Dehydration Testing and the Diagnosis of Meniere’s Disease: Case Report

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Abstract

A 34-year-old female presented to our clinic with complaints of hearing loss, tinnitus, and severe rotational vertigo. A test battery including complete audiometric testing, auditory brainstem response (ABR) audiometry, electronystagmography (ENG), electrocochleography (ECoG), and dehydration testing was performed. Monitoring of hearing sensitivity, speech understanding, and ECoG responses following administration of a dehydrating agent were sensitive measures of cochlear function and confirmed the diagnosis of Meniere’s disease. The induced changes also contributed to the therapeutic measures recommended.

Key Words: Dehydration testing, electrocochleography, (ECoG), Meniere’s disease

Meniere’s disease is an inner ear disorder first described in 1861 by Prosper Meniere (Pulec, 1984). Although its etiology remains unknown, Meniere’s disease is associated with the inability of the cochlea to regulate endolymph absorption, production, and/or circulation. Hallpike and Cairns (1938), on the basis of histologic evaluation of a temporal bone, found that as excessive endolymph accumulates in the membranous labyrinth, hydrops forms and distends Reissner’s membrane. This distortion is believed by many to cause the symptoms of Meniere’s disease.

Clinical diagnosis depends on the presence of four classic symptoms: (1) fluctuating, low-frequency sensorineural hearing loss that progresses to a flat configuration; (2) episodic rotational vertigo, with or without nausea and vomiting, that lasts several minutes or hours; (3) low-pitched or buzzing tinnitus that fluctuates with the hearing loss; and (4) aural fullness or pressure (Pulec, 1984). Symptoms may appear together or separately and in any order. This variability makes diagnosis and therapy difficult.

Patients presenting with Meniere’s symptoms undergo a variety of diagnostic procedures, because no single test has proven to be reliable diagnostically. Basic audiometric measures confirm the degree and configuration of hearing loss and rule out middle ear involvement. Auditory brainstem response (ABR) audiometry is used to exclude retrocochlear pathology. Electronystagmography (ENG) confirms the presence and localizes the site of vestibular disorder. Pressure testing (posturographic or ENG) can help identify perilymph fistula. The two procedures that contribute most information to the diagnosis of Meniere’s disease are electrocochleography (ECoG) and audiometric and electrophysiologic dehydration testing.

The following presentation describes the diagnostic protocol used at our facility to evaluate patients with suspected inner ear fluid disorder.

CLINICAL HISTORY

L M is a 34-year-old woman who had her first vertiginous episode 11 months prior to being seen at our clinic. She described that attack as a violent rotational vertigo accompanied by nausea and vomiting that lasted several hours. The episode remitted spontaneously and the following day she was quite well. She remained asymptomatic for 8 months and then
developed left-sided, low-pitched, “buzzing” tinnitus. In the next 4 months, she experienced five episodes of severe rotational vertigo, which was reliably accompanied by nausea, vomiting, diaphoresis, and pallor. The rotational component lasted up to several hours and was described as “like being on a ride at the fair.” These spells were frequently associated with a generalized increase in a feeling of left aural fullness and a marked increase in left-sided tinnitus. Sometimes, the increase in aural fullness and tinnitus were sufficient to give her advanced warning of the attacks. Her hearing fluctuated from day to day, but she felt it was generally worse around attacks of rotational vertigo. She had no dysarthria, dysphagia, visual change, numbness and tingling in the extremities, or focal motor weakness. There was no history of syncope or headache. Based on her history, LM was presumed to have otogenic vertigo. A gadolinium-enhanced MRI scan was obtained to rule out retrocochlear pathology, specifically acoustic neuroma. She was started on diuretics and a low-salt diet for presumptive diagnosis of Meniere’s syndrome. The following work-up was performed.

AUDILOGIC EVALUATION

Initial audiologic findings indicated normal hearing sensitivity for the right ear and a moderate, low-frequency loss rising to normal hearing above 2000 Hz in the left ear (Fig. 1). Immittance findings revealed normal middle ear function and acoustic reflexes at normal sensation levels. Based on patient history and audiologic results, audiometric and electrophysiologic (ECoG) dehydration studies and diagnostic audiovestibular studies were ordered.

Dehydration Protocol

Basic Audiometric Evaluation

Air-conduction pure-tone thresholds are obtained at octave and half-octave frequencies from both ears. A speech reception threshold (SRT) is obtained using Central Institute for the Deaf (CID) spondee words, and speech understanding for both ears is evaluated at 80 dB HL using a recorded version of the NU-6 word lists. Acoustic immittance measures are carried out, and if any abnormality is observed, bone-conduction pure-tone testing is performed at octave frequencies.

Electrocochleography

Extratympanic ECoG recordings are obtained in response to monaural alternating click stimulation using TIPtrode insert earphones. A horizontal recording montage is used, with the primary TIPtrode (+) at the contralateral ear and the secondary TIPtrode (-) at the ipsilateral ear, and a low forehead surface electrode serves as ground. Recording parameters are listed in Table 1. Amplitudes are measured from the prestimulus baseline to the peak of the sumating potential (SP) or action potential (AP), and an SP/AP ratio is calculated.

Dehydration Time Schedule

Baseline audiometric and ECoG recordings are obtained. A solution of 20 g of urea powder mixed in 4 oz of unsweetened fruit juice is then ingested by the patient. Serial ECoG recordings are obtained at 30-minute intervals for the next 1 ½ hours; serial audiograms are obtained at 1-hour intervals for the next 3 hours.

Table 1  ECoG Parameters

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
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<tr>
<td>Bandpass Filter</td>
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<td>Alternating</td>
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</tr>
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</table>
Results

Audiometric and electrophysiologic (ECoG) dehydration studies were performed 2 weeks following the initial audiogram. Baseline measures showed a low-frequency, rising sensorineural loss, a marked improvement in hearing sensitivity when compared to her initial test. Serial audiograms over a 3-hour period demonstrated essentially no change in threshold sensitivity; however, a 12 percent improvement in discrimination ability was observed (Fig. 2). Additionally, left ear SP/AP ratios decreased from a 0.52 preingestion level to 0.31 within the first 30 minutes post ingestion (Fig. 3).

Diagnostic studies were performed at a separate visit and included ABR audiometry, ENG, and pressure fistula testing. ABR results showed no evidence of retrocochlear pathology for either ear. Results of the ENG evaluation revealed low, left-beating positional nystagmus and a left unilateral weakness. No evidence of pressure sensitivity was observed in the fistula test.

Based on case history and test findings, the diagnosis of Meniere's disease was confirmed. LM was put on a salt-restricted diet and diuretic therapy and remained relatively asymptomatic for 4 months. She then experienced a cluster of typical vertiginous episodes, which were accompanied by nausea and tinnitus. When examined in the middle of this cluster, her audiologic reevaluation revealed a moderate, low-frequency sensorineural hearing loss for the left ear, with the right ear remaining normal. Left ear speech understanding had decreased from 92 percent to 72 percent.

To determine therapeutic benefits from diuretic therapy, dehydration testing was ordered and, once again, results were positive. Serial audiograms revealed a 10- to 15-dB threshold improvement, with speech understanding showing an improvement of 20 percent (Fig. 4). A dramatic change in the ECoG was also seen, with SP/AP ratios decreasing from 0.58 to 0.38 (Fig. 5).
LM continued rigorous salt-restriction and diuretic therapy. The vertiginous episodes remitted, and, when last seen, she had been vertigo-free for 6 weeks.

**COMMENT**

This case illustrates the effectiveness of dehydration testing in the diagnostic evaluation of a patient with suspected Meniere's disease. Audiometric and electrophysiologic testing suggest cochlear site of disorder, and positive dehydration results are consistent with the inner ear fluid imbalance associated with endolymphatic hydrops.

Incorporating ECoG into the diagnostic protocol for inner ear fluid imbalance has gained wide clinical acceptance. ECoG is a method of recording the stimulus-related potentials of the cochlea and auditory nerve. Following the presentation of a click stimulus, a repeatable, two-peaked waveform appears. The initial low peak or shoulder is identified as the summating potential, and represents the direct current (DC) activity generated within the cochlea. The second peak, identified as the action potential, represents the compound action potential generated by the auditory nerve.

Enlarged summating potential/action potential ratios have been observed in patients diagnosed with endolymphatic hydrops and Meniere's disease (Gibson et al., 1977; Coats, 1981; Ferraro et al., 1985; Staller, 1986). Increased intralabyrinthine pressure is suspected to alter cochlear mechanical properties and distorts basilar membrane vibration. This distortion causes an increase in the SP amplitude and, therefore, the SP/AP ratio. Although an increased ratio is present in the majority of patients with Meniere's disease, it is not observed in all patients diagnosed with Meniere's disease and is also found in patients diagnosed with perilymph fistula (Arenberg et al., 1988; Meyerhoff and Yellin, 1990).

The ability of a dehydrating agent to reverse the effects of endolymphatic hydrops was first demonstrated by Klockhoff and Lindblom (1966). Glycerol entering the bloodstream was thought to increase the osmolality of the blood and cause fluid to be extracted from the inner ear system. The fluid extraction would reduce endolymphatic hydrops, improve cochlear function, and result in improved hearing. In their study of patients suspected of having Meniere's disease, hearing was continuously monitored for 3 hours following ingestion of glycerol. These patients showed significant improvement in hearing sensitivity and speech understanding during the test period but a return to pretest levels within 24 hours.

Hearing improvement after dehydration using glycerol has been confirmed by other investigators and provides diagnostic and prognostic information in patients with Meniere's disease (Snyder, 1974; Thomsen and Vesterhauge, 1979; Sauer et al., 1980). A positive dehydration test is defined as a 10-dB improvement in pure-tone thresholds at two or more adjacent octave frequencies, and/or a 12 percent or greater improvement in speech understanding scores (Morrison et al., 1980; Van de Water and Arenberg, 1983). The use of urea instead of glycerol has also been successful without the debilitating side effects of nausea and vomiting (Van de Water and Arenberg, 1983; Van de Water et al., 1986).

The effects of dehydration on the ECoG response have been evaluated in several studies (Moffat et al., 1978; Coats and Alford, 1981; Dauman et al., 1988). After ingestion of a dehydrating agent, serial monitoring over a 90- to 120-minute period shows significant decreases in the SP/AP ratio in patients with Meniere's disease. This change is not observed in normal
controls (Ridenour et al., 1993). Dehydration ECoG testing, therefore, is a sensitive measure in the evaluation of patients with suspected Meniere's disease.

ECoG provides unique information that cannot be gleaned from other diagnostic measures. Although ENG testing discriminates peripheral from central site of disorder, it cannot differentiate eighth nerve from inner ear dysfunction. ABR eliminates retrocochlear concerns, but does not verify inner ear disorder. ECoG is sensitive to inner ear fluid imbalance, and incorporating it into dehydration testing successfully differentiates inner ear fluid disorders from other cochlear pathologies.

The processes responsible for audiometric and electrophysiologic changes that are induced by dehydration testing are not understood at this time. It is hypothesized that the decreased SP and improved SP/AP ratio is caused by shrinkage of the endolymphatic space induced by the dehydrating agent (Coats and Alford, 1981). This shrinkage could normalize cochlear mechanics and thereby improve responses. The changes observed in the patient presented, however, are not observed in all patients diagnosed with Meniere's disease. Dauman et al. (1988) speculate that the reason some patients do not exhibit audiometric and/or electrophysiologic shifts could be related to outer hair cell degeneration, the transient nature of the disease and the possibility of limited hydrops on the day of testing, or poor recording conditions.

The advantages of dehydration testing cannot be ignored. Such testing can easily be incorporated into a clinical test battery for the diagnosis of Meniere's disease. With the development of noninvasive ECoG recording techniques, ECoG can be adapted into evoked potential protocols. Using urea crystals rather than glycerol as the dehydrating agent minimizes the headaches and nausea experienced by patients.

Monitoring both hearing status and ECoG responses provides optimum test information, but requires a great deal of test time. Our results suggest that sufficient information may be obtained in a shorter period of time, monitoring only the ECoG response. Before this protocol is adopted, however, more information should be obtained concerning ECoG fluctuations in normal subjects. Ridenour et al. (1993) found that small but statistically significant changes can be observed in the SP/AP ratios of normal subjects. A minimum SP/AP shift for positive results should be determined before eliminating audiologic monitoring.

REFERENCES


