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Authors

Zeng, F G
Liu, S

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Speech Perception in Individuals With Auditory Neuropathy

Fan-Gang Zeng
Sheng Liu
University of California, Irvine

Purpose: Speech perception in participants with auditory neuropathy (AN) was systematically studied to answer the following 2 questions: Does noise present a particular problem for people with AN? Can clear speech and cochlear implants alleviate this problem?
Method: The researchers evaluated the advantage in intelligibility of clear speech over conversational speech in 13 participants with AN. Of these participants, 7 had received a cochlear implant. Eight sentence-recognition experiments were conducted to examine the clear speech advantage in 2 listening conditions (quiet and noise) using 4 stimulation modes (monaural acoustic, diotic acoustic, monaural electric, and binaurally combined acoustic and electric stimulation).
Results: Participants with AN performed more poorly in speech recognition in noise than did the normal-hearing, cochlear-impaired, and cochlear implant controls. A significant clear speech advantage was observed, ranging from 9 to 23 percentage points in intelligibility for all listening conditions and stimulation modes. Electric stimulation via a cochlear implant produced significantly higher intelligibility than acoustic stimulation in both quiet and in noise. Binaural hearing with either diotic acoustic stimulation or combined acoustic and electric stimulation produced significantly higher intelligibility than monaural stimulation in quiet but not in noise.
Conclusions: Participants with AN most likely derive the clear speech advantage from enhanced temporal properties in clear speech and improved neural synchrony with electric stimulation. Although the present result supports cochlear implantation as one treatment choice for people with AN, it suggests that the use of innovative hearing aids may be another viable option to improve speech perception in noise.

KEY WORDS: auditory neuropathy, binaural hearing, clear speech, cochlear implant, speech perception

Hearing loss can be classified by conductive versus sensorineural loss or peripheral versus central processing disorders. These different types of hearing loss produce different perceptual consequences and may require different treatment strategies for optimal performance. Auditory neuropathy (AN) is a recently described hearing disorder that has unique pathologies and perceptual consequences (Starr et al., 1991, 2003; Starr, Picton, Sininger, Hood, & Berlin, 1996). One main characteristic of AN is the disrupted auditory nerve activity with concurrently normal or nearly normal cochlear amplification function. Clinically, the disrupted auditory nerve activity is reflected by highly distorted or absent auditory brainstem responses, whereas the normal cochlear amplification function is reflected by the presence of otoacoustic emission and/or cochlear microphonics (Hood, Berlin, Bordelon, & Rose, 2003; Rance et al., 1999; Starr et al., 1996). The other main characteristic of AN is a significantly impaired capacity for temporal processing and difficulty in speech understanding, particularly in noise, that is disproportionate to the degree of hearing loss measured by pure-tone thresholds (Kraus et al., 2000; Rance, Cone-Wesson, Wunderlich, & Dowell, 2002; Rance, McKay, & Grayden, 2004; Zeng, Kong, Michalewski, & Starr, 2005; Zeng, Oba, Garde, Sininger, & Starr, 1999).

The prevalence of AN has been estimated to affect about 10% of infants who failed hearing screening (Rance et al., 1999). The etiologies of AN are diverse, involving drug agents (e.g., carboplatin), toxic/metabolic processes (e.g., hyperbilirubinemia and anoxia), infection (e.g., mumps), hereditary neuropathies (e.g., Charcot-Marie-Tooth syndrome), hereditary disorders affecting inner hair cells, and other unknown causes (Amatuzzi et al., 2001; Butinar et al., 1999; De Jonghe et al., 1999; Deltenre, Mansbach, Bozet, Clercx, & Hecox, 1997; Harrison, 1998; Salvi, Wang, Ding, Stecker, & Arnold, 1999; Shapiro, 2004; Starr et al., 2004). AN may result from a loss of inner hair cells (IHC), dysfunction of the IHC-nerve synapses, neural demyelination, axonal loss, or a possible combination of multiple sites. These pathologies may be mixed with the traditional cochlear loss involving outer hair cells and/or central processing disorders involving the brainstem and cortex, complicating the classification of AN (Rapin & Gravel, 2003). Because one possible neural mechanism underlying AN symptoms is desynchronized discharges in the auditory nerve fibers, it has been suggested that AN be termed "auditory dys-synchrony" (Berlin, Morlet, & Hood, 2003). Management of AN continues to be difficult as there is no standard treatment. Because conventional hearing aids have achieved limited success (Deltenre et al., 1999; Rance et al., 2002), cochlear implants are quickly becoming the choice of treatment for AN (Berlin et al., 2003; Buss et al., 2002; Miyamoto, Kirk, Renshaw, & Hussain, 1999; Peterson et al., 2003; Shallop, Peterson, Facer, Fabry, & Driscoll, 2001; Trautwein, Sininger, & Nelson, 2000).

It is important to explore alternative strategies that are much less invasive than cochlear implants but may benefit individuals with AN, particularly for those who have relatively mild AN. One effective means of improving speech intelligibility is to speak clearly (Picheny, Durlach, & Braida, 1985, 1986, 1989). When talkers are instructed to speak clearly, they usually produce more intelligible speech than they would when interacting in casual conversation. The higher intelligibility in clear speech than in conversational speech is likely a result of acoustic and phonetic differences between these two styles of speech. These differences include reduced speaking rate, increased energy in the 1000–3000 Hz range, enhanced temporal modulations, expanded voice pitch range and vowel space (Ferguson & Kewley-Port, 2002; Krause & Braida, 2002, 2004; Liu, Del Rio, Bradlow, & Zeng, 2004; Payton, Uchanski, & Braida, 1994; Uchanski, Choi, Braida, Reed, & Durlach, 1996). This line of research has recently received renewed interest as the clear speech advantage can be extended to the "fast" speaking rate (Krause & Braida, 2004) and the benefit has been demonstrated in diverse populations including those with learning disabilities, auditory neuropathy, and cochlear implants (Bradlow, Kraus, & Hayes, 2003; Kraus et al., 2000; Liu et al., 2004). Because a temporal processing deficit is a hallmark of AN (Zeng, Kong, et al., 2005; Zeng et al., 1999), the enhanced temporal properties in clear speech may be especially beneficial to individuals with AN.

The primary goal of the present study was to systematically compare performance between clear and conversational speech in participants with AN. So far, only one case study has reported the clear speech advantage in 1 participant with AN (Kraus et al., 2000). The clear speech advantage was defined as the intelligibility difference between clear and conversational speech under otherwise identical listening conditions. These listening conditions included speech perception in quiet and in noise, with and without cochlear implants, and monaural and binaural hearing. The secondary goal was to shed light on critical questions that have not yet been fully addressed in AN, including (a) Does noise present a particular problem? (b) Do cochlear implants help in quiet and in noise? and (c) Does binaural hearing (diotic acoustic stimulation or combined acoustic and electric stimulation) provide additional benefits in quiet and in noise?

Method

Participants

We studied 13 participants who had been clinically diagnosed with AN. Table 1 lists biographical and audiological information for these participants. The ages ranged from 9 to 41 years, with a mean of 24. Eight were female and 5 were male. The hearing loss in terms of pure tone thresholds ranged from nearly normal (25 dB in AN10's left ear) to profound (AN23 and AN24), with an average of 55 dB HL for the left ear and 54 dB HL for the right ear. Different from the high frequency loss configuration typically seen in most elderly persons, many of the participants with AN had low-frequency or flat hearing loss, including 4 participants who had essentially normal hearing at high frequencies (>2000 Hz in AN7, AN10, AN13, and AN16). Open-set speech recognition in quiet ranged from 0% to 90% correct, with a mean score of 29% for the left ear and 42% for right ear, which was much lower than expected by the average moderate pure-tone hearing loss. All participants had measurable otoacoustic emission and/or cochlear microphonics, but absent acoustic reflex and/or auditory brainstem responses. When tested, the participants

Table 1. Information on participants with auditory neuropathy (AN).

Participant	Age	Gender	PTA (L)	PTA (R)	Speech (L)	Speech (R)	Listening conditions
AN5	21	F	64	68 (CI)	0	DNT	1a, 3a, 3b, 4a, 4b
AN7	27	M	45	55	40	DNT	1a, 1b
AN10	26	M	25	33	90	64	1a, 1b, 2a, 2b
AN13	29	F	31	40 (CI)	60	60	1a, 3a, 3b, 4a, 4b
AN16	18	F	29	55	16	20	1a
AN17	18	F	51	50	12	44	1a, 2a
AN18	19	M	44	40	25	20	1a, 1b, 2a
AN19	12	F	68	43	68	64	1a, 1b, 2a, 2b
AN20	36	F	73 (CI)	66	0	24	1a, 3a, 3b
AN22	39	F	78	NA (CI)	DNT	DNT	3a, 3b, 4a, 4b
AN23	41	F	93	NA (CI)	16	DNT	3a, 3b
AN24	9	M	NA (CI)	93	0	DNT	3a, 3b
AN25	21	M	NA (CI)	NA (CI)	20	DNT	3a
M	24		55	54	29	42	

Note. Age refers to the age of the participant at the time of the experiments; PTA = pure-tone threshold average in dB HL measured presurgically at all tested frequencies from 125 to 8000 Hz; Speech refers to the percentage correct scores from the Hearing in Noise Test sentences obtained clinically; NA = not available; DNT = did not test; CI = cochlear implant. Listening conditions refer to 1a (monaural acoustic stimulation in quiet), 1b (monaural acoustic stimulation in noise), 2a (diotic acoustic stimulation in quiet), 2b (diotic acoustic stimulation in noise), 3a (monaural electric stimulation in quiet), 3b (monaural electric stimulation in noise), 4a (binaurally combined acoustic and electric stimulation in quiet), and 4b (binaurally combined acoustic and electric stimulation in noise).

always had evoked cortical potentials but with a delayed N100 component (12 of 13 cases, except for AN25 who was not tested) and normal brain imaging results (6 of 13 cases). Two participants (AN13 and AN18) had accompanying peripheral neuropathy as determined by standard neurological tests (Starr et al., 1996).

Seven of the 13 participants with AN received cochlear implantation, 1 of whom received bilateral cochlear implants (AN25). Table 2 shows additional information regarding these cochlear-implant users. Age at onset of deafness, as reported by the participants, ranged from 3 to 39 years of age. Six participants received the Nucleus device and 1 received the MED-EL device, with the duration of implant use ranging from 1 to 14 years.

The participants were coded in the same way as in previous studies (Zeng, Kong, et al., 2005; Zeng et al., 1999). Because of the varied and limited availability of participants with AN, we were not able to collect data from all experimental conditions in all participants. The availability was particularly limited for participants with AN who had cochlear implants. Table 1 also lists the listening conditions for each participant.

Table 2. Additional information on 7 participants with AN and cochlear implants.

Participant	CI age (yr)	Deaf duration (yr)	Etiology	CI use (yr)	Device	Strategy
AN5	20	16	Unknown	1	MED-EL	CIS
AN13	27	10	Unknown	2	N24	ACE
AN20	35	18	Hereditary	1	N24	ACE
AN22	38	18	Unknown	1	N24	ACE
AN23	39	25	Hereditary	2	N24	ACE
AN24	3	3	Unknown	6	N24	ACE
AN25 (L/R)	7/17	7/17	Hyperbili-rubinemia	14/4	N22/N24	SPEAK/ACE

Note. Participant AN25 received bilateral cochlear implants, with the left ear receiving the Nucleus-22 device and the right ear receiving the Nucleus-24 device. CI age = age at which the surgery was performed. Duration of deafness was self-reported. Etiology included a hereditary cause in 2 participants, hyperbilirubinemia in 1 participant, and was unknown in 4 participants. CI speech processing strategies included continuous-interleaved sampling (CIS) for the MED-EL device, spectral peak extraction (SPEAK) for the Nucleus-22 device, and advanced combination encoding (ACE) for the Nucleus-24 device.

Stimuli

The stimuli used included speech sentences recorded in clear and conversational speech styles. These sentences were modified from the original Bamford-Kowal-Bench (BKB) sentences used for British children (Bench & Bamford, 1979). Many of the BKB sentences were used by the Hearing in Noise Test (HINT) sentences (Nilsson, Soli, & Sullivan, 1994). The modified BKB sentences were recorded in the Phonetics Laboratory at Northwestern University (Bradlow et al., 2003) and further segmented and edited in the Hearing and Speech Research Laboratory at University of California, Irvine (Liu et al., 2004) using a 150-Hz high-pass filter to remove occasional breathing noise. Only sentences recorded by a female were used in the present study. Acoustic information regarding this female talker was presented by Liu et al. (2004), including an average sentence duration of 3.3 s for clear speech and 1.5 s for conversational speech. The 144 sentences were separated into 18 lists, each containing 8 sentences and with an average of 26 key words. We noted that the lists were half the size of the typical lists, consequently increasing the expected amount of variability in the data for each listener. The sentences in each list were either clear or conversational speech. In experiments involving background noise, the sentences were individually mixed with a speech-spectrum-shaped noise at signal-to-noise ratios (SNRs) from 0 to 15 dB in 5 dB steps. The speech-spectrum-shaped noise was produced by passing white noise through a 10th-order linear predictive coding (LPC) spectral envelope filter (derived from combined clear and conversational speech sentences). The sentences were presented at the most comfortable loudness level (>60 dBA) in both quiet and noise conditions. The noise level was varied to produce different SNRs. A 1000-Hz pure tone with its root mean square (RMS) level identical to the normalized RMS level in speech and noise stimuli was used as the calibration signal during all phases of the experiments.

Procedure

All participants were tested in a double-walled, sound-treated booth (Industrial Acoustics Company, Inc.). All participants were presented with stimuli from a speaker placed 0.75 m away. A sound level meter was used to measure the signal and noise levels at the participant's position. Monaural listening conditions were tested followed by binaural listening conditions. In the monaural condition with acoustic hearing, one ear was plugged with a MAXLITE hearing protector (Howard Leight Industries), producing 34, 38, and 45 dB attenuation at 125, 1000, and 8000 Hz, respectively. In the monaural condition with electric hearing, the implant was turned off when the ear with neuropathy was tested; conversely the ear with neuropathy was plugged when the ear with an implant was tested.

Several precautions were adopted in the test procedure. First, clear and conversational speech sentences were presented in a randomized order in each testing session to minimize the session-to-session variability. Second, to avoid potential repetition effects on intelligibility, each sentence was used only once for a given participant over the course of the entire experiment. Third, to familiarize the participants with the test materials and procedures, a short session with five sentences in quiet was conducted at the beginning of each test session.

For formal data collection, the participants were asked to type the sentence presented via a keyboard and were instructed to double-check the spelling before entering the answer. A computer program automatically calculated the recognition accuracy score based on the number of the key words correctly identified. Each experimental condition had eight sentences, with each sentence containing three or four key words, and took about 5 min to complete. The reported result was the averaged score from these 8 sentences.

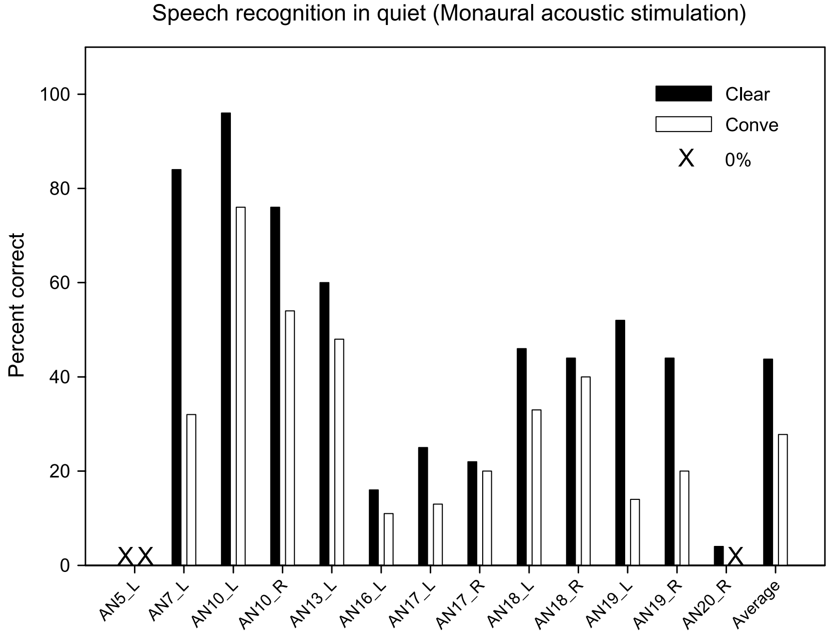
Results

We present the data according to stimulation mode: (a) monaural acoustic stimulation, (b) diotic acoustic stimulation, (c) electric stimulation via a cochlear implant, and (d) binaurally combined acoustic and electric stimulation. In each section, the data in quiet are presented first followed by the data in noise. The focus of the data analysis is to compare the performance between clear and conversational speech. Additional data analysis is in the Discussion section to test the following specific hypotheses: (a) noise poses a significantly greater problem in participants with AN than in participants with normal hearing, (b) electric stimulation produces better intelligibility than acoustic hearing, and (c) binaural stimulation produces better intelligibility than monaural stimulation.

Experiment 1: Monaural Acoustic Stimulation

Figure 1 shows percentage correct scores of clear speech (filled bars) and conversational speech (unfilled bars) obtained in quiet from 9 individual participants with AN, with scores from both left and right ears being obtained in 4 participants with AN (AN10, AN17, AN18, and AN19). The cross symbol ("X") represents a score of 0% correct, which was obtained in AN5's left ear for both clear and conversational speech and in AN20's right ear for conversational speech only.

Figure 1. Speech recognition in quiet using monaural acoustic stimulation from 13 ears in 9 participants with auditory neuropathy (AN). The filled bars represent the clear speech score, whereas the open bars represent the conversational speech scores. The cross ("X") symbols represent 0% intelligibility. The rightmost bars show the average data. The difference between the filled and open bars represents the clear speech advantage. The same format is be used in the remaining figures.



On average, the clear speech produced 44% correct intelligibility (*SD* = 30%), whereas the conversational speech produced 28% correct intelligibility (*SD* = 22%). The 16 percentage point clear speech advantage was significant, *F*(1, 12) = 14.3, $p < .01$.

Figure 2 shows percentage correct scores of clear speech (filled bars) and conversational speech (unfilled bars) obtained in noise at 0, 5, 10, and 15 dB SNRs (different panels) from 6 ears in 4 participants with AN. The cross symbol ("X") represents a score of 0% correct. With the exception of two cases (AN19_R at 5 dB SNR and AN18_L at 10 dB SNR), all remaining 12 cases showed a clear speech advantage. Because of the limited number of participants tested at each SNR, the clear

Figure 2. Speech recognition in noise using monaural acoustic stimulation. The four panels represent signal-to-noise ratios (SNRs) at 0, 5, 10, and 15 dB, respectively.

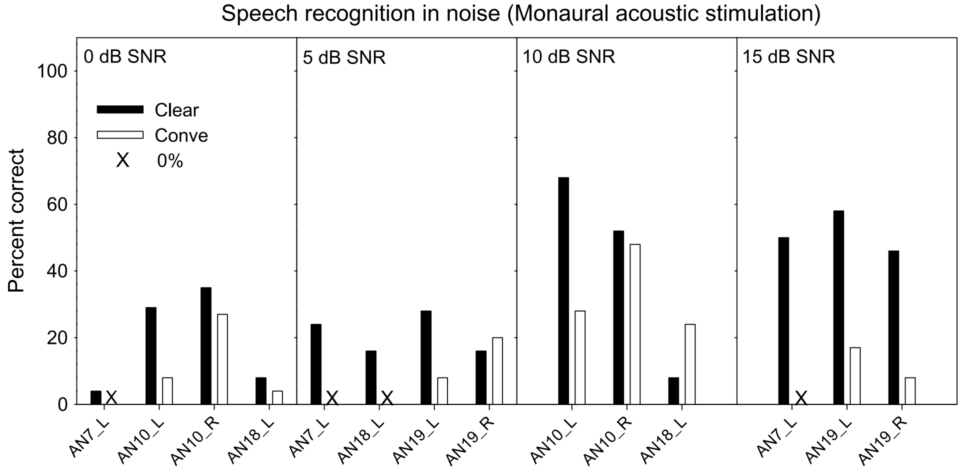
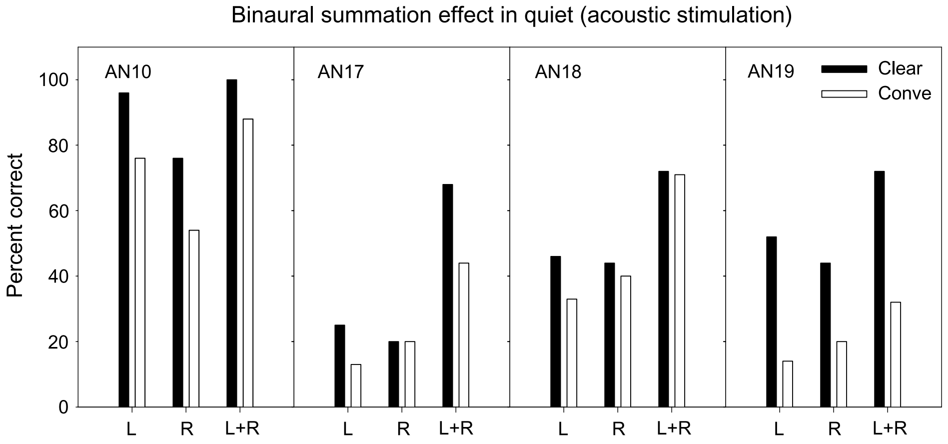


Figure 3. Speech recognition in quiet using diotic acoustic stimulation. The four panels represent intelligibility data collected in participants AN10, AN17, AN18, and AN19, respectively. L = left ear stimulation; R = right ear stimulation; L + R = diotic stimulation.



speech advantage was averaged across all noise conditions, resulting in a significant difference of 18 percentage points between the clear and conversational speech scores, $F(1, 13) = 12.0, p < .01$.

Experiment 2: Diotic Acoustic Stimulation

Figure 3 shows percentage correct scores of clear speech (filled bars) and conversational speech (unfilled bars) obtained in quiet from the left ear, the right ear, and both ears in 4 participants with AN (AN10, AN17, AN18, and AN19). Averaged across all ear conditions, clear speech produced 60% correct intelligibility (SD = 25%), whereas conversational speech produced 42% correct intelligibility (SD = 22%). The 18 percentage point clear speech advantage was significant, $F(1, 11) = 21.5, p < .01$. Stimulation mode was a significant factor, $F(2, 8) = 16.9$, $p < .05$, with diotic stimulation producing significantly better performance than the monaural left or right ear stimulation: 78% for diotic stimulation versus 55% and 46% for left- and right-ear stimulation in clear speech perception; 59% for diotic stimulation versus 34% and 34% for left- and right-ear stimulation in conversational speech perception ($p < .05$). There was no significant difference in scores between left and right ear stimulation ($p > .05$). The average binaural summation effect was 28 percentage points for clear speech and 25 percentage points for conversational speech.

Figure 4 shows percentage correct scores of clear speech (filled bars) and conversational speech (unfilled

Figure 4. Speech recognition in noise using diotic acoustic stimulation. The four panels represent intelligibility data collected in participant AN10 at 0 and 10 dB SNRs (left two panels) and AN19 at 5 and 15 dB SNRs (right two panels), respectively.

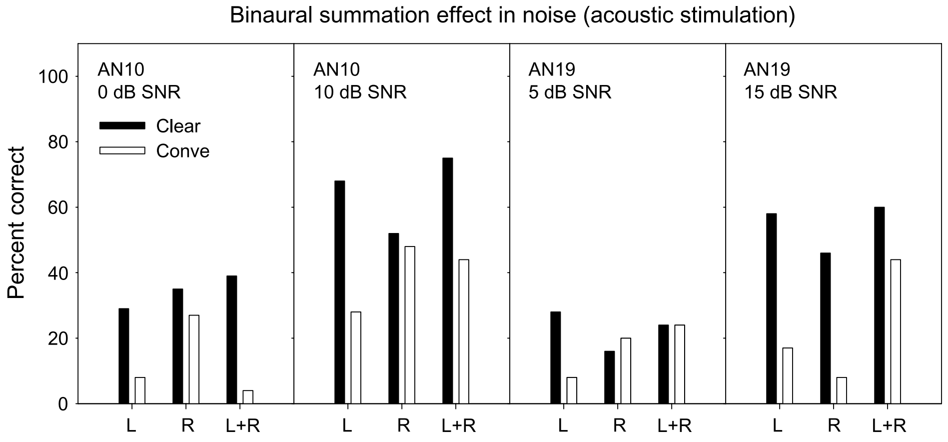
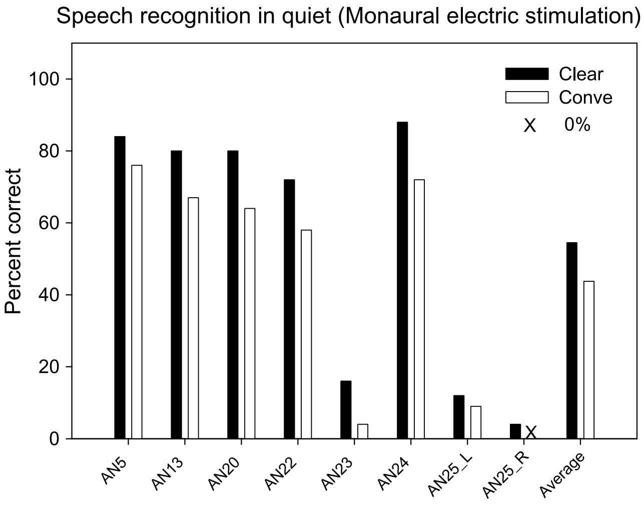


Figure 5. Speech recognition in quiet using monaural electric stimulation from 8 ears in 7 participants with AN.



bars) obtained in noise at two SNRs in 2 participants (AN10 and AN19). An analysis of variance (ANOVA) showed a significant effect for speech style, $F(1, 3) = 13.9$, $p < .05$, but not for binaural summation, $F(2, 6) = 1.7$, $p = .25$. The clear speech advantage was 21 percentage points when averaged across three stimulation modes and all noise conditions.

Experiment 3: Monaural Electric Stimulation

Figure 5 shows percentage correct scores for clear speech (filled bars) and conversational speech (unfilled bars) obtained in quiet from 8 implanted ears in 7 individual participants with AN, including AN25 who had received bilateral cochlear implants. The cross symbol ("X") represents a score of 0% correct, which was obtained only with conversational speech in AN25's right ear. Note that 3 participants (AN5, AN13, and AN20) had comparable acoustic-stimulation data (see Figure 1), allowing us to test the hypothesis that electric stimulation provides a significant advantage over acoustic stimulation in participants with AN. This hypothesis is explicitly tested in Experiment 4 and discussed in the Discussion section. On average, clear speech produced 55% correct intelligibility (SD = 37), whereas conversational speech produced 44% correct intelligibility (SD = 33%). The 11 percentage point clear speech advantage was significant, $F(1, 7) = 34.7$, $p < .01$.

Figure 6 shows percentage correct scores of clear speech (filled bars) and conversational speech (unfilled bars) obtained in noise at 0, 5, 10, and 15 dB SNRs (different panels) from 6 participants with AN who had cochlear implants. The cross symbol ("X") represents a score of 0% correct. With the exception of 1 case (AN24 at 15 dB SNR), all remaining 14 cases showed a clear speech advantage. Averaged across all noise conditions, the clear speech advantage was significant at 20 percentage points, $F(1, 14) = 49.3$, $p < .01$.

Experiment 4: Binaurally Combined Acoustic and Electric Stimulation

Figure 7 shows percentage correct scores of clear speech (filled bars) and conversational speech (unfilled bars) obtained in quiet from acoustic stimulation (AS), electric stimulation (ES), and binaurally combined stimulation (AS + ES) in 3 participants with AN. The cross ("X") represents a score of 0% correct. On average, the clear

Figure 6. Speech recognition in noise using monaural electric stimulation. The four panels represent SNRs at 0, 5, 10, and 15 dB, respectively.

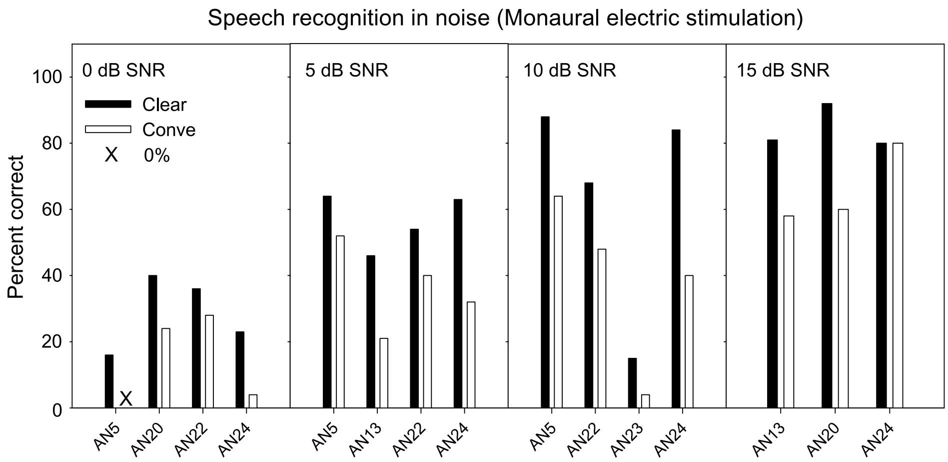
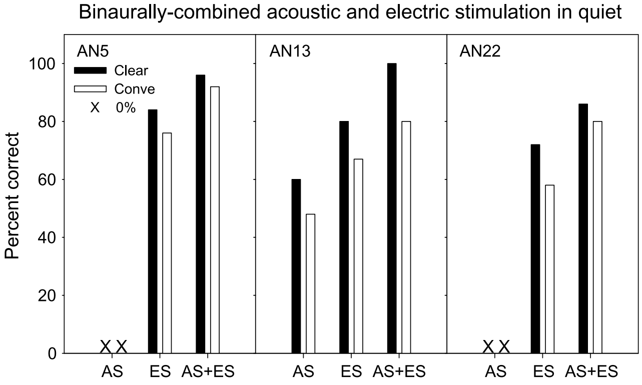


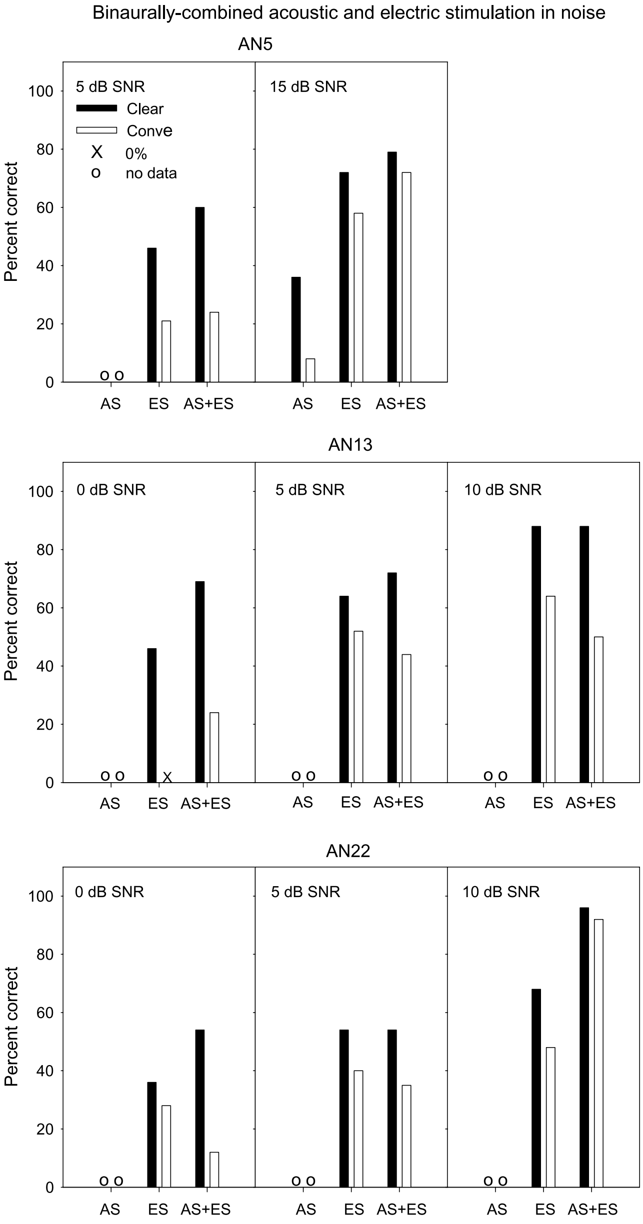
Figure 7. Speech recognition in quiet using binaurally combined acoustic and electric stimulation in 3 participants with AN, with the three panels representing AN5, AN13, and AN22, respectively. AS = acoustic stimulation; ES = electric stimulation (i.e., cochlear implant); AS+ES = combined stimulation.



speech produced a significant advantage of 9 percentage points, $F(1, 8) = 15.4$, $p < .01$. The stimulation mode was also a significant factor, $F(2, 4) = 12.8$, $p < .05$, with the binaurally combined stimulation producing significantly better performance than electric stimulation, and electric stimulation better than acoustic stimulation ($p < .05$). The averaged percentage correct scores were 95%, 79%, and 20% for the combined, electric, and acoustic stimulation, respectively, in clear speech perception and were 84%, 67%, and 16% for the combined, electric, and acoustic stimulation, respectively, in conversational speech perception. We also note that while acoustic stimulation alone produced 0% intelligibility, it improved the performance of electric stimulation alone by 12–22 percentage points when combined with the cochlear implant in 2 participants with AN (AN5 and AN22).

Figure 8 shows percentage correct scores of clear speech (filled bars) and conversational speech (unfilled bars) obtained in noise from acoustic stimulation (AS), electric stimulation (ES), and binaurally combined acoustic and electric stimulation (AS + ES) in 3 participants with AN. The cross ("X") represents a score of 0% correct and the circle ("O") represents conditions where no data were collected. Because the data from the AN ear were collected in only one condition (AN15 at 15 dB SNR), an ANOVA was performed to compare clear and conversational speech recognition performance between the electric and combined stimulations. The analysis showed a significant clear speech advantage of 23 percentage points, $F(1, 7) = 68.7$, $p < .01$, but an insignificant binaural summation effect, $F(1, 7) = 2.7$, $p > .05$. We note that this insignificant binaural summation effect in noise is different from the significant binaural summation effect in quiet. We discuss this difference later.

Figure 8. Speech recognition in noise using binaurally combined acoustic and electric stimulation. The two panels in the top row represent data from participants AN5 at 5 and 15 dB SNRs; the three panels in the middle row represent data from participant AN13 at 0, 5, and 10 dB SNRs; and the three panels in the bottom row represent data from participant AN22 at 0, 5, and 10 dB SNRs. The circle ("O") symbols represent conditions where no data were collected.



Discussion

The primary goal of the present study was to compare performance between clear and conversational speech perception in participants with AN. The present data unequivocally demonstrated a significant clear speech advantage in this participant population, regardless of listening conditions and stimulation modes. The intelligibility difference between clear and conversational speech was 16 percentage points in quiet and 18 in noise with monaural acoustic stimulation (Experiment 1), 18 in quiet and 21 in noise with diotic acoustic stimulation (Experiment 2), 11 in quiet and 20 in noise with monaural electric stimulation (Experiment 3), and 9 in quiet and 23 in noise with binaurally combined acoustic and electric stimulation (Experiment 4). The secondary goal of the present study was to address the following unsettled problems in auditory neuropathy.

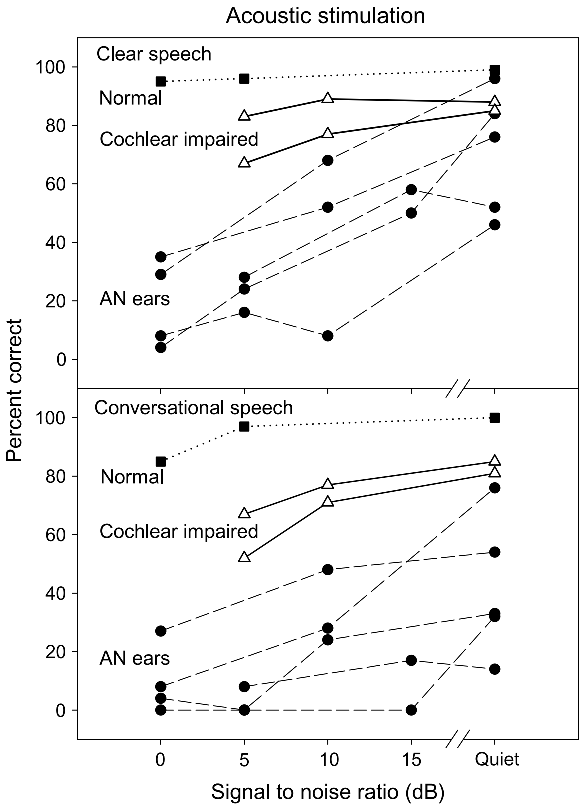
Does Noise Pose a Particular Problem?

The answer is "yes." Figure 9 shows speech recognition as a function of SNR in three participant populations. The dotted line represents our previously reported data in normal-hearing participants listening to the same stimuli as in the present study (i.e., the "Female" condition in Figure 4 of Liu et al., 2004). The solid lines represent data in 2 listeners with cochlear impairment from Payton et al. (1994; their Table V). The dashed lines represent individual data in 4 participants with AN in the present study (data in quiet are from Figure 1 and data in noise are from Figure 2; note that AN10's left and right ears were tested, resulting in five curves).

First, we can demonstrate this particular difficulty in speech recognition in noise by comparing performance between normal-hearing participants and participants with AN. Liu et al. (2004) showed that noise had no detrimental effect on clear or conversational speech perception at 5 dB SNR but only decreased conversational speech perception by 15 percentage points at 0 dB SNR. In contrast, the present AN data show that noise decreased the performance in quiet significantly at all SNRs, $F(1, 13) = 30.9$, $p < .05$. The most striking finding was that the two participants with AN who performed the best in clear speech perception in quiet (AN7 = 84% and AN10's left ear = 96%) had a drop in performance by 80 and 67 percentage points at 0 dB SNR, respectively.

Second, participants with AN seem to have more difficulty in speech perception in noise than do participants with cochlear impairment. Payton et al. (1994) measured intelligibility of nonsense sentences in 2 participants with cochlear impairment under listening conditions similar to those in the present study (quiet or anechoic, 5.3 and 9.5 SNRs with the noise being speech-spectrum shaped). Different from the relatively moderate hearing loss (44 dB PTA) in the present 4 participants with AN, the 2 participants with cochlear impairment in Payton et al.'s study had more severe hearing loss (75 and 77 dB PTA at octave frequencies from 250 to 8000 Hz, respectively). Despite the greater hearing loss and overall lower performance in quiet than the normal-hearing listeners, the 2 participants with cochlear impairment were able to maintain similar performance between the quiet and noise conditions, particularly for clear speech. These comparative data, combined with previous results (Kraus et al., 2000; Starr et al., 1996; Zeng, Oba, Garde, Sininger, & Starr, 2001), are consistent with the subjective complaint by the participants with AN of extreme difficulty when listening in noise.

Figure 9. Comparing speech recognition performance as a function of SNRs using monaural acoustic stimulation for clear speech (top panel) and conversational speech (bottom panel). The dotted line with squares represents average performance from 5 normal-hearing participants (Liu et al., 2004). The solid lines with triangles represent individual data from 2 participants with cochlear impairment but not AN (Payton et al., 1994). The dashed lines with circles represent individual data from 4 participants with AN (five ears) in the present study.

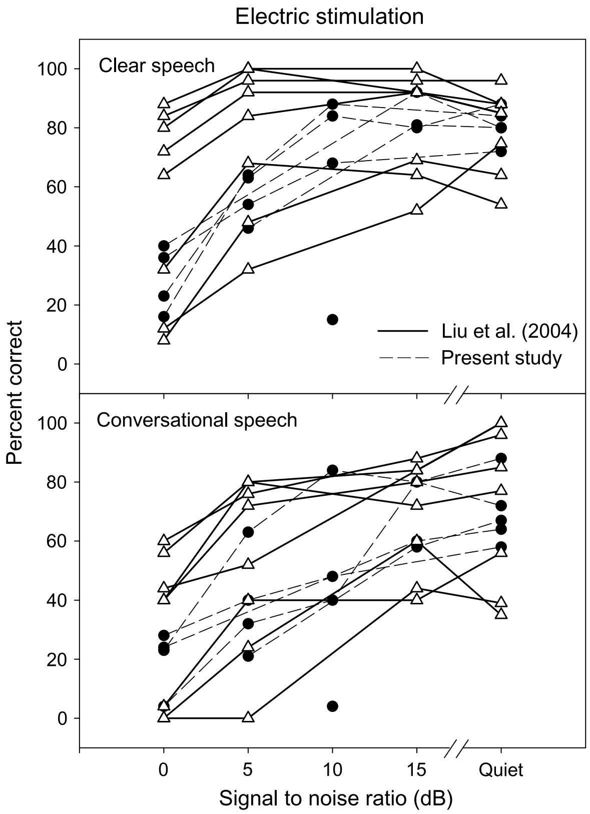


The physiological and perceptual mechanisms underlying this extreme difficulty remain unclear. Several psychophysical studies have demonstrated poor temporal and spectral processing in participants with AN (Rance et al., 2004; Starr et al., 2003; Zeng, Kong, et al., 2005; Zeng et al., 1999). In particular, Zeng, Kong, et al. (2005) found that participants with AN exhibited not only 10–20 dB excessive simultaneous masking for detection of tones in noise, but also had prolonged threshold elevations in both backward and forward masking. At a physiological level, the observed excessive masking may be due to either loss of inner hair cells (also called dead regions in the cochlea) or loss of spike synchrony resulting from damaged nerve fibers (Harrison, 1998; Moore, 2004; Starr et al., 1996; Wang et al., 1997). At a functional level, the excessive masking contributes directly to the extreme difficulty of understanding speech in noise because the perceptual SNR would be much lower than the physical SNR in participants with AN.

Does the Cochlear Implant Help?

The answer is "yes." Figure 10 shows individual speech recognition performance as a function of SNR in 8 typical cochlear implant users, that is, without AN (solid lines with triangles; data are from Liu et al., 2004) and in 5 cochlear implant users with AN (dashed lines with circles; data in quiet are from Figure 5 and data in noise are from Figure 6 in the present study). Except for one data point (10 dB SNR for AN23, the lone circle on both panels), the present data show that the implant users with AN performed similarly to the average implant users without AN (poorer than the best implant users without AN but better than the poor implant users without AN). On average, the implant users with AN achieved 81% and 67% correct scores in clear and conversational speech perception in quiet, respectively, and were able to maintain this level of performance in noise (84% and 66%, respectively) at 15 dB SNR ($p > .05$). This level of performance was much better than the AN ears without the implant (see Figure 9) at comparable SNRs.

Figure 10. Comparing speech recognition performance as a function of SNRs using monaural electric stimulation for clear speech (top panel) and conversational speech (bottom panel). The solid lines with triangles represent individual data from 8 participants with cochlear implants but not AN (Liu et al., 2004). The dashed lines with circles represent individual data from 5 cochlear-implant users with AN in the present study. The lone circle on both panels represents data from AN23, who was tested at 10 dB SNR only.



Additionally, the implant benefit is evident by significantly better performance in the implanted ear than in the nonimplanted ear in 3 participants with AN who had been tested using the same stimuli (see Figure 7). This demonstration of the implant benefit is valid only if the two ears in the same participant with AN have similar pathology and degree of hearing loss. Finally, strong evidence for the implant benefit is provided by direct comparison between presurgical and postsurgical performance in the same ear. For example, in AN7, consonant and vowel recognition scores improved from 32% and 16% correct presurgically to 72% and 54% correct postsurgically; in AN20, word recognition improved from 0% presurgically to 80% correct postsurgically.

How does the cochlear implant improve AN performance? The answer is closely tied to the apparent mechanisms underlying AN. If the site of lesion is in the inner hair cells or the synaptic transmission, then the cochlear implant ought to be an effective treatment for AN, as it bypasses both the inner hair cell and the synapse to directly stimulate the auditory nerve fibers with electric currents. If the site of lesion is related to nerve damage, then the cochlear implant can still be effective because electric stimulation provides much more synchronized neural firing than acoustic stimulation, thus possibly overcoming, at least partially if not fully, the neural desynchrony problem (Dynes & Delgutte, 1992; Javel & Shepherd, 2000; Litvak, Delgutte, & Eddington, 2001). Successful restoration of electrically evoked auditory brainstem potentials in implant users with AN has certainly supported these two possibilities (Buss et al., 2002; Peterson et al., 2003; Shallop et al., 2001; Trautwein et al., 2000). On the other hand, if AN involves extensive loss of neurons, then the cochlear implant would be less effective (Starr et al., 2003).

Are Two Ears Better Than One?

The answer is "it depends." The binaural summation effect was significant up to 28 percentage points in quiet (see Figure 3) but became insignificant in noise for the participants with AN without cochlear impants (see Figure 4). Similarly, the binaurally combined acoustic and electric stimulation yielded significantly better speech scores in quiet backgrounds, but not in noise backgrounds, for the participants with AN who had cochlear implants. Acoustic stimulation of the nonimplanted ear did not significantly improve performance above that achieved by electric stimulation alone. This result of participants with AN differs from the perceptual results in similar listening conditions with typical implant users without AN. In other studies, combined acoustic and electric stimulation seems to show stronger enhancement of speech perception in noise than in quiet (Ching, Incerti, & Hill, 2004; Kong, Stickney, & Zeng, 2005; Turner, Gantz, Vidal, Behrens, & Henry, 2004; von Ilberg et al., 1999).

Why then does the binaural summation effect become insignificant in noise in the present study? The answer is probably related to both the psychophysical capabilities in participants with AN and the nature of speech perception in acoustic and electric hearing. Psychophysically, participants with AN typically have elevated thresholds and poor pitch discrimination at low frequencies, excessive masking in noise, and cannot use low-frequency interaural timing difference cues (Zeng, Kong, et al., 2005). In quiet, the two ears in participants with AN probably provide independent information, producing a significant binaural summation effect. Because of excessive masking, the noise renders the information provided by acoustic stimulation relatively useless compared with the information received by electric stimulation. Several studies have convincingly demonstrated the benefit of combining low-frequency acoustic stimulation with electric stimulation, but no study has shown a similar benefit effect from combining high-frequency acoustic stimulation with electric stimulation. The benefit can be understood from the complementary nature of low-frequency and high-frequency speech cues in acoustic and electric hearing (Kong et al., 2005; Turner et al., 2004). Kong et al. (2005) showed in 3 participants who used a hearing aid in one ear and a cochlear implant in the other ear that the hearing aid, by itself, provided no speech intelligibility but significantly enhanced the contralateral cochlear implant speech performance in noise. Low-frequency speech cues may not contribute directly to intelligibility, but are critical to voice pitch perception, speaker identification, and source localization, thus contributing indirectly to speech intelligibility in noise (Zeng, Nie, et al., 2005). Unfortunately, poor pitch perception at low frequencies and excessive masking appear to have significantly reduced the benefit of combined acoustic and electric hearing in speech perception in noise.

Treatment Strategies

At present, cochlear implants appear to be the treatment of choice for participants with AN (Berlin et al., 2003). The present results certainly provided evidence for the effectiveness of electric hearing in alleviating speech perception difficulties, particularly in noise. However, additional benefits through combined acoustic and electric hearing appear to be limited to listening in a quiet condition only, as opposed to greater benefits of combined hearing observed in noise than in quiet in typical cochlear-implant users without AN. These typical implant users combined low-frequency acoustic hearing with electric hearing via either a short-electrode implant in the same ear or a conventional implant in the other ear (Ching et al., 2004; Kong et al., 2005; Turner et al., 2004; von Ilberg et al., 1999).

Perhaps more importantly, the present study shows an overall clear speech advantage across a wide range of listening conditions and modes, suggesting that innovative hearing aids that can convert conversational speech into clear speech can benefit participants with AN. There have been limited reports on the success of applying conventional hearing aids to participants with AN (Deltenre et al., 1999; Rance et al., 2002), but the clinical management has appeared to shift toward cochlear implantation (Berlin et al., 2003; Peterson et al., 2003).

Because temporal processing is significantly impaired in participants with AN, we suggest that amplitude compression should be avoided if at all possible because it reduces the amount of temporal modulation in the signals sent to the ear. Instead, linear amplification should be considered. The present speech perception results, together with psychophysical results (Rance et al., 2004; Zeng, Kong, et al., 2005; Zeng et al., 1999), suggest several innovative signal processing schemes that may benefit speech performance in participants with AN. One scheme may actually expand temporal modulation because one of the features in clear speech is enhanced amplitude modulation (Krause & Braida, 2004; Liu et al., 2004). Another scheme may filter out low-frequency signals and/or shift them to high-frequency regions based on the psychophysical observations that participants with AN have extremely poor pitch perception at low-frequencies but relatively normal pitch processing at high frequencies. We note that this frequency transposition is in the opposite direction of most frequency transposition methods documented in the literature. The traditional methods typically transpose high-frequency signals to the low-frequency region to solve the audibility problem at high frequencies in elderly listeners (Turner & Hurtig, 1999). Future treatment strategies will rely on physiological, psychophysical, and speech evaluations to achieve optimal options, including both cochlear implants and hearing aids for people with AN on an individual basis.

Summary

The present study compared perception performance between clear and conversational speech in participants with AN. The listening conditions included quiet and noise, while the stimulation modes included monaural acoustic, diotic acoustic, monaural electric, and binaurally combined acoustic and electric stimulation. Consistent with previous studies, the present study found a significant clear speech advantage in both listening conditions with all stimulation modes. The present study also showed a significant cochlear implant advantage for speech performance in both quiet and noise listening conditions. However, different from previous results in participants without AN, the present study found a binaural summation effect in quiet, but not in noise. This inconsistent result is interpreted as a reflection of the unique perceptual processing deficits in participants with AN, namely, poor pitch processing at low frequencies, excessive masking in noise, and inability to process interaural timing information. Although the present result supports cochlear implantation as an effective treatment option, it also suggests innovative hearing aids incorporating temporal envelope enhancement, low-frequency filtering, and high-frequency transposition as an alternate treatment option.

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Contact author: Fan-Gang Zeng, 364 Med Surge II, University of California, Irvine, CA 92697. E-mail: fzeng@uci.edu