

## Amplification Options in Children with Auditory Neuropathy Spectrum Disorder

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### Abstract

Auditory neuropathy spectrum disorder (ANSD) is a specific form of hearing loss characterized by the presence of normal or near-normal Otoacoustic Emissions (OAEs) but the absence of normal middle ear muscle reflex and severely abnormal or completely absent auditory brainstem response (ABRs). Amplification is considered a dilemma in such disorder with a lot of debates. This article will review hearing aids and cochlear implant as options for management in ANSD.

**Keywords:** Auditory neuropathy spectrum disorder; Hearing aids; Cochlear implant

### Introduction

Auditory Neuropathy spectrum disorder (ANSD) is a specific type of hearing loss and constitute about 10-15% of children diagnosed with sensorineural hearing loss [1-3]. Clinically, those patients exhibit normal outer hair cells function as measured by robust otoacoustic emissions (OAEs) and cochlear microphonics (CM) which invert with reversal of the stimulus polarity and abnormal acoustic reflexes (AR) [4-6]. However, the neural synchrony is deficient as revealed by absent or profoundly abnormal auditory brainstem response (ABR) [1].

The most common causes of ANSD in children include: perinatal conditions (e.g; hypoxia, hyperbilirubinemia), infectious disease (such as mumps, meningitis), immune disorders or genetic causes whether syndromic (e.g. Charcot-Marie-Tooth disease) or non-syndromic (e.g. Otoferlin (OTOF) genetic mutations); head injury (e.g. Shaken baby syndrome) [7,8].

Starr et al., [9] suggest that the clinical picture of ANSD accompanying disorders of distal components of the auditory periphery (terminal dendrites, inner hair cells, synapses) be designated as a distal AN or type II AN. On the otherhand, ANSD with involvement of the ganglion cells, axons, and proximal dendrites be designated as a proximal ANSD or type I ANSD.

Hearing levels in ANSD are quite variable ranging from normal hearing up to total hearing loss. However, the major physiologic deficit in ANSD is dys-synchronous firing of the auditory nerve and auditory brainstem neural pathways [10-12]. Such a neural dys-synchrony apparently leads to significant perceptual deficits, particularly in temporal coding of acoustic information (i.e., temporal resolution) [13,14], which in turn leads to poor speech perception, especially in noisy situations. In addition, the abnormal pattern of neural firing in the subcortical auditory pathways can disrupt normal central auditory maturation with increased risk for cognitive impairments [11,15].

Those patients typically have poor results in measures of speech perception test than their behavioral audiograms would predict

[16,17]. Speech understanding in ANSD patients is worsen in the presence of competing signals, such as background noise [11,12,18]. Patients with ANSD also have deficits in frequency discrimination [16]. The deficits could be due either to impaired tonotopic cochlear representation or due to impaired temporal precision and synchronization, which could account for speech recognition deficits that are disproportional to the hearing loss. Gabr [19] reported impaired frequency discrimination in individuals with AN/AD specially at 1000 Hz which is very important speech frequency lying in the middle of the most sensitive part of the audible frequency range the speech spectrum [20].

### Management

Management of ANSD showed a continual controversy and represents a great challenge to audiologists. ANSD is a disease of dys-synchrony with fluctuation in pure tone thresholds and poor speech perception scores which are not corresponding to the levels of hearing loss [21,22]. Moreover, the severity of dys-synchrony is not related to the severity of the hearing loss and cannot be characterized easily with behavioral measures [23,24].

Given these physiologic and perceptual vulnerabilities, some interventions are used to ameliorate the negative effects of ANSD. Among these interventions are the hearing aids (HAs) and cochlear implants (CI). There is a general recommendations that children diagnosed with ANSD can be treated with appropriate hearing aid (HA) amplification when reliable behavioral thresholds demonstrate a significant hearing loss [25]. When those patients showed poor progress in speech and language development despite appropriate amplification, cochlear implant (CI) should be considered [26,27].

Audiological management of infants with ANSD is often dependent on behavioral hearing thresholds using standard audiometry [1]. This is because electrophysiological methods (ABR, ASSR) are compromised in the majority of cases with the dys-synchronization and do not predict auditory detection thresholds. Behavioral assessment of pure-tone thresholds using developmentally appropriate, conditioned test procedures such as visual reinforcement audiometry (VRA), or conditioned orientation reflex (COR) audiometry. For very

young or developmentally-delayed infants, behavioral observation audiometry (BOA) may be used to observe the infant's reflexive response to sound, however, results should not be interpreted as representing behavioral thresholds or minimal response levels. Speech reception, recognition or detection measures can be used depending on the age and vocabulary of the patients. Obligatory cortical auditory evoked potentials to speech or speech-like signals can be used as an objective clinical tool for predicting speech recognition performance in young children with ANSD [17,28].

It has to be noted that the presence of additional cognitive impairments in some children with ANSD may result in a prolonged and more complicated process of threshold determination. So, amplifications might be postponed until we get a reliable hearing threshold. This results in delays in amplification and greater amount of time without adequate audibility of speech signal [29,30].

### Hearing aids

When an infant or young child with ANSD demonstrates elevated pure-tone and speech detection or recognition thresholds with consistent test-retest reliability, hearing aid fitting should be considered as soon as possible and a trial use of amplification should be offered [31].

The presence of residual auditory function was a considerable factor limiting early enthusiasm for cochlear implantation (CI) in the ANSD population as many patients have mild-moderate audiometric thresholds. Moreover, a subset of ANSD patients may exhibit auditory brainstem response (ABR) responses with age as a result of neuromaturation [32]. For these reasons, hearing aids (HAs) were initially recommended as a primary management strategy in this population [33].

Arguments have been made against hearing aid use in children with ANSD as long as OAEs are present to preserve OHC function and postpone HA fitting till OAEs disappear. Other audiologists recommended the restoration of audibility through HAs with proper gain and frequency prescription depending on patients' behavioral thresholds as in other cases with sensorineural hearing loss [31]. Early trials of clinical management of ASND focused on the use of low gain hearing aids or FM systems; low gain hearing aids in one ear only; or the avoidance of hearing aid use altogether [34].

Zeng et al. [35] suggested that because temporal processing appears to be affected in patients with AN, amplitude compression should be avoided and linear amplification is considered. Another method proposed to expand the amplitude of the temporal modulations. Adjusting the release time for compression to a slower rate allows for greater amplitude differences in the temporal modulations. These enhanced amplitude differences help provide salient cues for consonant and vowel perception. The enhanced amplitude difference also increases perception of speech events, such as voice onset time, burst, and transitions, and helps to maximize signal-to-noise ratios [35-37]. Another recommendation was to filter out low frequency signals or shift them to the high frequency ranges via frequency transposition [35] and temporal envelope enhancement [38].

Some infants and children with ASND may show a significant improvement in auditory function, including "recovery" from ANSD [32,39]. So, careful monitoring of their auditory function by ABR and behavioral response by conditioned test procedures is required to adjust and modify amplification as needed [26].

The hearing aid fitting alone, of course, is not the end and several factors should be considered to get successful hearing aid fitting. This included: the age at diagnosis, treatment, technology and regularity of HA use, proper rehabilitation and speech therapy programs, family involvement, and the child's other diagnoses or developmental delays all play into the overall picture.

### Cochlear implants

It was initially thought that cochlear implantation would be of no benefit in ASND due to its association with nerve degeneration secondary to processes such as demyelination and axon impairment. However, [40] consider the idea that cochlear implantation might be an effective intervention as electrical stimulation of demyelinated nerves in mice resulted in a measurable auditory brainstem response (ABR) waveform. It has also been shown that if a nerve fiber is stimulated electrically, both the growth of discharge rate and the maximum rate achievable are greater than when stimulating the nerve acoustically. In addition the timing of the nerve response to electrical stimulation seems to be more precise and repeatable than it is for acoustic stimulation [41]. More recent studies have shown that many children with ANSD benefit from cochlear implantation [32,42,43].

Many studies demonstrated similar postoperative performance after implantation between children with ANSD and those with sensorineural hearing loss (SNHL) [35,44]. Based on this evidence, cochlear implantation has been advocated as a feasible treatment for AN. However, some reports show poorer or variable outcomes which were attributed to uncertainty of the lesion location outcomes [eg: 45,46].

Once implanted, programming speech processor should be customized in individual manner depending on each child condition. Many children can do well with the default parameters of software, but others need to be given other fitting considerations in their mapping. Among these consideration is slowing the stimulation rate. This procedure is easy and allow longer refractory period for the auditory nerve fibers with subsequent enhancement of neural synchrony [33]. Another important parameter is widening the pulse width to get adequate stimulation. However, we should avoid too much widening to avoid compromising spectral and temporal resolution [47].

Another thing that could be done is trying to measure the loudness growth. However, lack of auditory experience with sounds or very young age in some children may compromise this technique. The use of picture corresponding to sounds varying from very soft to loud sounds can be helpful to scale loudness and optimize the true electrical dynamic range of that child [48]. Pitch ranking, aims to replicate the tonotopic organization of the cochlea to take full advantage of that to eliminate some channels that are perceptually the same. Finally, can use strategies that create virtual channels when children can participate more in testing and the audiologist can get more information from them [49].

All of these techniques can be implemented through the programming software, but they are not the default settings. It certainly will take more time to create a program when you are manipulating those defaults.

Many studies reported significant hearing improvement in hearing discrimination, pure tone detection thresholds, and recognition of words and sentences in ANSD patients fitted with CI. Moreover, Peterson et al., and Yamaguti reported improvement in sentence

understanding after electrical stimulation of the auditory nerve through the use of CI in those patients.

## Conclusions

ANSD is a complicated pathology and no single approach can be used for the management of all children. Some children will benefit from hearing aids after short term or long term use. However, careful monitoring of the child's progress in auditory communication is needed. Many children will require cochlear implantation and clinical experience showed dramatic improvement in speech perception ability. In addition to standard CI criteria for children, special consideration children with ANSD include: First, CI consideration with stable auditory test results with evident permanent ANSD. Second, Auditory nerve sufficiency should be obtained by appropriate imaging techniques prior to implantation. Third, for children with ANSD who do not demonstrate good progress in speech understanding ability and aural/auditory language development, CI should be considered, regardless of behavioral audiometric thresholds.

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