

Cochlear Implantation in An Adult with Auditory Neuropathy Spectrum Disorder: A Preliminary Outcome Report

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ABSTRACT

Cochlear implantation has emerged as an effective management option for individuals with Auditory Neuropathy Spectrum Disorder, although its outcomes are strongly influenced by the underlying site of lesion. We report our experience with the cochlear implantation of a 25-year-old female with adult-onset auditory neuropathy, six months post-surgery. With tailored auditory training combined with periodic Map optimisation, subject demonstrated substantial improvements in speech recognition in both quiet and noise, along with marked enhancement in overall quality of life. Our findings corroborate existing evidence on potential benefits of cochlear implantation in improving auditory performance and patient-reported outcomes in auditory neuropathy spectrum disorder. The case further underscores the importance of individualised post-implant rehabilitation and programming strategies, particularly in cases with less predictable prognostic indicators.

Keywords: Auditory Neuropathy, Aural Rehabilitation, Cochlear Implantation

INTRODUCTION

Auditory Neuropathy Spectrum Disorder (ANSD) refers to a group of hearing disorders associated with altered functioning of the auditory nerve or its synaptic junction, characterised by abnormal or absent Auditory Brainstem Responses (ABR) despite preserved cochlear outer hair cell function.¹ Individuals typically present with speech perception difficulties disproportionate to their hearing thresholds.

ANSD subtypes are broadly classified based on the site of lesion into: (a) presynaptic disorders (inner hair cells and/or ribbon synapses), (b) post-synaptic disorders (auditory nerve structures), and (c) central pathway disorders involving the auditory brainstem.² Identification of the subtype can provide significant insight into treatment outcomes, but requires a comprehensive test battery including audiometry, speech perception testing, Oto-acoustic Emissions (OAEs), ABR, Electrocochleography (ECoG), supplemented by genetic testing where feasible.

Hearing aid outcomes in ANSD are often limited and variable, whereas cochlear implants (CI) have emerged as a more consistent management option by bypassing dysfunctional sensory and synaptic mechanisms.^{1,3} However, outcomes in post-synaptic subtypes remain less predictable.^{1,2,4} In this

preliminary outcome report, we present our experience with cochlear implantation in an adult with late-onset ANSD.

CASE REPORT

A 25-year-old female presented with progressive hearing loss since age 13. Hearing aids fitted at 15 years provided no benefit, leading to social withdrawal and discontinuation of education. Audiological evaluation revealed bilateral moderately severe

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sensorineural hearing loss with word recognition scores below 20% in quiet. ANSD was diagnosed based on absent ABRs with robust OAEs and poor speech perception. Extra-tympanic ECochG showed only cochlear microphonics with absent summing and action potentials. No neurological or radiological abnormalities were detected, and genetic testing was not performed

Cochlear Implantation

As subject had slightly better low frequency residual hearing in the right ear, she underwent cochlear implantation in the left ear via cortical mastoidectomy and posterior tympanotomy, with full electrode insertion confirmed on C-arm X-Ray. Intraoperatively, electrode impedances were normal, but Electrically-evoked compound action potentials (ECAPs) and electrical stapedial reflexes were absent.

Post-implant Auditory Habilitation

CI activation was performed after 3 weeks using behavioural-response based mapping. Over 6 months, two additional mapping sessions were conducted at intervals of 1 and 3 months, which included Map level and Map parameter optimisation. The patient underwent 10 weeks of auditory training based on Boothroyd's framework, which focuses on minimising the deficits induced by hearing loss through sensory management (through optimum fitting of hearing device), instruction (on device usage and environmental modification), perceptual training (to improve auditory skills) and counselling (to enhance social participation, emotional management and remedial strategies).⁵

Outcome Report

Table 1: Comparison of Pre-implant and post-implant outcome measures in various domains

Outcome Measure	Pre-implant		Post-implant (6 months)	
	In Quiet	In Noise	In Quiet	In Noise
Word Recognition	< 20%	Not measurable	75%	50%
Sentence Recognition	Not measurable	Not measurable	70%	60%
CIQOL-10 Global Score	14.86 ± 10.96		58 ± 6.59	
ECAPs	Absent		Present (up to 6 electrodes)	
Psychosocial Functioning	Social withdrawal		Improved motivation, mood, and social engagement	

At 6 months, ECAPs were detectable on up to six electrodes. Word recognition improved to 75% in quiet and 50% in noise, while sentence scores improved to 70% in quiet and 60% in noise. The most notable improvement was observed in the

Cochlear Implant Quality of Life (CIQOL)-10 global score, which increased from 14.86 ± 10.96 pre-implant to 58 ± 6.59 post-implant. Additionally, the subject and her family reported significant improvements in motivation, general mood, and social engagement.

DISCUSSION

Cochlear Implantation has been shown to yield meaningful gains in auditory performance in individuals with ANSD despite outcomes being less predictable in comparison to typical cochlear implant recipients with cochlear pathology.^{3,6} Some of the predictors of better outcomes reported are post-lingual onset, pre-synaptic pathology and preserved cochlear nerve integrity, while post-synaptic lesion and cochlear-nerve deficiency are associated with poorer outcomes.^{3,4}

In the reported case, the absence of sufficient clinical evidence on the site of lesion made it difficult to predict treatment outcome pre-operatively. However, post-operatively remarkable improvements in her speech perception scores from <20% to 75% in quiet and 50% in noise is consistent with some of the more encouraging outcome reports in literature.^{1,4,6}

Furthermore, benefit of cochlear implantation cannot be quantified by audiological measures alone as the multidimensional impact of this treatment can yield variable perceived benefit.⁷ Subject as well as family members in the present study reported notable improvements in her psychosocial function and mood which was quantitatively reflected in the surge in her CIQOL-10 global scores from 14.86 ± 10.96 pre-operatively to 58 ± 6.59 post-operatively. This is in alignment with most reported data on qualitative measure of benefit post-cochlear implantation, even though literature reporting CI-QOL data exclusively of individuals with ANSD is sparse.^{7,8}

CONCLUSION

Our preliminary findings suggest that cochlear implantation in adults with ANSD, combined with individualised auditory rehabilitation, can significantly improve auditory performance and quality of life. We emphasize the importance of comprehensive pre-implant counselling and expectation setting, along with tailored post-implant care, particularly in cases with uncertain prognostic indicators. A multicentric study with a larger sample size is warranted to understand the multidimensional outcome of CI in adults with ANSD across various subtypes.

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