CASE REPORT

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Posterior Semicircular Canal Dehiscence and Auditory Neuropathy in a 22-month-old Male Baby

Görkem ERTUĞRUL[a]

[a]Department of Audiology, Hacettepe University Faculty of Health Sciences, Ankara, TURKEY

ABSTRACT Semicircular canal dehiscence (SCD) is usually seen in children, but it rarely occurs in adults. Posterior SCD in children is scarcely reported in the literature. This study reports the presence of posterior SCD and comorbid auditory neuropathy in a 22-month-old male baby. The video head impulse test was normal, but cervical vestibular-evoked myogenic potential test supported SCD on the left side. Also, temporal CT of the patient revealed posterior SCD. Cochlear microphonic was detected bilaterally by the Auditory Brainstem Response test. Posterior SCD may cause different audio-vestibular signs. Early vestibular assessment in children with congenital hearing loss, especially auditory neuropathy, is suggested because knowing the additional problems associated with the vestibular function will ease the prognosis of vestibular disorder and protect the children's balance functions.

Keywords: Child; semicircular canals; vestibular diseases; auditory neuropathy

Semicircular canal dehiscence (SCD), which is characterized by conductive hearing loss depending on the third window effects especially in low frequencies, vertigo, positive Tullio phenomenon (nystagmus induced with loud sound), and Hennebert signs (nystagmus induced with pressure), is a rare vestibular condition in children.[1] Commonly, the dehiscence occurs in the superior semicircular canal in both adults and children. In children, the prevalence of superior SCD and posterior SCD was 3.3% and 2.1%, respectively.[2] Furthermore, the prevalence of SCD was higher in children <3 years (2.3%) than in children >3 years (1.5%).[2]

The etiology of SCD is unknown. Some studies have asserted that a developmental defect may cause SCD, and head trauma or increased intracranial pressure from the temporal lobe may reveal the clinical symptoms during adulthood.[3] The superior semicircular canal of children is significantly thinner than that of adults.[4]

The aim of this case report is to show the presence of posterior semicircular canal dehiscence and comorbid auditory neuropathy in a 22-month-old male baby.

CASE REPORT

A 22-month-old male baby with congenital bilateral profound sensorineural hearing loss (SNHL) was referred to our hospital. According to patient history received from the parents, the patient failed to pass newborn hearing screening at another center and was diagnosed with profound SNHL at two months of age. He started using hearing aids in both ears until the age of eight months and then gave up using the aids regularly because of his speech and language skills did not develop. Since then, the patient started to be followed in our hospital. According to the parents, the gross motor development (holding head, sitting without support, and independent walking) of the baby was within normal durations. Written and verbal

Correspondence: Görkem ERTUĞRUL
Department of Audiology, Hacettepe University Faculty of Health Sciences, Ankara, TURKEY
E-mail: ertugrulgorkem@gmail.com

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consent was taken from the parents for the publication of this report.

Audiological assessments with and without the hearing aids of the patient were performed, and his behavioral responses to auditory stimulus were observed by Visual Reinforcement Audiometry in the free field. The patient was not interested in pure tones frequency-modulated (FM) stimulus, and the speech awareness threshold was absent at 75 dB HL, but he showed some responses to frequency-specific speech sounds (/ba/, /sh/, /s/) at approximately 65 dB HL. The patient's middle ear functions were within normal limits, as determined by acoustic immitancemetry. The ipsilateral and contralateral acoustic reflexes and TEOAEs were bilaterally absent. Diagnostic ABR (*Vivosonic® Integrity ABR system*, Canada) was performed during the patient's natural sleep. No bilaterally V wave in the click and 500 Hz tone burst stimulus at 99 dB nHL was reported. However, cochlear microphonic was observed on both sides during the recordings at the rarefaction and condensation polarities (Figure 1).

Spontaneous nystagmus and gaze nystagmus were absent, and oculomotor tests (ocular pursuit, saccade, optokinetic nystagmus) were normal. Vestibulo-ocular reflex (VOR) gains were measured by the video Head Impulse Test (*Synapsis® vHIT Ulmer*, France) and the mean gains of VOR for each SSCs were within the normal limit (>0.80) and saccades were not observed (Table 1). The air-conducted cVEMP test (*Otometrics-ICS Chartr® EP 200*, Denmark) using air conduction 500 Hz tone burst stimuli was performed. Although P1-N1 latencies (P1 11.17 msec, N1 19.17 msec for the right side and P1 11.58 msec, N1 20.17 msec for the left side at 95 dB nHL) on both sides were close to normal limits, amplitudes of P1-N1 on the left side (99.26 µV at 95 dB nHL) were quite higher than those of the right side (42.36 µV at 95 dB HL) but lower than the normal limits.[5] The cVEMP results were presented in Table 2. In addition, P1-N1 was obtained at 65 dB nHL on the left side (Figure 2). Computed tomography (CT) of the temporal bones of the patient revealed posterior SCD on the left side, although inner ear structures and the 8th cranial nerve were bilaterally normal (Figure 3). Ocular VEMP and the caloric test could not be performed because the patient was crying and became agitated.

The patient was a candidate for cochlear implantation because he had bilateral auditory neuropa-

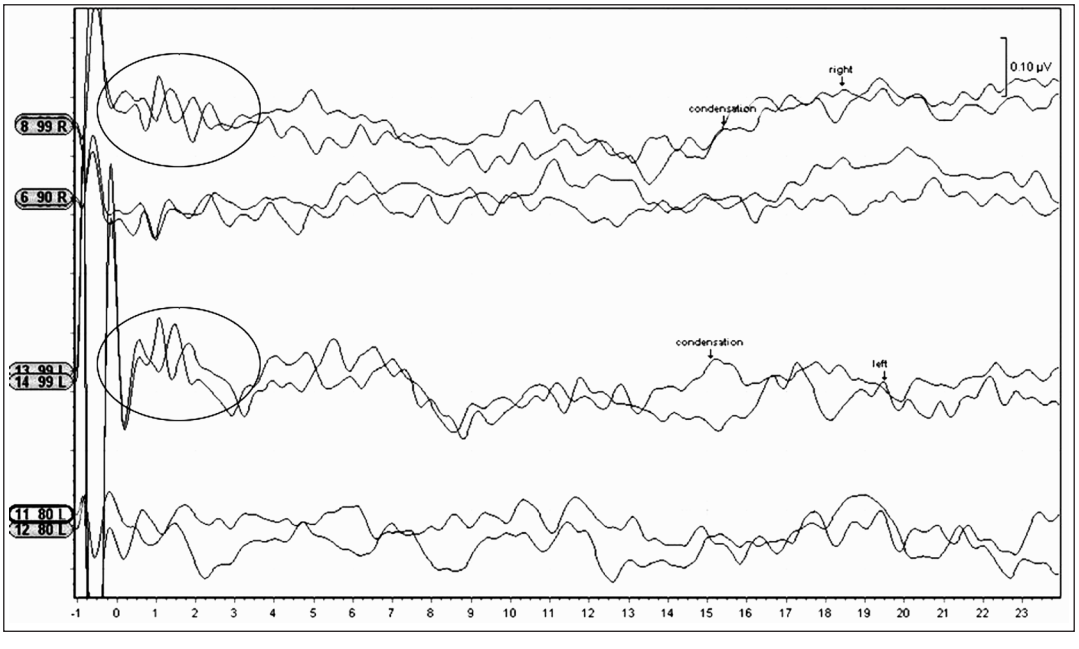

FIGURE 1: ABR test results. Cochlear microphonic is present on both sides.

TABLE 1: Findings of vHIT test.

VOR Mean Gain	Right	Left
Anterior	1.09	1.13
Lateral	0.94	0.91
Posterior	1.03	1.04

thy and no improvement in speech and language skills with hearing aids. In terms of posterior SCD, no intervention was suggested by the otolaryngologist. Maturational changes in the dehiscence would be followed up by the VEMP test.

DISCUSSION

A 22-month-old male baby with posterior SCD and auditory neuropathy has been described in this case report. The prevalence of superior SCD was 11.9%, 4.9%, 2.8%, and 0% in children <6 months, 6-11 months, 12-35 months, and 3-7 years, respectively, while the rate of posterior SCD was 16.7%, 2.4%, 1.4%, and 0% for the same age groups.[6]

Semicircular canal dehiscence may cause different audiovestibular signs, even if in cases with posterior SCD. Although most patients have Tullio's

TABLE 2: Findings of cVEMP test.

Stimulus Intensity	Ear	P1 latency (msec)	N1 latency (msec)	P1-N1 amplitude (μV)	EMG (μV)
95 dB HL	R	11.17	19.17	42.36	54
(126 dB SPL)	L	1.58	20.17	99.26	54
85 dB HL	R	11.08	17.42	25.19	55
(116 dB SPL)	L	11.50	18.67	64.21	51
75 dB HL	R	11.17	17.08	21.18	50
(106 dB SPL)	L	10.92	17.67	44.63	50
65 dB HL	R	NR	NR	NR	50
(96 dB SPL)	L	10.42	16.17	27.53	51

Right (R); Left (L); No Response (NR).

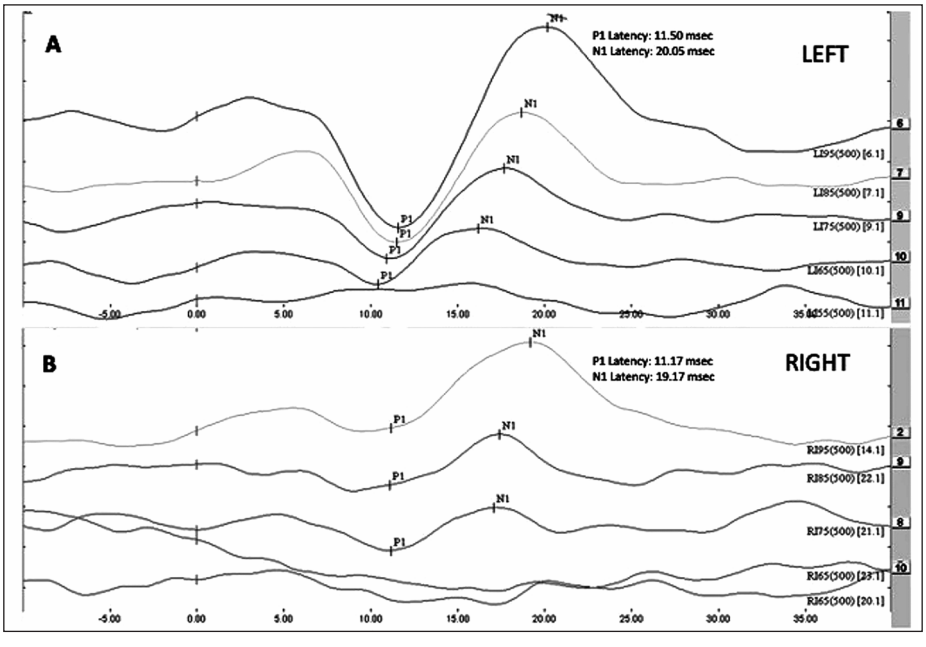

FIGURE 2: cVEMP test results. **A)** Left cVEMP. **B)** Right cVEMP. This figure shows that for each intensity level on the left side, all P1-N1 amplitudes are graphically higher than those of the right.

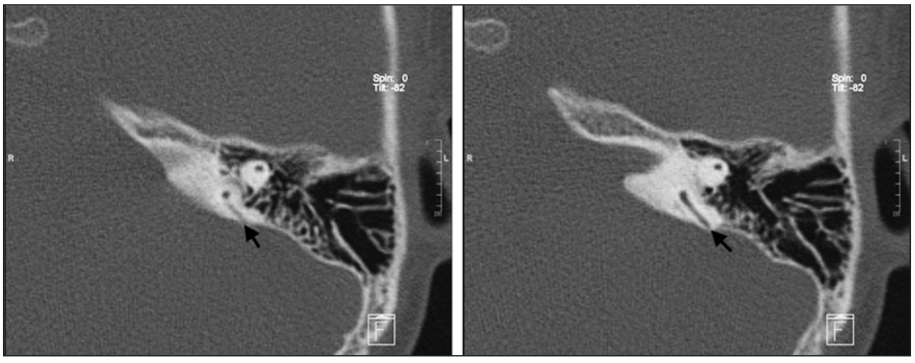

FIGURE 3: CT scans of the left temporal bone, both scans show the dehiscence of the inferior part of the posterior semicircular canal.

phenomenon, Hennebert's sign, dizziness, conductive hearing loss because of the third window effect, some patients may have sensorineural or mixed hearing loss.[7] In the present study, according to the patient's parents, the patient did not have vestibular symptoms such as vertigo and dizziness. In addition, Tullio's phenomenon and Hennebert's sign were not found by the clinician during the vestibular assessment.

A previous radiological study mentioned that the posterior canal bone is extremely thicker than other semicircular canals, and the prevalence of dehiscence decreases with increasing age because of the development of the temporal bone.[6] The development of semicircular canals begins with the occurrence of a superior semicircular canal, followed by the posterior and lateral canals. After the membranous labyrinthine has reached the adult size, ossification starts at the cochlea and semicircular canals.[1] However, the etiology of dehiscence was considered independent of the ossification process of the temporal bone because the prevalence of posterior SCD was lower than that of superior SCD despite the developmental order of SSCs.[1] By contrast, the thickness of the otic capsule in children continues until three years, and the dehiscence may result from failure of temporal bone development during the postnatal term, and head injuries or changes in intracranial pressure may induce vestibular symptoms in patients.[4]

Commonly, the presence of the air-bone gap in an audiogram is observed in superior SCD.[1,3] However, the air-bone gap was not seen in this patient because he was too small and air-bone conduction thresholds were not detected. The patient did not want to wear the insert earphones and bone conduction vibrator and was not interested in pure tone stimulus during the free-field test conducted using speakers. The patient showed some responses at nearly 65 dB HL during the speech audiometry test.

As an unexpected finding, bilateral cochlear microphonic was observed in this case, and thus bilateral neuropathy was present in this patient. Auditory neuropathy is defined as a synchronization problem associated with auditory signal transmission from the inner ear to the brain.[8] The characteristics of auditory neuropathy are known with the presence of normal outer hair cell functions (OAEs) and cochlear microphonics, despite the absence of ABR. The etiology and mechanism of auditory neuropathy have not yet been understood. *DFNB9* and *OTOF* gene mutations result in the bilateral auditory neuropathy in children.[8] In a previous study, vestibular impairment was reported in adults with polyneuropathy, and the gains of the horizontal VOR were reduced in two-thirds of patients (in 27 of 37 patients).[9] Moreover, 9 of 14 patients with auditory neuropathy had a caloric weakness, and other studies stated that cVEMP response in a patient with auditory neuropathy may be absent or abnormal.[10-12] Peripheral neuropathy may progressively affect vestibular functions and cause vestibular neuropathy.[10,13] However, there is no reported case with auditory neuropathy comorbid to SCD in the literature. The author believes that the presence of auditory neuropathy does not depend on posterior SCD, because the pathophysiology of both disorders is different.

The patient was a cochlear implant candidate, and his vestibular function might decrease after the implantation. Early diagnosis of posterior SCD, in

this patient, provided important information about his balance skills in the preoperative term. However, balance and vestibular function may be affected by some children after cochlear implantation.[14]

As a clinical procedure, vestibular assessment for each patient with hearing loss should be performed, even if they have no complaints, as hearing loss is a risk factor for vestibular hypofunction.[15] Thus, the author suggests performing early vestibular assessment in children with permanent hearing loss. In the early stages of life, knowing the additional problems associated with the vestibular function will ease the prognosis of vestibular disorder and protect the children's balance functions.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

This study is entirely author's own work and no other author contribution.

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