Co-Occurrence of Hearing Loss and Autism Spectrum Disorder (ASD): Exploring Cochlear Implantation Outcomes in the ASD Population

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Background

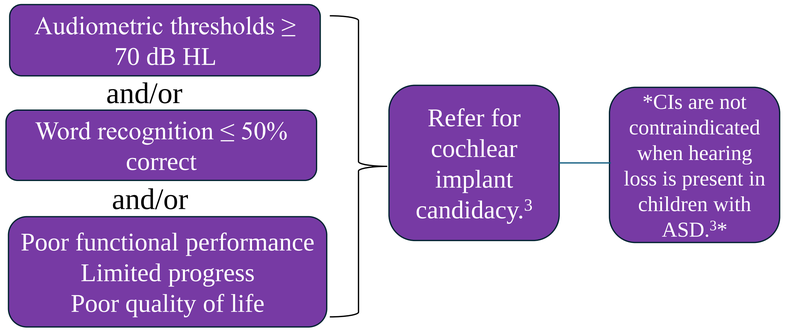
What is ASD?

ASD is a neurological and complex developmental disorder characterized by impaired social interaction, atypical communication and restricted, repetitive, and stereotypical patterns of behaviors, activities and interests.[1] ASD is classified as a "spectrum" disorder because there is a significant variation in the type and severity of symptoms that individuals experience.[2]

Pediatric Cochlear Implant (CI) Candidacy

CI candidacy varies across clinics but typically falls within this range noted by the FDA:[3]

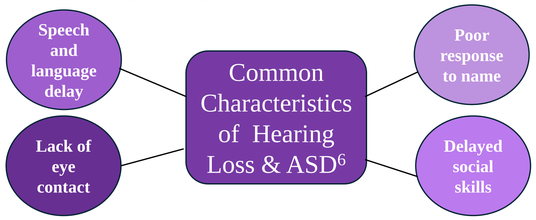
1) Infants with bilateral profound hearing loss at 9 or 12 months, depending on manufacturer.
2) Children with bilateral severe-to-profound hearing loss who are older than 24 months and up to age 18.
3) Minimal benefit from hearing aids.
4) No anatomical or surgical contraindications.



The primary objective of this literature review is to evaluate the current research available on CI outcomes in the ASD population to determine the effectiveness of CIs on improving communication/language skills and quality of life in individuals with ASD and hearing loss.

Diagnostic Overshadowing

Diagnostic overshadowing exists across all healthcare domains and is defined as the "attribution of symptoms to an existing diagnosis rather than a potential comorbid condition".[4] Over 1 billion people are estimated to experience disability. Individuals with disabilities are at greater risk of diagnostic overshadowing.[5] Hearing loss and ASD can share many behavioral characteristics which can contribute to diagnostic overshadowing and delay early intervention services.



Current Outcomes

- Profound bilateral hearing loss is estimated at a prevalence rate of 3.5% among the ASD population, **10x** higher than the prevalence of hearing loss within the general population.[7]
- In 21% of children, a diagnosis of hearing loss was delayed by several years following the diagnosis of ASD.[8]
- Conversely, the diagnosis of hearing loss may obscure recognition of autistic behaviors as ASD and delay an ASD diagnosis by up to 5 years following the hearing loss diagnosis.[9]

"Without his implant, he was stuck in his own little world, no sound, no eye contact with others. The implant brought his personality out to us".[10]

- Median aided soundfield thresholds measured at 30 dB HL.[11]
- 45% of consistent device users developed measurable open-set speech perception (PBK and CNC) by an average of 4.5 years.[10]
- **100%** of parents reported they would recommend a CI to families in a similar situation.[6]
- 86% of participants showed improvement in social engagement via parental survey responses.[10]
- The top three reported improvements after implantation include:[6]

1. Name recognition

2. Response to verbal requests

3. Enjoyment of music

9/15 *or 60% of participants saw an increase in expressive vocabulary scores.[6] Pre-operatively, speech expression was categorized as 0-1 (no vocalization or some vocalizations). Post-operatively, speech expression was categorized as 3 or 4 (using simple phrases/commands or able to produce sentences).*

Recommendations

- Pure tone audiometry (unaided and aided).
- Speech discrimination evaluated in quiet using standardized methods such as Early Speech Perception (ESP) test, Consonant Nucleus Consonant (CNC) word lists, Multisyllabic Lexical Neighborhood Test (MLNT), or the Phonetically Balanced Kindergarten (PBK) test.[6]
- Parental surveys. [6, 10]

Research Limitations

- Standard audiologic and speech perception measurements may not capture improvements in quality of life. There are currently no validated measures of quality of life for implanted children with additional disabilities (e.g. ASD).[10]
- Small sample sizes which included participants with a variety of comorbidities.
- Periods of device non-use in participants.
- Not all participants underwent a comprehensive speech and language evaluation (pre- and post- implantation).
- Participants across studies were in different geographical areas, school districts, etc., impacting access to therapeutic services.
- Lack of data on multilingual CI users with ASD.
- No comparison to normal hearing peers with ASD.[10]
- Limited family response to follow-up questionnaires.[10]

Clinical Implications

- Highlights the importance of educating multidisciplinary team members on the significance of audiological evaluation in differential diagnosis for patients undergoing evaluations for ASD.
- If audiologically and surgically candidates, patients with ASD and hearing loss can benefit from CIs.
  - Although CIs may provide access to spoken language, goals of implantation vary.
- Emergence of measurable open-set speech perception occurred on average 4.5 years post-CI[10] suggesting children with ASD and CIs need continued, long-term therapeutic services to achieve open-set speech perception compared to their peers with hearing loss and no other complicating conditions.
- Emphasizes the importance of modified audiological testing when assessing children with (suspected) ASD.
- Underscores the need for additional research on validating CI outcome measures meant for individuals with ASD and disabilities in general.

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