International Journal of Health Sciences and Research
Vol.13; Issue: 4; April 2023
Website: www.ijhsr.org
Case Report *ISSN: 2249-9571*

Auditory Neuropathy Spectrum Disorder: Diagnosis, Decision and Consequence

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DOI: https://doi.org/10.52403/ijhsr.20230428

ABSTRACT

Auditory Neuropathy Spectrum Disorder (ANSD) is a type of disorder where the signal transmission from auditory nerve to brain is often impaired thus affecting the speech perception. The prevalence of ANSD is quite variable. It is often characterized by the presence of otoacoustic emissions with abnormal or absent Auditory evoked brainstem response. Early stages of hearing testing and appropriate diagnosis can aid in early intervention. Delay in diagnosis can delay the intervention process worsening the auditory perception in noise and quiet conditions thus curbing the academic, social and occupational functioning of an individual. Hearing loss in individuals with ANSD can vary from normal to profound and thus the choice of amplification should be decided considering the degree of hearing loss. Comprehensive audiological evaluation and best choice of management along with auditory training can help improve the speech perception in individuals with ANSD. The present study outlines the audiological findings of a 12-year-old boy who had a delay in diagnosis of ANSD. It also summarizes how choice of management can impact hearing and auditory perception over years thus highlighting the need for consideration of hearing thresholds during choice of amplification.

Keywords: Auditory Neuropathy Spectrum Disorder (ANSD), Hearing aids, Early identification, Test battery, Amplification

INTRODUCTION

Auditory Neuropathy Spectrum Disorder (ANSD) is a type of hearing disorder that encompasses a range of hearing disorders in which sound enters the inner ear normally but the transmission of signals from the inner ear to the brain is impaired. A dysfunction at any level of the auditory pathway may disrupt the coding of acoustic features, especially temporal features. Based on the site of dysfunction, ANSD can be subdivided into presynaptic and postsynaptic. ANSD is often idiopathic, however a wide variety of pre- and post-natal factors including prematurity, cochlear malformation, damage to auditory nerve, genetic mutations, infections could also be possible causes [1]. The estimated incidence of ANSD identified through Universal Newborn Hearing Screening (UNHS) in well babies is 0.09/1000 live births [2]. Audiological indications of ANSD includes difficulty in perception of speech in noise, hearing threshold varying from normal to profound, disproportionate speech perception scores and hearing thresholds, presence of robust otoacoustic emissions and presence of cochlear microphonics in Auditory Brainstem Response. Based on the severity of ANSD, presence of otoacoustic

emissions and presence of P1-N1 & P2-N2 complex in Cortical Auditory Evoked Potential becomes questionable. Management options for ANSD typically involve two technologies, hearing aids (low power) and cochlear implantation. However, FM technologies have recently shown to have significant benefit in speech perception for individuals with ANSD [3]. Decision making on the management option should be made considering the hearing threshold, outer hair cell functioning and the degree of dys-synchrony. However, recommending low power hearing aids with auditory training can be provided as a first step in the line of management. Here we present a patient diagnosed with Auditory Neuropathy Spectrum Disorder highlighting the impact of the amplification system in progression of the degree of dys-synchrony worsening the speech perception because of acoustic overstimulation.

CASE REPORT

A twelve-year-old male presented with the concern of difficulty in understanding speech. At 8 years of age the patient was diagnosed to have mild conductive and moderate conductive hearing loss in right and left ears respectively using a subjective test (Pure tone audiometry) and was under medical intervention for the same. At 10 years of age, the child underwent electrophysiological audiological assessment which revealed absent Auditory evoked brainstem responses in rarefaction polarity and present Otoacoustic emissions. The child was fitted with high gain hearing aids in both ears based on ABR findings and has been using the same for the past 2 years. However no significant benefit was reported. Medical history revealed that the child was diagnosed with Attention Deficit Hyperactivity Disorder (ADHD) and mild receptive speech delay. He was also known to have complaints of anger and aggressiveness for which he was reported to be upon medication under Psychiatrist advice for the past 4 years. He also had concerns with his flow of speech with complaints of stuttering and tachyphemia.

AUDIOLOGICAL AND SPEECH PROFILE

The patient was subjected for a detailed audiological evaluation. Pure Tone Audiometry (PTA) testing revealed Severe to profound hearing loss in both ears (PTA average >90 dBHL). Speech Audiometry testing could not be done because of audiometric limitations. To rule out middle ear dysfunction, tympanometry was performed which revealed 'A' type tympanogram in both ears with absent reflexes indicating normal middle ear functioning. Absent reflexes could be attributed to the presence of severe to profound hearing loss in both ears. Electrophysiological test findings revealed presence of clear and replicable cochlear microphonics with no replicable peak V in low rate (11.1/sec) using rarefaction and condensation polarity. Cochlear microphonics was absent when the polarity was shifted to alternating, which confirmed the presence of cochlear microphonics in rarefaction and condensation polarity (Figure 1). Distortion Product Oto Acoustic Emission testing and Cortical Auditory Evoked Potential testing revealed Absent DPOAEs and absence of P1-N1 & P2-N2 complex respectively (Figure 2). Hearing Aid Trial was performed using the patient's own hearing aid and it revealed the aided responses were within the speech spectrum, however the speech perception score was poor (50%). FM trial was performed in both quiet and noisy environments which revealed 66% of Speech Identification Score in both quiet and noise conditions. Even though speech perception scores through FM trial are better in comparison with Hearing aids it still indicates moderate difficulty in speech perception. With Severe to Profound hearing loss in both behavioral and objective testing and poor hearing aid benefit for speech perception, the child was identified to be a candidate for Cochlear implantation. He was advised on the same

and was counseled on importance of auditory training post audiological management.

Figure 1 shows the Auditory brainstem responses of the subject.

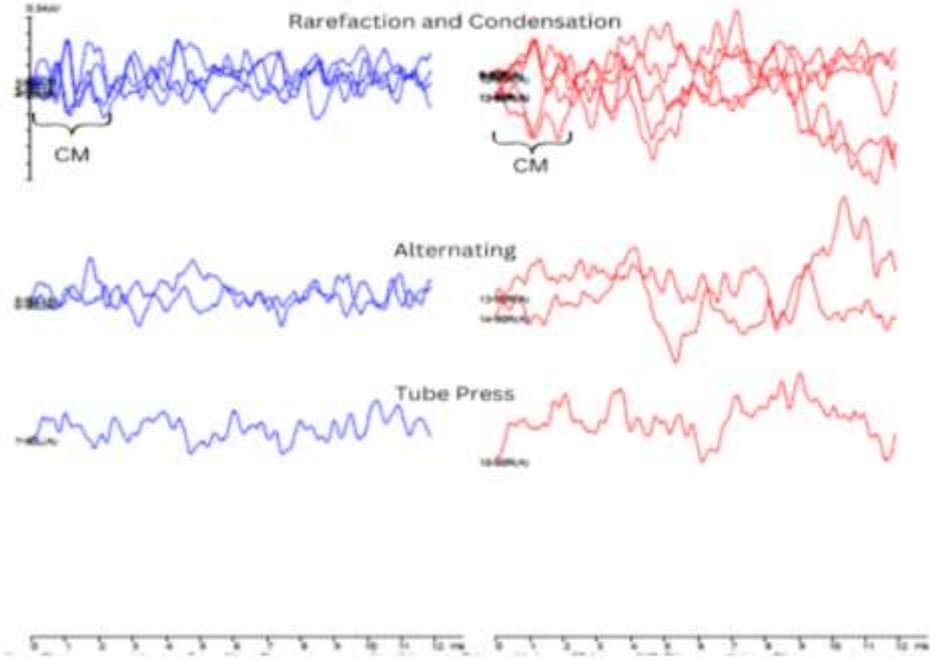

Figure 1 Auditory brainstem responses obtained from subject. Clear replicable cochlear microphonics (CM) is noted in both ears in both rarefaction and condensation polarity. No CM noted in alternating polarity or responses recorded using tube press method. Red indicates responses from the right ear, blue indicates responses from the left ear.

Figure 2 shows the unaided cortical auditory evoked potentials of the subject.

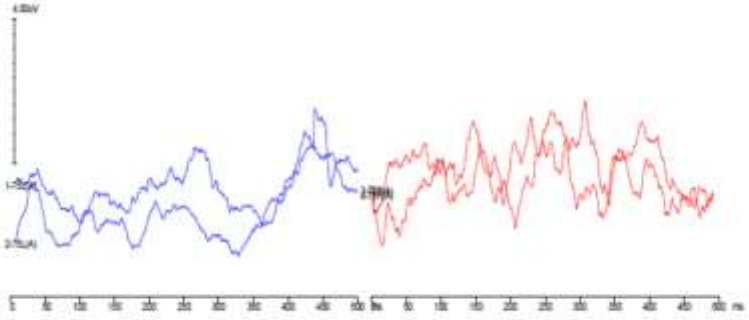

Figure 2 Unaided cortical auditory evoked potentials (CAEPs) of the subject. No CAEPs were recorded in both ears. Red indicates responses from the right ear, blue indicates responses from the left ear.

Speech & language evaluation was conducted and the patient was observed to show signs of stuttering & cluttering. His speech was observed to be dysfluent with presence of stuttering like dysfluencies such as blocks, revisions, part-word, whole-word & phrase repetitions of 3-5 iterations. His rate of speech was observed to be increased with articulatory errors. Truncation of words & sentences, frequent shifts in topics, lack of prosody with poor self-monitoring skills were noted. On behavioral observation, features of hyperactivity were also noted. Formal evaluation was conducted with appropriate test tools and the patient was diagnosed with Cluttering associated with Stuttering of Moderate severity and Expressive Language Disorder.

DISCUSSION

The auditory difficulties and complaints associated with ANSD are vast, necessitating a distinct pattern of audiometric tests for diagnosis. Audiologists play a major role in diagnosis of AN through a comprehensive audiological test battery. Test battery includes Pure tone Audiometry, Immittance Audiometry, Otoacoustic emissions, Brainstem evoked and Cortical Evoked potentials, Speech perception in noise and additional temporal resolution or central auditory processing tests as per the indication.

Audiograms obtained through behavioral testing are often disregarded, as audiometric thresholds are known to vary in individuals with ANSD and objective tests are considered reliable in testing young children. Hence, the primary indicator considered for ANSD are presence of robust OAEs with abnormal or absent Brainstem evoked potentials. The present study summarizes the audiological findings of a 12-year-old male child who had concerns with difficulty understanding speech (Table 1). He had undergone a detailed assessment protocol. On reviewing his previous medical records, it was noted that the child was diagnosed to have severe to profound hearing loss based on objective electrophysiological (ABR) testing and was fitting with high gain power BTE hearing aids in spite of presence of DPOAES in both ears and behavioral audiometric results indicating mild and moderate conductive hearing loss in right and left ear respectively. Even though Brainstem evoked potentials are absent, the presence of OAEs at the age of 8 years is an indicator for near normal to normal inner ear health [4]. However, the child was fitted with high power hearing aids. The current audiological findings indicated severe to profound hearing loss with absent DPOAEs in both ears. The use of high-power hearing aids could have resulted in acoustic overstimulation resulting in deterioration of DPOAEs and behavioral audiometric thresholds over years which is supported by literature [5]. Although amplification seems successful in few ANSD cases, appropriate fitting strategy should be determined considering the results of all tests including behavioral tests to avoid detrimental results. Hence, it is suggested that even in young children in addition to objective tests, informal hearing assessment, that is, looking for child's behavioral responses for environment sounds and behavioral audiometric thresholds should be considered in hearing aid fitting rather than fitting based only on objective test results. The presence of CM which is another indicator for ANSD can be appreciated if ABR was recorded in all polarities. Previous objective testing included auditory evoked responses obtained in only one polarity. Hence, the appreciation of cochlear microphonics and its confirmation were difficult as this could usually be confused with artifacts resulting in missing of ANSD diagnosis. Thus, it is suggested to follow a detailed protocol while recording auditory evoked responses for early identification of ANSD which includes responses obtained in both rarefaction, condensation polarity and a confirmatory response through responses recorded using alternating polarity and performing a tube-press method. Middle ear testing consisting of Tympanometry and Reflex audiometry could also supplement in the process of diagnosis. Even though absent stapedial reflexes are not a primary marker for ANSD, the presence of stapedial reflexes would help in ruling out ANSD [6]. Additional tests evaluating the speech perception can also be performed to obtain an overall picture of the speech perception in individuals with ANSD. The tests can include Speech perception noise test and Gap detection test. Early ANSD diagnosis and appropriate management could have prevented further worsening of hearing thresholds and thus the speech perception.

Speech perception can be adversely affected due to reduced sensitivity to follow fast and slow temporal modulations which can be identified through special tests such as Gap detection and Frequency discrimination

tests. Decreased sensitivity over temporal cues can in turn have an effect in monitoring speech. Auditory temporal processing was suggested to be critical in fluent speech production and thus literature has revealed auditory temporal deficits in children with stuttering [7]. The diagnosis of stuttering and cluttering in this subject contributes to the factor that ANSD can have a role in disrupting fluency due to interference in the auditory feedback loop. Appropriate clinical management including auditory and temporal training for ANSD can help in alleviating speech related concerns. However, this could not be noted in this patient as the patient did not visit for regular follow ups for both speech and audiological intervention.

Table 1 Summary of Audiological test protocol followed for the present ANSD case.

	Test	**Criterion**	**Current case findings**
Behavioral	Pure Tone Audiometry	Variable - can range from normal to profound hearing loss; Can have better thresholds which may worsen over time; Fluctuating hearing thresholds	Severe to Profound Hearing loss
	Speech Recognition test	Variable but generally poor speech perception scores	CNT due to audiometric limitations; Informal observation - child was not able identify words in open & closed set presentation.
	Speech perception test in noise	Poor speech perception scores	CNT
Physiological	Tympanometry	'A' type tympanogram	'A' type tympanogram
	Reflex Audiometry	Absent stapedial reflexes; Can rule out ANSD if reflexes are present	Absent stapedial reflexes
	Distortion Product Otoacoustic emissions (DPOAEs)	Robust DPOAEs during initial phases of ANSD Can diminish with worsening auditory thresholds	Absent DPOAEs
Electro-physiological	Auditory evoked Brainstem Responses in rarefaction, condensation & alternating polarity followed by tube-press method	Presence or absence of Jewett peaks with presence of Cochlear microphonics	Replicable cochlear microphonics (CM) in rarefaction & condensation polarity Absent CM in alternating polarity & tube press method Absent Jewett peaks in all 3 polarities
	Cortical evoked auditory potentials	Presence or absence of peaks based on severity of speech perception difficulty	Absent P1-N1 & P2-N2 complex
Temporal processing test	Gap detection test	Poor scores	CNT

Note: ANSD - Auditory Neuropathy Spectrum Disorder; CM - Cochlear microphonics; CNT - Could not be tested

CONCLUSION

Early diagnosis and appropriate intervention can result in better outcomes or at least can stall the worsening of symptoms. It requires sound knowledge in correlation of tests for early diagnosis thus aiding in appropriate early intervention. Considering the behavioral responses is primary in hearing aid fitting. Consequences of compromised hearing and auditory perception deficit due to auditory dys-synchrony could be the contributing factors for delayed speech & language development associated with stuttering & cluttering. Hence, parental guidance and intensive auditory based intervention is needed for better outcomes.

Declaration by Authors

Acknowledgement: None

Source of Funding: None

Conflict of Interest: The authors declare no conflict of interest.

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How to cite this article: Mathew Nishanth A, Jaya Lakshmi P B, Anusha Rao S et.al. Auditory neuropathy spectrum disorder: diagnosis, decision and consequence. *Int J Health Sci Res.* 2023; 13(4):223-228.
DOI: *https://doi.org/10.52403/ijhsr.20230428*

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