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Cochlear Implantation in Children with Additional Disability: A Closer Look at Cognitive Delay

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Introduction

As hearing impairment is frequently associated with additional disability, some cochlear implant teams have been challenged when considering these children as possible candidates for implantation, particularly those with significant cognitive delay. It has been suggested that 30 to 40% of children with sensorineural hearing loss have additional disability such as learning difficulty, mental handicap, Attention Deficit Hyperactivity Disorder, cerebral palsy, emotional disorders as well as other psychiatric disorders [1,2,3] . In 2008, the Gallaudet Research Institute reported associated disabilities in children with hearing loss, based on data from more than 30.000 children in the United States. Nearly 40% of the children were known to have one or more disabilities in addition to hearing loss. Visual impairment or blindness was observed in 5.4% of children, developmental delay in 4.8%, learning disability in 8.3%, orthopaedic impairment including cerebral palsy in 4.4%, mental retardation in 8.7%, emotional disturbance in 2% and autism in 1.6%. Other conditions such as traumatic brain injury, accounting individually for less than 1% of the total, were observed in 14.6% of the children [4].

It is unclear which child with associated disability will benefit from a cochlear implant and what degree of benefit should be expected. Measurement of success may be more subjective than objective measurement of speech perception outcome or auditory perception since some or many of these children may never achieve language or speech perception skills or be able to be tested accurately. In a child with cognitive impairment, the auditory information provided via a cochlear implant may not be as meaningful as he or she may not be able to process, analyze and organize the information presented. Awareness to the surrounding environment, reaction to dangerous situations and recognition of a parent's voice may be the only accomplished gain in this population and are considered by many to be positive reasons to consider implantation. Combined analyses of the results of previous studies are difficult because of the small number of subjects per study, broad inclusion criteria with significant individual differences and the use of several different tools to measure outcomes (see **Table 1**). The general use of the term "additional disability" can be very confusing as many children in the literature with "additional disability" have normal cognition and the challenges they face post implantation differ from those who have cognitive delay. As well, cognitive delay can range from mild to severe. It is this latter population that challenges cochlear implant teams the most.

Table 1. Summary of previous studies

Author (year)	Number of subjects (additional disability)	Main Outcome Measures
Nikolopoulos et al. (2008)[a]	108 (none); 44 (language and communication); 18 (cognitive); 21 (visual); 8 (physical); 13 (behavioural); 7 (vocal tract); 4 (autism); 2 (oral/facial)	Speech Intelligibility Rating Scale
Berretini et al. (2008)	3 (cerebral palsy); 10 (mental retardation); 2 (autism); 4 (attention deficit and hyperactivity); 3 (language and learning); 1 (epilepsy)	Speech perception test by Geers and Moog [6] and parent's questionary
Daneshi et al. (2007)	13 (mental retardation); 20 (learning); 15 (attention deficit and hyperactivity); 5 (cerebral palsy); 3 (visual); 4 (autism)	Persian auditory perception test
Holt et al. (2005)	50 (none); 19 (cognitive)	IT-MAIS; GAEL-P; Mr. Potato Head Task; PSI; PPVT-III; RDLS
Wiley et al. (2005) [b]	3 (cerebral palsy); 3 (motor); 3 (learning); 3 (behaviour); 1 (cognitive); 6 (language); 2 (visual)	Families' perceived benefits (open ended and closed ended questions)
Hamzavi et al. (2000) [c]	3 (learning); 5 (intellectual); 3 (motor); 2 (autism); 1 (hyperactivity); 3 (psychomotor); 1 (hemiparesis); 1 (autoagression); 1 (visual)	EARS
Waltzman et al. (2000) [d]	7 (motor); 3 (oral-motor); 3 (language); 2 (visual); 2 (learning); 7 (cognition); 3 (attention deficit and hyperactivity); 1 (autism); 1 (dyspraxia)	ESP; NU-CHIPS; GASP; PBK; MLNT; LNT; common phrases sentence test
Pyman et al. (2000)	55 (none); 11 (motor and cognitive); 6 (cognitive); 3 (motor)	Five point scale of speech perception levels

[a] 28 subjects had more than one additional disability
[b] 9 subjects had more than one additional disability
[c] 6 subjects had more than one additional disability
[d] 6 subjects had more than one additional disability; no disability was reported on 3 patients (one had CHARGE Association, one had USHER's Syndrome and the other one had Waardenburg's Syndrome)

IT-MAIS: Infant-toddler meaningful auditory integration scale
GAEL-P: Grammatical Analysis of Elicited Language: Pre-Sentence Level Test
PSI: Pediatric Speech Intelligibility Test
PPVT-III: Peabody Picture Vocabulary Test, Third Edition
RDLS: Reynell Developmental Language Scales
EARS: evaluation of auditory responses to speech
ESP: Early Speech Perception Test
NU-CHIPS: Northwestern University Children's Perception of Speech Test
GASP: Glendonald Auditory Screening Procedure
PBK: Phonetically Balanced Kindergarten Word
MLNT: Multisyllabic Lexical Neighborhood Test
LNT: Lexical Neighborhood Test

Review of the literature

Daneshi et al. studied 398 cochlear implanted prelingually deaf children retrospectively in order to identify the frequency of additional disability [5]. A two stage process (screening and comprehensive psychological assessment) identified additional disability in 60 (15%) of the prelingual deafened children. Severe mental retardation was considered a contraindication for implantation and these patients were not included in the analyses. The Persian auditory perception test was applied preoperatively and one year after device activation. When analyzing the different additional disabilities in separated groups of the 60 patients, some diversity was observed. The pre and post mean scores were significantly different in children with mild mental retardation-8 patients (p < .012), moderate mental retardation-5 patients (p < .043), learning disability-20 patients (p < .0001), attention deficit disorder-15 patients (p < .000) and cerebral palsy-5 patients (p < .043). Such differences were not statically significant in children with congenital blindness-3 patients (p < .102) and autism-4 patients (p < .068). Children with cerebral palsy showed maximum improvement whereas those with autism showed the minimum amount of improvement.

Hamzavi et al., in a case series report, examined the speech perception skills of 10 children with associated disabilities who received a cochlear implant [6]. Before cochlear implantation none of the children showed evidence of speech recognition or production. Following implantation half of the children achieved some level of word recognition and production and 4 children demonstrated changes in their behaviour, such as happiness with noise. Only one child was considered as no success due to absence of reaction to sound regardless of intensive therapy. Although this article was one of the pioneers to present subjective and objective outcomes on cochlear implanted children with other special needs, the small number of subjects, the wide range of disability of the population studied and the definition of success precludes meaningful conclusion. On a case by case review, some children did make improvement when tested with the Evaluation of Auditory Responses to Speech Test which resulted in the authors concluding that providing multi-handicapped children with cochlear implants results in benefit for both the child and the parent.

Wiley et al. determined families' perceived benefits of cochlear implants in children with multi-handicaps [7] . A questionnaire with open-ended and closed-ended questions was developed for this purpose. Interviews were performed on 15 families of 16 children. Only one child of the 16 studied was classified as having a cognitive delay. The remainder were classified as either having cerebral palsy, motor delay, learning disability, behaviour problems language disability or a combination of learning and behaviour problems. Improvement on awareness to environmental sounds was reported by 94% of the families, development of speaking skills by 88%, interaction with peers by 69%, communication of needs by 88%, attention at school by 60%, attention at home by 82%, work with siblings by 71% and work with classmates by 74%. Interestingly, families of children who had their implant for more than 2 years perceived more benefits than the families of children who had their implant for less than 2 years. All families stated that with they were to make the decision again, they would choose to have their child implanted. In this study 34 families met the inclusion criteria but only 17 (50%) were still enrolled in the study center therapy. All families were invited to participate in the study; the 17 enrolled on the therapy were contacted by the therapist and the other 17 were contacted by email. Only 2 families contacted by email were willing to participate. This represents a bias since the participants in the study are likely the most motivated families, who were still bringing their child to therapy.

Berretini et al. studied 23 cochlear implant recipients with additional disabilities from a cohort of 123 profoundly deaf children [8]. Preoperative and postoperative results of a six category speech perception test [9] were analyzed. The questionnaire proposed by Wiley [7] evaluated the parents' perceived benefits after implantation. Preoperatively, 74% of the children were in the lowest categories (0 and 1) on speech perception test and none attained category six. Postoperatively, 53% attained category six and only 13% remained in categories 0 and 1. Improvement on awareness to environmental sounds, child's speaking skills and interaction with peers was observed by 100%, 74% and 96% of families respectively. It was reported by 96% of the families that their child was more likely to communicate their needs, 100% reported that their child was more attentive at home and at school, and 100% found that their child worked better with siblings and classmates. Only one family stated that if they if they were to make the decision again, they would choose not to have their child implanted. Ten of the 23 children were classified as being mentally retarded. In the following areas; improved awareness to environmental sounds, improved interaction with peers, more likely to communicate wants/needs, more attentive and interested in school and home and gets along better with siblings/classmates, parental perception of improvement was 100% in these 10 patients. Seven out of 10 parents perceived improved development of speaking skills in this population as well. Subjective parental perspective does have it's limitations from a research perspective, but should not be totally discounted. This study assessed both subjective and objective outcomes and when both were analysed it was appreciated that 8 out of 9 children with poor speech perception outcomes (0 to 4) had high scores on families' perceived benefits. The authors reported that benefit can be achieved regardless of scores on speech perception tests, i.e. 8 out of 9 children with poor speech perception scores scored high with regard to the perceived benefit scores. They concluded that the presence of additional disabilities was not a contraindication for cochlear implantation and that not all deaf children with multiple impairments are good candidates. Further more homogenous and studies of greater numbers are required.

Nikolopoulos et al. [10] studied 67 implanted children with additional disabilities and compared them with 108 age equivalent children without disability. Speech intelligibility was assessed five years following implantation. Connected intelligible speech was observed in 47 (70%) of children with additional disabilities and 104 (96%) in the control group. More detailed analyzes showed that the quality of the speech intelligibility was markedly different between the groups. Only 11 (16%) of the children with additional impairments were intelligible to all or to people with little experience while in the control group 66 (61%) achieved these highest categories of speech skills ($p < .000001$). Analysis of the specific disabilities revealed that language and communication disorders were individually the most significant contributing factor to outcome followed by physical impairment, cognitive impairment and autism. The number of additional disabilities had the strongest correlation with the speech intelligibility results. Those with more than one additional disability besides hearing loss were less likely to develop intelligible speech. Compared to other studies this one has a large number of subjects (67) a control group (108) and a long follow-up (5 years). However, the separated analysis of different disabilities is compromised by small numbers (only 4 were autistic for example).

Waltzman et al. studied speech perception skills of 29 children with additional disabilities who received cochlear implants [11]. Relative to a control group of children without additional disabilities reported in a previous study [12], fewer of the additional disabled children were able to complete the speech perception tests given. When they could be tested, their average scores were lower than those of the children without additional impairments. The authors reported that children with associated disabilities obtained demonstrable benefit from implantation based on observations of increased social interaction and “connectedness” to the environment after implantation. No measurement of these subjective benefits was performed and the conclusion was based on the impression of the authors. In all of the areas tested, no data was prevalent both preimplanation and postimplantation for many of the patients.

Pyman et al. examined the speech outcome improvement of 75 pediatric cochlear implant recipients [13]. Children were classified according to etiology of deafness and according to the presence of motor and/or cognitive delays. Of the 75 children, 20 presented delayed milestones (17 had cognitive impairment and 14 had motor impairment). The authors found an association between cytomegalovirus as cause of deafness and presence of both motor and cognitive delays ($p < .001$). The etiology of deafness had no significant influence on speech perception development. On open-set word recognition, the children with motor and/or cognitive delays never reached the level of performance of the group without additional impairments, even after 4 years of implant use.

Holt et al. retrospectively analyzed speech and language development of 19 cochlear implanted children with mild cognitive delay and 50 cochlear implanted children with normal cognition [10]. In order to control the range of disabilities children with other diagnosis besides hearing loss and cognition impairment were excluded. All children demonstrated significant improvement on a battery of speech perception and receptive and expressive language skills on every test administered. Children with cognitive delays had significantly lower scores than typically developing children on two of the three measures of receptive and expressive language and had significantly slower rates of auditory-only sentence recognition development. The authors concluded that benefit was achieved in the areas studied in the patients with mild cognitive delay.

Discussion

Previous studies that measured speech perception outcomes in cochlear implant recipients with additional disabilities were able to show improvement comparing the results pre and post operatively [5, 6, 8]. Whenever the parent's perspective was analysed a significant benefit from the cochlear implantation was reported [7, 8]. In fact only affirmative answers were found in the study where the families were questioned. As well, if they had to make the decision again they all would have the surgery undertaken [7]. Parents of children with disabilities feel gratified and reassured of their decision to implant when their child becomes more socially involved, turns the head to the sound, smiles when listening to their voices or dances when there is music, even if expressive or receptive auditory skills are not improved. This raises the question: what is the appropriate measurable benefit? Historically there has been a tendency to assess the objective scores on speech or auditory perception tests. More recently the subjective improvement that the families are able to report have gained wider acceptance.

Whenever outcomes following cochlear implantation were compared to children without additional disabilities significant differences were observed. Speech perception tests showed lower scores from the disabled group when compared to the non-disabled group, but gains are made [10, 11, 12, 13, 14] .

When cognitive delay is teased out from the papers in the literature overall, a small number of patients per study is found. Of the 8 studies listed in **Table 1** from the literature related to cochlear implantation and "additional disability" 92 out of the 294 patients studied (31%) had cognitive delay which ranged from mild to severe. Generally these patients have not been formally tested by a psychologist and the testing pre and post implantation is not standardized. This does not allow any combining of the data. Outcomes for children with the same diagnosis can vary considerably which also makes comparison even more difficult. As the cognitive function of children with autism can very considerably, these patients were not part of the cognitive delay group above.

Trimble et al.[15] recently published an interesting paper related to speech perception outcome in multiply disabled children following cochlear implantation. The aim of the 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. The disability of the 58 patients studied was not divulged as the focus of the paper was related to utilizing the Graded Profile Analysis (GPA) Score and the functional disability of the patient via the Modified Battelle Development Inventory Screening Test and compare this with the findings of the post implantation Pediatric Ranked Order Speech Perception (PROSPER) test. The functional disability score significantly predicted high and low speech perception scores while the GPA score was not significantly associated with speech perception scores. This approach clearly allows a more objective assessment and combining this methodology with a valid subjective test would be ideal.

Conclusions

Children with additional disabilities besides hearing loss do benefit from cochlear implantation. This benefit can be objectively measured through speech/auditory perception tests and/or subjectively through the therapist's or parent's perception and quantified through a questionaire. However, further studies with standardized testing parameters in a multi-centred setting to allow greater numbers are required. Children with cognitive delay, particularly mild delay do improve on objective testing, but at a slower rate which is not unexpected. Gains are made.

Subjective benefit seems to be achieved in almost every case. The use of a valid subjective measurement in combination with an objective measurement would be beneficial. It would allow the ability to predict which candidates will be successful and to what degree gains in auditory/speech perception are made as well as benefits to the quality of life in this patient population including that of their families. It may also assist in predicting which candidate may become a non user. These findings would be considerably helpful to patients, parents and cochlear implant teams.

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