Functional Hearing Deficits in Listeners with Auditory Neuropathy Spectrum Disorder

Gary Rance

Abstract

Auditory neuropathy spectrum disorder (ANSD) is a form of auditory dysfunction in which peripheral processes can be normal, but neural transmission through the VIIIth nerve and auditory brainstem is disrupted. The perceptual consequences of ANSD are distinct from those of cochlear hearing loss and most commonly include distortion of temporal (timing) cues and altered frequency discrimination. These basic processing deficits can, in turn, affect functional hearing, resulting in severe impairment of speech perception particularly in the presence of background noise. This article will address the mechanisms underlying ANSD, illustrate it's effects in a detailed case study and show evidence for the use of personal FM devices in affected listeners.

Keywords

auditory neuropathy spectrum disorder, temporal processing, FM-systems, auditory brainstem response

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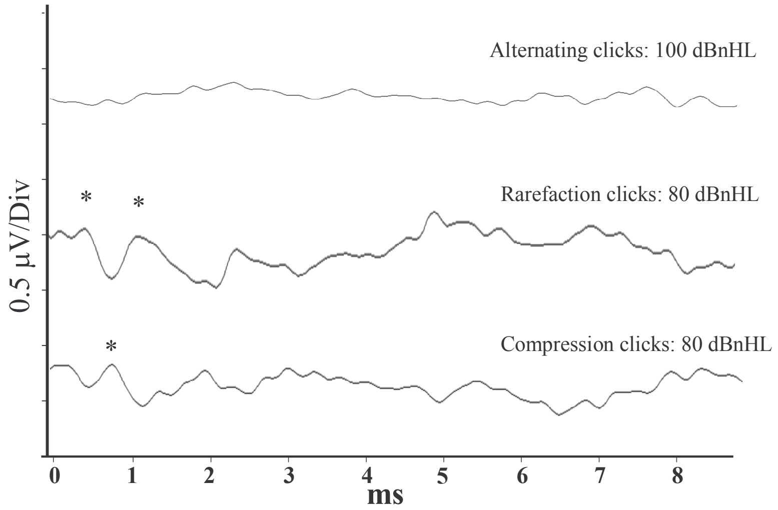
Introduction

Auditory neuropathy spectrum disorder (ANSD) is an umbrella term use to describe a range of pathologies which result in disordered activity in the auditory nerve and central auditory pathways despite apparently normal peripheral function. The clinical pattern which distinguishes ANSD from cochlear-type hearing loss is the demonstration of cochlear (outer hair cell) function through otoacoustic emission (OAE) and/or cochlear microphonic (CM) potentials in conjunction with absence or severe abnormality of the auditory brainstem response (ABR). See Figure 1. Disruption of the ABR in such cases is thought to be either the result of “pre-synaptic” abnormality, specifically affecting the cochlear inner hair cells and/or the release of neurotransmitters (and producing inefficient peripheral triggering of the ABR) or the result of “post-synaptic” or neural abnormality affecting the auditory nerve and it’s brainstem connections (McMahon et al. 2008; Starr et al., 1996). In the latter case, the ABR may be unrecordable if there are insufficient neural elements to produce a response large enough to be recorded on the scalp, or if the function of these elements (in particular their ability to produce synchronised activity) is compromised.

Figure 1. Averaged EEG tracings obtained following acoustic click stimuli presented to the right ear. The top tracing shows no recordable potentials to alternating stimuli at maximum levels (100 dBnHL). The middle and lower tracings show repeatable cochlear microphonics but absent neural responses to unipolar stimuli at 80 dBnHL. Asterisks denote the positive peaks in the CM waveform.



Temporal Processing

Disruption of auditory neural function in listeners with ANSD has been shown to particularly affect the perception of temporal cues (Rance et al., 2004; Rance et al., 2010; Zeng et al., 2005). In cases where the ABR is absent due to desynchronized neural conduction, temporal resolution (the ability to perceive changes in auditory signals over time) is compromised through a smearing of the central representation of the signal. Where the ABR is absent due to reduced neuronal population, timing cues may be preserved in the neural firing, but the low overall amplitude of the response may cause them to be lost in the spontaneous neural activity (Zeng et al., 2005).

The limit of the auditory system's ability to accurately encode or "resolve" rapid temporal changes has most commonly been measured using "gap detection" tasks where the smallest identifiable silent period in a continuous signal is determined. Findings in listeners with ANSD have been consistently poorer than for matched controls. Normal subjects for example, typically show gap detection thresholds of <5 ms where AN/AD listeners with impaired temporal processing usually require silent periods of 10-20 ms (Rance et al., 2010; Starr et al., 1991; Zeng et al., 2005).

Temporal resolution can also be assessed by determining the listener's ability to detect amplitude changes (occurring at different rates) in a continuous signal. This task is a measure of temporal processing in so far as it requires that the auditory pathway encode stimulus envelope variations occurring over time. Amplitude modulation (AM) detection has typically been very poor in listeners with ANSD, particularly for rapidly changing signals which pose the greatest temporal challenge to a compromised auditory system (Rance et al., 2004; 2010; Zeng et al. 2005). Amplitude modulation occurring at rates in excess of 100 Hz for example, while readily detectable to listeners with normal hearing or cochlear hearing loss, are often imperceptible to subjects with ANSD.

Fine Temporal Processing

Amplitude modulation and gap detection tasks, while effective in demonstrating deficits in the perception of overall envelope cues, offer a relatively broad measure of temporal resolution. "Fine temporal processing", reflecting more subtle disruptions of the neural code, can be investigated using frequency discrimination tasks which seek the smallest perceptible frequency change for low (eg. 500 Hz) and high frequency tones (eg. 4 kHz). Discrimination of low frequency sounds in normally hearing listeners is enhanced by temporal cues through "phase locking" where the frequency of the stimulus waveform is reflected in the pattern of neural firing. Phase locking however, requires a high degree of temporal precision and studies involving listeners with ANSD have tended to show impaired discrimination for stimulus frequencies <2 kHz (Rance et al. 2004; Rance et al., 2010; Zeng et al. 2005).

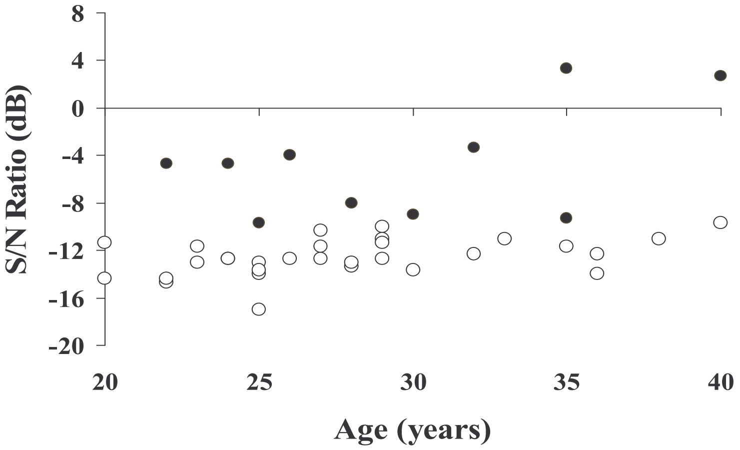
Speech Perception

Impaired speech understanding is the major functional consequence of ANSD. Almost all affected adults have shown performance levels poorer than expected given their audiometric (sound detection) levels (Rance et al., 2008; Starr et al., 1996; Zeng & Liu, 2006). Results in children have been more variable, but many reported cases have demonstrated little or no capacity to understand speech, even in quiet listening conditions, despite (in many cases) enjoying complete access to the speech spectrum (Rance et al., 2004; 2007). In contrast to the findings for populations with cochlear hearing loss (MacArdle et al., 1999; Rance et al., 2002; Yellin et al., 1989), perceptual ability in listeners with ANSD appears to be only weakly related to the behavioural audiogram, but is highly correlated with temporal processing ability (Rance et al. 2004; Zeng et al., 1999; 2005). As such, it appears the level of distortion introduced into the auditory system is the major limiting factor.

Listening in Noise

Discrimination of speech signals in the presence of background noise is a particular problem for listeners with ANSD (Kraus et al., 2000; Rance et al., 2007; Rance et al., 2008; Shallop, 2002). In some cases, ANSD listeners who seem able to cope reasonably well with listening in quiet, show negligible perceptual ability in even low levels of background noise. An example of these "figure-ground" difficulties can be seen in Figure 2 which shows Speech Reception Threshold levels (i.e. the signal-to-noise ratio at which the listener could correctly identify 50% of the items on closed-set testing) for a group of subjects with ANSD (filled circles) and a control cohort (unfilled circles). In this case, the control subjects on average, could meet the target criterion when the SNR was -12.6 dB (that is, when the noise was 12.6 dB higher then the speech). Mean SNR for the ANSD listeners in contrast, was -4.6 dB indicating that they needed the noise to be ≈8 dB lower before they could reach the same performance level.

Figure 2. Speech reception threshold data for the ADSPON (Adaptive Spondees Test) adapted from the data published in Rance et al., 2008. The filled data points show the findings for subjects with ANSD (N=10) due to Friedreich ataxia. Open data points show results for a control group of 30 subjects. Audiometric testing for each of the participants showed normal sound detection.



The link between ANSD and speech-figure/ground issues has not been examined in detail, but psychophysical testing has examined masking effects for tonal stimuli in both "simultaneous" and "forward/backward" masking experiments.

Simultaneous Masking: Listeners with ANSD typically show extreme masking effects for competing sounds presented at the same time as the signal (Kraus et al., 2000; Rance et al., 2004; Zeng et al., 2005). Results have varied across studies, but broadly, ANSD subjects require 10-20 dB less noise to obscure a target tone than normally-hearing controls. The mechanisms underlying this phenomenon are uncertain, but one possibility is simply that a temporally distorted signal may be less distinct in noise at low sensation levels (Zeng et al., 2005).

Forward& Backward Masking: Temporal masking studies involving the detection of brief tones presented in close proximity (before and after) a masking stimulus have also shown abnormal results in ANSD listeners (Kraus et al., 2000; Zeng et al., 2005). In affected subjects, tones separated from the noise by as much as 100 ms are more difficult to perceive whereas normally-hearing listeners typically show only limited masking effects for targets more than 20 ms from the masker (Zeng et al., 2005). As such, ANSD listeners suffer an impaired ability to separate sounds occurring successively. In a real-world listening context, this may translate as an inability to use the brief quiet periods, that occur in fluctuating background noise, to access the speech signal.

Case Study

To illustrate the effects of ANSD on basic auditory processing and functional hearing, findings for a "typical" child are presented. Also included in this case presentation are the results of a personal FM-device fitting designed to maximise his ability to communicate in real-world listening situations.

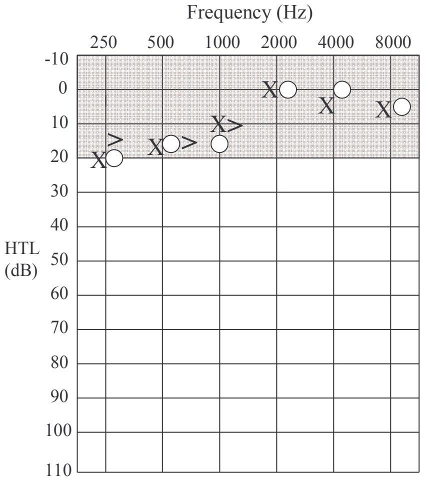
History & Clinical Findings

Subject A is an 8 year old boy who was born at full-term and showed no risk factors for peripheral hearing loss. Genetic testing did however reveal that he was homozygous for a GAA expansion of intron 1 of the FXN gene indicating the presence of Friedreich Ataxia (FRDA) – a mitochondrial neurodegenerative disease affecting both motor and sensory systems. At the time of testing he was still ambulant but was beginning to show signs of gait disturbance and coordination difficulties. His speech and language development were age-appropriate and Fisher-Atkin Articulation Testing revealed normal speech production. All of the auditory assessments described in this report were obtained within a three month time period.

Electrophysiologic assessment revealed the ANSD result pattern with absent auditory brainstem responses to acoustic clicks at maximum presentation levels [100 dBnHL] in each ear (Figure 1). Repeatable cochlear microphonic potentials were obtained to unipolar stimuli indicating the presence of cochlear (outer) hair cell function bilaterally. The ANSD diagnosis was confirmed by the presence of high amplitude distortion product otoacoustic emissions indicating normal function of the cochlear amplifier in both ears.

Sound detection for Subject A was relatively normal. Conditioned audiometric testing revealed mild, low frequency losses bilaterally (Figure 3). While individuals with ANSD can show any audiometric configuration, this pattern (elevated low frequency thresholds) is not uncommon and is demonstrated in approximately 30% of cases (Sininger & Oba, 2001).

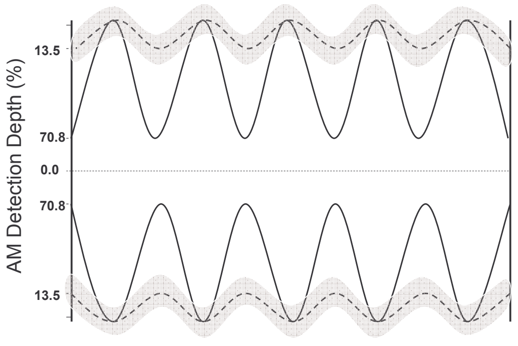
Figure 3. Hearing threshold levels (in dBHL) for the left (X) and right (O) ears.



Temporal Processing

Subject A showed abnormal temporal resolution bilaterally. Where normally-hearing control subjects can detect AM depths of approximately 10-15% (of the total amplitude), Subject A required a depth of >70% before becoming aware of the level fluctuation (Figure 4). This result is consistent with findings for other ANSD listeners and suggests a severely impaired ability to track rapidly occurring envelope changes in auditory stimuli.

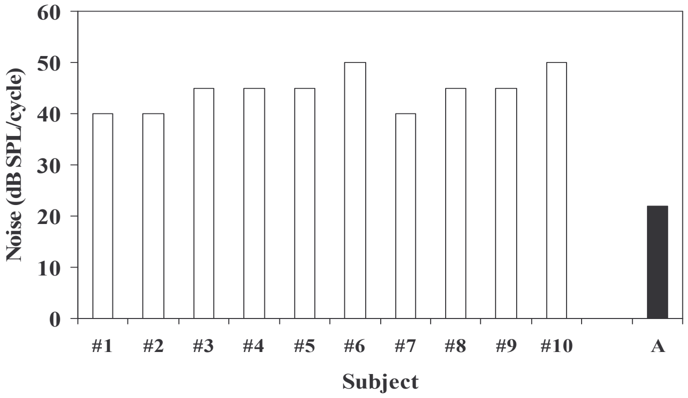
Figure 4. Amplitude modulation detection thresholds reflecting the minimum perceptible modulation depth for a white noise stimulus, sinusoidally modulated at a rate of 150 Hz. The shaded area represents the normal detection range (mean±1 SD) from a cohort of normally hearing, school-aged children (Rance et al., 2004). The solid line shows the detection threshold for Subject A.



Listening in Noise

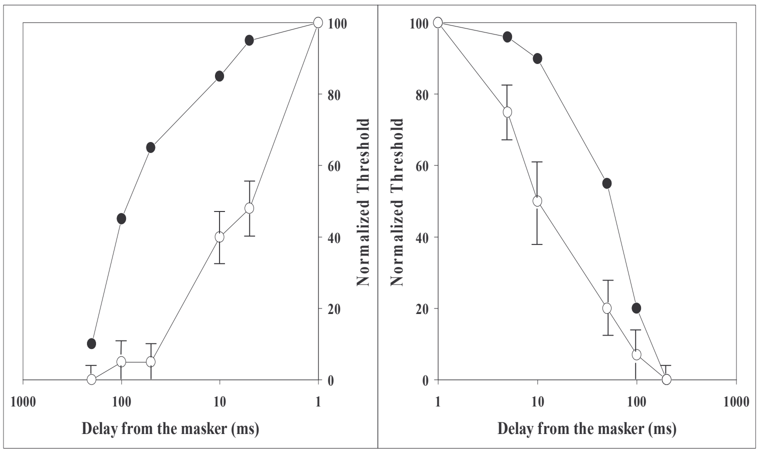
Consistent with his temporal processing deficit, Subject A also showed impaired figure/ground discrimination. The masking effect of simultaneous noise was determined by establishing the minimum dB-level of masking required to obscure a target tone. As can be seen in Figure 5, the signal in his case was masked by approximately 20 dB less noise than was required for a group of age-matched control subjects.

Figure 5. Minimum white-noise masker levels required to obscure a 1 kHz tone presented at 70 dBSPL. Unfilled bars represent the findings for a control group of normally hearing children aged 7-10 years at assessment. The filled bar shows the masking threshold for Subject A.



The effect of masking on non-simultaneous stimuli was also abnormal. In both forward and backward masking experiments, Subject A struggled to perceive a target tone presented within 50 ms of a loud masking stimulus (Figure 6). Age-matched controls in contrast, were relatively unaffected once the signal was more than 10 ms from the masker. As discussed previously, temporal masking deficits of this degree are likely to affect speech perception in environmental noise. Also of concern is the possibility that louder (vowel) components within the speech signal itself may mask perception of softer phonemes. The burst of energy involved in stop-consonants for example may occur less than 50 ms from the vowel and may be obscured in listeners (such as Subject A) unable to efficiently separate sounds occurring successively.

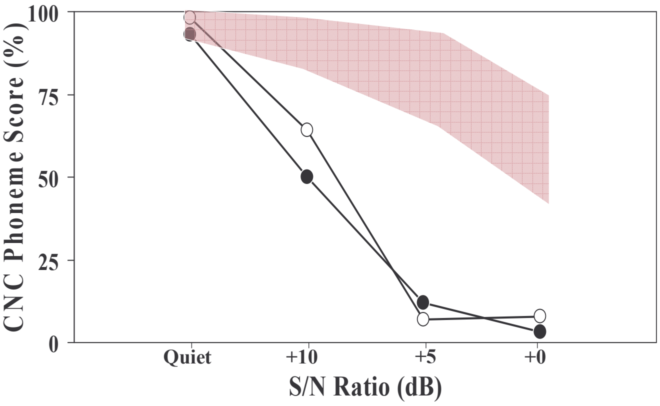
Figure 6. Forward (right panel) and backward (left panel) masking levels. Data points represent normalized thresholds for a 1 kHz tone presented at various time intervals in relation to a white noise masker (see Zeng et al., 2005 for details). Findings for Subject A are represented by the filled data points. Mean thresholds ± 1 SD for a group of 5 age-matched control subjects are represented by the unfilled points.



Speech Perception

Despite the previously described processing abnormalities, Subject A showed normal speech perception ability – at least in optimal (quiet) listening conditions. As can be seen in Figure 5, he could correctly identify and imitate close to 100% of the target phonemes in open set speech perception testing. Speech perception in background noise however, posed a significant challenge. As shown in Figure 7, even at relatively high signal to noise ratios [SNRs] his perceptual ability deteriorated dramatically. This result, which is consistent with findings for other subjects with ANSD due to Friedreich ataxia (Rance et al. 2008), represents a significant communication challenge when we consider that the SNR in a typical school classroom is around 0-5 dB (Crandell & Smalldino, 2000).

Figure 7. Open set speech perception (CNC word) scores for speech in four listening conditions: quiet (no competing signal) and speech in the presence of background noise (4 talker babble) at +10 dB, +5 dB and +0 dB signal to noise ratios. The filled data points show results for Subject A's right ear, and the unfilled are for his left ear. The shaded region is the performance range for a cohort of normally hearing, school-aged children (Rance et al., 2007).



Management

In an attempt to ameliorate Subject A's difficulties with listening in noise, he was fit with a personal FM-listening device. A Phonak Inspiro FM transmitter was paired with bilateral iSense ear-level receivers. On the day of fitting he underwent formal speech perception assessment comparing his unaided and aided (FM) performance. Testing was carried out in the free field with Subject A positioned between two loudspeakers - the speaker in front presenting speech stimuli calibrated to reach his head at 65 dBSPL and the rear speaker providing background noise at the same level (0 dBSNR). For aided testing the FM microphone was suspended 20 cm in front of the "speech" loudspeaker to replicate the typical device distance in everyday use (when worn at the lapel).

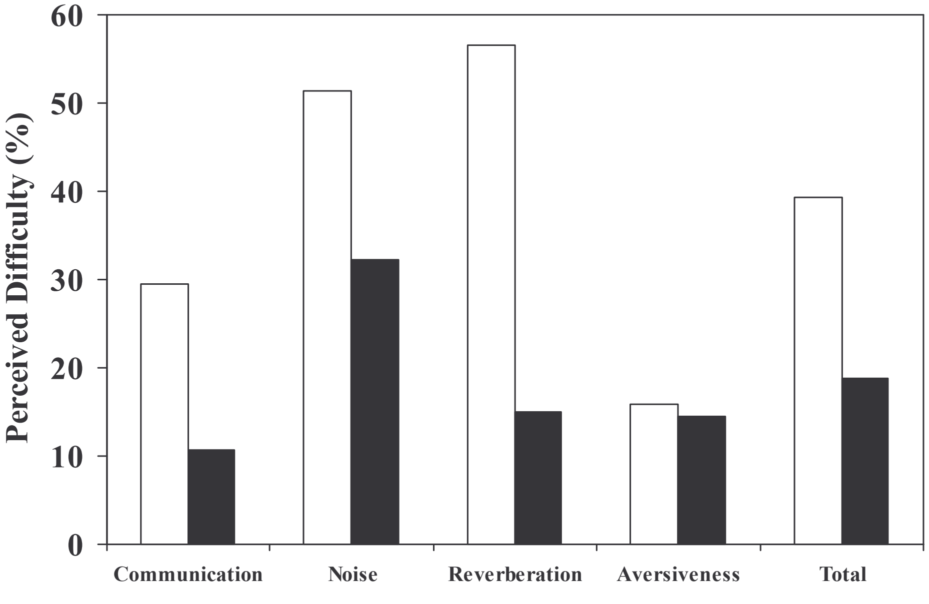
Closed-set (Adaptive Spondee) testing sought the signal to noise ratio at which the child could correctly identify 50% of test items (speech reception threshold [SRT]) in each condition (see Rance et al., 2007 for test details). Subject A's unaided SRT was -11 dB and aided was -24.33 dB which represented an improvement of 13.33 dB when using the FM-device.

His open-set speech perception performance was also significantly aided by the FM-system, improving from a phoneme correct score of only 48.0% in the unaided condition to 82.67% when wearing the device.

Day-to-day listening ability was then investigated across a six week device trial. A balanced (ABBA) design was used with Subject A completing a hearing disability questionnaire (Abbreviated Profile of Hearing Aid Benefit [APHAB]) prior to device fitting (unaided1), after 2 weeks device use (aided1), after a further two weeks use (aided2) and finally after 2 weeks non-use (unaided2). The APHAB explores 4 aspects of auditory function: communication difficulty, effect of background noise, effect of reverberation and aversiveness to sound. In each category a percentage score (representing the proportion of situations in which the individual perceived a difficulty) is generated.

FM device use produced significant listening and communication improvements for Subject A. APHAB scores across the 4 data collection points were; Unaided1: 48.6%; Aided1: 17.3%; Aided2: 17.3%; Unaided2: 46.9%. Figure 8 shows combined unaided and aid scores across the four listening categories. Significantly, Subject A reported FM-aided improvements in listening in noisy situations (including reverberant environments) and general communication. His aversiveness to sound was equally low in both unaided and aided conditions reflecting the fact that loudness growth is typically unaffected by ANSD (Zeng et al., 2005).

Figure 8. APHAB hearing disability questionnaire results for unaided (unfilled bars) and aided (filled) conditions.



Postscript: At the time of writing (9 months post trial) Subject A continues to wear the FM-device full-time at school and in challenging listening situations elsewhere. His teachers and parents report significant improvements in general listening, behaviour and social engagement.

Summary

Temporal processing disruption associated with ANSD has significant consequences for functional hearing. This is particularly evident for speech perception in the presence of background noise. The use of hearing devices which improve SNR (including personal FM-systems) do however offer a means of maximising communication in everyday listening situations.

Acknowledgements

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FIGURE LEGENDS

Figure 1. Averaged EEG tracings obtained following acoustic click stimuli presented to the right ear. The top tracing shows no recordable potentials to alternating stimuli at maximum levels (100 dBnHL). The middle and lower tracings show repeatable cochlear microphonics but absent neural responses to unipolar stimuli at 80 dBnHL. Asterisks denote the positive peaks in the CM waveform.

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