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Cochlear implantation in deaf children with associated disabilities: Challenges and outcomes

Key Words
Cochlear implant
Sensorineural hearing loss
Multiply impaired deaf children
Quality of life
Qualitative benefits
Questionnaire

Abbreviations
SNHL: Sensorineural hearing loss
CI: Cochlear implant
IQ: Intelligence quotient

Abstract
The issue of cochlear implantation in deaf children with associated disabilities is an emerging subject. Currently, there is no consensus on whether to implant children with multiple impairments; moreover, it may be difficult to evaluate these children with standard tests pre- or post-implantation. In addition, these children often have poor speech perception and language skills, making assessment more difficult. Despite these factors, these children often receive important benefits in daily life, with an overall improvement in quality of life. In the present study, post-implant outcomes of 23 profoundly deaf children with neuropsychiatric disorders were analysed, using objective measures of speech perception, and a questionnaire administered to the parents, aimed at evaluating the benefits in daily life after implantation. The results were quite variable, but overall positive, in terms of speech perception, communication abilities, and improvement in quality of life. The findings add an additional piece of evidence to support the effectiveness of cochlear implantation in these special cases.

Sumario
El tema de la implantación coclear en niños sordos con discapacidades asociadas es nuevo. No existe consenso en este momento sobre cuándo debe implantarse un niño con impedimentos múltiples; por otra parte, puede ser difícil evaluar a estos niños con las pruebas estándar, antes o después de la implantación. Además, tienen pobre tanto su percepción del lenguaje como sus habilidades lingüísticas, por lo que la evaluación es aún más difícil. A pesar de estos factores, estos niños frecuentemente reciben beneficios importantes en la vida diaria con una mejoría general en su calidad de vida. En el presente estudio, se analizaron los resultados post-implantación de 23 niños con sorderas profundas y trastornos neuro-psiquiátricos, usando mediciones objetivas de la percepción del lenguaje y un cuestionario aplicado a los padres, para valorar los beneficios en la vida diaria después de la implantacicœn. Los resultados fueron bastante variables pero en general fueron positivos, en términos de percepción del lenguaje, de habilidades comunicativas y de mejor calidad de vida. Los hallazgos agregan una evidencia más para apoyar la efectividad de la implantación coclear en estos casos especiales.

Approximately 30–40 percent of children with sensorineural hearing loss (SNHL) have additional disabilities (Filipo et al, 2004; Fortnum et al, 2002; Hamzavi et al, 2000; Holden-Pitt & Albertorio 1998; Lesinski et al, 1995). These children are more challenging for professionals in terms of audiological diagnosis, amplification, cochlear implantation, and rehabilitation. The issue of cochlear implantation in children with associated disabilities is an emerging one, and with expanding criteria for cochlear implants (CIs) and growing longitudinal experience, many centres are now making CI technology available to deaf children with additional disabilities, which may influence post-implant outcomes (Donaldson et al, 2004; Filipo et al, 2004; Fukuda et al, 2003; Hamzavi et al, 2000; Holt & Kirk 2005; Lesinski et al, 1995; Pyman et al, 2000; Waltzman et al, 2000; Wiley et al, 2005).

Currently there is no consensus in the literature or among CI centres on whether to implant children with multiple impairments. When deafness is the sole disability, the decision of implantation should be based on the expected benefits; however, what constitutes benefit is still an open question for individuals with additional disabilities, and it is unclear whether benefit should be considered in terms of speech and language gains or in terms of psychosocial development and improved quality of life. In fact, these children often achieve lower scores on measures of speech and language development than deaf children without additional disabilities, and only a subset of them achieve open-set recognition abilities and oral language skills (Donaldson et al, 2004; Hamzavi et al, 2000; Holt & Kirk 2005; Lesinski et al, 1995; Pyman et al, 2000; Waltzman et al, 2000). Despite this, there is a general report of improved quality of life and increased connectedness and interest in the environment and social interactions (Filipo et al, 2004; Fukuda et al, 2003; Hamzavi et al, 2000; Waltzman et al, 2000; Wiley et al, 2005).

To this regard, Pyman et al (2000) evaluated speech perception development in 20 deaf children with motor and/or cognitive delays out of a group of 75 paediatric CI recipients. They reported that these children progressed slower than children without additional impairments, and some of them never achieved open-set speech recognition abilities.

In the same year, Waltzman et al studied speech perception abilities in 29 deaf children with a wide range of additional disabilities who received a CI. Relative to a control group of

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individuals without additional disabilities who received a CI, fewer of the children with multiple disabilities were able to complete the speech perception tests, and when they could, their average scores were lower than those of the children without additional disabilities. The results of the study suggested that the children with multiple disabilities obtained demonstrable benefits from implantation, based on anecdotal observations of increased social interaction and connectedness to the environment (Waltzman et al, 2000).

Hamzavi et al (2000) examined speech perception abilities and auditory behaviour in ten children with multiple impairments who received a CI, by means of the evaluation of auditory responses to speech (EARS) test battery in German. The results showed a wide variability of outcomes, reflecting the heterogeneity of the impairments associated to deafness: Nine out of ten children obtained some kind of benefit from a CI, and six out of ten also achieved some level of word recognition and production after three years of implant use (Hamzavi et al, 2000). More recently, Fukuda et al (2003) reported language development and overall benefits after implantation in a single paediatric case with deafness and moderate developmental delays. In 2004, Filipo et al conducted a study on 18 deaf children with multiple impairments who received a CI; these children presented not only neuropsychiatric defects, but some of them were affected by other 'special' conditions such as visual impairment and bilingualism. The study concluded that, in these special cases, the CI improved the child's quality of life, increasing both speech perception and communication skills as well as self sufficiency (Filipo et al, 2004).

In 2005, Wiley et al analysed the perceived qualitative benefit of 16 paediatric CI recipients with additional disabilities after at least six months of CI use by means of a questionnaire specifically developed for CI children with multiple impairments. They found that the majority of the children wore the device consistently, had greater awareness to environmental sounds, made communication progress, and were more attentive and interested in the world around them (Wiley et al, 2005).

Despite these reported encouraging results, there are some severe conditions associated to deafness for which the reported benefits after implantation are poor or minimal and may be considered contraindications to the CI procedure.

In 2000, Hamzavi et al reported minimal benefit in implanted children with severe learning difficulties, psychomotor retardation, and autism. Included in this study was one child affected with autism, blindness, sensorimotor integration disturbances, and auto aggression. They concluded that these conditions may represent contraindications to CIs. Donaldson et al (2004) studied the outcomes of six children using a CI who had an associated diagnosis of autism spectrum disorder. Although most of the parents reported positive benefits after implantation, including changes in behaviour and communication skills and increased awareness to the environment, only one child affected with a mild autism spectrum disorder achieved spoken language. The authors concluded that oral communication is not likely to be a realistic goal for implanted children who present with a diagnosis of autism spectrum disorder.

The evidence gathered so far is quite sparse and does not provide clinicians consistent procedures for decision making and for the measurement of outcomes. Therefore, establishing and quantifying the benefits after implantation in deaf children with additional disabilities is a critical issue, as formal tests currently used to evaluate speech perception and language abilities may be unsuitable for this population. Moreover, there are few standardized methods available to measure benefit in daily life (Waltzman et al, 2000).

The goal of the present study was to comprehensively examine the benefits of CI use in a sample of profoundly deaf children with multiple handicaps and to investigate the correlation between objective post-implant outcomes and perceived benefits in daily life. Testing included pre- and post-implant communicative behaviour and speech perception skills with both direct and indirect procedures that included a questionnaire administered to the parents. There was concern that the application of conventional tests of speech perception may result in inappropriate estimation of the effects of CIs in terms of quality of life and educational and social gains. In the current study, it was assumed that the approach of combining speech perception data with the subjectively perceived benefits might prove to be a more adequate method for measuring the outcomes in this special category of CI recipients. In addition to evaluating the results in the entire sample, the results in a subgroup of ten patients affected by mental retardation were also evaluated in order to evaluate the impact of this disability on CI outcomes.

Methods

Participants

The experimental sample consisted of 23 paediatric CI recipients with additional neuropsychiatric disabilities, from a cohort of 123 profoundly deaf children (18.6%), referred to the Audiology and Phoniatric Service of Treviso Hospital (University of Padua) and to the ENT Unit of the University of Pisa for cochlear implantation between 1999–2005. The group included 10 males and 13 females, with a mean age of 6.3 years (range 2.3–17 years) at time of implantation.

The mean pre-operative unaided PTA (pure-tone average between 500, 1000, and 2000 Hz) was 112 dB HL (range 98–125 dB HL). Pure-tone thresholds in sound field were used for children under the age of three years and with non-cooperative patients (ear-specific pure-tone thresholds could not be obtained). For the remaining patients, the hearing threshold in the implanted ear was considered for the study. The thresholds beyond the audiometer limits (120 dB HL) were assigned a value of 125 dB HL. All of the children had worn appropriate hearing aids and received speech therapy before the evaluation for a CI.

The CI models used included the Nucleus 24 M-K (Cochlear) in two children, the Nucleus 24 Contour (Cochlear) in 12 children, the Nucleus 24 Contour Advance (Cochlear) in two children, the Clarion Hi Res (Advanced Bionics) in three children, the Clarion CII (Advanced Bionics) in one child, and the MED-EL C40+ in three children. None of the participants had inner ear malformations, and all of the patients received a complete insertion of the electrode array.

All of the children underwent comprehensive clinical, neurological, and neuropsychological testing during the cochlear implant candidacy evaluation or at subsequent appointments (in the occasion of post-implant referrals). According to the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) diagnostic criteria, the associated neuropsychiatric disorders were classified into the following categories: mental retardation, language and learning disorders, pervasive developmental disorders (PDD-autistic spectrum disorders), and behavioural and mood disorders (including attention deficit and hyperactivity disorder (ADHD), oppositional defiant disorder (ODD), depression, and bipolar disorders). We also included known neurological syndromes with central nervous system (CNS) involvement (cerebral palsy, TORCH encephalopathies, epilepsy, CNS malformations), and new syndromes (currently on record) that may be associated to deafness (such as deafness-dystonia peptide (DDP) syndrome,and deafness associated to leukoencephalopathies).

The primary neuropsychiatric disabilities in our sample were cerebral palsy in three cases, mental retardation in ten cases, autistic spectrum disorder in two cases, attention deficit and hyperactivity disorder in four cases, language and learning disorders in three cases, and epilepsy in one case. In 91% of the children (21/23) the associated disability was diagnosed before implantation. In one patient implanted at the age of two years and six months, a pervasive developmental disorder was diagnosed after surgery; in another patient, implanted at the age of four years, mild mental retardation and ADHD were diagnosed after implantation. In 39% of the children (9/23) two or more disabilities associated to deafness were present.

Table 1 includes a summary of the main characteristics of the entire sample, including aetiology, additional disability/disabilities, age at implantation, CI device, duration of follow-up, and pre-implant PTA.

Material and procedures

CI outcome was evaluated by comparing the pre- and post-operative speech perception skills, and by means of a questionnaire, administered to the parents, aimed at evaluating important aspects of their child's adaptive and communicative behaviour, such as the child's overall mode of communication before and after implantation and the changes in daily life after implantation.

The post-implantation results for the entire group were analysed first. Then, the outcome of the ten children with associated mental retardation in the sample was assessed separately.

The mean interval between CI stimulation and the post-operative evaluation was 2.5 years, with a range of 1–5 years.

Evaluation of speech perception skills

All of the children underwent pre-and post operative standardized tests of speech perception and were then assigned to Geers and Moog's six categories of speech perception skills (Geers & Moog, 1991), as described in Table 2. A categorical approach to the evaluation of speech perception skills was selected to better compare performance among subjects with different levels of speech perception abilities.

Four different speech perception tests in Italian were used to evaluate the children's speech perception skills:

1. PCAP (*Prime categorie percettive: First speech perception categories*), which analyses the ability to identify words with a different duration pattern and with a significantly different spectral pattern (in closed set) (Arslan et al, 1997).
2. TIPI 1 (*Test di identificazione di parole infantile 1: Test of identification of words for children 1*), which analyses the ability to identify words that differ for one vowel or one consonant (Arslan et al, 1997). The child has to choose between four different words (closed set).
3. 3.TIPI 2 (*Test di identificazione di parole infantili 2: Test of identification of words for children 2*), which analyses the ability to identify words that differ for one vowel or one consonant (Arslan et al, 1997). It differs from TIPI 1 because the child has to choose between six different words, instead of four (closed set).
4. Lists of phonetically balanced bisyllabic words, which analyse the ability to recognize words in an open set (AAVV, 1997).

Speech perception tests were administered by a speech therapist at the two CI centres involved in the study. The speech material was administered by live voice, at 70 dB HL in quiet, in auditory alone condition.

Questionnaire

Post-operatively, parents were asked to complete a questionnaire adapted to Italian from the questionnaire proposed by Wiley et al (2005) (Kluwin & Stewart, 2000). The questionnaire assessed CI use and recorded the parents' judgement about the perceived benefits after cochlear implantation in terms of speech perception, social interaction, communication, the improvement in the overall mode of communication, as well as the general opinion of the CI procedure.

To investigate the use of the CI, parents were asked what percentage of time during a typical day their child used the implant. The parents could choose between four different options: (1) for more than 75% of the time, (2) for 50–75% of the time, (3) for 25–50% of the time, and (4) for less than 25% of the time.

To investigate the overall judgment about the impact of the CI procedure on their child, parents were asked what they would choose if they had to make the decision for a CI for their child again; they were also asked if they would suggest a CI to the parents of a child with disabilities similar to their child.

The results of the questionnaire are reported in Table 3. Parental responses were assigned a score regarding the perceived benefits to obtain quantitative and comparable results. Each response was rated according to a four point scale (from 1 to 4, as shown in Table 3); therefore, the total score for each patient could range from a minimum of 7 to a maximum of 28 (see Table 3).

To analyse the overall mode of communication, communication skills were classified into five different categories, ranging from non-verbal to oral language use, as reported in Table 4. Parents were asked to describe their child's communication mode before and after implantation, according to this five point scale.

Analysis of data

A speech perception score greater than or equal to 4 was considered good, because it meant that the child has access to fine spectral information. The cut-off for the communication mode was a score ≥ 3: the child uses oral language to communicate. For the perceived benefit, a score ≥ 19 was considered good; this is superior to 66% of the maximum score.

A correlation analysis was carried out between the post-implant speech perception categories (from zero to six according to Geers and Moog) (Geers & Moog, 1991) and the post-implant communication modes and perceived benefits scores, using the

Table 1. Clinical characteristics of the sample.

Patient, Gender	*Aetiology*	*Main associated disabilities*	*Age at implantation (years)*	*Cochlear implant device*	*Duration of follow up (years)*	*Pre-operative PTA*
Patient 1, f	Unknown	Cerebral palsy	2.3	Nucleus 24 Contour	2	125 dB HL
Patient 2, f	Congenital cytomegalovirus	Cerebral palsy	7	Nucleus 24 Contour	1	117 dB HL
Patient 3, f	Unknown	Cerebral palsy	6	Nucleus 24 Contour	2	112 dB HL
Patient 4, f	Peri-natal asphyxia	Mild mental retardation and movement disorder	4.6	Nucleus 24 Contour Advance	1	107 dB HL
Patient 5, f	Unknown	Moderate mental retardation	17	MED-EL C40+	3	107 dB HL
Patient 6, m	Congenital cytomegalovirus	Severe mental retardation, motor delay	3.8	Clarion HiRes	2	125 dB HL
Patient 7, f	Congenital cytomegalovirus	Mild mental retardation, motor delay	2.6	Nucleus 24 Contour	4	125 dB HL
Patient 8, f	Congenital cytomegalovirus	Mild mental retardation, muscle hypotonia	2.9	Clarion HiRes	1	98 dB HL
Patient 9, m	Congenital rubella	Mild mental retardation, psycosis	12	Clarion HiRes	1	108 dB HL
Patient 10, m	Short syndrome	Mild mental retardation, attention deficit and hyperactivity disorder	4	Clarion CII	3	108 dB HL
Patient 11, m	Peri-natal asphyxia	Mild mental retardation, attention deficit and hyperactivity disorder	6	Nucleus 24 Contour	4	103 dB HL
Patient 12, f	Unknown	Mild mental retardation	10.4	MED-EL C40+	4	100 dB HL
Patient 13, f	Connexin 26	Mild mental retardation	6	Nucleus 24M	4	110 dB HL
Patient 14, m	Unknown	Autistic spectrum disorder	6	Nucleus 24 Contour	3	122 dB HL
Patient 15, f	Leukoencephalopathy of unknown origin	Autistic spectrum disorder	2.6	Nucleus 24 Contour	2	107 dB HL
Patient 16, m	Connexin 26	Attention deficit and hyperactivity disorder	2.7	Nucleus 24 Contour	2	115 dB HL
Patient 17, m	Unknown	Attention deficit and hyperactivity disorder	17	MED-EL C40+	1	107 dB HL
Patient 18, m	Connexin 26	Attention deficit and hyperactivity disorder	13	Nucleus 24 Contour	4	107 dB HL
Patient 19, m	Unknown	Attention deficit and hyperactivity disorder	4.10	Nucleus 24 Contour	3	103 dB HL
Patient 20, m	Leukoencephalopathy of unknown origin	Language disorder	4.2	Nucleus 24 Contour	3	117 dB HL
Patient 21, f	High prematurancy	Language disorder and hyperactivity	4	Nucleus 24 Contour Advance	1	98 dB HL
Patient 22, f	Congenital cytomegalovirus	Epilepsy	3	Nucleus 24 Contour	2	115 dB HL
Patient 23, f	Congenital rubella	Language and learning disorder and hyperactivity	7	Nucleus 24M	5	125 dB HL

Note: PTA is pure-tone average between 500, 1000, and 2000 Hz.

Table 2. The speech perception categories by Geers and Moog (1991).

Speech perception category	
0	No pattern of speech perception
1	Some pattern of speech perception
2	Some word identification (duration pattern)
3	Consistent word identification (significant spectral differences)
4	Identification of words that differ for one vowel
5	Identification of words that differ for one consonant
6	Open set recognition of words

Spearman function. A correlation analysis, using the Spearman function, between aetiology and outcomes, in terms of speech perception skills, perceived benefits, and communication mode improvement, was also completed. Finally, in the group of the implanted children with associated mental retardation, a correlation analysis, using the Spearman function, between the degree of mental retardation and the results was performed in terms of speech perception skills, perceived benefits, and communication mode improvement. A score was attributed to each category of mental retardation degree, ranging from 1 to 3 (mild mental retardation = 1, moderate mental retardation = 2, severe mental retardation = 3).

Results

Table 5 summarizes the post-implantation results of the entire sample, with regard to speech perception categories, communication modes, and perceived benefits. In this Table, pre-implant speech perception categories and communication modes are also reported (see Table 5).

Speech perception skills, according to Geers & Moog's categories

Pre- and post-operatively, children's speech perception skills were classified according to Geers and Moog's speech perception categories (Geers & Moog, 1991). In Figure 1 (a), the pre- and post-operative speech perception categories of the entire sample are reported. Pre-operatively, 74% of the children were in the lowest categories (0–1), 26% were assigned to categories 2–5, and none of the children attained category six. Post-operatively, children improved their speech perception skills, as 53% attained category six and only 13% were still at categories 0–1. For two children, the post-operative testing was difficult, and they were unable to complete the task due to the severity of their associated disabilities (see Figure 1 (a)).

In the group of ten children with associated mental retardation, pre- and post-operative speech perception skills were classified as follows. Pre-operatively, 60% of the patients were assigned to categories 0–1, 40% to categories 2–5, and no children were assigned to category six. Post-operatively, 50% of the children achieved category six, and only 20% were still classified into the lowest categories (0–1) (see Figure 1(b)). For one child the post-operative evaluation was difficult, due to the severity of her disability.

Survey responses

General aspects regarding the use of the CI and the parents' overall judgement about the CI procedure

All of the children of this study are CI users: 22/23 children used their implant for more than 75% of the time during a typical day, and 1/23 used it for 50–75% of the time during a typical day. Among the implanted children with associated mental retardation, 10/10 children used their implant for more than 75% of the time during a typical day.

Nearly all of the families (22/23) stated that if they were given the option to have their child implanted again, they would choose to implant; however, one family was unsure on this point. All of the families would suggest an implant for a child with disabilities similar to their child.

Perceived benefits

Test scores of the perceived benefits for each child of the entire sample are summarized in Table 5, and 6 (a) and (b). One hundred percent of the families indicated an improved awareness to environmental sounds, 74% indicated improvements in the child's speaking skills, and 96% reported improved interaction with peers. Ninety-six percent of the families reported that their child was more likely to communicate his/her wants/needs, 100% reported that he/she was more attentive/interested at home and at school, and 100% found that he/she worked better with siblings/classmates (see Table 6 (a)).

The reported benefits by parents of the 10 implanted children with mental retardation were similar to those of the total sample. In fact, 100% of the families indicated an improved awareness to environmental sounds; 70% reported improvement in developing speaking skills; 100% found improvement in interaction with peers; and 100% indicated that their child was more likely to communicate his/her wants/needs, that he/she was more atten-

Table 3. Perceived benefits and assigned scores.

Perceived benefits	*I strongly agree*	*I agree*	*I disagree*	*I strongly disagree*
Improved awareness to environmental sounds	4	3	2	1
Developed speaking skills	4	3	2	1
Improved interaction with peers	4	3	2	1
More likely to communicate wants/needs	4	3	2	1
More attentive and interested at school	4	3	2	1
More attentive and interested at home	4	3	2	1
Gets along better with siblings/classmates	4	3	2	1

Table 4. Definition of communication modes and assigned scores.

Score	*Mode of communication categories*
1	Behaviour: Child uses crying, facial expressions, vocalization, and some gestures to communicate.
2	Gestures: Child uses gestures to communicate (no sign language).
3	Augmentative communication*: Child exclusively uses augmentative communication.
4	Gestures + oral language: Child uses a combination of gestures and spoken words to communicate.
5	Oral language: Child exclusively communicates orally.

*None of the children enrolled in the study used the Italian sign language. Two children used augmentative communication; therefore, in category 3 sign language was substituted (which was present in the original version of the paper) with augmentative communication.

tive/interested at home and at school, and worked better with siblings/classmates (see Table 6 (b)).

Communication skills

Before implantation, 69% of the patients of the entire sample had a main communication mode of behaviours or gestures, and only 28% used oral language (either exclusively or associated to gestures). After the CI, the percentage of children using solely behaviours or gestures dropped to 28%, and the percentage using oral language (either exclusively or in association to gestures) increased to 69%. One patient used augmentative communication both before and after implantation.

Among implanted children with associated mental retardation, 70% of parents indicated behaviours or gestures as the main pre-implant communication mode, and 30% indicated oral language and gestures. Post-operatively, the percentage of children using oral language (either exclusively or associated to gestures) increased to 70%.

Analysis of data

The correlation (Spearman non-parametric correlation) between post-implant speech perception categories and post-implant communication modes was statistically significant (Rho by Spearman $= 0.739$, $p \leq 0.00$). Moreover, both post implant speech perception categories and communication modes were significantly correlated with post-implant perceived benefit scores (Rho $= 0.798$ $p \leq 0.00$, Rho 0.628, $p \leq 0.01$, respectively).

In addition, children who had poor post-implant improvement in speech perception and communication skills achieved a good score with regard to the perceived benefits; in fact, eight out of nine children with poor speech perception outcomes (speech perception category ≤ 4) scored quite high (score ≥ 19) with regard to the perceived benefits, and six out of seven children with poor communicative outcomes (communication mode score ≤ 3) achieved a score ≥ 19 for perceived benefits (see Table 5).

The results showed no significant correlation between aetiology and outcome. Specifically, there was no significant correlation between aetiology and post-CI speech perception category (Rho by Spearman -0.1) between aetiology and post-implant communication mode (Rho by Spearman -0.05), or between aetiology and post-implant perceived benefits score (Rho by Spearman 0.1).

Table 5. The results of the entire sample.

Patients	*Pre-operative speech perception category*	*Post-implant speech perception category*	*Pre-operative communication mode (score)*	*Post- implant communication mode (score)*	*Post-implant perceived benefits score*
Patient 1, f	0	6	2	5	28
Patient 2, f	1	Difficult to evaluate	3	3	28
Patient 3, f	1	6	4	5	28
Patient 4, f	0	1	1	1	26
Patient 5, f	4	5	4	4	21
Patient 6, m	0	1	1	2	20
Patient 7, f	0	6	2	5	27
Patient 8, f	1	Difficult to evaluate	2	2	20
Patient 9, m	2	2	4	4	20
Patient 10, m	2	6	2	4	27
Patient 11, m	3	6	4	5	28
Patient 12, f	1	6	2	4	25
Patient 13, f	1	6	2	4	25
Patient 14, m	0	3	2	2	20
Patient 15, f	0	1	2	2	18
Patient 16, m	0	2	1	1	21
Patient 17, m	5	5	5	5	21
Patient 18, m	1	6	2	4	27
Patient 19, m	0	6	1	5	27
Patient 20, m	0	6	2	4	28
Patient 21, f	4	6	5	5	28
Patient 22, f	0	6	2	5	28
Patient 23, f	0	4	2	4	26

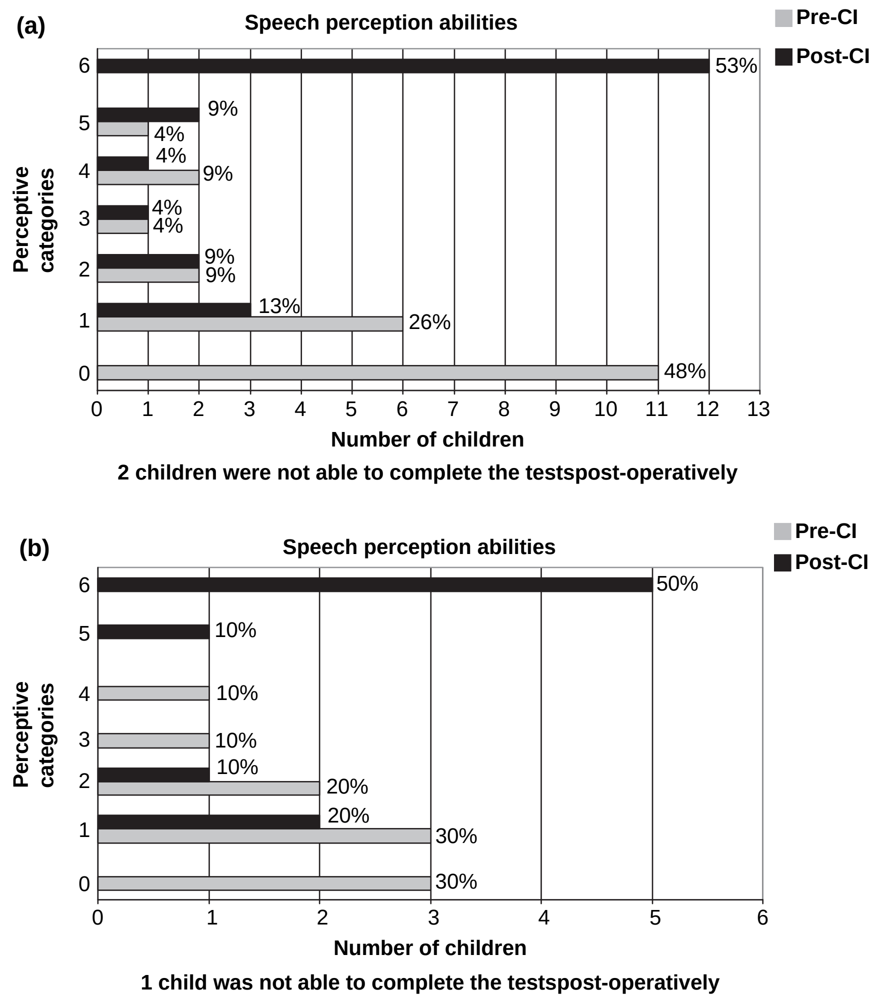

Figure 1. (a) Pre- and post-implantation Geers and Moog's (1991) speech perception categories (entire sample). (b) Pre- and post-implantation Geers and Moog's (1991) speech perception categories (children with mental retardation).

In the group of deaf children with associated mental retardation, the results showed no significant correlation between the degree of mental retardation and post-implant speech perception categories (Rho by Spearman −0.16), post-implant communication mode (Rho by Spearman −0.2), or post-implant perceived benefits score (Rho by Spearman −0.3).

Discussion

Hearing loss often coexists with other impairments, whether presenting as a syndrome or as a constellation of disabilities; therefore, the issue of submitting deaf children with multiple handicaps to a CI is a complex and emerging topic for CI centres (Filipo et al, 2004; Fortnum et al, 2002; Hamzavi et al, 2000; Holden-Pitt & Albertorio 1998; Lesinski et al, 1995). Every centre has a percentage of recipients with additional disabilities, which may affect post-implant outcomes. The reported percentages are variable, ranging from 11% to 19% (Filipo et al, 2004; Hamzavi et al, 2000; Lesinski et al, 1995; Waltzman et al, 2000). In the current study, the implanted children with additional disabilities represented 18.6% of a cohort of the 123 deaf children receiving a CI at the two Italian CI centres that collaborated in this study.

The presence of additional disabilities poses special problems concerning both the pre-operative assessment and the post-operative rehabilitation and follow-up. A multidisciplinary approach is needed, and the pre-operative neurological and neuropsychiatric observation must be particularly accurate and prolonged (Chilosi et al, 2005). Despite this, the early diagnosis of some neuropsychiatric disorders, such as learning disabilities, mental retardation, soft neurological signs, and autistic spectrum disorders, may be difficult in very young children because symptoms may not be apparent or can be missed. As the age at implantation is becoming younger, this is one of the most critical problems to be considered during the pre-operative counselling of very young children, especially under the age of 24 months. Parents should be informed that in about one third of cases, there may be additional disabilities that are not diagnosed until the child is older. These additional diagnoses may impact the child's performance with the CI (Wiley et al, 2005).

In two out of the 23 patients reported in the present study, the additional disabilities were diagnosed only after implantation. One is the case of a young girl, submitted to a CI at two years six months of age, who exhibited the signs of a PDD some months

Table 6. (a) The summary of the answers of the parents regarding the perceived benefits (entire sample).

	*Improvements**	*No improvements***
Perceived benefits (Wiley et al, 2005)		
Improved awareness to environmental sounds	23 (100%)	0 (0%)
Developed speaking skills	17 (74%)	6 (26%)
Improved interaction with peers	22 (96%)	1 (4%)
More likely to communicate wants/needs	22 (96%)	1 (4%)
More attentive and interested at school	23 (100%)	0 (0%)
More attentive and interested at home	23 (100%)	0 (0%)
Gets along better with siblings/classmates	23 (100%)	0 (0%)

(b) The summary of the answers of the parents regarding the perceived benefits (children with mental retardation).

	*Improvements**	*No improvements***
Perceived benefits		
Improved awareness to environmental sounds	10 (100%)	0 (0%)
Developed speaking skills	7 (70%)	3 (30%)
Improved interaction with peers	10 (100%)	0 (0%)
More likely to communicate wants/needs	10 (100%)	0 (0%)
More attentive and interested at school	10 (100%)	0 (0%)
More attentive and interested at home	10 (100%)	0 (0%)
Gets along better with siblings/classmates	10 (100%)	0 (0%)

*improvement is intended as a score of 3 or 4
**no improvement is intended as a score of 2 or 1

after implantation; the second is the case of a child, implanted at age four, who was diagnosed with mild mental retardation and attention deficit disorder after implantation.

Currently, there is no consensus on whether to implant a child with additional special needs. The prognostic factors of these special cases are also not well understood; therefore, it is problematic to predict results. To this regard, the studies of the literature are scarce, often limited to one or a few heterogeneous cases with a wide array of associated disabilities, making it problematic to determine which aspects of each disability impacts CI outcome. The current literature indicates that after implantation, the majority of children with multiple disabilities make progress; however, it is at a slower pace, and these children attain lower levels of communication than children without additional disabilities (Donaldson et al, 2004; Fukuda et al, 2003; Hamzavi et al, 2000; Holt & Kirk, 2005; Pyman et al, 2000; Waltzman et al, 2000; Wiley et al, 2005). Some of these children may never reach open-set recognition abilities or oral communication (Donaldson et al, 2004; Hamzavi et al, 2000; Waltzman et al, 2000; Wiley et al, 2005); they may, however, obtain some useful benefits from cochlear implantation, including the ability to recognize words from a closed set without lip-reading and improved open-set speech perception with the aid of lip-reading (Pyman et al, 2000). Moreover, these children may generally achieve important gains in daily life, derived from the greater access to the surrounding environment provided by the implant, with an overall improvement in quality of life (Donaldson et al, 2004; Filipo et al, 2004; Fukuda et al, 2003; Hamzavi et al, 2000; Holt & Kirk 2005; Lesinski et al, 1995; Pyman et al, 2000; Quaranta et al, 2004; Waltzman et al, 2000; Wiley et al, 2005).

The effects of CI use in the current group of patients were quite variable, but generally positive. All the children used their CI consistently, and speech perception testing yielded a overall improvement in speech perception abilities. Before implantation, most of the children were classified in the speech perception categories zero (48%) or one (26%) according to Geers and Moog's speech perception categories (Geers & Moog, 1991); and after implantation, 53% of children achieved open-set recognition abilities (speech perception category six).

In the survey, parents reported positive benefits after implantation. In fact, from the analysis of the questionnaires, it was evident that most of the children improved their communication skills, even if only 35% achieved an exclusively oral mode of communication. All of the children manifested a growing awareness to environmental sounds and were more attentive/ interested at home and at school; most of them were better able to communicate their needs/wants and to communicate in general. Moreover, most of the parents, when asked what they would choose if they had to make the decision for a CI for their child again, answered that they would choose a CI again. Although the obtained gains in terms of speech perception and communicative skills were lower in comparison to those generally achieved by implanted children with hearing impairment as the sole disability, satisfactory results were achieved by almost all of the children in terms of connectedness and interest in the environment, with an overall improvement in quality of life. This is an important result that agrees with other findings from the literature.

Interestingly, correlation analysis showed that children with better post-implant perceptual abilities significantly improved their communicative and social skills; nevertheless, children with poor speech perception and communicative outcomes achieved gains in daily life leading to an improvement of the overall quality of life.

As shown, the results of this study are quite variable with regard to speech perception and communication skills and to benefits in daily life. This may be in part related to the heterogeneity of the sample. In fact, the age at implantation (2.3 to 17 years) and the pre-implantation speech perception abilities and communication skills were quite variable, and the duration of the follow up was very different between patients, ranging from one to five years. All of these aspects are known to be important in influencing CI outcomes (Svirsky et al, 2004; Waltzman et al, 2005) and should be considered when evaluating the current results.

Another aspect to consider is the heterogeneity of the disabilities associated to deafness. In the current study, the sample was similar to that of most of the aforementioned studies. The children were affected by a variety of disabilities, leading to a variety of outcomes with the CI.

No significant correlation was observed between specific aetiologies and post-implant outcomes in the current study. This is probably because the same aetiology may be associated with different disabilities, with a different impact on the outcome. It is worth noting that in our sample of deaf children with multiple impairments, three patients carried connexin 26 mutations, which are generally described in patients with nonsyndromic hearing loss.

In the present study, the outcome of a subgroup of ten children with mental retardation was also evaluated. Mental retardation is one of the disabilities most frequently associated to deafness, and it is well known to be correlated to language abilities both in normal-hearing and in implanted deaf children (Wechsler, 1974; Zimmerman, 1972, Dawson et al, 2002; Geers et al, 2002; 2003a,b). To this regard, Nikolopoulos et al (2004a,b) showed that the cognitive level and the learning style were among the most significant predictors of CI outcome in a sample of deaf children without additional disabilities, which could explain the variable results observed after cochlear implantation. Geers et al (2003a,b) reported a significant positive correlation between the performance intelligence quotient (PIQ) and speech perception skills in children with CIs; whereas Tobey et al (2003) demonstrated a positive correlation between PIQ and speech production skills. Several studies show that mental retardation or cognitive delay can have post-operative effects on communication in children with a CI, which further supports a positive relationship between the level of cognitive function and measures of speech and language ability for deaf children receiving a CI (Fukuda et al, 2003; Hamzavi et al, 2000; Lesinski et al, 1995; Pyman et al, 2000; Waltzman et al, 2000).

In the current sample of children with mental retardation, satisfactory results overall were obtained. All of the children were reported to use the implant consistently, 50% achieved open-set recognition abilities, and most of the patients had communicative benefits, even if only two out of ten achieved an exclusively oral mode of communication.

The presence of a clear correlation between degree of mental retardation and outcome could not be demonstrated, most likely because of the small number of participants and the wide heterogeneity of the subjects; the age at implantation varied from 2.6 to 17 years, the follow-up duration ranged from 1 to 4 years, and the degree of mental retardation ranged from mild (eight out of ten children) to moderate or severe (two out of ten children). Moreover, in seven out of children mental retardation was accompanied by other disabilities, such as motor disorders or behavioural and emotional disorders (Table 1).

Holt and Kirk (2005), in a recent paper on speech and language development of 19 implanted children with mild mental retardation, reported results similar to those recorded in this study. Their results were quite variable; however, they could not identify any characteristics that distinguished lower from higher performers.

From the analysis of the literature as well as from the results of the present study, it appears that one of the most important issues concerning these special cases is how to evaluate the benefits achieved after implantation. In fact, these children may be difficult to test with standard speech perception and language tests, and, when obtainable, results in terms of speech perception and language skills are often poorer than for implanted children without additional disabilities. The findings of the current study show that despite this difficulty, these patients may achieve measurable benefits in daily life, derived from the greater access to the surrounding environment provided by the implant. These improvements are often cited as anecdotal reports and not objectively measured. Only a few studies have focused on the outcome from the broader perspective of quality of life or the effects on the child and family from the viewpoint of the parents (Archbold et al, 2002; O'Neill et al, 2004).

Although parents' judgment may introduce some bias, it has the potential to add important information regarding the child's function in everyday situations. The position of the parents in the household uniquely places them to assess the impact of implantation in the context within which the child grows up. This broader view can complement the assessment measured by professionals at the implant centre, by means of standardized tests of speech perception and production. It may also be useful where more formal measures are inappropriate, which is sometimes the case of deaf children with additional disabilities (Vidas et al, 1992).

For these reasons, we attempted to compile a questionnaire aimed at investigating important aspects in everyday life such as use of implant, awareness to environmental sounds, social interaction, familial relationships, behavioural changes, and overall communication modalities. We believe that the results of the questionnaire, in association with speech perception tests, may offer a broader view on the effective benefits obtained in these difficult cases.

This overall evaluation may provide further information about prognostic factors and thereby support pre-operative counselling.

Conclusions

The results of the current study suggest that deaf children with additional disabilities may achieve some benefits from cochlear implantation, which is consistent with the available literature.

The current study attempted to define clinical procedures for the evaluation of CI use in deaf children with special needs. The results attest to the importance of a multidisciplinary approach of combining both quantitative and qualitative standardized measures. This combination helps to address the variability of the possible outcomes and to evaluate the overall post-implant gains in daily life. The present study was based on a small number of heterogeneous cases; therefore, further studies including multiple centres and wider, more homogeneous samples are needed in order to develop standardized measures to evaluate the overall outcomes and to better define the prognostic factors and expected benefits in this population.

In conclusion, the presence of additional disabilities is not a contraindication for cochlear implantation; however, not all deaf children with multiple impairments are to be considered good candidates. Their outcomes may be limited by their additional deficits, but these limitations do not appear to preclude CI benefit in speech perception, communication gains, or overall improvement in quality of life. However, parents must be advised on realistic expectations of what their child will be able to accomplish with a CI by providing them with appropriate counselling from a multidisciplinary team.

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