Audiological Considerations for Children with Cerebral Palsy, Deaf-Blindness, and Autism Spectrum Disorder

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Background

Research suggests that 30-40% of children with permanent hearing loss have co-occurring disabilities (Fitzpatrick, Lambert, Whittingham, & Leblanc, 2014). According to Roush, Holomb, Roush, & Escolar (2004), the most common diagnoses which occur with hearing loss include learning disorders, intellectual disabilities, attention disorders, visual impairment, and cerebral palsy. Such conditions can make conventional methods of diagnosis and management of hearing loss more difficult compared to working with typically developing deaf and hard of hearing children.

Objective

The goal of this systematic review was to evaluate the available evidence regarding alternate methods for assessment and management of hearing loss (HL) in children with three of the most commonly occurring comorbidities: Cerebral Palsy (CP), Deaf-blindness, and Autism Spectrum Disorder (ASD).

Methods

Electronic search of CINAHL and Embase databases, using the following search terms:

- Key search terms included variations on (1) hearing loss, (2) childhood, (3) Cerebral Palsy, (4) Autism Spectrum Disorder, (5) Deaf-blindness, (6) audiological assessment, (7) treatment, and (8) outcomes.
- Full search terms available upon request.

Each step in the systematic review process was independently conducted by the three authors. No less than 20% of articles were double-reviewed for each step, and inter-rater reliability was calculated.

- Title/abstract review (reliability = 98%)
- Full text review of included articles, assessing relevance and adherence to inclusion/exclusion criteria (reliability = 94%)
- Quality appraisal of included articles (reliability = 100%)
- Data extraction from included articles

Inclusion Criteria

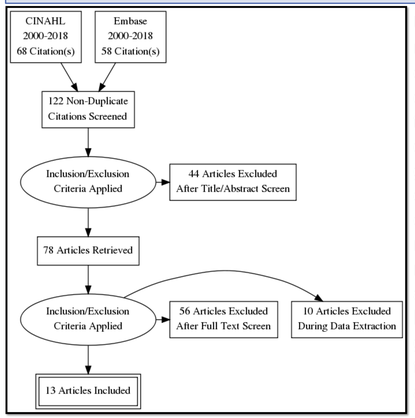
- Children with congenital or late onset/progressive HL due to a genetic disorder
- Children with CP, Deaf-blindness, and/or ASD
- Children under 21 years of age
- Journal articles and book chapters

Exclusion Criteria

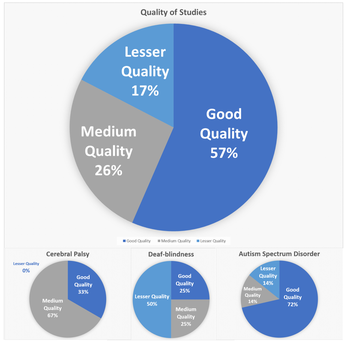
- Adults with hearing loss
- Auditory Neuropathy Spectrum Disorder
- Children with multiple other/severely debilitating disorders
- Papers written before 2000
- Papers written in a language other than English

Results

Review and Quality Appraisal



Upon reviewing the quality of 23 articles using the Joanna Briggs Institute checklists, 10 articles were excluded for data extraction. It was determined that the studies which addressed CP and deaf-blindness studies were not generalizable nor good quality. For example, the deaf-blind articles focused only on Usher syndrome. The 13 remaining articles consisted of 7 regarding Autism Spectrum Disorder, and 6 which discussed children with hearing loss and multiple disabilities in general. Autism Spectrum Disorder was then the primary focus for our data extraction, as they provided relevant and good quality data.



Data Extraction

	Donaldson, Heavner, & Zwolan (2004)	Eshragi et al., (2015)	Fitzpatrick, Lambert, Whittingham, & Leblanc (2014)	Hansen, S. (2018)	Mikic et al. (2016)	Thompson & Yoshinaga-Itano (2014)	Valero et al. (2016)
Assessment	Authors made no recommendations regarding alternate methods of assessment.	Communication and language difficulties can make conventional assessment methods more challenging.	Children with ASD tend to be less reliable and give more variable responses during behavioral audiologic testing. Therefore, electrophysiological measurements (like Auditory Brainstem Response) are valuable.	Found a lack of modified assessment tools for children with ASD and hearing loss dual diagnosis.	The development of auditory perception and speech intelligibility were assessed using the Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) SIR at 5 year follow up	Recommended: a predictable routine, social stories, practice with inserts, explanation for all parts of the assessment, modeling and practice for all aspects of testing, allowing the child to watch a hearing test of a parent or sibling.	Authors compared assessing children with ASD to assessing non-English speaking children. There needs to be more validated tools for assessing hearing loss in children with ASD.
Management	Cochlear implants	Cochlear implants	Hearing aids, FM systems, and/or cochlear implants	Found a lack of modified intervention methods and advocated for an interdisciplinary approach due to lack of specialists in both subjects.	Cochlear implants	Recommended using pictures, videos, models and demonstrations for earmold impressions and hearing aid fittings.	Cochlear implants
Outcomes	All 7 children demonstrated progress when compared to pre-operative scores. Parents reported greatest improvement for enjoyment of music.	Per parent report, the top three areas of improvement post-implantation were (1) response to name, (2) awareness of environment, and (3) enjoyment of music.	9/17 children included in the study	None reported	Significantly slower skill development for children later diagnosed with ASD compared to neurotypical children	Children with ASD and D/HH diagnoses will have better fittings and outcomes if a routine is used and they knew what to expect.	16/22 children remained compliant with cochlear implant use, however, not all of them went on to develop appropriate spoken language skills.

Discussion

Based on this systematic review, it is clear that there is a lack of evidence regarding the assessment and management of hearing loss in children with cerebral palsy and deaf-blindness. Considering the large number of children with hearing loss and other disabilities, there needs to be more research to validate alternate methods for evaluating and treating hearing loss in this population, as well as to identify expected outcomes.

Regarding children with hearing loss and Autism Spectrum Disorder, there is more available evidence.

- Assessment – children with ASD can be difficult to test using conventional methods due to their language and communication difficulties. Because of this, it is important to use a predictable routine, and allow the child the opportunity to rehearse this routine. However, children with ASD are often highly variable during behavioral testing. Therefore, it is helpful to use electrophysiological tests to verify behavioral results.
- Management – children with ASD regularly receive hearing aids, FM systems and cochlear implants as treatment for varying types and degrees of hearing loss.
- Outcomes – though children with ASD often do not achieve the same levels of progress in speech and language as their typically developing peers, they can be expected to benefit from amplification and/or implantation. Many parents report subjective improvement in attention and behavior following treatment for hearing loss.
- It is important to emphasize to parents that appropriate management of hearing loss *will not* eliminate a child's diagnosis of ASD.

This data suggests that ASD is by no means a contraindication to amplification and/or implantation. Overall, more research is needed to create validated methods for diagnosing and managing hearing loss in children with ASD.

Key References

Handout of full reference list available upon request.

Disclosures/Acknowledgements

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