



# Audiologic Management and Outcomes of Children with Auditory Neuropathy Spectrum Disorder (ANSD)

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## Background

Auditory Neuropathy Spectrum Disorder (ANSD) is clinically characterized by abnormal Auditory Brainstem Response (ABR) wave morphology and normal outer hair cell function with present cochlear microphonic and/or otoacoustic emissions (OAEs) (Starr et al, 1996). This combination of features makes electrophysiologic prediction of functional hearing and outcomes of amplification difficult to predict (Berlin et al, 2010). Hearing ranges from normal to profound loss in one or both ears, and with equally variable speech performance that is often disproportionately poor relative to the degree of hearing loss (Rance et al, 1999). There is no current international standard of management for this population. Clinical interventions vary between no intervention, hearing aids, and cochlear implants. The purpose of this systematic review is to evaluate the current literature for management strategies for ANSD internationally and their associated outcomes.

## Clinical Question

What are the current recommended audiologic management strategies and related outcomes for children diagnosed with Auditory Neuropathy Spectrum Disorder (ANSD)?

## Methods

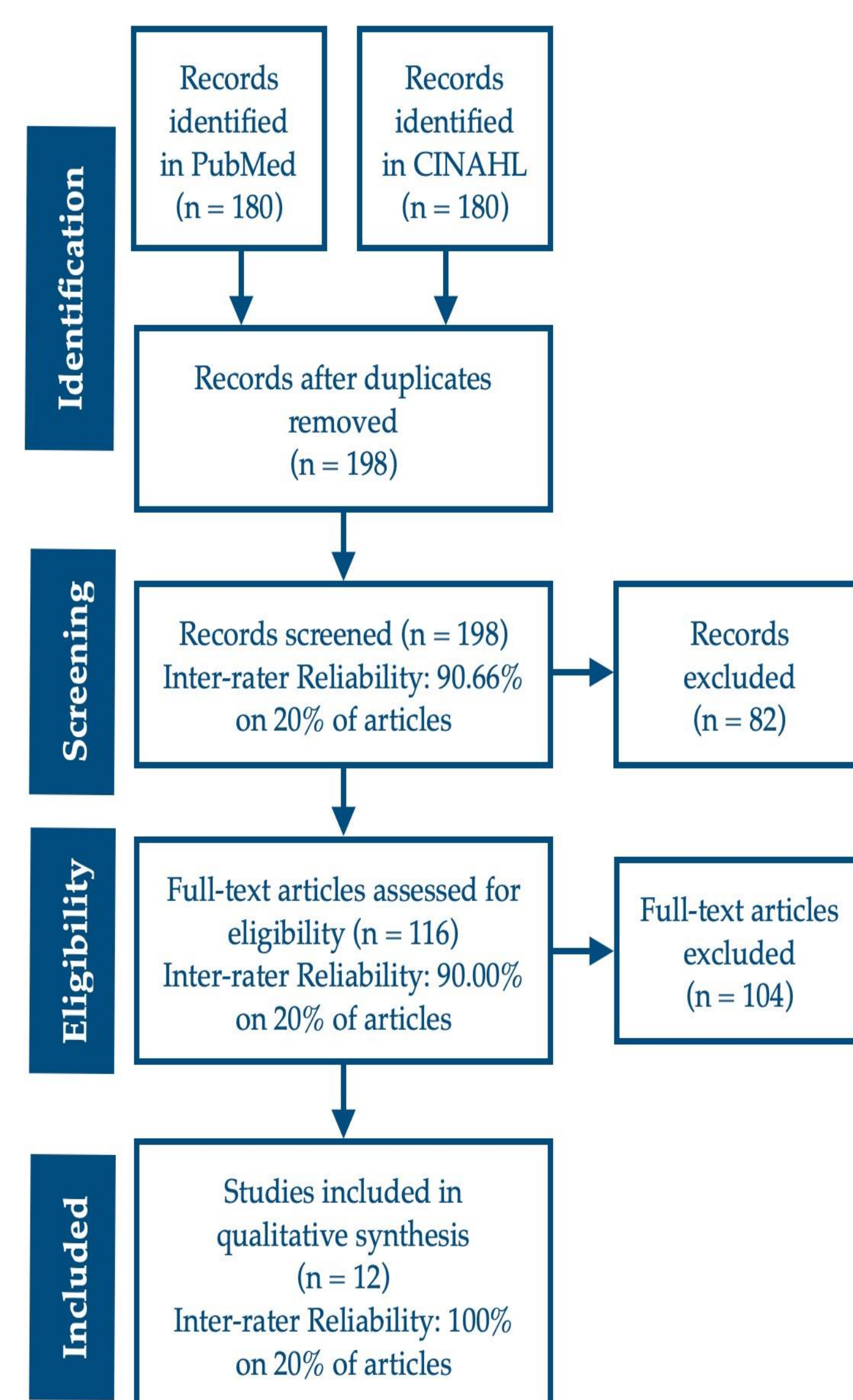
### Search Criteria

- Inclusion**
  - Expert opinions
  - Clinical Practice Guidelines
  - Conference presentations
  - Qualitative research
  - Quantitative research
  - Participants ≤ 18 years old
- Exclusion**
  - Literature reviews
  - Animal studies
  - Case studies <5 subjects
  - Participants > 18 years old
  - Participants with multiple disabilities
  - Non-English
  - Diagnosis only

### Key Search Terms

- Management/intervention
  - Children
  - Auditory neuropathy spectrum disorder
- \*and related synonyms

## Search Outcomes



## Results

Author	Study Type	Sample Size	Interventions	Assessment Tools	Results & Findings
Breneman et al., (2012)	Retrospective Cohort Study	N = 70  (35 ANSD 35 SNHL)	ANSD + CI vs. SNHL + CI	- Questionnaires (ITMAIS/MAIS, LittIEARS) - ESP & GASP - Closed Set (NU-CHIPS, WIPI) - Open Set (MLNT, LNT, CNC) - Sentences (HINT-C, AzBIO, BKB-SIN)	- Children with ANSD received a cochlear implant after showing limited improvement in auditory and spoken language skills after 3-6 months of HA use. - Once implanted, no significant difference in performance on word recognition or parental perceived benefit was noted compared to SNHL + CI peers. - Children with ANSD are equal candidates for cochlear implantation when compared to children with SNHL.
Rance et al., (2009)	Retrospective Cohort Study	N = 30  (10 ANSD + CI 10 SNHL+ CI 10 ANSD + HA)	ANSD + HA vs. ANSD + CI	- Receptive Language (PPVT: LQ) - Speech Production (DEAP)	- 9 of the 10 ANSD + CI subjects had similar language outcomes to the general pediatric CI population. ANSD + HA group language outcomes were comparable to that of the ANSD + CI group. - These results indicate that ANSD children should not automatically be considered for a CI. - Young ANSD children should be considered for a CI as soon as "stable" hearing thresholds in severe to profound range can be established. If hearing thresholds in normal to moderate, degree of functional hearing should be assessed with formal speech perception testing (4yo+). - However, this study shows age at implantation is a significant factor that impacts language.
Runge et al., (2011)	Controlled Clinical Trial	N = 9  (All bilateral ANSD with Unilateral CI)	All ANSD Unilateral CI + HA vs. Unilateral CI only	- Speech Perception (CRISP Jr.)	- Statistically significant overall improvement in SRT with amplification on the contralateral ear. - ANSD patients with one CI benefit from amplification on the contralateral ear (especially when considering candidacy for a sequential CI).
Jeong et al., (2007)	Retrospective Cohort Study	N = 21  (9 ANSD + CI 12 SNHL + CI)	After ANSD + low-gain HAs: ANSD + CI vs. SNHL + CI	- Receptive Language (CAP) - Open Set (Monosyllabic Words for phonemes, Common Phrases) - ECAP	- All ANSD children trialed low-gain HAs and did not show spontaneous improvement in hearing thresholds or speech perception. - After no perceived benefit, all ANSD children were implanted and showed improvement (no difference when compared to children with SNHL + CI).
Walker et al., (2016)	Longitudinal Cohort Study	N = 34  (12 ANSD 22 SNHL)	ANSD + HA vs. SNHL + HA	- Speech & Language (GFTA-2, PPVT-4, Vineland Adaptive Behavior Scales-II, CASL) - Speech Perception (PBK) - Questionnaire (LittIEARS)	- No significant difference between ANSD & SNHL on language articulation measures. - Although not statistically significant, a trend that ANSD + HA group did worse in background noise than SNHL + HA group was found. - ANSD children received a HA trial if behavioral thresholds are reliable and sufficiently high enough to impede speech perception at conversational levels.
Daneshi et al., (2018)	Retrospective Cohort Study	N = 136 All severe-profound bilateral ANSD HL	ANSD + CI  (Group 1: ≤ 24 mos Group 2: > 24 mos)	- Receptive Language (CAP) - Speech Production (SIR)	- At 1-year and 2-year post-op, there was a significant improvement in CAP results for both groups. - A 1-year post-op, there was a significant improvement in SIR results for both groups. - At 2-year post-op, there was <i>not</i> a significant improvement in SIR results for both groups. - All children with ANSD benefitted from cochlear implantation.

\* This chart includes only the highest quality articles. Six lesser quality articles are not discussed in detail. Of those, five articles are retrospective cohort studies and one is a case control study.

## Discussion

The evidence reviewed shows current clinical practices continue to include a variety of interventions. There is evidence of benefit from both cochlear implants and hearing aids. Some intervention compared to no intervention is beneficial; however, there is no standardized intervention protocol. The strength of clinical implications are not clear because effect size was not noted in any of the analyzed articles and heterogenous outcome measures complicates comparison across studies. Children with ANSD can have positive outcomes through varied interventions and recommendations for management appear to be patient-dependent. The goal of this review was to focus specifically on ANSD management and outcomes. One limitation of this study is excluding participants with multiple disabilities, which often co-occur with ANSD. It is possible that co-occurring disabilities could affect management of this patient population. Two areas of future research could assess 1) management of children with ANSD and co-occurring disabilities, and 2) patient characteristics that indicate specific management guidelines.

## References

References available upon request.  
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## Disclosures

The authors have no conflicts of interest to report.