



Clinical Manifestations of Superior Semicircular Canal Dehiscence

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Objectives/Hypotheses: To determine the symptoms, signs, and findings on diagnostic tests in patients with clinical manifestations of superior canal dehiscence. To investigate hypotheses about the effects of superior canal dehiscence. To analyze the outcomes in patients who underwent surgical repair of the dehiscence. **Study Design:** Review and analysis of clinical data obtained as a part of the diagnosis and treatment of patients with superior canal dehiscence at a tertiary care referral center. **Methods:** Clinical manifestations of superior semicircular canal dehiscence were studied in patients identified with this abnormality over the time period of May 1995 to July 2004. Criteria for inclusion in this series were identification of the dehiscence of bone overlying the superior canal confirmed with a high-resolution temporal bone computed tomography and the presence of at least one sign on physiologic testing indicative of superior canal dehiscence. There were 65 patients who qualified for inclusion in this study on the basis of these criteria. Vestibular manifestations were present in 60 and exclusively auditory manifestations without vestibular symptoms or signs were noted in 5 patients. **Results:** For the 60 patients with vestibular manifestations, symptoms induced by loud sounds were noted in 54 patients and pressure-induced symptoms (coughing, sneezing, straining) were present in 44. An air-bone on audiometry in these patients with vestibular manifestations measured (mean \pm SD) 19 \pm 14 dB at 250 Hz; 15 \pm 11 dB at 500 Hz; 11 \pm 9 dB at 1,000 Hz; and 4 \pm 6 dB at 2,000 Hz. An air-bone gap 10 dB or greater was present in 70% of ears with superior canal dehiscence tested at 250 Hz, 68% at 500 Hz, 64% at 1,000 Hz, and 21% at 2,000 Hz.

Similar audiometric findings were noted in the five patients with exclusively auditory manifestations of dehiscence. The threshold for eliciting vestibular-evoked myogenic potentials from affected ears was (mean \pm SD) 81 \pm 9 dB normal hearing level. The threshold for unaffected ears was 99 \pm 7 dB, and the threshold for control ears was 98 \pm 4 dB. The thresholds in the affected ear were significantly different from both the unaffected ear and normal control thresholds ($P < .001$ for both comparisons). There was no difference between thresholds in the unaffected ear and normal control ($P = .2$). There were 20 patients who were debilitated by their symptoms and underwent surgical repair of superior canal dehiscence through a middle cranial fossa approach. Canal plugging was performed in 9 and resurfacing of the canal without plugging of the lumen in 11 patients. Complete resolution of vestibular symptoms and signs was achieved in 8 of the 9 patients after canal plugging and in 7 of the 11 patients after resurfacing. **Conclusions:** Superior canal dehiscence causes vestibular and auditory symptoms and signs as a consequence of the third mobile window in the inner ear created by the dehiscence. Surgical repair of the dehiscence can achieve control of the symptoms and signs. Canal plugging achieves long-term control more often than does resurfacing. **Key Words:** Vertigo, superior semicircular canal dehiscence syndrome, labyrinth, oscillopsia, autophony.

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INTRODUCTION

A syndrome of vertigo and oscillopsia induced by loud sounds or by stimuli that change middle ear or intracranial pressure has recently been identified.^{1–4} Patients with this syndrome can have a Tullio phenomenon (eye movements induced by loud noises) or a Hennebert sign (eye movements induced by pressure in the external auditory canal). Valsalva maneuvers can also elicit vestibular symptoms and signs in patients with superior canal dehiscence. The eye movements evoked by these stimuli typically align with the plane of the dehiscent superior semicircular canal. Confirmation that the bone overlying the superior semicircular canal is dehiscent in these patients has been provided by findings at the time of surgical

repair of the abnormality.^{1,2,5,6} Symptoms and signs resolve, in most cases, after surgical correction.

The dehiscence of bone overlying the superior canal in patients with this syndrome has been identified with high-resolution computed tomography (CT) scans of the temporal bones.^{1,2,7-9} Conventional temporal bone CT scans that are performed with 1.0 mm collimation with images displayed in the axial and coronal planes have relatively low specificity (high number of false-positives) in the identification of superior canal dehiscence because of the effects of partial volume averaging. The specificity and positive predictive value of these scans is improved when 0.5 mm collimated helical CT scans are performed with reformation of the images in the plane of the superior canal.⁷

Vestibular-evoked myogenic potentials (VEMP responses) have been shown to be useful in the evaluation of patients suspected of having symptoms and signs caused by superior canal dehiscence.⁸⁻¹⁰ Patients with superior canal dehiscence syndrome have a lower than normal threshold for eliciting the VEMP response in the affected ear. VEMP responses are short-latency relaxation potentials measured from tonically contracting sternocleidomastoid muscles that relax in response to ipsilateral presentation of loud clicks.^{11,12} These responses are thought to be of vestibular origin because they disappear after vestibular neurectomy and are still present in deaf patients with preserved vestibular function.^{11,13,14} The inferior vestibular nerve has been implicated in the responses because all patients who developed posterior canal benign paroxysmal positional vertigo (BPPV) after vestibular neuritis were noted to have intact VEMP responses, whereas these responses were absent in most patients after vestibular neuritis who did not develop BPPV.¹⁵ Single-unit recordings from vestibular-nerve afferents in the guinea pig support the hypothesis that VEMP responses originate from the sacculus.^{16,17}

Auditory manifestations of superior semicircular canal dehiscence have also been described. The Weber tuning fork test (512 Hz) typically lateralizes to the affected ear. Patients can experience symptoms such as hearing their pulse or hearing their eye movements.^{2,9} An increased sensitivity to bone-conducted sounds appears to be the mechanism responsible for these symptoms. Bone conduction thresholds on audiometry can be less than 0 dB normal hearing level (NHL). Therefore, an air-bone gap can exist even when air conduction thresholds are normal.^{10,18}

Four patients with dehiscence of bone overlying the superior canal with air-bone gaps in the affected ears that were greatest at lower frequencies have been described.¹⁸ Three of these patients had undergone stapedectomy before identification of superior canal dehiscence. The air-bone gap was unchanged postoperatively. Each patient had an intact VEMP response from the affected ear, a finding that would not have been expected based on exclusively a middle ear cause of conductive hearing loss (HL). Resurfacing of the superior canal through a middle fossa approach was performed in one of these four patients. Postoperatively, his vestibular symptoms were relieved, and his air conduction thresholds were improved.

Another one of these four patients had exclusively auditory and no vestibular symptoms or signs.

A recent report has described eight patients (10 ears) with air-bone gaps principally in the lower frequencies who were noted to have superior semicircular canal dehiscence on high-resolution temporal bone CT scans.¹⁹ Middle ear exploration had been performed and did not reveal any abnormality in 6 ears. Stapedectomy had been performed in three ears and ossiculoplasty in two ears, but the air-bone gap was unchanged postoperatively. None of these patients had vestibular symptoms or signs.

The purpose of this study was to determine the demographic features as well as the incidence of specific symptoms and signs in a large population of patients with dehiscence of bone overlying the superior semicircular canal. Audiograms and VEMP responses in these patients were reviewed to evaluate the abnormalities on these studies that are seen in association with superior canal dehiscence. Three hypotheses were evaluated in this study: 1) superior canal dehiscence will have effects on vestibular and auditory function occurring as a result of the third mobile window created by the dehiscence; 2) the threshold for eliciting a response for VEMP response is less in ears with superior canal dehiscence; and 3) symptoms in patients with superior canal dehiscence can be controlled with surgical repair of the dehiscence.

METHODS

Clinical manifestations of superior semicircular canal dehiscence were studied in patients identified with this abnormality over the time period of May 1995 to July 2004. Criteria for inclusion in clinical series were identification of the dehiscence of bone overlying the superior canal confirmed with a high-resolution temporal bone CT⁷ and the presence of at least one sign on physiologic testing indicative of superior canal dehiscence. These signs included eye movements evoked by sound or pressure stimuli or a VEMP threshold in the affected ear 85 dB or less NHL. The specific stimuli used to evaluate for evoked eye movements were pure tones at frequencies of 250 to 4,000 Hz and intensities of 100 to 110 dB NHL (administered for a period of 5 sec); Valsalva maneuver against pinched nostrils (inspiratory Valsalva); Valsalva against a closed glottis; and positive-negative pressure insufflation of air into the external auditory canal through a pneumatic otoscope. There were 65 patients who qualified for inclusion on the basis of these criteria. Vestibular manifestations were present in 60 of these patients, and there were 5 patients who had exclusively auditory manifestations without vestibular symptoms or signs but with an abnormal VEMP threshold and with dehiscence of bone overlying the affected superior canal identified on high-resolution temporal bone CT scan.

The clinical records, audiograms, and VEMP responses were reviewed to evaluate and classify the abnormalities associated with this disorder. In no case was there any sign of elevated intracranial pressure such as papilledema. Cranial imaging including magnetic resonance imaging and CT scans did not reveal any enlargement of the cerebral ventricles. No patient displayed signs or symptoms of a more generalized abnormality of bone metabolism such as osteoporosis.

This study was a review of existing clinical data with patient identifiers removed. It qualified for exemption from an institutional review board protocol based on United States Department of Health and Human Services criteria 45 CFR 46.101(b4).

The determination that the study was exempt for a protocol requirement was made by the institutional review board.

The initial symptoms and signs that led to the identification of this syndrome were sound or pressure induced vertigo and oscillopsia as well as eye movements in the plane of the superior canal evoked by these stimuli. These patients were noted to have dehiscence of bone overlying the superior canal identified on temporal bone CT scanning. As the understanding of the effects of superior canal dehiscence increased, it was recognized that patients can have auditory manifestations of dehiscence without accompanying vestibular symptoms or signs. The identification of these patients with auditory symptoms alone was prompted by findings of an air-bone gap on audiometry with normal acoustic reflex responses, intact responses to VEMP responses, and no ossicular abnormality noted on middle ear exploration.

Audiometry

Pure-tone audiometry was performed over the frequency ranges of 250 to 8,000 Hz for air conduction and 250 to 4,000 Hz for bone conduction. Testing was performed in a soundproof booth. Appropriate masking was used for bone conduction and, when needed, for air conduction.

VEMP Responses

The procedures used in the recording of VEMP responses were similar to those described previously.⁸⁻¹¹ The vestibulocollic reflex was evoked by rarefaction clicks delivered unilaterally to the ear by a pair of headphones (Telephonics Corp., New York, NY). The stimulus parameters were as follows: duration 0.1 millisecond; stimulation rate, 10 Hz; loudness, 60 to 103 dB NHL. The intensity (loudness) of the clicks was the sound pressure level referenced to the threshold for persons with normal hearing (NHL) and not referenced to the hearing level in an individual patient. Patients elevated their head to activate the sternocleidomastoid muscles during the recordings. The electromyography signal was recorded from surface electrodes, which were applied overlying the sternocleidomastoid muscle halfway between the mastoid and the clavicle. Reference electrodes were placed overlying the medial third of the clavicle along its anterior border. The ground electrode was located on the sternum. The analysis time after each click was 80 milliseconds. The signal was bandpass-filtered from 20 Hz to 2 kHz. A total of 128 sweeps were averaged, and the responses were reproduced in a second turn. The delivery of the sound stimulus and the analysis of the responses were performed using a Nicolet Spirit (Hong Kong) evoked potential system. The threshold for appearance of the characteristic p13/n23 waveform of the VEMP response was measured as has been previously described.⁸⁻¹¹ Control data were obtained from the results of VEMP testing in patients who, from a complete neurologic work-up and other tests such as audiometry, were determined not to have an inner ear disorder.

Recording of Eye Movements

Three-dimensional eye movements evoked by sound and pressure stimuli were recorded using magnetic search coils as has been described previously.³ Subjects gave informed consent for scleral search coil recordings through a protocol approved by the institutional review board. Horizontal, vertical, and torsional components of the movement of one or both eyes were recorded using a dual search coil embedded in a silicone annulus that was placed around the cornea (Skalar, Delft, The Netherlands). The eyes were anesthetized with topical proparacaine before placement of the search coil. The magnetic fields generating the voltages in the search coils were produced by three orthogonal pairs of coils, with a diameter and a distance of 1.02 m. The techniques for search coil calibration and recording have been described

previously.²⁰ Angular position signals were low-pass filtered with an analogue, single-pole, Butterworth, antialiasing filter with a 3 dB bandwidth of 100 Hz and then digitally sampled at 500 Hz. Eye and head velocities were derived by digital differentiation of the coil position signals with a fifth order finite-impulse-response filter limited to 30 Hz.

Temporal Bone CT Scans

During the period from May 1995 to December 1998 when patients were initially being identified with this syndrome, conventional temporal bone CT scans were performed with the following parameters: 1.0 mm collimation in the axial and coronal planes, 1.0 mm table increment, 330 mAs, 120 kVp. There were 12 patients in this series who underwent temporal bone CT scans with this technique. All of these patients had eye movements evoked by loud tones or by pressure stimuli, and the directional features of these evoked eye movements corresponded to those expected based on activation of the superior canal in the affected ear.^{1,3} Patients who were identified subsequent to December 1998 underwent thin-section helical CT of the temporal bones.⁷ Single and helical sections were obtained with 0.5 mm collimation in the axial plane and reformation of images in the plane of the superior canal. These high-resolution temporal bone CT scans have been shown to have a sensitivity of 100%, specificity of 99%, positive predictive value of 93%, and negative predictive value of 100% in the identification of superior canal dehiscence.⁷ The specificity and positive predictive value of the high-resolution scans are substantially greater than those noted with conventional temporal bone CT scans.^{7,21}

RESULTS

The symptoms and signs that are associated with superior canal dehiscence can be better understood from a case presentation. This patient's history, findings on examination, and test results were typical of those patients with vestibular manifestations of superior canal dehiscence.

Case Presentation

A 39-year-old man presented with a history of a clogged sensation in his left ear and a feeling of unsteadiness when he coughed or sneezed. These symptoms began approximately 5 months before his presentation and were not associated with any specific event or illness. He was the driver of a car involved in an automobile accident about 2 months after the initial onset of his symptoms. Although he did not hit his head and did not lose consciousness as a result of the accident, he was moved backward and forward in a violent fashion as a result of the impact. He began to experience a new constellation of symptoms after the automobile accident. He noted oscillopsia (the apparent motion of objects in his visual surround) when he coughed or sneezed and when he heard loud noises in his left ear. He also began to hear his heartbeat in his left ear, his voice sounded louder in his left ear, and he noted that he heard the impact of his foot loudly in his left ear when he ran.

On examination under Frenzel lenses, tones of 500 to 2,000 Hz at intensities of 100 to 110 dB NHL in the left ear evoked a nystagmus with slow phases that had vertical and torsional components (Fig. 1). The slow-phase motion of the eye was upward (vertical component) and also involved motion of the superior pole of each eye to the right

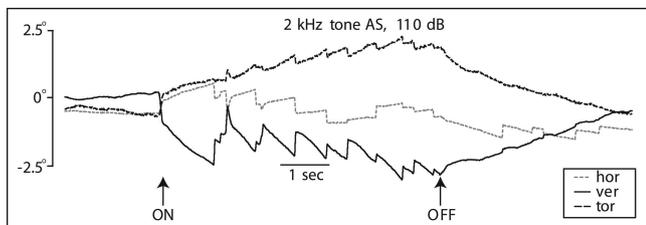


Fig. 1. Horizontal (hor), vertical (ver), and torsional (tor) eye positions were recorded with scleral search coils and plotted against time for the left eye in a 39-year-old man with left superior canal dehiscence syndrome. A 2 kHz tone at 110 dB was presented to the left ear. The tone began at the arrow labeled "On" and was turned off at the arrow labeled "Off." Positive direction for the hor, ver, and tor axis is defined as left, down, and clockwise (rotation of the superior pole of the patient's eye toward his right side). These directional conventions are standard for the field of three-dimensional eye movement analysis and represent axis orientations defined by the right-hand rule. In response to the tone presented to the left ear, the patient developed nystagmus with slow phase components that were principally directed upward and clockwise. The directional characteristics of this nystagmus are those expected based on excitation of the left superior semicircular canal.

(torsional component). Valsalva against a closed glottis led to eye movements that were in the same plane but opposite in direction (Fig. 2). Release of the glottic Valsalva caused reversal of the evoked eye movements. These vertical-torsional eye movements are in the plane of the left superior semicircular canal with responses to tones and to the release of the glottic Valsalva maneuver corresponding to excitation of the left superior canal and responses to the glottic Valsalva maneuver itself corresponding to inhibition of the left superior canal.

Weber tuning fork testing (512 Hz fork) lateralized to the left ear. Rinne tuning fork testing was positive (air conduction > bone conduction) bilaterally. The remainder of the neurotologic examination was normal.

The patient's audiogram revealed an air-bone gap that was greatest for the low frequencies of the left ear

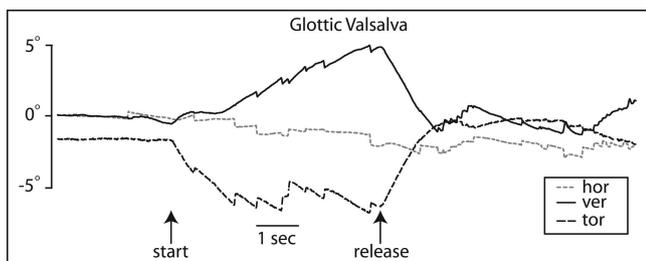


Fig. 2. Response to glottic Valsalva maneuver in same patient as Figure 1. The patient was instructed to take a deep breath and strain down against a closed glottis. This maneuver began at the arrow marked "start," and the patient released the pressure at the arrow marked "release." Directional conventions for the eye movements are the same as those described in Figure 1. During the glottic Valsalva maneuver, the patient has a nystagmus with slow phase components that were principally downward and counterclockwise. The directional characteristics of this nystagmus are those expected based on inhibition of the left superior semicircular canal. On release of the pressure from the Valsalva maneuver, the patient developed eye movements and a nystagmus with slow phase components that were upward and clockwise (corresponding to excitation of the left superior canal).

(Fig. 3). His VEMP threshold for the left and right ears were 80 and 100 dB NHL, respectively. A high-resolution temporal bone CT scan revealed dehiscence of bone overlying the left superior semicircular canal (Figs. 4 and 5). Bone overlying the right superior canal was intact.

Clinical Manifestations in Patients with Vestibular Symptoms and Signs

Vestibular manifestations of superior canal dehiscence were identified in 60 patients (41 males and 19 females). The age range at the time of diagnosis was 13 to 70 (median = 41; mean = 43) years. The right ear alone was affected in 27 patients and left ear alone in 23 patients. There were 10 patients with vestibular symptoms, signs, and CT findings indicative of bilateral superior canal dehiscence. Vestibular symptoms induced by loud sounds were noted in 54 (90%) patients, and pressure-induced symptoms (coughing, sneezing, straining) were present in 44 (73%). There were 40 (67%) patients who had both sound- and pressure-induced symptoms. Symptoms indicative of hyperacusis for bone-conducted sounds (described as "conductive hyperacusis")⁹ were present in 31 (52%) patients. Conductive hyperacusis included symptoms such as hearing eye movements in the affected ear and hearing the impact of the feet during walking or running. There were 36 (60%) patients with autophony in the affected ear.

Vestibular signs associated with superior canal dehiscence were present in 57 of these 60 (90%) patients. The vestibular signs in these 57 patients could be grouped into four categories. Sound-evoked eye movements were noted in 46 (82%) patients. A sound-induced tilt of the head in the plane of the superior canal was noted in 11 (20%) patients. There were 42 (75%) patients with eye movements induced by Valsalva maneuvers. Eye movements evoked by pressure in the external auditory canal were noted in 26 (45%) patients. A sign in only one of these four categories was noted in 17 (27%) patients. Signs in two categories were noted in 16 (29%) patients. Signs in three categories were noted in

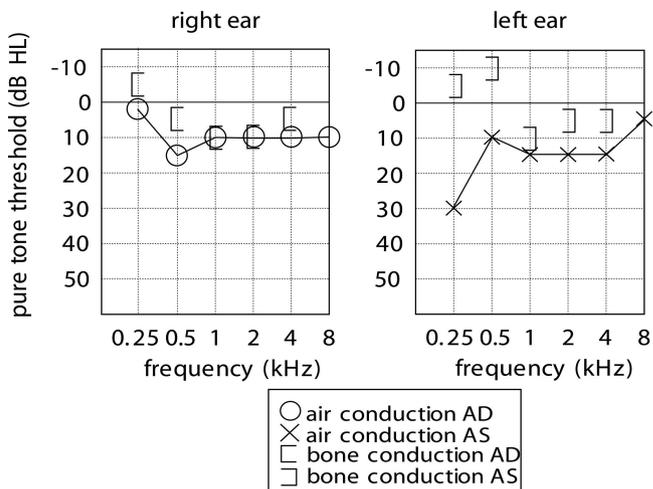


Fig. 3. Pure-tone audiogram in 39-year-old man with left superior canal dehiscence syndrome. Bone conduction thresholds were measured while giving masking noise to the contralateral ear.

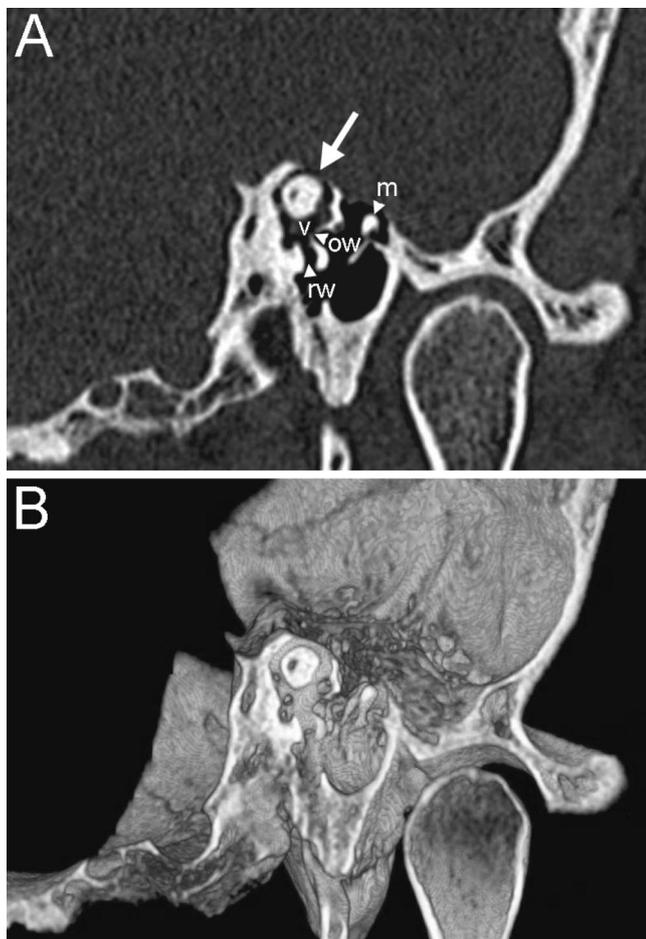


Fig. 4. Computed tomography scan reconstructions in two (A) and three (B) dimensions passing through the plane of the dehiscent left superior canal (SC) in the 39-year-old man whose history is described in text. Dehiscence of the SC into the middle cranial fossa (arrow). Scans were acquired on a Toshiba Aquilion 16-detector scanner in step-scan mode (4 detectors used per rotation) with no overlap of scans. Scan time with 1 rotation/sec was approximately 30 seconds. The beam was collimated to 0.5 mm with settings of 120 kV and 300 mA. A bone algorithm filter was applied before image reconstructions. A high-resolution temporal bone image volume dataset was constructed by selecting a small field of view that included only the temporal bone (180 mm). For multiplanar reconstructions, this image dataset was exported to a graphical workstation and manipulated with Vitrea 2 Imaging Software. The dataset was rotated and sliced so that the plane of the image corresponded to the plane of the bony SC. Thickness of the image in A is approximately 0.2 mm; in B, an additional 25 mm of temporal bone posterior to the SC is included for orientation. v = vestibule, m = malleus head, ow = oval window, rw = round window.

20 (36%) patients. Signs in all four categories were noted in four (7%) patients.

One patient, a 60-year-old man, initially presented with bilateral conductive hearing loss and no vestibular symptoms or signs. His acoustic reflex responses were intact, which prompted investigation for superior canal dehiscence. The threshold for evoking a VEMP response was reduced in both ears (75 dB bilaterally), and a high-resolution CT scan of the temporal bones revealed dehiscence of bone overlying both superior canals. He began to develop sound-induced vertigo and eye movements in the

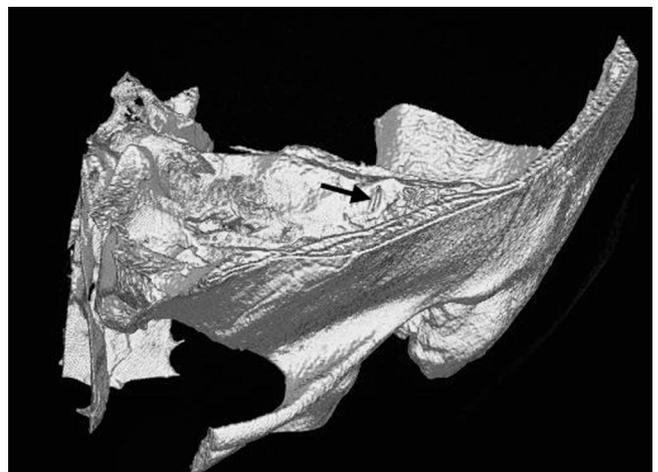


Fig. 5. Computed tomography images from reconstructions in the plane of the left superior canal in the same patient as in Figure 4. Image data were acquired and reconstructed as described in legend to Figure 4. Arrow points to dehiscence.

plane of the superior canal evoked by loud tones in each ear 2 years after the diagnosis of superior canal dehiscence was made on the basis of his auditory findings.

There were three patients with vestibular symptoms who did not have specific signs of dehiscence on clinical examination. Each of these patients had dehiscence of bone overlying the affected superior canal noted on high-resolution temporal bone CT scan. The first patient, a 59-year-old woman, had vertigo induced by loud noises in the left ear. She had undergone prior resurfacing of the left superior canal through a middle fossa approach at an outside institution. She continued to have sound-induced symptoms and hyperacusis postoperatively, but there were no eye movements evoked by sound or pressure stimuli. Temporal bone CT scan showed that the bone graft had migrated and was not covering the superior canal dehiscence on the left side. The bone overlying the right superior canal was intact. The second patient, a 49-year-old man, had autophony and fullness in his left ear and vertigo with pressure in the left external canal but no evoked eye movements. Temporal bone CT scan showed left superior canal dehiscence. The third patient, a 48-year-old man, had vertigo with loud noises in his left ear without evoked eye movements. Temporal bone CT scan showed left superior canal dehiscence. In each of these patients, the threshold for a VEMP response in the affected ear was abnormally low (75–80 dB NHL).

There were 10 patients with vestibular manifestations of bilateral superior canal dehiscence. The symptoms and signs were equal for the right and left ears in 4 of these 10 patients. For these four patients with equal findings bilaterally, eye movements were evoked by sound stimuli delivered to each ear but not by pressure stimuli in either external auditory canal in one patient. Eye movements were induced by pressure in the external auditory canals in both ears but not by sound stimuli in either ear in two patients. Eye movements were induced by both sound in each ear and by pressure in the external auditory canals in one patient. There were four patients with bilat-

eral dehiscence in whom the symptoms and signs were present in both ears but more severe in one ear than in the other. Sound-induced eye movements from each ear but not pressure-induced eye movements were noted in two of these patients. Pressure-induced eye movements from each external auditory canal but not sound-induced eye movements were noted in one patient. There were two patients who had sound- and pressure-induced eye movements from one ear but no evoked eye movements from the contralateral ear, although in each patient a high-resolution CT scan of the temporal bones demonstrated bilateral dehiscence.

Nine of the patients with vestibular manifestations of superior canal dehiscence had undergone prior middle ear exploration for presumed perilymphatic fistula before the identification of superior canal dehiscence. There were three patients who had undergone a stapes replacement procedure, without improvement in their air-bone gap, before referral to our center, where a diagnosis of superior canal dehiscence was made.

There were 14 patients who described a specific event or occurrence that was associated with the onset of symptoms. Head trauma was the cause in eight patients, and activities resulting in changes in middle ear or intracranial pressure (such as weight lifting, coughing, straining) were identified by six patients as being responsible for initiation of symptoms. No precipitating event associated with the onset of symptoms could be identified in 46 patients. Their symptoms appeared to begin gradually, making it difficult for them to establish a precise time of onset.

Clinical Manifestations in Patients with Exclusively Auditory Symptoms and Signs

Auditory manifestations of superior canal dehiscence (without vestibular symptoms or signs) were identified in five patients (3 males and 2 females). The age range of these patients at the time of diagnosis was 30 to 78 (median = 54; mean = 53) years. The right ear alone was affected in one patient and the left ear alone in three patients. There was one patient with auditory symptoms and signs and CT findings indicative of bilateral superior canal dehiscence. Symptoms indicative of hyperacusis for bone-conducted sounds were present in three patients, and there were three patients with autophony in the affected ear. One patient had undergone a stapes replacement procedure before the identification of superior canal dehiscence. His air-bone gap was unchanged after stapes replacement. There were no precipitating events associated with the onset of auditory symptoms in any of these patients. Each of the six ears affected with superior canal dehiscence had an abnormally low VEMP threshold (≤ 85 dB NHL).

Audiometry

Audiograms with information suitable for analysis of an air-bone gap were available for 47 ears with superior canal dehiscence in patients with vestibular manifestations of this disorder. The air-bone gap (mean \pm SD) was greatest at the lower frequencies and measured as follows: 19 ± 14 dB at 250 Hz; 15 ± 11 dB at 500 Hz; 11 ± 9 dB at 1,000 Hz; 4 ± 6 dB at 2,000 Hz; 4 ± 8 dB at 4,000 Hz. An

air-bone gap 10 dB or greater was present in 33 ears with superior canal dehiscence at 250 Hz, 32 ears at 500 Hz, 30 ears at 1,000 Hz, and 10 ears at 2,000 Hz.

Audiograms with information suitable for analysis of an air-bone gap were available for six ears with superior canal dehiscence in patients with exclusively auditory manifestations of this disorder. As for the patients with vestibular manifestations of dehiscence, the air-bone gap was greatest at the lower frequencies and measured as follows: 17.5 ± 9.4 dB at 250 Hz; 14 ± 11 dB at 500 Hz; 10 ± 6 dB at 1,000 Hz; 1 ± 3 dB at 2,000 Hz; 5 ± 9 dB at 4,000 Hz. An air-bone gap 10 dB or greater was present in five ears with superior canal dehiscence at 250 Hz, four ears at 500 Hz, three ears at 1,000 Hz, and none at 2,000 Hz. There was no difference in the measured air-bone gap at any frequency between the patients with vestibular symptoms and those with exclusively auditory manifestations of superior canal dehiscence (*t* test; $P > .5$ at all frequencies).

VEMP Responses

VEMP testing was performed in 51 ears with superior semicircular canal dehiscence, 30 unaffected ears in patients with superior canal dehiscence in the opposite ear, and 60 ears in 30 patients who had undergone VEMP testing for other reasons and who did not have symptoms or signs suggestive of superior canal dehiscence (controls). The threshold (mean \pm SD) for the affected ears was 81 ± 9 dB NHL. The threshold for the unaffected ears was 99 ± 7 dB, and the threshold for the control ears was 98 ± 4 dB. Comparisons between groups were made using a nonparametric statistical test, the Mann-Whitney rank sum test. The affected ear thresholds were significantly different from both the unaffected ear and normal control thresholds ($P < .001$ for both comparisons). There was no difference between the unaffected ear and normal control thresholds ($P = .2$). The distribution of thresholds in these groups of patients is shown in Figure 6.

Surgical Repair of Dehiscence

There were 20 patients (11 males, 9 females; age 27–64) who underwent surgical repair of superior canal dehiscence through a middle cranial fossa approach. Dehiscence was present in the left superior canal alone in 11 patients and the right superior canal alone in 6. There were three patients with signs of bilateral superior canal dehiscence on high-resolution temporal bone CT scan. In each case, symptoms and signs predominantly localized to one ear (left ear in 2 and right ear in 1), and surgery was performed on the more symptomatic ear.

Each of the 20 patients in this surgical series was debilitated by symptoms associated with superior canal dehiscence. These symptoms involved sound-induced vertigo and oscillopsia in 17 (85%), pressure-induced vertigo and oscillopsia in 16 (80%), conductive hyperacusis in 12 (60%), and chronic disequilibrium in 12 (60%). Eye movements were evoked by tones in the affected in 14 (70%) patients, by Valsalva maneuvers in 12 (60%) patients, and by pressure in the external auditory canal in 7 (35%) patients.

VEMP Threshold

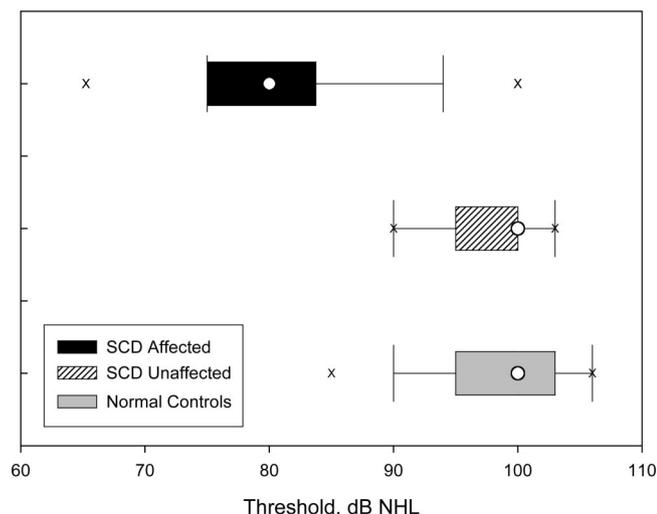


Fig. 6. Thresholds for evoking a vestibular-evoked myogenic potentials (VEMP) response in ears affected with superior canal dehiscence, unaffected ears in patients with superior canal dehiscence in the opposite ear, and normal control ears. Each box represents the 25th to 75th percentile range, the vertical bars represent the 10th to 90th percentile range. The two outer Xs represent the 5th and 95th percentile range, and the middle circle denotes the median value.

The surgical procedure in nine of the patients involved plugging the canal using a procedure similar to that described for posterior canal occlusion in patients with intractable BPPV.²² Fascia and bone pate were placed in the lumen of the superior canal, thereby plugging the canal. The plugged canal was then covered with a cortical bone graft harvested from the inner surface of the middle fossa bone flap. The surgical procedure in 11 patients involved resurfacing the canal (without plugging the lumen). Fascia was placed over the canal but not packed within the lumen. The fascia was then covered with a cortical bone graft, and fibrin glue was used to seal the area and hold the graft in position. The auditory brainstem response, facial nerve, and somatosensory evoked potentials were monitored in all of the surgical procedures. In no case was there any change in the neural responses during the procedure. All patients received Decadron, which was tapered gradually, typically over the initial 2 weeks after the procedure.

The initial surgical approach devoted for the treatment of this condition involved plugging the superior canal through a middle cranial fossa approach. The second patient developed moderate-severe sensorineural hearing loss 7 days after a revision plugging procedure. The resurfacing technique was then used in an attempt to minimize the risk of hearing loss.

Resurfacing of the superior canal led to complete resolution of symptoms in 7 of the 11 patients who underwent this procedure. Four of the patients undergoing canal resurfacing had initial resolution of symptoms but then developed recurrent vestibular symptoms and signs at 3 to 6 months after the surgical procedure. These re-

currences led us to use the plugging procedure in subsequent cases.

Complete resolution of symptoms and signs has been achieved in eight of the nine patients who underwent canal plugging. There was one patient who described her symptoms as being "90%" relieved after a plugging procedure. She had experienced sound- and pressure-induced vertigo and eye movements before the plugging procedure. The sound-induced eye movements were resolved after the plugging procedure, but she continued to have eye movements evoked by pressure in the external auditory canal, although these pressure-induced eye movements were decreased in amplitude compared with preoperative responses.

Two patients had moderate to severe sensorineural hearing loss in the operated ear after a revision procedure to repair superior canal dehiscence. In each case, this hearing loss was delayed in onset relative to the surgery. In each case, this hearing loss was delayed in onset relative to the surgery. The first patient, a 41-year-old woman, experienced pressure-induced vertigo and oscillopsia and disabling disequilibrium from left superior canal dehiscence. She underwent plugging of the left superior canal through a middle cranial fossa approach. Her symptoms were completely relieved for about 2 months. She then experienced a return of oscillopsia induced by pressure in the left external auditory canal and of disequilibrium. The left middle fossa in the region of the dehiscence was surgically re-explored. The plug was noted to be intact, but a further region of dehiscence in bone of the superior canal was identified anterior and posterior to this plugged region. The canal was packed with fascia and bone dust on both sides of the previous plug. Her hearing was normal initially, but she experienced a sudden decline in acuity and speech understanding in the left ear on the seventh postoperative day.

The second patient who experienced hearing loss after surgery to repair superior canal dehiscence was a 59-year-old woman who was debilitated by sound-induced vertigo and oscillopsia from left superior canal dehiscence. She underwent resurfacing of the superior canal through a middle fossa approach at an outside institution. Her symptoms were improved for approximately 2 weeks after surgery but then returned and were as severe as they were before this procedure. She underwent re-exploration of the left middle fossa. The previously placed bone graft was noted to have migrated and was no longer covering the canal. The membranous canal was covered with fascia, and another bone graft was placed. Her vestibular symptoms were relieved after this procedure. Her hearing was initially intact but dropped suddenly about 1 week after surgery. Improvement in hearing occurred over time and she has a moderate to severe hearing loss in the left ear.

There have been no other cases of HL after these surgical procedures for superior canal dehiscence. There were no other complications in this series of 20 patients.

DISCUSSION

Overview of Superior Semicircular Canal Dehiscence Syndrome

This report presents the clinical findings in 65 patients with vestibular or auditory abnormalities related to

superior canal dehiscence. Most of the patients in this series (60 of the 65, 92%) had vestibular symptoms attributable to superior canal dehiscence. Specific vestibular signs associated with superior canal dehiscence were present in 57 of these 60 (95%) patients. These signs included eye movements evoked by stimuli such as loud tones, Valsalva maneuvers, or pressure in the external auditory canal. Some patients also had a brisk motion of the head in the plane of the superior canal in response to loud tones. Chronic disequilibrium, presumably arising from the effects of abnormal vestibular stimulation on balance mechanisms, was also a common complaint.

There were five patients in this series that had auditory manifestations of superior canal dehiscence but no vestibular symptoms or signs. These auditory symptoms included hypersensitivity to bone conducted sounds and autophony. The physiologic basis for differences in the specific symptoms and signs experienced by patients with superior canal dehiscence is not presently known.^{18,19,23}

The symptoms and findings in these patients define a clinical syndrome of abnormalities in vestibular and auditory function resulting from superior canal dehiscence. Before the discovery of superior canal dehiscence as the underlying cause for these abnormalities, some of these patients had undergone middle ear exploration for presumed perilymphatic fistula. Others had undergone stapes replacement, although there was no improvement in their air-bone gap postoperatively.^{18,24} Clinical studies before the description of superior canal dehiscence syndrome identified patients with the Tullio phenomenon and a lower threshold for the VEMP response in the affected ear, but the cause of these findings was unknown.^{25,26} It is likely that the patients in these earlier studies had superior canal dehiscence.⁹

Many patients with superior canal dehiscence have only mild to moderate symptoms. Treatment options should be considered on the basis of the character and severity of the patient's symptoms. For patients with primarily sound-induced symptoms, avoidance of loud noises may be sufficient to prevent the clinical manifestations. A tympanostomy tube can be beneficial for patients with symptoms arising mainly from pressure in the external auditory canal.

Surgical repair of the dehiscence should be reserved for patients who are debilitated by their symptoms. Vestibular manifestations are usually the most troubling and are the ones for which surgical correction has been shown to be beneficial. The middle cranial fossa approach has been used in this series. It offers direct visualization of the dehiscence, and the degree of dural retraction required to expose the superior canal is considerably less than that required in middle fossa approaches to the internal auditory canal. Plugging of the superior canal through a transmastoid approach could possibly be an alternative approach. The tegmen is often low lying in these cases, which could compromise access to the full extent of the dehiscence in the transmastoid approach.

This surgical series began with repair of the dehiscence achieved through canal plugging. A resurfacing procedure was then developed after one patient developed delayed sensorineural hearing loss after a revision plug-

ging procedure. Although the resurfacing procedure has achieved complete relief of symptoms, recurrent symptoms and signs have developed in 4 of 11 patients who underwent the procedure. In contrast, complete relief of symptoms was achieved and maintained in eight of the nine patients who underwent canal plugging. The findings from this series and from a recent presentation of two other patients who underwent surgical repair of superior canal dehiscence²⁷ indicate that canal plugging is more likely to achieve enduring control of symptoms.

The findings in this series suggest that the risk of HL is not dependent on whether canal plugging or resurfacing is used because one patient from each of these groups developed delayed hearing loss after the procedure. Hearing loss occurred in the two patients who underwent revision procedures. The cause of the hearing loss cannot be determined with certainty. There may have been adhesions between the membranous canal and the middle fossa dura, although none were noted at the time of surgery. A tear in the membranous canal as a consequence of retraction or disruption of these adhesions would have been expected to lead to hearing loss immediately or shortly after the procedure. Instead, these patients had intact hearing initially after surgery and developed delayed sensorineural hearing loss in the operated ear. Chemical labyrinthitis or secondary endolymphatic hydrops are other possible causes. There were no patients in this series that had HL after a primary (not revision) procedure to repair superior canal dehiscence.

Two recent studies provide further insight into the clinical manifestations of superior canal dehiscence. A patient with head-movement-induced oscillopsia and bilateral superior canal dehiscence has recently been reported.²⁸ This patient had diminished function in both superior canals. The impairment of the vertical vestibulo-ocular reflex (VOR) that should have arisen from the superior canals is thought to be responsible for the patient's vertical oscillopsia and impaired vision during locomotion. In a study of four patients with superior canal dehiscence syndrome, vertical eye movements were recorded with electro-oculography when responses to 128 click stimuli from the affected ear were averaged.²⁹ Such responses were not present in normal subjects.

Mechanism

The mechanism responsible for the vestibular symptoms and signs in these patients is related to the effects of the dehiscence on pressure transmission in the labyrinth. The dehiscence creates a "third mobile window" into the inner ear, thereby allowing the superior canal to respond to sound and pressure stimuli.¹ The evoked eye movements in these patients typically align with the affected superior semicircular canal.

Ampullifugal (excitatory) deflection of cupula in the superior canal results from loud sounds, positive pressure in the external auditory canal, and the Valsalva maneuver against pinched nostrils. The evoked eye movements resulting from these stimuli are conjugate (relatively equivalent for both eyes) and has slow phase components that are directed upward with torsional motion of the superior pole of the eye away from the affected ear. Conversely,

negative pressure in the external canal, Valsalva against a closed glottis, and jugular venous compression cause ampullipetal (inhibitory) deflection of the cupula in the superior canal. The evoked eye movements resulting from these stimuli are downward with torsional motion of the superior pole of the eye toward the affected ear (i.e., in the same plane but opposite in direction to those resulting from the excitatory stimuli). These eye movement findings have been documented with three-dimensional search coil techniques and can be observed on clinical examination.³ Frenzel lenses should be used when these observations are being made in the clinic because visual fixation can lead to suppression of the evoked eye movements.

A longer length of dehiscence overlying the superior canal (≥ 5 mm) can lead to dysfunction in the affected canal.^{3,4} The function in individual semicircular canals is evaluated by measuring the three-dimensional VOR evoked by rapid, high-acceleration, transient head movements ("head thrusts") in the planes of each of the canals.^{30,31} Hypofunction in an individual canal leads to a deficient VOR evoked by head thrusts that would result in an excitatory response from the canal if its function were intact. Quantitative evaluation of the three-dimensional VOR in response to head thrusts has demonstrated that the VOR evoked by excitation of the affected superior canal often has reduced gain when the dehiscence has a length of 5 mm or more. This reduced function in the canal may be caused by compression of the membranous canal by the overlying dura and temporal lobe. The eye movements evoked by sound and pressure stimuli in these patients may not align with the plane of the superior canal when function in the canal is reduced.^{3,4}

Sound-induced vestibular responses were initially documented in experimental studies over 75 years ago. In initial studies by Tullio,³² later elaborated by Huizinga³³ and Eunen et al.,³⁴ fenestration of individual semicircular canals in pigeons led to sound-evoked eye and head movements in the plane of the fenestrated canal. Clinical studies initially identified the Tullio phenomenon in patients with congenital syphilis. Histopathologic studies of the temporal bone in these cases showed gummatous osteomyelitis and labyrinthine fistulae.³⁵ Hennebert³⁶ noted that pressure in the external auditory canal in some patients with congenital syphilis caused vestibular symptoms and signs. It is presumed that the labyrinthine fistulae that result from congenital syphilis are responsible for the Tullio phenomenon and Hennebert sign in these patients.

An animal model of superior canal dehiscence has been developed by fenestration of the bone covering the uppermost portion of the superior canal, where it protrudes into the mastoid bulla, in chinchillas.³⁷ Single-unit extracellular recordings from vestibular-nerve afferents were then made to determine the effects of this fenestration on afferent responses to pressure and sound stimuli. Superior canal afferents always responded to external ear canal pressure changes after fenestration and so did 36% of horizontal canal afferents and half of otolith afferents.³⁷ Responses to acoustic stimuli were recorded from afferents in the superior division of the vestibular nerve before and after fenestration of the superior canal.³⁸ Afferents

from all vestibular end organs encountered could respond to acoustic stimuli, even before fenestration. The effects of superior canal fenestration were most pronounced for afferents innervating this canal. Two response patterns to acoustic stimuli were seen: rapid phase locking and slow tonic changes in firing rate. Phasic responses principally occurred in irregularly discharging afferents and tonic responses in regularly discharging afferents. Fenestration lowered the thresholds for acoustic stimulation in superior canal afferents with phasic responses and increased the magnitude of tonic responses. The findings provide a basis for understanding the alignment of the evoked eye movements in patients with superior canal dehiscence.

Auditory Manifestations of Superior Canal Dehiscence

The audiograms in these patients show that superior canal dehiscence is associated with an air-bone gap of 10 dB or greater at lower frequencies for most ears with this disorder. This air-bone gap is not caused by a disorder in middle ear conductive mechanisms because stapedius reflex responses are intact in these patients and middle ear explorations (performed prior to the recognition of superior canal dehiscence) have commonly revealed no abnormality.^{18,19} It is likely that the same mechanism responsible for the vestibular abnormalities also underlies the auditory manifestations. The third mobile window allows acoustic energy to be dissipated through the dehiscence.¹⁸ Direct experimental evidence in support of this mechanism has been provided from studies performed in a chinchilla model of superior canal dehiscence.²³ The sound-induced velocity measured within the perilymph or endolymph of a superior canal dehiscence demonstrated sound flow through the dehiscence.²³ Measurements of the cochlear potential showed that superior canal dehiscence causes an increase in the bone-conducted sound.²³

Laser-Doppler vibrometer measurements of sound-induced umbo velocity in patients with superior canal dehiscence have revealed hypermobility.²³ The pattern of changes in velocity and angle on laser-Doppler vibrometry in these patients are similar to those noted in ears with ossicular interruption.

The acoustic effects of superior canal dehiscence include increased sensitivity to bone-conducted sounds (conductive hyperacusis) and autophony. The Weber tuning fork test typically lateralizes to the affected ear, and patients may hear a tuning fork placed at a site remote to the skull such as the lateral malleolus of the ankle.⁹

Recognition of the acoustic effects of superior canal dehiscence is important in preventing the interpretation of an air-bone gap on audiometry in these patients as being caused by a middle ear abnormality. Earlier studies have described "inner ear conductive HL" in patients with a conductive hearing impairment on audiometry but for whom there is no evidence of tympanic membrane or ossicular abnormality.³⁹ Superior canal dehiscence is a cause of inner ear conductive HL.^{18,19} Patients with intact acoustic reflex responses and an air-bone gap on audiometry should undergo further investigation for superior canal dehiscence, such as a high-resolution CT scan of the

temporal bones, before contemplating a middle ear exploration.

Further studies will be required to determine why some patients with superior canal dehiscence have exclusively vestibular abnormalities, some exclusively auditory effects, and others both vestibular and auditory manifestations. Factors that may contribute to differences in the auditory or vestibular manifestations of superior canal dehiscence include whether the cochlear aqueduct is patent and the relative compliance of the round window membrane. The chinchilla model of this abnormality offers opportunities for an increased understanding of the physiologic effects of dehiscence.^{23,37,38} Continued longitudinal follow-up of patients with the disorder may also provide additional insights. One patient in this series had exclusively auditory manifestations at the time superior canal dehiscence was identified but later developed vestibular symptoms and signs.

VEMP Responses in Superior Canal Dehiscence

VEMP responses have a lower threshold in superior canal dehiscence.^{8–10} The findings in this large clinical series establish that evaluation of VEMP responses has considerable diagnostic utility in patients in whom superior canal dehiscence is suspected. The VEMP thresholds measured in this study in ears affected with superior canal dehiscence are comparable with those reported in other studies, as are the control values for normal responses.^{8–10} The mechanism responsible for the lowered VEMP threshold in these patients is likely to also be related to the lowered impedance for transmission of sound and pressure stimuli created by the third mobile window. Thus, the VEMP stimulus in an ear with superior canal dehiscence results in larger activation of the sacculus than a comparable stimulus in an ear without dehiscence. Lowered thresholds for VEMP responses have also been reported in patients with enlarged vestibular aqueduct.⁴⁰

The findings in this study indicate that the assessment of VEMP responses can be particularly useful in the search for the cause of an air-bone gap on audiometry. Conductive HL caused by a middle ear abnormality typically results in an absence of a VEMP response to click stimuli in the affected ear. The presence of a VEMP response elicited at an abnormally low threshold, as noted for the patients in this study, in the setting of conductive HL provides powerful evidence that middle ear mechanisms are not responsible for the air-bone gap.

Rarefaction clicks delivered through headphones were used as the stimulus for VEMP responses in this study. Bone-conducted tone bursts and clicks have been shown to evoke myogenic potentials of vestibular origin when recordings with surface electrode electromyography are made overlying the sternocleidomastoid muscle.⁴¹ A VEMP response is elicited bilaterally when bone-conducted sounds are presented both behind the healthy and the affected ear in patients with unilateral superior canal dehiscence.⁴² The amplitude of the VEMP response is larger on the affected side than on the healthy side.

Etiology of Superior Canal Dehiscence

Temporal bone CT studies have shown that the thickness of bone overlying the intact superior canal in patients with unilateral dehiscence is significantly thinner than in patients without superior canal dehiscence.⁴³ In a histologic study of 1,000 temporal bones from 596 adults, dehiscence or thinning (thickness of bone measuring ≤ 0.1 mm) of the superior canal in the floor of the middle cranial fossa was identified in nine specimens (approximately 1%). The abnormalities were typically bilateral. The findings from both the temporal bone CT study and the histologic study indicate that superior canal dehiscence most likely has a congenital or developmental etiology.

Symptoms and signs do not, however, appear to be present early in life. The youngest patient in this series was 13 at the time the diagnosis was made, which was a few months after he noted the onset of symptoms. Patients were commonly middle aged at the time the clinical manifestations began to appear and the diagnosis was made. It is possible that patients with this disorder fail to develop a normal thickness of bone overlying the superior canal and that this abnormally thin layer of bone becomes disrupted either by a traumatic event or by pressure from the overlying temporal lobe or dura. Such effects could develop gradually over the course of time.

CONCLUSIONS

Dehiscence of bone overlying the superior canal has been shown to have effects on vestibular and auditory function. The vestibular symptoms include vertigo and oscillopsia induced by loud sounds and by maneuvers that change middle ear or intracranial pressure. The vestibular signs include eye movements evoked by these stimuli. The plane of these eye movements typically aligns with the plane of the affected superior canal, indicating activation of the VOR by these stimuli. Patients may also experience head movements in the plane of the superior canal in response to loud sounds from activation of the vestibulo-collic reflex. High-resolution temporal bone CT scans have confirmed the presence of a dehiscence.

Auditory manifestations of superior canal dehiscence include hyperacusis for bone conducted sounds and autophony. An air-bone gap on pure tone audiometry for low frequency sounds is often observed in the affected ear.

The auditory and vestibular manifestations of superior canal dehiscence can be understood in terms of the creation of a third mobile window by the dehiscence. A similar mechanism likely accounts for the lower threshold for VEMP responses in an ear affected with superior canal dehiscence.

The symptoms in some patients with superior canal dehiscence can be controlled by avoidance of provocative stimuli. For patients who are debilitated by the vestibular manifestations, plugging of the superior canal performed through the middle fossa approach has been shown to be effective in achieving long-term resolution of vestibular symptoms and signs.

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