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ORIGINAL ARTICLE

Vestibular evoked myogenic potential (VEMP) in patients with auditory neuropathy: Auditory neuropathy or audiovestibular neuropathy?

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Abstract

Conclusion: Our results suggest that isolated auditory or vestibular involvement is unlikely and in fact audiovestibular neuropathy can better explain auditory neuropathy. *Objective:* The purpose of this study was to investigate saccule and related neural pathways in auditory neuropathy patients. *Methods:* Three males and five females diagnosed with auditory neuropathy were included in this prospective study. Patients' ages ranged from 21 to 45 years with a mean age of 28.6 ± 8.1 years and the history of disease was between 4 and 19 years. A group of 30 normal subjects served as the control group. The main outcome measures were the mean peak latency (in ms) of the two early waves (p13 and n23) of the vestibular evoked myogenic potential (VEMP) test in patients and controls. *Results:* Of the 8 patients (16 ears), normal response was detected in 3 ears (1 in right and 2 in left ears). There were unrepeatable waves in four ears and absent VEMPs in nine ears.

Keywords: *Auditory neuropathy, vestibular evoked myogenic potential, vestibule, neural hearing loss*

Introduction

Auditory neuropathy (AN) is an uncommon hearing disorder characterized by an absent or severely abnormal auditory brainstem response (ABR), with preservation of the cochlear microphonics (CM) and otoacoustic emissions (OAEs) [1]. Clinically, AN is defined as (1) hearing loss, usually bilateral, of any degree; (2) normal outer hair cell function as evidenced by the presence of OAEs and/or CM; (3) abnormal evoked potentials beginning with wave I of the ABR; (4) poor speech perception; and (5) absent acoustic reflexes to the ipsilateral and contralateral tones at a 110 dB hearing level [2]. The overall incidence rate as reported varies from 0.54% to 11% of the hearing-impaired population [3,4]. The pathophysiology of AN has been considered to involve an abnormality of the peripheral auditory system localized to the inner hair cells, to the eighth cranial nerve, or to the synapse between them [2,5]. Although different neurological disorders can cause AN with or without associated peripheral neuropathy, it is estimated that approximately half of all cases are idiopathic [6].

Vestibular evoked myogenic potential (VEMP) testing is a resource for measuring the function of the saccule and inferior vestibular nerve. Loud monaural clicks evoke an initial inhibitory potential (p13-n23) in the tonically contracted ipsilateral sternocleidomastoid muscle (SCM). This test has seen growing popularity in clinical assessment of vestibular integrity in a number of different disorders such as acoustic neuroma, high and low tone sensorineural hearing loss, brainstem lesions, and vestibular neuropathy.

The use of the VEMP test for vestibular integrity assessment of AN has only recently been reported, and there are fewer than eight cases documented in the literature [7–9]. The aim of this paper was to investigate saccule and related neural pathways in AN

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patients. Eight patients (16 ears) with AN were tested for VEMPs. Results were compared with those of 30 healthy controls (60 ears).

Material and methods

The present study was approved by the Institutional Review Board and each patient gave informed consent. From May 2007 to April 2008, three males and five females diagnosed with AN were included in this study. AN was diagnosed based on the criteria described above [2]. A group of 30 normal subjects (15 males and 15 females, aged 18–35 years, with a mean of 27 years of age) with no history of neurologic disease or auditory and vestibular disorder, selected on a case-matched basis for age and sex, served as the control group. All the subjects and patients underwent a thorough history and physical examination (i.e. neurological and systemic).

Audiometric tests

All patients and controls underwent standard pure-tone air- and bone-conducted audiometry (250/8000 Hz) and speech reception threshold (SRT) testing. Standard tympanometry and acoustic reflex testing to pure tone stimuli from 500 to 4000 Hz were also performed.

Distortion product otoacoustic emissions (DPOAEs) test

DPOAEs were recorded and analyzed using the Madsen Capella system. Two simultaneous pure-tone signals with frequency ratio (f2:f1) 1.2 at 65 dB SPL were presented at 500, 1000, 2000, 3000, and 4000 Hz. The frequency of f2 was changed in half octave steps from 750 to 6000 Hz. DPOAEs were plotted on a DP-gram and were accepted as normal if they had an amplitude of at least 5 dB above the noise level.

Auditory brainstem response test

ABRs were recorded on the Biologic system using an electrode montage of forehead (CZ) to the ipsilateral mastoid (test ear) and ground to contralateral mastoid (non-test ear). The amplifier bandpass was 100–3000 Hz. Alternating polarity click stimuli were presented monaurally at a rate of 11.1 Hz at 90 dB nHL. Averaged responses to 1024 clicks were collected in each of 2 runs. Reproducible components were discerned and latencies were measured for each.

Vestibular evoked myogenic potential test

The subjects were seated in a chair with their back against the back support of the chair. The subject had to flex the head approximately 30° forward and rotate it approximately 30° to each side for recording of potentials from the opposite side. To contract the SCM during the VEMP test, a feedback method was applied based on the use of a blood pressure manometer with an inflatable cuff (Welch-Allyn, Skaneateles Falls, NY, USA) between the patient's hand and jaw [9]. The subject pushed with his or her head against the hand-held cuff to generate a cuff pressure of 40 mm Hg. The responses were obtained from each side separately using tone bursts of 500 Hz (95 dB nHL, rise/fall time 1 ms, plateau time 2 ms, repetition rate 5 Hz), which were delivered unilaterally with insert earphones ((Telephonics TDH-49P). VEMPs were recorded with an Interacoustic audiometer model EP25 with a two-channel averaging capacity. The acoustically evoked VEMP responses were amplified, bandpass filtered (10 Hz–1.2 kHz), and averaged. Each side was subjected to 2 runs of 100 tone bursts, which were averaged. Analysis sweep time was 100 ms. The two averaged records were compared at the end of each test and had to be perfectly coincident in order to eliminate potentials artifacts. We measured the mean peak latency (in ms) of the two early waves (p13 and n23) of the VEMPs, since these potentials were of saccular origin [10]. p13 is the first positive peak of VEMPs and n23 the first negative peak following p13. We defined the latency as the time from the onset of the stimulus to the peak. We also measured peak to peak amplitude.

Result

Patients' ages ranged from 21 to 45 years with a mean age of 28.6 ± 8.1 years and the history of disease was between 4 and 19 years. All patients and controls had a normal tympanometric examination. Acoustic reflexes were detected in controls and were absent in patients. Neurological and systemic examinations were normal in patients and controls. DPOAEs and ABRs were normal in all audiologically normal subjects. VEMP response to tone burst was displayed in all healthy ears. These healthy controls demonstrated short latency waves to stimuli during tonic neck flexor activation. Mean latencies of p13 and n23 in this group were 14.47 ± 1.19 ms and 23.79 ± 3.36, respectively. The mean p13-n23 amplitude was 114.09 ± 35.54 μV.

All patients had bilateral involvement. The patients had few complaints that could be associated with their

vestibular involvement. Test results are summarized in Table I for right ears and Table II for left ears. No DPOAE abnormalities were found in any of the patients' ears. No ABR responses were found in left ears. Results of ABR on right ears showed that only two patients had prolonged I–V interpeak latencies.

VEMPs displayed normal responses in three ears (one in right and two in left ears) (Figure 1) and abnormal responses in all others including unrepeatable waves in four ears and absent VEMPs in nine ears. There were no different audiovestibular symptoms between the subjects with normal VEMP results and those with abnormal results.

Discussion

In the literature the occurrence of vestibular involvement in this rare auditory dysfunction remains unknown, since the vestibular tests are not carried out on a routine basis in these patients. In the primary study of 10 AN patients, Starr et al. explained horizontal nystagmus on lateral gaze in three patients and absent responses to caloric tests (ENG) in another one [1]. Fujikawa and Starr [11] emphasized that asymptomatic vestibular disorders are common in patients with AN when a peripheral neuropathy is also present. They suggested that the reason for the abnormal vestibular test results was likely a neuropathy of the vestibular nerves and vestibular neuropathy in patients with auditory neuropathy is a late manifestation.

Advances in the accurate measurement of sacculocollic pathway function have led to the recent identification of an impaired sensory or neural response involving the inferior vestibular nerve and/or its end organ, the saccule, in AN. These abnormalities occurred in 13 of the 16 patients' ears in this study. These results provide evidence that certain types of vestibular neuropathies are associated with a neuropathy of the auditory nerves. A review of the literature revealed eight examples of this type of association [7–9]. Sheykholeslami et al. [7] reported no response on left ear stimulation and a biphasic response with normal latency and amplitude on right ear stimulation of short tone-burst VEMPs in one patient with AN. In another study by these authors,

Table I. Details of the eight patients with auditory neuropathy, auditory brainstem responses (I–III and I–V interpeak latency and wave V latency), distortion product otoacoustic emissions (DPOAEs), and vestibular evoked myogenic potentials (VEMPs, p13 and n23 latencies) for right ears are shown.

			PTA-AC (dB) / PTA-BC (dB)					ABR latency (ms)				VEMP latency (ms)	
Patient no.	Age (years)	Sex	500 (Hz)	1000 (Hz)	2000 (Hz)	4000 (Hz)	SRT(dB)	I–III	III–V	V	DPOAE	p13	n23
1	28	F	80	75	50	80	CNT	...	...	7.06	Normal	...	...
			75	70	45	70							
2	21	M	60	60	20	70	80	...	...	...	Normal	...	...
			60	55	10	65							
3	45	M	70	70	70	70	70	...	...	...	Normal	15.33	25
			60	60	60	60							
4	25	F	50	50	20	30	60	...	...	8.48	Normal	...	...
			40	40	10	20							
5	24	F	110	110	110	110	CNT	...	...	...	Normal	...	...
			–	–	–	–							
6	28	F	70	40	40	40	65	...	...	...	Normal	...	...
			65	40	30	40							
7	22	F	60	60	70	70	70	...	...	...	Normal	...	...
			60	60	60	60							
8	36	M	60	60	50	70	55	...	...	...	Normal	...	...
			50	55	40	70							

SRT, speech reception threshold; CNT, could not test.

Table II. Pure tone and speech audiometry, auditory brainstem responses (I–III and III–V interpeak latency and wave V latency), distortion product otoacoustic emissions (DPOAEs), and vestibular evoked myogenic potentials (VEMPs, p13 and n23 latencies) for left ears of the eight patient with auditory neuropathy.

	PTA-AC (dB)					ABR latency (ms)				VEMP latency (ms)	
	PTA-BC (dB)										
Patient no.	500 (Hz)	1000 (Hz)	2000 (Hz)	4000 (Hz)	SRT (dB)	I–III	III–V	V	DPOAE	p13	n23
1	80	70	60	60	CNT	...	...	...	Normal	14.33	26.67
	–	60	55	55							
2	80	60	65	65	CNT	...	...	...	Normal	...	...
	60	60	60	60							
3	60	60	60	60	65	...	...	...	Normal	14	25.67
	50	50	50	50							
4	60	40	20	20	55	...	...	...	Normal	...	...
	50	30	10	10							
5	100	100	100	100	CNT	...	...	...	Normal	...	...
	–	–	–	–							
6	60	40	10	10	90	...	...	...	Normal	...	...
	50	30	0	0							
7	60	40	20	20	60	...	...	...	Normal	...	...
	55	35	10	10							
8	80	60	60	60	CNT	...	...	...	Normal	...	...
	65	60	55	50							

SRT, speech reception threshold; CNT, could not test.

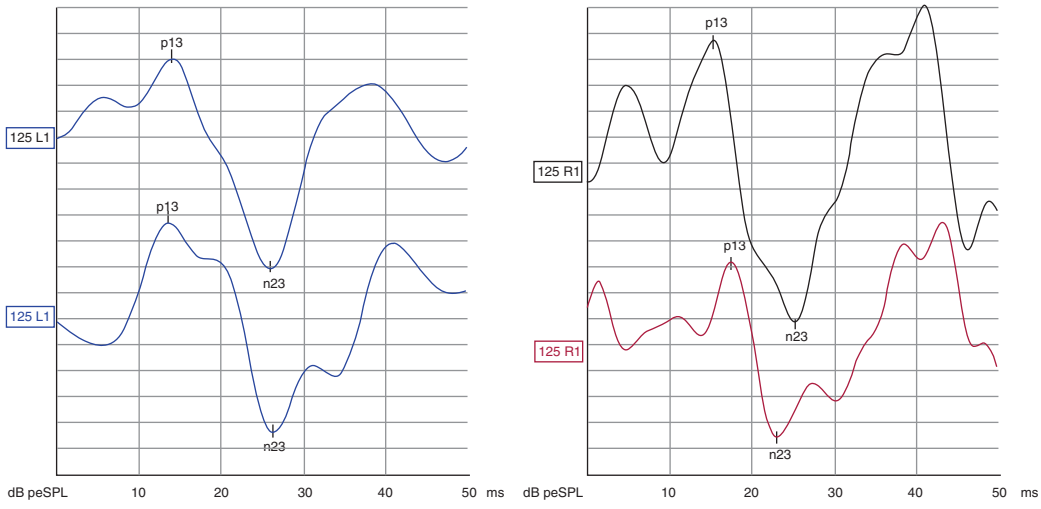

Figure 1. Patient no. 3, showing bilateral auditory neuropathy and positive VEMP responses. Latencies for first positive waves on the left side (A) were 14 ms (p1) and on the right side (B) were 15.33 ms (p1). Latencies for first negative waves on the left side (A) were 25.67 ms (n1) and on the right side (B) were 25 ms (n1).

VEMPs were abolished in all three patients (all female; 57, 61, and 71 years old) [9]. In other study by Akdogan et al. [8], VEMPs were detected in three of six ears in three children with AN. In this study p13 latencies were 10.8, 11.4, and 10.8 ms and n23 latencies were 16.8, 18, and 18.3 ms, respectively. Normal value of latencies in this study was accepted as 11.3 ms for p13 and 18.2 ms for n23. In our study we measured latencies of p13 and n23 for comparison among patients and subjects and site

of lesion assessment [12], because prolonged VEMP latencies, especially prolonged p13, would strongly suggest lesions in the retrolabyrinthine (vestibulospinal tract) [12]. Like this result, some investigators believe that inner hair cells and/or synapses linking inner hair cells to the dendrites of auditory nerve fibers were impaired in the auditory part too. Raw peak to peak amplitudes of potentials are less reproducible and displayed wider variations than latencies did. The response amplitude was also dependent on the muscle tension, contraction level, and patient compliance.

In our study, like others, we suggest that the lack of vestibular symptoms reflects both the bilateral distribution of the disorder and the slow progression of vestibular neuropathy. In addition, the results demonstrated a centrally compensated decrease in the response of the vestibular end organs, which was associated with hearing loss. These findings imply that a subclinical well-compensated malfunction of the vestibular system is associated with the auditory destruction.

In various studies in this field, the vestibular system was tested using a mixture of vestibular tests including bithermal caloric test, eye movement recording tests, rotation test, ENG, and VEMP. Each of these tests examines only a part of the vestibular system and it is not correct to state that the whole vestibular system is normal when using some of these tests. The results of these studies provide further evidence that a vestibular neuropathy is associated with AN. With regard to the results obtained in this study, the authors recommend that complementary studies are carried out to evaluate the superior vestibular nerve route and caloric tests. In further studies, the new disorder in the path of the superior vestibular nerve in these patients may be found.

In other words, AN and vestibular neuropathy may be two limits of inner ear and cranial nerve VIII involvement. In addition to the anatomic proximity of the vestibule to the auditory system, the great similarity in cochlear and vestibular hair cell ultrastructures and the common arterial blood supply of the cochlea and vestibular end organs via the same end artery all support the possibility of vestibular, especially saccular and inferior vestibular nerve, deterioration associated with the cochleoauditory pathogens. Therefore we propose that isolated auditory or vestibular involvement is unlikely and in fact audiovestibular neuropathy can better explain this entity.

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Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

References

[1] Starr A, Picton TW, Sininger Y, Hood LJ, Berlin CI. Auditory neuropathy. Brain 1996;119:741–53.

[2] 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.

[3] 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.

[4] Kumar UA, Jayaram MM. Prevalence and audiological characteristics in individuals with auditory neuropathy/auditory dys-synchrony. Int J Audiol 2006;45:360–6.

[5] Starr A, Michalewski HJ, Zeng FG, Fujikawa-Brooks S, Linthicum F, Kim CS, et al. Pathology and physiology of auditory neuropathy with a novel mutation in the MPZ gene (Tyr145 →Ser). Brain 2003;126:1604–19.

[6] Starr A, Sininger YS, Pratt H. The varieties of auditory neuropathy. J Basic Clin Physiol Pharmacol 2000;11:215–30.

[7] Sheykholeslami K, Schmerber S, Habiby Kermany M, Kaga K. Sacculo-collic pathway dysfunction accompanying auditory neuropathy. Acta Otolaryngol 2005;125:786–91.

[8] Akdogan O, Selcuk A, Ozcan I, Dere H. Vestibular nerve functions in children with auditory neuropathy. Int J Pediatr Otorhinolaryngol 2008;72:415–19.

[9] Sheykholeslami K, Kaga K, Murofushi T, Hughes WD. Vestibular function in auditory neuropathy, Acta Otolaryngol 2000;120:849–54.

[10] Vanspauwen R, Wuyts FL, Van de Heyning PH. Improving vestibular evoked myogenic potential reliability by using a blood pressure manometer. Laryngoscope 2006;116:131–5.

[11] Fujikawa S, Starr A. Vestibular neuropathy accompanying auditory and peripheral neuropathies. Arch Otolaryngol Head Neck Surg 2000;126:1453–6.

[12] Murofushi T, Shimizu K, Takegoshi H, Cheng PW. Diagnostic value of prolonged latencies in the vestibular evoked myogenic potentials. Arch Otolaryngol Head Neck Surg 2001;127:1069–72.