

## CASE REPORT

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### **Cochlear implantation in a patient with auditory neuropathy/dyssynchrony: A Case Report**

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Auditory neuropathy/dyssynchrony is a hearing disorder characterized by poor speech discrimination that is disproportionate to the degree of hearing loss, an abnormal or absent auditory brainstem response in the presence of normal otoacoustic emissions and cochlear microphonics, absent acoustic reflexes, absent efferent suppression of otoacoustic emissions, and abnormal masking level difference scores.

In this study, the preoperative and postoperative audiologic test results of an adult patient with auditory neuropathy/dyssynchrony who underwent cochlear implantation are presented.

Visually detected electrical stapedius reflexes and the results of neural response telemetry were noted intraoperatively.

A statistically significant improvement in hearing and speech perception occurred after cochlear implantation.

The results suggest that cochlear implantation can be performed to overcome dyssynchronization and enable rehabilitation.

Auditory neuropathy/dyssynchrony (AN/AD) is a hearing disorder characterized by poor speech discrimination disproportionate to the degree of hearing loss, abnormal or absent auditory brainstem response in the presence of normal otoacoustic emissions and cochlear microphonics, absent acoustic reflexes, abnormal masking level difference scores, and absent efferent suppression of otoacoustic emissions.<sup>(1-3)</sup> The results of cochlear implanted patients with AN/AD have been reported in the literature.<sup>(4-12)</sup> Rance and colleagues reported a case of poor speech identification performance after cochlear implantation<sup>(4)</sup> and Miyamoto and colleagues reported on a patient with AN and Friedreich's ataxia in whom the benefit derived from a CI was modest and who demonstrated poor postsurgical open set speech understanding<sup>(5)</sup>. However, Trautwein and colleagues described a child with AN whose speech perception improved after CI placement<sup>(6)</sup>. In other studies, Shalloo and colleagues reviewed the results of cochlear implantation in 5 children with AN<sup>(7)</sup> and Buss and colleagues reported on 4 children with AN who received a CI<sup>(8)</sup>. The results of those 2 investigations<sup>(7,8)</sup> showed that all subjects received a measurable benefit from cochlear implantation. In addition, the presurgical behavioral thresholds for speech and tones (which were achieved with hearing aids) improved to a statistically significant degree after CI placement. Madden and colleagues reported that 4 patients who received a CI demonstrated improvement in auditory and verbal performances in varying degrees<sup>(9)</sup>. The results of 3 adults and 1 child who received a CI were reported by Lesinski-Schiedat and colleagues<sup>(10)</sup>. Two of the adults and 1 child studied by those investigators demonstrated better word recognition after CI placement. One of the adult subjects, who was prelingually deafened, obtained sound identification only after having received a CI<sup>(10)</sup>. Peterson and colleagues compared postimplant progress in the speech perception of 10 children diagnosed with AN/AD with that of 10 children whose hearing impairment resulted from other causes<sup>(11)</sup>. The results of that study revealed no statistically significant differences in CI benefit between the 2 groups. Four children with AD/AN reported by Ciprut and Akdas demonstrated a statistically significant improvement in auditory perception, communication skills, and speech perception after cochlear implantation<sup>(12)</sup>.

The following case report describes an adult patient with AN/AD who received a CI. A review of the patient's performance in speech perception before and after cochlear implantation is presented.

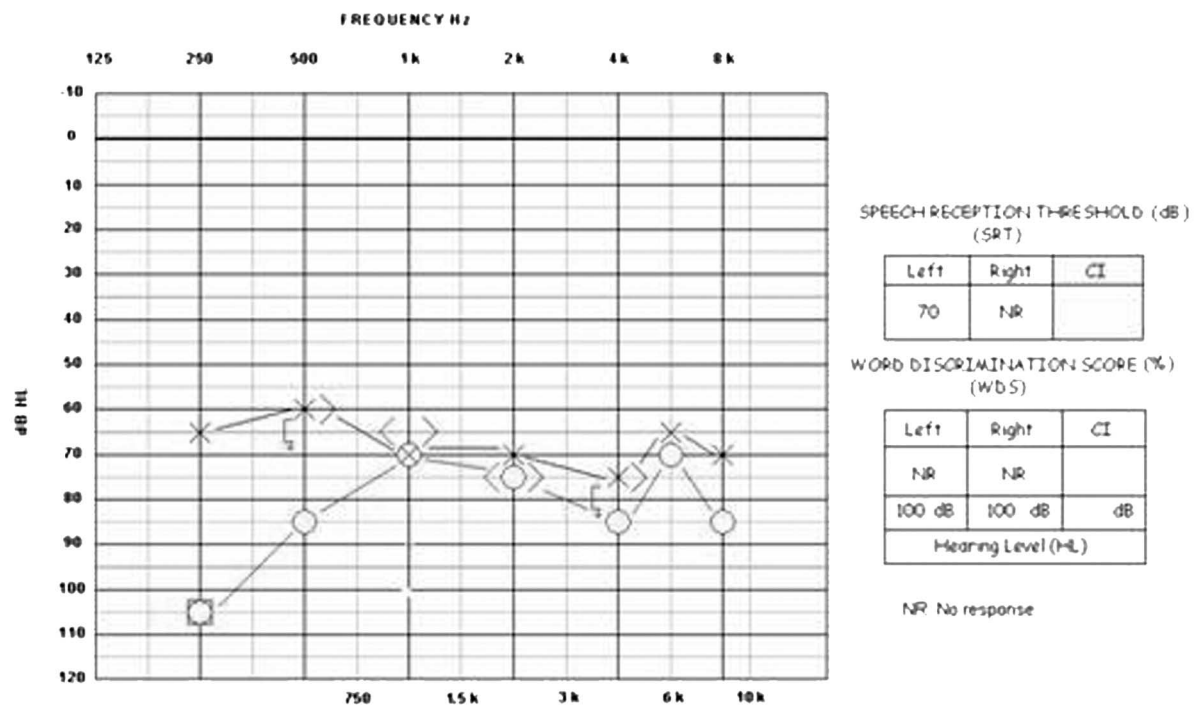
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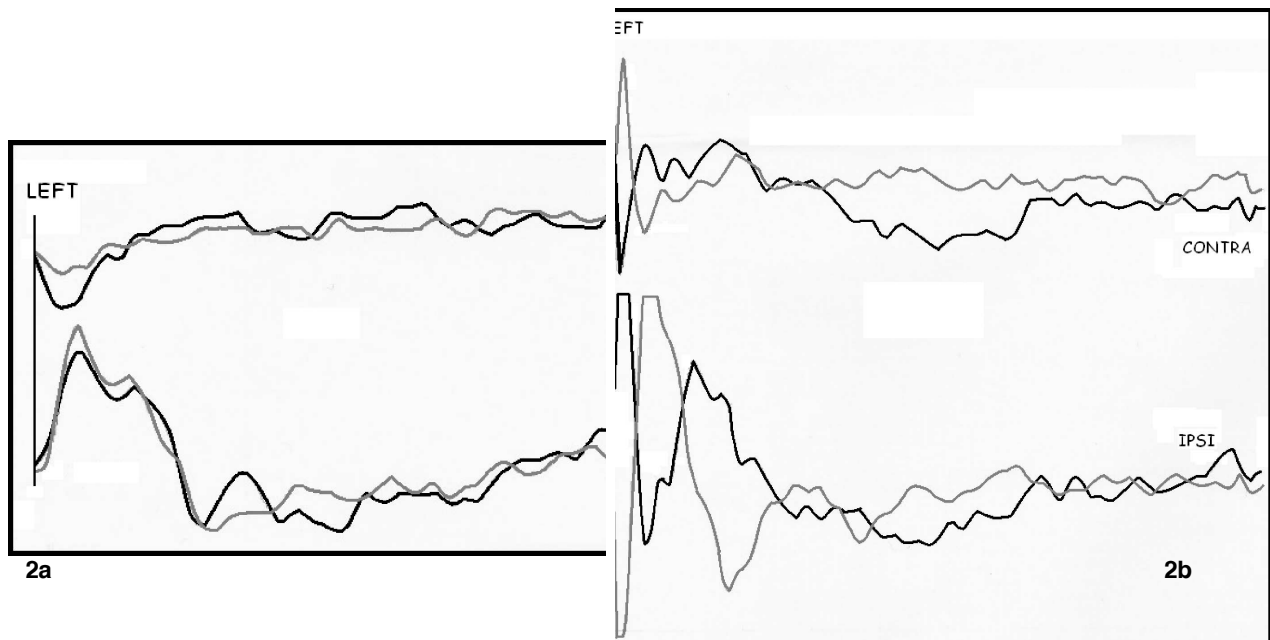
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The patient was a 52-year-old man who was referred to our clinic for the evaluation of candidacy for a CI. At the age of 11 years, he sustained a bilateral sensorineural hearing loss, but his medical history was otherwise unremarkable. His major complaint was poor speech discrimination. When the patient was 16 years old, hearing aids were prescribed, but he did not use them when no benefit was noted. He used lip-reading for verbal communication.

The results of an audiologic-evaluation revealed severe sensorineural hearing loss in the right ear, moderate sensorineural hearing loss in the left ear, and no measurable word discrimination score in either ear. The speech reception threshold (SRT), which could not be obtained for the right ear, was 70 dB HL for the left ear. The speech detection threshold was 70 dB HL for the right ear. Figure 1 shows the results of the patient's audiogram before cochlear implantation. During an immittance-metric evaluation, a type A tympanogram was obtained for both ears. The results of tympanometry showed that in both ears, the patient's normal middle ear function was within normal limits. There were no ipsilateral or contralateral acoustic reflexes, and transient-evoked otoacoustic emissions were absent bilaterally. There was no auditory brainstem response to clicks with alternating polarity, even at the maximum intensity levels. Cochlear microphonics were bilaterally observed via the comparison of condensation and rarefaction polarities. Figure 2 (a and b) shows the auditory brainstem response recordings in which a neural response is absent and cochlear microphonics are present. The results of radiologic studies, computed tomography, and magnetic resonance imaging were within the normal range, which indicated that there was no retrocochlear pathologic condition in either ear. The patient was diagnosed as having AN/AD. Promontory electrical stimulation revealed auditory sensation in both ears in response to electrical stimulation.



**Figure-1:** Audiogram results before cochlear implantation. Right-ear and left-ear hearing thresholds before implantation are marked with "O" and "X," respectively.



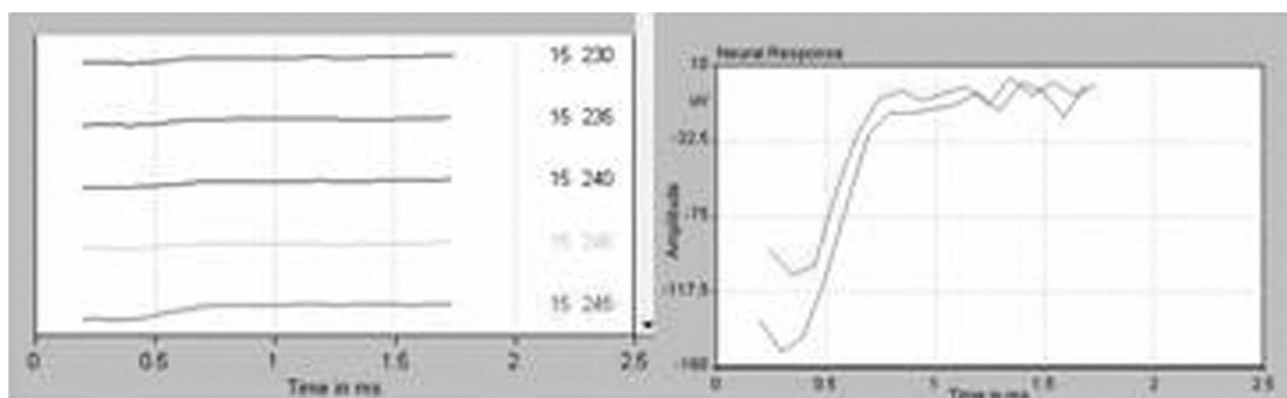
**Figure-2a:** Auditory brainstem response is absent when tested in alternating polarity at the maximum intensity level of 95 dB nHL. **b:** The overlaying of rarefaction and the condensation clicks indicate the cochlear microphonics

After the patient had undergone audiologic and otologic assessment, he was selected as a candidate for cochlear implantation. A Nucleus CI24R (CS) device (Cochlear Corporation, Sydney, Australia) was implanted in his left ear, which was chosen for implantation because it was the better ear for speech reception and because the patient reported more natural hearing in the left ear during promontory stimulation.

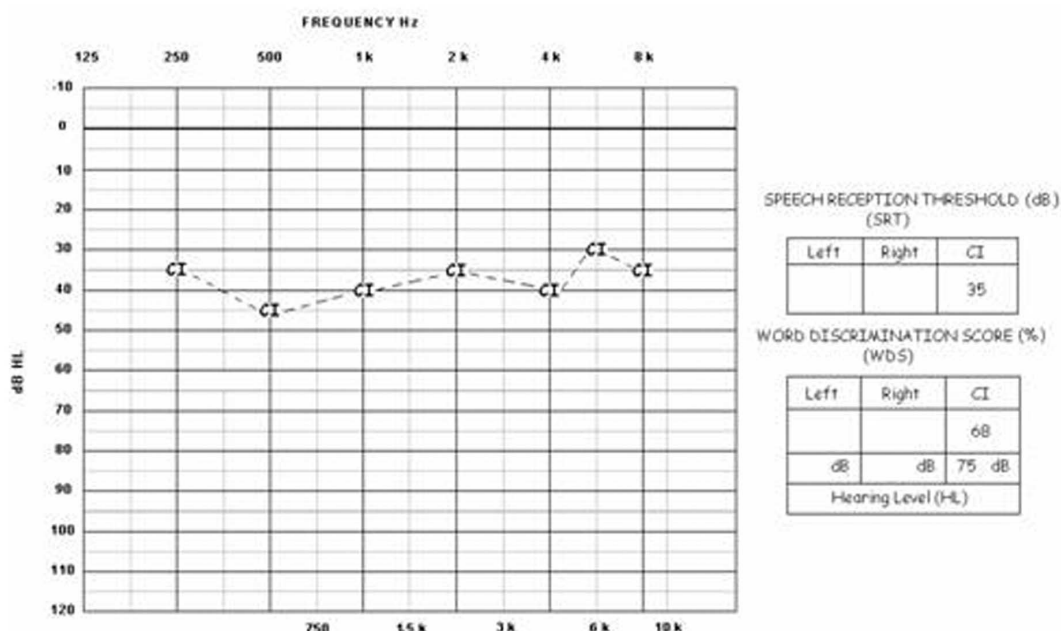
Full insertion of all electrodes was achieved during surgery. Visually detected electrical stapedius reflexes and electrically evoked compound action potentials were obtained intraoperatively by means of the Neural Response Telemetry (NRT) system (NRT version 3.0,

Cochlear Corporation). The synchronous activity of the auditory nerve was shown via neural response telemetry, which revealed results similar to those of patients without AN. Figure 3 shows the intraoperative NRT recording for electrode <sup>15</sup>.

The patient was fitted with a Nucleus ESPrit 3G speech processor (Cochlear Corporation, Sydney, Australia). He used an Advanced Combination Encoders 900 Hz map in the monopolar 1+2 mode with a pulse width of 25  $\mu$ s. His latest sound field warble tone thresholds with the implant were between 30 and 35 dB across the audiometric frequency range (Figure 4).



**Figure-3:** Results from intraoperative testing with the Neural Response Telemetry (NRT) system (NRT version 3.0, Cochlear Corporation, Sydney, Australia) for electrode 15.



**Figure-4:** The hearing thresholds in the sound field with the cochlear implant.

Speech perception was measured with a battery of closed and open set speech tests for the Turkish language. Preoperatively, only open set speech tests were administered to the patient in free-field under the best aided conditions. Postoperative closed set and open set speech perception tests were also conducted.

Open set word recognition was assessed with a trisyllabic words test, an everyday sentence test, and a monosyllabic words test. The trisyllabic words test consisted of 40 trisyllabic words. The patient was asked to repeat each word, and the test was scored as the percent of words that were correct. The monosyllabic words test consisted of 6 lists with 25 monosyllabic words in each list. The patient was asked to repeat each word, and the test was scored as the percent of words that were correct<sup>(13)</sup>. The everyday sentence test was an open set test consisting of 75 of the sentences used most frequently in everyday language. The patient was expected to correctly repeat the sentence that the tester had read to him, and the answer was recorded by the tester. The test score was based on percent of entirely correct sentences<sup>(14)</sup>.

In closed set testing, phoneme recognition of the consonant at the beginning of the monosyllabic words test and at the beginning of the trisyllabic words test was evaluated<sup>(14)</sup>. In phoneme recognition at the beginning of the monosyllabic words test, the task was to determine the consonant at the beginning of the monosyllabic word. There were 25 items, each of which had 3 choices. The

choices were the same monosyllabic words, but only the consonant at the beginning of the words was different. The tester read a word, and the patient was expected to identify that word from among the 3 choices on the sheet. In phoneme recognition at the beginning of the trisyllabic words test, the task was to discriminate the consonant at the beginning of the trisyllabic word. There were 25 items, each of which had 3 choices. The choices were the same trisyllabic words, but only the consonant at the beginning of the words was different. The tester read a word, and the patient was expected to identify that word from among the 3 choices on the sheet.

All tests were administered via a monitored live voice at a 70-dB sound pressure level from a loudspeaker at zero degrees azimuth in Industrial Acoustics Company (IAC) sound-treated rooms with an Interacoustic AC5 audiometer. The patient was tested under the best aided conditions before cochlear implantation and with his speech processor set at his preferred listening level postoperatively. The Table illustrates the speech perception test results obtained preoperatively and 1 month, 6 months, and 1 year after cochlear implantation. After 6 months of cochlear implant use, the patient could understand open set speech tests. His speech test results improved over time. After 1 year of cochlear implant use, he could understand speech without lip-reading, and at the time of this writing, he can communicate via telephone.

Table: Speech perception test results after implantation

Speech Test Administered	Preoperative Result	Postsurgical 1-mo Result (%)	Postsurgical 6-mo Result (%)	Postsurgical 1-y Result (%)
Closed set monosyllabic words test	NT	100	100	100
Open set 3 syllabic words test	NR	68	84	100
Open set monosyllabic words test	NR	4	48	68
Open set sentences test	NR	24	68	84

NT, Not tested; NR, no response.

## DISCUSSION

Cochlear implantation in postlingually deafened adults is a generally accepted treatment, and those patients usually achieve a substantial benefit from implantation.

However, some authors have reported a less-than-adequate outcome after cochlear implantation in patients with AN<sup>(4,5)</sup>. In contrast, other investigators have noted that patients with AN have benefitted substantially from cochlear implantation<sup>(6-9,11)</sup>. The wide range of outcomes after cochlear implantation might reflect the multiple pathologic conditions that cause the symptoms of AN. Possible sites that may be affected include inner hair cells, the synapse between the hair cells and the auditory nerve; and/or the auditory nerve fibers (myelin and axonal impairments)<sup>(15)</sup>. Harrison proposed scattered inner hair cell lesions as a possible cause of AN<sup>(16)</sup>. It has also been suggested that the success of cochlear implantation may depend on the site of the lesion. For example, if a lesion is in the auditory nerve, then the CI will not transmit auditory signals to the higher auditory pathway structures. If the AN involves only the inner hair cells and/or the synapse with afferent nerve fibers, then the patient has a greater chance of benefitting from cochlear implantation<sup>(4)</sup>.

Improvement in hearing and speech perception was obtained in our patient after cochlear implantation. These results are comparable to those in postlingual CI users without AN/AD<sup>(17,18)</sup>. Although the pathophysiology of AN/AD in our patient remains unknown, the absence of accompanying peripheral neuropathies and the benefit obtained in speech understanding after cochlear implantation suggest that the site of the lesion was not in the auditory nerve but was instead in the inner hair cells and/or in the synapse with the afferent fibers.

Although the results of a single case are presented in this article, we suggest that electrical stimulation via a CI can be a treatment option for patients with AN/AD and may help to the habilitation process for some AN cases. However, the decision to provide a CI for a patient with AN/AD should include an assessment of the

pathophysiologic characteristics of AN in that individual. Children or adults diagnosed with AN/AD should be evaluated for concomitant neuropathies as well. Until the underlying causes for AN/AD are fully understood, the decision to provide a CI for a patient with AN/AD should be made carefully and on an individual basis

## REFERENCES

1. Sininger Y, Hood LJ, Starr A, Berlin CI, Picton TW. Hearing loss due to auditory neuropathy. *Audiology Today* 1995;7:10-3.
2. Starr A, Picton TW, Sininger Y, Hood LJ, Berlin CI. Auditory neuropathy. *Brain* 1996;119:741-53.
3. Berlin C, Hood L, Rose K. On renaming auditory neuropathy as auditory dys-synchrony. *Audiology Today* 2001;13:15-7.
4. Rance G, Beer DE, Cone-Wesson B, Shepherd RK, Dowell RC, King AM, et al. Clinical findings for a group of infants and young children with auditory neuropathy. *Ear Hear* 1999;20:238-52.
5. Miyamoto RT, Kirk KI, Renshaw J, Hussain D. Cochlear implantation in auditory neuropathy. *Laryngoscope* 1999;109:181-5.
6. Trautwein PG, Sininger YS, Nelson R. Cochlear implantation of auditory neuropathy. *J Am Acad Audiol* 2000;11:309-15.
7. Shallop JK, Peterson A, Facer GW, Fabry LB, Driscoll CL. Cochlear implants in five cases of auditory neuropathy: postoperative findings and progress. *Laryngoscope* 2001;111:555-62.
8. Buss E, Labadie RF, Brown CJ, Gross AJ, Grose JH, Pillsbury HC. Outcome of cochlear implantation in pediatric auditory neuropathy. *Otol Neurotol* 2002;23:328-32.
9. Madden C, Rutter M, Hilbert L, Greinwald JH Jr, Choo DI. Clinical and audiological features in auditory neuropathy. *Arch Otolaryngol Head Neck Surg* 2002;128:1026-30.

10. Lesinski-Schiedat A, Frohne C, Hemmaouil I, Battmer RD, Lenarz T. [Subjective deafness in case of peri-synaptic audiopathy. Isolated defects of the inner haircells?] *Laryngorhinootologie* 2001;80:601-4.
11. Peterson A, Shallop J, Driscoll C, Breneman A, Babb J, Stoeckel R, et al. Outcomes of cochlear implantation in children with auditory neuropathy. *J Am Acad Audiol* 2003;14:188-201.
12. Ciprut A, Akdas F. Cochlear implantation in four auditory dys-synchrony cases: preimplant audiological results and postimplant progress. In: *Proceedings of the Eighth European Symposium on Pediatric Cochlear Implantation*; March 25-28, 2006; Venice, Italy. Abstract [FP 62]
13. Aksit M. Preparation of Isophonic Monosyllabic Word Lists for Speech Discrimination [M.S. thesis]. Marmara University, Institute of Health Sciences, Istanbul Turkiye;1994.
14. Derinsu U, Gence Gumus Z, Akdas F. Standardization of the speech tests used with cochlear implant patients. *Marmara Medical Journal*, 2007;20:18-23.
15. Starr A, Sininger YS, Pratt H. The varieties of auditory neuropathy. *J Basic Clin Physiol Pharmacol* 2000;11:215-30.
16. Harrison RV. An animal model of auditory neuropathy. *Ear Hear* 1998;19:355-61.
17. Cohen NL, Waltzman SB, Shapiro WH. Telephone speech comprehension with use of the nucleus cochlear implant. *Ann Otol Rhinol Laryngol Suppl* 1989;142:8-11.
18. Schindler RA. Personal reflections on cochlear implants. *Ann Otol Rhinol Laryngol Suppl* 1999;177:4-7.