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Psychoacoustics and Speech Perception in Auditory Neuropathy

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Auditory neuropathy (AN) is typically associated with hearing loss, the presence of otoacoustic emissions, and the absence of evoked potentials. Functional deficits in people with this disorder are poorly appreciated, however. They often demonstrate a speech recognition deficit, particularly in noise, that is disproportional to the amount of pure-tone hearing loss if it were sensory in nature. Conventional hearing aids are less effective than expected in alleviating their speech recognition problems in daily life. The causes of AN are not clear and may be related to mechanical, chemical, or neuronal deficiency (or a combination of these factors) in the inner hair cell, the synapse between the inner hair cell and the auditory neurons, or the auditory neurons (Harrison, 1998; Starr, Picton, Sininger, Hood, & Berlin, 1996). This chapter describes a psychophysical approach to the description of functional processing deficits in AN.

There are three goals in psychophysical studies of AN. The first is to characterize the functional capabilities in people with auditory neuropathy. The emphasis here is to understand why these people often complain that they can hear sounds but cannot understand speech. The second goal is to develop behavioral tests that can help delineate underlying physiological mechanisms and differentiate hearing loss of different origins (e.g., damage to outer hair cells, inner hair cells, and the auditory nerve). The third goal in psychophysical studies of auditory neuropathy is to provide guidance for designing auditory prostheses and rehabilitation strategies that best fit the residual processing capabilities in these people.

We have collected extensive psychophysical data for intensity, frequency, and temporal processing in subjects diagnosed with AN. Our results indicate that although there are abnormalities in intensity and frequency processing, the impaired temporal processing is likely the major reason for the poor speech recognition in AN subjects. This conclusion receives further support from a simulation of the temporal processing impairment in listeners with normal hearing who can produce psychophysical and speech recognition deficits similar to those seen in AN subjects.

SPECIFIC STUDIES

Subjects

We studied 10 AN subjects, including one with unilateral neuropathy. We also studied three control subjects including (a) the healthy ear in the subject with unilateral neuropathy, (b) one subject with cochlear impairment who has an atypical low-frequency hearing loss configuration, and (c) a group of six subjects with normal hearing, including three female and three male subjects, ages 27 to 35 years old; all had pure-tone air- and bone-conduction thresholds of 20 dB HL or better for the octave frequencies between 250 and 8000 Hz.

Table 8–1 lists audiologic and neurologic test results of the subjects with AN and of control subjects. Figure 8–1 shows pure-tone thresholds as a function of frequency for the subjects with AN and the control subjects. The subjects with AN exhibited a wide range of hearing loss; on average, they had a moderate (60 dB) hearing loss in the low frequencies but only a mild hearing loss (30–40 dB) in the high frequencies. Subjects with AN had normal measures of cochlear outer hair cell functions (i.e., otoacoustic emissions, cochlear microphonics, or both were present in all subjects). No subjects with AN demonstrated wave I on auditory brain stem potentials, and all who were tested showed absent acoustic middle ear muscle reflexes to tones up to and including 110 dB HL. Brain imaging results were normal in the six subjects with AN who were tested. The control ear from the subject with a unilateral auditory neuropathy (N-AN8) had normal pure-tone thresholds, present otoacoustic emissions, cochlear microphonics, auditory brain stem potentials, and 100%-correct word recognition.

The subject with cochlear impairment in this study had a low-frequency hearing loss and near-normal hearing at higher frequencies. This configuration was chosen to mimic the loss of average auditory neuropathy subjects and was different from the usual high-frequency-sloping hearing loss typically seen in subjects with cochlear loss. His

Table 8–1. Audiologic and neurologic test results of subjects with auditory neuropathy and control subjects.

Subject	Age	Gender	PTA (dB)	Speech (%)	Otoacoustic Emission (dB)	Cochlear Microphonic	ABR	Acoustic Reflex	Imaging	PN
AN1	50	F	32	56	TEOAE = 11.8	Present	Abnormal	Absent	DNT	Yes
AN2	53	F	15	DNT	TEOAE = 8.9	DNT	Abnormal	DNT	DNT	Yes
AN3	19	F	72	12	DPOAE = 3–15	DNT	Absent	Absent	Normal MRI	Yes
AN4	23	F	38	0	TEOAE = 5.4	Present	Absent	Absent	Normal MRI	No
AN5	17	F	62	0	Absent	Present	Absent	Absent	Normal CT	No
AN6	37	M	55	CNT	DPOAE = 4–13	DNT	Absent	Absent	Normal MRI	Yes
AN7	27	M	45	40	TEOAE = 12.3	Present	Abnormal	Absent	DNT	No
AN8	10	M	82	0	DPOAE = 3–10	Present	Abnormal	DNT	DNT	No
AN9	13	M	63	0	TEOAE = 12.6	Present	Absent	Absent	Normal	No
AN10	22	M	30	64	TEOAE = 11.7	DNT	Absent	DNT	Normal PET	No
Average	27.1		49.4	21.5						
CHL	40	F	60	84	DPOAE = 5 dB @ 6,000 only	DNT	Present	DNT	DNT	No
N-AN8	10	M	7	100	TEOAE = 10.3	Present	Present	DNT	DNT	No

Note. F = female; M = male; PTA = pure-tone average threshold at 500, 1000, and 2000 Hz; CNT = could not test; DNT = did not test; TEOAE = transient-evoked otoacoustic emission; DPOAE = distortion product otoacoustic emission; ABR = auditory brain stem response; MRI = magnetic resonance imaging; CT = computer tomography; PET = positron emission tomography; PN = peripheral neuropathy.

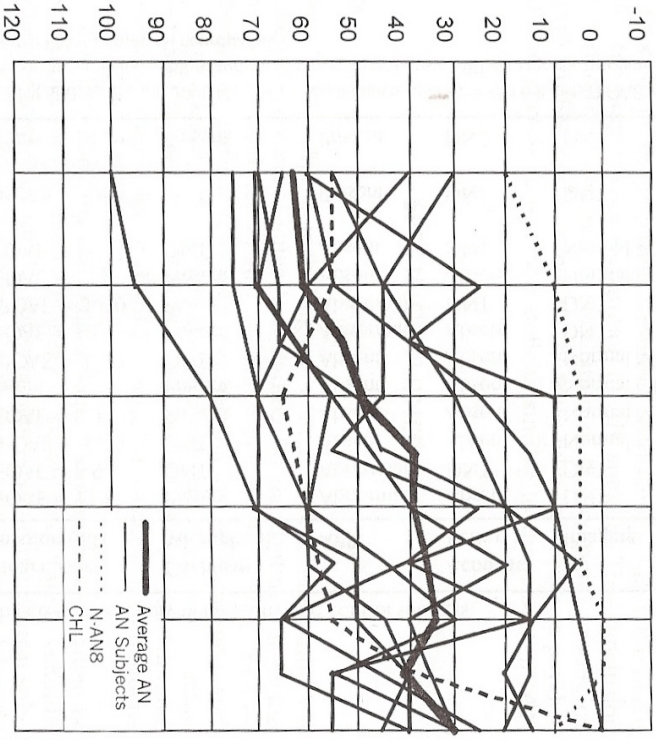


Figure 8–1. Audiogram. Pure-tone thresholds are plotted as a function of frequency. Narrow lines indicate individual subjects with auditory neuropathy, and the wide line indicates the average of these subjects. The long dashed line indicates the control subject with a cochlear hearing loss, and the dotted line indicates normal ear of the unilateral auditory neuropathy case (N–AN8).

low-frequency hearing loss generally was within 20 dB of the average hearing thresholds of AN subjects. The subject with cochlear impairment had no otoacoustic emissions except at the 6000 Hz where the hearing threshold was normal, and all components of the auditory brain stem potential were identified. His 84% word recognition was consistent with his moderate pure-tone loss. In contrast, word recognition scores among subjects with AN ranged from 0 to 64% with an average of 22%—

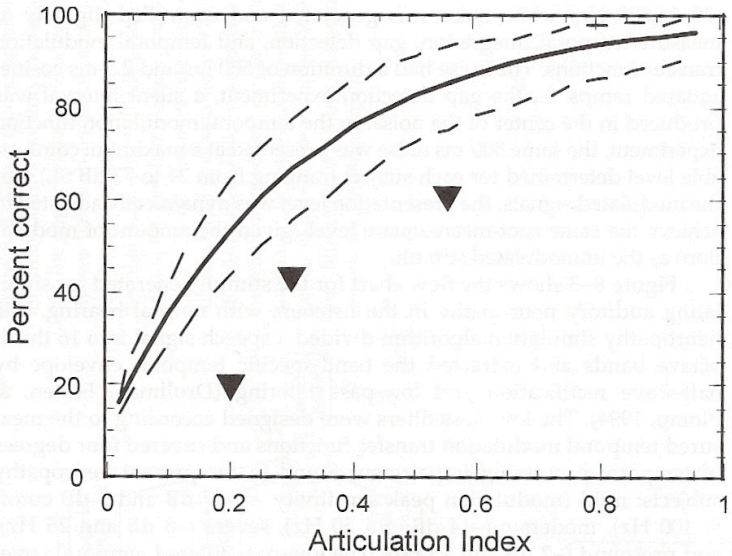


Figure 8–2. Articulation index (AI) analysis on speech recognition deficits in people with auditory neuropathy. The solid line and surrounding dashed lines indicate the expected relationship between AI (abscissa) and speech discrimination scores (ordinate) as determined by Dubno and Alstrom (1995). The actual relationships for three subjects with auditory neuropathy are indicated by the closed triangle symbols.

significantly lower than what would be expected from their pure-tone hearing loss. Figure 8–2 shows an estimate of the phoneme recognition scores that should be expected from the pure-tone threshold measurement (Dubno & Ahlstrom, 1995). A simplified version of the articulation index (AI; Mueller & Killion, 1990) was applied to the phoneme recognition scores of three subjects with auditory neuropathy. This showed that the actual phoneme recognition scores of these subjects were at least two standard deviations poorer than the AI-predicted scores.

Stimuli

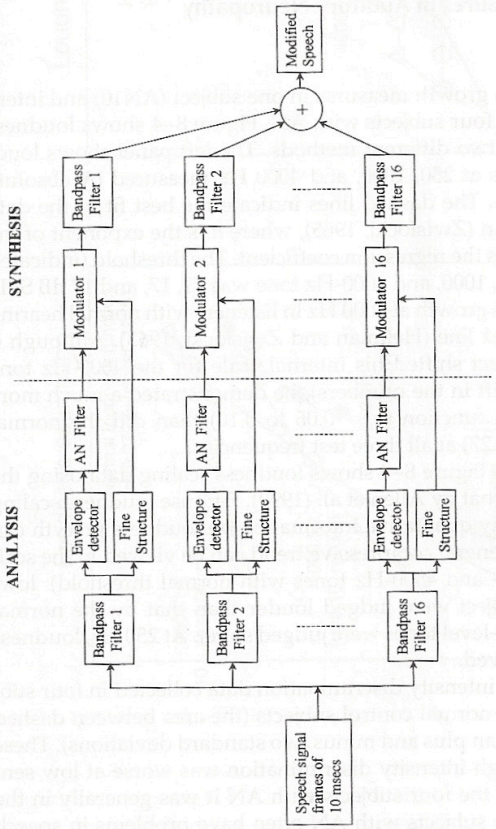
In the intensity and frequency processing experiments, tonal stimuli of 200 ms in duration were generated digitally using TDT System II (Tucker-Davis Technology). In the temporal processing experiments, a broadband

(20–14,000 Hz) white noise was generated and controlled digitally to measure temporal integration, gap detection, and temporal modulation transfer functions. The noise had a duration of 500 ms and 2.5-ms cosine-squared ramps. In the gap detection experiment, a silent interval was produced in the center of the noise. In the temporal modulation function experiment, the same 500-ms noise was presented at a maximum comfortable level determined for each subject (ranging from 29 to 77 dB SL). For the modulated signals, the presentation level was dynamically adjusted to achieve the same root-mean-square level (given the amount of modulation) as the unmodulated stimuli.

Figure 8–3 shows the flow chart for the stimuli generated for simulating auditory neuropathy in the listeners with normal hearing. The neuropathy simulation algorithm divided a speech signal into 16 third-octave bands and extracted the band-specific temporal envelope by half-wave rectification and low-pass filtering (Drullman, Festen, & Plomp, 1994). The low-pass filters were designed according to the measured temporal modulation transfer functions and covered four degrees of temporal processing impairment found in the present neuropathy subjects: mild (modulation peak sensitivity = –17 dB and 3 dB cutoff = 100 Hz), moderate (–14 dB and 50 Hz), severe (–8 dB and 25 Hz), and profound (–2 dB and 15 Hz). The low-pass-filtered temporal envelopes were used to modulate the amplitude of the fine-structure of the original acoustic signal, resulting in a temporally smeared acoustic waveform.

Procedure

We used standard procedures to collect and analyze all audiologic and neurologic data. Pure-tone averaged thresholds were based on thresholds at frequencies of 500, 1000, and 2000 Hz. Word recognition was based on NU–6 (1/2 list) test materials. This word test could not be performed on subject AN6 because of his nonnative English-speaking status. Auditory brain stem responses were recorded between vertex and the stimulated ear and identified as either absent (no definable waveforms) or abnormal (presence of only wave V). Acoustic reflexes were measured for pure-tone stimuli at 500, 1000, 2000, and 4000 Hz, presented ipsilaterally, contralaterally, or both to the stimulated ear using a Grason-Stadler GSI 33 middle ear analyzer. Otoacoustic emissions were measured with a V5 ILO92 OAE system by Otodynamics Ltd. and reported as the dB value above the noise floor. The cochlear microphonic was measured from auditory brain stem responses averaged to separate presentations of condensation and rarefaction clicks. All psychophysical tests used a three-alternative, forced-choice procedure to measure the threshold that resulted in a 70.7% correct response.



AN Filters:

Degree of Auditory Neuropathy	Cutoff frequency (Hz)	Gain (db)
Normal	238 Hz	0db
Mild	100 Hz	-3 db
Moderate	50 Hz	-6 db
Severe	25 Hz	-12 db
Profound	15 Hz	-18 db

Figure 8–3. Signal processing chart for simulations of auditory neuropathy. The degree of auditory neuropathy was based on temporal modulation transfer functions (see text and the table on the left). Because normal-hearing listeners had a peak sensitivity of −20 dB and mild neuropathy subjects had a peak sensitivity of −17 dB, a 3-dB attenuation was applied to the temporal envelope signals in the simulation. Examples of simulations can be found on the following Web site: http://www.com.uci.edu/hesp/simulations.html#neuropathy.

RESULTS

Experiments were conducted to identify the nature of loudness growth, intensity discrimination, frequency discrimination, temporal integration, gap detection, and temporal modulation transfer function in subjects with AN. An acoustic simulation of speech perception was developed based on the measured psychophysical data in auditory neuropathy subjects. The simulation produced similarly impaired temporal and speech processing results in subjects with normal hearing. Data in temporal processing from eight subjects were reported previously (Zeng, Oba, Garde, Sininger, & Starr, 1999).

Psychophysical Measures in Auditory Neuropathy

Intensity Processing

We collected loudness growth measures in one subject (AN10) and intensity discrimination in four subjects with AN. Figure 8–4 shows loudness growth measured by two different methods. The left panel shows loudness growth functions at 250, 1000, and 4000 Hz measured by absolute magnitude estimation. The dashed lines indicate the best fit to the data using a power function (Zwislocki, 1965), where n is the exponent of the power function and r is the regression coefficient. The threshold (indicated by arrows) for the 250, 1000, and 4000-Hz tone was 71, 17, and 17 dB SPL, respectively. Loudness growth at 1000 Hz in listeners with normal hearing is depicted as the solid line (Hellman and Zwislocki, 1963). Although it appears that the subject shifted his internal scale for the 4000-Hz tone (an overall vertical shift in the numbers), he demonstrated a much more compressive loudness function (n = 0.06 to 0.10) than did the normal control subjects (n = 0.27) at all three test frequencies.

The right panel in Figure 8–4 shows loudness scaling data using the identical approach to that by Allen et al. (1990). Because loudness scaling is not a ratio scale, only qualitative information on loudness growth can be inferred. Still, the general compressive trend can be viewed in the scaling data (for the 1000- and 4000-Hz tones with normal threshold): low-level tones in this subject were judged louder than that by the normal controls, whereas high-level tones were judged softer. At 250 Hz, loudness recruitment was observed.

Figure 8–5 shows intensity discrimination data collected in four subjects with AN and the normal control subjects (the area between dashed lines represents the mean plus and minus two standard deviations). These data show that although intensity discrimination was worse at low sensation levels in two of the four subjects with AN it was generally in the normal range. Because subjects with AN often have problems in speech

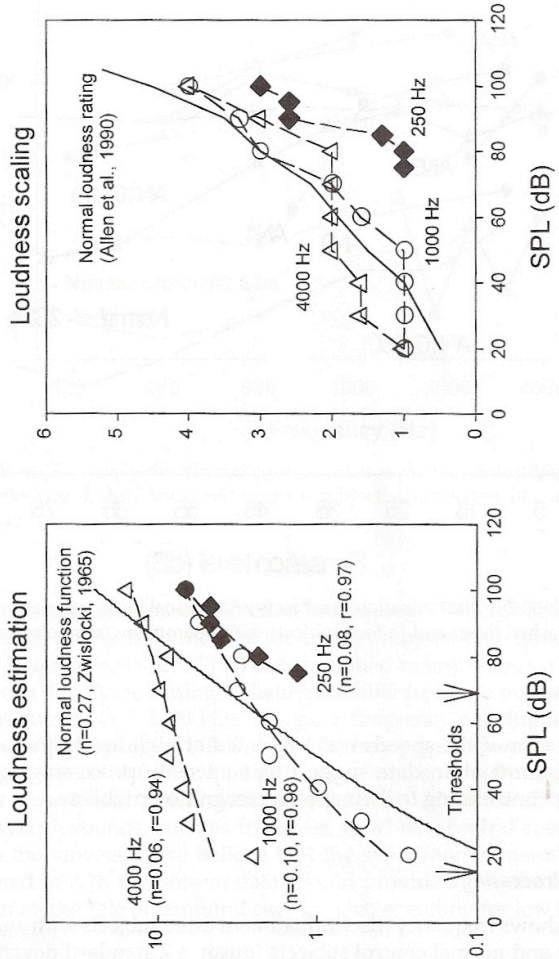


Figure 8–4. Loudness growth in one subject with neuropathy (AN10). Normal loudness growth is indicated by the solid line.

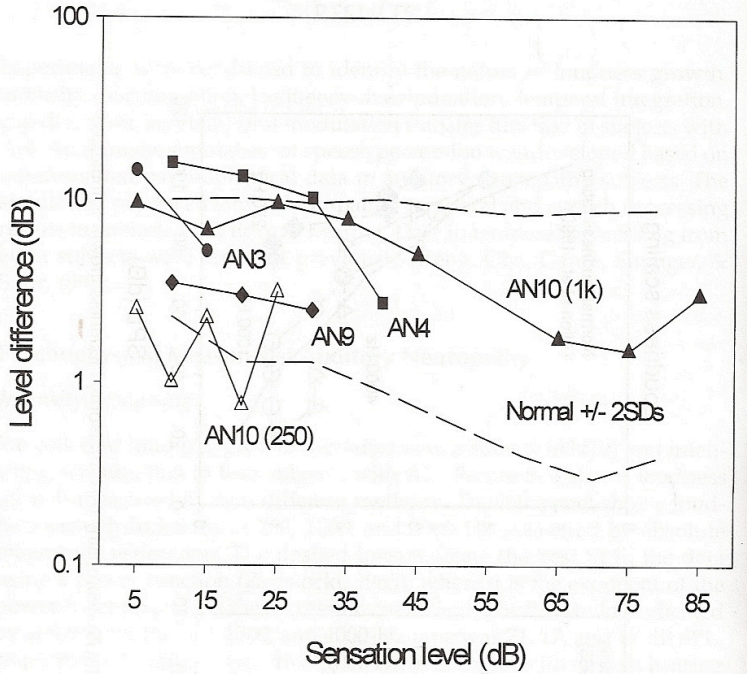


Figure 8–5. Intensity discrimination in subjects with normal hearing (mean ± 2 SD, marked by dashed lines) and in four subjects with neuropathy (symbols and solid lines).

recognition even if the speech was presented at high levels, the present intensity discrimination data suggest that intensity processing is not a major factor contributing to their speech recognition problems.

Frequency Processing

Figure 8–6 shows frequency discrimination in three subjects with auditory neuropathy and normal control subjects (mean ±2 standard deviations). For listeners with normal hearing, the difference limen (y-axis) increases as a monotonic function of frequency (x-axis). They require several Hz to notice a difference in pitch for frequencies less than 1000 Hz and several hundreds of Hz at 8000 Hz. Interestingly, the three subjects with auditory neuropathy showed a totally different pattern of results; the difference

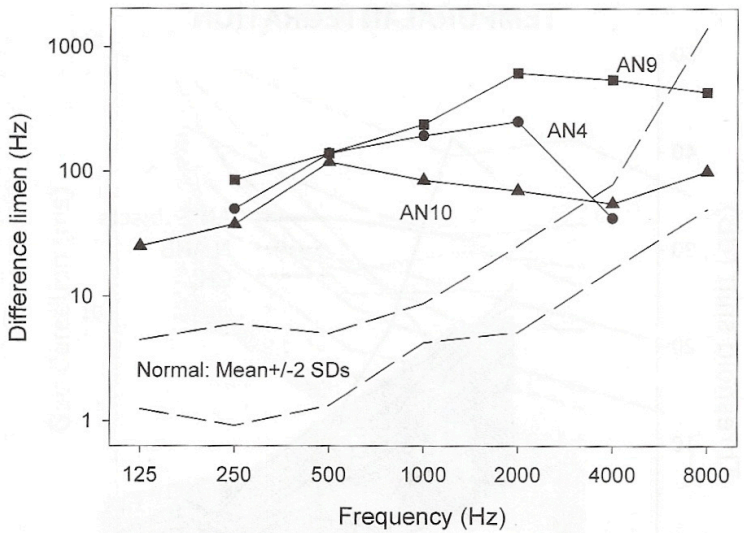


Figure 8–6. Frequency discrimination in subjects with normal hearing (mean ± 2 SD, marked by dashed lines) and in three subjects with neuropathy (symbols and solid lines).

limen was a nonmonotonic function of frequency. Their difference limens were much poorer than those of the control subjects with normal hearing at low frequencies (< 2000 Hz) but continued to improve as a function of frequency, finally becoming indistinguishable from the normal values at high frequencies (> 4000 Hz). The poor frequency discrimination in the middle frequency region (1000–3000 Hz) may pose some problem for discerning the second formant frequencies of two spectrally closely spaced vowels but should not prevent subjects with AN from distinguishing other speech sounds, such as fricatives, in which spectral cues are not as fine as the vowels. If we believe that the synchronous neural activity is disrupted in AN, the present data should provide an interesting demonstration on the role of temporal cues in pitch encoding for low frequencies (< 2000 Hz).

Temporal Processing

Figure 8–7 shows that detection thresholds for listeners with normal hearing (the shaded area) decrease at a rate of about 3 dB per doubling of

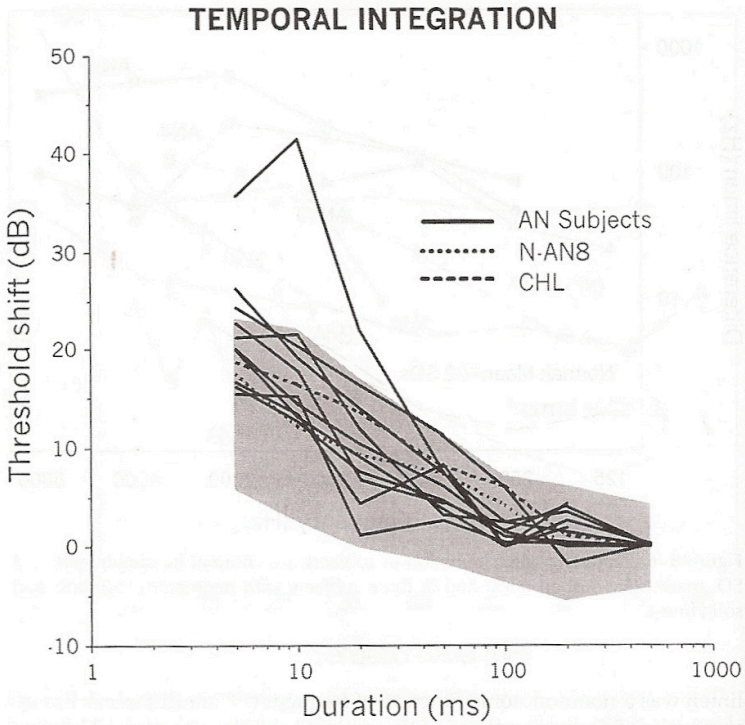


Figure 8–7. Temporal integration functions. Threshold shifts (y-axis) refer to the difference in dB between detection thresholds for noise bursts of different durations (x-axis) and that for the detection threshold at the longest duration (500 ms). Normal control data are represented as the shaded area (mean ± 2 SD). Neuropathy data are represented by solid lines. The dashed line represents the cochlear-impaired case, and the dotted line represents the healthy ear of the unilateral case.

signal duration for durations up to 100–200 milliseconds. Figure 8–7 also shows (with the exception of AN3, the original case in the Starr et al. 1991 study) that subjects with AN, as well as the two control subjects, showed normal or nearly normal temporal integration functions. Although subject AN3 had a much steeper slope of −8 dB, the remaining nine subjects with AN had an averaged slope of −4 dB, much closer to the normal value. Thus, the results from the detection of short-duration sounds apparently could not explain the poor speech recognition in subjects with neuropathy.

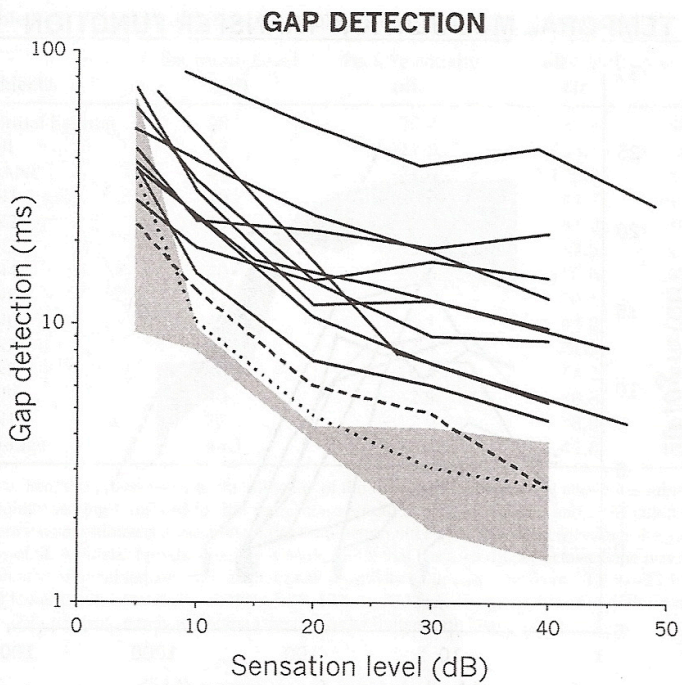


Figure 8–8. Gap detection thresholds. Detection thresholds (y-axis) are plotted as a function of sound presentation level (dB SL). Normal control data are represented as the shaded area (mean ± 2 SD). Neuropathy data are represented by solid lines. The dashed line represents the cochlear-impaired case, and the dotted line represents the healthy ear of the unilateral case.

Figure 8–8, on the other hand, shows that detecting short, silent intervals, or gaps, in acoustic signals was uniformly impaired in the subjects with AN. In both the listeners with normal hearing (shaded area) and the unilateral control, gap detection thresholds improved from 20–30 ms at low sound levels to 2–3 ms at high sound levels. The subject with cochlear impairment had slightly elevated gap detection thresholds at moderate sound levels but reached the normal range of values at the highest sound level, a pattern similar to previous studies employing listeners with cochlear impairment (Fitzgibbons & Gordon-Salant, 1987; Florentine & Buus, 1984; Moore & Glasberg, 1988). In contrast, all subjects with AN still had large deficits at the highest sound level; their gap detection thresholds were 2 to 25 times greater than normal.

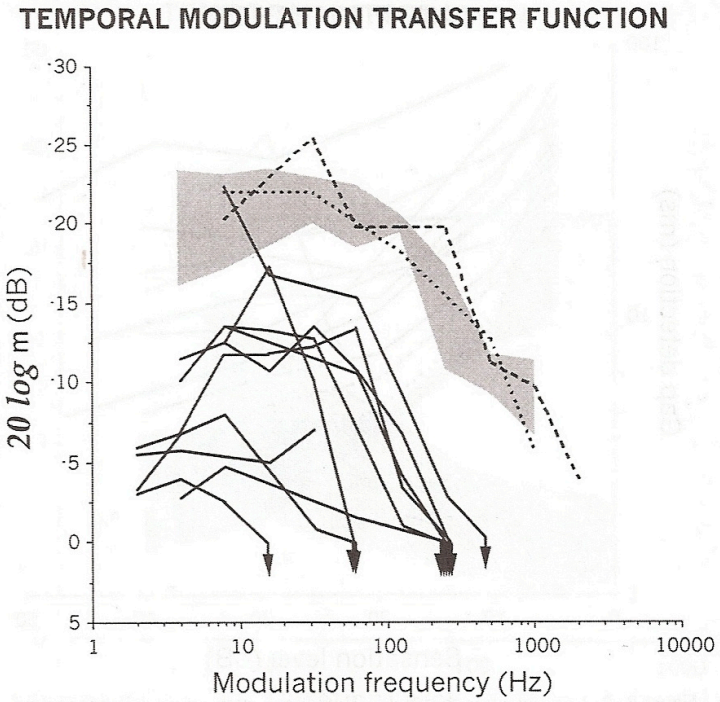


Figure 8–9. Temporal modulation transfer functions. Modulation detection thresholds (y-axis) represented as 20*log*(m) are plotted as a function of modulation frequency (x-axis). Arrows represent the fact that subjects could not reliably detect the presence of even a 100% modulated noise. Normal control data are represented as the shaded area (mean ± 2 SD). Neuropathy data are represented by solid lines. The dashed line represents the cochlear-impaired case, and the dotted line represents the healthy ear of the unilateral case.

Figure 8–9 shows the measured sensitivity to slow and fast temporal fluctuations (i.e., modulation transfer functions, see Viemeister, 1979). We modeled the modulation transfer function as a first-order Butterworth low-pass filter (Table 8–2). The listeners with normal hearing showed a low-pass function, being most sensitive (peak sensitivity = −20.4 dB) to slow temporal fluctuations and becoming less sensitive as the fluctuation rate was increased (3 dB cutoff frequency = 247 Hz). Both the unilateral and control subjects with cochlear impairment showed modulation transfer functions that were virtually indistinguishable from the normal

Table 8–2. Summary of temporal modulation transfer function parameters.

Subjects	Sensation Level (dB)	Peak Sensitivity (dB)	3-dB Cutoff (Hz)	*r*
Normal hearing	40	−20.4	237.8	0.97
CHL	37	−21.8	234.8	0.95
N-AN8	41	−21.6	175.2	0.99
AN1	40	−20.1	41.7	0.98
AN2	29	−13.8	51.2	0.99
AN3	49	−5.8	32.9	0.79
AN4	40	−3.6	106.6	0.88
AN5	42	−3.6	14.1	0.95
AN6	45	−6.1	44.8	0.82
AN7	39	−12.4	72.3	0.97
AN8	40	−16.4	14.3	0.87
AN9	40	−12.9	38.8	0.93
AN10	77	−12.9	38.8	0.90
Average	44.1	−10.8	45.6	0.91

Note. Sensation level refers to the dB value of the noise presentation level above the subject's absolute hearing threshold for the same noise stimulus. Peak sensitivity and 3-dB cutoff frequency were estimated using a first-order Butterworth filter. The coefficient (*r*) reflects the goodness of fit. A different model (Formby & Muir, 1988) that has a −3 dB per octave slope was also evaluated and yielded generally higher peak sensitivity values (ranging from −3.9 to −23.1 dB) and lower cutoff frequencies (ranging from 4 Hz to 120 Hz). The goodness of fit of this model (r = .72–.98) was slightly worse than the first-order Butterworth filter.

low-pass function. In contrast, all subjects with AN showed impaired sensitivity to both slow and fast temporal fluctuations. The average peak sensitivity at low modulation frequencies was −10.8 dB and the average 3-dB cutoff modulation frequency was 45.6 Hz. These values were about one third (−10.8 vs. −20.4 dB) and one fifth (45.6 vs. 247 Hz), respectively, of the corresponding values obtained for our control listeners.

Simulations of AN

The temporal modulation transfer function measured for the subjects with AN allowed development of simulations of auditory neuropathy in listeners with normal hearing (Drullman et al., 1994). The detailed method of simulation was described earlier (see Figure 8–3). Figure 8–10 (top panel) shows simulation results of gap detection thresholds, which increased monotonically from about 2–20 ms as the severity of the simulated AN was increased from "mild" to "profound." The actual gap thresholds from subjects with AN generally were within two standard deviations of the

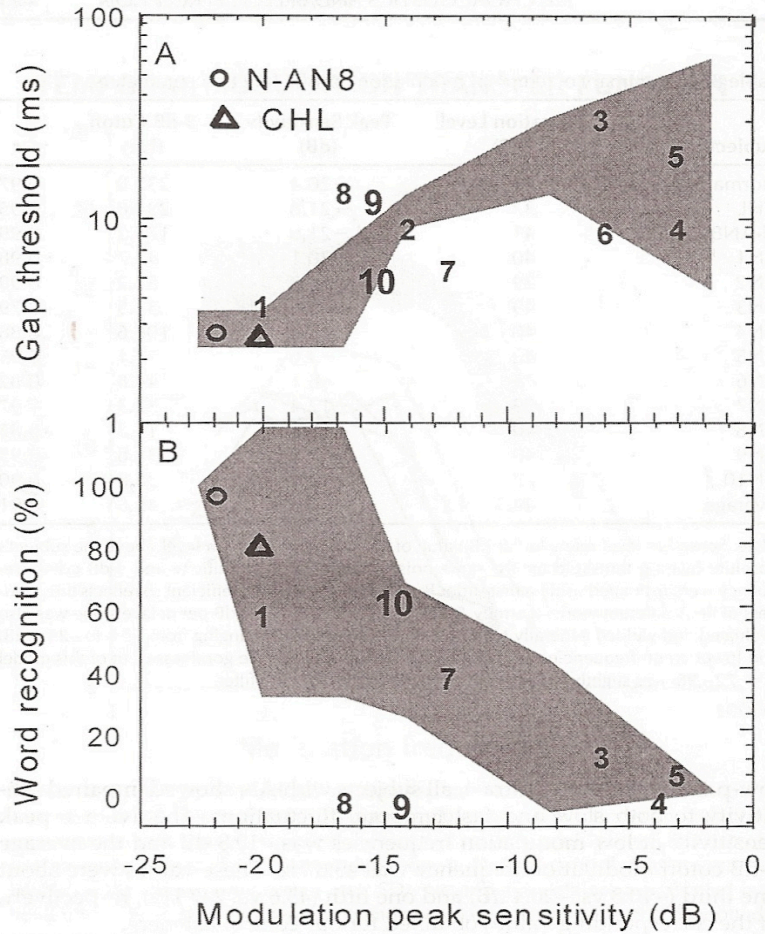


Figure 8–10. Simulations of auditory neuropathy in normally hearing listeners. **Top:** Gap detection thresholds at 40 dB SL. The gap detection threshold is plotted as a function of the severity of the auditory neuropathy, represented as the peak sensitivity of the temporal modulation transfer function (x-axis). The shaded area represents the mean ± 2 SD from normally hearing listeners who performed gap detection by listening to temporally smeared waveforms (see text for details). The digits denote gap detection thresholds measured for each of the subjects with auditory neuropathy also tested at 40 dB SL (from Figure 8–8). The symbols denote two control subjects (also from Figure 8–8). Because the listeners with normal hearing and the two control subjects all produced peak sensitivity values of about −20 dB, their x-coordinates were shifted by 1–2 dB to avoid overlap. **Bottom:** Word recognition from listeners with normal hearing who listened to both unprocessed words and temporally smeared speech sounds simulating various degrees of auditory neuropathy (see text for details). Symbols are identical to that in Figure 8–10A.

simulated thresholds. For comparison, the two control conditions also are shown for the healthy ear of the unilateral patient with AN (circle) and for the listener with cochlear impairment (triangle).

Figure 8–10 (bottom panel) shows results of simulated word recognition, which decreased monotonically to a 50%-correct level when the modulation detection threshold was elevated by about 5 dB from the normal value of –20 dB and reached 0%-correct level with about 15 dB elevations. Actual word recognition scores from eight neuropathy and two control subjects are shown in the same fashion as in the top panel of Figure 8–10. With two exceptions (AN8 and AN9), poor word recognition was consistent with the degree of impaired temporal processing in neuropathy subjects. The exceptional subjects had more severe hearing loss, suggesting that additional factors such as audibility may have contributed to the poor speech recognition in these two subjects. Adding audibility to modulation measures (temporal factors) allows excellent prediction of word recognition in patients with auditory neuropathy. The following equation reveals an $r = .93$ and $p = .01$.

$$\text{word\%} = 49.5 - .99 * \text{PTA} - 2.02 * \text{TMTF(dB)} + 0.04 * \text{TMTF(Hz)}$$

where PTA is pure-tone-averaged threshold and TMTF represents measures in temporal modulation transfer functions.

DISCUSSION

The present tests have revealed a severe temporal processing impairment in subjects with auditory neuropathy, in contrast to the relatively normal temporal processing often associated with hearing disorders due to cochlear damage (Bacon & Gleitman, 1992; Bacon & Viemeister, 1985; Fitzgibbons & Gordon-Salant, 1987; Florentine & Buus, 1984; Formby & Muir, 1988; Moore & Glasberg, 1988; Moore, Shailer, & Schooneveldt, 1992). To assess whether a lack of audibility is a confounding factor, a correlation was computed between subjects' averaged pure-tone thresholds (column 4 in Table 8–1) and their peak sensitivities to slow fluctuations (column 3 in Table 8–2). Only an insignificant correlation ($r = .23$) was found between the pure-tone average threshold and the peak sensitivity. In addition, a high-pass filter at 1000 Hz (135 dB/octave) was applied to simulate the low-frequency hearing loss in a subject with normal hearing. This produced no significant effect on modulation transfer functions (–17 vs. −16 dB, –19 vs. −19 dB, –17 vs. −17 dB, –17 vs. −18 dB at modulation frequencies 4, 8, 16, 32 Hz, respectively). These analyses suggest a true temporal processing deficit in AN, rather than a byproduct of hearing loss due to limited bandwidths at low sensation levels. This conclusion

received additional support from the simulation results that the degree of temporal processing impairment can account for the abnormal speech recognition observed in subjects with neuropathy. The present finding of a close coupling between temporal and speech processing deficits complements the recent emphasis on speech recognition using temporal cues in general (e.g., Shannon, Zeng, Wygonski, Kamath, & Ekelid, 1995; Van Tasell, Soli, Kirby, & Widin, 1987) and amplitude modulations in particular (Arai & Greenberg, 1998; Greenberg & Arai, 1998).

Desynchronous Neural Activity

Although the exact physiologic process underlying AN is not clear, there is evidence linking the observed temporal processing impairment to demyelination in the auditory nerve. For example, the failure to detect the evoked auditory brain stem responses in these patients has been related to the loss of discharge synchrony secondary to demyelination of the auditory nerve (Kalaydjieva et al., 1998; Starr et al., 1998). Demyelinated nerve fibers have slowed conduction velocities, which vary as a function of the extent of demyelination in each fiber, resulting in disrupted discharge synchrony both within a neuron and across a neural population.

Figure 8–11 presents a phenomenologic model of the disrupted synchronous neural activity and its account for the present psychophysical data. It could be assumed that the main effect of the desynchronous activity is a smeared temporal representation of the acoustic stimulus (see the difference between the sharp waveform in the physical representation and its smeared version in the internal representation). If the listening task was to detect merely either presence (top trace) or absence (bottom trace) of a sound, as in the case of the temporal integration experiment, then this smeared representation would not present a difficult perceptual problem. If the task was to discriminate two different waveforms, however, one with a gap (top trace) and one without gap (bottom trace), then the smearing in the internal representations would result in a much more difficult perceptual task. A quantitative prediction of the psychophysical data is not possible at present and requires much better understanding of the exact physiologic mechanisms of auditory neuropathy (see Harrison, Chapter 4, and Starr, Picton, and Kim, Chapter 5, in this book).

Temporal Processing Deficits

Temporal processing deficits have also been observed in elderly listeners (Gordon-Salant & Fitzgibbons, 1993), patients with multiple sclerosis (Levine, 1993), and children with learning disabilities (Kraus et al., 1996; Merzenich et al., 1996; Tallal & Piercy, 1973; Tallal et al., 1996; Wright et al., 1997). Similar to the present results, those previous studies also found a

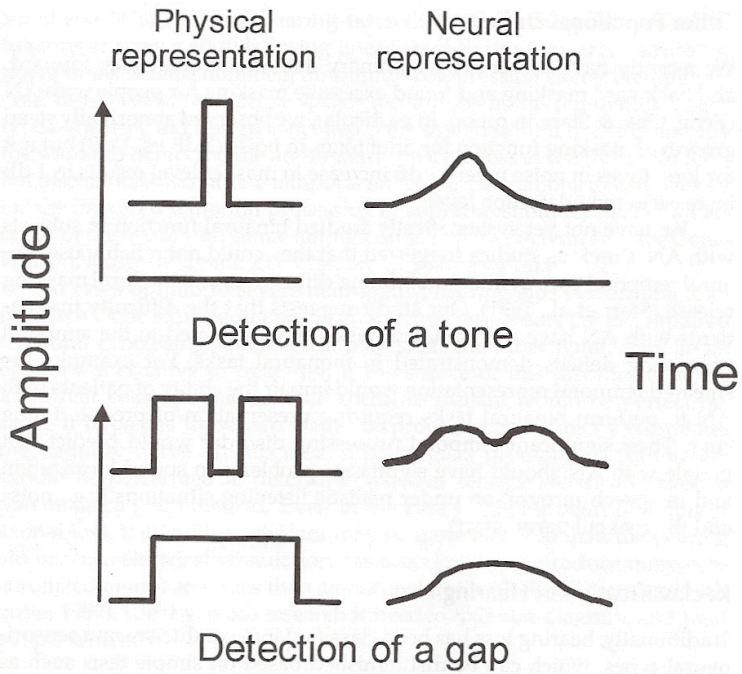


Figure 8–11. A phenomenologic model of auditory neuropathy. This simple model assumes that desynchronous neural activity results in a smeared internal representation of a physical stimulus. The smearing does not affect the detection of a tone (top panel) because the task requires only an all-or-none decision. The smearing can cause a major problem in gap detection (bottom panel) if the task requires finer discrimination of two different waveforms, however.

close relationship between temporal processing and speech recognition deficits, despite a peripheral origin of the temporal processing impairment in auditory neuropathy and a central origin in the other cases. Our study suggests that simple behavioral gap and temporal modulation detection tasks, when used in combination with other audiologic and neurologic tests, can distinguish the extent of temporal processing impairment due to auditory neuropathy in those communication disorders. For example, abnormal temporal processing may reveal the presence of disordered auditory nerve synchrony when there is a concomitant cochlear hearing loss, as occurs in aging.

Other Functional Deficits

We recently have presented preliminary data in simultaneous, forward, and backward masking and found excessive masking for people with AN (Zeng, Oba, & Starr, in press). In particular, we observed abnormally steep growth of masking function for brief tones in noise (2 dB vs. 1 dB) but not for long tones in noise where 1 dB increase in masker level results in 1 dB increase in tone-detection level.

We have not yet systematically studied binaural function in subjects with AN. Previous studies suggested that they could not reliably use binaural temporal cues in interaural timing differences and binaural masking release (Starr et al., 1991). Our study suggests that the difficulty that patients with AN have with binaural tasks may be related to the temporal processing deficits demonstrated in monaural tasks. For example, the smeared temporal representation would impair the ability of patients with AN to perform binaural tasks requiring preservation of precise timing cues. Their significant temporal processing disorder would predict that people with AN should have significant problems in sound localization and in speech recognition under realistic listening situations (e.g., noise and the cocktail party effect).

Reclassification of Hearing Loss

Traditionally, hearing loss has been classified into conductive and sensorineural types, which can be distinguished based on simple tests such as air- versus bone-conduction thresholds. Sensorineural hearing loss can be further classified into cochlear and retrocochlear loss, with the cochlear loss typically demonstrating loudness recruitment and no tone decay. Retrocochlear loss typically refers to the presence of acoustic tumors. Where AN will fall in this classification scheme will depend on further studies isolating the site of lesion. If AN is mainly due to demyelination in the auditory nerve, then it would be best classified into the retrocochlear loss category. If it is mainly due to inner hair cell loss or a dysfunction in the synaptic transduction, then it would be better classified as cochlear loss. Both Starr and Harrison present detailed analysis of both scenarios in this volume. In any case, AN clearly presents a type of hearing loss that is distinct from outer hair cell loss. As we gain additional knowledge about AN, traditional hearing loss classification will have to be modified and refined.

Rehabilitation Strategies

Our results also bear on the failure of conventional hearing aids to help people with AN, who often complain: "I can hear you, but I cannot under-

stand you." Conventional hearing aids either do not change temporal fluctuations of speech sounds (using linear amplification) or even reduce the fluctuations when a nonlinear amplitude-compression circuit is employed (Van Tasell, 1993). To improve speech recognition in this population, a new type of hearing aid design is needed. This design should not only amplify the sound to overcome the audiometric hearing loss at the threshold level but also should accentuate temporal envelope fluctuations to compensate for the impaired temporal processing at suprathreshold levels. For an account of the use of traditional amplification in patients with AN, see Cone-Wesson, Rance, and Sininger, Chapter 12, in this volume.

In cases of mild to severe neuropathy, hearing aids performing temporal envelope expansion may be enough to compensate for the impaired temporal processing. Cochlear implants, on the other hand, may be an effective alternative to treat AN. If the source of this disorder is at the inner hair cells or synapse, then cochlear implant would be perfect because it bypasses these two stages of processes and directly stimulates the auditory nerve. In this case, a presurgical promontory stimulation should be performed to determine whether reliable electrically evoked potentials can be obtained. Even in the case of neural demyelination or axonal loss, the cochlear implant may be more effective than the hearing aid because electrical stimulation has been known to produce more synchronized neural activities than any acoustic stimulation (Dynes and Delgutte, 1992). Clearly, more research is needed to better diagnose and treat people with AN.

SUMMARY

Our data show that people with AN uniformly have a deficit in temporal processing and poor frequency discrimination at low and moderate frequencies while having relatively normal intensity processing and normal frequency discrimination at high frequencies. Correlation analysis suggests that the severe impairment in temporal processing is likely the major factor contributing to poor speech recognition in people with this disorder. This conclusion is further supported by a simulation of this temporal processing impairment, which produce similar speech recognition deficits in listeners with normal hearing. Our data not only demonstrate the importance of neural synchrony for auditory perception, but also provide guidance for better diagnosis and treatment of auditory neuropathy.

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