

# Background

Auditory Neuropathy Spectrum Disorder is a sensorineural hearing loss (SNHL) characterized by an impairment of the auditory nerve. This generally means that while sound is able to travel through the outer, middle, and inner ear, it is unable to successfully reach the brain. Today, cochlear Implants and hearing aids have both become common interventions for children with ANSD. The primary goal of any hearing loss intervention in children is to foster speech and language development. Thus, the intent of this systematic review was to answer the question: In children with ANSD, how does amplification compared to cochlear implantation affect speech and language outcomes?

# Objective

The purpose of this systematic review was to investigate differences in speech and language outcomes between hearing aids and cochlear implants in children with Auditory Neuropathy Spectrum Disorder (ANSD)

# Methods

The three authors performed a literature search from three major databases; PubMed, Ovid, and MedlinePlus, using the following search string: (ANSD OR CND OR Auditory Neuropathy OR Cochlear Nerve Deficiency) AND (children OR pediatric OR school) AND (amplification OR Hearing aids OR hearing amplification OR acoustic stimulation) AND (cochlear implant\* OR electric stimulation). The strategy yielded 194 articles across the three databases searched. Articles were then initially screened based on title and year of publication. Any articles that clearly did not meet inclusion criteria, based on title or review of the abstract, were deleted. This resulted in 119 articles that were then checked for duplicates. Once duplicates were removed, 75 articles remained to be further evaluated.



# Interventions for Speech and Language Outcomes for Children with ANSD: A Systematic Review Jordan Potosky, B.A., Megan Frey, B.S., & Lisa Copeland, B.A. Division of Speech and Hearing Sciences, The University of North Carolina at Chapel Hill

## Results

### Inter-rater Reliability

The abstracts of 75 articles were carefully reviewed, independently, by the three authors. Studies including speech and language outcomes of this population of children were included. Exclusion criteria included children with comorbidities, non-English speaking populations, and studies that only considered academic achievement outcomes. The researchers compared which articles they reviewed that met the inclusion criteria and found a 80% inter-rater reliability. After discussion about each article in question, a final 11 articles were chosen for the systematic review.

### Critical Appraisals

Critical appraisals were independently completed by three researchers. One third of the articles were evaluated by each researcher to measure inter-rater agreement using LEGEND Appraisal Forms from Cincinnati Children's Hospital (Cincinnati Children's Medical Center, 2011). The researchers focused on the design of the study, population, sample size, results, and limitations to determine level of evidence. Results of the evidence appraisals were crossed checked by one other researcher. Each article was assigned a "lesser quality" or "good quality" rating based on the type of study appraised. Seven articles were rated as "lesser quality" and four articles as "good quality."

## Data Extraction

Comparing results across studies was difficult due to the heterogeneity of the populations. Participants and methodology, including speech perception measures used, varied greatly across studies. Data extraction focused on the populations considered, sample size, age at intervention, type of intervention, type of control/comparison group, speech outcome measures used, and general findings. Each researcher was responsible for extracting data from 3-4 articles, and the data was cross-checked by at least one other researcher.

	DECICN		NT		
	DESIGN	POPULATION	N	KESULIS	<b>EVIDENCE</b> <b>QUALITY</b>
<b>BERLIN ET AL.</b> (2010)	Case Review	Children with ANSD	260	No analysis completed	Lesser
<b>DEAN ET AL.</b> (2013)	Case Review	Children with ANSD and CI	27	Bilateral CI users were better performers than unilateral CI users. Better pre-CI PTAs correlated with better post CI speech perception	Good
<b>KUTZ ET AL. (2011)</b>	Case Review	Children with ANSD and CI	9	Higher pre-implantation speech scores may be an indicator of higher post-implantation speech scores	Lesser
SHALLOP ET AL. (2015)	Case Review	Children with ANSD and CI	5	CIs are an appropriate intervention for ANSD	Lesser
<b>RANCE ET AL.</b> (2008)	Cohort Study	Children with ANSD and CI / Children with ANSD and HA	20	No difference, p-value not provided	Lesser
KANG ET AL. (2010)	Cohort Study	Children with ANSD and CI / Children with SNHL and CI	21	No statistical analysis completed	Lesser
<b>TEAGLE ET AL.</b> (2010)	Cohort Study	Children with ANSD and CI / Children with ANSD and HAs	102	Due to heterogenous nature of ANSD, performance of speech perception varies among children with CIs and HAs. No statistical analysis completed.	Good
ZENG ET AL. (2006)	Cohort Study	Children with ANSD and CI	13	No statistical analysis completed	Lesser
PETERSON ET AL. (2003)	Cohort Study	Children with ANSD and CI / Children with SNHL and CI	10	No statistical analysis completed	Lesser
BRENEMAN ET AL. (2012)	Cohort Study	Children with ANSD and CI / Children with SNHL and CI	35	No differences found using ANOVA	Good

The clinical evidence determining the differences in speech and language outcomes between CI and HA interventions in the ANSD population is limited. It is important to note that studies included in our review varied in type of speech perception and language measure making it challenging to draw conclusions across studies. Stronger evidence is needed to demonstrate any important differences in cochlear implant benefit compared to hearing aid benefit as it pertains to speech and language outcomes in children with ANSD.

While the evidence supporting positive outcomes for children with ANSD that use hearing aids is limited, our review suggests that some children with ANSD receive benefit from hearing aids with improved speech perception scores. Similar findings were noted in the population of children with ANSD who received a cochlear implant.

Roush et al (2009) reported that children with ANSD with thresholds within the severe to profound range may benefit from cochlear implantation with improved speech perception scores as compared to speech perception scores using hearing aids, while Rance et al. (2002) found no correlation between speech perception scores and pure-tone thresholds in a population of children with ANSD. While one child with ANSD and a more severe hearing loss may benefit from hearing aids, another child with ANSD and a mild hearing loss may not. This creates a challenge for the audiologist when determining intervention strategy for children with all degrees of hearing loss.

Further research is needed to address the methodological issues related to the studies we reviewed. Considering the variety of etiologies and characteristics in children with ANSD, there is a need to create studies designed with a more homogeneous group to more accurately assess the impact of interventions on speech and language outcomes.

References available upon request. Jordan Potosky: jordan\_potosky@med.unc.edu Megan Frey: megan\_frey@med.unc.edu Lisa Copeland: lisa\_copeland@med.unc.edu

# **Disclosures and Acknowledgements**

# Discussion

# References

• The researchers have no financial or intellectual conflicts of interest. • This systematic review was completed as a project for SPHS 701 Introduction to Research Methods, under the guidance of Dr. Linda Watson & Dr. Jessica Steinbrenner.