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Outcomes and benefits of pediatric cochlear implantation in children with additional disabilities: a review and report of family influences on outcomes

Ivette Cejas[1]
Michael F Hoffman[2]
Alexandra L Quittner[2]

[1]Department of Otolaryngology, University of Miami Miller School of Medicine, [2]Department of Psychology, University of Miami, Miami, FL, USA

Abstract: The number of children with hearing loss with additional disabilities receiving cochlear implantation has increased dramatically over the past decade. However, little is known about their auditory and speech and language development following implantation. The purpose of this review is to evaluate the effects of cochlear implantation on the most common genetic and developmental disorders in children with hearing loss. Benefits of cochlear implantation for children with autism spectrum disorder, developmental delay, CHARGE syndrome, cerebral palsy, learning disorders, Usher syndrome, Waardenburg syndrome, and attention deficit/hyperactivity disorder are reviewed. Our review indicates that children with hearing loss and additional disabilities benefit from cochlear implantation, especially when implanted early. Thus, early interventions seem as important for these children as for deaf children without additional disabilities. Comparisons of outcomes across these disabilities indicate that children with little to no cognitive impairment (eg, Waardenburg sydrome, attention deficit hyperactivity disorder) have better outcomes than those with greater deficits in intellectual functioning (eg, autism, CHARGE syndrome). In addition, parents of children with hearing loss and additional disabilities report higher levels of parenting stress and greater child behavior problems than those without comorbid diagnoses. However, these parents are as sensitive when interacting with their children as parents with typically developing children using cochlear implantation. Given these results, it is critical to evaluate these children's developmental milestones to provide early implantation and intervention, appropriately counsel families regarding realistic expectations for the implant, and facilitate family adaptation.

Keywords: cochlear implantation, outcomes, comorbid disorders, speech and language development, auditory skills, childhood deafness

Introduction

A cochlear implant (CI) stimulates the auditory nerve, bypassing the defective cochlea, and provides auditory information to the developing brain, thus improving oral language.[1,2] They are currently approved for individuals with bilateral severe to profound sensorineural hearing loss. They are now widely used for children, with a growing body of evidence suggesting they are beneficial for speech and language development, as well as joint attention, symbolic play, and behavioral regulation.[3–5] Initially, CIs were not recommended for children with hearing loss and additional disabilities.[6–8] However, 30%–40% of children currently receiving CIs have a comorbid disorder, with mixed evidence of their potential benefits.[9]

Correspondence: Ivette Cejas
Barton G Kids Hear Now Cochlear Implant Family Resource Center, University of Miami, 1120 NW 14 Street, CRB Fifth Floor, Miami, FL 33136, USA
Tel +1 305 243 9199
Fax +1 305 243 2009
Email icejas@med.miami.edu

The purpose of this review is to evaluate the effects of CIs on the most common genetic and developmental disorders in this population and provide guidance on the needs for future research. There were no articles comparing normally hearing children with developmental or genetic disorders to CI users available to review. Notably, children with additional disabilities, but no hearing loss, are typically not referred to or evaluated by CI centers, and their trajectory of early intervention services is substantively different. We have organized this review based on the extent of developmental delays (DDs) associated with each disorder, from most severe (atypical) to least severe (typical). Each section provides a definition of the disorder, an estimate of its prevalence among the general population and in children with hearing loss, and a review of published outcomes.

Autism spectrum disorder

Autism spectrum disorder (ASD) is a developmental disability that causes significant social, communication and behavioral challenges. Severity is assessed on a continuum and includes children previously diagnosed with Pervasive Developmental and Asperger's Disorder. ASD is characterized by impairment in social interactions, atypical communication, and restrictive, repetitive behaviors. Epidemiological studies found the prevalence of cognitive impairment in children with ASD to range between 50% and 70%.[10,11] According to the Centers for Disease Control, one in 68 children are identified as having some form of ASD, and males have a greater likelihood of developing ASD than females (5:1; see Table 1).[12] Comorbidity rates of hearing loss

in children with ASD are high and increasing, with estimates of children having both ranging from 1% to 6%.[13] There is a disproportionate number of children with profound hearing loss who have a coexisting diagnosis of ASD (35.4%). In 2009–2010, the Annual Survey of Deaf and Hard of Hearing Children and Youth indicated that one in 59 children with hearing loss also receive services for ASD.[14]

In the largest, nationally representative study of young children receiving CIs (Childhood Development after Cochlear Implantation; CDaCI),[15] eight of 188 deaf children were diagnosed with an ASD.[16] Comparisons of receptive and expressive language skills and behavior problems before to 3 years after implantation indicated that those with an ASD had lower initial language scores but similar rates of externalizing behavior problems than those who were developing typically. Over time, children with ASD improved in both receptive and expressive language, but at half the rate of those who did not have a comorbid diagnosis. Increases in externalizing behaviors were also observed over the 3-year period in the ASD group compared with the non-ASD group. This is consistent with a more recent national study in Ireland.[17]

Improvements in speech and language skills were also reported in a small descriptive study of children with ASD and hearing loss.[18] Four of the 14 children used verbal communication. In a retrospective chart review of six children aged 3–16 years with ASD and hearing loss, positive changes postimplantation were noted in responsiveness to sound, speech perception, interest in music, vocalizations, and eye contact.[19] Further, five of six parents would have recommended a CI for a family in a similar situation.

Table 1 Prevalence data for common comorbid disorders with hearing loss

Comorbid condition	Prevalence of disorder in all children	Sex differences	Prevalence of hearing loss	Prevalence of an additional disability in children with hearing loss
Autism spectrum disorder	One per 68[12]	5:1 male to female ratio[12]	1%–6% have some degree of sensorineural hearing loss[100]	4%[101]
Developmental delay	One per 6[20]	1.3 male to female ratio[20]	10.2% have some degree of sensorineural hearing loss[77]	8.8%[14]
CHARGE syndrome	One per 9–10,000[102]	Equal sex representation[44]	34%–38% have severe to profound hearing loss,[43,44] 75% have some degree of hearing loss	NR
Cerebral palsy	One per 323[103]	1.3: male to female ratio[103]	12%–25% have some degree of sensorineural hearing loss[53]	NR
Learning disorder	50–150 per 1,000[20]	2–3:1 male to female ratio[20]	NR	7.20%[14]
Usher syndrome	10 per 100,000[62]	Equal sex representation[104]	100% have some hearing loss[104]	3%–6%[105]
Waardenburg syndrome	One per 42,000[70]	Equal sex representation[70]	35%–78% of children with WS I and 55%–91% of children with WS II[71,72]	2%–5%[73,77]
ADHD	50 per 1,000[20]	2:1 male to female ratio[20]	NR	5.4%[14]

Abbreviations: NR, not reported; ADHD, attention deficit/hyperactivity disorder; WS, Waardenburg syndrome.

In sum, although few studies have been conducted comparing the outcomes of cochlear implantation in children with versus without ASD, the results are generally positive in terms of speech and language outcomes, and it seems critically important to provide this auditory stimulation. Given the challenge of "diagnostic overshadowing", in which ASD may be overlooked in children with hearing loss, health care providers should closely monitor developmental milestones to identify those at increased risk for ASD or those who might benefit from a more in-depth assessment.

Developmental delay

In young children, DDs are characterized by a failure to reach expected developmental milestones, including motor and intellectual functioning.[20] This is a common diagnosis for children under the age of 5 years, for whom standardized testing is not possible. DDs occur frequently in children with hearing loss, who lack auditory input and communicative skills. Children with DD may have severe or more mild impairments as shown in Figure 1; however, we have reviewed studies of children with various levels of hearing loss and DD in the following text.

Children with DD experience impairments in adaptive functioning, such as communication, social skills, academic achievement, and independence. DD is usually caused by traumatic brain injury, genetic disorders (eg, CHARGE syndrome), viruses (cytomegalovirus), or abnormalities in the brain. DD has an overall prevalence of one in six children and approximately 15% of all those aged 3–17 years have one or more developmental disabilities.[21] Males are affected more often than females. Studies estimate that DDs occur in approximately 23%–34% of children receiving CIs.[22]

Seven studies focusing on children with CIs and DD reported mixed evidence of improvements in auditory and speech and language outcomes. Results varied based on severity of impairment. Amirsalari et al compared 28 children with mild DD, based on gross motor functioning, with 234 children with CIs but without DD, before and 2 years after implantation.[23] Children in both groups demonstrated significant progress on the Categories of Auditory Perception and

Speech Intelligibility Rating from pre to post implantation. No significant differences were found between the groups on the Categories of Auditory Perception or Speech Intelligibility Rating 2 years following implantation.

In contrast, Holt and Kirk reported on a group of 19 children with mild cognitive delays (mean age at implantation 37.7±14.6 years) and 50 without delays (mean age at implantation 28.8±13.2 years).[24] Measures of receptive and expressive language and sentence recognition, averaged across 2 years postimplantation, showed that children with mild cognitive delays scored significantly lower than their non-delayed peers. Hiraumi et al also found that eleven children with DD had lower speech perception skills than 24 non-delayed children 2 years after CI surgery.[22]

Although the literature on auditory perception and speech intelligibility in children with mild delays is mixed, several studies have found that a greater degree of DD is associated with worse outcomes. Pyman et al retrospectively examined the speech perception outcomes for 75 pediatric CI users (mean age at implantation 3.2±1.2 years) to determine the effects of etiology of deafness.[25] Results showed that the presence of motor and/or cognitive delays was associated with slower high-level speech processing ability. Wiley et al similarly compared 14 children with DD (mean age at implantation 17 months) with 21 children without DD (mean age at implantation 16 months).[26] Children with DD were classified as either severely or mildly delayed. Results 1 year postimplantation showed that children with mild delay (ie, a developmental quotient of ≥80) made similar auditory skills improvement as those without DD. Children with more severe delay (ie, developmental quotient <80) were much slower in making these improvements. In a more recent study, children with DD (mean age at implantation 41.4 months) demonstrated gains in receptive language, self-care, and social functioning 1 year after implantation.[27]

In addition, Edwards et al found that those with versus without DD (n=11 with DD; n=21 without DD) had significant delays in speech intelligibility and perception 1 and 2 years after surgery.[28] Post hoc analyses showed that children with severe DD had poor outcomes, but those with

Figure 1 Various disabilities discussed in this paper placed on a continuum from minimal potential benefit to maximum potential benefit based on a review of the literature.
Abbreviations: ADHD, attention deficit/hyperactivity disorder; CI, cochlear implant; CP, cerebral palsy; DD, developmental delay.

mild delays showed demonstrable progress. In general, retrospective chart reviews have indicated that children with DD demonstrate wide variability in speech and language outcomes following implantation.[26,27,29–38]

Overall, results were mixed, with children who had more severe DD lagging further behind than those with milder DD. Findings from individual studies were likely influenced by the inclusion and exclusion criteria for DD, how it was assessed, and which outcome measures were utilized. However, it is clear that children with DD do improve following cochlear implantation, although their rate of change may be slower than expected for typically developing deaf children.

CHARGE syndrome

CHARGE syndrome is a rare genetic pattern of birth defects, which occurs in about one in every 9–10,000 births worldwide (see Table 1). It is an extremely complex syndrome, involving extensive medical and physical difficulties that vary across children. Infants with CHARGE syndrome are often born with severe birth defects, including coloboma (eg, visual defects causing vision loss), malformed cochlea, complex heart defects, and intellectual impairment.[39] Approximately 50% of children with CHARGE syndrome have typical intellectual functioning, 25% have moderate functioning, and 25% have very poor functioning.[40,41] The incidence of severe to profound hearing loss ranges from 34% to 38%, with up to 75% of patients having some degree of hearing loss.[42,43]

In a study of eleven children with CHARGE syndrome implanted at 1–3.5 years of age and followed for 1–15 years, all showed varying degrees of auditory benefit. After cochlear implantation, most continued to show a mild hearing loss (eg, 30–45 dB). Note, however, improvements were found on a parent report measure of auditory skill development.[44] The majority of children were using manual communication and were enrolled in special education classrooms.

Two studies that each included six children with CHARGE syndrome, with a mean age at implantation of 4.9–5.9 years, measured speech perception and auditory performance.[42,45] In both studies, children showed variable but improving performance depending on age and time since implantation. Children with CHARGE syndrome who had more severe intellectual disabilities showed poorer performance than the average. Five children with CHARGE syndrome, who had a mean age of 37 months at time of implantation, were followed for 1–4.5 years. Two used verbal communication, one began using oral language, and two did not develop any communicative skills.[43]

Three case studies of children with CHARGE syndrome indicated that children showed benefits in terms of detection of sound, recognition, and identification of environmental sounds, and in some children, a demonstrable improvement in receptive language.[39,46,47] Although improvements in auditory perception have been documented in children with CHARGE postimplantation, their oral language skills appear to be limited. A comprehensive early intervention program is needed to follow up on the complex issues these children face.[16]

Cerebral palsy

Cerebral palsy (CP) is a group of disorders that affects the development of movement, balance, and posture, and is the most common motor disability in childhood. "Cerebral" means having to do with the brain and "palsy" means weakness or problems using the muscles. CP is caused by abnormal brain development affecting control of children's muscles.[48] Although all individuals with CP have problems with movement and posture, the severity of symptoms vary from person to person, ranging from mild difficulties with ambulation and fine motor control to use of assistive devices to walk and lifelong care. CP does not get worse over time, although symptoms often worsen over a person's lifetime. It is frequently associated with other comorbidities, such as intellectual disability, seizures, problems with speech and hearing, and changes in the spine (eg, scoliosis). Cognitive impairment has been found in 2%–44% of children with CP; however, the prevalence increases when epilepsy is present.[49] Furthermore, among children with severe CP, approximately 97.7% have a profound mental impairment. The prevalence of CP is approximately one in 323 children in the USA, with 12%–25% experiencing some level of hearing loss (see Table 1).[48–50]

In a small study comparing children with CIs and CP with those with CIs alone (n=8; age and sex-matched pairs), audiologic outcomes were mixed.[51] Within the CP group, four children performed similarly to their matched peers on measures of receptive and expressive language, speech perception, and social maturity. These children had less severe cognitive and motor impairment. The remaining four children in the CP group had a more severe degree of impairment and demonstrated slower rates of improvement in speech and auditory perception over time compared with the control group. Cruz et al also found that children with CP (n=4) develop language at a slower rate than CI children without additional disabilities.[16] The CP group also displayed higher rates of externalizing behaviors (eg, aggression) than typically developing children prior to implantation.

However, externalizing behaviors decreased over time at a rate similar to that in the control group.

Steven et al evaluated the speech recognition scores of 36 children with CP 2 years after implantation without a control group.[52] Results showed that children with mild cognitive disabilities displayed speech recognition scores that were significantly higher than those with severe impairments. Similarly, Bacciu et al reported on the auditory perception and speech intelligibility of five children with CP and CIs (mean age at implantation 91.6±9.7 years).[53] All five children demonstrated substantial improvements in auditory perception and speech intelligibility at least 1 year after device activation.

In conclusion, results were mixed, with children who had more severe CP developing language at a slower rate than those with milder CP. The findings across studies are similar to those for children with DD. It is evident that children with CP do improve following cochlear implantation, although expectations for development may need to be adjusted based on the degree of impairment. Furthermore, specific accommodations are often needed to assist the child with CP in wearing the implant device. A physical or occupational therapist should be consulted when making these adjustments.

Learning disorder

Learning disorder (LD), also known as "specific learning disorder" in the *Diagnostic and Statistical Manual of Mental Disorders,* Fifth Edition (DSM-5), is a neurodevelopmental disorder characterized by persistent difficulties learning core academic skills.[20] Key academic skills include reading single words accurately and fluently, reading comprehension, written expression and spelling, calculation, and mathematical reasoning. LD occurs due to an interaction of genetic, epigenetic, and environmental factors that affect the brain's ability to perceive or process verbal or nonverbal information efficiently and accurately. In contrast to talking and walking, which are acquired developmental milestones that occur with maturation of the brain, academic skills must be explicitly taught and learned.[20] LDs disrupt the acquisition of learning skills, despite having an adequate opportunity to learn.

The prevalence of LDs across academic domains of reading, writing, and mathematics is 5%–15% among school-aged children (see Table 1).[20] LDs are more common in males than females (estimates range from 2:1 to 3:1). Deafness has been linked to increased rates of academic difficulties, particularly the acquisition of reading skills.[54] The Annual Gallaudet Survey reported a national average of 7.2%.[14]

Studies have shown a mixed pattern of outcomes. Isaacson et al matched five post-meningitic children with CIs and LDs (mean age at implantation 5.4 years) to non-LD peers (mean age at implantation 5.2 years).[55] In general, children with hearing loss and LDs made substantial improvements postimplantation; however, their progress was slower and less consistent in speech perception, speech-reading, and receptive vocabulary than the non-LD group over 30 months postimplantation. In contrast, Cruz et al compared seven children with LDs with 157 children without additional disabilities, and found no significant differences in the rate of language growth between groups pre to 36 months postimplantation.[16] However, the LD group had worse language scores prior to implantation. It is important to note that externalizing behavior problems decreased over 3 years postimplantation in this group. Positive outcomes from preimplantation to postimplantation were also reported in a sample of 20 children with CIs and LDs 1 year after implantation.[56] However, there was no typically developing deaf comparison group. Two case reports also reported improvements in speech intelligibility and auditory perception.[26,57]

Studies of children with hearing loss and LD, which documented later implantation (eg, after 10 years of age) have consistently reported worse language outcomes and furthermore their reliance on manual forms of communication.[58–60] Both the severity of the learning disability and length of auditory deprivation are key factors in explaining the variability of these speech and language outcomes. In sum, these children should be implanted as early as possible and should receive early intervention and ongoing monitoring of their academic functioning.

Usher syndrome

Usher syndrome is an autosomal recessive, genetic disorder that affects both hearing and vision, with three clinical subtypes. It occurs in 3%–6% of children with congenital deafness and may account for 50% of the deaf-blind population (National Institute of Child Health and Human Development, 2008; see Table 1).[61] In the typical population, Usher syndrome occurs in ten of every 100,000 people.[62] Usher syndrome type 1 is the most common and severe, characterized by profound hearing loss, vestibular dysfunction, and retinitis pigmentosa. Retinitis pigmentosa leads to degeneration of the rods of the retina causing impaired night vision, progressive narrowing of the visual field, and loss of peripheral vision. Type 2 is also common and is associated with moderate to severe hearing loss, retinitis pigmentosa,

and normal vestibular function. Type 3 has a prelingual or post-lingual onset of progressive hearing loss, progressive loss of vestibular function, and occasionally, late onset of retinitis pigmentosa. Little is known about the prevalence of cognitive impairments in children with Usher syndrome. One study reported that 15% of their sample had some degree of intellectual impairment.[63] Another study found similar levels of cognitive performance when comparing children with Usher syndrome and CIs to children with hearing aids.[64]

A multisite study of children receiving CIs from 1991 to 2010 identified 712 children; 26 (3.7%) of these children also had Usher syndrome type 1.[65] Mean time to follow-up was 7.8 (1–16) years. Speech perception outcomes for those with Usher syndrome indicated that 92% were able to achieve some level of open-set speech perception on age-appropriate tests and over half of these children were also oral communicators.

In a study at two CI centers, nine children with Usher syndrome type 1, implanted between age of 2 and 15 years, completed measures of speech perception and CI usage.[61] Outcomes were measured 1–2 years postimplantation. Six of nine children could discriminate speech sounds without lip reading, and three of nine scored well on an open-set word recognition task. Those who were implanted earlier, prior to the age of 3 years, performed significantly better than those implanted later. In a second study, of seven children with Usher syndrome type 1 and aged 7–16 years performed better on a working memory task (ie, sentence completion and recall) and nonword discrimination task than typically developing children with CIs.[64] No differences were found between these groups with regard to phonologic working memory.

In a study of 14 children and adults with Usher syndrome type 1, followed for 24–120 months, five of seven implanted before the age of 10 years improved significantly over time on a speech perception test.[66] Earlier implantation led to better performance. No relationship was found between gene mutation and audiologic outcomes. In a subsequent analysis of the same sample, children and adults with Usher syndrome using CIs reported better health-related quality of life scores on a hearing-specific questionnaire than those who were not implanted.[67] Overall, children with Usher syndrome type 1, which is the most severe form, performed similarly to typically developing children with CIs. Thus, early identification and implantation is recommended.

Waardenburg syndrome

Waardenburg syndrome (WS) is a group of genetic conditions that may cause hearing loss and changes in the pigmentation of the hair, skin, and eyes.[68] Children with WS often have extremely pale blue eyes or different colored eyes; in some cases, one eye may contain segments of two different colors. WS is also distinguished by a white forelock of hair or premature graying. There are five clinical subtypes of WS, each categorized by different physical characteristics. Types I and II are the most common and have similar features, with the exception of widely spaced eyes in those with type I. Limited evidence exists for a link between WS and cognitive impairment. One study reported delayed development or poor school performance in nine of 52 children with WS.[69] WS is caused by mutations in the EDN3, MITF, PAX3, SNAI2, and SIX10 genes and is inherited in an autosomal dominant pattern.

Estimates of prevalence are one in every 42,000 births, with equal sex representation.[70] Sensorineural hearing loss occurs in 35%–78% of children with type I WS and 55%–91% of children with type II WS.[71,72] WS accounts for 2%–5% of all cases of congenital hearing loss.[73] Approximately one in five children with WS has a hearing loss that warrants some type of amplification (see Table 1).[74] When evaluating a child with WS for implantation, extensive radiologic screening should be conducted. Several studies have described structural abnormalities of the inner ear that may affect the surgical procedure, such as vestibular aqueduct dilatations and semicircular malformations.[75–77]

A review of the literature revealed nine articles that specifically studied cochlear implantation in deaf children with WS (see Table 2). Overall, the majority of studies found improved auditory performance and speech intelligibility. Three studies compared outcomes in implanted children with and without WS. Amirsalari et al compared six young deaf children with WS (mean age 26±15.8 months) and 75 deaf children without WS (mean age 54.5±14.8 months) 1 year following implantation.[78] Age at implantation was 2.2 years for the WS group; however, these data were not reported for the control group. All of the children with WS demonstrated significant improvement in auditory perception and speech intelligibility from preimplantation to postimplantation. However, the WS group had lower speech intelligibility than the control group. In contrast, no differences were found on auditory performance. Note that the WS group was 2 years younger than the control group and it is unclear whether chronological age was controlled for in the analyses. In a second comparison study, a small group of WS children (n=5; mean age at implantation 30.6±9.7 months) were compared with 261 non-syndromic children (mean age at implantation 36.7±18.6 months) and no differences were found between groups with regard to measures of auditory perception or speech performance.[79]

Table 2 Studies in children with cochlear implantation and additional disabilities

Reference	Date	Sample (n)	Mean age at CI (months)	Mean length of CI use (months)	Outcome measures	Results
Autism spectrum disorder						
Hayman and Franck[106]	2005	2	Child 1, 12 months; child 2, 135 months	Child 1, 30 months; child 2, N/A	ESP, MLNT	Child 1 had increased social/ emotional responsiveness and nonverbal communication. Child 2 was in a total communication class for children with CIs
Robertson[17]	2013	10	51 (SD 41)	N/A	CAPS, SIR	6 of 10 children used their CI consistently. One child used spoken language as their main form of communication, 3 used some speech and some signs, and 6 used nonverbal communication
Beers et al[107]	2014	Literature review	N/A	N/A	N/A	Children with ASD and CIs displayed improved social communication, behavioral and environmental awareness, increased vocalizations, eye contact, and reaction to music after implantation
Meinzein-Derr et al[18]	2014	14; 10 hearing aid controls	33.6 (SD 21.4)	N/A	PLS-4, CELF	4 children with ASD achieved oral communication skills postimplantation. 8 children with ASD used a form of augmented communication (eg, Picture Exchange Communication System)
Donaldson et al[19]	2004	7	63.4 (SD 31.5)	25	MAIS, IT-MAIS, GASP-W, GASP-S, PPVT-III, EVT, CDI	Children with ASD experienced increases in vocalization, eye contact, use of sign language, reaction to sound, and response to requests following implantation
Fitzpatrick et al[108]	2014	17	N/A	N/A	Retrospective chart review	Higher prevalence of hearing loss in children with ASD than in general population. Audiologic profiles of children with ASD were highly variable
Developmental delay						
Fukuda et al[35]	2003	1	56 (SD N/A)	Assessed at 6, 9, 12, 15, and 24	KSPD, Type 67 Test Battery	The child achieved monosyllable speech ability and audiometric results similar to children without ADs
Holt and Kirk[24]	2005	19; 50 CI controls	DD 37.7 (SD 14.6); non-DD 28.84 (SD 13.2)	12	IT-MAIS, GAEL-P, Mr Potato Head Task; PSI, PPVT-III, RDLS	Children with and without DD had significant improvements in speech and language. Children with DD performed significantly worse on tests of expressive and receptive language
Edwards et al[28]	2006	11; 21 CI controls	28 (SD N/A)	24	SIR, IT-MAIS, E2L	Children with DD were delayed in speech perception or speech production. Children with mild DD made progress in areas of speech perception and speech intelligibility 1–2 years postimplantation
Meinzen-Derr et al[109]	2011	15; 15 DD controls	21 (SD N/A)	12	PLS-4	Children with ADs had significantly lower receptive and expressive language quotients than children without ADs

(Continued)

Table 2 (*Continued*)

Reference	Date	Sample (n)	Mean age at CI (months)	Mean length of CI use (months)	Outcome measures	Results
Amirsalari et al[78]	2012	28: 234 CI controls	N/A	24	CAP, SIR	No significant differences were found between children with motor DD and children without motor DD in CAP and SIR Children with motor DD showed significant improvement on CAP and SIR scores following implantation
Eze et al[110]	2013	Literature review	N/A	N/A	N/A	Children with mild DD had comparable outcomes to children with without DD All parents of children with DD stated that they would choose implantation for their child again
Hiraumi et al[22]	2013	11; 24 CI controls	37 (SD N/A)	24	CV, PPT	Preoperative DD negatively affected postimplant speech perception outcomes Children with mild DD achieved speech perception scores comparable to children without DD
Wakil et al[27]	2014	21	56.4 (SD 2.9)	135.6	IT-MAIS, ESP, GASP, PBK, HINT	Severe DD, limited improvement from CI, although some level of basic auditory skills (sound awareness, association of meaning with sounds, and vocalization behavior) developed from CI use; mild/moderate DD, developed open-set speech recognition; suggests level of DD has impact on effect of CI
CHARGE syndrome						
Bauer et al[42]	2002	6; 5 CI controls	70 (SD 40)	36	ESP, GASP, IT-MAIS	3 of the 5 children demonstrated improved speech perception All children with CIs experienced improved quality of life after implantation
MacArdle et al[111]	2002	4	76.3 (SD 35.7)	48	LSQ-E, MPT, MJWL, SIR	All children showed improvement in receptive language scores postimplantation All children displayed unintelligible speech postimplantation
Au et al[47]	2004	1	30 (SD N/A)	Assessed at 12, 24, 36, and 48 months	RDLS-Cantonese version	Speech perception gradually improved after implantation
Buchman et al[112]	2004	28	66 (SD 59)	29	ESP, PBK	Children with inner ear malformations were slower to develop speech perception than children without malformations
Lanson et al[44]	2007	10	26.4 (SD 10.8)	43.6	IT-MAIS, 9 children tested	All children showed improvements on the IT-MAIS postimplant All children demonstrated increased awareness of environmental sounds and increased vocalizations
Southwell et al[39]	2010	3	Child 1, 17 years; child 2, 48 months; child 3, 36 months	6.3	HINT-C	All children reported significant improvements in hearing skills following implantation

(*Continued*)

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Table 2 (*Continued*)

Reference	Date	Sample (n)	Mean age at CI (months)	Mean length of CI use (months)	Outcome measures	Results
Ahn and Lee[45]	2013	6	58 (SD 40)	58.4	IT-MAIS, CAP, SIR	Language and speech improved in 5/6 children over time Children with CHARGE syndrome showed varying but steady improvement in auditory performance
Cardoso et al[113]	2013	1	52	41	IT-MAIS, MUSS	The child reached highest category of auditory perception The child's IT-MAIS score rose from 5% preimplant to 90% postimplant
Ricci et al[43]	2014	5	37 (SD 16)	29	ESP, IT-MAIS	2 children used oral language as the sole mean of communication; one child started using oral language as main mode of communication 2 children did not develop linguistic abilities
Cerebral palsy						
Bacciu et al[53]	2009	5	91.6 (SD 24.3)	28.2	ESP, GASP, SIR	All children achieved improved SIR scores 1 year after CI All children improved in speech perception and speech intelligibility over time
Steven et al[52]	2011	36	42.6 (SD 23.2)	55.2	SRS	The degree of cognitive impairment was a strong predictor of outcomes postimplantation More severe cognitive impairment led to poorer outcomes, but these children still showed improvement in speech perception and production
Byun et al[51]	2013	8; 8 CI controls	21.6 (SD 5.3)	24	CAP, K-Ling, SELSI	4 children with CP had speech and language outcomes comparable with age-matched and sex-matched peers 4 children with severe CP did not produce meaningful speech by 24 months
Learning disability						
Issacson et al[114]	1996	5; 5 CI controls	LD 64.2 (SD 17.4); non-LD 62 (SD 18.4)	6, 12, 18, 24, and 34 months	DAT, MTS, SERT, WIPI, GASP, MAC, CID, Iowa Medial Vowel and Medial Consonant Tests, NU No 6, PPVT, K-ABC	Children with LD showed speech and language benefits from CI, but their progress was slower and more inconsistent
Usher syndrome						
Hinderink et al[115]	1994	4	USH 249 (SD 76.2); no-USH 258.6 (SD 101.3)	24	MST, AN, Gestel-Nijmegen Implant Questionnaire	All children demonstrated improvements in speech perception and pattern recognition Children with USH achieved speech perception scores comparable with the control group
Jenison et al[116]	1995	2	N/A	36	ESP, NU-CHIPS, GASP	Both children use their CIs full-time and have shown improvement in speech perception skills Both children showed a plateau in improvement during the decline of their vision

(*Continued*)

Table 2 (*Continued*)

Reference	Date	Sample (n)	Mean age at CI (months)	Mean length of CI use (months)	Outcome measures	Results
Damen et al[67]	2006	7; 2 USH controls	75.6 (SD 28.8)	110	NCIQ, SF12, EHL	Children demonstrated increased speech perception abilities after implantation Children expressed greater independence following implantation
Liu et al[61]	2008	9	64.8 (SD 37.2)	24	IT-MAIS, ESP, NU-CHIP, MLNT, LNT, PBK, CAP, QACIU	Children with USH type 1 exhibited improvement in speech and auditory perception Children with severe visual impairment benefited most from implantation
Henricson et al[64]	2012	7; 33 CI controls; 43 hearing aid controls; 120 normal hearing controls	13 (SD 14)	110	VMPT, NWD, PR, PI, SDM, WM, Word Spotting	Children with USH type 1 performed similarly to the control group on cognitive tests, but demonstrated poorer phonologic working memory
Jatana et al[65]	2013	26	39.6 (SD 37.2)	93.6	ESP, MLNT, LNT, PBK, CNC, HINT	The majority of children with USH achieved some level of open-set speech perception 69.2% of children were using oral or primarily oral communication by time of last follow-up
Waardenburg syndrome						
Sugii et al[77]	2000	1	48 (SD N/A)	24	Ling Sound Test; Open-set word testing	The child achieved 58% on open-set word testing with auditory cues 2 years postimplantation Speech and language abilities were delayed
Daneshi et al[80]	2005	6	79 (SD 54.1)	42.8	PAPT/HI, Persian Spondee words test; CAP, SIR	Children with WS demonstrated significant improvement in speech perception and speech intelligibility All children eventually transferred to regular educational settings
Migirov et al[82]	2005	5	58.1 (SD 73.2)	52.8	ESP, IT-MAIS	4 children with WS achieved open-set recognition of monosyllabic words, and all 5 children achieved open-set recognition of 2-syllable words Children with WS achieved speech perception outcomes comparable to implanted children without WS
Cullen et al[73]	2006	6	37 (SD 20)	69	ESP, PBK	All children achieved some degree of closed-set and open-set speech Overall, children with WS performed extremely well following implantation
Pau et al[76]	2006	13	N/A	12	Melbourne Speech Perception Categories	4 children had EABR and lower speech perception scores than the other patients with normal EABR; children were diagnosed with auditory neuropathy

(*Continued*)

Table 2 (*Continued*)

Reference	Date	Sample (n)	Mean age at CI (months)	Mean length of CI use (months)	Outcome measures	Results
Deka et al[81]	2010	4	63.9 (SD 52)	12	CAP, MAIS, SIR	WS group achieved postimplant scores comparable to children without WS All children with WS significantly benefitted from CI
Kontorinis et al[75]	2011	25	70.8 (SD 61.2)	99.6	HSM sentence test, FMT, CAP	9 of 25 patients achieved monosyllabic scores better than 80%
Amirsalari et al[78]	2012	6; 75 CI controls	26 (SD 15.8)	12	CAP, SIR	Children with WS showed significant improvement postimplantation on CAP and SIR WS group had lower SIR scores than control group
De Sousa Andrade et al[79]	2012	7; 261 CI controls	30.6 (SD 9.7)	57.7	IT-MAIS, CAP, MUSS, SIR	Speech perception skills slightly worse in children with WS versus children without WS All children with WS showed significant improvements in speech perception over time
Attention deficit/hyperactivity disorder						
Pundir et al[88]	2007	2	Child 1, 72 months; child 2, 84 months	9	ISLD	Both cases improved after medication was used to address ADHD-related issues
Moreno Torres et al[87]	2010	1	18 (SD N/A)	Dec-60	LAQ, TNI-2, PPVT-III	After 5 years of implant use, the child scored at the mean for children of the same auditory age on expressive and receptive language tests
Retrospective reviews collapsing disabilities						
Hamzavi et al[60]	2000	10	47.2 (SD 19.3)	1, 3, 6, 12, 18, 24, and 36 months	EARS, MTP	9 of 10 children showed significant improvement in their ability to make speech sounds Children without DD reached a greater mean speech perception level than children with DD at each assessment point postimplant
Pyman et al[25]	2000	20; 55 CI controls	38.4 (SD 14.4)	12	Classification of Performance Levels	Children with DD developed closed set speech perception and open-set speech perception with the aid of lip-reading
Waltzman et al[57]	2000	29	50.4 (SD 30)	28.8	ESP, NU-CHIPS, GASP, PBK, MLNT, LNT, Common Phrases sentence test	Children with DD benefited from CI, but the development of auditory and linguistic skills was slower and less stable than children without ADs 59% of the children with ADs used oral communication postimplant
Rajput et al[83]	2003	85; 21 CI controls	48 (SD 20.4)	N/A	RLIM, SIR	Children with syndromal disease had lower receptive language and speech intelligibility scores than other disability groups postimplant
Dettman et al[33]	2004	22; 27 CI controls	DD 29.1; non-DD 26.4 (SD N/A)	37.3	PVT, PBK, CNC, BKB	Children with severe DD had poorer speech perception than those with no DD or mild DD All children showed some benefit from CI

(*Continued*)

Table 2 (*Continued*)

Reference	Date	Sample (n)	Mean age at CI (months)	Mean length of CI use (months)	Outcome measures	Results
Filipo et al[34]	2004	18	76.8 (SD 3.1)	34.7	Listening Progress Profile; Test Abilita Percettive; Test Identificazione Parole Infantili; Bi-Trochee-Polysyllabic word test; Test Abilita Uditive Varese; Common Protocol; Erber's categories	All children showed improved listening and communication skills following implantation Deaf-blind patients achieved the best outcomes
Wiley et al[59]	2005	16	58.8 (SD 56.4)	56.4	Open-ended and close-ended questions	All children had greater awareness of environmental sounds, made progress in speech, and were more attentive following implantation
Wiley et al[58]	2006	45	GJB2+ 23 (SD N/A); GJB2– 26 (SD N/A); GJB2 NT 55 (SD N/A)	33.5	N/A	62% of GJB2+ children were oral communicators; 67% of GJB2– children were oral communicators; 38% of GJB2 children were oral communicators
Daneshi and Hasanzadeh[56]	2007	60	68 (SD 32.2)	12	Revised Leiter International Performance Scale, PAPT/HI	Auditory perception improved in all disability groups Children with CP showed fastest improvement in auditory perception and children with ASD showed slowest improvement
Edwards et al[28]	2007	Literature review	N/A	N/A	N/A	Children with ADs, especially a mild cognitive delay, experienced numerous benefits with CI, but those with more severe disabilities showed slower rate of progress
Berrettni et al[31]	2008	23	75 (SD 52)	30	PCAP, TIPI 1, TIPI 2	Following implantation, 53% of children attained the highest category of speech perception All children with CI and DD showed improvement in environmental awareness, speaking skills, and interaction with peers
Nikolopoulos et al[37]	2008	67; 108 CI controls	ADs 40.8 (SD N/A); No ADs 39.6 (SD N/A)	60	SIR	Children with DD had similar performance on measures of speech intelligibility
Wiley et al[26]	2008	14; 21 CI controls	ADs 17; No ADs 16	12	ASC	Children with DD achieved the same rate of auditory skills as children without DD Children with more severe cognitive disabilities improved at half the rate than children with mild cognitive delays
Meinzen-Derr et al[110]	2010	20	24 (SD N/A)	28	PLS-4, RI-TLS	Nonverbal cognition, duration with CI, age at hearing loss diagnosis, and number of different therapies were related to post-CI language skills

(*Continued*)

Table 2 (*Continued*)

Reference	Date	Sample (n)	Mean age at CI (months)	Mean length of CI use (months)	Outcome measures	Results
Beer et al[20]	2012	23; 23 CI controls	24 (SD 10)	12	IT-MAIS, PLS-4, Vineland-II	Children with ADs made progress in functional auditory skills within 1 year of implant use Degree of loss, age, and age at implantation did not correlate with progress in functional auditory skills in children with ADs
Birman et al[117]	2012	29; 65 CI controls	N/A	12	CAP	Children with ADs had slower progress and poorer results than those without ADs Children with ADs showed greater social engagement/interaction and self-sufficiency over time
Cruz et al[16]	2012	157; 31 CI controls	30.1 (SD 15.1)	Assessed at 12, 24, and 36 months	RDLS	Children with ADs have slower rate of growth in oral language than those without ADs; slowest progress was for children with autism Some children with ADs had increases in externalizing behaviors compared with the control group
Wiley et al[118]	2012	6	41.7 (SD 45.8)	12	PLS-4	Children with ADs demonstrated improvement in spoken language
Broomfield et al[30]	2013	38	39 (SD N/A)	115.5	BKB, SRS	At follow-up, 82% of children were using their CI full time Variable outcomes were found within the syndromic group
Cupples et al[32]	2013	34; 85 hearing aid controls	17.5 (SD 7.5)	>10	PLS-4, PPVT-4, DEAP, CDI, PEACH, SIR	Children with ASD, CP, and/or DD had lower receptive and expressive language than those without DD. All children showed improvements in auditory performance after implantation
Janeschik et al[119]	2013	115; 148 CI controls	62.1 (SD N/A)	65.4	Mainzer and Gottinger Test for Speech Comprehension in Childhood	Children with syndromal diseases performed significantly worse on all audiologic tests when compared with children with other etiologies

Abbreviations: ESP, Early Speech Perception test; MLNT, Multisyllabic Lexical Neighborhood Test; CAP, Categories of Auditory Performance; SIR, Speech Intelligibility Rating; PLS-4, Preschool Language Scales, Fourth Edition; CELF, Clinical Evaluation of Language Fundamentals; MAIS, Meaningful Auditory Integration Scale; IT-MAIS, Infant-Toddler-MAIS; GASP-W, Glendonald Auditory Screening Procedure for Words; GASP-S, Glendonald Auditory Screening Procedure for Sentences; PPVT-III, Peabody Picture Vocabulary Test; EVT, Expressive Vocabulary Test; CDI, MacArthur-Bates Communicative Development Inventory; KSPD, Kyoto Scale for Psychological Development; GAEL-P, Grammatical Analysis of Elicited Language Pre-Sentence Level Test; PSI, Pediatric Speech Intelligibility Test; RDLS, Reynell Developmental Language Scales; CV, Consonant-Vowel Syllable Perception Test; PPT, Phrase Perception Test; PBK, Phonetically Balanced Monosyllabic Word List -Kindergarten; HINT, Hearing in Noise Test; E2L, E2L Toy Test; LSQ-E, Listening Skills Questionnaire-Environmental sounds; MPT, Manchester Picture Test; MJWL, Manchester Junior Word Lists; HINT-C, Hearing-in-Noise Test; MUSS; Meaningful Use of Speech Scale; SRS, Modified Speech Reception Score; K-Ling, Korean Version of the Ling's Stage; SELSI, Sequenced Language Scale for Infants; DAT, Discrimination After Training; MST, Monosyllable, Trochee, Spondee Word and Stress Test; SERT, Sound Effects Recognition Test; WIPI, Word Intelligibility by Picture Identification Lists; MAC, Minimal Auditory Capability Battery; CID, Central Institute for the Deaf Everyday Sentences; NU No 6, Iowa Medial Vowel and Medial Consonant Tests; Northwestern University Auditory Test No 6; K-ABC, Kaufman Assessment Battery for Children; AN, Antwerp-Nijmegen Test Battery; NU-CHIPS, Northwestern University Children's Perception of Speech; NCIQ, Nijmegen Cochlear Implant Questionnaire; SF12, Standard Medical Outcome Study Short-Form 12; EHL, Equivalent HL; LNT, Lexical Neighborhood Test; QACIU, Assessment of Cochlear Implant Use; VMPT, Visual Matrix Pattern Test; NWD, Nonword Discrimination; PR, Phonological Representations; PI, Phoneme Identification; SDM, Semantic Decision Making; WM, Word Mobilization; CNC, Consonant-Nucleus-Consonant words test; PAPT/HI, Persian Auditory Perception Test for the Hearing Impaired; FMT, Freiburg Monosyllabic Test; ISLD, Integrated Scales of Language Development; LAQ, LittlEars Auditory Questionnaire; TNI-2, Test of Nonverbal Intelligence-2; EARS, Evaluation of Auditory Response to Speech; MTP, Monosyllable Trochee Polysyllable Test; RLIM, Receptive Language Improvement score; PVT, Picture Vocabulary Test; BKB, Bamford-Kowal-Bench; PCAP, First Speech Perception Categories; TIPI 1, Test of Identification of Words for Children 1; TIPI 2, Test of Identification of Words for Children 2; ASC, Auditory Skills Checklist; RI-TLS, Rossetti Infant-Toddler Language Scale; Vineland-II, Vineland Adaptive Behavior Scale, Second Edition; DEAP, Diagnostic Evaluation of Articulation and Phonology; CDI, Child Development Inventory; PEACH, Parent Evaluation of Aural/Oral Performance of Children; EABR, electric auditory brainstem response; ADs, additional difficulties; CI, cochlear implant; N/A, not available; SD, standard deviation; ASD, autism spectrum disorder; DD, developmental delay; CP, cerebral palsy; LD, learning difficulty; USH, Usher syndrome; ADHD, attention deficit/hyperactivity disorder; WS, Waardenburg syndrome.

In a sample of older children, Kontorinis et al compared outcomes in 25 deaf children with WS (mean age at implantation 5.9 years) with 50 children who had non-syndromic deafness (mean age at implantation not reported).[75] No significant differences were found between groups 8 years postimplantation on measures of auditory perception. However, the authors reported that four of 25 deaf children with WS showed little progress following implantation.

The remaining six studies reported implantation outcomes without a control group.[30,73,76,77,80–82] All but one study found improvements in speech and language skills following implantation. Additionally, Broomfield et al compared outcomes across 38 children with different causes of syndromic deafness, 10 of whom had WS. Significant variation was found between and within syndromic groups. Retrospective reviews, which collapsed across disabilities, included children with WS, but did not analyze their outcome data separately.[32–34,57,83]

In sum, this review indicates that deaf children with WS who are implanted develop similar auditory perception and speech intelligibility skills as those with typical sensorineural hearing loss.

Attention deficit/hyperactivity disorder

Attention deficit/hyperactivity disorder (ADHD) is a neurodevelopmental disorder characterized by a persistent pattern of inattention and/or hyperactivity-impulsivity that interferes with daily functioning.[20] There are three primary classifications of ADHD predominantly inattentive presentation, which includes off-task behavior, lack of persistence, difficulty sustaining attention, and general disorganization; predominately hyperactive/impulsive presentation, which includes excessive motor activity, talkativeness, and impulsive behavior; and a combination of the above symptoms (inattentive and hyperactive behaviors). ADHD has a strong genetic component and is characterized by delayed development of the prefrontal cortex, which affects inhibition and self-regulation.[84] It is also associated with significant academic and social impairment.[20] Children with ADHD often experience cognitive difficulties, and perform poorly on tests of attention, executive functioning, memory, and processing speed.[20] Baker et al found that children with DD were three times as likely to have an ADHD diagnosis than typically developing children.[85]

Population estimates indicate that ADHD occurs in approximately 5% of children and is more common in males than females (ratio 2:1; see Table 1).[20] The Annual Survey of Deaf and Hard of Hearing Youth conducted by Gallaudet University reported similar prevalence rates of ADHD (3.4%–6.8%); however, several studies have reported higher rates of attention and impulsivity in children with hearing loss.[4,14,86]

Outcomes of implantation in children with ADHD were reported in six studies that aggregated disabilities. The majority of these studies found that these children benefitted from implantation. Cruz et al compared 12 children with ADHD with 157 children without additional disabilities.[16] No significant differences were found in the rate of language growth preimplantation to 36 months postimplantation, suggesting children with ADHD develop language at a similar rate as typically developing children with CIs. Despite the improvements in language skills, these children evidenced increasing rates of externalizing behavior problems over time in comparison with children with CIs.

In general, retrospective chart reviews showed that children with ADHD improved significantly postimplantation, as measured by speech perception and communicative ability.[31,56–58] Similarly, case studies have also reported positive language outcomes for children with ADHD and hearing loss. Moreno-Torres et al described the language outcomes for a 9-year-old girl implanted at 18 months and diagnosed with ADHD-inattentive type at 3 years of age.[87] Results showed that some aspects of oral language (eg, acquisition of verbal morphemes) occurred within the typical timeframe of a CI user, but other domains were delayed, such as the number of lexical and grammatical errors, and mean length of utterance. In contrast, children with ADHD and comorbid learning or intellectual disabilities had worse performance across both retrospective reviews and case studies.[31,60,88]

In conclusion, there is consistent evidence that children with ADHD and hearing loss benefit from cochlear implantation. Note, however, these children have difficulties with visual selective attention and externalizing behaviors, which may negatively impact the efficacy of cochlear implantation. Therefore, it may be important to assess these behaviors and incorporate parent training to target these issues.[4,89] However, identification of ADHD in children is complex because attention difficulties are common at young ages and definitive diagnosis is often not made until the child enrolls in school.

Family influences on CI outcomes

The studies reviewed above focused almost exclusively on auditory and speech and language outcomes, ignoring the larger family system in which these children develop.[90] In the

CDaCI study, the largest longitudinal study of young children receiving CIs, two key parental factors were examined that affect outcomes, ie, parenting stress and maternal sensitivity (MS). The CDaCI study enrolled 188 deaf children of hearing parents prior to implantation and 97 age-matched normally hearing children, with assessments every 6 months over 3 years.[15] The average age of the deaf children prior to implantation was 26.7 months and in the hearing comparison group was 27.5 months. All of the deaf children had severe to profound hearing loss and parents committed to educating their children in spoken English.

Parenting stress

We measured both general and context-specific parenting stress using the Parenting Stress Index and the Family Stress Scale.[89,91,92] At baseline, prior to implantation, we found that parents of deaf children reported similar levels of general stress but higher levels of deaf-specific stress. Highest ranked stressors for parents of children with hearing loss were communication, finances, and educational concerns. In contrast, parents of hearing children ranked finances, disciplines, and safety as their top three stressors.[89] Hearing status was significantly related to parenting stress by way of language delays and externalizing child behavior problems.

Levels of parenting stress may be even higher when raising a child who is deaf with an additional disability. To test this hypothesis, we compared parents of typically and atypically developing deaf children on both of these measures. Prior to implantation, these two subgroups did not differ in levels of generic or context-specific stress [Parenting Stress Index ($t(145) = -0.40$, $P > 0.05$; Family Stress Scale $t(158) = -0.30$, $P > 0.05$]. At the assessment 36 months after cochlear implantation, significant differences in parenting stress were found between these subgroups on both measures, with parents of children having additional disabilities reporting higher levels of stress [Parenting Stress Index, $t(132) = -3.1$, $P < 0.05$, $r = 0.35$; Family Stress Scale, $t(157) = -3.1$, $P < 0.05$, $r = 0.31$].

Maternal sensitivity

In addition, we videotaped parent–child interactions in both the deaf and hearing groups in the CDaCI study every year for 4 years to evaluate levels of MS. This construct is well-validated and has been used in several large national studies such as the National Institute of Child Health and Human Development Early Childcare Study.[93,94] MS was scored by rating videotapes of structured and unstructured parent–child interactions on the following dimensions:

sensitivity/responsivity, respect for child autonomy, positive regard, and hostility (reverse-coded). At baseline, parents of deaf children were less sensitive on the MS composite than parents of hearing children and this difference was observed across the 4 years postimplantation.[95] Further, after accounting for early hearing experience and child and family demographics, MS predicted growth in oral language over 4 years, and the magnitude of this effect was similar to that observed for age at implantation. Specifically, children of parents with higher MS had only a 1.3-year language delay 4 years post-CI compared with a 2.7-year delay in children of parents with low MS.

An examination of MS in the typically and atypically developing children revealed no significant differences at baseline [$t(183) = 0.8$, $P > 0.05$] or 3 years after implantation [$t(172) = 1.1$, $P > 0.05$]. Thus, despite the challenges of managing a child with a CI who has other disabilities, these parents were equally sensitive when interacting with their children. Given that MS plays a strong role in the development of oral language, the family resilience evidenced by these parents argues strongly for the importance of early implantation.

Conclusion

Our review indicates that children with hearing loss and additional disabilities benefit from cochlear implantation, and similar to typically developing deaf children, earlier implantation is better.[96] The majority of articles have focused on auditory performance and speech and language development. Overall, these studies showed that children's speech intelligibility and auditory perception improved following implantation, despite a slower rate of growth. A few studies also reported positive outcomes for self-care, social functioning, and quality of life. In the CDaCI study, we found that parents of children with deafness and additional disabilities reported higher levels of parenting stress and child behavior problems than those without comorbid diagnoses. On the positive side, these parents were as sensitive when interacting with their children. This suggests that early interventions to reduce family stress and foster positive parent–child interactions are needed.

Comparisons of outcomes across these disabilities indicated that children with little to no cognitive impairment (eg, WS, ADHD) had better outcomes than those with greater deficits in intellectual functioning (eg, autism, CHARGE syndrome). We have illustrated these outcomes by disability and potential benefit in Figure 1. Given these results, it is critical to evaluate these children's developmental milestones to provide early implantation and intervention, appropriately

counsel families regarding expectations for the implant, and facilitate family adaptation.[16,97]

Prior research in this area has been plagued by a number of methodologic limitations, which have limited our understanding of the potential benefits of implantation for this population. These studies have relied on case reports, retrospective chart reviews, and prospective investigations with small samples. Future studies with limited samples should report effect sizes, given that they are likely underpowered to find statistically significant effects. There is also a need for multisite, collaborative studies that utilize the same outcome measures and evaluate larger numbers of children with hearing loss and additional disabilities. In addition, a broader array of outcomes should be examined to identify potential benefits of implantation, such as improved attention, behavioral regulation, social functioning, school performance, and health-related quality of life.[4,98,99]

In conclusion, we strongly advocate for early cochlear implantation in children with hearing loss and additional disabilities. CI centers should make accommodations in their protocols for evaluation, CI programming, and rehabilitation services for these children and their families. These services are best provided by multidisciplinary teams, which include social workers and psychologists who can assist with both assessment and the intervention process.

Disclosure

The authors report no conflicts of interest in this work.

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