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Hearing loss in multiple sclerosis: localization of the auditory pathway lesion according to electrocochleographic findings

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Abstract

Multiple sclerosis is known to affect the myelin of the auditory pathway resulting in acute hearing loss. Two cases of sudden deafness due to multiple sclerosis have been evaluated by conventional audiometry, brainstem auditory evoked response audiometry and transtympanic electrocochleography. The abnormalities of the compound action potential in both patients (enhanced latency, abnormal adaptation using fast stimulus rate) and the normal receptor potentials (cochlear microphonic, summating potential), as well as the absence of brainstem responses suggest a disturbance of synchronization at the level of the first auditory neurone. The electrocochleography provides valuable information for the topodiagnosis of this and other neural hearing losses, especially in the absence of reliable brainstem responses.

Key words: Multiple sclerosis; Hearing loss, sensorineural; Audiometry, evoked response

Introduction

Impairment of the vestibular and auditory pathways is known to be rare in cases of multiple sclerosis (MS). If this occurs, especially when other symptoms of brain stem demyelination are present (McAlpine *et al.*, 1955), it results in provoked or spontaneous nystagmus or sudden hearing loss (Lehnhardt, 1975).

In such cases, negative recruitment, tone decay and impaired dichotic speech discrimination suggest a neural lesion. Myelin degeneration of central nerve fibres has been postulated (Miller *et al.*, 1969; Lehnhardt, 1975). Brainstem auditory evoked potential (BAEP) findings point to a lesion of the most caudal segment of the auditory pathway or even of the hearing nerve, itself (Hopf and Maurer, 1983; Fischer *et al.*, 1985). Electrocochleography provides further information regarding the impairment of the first auditory neurones.

This paper presents additional electrocochleographic findings in order to localize the lesion in the auditory pathway of two patients with definite multiple sclerosis.

Electrocochleography (ECoG) was performed transtympanically under local anaesthesia according to the method described by Aran (1973) and Gibson *et al.* (1983). The WESTRA Electronic ERA device QS/2 was used as the stimulator and averager. Alternated clicks in 10 dB steps, averaged 500 times, served as stimuli for the compound action potential (CAP). In addition to the standard stimulus rate of 20 clicks per second, 100 clicks per second were also used to study the adaptation of CAP to fast stimulus rate. Further, cochlear microphonics (CM) were elicited by 6 msec, 2 kHz tone bursts with 2 msec rise and fall in 10 dB steps. In order to eliminate the DC-components (summating and compound action potential), initial condensation and rarefaction responses were subtracted on-line and 500 sweeps were averaged. Electromagnetic artefacts were separated from CM responses by introducing a time lag, by using a 1 m long tube between the transducer and examined ear.

Case reports

Case 1

A 39-year-old male with diagnosed definite MS since 1975, suffered with fluctuating neurological symptoms including ataxia and diplopia. In his cerebrospinal fluid oligoclonal bands of IgG had already been found in previous examinations. Magnetic resonance imaging showed paraventricular and brain stem lesions typical of MS.

Four weeks before our examination an acute hearing loss on the right with tinnitus occurred (Figure 1). An elevated average pure-tone threshold of 60 dB, combined with rapid tone decay, negative recruitment and poor speech discrimination suggested a neural hearing loss. No stapedial reflexes could be obtained from the right ear. Spontaneous nystagmus to the right and normal bithermal caloric tests were observed. The BAEP revealed normal thresholds and interpeak latencies on the left, but no responses at all on the right. Even wave I was absent on the affected side.

In transtympanic ECoG, a prolonged click evoked CAP latency was observed (Figure 2, left). Receptor potentials (cochlear microphonic and summating potential) appeared to be normal (Figures 2 and 3, left). This finding suggested a lesion of the hearing nerve, itself.

The patient was treated with steroids and a remission occurred (Figure 1). Most of the audiometric findings returned to normal. The only negative result which persisted, was increased tone decay. The stapedial reflex could be elicited at normal threshold, and speech

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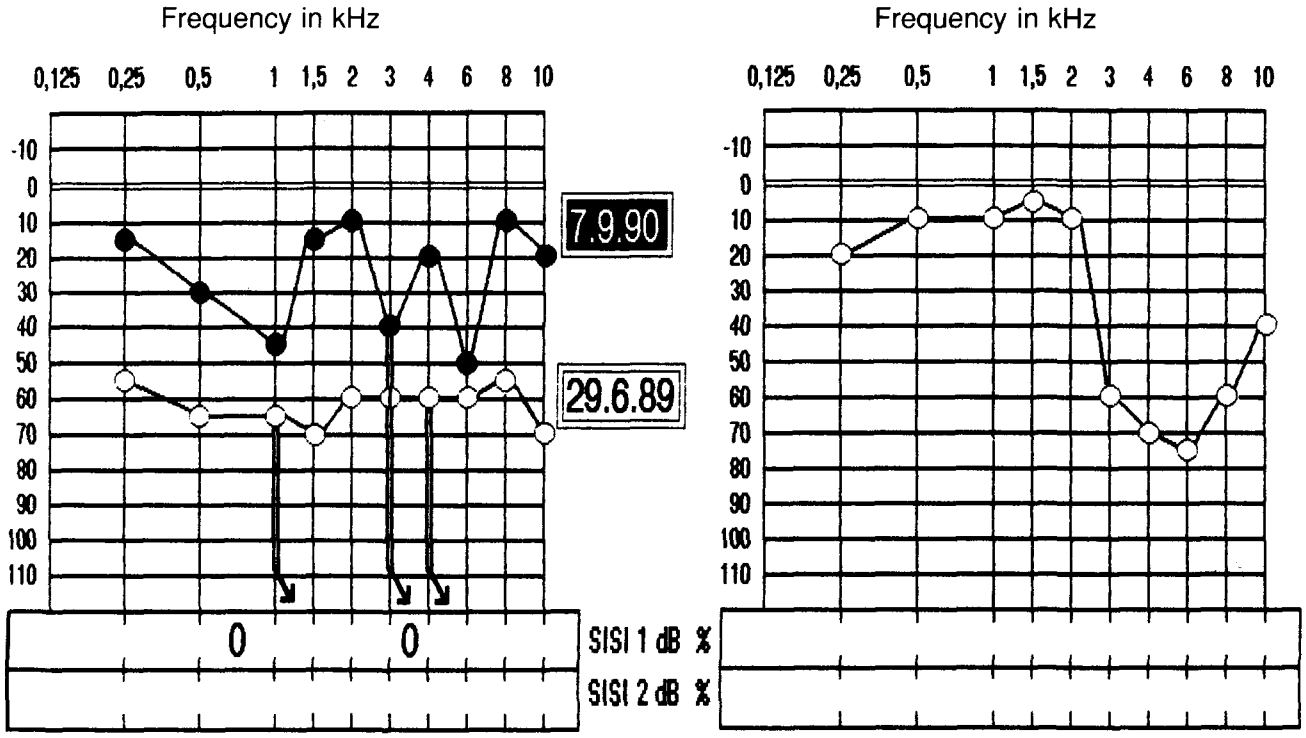

Right ear

Left ear

Impedance

29.6.89

Right			Left	
contralateral				
dB HL	dB SL		dB HL	dB S
90	80	0,5 kHz	—	—
90	85	1 kHz	—	—
90	85	2 kHz	—	—
100	45	4 kHz	—	—
ipsilateral				
—	—	0,5 kHz	110	10(
—	—	1 kHz	—	—
—	—	2 kHz	—	—

7.9.90

Right			Left	
contralateral				
dB HL	dB SL		dB HL	dB SL
90	80	0,5 kHz	85	55
90	85	1 kHz	90	45
90	85	2 kHz	85	75
100	45	4 kHz	95	75
ipsilateral				
85	55	0,5 kHz	110	100
85	40	1 kHz	—	—
80	70	2 kHz	—	—

FIG. 1

Pure tone threshold and stapedial reflexes after acute hearing loss on the right. SISI 0 per cent, tone decay and missing stapedial reflex suggest a retrocochlear origin. One year later, recovery of the pure tone threshold on the right, as well as of the stapedial reflex.

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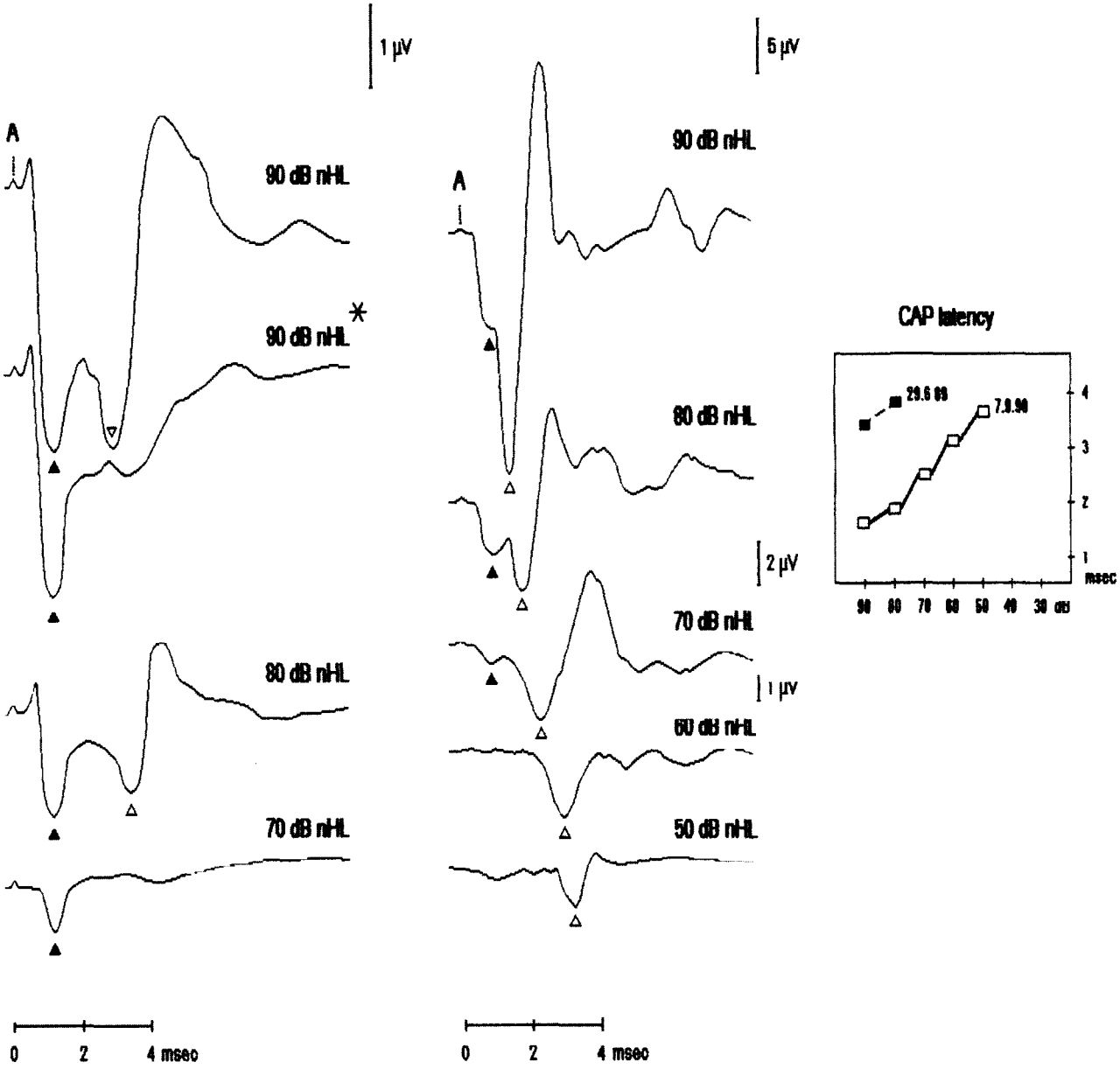

FIG. 2

Click-evoked transtympanic response from the right ear. Left trace: Note the delayed compound action potential (CAP, △) and the normal summating potential amplitude and threshold (SP, ▲) at standard stimulus rate (20 clicks per second). Extreme abolition of the CAP at fast stimulus rate (*), whereas the presynaptic SP remains unchanged. Right trace: the same ear one year later: normal waveforms for the CAP and SP, better CAP threshold, normal intensity-latency function (right).

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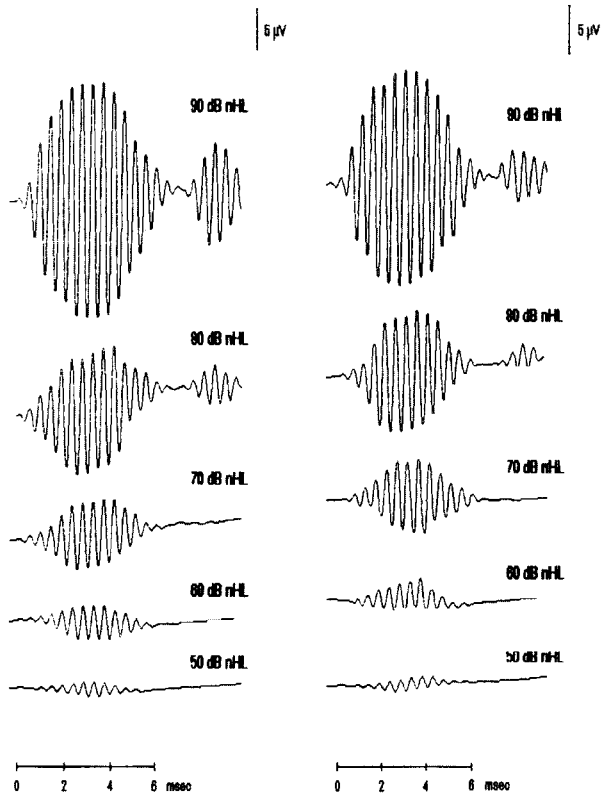

FIG. 3

2 kHz tone burst evoked cochlear microphonics with normal threshold on the same ear as in Figure 2: no evidence of hair cell damage during hearing loss (left trace) and after recovery one year later (right trace).

discrimination recovered as well. Normal BAEP threshold, morphology and interpeak latencies were obtained on both sides. Evoked otoacoustic emissions were present.

ECoG revealed a normal CAP waveform, of normal latency (Figure 2, right). Cochlear microphonics remained unchanged (Figure 3, right).

Case 2

This 55-year-old male was hospitalized because of indefinite neurological disturbances probably due to chronic alcoholism. Two days following admission, he developed an acute unilateral hearing loss (Figure 4). The pure-tone audiogram revealed a 'dead ear' on the right and a 'sensory' hearing loss on the left. Stapedial reflexes were absent on both sides. His baseline hearing thresholds were unknown and he was unable to provide any information due to his delirious confusion. No spontaneous nystagmus was observed and both vestibular organs did not respond to bithermal caloric stimulation. Reliable BAEP responses could not be obtained.

In the ECoG, the receptor potentials (cochlear microphonic and summating potential) were normal, but the CAP was delayed (Figure 5). Although these findings suggested a neural impairment, a right tympanotomy was performed to rule out the presence of perilymphatic fistula. No fistula was found.

Two days after the operation, an acute hearing loss occurred, now on the left (Figure 4). Unfortunately, no further neurological evaluation was possible at this time because the patient developed Korsakoff's syndrome with psychotic signs due to the dramatic bilateral hearing loss. Psychiatric assistance was necessary.

Two weeks later, cerebrospinal fluid examination and magnetic resonance imaging could be performed: pathological IgG quotients in CSF and typical paraventricular changes in MR confirmed a definitive diagnosis of a delayed form of multiple sclerosis.

Discussion

Sudden hearing loss is a rare manifestation of multiple sclerosis. Its onset occurs within the first years of the disease (Daugherty *et al.*, 1983), although latent hearing loss, as in our first patient, has also been described (Daugherty *et al.*, 1983; Franklin *et al.*, 1989).

Acute and extensive demyelination of the auditory pathway is the pathological correlate of hearing dysfunction. Several BAEP studies in patients with definite MS, with or without hearing loss, revealed pathological response patterns of the waves III and V (Chiappa *et al.*, 1980; Hopf and Maurer, 1983; Fischer *et al.*, 1985). Additionally, these same authors described the occasional absence or prolongation of wave I as being associated with MS. They suggest, that demyelination can be localized to the most peripheral parts of the auditory pathway or to the hearing nerve, itself.

Schlesinger (1913, cited by Lehnhardt, 1975) found microscopically lesions of the hearing nerve in patients with MS. In contrast to these findings, a large zone of demyelination of the ventral cochlear nucleus has been described at necropsy in a patient with acute MS and deafness (Brock and Gagel, 1933, cited by Noffsinger *et al.*, 1972). Dix (1965) observed the same site of lesion close to the right inferior cerebellar peduncle including the cochlear nuclei.

The presented electrocochleographic findings in both our cases, suggest a lesion of the peripheral cochlear nerve, assuming the CAP is considered to be generated at the habenula perforata. This is in agreement with the reports of Lang (1967), Pollock *et al.* (1977) and Hopf and Eyshold (1978), who described an involvement of the peripheral nervous system, including other cranial nerves.

In some cases, ECoG may be the only clinical tool indicating the site of lesion for a sudden hearing loss, because it enables the differentiation between sensory and neural hearing impairment. If the electrocochleographic findings in our second case had been correctly interpreted, unnecessary surgery would have been avoided.

This report strongly indicates that the clinical value of ECoG should not be limited to the diagnosis of endolymphatic hydrops. This is an important statement both from clinical and neuro-otological points of view. Transtympanic electrocochleography provides valuable topodiagnostic information, not only in cases of sensory hearing loss or hydrops, but also in retrocochlear lesions such as those associated with multiple sclerosis or acoustic neuromas.

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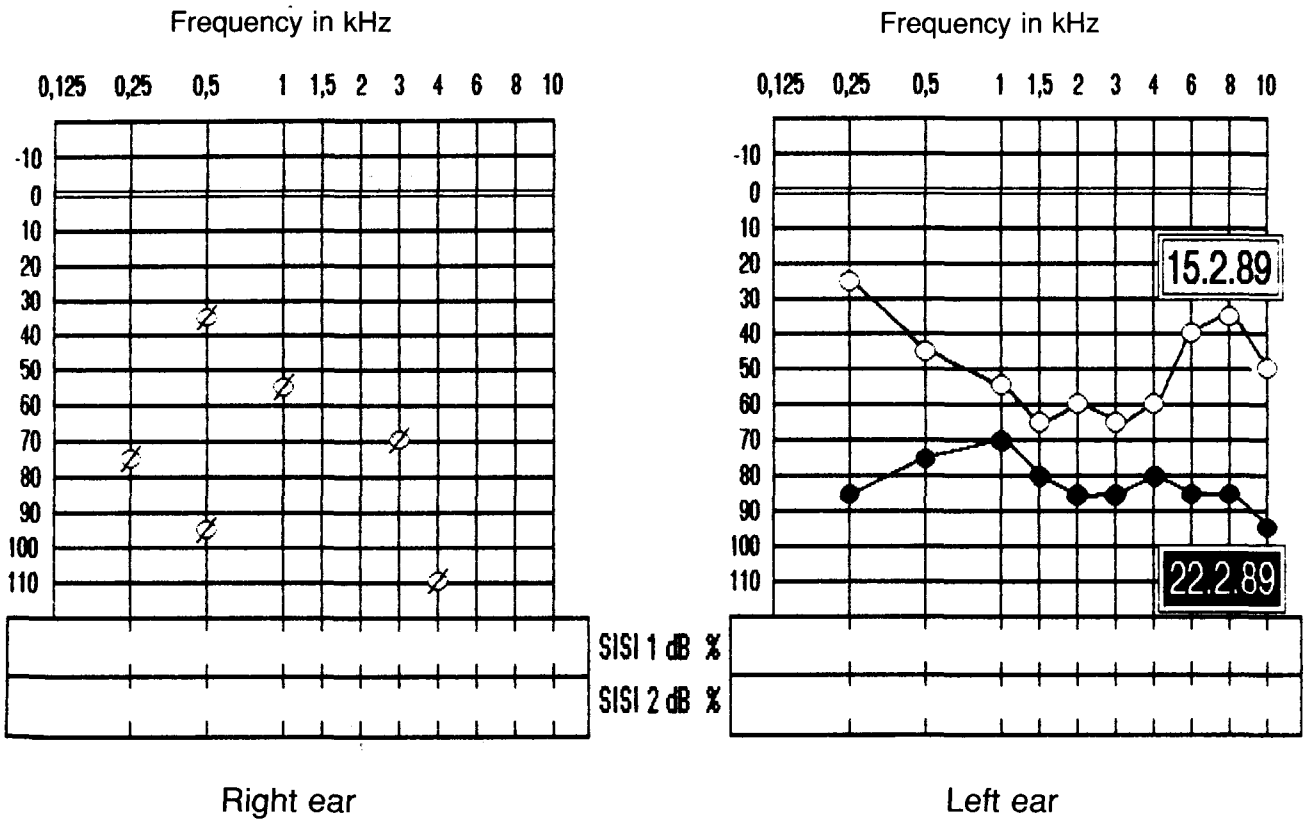

Impedance

Right			Left	
contralateral				
dB HL	dB SL		dB HL	dB SL
110	65	0,5 kHz	—	—
100	45	1 kHz	—	—
105	45	2 kHz	—	—
—	—	4 kHz	—	—
ipsilateral				
—	—	0,5 kHz	—	—
—	—	1 kHz	—	—
—	—	2 kHz	—	—

FIG. 4

Pure tone threshold and stapedial reflexes of the second patient. No detectable thresholds on the right side after masking the left. Some days later, hearing loss on the left increased.

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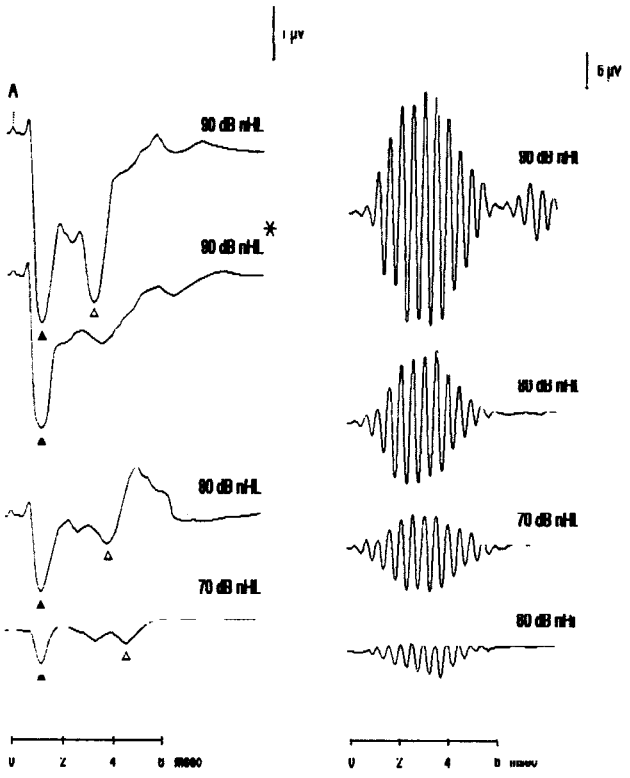

FIG. 5

Electrocochleography on the second patient after acute deafness on the right. Left trace: Normal click evoked summating potential (▲) and delayed compound action potential (△). Right trace: normally configurated toneburst evoked cochlear microphonics at 2 kHz: no evidence of a 'dead cochlea'. Compare these findings with those of the first patient (Figures 2 and 3).

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