J Am Acad Audiol 19:204–209 (2008)

Delayed Endolymphatic Hydrops: A Case Study

DOI: 10.3766/jaaa.19.3.3

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Abstract

Delayed endolymphatic hydrops (DEH) is an unusual variation of Ménière's disease characterized by episodic vertigo that develops some time after the onset of a profound, typically unilateral sensorineural hearing loss. This case study describes a 48-year-old male who presented with complaints of episodic vertigo and disequilibrium 15 years following the onset of unilateral sensorineural hearing loss. The patient's history, audiologic findings, and vestibular evaluation led to the diagnosis of DEH. The case highlights the diagnostic and treatment challenges associated with this condition and focuses attention on principles that guide the audiologist in collecting evidence that aids in solving these challenges.

Key Words: Delayed endolymphatic hydrops, peripheral vestibulopathy, unilateral deafness

Abbreviations: DEH = delayed endolymphatic hydrops; VEMP = vestibular evoked myogenic potential; VNG = videonystagmography

Sumario

El hidrops endolinfático retardado (DEH) es una variante inusual de la Enfermedad de Ménière, caracterizada por vértigo episódico que se desarrolla en el tiempo luego del inicio de una hipoacusia sensorineural unilateral típica. Este estudio de caso describe una varón de 48 años que presentó quejas de vértigo episódico y desequilibrio, 15 años después del inicio de una hipoacusia sensorineural unilateral. La historia del paciente, los hallazgos audiológicos y la evaluación vestibular llevaron al diagnóstico de DEH. El caso destaca los retos diagnósticos y terapéuticos asociados con esta condición y concentra su atención en los principios que guían al audiólogo en la recolección de evidencia que ayude a resolver estos retos.

Palabras Clave: Hidrops endolinfático retardado, vestibulopatía periférica, sordera unilateral

Abreviaturas: DEH = hidrops endolinfático retardado; VEMP = potencial miogénico vestibular evocado; VNG = videonistagmografía

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Audiologists are well familiar with Ménière's disease, also known as idiopathic endolymphatic hydrops. Patients with the classic symptoms of unilateral fluctuating sensorineural hearing loss and tinnitus, spontaneous and episodic attacks of vertigo, and often a sensation of aural fullness are encountered in most otologic/audiologic practices. Delayed endolymphatic hydrops (DEH) is a less common variation of Ménière's and was first described by Nadol, Weiss, and Parker (1975) and Wolfson and Leiberman (1975). The condition is characterized by episodic vertigo that develops some time after the onset of severe to profound sensorineural hearing loss. Associated symptoms of ipsilateral aural fullness and increased tinnitus are common.

The purpose of this article is to describe the results of an audiologic and vestibular evaluation of a patient who presented with symptoms of DEH. The case highlights the diagnostic and treatment challenges associated with this condition and focuses attention on principles that guide the audiologist in collecting evidence to aid in solving these challenges.

CASE REPORT

A 48-year-old male was referred to Mayo Clinic Florida for an evaluation of episodic vertigo. He was a musician (drummer) and a building supervisor. Beginning 15 years earlier, he became aware of a slowly progressive hearing loss in the left ear of unknown etiology. He had several otolaryngology consults over the years that confirmed the idiopathic nature of the hearing loss. Three years prior to his visit, he experienced some motion-provoked vertigo and complained of tinnitus and hearing loss in the left ear. An audiological evaluation at that time revealed a profound sensorineural hearing loss in the left ear. His dizziness symptoms resolved, in part through vestibular rehabilitation.

Eight months prior to this evaluation, the patient began experiencing episodes of severe dizziness lasting between 10 and 15 minutes to several hours. During the episodes, left-sided tinnitus and aural fullness were prominent features, along with nausea, fatigue, and excessive sweating. The dizziness was described as "moving on a small boat over waves." His vision would jerk back and forth very fast, and his wife observed that his eyes bounced side to side. His episodes were becoming more frequent (once/week), leaving him with residual imbalance and light-headedness. These symptoms were disabling to the patient, prompting the present evaluation.

A neurologic evaluation was completed by a staff neurologist. A right Horner's syndrome was appreciated, consistent with a recent cervical sympathetic chain schwannoma resection. This was not considered related in any way to the patient's complaints of dizziness. Neurologic opinion was that the dizziness localized to the vestibular periphery. No other neurologic deficits were noted.

Otologic evaluation was unrevealing. External ears, external auditory canals, and tympanic membranes were normal bilaterally under binocular microscopic examination. Nose, mouth, oropharynx, hypopharynx, larynx, head, and neck assessments were all unremarkable.

The patient's pure-tone audiogram is shown in Figure 1. There was essentially normal hearing in the right ear with a moderate sensorineural notch at 4000 Hz, consistent with the patient's noise-exposure history. Word recognition was excellent. On the left side, a profound sensorineural hearing loss was present. Tympanometry was within normal limits. Acoustic reflexes were present in the right, with no reflex decay noted. Reflexes were absent with sound stimuli presented to the left ear.

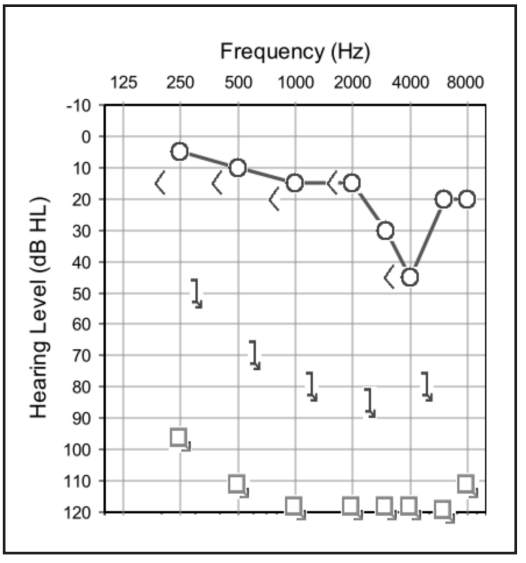

Figure 1. Pure-tone audiometry for the right and left ears.

The vestibular assessment demonstrated a persistent 3–6 degree/sec right-beating spontaneous nystagmus that suppressed with visual fixation. Bilateral, bithermal caloric responses (ICS Medical Chartr VNG) are shown in Figure 2 and demonstrated a left unilateral weakness (88%) with a strong directional preponderance to the right (86%). Rotary chair slow harmonic acceleration testing (Micromedical Spectrum Technologies, Rotational Chair Model 300, System 2000 4 channel) demonstrated low gain and a phase lead below 0.16 Hz, with a strong asymmetry to the left (Figure 3).

Neurocom computerized dynamic posturography sensory organization test results were just below age-adjusted normal limits (score = 68, normal limits >69), with difficulty on condition 5 (eyes closed, standing on an unstable platform). There was a strong tendency to carry the center of mass over the left leg.

Vestibular evoked myogenic potentials (VEMPs), as measured using the head-lifting method without muscle monitoring, were absent bilaterally. (For parameters used to record VEMP, see Figure 3 in Lundy, Zapala, and Olsholt, in this issue.) An absent VEMP on the left would suggest involvement of the inferior vestibular nerve and the saccule. VEMP presence or absence is not directly affected by the amount of sensorineural hearing loss and may be measured in deaf subjects (Halmagyi and Colebatch, 1995; Tribukait et al, 2004). The absent VEMP on the right was a solitary finding implicating right-side saccule or inferior vestibular nerve involvement. However, during a comprehensive vestibular evaluation, multiple tests are run. As a general rule, the more tests that are run, the more likely an isolated abnormal test result may occur just by chance. Consequently, without an additional line of evidence for right-sided involvement, this test was considered of equivocal significance and may reflect a false positive result.

Contrast-enhanced magnetic resonance imaging (MRI) studies showed some enhancement of the left cochlea and vestibule. This was interpreted as consistent with an inflammatory process in the left labyrinth.

In summary, these results were consistent with an acute or fluctuating, partially compensated vestibular weakness on the left side, involving structures innervated by the superior vestibular nerve branch (caloric asymmetry) and the inferior vestibular nerve branch (absent VEMP). Evidence for a partially compensated vestibulo-ocular reflex included the spontaneous nystagmus, which was present only with visual fixation removed, and the strong directional preponderance on the caloric and rotary chair tests. Evidence for a partially compensated vestibulospinal reflex included the poor performance on the sensory organization test and the tendency to carry the center of mass over the left leg (in the absence of a co-occurring orthopedic or neurologic cause). Evidence for a fluctuating vestibular deficit might be the direction-fixed positional

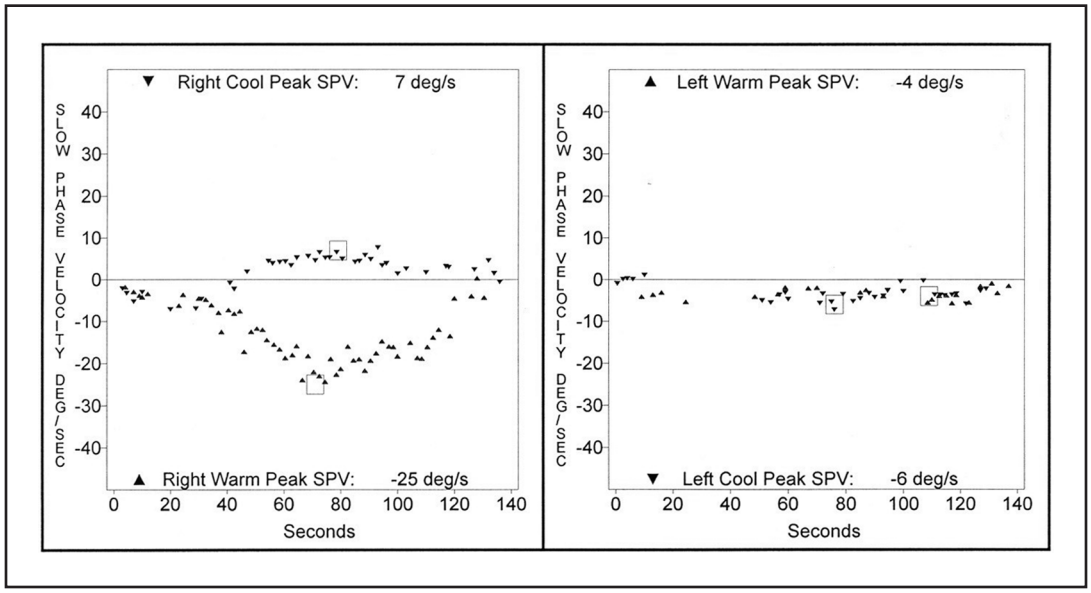

Figure 2. Bithermal water caloric results indicating an 88 percent left weakness. SPV = slow phase velocity.

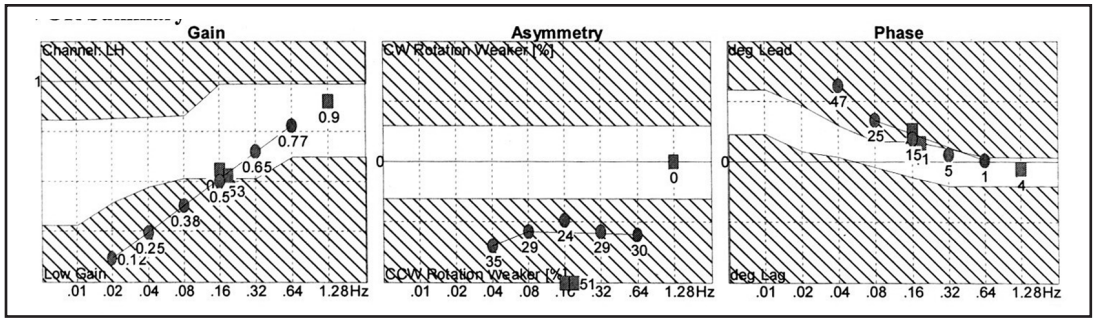

Figure 3. Vestibulo-ocular reflex testing using a rotary chair, showing significantly reduced gain (far left of graph), asymmetry to the left (middle of graph), and a phase lead (far right of graph). CW = clockwise; CCW = counterclockwise.

nystagmus that beat toward the weak ear. This suggests Bechterew's phenomenon (Shepard and Telian, 1996)—where a new injury to an ear with a previously compensated vestibular weakness produces a nystagmus that beats paradoxically toward the damaged ear. Fluctuating vestibular lesions can produce this effect.

Based on the history and the testing results, the patient was diagnosed with DEH. As a preliminary step, it was recommended that he decrease his salt intake to between 1500 and 2000 mg/day and increase his level of physical activity to improve compensation. He already had a prescription for diazepam for use during episodes. He is being followed to determine if these first-line treatments are effective.

DISCUSSION

Ipsilateral DEH can produce episodic vertigo with a time course similar to that of Ménière's disease. The vertigo can occur several years after the onset of hearing loss in that ear (Nadol et al, 1975; Huang and Lin, 2001). The cause of hearing loss varies. Head trauma (acoustic and physical), viral etiology (measles, mumps, rubella, influenza), acoustic trauma, early childhood deafness, mastoiditis, meningitis, diphtheria, and sudden idiopathic deafness are all associated with the condition (Hicks and Wright, 1988; Ylikoski, 1988; Schuknecht et al, 1990). Although the source of the hearing loss may be varied, patients with DEH all experience Ménière's-type vertigo. That is, they experience attacks of true vertigo lasting between several minutes to several hours, often associated with nausea, emesis, ipsilateral aural fullness, and increased tinnitus.

Schuknecht (1978) has proposed that DEH could be further classified into ipsilateral and contralateral forms. Ipsilateral DEH is characterized by profound unilateral or bilateral hearing loss followed by the onset of episodic vertigo of the Ménière's type. Contralateral DEH involves profound unilateral hearing loss and vestibulopathy, with the co-occurring onset of fluctuating hearing loss in the opposite ear (with or without episodic vertigo). There is little consensus about the occurrence of bilateral DEH. However, Ménière's disease evolves into bilateral involvement eventually in approximately 45 percent of patients (Schessel et al, 2004). Histopathologic studies of patients with DEH have shown severe endolymphatic hydrops involving the cochlea, saccule, and utricle, with destruction of sensory epithelium and in some cases, a blocked endolymphatic sac (Schuknecht et al, 1990).

Pollak (2004) compared audiovestibular abnormalities between patients with Ménière's disease and patients with DEH. The study determined that the prevalence of vestibular abnormalities was similar between both groups of patients, but the duration of hydropic symptoms was shorter in the DEH group. Pollak speculates that the DEH patients' symptoms were related to a primary insult that caused severe damage to endolymph resorption structures, which in turn led to a faster decompensation of their capacities following the onset of hydrops.

Young, Huang, and Cheng (2002) studied caloric and VEMP tests in 20 DEH patients, hoping to correlate the incidence of abnormal VEMP studies with the incidence of saccule atrophy found in studies demonstrating histopathologic changes in DEH patients. They found evidence for residual vestibular function in all of their cases (based on VEMP test data) and suggest

that only those ears without both VEMPs and caloric responses can become free of episodic vertigo.

In the case described here, the vestibular assessment indicated a partially compensated left peripheral vestibulopathy, involving both the inferior and superior vestibular nerves, consistent with the history of a recent vertiginous episode. The presence of profound sensorineural hearing loss in the left ear indicated involvement of all branches of cranial nerve VIII. The time course of auditory and vestibular symptoms was consistent with DEH. In this case, electrocochleography was not used as part of the diagnostic protocol but could have added additional information, especially with regard to possible bilateral DEH or progression of hydrops.

Treatment of DEH follows the treatment strategies for idiopathic endolymphatic hydrops. In most cases, the preexisting severe to profound hearing loss removes hearing preservation concerns from management consideration, simplifying decisions to some extent. Goebel (2000) suggests that three questions need to be answered before treatment for dizziness can be planned. They are as follows:

1. Is this a peripheral labyrinthine or eighth nerve disease, central vestibular disease, or diffuse disease?

2. If labyrinthine, is this unilateral or bilateral disease?

3. Is this acute isolated disease, acute recurrent disease, or chronic disease?

Once a unilateral peripheral labyrinthine focus has been established, the time course of symptoms determines the treatment strategy. Unilateral peripheral vestibular lesions that are chronic (i.e., stable) and causing symptoms are amenable to central adaptation and compensation processes. In contrast, unilateral peripheral lesions that are unstable, as in recurrent Ménière's episodes, are less likely to be amenable to central adaptation and compensation processes. Most treatments for Ménière's disease or DEH focus first on symptomatic treatment during the acute onset of each episode. However, the long-term goal is to stabilize the unstable vestibular lesion, even if this requires ablation. Only then can central adaptation and compensation processes take hold. Therefore, otologists may see little to lose in surgically destroying residual vestibular function in the DEH ear that is causing debilitating symptoms.

Accordingly, several authors advocate surgical management of DEH (Hicks and Wright, 1988; Huang and Lin, 2001). Labyrinthectomy is highly effective but the most destructive surgical option. Endolymphatic sac surgery is thought to be less debilitating than labyrinthectomy and may preserve cochlear implant candidacy in the involved ear should the other ear become severely hearing impaired in the future. Aminoglycoside ablation (such as transtympanic gentamicin administration) is probably the least traumatic and can be performed in the clinic setting (Bauer et al, 2001). All of these procedures are designed to turn an unstable vestibular end organ into a stable but weakened end organ.

In this case the most benign treatment, salt restriction, had not yet been tried. This more conservative approach was attractive because it is nondestructive. Goebel's second question bears on this treatment option. Recall that the VEMP was absent bilaterally and that it is possible for DEH to evolve into bilateral disease. Even though the MRI did not show enhancement of the normal right ear, there is a chance that the right ear might become active in the future. From this perspective, an ablative procedure might be premature, particularly if salt restriction is effective. Over time, the right ear will declare itself. A retest of the VEMP might indicate that the absent response on the right was a false positive. Monitoring changes in electrocochleography may also be helpful in detecting early hydrops. If the right ear remains uninvolved, additional medical or surgical treatment may be warranted if the patient's symptoms continue or worsen.

CONCLUSION

Delayed endolymphatic hydrops is a variation of Ménière's-type vertigo, which occurs some time following the onset of a profound sensorineural hearing loss. This article describes a case of apparent ipsilateral DEH, where the left ear has a profound sensorineural hearing loss and an incomplete, unstable left peripheral vestibulopathy. Unilateral DEH may evolve

into bilateral disease in some cases. Thus, monitoring VEMP responses and electrocochleography changes in the uninvolved ear may be useful in detecting the early beginnings of bilateral DEH. Management of this patient's symptoms involved simple, noninvasive treatment in an effort to stabilize vestibular function. With close follow-up over time, the effectiveness of conservative management, risk of bilateral involvement, and necessity of more aggressive management options will become clear.

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