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Behavioral Outcomes in Children with Autism Spectrum Disorder and Cochlear Implants

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A capstone research project submitted to the Graduate Faculty in Audiology in partial fulfillment of the requirements for the degree of Doctor of Audiology, The City University of New York

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This manuscript has been read and accepted for the Graduate Faculty in Audiology in satisfaction of the capstone research requirement for the degree of Doctor of Audiology, Au.D.

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THE CITY UNIVERSITY OF NEW YORK

Abstract

BEHAVIORAL OUTCOMES IN CHILDREN
WITH AUTISM SPECTRUM DISORDER
AND COCHLEAR IMPLANTS

by

Patricia K. Herz

Adviser: Professor Dorothy Neave-DiToro, Au.D. CCC-A, F-AAA

The purpose of this literature review was to establish what is currently known about behavioral outcomes of children with Autism Spectrum Disorder (ASD) and cochlear implants (CIs). Sixteen articles were included in this review. The areas investigated were behavioral changes postimplantation, as determined by subjective and objective measures, mode of communication used by children with ASD and CIs, and effects of additional comorbidities on these outcomes. Considerations for CI use for children with ASD include potential hypersensitivity to auditory stimuli, inability to complete performance measures, and markedly slower progress than their typically developing peers. Many of the studies included in this review show that children with ASD and hearing loss can benefit from CI use. On subjective measures, improvements in reaction to sounds, name, and music, as well as reduced anxiety were reported by parents or caregivers, while objective measures showed no change, or an increase in behaviors postimplantation. The greatest number of children used a combination of communication modalities (37.4%), followed by oral language (34.1%). The results of this literature review indicate that the behavioral aspects of CI in children with ASD require further investigation.

ACKNOWLEDGEMENTS

I would like to express immense gratitude to my adviser, Dr. Dorothy Neave-DiToro for her guidance and encouragement throughout this endeavor. Thank you for your patience and support. This project could not and would not have come to fruition without your help.

To my mother: thank you for being my role model, backbone, and greatest supporter in one. You have shown me the true meaning of perseverance. Everything I am, I am because of you. Everything I do, I do for you. This capstone is dedicated to you.

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Introduction:

Autism Spectrum Disorder (ASD) is an increasingly prevalent developmental disability. The Centers for Disease Control and Prevention (CDC)'s Autism and Developmental Disabilities Monitoring (ADDM) Network indicates that as of 2016, the prevalence of ASD as 1 in 54 children aged 8 years in the United States, with prevalence being 4.3 greater in boys than girls (Maenner et al., 2016). The Annual Survey of Deaf and Hard of Hearing Children and Youth collected by Gallaudet Research Institute in 2010 revealed that, "higher rates of autism are reported to occur in children with hearing loss when compared to typically developing hearing peers" (Szymanski et al., 2012). According to the Gallaudet Research Institute, "as many as 40 percent of children with hearing loss exhibit an additional disability and estimates the prevalence of ASD among children who are deaf or have hearing loss to be 1 in 59" (Clason, 2017).

"Autism […] refers to a range of conditions characterized by challenges with social skills, repetitive behaviors, speech and nonverbal communication, as well as by unique strengths and differences" (What is Autism, 2012). It is considered a "developmental disorder" because symptoms typically arise within the first two years of life (Autism Spectrum Disorder, n.d). Children with ASD are often more or less sensitive than other people to sensory input, such as light, noise, clothing, or temperature (Autism Spectrum Disorder, n.d.). As the rate of diagnosed cases of ASD continues to climb, so does the likelihood that a child with a peripheral hearing loss may also have ASD. However, Fitzpatrick et al. state that, "it is difficult to estimate the true change in prevalence as the increase may be partly related to improved awareness and access to services, which in turn affect the ability of data collection sites to identify ASD" (2014).

At present, cochlear implants (CI) are commonly being used for young children with profound hearing loss (Cruz et al., 2012). A cochlear implant is a device that is used to "bypass

damaged portions of the ear and directly stimulate the auditory nerve" (NIDCD, 2018). These devices help children with significant degrees of hearing loss, who are not benefitting from hearing aids to hear. Cochlear implants are comprised of a portion that must be surgically implanted, as well as a portion that sits behind the ear. The external portion has a microphone to pick up sounds, which are then processed and sent via a cable up to a headpiece called the coil. Within the coil is a magnet, which serves to keep the internal and external components aligned. Within the internal component, an antenna picks up the information, and that information gets sent to the receiver stimulator, which analyzes the information and sends it to the electrode array in the cochlea. A diagram of a cochlear implant can be seen in Figure 1.

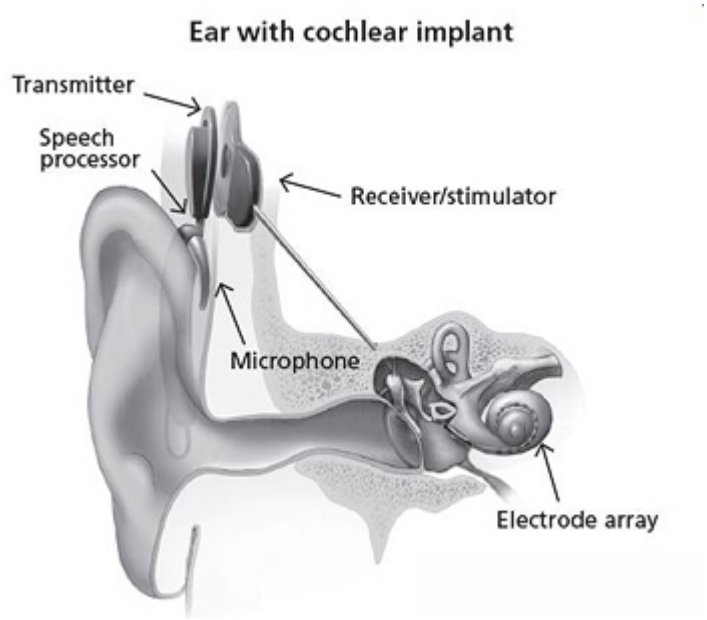

Fig. 1 *Ear with Cochlear Implant* **Source:** (NIDCD, 2018)

According to the American Speech-Language-Hearing Association, the Food and Drug Administration has approved cochlear implantation in children beginning 12 months of age, "and many children younger than 12 months of age have been implanted off protocol" (ASHA, 2004). Researchers have found that children achieve better receptive and expressive language outcomes when they are implanted before 24 months of age (Nicholas & Geers, 2013, May-Mederake,

2012, as cited by Mikic et al., 2016). Early identification of hearing loss due to newborn hearing screenings combined with the better outcomes achieved from early implantation has resulted in children with significant hearing loss being implanted before being identified with comorbidities such as ASD (Johnson & Wiley, 2009, as cited by Cejas et al., 2015). The average age of diagnosis of ASD in the Unites States (U.S.) is greater than 4 years of age (American Psychiatric Association, 2016), making it unlikely that a child will be diagnosed with ASD preoperatively if they are identified with hearing loss at birth.

While children with ASD continue to be implanted, little is currently known about the outcomes of cochlear implantation for this population. Research has consistently shown deaf children and children with developmental disabilities to have higher rates of behavior problems than their typically developing peers, in terms of both internalizing (e.g. anxiety, sadness) and externalizing behaviors (e.g. inattention, aggression) (Cruz et al., 2012). Previous researchers have stated, "Due to the limited number of reports published on cochlear implantation in children with ASD, post-implant success of children with ASD cannot be predicted" (Beers et al., 2014). Various technological options must be evaluated for the treatment of hearing loss in children with ASD, including cochlear implants. Despite the risk of negative behavioral outcomes, and aversion to increased auditory stimuli being commonly seen in children with ASD, they are still being implanted.

Pediatric Audiometry

Regardless of whether a diagnosis of ASD has been made, to qualify for CIs, children must be diagnosed with significant hearing loss. In order to receive a diagnosis of hearing loss, a child must have their hearing tested. There are multiple methods of evaluating children's hearing, which are dependent on both their age and their capabilities. Behavioral responses are

the gold standard when it comes to audiometry, but there are instances where results can only be obtained through objective measures. Infants are screened at birth via either transient evoked otoacoustic emissions (TeOAEs), or auditory brainstem response (ABR) to detect hearing loss. Otoacoustic Emissions (OAEs) are used to assess cochlear outer hair cell function. According to the American Academy of Audiology (AAA), "Although not a direct measure of hearing, OAEs provide information about the status of the auditory periphery and, in the absence of middle ear disorder, the likelihood of sensory hearing loss" (2018). This is done by sending in a stimulus and measuring the resulting echo that is generated by the outer hair cells of the cochlea for individuals with normal or near normal hearing. Present TeOAEs are consistent with no more than a mild hearing loss.

ABR screenings, and further diagnostic ABRs are another objective method of evaluating hearing status in children. ABRs are performed while the child is sleeping naturally or sedated, depending on their age and activity level. This type of evaluation is done using electrodes placed on the child's scalp or forehead, and behind each ear, and headphones or a bone oscillator are used to produce the sounds. The electrodes measure the brain's response to those sounds in the form of five waveforms. The presence, absence, amplitude, and latency of these waveforms are interpreted to determine if the child has hearing loss, and if so, what degree and configuration of hearing loss is present in each ear. In some cases, if the child begins to wake up or become restless, multiple sessions may be required to obtain all of the necessary thresholds to determine the degree and type of hearing loss.

The simplest form of behavioral audiometry is Behavioral Observation Audiometry (BOA). For typically developing children, this is performed until around 6 months of age (AAA, 2018). This type of testing does not provide exact thresholds of hearing sensitivity, but rather is

a measure of children's responses to sound. Typically, a stimulus is presented from soundfield speakers, and the audiologist will observe some change in behavior. This can come in the form of a startle response, the initiation or cessation of sucking behavior during nursing, being bottle fed or while using a pacifier, or any other noticeable shift in their demeanor that is time-locked to the stimulus. This is the least reliable and precise type of subjective testing in audiometry.

The next type of audiometry used in pediatric populations is Visual Reinforcement Audiometry (VRA). This type of testing is a conditioned response procedure that allows for an estimate of frequency specific thresholds (AAA, 2018). This type of testing is typically utilized for children between the developmental ages of 5 - 24 months. Through this method, children are conditioned that every time they detect a sound is presented, they will see a reinforcer (e.g. a light up toy, or video clip) appear. Once conditioned, the audiologist will present the stimulus, and see if the child seeks the reinforcer by turning their head or shifting their gaze in its direction. This can be done at lower and lower presentation levels to find the softest sound that the child will respond to. This testing provides, "Estimation of hearing thresholds based on minimum response levels (MRLs) that have a close relationship with perceptual thresholds" (AAA, 2018). This can be done with both tonal and speech stimuli to get frequency-specific responses, as well as a speech detection threshold (SDT) in the soundfield. As this is often done through speakers in the soundfield, it will not provide responses from each ear individually. If testing can be performed under headphones or insert-earphones, then ear-specific responses may be able to be obtained.

For children who are between 2 and 5 years of age, developmentally, the test method ordinarily used is Conditioned Play Audiometry (CPA). Under this method of testing, children are conditioned to perform a task every time a sound is presented. For example, every time the

stimulus is presented, the child will put a block on a tower. This is also done at progressively softer intensities until the child's threshold is obtained for as many frequencies as possible. If being done under headphones or insert-earphones, typically the audiologist will alternate between the two ears so responses can be compared at each frequency due to possible fatigue setting in. Testing also usually is prioritized to obtain a threshold for a low and a high frequency in order to measure sloping or rising hearing loss. Once children are older than 5 years developmental age, they can often participate in standard methods of audiometry (AAA, 2018).

Speech audiometry is also variable depending on children's developmental age, and capabilities. For children where VRA is used, SDTs can be obtained, as aforementioned. Some children who participate in VRA, and those who participate in play, can be tested to obtain speech reception thresholds (SRT) as well, where two-syllable words are repeated back until a threshold is obtained. Children who have very unclear speech, or who are non-verbal, can point to corresponding images of test words being presented (AAA, 2018). This is done at progressively softer levels until they are unable to repeat or select the correct word for a certain number of trials, or stop responding altogether (e.g. 4 out of 6 responses are incorrect). The last intensity level where they are able to provide the necessary amount of correct responses is deemed their SRT. Word recognition scores are also measured on children who can participate and repeat back a series of monosyllabic words to obtain a percentage score. These scores often help determine how well children with hearing loss will do with amplification.

Diagnosis of Children with Hearing Loss and ASD

Audiological assessment can be difficult in children with ASD, and test-retest reliability can be poor when they are tested behaviorally. The aforementioned methods of testing may not yield consistent results in this population. Additionally, when children have both ASD and a

hearing loss, the diagnosis of one often leads to a delay in diagnosing the other (Beers et al., 2014). In a systematic review by Beers et al. (2014), 33 articles were reviewed to establish the relationship between ASD and peripheral hearing loss. These 33 articles included 1 systematic review of Cohort studies, 2 Individual Cohort studies, 3 systematic reviews of Case-control studies, 13 Individual Case-control studies, 8 Case studies, and 6 Expert opinions. When discussing audiological evaluation in children with ASD, it was noted that test settings and procedures often have to be modified when assessing children with ASD, because they can be unresponsive to stimuli, and may become upset in the soundbooth. Children with ASD may resist wearing headphones, insert earphones or a bone oscillator, or having a probe inserted into their ear for testing impedance or otoacoustic emissions. Tone-burst ABR is an objective way to approximate the audiogram in cases where it is not possible to obtain reliable behavioral responses to stimuli (Beers et al., 2014).

Some typical signs of autism include speech-language delays, regression of developmental milestones at 18 – 24 months of age, reduced eye contact, tactile defensiveness, and repetitive and self-stimulating behaviors (Beers et al., 2014). Children who demonstrate avoidance of sensory stimuli have been seen to engage less in novel play environments and show reduced independence in self-care at home (Beers et al., 2014). As aforementioned, hyper-responsiveness to auditory stimuli may also be evident in children with ASD, and may be taken into account when testing, and fitting amplification. Beers et al. define hyper-responsiveness as "the pattern of exaggerated behavioral reactions to sensory stimuli often displayed by children with ASD and is an umbrella term that includes hyperacusis, hypersensitivity, sensory defensiveness, sensory modulation dysfunction, aversion, avoidance, hyperarousal and lack of habituation to sensory stimuli" (Beers et al., 2014).

An article by Szarkowski et al. (2014) outlines the difficulty of diagnosis when hearing loss and ASD are comorbid. Early intervention is critical for both of these diagnoses, and if ASD is missed in a child with hearing loss, or vice versa, they will not receive the services they need. "When hearing status is determined early in life, efforts to intervene and promote children's development are often focused on addressing issues known to commonly arise in children who are [deaf/hard of hearing], such as speech-language communication therapy" (Szarkowski et al., 2014). This may decrease the amount of attention given to children's other behaviors, and therefore could delay the diagnosis of ASD. This delay can then have a negative impact on overall development and language, especially social development. Another difficult factor in this diagnosis is the similarity of symptoms observed between ASD and hearing loss. Some examples of these include: "overall language delays and difficulties with particular areas of language functioning, delayed theory of mind, failure to respond to one's name, and pragmatic language difficulties" (Szarkowski et al., 2014). According to Peters, Remmel, and Richard, the term theory of mind (ToM) refers to, "the understanding of mental and emotional states such as desires and beliefs that allows individuals to predict and explain the behaviors of others. ToM first emerges in typically developing children as early as 15 to 18 months in the form of understanding of intentional behavior" (2009). However, while deaf children of hearing parents may show delayed ToM, the delay is not as significant or persistent as in children with ASD. Children with both ASD and hearing loss may present with similar symptoms as children who can hear, but still have ASD. Examples of such symptoms are reduced levels or avoidance of eye contact, reduced use of gesture and joint attention, and problems with turn taking, among others (Szarkowski et al., 2014).

However, some symptoms can help distinguish between the diagnoses. While patterns of

repetitive behavior or restricted interests are typical for children with ASD, they are usually not seen among typically developing children who are deaf or hard of hearing (Szarkowski et al., 2014). These can sometimes be seen in children with developmental delays with hearing loss, but not with ASD. Additionally, typically developing children who have hearing loss may have some atypical sensory responses or hyper-/hypo-sensitivities, similar to children with ASD, but less persistent (Szarkowski et al., 2014). Szarkowski et al. also stated that, "The presence of [hearing loss] may confound results of standardized tests used to evaluate children for ASD, leading to either under or over identification" (2014). They went on to say that the Autism Diagnostic Observation Schedule, Second Edition (ADOS-2), which is considered a "gold standard" in assessing ASD, directly states that it was not intended for use for children with sensory impairments (e.g. deafness).

Another interesting aspect in the diagnosis of ASD in children with hearing loss is the age of implantation for cochlear implants. The criteria of eligibility for cochlear implantation continue to grow broader, and the age of implantation has gotten younger. With children receiving CIs at increasingly younger ages, the diagnosis of ASD is often not made until after implantation, when it becomes evident that something more than a hearing loss is present, as evidenced by less progress than expected postimplantation. "Although the age of diagnosis of ASD is falling, most children are not diagnosed until approximately the age of 3 and most children will have already received their Cl by then" (Fountain et al., 2011, as cited by Robertson, 2013). Therefore, regardless of whether or not children with ASD may be contraindicated for CI use later on, they may have been implanted before a formal diagnosis has been made.

Considerations for Children with ASD and CI

Due to their unique characteristics, children with ASD have some additional needs when it comes to being fit with any amplification device, including a cochlear implant. Hyper-responsiveness to auditory stimuli, as mentioned above, is not uncommon in children with ASD, and should be considered when making recommendations for, or programming amplification, as well as planning behavioral intervention, in order to facilitate acceptance of new technology (Beers et al., 2014). As some children with ASD experience increased perception of loudness, and display hyper-responsive behaviors to auditory stimuli, initial output levels for any amplification device may be limited in order to foster acceptance of new equipment. Additionally, as cited in Beers et al., (2014), Daneshi et al. found that, "children with ASD showed limited development in auditory perception following implantation compared to deaf patients with other secondary disabilities." Success of children with ASD postimplantation is unclear largely due to the limited research in this area. Preoperative counseling is necessary in order to set realistic expectations with families or caregivers (Beers et al., 2014).

In a study by Cupples et al., (2013) the speech, language, and functional auditory outcomes of 119 3-year-old children with hearing loss and additional disabilities were evaluated by means of direct assessment, as well as caregiver report. Of these 119 children, 9 had ASD. According to Cupples et al., "Approximately 20–40% of children born with hearing loss also have significant additional disabilities that might prevent them from reaching their full potential in regard to speech, language, cognitive, or social-communicative outcomes" (2013). Within the study, children were divided into two groups, Group A included children with ASD, cerebral palsy (CP), and/or developmental delay (DD), while Group B was composed of children with vision or speech output impairments, syndromes not entailing DD, or medical disorders.

Children in this study were evaluated using the PLS-4 to provide a formal assessment of children's overall receptive and expressive language abilities, the PPVT-4 (Peabody Picture Vocabulary Test Fourth Edition) measures receptive language, particularly vocabulary, and the phonology subtest of the DEAP (Diagnostic Evaluation of Articulation and Phonology) to quantitatively measure children's speech production ability. Report-based evaluations used were the Child Development Inventory (CDI), to obtain caregiver report of participants' receptive and expressive language abilities, the Parent Evaluation of Aural/Oral Performance of Children (PEACH), to obtain a measure of participants' functional auditory performance in everyday situations as reported by their caregivers, and a Speech Intelligibility Rating (SIR), as ascertained by a research speech-language pathologist.

Cupples et al. report that, "Children with ASD, CP, and/or DD were more often than not unable to cope with the task demands, whereas children with vision or speech output impairments, various syndromes not entailing DD, or medical disorders achieved consistently high rates of completion (75% or better)" (2013). The results of this study revealed that the children in Group A, which included those with ASD, performed lower in terms of receptive and expressive language outcomes than children in Group B (other disabilities) on both the PLS-4 and CDI. Also, Group A children also received poorer caregiver ratings on the PEACH, and poorer clinician ratings of speech intelligibility. When controlling for all demographic variables apart from disability group, Cupples et al. (2013) found that disability group accounted for significant unique variance in receptive and expressive language outcomes on both the PLS-4 and the CDI, and that, in Group A, degree of hearing loss was not an important correlate of language outcomes. They went on to state that, "It is possible that for children with ASD, CP, and/or DD, the additional disability itself was of paramount importance in determining their

capacity to acquire language skills" (Cupples et al., 2013). It was noted that children with ASD were less likely to cope with the demands of formal testing than children in any other disability category, and they attained the lowest average scores on both receptive and expressive language scales of the PLS-4 and CDI. However, since only 9 children with ASD took part in this study, they could not be considered as a separate group for analysis.

In 2013, Özdemir et al. evaluated etiologic factors on non- and limited use of CI in children. "Limited use" is used for the recipients who used the implant for fewer than 2 hours a day while awake, and "non-use" is used for complete rejection of implant usage of implant. The Listening progress profile (LiP) and Meaningful Auditory Integration Scale (MAIS) tests were also utilized to analyze the auditory performances of the patients. Out of 413 CI recipients (200 males, 213 females) under the age of 16, 12 children patients (7 male and 5 female patients; age range, 5– 13 years) were limited and non-users of cochlear implants, with a follow-up of at least 24 months. None of these children had experienced device failure or any medical or surgical complication. Of these 12 patients, 4 were nonusers and 8 were limited users. The factors that could have caused this usage problem (e.g. presence of additional disabilities, and family interest) were investigated. This revealed that 3 of the limited users, and 1 of the non-users had ASD. Özdemir et al. state that, "Although additional disabilities (mild/moderate mental retardation, attention deficit/hyperactivity disorder, cerebral palsy, learning disability, congenital blindness, autism spectrum disorder) are not a contraindication for cochlear implantation, especially autistic and congenitally deaf-blind patients show limited development" (2013). This study suggested that children with additional disabilities who receive CIs may experience a great deal of failure because of the characteristics of their disability, or due to a lack of unique and specific rehabilitation needed for them to progress with their CI.

In sum, children with ASD may experience many difficulties prior to cochlear implantation, which persist postoperatively. Cupples et al. (2013), reported that children with ASD and hearing loss were more likely to be unable to complete assessments, scored lower on receptive and expressive language tasks when they could be completed, and also received worse results on caregiver report than other children with hearing loss and additional disabilities, suggesting that ASD plays more of a role in outcomes with CIs than the hearing loss itself does. Finally, Özdemir et al. (2013) found that of 12 children that were limited or non-users of CIs, 4 had a diagnosis of ASD. Therefore, while these children are still being implanted, appropriate preoperative counseling is critical for helping families understand the higher risk of limited or non-use of cochlear implants, especially by children with autism, as well as a potential for limited development or progress from cochlear implantation.

Purpose of Current Study:

The research described above outlines the diagnoses of hearing loss and ASD together, as well as considerations for CI use in this population. The literature provides evidence that, in many cases, diagnosis of ASD may be mistaken for hearing loss, and vice versa, as well as the fact that the diagnosis of one may cause a delay in diagnosing the other. It was made critical that diagnoses be made as soon as possible so appropriate intervention can be provided for these children. In terms of the considerations regarding cochlear implantation in children with ASD and hearing loss, the most pertinent findings were that children with ASD do not show as much progress in language outcomes as those children with hearing loss and other comorbidities.

If anything can be concluded from the aforementioned research, it is that degree of success with CIs in children with ASD and hearing loss is highly variable between studies, and between individuals. Since ASD is a spectrum disorder, children range greatly in their

performance on language tasks, and in their ability to complete them. Given the lack of definitive research on behavioral outcomes of children with ASD and hearing loss, and the limited results of studies with small numbers of participants, further research is required in order to establish if there is a general trend in behaviors postimplantation, as well as if there is a pattern of progress that differs greatly from that of typically developing children who use CIs, or those with CIs and other comorbidities. Therefore, this systematic review sought to encompass current research, specifically regarding behavior outcomes of this population, in order to draw further conclusions.

The purpose of this systematic review is to evaluate whether or not cochlear implantation has a significant effect on the behaviors of this population postimplantation, as compared to typically developing children with CIs. Such behaviors as aggression, lack of joint attention, and resistance to use of amplification were examined, among others. The research questions are defined as follows: (1) Given that some children with ASD are prone to aversion to certain stimuli, what are the behavioral outcomes, for those with cochlear implants as measured both subjectively and objectively? (2) Are there significant differences in outcomes for children with ASD after cochlear implantation, versus children with hearing loss with or without other comorbidities: Is there a general trend? (3) What mode of communication do children in this population utilize? Hypotheses regarding these research questions include that children with ASD will have substantially more of the aforementioned behaviors postimplantation, as compared to the children with hearing loss without ASD. However, these may be maturational rather than being attributed to cochlear implantation. These children are also hypothesized to make progress with CI, but more slowly than their typically developing peers due to their developmental disability, and more slowly than those children with CIs and other comorbidities.

Lastly, these children are hypothesized to use a non-verbal mode of communication over oral language. The following summary of literature outlines the current findings in the field of hearing loss in children with ASD regarding: objective and subjective measures of evaluating language and behavior in this population, and how outcomes compare to typically developing children with CIs, mode of communication used by children with ASD and CIs, and effects of other comorbidities on outcomes.

Methods:

Search Criteria

To find the research included in this systematic review, all EBSCOhost databases were searched, as well as the PubMed database. Articles were found using search terms "Autism" and "Cochlear Implant." The criteria were further narrowed by accepting only articles that were peer-reviewed, and for which the full text was available. The timeframe for this research was set for articles published between 2008 and 2019. Only articles written, or translated, in English were accepted. This search criteria yielded 92 articles; 60 articles were found on PubMed, and 32 articles were found in the EBSCOhost databases. Once duplicates were removed, 22 articles remained on PubMed, and 21 on EBSCOhost. The abstracts from these articles were analyzed to determine their relevance to the current study, and 7 were excluded. Repeated literature searches with the same criteria were completed to maintain that all of the most current articles were included. These searches yielded two additional studies that were applicable, and thus they were included in the research to be evaluated more thoroughly to determine final acceptance into the systematic review. A chart illustrating the flow of this literature search can be seen in Figure 2.

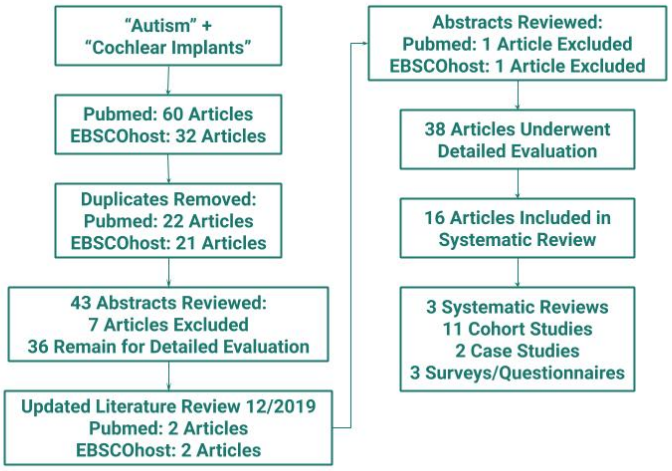

Fig. 2 *Flow Chart of Systematic Review*
Breakdown of articles included in this systematic review.
Note: Some articles contained multiple types of research,
and have been categorized accordingly.

Research Selection

From the search criteria outlined above, 38 articles underwent a detailed evaluation to determine if they were accepted into the systematic review. Articles were included if they pertained to behavioral outcomes in children with ASD, who wear CIs. Studies that focused solely on speech and language outcomes, academic performance, or other non-behavioral outcomes were excluded. The systematic review comprises 16 total articles. Some of the articles included fall into more than one category, as they complete multiple types of research. As such, this review contains: 11 cohort studies, 3 literature reviews, 2 case studies, and 3 surveys/questionnaires.

Data Analysis

Information was extracted from the included articles, analyzed for patterns, and ultimately summarized in a narrative format. The purpose of this systematic review is to identify if cochlear implants consistently lead to a change in behavioral outcomes for children with ASD and peripheral hearing loss, what means of measuring benefit are used, what mode of communication this population uses, and the impact of additional comorbidities on these results. Due to the variable study designs, inclusion/exclusion of subjects, evaluation methods, and measured outcomes, a systematic review, and not a meta-analysis, was selected as the format for the current study.

Results:

Objective Outcome Measures

As discussed above, children with ASD often cannot complete outcome measures, or diagnostic testing. Additionally, there are no measures standardized for children on the spectrum with comorbid hearing loss. As such, many of the articles included in this paper used different objective measures to measure outcomes, with some, though little, overlap. The simplest objective measure of if a child with ASD is progressing with their device may be whether or not they use it. In a study by Rodriguez-Valero et al., 22 children with CIs who were subsequently diagnosed with ASD were selected and examined via retrospective chart review with regard to compliance with their device, and mode of communication used (2016). On average, these children were diagnosed with ASD two years after receiving their CIs. The researchers note that, "There is a range of level of disabilities in ASD, with some relatively minor social communication difficulties through to severe language, cognitive, and behavioural difficulties" (2016).

In terms of compliance with their devices, 16 (72.7%) use their cochlear implants, 13 (59.1%) had periods of intermittent CI use, and 6 (27.2%) went on to become non-users (Rodriguez-Valero et al., 2016). Of note, four of the six children who became non-users had documented learning disabilities, indicating their ASD symptoms were more pervasive. Interestingly, two children presented with hyperacusis, however both children continued to be compliant with device use. As most of the cohort did consistently use their device, the researchers report that patient compliance with CIs with comorbid ASD is acceptable, though variable, and appears to change in accordance with the degree of ASD symptoms. They state directly that, for this population, "The main factor that appeared to affect usage in this context

was the severity of the disorder" (Rodriguez-Valero et al., 2016).

Robertson (2013) also looked at device compliance in a cohort of 10 children with ASD and CIs. Within this cohort of 10 children, many had other comorbidities in addition to ASD. These comorbidities included intellectual disability and cerebral palsy, and epilepsy. Within the cohort, six children use their implants consistently, two use them inconsistently, and the final two are non-users of their devices. Of the two who became non-users, one had no additional comorbidities, and the other had cerebral palsy and an intellectual disability. Robertson states that for one of these children, "there was evidence that the auditory stimulation was exacerbating her behaviour problems. Outcomes were clearly related to the severity of the autism, and it can be difficult to predict severity and to counsel families appropriately regarding realistic expectations" (2013). In both of these studies, the majority of the participants consistently used their devices, despite comorbid ASD. However, they both note that compliance is correlated with the severity of ASD symptoms a child presents with on an individual basis.

For receptive and expressive language outcomes, three papers used the CAP (categories of Auditory Performance) and the SIR (Speech Intelligibility Rating). The first of these measures, the CAP, is used to, "classify auditory receptive ability into eight performance categories that increase in difficulty (from 'no awareness of the environment' to the 'use of the telephone with known users')" (Mikic et al., 2016). Whereas the SIR rates speech intelligibility on a 5-point scale ranging from unintelligible to speech that is intelligible to all listeners, and is more complex in nature.

In 2016, Mikic et al. observed 14 children with cochlear implants, and evaluated their auditory perception and speech intelligibility via the CAP and SIR. The children included in the study had congenital bilateral profound sensorineural hearing loss, and received little-to-no

benefit from amplification. They were all subsequently implanted between 12 and 18 months of age. Of these 14 children, four had the comorbid diagnosis of ASD (Mikic et al., 2016). The purpose of this study was to compare the scores on the aforementioned measures between those children who are typically developing and receive CIs, with those children with ASD and CIs.

The children were initially evaluated at two years of age, and then annually until age 6 years. Results revealed that the typically developing children made steady progress, as was expected. For the children with ASD, scores on the CAP improved at a much slower rate. Researchers found that, depending on the child, at six years of age some were just reaching the point where they could discriminate speech sounds, or identify sounds in their environment (Mikic et al., 2016). Additionally, scores on the SIR were at most a two out of five, even at six years of age and with speech and hearing therapy. The authors report that postimplantation outcomes deaf children with ASD are both uncertain and unpredictable. In this study, none of the subjects developed expressive speech that was generally intelligible as categorized by the SIR. The researchers state that, "In this study, communication skills, receptive and expressive language development were strongly affected by a degree of autistic features expression" (Mikic et al., 2016).

In the aforementioned study by Robertson (2013), these measures were also used to complete a case study on 1 of their participants. With regard to his performance, at seven years postimplantation, it was reported that the child was able to understand common phrases, and that his speech remained unintelligible. While this progress may seem limited, it is an improvement from his lack of awareness to sounds and his lack of any spoken language before receiving his CI.

Nasralla et al. used the CAP as well as the CL (Categories of Language) to measure

postoperative language outcomes in 14 children with CIs and additional disabilities (2018). Of these 14, 4 had ASD. The children with ASD also had other comorbidities, including cognitive delays and ADHD (Attention Deficit Hyperactivity Disorder). The CL is a similar measure to that SIR, in that it ranges from a rating of 1, where the child does not speak at all, to a rating of 5 where the child produces connected and more complex speech. Three of the four children with ASD in this study showed very limited progress on both the CAP and CL when they were evaluated postoperatively. The time of evaluation was not set across the children, however ranging from 9 months to 4 years 11 months of CI use, these three children scored in the lowest category for both CAP and CL scales. The fourth child with ASD, after a period of 9 years and 8 months of CI use, scored in category 4 and 5 for the CAP and CL, respectively, indicating much more advanced auditory perception and expressive language skills than the remainder of this group (Nasralla et al., 2018).

The final study that used the SIR is one by Cupples et al. (2013), described previously in this paper. This study is one including nine children with ASD and hearing loss, who are grouped with children with cerebral palsy, and developmental delay. The results of this study again correlated with those described above, in that the children in this group scored poorer on the SIR consistently, as compared to the other group of children with hearing loss and various other disabilities or medical conditions. This study, however, does not separate children who use hearing aids from those who have CIs.

In a retrospective study by Eshraghi et al. (2015), speech perception and expression scores in children with ASD and CIs were compared to those of typically developing children with CIs. Rather than the CAP and SIR, in this study, speech discrimination was evaluated in quiet using the Early Speech Perception (ESP) test, Multisyllabic Lexical Neighborhood Test

(MLNT), or the Phonetically Balanced Kindergarten (PBK) test. This study population consisted of 15 children with ASD, and 15 CI users with no comorbidities who served as controls.

Regarding speech perception, in the ASD group, 12 out of 14 children with previous data available to use for comparison, were in the lowest categories (0 - 1). Postoperatively, most children with ASD significantly improved their speech perception skills, with 10 out of 15 having a speech perception category of 3 or 4, while 4 out of 15 were rated in category 1, and none remained in category 0. For speech expression, 13 out of 14 children were in the lowest categories (0 - 1) preoperatively. Postoperatively, children in the ASD group significantly improved their expressive vocabulary: 9 out of 15 scored in a category of 3 or 4, (could communicate using simple phrases and some sentences), 5 out of 15 remained in category 1, and none remained in category 0. While the children with ASD made significant improvements, these improvements were not as great as those seen in the control group.

With specific regard to behavioral outcomes, two studies used objective measures as means of measuring the benefit of implantation. In a longitudinal study by Cruz et al. (2012), the effects of cochlear implantation on language and behavioral outcomes in children with and without additional disabilities were examined three years postimplantation. The study included 188 deaf children, 157 of which had no comorbidities, while 31 children had diagnoses of additional disabilities. Of these 31 children, 8 had ASD. These children were evaluated on their oral language, both receptive and expressive, and their behavioral outcomes, both internalizing and externalizing. Their oral language was evaluated via the Reynell Developmental Language Scales (RDLS), while their behavioral outcomes were evaluated via the Child Behavior Checklist (CBCL). Internalizing behaviors were measured on four subscales: Emotional Reactivity, Anxious/Depressed, Somatic Complaints, and Withdrawn. Externalizing behaviors

were measured on two subscales: Attention Problems and Aggressive Behavior.

According to this article, research has consistently reported that deaf children and children with developmental disabilities have higher rates of behavior problems than children without disabilities, and children with sensorineural hearing loss also show higher rates of externalizing behavior problems, (e.g. inattention and aggression) than children with normal hearing. These past findings would suggest that, "language influences behavior problems by limiting the child's ability to effectively communicate with others, or by affecting emotional and behavioral regulation" (Cruz et al., 2012).

Within the study, children with ASD had the lowest language scores prior to cochlear implantation as compared to any other disability included. Additionally, three years postimplantation, these children still scored lower on language measures than children with other disabilities (e.g. Children with ADHD had oral language scores similar to typically developing children using CIs). While all children improved their oral language, children with ASD improved at half the rate of the Deaf group. In terms of behaviors, children who received a CI, but had no other diagnoses evidenced no change in their internalizing behavior problems and a decrease in their externalizing behavior problems, whereas all children with a CI and additional disabilities evidenced higher rates of externalizing behavior problems three years postimplantation, and no change in internalizing behaviors.

In a study by Johnson et al. (2008), two children with CIs were evaluated on receptive and expressive language skills via the Reynell Developmental Language Scales and the MacArthur-Bates Communicative Development Inventories at multiple points in time: before implantation (baseline), and at 12 months postimplantation. Additionally, the two children were observed in regard to their joint attention and symbolic play through coded video recordings at

baseline, 6 months postimplantation, and 12 months postimplantation. One of the two children selected for these case studies has ASD.

Both joint attention and symbolic play are crucial in developing language and social skills. Johnson et al state that joint attention refers to a child's ability to, "share attention with both a partner and an object or event, allowing the child to observe how others assign meaning to new objects and situations" (2008). Without auditory stimulation or access, deaf children may experience delays in joint attention. Symbolic play is a skill in which children use one item to represent another (e.g. a block represents a car) during play. The combination of these two tasks is being used as a means of observing the process of language acquisition in the children included in this study up to 12 months postoperatively.

The child with ASD included in this study was diagnosed with hearing loss at 2, and was implanted at the age of 4 years 4 months. He was diagnosed with ASD 3 months postimplantation. At baseline, this child presented with an age-equivalent of 16 months on the RDLS and CDI with regard to receptive language. Expressive language could not be assessed for the RDLS, as the child could not continuously attend to the task. However, for the expressive language tasks on the CDI, the child had an age equivalent of 17 months. At 12 months postimplantation, his scores on the RDLS placed him at age 23 and 22 months for receptive and expressive language, respectively. For the CDI, scores similarly increased, placing him in the range of 23 to 24 months of age. However, when observing his play in the video recorded sessions with his mother, he, "showed a pattern typical of children with ASD. He spent most of his time focusing on objects instead of engaging with his parent, a behavioral pattern that became more evident at 6 and 12 months [postimplantation]" (Johnson et al., 2008). Some improvements in his responsiveness to his mother's communication attempts were noted.

Additionally, the child made gains in symbolic play and sustained attention. Ultimately, at 12 months postimplantation, the child made no attempt to attain joint attention with his mother, as is common in children with ASD regardless of hearing status (Johnson et al., 2018).

Objective measures have been used to measures language outcomes, device compliance, and behavioral changes in children with ASD and CIs. Regarding device compliance, the researchers included here both found that a majority of the children in their studies consistently used their devices. However, they also both noted that children who did not use their devices had a more severe display of symptoms, and their behavior caused decreased compliance. With regard to speech and language outcomes, progress was variable. What progress was made, was made slowly, and rates half that of children with CIs and no comorbidities in some cases (Cruz et al., 2012). Again, these researchers note that more limited progress was made in children whose ASD symptoms were more pervasive. And lastly, and most importantly for the purpose of this paper, the two studies who used objective measures to determine changes in behavior postimplantation revealed increases in externalizing behaviors in one case, and no change in autistic behaviors in the other (Cruz et al., 2012; Johnson et al., 2018).

Subjective Outcome Measures

For the very same reasons mentioned above, many researchers utilize subjective measures to both corroborate and supplement outcomes obtained through objective means. Many studies included in this review relied on parental reports or surveys to determine perceived benefit of these devices. In 2018, Lachowska et al. conducted a retrospective study analyzing 6 children with ASD who wore CIs, in order to assess reaction to music and sound, spoken child's name, and requests, and administer a questionnaire for parents. There were 6 questions on the parental questionnaire, including: "Does the child respond to his/her name in quiet with auditory

cues only (no visual cues)?", "Is the child's behavior affected while wearing his/her sound processor?" and, "[Have] the family interactions with the child and within the family benefited from implant?" (Lachowska et al., 2018). Responses to the parental survey shed a more positive light on CI use in this population. Improvements were more commonly seen with regard to response to their name and environmental sounds. Responses revealed that, "most of the children presented reduced anxiety when wearing the sound processor. Amount of eye contact was the least improved factor in this study. In two cases the behavior did not change despite the processor on but at the same time no increased hyperactivity associated with daily use cochlear implant was observed" (Lachowska et al., 2018). Perhaps most importantly, all families reported benefits in their child's personal interaction with family members, and within the family postimplantation. Therefore, the results of this study support the conclusion that while a CI does not definitively allow children with ASD to develop speech and language, they may enhance quality of life for this population.

To a similar result, in the aforementioned study by Eshraghi et al. (2015), a parental survey of 39 subjective questions evaluating CI benefits was administered to parents of children in the ASD group by telephone interview, focusing on three core characteristics of ASD: 1) communication skills, 2) behavior, and 3) interaction with others, as well as asking about overall device satisfaction. For this survey, 13 families of children in the ASD agreed to participate. Their responses revealed that the top three improvements after CI were name recognition, response to verbal requests, and enjoyment of music, while use of eye contact was least improved. Eshraghi et al. further stated that, "the most improved aspects after CI were attending to other people's requests and conforming to family routines. Awareness of the child's environment is the most highly ranked improvement attributed to the CI" (2015). When asked,

12 out of 13 parents said the success with CI was better than they had expected, and they would recommend CI to another family member in a similar situation. Overall, while children with ASD may not make the same strides in language and communication skills as typically developing children who use CIs, they are still able to receive benefits in various other areas.

Nasralla et al. (2018) sought to create a comprehensive parent questionnaire for the families of children with CISs other comorbidities. This survey contained questions regarding type of disability, amount of use of the CI, interparental and familial support, and professional support. The questionnaire also included questions addressing advantages of the CI, and their child's communication skills (including mode of communication). With specific focus on behavior, there were questions on social-emotional abilities, and, "interests, behaviors, temperament, family and social interactions, independence during activities of daily life (ADL), adaptive potential, self-control, openness to experiences, and learning styles" (Nasralla et al., (2018). Of import, parents were also asked if their expectations were met in terms of their child's progress with their CI, and if they would recommend implantation to another family in a similar situation to their own.

On the whole, the results of this questionnaire were positive. Postimplantation, parents of children with disabilities reported that their children have improved with regard to communication, sociability, and adaptability. Increased reaction to sound led to increases in name recognition and eye contact in many participants. This in turn led to increases in enjoyment from music, and engagement in school activities (Nasralla et al., 2018). Increases in expressive communication were seen in nearly all children, including three of the four with ASD. 84 percent of children participated in eye contact after being implanted, up from 56 percent

preoperatively. This facilitated significant improvements in playing with other children, and adaptiveness to family routines, as well as to novel situations.

However, six of the families included in this study reported they were frustrated with the amount or type of results seen postoperatively. These included three of the four families of children with ASD. In terms of why they were frustrated, "one was the mother of a child who was diagnosed 3 years after cochlear implantation, while two did not accept their children's diagnoses and expected more verbal fluency" (Nasralla et al., 2018). The researchers note that as the age of implantation is very low, parents aren't given the chance to develop realistic expectations for their child's performance with a CI, because other developmental issues or disorders may not be known at that time. This was especially evident for those children with ASD, and negatively impacted the family's perception of the CI and its benefits.

Nasralla et al. (2018) state that, "the main and most exciting revelation of this study came through cases involving more serious conditions, when the mothers of several patients revealed that the implants made their children happier." Every family included, even those who were frustrated with some of the results, responded that they would recommend CIs to other families of children with additional disabilities. The researchers state that the benefits seen in both communication and socio-emotional development improve quality of life in these children, and their families.

Other researchers make note of subjective behavioral measures or changes in less explicit ways. For example, Cupples et al. (2013) note that children in ASD group received poorer parental ratings on the PEACH. In the aforementioned study by Mikic et al. (2016), what the researchers say of behavior, they say anecdotally in reference to past research. This research revealed that children with ASD showed improvements in response to requests, eye contact, and

behavior on the whole (Donaldson et al., 2004, as cited by Mikic et al., 2016). However, they do not explicitly report behavioral changes in their own participants postimplantation. In a study by Yamazaki et al. subsequently discussed in this paper, researchers state that while children with ASD, CMV, and CIs, "could not understand the meaning of spoken words or sentences, their mothers thought that CI was effective, because behaviors and family interactions of deaf children with autistic spectrum disorders were improved to some extent after the implantation" (2011).

The subjective measures used to evaluate benefit of CIs in children with ASD, and their changes in behavior illuminate the positives in many cases. Something as simple as children responding to sounds, or to their name, can greatly benefit the family's perception of the use of a CI. In the studies detailed in this section, responsiveness to sounds and requests led to participants being better able to attend to their therapies, their schooling, and to the routines of their families. However, it is important to note that parents do often feel frustration in their child's lack of expected progress if the diagnosis of ASD was not known prior to implantation, as seen in Nasralla et al., 2018). As seen in the objective measures as well, eye contact is least affected by CIs, though some research has reported improvements. In terms of subjective benefit, these changes in a child's reaction to sounds or communication attempts by his parents can increase quality of life for both himself and the family as a whole.

Mode of Communication

For many parents of children with hearing loss, oral or spoken language is the end goal. However, for children with ASD and hearing loss, this goal may not be realistic. The likelihood of children with ASD and CIs developing oral language is comparable to that of children with ASD alone. Many studies done within this population report highly variable results with regard to preferred mode of communication. Many of the studies included here categorize modes of

communication differently, in that they don't all include the same categories. Oral language, sign language, and combinations of communication modalities are addressed in many studies. Reliance on behavior, gesture, or non-verbal means of communication are addressed in others, while some studies report a complete lack of communicative abilities or intent, or that the children or their families are undecided as to what the primary mode of communication is.

Somewhat unique to this population is the use of Augmentative and Alternative Communication (AAC). According to Lloyd et al. AAC is defined both as, "The supplementation or replacement of natural speech and/or writing using aided and/or unaided symbols" and "The field or area of clinical/educational practice to improve the communication skills of individuals with little or no functional speech" (1997, as cited by the AAC Institute, n.d.). One of the most well-known methods of AAC used for children with ASD is PECS (Picture Exchange Communication System). PECS is described as consisting of six phases, with the initial phase, "teaching an individual to give a single picture of a desired item or action to a 'communicative partner' who immediately honors the exchange as a request" (PECS, n.d.) The phases get gradually more complex, and include picture discrimination, and well as sentences and questions. As many children with ASD do not develop spoken language, it is unsurprising that some children with ASD and comorbid hearing loss also use PECS or other AAC systems.

A graph representing mode of communication as reported in nine of the studies included in this review can be seen in Table 1. Of note, two of these studies (Cruz et al., 2012; Cupples et al., 2013) had children in groups containing more than one disability and did not explicitly document the mode of communication for their participants with ASD. As such, the communication modality of their entire group was documented in Table 1. Three of the studies reporting mode of communication are highlighted below.

In a 2014 study by Meinzen-Derr et al., 24 children with both ASD and permanent hearing loss were examined in terms of degree and etiology of hearing loss, use and type of amplification, and language and communication. Most of the children included in this study had severe to profound hearing loss (16 children), whereas less had mild to moderate or unilateral hearing loss (8 children). For this particular sample of children, "Nine [...] children with a dual diagnosis of hearing loss and ASD used speech as their mode of communication (oral communicators). Nine children used a combination of sign language and behavior, while 6 children used only behavior for communication" (Meinzen-Derr et al., 2014). In this study, behavior refers to the children acting out as their method of communicating wants and needs. When specifically looking at children with cochlear implants, eight used augmentative communication strategies, such as PECS (Picture Exchange Communication System).

Of the 24 children in this study, 14 received CIs. Of those 14, two became non-users. These 14 children also varied in their method of communication, in that two used oral language only, and the remaining children used some combination or oral language, sign language, or behavior to communicate. On language and communication tasks, Meinzen-Derr et al. found that "Outcomes and expectations for children with ASD and cochlear implants are as variable as outcomes and expectations for children with ASD who have normal hearing. Thus, the severity of autism may also contribute to communication outcomes" (2014). The researchers go on to say that forming a groundwork for language by any means is essential in this population, rather than simply focusing on word recognition scores postimplantation. This research revealed that some children developed speech after an augmentative communication system had been implemented. This furthers their notion that "Focusing only on an oral or signing approach often is insufficient at helping a child make progress with the core communication deficits commonly seen in ASD"

(Meinzen-Derr et al., 2014). CIs can provide auditory stimulation necessary for children with ASD and hearing loss to develop oral language, pending that it is a method of communication they can operate in, with or without other supports.

In the study by Robertson (2013), the communication modalities of the ten participants with ASD and CIs were also documented. From these ten, four have some extent of oral language, though only one uses it alone, and the other three use it in combination with sign language. Robertson (2013) reports that these children are happy with their implants. Five of these children rely solely on AAC to communicate, and the final child uses only sign language to communicate. The author notes that the results of this study related to severity of ASD symptoms. Robertson claims that it is possible for children in this population to receive benefit from CIs and achieve some spoken language skills. However, she notes that, "other children with ASD and CI will rely on non-verbal means as their primary mode of communication, and some may reject the speech processor completely" (Robertson, 2013). Ultimately, the author notes that special regard must be given to a child's display of symptoms, particularly sensory sensitivities, as these can affect use and tolerance of the device, and subsequently, the child's communication modality.

In the study by Rodriguez-Valero et al. described above, the communication modalities of the 22 participants were also detailed (2016). Researchers report that 13 of their participants had some extent of verbal communication skills, while seven relied on non-verbal communication modalities (including sign language and AAC). They report that a majority of their population used at least two different modes of communication (12 out of 22 children). However, Rodriguez-Valero et al. also note that two of the children who participated in this study presented with a, "complete deficit in communication, with neither of them using their CI"

(2016). They go one to note that these two children had more severe ASD symptoms than the other children in their cohort, and their outcomes were thus affected.

The other studies that documented communication modality also found mixed results. Across these studies, the results of 182 children (94 with ASD) were recorded. Those who used only one mode of communication modality were only recorded in that category in the following table. Those who used any combination of communication modality were recorded in the 'combination' category. Some studies included additional categories for participants or caregivers who were undecided as to the primary modality used, those participants who had a complete lack of communication, or for those whose results were not explicitly recorded. In the example of Eshraghi et al. (2015), researchers stated that nine children with ASD and CIs had oral language, and five relied on non-verbal communication methods, which remained unspecified. They also noted that eight of these children used sign language to some extent, though they failed to mention which of these eight overlapped with those who used oral language or another non-verbal method. As such, those five participants were placed in the 'other' category.

From this sample of children across studies, 62 of the 177 (35%) used oral language as their sole communication modality. This encompasses roughly one third of the total population. Sign language encompasses a much smaller percentage, with only 13 participants (7.3 %) utilizing it alone, and behavior or other non-verbal means of communication account for only 6 children (3.4%) in this population. Children using only AAC were only 6 in number (3.4%). The largest group was those children who used a combination of at least two communication modalities. This group consisted of 69 children (40%). These numbers may be influenced by the presence of children with disabilities other than ASD, as they number 88 (48.4%). However,

when the two studies with those children were excluded, and the remaining data reanalyzed, the combination category still accounted for 23 out of 72 children (31.9%), making it the most prevalent communication modality. It was followed by oral language (26.4%). Behavioral or other non-verbal language, sign language, and AAC each accounted for 8.3%, and the remaining children were in the 'other' category (26.4%). All studies report high variability, with many stating that realistic expectations must be addressed with parents of these children, as spoken language is not a guaranteed outcome.

Table 1. Mode of Communication

	Mode of Communication					
	Oral Language	Sign Language	Behavior/ Non-verbal/ Gestural	Aug. Comm.	Combination	Other
Study	**N**	**N**	**N**	**N**	**N**	**N**
Cruz et al.[a]	8 (26%)	7 (23%)	N/A	N/A	7 (23%)	9 (29%)
Cupples et al.[b]	35 (47.3%)	-	N/A	N/A	39 (52.7%)	N/A
Eshraghi et al.[c]	9 (60%)	(8 [61.5%])	-	N/A	N/A	5 (33.3%)
Lachowska et al.	1 (16.7%)	-	-	N/A	N/A	5 (83.3%)
Meinzen-Derr et al.[d]	2 (14.3%)	-	2 (14.3%)	(8 [57.1%])	7 (50%)	N/A
Nasralla et al.	1 (25%)	1 (25%)	2 (25%)	N/A	N/A	N/A
Robertson	1 (10%)	1 (10%)	N/A	5 (50%)	3 (30%)	N/A
Rodriguez-Valero et al.[e]	5 (22.7%)	2 (9.1%)	-	1 (4.5%)	12 (54.5%)	2 (9.1%)
Yamazaki et al.	-	2 (100%)	-	N/A	-	N/A
Total = 182[f]	62 (34.1%)	13 (7.1%)	4 (2.2%)	14 (7.7%)	68 (37.4%)	21 (11.5%)

Mode of communication reported by number of children, and percentage of respective total population reported by study. Categories except 'Combination' denote that the child uses only that communication modality. The 'Other' category represents unspecified modes of communication, or complete lack of communication.
Abbreviations: Aug. Comm., Augmentative Communication; CP, cerebral palsy; DD, Developmental Disabilities
a. Combined results for all children with disabilities, and do not separate ASD; 8/31 have ASD
b. Combined results for ASD, CP, & DD; 9/74 have ASD. Only asked oral vs. sign language, or combination;
c. Children noted to use sign language from 'sometimes' to 'always.' Combinations of modality not reported.
d. Rodriguez-Valero et al. report 2/22 children have a complete deficit in language
e. Augmentative Communication was not included in the Combination category in Meinzen-Derr et al.
f. Only 94/182 participants included studies are diagnosed with ASD. However, as they are included in groups with children with other comorbidities, and their specific mode of communication is not detailed in their respective studies, the results from their entire groups are included here.

Comorbid Diagnoses

In addition to the symptoms of ASD having great effect on CI outcomes, the presence of cognitive impairments or additional comorbidities may exacerbate any issues, or further stunt progress. Beers et al., state that, "Approximately 80% of children with autism have some degree of cognitive impairment" (2014). Furthermore, the existence of ASD does not preclude children from having other disabilities; many studies included in this review have children with ASD and additional comorbidities. In a study by Motegi et al. (2019), 13 children with cochlear implants were evaluated using the Enjoji Scale. Of these children, five had no comorbid diagnoses, four had an intellectual disability (ID) and no other comorbidities, and the remaining four children had both an ID and ASD. The goal of this study was to analyze the changes in degree of developmental delays in children with hearing loss postimplantation, and compare the results between those children with or without ID and ASD, as children with both developmental disabilities and hearing loss are likely to have deficits in more areas than only speech and language. The Enjoji Scale of Infant Analytical Development is a questionnaire used to evaluate developmental milestones. This questionnaire is divided into 6 categories: language development, intelligence development, emotional development, social behavior development, manual activity development, and locomotor activity development (Motegi et al., 2019). This tool also includes questions in, “skill categories for the assessment of typical development (e.g., hearing, speech and language, intellectuality, emotion, social behavior, gross motor function, fine motor function, and vision)” (Motegi et al., 2019).

The children were all provided with aural habilitation and attended schools for the deaf or hard of hearing. The children were evaluated at various time intervals from 1 month to 84 months old. The degree of developmental delay for each of the aforementioned categories was

defined as the difference in months between developmental and chronological age. The results of this research were such that children who had CIs, but no other developmental disabilities showed delays in language and intelligence development (which are defined for the purposes of this study as utterance ability, and language perception ability, respectively). These delays recovered over time with CIs, even to near the range of normal for language development.

For the children with CIs and also ID, there the same delays were present. However, unlike the control group, the language delay worsened significantly at 6 months postimplantation, and was maintained at 12 months postimplantation, while intelligence development was also significantly delayed at 12 months postimplantation. Finally, in the group of children with both ASD and ID, delays in language and intelligence development worsened at 6 months and again at 12 months postimplantation. Additionally, for this group of children, there were significant delays in emotional development (as measured by interpersonal emotional ability), and social behavior development (as measured by development in living ability) (Motegi et al., 2019). Both of which maintained or worsened postoperatively.

The researchers in this study note that the children with ASD and ID who received CIs had significant developmental delays in emotional and social behavior development, in addition to the language and intelligence delays seen in both the control group, and those children with CIs and ID alone. They go on to state that their results are a, "reflection of the features of ASD, which is characterized by impaired social interaction, atypical communication, and repetitive, restrictive behaviors" (Motegi et al., 2019). Ultimately, the researchers conclude that CIs do not inherently improve those features intrinsic to ASD itself, and that the addition of ID does not alter these results.

In 2014, Fitzpatrick et al. completed a retrospective chart review on patients with hearing

loss and ASD. Seventeen children were selected for examination. The intended purposes of this research were to describe audiologic presentation, and examine the recommendations given for these children, including amplification. Such factors as age at diagnosis of hearing loss and ASD, etiology, type and severity of hearing loss, other disabilities, audiologic results and recommendations, type of amplification used, recommendations made at time of diagnosis, and consistency in use of amplification were examined (Fitzpatrick et al., 2014). These researchers found that, "ASD was generally diagnosed much later than hearing loss at a median age of 51.5 months" and that, "only one child [...] received a diagnosis of ASD (age 48 months) prior to hearing loss identification at 79.8 months" (Fitzpatrick et al., 2014). In terms of etiology of hearing loss, it was highly variable, which agrees with the research discussed previously in this review. Also in agreement with the aforementioned research is the fact that these researchers found these children to have highly variable capabilities in terms of behavioral audiologic testing. Therefore, many different test techniques were used to arrive at a diagnosis (e.g. VRA, CPA, and ABR).

Of the 17 children included in this study, six went on to receive CIs. Five of these six had additional documented disorders or developmental delays, including CMV (cytomegalovirus), ANSD (Auditory Neuropathy Spectrum Disorder), and global developmental delays. Of those children, the data available during the research period showed that four used their implant(s) consistently, one used their implant(s) inconsistently, and the last child did not use their implant(s) at all. After being fit with amplification - CI or hearing aids - four of these 17 children still required to be tested via objective measures, as their behavioral capabilities did not improve to a point where more traditional behavioral audiometry could be utilized. In terms of behavioral benefit to cochlear implantation for this population, the researchers state, "Our

findings are consistent with a study investigating the benefits of cochlear implants for seven children with ASD that described positive behavioral changes including improvement in such areas as reaction to music and sounds, vocalizations, and eye contact" (Donaldson et al, 2004, as cited by Fitzpatrick et al., 2014). They also cite a need for diagnostic testing created specifically for this population.

Yamazaki et al. (2011) compared the outcomes of 11 children with CMV and CIs to that of 14 children with CIs and no other comorbidities. Hearing loss is a common symptom of congenital CMV, and is typically late-onset and progressive in nature (Yamazaki et al., 2011). It also can lead to neurodevelopmental disorders, including intellectual disabilities, and pervasive developmental disorder; the latter being classified as ASD under the DSM-V (Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition). In the group of children with CMV, two also had the comorbid diagnosis of ASD, with additional motor dysfunction. These children were evaluated with regard to hearing threshold, open- and closed-set speech discrimination tasks, and language development using the Kyoto Scale of Psychological Development (K-test). According to Yamazaki et al., "In the K-test, three categories of a child's development including Postural-Motor (P-M; fine and gross motor functions), Cognitive-Adaptive (C-A; non-verbal reasoning or visuospatial perceptions), and Language-Social (L-S; interpersonal relationships, socializations, and verbal abilities) are assessed separately" (2011). Only C-A and L-S are addressed in this paper. Scores are in the form of developmental quotients (DQ), with developmental delay defined as DQ below 80.

Of these four outcome measures, hearing threshold was the most comparable between groups. Yamazaki et al. state that, "In autistic [children with CMV and CIs, who showed the most devastating results, all three abilities other than hearing threshold were severely affected"

(2011). In the open- and closed-set discrimination tasks, the two children with ASD and CMV performed significantly poorer than those children with CIs and no comorbidities, as did two children with CMV and mental retardation. The other children with CMV and additional disabilities had comparable scores to that of the control group. On the K-test, the L-S DQs of both groups improved significantly postimplantation, while the increase in L-S DQ was significantly smaller in the group with CMV and additional disabilities than the control group. However, for the children with ASD, "postoperative L-S DQs did not improve and never exceeded 40, indicating that their language development remained severely retarded even after the implantation" (Yamazaki et al., 2011). Similar results were also obtained with regard to C-A DQs. The researchers also reported that L-S DQ and C-A DQ were correlated, with the exception of the children with ASD, in that their developmental delay in the language-social domain was more severe than in the cognitive-adaptive domain pre- and postimplantation.

In all, postimplantation, improvements in language development were seen in many of the children with CMV and additional disabilities, as well as the typically developing children, with the former progressing at a slower rate. While the developmental ages of two children with CMV and ASD increased post CI, their language and social development showed little improvement, remaining severely impaired. Both of these children relied on sign language, while other children in this group, whose L-S DQs were higher, went on to develop oral language (Yamazaki et al., 2011). Therefore, the researchers remark that children with CMV and ASD, who have hearing loss, should undergo further testing preoperatively. Yamazaki et al. state that evaluating, "cognitive development and autistic tendency [...] might be useful for determination of an appropriate goal for language rehabilitation [and] choice of communication mode, which is critical to achieve best language outcomes and improve both children's and

family's satisfaction with CIs" (2011).

The addition of comorbidities can also have an effect on communication modality. In the study by Robertson (2013) described previously five children had an intellectual disability, two with additional comorbidities (i.e. cerebral palsy, and visual impairment). Four out of these five relied on AAC in the form of PECS, while the fifth child used a combination of speech and sign language. None used solely oral language (Robertson, 2013). In the aforementioned study by Nasralla et al. (2018), all four children with ASD had other comorbidities. Two children had ADHD, one utilizing sign language and the other behavioral means of communication. The other two had cognitive impairments. One child, whose cognitive impairment was severe, also relied on behavioral communication, while the other, with a mild cognitive impairment, developed oral language (Nasralla et al., 2018).

In sum, many of the studies done on children with ASD and CIs include participants in which other diagnoses are present. In keeping with the results detailed in the previous sections, these children had variable outcomes in device compliance, and communication modality. Of particular note, in those studies where children with ASD, CIs, and another disability were compared to children with ASD and only the additional comorbidity, the children with ASD performed worse on language outcomes, had additional behavioral problems, or made the least amount of progress over time (Motegi et al., 2018, Yamazaki et al., 2011). ASD, in the presence of another disability, or on its own, appears to drastically impact most CI outcome measures used in the collective research detailed herein.

Literature Reviews

Presently, there are few literature reviews that have been done using research on children with CIs and ASD. As these reviews discuss overarching results, and do not always specifically

specify methods, they are discussed in a separate section in this paper. Three literature reviews are outlined herein. In a review by Cejas et al. (2015), the outcomes and benefits of cochlear implantation in children with additional disabilities were analyzed. This research notes that initially, cochlear implantation was not being done for children who had comorbid disabilities alongside hearing loss. Whereas more recently, as cited in Cejas at al. (2015), Johnson and Wiley found that, "30%–40% of children currently receiving CIs have a comorbid disorder, with mixed evidence of their potential benefits." This review included children with ASD, developmental delay, CHARGE syndrome, cerebral palsy, learning disorders, Usher syndrome, Waardenburg syndrome, and attention deficit hyperactivity disorder. Once analyzed separately, comparisons were made across the research pertaining to each group.

In terms of the research compiled on children with ASD and CIs, this systematic review included eight articles on the topic. Of the studies analyzed, various objective and subjective measures were utilized to analyze the outcomes of this population. Such objective data included speech and language outcomes, communication style used by children in this population, retention of the devices, as well as changes in behaviors. Examples of objective measures used are the Early Speech Perception Test (ESP), and the Multisyllabic Lexical Neighborhood Test (MLNT). The ESP is a test of speech perception for children who are profoundly deaf, beginning at age three. It is used to measure the benefit with regard to speech perception provided by the child's hearing aid(s) or cochlear implant(s) (Supporting Success, n.d.). The MLNT is an open-set word list used for children who wear CIs, as a means of measuring spoken word recognition. This measure is based on the Neighborhood Activation Model (NAM), which organizes words into, "'similarity neighborhoods' based on their frequency of occurrence in the language and the number of phonemically similar words, or neighbors, within the lexical

neighborhood" (Auditec, 2015). Examples of the subjective measures used for these studies are parent/caregiver report.

Research contained in this review includes an article by Fitzpatrick et al., (2014), which suggests that there is a, "Higher prevalence of hearing loss in children with ASD than in [the] general population" (as cited by Cejas et al., 2015). This retrospective chart review also notes that children with ASD had highly variable results on audiologic measures, indicating there is no specific symptom array that accompanies the disorder. The remaining research focuses on the outcomes of these children postimplantation.

As cited in Cejas et al. (2015), a study by Cruz et al., previously detailed in this review, contained 15 children who received CIs and were diagnosed with ASD. The study examined receptive and expressive language skills and behavior problems before implantation to three years postimplantation. The results bore that children ASD had lower language scores, and similar rates of externalizing behavior problems pre-implantation when compared to their peers with CIs but without ASD. However, postimplantation the results showed that children with ASD improved in receptive and expressive language at half the rate of their typically developing counterparts with CIs. Additionally, and most importantly for the purposes of this review, "Increases in externalizing behaviors were also observed over the 3-year period in the ASD group compared with the non-ASD group" (Cruz et al., 2012, as cited by Cejas et al., 2015).

Another article discussed in this review was one containing 10 children with ASD and CIs, in which use of their processors, and selected mode of communication are examined. The results of this study revealed that six of the ten participants used their CIs consistently, one child used spoken language as his mode of communication, 3 used speech and sign language in tandem, while the remaining six were non-verbal (Robertson, 2013, as cited by Cejas et al.,

2015). These results align with the aforementioned study in that language progress is variable and prolonged in children with ASD and CIs. Another study focusing on speech and language progress was one containing 14 children with CIs and ASD, and 10 hearing aid controls (Meinzen-Derr et al., 2014, as cited by Cejas et al., 2015). This study revealed that four of the children with ASD utilized oral communication skills postimplantation, while eight used alternative augmentative communication (AAC) strategies, such as Picture Exchange Communication System (PECS). The overarching result of this study was that improvements in speech and language skills were seen in this population as measured by the Clinical Evaluation of Language Functions (CELF).

Further research revealed improvements in various behaviors postimplantation. A retrospective chart review on six children with ASD and CIs reports, "positive changes postimplantation [...] in responsiveness to sound, speech perception, interest in music, vocalizations, and eye contact" (Donaldson et al., 2004, as cited by Cejas et al., 2015). They go on to report increases in patients' use of sign language, and their responsiveness to requests postimplantation. The parental report section of this research reveals that five of the six parents would recommend cochlear implantation to other families of children with ASD and hearing loss. Another studying lending itself to these results is one by Hayman and Franck, comprised of two case studies, one of which led to the child with ASD and CIs experiencing increased social/emotional responsiveness, as well as nonverbal communication (as cited by Cejas et al., 2015). And lastly, a systematic review by Beers, also discussed in the current systematic review, found that, "Children with ASD and CIs displayed improved social communication, behavioral and environmental awareness, increased vocalizations, eye contact, and reaction to music after implantation" (as cited by Cejas et al., 2015). Thus, the research encompassed in this systematic

review suggests improvements in outward behaviors in children with ASD and hearing loss post cochlear implantation.

The aforementioned systematic review and the research therein reveal that children with ASD and CIs experience improvements in speech and language measures, as well as some behavioral responses to sounds and/or social interaction. When comparing the outcomes of children with CIs and other comorbidities, "these studies showed that children's speech intelligibility and auditory perception improved following implantation, despite a slower rate of growth" (Cejas et al., 2015), as compared to their typically developing peers with CIs. More specifically, this body of research led to the conclusion that children whose comorbid diagnoses entailed a lesser cognitive impairment, or no cognitive impairment (e.g. ADHD) had better outcomes than those whose comorbidities entail more deficits in intellectual functioning, including ASD.

In a systematic review by Cosetti and Waltzman (2012), the numerous variables affecting CI performance were examined. Such factors included changes in CI technology, patient physiology (e.g. age at implantation), medical or surgical issues, education/rehabilitation environment, social factors (e.g. socioeconomic status), and the presence of multiple disabilities. When examining comorbidities, including ASD, the research contained in this systematic review bore similar results to those discussed above. Across studies, researchers found that given the spectrum of symptoms and capabilities seen in persons with multiple disabilities and deafness who use CIs, performance postimplantation cannot be easily generalized or predicted. However, "evaluation of postimplantation outcomes, including speech perception, receptive and expressive language development, social interaction, environmental awareness, and quality of life suggest that cochlear implantation in patients with multiple disabilities can lead to substantial benefit"

when discussing multiple disabilities broadly (Cosetti & Waltzman, 2012). Similar again to the results of the aforementioned research is the fact that speech perception scores remained lower and took longer to progress in this group than for typically developing children with CIs. A study by Berrettini et al. discussed in this review included 23 children with CIs and additional diagnoses (e.g. cerebral palsy, mental retardation, epilepsy, attention deficit and hyperactivity), including ASD (as cited by Cosetti & Waltzman, 2012). Benefits were seen via improved speech perception scores. Additionally, a parental questionnaire revealed further subjective benefit. "100% of parents reported increased awareness of environmental sounds, 96% indicated improved interaction with peers, and 74% noted improvement in speaking skills. In addition, the percentage of patients using oral language increased from 28% (before surgery) to 67% after surgery" (Berrettini et al., 2008, as cited by Cosetti and Waltzman, 2012). Research by Meinzen-Derr et al. (2010, as cited by Cosetti & Waltzman, 2012) found that children with CIs and developmental disabilities, such as ASD, had significantly lower rates of receptive and expressive language when compared to age- and cognition-matched controls. These researchers go on to state that benefits were seen through quality of life changes and improvements in environmental awareness and social interaction.

This systematic review also contains research pertaining specifically to children with ASD and CIs, and states that the outcomes are highly variable in nature, and some studies have found minimal speech and language benefit postimplantation (Donaldson et al., 2004, as cited by Cosetti & Waltzman, 2012). A study by Daneshi and Hassanzadeh (2007), as seen in the research detailed above, highlights improved quality-of-life, including "increased responsiveness to sound, improved eye contact, greater attempt at vocalizations, and increased environmental awareness" (as cited by Cosetti and Waltzman, 2012). However, they, too, note minimal

improvement in communication skills. Of importance is the emphasis laid on counseling parents of children with ASD regarding realistic expectations for developing language postoperatively.

The final systematic review to be discussed is one previously mentioned in this paper. In this systematic review by Beers et al, (2014), the composite results revealed that, "Positive benefits have been reported following the cochlear implantation in children with ASD, which include improvements in social communication, [behavior] and environmental awareness, as well as increased vocalizations, eye contact and reaction to music" (2014). However, as cited in Beers et al., (2014), Daneshi et al. found that, "children with ASD showed limited development in auditory perception following implantation compared to deaf patients with other secondary disabilities." The success of children with ASD postimplantation is unclear largely due to the limited research in this area. Preoperative counseling is necessary in order to set realistic expectations with families or caregivers (Beers et al., 2014).

The literature reviews included in this paper made similar conclusions to one another, and to multiple other studies included herein, with regard to the outcomes of CI use in this population. All three bodies of research revealed that children with ASD can make gains in language, both receptive and expressive, postimplantation. However, given the nature of ASD, the rate at which these improvements are made can be significantly slower than those of typically developing children with CIs. Additionally, many of the studies encapsulated in these reviews reveal subjective benefits perceived by parents or caregivers of these children. Such subjective benefits as increased social interaction with family members, and responsiveness to requests. In general, despite slow or limited language progress, the subjective behavioral changes show benefit for implantation of children with ASD.

Discussion

Behavioral Benefits of CI for Children with ASD

A host of studies, with varying results, have been discussed in this paper. The population, research type, outcome measures, and main conclusions of each study are summarized in Table 2. While considerations regarding cochlear implantation in children with ASD have been raised, research has shown benefits from CI use in this population. Benefits have been seen particularly in those studies using subjective measures, including parental surveys or reports. According to the study by Beers et al., "Positive benefits have been reported following the cochlear implantation in children with ASD, which include improvements in social communication, [behavior] and environmental awareness, as well as increased vocalizations, eye contact and reaction to music" (2014). Cruz et al. (2012) state in their study that children with additional disabilities can make significant gains in receptive and expressive language with a CI, although their growth is likely to not be as rapid as typically developing deaf children. Other subjective benefits have been reported in reaction to sound, name, and requests. Parents across studies have reported that children are better able to adapt to their routines, interact with therapy or school activities, and display reduced anxiety when using their CI. These subjective benefits led to better parent satisfaction with CIs, and in studies where asked, they reported that they would recommend implantation to families under similar circumstances (Eshraghi et al., 2015; Nasralla et al., 2018). Additionally, device compliance was found to be generally good in this population, even in the presence of hyperacusis in some cases (Rodriguez-Valero et al., 2016). Perhaps most importantly, as reported previously, parents reported that their children were happier when they were wearing their CIs (Nasralla et a., 2018).

Behavioral Considerations for Cochlear Implantation in Children with ASD

While the subjective measures provided insight to the many benefits perceived by parents and families of children with ASD and CIs, the objective measures used brought light to the outcomes of these children as compared to typically developing children, as well as those with other comorbidities. Consistently, across outcome measures, and across studies, children with ASD and CIs performed far poorer, progressed at a much slower rate, or progressed to a much lesser extent than their typically developing peers. The most comprehensive objective measure of behavior, the CBCL, as used by Cruz et al. (2012) showed an increase of externalizing behaviors postoperatively, while typically developing children showed the opposite. Johnson et al. (2008) also found no change in those behaviors intrinsic to ASD with use of a CI. Eye contact was repeatedly reported to be the least improved behavioral outcome (Eshraghi et al., 2015; Lachowska et al., 2018). These findings are in agreement with the hypotheses posed previously. Also in keeping with those hypotheses are the findings that children with ASD performed more poorly than those children with CIs and other comorbidities. Moreover, children with ASD and another comorbidity displayed the same pattern of results as children with ASD and CIs alone. Those children with ASD and other comorbidities performed still poorer on objective measures, even when compared to children with CIs and the same additional comorbidity, as seen in the study by Motegi et al. (2018). The addition of another comorbidity did not change the general trend of results seen in children with ASD.

With regard to the mode of communication used by this group, results were once again highly variable. The majority of children whose preferred mode of communication was reported utilized more than one mode of communication, and some children did not develop any spoken language. The composite researchers reported poorer oral language outcomes, and device

compliance for children in whom the hallmark symptoms of ASD were more severe. While this does not preclude them from implantation, it necessitates the need for counseling families about prospective outcomes and setting realistic expectations for mode of communication postoperatively. As in children with ASD alone, spoken language is not guaranteed.

Table 2 Main Conclusions of Studies

Author	Date	Research Design	Sample Size	Outcome Measures	Conclusions
Beers et al.	2014	Literature Review	N/A	N/A	Improvements in social communication, behavior environmental awareness; increased vocalizations, eye contact, reaction to music postimplantation
Cejas et al.	2015	Literature Review	N/A	N/A	Improvements in speech and language measures, some behavioral responses to sounds, social interaction postimplantation
Cosetti & Waltzman	2012	Literature Review	N/A	N/A	Significant variability in outcomes; some research shows minimal speech and language benefit; Quality of life improvements generally seen: increased responsiveness to sound, eye contact, vocalization attempts, environmental awareness
Cruz et al.	2012	Cohort Study	188 children with CIs; 8 with ASD	RDLS; CBCL	Children with ASD made slowest progress in language; behavior problems increased significantly
Cupples et al.	2013	Cohort Study	119 children with HL children; 9 with ASD	PLS-4; PPVT-4 DEAP; CDI; PEACH; SIR	Children with ASD were less likely to participate fully in formal testing; lowest average scores on receptive/expressive language scales; poorer caregiver ratings on the PEACH and CDI
Eshraghi et al.	2015	Cohort Study; Survey	30 children with CIs; 15 with ASD	ESP; MLNT; PBK; Parental Survey	Improvements in speech, less than controls; on questionnaire top three improvements: name recognition, response to verbal requests, and enjoyment of music, least improved: eye contact
Fitzpatrick et al.	2014	Cohort Study	17 children with HL and ASD; 6 with CIs	Age at diagnosis of HL and ASD; etiology/ severity/type of HL; other disabilities; audiologic results/ recommendations; type/consistency in use of amplification;	Highly variable audiologic presentation, highly variable etiology of HL, variable performance on testing, variable use of amplification; behavioral benefits of CIs: reaction to music and sounds, vocalizations, and eye contact
Johnson et al.	2008	2 Case Studies	2 children with CIs; 1 with ASD	RDLS; CDI; Joint Attention Task; Symbolic Play Task	Expressive and receptive language scores increased; improvements in symbolic play; behaviors typical of ASD increased postimplantation: lack of joint attention
Lachowska et al.	2018	Cohort Study; Survey	6 children with CIs ASD	Reaction to music and sound, Ling's six sounds test, onomatopoeic word test, reaction to spoken child's name, response to requests, parental survey	Receptive/expressive language development very delayed; most children showed reduced anxiety with CI; no change in behavior, no increased hyperactivity with daily use in 2 cases; benefits in personal interaction with family members

Meinzen-Derr et al.	2014	Cohort Study	24 children with HL and ASD; 14 with CIs	ADOS; GARS; RGDS; CELF; PLS-4; VABS;	Outcomes for children with ASD and cochlear implants are as variable as for children with ASD and normal hearing; severity of autism contributes to communication outcomes
Mikic et al.	2016	Cohort Study	14 children with CIs, 4 with ASD	CAP; SIR	Significantly slower progress on CAP seen in ASD group, little progress seen on SIR
Motegi et al.	2018	Cohort Study	13 children with CIs; 4 with ASD & ID, 4 with ID, 5 with only CI	Enjoji Scale	Significant delays in language, intelligence, emotional and social behavior development that persist with time seen in the ASD and ID group
Nasralla et al.	2018	Survey	14 families of children with CIs; 4 with ASD	Parental Survey; CAP, CL	Parents disagree with ASD diagnosis/are frustrated by lack of progress/desired results; some behavioral improvements noted: adaptation to routines; 3/4 children scored in category 1 on the CAP and CL postoperatively, the 4th scored in category 5 and 4, respectively.
Robertson	2013	Cohort Study; Case Study	10 children with CIs and ASD	CAP; SIR	4 children developed some functional spoken language, the rest use non-verbal methods and are happy with their implants; 2 became non-users; Outcomes' related to the severity of ASD; Patient in case study made slow and limited improvement on SIR and CAP
Rodriguez Valero et al.	2016	Cohort Study; Survey	22 children with CIs and ASD	Device Compliance	16/22 (72.7%) use their cochlear implants; 13/22 (59%) had periods of intermittent CI use; 6 non-users; Researchers deemed acceptable compliance with CIs in children with ASD, but results vary with degree of symptoms
Yamazaki et al.	2011	Cohort Study	25 children with CIs; 11 with CMV; 2 with ASD and CMV	Hearing threshold; closed-set infant word discrimination; open-set monosyllabic word discrimination; K-test	Children with CMV& ASD had lowest scores/made least progress on all measures; could not understand spoken words/ sentences; mothers felt CIs effective: behaviors/family interactions improved; developmental age increased; language development remained severely delayed

Studies categorized by author, year of publication, type of research, population, outcome measures, and main conclusions.
Abbreviations: RDLS, Reynell Developmental Language Scales; CBCL, Childhood Behavior Checklist; PLS-4, Preschool Language Scale Fourth Edition; PPVT-4, Peabody Picture Vocabulary Test Fourth Edition; DEAP, Diagnostic Evaluation of Articulation and Phonology; CDI, Child Development Inventory; PEACH, Parent Evaluation of Aural/Oral Performance of Children; SIR, Speech Intelligibility Rating; ESP, Early Speech Perception; MLNT, Multisyllabic Lexical Neighborhood Test; PBK, Phonetically Balanced Kindergarten Test; HL, Hearing Loss; ADOS, Autism Diagnostic Observation Schedule; GARS, Gilliam Autism Rating Scale; RGDS, Revised Gesell Developmental Schedules; CELF, Clinical Evaluation of Language Fundamentals; VABS, Vineland Adaptive Behavior Scales; CAP, Categories of Auditory Performance; ID, Intellectual Disability; The Enjoji Scale, The Enjoji Scale of Infant Analytical Development; CL, Categories of Language; CMV, Cytomegalovirus; K-test, Kyoto Scale of Psychological Development

Limitations

The body of research was limited in more ways than one. Many of the authors of these studies report the need for outcome measures appropriate for children with ASD and sensory impairments. As mentioned previously in this paper, the ADOS, which is the gold standard for diagnosing children with ASD, is not approved for children with hearing loss. Therefore, it is not surprising that methods of diagnosis are variable, and there were many different measures used across these studies. There is also the challenge presented by children with ASD being more likely unable to complete standard diagnostic testing, in terms of audiometry and other areas of development. While parental report is highly valuable, and audiometry can be done without the child's participation if need be, behavior and its progression post CIs cannot be measured with a child under sedation.

Another limitation of this literature review is the limited number of participants, even across studies. Some studies had sample sizes of children with ASD numbering two or four. The total number of children with ASD and hearing loss whose results were analyzed in this paper was 126. Of these, between 96 and 105 used cochlear implants. A greater body of research, using the same or similar outcome measures, needs to be completed to confirm or expand upon the results detailed above.

Suggestions for Future Research and Resources

Future research should be done in order to adapt outcome measures used for children with ASD, such as the ADOS, for children with hearing loss and other sensory impairments. Additional research needs to be done in children with ASD and hearing loss, who present with auditory, or other sensory sensitivities. Specifically, how being implanted affects the extent and manifestations of hyperacusis and the device compliance. While two children in this literature

review had documented hyperacusis and consistently used their devices, that results cannot be generalized to the larger population of children with those symptoms.

Educational materials for parents must be made in order to relay information regarding the overlap in symptoms of hearing loss and ASD, and the likelihood of having both. Additionally, the increasingly younger age of implantation does not always leave time for any and all developmental disorders to make themselves known. Therefore, when they do appear postoperatively, parents may be dissatisfied with the more limited progress their child may make with their device. A cochlear implant can only address the auditory deficits that a child has but will not ensure that a child will be otherwise typically developing. Appropriate counseling must be used to ensure that parents have realistic expectations preoperatively, and also when or if a subsequent diagnosis is made. Parents should be made aware that a CI can help their child to hear, and can have very positive effects on that child's development, but it will not remove the diagnosis of ASD.

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