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.


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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 caloric test, 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
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 TymStar middle ear analyzer (Graison-Stadler, a division of ViaSys 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 (e.g., <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. To activate the SCM muscle during the recording of the VEMP, the patient was asked to turn his or her head toward the contralateral side of the ear being tested. For younger children, an assistant was used to attract the patients’ attention, allowing them to make a head turn. Ongoing EMG activity of the SCM muscle was visually monitored on an oscilloscope to ensure that sufficient muscle contraction (e.g., 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 (Osmotic Research Inc, Elk Grove Village, Illinois). Electromyographic signals were amplified (∗2000), bandpass filtered (30-1300 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. The statistical analysis was performed using the software program the use of the post-test analysis. The VEMP amplitude and P1 and N1 latencies were measured at stimulus 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 statistical analysis was performed using a t-test for each group.
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 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.

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,
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.

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, 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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