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Cochlear Implantation Can Improve Auditory Skills, Language and Social Engagement of Children With Autism Spectrum Disorder

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Objective: To review outcomes of cochlear implantation (CI) in children diagnosed with autism spectrum disorder (ASD).

Study Design: Retrospective case review and parent survey.

Setting: Tertiary care children’s hospital.

Patients: Thirty children with ASD who underwent CI between 1991 and 2018. Mean age at CI = 3.5 years (0.8–11.8), mean age at diagnosis of ASD = 5.1 years (2.0–15.0) (22/30 diagnosed after CI), mean follow-up = 10.5 years (1.4–21.6). Parents of 7 children returned a survey.

Intervention: Unilateral or bilateral cochlear implantation.

Main Outcome Measures: Speech perception; expressive communication mode; educational placement; social engagement; consistency of CI use; parent survey of child behavior change.

Results: Thirty-three percent of all and 45% of the 22 consistent device users developed measurable open-set speech perception by an average of 4.5 years of device use. Educational placement at last follow-up included 13% mainstreamed without interpreter, 50% Special Education

programs, 10% therapeutic residential or day programs, 23% total communication programs, and one home schooled. Spoken language alone was used by 31% and spoken plus sign by 14%, with the remainder using sign alone, augmentative communication devices or no mode of communication. By parent report, 86% showed improvement in social engagement compared to pre-CI. Survey results showed the behaviors most frequently ranked as most affected by CI were communication and attention, while awareness of environment had the lowest (most affected) mean ranking.

Conclusions: Findings support a growing body of literature that cochlear implantation has the potential to improve auditory skills, language, and enhance social engagement in some deaf children with autism spectrum disorder.

Key Words: Augmentative and alternative communication—Autism spectrum disorder—Cochlear implantation—Educational placement—Language—Social engagement.

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Children with autism spectrum disorder (ASD) have been reported to have a higher prevalence of sensorineural hearing loss (SNHL) than children without ASD (1,2).

Conversely, children with SNHL have been reported to have a higher rate of ASD than those with normal hearing (3,4). Autism spectrum disorder is a complex developmental disorder characterized by impaired social interaction and communication as well as restricted, repetitive, and stereotyped patterns of behavior. The number of children diagnosed with ASD has increased over the past two decades, with recent studies estimating a prevalence of 1 in 88 children in the United States and 1 in 161 children worldwide (5,6). The reason for an increased rate of co-occurrence of SNHL and ASD is unknown, although developmental insults affecting both neurocognitive and auditory development have been theorized (3,7).

Cochlear implantation (CI) is the only effective treatment of SNHL for which amplification does not provide adequate access to spoken language. Cochlear implants have enabled spoken language and mainstream

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education for many children with significant bilateral SNHL. CI has been demonstrated to increase the rate of both spoken and sign language acquisition for children in oral and total communication classrooms (8). In addition, for children unable to develop receptive and expressive language, CI may improve child-caregiver engagement, thereby positively influencing quality of life (7). Understanding the range of outcomes of implanted children with ASD is important for counseling of parents and professionals, including educators, to ensure that implanted children with ASD receive appropriate support and educational services. The aim of this study is to describe long-term outcomes of CI in a large series of children with ASD treated at a single tertiary care children's hospital.

MATERIALS AND METHODS

Institutional Review Board approval was obtained for retrospective clinical review and parental survey of children diagnosed with ASD among 1,649 children implanted between 1991 and 2018 (IRB study/protocol #2014–15875). Electronic medical records were reviewed for age and sex, electrode insertion and eighth nerve anatomy, etiology of hearing loss, age at ASD diagnosis, additional comorbidities, measures of auditory skills including Meaningful Auditory Integration Scale (MAIS) and speech perception tests, interval at which open-set skills was first measured, consistency of device use, communication mode, educational placement, and post-CI parental report of improved social engagement. Device use was often based on parent report as data logging was not consistently available. Improved social engagement from pre-CI was determined primarily from notes routinely made by the child's cochlear implant audiologist based on discussion with the parent. Communication with others by spoken and/or sign language, and use of Augmentative Alternative Communication (AAC) Picture Exchange System (PECS) was also abstracted.

A parental survey of behavior after CI was mailed to the home. The survey was a modified version of a University of Michigan survey created to evaluate impact of CI on behavior and family interactions of children with ASD (9).

Patient Characteristics

Thirty children with ASD were identified (Table 1). All had full electrode insertion(s) and none had cochlear nerve deficiency. Mean length of follow up was 10.5 years (1.4–21.6). The majority (68%) were male. The average age at first CI was 3.5 years (0.8–11.8). Fourteen (47%) were unilaterally implanted and one continued to use an aid in the non-implanted ear. Five (16%) received bilateral simultaneous CIs, and 11 (37%) underwent bilateral sequential CIs at an average interval of 3.6 years (0.4–12.3). One (#25) had measurable open-set speech discrimination pre-CI. Average age at diagnosis of ASD was 5.1 years (2.0–15.0). Twenty-two (73%) were diagnosed after CI. Eight (27%) were diagnosed with ASD prior to CI, of which one (#29) had spoken language pre-CI. The remaining seven children diagnosed with ASD pre-CI had no spoken language and an average age of 6.3 years (2.8–11.8) at first CI. Etiology of SNHL was known in four patients (#4-GJB2; #13-Pendred, #22-Waardenburg, #23-Usher), and unknown in the majority (87%).

Thirteen patients had additional diagnoses likely to impact CI outcome, including 9 (30%) with additional behavioral or psychiatric diagnoses and three (#5,9,11) with cognitive impairment of varying degrees.

RESULTS

Individual outcomes are presented in Table 2. Thirty-three percent (10/30) of all patients and 45% (10/22) of consistent device users developed measurable open-set speech perception (PBK, CNC) by an average of 4.5 years of device use (0.3–9.1). Four (#8,10,16,20) developed closed-set recognition of monosyllable words (ESP category 4). MAIS scores were the only available outcome measure for 15 children. These scores improved for seven children. Six children had no change in MAIS scores despite significantly improved auditory thresholds. One child (#5) with a profound cognitive impairment secondary to degenerative neurologic disease, had no measurable auditory improvement.

At last follow up, 4 (13%) were mainstreamed (two graduated mainstream high school) without an interpreter. Fifteen (50%) were enrolled in Special Education programs, five with a sign interpreter. Three (10%) were enrolled in a therapeutic program (residential or day) for deaf children with behavior or psychiatric disorders that focuses on teaching life skills. Seven (23%) were enrolled in Total Communication (TC) programs. One (#14) was home schooled and lost to follow up after 1.5 years. Three children (#2,26,27) received Applied Behavioral Analysis (ABA), an intensive ASD intervention, either in school or privately.

Primary expressive communication mode with others at most recent follow up was spoken language alone for 9 (31%); spoken and sign for 4 (14%), and sign alone for 4 (14%). An augmentative communication device was primarily used for communication by four children (#11,22,26), of whom one (#2) also used limited sign. Eight (28%) had no mode of communication. For the child lost to follow-up, communication mode is unknown.

Twenty-two children (73%) were consistent CI users; five (17%) inconsistent, and three (10%) nonusers. The nonusers (#5,6,19) were never consistent users and became nonusers within 2 to 4 years.

Based upon parent report to the audiologist, 25 of 29 (86%) children had improvement in social engagement compared to pre-CI condition. Of those with no increased engagement (#5,6,18,22), three were inconsistent or nonusers, none of whom developed measurable speech perception. None of the four have language (spoken or sign) and one uses an AAC Picture Exchange Communication System (PECS).

Survey Results

Surveys were returned by seven families. Average age at first CI for this group was 3.6 years (1.17–11.83). Five (71.4%) reported consistent device use (>8 h/day), one fluctuating use, and one nonuse. These characteristics are similar to the overall group. Table 3 provides mean

TABLE 1. Individual subject characteristics of 30 implanted children with autism spectrum disorder (ASD)

Subject	Sex	Age ASD Diagnosis (Years)	Age First CI (Years)	Laterality	Additional Diagnoses
1 ^b	M	3.0	1.9	Bilateral	Tuberos sclerosis, Seizures
2	M	2.5 ^a	3.0	Left	
3	F	9.0	1.8	Right	
4 ^b	F	2.6	1.6	Bilateral	
5	M	15.0	5.1	Right	Degenerative neurologic disease, Leukodystrophy, Dementia, Seizures, Profound cognitive impairment
6	M	4.1 ^a	6.2	Right	
7	M	4.0	1.8	Bilateral	
8	M	3.5	1.5	Right	
9	M	14.9	3.3	Right	Disruptive behavior disorder, Moderate cognitive impairment
10	M	2.0	1.2	Right	Disruptive behavior disorder
11 ^b	F	2.5	1.8	Bilateral	Mild cognitive impairment
12	M	n/a ^a	5.5	Left	
13	F	n/a	1.2	Bilateral	Towns Brock Syndrome, Cerebritis, Seizure disorder, hypothyroid
14	M	2.1 ^a	6.2	Right	
15	F	2.5 ^a	2.8	Bilateral	ADHD, Disruptive behavior disorder
16 ^b	M	9.1 ^a	11.8	Right	ADHD
17	F	6.0 ^a	6.6	Bilateral	
18 ^b	F	5.7	3.7	Bilateral	Anxiety
19	M	n/a	0.8	Bilateral	
20	M	4.1	2.5	Right	ADHD, OCD, Coarctation aorta, Ventricular septal defect, Heart block
21	M	n/a	1.3	Bilateral	
22	F	n/a ^a	7.7	Right	
23	M	n/a	1.4	Bilateral	Anxiety, Oppositional defiant disorder
24 ^b	M	4.0	3.3	Left	Tourette syndrome
25	M	11.6	7.2	Left	Long QT syndrome, Heart transplant
26	M	2.8	2.1	Bilateral	
27	M	n/a	1.5	Bilateral	ADHD, Extreme prematurity, 22q11.2 duplication syndrome
28	M	2.8	1.0	Bilateral	
29	F	2.0 ^a	8.5	Bilateral	
30	F	2.5	1.3	Bilateral	

^aASD diagnosed prior to CI.

^bParent survey completed.

ADHD indicates attention deficit hyperactivity disorder; CI, cochlear implant; F, female; M, male; OCD, obsessive compulsive disorder. n/a, Data not available.

parental responses regarding behavior and communication, and interpersonal interaction. For the majority of behaviors, mean parent ratings were improved postimplantation, though this must be interpreted with caution given the small number of families responding. Table 4 provides mean parental ranking of behaviors affected by CI. There was extreme variability in ranking of affected behavior, with no single behavior consistently ranked first. Communication and attention were ranked by two families each as the behaviors most affected by CI, with awareness of environment and ability to succeed ranked as most affected by one family each. None of the other behaviors were ranked as the #1 most affected behavior by any of the six families who responded to this question. Awareness of environment had the lowest (most affected) overall mean ranking, while emotional needs of the child had the highest (least affected) mean ranking.

Parents were also asked if they would recommend CI to other parents with a similar child. Three would

recommend “without hesitation”; four with reservations. When asked if they would still choose CI for their own child, 6 of 7 parents responded, of whom 5 would implant “without hesitation”; one with “a little hesitation.”

DISCUSSION

The opportunity to hear may provide implanted children who have complicating conditions such as ASD with a range of benefits, especially if implantation is done early in life and coupled with necessary behavioral therapy to support auditory skill and language development (7). No methods exist to accurately predict future cognitive ability and language of young children with or without ASD, including those with normal hearing. The rationale for early intervention is to change the child’s trajectory of development when delays are present (10–13). Some of the skills that predict language outcome in

TABLE 2. Individual outcomes of cochlear implantation for 30 children with ASD

Subject	Follow Up (Years)	Most Recent Audiology Measures	Post-CI Expressive Communication	Education	Increased Engagement (Yes/No)
1 ^a	18.8	PBK 60%	Oral + sign	Special Ed + Sign	Yes
2	3.3	MAIS 63%	Sign + AAC	TC program with ABA	Yes
3	19.8	CNC 52%	Oral	Special Ed	Yes
4 ^a	6.0	MAIS 53%	None	Special Ed	Yes
5	9.0	^b	None	Special Ed ^e	No
6	11.3	MAIS 35% @ 3 years ^b	None	Therapeutic Program ^d	No
7	12.6	CNC 89%	Oral	TC program	Yes
8	17.3	ESP (standard): Category 4 ^c	None	Therapeutic Program ^d	Yes
9	14.3	MAIS 25%	Sign	Special Ed + Sign	Yes
10	10.1	ESP (low verbal): Category 4 ^c	Sign+ Oral	TC program	Yes
11 ^a	6.7	MAIS 75%	AAC	TC program	Yes
12	14.9	MAIS 45%	None	Therapeutic Program ^d	Yes
13 ^a	12.7	MAIS 75%	Sign	Special Ed + Sign	Yes
14	1.5	MAIS 83%	Unknown	Home school	Unknown
15	10.3	MAIS 68% ^c	None	Special Ed	Yes
16 ^a	21.6	ESP (low verbal) Category 4 ^c	Sign	Special Ed + Sign	Yes
17	18.9	CNC 68%	Oral	Mainstreamed through high school	Yes
18 ^a	5.7	MAIS 33% @ 4 years ^b	None	Special Ed	No
19	15.3	PBK 72%	Oral	Mainstreamed	Yes
20	15.0	ESP (standard): category 4	Oral	Special Ed	Yes
21	7.3	PBK 84%	Oral + sign	TC program	Yes
22	2.3	MAIS 7% ^c	AAC	Special Ed + Sign	No
23	19.9	CNC 84%	Oral	Mainstreamed through high school	Yes
24 ^a	14.1	PBK 56%	Oral + sign	Special ed + Sign	Yes
25	6.6	PBK 40%	Oral	Special Ed	Yes
26	1.4	MAIS 83%	AAC	Special Ed (ABA)	Yes
27	12.6	MAIS 80%	Sign	TC program (ABA privately)	Yes
28	2.0	MAIS 55%	None	TC program	Yes
29	2.2	PBK 68%	Oral	Mainstreamed	Yes
30	2.8	MAIS 88%	Oral	Special Ed	Yes

^aParent survey completed.

^bNonuser after limited use (#5 discontinued after 2 years, #6 and 18 after 4 years, although follow up with CI program continued).

^cInconsistent device use.

^dTherapeutic program, residential or day, for deaf with behavior disorders.

^e+program for children with profound cognitive impairment.

+Sign, sign interpreter.

AAC indicates augmentative and alternative communication device; ABA, Applied Behavioral Analysis; CNC, Consonant Nucleus Consonant Monosyllabic Word Test; ESP, Early Speech Perception Test; MAIS, Meaningful Auditory Integration Scale; PBK, Phonetically Balanced Kindergarten Word Test; Special Ed, Special Education classroom; TC, total communication.

hearing children with ASD include early language and verbal ability, nonverbal cognitive ability, and joint attention, skills which impact outcomes in the general population of pediatric CI recipients (14–16). In the case of children with hearing loss, improved hearing with access to spoken language may enhance their cognitive and communication potential, including those with ASD. In addition, hearing may improve quality of life and safety by enabling awareness of environmental sounds and increasing child-caregiver engagement. These are benefits often valued by families of children unable to develop language.

Children with normal hearing and ASD have a less linear trajectory of spoken language development than children in the general population (17). Our study, the

largest series with the longest follow up (mean 10.5 years) reported to date, adds to the growing body of evidence that implanted children with ASD may develop speech perception and language skills, in addition to benefiting from improved auditory awareness and caregiver/family engagement. One third of our patients developed open-set speech perception. However, emergence of measurable open-set speech perception occurred on average 4.5 years post-CI, far later than usual. Delayed emergence of skills has been reported in implanted children with other complicating conditions (18–20). In addition, the majority of nonmedically complex children who receive a CI before 3 years of age develop measurable open-set speech perception, varying from 93.5% for those implanted under 1 year of age to

TABLE 3. Mean parental survey responses for seven children^a

Parent Survey	Pre-CI	Post-CI
Behavior and communication		
Reacts to sounds	1.0	4.0
Has adverse reactions to sounds	1.0	2.9
Wears hearing device	3.0	3.0
Reacts or responds to visual stimuli in environment	3.3	4.2
Vocalizes	2.8	4.0
Makes eye contact	2.3	4.2
Uses sign language	2.8	3.5
Recognizes name	1.7	4.0
Complies to family routine	2.3	3.5
Responds to verbal requests (with sign)	2.5	3.7
Responds to verbal requests (without sign)	1.2	2.8
Enjoys music	1.0	3.3
Disruptive behavior/tantrums	3.7	3.5
Behaves poorly in school	3.0	2.6
Uses "self-stimulating" behavior	4.0	3.8
Transitions easily between activities	1.8	3.7
Attends to objects	3.2	3.7
Attends to people	2.0	3.8
Interpersonal Interactions		
Siblings talk to child	3.5	4.3
Siblings play with child	2.8	4.0
Child taken to family gatherings	3.3	4.0
Others are interested in child's progress	4.2	4.3
Child is making progress at school	3.0	4.0
Child conforms to family routine	3.1	3.5
Family supports use of hearing device	4.0	3.4
Child complies to family routine (meals, bedtime)	3.3	4.3
Comfortable taking child to public places	3.0	4.1
Other children like to play with child	2.4	3.3

^aNot all questions were answered by all parents, with n ranging from 4 to 7.

Scoring: 1 = Never, 3 = Sometimes, 5 = Always.

84.7% for those implanted after 2 years (21). These rates are similar for children with complex medical histories, which includes ASD, if implanted under 2 years of age but drops to 50% in those implanted after 2 years but still before 3 years of age. Twelve of the 30 children with ASD in this study were implanted at ages later than 3 years, some considerably older, so a direct comparison to "expected" rates of open-set speech perception development in non-ASD implanted children is difficult. However, it is clear that rates are lower than in the "normal" CI population.

Twenty-five to 30% of normal hearing children with ASD do not develop spoken language as a means of communication (15,22,23). Therefore, a significant incidence of limited or absent spoken language in children with co-occurring deafness and ASD would not be surprising. Several authors have reported that very few implanted children with ASD develop even limited spoken language (9,24,25). More recently, Valero et al (26) and Eshraghi et al. (27) reported, respectively, that 27% and 60% used spoken phrases or sentences, and 18% and

TABLE 4. Mean parental ranking of behaviors most to least affected by implantation^a

Behavior	Mean Ranking	Number Ranking as #1 ^b
Awareness of environment	2.67	1
Communication	3.67	2
Attention	4.00	2
Ability to succeed	5.17	1
Overall family interaction	5.50	0
Behavior	5.67	0
Social interaction	6.00	0
Education	6.60	0
Sibling interaction	6.67	0
Emotional needs of child	6.83	0

^a1 = most affected – 10 = Least affected.

^bOnly six families completed this item.

7%, respectively, used single words. In our series, 45% (13/29) used spoken language to some degree, either exclusively or in combination with sign. Only four with open-set skills were oral communicators with mainstream educational placement. Again, to compare to another population of implanted children, 90.3% of non-medically complex children implanted under 1 year of age have been found to use oral communication exclusively (21). This rate drops to only 35.0% in those implanted between 2 and 3 years of age. For children with complex medical histories, the rate of oral communication exclusively is only 8.3% if implanted between 2 and 3 years of age. Interestingly, two of the four children in the current study who were mainstreamed using oral communication were implanted after 3 years of age (6.6 and 8.5 years of age at first CI), thus comparing very favorably to non-ASD CI children.

Although comprehensive speech and language evaluations were not available to characterize the children in this series, a limitation of this study, it was clear from the medical record that, with the exception of the four children who were mainstreamed, language levels (spoken or sign) were not age appropriate and often quite limited. Eleven did not use spoken or sign language in their communication with others. Of these children, eight had little communication other than pointing to make their needs known while three use a Picture Exchange Communication System (PECS) in which the child touches a single picture, in a book or on a computer tablet, to indicate an item or activity they desire (28).

Many of the children in this series did not use sign language despite early sign language exposure. Many were placed in special education programs to better address ASD, and/or cognitive impairment, with a minority requiring support of a sign interpreter. However, two children (#16, 27), ages 6 and 12 years at CI who used only natural gesture pre-CI, developed sign language as their primary communication mode post-CI. It is possible that the development of this sign language ability was facilitated by their CIs, as has been reported in deaf children without this diagnosis (8).

Standard audiologic and language measures may not capture improvement in quality of life related to CI for children with multiple disabilities and complicating medical conditions. Unfortunately, validated measures of quality of life for implanted children with additional disabilities are not available. However, in our series and others, it was clear that increased social engagement was perceived by parents (24,27). One parent whose child (#24) uses spoken language and sign reported “Without his implant, he was stuck in his own little world, no sound, no eye contact with others. The implant brought his personality out to us.”

Prior authors have noted significant variability in consistency of device use among children with ASD as well as temporary periods of nonuse (24,26,27). Challenges achieving consistent device use are to be expected in children with ASD in light of the tactile and auditory sensory issues associated with this condition (29–33). However, despite these challenges, the majority of children in our series continue to use their CI, including those with little or no spoken or sign language. Based upon the parental surveys, benefit may also be surmised from the lack of regret of having their child implanted, willingness to recommend a CI for other children with ASD, and reported enhanced social engagement. The parent of one child (#13) with no measurable speech perception and limited sign reported “She was absent. The implant brought her to us.” Regarding behaviors most affected by CI, the parents who completed the survey, in this series and the earlier Eshraghi and Donaldson series, most often noted communication and awareness of the environment as most impacted by CI (9,27).

There are a number of limitations to this study. For unknown reasons, only seven families returned the parental survey. Objective or qualitative measures of ASD severity and cognitive ability were not available, which is a significant limitation because each impacts language and educational placement. The children were in different school districts with varying services available to children with ASD and hearing loss, making it difficult to separate language improvement due to CI from those more influenced by educational support. And, as noted above, comprehensive speech and language assessments to evaluate language outcome were not available. In addition, we had no access to a group of normal-hearing children with ASD from whom a matching sample could be obtained to compare to our study population.

The average age of ASD diagnosis in hearing children is age 4 to 4.5 years, with diagnosis as young as 2 years (34,35). Average age of diagnosis in our series was 5 years (range 2–15), which is similar to the mean age of ASD diagnosis in other series of CI recipients (25,26). Older age of diagnosis of ASD in implanted children is not surprising since speech and language delay may be attributed to hearing loss alone. Monitoring by CI team members, who recognize slower than expected progress and behaviors atypical of young implanted deaf children, may be the reason ASD diagnosis is not more significantly delayed compared to that of hearing children.

CONCLUSION

Language outcome and school placement is impacted significantly by ASD, with few children achieving language and behavior permitting mainstream education. However, our findings support a growing body of literature that CI has the potential to improve auditory skills, language and enhance social engagement in at least some children with ASD.

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