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The Role of Educational Audiologists to Support Students with Auditory Neuropathy Spectrum Disorders

Auditory neuropathy spectrum disorder (ANSD) has been described as "…a range of hearing impairments characterized by deteriorated speech perception, despite relatively preserved pure-tone detection thresholds" (De Saiti et al, 2020). The classic assessment profile includes abnormal auditory brainstem responses (ABRs) but the presence of acoustic emissions (OAEs). De Saiti et al go on to suggest that "these electrophysiological characteristics have led to the hypothesis that ANSD may be caused by various dysfunctions at the cochlear inner hair cell (IHC) and spiral ganglion neuron (SGN) levels, while the activity of outer hair cells (OHCs) is preserved, resulting in discrepancies between pure-tone and speech comprehension thresholds". It was first described in the clinical literature by Starr and colleagues in 1996, who used the new technology in OAE testing to demonstrate that significant auditory dysfunction could occur in the presence of functioning outer hair cells in the cochlea. While our definitions describes the clinical assessment profile, they do not capture the range of functional presentations and communication outcomes, from children who have very good speech and language communication to those who appear to receive no benefit whatsoever from auditory information. An individual's auditory performance can vary greatly across contexts (e.g., quiet vs. noise) and even from day to day. Add in the fact that AN can be overall stable, progressive, or, in some cases, improve over time, and the challenges in managing ANSD are clear.

Even today, they remain amongst the most challenging students because of their behavioral variability, the fact that the student's audiogram is of little to no help in guiding an intervention plan, and the lack of clear clinical guidance on management (and perhaps its unavailability). In fact, the advice given to families and educators is essentially to try "everything" and see what

works. Educators are very comfortable with the idea that understanding student needs requires flexibility, patience, careful assessment (and frequently, creativity). Still, they need to understand first what ANSD is and, second, that the information educational audiologists and teachers of the deaf have been giving them about sensorineural and conductive hearing loss may not apply to ANSD. Educational audiologist Sandra Vandenhoff, PhD, recently created an excellent analogy to understand ANSD using the example of The Wave in a sports stadium. My own analogy for my York students has been to describe ANSD as driving on a large highway in Toronto, where you may be driving along smoothly at the speed limit with traffic, then suddenly all traffic grinds to a halt. A minute later, things speed up a bit, then stop again, yet somehow cars in the lane beside you are moving at the speed limit while you wait, stopped in your lane, for no apparent reason whatsoever. Both analogies emphasize the role of timing in the efficient flow of information, something which is much more difficult to understand than frequencies, decibels and the speech banana.

The “spectrum” of ANSD

The traditional classification system for degree of hearing loss (e.g mild, moderate, moderately severe, etc.) is not generally used for ANSD; however, audiologists, families and school staff who have supported these students likely have a sense of functional “degree” of auditory neuropathy in terms of impact on speech, language and communication development. Profiles of 3 students with whom I have worked over the years demonstrate the breadth of the spectrum and the need for school staff to keep an open mind when considering educational options.

Student A. The first student was 8 years old with a congenital severe to profound hearing loss, diagnosed primarily by ABR at 3 years of age. The student was fit with hearing aids but had never tolerated them, and generally showed no response to sound with amplification. Curiously, however, there were reliable reports from parents and school staff of the child responding to something like a bird chirping without hearing aids, surely impossible with an audiogram in the profound range. The student communicated through Signed English, but had no intelligible speech. Eventually, the astute and patient clinical audiologist at the student’s residential facility obtained an audiogram showing reliable responses at 20 dB across frequencies. The immediate question was “what is the appropriate educational placement and communication methodology for a student who has both an audiogram showing thresholds at 100 dB and an audiogram showing thresholds at 20 dB”? In fact, the school for the deaf's admission criteria made the student ineligible for placement there. However, the educational audiologist on the school team helped everyone understand ANSD and its implications. Careful functional assessment by the educational audiologist and teachers of the deaf and hard of hearing provided clear evidence that in fact, the proposed placement at the school for the deaf with a communication methodology of Signed English, was the most appropriate, and an exception to the rule would need to be made. Unlike for students with “regular” sensorineural hearing loss, there were no expectations that this student would use hearing aids or an FM system. The student’s access to language was exclusively visual; auditory information appeared to have no meaning for this student.

Student B. This student was 12 years old when we first met, with an audiogram indicating a flat 60 dB hearing loss since diagnosis at age 4. He had been fitted with hearing aids, which he refused to wear; however, he also had an FM system with headphones at school that he found helpful. Because this was in the early days of ANSD diagnosis, the student had not previously received OAE or ABR testing. There was significant confusion at school because they did not present as a student with 60 dB hearing loss who did not wear hearing aids. The educational audiologist

conducted functional audiological and speech-perception testing and found that the student responded to pure tones at 20 dB but had unaided speech-perception scores of 48% at a conversational voice level. Aided speech perception scores were actually poorer than unaided, at 40%, results which made no sense in the context of a 60 dB hearing loss. Scores with the FM system alone were 76%. The student received a subsequent diagnosis of ANSD. They were also seen by the school psychologist who identified a severe learning disability. The student was ultimately recommended for placement in a congregated class for severe learning disabilities, with provision of the FM system. Support services through the Hearing Department were reduced to being "on call" for equipment problems, but otherwise the student did very well in the new class. This student appeared to use auditory information relatively well, particularly with an FM system alone, although they demonstrated significant literacy learning challenges.

Student C. This student was diagnosed with severe to profound hearing loss at 10 years of age despite having no history of hearing loss and demonstrating essentially normal speech and language skills. The diagnosis was quickly refined to ANSD, and the student was fitted with hearing aids. Functional auditory assessment by the educational audiologist using the Ling 6-sound test and other speech-perception assessments indicated that the student had essentially no ability to discriminate speech with hearing alone, which did not seem to match their use of spoken language for communication. The educational audiologist went on to compare speech perception through auditory, visual, and combined modalities. These results indicated that although the student achieved 4% speech discrimination using audition alone and 20% using vision alone (i.e., only lipreading), when audition and vision were combined, their speech perception score rose to 68%. Although the student was unable to make meaningful use of sound alone, the combination of listening and speechreading somehow created useful access to spoken language. The student ultimately received bilateral cochlear implants and continued to receive direct support from the teacher of the deaf while remaining in a regular classroom at his neighborhood school. The student's speech perception scores with hearing aids and with cochlear implants suggest limited to no benefit, yet the student consistently used cochlear implants and an FM system every day at school and communicated very well in spoken language.

The role of the educational audiologist

In all 3 cases, the educational audiologist was crucial in four areas:

i. helping school staff understand the nature of the problem. Because ANSD is rare, school staff (including teachers of the deaf) may not have encountered a student with this profile previously, and maybe confused that their prior knowledge, experience and strategies with hearing loss does not seem to apply or work here

ii. conducting the kinds of functional speech perception assessments that help tease out what is happening in the classroom. Because we cannot rely on the audiogram to predict functional outcomes, we must employ a variety of assessment tools including functional speech perception under different conditions, teacher checklists, questionnaires, interviews, student and classroom observations, etc. Use of a range of functional assessments helps provide a clearer picture of the student in a variety of different contexts

iii. providing strategies and recommendations along the road to trying "everything". While we often do need to try a variety of different approaches, we can't (and shouldn't) try them all at once.

There needs to be a systematic approach, integrated with the clinical audiologist's recommendations, to properly evaluate a range of options. My own approach is to focus on evaluating the extent to which the student can (or cannot) make meaningful use of auditory information, and proceed from there.

iv. providing evidence-based tools to measure progress or lack thereof. For example, while hearing aids are often recommended (Walker et al., 2016), research shows relatively poor success for many of these students (De Siati et al., 2020; Ramanathan et al., 2023; Swain & Prusty, 2026). Morlet et al. (2023) found that only 5% (six patients) received significant benefit from their hearing aids for speech and language development. The authors concluded that hearing aids were not a viable option for most infants and children with ANSD.Statistically true, but hearing aids *did* make a significant difference for these 6 children, so we need to ensure we have an evidence-based, functional, real-world approach to measuring benefit. For example, if parents or school staff report that the student “refuses” to wear hearing aids, educational audiologists can explore this issue to determine if it is truly a rejection of amplified sound because of the ANSD, or if there had never been a consistent approach to implementing hearing aid use to provide a fair trial. If hearing aids have truly not been beneficial, this needs to be clearly documented to support referral to a cochlear implant program, as efficacy of cochlear implants for many students has been demonstrated (Myers & Nicholson, 2021).

v.

Educational audiologists can work at the intersection of audiology and education, providing an opportunity to support the development of a systematic, evidence-based, collaborative approach for students with ANSD. As professionals who often meet and have good relationships with families, educational audiologists can also support families in understanding how we are approaching the problem to create an educational plan that will support communication and academic success. Educational audiologists play a crucial and uniquely positioned role in supporting students with ANSD.

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