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From cochlear microphonics to cortical silence: Audiological progression in auditory neuropathy spectrum disorder

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Abstract
Auditory Neuropathy Spectrum Disorder (ANSD) is a sensorineural hearing loss characterized by impaired neural synchrony despite preserved outer hair cell function. It presents with absent or abnormal Auditory Brainstem Response, present otoacoustic emissions, and cochlear microphonics. This case study reports a 22-year-old female with progressive Auditory Neuropathy Spectrum Disorder. Initial testing showed minimal hearing loss, robust otoacoustic emissions, present cochlear microphonics, and intact cortical responses. During the follow up testing, the patient's speech perception scores declined, acoustic reflexes and cortical responses were absent indicating deterioration in the central auditory system. The case highlights the importance of comprehensive audiological assessment including cortical measures, in diagnosing and managing the progressive Auditory Neuropathy Spectrum Disorder.

Keywords: Dyssynchrony, neuropathy, cortical test, ANSD, progressive loss

Introduction
Auditory Neuropathy Spectrum Disorder (ANSD) is a Sensorineural hearing loss caused by the dyssynchronous excitation of signals along the auditory pathway or cochlear nerve deficiency. Although the causes for ANSD is still unclear, the pathophysiology is based on the loss or dysfunction of the Inner Hair Cells (IHCs) and/or its synapses or abnormal spiral ganglion neurons or cochlear nerve deficiency with aplasia or hypoplasia of the cochlear nerve. ANSD is characterised by presence of Oto Acoustic Emission (OAE) with absent or abnormal Auditory Brainstem Response (ABR). In some cases, presence of cochlear microphonics is also observed which does not correlate with the ABR or behavioural thresholds [1]. They also exhibit poorer speech discrimination scores especially in noise as opposed with the behavioural pure tone threshold which is varied between mild to profound hearing loss. The middle ear reflexes are usually undetectable or of higher threshold in individuals with ANSD [2]. The onset of ANSD is divided across age as early onset where the onset is early in life (infancy to childhood) or develop in their adolescence or early adulthood [3]. The mean age of onset was reported to be 21.03 years of age for adolescence and adults [1]. Detailed Audiological test battery inclusive of Pure Tone Audiometry, Immittance Audiometry, Oto Acoustic Emissions and Auditory Brainstem Response is considered as the essential aspect for diagnosing ANSD. Additionally Late Latency Response (LLR) can also be performed to measure the integrity of the auditory system beyond the level of brainstem. Management of ANSD typically involves hearing aids as the first line of management. Cochlear Implantation can also be performed for individuals with limited benefit from hearing aids. Recently FM technologies also have shown to provide speech perception benefits for individuals with ANSD. This case study discusses the degree of progression of the degree of dys-synchrony over time and its impact in audiological findings.

Case History
A 22-year-old female presented with a complaint of difficulty in understanding speech in noisy environment and tinnitus perception in both ears for a period of 5 months.

She had undergone audiological testing and radiological assessment before 2 months for the same concern. Results revealed both ears hearing sensitivity within normal limits in Pure Tone Audiometry (PTA) and Both ears 'A' type tympanogram. Magnetic Resonance Imaging (MRI) of the brain was performed which revealed the appearance and intensity of the brain are normal. The patient was referred to Madras ENT Research Foundation P(Ltd.) for an ENT consultation as the concern was not resolved, and the patient was advised for a detailed audiological test battery to explore the potential underlying cause for the same and to obtain a more comprehensive understanding of her auditory difficulties.

2.1 Audiological Profile

The patient underwent a comprehensive audiological evaluation. Pure Tone Audiometry revealed minimal hearing loss in the right ear (PTA-21.66 dBHL), hearing sensitivity within normal limits in the left ear (PTA-13.33 dBHL). Speech audiometry findings revealed Speech Recognition Threshold (SRT) at 30 dBHL, Speech Identification score of 80% and Uncomfortable Level (UCL) was obtained at 100 dBHL in right ear and SRT at 25 dBHL, SIS score of 80%, UCL was obtained at 100 dBHL in the left ear. Tympanometry yielded Type 'A' tympanograms in both ears across visits, suggesting normal middle ear function, and ipsilateral acoustic reflexes were present. Distortion Product Otoacoustic Emissions (DPOAE) were robust in both ears, indicating intact outer hair cell function. Auditory Brainstem Response (ABR) testing showed clear and replicable cochlear microphonics (CM) in both ears, with phase reversal between rarefaction and condensation polarities. However, Peak I could not be obtained due to CM dominance, while Peak III was robust (4-5 ms), and Peak V was present but of low amplitude (6-7 ms). Cortical Auditory Evoked Potentials (CAEPs) initially revealed the presence of the P1-N1 and P2-N2 complexes within normal latency in both ears. Tinnitus Matching and Masking tests showed a matched tone at 1 kHz in the left ear, while no match was found in the right. Masking in the left ear failed to produce residual inhibition. The Tinnitus Handicap Inventory score indicated a severe handicap (Grade 4). Speech-in-noise testing demonstrated poor performance with scores of 48% (right) and 64% (left) at 0 dB SNR, 80% and 72% at +10 dB SNR, and significantly reduced scores of 12% and 32% at-10 dB SNR, respectively. The Gap Detection Test using a three-interval forced-choice paradigm revealed the patient's inability to detect temporal gaps, suggesting poor temporal resolution. An FM system trial resulted in improved speech perception, with SIS scores of 85% in quiet and 75% in noise.

During a follow-up evaluation, PTA revealed a progression to mild sensorineural hearing loss in the right ear (PTA: 28.33 dBHL) and minimal hearing loss in the left (PTA: 18.33 dBHL). Speech audiometry showed a decline, with SRTs of 35 dBHL (right) and 30 dBHL (left), and SIS scores of 70% and 72%, respectively. UCL remained stable at 100 dBHL in both ears. Reflexometry in this session revealed absence of both ipsilateral and contralateral reflexes bilaterally. Although DPOAEs remained robust, repeat CAEP testing failed to elicit the P1-N1 and P2-N2 complexes in either ear, indicating a deterioration in cortical auditory processing, correlating with the patient's continued difficulty in speech perception (as shown in figure 1).

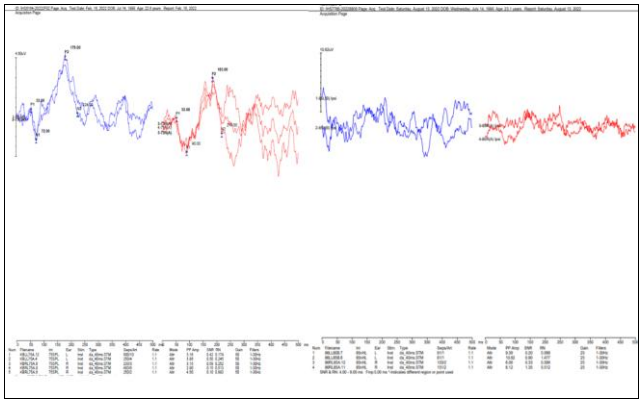

Fig 2: Cortical auditory evoked potentials (CAEPs) of the subject. Red indicates responses from the right ear; blue indicates responses from the left ear.

Discussion

Auditory Neuropathy Spectrum Disorder (ANSD) is a complex auditory disorder that requires a comprehensive diagnostic approach. Traditional diagnostic protocol includes Pure Tone Audiometry (PTA), Impedance Audiometry, Acoustic Reflexes, Otoacoustic Emissions (OAE), and Brain Stem Evoked Response Audiometry (BERA). In the present case, addition of cortical measure, Cortical Auditory Evoked Potential (CAEP) provided a clearer diagnostic picture. Although ABR results remain unchanged over a period of six months, the P1-N1 and P2-N2 complexes that were observed in the initial visit were absent during the follow up visit indicating deterioration at cortical level. This suggests that CAEP can be a valuable addition to the ANSD diagnostic battery, especially for tracking disease progression [4]. A characteristic finding in the later stages of ANSD is the absence of acoustic reflexes, despite the presence of Type 'A' tympanograms [5]. This was mirrored in the current case, where initial ipsilateral reflexes were present but became absent over time, while tympanometry remained unchanged. The presence of cochlear microphonics (CM) with absent ABR waveforms especially when confirmed using alternating stimulus polarities is widely regarded as a reliable indicator of ANSD. Literature supports the diagnostic value of CM, with studies such as Shi *et al*. (2012) documenting its presence in patients with absent ABRs [6]. ANSD patients typically show preserved OAEs and CM, absent or highly distorted ABRs, and lack of acoustic reflexes. Audiometrically, they may present with permanent or fluctuating hearing loss, often with a flat or ascending configuration and marked speech perception difficulties in noise [7]. Though OAEs may be initially present, they can diminish over time. Additional findings such as absence of the OAE suppression effect due to efferent auditory pathway dysfunction also support an ANSD diagnosis. CM presence remains the most definitive diagnostic marker, often persisting longer and appearing more prominently than in normal individuals. CM durations can extend to 4-6 ms in ANSD patients, potentially leading to misinterpretation as brainstem activity unless verified through stimulus polarity reversal [8].

Conclusion

These findings highlight the importance of detailed electrophysiological assessment like CAEP in diagnosing and monitoring of Auditory Neuropathy Spectrum Disorder as it increases the diagnostic sensitivity. Also, in this patient

the progression of the condition was observed within 6 months of duration highlighting the need for early identification and timely intervention.

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