Assessing the Stage of Ménière’s Disease Using Vestibular Evoked Myogenic Potentials

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Background: The staging of Ménière’s disease that was proposed by the American Academy of Otolaryngology–Head and Neck Surgery in 1995 was based on the arithmetic mean of the pure-tone thresholds at 0.5, 1.0, 2.0, and 3.0 kHz using the worst audiogram during the 6-month interval before treatment. The stages were classified as follows: stage I, a 4-tone average of less than 26 dB; stage II, 26 to 40 dB; stage III, 41 to 70 dB; and stage IV, more than 70 dB.

Objective: Because the saccule, next to the cochlea, is the second most frequent site of hydrops formation, we wanted to find out whether vestibular evoked myogenic potential (VEMP) responses can reflect the stage of Ménière’s disease.

Design: Prospective study.

Setting: Tertiary university hospital.

Patients: Forty patients (23 men and 17 women; mean±SD age, 43±12 years) with unilateral definite Ménière’s disease were enrolled in the study.

Main Outcome Measures: The interaural amplitude difference (IAD) ratio, which was defined as the difference of the amplitudes of peak p13 to peak n23 (p13-n23) in the right (R) and left (L) ears divided by the sum of p13-n23 in both ears (R-L/R+L), was measured, and the stages of the disease were compared with the types of hearing loss, the percentage of unilateral weakness of caloric response, and the IAD ratio, respectively.

Results: Six ears were classified as stage I: the VEMPs were normal in 5 and augmented in 1, with a mean±SD IAD ratio of −0.02±0.20. Twelve ears were classified as stage II: the VEMPs were normal in 7, augmented in 2, depressed in 1, and absent in 2, with an IAD ratio of −0.12±0.39. Seventeen ears were classified as stage III: the VEMPs were normal in 10, depressed in 4, and absent in 3, with an IAD ratio of −0.30±0.30. Five ears were classified as stage IV: the VEMPs were normal in 2, depressed in 1, and absent in 2, with an IAD ratio of −0.54±0.43. A comparison of the IAD ratio and the stage of Ménière’s disease demonstrated a significant relationship (P<.05, 1-way analysis of variance test), whereas there was no significant relationship noted between the percentage of unilateral weakness of caloric response and the stage of disease.

Conclusion: The IAD ratio of VEMPs correlates with the stage of Ménière’s disease and can be used as another aid to assess the stage of Ménière’s disease.

Surgery in 1995. Sixteen right ears and 24 left ears were affected. The diagnostic criteria, as well as the staging of Ménière’s disease, were based on the guidelines proposed by the American Academy of Otolaryngology–Head and Neck Surgery in 1999. Sixteen right ears and 24 left ears were affected. A detailed history was obtained from each patient, and all patients underwent a physical examination and a battery of audiometric tests, including pure tone audiometry, a caloric test, and a VEMP test, after giving informed consent. The 4-tone average was calculated from 4 frequencies (0.5, 1.0, 2.0, and 3.0 kHz) on the worst audiogram during the 6-month interval before treatment. The caloric test was performed using a bithermal method with electronystagmographic recordings. The unilateral weakness represents the percentage of difference of the maximum slow-phase velocity between lesioned and healthy labyrinthine responses divided by the sum of the maximum slow-phase velocity in both ears.

The sternocleidomastoid muscle was selected as the target for recording the VEMPs by short-tone burst (95-dB hearing level, 300 Hz; rise-fall time, 1 millisecond [ms]; and plateau time, 2 ms) stimulation to 1 ear and was activated by the patient raising his or her head in a supine position during the examination. All patients were instructed to keep steady muscle contraction throughout the entire test to fit the electromyographic activation criteria (50-200 µV) of the sternocleidomastoid muscle. Electromyographic signals were amplified and bandpass filtered between 30 and 3000 Hz at a stimulation repetition rate of 5 Hz. Analysis time for each stimulus was 50 ms, and responses up to 200 stimuli were averaged for each test. Two tests were performed consecutively on the same ear to verify reproducibility. The detailed procedures were described elsewhere.

The mean latencies of peaks p13 and n23 and the amplitudes of peak p13 to peak n23 were measured. The interaural amplitude difference (IAD) ratio was defined as the difference of the amplitudes of peak p13 to peak n23 in the right (R) and left (L) ears divided by the sum of p13-n23 in both ears (R-L/R+L). In our laboratory, the mean±SD IAD ratio determined in 10 normal volunteers was 0.13±0.10. Therefore, when the IAD ratio exceeded 0.33 (mean±2 SD), the condition was labeled as either an augmented VEMP or a depressed VEMP depending on whether the amplitude of the lesioned side was greater or less than that of the opposite side (Figure). Then, the stages of the disease were compared with the types of hearing loss, the percentage of the unilateral weakness of the caloric response, and the IAD ratio of the VEMP, respectively.

Among 40 ears, the Ménière’s disease was staged as follows: stage I in 6 ears, stage II in 12 ears, stage III in 17 ears, and stage IV in 5 ears. The audiograms demonstrated low-tone hearing loss in 14 ears, high-tone hearing loss in 7 ears, peak-type hearing loss in 7 ears, and flat-type hearing loss in 12 ears (Table 1). Low- or high-tone hearing loss was found in 5 (83%) of the stage I ears and in 9 (75%) of the stage II ears. In contrast, peak- or flat-type hearing loss was found in 10 (62%) of the stage III ears and in 5 (100%) of the stage IV ears, indicating a significant relationship between the types of hearing loss and the stage of Ménière’s disease (P<.05, Kruskal-Wallis test).

In our laboratory, the normal limit of the caloric response is within 25% of the unilateral weakness. Therefore, 29 ears (72%) revealed a normal caloric response and 11 ears (28%) demonstrated a reduced caloric response. The prevalence of normal caloric response in stages I to IV was 100%, 67%, 76%, and 40%, respectively (Table 2). The mean±SD percentage of unilateral weakness in stage I was −14%±40%, compared with −15%±64% in stage II, 7%±27% in stage III, and 41%±30% in stage IV, showing a nonsignificant relationship with the stage of the disease (P=.37, 1-way analysis of variance test).

Testing revealed that the VEMPs were normal in 24 ears (60%), augmented in 3 ears, depressed in 6 ears, and
absent in 7 ears (18%) (Table 3). Excluding the absent VEMPs, the mean ± SD latencies of p13 and n23 in 33 ears with positive VEMPs were 15.2 ± 2.5 ms and 21.6 ± 2.5 ms, respectively, compared with those of the contralateral healthy ears, which were 15.4 ± 2.7 ms and 22.0 ± 2.8 ms, respectively, demonstrating a nonsignificant difference (p13, P = .76; n23, P = .54, paired t test).

Of the 6 stage I ears, the VEMPs were normal in 5 and augmented in 1 (IAD ratio, −0.02 ± 0.20); of the 12 stage II ears, the VEMPs were normal in 7, augmented in 2, depressed in 1, and absent in 2 (IAD ratio, −0.12 ± 0.39); of the 17 stage III ears, the VEMPs were normal in 10, depressed in 4, and absent in 3 (IAD ratio, −0.30 ± 0.30); and of the stage IV ears, the VEMPs were normal in 2, depressed in 1, and absent in 2 (IAD ratio, −0.54 ± 0.43). Comparing the IAD ratio in each stage of Ménière’s disease showed a significant difference (P < .05, 1-way analysis of variance test).

### COMMENT

The VEMPs were positive in 33 cases (82%) of Ménière’s disease in the present study. Variations between in the incidence of positive VEMPs in our study and others, such as the 54% positive rate reported by deWaele et al and the 65% positive rate reported by Murofushi et al, can be explained by the different stages of Ménière’s disease in the study patients. However, the ears of all patients with Ménière’s disease and positive VEMPs demonstrated normal latencies of p13 and n23. Therefore, before the VEMP results in patients with Ménière’s disease are evaluated, it is important to stage the disease.

Previously, the staging system for Ménière’s disease was based on either the type of hearing loss or the 4-tone average. Low-tone hearing loss develops during the early stage of Ménière’s disease, and as the disease progresses the higher frequencies are affected. Paparella et al proposed that a peak-type hearing loss occurs after some time has elapsed. However, Friburg et al suggested that flat-type hearing loss is the most common type after 15 years of follow-up. In the present study, we also found that types of hearing loss correlated with the stage of Ménière’s disease (Table 1). Therefore, hearing is the variable that is most readily measured, as well as the variable that is most related to the natural course of Ménière’s disease.

In addition to hearing loss, a reduction in caloric response is observed in patients with Ménière’s disease. Rizvi found that in the late stages of Ménière’s disease, there might be severe dilatation or collapse of the ampullary walls, which would interfere with the cupular movement, resulting in a poor caloric response. Meanwhile, the vestibular sense organs may also undergo severe atrophic changes. After repeated ruptures of the labyrinthine membrane, partial or total collapse of the membranous labyrinth occurs, a condition that Merchant and Schuknecht termed vestibular atelec-tasis. These histopathologic findings suggest why spontaneous relief of vertigo occurs in the late stage of Ménière’s disease. Nevertheless, hydrops develops less often in the semicircular canal. Stahl and Bregman found that the caloric response was reduced in 65% of 300 patients with Ménière’s disease. They also observed that the number of patients with reduced caloric excitibility did not increase with increased duration of the disease. Similarly, our results also indicate that the percentage of unilateral weakness fails to correlate with the stage of Ménière’s disease (Table 2). In contrast, the percentage of unilateral weakness was negative in 1 patient with stage I disease and in 3 patients with stage II disease, implying an “irritative” state and is usually indicative of a functional disturbance in the early stages of Ménière’s disease.

In addition to hearing and caloric tests, the recently developed VEMP test can be used to evaluate inner ear function other than cochlear or ampullary. It gives clinicians another tool that they can use to assess saccular abnormalities. Because the saccule is the second most frequent site of hydrops formation, and because severe hydrops is observed most frequently in the saccule, the normal, augmented, depressed, or absent VEMPs demonstrated in our study may reflect the pathologic findings in the saccule.

The VEMPs were normal in 5 (83%) of the stage I ears, indicating that the sacculeolocic reflex retains normal velocity conduction in the earliest stage of Ménière’s disease. Because VEMP amplitude has been correlated to the intensity of acoustic stimulation, augmented VEMPs can be explained as dilatation of the saccular hydrops extending to press against the footplate, as this action enhances the sensitivity of the saccular macula to loud sound. In contrast, the Tullio phenomenon has been associated with an abnormally low threshold for click-evoked vestibulococ-locic response as well as with dehiscence of the superior semicircular canal. Kwes suggested that sound stimulates the vestibular neuroepithelium by means of a dilated saccular wall that is in contact with the footplate. However, our 3 patients with augmented VEMPs presented neither a positive Tullio phenomenon nor a positive Hennebert sign, pos-

### Table 2. Staging of Ménière’s Disease vs Caloric Response*

<table>
<thead>
<tr>
<th>Unilateral Weakness, %</th>
<th>Stage I</th>
<th>Stage II</th>
<th>Stage III</th>
<th>Stage IV</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤25</td>
<td>6</td>
<td>8</td>
<td>13</td>
<td>2</td>
</tr>
<tr>
<td>26-50</td>
<td>0</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>51-75</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>&gt;75</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Mean ± SD</td>
<td>−14 ± 40</td>
<td>−15 ± 64</td>
<td>7 ± 27</td>
<td>41 ± 30</td>
</tr>
</tbody>
</table>

*The study showed a nonsignificant relationship between the unilateral weakness and the stage of Ménière’s disease (P = .37, 1-way analysis of variance test).

### Table 3. Staging of Ménière’s Disease vs Vestibular Evoked Myogenic Potential (VEMP) Results*

<table>
<thead>
<tr>
<th>VEMP Results</th>
<th>Stage I</th>
<th>Stage II</th>
<th>Stage III</th>
<th>Stage IV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>5</td>
<td>7</td>
<td>10</td>
<td>2</td>
</tr>
<tr>
<td>Augmented</td>
<td>1</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Depressed</td>
<td>0</td>
<td>1</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Absent</td>
<td>0</td>
<td>2</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>IAD ratio, mean ± SD</td>
<td>−0.02 ± 0.20</td>
<td>−0.12 ± 0.39</td>
<td>−0.30 ± 0.30</td>
<td>−0.54 ± 0.43</td>
</tr>
</tbody>
</table>

*The study showed a significant relationship between the interaural amplitude difference (IAD) ratio and the stage of Ménière’s disease (P < .05, 1-way analysis of variance test).
A dilated saccule with an atrophied saccular macula, which was described in one histopathologic study of Ménière’s disease, could be an explanation for depressed VEMPs. Absent VEMPs in stage IV disease could result from the saccular wall collapsing onto the otolithic membrane, as observed in a temporal bone histopathologic study by Fraysse et al, who reported that the saccular membrane was collapsed on the sensory epithelium in a 75-year-old woman with Ménière’s disease. However, further temporal bone studies in patients with abnormal VEMPs are required for confirmation of these hypotheses.

In the earliest stage (stage I) of Ménière’s disease, fluctuating hearing loss or augmented VEMPs may be attributed to a mechanical, biochemical, or some other reversible cause. However, in the latest stage (stage IV), there are permanent morphological changes in the sense organs, including loss of hair cells accompanied by collapse of the Reissner membrane onto the organ of Corti, resulting in a flat-type hearing loss with a mean hearing level of more than 70 dB. Furthermore, loss of saccular macula associated with collapse of the saccular wall onto the otolithic membrane is thought to be responsible for a depressed-type VEMP, with an IAD ratio of up to −1. Okuno and Sando suggested that the severity of hydrops correlates to the severity of hearing loss. In the present study, the IAD ratio of the VEMPs increased significantly according to the stage of Ménière’s disease (P = .04) (Table 3). Therefore, the IAD ratio of VEMPs, like the 4-tone average of hearing, shows promise in facilitating the staging of Ménière’s disease. In other words, besides the hearing test, the VEMP test provides another aid for evaluating the stage of Ménière’s disease.

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