

Cochlear implantation in postsynaptic auditory neuropathies are variable but generally good with patients showing improvements in hearing thresholds and speech perception.

Cochlear Implantation Outcomes in Post Synaptic Auditory Neuropathies: A Systematic Review and Narrative Synthesis D. Chaudhry, A. Chaudhry, J. Muzaffar, P. Monksfield, M. Bance

INTRODUCTION

Auditory neuropathies (ANs) can be broadly classified by the anatomical locus of dysfunction. These divisions include presynaptic disorders, postsynaptic disorders, and central neural pathway disorders. **Figure 1** demonstrates a schematic representation of these divisions. Although cochlear implantation (CI) outcomes in patients with AN are variable, most patients seem to benefit with improvements across their speech perception, language development, and communication. The observed efficacy of CI seems to be closely related to the locus of the lesion. In presynaptic ANs, outcomes are invariably good. CI outcomes in postsynaptic AN have been reported as much more variable. This is partially explained by the wide array of etiologies classified as postsynaptic ANs, compounded by their relative rarity and limited published data.

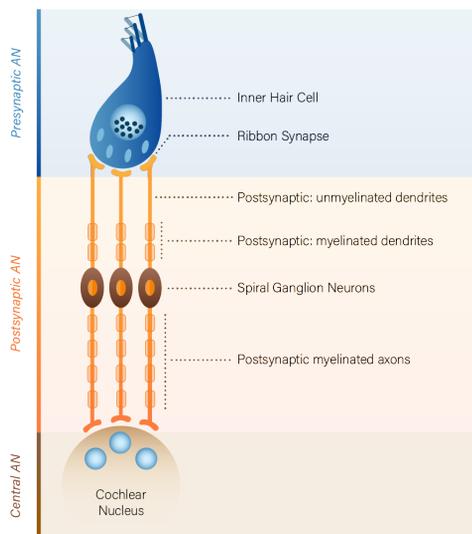


Figure 1. Overview of the peripheral auditory system showing the presynaptic, postsynaptic, and central sites of lesions associated with auditory neuropathy .

OBJECTIVES

The aim of this review was to collect and synthesize available literature on CI outcomes in patients with postsynaptic ANs. Pooling this data may lead to more reliable estimations of cochlear implant efficacy on the basis of the etiology of the AN and subsequently enable improved patient counseling and management.

Population: Children or adults with postsynaptic ANs | **Intervention:** CI (with or without auditory training, rehabilitation, or acoustic hearing aids) | **Comparison:** No comparison group | **Outcomes:** Primary outcomes were pre-implantation versus post-implantation audiometric outcomes. Secondary outcomes included intraoperative and postoperative adverse events, use of cochlear implant at follow-up, and patient reported outcome measures (PROMs).

MATERIALS and METHODS

Systematic review and narrative synthesis.

Databases searched: MEDLINE, PubMed, EMBASE, Web of Science, Cochrane Collection and ClinicalTrials.gov. No limits placed on language or year of publication.

RESULTS

Searches identified **98 studies in total**, of which **14 met the inclusion criteria** reporting outcomes in **25 patients** with at least 28 CIs. Of these:

- 4 studies focused on Charcot-Marie-Tooth disease (CMT)
- 3 on Brown-Vialetto-Van-Laere syndrome (BVVL)
- 2 on Friedreich Ataxia (FRDA)
- 2 on Syndromic dominant optic atrophy (DOA+)
- 2 on Cerebellar ataxia - areflexia - pes cavus - optic atrophy - sensorineural hearing loss (CAPOS) syndrome
- 1 on Deafness-dystonia-optic neuropathy (DDON) syndrome.

All studies were Oxford Centre for Evidence Based Medicine (OCEBM) grade IV. Overall trend was towards good post-CI outcomes with 22 of the total 25 patients displaying modest to significant benefit.

CONCLUSION

Hearing outcomes following CI in postsynaptic ANs are variable but generally good with patients showing improvements in hearing thresholds and speech perception. Further research is required to understand AN pathophysiology and develop better diagnostic tools for more accurate identification of lesion sites. Multicenter longitudinal studies with standardized comprehensive outcome measures including health-related quality of life data will be key in establishing a better understanding of short and long-term post-CI outcomes.

