Outcomes in Children with Additional Disabilities Following Cochlear Implantation: A Systematic Review

Rebecca M. Tuchman

The Graduate Center, City University of New York

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OUTCOMES IN CHILDREN WITH ADDITIONAL DISABILITIES FOLLOWING COCHLEAR IMPLANTATION: A SYSTEMATIC REVIEW

by

REBECCA TUCHMAN

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

2019
Outcomes in Children with Additional Disabilities Following Cochlear Implantation: A Systematic Review

by

Rebecca Tuchman

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

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ABSTRACT

Outcomes in Children with Additional Disabilities Following Cochlear Implantation: A Systematic Review

by

Rebecca Tuchman

Advisor: Adrienne Rubinstein, Ph.D.

Background: Thirty percent of children with hearing loss have an additional disability. These children may be difficult to test according to standard audiologic behavioral test protocols. Additionally, progress within this population may present differently than in children with no additional disability. Currently, no evidence-based protocol exists for assessing cochlear implant benefit and outcomes in this population.

Objective: The purpose of this investigation is to perform a systematic review on the outcomes of cochlear implantation in children with additional disabilities. Specifically, this study focused on areas of function assessed, outcome measures used, and evidence of benefit observed.

Methods: A comprehensive search was conducted utilizing the databases MEDLINE/PubMed, OneFile, ProQuest Nursing & Allied Health Source. The keywords used to identify relevant studies included "pediatric," "special needs," "developmental disabilities," "functional assessment," "outcomes," "benefit." The keywords "cochlear implantation" or "cochlear implant" were present
throughout all searches.

**Results:** Included in this study were 24 articles. The results revealed that despite wide variability among the studies, some benefit was observed in children with cochlear implantation and additional disabilities in the areas of auditory skills and speech perception, receptive and expressive language, and adaptive behaviors.

**Discussion:** Many challenges arose when studying this population. Limited experimental control as well as wide variability in disability type were major issues noted throughout this review. However, overall children with cochlear implantation and additional disabilities showed some improvement in all areas, although they still did not perform as well as children with cochlear implantation and no additional disabilities, or normally hearing peers matched according to age and cognitive abilities.

**Conclusions:** Research in this area is challenging due to the limitations involved in the ability to produce randomized, double blind studies to determine value of cochlear implantation in this population. Cognitive ability is a strong, but not the only, predictor of performance. Although on average the lower the cognitive ability, the lower the post implant performance, there was much variability among participants, adding to the challenge of deciding whether to implant such a child. There is some evidence to support the implantation of children with additional disabilities, however, more research is recommended involving more multicenter collaborations to increase the participant pool and to isolate individual disabilities to establish performance. Research should continue to explore use of alternative assessments such as quality of life measures.
ACKNOWLEDGMENTS

I would like to express my deepest gratitude to my capstone advisor, Dr. Adrienne Rubinstein, for her endless patience, generous guidance and unending support she provided throughout the completion of this project and my graduate study. Her dedication and advice were invaluable. I must also thank the CUNY audiology faculty as well as my clinical supervisors for sharing their knowledge throughout my time in the Au.D. program.

Finally, and most importantly, I would like to thank my family for their immense love, caring, understanding, and support all these years. To my parents, who throughout my life never doubted that I could achieve whatever I set my mind to and instilled within me my core values. To my in-laws, without whom the combination of raising children and completing my degree would have been impossible. To my husband Daniel for acting as my rock and ultimate supporter, and for his endless patience and understanding as I completed this degree. And to my sons, Yisrael and Shmuel, for giving me a reason to smile every single day, no matter what.
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INTRODUCTION

Cochlear implantation has been widely accepted in the field of pediatric audiology as a successful and often recommended method of intervention for children with severe to profound hearing loss. When conventional hearing aids do not provide sufficient benefit to facilitate the development of speech and language, cochlear implants (CI) can provide children with the auditory access they need. Additionally, CIs are associated with better academic achievement, improved quality of life, and better employment status (Vinceti et al., 2014).

Although cochlear implantation is a relatively minor surgery, its consequences are critical and irreversible. CI surgery typically destroys the residual hearing that a patient may have (although advances have been made in recent years in preserving some residual hearing (Miranda et al., 2014)). Additionally, the recipient hears through electrical stimulation, a completely different experience than the typical electro-acoustic mechanism that the body naturally uses. This differs from the way hearing aids digitally amplify sound, utilizing the remaining hearing in a patient. The CI recipient must essentially “relearn” how to hear, making sense of the electrical signals sent from the implant and translating that into sound and more importantly, speech. This often involves intensive aural rehabilitation therapy, even for those individuals who lose their hearing post-lingually.

Due to the serious and permanent implications of cochlear implantation, candidacy guidelines have been developed to limit the surgery for those who meet appropriate criteria. Candidacy guidelines stipulate the degree of hearing loss required, as well as require the potential recipient to demonstrate insufficient benefit after undergoing a trial with appropriately fit acoustic amplification and aural habilitation. Additionally, candidates must undergo a medical workup, complete with imaging of their cochlea and cochlear nerve to rule out any
contraindications for cochlear implantation. Common contraindications for cochlear implantation are outlined by Vinceti et al. (2014) and include the following: absence of cochlear development; aplasia or absence of the acoustic nerve; deafness due to lesions of the central auditory pathway; medical conditions or developmental delays that would severely limit participation in aural habilitation; massive cochlear ossification that prevents electrode insertion. Families must also demonstrate the ability and commitment necessary for attending ensuing audiologic and aural rehabilitation appointments and implementing therapeutic recommendations in the home. Failure to satisfy any of these criteria may preclude candidates from ultimately undergoing cochlear implantation.

Additional disabilities (AD) present in approximately 30-40% of children with sensorineural hearing loss (Berrettini et al., 2008). AD in children with sensorineural hearing loss can include developmental delay, visual and spatial disorders, cerebral palsy, autism, attention deficit and hyperactivity disorder, physical and gross motor delays, and speech and language delays. They can be due to a variety of causes including, but not limited to, hypoxia at birth, CMV, meningitis, epilepsy, brain trauma, prenatal complications, or syndromes related to hearing loss, such as fetal alcohol syndrome (FAS), CHARGE syndrome, Downs Syndrome, or Fragile X Syndrome (Corrales et al., 2013). In the past, the presence of ADs had been a contraindication for cochlear implantation, due at least in part to limited potential for speech and language and other medical or educational concerns. With advances in available technology, the increasing benefits of implantation have been demonstrated, and as candidacy guidelines expand more children with ADs are receiving CIs (Zaidman-Zaiti et al., 2015).

Traditionally, benefit of cochlear implantation in children is assessed using a variety of accepted standardized measures, including aided narrow-band noise thresholds, correct
identification of the Ling-6 sounds, as well as performance on open and closed set speech testing, such as the WIPI, NU-Chips, ESP or HINT-C. Additionally, speech and language development is viewed as the gold standard for assessing benefit from cochlear implantation (Hayward et al., 2013). CI recipients are followed closely by their implant centers, and various assessment measures are given repeatedly and monitored over time to track the patients’ progress.

These standardized measures may not be fitting for children with AD. Aided thresholds may be inappropriate or impossible to obtain from children who are difficult to test due to their cognitive or physical delays. Additionally, the development of speech and language may be an unrealistic standard when applied to children with AD. Many of these children begin with a limited potential for speech and language development and may never develop normal speech and language. For example, many children with cerebral palsy or autism rely on non-verbal, augmentative communication methods, even in the absence of hearing loss. Therefore, a child with AD who receives a cochlear implant may still not develop language due to underlying cognitive or motor potential. When an additional disability is combined with hearing impairment, it may be unclear whether present speech and language delays should be attributed to lack of benefit from implantation, or to non-auditory delays related to the AD. The difficulty in assessing benefit in these children is compounded by the fact that many of them are specifically excluded from studies on cochlear implantation due to their disabilities, as pointed out by Meinzen-Derr et al. (2010)

To contrast this, greater potential for success with cochlear implantation in this population may be linked to the phenomenon known as the "pseudo handicap effect." It refers to a case in which one disability, combined with a second disability, interact to increase the overall
disability an individual to a greater degree than expected. For example, hearing loss may act to exacerbate the disabilities attributable to a child's other disabilities. If the hearing loss can be addressed, a reversal may take place, and the overall effect of AD may be mitigated (Corrales et al., 2013). When applied to cochlear implantation, the pseudo handicap effect may imply that early and effective implantation can address the hearing handicap in the child as well as grant them greater potential to deal with their other challenges than had the hearing loss gone unaddressed.

In recent years, more studies have emerged that focused on finding appropriate measures for assessing benefit in the AD population. The focus has shifted somewhat to qualitative benefits, such as behavioral improvements, activities of daily living, quality of life, social functioning, and parental or familial perception of benefit. Measures that focus more on behavioral outcomes may be more appropriate for use in individuals with disabilities, especially in populations where speech and language development are heavily impaired by the presence of a disability. However, no comprehensive guidelines have been established to facilitate appropriate recommendations and rehabilitation plans. Unlike speech and language development or auditory skills, a comprehensive test battery of this nature has never been widely accepted and implemented for use in these cases.

The lack of evidence-based practice for these cases is concerning. Considering the high comorbidity of hearing loss and developmental disabilities, it is important to be able to provide evidence of the benefit these children are receiving through cochlear implantation and to reach some consensus to better inform best practice among audiologists and other professionals, as well as parents, involved in the child's education and development. As Cruz et al. (2013) notes, there are no current candidacy guidelines specifically for children with additional disabilities.
Evidence-based practice in this area must address a multitude of considerations, such as how early implantation should be implemented, the type of educational environment best suited for this child, and the realistic expectations for such a child. Additionally, as caregivers are likely dealing with a plethora of other medical or developmental issues, concrete evidence can inform the level of priority of this intervention in relation to additional concerns.

Cochlear implantation guidelines recommend that the surgery be performed as early as possible. The current FDA guidelines approve implantation for children as early as 12 months of age, but some centers will perform implantation as early as six months. Studies have demonstrated that children who receive implantation before two years of age are projected to perform better than those who receive it after, due to the nature of neural pathway formation in the auditory cortex. Although some disabilities are apparent at birth, many, such as developmental delay, autism, or language disorders, are not diagnosed until the child is slightly older. In these cases, a child may have already undergone cochlear implantation. As a result, the presence of an additional disability no longer factors into the question of candidacy, however it can play a role in determining the most appropriate rehabilitation plan for the child, based on outcomes data gathered from other children with cochlear implantation and AD. Additionally, it can serve as an important counseling tool when discussing what progress they may realistically expect from their child as they deal with the implications of a new diagnosis.

The goal of the present review is to examine studies that explore the benefits of cochlear implantations in children with comorbid developmental disabilities. Specifically, this review focused on how studies answered the following three questions:

1. What areas of daily function have been looked at to assess benefit from CIs in the special needs population?
2. What outcome measures have been used to assess benefit of CIs in the special needs population?

3. Has there been measurable benefit seen as a result of cochlear implantation in this population, and in what areas?
METHODS

A systematic search of the literature was performed in June 2018 using the search engine OneSearch to identify relevant studies. OneSearch combs multiple databases including: MEDLINE/PubMed, OneFile, ProQuest Nursing & Allied Health Source. The search was limited to peer reviewed articles available in English and published between 2005 to the present.

The following key words were applied in various combinations: "pediatric," "special needs," "developmental disabilities," "functional assessment," "outcomes," "benefit." The keywords of "cochlear implantation" or "cochlear implant" were always present throughout the literature search.

The following criteria were applied to exclude articles, at first through an initial review of titles and abstract, and subsequently through an in depth review of the articles: articles relating to deaf culture or decision making prior to implantation, articles relating to cochlear implantation in populations other than special-needs children, articles relating to neuroplasticity post-implantation, articles examining other independent variables (such as bilateral versus unilateral implantation or simultaneous versus sequential implantation) and articles with no full text version available. Due to the nature of this review and its focus on how benefit has been defined in various studies as well as examining the different outcome measures used, levels of evidence were not included as an inclusion or exclusion criteria.

Articles that fit the above criteria were then read in completion and included based on their relevance to the research questions.
RESULTS

Retrieval process

Thirty five articles were initially identified using the keywords and databases previously described. Figure 1 is a flow chart that summarizes the search process for the identification of articles used in this study.

Figure 1: Article retrieval process

*Reasons for exclusion: Full text not available, articles that did not have outcome measures, previous systematic reviews, articles about cochlear implantation not related to additional disabilities.

Before discussing the results, it should be noted that the studies included in this review varied widely in terms of both study design and participant types.

Study Design

The majority of studies included in this review were retrospective. This is due to the limited number of subjects available for studies of this nature. The studies included a wide variety of within subject, between subject and mixed designs, in addition to one study featuring two case studies. Thirteen studies included a comparison group. Of these studies, 11 included a
comparison group with hearing loss and no additional disability, whereas two studies compared the experimental group with a group of cognitive and age matched children without hearing loss. This is significant because children with AD may have different “cognitive potential,” than typically developing children, thus matching for this variable can help to avoid its potential confounding effects. In other cases, however, differences in cognitive ability was the independent variable, i.e., poorer cognitive ability was the additional disability. Eleven studies included repeated measures pre- versus post- cochlear implantation. In addition, 7 studies examined the correlations between two measures and whether specific factors could act as correlational predictors of outcomes.

**Participant Characteristics**

Table 1 summarizes characteristics of the participants in both the experimental and comparison groups in the studies assessed. There was further variety in the experimental group regarding the definition of “additional disabilities”. The studies focused on a large variety of disabilities, with some including a broad range within a single study, and others focusing on specific disability. For example, Amisalari et al. (2010) and Eshraghi et al. (2015) focused exclusively on motor developmental delays, and autism spectrum disorder, respectively. Holt et al. (2005) restricted his experimental group only to those with mild cognitive delay. On the other hand, Beer et al. (2012) listed eleven different special needs conditions, and Rafferty et al. (2013) defined his population under the broad term of “complex needs” and included children with such disabilities as developmental delay, autism spectrum disorder, and language disorders. There was also a variety in the details provided in operational definitions. For example, 3 studies included participants with “additional disabilities,” but did not specify how they defined it in
their inclusion criteria. On the other hand, Meinzen-Derr et al. (2011) and Meinzen-Derr et al. (2013) reported that they required all participants to be evaluated and diagnosed by a developmental pediatrician before being admitted into the study. Figure 2 details the 26 disability types studied and in how many studies each type was included. Note that 15 of the disabilities are only included in a single study. The most frequent disability studied was autism spectrum disorder. In addition, different degrees of a disability were also compared (e.g. Berrettini et al., 2006, Wakill et al., 2014)

Figure 2: Number of studies including a particular disability
Table 1: Participant Characteristics

<table>
<thead>
<tr>
<th>Authors (year)</th>
<th>Disabilities/Delays Included</th>
<th>Number of Subjects *</th>
<th>Comparison Group</th>
<th>Age at Implantation (Mean)</th>
<th>Length of Implantation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amisalari et al (2010)</td>
<td>Motor Developmental Delay (Excluding Severe motor delays)</td>
<td>n=28</td>
<td>n=234 CI without motor delay, same surgeon, electrode array</td>
<td>3.54 (Experimental) 4.22 (Control)</td>
<td>Followed for one year after implantation</td>
</tr>
<tr>
<td>Beer et al (2012)</td>
<td>Autism Spectrum Disorder Goldenhaar syndrome Cerebral Palsey Prematurity BOR syndrome Blindness, Motor delay, CHARGE Syndrome, VATER Syndrome, Leigh's disease Robinow Syndrome</td>
<td>n=23</td>
<td>n=23 CI with no AD</td>
<td>24 months (Experimental) 22 months (control)</td>
<td>6 months or 12 months (matched in control)</td>
</tr>
<tr>
<td>Authors (year)</td>
<td>Disabilities/Delays Included</td>
<td>Number of Subjects *</td>
<td>Comparison Group</td>
<td>Age at Implantation (Mean)</td>
<td>Length of Implantation</td>
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<tr>
<td>Berrenttini et al (2006)</td>
<td>Attention deficit hyperactivity disorder, Pervasive Developmental Disorder/autism Learning disability Cerebral palsy</td>
<td>n=23</td>
<td></td>
<td>Range 2.3-17 years</td>
<td>mean= 2.5 years</td>
</tr>
<tr>
<td>Birman et al (2012)</td>
<td>Developmental Disabilities</td>
<td>n=23</td>
<td>n=23 CI with no AD</td>
<td>Range 0-16 years</td>
<td>12 months</td>
</tr>
<tr>
<td>Cruz et al (2012)</td>
<td>Attention deficit hyperactivity disorder, Pervasive Developmental Disorder/autism Learning disability Cerebral palsy</td>
<td>n=31</td>
<td>n= 157 CI with no AD</td>
<td>28.55 months (experimental)</td>
<td>Assessed annually for 3 years post implantation</td>
</tr>
<tr>
<td>Edwards et al (2006)</td>
<td>Developmental Delay</td>
<td>11</td>
<td>21 CI with no AD</td>
<td>2.4 years</td>
<td>2 years</td>
</tr>
<tr>
<td>Authors (year)</td>
<td>Disabilities/Delays Included</td>
<td>Number of Subjects</td>
<td>Comparison Group</td>
<td>Age at Implantation (Mean)</td>
<td>Length of Implantation</td>
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<tr>
<td>Eshraghi et al (2015)</td>
<td>Autism Spectrum Disorder</td>
<td>n=15</td>
<td>n=15 with CI with no AD, matched by age of implantation, and years of usage</td>
<td>3 years</td>
<td>8.3 years</td>
</tr>
<tr>
<td>Hiraumi et al (2013)</td>
<td>Developmental Delay (diagnosed preoperatively)</td>
<td>n=11</td>
<td>24 with CI with no AD, age and pre-op hearing level, cause of deafness, implant device and coding strategy not significantly different</td>
<td>37 months (group specific N/A)</td>
<td>2 years</td>
</tr>
<tr>
<td>Holt et al (2005)</td>
<td>Mild cognitive delay</td>
<td>n= 19</td>
<td>n=50 CI without cognitive delay</td>
<td>38 months (experimental) 29 months (control)</td>
<td>1 year</td>
</tr>
<tr>
<td>Authors (year)</td>
<td>Disabilities/Delays Included</td>
<td>Number of Subjects *</td>
<td>Comparison Group</td>
<td>Age at Implantation (Mean)</td>
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<tr>
<td>Johnson et al (2008)</td>
<td>Global Developmental Delay and Autism Spectrum Disorder</td>
<td>2 case studies</td>
<td>Subject1: 3 years, Subject 2: 4 years</td>
<td></td>
<td>12 month</td>
</tr>
<tr>
<td>Lanson et al (2007)</td>
<td>CHARGE Syndrome</td>
<td>n=10</td>
<td>2.75</td>
<td></td>
<td>3 months to 7 years</td>
</tr>
<tr>
<td>Meinzen-Derr et al (2010)</td>
<td>Additional Disability</td>
<td>n=20</td>
<td></td>
<td>23.9 months</td>
<td>12 months or longer</td>
</tr>
<tr>
<td>Meinzen-Derr et al (2011)</td>
<td>Cognitive and motor Delays</td>
<td>n=15</td>
<td>n=15 children with normal hearing, pair-matched re: age and cognitive abilities, No significant difference in maternal education, family income</td>
<td>21 months (group specific not available; participants matched within 12 months)</td>
<td>10 months to 68 months</td>
</tr>
<tr>
<td>Meinzen-Derr et al (2013)</td>
<td>Developmental Disability</td>
<td>n=23</td>
<td>n=7 no hearing loss, with developmental/motor disability matched</td>
<td>30 months</td>
<td>28.5 months</td>
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<tr>
<td>Authors (year)</td>
<td>Disabilities/Delays Included</td>
<td>Number of Subjects</td>
<td>Comparison Group</td>
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<tr>
<td>Nikolopoulos et al (2008)</td>
<td>8 etiologies including Cognitive delay, Behavioral problems, Pervasive Developmental Disorder/autism Learning disability</td>
<td>n=67</td>
<td>n=108 CI with no AD, with same implant as exp. group</td>
<td>3.4 years (experimental) 3.3 years (control)</td>
<td>5 years</td>
</tr>
<tr>
<td>Rafferty et al (2013)</td>
<td>Mostly developmental delay with learning disabilities</td>
<td>19</td>
<td>n=230</td>
<td>3.5 years (experimental) 4.1 years (control)</td>
<td>12 months</td>
</tr>
<tr>
<td>Robertson (2013)</td>
<td>Autism Spectrum Disorder</td>
<td>10</td>
<td>Range 2-13 years old. (all under the age of 4 years except two).</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>Authors (year)</td>
<td>Disabilities/Delays Included</td>
<td>Number of Subjects *</td>
<td>Comparison Group</td>
<td>Age at Implantation (Mean)</td>
<td>Length of Implantation</td>
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<tr>
<td>Trimble et al (2008)</td>
<td>Multiply Disabled including developmental delay, CP, language disorder, deaf/blind and learning disability</td>
<td>58</td>
<td></td>
<td>Under 18 years</td>
<td>At least 6 months</td>
</tr>
<tr>
<td>Wakil et al (2014)</td>
<td>Cognitive delay</td>
<td>13 with severe delay</td>
<td>8 with mild delay</td>
<td>median=4.3 years</td>
<td>11.3 years,</td>
</tr>
<tr>
<td>Wiley et al (2005)</td>
<td>Visual impairment Mild motor disabilities Cognitive disabilities Specific learning disabilities Behavioral disorders Language disorders</td>
<td>15 families (16 children)</td>
<td>N/A</td>
<td>mean= 4 years range= 13 months-14 years</td>
<td>mean= 3 years Range=0.5-8 years</td>
</tr>
<tr>
<td>Authors (year)</td>
<td>Disabilities/Delays Included</td>
<td>Number of Subjects *</td>
<td>Comparison Group</td>
<td>Age at Implantation (Mean)</td>
<td>Length of Implantation</td>
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<td></td>
<td>(experimental) 16</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(control)</td>
<td></td>
</tr>
<tr>
<td>Wiley et al (2012)</td>
<td>Cognitive and motor delays</td>
<td>6</td>
<td>N/A</td>
<td>13.8-134 months</td>
<td>12 months</td>
</tr>
<tr>
<td>Zaidman-Zait et al (2015)</td>
<td>Additional Disabilities</td>
<td>23 families</td>
<td>N/A</td>
<td>40.34 months</td>
<td>9 months or more</td>
</tr>
<tr>
<td>Zaidman-Zait et al (2017)</td>
<td>Autism Spectrum Disorder, Cerebral Palsy, Developmental delay</td>
<td>n=43 parents of children with additional disability</td>
<td>n= 49 parents of CI recipients (TD)</td>
<td>23.8 months (experimental) 27.1 months (control)</td>
<td>6 months or more</td>
</tr>
</tbody>
</table>
Areas of Function Studied to Assess Benefit

The first research question addressed the areas of function that have been studied to determine if pediatric patients were receiving benefit from their CIs. Five different areas of function were identified upon reviewing the studies: a. Auditory Skills and Speech Perception, b. Receptive and Expressive Language, c. Adaptive Behaviors d. Mode of Communication e. Other (Implant Use and Health Related Quality of Life, which were each examined by one study).

Regarding auditory skills and speech perception, measures included a variety of different skills and behaviors. For example, Rafferty (2013) used repeated measures to examine how these children performed pre-implantation and 12 months post implantation activation with measures that included Categories of Auditory Perception (CAP), Meaningful Auditory Information Scale (MAIS), Listening Progress Score (LiP), and Meaningful Use of Speech Scale (MUSS). The MAIS was used in many studies. It is designed to measure behaviors such as spontaneous responses and alerting to sound, vocalization behavior and deriving meaning from sound.

To assess higher level communication skills, Receptive and Expressive Language function was included in some studies. For example, five studies used the Preschool Language Scale (PLS-4) to assess receptive and expressive language. However in several of these studies, a floor effect for this scale was noted. To address this issue, the studies calculated language quotients by dividing the age-equivalent score by the child’s chronological age at the time of testing and multiplying by 100 (Beer et al., 2012, Meinzen-Derr et al., 2011).

Adaptive Behaviors refer to any outcome measure that looked at social interaction, cooperation, attention, or skills of daily living (e.g. self-care). Cruz et al. (2012) used the Child Behavior Checklist (CBCL), a validated behavior checklist that assesses the intensity of
various behaviors to assess children pre and post implantation. The CBCL assesses children on two scales; Internalizing and Externalizing Behavior Problems, in addition to a Sleep Problem scale. The Internalizing scale consists of four subscales: Emotional Reactivity, Anxious/Depressed, Somatic Complaints, and Withdrawn. The Externalizing scale contains two subscales: Attention Problems and Aggressive Behavior. Two studies utilized the Pediatric Evaluation of Disability (PEDI) which include content in the domains of self-care, mobility, toileting, and social cognition. Johnson et al. (2008) assessed joint attention and symbolic play by analyzing play in two case studies.

To a lesser extent, mode of communication, implant use, and health-related quality of life were also considered. Mode of communication refers to whether the child developed spoken language or used a form of nonverbal communication post implantation. Robertson (2013) focused exclusively on the autism spectrum disorder population and Berrettini et al. (2006) studied communication mode in two ways, by examining the actual form of communication and also by determining if this correlated with other measures. Other measures included implant use and health related quality of life, which examines the quality of life of patients as it relates to their physical and emotional wellbeing (Robertson, 2013 and Zaidman-Zait et al., 2017, respectively).

Many of the studies looked at more than one area of function and fell within the first three categories. Of the 24 studies included in the review, 15 studied receptive and expressive language, 13 included speech and auditory perception measures, and 9 investigated adaptive behaviors. Mode of communication was assessed in 2 studies. The remaining categories (implant use and health related quality of life) were each included in only one study.
Test Measures

For the 24 studies included, no fewer than 24 different outcome measures were used to measure benefit from cochlear implantation in the pediatric special needs population across the 5 areas assessed. As noted in the previous section, many studies focused on more than one area of benefit, and several studies used several different measures for one area of benefit. Table 2 lists the number of studies using each outcome as well as the area of benefit to which they corresponded, whereas Table 3 identifies the outcomes measure(s) used in each study, and the areas of function assessed as described by the investigators.

It is important to note that two of the outcome measures listed represent more than one specific measure. For example, if a study used a questionnaire or interview, they were recorded as having used the “parent questionnaire/survey/interview” outcome measure, even though all five studies used five different formats. It is also important to note that parent questionnaire refers to non-standardized formats developed by the researchers for the purposes of their studies. Validated and standardized interviews or questionnaires, such as the PEDI, were included as separate measures. The other outcome representing more than one measure is labeled as “Other speech tests” referring to speech perception tests in languages other than English and speech perception test results which were recovered through a chart review and were not specified. Therefore, the actual total number of outcome measures used is higher than 24.

The large number of outcome measures is at least in part reflective of the absence of a gold standard for this population. Additionally, it is likely reflective of the difficulty of having participants with AD participate in standard behavioral procedures (such as speech recognition testing), 14 of the outcome measures were inventories or checklists filled out by the parents, (IT-MAIS, PEDI, etc.), while only 10 were assessments filled out by the researcher or clinician.
<table>
<thead>
<tr>
<th>Outcome Measure</th>
<th>Area of Function</th>
<th>Number of studies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preschool Language Scale (PLS-4)</td>
<td>Receptive and Expressive Language</td>
<td>5</td>
</tr>
<tr>
<td>Parental Questionnaire/Survey/Interview</td>
<td>Receptive and Expressive Language</td>
<td>5</td>
</tr>
<tr>
<td>Other speech perception tests (Italian, Japanese, Unspecified)</td>
<td>Auditory Skills/Speech Perception</td>
<td>4</td>
</tr>
<tr>
<td>Categories of Auditory Performance (CAP)</td>
<td>Auditory Skills/Speech Perception</td>
<td>3</td>
</tr>
<tr>
<td>Speech Intelligibility Ratings (SIR)</td>
<td>Receptive and Expressive Language</td>
<td>3</td>
</tr>
<tr>
<td>Infant Toddler Meaningful Auditory Integration Scale (IT-MAIS)</td>
<td>Auditory Skills/Speech Perception</td>
<td>4</td>
</tr>
<tr>
<td>Reynell Developmental Language Scale</td>
<td>Receptive and Expressive Language</td>
<td>2</td>
</tr>
<tr>
<td>Pediatric Evaluation of Disability Inventory</td>
<td>Adaptive Behavior</td>
<td>2</td>
</tr>
<tr>
<td>Vineland Adaptive Behavior Scale</td>
<td>Adaptive Behavior</td>
<td>1</td>
</tr>
<tr>
<td>Child behavior checklist</td>
<td>Adaptive Behavior</td>
<td>1</td>
</tr>
<tr>
<td>Early Speech Perception test</td>
<td>Auditory Skills/Speech Perception</td>
<td>2</td>
</tr>
<tr>
<td>Multisyllabic Lexical Neiborhood Test (MLNT)</td>
<td>Auditory Skills/Speech Perception</td>
<td>1</td>
</tr>
<tr>
<td>Phonetically Balance Kindergarten Test (PBK)</td>
<td>Auditory Skills/Speech Perception</td>
<td>2</td>
</tr>
<tr>
<td>MacArthur-Bates Communicative Developmental Inventories</td>
<td>Receptive and Expressive Language</td>
<td>1</td>
</tr>
</tbody>
</table>
Of the 12 outcome measures that assessed auditory skills and speech perception, 6 were questionnaires and/or checklists, such as the IT-MAIS or MUSS, and three were audiological test batteries, such as the MLNT and PBK. One study also used other/unspecified measures, such as results from chart reviews and speech testing in a language other than English. Nikolopoulos et al. (2008) supported their use of the SIR, which was also used in two additional studies by stating that it is an objective test measure that can be readily applied to large groups of young deaf children over time regardless of participant age and abilities, and additionally has been
shown to be reliable between observers.

Six measures looked at receptive and expressive language. Four of those were clinician-administered assessments, such as the PLS-4, and 2 of those were questionnaires or checklists filled out by the parent. Administration by the clinician is important, especially in those studies that compared groups, as it increases the consistency and validity of the scoring and may reduce bias. Additionally, parents may be more susceptible to the Rosenthal effect, and perceive improvement or change where it does not exist. The PLS-4 was the most commonly used test measure as well. According to Meinzen Derr et al. (2011), this assessment tool:

targets skills that are important precursors for language development (e.g. attention to speakers, appropriate object play), comprehension of basic vocabulary, concepts, grammatical markers, and the ability to understand complex sentences and make comparisons and inferences. Vocal development and social communication, naming common objects, the use of concepts that describe objects, express quantity, prepositions, grammatical markers, sentence structures, and examines pre-literary skills (i.e. phonological awareness tasks, ability to tell a short story in sequence). (p. 795)

Five measures were utilized in the analysis of adaptive behavior. All but one were parent-based interviews, checklist, or questionnaires. The only one performed by a clinician was the analysis of joint attention and symbolic play for two case studies (Johnson et al., 2008). Although having the parent fill out the interview may invite different biases, as well as inconsistent scoring across subjects, for this specific area it is almost necessary, since only the child’s caregiver can satisfactorily answer questions about behavioral tendencies and give accurate comparisons of the child before and after implantation. Areas covered in these questionnaires included communication, daily living skills, and socialization, joint attention,
behavior, cognition, functional mobility, among other areas.

Chart reviews were used to assess frequency of implant use and main mode of communication in Robertson (2013). In order to measure the health-related quality of life in the pediatric CI population, Zaidman-Zait et al, (2017) had parents of children both with and without AD fill out the KINDL survey.
Table 3: Outcome measures used in each study

<table>
<thead>
<tr>
<th>Study</th>
<th>Areas of Functionality Assessed</th>
<th>Outcomes Measures</th>
</tr>
</thead>
</table>
2. Auditory Perception                                           | 1) Speech Intelligibility Ratings (SIR)                                           |
|                        |                                                                      | 2) Categories of Auditory Perception (CAP)                                        |
2) Receptive and Expressive Language  
3) Adaptive Behavior (motor, social, daily living and communication skills) | 1) Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS) (Clinician-Parent Interview)  
2) Preschool Language Scale (PLS-4)  
3) Vineland Adaptive Behavior Scale (2nd edition) (Semi structured interview) |
2. Overall Communication Behavior                                  | 1) Four Italian speech perception tests                                           |
|                        |                                                                      | 2. Questionnaire for parents (CAP)                                                |
2. Communication Skills  
3. Behavior,  
4. Interaction with others| 1. Early Speech Perception (ESP), Multisyllabic Lexical Neighborhood Test (MLNT), or Phonetically Balanced Kindergarten (PBK)  
2-4. Parental Survey Consonant–vowel (CV) Syllables and Short Sentences (Japanese) |
| Holt et al, (2005)     | 1. Auditory Skill  
2. Word and Sentence Recognition  
3. Word Recognition  
4. Receptive and Expressive Language | 1. IT MAIS  
2. GAEL-P, Mr. Potato Head Task  
3. PSI  
4. Reynell Developmental Language Scale |
<table>
<thead>
<tr>
<th>Study</th>
<th>Areas of Functionality Assessed</th>
<th>Outcomes Measures</th>
</tr>
</thead>
</table>
2. Joint Attention  
and the MacArthur-Bates Communicative Development Inventories (CDI)  
2-3. Analyzed using clinical coding schemes |
| **Lanson et al, (2007)**      | Auditory Benefit                                                   | IT-MAIS                                                                            |
| Meinzen-Derr et al, (2011)    | Language Skills (as compared to cognitively matched hearing peers)  | Pediatric Evaluation of Disability inventory?                                     |
2. Receptive and Expressive Language                               | PLS-4                                                                              |
2. Meaningful Auditory Information Scale (MAIS)  
3. Listening Progress Score (LiP),  
4. Meaningful Use of Speech Scale (MUSS) | |
| Robertson (2013)              | 1. Implant Use  
2. Mode of communication                                         | Case Reviews                                                                      |
ITMAIS, GASP, PBK HINT             |
| Wakil et al (2014)            | Auditory Abilities                                                  | Parent interview (developed by researchers)                                       |
| Wiley et al (2005)            | Perceived Benefits as reported by parents                          | Auditory Skills Checklist (ASC)                                                   |
4. Preschool Language Scales (PLS-4) |
2. Mobility  
3. Social Function  
4. Expressive and Receptive Language | Survey and Interview                                                             |


| | 2. Parental Perception of benefit | 2. Open ended questions and perceived benefits scale. |


Benefit from Cochlear Implantation

The third research question addressed whether the studies found benefit from cochlear implantation in the pediatric AD population. In order to efficiently evaluate the findings, the studies were divided into the areas of function they included, and each area of function was assessed individually. Table 4 summarizes the statistical measures used in each study, in addition to the results obtained.
Table 4: Statistical Measures and Results

<table>
<thead>
<tr>
<th>Study</th>
<th>Statistical Measures</th>
<th>Inferential Statistical Findings</th>
<th>Other findings and Conclusions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amisalari et al (2010)</td>
<td>Paired and unpaired T-tests used to compare effects of cochlear implantation on the speech perception measures, which looked at group difference.</td>
<td>Significant improvement from pre to post-implantation in speech perception tests. No significant difference in degree of improvement between groups, after they controlled for cognitive skill.</td>
<td>Children with motor delay can benefit from cochlear implantation.</td>
</tr>
<tr>
<td>Beer et al, (2012)</td>
<td>A two-way ANOVA with one repeated measure was used to compare pre- to post-CI scores in functional auditory skills between the AD and No-AD groups. Language and adaptive behaviors were analyzed descriptively due to the small sample size.</td>
<td>Both groups improved significantly on IT-MAIS, however, the control group made more progress. Younger chronological age and earlier age at implantation were significantly correlated with larger gain in IT-MAIS scores for the control group only.</td>
<td>Most children with ADs improved in auditory skills. Progress seen in children in the AD group for receptive but not expressive language but their language quotients were lower than the No-AD group. Children with ADs made progress in daily living skills and socialization skills. Children with ADs who did not make progress in language, did show progress in adaptive behavior.</td>
</tr>
<tr>
<td><strong>Berrenttini et al, (2006)</strong></td>
<td>Spearman function to analyze correlation between: 1. postimplant speech perception categories and post-implant communication modes (oral vs. gestures and perceived benefits scores 2. etiology and outcomes, in terms of speech perception skills, perceived benefits, and communication mode improvement. 3. degree of cognitive delay and the same outcomes.</td>
<td>Significant correlation found between post-implant speech perception categories and post-implant communication modes. Both post implant speech perception categories and communication modes were significantly correlated with post-implant perceived benefit scores. No significant correlation between etiology and outcomes. Specifically, there was no significant correlation between etiology and post-CI speech perception category, post-implant communication mode post-implant perceived benefits score. No significant correlation between degree of cognitive delay and outcomes.</td>
<td>Varied results, however overall improvement seen. Degree of impairment could not predict outcomes.</td>
</tr>
<tr>
<td><strong>Birman et al, (2012)</strong></td>
<td>Mann Whitney and (x^2) test to distinguish between the experimental and comparison group on speech perception scores.</td>
<td>Experimental group performed significantly worse than control groups in CAP score categories and median CAP scores.</td>
<td>Children with developmental delay do not perform as well as children without developmental delay on tests of auditory perception.</td>
</tr>
<tr>
<td><strong>Cruz et al, (2012)</strong></td>
<td>Multilevel modeling techniques were used to predict oral language and behavior problems using time and group as predictors.</td>
<td>Children in the AD group had a slower rate of change compared to children in the comparison group for receptive and expressive language.</td>
<td>Comparison group’s externalizing behavior problems decreased over time while these problems increased in the AD group. However, findings support the use of cochlear implant given the improvement in expressive and receptive language.</td>
</tr>
<tr>
<td>Authors</td>
<td>Methodology</td>
<td>Findings</td>
<td>Comments</td>
</tr>
<tr>
<td>-------------------------</td>
<td>------------------------------------------------------------------------------</td>
<td>-------------------------------------------------------------------------------------------</td>
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</tr>
<tr>
<td>Edwards et al, (2006)</td>
<td>T tests used to compare groups step-wise multiple linear regression, ordinal regression.</td>
<td>Significant differences in performance between experimental and comparison group. Degree of developmental delay was the most significant predictor of SIR and Speech perception.</td>
<td>Descriptively, large differences were found between mildly and severely developmentally delayed in speech intelligibility and speech perception.</td>
</tr>
<tr>
<td>Eshraghi et al, (2015)</td>
<td>Sign tests used to evaluate whether the ordinal outcome had improved in a significant amount of pairs in each of the groups after implantation. Fisher’s exact tests used to assess whether the proportions of significant improvement differed between control and experimental groups.</td>
<td>The perception and expression scores significantly improved after implantation in a significant amount of pairs in both groups. More significant improvement is observed in the control group than in ASD group in both speech perception and speech expression.</td>
<td>Parents noted the most improvement in awareness of environment, potential for education, communication, and family interaction for children with ASD/PPD.</td>
</tr>
<tr>
<td>Hiraumi et al, (2013)</td>
<td>T test for between group measures. Correlation analysis was conducted between the pre-operative developmental quotient for the cognitive–adaptive area (DQCA) and speech perception scores using a Pearson’s correlation coefficient and the partial correlation coefficient.</td>
<td>Speech perception scores in the non-delayed group were significantly higher than those in the delayed group. After controlling for age at the time of implantation and average pre-op aided hearing level, the relationship between the DQCA scores and the speech perception scores was weak.</td>
<td>Large intersubject variability, thus difficult to assign a specific cognitive score for candidacy.</td>
</tr>
<tr>
<td>Holt et al, (2005)</td>
<td>Two-way ANOVA) with one repeated measure.</td>
<td>Significant improvement was found for both groups across all measures. Significant group difference was present only on tests of productive language skills and receptive vocabulary skills.</td>
<td>Deaf children with cognitive impairment benefit from cochlear implantation.</td>
</tr>
<tr>
<td>Johnson et al, (2008)</td>
<td>Case Studies</td>
<td>n/a</td>
<td>Both participants showed progress for expressive and receptive language, and demonstrated joint attention</td>
</tr>
</tbody>
</table>
Lanson et al, (2007) | No statistical analysis performed | n/a | Limited degree of auditory benefit, but parental reports of improve connectivity to environment and increased ability to develop communication skills. 

Meinzen-Derr et al, (2010) | Linear regression models used to analyze independent factors related to language skills while adjusting for potential confounders. | Children with cochlear implants and AD had significantly lower receptive and expressive language quotients compared to their hearing peers of the same age and nonverbal cognitive abilities. | Children with additional disabilities appeared to have significant delays in their language development that were disproportionate to their nonverbal “cognitive potential.” 

Meinzen-Derr et al, (2011) | Categorical variables: McNemar’s Chi-square. Continuous variables : WilcoxonSignRank test. Correlations between language quotients and nonverbal cognitive quotients: Spearman correlation coefficient. | Language was highly correlated with nonverbal cognitive abilities; however CI group still scored significantly lower on receptive and expressive language scores than hearing controls (at least 20 points). Language quotients of CI group were not matching their cognitive quotients. | Children with additional disabilities appeared to have significant delays in their language development that were disproportionate to their nonverbal “cognitive potential.”
<table>
<thead>
<tr>
<th>Study</th>
<th>Methodology</th>
<th>Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Meinzen-Derr et al, (2013)</td>
<td>Linear multiple regression models use to analyze differences in scores between group across the three PEDI domains.</td>
<td>Children with C and AD had significantly lower social functioning stand median scores than cognitively and age matched peers. The significance disappeared after controlling for nonverbal cognitive abilities and language level. Among children with CI and AD, age at implant and duration with device were not associated with PEDI scores. Less functional independence and poorer social functioning in the AD group, but similar results in self care and mobility.</td>
</tr>
<tr>
<td>Nikolopoulos et al, (2008)</td>
<td>Chi-square test and rank correlation coefficients</td>
<td>Significant difference in speech quality between controls and experimental group. The number of additional disorders had the strongest correlation with speech quality. A majority with AD developed connected intelligible speech 5 years after implantation but a proportion did not develop any speech. A majority of children with additional disorders developed connected intelligible speech, although a significant proportion did not develop any speech at all. The number and type of additional disabilities was the most strongly correlated factor.</td>
</tr>
<tr>
<td>Rafferty et al, (2013)</td>
<td>Means and ranges reported in comparison to control group</td>
<td>No significance measured Improvement in all outcome measures though less in those with AD than those without AD. Development of oral language may not be a realistic goal.</td>
</tr>
<tr>
<td>Robertson (2013)</td>
<td>No statistical analysis (Review of outcomes, examining processor use and mode of communication).</td>
<td>n/a Outcomes were highly variable, found to be related to severity of autism.</td>
</tr>
<tr>
<td>Study</td>
<td>Methodology</td>
<td>Findings</td>
</tr>
<tr>
<td>-------------------------------</td>
<td>-----------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Trimble et al, (2008)</td>
<td>T-test, chi square, logistical regression.</td>
<td>The functional disability score significantly predicted high or low speech perception scores and had excellent discrimination ability. The GPA (a functional disability assessment) score was not significantly associated with speech perception scores and demonstrated no ability to discriminate postimplant speech perception scores. Newly developed functional disability assessment found to be predictive of post-implant outcomes and should be used as adjunct to traditional measures.</td>
</tr>
<tr>
<td>Wakil et al (2014)</td>
<td>Data presented primarily descriptively (due to small sample size) using means, medians, or proportions as appropriate. For the group with severe delay, pre- and post-implant results were tested for statistical significance with a Student’s t-test. Differences between communication mode for the two groups (severe versus mild-moderate delay) were tested for significance using ( \chi^2 ) analysis.</td>
<td>Comparison of pre- and post-implant results in the severely delayed group at the most recent IT-MAIS testing for the group showed a significant improvement in score. Communication mode (oral versus non-oral) as a function of category of delay was significantly different between the two groups of children. Children with severe or complex developmental delay demonstrated relatively limited progress in auditory abilities despite several years of cochlear implant use. Children with mild to moderate developmental delay demonstrated skills consistent with results reported for implanted children without additional disabilities.</td>
</tr>
<tr>
<td>Wiley et al (2005)</td>
<td>Descriptive results reported as percentages (parent interview).</td>
<td>Majority of participants wore devices consistently, made some communication progress, had greater awareness of environmental sounds, and were more interested and attentive to their environments.</td>
</tr>
<tr>
<td>Author(s)</td>
<td>Methodology</td>
<td>Findings</td>
</tr>
<tr>
<td>-----------</td>
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</tr>
<tr>
<td>Wiley et al, (2008)</td>
<td>Repeated measures analysis, test of parallelism to compare group differences.</td>
<td>Children with AD had the same rate of auditory skills progress as children without AD, however children with AD appeared to have a lower baseline skills set. Children with a developmental quotient of less than 80 had half the rate of progress of children with a DQ of at least 80.</td>
</tr>
<tr>
<td>Wiley et al, (2012)</td>
<td>Friedman's test to analyze changes in score over time; repeated measures to assess correlation between nonverbal cognition and language levels.</td>
<td>The scaled score for the Mobility domain of the PEDI improved significantly. Median language ages increased significantly over the first year, but not fast enough to impact language quotients significantly. Nonverbal cognition was also significantly correlated with change in receptive language age, but not with language quotient.</td>
</tr>
<tr>
<td><strong>Zaidman-Zait et al. (2017)</strong></td>
<td>Profile analysis: includes tests of flatness, parallelism, a level test, and pair-wise comparison).</td>
<td>Significant differences for parallelism, level test, flatness test. Bonferroni revealed significant differences between three out of six subscales (all p’s ≤ 0.001). Children in the CI-DD group had lower scores on the self-esteem, friend, and school HRQoL subscales than Children with CI-TD.</td>
</tr>
</tbody>
</table>
Auditory Skills and Speech Perception

Overall, in the 12 studies that assessed auditory skills and speech perception, the majority found general benefit from CIs. Beer et al. (2012), for example, found that on IT-MAIS assessments administered pre and post implantation, all children showed significant improvement, however the comparison group made more progress than children with AD. They also found that in addition to presence of disabilities, earlier age at implantation was significantly correlated with better outcomes.

Birman et al. (2012) drew exclusively negative conclusions regarding benefit, specifically that children with AD had significantly lower CAP scores than typically developing children with cochlear implantation. However this was a between-subjects design study. Judging benefit by such a metric reflects unrealistic expectations, considering that experimental group may have started at a lower overall baseline than the control group, as noted in Rafferty et al. (2013). A more valid model for measuring benefit was executed in the mixed designs studies, such as Eshragi et al. (2015) and Rafferty et al. (2013). Rafferty et al. (2013) compared IT-MAIS scores between children with a variety of complex needs and typically developing children. Measures were performed pre-implantation and 12-months post implantation. The IT-MAIS was filled out by parents and local teachers of the deaf at both intervals. Although only means were compared and no statistical measures were done, the study showed a distinctive difference between the typically developing group and the group with complex needs, demonstrating that the group with complex needs was not performing at the same level as the typically developing group post implantation. However, the study also noted that the two groups had different baselines, and if each were measured according to their baseline, the rate of progress of each
group would be similar. Eshragi et al. (2015) used statistical measures to demonstrate that significant improvement was observed for standardized speech perception measures in both typically developing children and children with autism spectrum disorder. Additionally, this study used a Fisher’s exact test to compare the rate of improvement between the subjects. A statistically significant difference was found in the rate of improvement between the ASD group and the typically developing group. While two studies agree that the degree of improvement was not equal between groups, Eshragi et al. (2015) found differences in the rate of improvement as well. These findings agree with those of Birman et al. (2012) that children with AD may not perform at the same skill level as children without AD, but progress seen on repeated measures lead them to draw a different conclusion regarding benefit.

Amisalari et al. (2011) found no differences between the experimental and comparison group, however, the experimental group in this study consisted exclusively of children with motor delays and excluded children with more severe delays or cognitive delays. This suggests that outcomes may be related to severity of disability, and is supported by Wiley et al. (2008), who first compared outcome measures based on presence of disability, then by developmental quotient (>80 vs <80). Although both comparisons produced significant differences in outcomes, the low developmental quotient in the AD group appeared to have skewed the analysis when comparing by presence of disability. Both the experimental and comparison groups made progress, however, the progress was significantly influenced by subjects’ developmental quotient. Length of implantation proved to be an important variable in one study. Although Holt et al. (2005) did not find significance in auditory skills between children with AD and a comparison group, they did find an interaction between the presence of a cognitive impairment and length of device use for auditory-only sentence recognition. According to the results,
children with mild cognitive delays needed more experience using their implants to achieve similar sentence recognition scores to those of typically developing children.

Receptive and Expressive Language

Findings for benefit in the functional category of receptive and expressive language were similar to those of auditory skills and speech perception. Overall benefit was observed, but at a slower rate and degree when compared to comparison groups of children with hearing loss and no AD. Holt et al. (2005) completed a retrospective study on language measures taken at 6 month intervals post implantation and compared children with mild cognitive delay to a control group of children with no cognitive impairment, finding that the differences between the two groups was most apparent when measuring higher level language skills. Meinzen-Derr et al. (2010) found that early measures of nonverbal cognition were predictive of language outcomes on the PLS-4 in implanted children more than the actual diagnosis. However, using the same measure, Meinzen-Derr et al. (2011) found that when compared with children matched according to both age and cognitive abilities, children with AD and hearing loss still underperformed, suggesting that their language delays were not solely due to their developmental delays and they were not reaching their full language potential with cochlear implantation. Cruz et al. (2012) performed a three-year longitudinal study and found that children with comorbid disabilities in addition to hearing loss did not have lower baseline language levels compared with typically developing children. However, post hoc analysis revealed that there was in fact a statistical difference among these two groups when children with a diagnosis of ADHD were excluded from the experimental group. Additionally, this study found that only the group with pervasive developmental disorder (PDD) had a slower rate of growth on
annual receptive and expressive language measures over a three-year period, progressing at about half the rate of typically developing children and children with other AD.

**Adaptive Behavior**

Although studies that examined adaptive behavior noted either a positive or negative change post implantation, maturation may have been a strong confounding factor in these observations. Cruz et al. (2012) found increased behavioral problems in the children with disabilities post implantation, in contrast to the improved behavior observed in the typically developing children after undergoing cochlear implantation. However, again the issue might be with comparing two inherently different groups. For instance, consider how the reported behavioral issues in the first group would compare with a control group of cognitively matched peers without hearing loss. Cruz also pointed out that the study began with an average participant age of 28 months, which is before many of these diagnoses were even made. The typical problem behaviors within this population may naturally emerge as the study followed them longitudinally introducing a maturation effect. In a study focusing exclusively on children with autism, Eshragi et al. (2015) found that significant improvements were seen in family interaction, potential for education, communication, and awareness of environment. This study also noted that their results clashed with those of Cruz et al. (2012), and suggested that the prior findings may be attributable to the fact that the children in their study were young when they received their implantations and may not have exhibited any behavior problems at that point in time. The authors pointed out that the behavioral problems may be more a reflection of maturation and emerging developmental delays that an affect of cochlear implantation.

Meinzen-Derr et al. (2013) compared PEDI scores between age and cognitively matched
peers with and without hearing loss. The PEDI is a standard test used in assessing disabilities. The study found that children in the hearing loss group had significantly lower scores, indicating greater impairment than those without hearing loss. However, when controlling for receptive language abilities, the difference was no longer significant, suggesting residual handicap due to the hearing loss despite cochlear implantation. Overall, parents reported improvement in their children’s adaptive behavior after cochlear implantation.

Through parent interviews conducted by Wiley et al. (2008) parents reported improvement in environmental awareness, communication, and engagement with their surroundings. Family interaction was also an area in which improvement was noted (Zaidman-Zait, 2015). However, it is important to note that although Zaidman Zait et al. (2015) reported that families reported increased awareness of environmental sound, enjoyment of music, and overall benefit in family interactions, their outcome measures involved mailed out surveys that families needed to mail back. Parents who were driven to respond to these surveys may only reflect the more motivated ones out of a much larger population, resulting in biased results as a result of differential subject selection. Similarly, Wiley et al. (2005) noted that although families noted several benefits related to their child’s CI, respondents were made up of families currently engaged in the associated therapy center and two parents who responded to a mailed out survey, likely representing the more motivated and engaged families and introducing a response bias.

**Communication Mode**

Of all the studies found through systematic review, only Robertson (2013) and Berrettini et al. (2008) studied implant use and mode of communication. Development of spoken langue is typically the goal for children who are implanted. It follows
that it is important to assess how many children with AD develop spoken language with implantation. Robertson (2013) exclusively focused on children with autism spectrum disorder. The study was further limited by a small sample size of only ten children, all of whom were implanted at age two years or older. The fact that only one child used spoken language as their main form of communication therefore cannot be generalized to a broader population, even among other children with autism. Great variability was found in the results. Six of the children included never developed any form of spoken language. Three used a mix of sign language and spoken language, and, as mentioned previously, one used mainly spoken language. The lack of development of spoken language may be largely due to the late implantation age. Prior research has shown that implantation before two years of age is associated with better outcomes among typically developing children. The small sample size combined with late implantation ages cast doubt on the validity of these outcomes.

Berrettini et al. (2008) looked at communication mode in two ways, first examining the overall change in participants main form of communication pre and post implantation and further examining the correlation of post-implant communication mode with post-implant speech perception categories and post-implant perceived benefit. The study found an increase in patients who used oral language (from 28% to 69%) and a decrease in patients who had a main communication mode of gestures or behaviors (from 69% to 28%) One patient used augmentative communication both pre and post implantation. The study found statistical significance for both correlations. Additionally, no statistical significance was found between degree of disability or delay and communication mode.

Other Areas
Zaidman-Zait et al. (2017) compared the health-related quality of life (HRQoL) between children with AD and CIs and typically developing children with CIs, by having parents fill out HRQoL questionnaires for their children. Their control group included children with cerebral palsy, autism spectrum disorder, and developmental disabilities. They found that children with AD had lower overall HRQoL than the typically developing children, and more specifically had lower scores on the self-esteem, friend, and school HRQoL subscales. However, when asked about perceived benefits from the cochlear implantation, the parents for both groups of children indicated a strong perception of benefits from the implant in terms of their child’s improved language, communication, and interaction, increased connection with the social environment, and increased confidence. The study did not address whether the experimental groups HRQoL was comparable to those of children with AD without hearing loss. We do not know from this study if children with developmental disabilities typically have a lower HRQoL than typically developing children, regardless of hearing loss. This is another study in which a control group of children with only AD but no hearing loss may have strengthened the study by providing a realistic comparison.

Robertson, (2013) was also the only study to focus on implant use. Again, he focused solely on individuals with Autism, where their sensory processing issues might make this area a challenge. However, all but two were recorded as “consistent users.” It was noted that one of the inconsistent users did not wear her hearing implant because the sensory input caused her distress. More studies of a larger scope would need to be conducted to determine if this outcome is representative of the broader population it attempts to represent.

Factors correlated with outcomes
Several factors were correlated with outcomes in children with AD. One key predictor of auditory skills and speech perception found in a number of studies was the degree of developmental delay. Wiley et al. (2008) found that although there was no significant difference in the progression between children with or without disabilities (although initial baseline was lower in the group with disabilities), when categorized by developmental quotient (DQ), children who fell below normal (DQ<80) had half the rate of progress of children above normal (DQ>80), suggesting that developmental quotient may be a more accurate predictor of progress than solely the presence of a disability. Hiraumi et al. (2012) similarly found that developmental quotient was correlated with post implantation speech perception scores, however noted that the correlation was weak after controlling for various other factors. Nevertheless, Edwards et al. (2006) found a correlation between measures of cognitive and developmental function and speech, and included children with more severe disabilities than those included in the Hiraumi et al. (2012) study. Trimble et al. (2008) found that functional disability scores significantly predicted high and low speech perception scores, even after controlling for chronological age, age at activation, and duration of implant use. Cruz et al. (2008) found correlations between disability type and outcomes in expressive and receptive language. Similar to the previous findings, the degree and type of delay played a significant role in whether benefit was observed in receptive and expressive language and to what degree. (Edwards 2006, Cruz et al., 2012, Nikolopoulos et al., 2008).

Bilateral implantation was another factor discussed in studies as a possible predictor of benefit from cochlear implantation among children with AD. Eshragi et al. (2015) found that children implanted bilaterally showed the most improvement in speech perception. However, the study noted that all these participants were implanted sequentially, which may have resulted in
differential subject selection, as the children performing well with their initial implant were more likely to go ahead with a second surgery.

Age at implantation has been shown to be an important predictor of CI benefit in the general population. Trimble et al. (2008) supported this finding among individuals with AD in addition to chronological age and duration of implant use, concluding that all these factors contributed to speech perception scores. However, Beer et al. (2012) found that younger age at implantation and younger cognitive age was only associated with greater improvement in functional auditory skills in the first year of implantation for children without AD, but was not predictive of improvement for children with AD. Meinzen Derr et al. (2013) also found no correlation between age of implantation and outcomes for functional skills.
DISCUSSION

The general goal of the present study was to analyze benefit in the pediatric cochlear implantation population when AD are present. This study focused on three main issues as they related to children with cochlear implantation and AD: areas of function assessed, outcome measures used, and evidence of benefit observed. The 24 articles included in this review demonstrate the wide variability in this area of research and highlight the many challenges encountered when attempting to answer these questions.

Unlike cochlear implantation for otherwise typically developing children and adults with hearing loss, there is currently no protocol or gold standard for assessing the population with AD. With reference to the first issue, results of the investigation revealed several different areas that were used to assess benefit from cochlear implantation. The most common areas of function included in the studies were auditory skills and speech perception, and expressive and receptive language. These types of assessments are similar to what is performed in children without AD. Recognizing that tests of language and auditory skills may lack validity among this population due to inherent language delays, several studies looked at the effects of cochlear implantation on adaptive behavior, including environmental awareness, engagement and social functioning. Other areas included CI use, mode of communication, and health-related quality of life, although they were considered much less frequently.

There were many different outcome metrics used for determining benefit even within the same area of function, and within a particular study a number of different types of assessments were often made. A variety of measures to assess benefit from cochlear implantation was used, including objective audiological tests, parent interviews, and checklists. This further highlights the variability among these studies and lack of a gold standard for measuring progress within this
population. While some studies used standard tests including SIR or PBK word lists, the majority of the studies used some form of observational checklist or parental questionnaire. This reflects the challenges of performing formal behavioral testing with the special needs population, a problem not typically encountered with typically developing children who are capable of sitting through more standard types of testing.

Regarding the third question on the benefit of CI obtained in this population, although results tend to reveal positive findings, many complications arise when researching this area and interpreting the results. The limited available population often results in studies with smaller sample sizes. The small sample size can be seen in studies that performed chart reviews, where the size of the experimental group was highly disproportionate to the comparison group. For example, Amisalari et al. (2010) included 28 children in the experimental group and 234 in the comparison group, while Cruz et al. (2008) included an experimental group of 31 and a comparison group of 157. Such discrepancies also highlight the complications pertaining to experimental control and selection of participants, which will be discussed later in greater detail. The problem of limited sample size is compounded in repeated measure studies due to mortality effects; in many studies, it was noted that patients were lost to follow-up and thus were not included in the final analysis. When a sample size is so limited, exclusion criteria often cannot be too specific. The term “additional disabilities” encompasses many different types of disabilities and was reflected in the varied types of participants often included in a single study.

Another limitation is that whereas some of the studies listed many types of disabilities in their participant pool, others did not specify the types of disabilities, but used vague descriptions such as “complex needs,” or “additional disabilities.” There was a much smaller number of studies which did focus exclusively on a single disorder, such as autism spectrum disorder or
cognitive or motor delays. One study only included mild cognitive delay, while another study compared children with mild and severe cognitive impairments. There was insufficient literature recovered, however, about any individual disability to warrant a review of a single disability. As a result, these results should be interpreted cautiously when generalized to any individual with special needs.

Several studies touched upon a potential solution to this variability. These studies noted that the degree and severity of cognitive delay or disability could predict benefit from cochlear implantation more so than presence of disability, suggesting that using cognitive ability or developmental quotients as an independent variable could be a more useful and reliable strategy. According to these studies, the disability type is less important that its impact on cognitive abilities. Other factors, such as age at implantation and bilateral implantation have been proven to correlate with CI benefit among otherwise typically developing children with hearing loss. Those correlations were either not present or noted to be much weaker among children with AD.

Research design was highly varied among the studies. The majority of articles included a comparison group. The studies without a comparison group did not address the potential for a maturation effect. It is difficult to explain any progress observed in cochlear implantation recipients with special needs without the context of a comparison group. Almost all the studies that included a comparison group included typically developing children with cochlear implantation as the comparison population. Only two studies compared the experimental group to a group of children without hearing loss, matched for age and cognitive abilities. It is important to consider whether a cohort of cognitively matched peers serves as a better comparison group, controlling for presence of disability and reflecting more of the residual disability due to hearing loss. None of the studies compared children with AD and cochlear
implantation with children with AD who were CI candidates and did not receive cochlear implantation. Examining these two groups may give a better idea of whether cochlear implantation provides additional benefit. However, when considering reasons why these children did not receive implants, the possibility of a differential participant selection effect seems inevitable. In the matching of experimental and comparison groups, many potential variables have been mentioned or considered in the literature. Below is a list of many of the variables noted within the 24 studies reviewed. Each of these variables, some of which were controlled for in various studies, could have confounding or interactive effects on CI benefit. Many of these factors have been linked to success with cochlear implantation in the general population. All speak to the challenge of designing and executing a tightly controlled empirical study in this area.

- **Child attributes**
  - Age at time of study
  - Gender
  - Etiology
  - Unaided PTA in better ear

- **Family Attributes**
  - Marital status
  - Maternal education
  - Family income
  - Medical insurance

- **CI Attributes**
  - Model
- Processor
- Processor strategy
- Number of active electrodes/Array
- Unilateral/bilateral implantation
- Simultaneous/sequential implantation
- Frequency of use
- Surgeon

- Training
  - Number of therapy sessions
  - Weekly hours in therapy
  - Communication method used
  - Communication methods committed to by parents
CONCLUSIONS

Despite the numerous challenges that arise in research on this topic, there does seem to some consensus on the potential for benefit due to cochlear implantation in children with AD. A common theme observed throughout the studies was that children with AD do benefit in the areas of auditory skills, speech and language, and adaptive behaviors as a result of cochlear implantation, however on a smaller scale or slower rate than children without AD who have received cochlear implantation. The improvement from before to after implantation appears to be greater than what would be expected from maturation effects alone. These results are consistent with those found in a literature review completed by Palmeiri et al. (2014).

Although widely accepted as the gold standard for assessing successful cochlear implantation among typically developing children, the validity of using speech and language measures as an indicator of CI benefit in the special needs population is questionable. Speech and language development across the span of developmental disabilities is highly varied. Demonstrating that a child with autism spectrum disorder or developmental delay does not reach equivalent language levels of their typically developing peers with cochlear implantation does not necessarily indicate lack of benefit from cochlear implantation. Many children with autism, for example, may not reach these levels even in the absence of hearing loss. Studies using cognitively matched peers without hearing loss likely gives more information regarding whether children with additional needs are reaching their language potential. This protocol was only found in two of the studies. Findings from these studies do support the notion that speech and language measures were not matching those of their cognitively matched peers, indicating residual disability due to the hearing loss.

Additionally, more assessments should analyze the health-related quality of life for these
children. Increasing the quality of life for individuals with multiple disabilities is frequently a prime goal for therapeutic and medical intervention, including cochlear implantation. Having a measure that directly assesses this area may provide more information of the efficacy of such intervention.

An interesting result found in several studies addressed the possibility of replacing the independent variable of “presence of additional disability” with “cognitive ability”. These studies found that cognitive ability prior to cochlear implantation was a more reliable predictor of outcomes than just the presence of an additional ability alone.

All of this information is critical in the evaluation and counseling of families of children with hearing loss. While the presence of an additional disability should not disqualify the child as a candidate for cochlear implantation due to the strong evidence of benefit, realistic expectations must be clarified, especially in the cases of severe cognitive impairment. While speech and language development may be the goal for children without AD, other areas of achievement should be emphasized with parents of children with AD, such as behavior, environmental awareness and social engagement, and education.

Additionally, more standardized measures should be developed specifically for this population. Considering that the prevalence of AD in the hearing loss population is about 30%, it is necessary to develop realistic and appropriate measures for parents and clinicians to assess benefit from cochlear implantation. The wide variety of outcome measures included in this review highlight the absence of such measures.
SUMMARY

Research in this area is challenging due to the limitations involved in the ability to produce randomized, double blind studies to determine value of CI in this population.

Cognitive ability is a strong, but not the only, predictor of performance. Although the lower the cognitive ability, the lower the post implant performance, there is much variability among participants, adding to the challenge of deciding whether to implant such a child.

There is some evidence to support the benefit of implantation of children with AD, however, more research is recommended involving more multicenter collaborations to increase the participant pool and to isolate individual disabilities to establish performance.

Research should continue to explore use of alternative assessments such as quality of life measures.
REFERENCES


