluate the degree of parent satisfaction (O'Neill et al. 2004; Wiley et al. 2005; Lin & Niparko 2006). Questionnaires administered both before and after implantation try to evaluate the benefits derived from cochlear implants.

These questionnaires and other recent studies report that, for deaf children with additional disabilities, improvement in speech perception, speech intelligibility, and verbal skills after implantation is slow and sometimes modest (Hamzavi et al. 2000; Pyman et al. 2000; Waltzman et al. 2000; Fukuda et al. 2003; Holt & Kirk 2005; Edwards 2007; Nikolopoulos et al. 2008; Wiley et al. 2008), thus requiring longer follow-up periods (Vlahovic & Sindija 2004; Nikolopoulos et al. 2008). Nevertheless, cochlear implants also provide benefits regarding increased connectivity and interest in the environment, social interaction, and improvement in other psychological skills that contribute to significantly improving their quality of everyday life (Hamzavi et al. 2000; Waltzman et al. 2000; Donaldson et al. 2004; Filipo et al. 2004; Meinzen-Derr et al. 2010). Therefore, if only those scores, obtained in tests assessing speech perception and language development, are taken into account, the benefits of cochlear implants are likely to be underrated.

The results of the abovementioned studies point out not only the need to assess the improvements in sound and speech perception and language abilities, but also the need to assess cognitive functions and social and relational skills. This article presents a newly developed assessment tool that aims to quantify the changes caused by CI in deaf children with additional disabilities. More precisely, the tool is intended to measure changes in behavior during everyday activities that require specific neuropsychological and perceptual skills. These changes are eventually intended to provide an indication of the improvement in quality of life derived from CI.

MATERIALS AND METHODS

The new assessment tool has been developed based on the concept that an improvement in the quality of life of deaf children with cochlear implants derives from increased well-being in the following domains: physical, neuropsychological, and social (Hinderink et al. 2000). For each of these domains, we have taken into account the most relevant skills, and have

grouped them into five areas, as shown in Table 1. The new tool (henceforth referred to as the Deafness and Additional Disabilities questionnaire, [DAD questionnaire] or [DADQ] for short) evaluates the children's skills in these five areas. The questions have been chosen following the "Guttman scaling format" (Judd et al. 1991) and the "constructing Likert scales" criteria (Streiner & Norman 2003), and they have been phrased in a similar way using simplified language.

A number of questions have been selected from existing questionnaires (Chmiel et al. 2000; Donaldson et al. 2004; Nikolopoulos et al. 2004; Wiley et al. 2005; Barton et al. 2006; Damen et al. 2006; Stacey et al. 2006; Berrettini et al. 2008; Schorr et al. 2009) and from some parts of the Battelle Developmental Inventory Screening Test (Glascoe & Byrne 1993). We have subsequently adapted them to our own needs, taking into account the traits observed in children with multiple impairments without a hearing condition. Yet, we have phrased most of the questions based on our experience gained during clinical observations of these children and on observed improvements in everyday activities, as reported by parents during clinical interviews.

The DADQ has been designed as a closed-format questionnaire containing questions related to behaviors that can be found in everyday activities. These questions have been grouped into the five abovementioned areas and cover the physical, neuropsychological, and social domains. Each question is formulated as a statement with a 5-point response scale, referring to a certain behavior. The respondent is asked to report as many examples as they can recall for that behavior. From the descriptive answers and the number of examples, a therapist assigns a numerical score, corresponding to the reported frequency of the behavior: never = 0 (the child never showed this behavior), seldom = 1 (the respondent reported one or two examples), sometimes = 2 (three examples), often = 3 (four or more examples), always = 4 (the child always showed this behavior).

By independently summing the scores, partial scores can be obtained for each area. Their sum, that is, the global score, is indicative of the child's ability in carrying out everyday activities. The difference between these partial scores in subsequent tests can be used to track changes.

DAD Questionnaire

The DAD questionnaire (Appendix A, Supplemental Digital Content 1, <http://links.lww.com/EANDH/A85>) comprises the following five areas:

Perceptual Skills (DADQ-A) • Ten questions address perceptual skills, that is, use of the hearing aid or cochlear implant, perception and identification of environmental sounds, and speech perception in quiet and noisy environments. The

TABLE 1. The five areas of the Deafness and Additional Disabilities Questionnaire, together with their related domain, the type of skills assessed and the score ranges

Domain	Area	Skills	Range
Physical	DADQ-A	Perceptual skills	0–40
Physical	DADQ-B	Preferred communication mode	0–8
Neuropsychological	DADQ-C	Communicative behaviors	0–36
Neuropsychological	DADQ-D	Attention and memory skills	0–32
Social	DADQ-E	Social interaction, control of behavior, and self-government	0–56
Global	DADQ-Tot	Total score	0–172

TABLE 2. Possible answers to the DADQ B question, “What is the preferred way the child uses to communicate?”

Score	Communication Mode Using	Abbreviation
0	Behavior (e.g., smiles, cries, facial expressions)	B
1	Gestures	G
2	Gestures and vocalization	G+V
3	Sign language or augmentative and alternative communication	SL
4	Gestures associated to syllables	G+S
5	Words associated to gestures	G+W
6	Single words	SW
7	Association of words (i.e., putting two or more words together)	WA
8	Complete sentences	CS

DADQ, Deafness and Additional Disabilities Questionnaire. The abbreviations are those used in Figures 1 and 3.

following are a few example questions: “Answers if called in a noisy place?”, “Hears the noise of home appliances?”, “Realizes that someone is talking even if the child does not see him/her?”, and “Recognizes an animal by its cry?”

Preferred Communication Mode (DADQ-B) • This area contains only one question: “What is the preferred way the child uses to communicate?” The possible answers are attributed a score between 0 (use of behavior) and 8 (use of complete sentences). See Table 2 for a complete list.

Communicative Behaviors (DADQ-C) • Nine questions address the child’s communicative behaviors with familiar talkers and strangers, in terms of communicative intent, communicative efficiency, and vocal or gestural turn taking. A few example questions: “Respects the turns of communication?”, “Tries to correct him/herself if persons do not understand (using any communicative mode)?”, “Can communicate his/her needs (using any communicative mode)?”, or “Can communicate on the telephone?” By these questions we mean the ability to communicate at a level and complexity that corresponds to the child’s age and language development. For example, for very small children, with regard to communication on the telephone, the ability of greeting and of providing very short answers to simple questions was considered sufficient.

Attention and Memory (DADQ-D) • Eight questions are directed to focused and selective attention and to long-term memory skills. The following are a few example questions: “Able to focus attention on a task for a time sufficient to carry it out?”, “Able to keep his/her attention on a task in the presence of interference stimuli and to carry out the task?”, “Able to remember the meaning of high frequency words?”, and “Capable of retaining the rules of a simple game?”

Social Interaction, Control of Behavior, and Self-Government (DADQ-E) • Fourteen questions address the child’s behavior during interaction with family and strangers, and with siblings and peers, his/her control of emotional needs, and his/her independence during everyday activities. The following are a few example questions: “Acts independently during everyday activities?”, “Spontaneously starts an activity (e.g., chooses and propose games him/herself)?”, “Happy to be in somebody’s company?”, and “Interacts with family and siblings in an appropriate way?”

Participants

This study presents the results for 50 deaf children with additional disabilities. The children were selected for CI at the Otolaryngology and Cochlear Implant Centre of Pisa, the Audiology Unit of Treviso, Padua University Hospital, the Audiology Unit University of Ferrara, and the Audiology Unit University of Modena.

All children met the following criteria: (a) less than 18 years of age, (b) severe-to-profound deafness since prelingual age (congenital deafness or hearing loss sustained before 6 months of age), (c) had used hearing aids for at least 6 months, (d) had received at least 6 months of oral rehabilitation training before implantation, and (e) had shown significant additional disability. A few children had been suffering from progressive hypoacusis. All had limited benefits from hearing aids. The age at implantation ranged from 22 months to 13.3 years, with an average of 4.1 years and a median of 3.5 years. Twenty children (40%) were male and 30 (60%) were female.

Preoperatively all the children underwent a comprehensive audiological evaluation and a neuropsychiatric examination, as described by Chilosi et al. (2010). The diagnosed additional disabilities, based on the *Diagnostic and Statistical Manual of Mental Disorders IV* (American Psychiatric Association 2000), were classified into categories (Table 3, Additional Disability): intellectual disabilities (nonverbal intelligence quotient [IQ] ≤ 70 on the Leiter international scale); behavioral and mood disorders including attention-deficit hyperactivity disorder (on the basis of clinical evaluation in multiple contexts and rating scales questionnaires, such as Child Behavior Checklist total score > 70 and Conners rating scales); pervasive developmental disorders including autistic spectrum disorders (on the basis of clinical evaluation and Childhood Autism rating scale ≥ 30 or Autism Diagnostic Observation Schedule ≥ 7); neurological syndromes with central nervous system involvement including cerebral palsy (on the basis of clinical and instrumental evaluation, including cerebral magnetic resonance imaging), TORCH encephalopathy, epilepsy, and central nervous system malformations. For those children who had multiple diagnoses,

TABLE 3. Study characteristics

	N (%)
Gender	
Female	30 (60)
Male	20 (40)
Additional Disability	
Intellectual disability	29 (58)
Attention deficit without hyperactivity disorder	7 (14)
Cerebral palsy	6 (12)
Attention deficit with hyperactivity disorder	5 (10)
Pervasive developmental disorder	2 (4)
Epilepsy	1 (2)
Etiology	
Prematurity	11 (22)
Congenital cytomegalovirus	10 (20)
Perinatal asphyxia	6 (12)
Meningitis	3 (6)
Congenital rubella	1 (2)
Down syndrome	1 (2)
Waardenburg syndrome	1 (2)
Cochlea malformations	1 (2)
Toxoplasma infection	1 (2)
Unknown	15 (30)

TABLE 4. Additional study characteristics

	Age at Implantation	Hearing Age	Cognitive Level (Intelligence Quotient)	N (%)
Mean	4.1 yrs	3.0 yrs	Normal (>70)	6 (12)
Median	3.5 yrs	3.0 yrs	Mild cognitive delay (53–70)	32 (64)
Range	22 mos–13.3 yrs	6 mos–8.3 yrs	Moderate cognitive delay (38–52)	8 (16)
			Severe cognitive delay (23–37)	4 (8)

only the main one is reported. The IQ of the children ranged from 36 to 107, with a median of 64, and was also classified into cognitive levels according to the DSM-IV intervals (see Table 4, Cognitive Level [Intelligence Quotient]). As can be seen from Tables 3 and 4, 44 children had cognitive impairment, but only for 29 of them intellectual disability was the main diagnosis. More precisely, in our population there were 15 children with IQ lower than normal and a more severe nonintellectual disability (14 had mild disability: 7 attention deficit, of which 4 were with hyperactivity disorder and 3 without, 5 had cerebral palsy, 2 had pervasive developmental disorder; 1 had moderate disability: cerebral palsy).

In addition to the DADQ, the following tests were used to assess the children: P.C.A.P., TIPI1, TIPI2, and two-syllabic word lists (Arslan et al. 1997; Burdo 1997). The results of these tests were used as an Italian-language equivalent of the Early Speech Perception (ESP) test for evaluating speech perception on the six-level scale devised by Moog and Geers (1990). Moreover, the level of spontaneous language organization was defined according to a six-level rating system called Grid Analysis of Spontaneous Speech (GASS; Cipriani et al. 1993; Chilosi et al. 2007). The data obtained in these additional tests were used to evaluate the psychometric validity of the DADQ. Those tests were not intended to be part of the DADQ. The assessment of cognitive abilities was performed using Leiter international scales.

The DADQ, GASS, and ESP tests were administered twice, before and after CI, to evaluate whether significant improvements had taken place. However, in 13 cases (26%), the pre-CI skills could only be assessed retrospectively in face-to-face interviews, because the children were implanted before the development of the DADQ. At the time the post-CI tests were administered, the children had been using their cochlear implants for at least 6 months: their so-called hearing age ranged from 6 months to 8.3 years (average and median 3.0 years).

Statistical Methods

We calculated the difference between the post-CI and the corresponding pre-CI score for each patient and test. Wilcoxon signed-rank test and Spearman rank correlation coefficients were used for the statistical analysis of the results. Statistical significance was accepted at the $p < 0.05$ level.

The R program, by The R Foundation for Statistical Computing (Vienna, Austria), was used for computation of numerical data and for drawing the figures. The radar plot in Figure 4 was drawn with Microsoft Excel (Redmond, WA).

RESULTS

The box plots in Figures 1 and 2 summarize the results obtained by the DADQ, GASS and ESP tests of the 50 children. The boxes correspond to the lower and upper quartiles (25th

and 75th percentiles, respectively), the horizontal bold line represents the median, the whiskers correspond to the lowest datum still within 1.5 interquartile ranges of the lower quartile and to the highest datum still within 1.5 interquartile ranges of the upper quartile, and the circles represent outliers. The pairs of boxes in Figure 1 represent the results obtained before and after CI for the DAD questionnaire (areas A–E and total score), the GASS level and the ESP test score. To ensure that all results were plotted on the same scale, the scores in each category were normalized (i.e., divided by the respective maximum score) so that each bar has a maximum value of 100%. For example, a score of 30 for DADQ-A corresponds to a normalized score of 75% (30/40). The maximum scores for the DADQ are shown in the last column of Table 1; the maximum scores for the GASS and ESP were 5 and 6, respectively. For patients who were unable to complete the GASS-level measurement, a score of –1 was arbitrarily assigned (which yielded a negative normalized score). DADQ-B values are plotted on separate scales, to represent the actual categories, that is, communication modes, rather than the numerical scores (see Table 2 for a list of abbreviations).

Figure 2 shows the descriptive statistics of the differences between the score obtained by each patient at the post-CI assessment and the corresponding score obtained at the pre-CI assessment. The differences have been normalized for each DADQ area and test, as for Figure 1 (i.e., post-CI score minus pre-CI score, divided by the maximum score). The differences in scores are non-negative for each patient, which means that the majority of patients scored better at the post-CI assessment than at the pre-CI assessment. The improvements were statistically significant for all tests ($p < 0.0001$, Wilcoxon signed-rank test).

Table 5 shows whether there is correlation between the improvements recorded by the different tests and whether these improvements correlate with the children's age at CI, hearing age (age since CI), age at the post-CI test, and IQ. As pointed out in the Discussion section, the different tests often agreed in detecting improvements (positive correlation); however, only few of these improvements correlated with time-related factors and IQ. The reason why we report correlations with post-CI improvements, rather than with absolute post-CI scores, is that we have found that potentially confounding factors such as IQ and age have less influence on the former. Therefore, we believe that post-CI improvements are a more neutral indicator for evaluating benefits of CI for our population, which includes a wide range of ages and IQs. Moreover, items that are too complex for certain subjects in terms of age and cognitive level do not influence the results: for these items, subjects typically get the lowest score both at the pre-CI and the post-CI test; the difference in score is therefore zero, and zero differences are excluded from the calculation of the signed rank tests.

Table 6 shows the frequency distribution related to the usage of hearing aids (pre-CI) and cochlear implants (post-CI). Assuming that the scale of answers is linear (never = 0%,

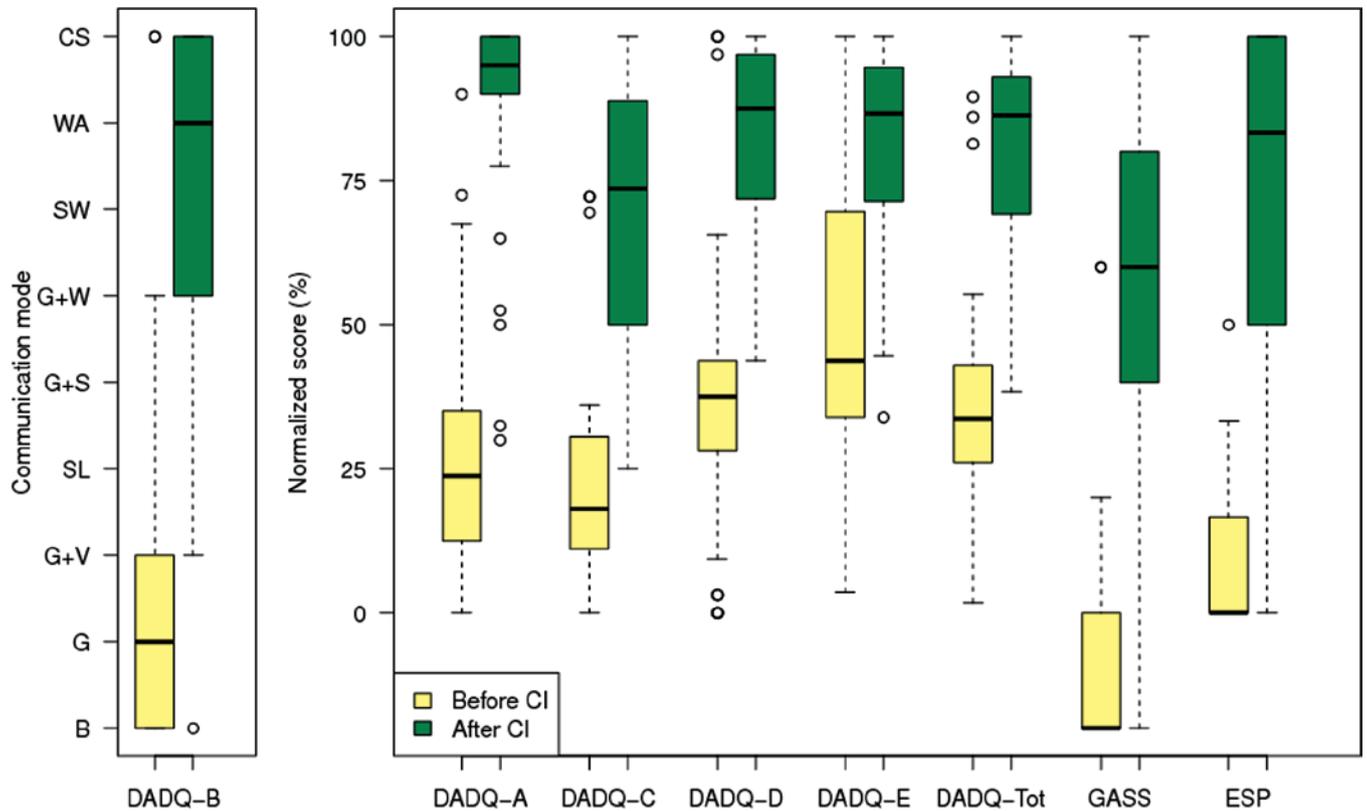


Fig. 1. Scores obtained before and after CI in the DADQ (areas A–E and total score), GASS level and ESP test. All the values are normalized, that is, they have each been divided by their respective maximum score, so that each bar has a maximum value of 100%. Subjects who were unable to complete the GASS-level measurement were assigned level –1, which corresponds to a negative normalized score. See Table 2 for the meaning of the labels of the DADQ-B scale. CI, cochlear implantation; DADQ, Deafness and Additional Disabilities Questionnaire; GASS, Grid Analysis of Spontaneous Speech; ESP, Early Speech Perception.

seldom = 25%, sometimes = 50%, often = 75%, always = 100%), the children used their implant on average 97.5% of the time; conversely, before CI, their average daily use of hearing aids was only 65.8%. These data are also represented as a box plot in Figure 3. Furthermore, Figure 3 shows the improvements related to certain skills for which parents have high expectations, such as communication mode (DADQ-B), the ability to communicate on the telephone at a level commensurate to their age, and speech intelligibility to familiar and unfamiliar listeners. DADQ-B was plotted separately to show the number of children for each communication mode (see Table 2 for a list of abbreviations). The radar plot in Figure 4 shows the median values, expressed as percentages of the maximum scores, obtained by the 50 children in the five areas of the DAD questionnaire, before and after CI. The overall improvement in the median scores is clearly shown by the change in size of the polygons.

The final questions of the DADQ are related to parent satisfaction. Of the 50 parents, 47 felt that they had received enough information from the CI center to make a decision regarding implantation, 48 parents would recommend a CI for a child with similar impairments, and 47 would opt again for implantation.

DISCUSSION

The results show significant improvements in all areas investigated by the DAD questionnaire and suggest that these deaf

children with additional disabilities have had benefits, after CI, on a wide spectrum of abilities, such as cognitive, social, relational, hearing, and linguistic skills (Wilcoxon $p < 0.0001$). As shown in Figure 4, the change in size of the polygons is indicative of the children's improvements in carrying out everyday activities after CI.

These data are in agreement with previously published results on implanted deaf children with additional disabilities. Using a questionnaire about changes in children's behaviors Vlahovic and Sindija (2004) found positive life changes, as reported by the parents of these children. Donaldson et al. (2004), in a survey addressing parents of deaf children with autism spectrum disorders, found positive benefits including changes in behavior and communication, increased awareness of the environment, reactions to music and sound, vocalization, eye contact, use of sign language, and response to requests. Filipo et al. (2004) stated that, in children with special needs, CI could improve quality of life, increasing both listening and communication skills, and self-sufficiency. Waltzman et al. (2000) found that, in addition to improved auditory and communication skills, social interactions and a general "connectedness" to the environment were visible on a daily basis. Wiley et al. (2005) analyzed the qualitative benefits, perceived by families, of the progress in communication of implanted children with additional disabilities, their awareness of environmental sounds, and their attentiveness and interest in the world around them. Berrettini et al. (2008) agreed to

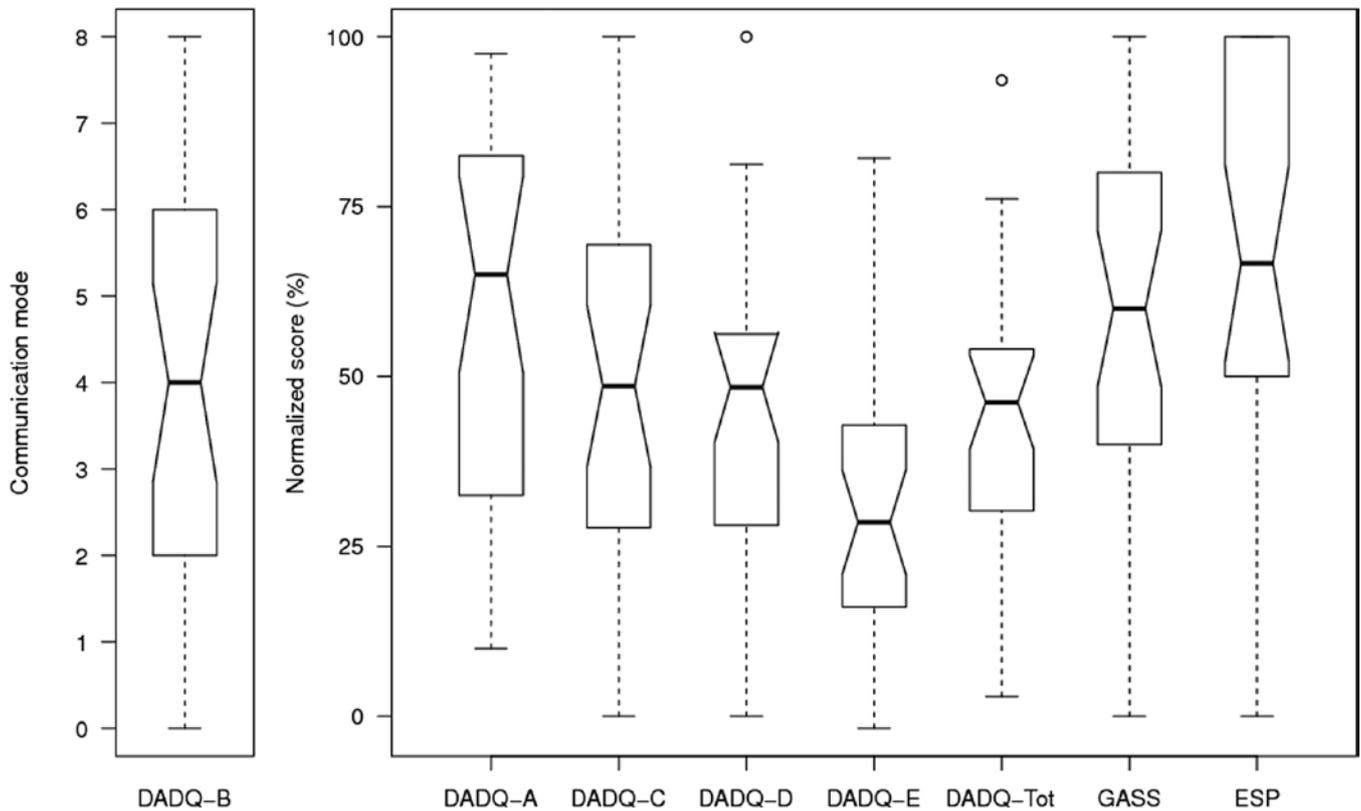


Fig. 2. Improvements in scores obtained after cochlear implantation at the DADQ (areas A-E and total score), GASS level and ESP test. All the values are normalized, that is, they have each been divided by their respective maximum score, so that each bar has a maximum value of 100%. DADQ, Deafness and Additional Disabilities Questionnaire; GASS, Grid Analysis of Spontaneous Speech; ESP, Early Speech Perception.

the fact that CI leads to positive results in terms of speech perception, communication abilities, and improvements in quality of life in such children.

As shown in Figure 2, in our questionnaire, the largest improvements were observed in perception skills (DADQ-A), followed by preferred communication mode (DADQ-B), communicative behaviors (DADQ-C), and attention and memory skills (DADQ-D). The scores of social interaction, control of

behavior, and self-government (DADQ-E) improved significantly less (Wilcoxon $p < 0.0001$). Moreover, we have also found negative correlation between post-CI improvements and pre-CI scores in the DADQ areas (Spearman $r \leq -0.46, p < 0.01$), which points out that there is a plateau effect at the upper levels of all DADQ areas: if skills are high in the beginning then they may remain high post-CI and changes may be minor.

TABLE 5. Values of the Spearman rank correlation coefficient and corresponding p values

	DADQ-A		DADQ-B		DADQ-C		DADQ-D		DADQ-E	
	Coefficient	p	Coefficient	p	Coefficient	p	Coefficient	p	Coefficient	p
DADQ-B	0.70	<0.0001	—	—	—	—	—	—	—	—
DADQ-C	0.66	<0.0001	0.82	<0.0001	—	—	—	—	—	—
DADQ-D	0.65	0.0001	0.58	0.0008	0.62	0.0002	—	—	—	—
DADQ-E	0.32	0.08	0.51	0.004	0.47	0.008	0.65	0.0001	—	—
DADQ-Total	0.79	<0.0001	0.81	<0.0001	0.81	<0.0001	0.87	<0.0001	0.73	<0.0001
Grid Analysis of										
Spontaneous Speech	0.65	<0.0001	0.89	<0.0001	0.75	<0.0001	0.51	0.004	0.39	0.04
Early Speech Perception	0.80	<0.0001	0.65	0.0001	0.66	<0.0001	0.47	0.009	0.29	0.12
Age at cochlear implantation	-0.22	0.26	-0.23	0.23	-0.13	0.51	-0.29	0.12	-0.43	0.02
Hearing age	0.31	0.1	0.47	0.01	0.45	0.015	0.34	0.08	0.36	0.06
Age at post-cochlear										
implantation	0.02	0.92	0.11	0.56	0.14	0.47	0.04	0.86	-0.02	0.94
Intelligence quotient	0.22	0.46	0.38	0.02	0.32	0.12	0.1	0.33	-0.03	0.40

DADQ, Deafness and Additional Disabilities Questionnaire.

Correlations between the improvements obtained in the various areas of the Deafness and Additional Disabilities Questionnaire, Grid Analysis of Spontaneous Speech level, Early Speech Perception score, age at cochlear implantation, hearing age, age at the post cochlear-implantation test, and intelligence quotient.

TABLE 6. Frequency of usage of hearing aids and cochlear implants

Usage Frequency	Hearing Aids, N (%)	Cochlear Implants, N (%)
Never	3 (6)	0 (0)
Seldom	8 (16)	0 (0)
Sometimes	5 (10)	0 (0)
Often	20 (40)	5 (10)
Always	14 (28)	45 (90)

The gains observed in language and communication skills (DADQ-B and DADQ-C) correlate positively with hearing age, that is, the time period elapsed between implantation and the post-CI test (Spearman $r \geq 0.45$, $p < 0.02$). Conversely, perception skills (DADQ-A), attention and memory skills (DADQ-D), and social interaction, control of behavior, and self-government (DADQ-E) show no correlation with hearing age (Spearman $r \leq 0.36$, $p > 0.05$). This finding is to be ascribed in part to the rehabilitative approach, together with the limited length of the follow-up. In the first phase after implantation, rehabilitation is focused on perceptive skills and development of oral language. Perceptive skills show an immediate, sharp improvement after CI; however, subsequent gains are slow to be achieved and require continuous rehabilitation. This latter phase is likely not detectable in our children because the relatively short follow-up period (3 years in average) did

not allow them to reach higher levels of speech perception such as open-set word recognition. However, language and communication skills do not show an initial boost, but improve more steadily. A further point of consideration is that slow or incomplete progress of language and communication skills may also be a result of limitations deriving from other underlying impairments, for example, severe dysarthria. Last, attention and memory skills are scarcely rehabilitated in these children and this may be the reason for the lack of correlation. Data in the literature show slow progress over time and the need for long follow-up times and for several years of rehabilitation to observe significant improvements in children with additional disabilities (Waltzman et al. 2000; Rajput et al. 2003; Vlahovic & Sindija 2004; Hans et al. 2010). Wiley et al. (2008) conclude that such children make progress in auditory skills but may not reach higher levels of identification and comprehension. Regarding the fact that hearing age does not significantly correlate with social interaction, control of behavior, and self-government skills (DADQ-E), we are in agreement with Filipo et al. (2004), who assert that family and social relationships tend to remain stable, because they depend on more numerous and complex variables, which can only partially be controlled. Furthermore, these skills are seldom rehabilitated. Regarding other age-related factors, age at CI negatively correlated with improvements in all areas of DADQ, which generally indicates that children who are implanted later improve less or more slowly than children who are implanted earlier. This result is

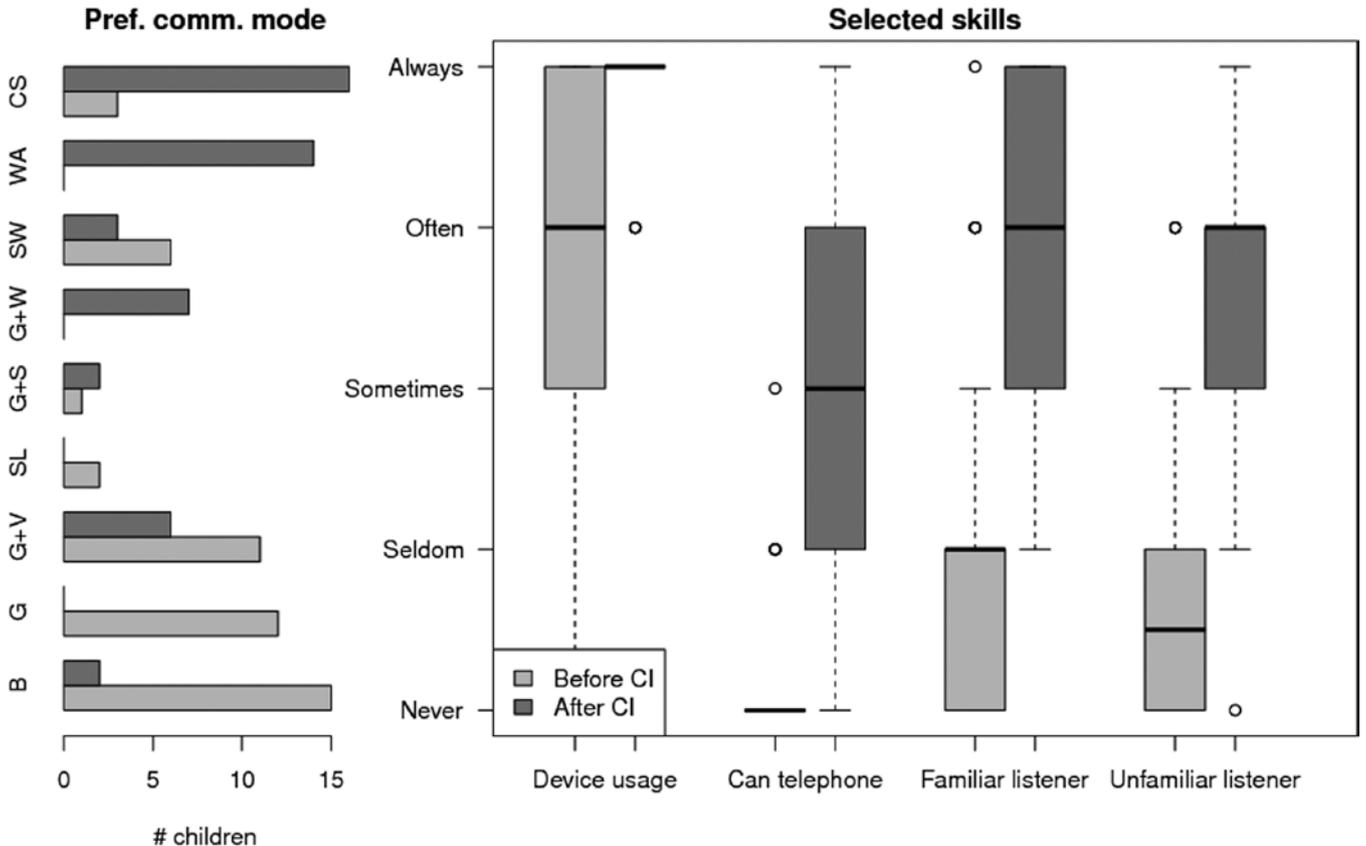


Fig. 3. Preferred communication mode (see Table 2 for the meaning of the labels of the Deafness and Additional Disabilities Questionnaire-B scale) and frequencies related to selected skills before and after implantation: device usage, ability to have an age-commensurate telephone conversation, and speech intelligibility by familiar and unfamiliar listeners. CI, cochlear implantation.

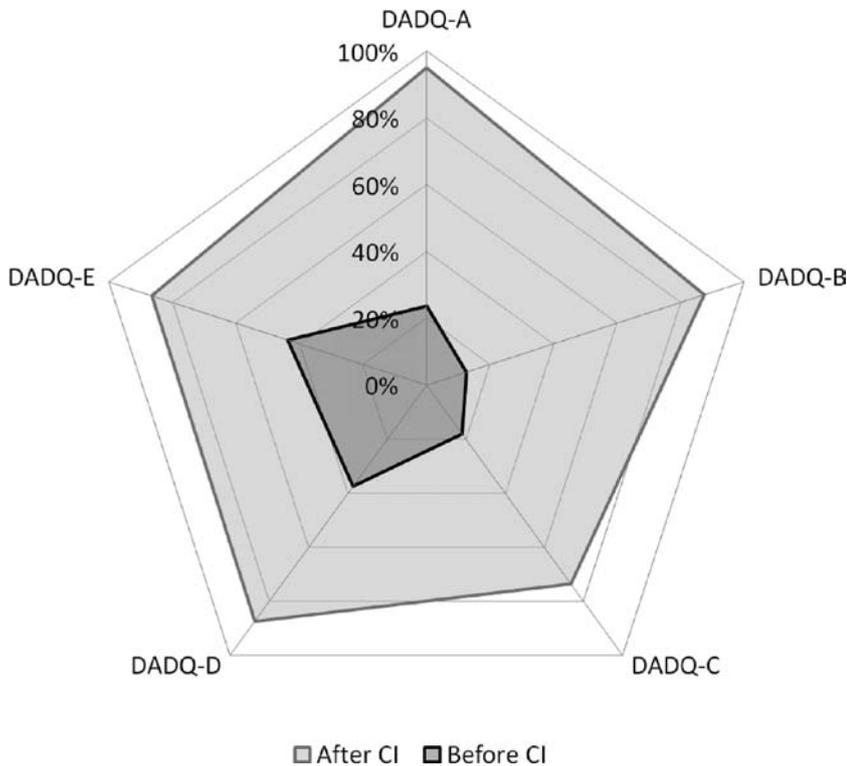


Fig. 4. Median values, expressed as percentages of the maximum scores, obtained by the 50 children in the five areas of the DADQ before and after CI. DADQ, Deafness and Additional Disabilities Questionnaire; CI, cochlear implantation.

in agreement with several studies that regarded as essential providing cochlear implants to children as early as possible (Kral et al. 2001; Sharma et al. 2005; Waltzman & Roland 2005). However, the negative correlation is only significant for DADQ-E (Spearman $r = -0.43$, $p = 0.02$). This could be again because of the lack of rehabilitation of those skills. Conversely, age at the post-CI test does not correlate with any improvement in the DADQ areas (Table 5).

The IQ correlates positively with the preferred communication mode (DADQ-B) and the GASS level (Spearman $r = 0.38$ and 0.58 , p values 0.05 and 0.02 , respectively), even after having accounted for variable hearing age by means of multiple regression ($p \leq 0.01$). This result may be directly related to the fact that obtaining high scores in these two tests requires higher levels of linguistic and cognitive skills, which are seldom attained by individuals with significant intellectual disability. It is worth noting that in the case of DADQ-C, which is also partly directed to language skills, the p value is approximately 0.1 , but the correlation coefficient ($r = 0.32$) clearly indicates a positive correlation. The fact that in our population most children had mild cognitive delay may have influenced the p values of correlation coefficients. Indeed, the Mann-Whitney test shows more significant differences between the post-CI improvements of the group of children with normal cognitive levels and the group with moderate or severe intellectual disability for all tests assessing language and communication skills: DADQ-B and DADQ-C ($p = 0.01$) and GASS ($p < 0.05$). Similar conclusions can be drawn from the comparison of the group of children having mild cognitive delay with those having moderate-to-severe cognitive delay and from corresponding statistical tests on absolute post-CI levels. Conversely, the other areas of the DADQ are not directed at the assessment of higher levels of skills, such as memory, attention, social interaction, etc. Therefore,

children with mild and moderate cognitive delay can, after suitable rehabilitation, also obtain good scores in these tests. We are presently including further patients in our study with the aim of drawing clearer conclusions on the effects of cognitive disability on CI.

Concerning preferred communication mode (DADQ-B), the number of children in each category, before and after CI, are shown in Figure 3 (labels are explained in Table 2): before CI, 82% of children used behavior, gestures, or vocalization to communicate and only 18% used oral language, whereas after implantation, 80% the children used oral language, of which 28% used association of words and 32% used complete sentences.

The literature reports fewer improvements in communication mode for these special cases. In the study by Wiley et al. (2005), only 12% of the children gained oral speech and language abilities, although all children broadened their communication skills. Vlahovic and Sindija (2004) and Donaldson et al. (2004) reported poor and slow improvements in speech development. Better results are reported in the study by Waltzman et al. (2000) in which 59% of special cases used oral language after CI.

Cochlear implants were more extensively used during waking hours than hearing aids were (Table 6 and Fig. 3): this provides major evidence of benefits perceived by the children. This result is in agreement with previous studies (Summerfield & Marshall 1995; Wiley et al. 2005; Berrettini et al. 2008; Bacciu et al. 2009; Hans et al. 2010). The remaining part of Figure 3 shows results related to the ability to communicate on the telephone at a level commensurate to their age and speech intelligibility to familiar and unfamiliar listeners. Before implantation, no child was able to use the telephone, whereas after CI, 14% were able to communicate on the telephone frequently and 22%

at all times (Fig. 3). Bacciu et al. (2009) reported that two out of five children with cerebral palsy were able to use the telephone with a familiar talker after CI. Concerning speech intelligibility, before CI only 16% of children were understood by familiar listeners and 8% by unfamiliar listeners, whereas after CI, 74% were understood by familiar listeners and 22% by all. This result is in agreement with the findings by Nikolopoulos et al. (2008) who reported that after CI 70% of children with additional disabilities developed connected intelligible speech, which was, however, intelligible to people with little or no experience of deaf person's speech in only 16% of cases. Also, Bacciu et al. (2009) found that there was an improvement in speech intelligibility over time with an increased use of CI, but only one out of five children had connected speech intelligible to all listeners.

Parents of deaf children who elect for implantation usually have the expectation that their child will develop intelligible speech. In many cases this goal was not achieved in our study, possibly because of the additional disabilities. Nevertheless, 93% of the parents were satisfied with the progress of their children and would recommend CI to parents with a similar child. In our opinion, this reflects the importance we devoted to counseling, before and after implantation, to set realistic expectations.

Last, the GASS level and ESP score in post-CI assessments were significantly higher than the corresponding values before CI (Wilcoxon $p = 0.003$ and 0.0001 , respectively). However, it has to be pointed out that 28 children were unable to complete the GASS-level measurement before CI (one of them could not complete it after CI also). In several cases, administering the ESP test and measuring the GASS level have been difficult. Regarding the ESP test, before implantation, 32 children did not show any sign of speech detection and were therefore assigned level 0 on the ESP scale (2 children had level 0 also after CI). However, interpreting whether the absence of response was because of an absence of detection, rather than other factors (such as behavioral deficit, impaired attention, lack of understanding of the task, etc.), has not always been straightforward. Yet, with the DADQ, it has been possible to obtain information about perceptual skills (DADQ-A) and preferred communication mode (DADQ-B) in all cases. Our test is based on the frequency with which parents observe certain types of behaviors during the everyday activities of their children. These data do not depend on the comprehension of an abstract task and, although being obtained indirectly, nevertheless allow the inference of speech-perception skills.

Correlation analysis between DADQ, ESP, and GASS shows positive correlation between tests that assess perceptive skills (DADQ-A versus ESP, Spearman $r = 0.80$, $p < 0.0001$) and between tests that assess perception and communication skills (any one of DADQ-A, DADQ-B, and DADQ-C versus GASS, Spearman $r \geq 0.65$, $p < 0.0001$). In other words, there is positive correlation between tests assessing similar skills. This allows the assertion that the DADQ can be a worthy alternative in cases where the usage of the GASS measure or the ESP test are not possible, or where there are doubts about the interpretation of collected data.

CONCLUSIONS

In recent years, the literature about deaf children with additional disabilities who received cochlear implants has increased substantially. Many studies have assessed the gains in specific

skills such as hearing and speech perception, language development, and speech intelligibility. Few studies focused on joint attention, symbolic play, self-sufficiency, and emotional, social, and family relationships. All these studies generally report a positive influence of CI on the assessed skills.

In our study, we observed that activation of the auditory sensory canal, previously absent or deficient, provided benefits not only for perception, communication, and language skills, but also for attention and memory skills, social interaction, and for control of behavior and self-government.

The newly developed DADQ has a simple-to-use format and it has been proven to be sufficiently sensitive for the detection of changes in each examined area. Therefore, we think that it can be a useful tool to assess improvements in deaf children with additional disabilities, even in cases in which such improvements are not detected by other tests routinely used in clinical practice. Although additional research is necessary to further refine the DADQ, this type of assessment is eventually intended to provide an indication of the improvement in quality of life derived from behavioral changes.

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