**Speech and Feature Perception in Children with Auditory Neuropathy/Dys-synchrony Type Hearing Loss: Implications for Management**

*Gary Rance*

### Introduction

Auditory neuropathy/dys-synchrony (AN/AD) is a term used to describe hearing loss affecting the transmission of neural signals in the auditory brainstem and beyond (Berlin, Hood and Rose 2001; Starr, Picton, Sininger, Hood and Berlin 1996). Identification of affected individuals is relatively straightforward, but devising appropriate management strategies remains a challenge as AN/AD can produce a pattern of perceptual deficits unlike those seen for other forms of permanent hearing loss.

### The AN/AD Result Pattern

The configuration of clinical findings that distinguishes AN/AD from peripheral (sensorineural) type hearing loss is the presence of pre-neural cochlear responses in conjunction with an inability to record evoked neural activity from the auditory brainstem. In affected patients, normal cochlear (outer hair cell) function is suggested by the presence of otoacoustic emissions and/or cochlear microphonics, and the auditory pathway dysfunction is indicated by the absence (or severe distortion) of potentials from the auditory nerve (compound action potential) and auditory brainstem (ABR).

Figure 1 shows an example of the typical electrophysiological findings for a subject with AN/AD type hearing loss. The top tracing in this case shows no recordable ABR to alternating polarity acoustic clicks at the maximum presentation level (100 dBnHL). The lower tracings also reveal no auditory brainstem potentials, but show clear cochlear microphonic responses to unipolar stimuli. The inconsistency between pre-neural (CM) and neural (ABR) findings is indicative of auditory pathway disorder in this case.

### Mechanisms

Disruption of auditory neural activity in individuals with AN/AD type hearing loss may be the result of abnormality at a number of possible sites, including the auditory nerve, brainstem, or central auditory pathways. Understanding the specific mechanisms underlying AN/AD hearing loss can guide the development of more effective management strategies.
cochlear inner hair cells, the synapse between the inner hair cells and cochlear nerve and the auditory nerve itself (Amatuzzi et al. 2001; Starr et al. 1996). ABR abnormality in such cases is thought to be the result of either a paucity of neural elements available to contribute to the volume-conducted response, or a disruption of the synchrony or timing of neural activity in the auditory brainstem (Rance 2005). In the former case, potentials generated in the auditory pathway may simply be too small to be recorded by scalp-sited electrodes. In the latter, the ABR may be unrecognizable, not because of a lack of activity, but because the timing of that activity is not sufficiently precise to allow extraction from within the EEG using signal averaging procedures (Clarke et al. 1961). As variations in the timing of responses to individual stimuli of only fractions of a millisecond are sufficient to disrupt the averaged ABR, it has been suggested that temporal inconsistency may be a factor in patients with the AN/AD result pattern (Kraus et al. 2000; Michalewski, Starr, Nguyen, Kong and Zenn 2005; Starr et al. 1991).

Speech Perception

Difficulty in speech understanding is a consistently reported consequence of AN/AD type hearing loss. Most affected adults show perceptual performance levels significantly worse than would be expected for their degree of hearing loss (Starr et al. 1996; Starr, Sininger and Pratt 2000; Zeng, Oba and Starr 2001). Results in children have been more varied, but again, a high proportion (≈50%) of reported cases have shown only limited capacity to understand speech despite (in many cases) enjoying complete access to the normal speech spectrum. That is, they can detect speech sounds but cannot make discriminations based on what they have heard. Figure 2 shows open-set speech perception scores (for word and sentence level materials) plotted against 3-frequency average hearing level for all of the children presented in the literature to date. As can be seen in these data, while some cases show perceptual abilities similar to their peers with sensorineural hearing loss (scoring above the dashed line in this case), many perform at only chance levels despite showing audiometric thresholds in the only mild-to-moderate hearing loss range.

Speech Perception in Noise

In addition to speech perception problems in favorable (quiet) listening conditions, extreme difficulties in background noise have been reported for both adults and children with AN/AD (Kraus et al. 2000; Shallop 2002; Starr et al. 1998). Each of the afore-mentioned studies has presented case examples showing excellent speech perception in quiet, but negligible perceptual ability at signal-to-noise ratios consistent with everyday listening conditions (+3 dB to +12 dB SNR).

Speech perception difficulties are certainly not restricted to AN/AD type hearing loss. Ears with significant sensorineural deficit also show greater noise effects than do normally hearing subjects for reasons thought to be related to a loss of precision in cochlear-level processing (Rance et al. 2007). The mechanisms underlying excessive noise effects in AN/AD type hearing loss are unclear, although there is psychophysical evidence that auditory signals in AN/AD subjects are more affected by simultaneous and non-simultaneous masking than normal listeners (Kraus et al. 2000; Vinay and Moore 2007; Zeng et al. 2005).

An example of the degree to which noise affects speech understanding in hearing impaired children can be seen in figure 3, which shows closed-set results for groups of school-aged subjects with AN/AD and SN type hearing loss. Also shown are findings for an age-matched cohort of children with normal hearing. In this experiment (the Adaptive Spondee Test [ADSPON]), signal-to-noise ratio is varied to determine the point at
which 79.4% spondee recognition is obtained. Hence a lower SNR on this scale represents a greater ability to cope with a competing signal. As can be seen in these data, speech perception in noise for children with AN/AD and SN type hearing loss was broadly similar and both groups were more affected by noise than their normally hearing peers.

Cochlear level and is mediated by the active processes of the outer hair cells, which amplify and sharpen the peaks of basilar membrane movement (Yates, Johnstone, Patuzzi and Robertson 1992). As such, frequency resolution in subjects with even mild degrees of cochlear hearing loss is significantly reduced (Sellick, Patuzzi and Johnstone 1982). As outer hair cell function appears “normal” in ears with AN/AD type hearing loss, it is not surprising to find that frequency resolution measured both physiologically (through otoacoustic emission [OAE] suppression tuning curves [Abdala, Sininger and Starr 2000]), and psychophysically (Cacace, Satya-Murti and Grimes 1983; Rance et al. 2004; Vinay and Moore 2007) has been normal in most reported cases.

Intensity-Related Perception

Discrimination of intensity cues also appears to be normal in AN/AD subjects. Zeng et al. (2005), for example, found that intensity difference limen (the smallest perceivable level difference between stimuli) for a group of AN/AD subjects were similar to those obtained for a normally hearing control group. In addition, these authors found that binaural intensity processing was unimpaired in their cohort of AN/AD subjects allowing normal sound lateralization percepts for stimuli presented simultaneously (at different levels) in each ear.

Processing of Temporal Cues

Apart from a reduction in signal audibility, disruption of timing-related cues is the major way in which auditory perception is affected in subjects with AN/AD type hearing loss. Significant deficits have been reported in this population for both monaural and binaural processing abilities.

Temporal Resolution: temporal resolution is the ability to perceive changes in auditory signals over time. In ears with sensorineural hearing loss, temporal processing is relatively normal (provided the stimuli are sufficiently audible; Moore 1995). Subjects with AN/AD in contrast, show clear deficits in their ability to “resolve” rapid stimulus changes. In gap detection tasks for example, where normally hearing subjects can perceive silent periods of < 5 msec in a continuous signal, individuals with AN/AD usually require gaps of 20 msec or more before they become aware of the change (Michalewski et al. 2005; Starr et al. 1991; Zeng, Oba, Garde, Sininger and Starr 1999; Zeng et al. 2005).
Amplitude Modulation Detection: temporal resolution can also be assessed by measuring a subject’s “temporal modulation transfer function (TMTF)”, which determines their ability to detect rapid amplitude fluctuations (at different modulation rates) in the level of a steady-state signal. TMTF findings for AN/AD patients have been variable, but have typically shown an impaired capacity to track fast, and even relatively slow (<10 Hz) amplitude envelope changes over time (Rance et al. 2004; Zeng et al. 1999; Zeng et al. 2005).

Forward and Backward Masking: temporal processing deficits in AN/AD patients have also been demonstrated in masking studies (Kraus et al. 2000; Zeng et al. 2005). Forward and backward masking experiments (examining the detection of brief tones in the proximity of masking stimuli) have indicated that perception of short signals is affected if they are presented within as much as 100 msec of a masker. Normally hearing subjects, in contrast, show only limited masking effects when the target is beyond ≈10–20 msec of the masker (Zeng et al. 2005). As such, AN/AD patients display a reduced capacity to separate sounds that occur successively.

Binaural Temporal Processing: in addition to showing monaural temporal processing deficits, AN/AD patients also demonstrate an impaired ability to integrate binaural temporal cues. Abnormal Masking Level Difference (MLD) results, for example, are a consistently reported finding (Berlin, Hood, Cecola, Jackson and Szabo 1993; Hood 1999; Starr et al. 1991; Starr et al. 1996). MLD assessment measures the release from masking obtained when a signal is presented out of phase with a competing signal in the contralateral ear. In contrast to normally hearing listeners who typically show an MLD ≈10 dB, AN/AD subjects tend to show little or no masking release, suggesting an inability to combine accurately the neural code from each ear.

Localization based upon binaural timing cues also appears to be disrupted in AN/AD patients. Despite the fact that affected individuals can use interaural intensity differences to make lateralization judgments (Zeng et al. 2005) a number of studies have found that even gross timing differences (> 0.5 msec between ears) are not interpreted as changes in sound direction (Starr et al. 1991; Kaga et al. 1996; Zeng et al. 2005).

Frequency Discrimination

Frequency discrimination is the ability to perceive changes in frequency (or pitch) over time or between stimuli. High frequency discrimination (≥ 4 kHz) is dependent upon the spatial arrangement of excitation along the basilar membrane (Sek and Moore 1995). Discrimination of lower frequency sounds is also influenced by tonotopic cochlear representation, but is enhanced by temporal information (Sek and Moore 1995) through “phase locking” (where the frequency of the stimulus waveform is reflected in neural firing patterns). Phase locking requires a high degree of temporal precision and so, not surprisingly, individuals with AN/AD type hearing loss show a reduced ability to use these cues and demonstrate severely impaired frequency discrimination (Rance et al. 2004; Zeng et al. 2005). Elevated frequency difference limen have been reported for AN/AD subjects at all frequencies in the audiometric range, but low frequency perception (which benefits most from phase locking) is most affected. Low tone difference limen in AN/AD cases are typically more than 10 times greater than those of normally hearing subjects (Rance et al. 2004; Zeng et al. 2005).

Temporal Processing and Speech Perception

While degree of temporal disruption is strongly correlated with overall speech perception ability in AN/AD patients (Rance et al. 2004; Zeng et al. 1999), the ways in which this form of processing abnormality affect perception (at the speech feature level) are yet to be fully considered. Kraus et al. (2000) have demonstrated that an inability to detect brief gaps in the speech signal affects the perception of brief vowel features such as third formant onset frequency. One might also expect discrimination of consonant place of articulation (which is based upon subtle differences in voice onset time) to be similarly affected. A reduced ability to perceive cues contained within the overall amplitude envelope (as suggested by the TMTF findings) might also be expected to have a negative impact on speech understanding (Shannon, Zeng and Kamath 1995; Turner, Souza and Forget 1995). Furthermore, an impaired ability to separate sounds occurring successively (as indicated by the forward and backward masking studies) may produce excessive intra-speech masking effects, where louder speech elements (such as vowels) could mask brief consonant information.

Management

The pattern of perceptual deficits in AN/AD type hearing loss is fundamentally different from that associated with sensorineural loss. As such, different manage-
ment strategies may be warranted. While the application of AN/AD-specific habilitation approaches in this population is yet to be thoroughly investigated, the appropriateness of hearing devices has been considered.

Amplification

The provision of hearing aids to AN/AD patients (particularly young children) has been a controversial issue. There are two main arguments against amplification in this population. The first is that amplified sound may cause some damage in these ears with evidence of normal cochlear outer hair cell function. The second is that conventional amplification is not designed to overcome the temporal distortion introduced into the system by the auditory pathway disorder. That is, the aided child will simply be presented with a loud, but equally distorted signal. The potential benefit offered by hearing aids relates to sound access. Many affected individuals present with significantly elevated hearing thresholds and as such, cannot detect speech and other signals at normal levels.

Outcomes for AN/AD patients managed with conventional amplification have been mixed. Some children (perhaps those with lesser degrees of temporal disruption) have responded well to hearing aids and have shown aided speech perception abilities (and overall levels of expressive speech and receptive language development) consistent with their sensorineural counterparts (Rance, Cone-Wesson, Wunderlich and Dowel 2002; Rance et al. 2007). In many youngsters and almost all affected adults, however, conventional amplification has been of little or no benefit (Rance 2005; Starr et al. 1996). As a result, amplification systems using signal processing strategies aimed at overcoming the particular processing deficits associated with AN/AD have been considered (Zeng et al. 2001). For example, processing algorithms that accentuate temporal cues and amplitude differences, or transpose spectral information into the high frequency region (where frequency discrimination is less affected) may, in the future, improve perceptual outcomes in some cases.

Cochlear Implants

Cochlear implantation is currently the management option of choice for AN/AD patients with audiograms in the severe/profound range or significant auditory processing difficulties. Most reported cases have shown normal device function and post-implant speech perception performance broadly consistent with their implanted sensorineural peers (Madden, Rutter, Hilbert, Greinwald and Choo 2002; Mason, DeMichele, Stevens, Ruth and Hashisaki 2003; Trautwein, Shallop, Fabry and Friedman 2001; Shallop 2002). These findings may seem counter-intuitive for a group of subjects with evidence of transmission disorder in the auditory brainstem and yet, of the >100 reported subjects, only a handful have shown poor outcomes (Gibson and Sanli 2007; Miyamoto et al. 1999; Rance et al. 1999). The ways in which perception is improved by the cochlear implant in ears with AN/AD is currently under investigation. Interestingly, most implanted subjects show recordable brainstem potentials to electrical stimulation, where previously to acoustic stimulation they had not. This suggests either an increase in the number of neural elements contributing to the ABR (perhaps as a result of by-passing a peripheral abnormality when stimulating the spiral ganglion directly) or an improvement in the synchrony of neural firing in the brainstem (perhaps reflecting the fact that the electrical stimuli are presented as a series of discrete pulses rather than analog signals). Whatever the explanation, it would seem that cochlear implantation offers a desirable management option for most patients with AN/AD type hearing loss.

Comparison of Long-Term Speech Perception Outcomes in Children Managed with Hearing Aids and Cochlear Implants

Despite the recent interest in management options for children with AN/AD type hearing loss, almost no data directly comparing long-term outcomes in subjects fitted with hearing aids and those receiving cochlear implants have been presented thus far. This concluding section describes speech perception findings from a recent comparative study carried out in our laboratory (Rance and Barker in press). The aim of this work was to evaluate perceptual skills (in quiet and noisy listening conditions) for a group of school-aged AN/AD children who had received a cochlear implant and to compare these findings with those of a matched cohort of young implantees with sensorineural type hearing loss and a group of AN/AD children managed with conventional hearing aids.

Subjects

Twenty children with AN/AD type hearing loss were included in this study. Ten of these had been fitted with cochlear implant devices (in one or both ears) once
severe/profound hearing loss had been established, or once it had become clear that they were obtaining little or no benefit from amplification. Lack of benefit was determined from formal speech perception testing (phoneme score < 40% on open set word testing in the best aided condition) and from the impressions of parents and clinicians. Mean age at implantation for this group was 33.3 ±16.9 months and each child had been a consistent device user for over two years at the time of the assessment. Average age at the time of testing was 89.4 ±42.1 months.

Electrical auditory brainstem response (EABR) assessment was carried out in each of these children at the time of the surgery or in the immediate post-operative period. Nine of the ten subjects showed robust potentials of normal latency and morphology consistent with the presence of synchronized neural activity in the auditory brainstem. In one child however, responses could not be elicited to electrical stimulation at any level on any channel (see Rance et al. 1999 for a detailed description of this subject).

The remaining ten AN/AD subjects were all long-term hearing aid users. Nine of the ten were fitted at a reasonably young age (< 10 months), and one child with only mild hearing loss was identified and aided at 16 months. All had been fitted bilaterally with behind-the-ear devices adjusted to the level of their behavioral audiogram according to the NAL prescriptions for gain and frequency response (Byrne and Dillon 1986). These subjects were selected (from our broader AN/AD cohort) to match the implanted AN/AD group for age at assessment (mean: 94.2 ±57 months). Three frequency average hearing levels in these cases ranged from 21.6 dB HL to 68.3 dB HL, and all were afforded complete access to the normal 70 dB SPL speech spectrum by their hearing aids.

A control cohort of implanted children with sensorineural hearing loss was also assessed. These subjects were selected to closely match the implanted AN/AD group for age at assessment (92.6 ±34.6 months), age at implantation (30.2 ±15.5 months) and hearing device configuration.

Procedures

Speech Perception in Quiet

Open-set speech perception testing in quiet (CNC words) was carried out for each subject. Stimuli were presented via a loudspeaker situated at a distance of one meter and calibrated to produce an average signal level of 70 dB SPL. Each child was tested in their optimal listening condition: bilateral hearing aids (AN/AD: n = 10), cochlear implant alone (AN/AD: n = 4; SN: n = 4), cochlear implant and a contralateral hearing aid (AN/AD: n = 3; SN: n = 3) or bilateral cochlear implant (AN/AD: n = 3; SN: n = 3).

Speech Perception in Noise

The adaptive sonde in noise test (ADSPON) was also undertaken with each child. Again, the test stimuli were presented in the free field with the child wearing their typical (optimal) device configuration. This closed-set task, which requires that the subject select from a set of four items, was carried out with varying levels of background noise, and the signal-to-noise-ratio at which they could correctly identify the spondees 79.4% of the time was determined (Rance et al. 2007).

Results and Interpretation

Speech Perception in Quiet

Speech perception findings (in quiet listening conditions) for the implanted AN/AD children confirm the previously reported observation that cochlear implantation can provide significant open set speech perception in this population (Madden et al. 2002; Mason et al. 2003; Trautwein et al. 2001; Shallop 2002). As can be seen in figure 4, nine of the ten children in this group showed CNC scores > 55%, which in each case represented a significant post-implant improvement in perceptual ability. One child (represented in grey in figure 4) failed to show useful hearing with his implant. He was distinct from his peers in that he was the only subject who showed no recordable EABR. As such, he resembles other (relatively uncommon) cases in the AN/AD literature who have presented with absent post-operative brainstem potentials and poor speech understanding (Gibson and Sanli 2007).

Despite the clear post-implant speech perception improvement observed in most cases, overall findings for the implanted AN/AD group in this study were poorer than those of the implanted sensorineural cohort. As can be seen in table 1, the mean CNC score for the matched SN children was almost 25% higher. A one way analysis of variance (comparing the 3 groups) showed a significant group effect (F(2, 29) = 6.34, p = 0.006), and post-hoc analysis (Tukeys) showed a significant difference between the implanted AN/AD and
SN groups. This result is inconsistent with the current literature, which has suggested that implanted AN/AD subjects perform as well as their sensorineural counterparts, although Zeng and Liu (2006) also found that AN/AD patients tend to be at the low end of the post-implant performance range.

Speech perception results for the aided AN/AD children in this study were variable, but most subjects were able to score at significant levels on the CNC word test (see Figure 4 for details). Results for this group were biased to an extent by the fact that most of the poorer hearing aid performers (in our broader AN/AD population) had already received a cochlear implant. Hence the subjects presented in this study tended to be the ones who had coped well with amplification. Nonetheless the findings do indicate that at least some aided AN/AD children show perceptual skills sufficient to support the development of oral speech and language. Furthermore, there was no difference in performance levels between the AN/AD groups indicating that in quiet listening conditions at least, some children with AN/AD type hearing loss can perceive speech as well with hearing aids as their implanted peers.

Table 1: Mean performance levels for the CNC Word and Adaptive Spondee tests for each of the subject groups.

<table>
<thead>
<tr>
<th>Subject Category</th>
<th>N</th>
<th>CNC Phoneme Score (%)</th>
<th>APSPON SNR (dB)</th>
</tr>
</thead>
<tbody>
<tr>
<td>AN/AD Aided</td>
<td>10</td>
<td>55.1 ± 24.8</td>
<td>2.0 ± 9.9</td>
</tr>
<tr>
<td>AN/AD Implanted</td>
<td>10</td>
<td>59.6 ± 20.6</td>
<td>0.1 ± 7.7</td>
</tr>
<tr>
<td>AN/AD Implanted (EABR)</td>
<td>9</td>
<td>65.5 ± 8.9</td>
<td>-2.11 ± 3.5</td>
</tr>
<tr>
<td>SN Implanted</td>
<td>10</td>
<td>83.1 ± 5.3</td>
<td>-2.7 ± 2.8</td>
</tr>
</tbody>
</table>

Speech Perception in Noise

Overall, children in the three subject groups were affected to similar degrees by the presence of background noise (Table 1). Analysis of variance showed no significant group effect (F(2, 27) = 1.04, p = 0.37) in the signal-to-noise ratios required to perceive spondee stimuli in speech-shaped broadband noise. There were, however, a number of outliers. Two children in the aided AN/AD group required that the speech be at least 20 dB higher than the competing signal before they could correctly identify the speech targets. As such they resemble the (mostly adult) AN/AD cases previously reported in the literature whose perception of acoustic cues was dramatically reduced by relatively low levels of background noise (Kraus et al. 2000; Shallop 2002; Starr et al. 1998).

One child in the implanted AN/AD group (again the subject who showed no post-operative EABR) also struggled in background noise. Where the rest of the implanted AN/AD cohort appeared no more affected by noise than their SN peers, this child, despite being able to carry out the closed-set discrimination task in quiet, could not cope at all with listening conditions equivalent to the average classroom environment (SNR ≈ 0–5 dB) and required signal-to-noise ratios of at least 20 dB.

Correlation of the findings for the two speech perception tests in this study is shown in Figure 4. A significant relationship was obtained (r = -0.76, p = 0.001) in-
indicating that those AN/AD subjects whose auditory pathway disorder significantly limited their open-set speech perception ability, were also the most affected by the presence of background noise.

Study Summary

The results of this investigation raise some important management issues. Cochlear implantation certainly offers the possibility of perceptual improvement and open-set speech discrimination in those individuals most affected by AN/AD type hearing loss. However, a significant proportion of children showing the AN/AD result pattern can do as well with conventional hearing aids as the average implantee and as such, it is not appropriate to assume that every newly diagnosed child will be a suitable CI candidate. Establishing appropriate cochlear implant selection criteria will require greater experience and subject numbers. The results of this study do however suggest that these criteria may need to be different for subjects with AN/AD from those used for children with sensorineural loss.

Conclusions

While identification of AN/AD type hearing loss in the pediatric clinic is now relatively straightforward, the diagnosis presents clinicians (and families) with a number of management issues. Clearly some affected children can cope well with conventional amplification, whereas others (with the same pattern of physiologic results) show little or no useful hearing without the aid of cochlear implantation. Determining the most effective way(s) to establish a child’s hearing capacity in infancy remains a significant challenge for the future.

References


Rance, G. and Barker, E. in press. Speech perception in children with auditory neuropathy/dys-synchrony managed with either hearing aids or cochlear implants. *Otology and Neurotology*.


