

Reflections on Auditory Neuropathy Spectrum Disorder: 2013

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Auditory neuropathy spectrum disorder (ANSD) is a fascinating audiological diagnosis. Fortunately, knowledge and publications (addressing ANSD) continue to increase exponentially. The hallmark audiological signs of ANSD are the presence of outer hair cells (represented by normal OAEs perhaps 75 percent of the time, or a normal cochlear microphonic response) and an absent/abnormal auditory brainstem response (ABR). ANSD is presumed to occur at the junction of the spiral ganglion cells and the auditory nerve (Nachman, 2012) and is most often suspected and diagnosed based on (see Picton, 2011 and Berlin, 2012) the child's birth and related medical history and the results from multiple behavioral and electrophysiologic tests (such as ABR, OAE, acoustic reflexes, word recognition scores, speech perception in noise, etc.). It appears that 90 to 95 percent of all cases of ANSD are bilateral (Picton, 2011). ANSD may be present in some 10 to 15 percent of all children with hearing loss and ANSD may be present in higher percentages (15 to 20 percent) among children with severe-to-profound hearing loss (Berlin, 2012 and Hood and Morlet, 2012).

It is particularly noteworthy that 90 to 95 percent of all people with ANSD will not have present acoustic reflexes. Of particular importance, Berlin (justifiably) argues all hearing screenings should include tympanometry, acoustic reflexes and OAEs. Indeed, it is important to note there is no such thing as congenital absence of the stapedial reflex (see Berlin, 2012). That is, when a clinician tests a child (or adult) with apparent normal hearing thresholds and no acoustic reflex, it can be argued these findings represent ANSD until proven otherwise! However, if the acoustic reflex has not been tested, the suspicion and diagnosis (of ANSD) will likely be missed. Further, ANSD can masquerade as auditory processing disorder (APD) in children with normal hearing thresholds and poor performance with word recognition—particularly in noise, and ANSD may masquerade as an acoustic neuroma in adults with similar presentations (normal hearing thresholds, poor performance in noise, absent/abnormal ABR).

Additionally, of significant importance and unfortunately less well known (among practicing otolaryngologists and audiologists) is the probability that the majority of children with ANSD may have additional intra-cranial anomalies. Specifically, Roche et al (2010) reported 118 children diagnosed with ANSD (who had either CT

and/or MRI scans). Of those, prematurity was the most common historic finding (reported in 42 percent of their subjects) and other less prominent "risk factors" included NICU admission, mechanical ventilation, and hyperbilirubinemia. Roche et al reported that of the children who received MRIs, some two-thirds had at least one intra-cranial abnormality and 41 percent had two or more abnormalities. Of the children who received CT scans, 55 percent had at least one abnormality and 45 percent had two or more.

Nachman (2012) reported a child with brainstem juvenile pilocytic astrocytoma (JPA) who presented with normal pre and post-natal history, but had failed a screening automated ABR. Diagnostic ABR and OAE suggested left unilateral hearing loss consistent with left ANSD. ENT evaluation revealed no remarkable findings and neuro-imaging was not ordered at that time. At 2 years, 8 months reliable behavioral hearing threshold results were determined to be within normal limits (WNL). Approximately one year later, a mild low frequency neural hearing loss was noted in the left ear, presumed to be ANSD. By age 4 years, 10 months the left hearing loss had progressed to a moderate low frequency hearing loss. At 4 years, 11 months the child was admitted to the hospital with debilitating headaches and vomiting. MRI (and intraoperative findings) revealed a JPA at the root entry zone of the eighth nerve—admittedly a very rare finding.

Of note, auditory (re)habilitation (AR) of children with ANSD has been (and clearly remains) controversial. That is, the recommendation of hearing aid amplification versus cochlear implantation is a difficult and likely pivotal decision—particularly for children with normal (or nearly normal) hearing, who are not progressing with regard to their speech and language skills. Of additional concern are the very rare-but-well-documented cases in which ANSD reversal (i.e. normalization) occurs in some children (Hood and Morlet, 2012). For the clinical team managing children with ANSD, the options are vast and the choices are of critical importance.

Nonetheless, as I reflect on and consider the ever-changing contemporary publications and findings related to ANSD in children, it's clear (to me) that "keeping up" is extremely time consuming and challenging. Given the contemporary publications cited here (and many others), I believe it makes sense (and I urge all audiologists) to incorporate acoustic reflex testing into *all* hearing screening batteries. I believe we must assuredly manage ANSD as a team member only. That is, the suspicion, diagnosis, and management of the child with ANSD mandates involvement of the speech-language pathologist, as well as the parents and the physician, to evaluate the child with regard to a global perspective. Finally, it seems to me, we (audiologists) should urge neuro-

imaging studies for all children diagnosed with ANSD, as many other professionals (pediatricians, otolaryngologists, family practitioners...) are likely to be unaware of the work cited above by Roche et al.

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For More Information, References, and Recommendations

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