Cochlear Implant Behavioral Outcomes for Children with Auditory Neuropathy Spectrum Disorder: A Mini-systematic Review

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ABSTRACT

Objective

The aim of this mini-systematic review was to evaluate the evidence reporting speech, language, and auditory behavioral outcome measures for children with a diagnosis of Auditory Neuropathy Spectrum Disorder (ANSD) who received cochlear implants (CI) prior to three years of age.

Design

A mini-systematic review of the literature supporting evidence-based practices was performed. Two databases were searched utilizing a search strategy derived from the PICO Framework. Peer-reviewed articles published between 2009-2019 evaluating children with a diagnosis of ANSD who were implanted prior to age three utilizing speech, language, and auditory behavioral outcomes were included. Four articles meeting inclusion criteria were critically appraised for reputable research design and risks of bias. Each of the four studies was assigned a level of evidence for effectiveness and quality assessment rating.

Results

Evidence supports cochlear implantation is an appropriate intervention for children with ANSD. Improvements in outcome performance were observed in all the included studies. Children with ANSD fit with CI can achieve similar outcomes to children with sensorineural hearing loss (SNHL) and CIs, despite the heterogeneity of ANSD.

Conclusions

These findings have implications for clinical practice and for future research with current CI technology for facilitating parent education, counseling, and realistic expectations for children with ANSD and CIs.

PICO QUESTION

What is the efficacy of CI for children with a diagnosis of ANSD utilizing speech, language, and auditory behavioral outcome measures?

Table 1. *PICO Framework Defined*

PICO Components	PICO Question Elements	PICO Concepts
Patient	Children with ANSD	Children diagnosed with ANSD and implanted prior to age three
Intervention	Cochlear implantation	Cochlear implantation of affected ear/ears
Comparison	Pre- and/or post-operatively	CI performance before and after surgery
Outcome	Speech, language, and auditory behavioral outcome measures	Do cochlear implants help children with ANSD improve on speech and auditory behavioral outcome measures?

Note. PICO Components, PICO Question Elements, and PICO Concepts are presented. P = Patient; I = Intervention; C = Comparison; O = Outcomes. Adapted from "Formulating Questions and Locating Primary Studies for Inclusion in Systematic Reviews," by C. Counsell, 1997, *Annals of Internal Medicine, 127,* p. 380-387 .

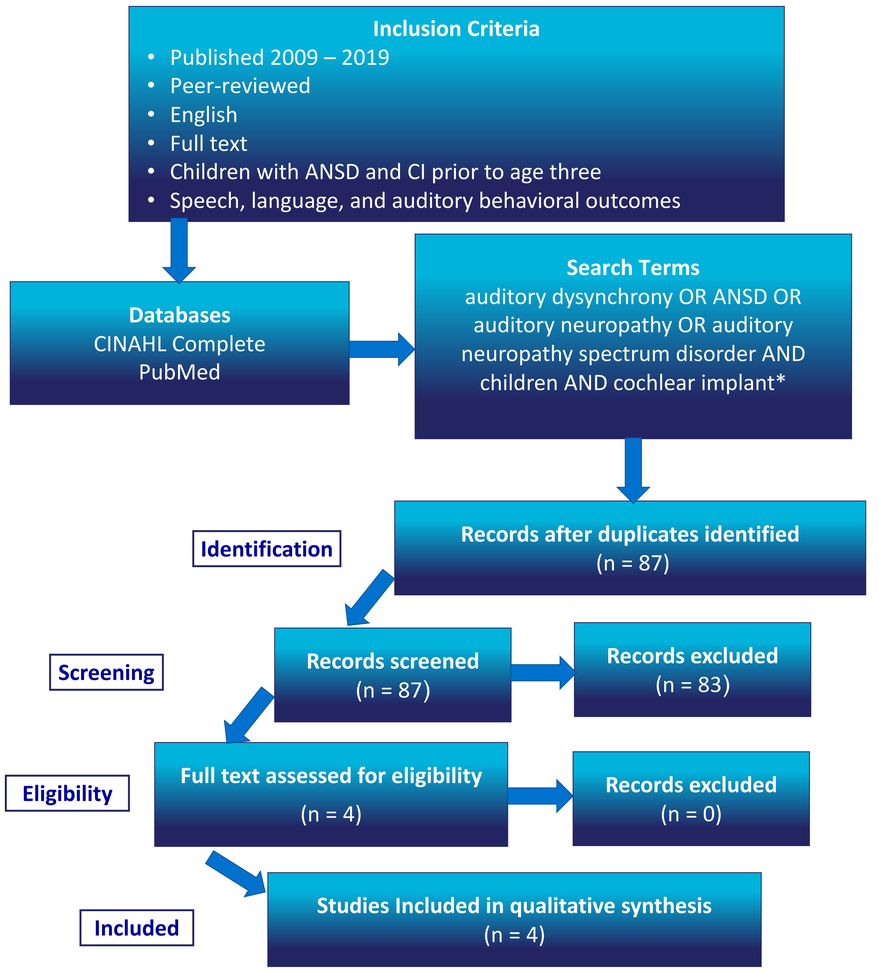

Figure 1. Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) Flow Diagram. Adapted from "Preferred Reporting Items for Systematic Reviews and Meta-analyses: The PRISMA Statement," by D. Moher, A. Liberati, J. Tetzlaff, D. G. Altman, & The PRISMA Group, 2009, *PLOS Med, 6*, e1000097.

RESULTS

Table 2. *Study Characteristics*

	Alzhrani et al. (2019)	Ching et al. (2013)	Daneshi et al. (2018)	Liu et al. (2014)
Design	Cohort	Cohort	Cohort	Case Series
N	58	47	136	10
Age Implanted (Mean months)	32.1	18.2	31.9	35.5
Auditory and Speech Measures	CAP, SIR	PEACH, DEAP, CDI, PPVT-4, PLS-4	CAP, SIR	CAP, IT-MAIS, MAIS, MLNT, LNT, SIR, MUSS
Performance Effect	No Effect	No Effect	Favorable	Favorable
GRADE	Moderate	Moderate	Moderate	Low
JBI Level	3	3	3	4

Note. Studies included were summarized in terms of design, number of participants, age at implantation in mean months, speech and auditory behavioral outcome measures with reportable data, performance effect, GRADE quality assessment rating, and level of evidence. N = number of participants; JBI = Joanna Briggs Institute; GRADE = Grading of Recommendations, Assessment, Development and Evaluations; CAP = Categories of Auditory Performance; MAIS = Meaningful Auditory Integration Scale; IT-MAIS = Infant-Toddler Meaningful Auditory Integration Scale; LNT = Monosyllabic Lexical Noise Test; MLNT = Multisyllabic Lexical Noise Test; SIR = Speech Intelligibility Rating; PLS-4 = Preschool Language Scale version 4; PPVT-4 = Peabody Picture Vocabulary Test version; DEAP = Diagnostic Evaluation of Articulation and Phonology; CDI = Child Development Inventory; PEACH = Parents Evaluation of Aural/Oral Functional Performance; MUSS = Meaningful Use of Speech Scale. Adapted from "GRADE: An Emerging Consensus on Rating Quality of Evidence and Strength of Recommendations," by G. H. Guyatt, A. D. Oxman, G. E. Vist, R. Kunz, Y. Falck-Ytter, P. Alonso-Coello, & H. J. Schünemann, 2008, *BMJ*, *336*, p. 924-926.; "JBI Levels of Evidence," by the Joanna Briggs Institute, 2014, Retrieved from https://joannabriggs.org/sites/default/files/2019-05/JBI-Levels-of-evidence_2014_0.pdf.

Table 3. *Risks of Bias*

	Alzhrani et al. (2019)	Ching et al. (2013)	Daneshi et al. (2018)	Liu et al. (2014)
Pre- & Post- Measures	-	-	+	-
Controlled for Comorbidities	-	+	-	+
Inconsistencies in Data Reporting	+	+	+	+
Auditory Progress	+	*	+	+
Recommend CI for ANSD	+	*	+	+
HA Trial Prior to CI	+	+	+	*

Note. Studies were assessed to examine the inclusion of pre- and post- operative speech and auditory behavioral outcome measures, control for comorbidities, risk factors affecting outcome performance, inconsistencies in data reporting, auditory progress of children with ANSD and CI, if CI was a recommended intervention option for children with ANSD, and if children included had a proper hearing aid trial prior to implantation. (+) = YES; (–) = NO; (*) = UNCLEAR; ANSD = Auditory neuropathy spectrum disorder; CI = cochlear implant; HA = hearing aid.

SUMMARY OF FINDINGS

Alzhrani et al. (2019)

- No significant differences in performance between ANSD-CI and SNHL-CI groups
- JBI Level 3 due to cohort design
- GRADE: Moderate quality and high risk of bias

Ching et al. (2013)

- No significant differences between ANSD-CI and SNHL-CI groups or ANSD-HA or ANSD-CI groups, respectively
- JBI Level 3 due to cohort design
- GRADE: Moderate quality and high risk of bias

Daneshi et al. (2018)

- Children with ANSD improved significantly with auditory and speech production skills with CI and was affected by age at implantation
- JBI Level 3 due to cohort design
- GRADE: Moderate quality and low risk of bias

Liu et al. (2014)

- Children implanted before 24 months may acquire better auditory and speech skills than those implanted after 24 months
- JBI Level 4 due to case series design
- GRADE: Low quality and high risk of bias

CLINICAL IMPLICATIONS

- Children with ANSD and CI can perform equally as well as children with permanent SNHL and CI on speech and auditory behavioral outcomes, suggesting that parents can be counselled on CI as a potential treatment option for a child with ANSD.
- Early diagnosis and early intervention can positively affect future CI success in terms of speech, language, and auditory abilities.
- Speech, language, and auditory behavioral outcomes for children with ANSD should ideally be frequently monitored and discussed on a case-by-case basis with a patient and family centered care approach.

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