EUROPEAN ARCHIVES OF OTO-RHINO-LARYNGOLOGY (2005) 262 -6:449-452.

Cochlear implantation in an adult patient with auditory neuropathy

Katada, A; Nonaka, S; Harabuchi, Y

Cochlear Implantation in an Adult Patient with Auditory Neuropathy

Akihiro Katada, Satoshi Nonaka and Yasuaki Harabuchi

Department of Otolaryngology—Head and Neck Surgery
Asahikawa Medical College
Midorigaoka Higashi 2-1-1-1, 078-8510, Japan

Corresponding author:
Akihiro Katada, M.D.
Department of Otolaryngology—Head and Neck Surgery
Asahikawa Medical College
Midorigaoka Higashi 2-1-1-1, Asahikawa, 078-8510
Japan
Tel: (81) 166-68-2554
Fax: (81) 166-68-2559
E-mail: katada@asahikawa-med.ac.jp

Abstract:

We present a case report of effective cochlear implantation for an adult patient with auditory neuropathy. A 34-year-old man developed bilateral hearing loss at approximately 10 years of age. His speech discrimination score was very severe despite only moderate sensorineural hearing loss. Absence of auditory brainstem responses (ABR) and preservation of distortion product otoacoustic emissions (DPOAE) were confirmed by our audiological examinations. After cochlear implantation, good responses for electrically evoked compound action potential (EAP) and electrically evoked ABR (EABR) were observed. Postoperatively, his audiological performance was significantly improved. We conclude that cochlear implantation can be a valid option for patients with auditory neuropathy.

Key Words:

Auditory neuropathy; Adult; Cochlear implantation; Electrically evoked compound action potentials (EAPs); Electrically evoked auditory brainstem responses (EABRs); Speech discrimination

Introduction:

Auditory neuropathy is defined as impairment of auditory neural function with preserved cochlear hair cell function [11]. Clinical features of auditory neuropathy include: (1) bilateral sensorineural hearing loss of any degree; (2) normal outer hair cell function evidenced by preservation of otoacoustic emissions (OAEs) and/or cochlear microphonics (CM); (3) abnormal auditory brainstem responses (ABRs); and (4) disproportionate difficulties in speech discrimination [1, 6, 11].

Conventional hearing aids are rarely effective in patients with auditory neuropathy, which makes intervention and auditory rehabilitation difficult in this patient population [1, 8, 10]. On the other hand, the efficacy of cochlear implantation for patients with auditory neuropathy is questioned because the site of lesion for patients with auditory neuropathy is still uncertain. Although initial reports of cochlear implantation in patients with auditory neuropathy recommended caution [5], more recent studies of children with auditory neuropathy have demonstrated benefit of cochlear implantation [8, 10, 14]. Furthermore, to our knowledge there are only a few formal published reports on cochlear implantation in adults with auditory neuropathy [4, 9].

The following case presentation details the cochlear implantation of an adult patient with auditory neuropathy, and may serve as a representative example of its potential benefits.

Case Report:

A 34-year-old man was referred to the Department of Otolaryngology—Head and Neck Surgery of Asahikawa Medical College for the examination of bilateral sensorineural hearing loss. His chief complaint was poor speech discrimination. He had developed bilateral hearing loss at approximately 10 years of age. He had used hearing aids for several years as a teenager. However, he had given up using hearing aids because of his limited improvement with speech discrimination. At the time of evaluation, he no longer wore hearing aids and was dependent on lip reading for verbal communication. He had no other significant medical history. His mother's pregnancy was normal, and his family history was negative.

His pure-tone audiogram revealed moderate-to-severe bilateral sensorineural hearing loss (Figure 1A). Unaided pure-tone average of the right side was 77.5 dB HL, and that of the left side was 53.8 dB HL. Tympanometry of both ears showed type A tympanogram. The stapedial reflexes of both ears were absent. He was administered speech discrimination testing using a standardized protocol, the Japanese Monosyllable Word List 67, which uses 20 monosyllables in semi-closed form. The word discrimination score of normal controls for this test reaches 100% in the range of between 30 and 40 dB hearing level. However, our patient's maximum discrimination scores were 0% in the right ear and 15% in the left ear, even at sound levels sufficient to overcome his hearing loss (Figure 1B).

Distortion product otoacoustic emissions (DPOAEs) were measured using an ILO292 (Otodynamics, UK) in both ears, and a normal response was noted in the lower

frequencies from 1000 to 2500 Hz (Figure 2). A click-evoked ABRs revealed no repeatable waveforms. Transtympanic promontory electrical stimulation revealed almost normal auditory perception bilaterally (Table 1), with gap detection thresholds obtained at 10 ms and 20 ms for the right and left ears, respectively. CT scan and MRI revealed no abnormality in either ear. There were no neurological findings noted on a complete physical exam, with the exception of his hearing loss. Studies of the motor and sensory nerve conduction velocity were completely normal.

He was implanted with a Nucleus CI24M device (Cochlear Corporation, Australia). The right ear, the worse-hearing ear, was chosen for implantation based on the level of pure-tone thresholds. The surgery proceeded with no complications, with a full insertion of the device. We performed intra-operative measurement of electrically evoked compound action potentials (EAPs) using Neural Response Telemetry System (NRT version 3.0; Cochlear Corporation, Australia). The cochlear implant was stimulated, and representative EAPs were recorded (Figure 3A). We also performed intraoperative measurement of electrically evoked ABRs (EABRs), and the representative EABRs are shown in Figure 3B.

Two weeks after the implantation, he was fit with the ACE processing strategy and monitored closely at three-month intervals. We repeated his speech recognition scores after implantation, using the Japanese vowel, consonant, word and sentence recognition tests. In the vowel and consonant recognition tests, we presented the patient with twenty vowel sounds and fifty-six consonant sounds, respectively, with and without lip-reading, and calculated the percentage of correct answers. Six months after implantation, the

vowel recognition score increased from 20% to 70%, and the consonant recognition score increased from 6% to 45%, both without lip reading. When compared to eleven other patients in our hospital implanted with the same device, his combined speech recognition scores improved equally well at six months after implantation. Nine months after cochlear implantation, the patient could understand simple words by telephone and was quite satisfied with his improvement in communication.

Discussion:

We have reported an adult case of auditory neuropathy treated by cochlear implantation. The diagnosis of auditory neuropathy was made due to sensorineural hearing loss, poor speech discrimination, absence of ABRs, and preservation of OAEs. His pure-tone audiogram showed a flat shape, and his hearing loss at high frequencies was about 70 dB HL. In our case, DPOAEs were preserved from 1000 to 2500 Hz, and diminished only in high frequencies above 3500 Hz. The presence of intact DPOAEs in low frequencies indicates that his absent ABR is not caused by usual cochlear pathology. Thus, we conclude that this is a case of auditory neuropathy.

In the previous reports, a number of etiologies and pathophysiologies are described for auditory neuropathy [12, 13]. These include: (1) part of generalized metabolic or toxic neuropathy; (2) genetic or hereditary factors; (3) immune or inflammatory disorders; and (4) infectious processes. However, Starr et al. reported that no etiology was defined in 48% of cases of auditory neuropathy [12]. Our patient's neurological examination was normal, without evidence of cranial or peripheral neuropathy, and no particular hereditary evidence was found in his family or past history. Therefore, we believe our patient has this sporadic form of auditory neuropathy.

The specific pathophysiology of auditory neuropathy is currently uncertain. Zeng et al. and Kraus et al. decribed the pathophysiology of auditory neuropathy [3, 15]. They suggested that auditory neuropathy represents a disruption of the synchronous activity of the auditory nerve, without affecting the function of the outer hair cells. The absence of ABRs in patients with auditory neuropathy may be caused by this impairment of neural

synchrony within the auditory pathways. Including our patient, adults with auditory neuropathy typically experience that they can hear people talking, but they cannot understand the words. This fact is thought to reflect the impairment of neural synchrony. This impairment may be closely related to poor speech discrimination and the small benefit of the conventional hearing aids in most of the patients with auditory neuropathy. In recent studies, Gibson et al. and Rea et al. described that electrophysiological tests were very useful for evaluation of patients with auditory neuropathy [2, 7]. In our patient, transtympanic promontory electrical stimulation testing before implantation revealed a normal auditory response bilaterally. This result suggested the possibility that the electrical stimulation was able to produce synchronous discharges of auditory pathways. In addition, the EAPs and EABRs testing of our patient showed good responses, which were typical of cochlear implantation patients without auditory neuropathy. His good responses to EAPs and EABRs suggest that the electrical pulses from the cochlear implant were able to produce synchronous activity of the auditory nerve. These facts predicted that cochlear implantation would be effective for our patient's auditory rehabilitation. Gibson et al. also reported that the auditory neuropathy patients who showed good responses to EABRs performed well with their cochlear implant [2]. In fact, speech recognition scores of our patient did improve after cochlear implantation, and the degree of improvement was as same as that of other patients without auditory neuropathy.

Although benefit from conventional hearing aid usage has been reported in a few cases of auditory neuropathy, most patients achieve limited improvement of speech discrimination with a hearing aid [1, 6]. Miyamoto et al. reported that not all cases of

auditory neuropathy are suitable for cochlear implantation [5]. On the other hand, recent papers have demonstrated that cochlear implantation was effective for many cases of children with auditory neuropathy [8, 10, 14]. Until now there have been few reports on cochlear implantation in adults with auditory neuropathy. To clarify the potential benefits of cochlear implantation for the adult patients with auditory neuropathy, further investigation will be required. However, Manson et al. reported achieving good results with three adult patients with auditory neuropathy using cochlear implantation [4]. Our case further shows that cochlear implantation can be effective for certain adults with auditory neuropathy. We assert that auditory neuropathy should not be considered a contraindication to cochlear implantation, especially in those patients who do not benefit from conventional hearing aids.

Acknowledgements:

We express our sincere thanks to Dr. Jeremy D Vos of the Vanderbilt University Medical Center for critically reviewing the final version of this manuscript.

References:

[1] Doyle KJ, Sininger Y, Starr A (1998) Auditory neuropathy in childhood. Laryngoscope 108: 1374-1377

[2] Gibson WPR, Sanli H (2002) Auditory neuropathy: the use of electrophysiological tests. In: Kubo T, Takahashi Y, Iwaki T (eds) Cochlear Implants: An Update. Kugler, The Hague, p53-58

[3] Kraus N, Ozdamar O, Stein L, Reed N (1984) Absent auditory brain stem response: peripheral hearing loss or brain stem dysfunction? Laryngoscope 94: 400-406

[4] Mason JC, De Michele A, Stevens C, Ruth RA, Hashisaki GT (2003) Cochlear implantation in patients with auditory neuropathy of varied etiologies. Laryngoscope 113: 45-49

[5] Miyamoto RT, Kirk KI, Renshaw J, Hussain D (1999) Cochlear implantation in auditory neuropathy. Laryngoscope 109: 181-185

[6] Rance G, Cone-Wesson B, Wunderlich J, Dowell R (2002) Speech perception and cortical event related potentials in children with auditory neuropathy. Ear Hear 23: 239-253

[7] Rea PA, Gibson WPR (2003) Evidence for surviving outer hair cell function in congenitally deaf ears. Laryngoscope 113: 2030-2034

[8] Shallop JK, Peterson A, Facer GW, Fabry LB, Driscoll CL (2001) Cochlear implants in five cases of auditory neuropathy: postoperative findings and progress. Laryngoscope 111: 555-562

[9] Sheykholeslami K, Kaga K, Kaga M (2001) An isolated and sporadic auditory neuropathy (auditory nerve disease): report of five patients. J Laryngol Otol 115: 530-534

[10] Simmons JL, Beauchaine KL (2000) Auditory neuropathy: a case study with hyperbilirubinemia. J Am Acad Audiol 11: 337-347

[11] Starr A, Picton TW, Sininger Y, Hood LJ, Berlin CI (1996) Auditory neuropathy. Brain 119: 741-753

[12] Starr A, Sininger YS, Pratt H (2000) The varieties of auditory neuropathy. J Basic Clin Physiol Pharmacol 11: 215-230

[13] Starr A, Sininger Y, Nguyen T, Michalewski HJ, Oba S, Abdala C (2001) Cochlear receptor (microphonic and summating potentials, otoacoustic emissions) and auditory pathway (auditory brain stem potentials) activity in auditory neuropathy. Ear Hear 22: 91-99

[14] Trautwein PG, Sininger YS, Nelson R (2000) Cochlear implantation of auditory neuropathy. J Am Acad Audiol 11: 309-315

[15] Zeng FG, Oba S, Garde S, Sininger Y, Starr A (1999) Temporal and speech processing deficits in auditory neuropathy. Neuroreport 10: 3429-3435

Legends:

Figure 1:

A) Pure tone audiogram at the first visit.

B) Speech audiogram with speaker method.

Figure 2:

Results of the distortion product otoacoustic emissions (DPOAEs) were plotted on the DP-gram. DPOAEs were recorded with F2/F1 ratio of 1.22. The residual noise levels were shown by the hatched area.

Figure 3:

A) The electrically evoked compound action potentials (EAPs) waveform recorded intra-operatively with Neural Response Telemetry (NRT) testing at variable current levels. These waves were recorded from the No. 20 electrode of the cochlear implant. The stimulation electrode was No. 17. The number of the probe current levels indicates the programming units of the NRT software.

B) Record of the intraoperative cochlear implant electrically evoked ABRs (EABRs). These waves were recorded from the No. 20 electrode of the cochlear implant. The number of the probe current levels indicates the programming units of the NRT software.

Table 1:

Results of the promontory stimulation test.

	100 Hz burst (□A)	200 Hz burst (□A)
Right ear		
threshold	7.0	13.0
maximum acceptable loudness	23.0	33.0
dynamic range	16.0	20.0
Left ear		
threshold	8.0	12.0
maximum acceptable loudness	13.0	17.0
dynamic range	5.0	5.0

Table 1

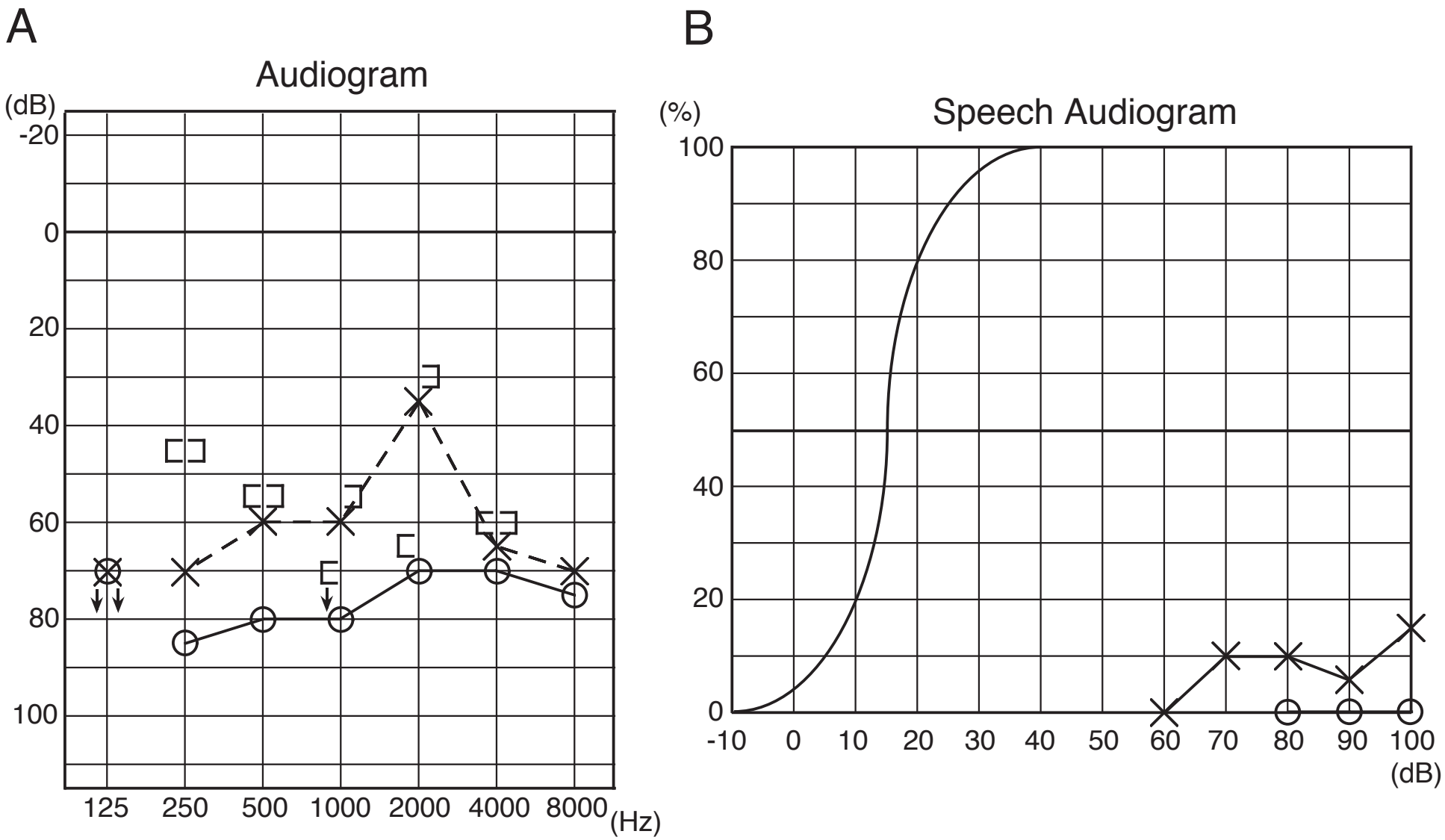
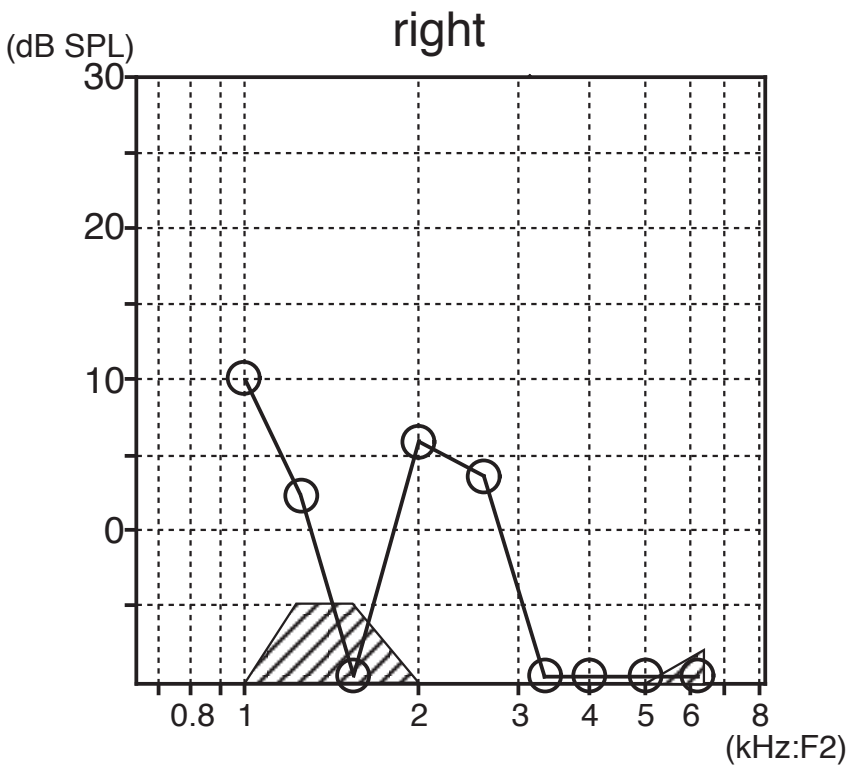

A
Audiogram
(dB)
-20
0
20
40
60
80
100
125
250
500
1000
2000
4000
8000
(Hz)
B
Speech Audiogram
(%)
100
80
60
40
20
0
-10
0
10
20
30
40
50
60
70
80
90
100
(dB)

Fig. 1


right
(dB SPL)
30
20
10
0
0.8
1
2
3
4
5
6
8
(kHz:F2)

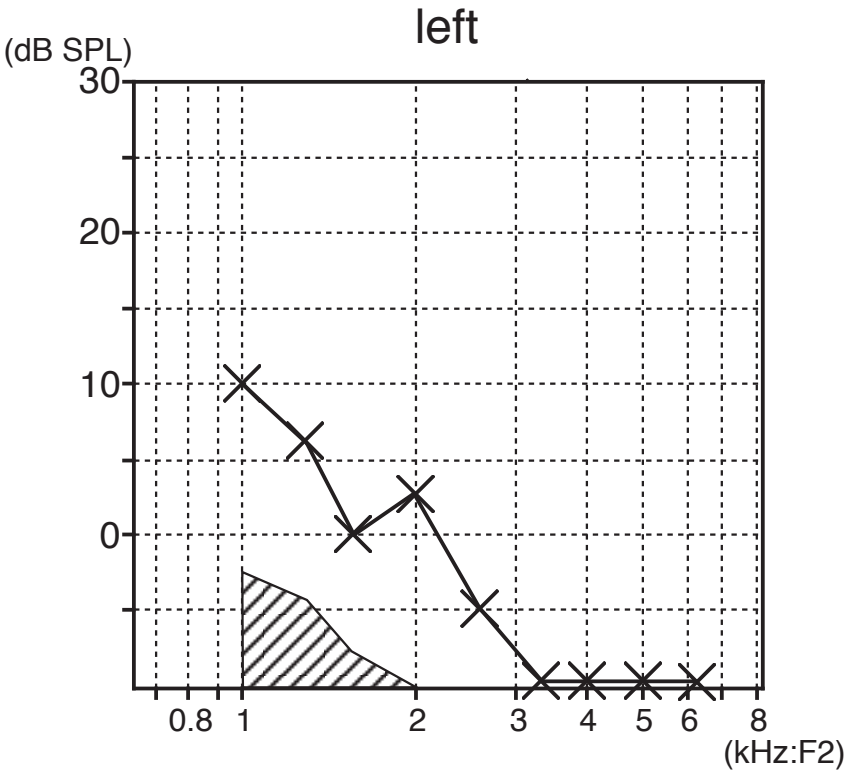

left
(dB SPL)
30
20
10
0
0.8
1
2
3
4
5
6
8
(kHz:F2)

Fig. 2

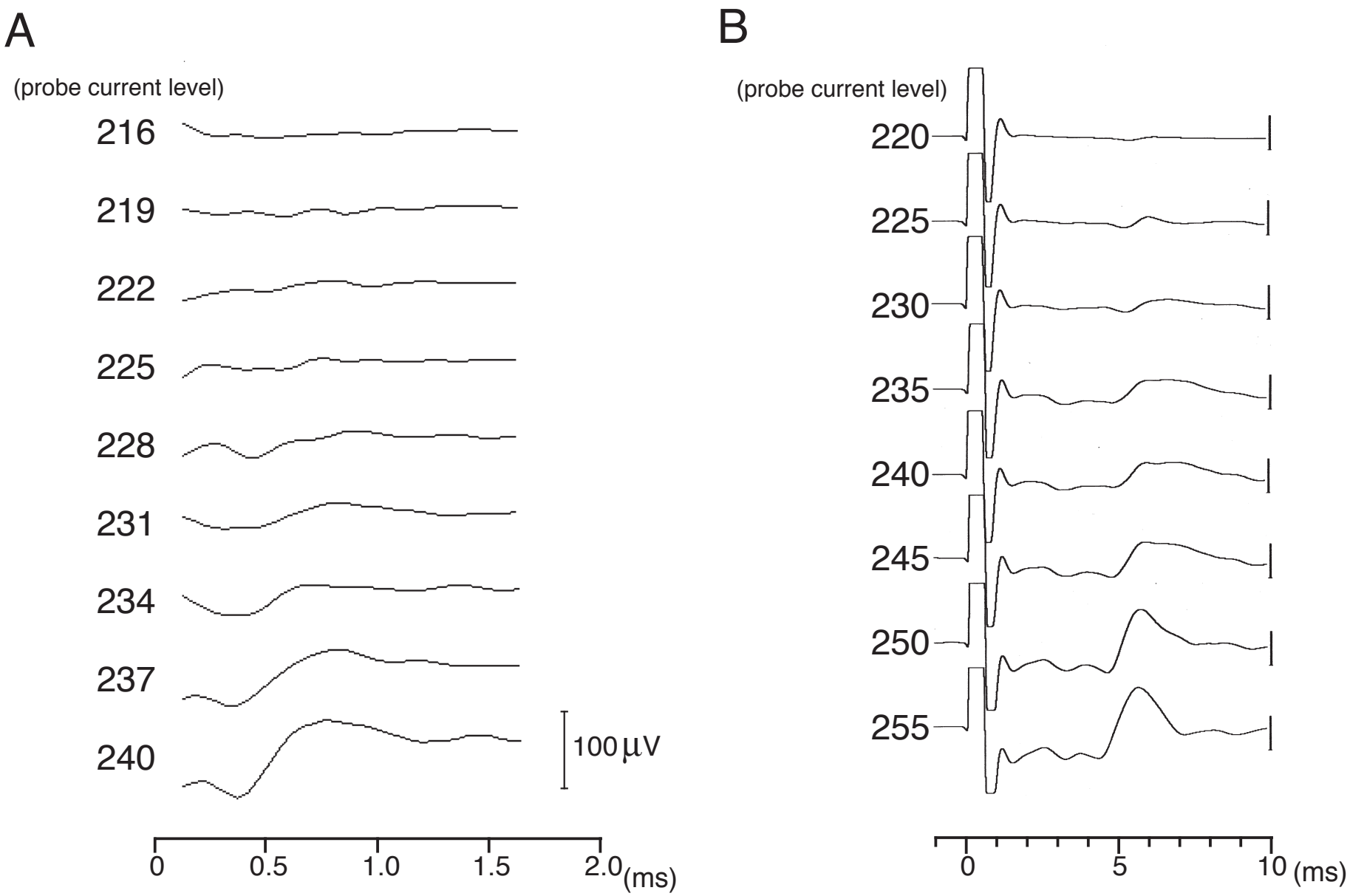

A
(probe current level)
216
219
222
225
228
231
234
237
240
100 µV
0
0.5
1.0
1.5
2.0
(ms)
B
(probe current level)
220
225
230
235
240
245
250
255
0
5
10
(ms)

Fig. 3