Assessment of Saccular Function in Children With Sensorineural Hearing Loss

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Objective: To investigate saccular function using vestibular evoked myogenic potentials in children with congenital or early acquired sensorineural hearing loss.

Design: Retrospective cohort study.

Setting: Pediatric tertiary referral center.

Patients: Twenty-three children with bilateral sensorineural hearing loss (severe to profound in 22 cases, moderate in 1 case) underwent evaluation of saccular function. Twelve pediatric subjects with normal hearing were included in the study as the control group.

Interventions: Otologic examination, computed tomography of the temporal bone, audiometry, tympanometry, and vestibular evoked myogenic potential testing.

Main Outcome Measure: Differences in threshold, amplitude, and P1 and N1 latencies of vestibular evoked myogenic potentials between children with normal-hearing and hearing-impaired children.

Results: Abnormal vestibular evoked myogenic potentials were found in 21 of 23 children (91%) with sensorineural hearing loss. The thresholds of vestibular evoked myogenic potential were significantly higher (P < .001) and the amplitudes were lower in children with sensorineural hearing loss than those in children with normal hearing. There were no differences in the P1 and N1 latencies between the 2 groups.

Conclusions: The impairment of saccular function, indicated by the abnormal findings in the vestibular evoked myogenic potential, is often associated with sensorineural hearing loss in the pediatric population. Although saccular dysfunction may create a vestibular deficit, its manifestations can vary and be easily overlooked in children. Considering the high percentage of abnormal findings in our study, we recommend that deaf and hard-of-hearing children undergo vestibular evaluation. Vestibular evoked myogenic potential testing is an easy and reliable procedure to evaluate saccular function in children.


The cochlea and the vestibule are the peripheral sensory organs of the auditory and vestibular system, respectively. They are anatomically and phylogenetically related. Disturbances of cochlear function, which can result in sensorineural hearing loss (SNHL), could accompany vestibular impairment because the cochlea and the vestibule share the continuous membranous labyrinth of the inner ear. On the one hand, owing to the success of newborn hearing screening and early intervention programs, congenital and early-onset SNHL in children is usually well managed. On the other hand, vestibular function deficits in hearing-impaired children are often overlooked and not thoroughly investigated, in contrast to the adult population. Early detection of peripheral vestibular dysfunction in the pediatric population not only can help clinicians and parents understand why children experience balance disturbances but also facilitate children’s learning of compensation strategies for balance control. There are a variety of reasons why vestibular evaluation is not routinely performed in the pediatric population. One of those is the lack of feasible and effective procedure(s) for clinical use. The standard procedures for vestibular evaluation in adults, such as electronystagmography (ENG) and calorictest, are challenging, if not impossible, when attempted in young children. In recent years, there has been a growing awareness of vestibular dysfunction in deaf children. Efforts to create child-friendly vestibular evaluation procedures have yielded important progress. One of these procedures is vestibular evoked myogenic potential (VEMP) testing. The VEMP test measures a vestibulo-spinal reflex mediated through the saccule and the inferior vestibular nerve, in which a loud auditory stimulus induces an ipsilateral inhibition of the tonic neck muscle activity recorded on...
em electromyography (EMG). Clinically, the VEMP test is used to evaluate the function of the saccule and inferior vestibular nerve and aid in the diagnosis of vestibular disorders. It is frequently used in adults, and many studies have demonstrated it to be reliable and effective for diagnosis of vestibular disorders such as Ménière’s disease, vestibular schwannoma, and superior semicircular canal dehiscence. Although the VEMP test is noninvasive, time saving, and well tolerated in children, limited VEMP studies have been done in pediatric populations, especially in hearing-impaired children.

This study assessed saccular function using the VEMP test in children with SNHL. We investigated the differences in VEMP measurements (threshold, latency, and amplitude) between children with normal hearing and hearing-impaired children. This study also evaluated the occurrence of saccular dysfunction in a cohort of hearing-impaired children, allowing proper counseling and recommendation for their parents.

METHODS

This retrospective study was approved by the institutional review board of the Children’s Hospital Boston, Boston, Massachusetts.

NORMAL SUBJECTS

Twelve children with normal hearing, aged 4 to 18 years, including 10 girls and 2 boys, were recruited to participate in this study. These children had no history of vestibular or neurological disorders. Normal hearing and middle ear function were verified by audiological evaluation.

PATIENTS

Twenty-three hearing-impaired children were included in this study. They ranged in age from 2 to 16 years and included 12 boys and 11 girls. All of them had bilateral SNHL, either congenital or early-onset. Detailed medical information is demonstrated in Table 1. All patients underwent imaging studies (computed tomography) to rule out any inner ear structural anomalies.

AUDIologic evaluation

Patients’ hearing losses were documented by behavioral (pure-tone) audiometry, which included air and bone conduction thresholds. Electrophysiological testing, such as auditory brainstem responses (ABRs) and otoacoustic emissions were also used when deemed necessary to establish or confirm the hearing loss. Tympanometry was performed to confirm normal middle ear pressure and mobility before VEMP testing. Audiometry and tympanometry were performed using a GSI 61 Audiometer and GSI TymppStar middle ear analyzer (Grason-Stadler, a division ofViaSys Healthcare Inc, Madison, Wisconsin), respectively. Both ABR and VEMP tests were performed using the NavigatorPro evoked potential system made by Bio-logic (Mundelein, Illinois).

The VEMP test was performed in a sitting position. Older children could sit upright by themselves while being tested, and younger children (eg, <4 years) would sit in their parents’ lap. The setup of the VEMP test was described in a previously published study. Briefly, surface (silver–silver chloride) electrodes were placed on the sternocleidomastoid (SCM) muscle, with the noninverting electrode on the upper third of the muscle belly and the inverting electrode on the muscle tendon just above the clavicle. A ground electrode was placed on the forehead. Ongoing EMG activity of the SCM muscle was visually monitored on an oscilloscope to ensure that sufficient muscle contraction (eg, EMG level of 60-100 µV) occurred during the acquisition.

The VEMP responses were elicited using clicks and 500-Hz tone bursts (with Blackman gating, and 2-cycle rise/fall and no plateau). Stimuli were presented monaurally to patients at a rate of 13/s via ER-3A insert phones (Omus Research Inc, Elk Grove Village, Illinois). Electromyographic signals were amplified (×2000), bandpass filtered (30-1500 Hz), and averaged (100-200 sweeps). To obtain VEMP thresholds, a stimulus level of 90 dB nHL (normal hearing level) was used as the default starting intensity. The stimulus intensity would decrease in steps of 10 dB or increase in steps of 5 dB depending on the presence or absence of VEMP, respectively. The lowest stimulus intensity at which a clear and repeatable biphasic (P1 and N1) wave was observed would be recorded as the VEMP threshold. If no reliable response was found, the VEMP would be considered as absent, and the threshold was recorded as 10 dB higher than the maximal intensity of the stimulus used for the purpose of statistical analysis. The VEMP amplitude and P1 and N1 latencies were measured at stimuli level of 90 dB nHL.

STATISTICAL ANALYSIS

The means (SDs) of VEMP thresholds, amplitudes, and P1 and N1 latencies were calculated for each group (children with normal hearing and hearing-impaired children). The VEMP thresholds, amplitudes, and P1 and N1 latencies were compared between groups using the Mann-Whitney U test. The two-tailed p value was used to determine statistical significance, with p < .05 considered significant. A Bonferroni correction was applied to control for multiple comparisons. The post hoc analysis was performed using the Kruskal-Wallis test.

Table 1. Demographic Information for 23 Children With Sensorineural Hearing Loss (SNHL)

<table>
<thead>
<tr>
<th>Patient No./Sex/</th>
<th>Age, y</th>
<th>Degree of SNHL</th>
<th>Etiology of SNHL</th>
<th>Habilitation and Communication</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M/5</td>
<td>Profound</td>
<td>GJB2 (connexin 26)</td>
<td>Binaural, CI</td>
<td></td>
</tr>
<tr>
<td>2/M/4</td>
<td>Profound</td>
<td>Meningitis</td>
<td>Monaural, CI</td>
<td></td>
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<tr>
<td>3/F/6</td>
<td>Profound</td>
<td>AN/AAD</td>
<td>Monaural, CI</td>
<td></td>
</tr>
<tr>
<td>4/F/12</td>
<td>Severe-profound</td>
<td>Unknown</td>
<td>Monaural, CI/HA</td>
<td></td>
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<tr>
<td>5/F/14</td>
<td>Severe-profound</td>
<td>Unknown</td>
<td>Binaural, CI</td>
<td></td>
</tr>
<tr>
<td>6/F/8</td>
<td>Profound</td>
<td>GJB2 (connexin 26)</td>
<td>Binaural, CI</td>
<td></td>
</tr>
<tr>
<td>7/F/2</td>
<td>Profound</td>
<td>Cogan syndrome</td>
<td>Binaural, CI</td>
<td></td>
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<tr>
<td>8/M/11</td>
<td>Profound</td>
<td>GJB2 (connexin 26)</td>
<td>Binaural, CI</td>
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<tr>
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<td>Unknown</td>
<td>Monaural, CI/HA</td>
<td></td>
</tr>
<tr>
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<td>Binaural, CI</td>
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<td>12/F/17</td>
<td>Profound</td>
<td>CMV</td>
<td>Monaural, CI</td>
<td></td>
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<tr>
<td>13/F/7</td>
<td>Profound</td>
<td>GJB2 (connexin 26)</td>
<td>Binaural, CI</td>
<td></td>
</tr>
<tr>
<td>14/F/6</td>
<td>Profound</td>
<td>CMV</td>
<td>Binaural, CI</td>
<td></td>
</tr>
<tr>
<td>15/F/5</td>
<td>Profound</td>
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<td>Binaural, CI</td>
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<tr>
<td>16/F/8</td>
<td>Severe-profound</td>
<td>Unknown</td>
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<td>Monaural, CI</td>
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<td>18/F/16</td>
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<tr>
<td>19/M/6</td>
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<td>GJB2 (connexin 26)</td>
<td>Binaural, CI</td>
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<td>20/M/4</td>
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<td>ASL</td>
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<td>Moderate</td>
<td>GJB2 (connexin 26)</td>
<td>Binaural, HA</td>
<td></td>
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</tbody>
</table>

Abbreviations: AN/AAD, auditory neuropathy/dysynchrony; ASL, American Sign Language; CI, cochlear implant; CMV, cytomegalovirus infection; HA, hearing aid. a GJB2 (connexin 26) mutations.
mal hearing and hearing-impaired children). All VEMP measurements underwent further statistical analysis (ie, Mann-Whitney test and analysis of variance), using SPSS statistical software (version 14.0; SPSS Inc, Chicago, Illinois).

RESULTS

ETIOLOGIES OF HEARING LOSS

Although most of the hearing-impaired children in our study had severe to profound SNHL, the etiology of the hearing loss varied. We were able to identify the causes of hearing loss in 13 patients. Among them, 7 had biallelic GJB2 (connexin 26) mutations, 3 had congenital cytomegalovirus infection, 1 had bacterial meningitis, 1 had Cogan syndrome, and 1 had auditory neuropathy and dysynchrony. The causes of hearing loss in the other 10 children remained unclear. All patients currently either wear hearing aid(s) or have cochlear implant(s), except 1 who uses American Sign Language as the primary communication mode.

VEMP OUTCOMES

Table 2 reports the means (SDs) for VEMP measurements, including thresholds, amplitude, and P1 and N1 latencies in children with normal hearing and children with SNHL. In children with SNHL and cochlear implant, the VEMP measurements were obtained in non-implanted ears and/or before the implantation. Of the total 23 hearing-impaired children, 21 were found to have abnormal VEMP findings. Specifically, VEMP thresholds were significantly higher (>10 dB, 2 SDs above normal) (P < .001) and amplitudes were lower (<10 µV), or the VEMP responses were absent altogether. However, the P1 and N1 latencies were similar in children with normal hearing and hearing-impaired children when VEMPs were present. Comparisons of each VEMP measurement showed that the differences in VEMP thresholds (P < .001) and amplitudes (P < .01) between the 2 groups were statistically significant, but the differences in P1 and N1 latencies were not (P > .05).

In the Figure, the graph on the left side shows typical VEMP responses from a child with normal hearing, of hearing loss in 13 patients. Among them, 7 had biallelic GJB2 (connexin 26) mutations, 3 had congenital cytomegalovirus infection, 1 had bacterial meningitis, 1 had Cogan syndrome, and 1 had auditory neuropathy and dysynchrony. The causes of hearing loss in the other 10 children remained unclear. All patients currently either wear hearing aid(s) or have cochlear implant(s), except 1 who uses American Sign Language as the primary communication mode.

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In the Figure, the graph on the left side shows typical VEMP responses from a child with normal hearing,
and the graph on the right shows VEMP responses from a patient with profound hearing loss. Considerable differences are noted in VEMP threshold and amplitude, whereas P1 and N1 latencies are similar between subjects with normal hearing and hearing-impaired children. These findings suggested peripheral vestibular deficit (ie, saccular impairment), whereas the inferior vestibular nerve was not compromised in our hearing-impaired subjects.

**COMMENT**

Current opinion on vestibular function in children states that routine and thorough examination of the vestibular system in hearing-impaired children is necessary.\(^1\)\(^2\)\(^3\)\(^4\)\(^14\) Several studies\(^2\)\(^4\)\(^14\) have demonstrated vestibular deficits in deaf children, although there is a wide range of incidence. Our study of saccular function by VEMP testing showed a high percentage of abnormality in hearing-impaired children. This finding may be attributed to the etiology of the hearing loss in our patients because each patient’s hearing loss was either congenital or early-onset, increasing the likelihood of the whole inner ear being affected. Unlike a previously published study,\(^3\) we did not find a clear relationship between the degree of hearing loss and the severity of saccular dysfunction.

Based on our clinical experience, the VEMP test proved to be a feasible and relatively easy method to conduct vestibular evaluation in children. The test usually takes only 15 to 30 minutes and is well tolerated by children at any age group. Low-frequency tone bursts, such as 500 Hz, seem to be better stimuli than clicks because they produce more robust VEMP responses. Moreover, less intensity of stimulus is needed for tone bursts than clicks to elicit clear VEMP responses, thus minimizing the exposure of the subjects to an unpleasant loud sound. We found that VEMP threshold and amplitude were the 2 measurements that were most sensitive to detect saccular dysfunction, although the variability of VEMP amplitude was relatively high primarily owing to variations of the ongoing EMG level. In contrast, the P1 and N1 latencies were not significantly different (P > .05) between children with normal hearing and hearing-impaired children. However, for both groups, the P1 and N1 latencies increased with age; no sex difference was noted in either group.

It remains unclear why many hearing-impaired children with abnormal VEMP outcomes do not have complaints of vestibular symptoms. Possible explanations include the following: (1) young children are not able to describe dizziness or vertigo to their parents and physicians, (2) saccular impairment alone is not enough to cause clinically significant vestibular disturbance, (3) chronic peripheral vestibular deficit may generate central compensation, and (4) less attention is paid to subtle manifestations of vestibular dysfunction by caregivers. Future studies with more comprehensive investigation, including a standardized balance questionnaire for the children and their parents or caregivers and other vestibular evaluation procedures, are needed to address these issues.

Our study found that most children with SNHL had reduced saccular function, consistent with other studies published previously. Although most of the past studies on VEMP in children reported only the presence or absence of VEMP responses or amplitude differences,\(^3\)\(^4\)\(^14\) our study adopted a more comprehensive approach, measuring all parameters of VEMP response (ie, threshold, amplitude, and P1 and N1 latencies). By doing so, we believe it increases the sensitivity of VEMP testing to detect minor changes of saccular function.

Although most hearing-impaired children may not be disabled by the saccular deficit, awareness of the status of vestibular function in this population is very important. A considerable portion of the hearing-impaired children may be considered as cochlear implant candidates, as in our study. In the process of cochlear implantation, the electrode wire inserted into the inner ear can produce permanent damage to the sensory structures of the cochlea and the vestibule.\(^16\)\(^19\) Considering that more children may have bilateral cochlear implants in the near future, potential vestibular dysfunction after the implant surgery is a cause for concern and needs to be addressed before and after surgery. Proper counseling and recommendations regarding this issue are imperative. Vestibular evaluation in hearing-impaired children is not only necessary and possible but also beneficial for clinical treatment. Correlation of vestibular function with the etiology of the hearing loss will improve counseling for all children with hearing loss and will better define many of these etiologies as well, including improved phenotype-genotype descriptions for those with genetic hearing loss. Further investigations into this exciting area of vestibular function are needed to help clarify these issues.

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**Author Contributions:** All of the authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. **Study concept and design:** Zhou, Kenna, Stevens, and Licameli. **Acquisition of data:** Zhou, Kenna, Stevens, and Licameli. **Analysis and interpretation of data:** Zhou, Kenna, and Licameli. **Drafting of the manuscript:** Zhou, Kenna, Stevens, and Licameli. **Critical revision of the manuscript for important intellectual content:** Kenna, Stevens, and Licameli. **Statistical analysis:** Zhou and Licameli. **Obtained funding:** Stevens. **Administrative, technical, and material support:** Stevens and Licameli. **Study supervision:** Kenna.

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REFERENCES