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Contrasting results of tests of peripheral vestibular function in patients with bilateral large vestibular aqueduct syndrome

Yu-Juan Zhou [1], Yong-Zhen Wu [1], Ning Cong [1], Jing Yu, Jun Gu, Jing Wang *, Fang-Lu Chi *

Department of Otology and Skull Base Surgery, Eye Ear Nose & Throat Hospital, Fudan University, Shanghai, China
Shanghai Auditory Medical Center, Shanghai, China
Key Laboratory of Hearing Science, Ministry of Health, Shanghai, China

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HIGHLIGHTS

- LVAS patients have lower VEMP thresholds, higher amplitudes, and shorter latencies.
- The otolithic organs in patients with LVAS are hypersensitive to sound or pressure.
- Patients with LVAS have semicircular canal hypofunction and disequilibrium.

ABSTRACT

Objective: To analyze and summarize the effect of bilateral large vestibular aqueducts in peripheral vestibular organ function.
Methods: Eighteen patients with bilateral large vestibular aqueduct syndrome (LVAS; Study Group) and 18 healthy volunteers (Control Group) were investigated using audiometry, caloric test, sensory organization test (SOT), and vestibular-evoked myogenic potential (VEMP) tests.
Results: All 18 patients (36 ears) exhibited sensorineural hearing loss. For cervical VEMP (cVEMP), the Study Group showed lower thresholds (Study Group vs. Control Group: 71.4 vs. 75.3 dB nHL; $p = 0.006$), N1 latencies (24.1 vs. 25.2 ms; $p = 0.026$) and shorter P1 (15.3 vs. 16.6 ms; $p = 0.003$), and higher amplitudes (400.7 vs. 247.2 μV; $p < 0.001$) than the Control Group. For ocular VEMP (oVEMP), the Study Group had lower thresholds (79.3 vs. 81.8 dB nHL; $p = 0.046$) and higher amplitudes (40.6 vs. 14.4 μV; $p < 0.001$) than the Control Group. Fourteen of 16 patients (87.5%) who completed caloric tests had abnormal results, and 10 of 18 patients (55.6%) exhibited abnormal results in SOTs.
Conclusions: The hyperfunction of vestibular test in otolithic organs and the hypofunction of vestibular test in semicircular canals, as well as the dysfunction in the balance test were demonstrated in patients with LVAS.
Significance: Our findings can help clinicians gain a better understanding of the characteristics of vestibular organ function in patients with LVAS, which can facilitate optimal targeted treatment.

1. Introduction

Carolo Mondini (1997) first discovered large vestibular aqueduct syndrome (LVAS) in 1791 after finding cochlear malformation in the temporal bone of a child. The vestibular aqueduct, filled with endolymph, is a small bony canal located between the vestibule and the posterior cranial fossa. The diameter of the vestibular aqueduct normally ranges between 0.4 mm and 1 mm in healthy people (Przewozny et al., 2015). In 1978, Valvassori and Clemis (1978) first described and named LVAS after finding the association between the large vestibular aqueduct and sensorineural hearing loss by observing 50 patients radiographically. The Valvassori criterion involves a diameter >1.5 mm at the midpoint in patients with LVAS. However, El-Badry et al. (2016) suggested that the Cincinnati criterion (midpoint 0.9 mm or operculum 1.9 mm) was more sensitive than the Valvassori criterion for diagnosing LVAS, after finding that only 81% of the ears of children with LVAS met the Valvassori criteria.

* Corresponding authors at: 83# FenYang Rd., XuHui District, Shanghai 200031, China.
E-mail addresses: jingwang6437@163.com (J. Wang), chifanglu@126.com (F.-L. Chi).
[1] These authors contributed equally to this work.

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A large vestibular aqueduct is often associated with hearing loss. Thus, patients with LVAS may have sudden, fluctuating, or progressive hearing loss, since childhood. Nevertheless, there are some controversies about the type of hearing loss involved. Three types, i.e., sensorineural hearing loss, mixed hearing loss, and conductive hearing loss have all been reported. Corticosteroid therapy and hyperbaric oxygen therapy may be a potential treatment for sudden sensorineural hearing loss in patients with LVAS (Gopen et al., 2011; Shilton et al., 2014). In a recent study, Nishio et al. (2016) found that restoration of SLC26A4 expression can reduce or prevent fluctuation in hearing. Usually, cochlear implantation is an effective approach for increasing auditory perception in patients with LVAS.

Besides experiencing hearing loss, patients with LVAS may also have vestibular dysfunction. A recent theory proposed by Zalewski et al. (2015) suggested that doctors should be aware of the high prevalence of vestibular dysfunction in patients with LVAS. Vestibular-evoked myogenic potential (VEMP) tests have conventionally been used to evaluate otolithic function in patients with vestibular disorders. The cervical VEMP (cVEMP) test is considered to reflect saccular function and inferior vestibular nerve input pathways, and it has been shown that saccular neurons have a strong projection to the sternocleidomastoid muscles (Pereira et al., 2015). The ocular VEMP (oVEMP) test can reportedly reflect utricular function and the functioning of the superior vestibular nerve input pathways, and it has been shown that utricular neurons have a strong projection to the inferior oblique muscle. The oVEMP is generated by the activation of utricular afferents and is mediated by a crossed otolith–ocular pathway (Heydari et al., 2015; Zhou et al., 2016).

However, systematic studies of peripheral vestibular organs function (particularly the function of the otolithic organs) in patients with LVAS are rare. In the present study, we analyzed the results of the cVEMP, oVEMP, caloric, and sensory organization tests (SOTs) in patients with LVAS, in order to help clinicians gain a better understanding of the characteristics of vestibular organ function in LVAS patients, which may facilitate use of the optimal targeted treatment.

2. Materials and methods

2.1. Subjects

This retrospective study included 18 patients who visited the Eye and Ear, Nose, & Throat Hospital of Fudan University, from May 2015 to June 2016. The patients were diagnosed with LVAS according to the Valvassori criteria (the vestibular aqueduct diameter >1.5 mm at the midpoint, in the axial view in a computed tomography scan) (Valvassori and Clemis, 1978). There were nine males and nine female patients, aged 7–27 years, with a mean age (±standard deviation [SD]) of 16.4 ± 5.7 years, in the Study Group. Additionally, 18 healthy volunteers, aged 13 to 29 years, with a mean age (±SD) of 18.7 ± 5.2 years, were recruited as the Control Group. Middle ear pathologies, such as otosclerosis and chronic otitis media, could result in absent VEMP responses elicited by air-conduction stimuli (Zhou et al., 2012). Hence, the patients and volunteers were required to have normal tympanic membranes on otoscopy for inclusion into the study. Those subjects who had external and middle ear diseases, or other internal ear diseases, were excluded. This study was approved by the institutional review board, and all of the patients provided written informed consent.

2.2. Vestibular-evoked myogenic potential test

All subjects were placed supine in a sound-proof examination room. Air-conducted sound (ACS) with a short tone burst (2-ms rise/fall time and 2-ms plateau time) at 500 Hz was used. The electromyography signals were amplified in a Bio-Logic Navigator PRO system (Natus Medical Inc., San Carlos, CA, USA). In the cVEMP test, according to the manual, a non-inverting electrode was placed over the upper sternum, an inverting electrode was placed on the middle of sternocleidomastoid muscle (SCM), the same as the sound stimulation side, and a common electrode was placed on the middle of contralateral SCM. The sites of the electrodes did not need to be changed when the test was repeated on the other side, because the electrodes on both sides of SCM can be switched between inverting and common electrodes automatically. Patients were asked to lift their heads when they heard a sound from the calibrated insert headphones. The waveforms we recorded using this method were inverted as compared with the commonly used methods (non-inverting: SCM, inverting: sternum, and common: forehead). Positive peaks display in a downward direction and negative peaks display in an upward direction.

In the oVEMP test, an inverting electrode was placed on the orbital margin below the eye, a non-inverting electrode 20 mm below inverting electrode on the cheek, contrary to the sound stimulation side. A common electrode was placed on the contralateral orbital margin below the eye, the same as the sound stimulation side. When repeat the test on the other side, we just need to move non-inverting electrode to the contralateral cheek, because the electrodes on both sides of orbital margin below the eyes can be switched between inverting and common electrodes automatically. During the test, patients were asked to gaze on a target about 30° above the visual horizontal line when they heard sound from the calibrated insert head phones.

The starting stimulus intensity was 95 dB nHL; then, the intensity was decreased in 5 dB nHL steps until the VEMP response was abolished. This process was repeated three times. The response threshold represented the stimulus intensity of the last characteristic waveform. The presence of cVEMP was recorded when the first positive–negative–positive peak (P1–N1–P2) appeared and the presence of oVEMP was recorded when the peaks of the first negative and positive biphasic wave (N1–P1) appeared. The P1 latency and N1 latency were defined from 0 ms to the corresponding maximum peak of the recorded waveforms. The vertical distance of the P1 and N1 was considered the amplitude.

2.3. Caloric test

The subjects were placed in a supine position, with the head flexed at 30°, in a darkroom. A constant flow of air (8 L/min) at 49 °C or 23 °C for 60 s was blown into each ear by means of a GN Otometrics Type air irrigator (Otometrics, Taastrup, Denmark). The ear was irrigated with cool air prior to irrigation with hot air, with a 5-min interval between the two irrigations. The maximum slow-phase velocities of nystagmus were calculated using a GN Otometrics Type 1068 system (Otometrics, Taastrup, Denmark). If the total response on both sides was <15°/s, it was regarded as bilateral canal paresis. A canal paresis (unilateral weakness) value ≥22% was considered to be an abnormal response.

2.4. Sensory organization test

The SOT was conducted by means of computerized dynamic posturography (EquiTest System, Neurocom Inc., Clackamas, OR, USA). This can be used to distinguish on which sensory system (somatosensory, visual, or vestibular) the subjects rely most for maintaining balance. The SOT employs six sensory conditions (tested three times each) by changing the surface and visual surroundings systematically. The summary data includes scores for each condition, scores for each sensory system, and a composite score (the weighted average of the scores of all conditions). Results