

Sound- and/or Pressure-Induced Vertigo Due to Bone Dehiscence of the Superior Semicircular Canal

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Objectives: To present symptoms, patterns of nystagmus, and computed tomographic scan identification of patients with sound- and/or pressure-induced vertigo due to dehiscence of bone overlying the superior semicircular canal. To describe anatomical findings and outcome in 2 patients undergoing plugging of the superior semicircular canal for treatment of these symptoms.

Design and Setting: Prospective study of a case series in a tertiary care referral center.

Patients and Results: Eight patients with vertigo, oscillopsia, and/or disequilibrium related to sound, changes in middle ear pressure, and/or changes in intracranial pressure were identified in a 2-year period. Seven of these patients also had vertical-torsional eye movements induced by these sound and/or pressure stimuli. The direction of the evoked eye movements could be explained by excitation or inhibition of the superior semicircular canal in the affected ear. Computed tomographic scans of the temporal bones identified dehiscence of bone overlying the affected superior semicircular canal in each case. Disabling disequilibrium in 2 patients

prompted plugging of the dehiscent superior canal through a middle cranial fossa approach. Symptoms were improved in each case. One patient developed recurrent symptoms requiring an additional plugging procedure and developed sensorineural hearing loss several days after this second procedure.

Conclusions: We have identified patients with a syndrome of vestibular symptoms induced by sound in an ear or by changes in middle ear or intracranial pressure. These patients can also experience chronic disequilibrium. Eye movements in the plane parallel to that of the superior semicircular canal were evoked by stimuli that have the potential to cause ampullofugal or ampullopetal deflection of this canal's cupula in the presence of a dehiscence of bone overlying the canal. The existence of such dehiscences was confirmed with computed tomographic scans of the temporal bones. Surgical plugging of the affected canal may be beneficial in patients with disabling symptoms.

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VESTIBULAR responses to sound and/or pressure transmitted to an inner ear were initially documented in studies during the first quarter of this century. The Tullio phenomenon is vertigo or other abnormal vestibular sensations accompanied by eye and/or head movements in response to sound. In the initial experimental studies by Tullio,¹ later elaborated by Huizinga² and Eunen et al,³ fenestration of individual semicircular canals in pigeons led to sound-evoked eye and head movements in the plane of these canals. These responses were abolished by application of cocaine to the ampulla of the fenestrated canal. It was further noted that sound-evoked eye movements could be produced without surgical interventions on the labyrinth when greater intensity

stimuli were used. Young et al⁴ subsequently showed that vestibular nerve afferents in the squirrel monkey respond to sound with phase-locking thresholds typically higher than 100-dB sound pressure level and rate-change thresholds 10 to 30 dB higher than intensities required for phase locking.

Clinical studies initially identified the Tullio phenomenon in patients with congenital syphilis. The histopathological features of the temporal bone in these cases was shown to be gummatous osteomyelitis and labyrinthine fistulae.⁵ Vestibular symptoms and/or eye movements evoked by sound have also been demonstrated in congenital deafness,⁶ Ménière disease,⁷ perilymph fistula,⁸ head trauma,^{7,9} Lyme disease,¹⁰ and cholesteatoma with semicircular canal erosion and fenestration.¹¹

PATIENTS AND METHODS

We identified 8 patients from April 1995 through July 1997 with vestibular symptoms evoked by sound and/or pressure applied to the external auditory canal. Seven of these patients also had eye movements in the plane of the affected superior semicircular canal evoked by these stimuli. Eye movements were documented with either video-oculography or, in 3 patients, magnetic field search-coil recordings. The temporal bone and labyrinth were examined with axial and coronal computed tomographic (CT) scans (Siemens Somatom Plus CT scanner, Siemens, Islin, NJ) using the following parameters: 1-mm slice thickness; 1-mm table incrementation; 330 mA; 120 kilovolt (peak); with a zoom of 3.5 and 6.

Three-dimensional binocular eye movement measurements were made using a dual search-coil technique. Patients gave informed consent for scleral search-coil recording through a protocol approved by the Joint Committee on Clinical Investigation of The Johns Hopkins University, Baltimore, Md. The head was centered in a 100-cm aluminum frame in which 3 magnetic fields were generated. The fields were orthogonal and oscillated at different frequencies. Details of the *in vitro* eye coil calibration procedure can be found in a previous study.¹⁷ Subjects wore an annulus that attached to the sclera with suction and moved with the eye. Contained within each annulus were 2 loops of wire in different orientations. A voltage was generated in proportion to the magnetic flux through the area enclosed by each loop. Signals from each loop were processed with a detector that determined the contribution of each magnetic field to the total voltage in the eye coil, thus specifying the orientation of the eye with respect to the frame. Eye position was sampled at 500 Hz. Data were analyzed using programs written in Matlab (The Math Works Inc, Natick, Mass) to calculate rotation vectors; they describe the axis about which the eye rotates, and the amount of rotation, relative to the straight ahead position in a head-coordinate frame. Rotation vectors were converted to degrees and plotted against time in the figures presented in this article.

The nomenclature for the description of torsional eye movements needs to be precisely defined. We have used terms that are referenced with respect to the patient's point of regard. This convention establishes a parallel between accepted methods for describing horizontal and vertical movements and those used for torsion. For example, a leftward eye movement refers to motion of the eyes to the patient's left, which when the examiner is standing in front of the patient, is to the examiner's right. Similarly, a conjugate clockwise torsional eye movement refers to intorsion of the superior pole of the patient's left eye and extorsion of the superior pole of the patient's right eye. Note that such an eye movement will appear counterclockwise to the examiner standing in front of the patient. All torsional eye movements in this study are described with respect to the patient's point of regard.

In pathological states closely resembling those that can result in the Tullio phenomenon, pressure in the ear canal with resultant movement of the tympanic membrane has been described to produce vestibular symptoms and signs. Hennebert¹² identified such findings in patients with congenital syphilis. Nadol¹³ described patients with long-standing Ménière disease who developed pressure-evoked vertigo and nystagmus. Temporal bone studies suggested that the cause was adhesions between the footplate of the stapes and structures in the membranous labyrinth such as the sacculus. Pressure in the external canal has also been reported to induce eye movements and vertigo in patients with perilymph fistula.¹⁴ Both the Tullio phenomenon and Hennebert sign occur together in some patients.¹⁵ Disruption of the bony labyrinth with concomitant development of a third "mobile window" into the vestibule has been conjectured to be a mechanism in such cases.¹⁶

REPORT OF CASES

CASE 1

For about the past 5 years, a 50-year-old man noted that objects in the visual surround seemed to move "like on a clockface" whenever he whistled or hummed a sound of a specific frequency (around 440 Hz). He also experienced vertical diplopia during these periods. Loud noises in the environment occasionally brought about the symptoms. He experienced no vestibular symptoms except under these specific circumstances. He denied hearing loss, tinnitus, or aural fullness. There was no history of otological disease or of ear surgery.

Examination revealed vertical-torsional movement of the eyes whenever he hummed or trilled a pitch with a frequency of about 440 Hz. Tonic motion of the eyes was upward and counterclockwise (intorsion of the superior pole of the right eye and extorsion of the superior pole of the left eye). Presentation of 430- to 500-Hz tones at 100-dB sound pressure level to the right ear evoked a similar eye movement. Tones outside of this frequency range, tones in the left ear, as well as positive or negative pressure insufflation in either external auditory canal resulted in neither an eye movement nor in a sensation of oscillopsia. There was no nystagmus after horizontal or vertical head shaking. The head thrust maneuver performed in the horizontal plane revealed a symmetric vestibulo-ocular reflex.

Figure 1 displays the vertical-torsional eye movements of the right eye recorded with a scleral search coil in response to a 450-Hz tone at 100 dB in the right ear. An upward, counterclockwise movement (intorsion of the superior pole of the right eye) began at a latency of 13 milliseconds after the onset of the tone. **Figure 2** shows vertical eye movements recorded from both eyes in response to the same tone. Upward movement of the right eye (ipsilateral to the stimulus) is greater than that of the left eye. Vertical displacement of the eyes is maintained during the period that the tone is sounded.

An audiogram revealed normal pure tone thresholds and speech discrimination scores bilaterally. Contralateral and ipsilateral acoustic reflexes were present

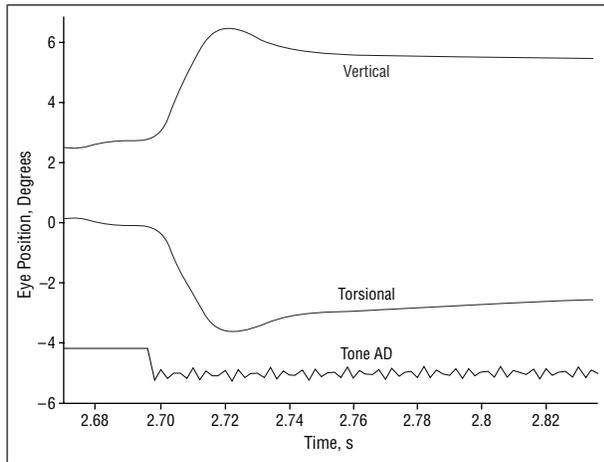


Figure 1. Vertical-torsional eye movements recorded from the right eye of patient 1 in response to a 450-Hz tone at 100 dB in the right ear. An upward, counterclockwise (intorsion of the superior pole of the right eye) occurs at a latency of 13 milliseconds after onset of the tone. AD indicates right ear.

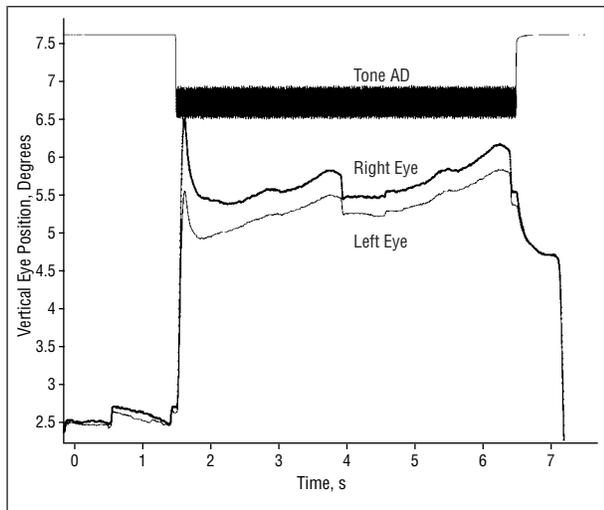


Figure 2. Vertical movements of the right and left eyes of patient 1 in response to a 450-Hz tone at 100 dB in the right ear. Both eyes move upward at the onset of the tone with elevation of the right eye being greater than that of the left. AD indicates right ear.

at normal levels. Neither eye movements nor vestibular symptoms were evoked by acoustic reflex testing. Results of cerebral magnetic resonance imaging (MRI) performed with and without gadolinium were normal. A CT scan of the temporal bones revealed a dehiscence of bone overlying the right superior semicircular canal. **Figure 3** shows 2 consecutive axial cut CT images that document dehiscence of the anterior aspect of the right superior canal along the floor of the middle cranial fossa. Results of serological tests for syphilis and Lyme disease were negative.

CASE 2

About 5 years before presentation a 27-year-old woman began to note oscillopsia induced by loud noises in the left ear. This symptom was initially noted when a telephone rang near to her left ear. She noted vertical movement of objects in the visual surround with these loud

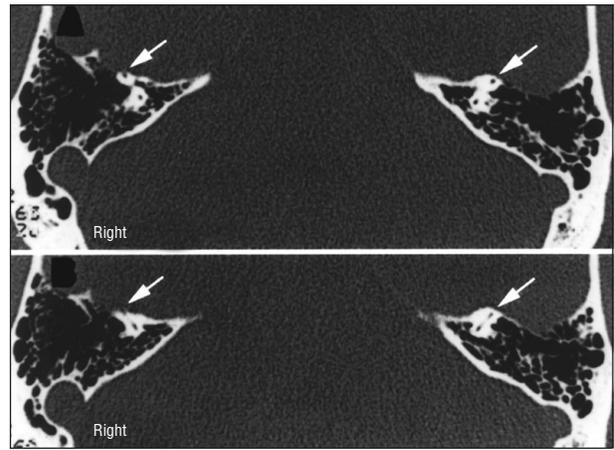


Figure 3. Consecutive 1.0-mm axial computed tomographic images showing dehiscence of the bone overlying the right superior semicircular canal (arrows) in patient 1.

noises. She also developed vertigo with Valsalva maneu-

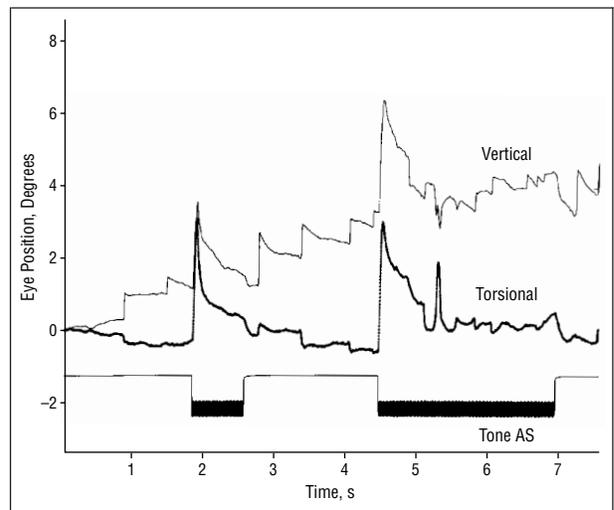


Figure 4. Vertical-torsional eye movements recorded from the left eye of patient 2 in response to a 500-Hz tone at 100 dB in the left ear. A stereotyped upward, clockwise movement of the left eye is noted at each onset of the tone. AS indicates left ear.

vers or pressure (applied by the patient or the examiner) in the left external auditory canal. Another otologist explored the left middle ear in 1993 to inspect for a perilymph fistula. The ossicular chain appeared normal and leakage of perilymph was not seen. Her symptoms were unchanged after this middle ear exploration. Her sound-evoked symptoms increased in severity over the next 2 years and she developed a persistent sense of disequilibrium and unsteadiness. She had no auditory complaints except for tinnitus evoked by leftward gaze.

On examination, the extraocular movements were intact. There was no spontaneous, gaze-evoked, or head shaking-induced nystagmus. Pure tones from 500 to 1500 Hz delivered through insert earphones at an intensity of 100 dB in the left ear produced an upward, clockwise movement of the eyes (**Figure 4**). Positive pressure insufflation into the left external canal induced a vertical-torsional nystagmus with slow phases upward and clockwise (**Figure 5**). This same

nystagmus was noted with Valsalva maneuver against pinched nostrils and there was a reversal of the nystagmus during the period after release. Valsalva maneuver against a closed glottis, jugular venous compression with pressure to the upper aspect of the neck in the region of the jugular foramen, and negative pressure insufflation of the left ear produced an oppositely directed nystagmus. Similar tones and insufflation of air in the right ear produced no eye movements or symptoms. A rapid neck movement to the left followed by return to midline position was noted at the onset of the tone.

Audiometry revealed normal pure tone thresholds and speech discrimination bilaterally. Electronystagmography showed symmetric caloric responses. Results of cerebral MRI were normal. Results of serological tests for syphilis and Lyme disease were negative. **Figure 6** shows coronal CT images of the temporal bones documenting a dehiscence of bone overlying the left superior canal. **Figure 7** shows 1.5-mm-thick axial CT images that iden-

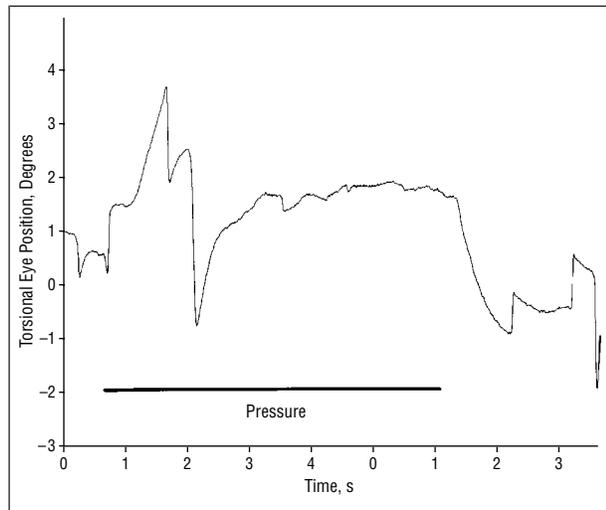


Figure 5. Torsional eye movements recorded in the left eye in response to positive pressure in the left external canal of patient 2. A clockwise movement (intorsion of the superior pole of the left eye) is noted with application of positive pressure through a pneumatic otoscope. This torsional deviation is maintained during the period of positive pressure and reverses when pressure is released.

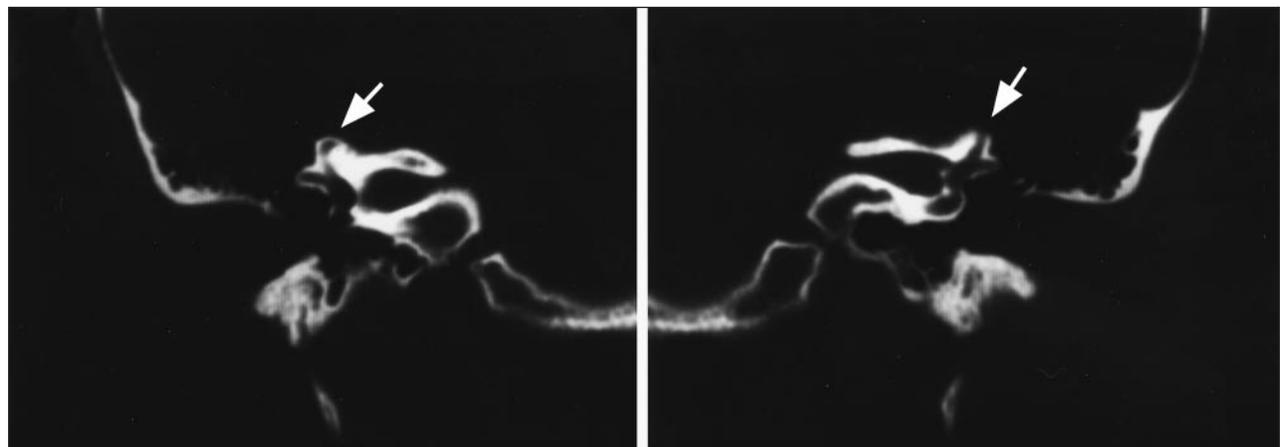


Figure 6. Coronal computed tomographic images at comparable levels of right (left) and left (right) temporal bones in patient 2. Left, An intact area of bone is noted overlying the right superior semicircular canal (arrow). Right, Dehiscence of bone is noted overlying the left superior semicircular canal (arrow).

tify absence of bone directly overlying the left superior canal.

Medical treatment with a low-salt diet, diuretics, and vestibular suppressants was ineffective at relieving her sound- and pressure-induced symptoms. Her most disturbing symptom was chronic disequilibrium that was exacerbated by head movements. She underwent plugging of the left superior semicircular canal through a middle cranial fossa approach. On elevation of the dura from the floor of the middle cranial fossa, a dehiscence of the superior canal in the area of the arcuate eminence was noted. There was an absence of bone overlying the membranous canal in this region (**Figure 8**). The superior canal was packed with a mixture of fascia, bone dust, and fibrin glue using a technique similar to that of Parnes and McClure.¹⁸ Cortical bone was placed over the plug. The bone was also eroded in the regions of the tegmen tympani and tegmen mastoideum (**Figure 9**). There was no evidence of granulation tissue or an active inflammatory process. Auditory brainstem response was monitored throughout the procedure and showed no change in wave form. Her hearing was unchanged (pure tone average, 10 dB; speech discrimination score, 100%) postoperatively. The symptoms and eye movements that were previously evoked by sound and pressure in the right ear resolved completely as did her sense of disequilibrium and unsteadiness. She continued to have nystagmus evoked by Valsalva maneuvers.

CASE 3

A 41-year-old woman noted the onset of disequilibrium and episodes of oscillopsia and vertigo provoked by pressure in her left ear beginning 2 years prior to her presentation. She associated the onset of these symptoms with significant straining and exertion while digging up a root in her garden. She reported no symptoms immediately following this activity, but 2 days later she began to experience unsteadiness and disequilibrium. She would lose her balance in situations that had never troubled her before such as walking around a corner. She could also elicit oscillopsia when applying pressure on her left tragus. Her evaluation and treatment prior to referral to

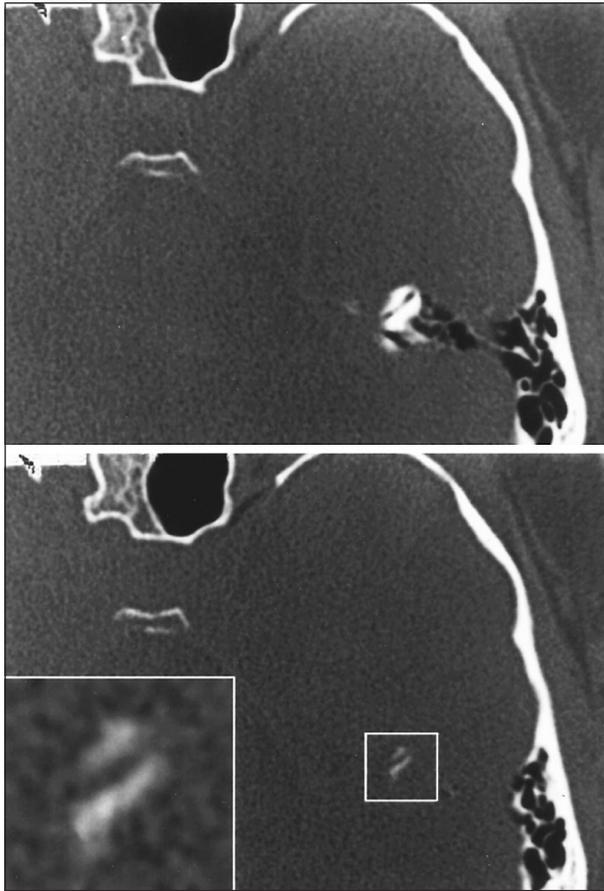


Figure 7. Axial computed tomographic images of the left temporal bone in patient 2. The bottom image is 1.0 mm superior to the top image. Bone is present anterior and posterior to the superior semicircular canal in bottom image, but there is a dehiscence of bone directly overlying the canal as shown in the inset.

our institution included a platform fistula test, the results of which were reportedly positive for the left ear followed by a left middle ear exploration that revealed no evidence of perilymph fistula. Her unsteadiness worsened despite empirical treatment with a combination product of triamterene and hydrochlorothiazide, acetazolamide, clonazepam, and lorazepam.

On the neuro-otological examination there was a right beating nystagmus after horizontal head shaking. A vertical-torsional nystagmus was evoked by positive pressure insufflation through a pneumatic otoscope on the left. Slow-phase components of the nystagmus were directed upward and clockwise. Similar stimuli delivered to the right ear did not lead to eye movements or symptoms. Tones in either ear, Valsalva maneuver against pinched nostrils or closed glottis, and jugular venous compression did not elicit eye movements.

An audiogram revealed normal pure tone thresholds and speech discrimination scores in both ears, and an auditory brainstem response was normal. Electrocochleography performed with transtympanic electrodes revealed normal potentials in response to tone bursts and clicks. Rotatory chair testing (velocity steps to 240°/s) with the head pitched downward such that the lateral canals were in the plane of rotation showed gain asymmetry suggestive of left labyrinthine hypofunction with right-



Figure 8. Operative photograph at the time of middle cranial fossa exploration in patient 2. The photograph is oriented from the surgeon's perspective sitting at the patient's head and looking onto the floor of the middle cranial fossa. The temporal lobe with dura intact is retracted superiorly and arrows indicate the dehiscence in the superior semicircular canal. The membranous canal is intact in this region.

ward slow phases evoked by leftward head rotations being 20% lower in maximum velocity than leftward slow phases evoked by rightward head rotations.¹⁹ Results of cerebral MRI were normal. Computed tomographic scan of the temporal bones revealed dehiscence of the bone overlying the left superior canal with images similar to those presented in patients 1 and 2.

The patient underwent plugging of the left superior semicircular canal through a middle cranial fossa approach. The dehiscent canal was packed in a manner similar to that in patient 2. Areas of bone erosion in the tegmen tympani and tegmen mastoideum were repaired with temporalis fascia and cortical bone harvested from the craniotomy bone flap. The patient had a left middle ear effusion and conductive hearing loss postoperatively that resolved within 6 weeks with return of hearing to the preoperative level. Her symptoms were completely relieved for about 2 months. She then experienced return of oscillopsia induced by pressure in the left external auditory canal followed several weeks later by return of the unsteadiness and disequilibrium that had been so disturbing to her preoperatively. The left middle fossa in the region of the arcuate eminence was surgically reexplored. The plug was noted to be intact but a further region of dehiscence in the bone overlying the superior canal was identified anterior and posterior to this plugged region. Fascia, bone dust, and fibrin glue were used to pack the canal on both sides of the previous plug. Auditory brainstem response was monitored throughout both plugging procedures and showed no change in morphologic features. Steroids were administered during the postoperative period. 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. Her steroid dosage was increased and therapy with acetazolamide was started for treatment of presumed endolymphatic hydrops resulting from



Figure 9. The dehiscent canal is packed with fibrin glue, bone dust, and fascia. A piece of cortical bone is placed over the plugged canal (black arrow). Dehiscences in the tegmen tympani and mastoidium are also noted (white arrow).

the surgical procedure. Pure tone thresholds and discrimination scores in the left ear improved initially but have continued to fluctuate. Her disequilibrium is less severe, as is her sense of imbalance induced by pressure in the external canal. Eye movements could not be induced by pressure in the affected ear during the first 3 months after this revision canal plugging procedure. A small eye movement in a direction similar to that before the revision procedure but of lower amplitude was noted during the fourth month after the procedure.

CASE 4

A 45-year-old woman began to note “vibrations” in her vision in association with loud noises in her right ear about 1 year prior to her presentation. She had experienced chronic disequilibrium and motion intolerance for several years before the onset of the sound-evoked symptoms.

Examination revealed intact extraocular movements with no spontaneous, gaze-evoked or head shaking–induced nystagmus. Tones of 250 to 1500 Hz administered at intensities of 90 to 110 dB in the right ear evoked upward and counterclockwise eye movements. Positive pressure insufflation in the right external canal and tragal compression induced similar eye movements. The external canals and tympanic membranes were normal. The Weber test for hearing at 512 Hz lateralized to the right ear. Results of the Rinne test were positive bilaterally.

The patient’s pure tone audiogram revealed a 20-dB conductive hearing loss at 250 and 500 Hz in the right ear. There was also a mild sensorineural hearing loss at 6000 and 8000 Hz in the right ear. Pure tone thresholds were lower than 10 dB for all frequencies in the left ear. The speech discrimination score was 100% bilaterally. Tympanograms were type A in each ear. A CT scan of the

temporal bones showed dehiscence of the bone overlying the right superior semicircular canal.

CASE 5

A 37-year-old man began to experience vestibular symptoms after a head injury in 1995. He fell off a bulldozer and hit the back of his head, resulting in a loss of consciousness for about 2 minutes. He has had headaches, constant disequilibrium, and vertigo evoked by loud noises or straining since that time. Loud noises produced an “eye flutter” that prevented him from focusing on stationary objects. He tended to stagger to the right if he was exposed to loud sounds while walking.

Examination revealed intact extraocular movements with no spontaneous, gaze-evoked or head shaking–induced nystagmus. Tones over a frequency range of 500 to 3000 Hz at intensities of 100 to 110 dB in the right ear evoked an upward and counterclockwise deviation of the eyes. Tones over the frequency range of 500 to 2000 Hz at intensities of 100 to 110 dB in the left ear evoked an upward and clockwise deviation of the eyes that was lower in amplitude than the eye movements evoked with tones in the right ear. Eye movements in the same direction as those evoked by sound, but lower in amplitude, were noted with positive pressure in the corresponding ear. No eye movements were noted in response to Valsalva maneuver against pinched nostrils or closed glottis.

An audiogram revealed a symmetrical, 15- to 20-dB conductive hearing loss at 500 and 1000 Hz in both ears. A mild to moderate high-frequency sensorineural hearing loss was also noted bilaterally. The tympanogram for each ear was type A. Caloric responses were normal in amplitude and without significant unilateral weakness or directional preponderance. Results of cerebral MRI and electroencephalography were normal. A CT scan of the temporal bones showed dehiscence of bone overlying both the left and the right superior semicircular canals.

CASE 6

A 33-year-old woman presented with a 7-year history of vertigo and disequilibrium. In January 1989 she had a week of marked imbalance and the sensation that the world was tilted. She had experienced arthralgias for several weeks before the onset of those vestibular symptoms and was diagnosed as having Lyme disease shortly after the onset of imbalance. She was treated with antibiotics intravenously, and recent Lyme titers have been negative. She has continued to experience episodes of disequilibrium in association with a sensation that the visual surround is tilting to the right. These sensations were substantially improved when she tilted her head to the left. Loud noises in her right ear worsened her unsteadiness and sensation of tilt. She has a history of migraine headaches that began at the age of 15 years.

Examination revealed an alternating hyperphoria on down and lateral gaze with the abducting eye higher. Clockwise beating torsional nystagmus was noted behind Frenzel lenses and on straight-ahead fixation. This

Table 1. Summary of Symptoms in Patients With Dehiscence of the Superior Semicircular Canal*

Patient/ Age, y/Sex	Affected Ear	Sound- Induced Symptoms	Pressure- Induced Symptoms	Chronic Disequilibrium
1/50/M	Right	+	-	-
2/27/F	Left	+	+	+
3/41/F	Left	-	+	+
4/45/F	Right	+	+	+
5/38/M	Both	+	+	+
6/36/F	Right	+	-	+
7/50/F	Right	+	-	-
8/41/M	Left	+	+	-

*Plus sign indicates positive; minus sign, negative.

torsional nystagmus was accentuated by hyperventilation. Presentation of a 2000-Hz tone in the right ear at intensities of 100 to 110 dB resulted in a substantial increase in her sense of tilting and vertigo. No eye movements were noted in specific relationship to the tones. Valsalva maneuvers and insufflation of air into the external canal produced neither signs nor symptoms.

An audiogram revealed normal pure tone thresholds and speech discrimination scores in both ears. Results of caloric testing were normal. Time constants and gains measured on rotatory chair testing were normal. A cerebral MRI scan revealed no abnormality. A CT scan of the temporal bones showed dehiscence of bone overlying the right superior semicircular canal.

CASE 7

Three months prior to presentation, a 50-year-old woman noted that noises such as a child crying or loud music produced oscillopsia and a spinning sensation. These symptoms went away when the noise ended but she continued to feel unsteady for several minutes afterward. A similar sensation could be brought on by vigorous coughing.

Examination revealed an upward and counterclockwise movement of the eyes in response to 1000- to 1500-Hz tones at intensities of 100 to 110 dB in the right ear. Neither eye movements nor symptoms were evoked by tones in the left ear. Valsalva maneuver against pinched nostrils resulted in a torsional nystagmus with slow phases in the counterclockwise direction. No eye movements were evoked by pressure in each external canal.

An audiogram revealed a moderate low-frequency sensorineural hearing loss in both ears. The speech discrimination score was 100% bilaterally. Caloric responses were normal. A CT scan of the temporal bones showed a dehiscence of bone overlying the right superior semicircular canal.

CASE 8

A 41-year-old man had experienced oscillopsia with loud noises in his left ear for as long as he could remember. He also had a history of migraine headaches and of unsteadiness occurring in association with the headaches.

Table 2. Summary of Signs in Patients With Dehiscence of the Superior Semicircular Canal*

Patient	Sound- Evoked Eye Movement	Sound- Evoked Head Tilt	Valsalva- Induced Eye Movement	Hennebert Sign	Postural Sway Induced by EAC Pressure
1	+	+	+	-	+
2	+	+	+	+	-
3	-	-	-	+	+
4	+	-	-	+	-
5	+	-	-	+	-
6	-	-	-	-	-
7	+	-	+	-	-
8	+	-	-	-	-

*EAC indicates external auditory canal; plus sign, positive; and minus sign, negative.

His migraines had been successfully controlled with the use of propranolol hydrochloride and acetazolamide.

On examination, upward and clockwise movement of the eyes was noted in response to 500- to 1000-Hz tones at 100 to 110 dB in the left ear. Similar tones in the right ear did not produce eye movements or symptoms. Positive and negative pressure applied in the external canals and Valsalva maneuvers did not evoke symptoms or signs. The tympanic membranes were mildly retracted.

The pure tone audiogram revealed a 10- to 15-dB conductive hearing loss for low frequencies in both ears. The tympanogram was type A bilaterally. Caloric responses were normal. Results of a cerebral MRI scan were normal. A CT scan of the temporal bones showed a dehiscence of bone overlying the left superior semicircular canal.

RESULTS

Signs and symptoms of sound- and pressure-induced eye movements in these patients are presented in **Table 1** and **Table 2**. No patient had a history of otological disease other than sporadic episodes of otitis media during childhood. The symptoms began in one patient (patient 5) after a closed head injury from a fall off a bulldozer and in another patient (patient 3) following straining from lifting and pulling. One patient (patient 6) had Lyme disease that was diagnosed and treated approximately 3 years prior to her development of vestibular symptoms.

The specific features of the induced eye movements are described in the "Report of Cases" section and accompanying search-coil records. Electronystagmography, performed in 5 patients, showed no localizing abnormalities. Results of rotatory chair testing, performed in 3 patients, were normal in 2 and showed gain asymmetries consistent with vestibular hypofunction in the affected ear in 1 patient. Audiometry showed normal pure tone thresholds and speech discrimination scores in 4 patients. The 1 patient with bilateral Tullio phenomenon (patient 5) had a 15- to 20-dB conductive hearing loss at 500 and 1000 Hz and a 20- to 30-dB high-frequency sensorineural hearing loss in each ear. Patient 4 had a 20-dB conductive hearing loss at 500 Hz in the affected

ear, and patient 8 had a 10- to 15-dB conductive hearing loss at 500 and 1000 Hz in both ears. Pure tone thresholds and discrimination scores were otherwise normal in these patients. Patient 7 had a mild low-frequency sensorineural hearing loss in both ears. Cerebral MRI with and without intravenous administration of gadolinium was performed in 6 patients and results were normal. Computed tomographic scans of the temporal bones revealed dehiscence of bone overlying the superior semicircular canal of the affected ear in each case. Fluorescent treponemal antibody titers were negative in these patients.

COMMENT

The symptoms and findings in these patients define a clinical syndrome resulting from dehiscence of bone overlying the superior semicircular canal. Manifestations of this syndrome include vertigo or oscillopsia induced by loud sounds and/or pressure that is transmitted to the inner ear from the external auditory canal or through Valsalva maneuvers. Seven of the 8 patients also had specific signs attributable to the canal dehiscence. These signs included eye movements in the plane of the affected semicircular canal evoked by sound or by maneuvers that result in pressure changes inside the labyrinth. These pressure-evoked signs were noted following Valsalva maneuvers or pressure transmitted via the external canal and tympanic membrane. Bone dehiscence of the affected superior semicircular canal was identified by CT scans in all 8 patients. One patient had sound-induced symptoms in the ear in which superior canal dehiscence was noted on CT scan but no sound- or pressure-evoked eye movements. We speculate that the thresholds for evoking eye movements in this patient were higher than those that could be safely used in clinical testing.

Five patients also experienced chronic disequilibrium that was the most disabling symptom in 2 of these patients. Since this symptom was alleviated by surgical plugging of the affected superior semicircular canal, it seems likely that it was due to the persistent effects of abnormal activation of vestibular receptors. An analogous situation might be the disequilibrium that can follow acute attacks of vertigo in Ménière disease or that can occur in some patients with benign paroxysmal positional vertigo.

Among the myriad of conditions that can cause dizziness and disequilibrium, this syndrome has specifically localizing signs and is a treatable form of vestibular disturbance. However, the identifying symptoms and signs may not be immediately apparent. Unsteadiness and persistent disequilibrium may be features of the syndrome that lead the patient to seek medical attention. Symptoms evoked by sound and/or pressure may only be identified on explicit questioning from the examiner. As indicated by the prior evaluations and interventions in the patients in this report, these symptoms may be thought to be caused by other processes such as perilymph fistula or psychosomatic illness. The 3-dimensional characteristics of the evoked eye movements provide a strong indication of a cause related to

the superior semicircular canal. The vertical-torsional eye movements that are characteristic of this syndrome are clearly noted on examination with Frenzel lenses when tones or pressure are administered. The diagnosis is confirmed with CT scanning of the temporal bones.

An opening in the bone overlying the superior semicircular canal creates a third mobile window into the inner ear that allows the canal to be responsive to sound and to changes in pressure in the membranous labyrinth. Similar symptoms can result from erosion of bone of the horizontal semicircular canal from cholesteatoma. The evoked eye movements in these patients are in the plane of the horizontal canal. Surgical fenestration of the horizontal canal that was formerly used for improvement of hearing in patients with otosclerosis can have similar effects. The relationship between planar and directional characteristics of the evoked eye movements and those known to be produced by excitation or inhibition of the superior semicircular canal help to define the pathophysiological features of the patients in this report.

In studies of the eye movements produced by selective stimulation of individual ampullary nerves, Cohen and colleagues²⁰⁻²² found that electrical stimulation of the superior semicircular canal ampullary nerve resulted in upward and torsional movement of both eyes. The torsional movement was such that the superior pole of the eye on the stimulated side was intorsion (directed nasally) whereas motion of the eye on the nonstimulated side was extorsion (directed temporally). With eyes in the primary position of gaze (centered in the orbit), the upward movement was greatest in the eye ipsilateral to the stimulated ampullary nerve. A similar disconjugacy of vertical eye movements (skew deviation) induced by angular rotations in the roll plane has been noted.²³ Eye movements recorded in our patients display these same features.

THE DIRECTION of torsional components of eye movements evoked by sound and pressure in patient 2 provides insight into the effects of these stimuli on the superior semicircular canal ampulla. Sound, positive pressure in the external canal, and Valsalva maneuver against pinched nostrils lead to a rise in middle ear pressure that, when transmitted to the vestibule, causes ampullofugal (excitatory) deflection of the superior canal cupula in the presence of a canal dehiscence (**Figure 10, A**). Valsalva maneuver against a closed glottis, jugular venous compression, and negative pressure in the external canal lead to inwardly directed pressure on the exposed area of membranous canal resulting in ampullopetal (inhibitory) deflection of the cupula (**Figure 10, B**). Sound and direct pressure-evoked eye movements were eliminated after plugging of the superior semicircular canal although a similar vertical-torsional nystagmus, of lower amplitude, was still noted with Valsalva maneuver against pinched nostrils. Persistence of this response may indicate that the plug is not completely effective at eliminating deflection of the cupula from a sustained change in middle ear pressure.

Other reports in the literature have described eye movements associated with the Tullio phenomenon or

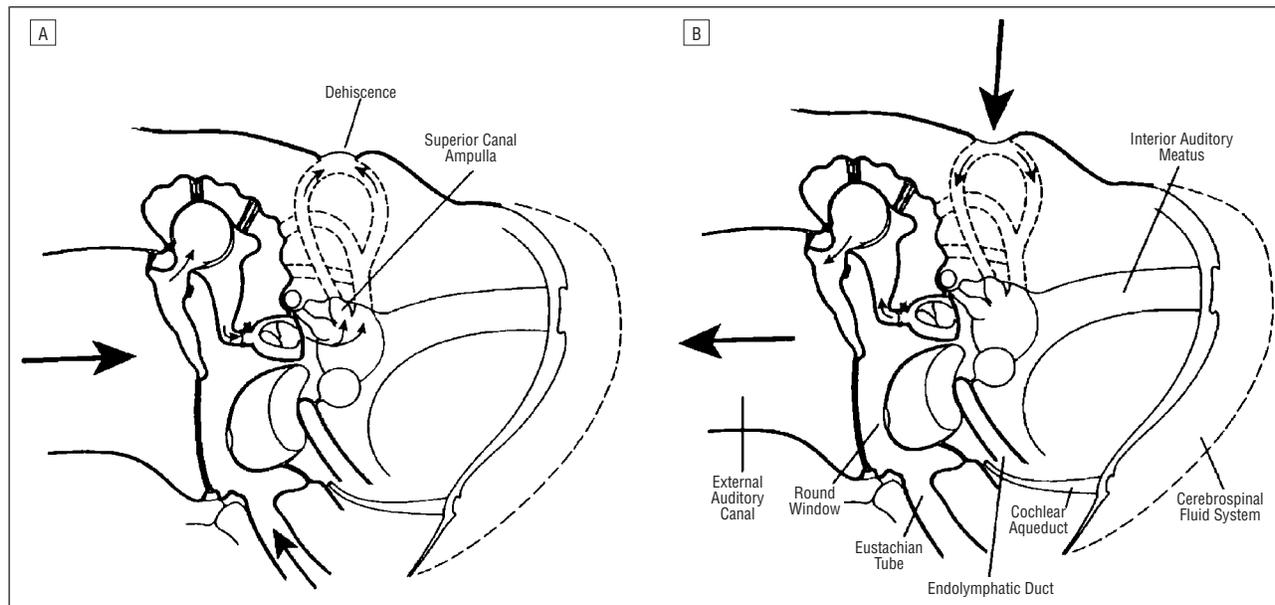


Figure 10. Schematic drawing of pressure changes in the middle and inner ear resulting from testing maneuvers used in this study.²⁴ A, Sound, positive pressure in the external canal, and Valsalva maneuver against pinched nostrils evoke pressure changes that result in expansion of the membranous canal with corresponding outward movement in the area of dehiscence. Such pressure within the membranous canal causes ampulofugal deflection of the superior canal cupula that results in excitation of vestibular-nerve afferents innervating the ampulla. B, Valsalva maneuver against a closed glottis, bilateral jugular venous compression, and negative pressure in the external canal result in inward movement in the area of dehiscence of the superior canal. Such pressure leads to ampulopetal deflection of the cupula and inhibition of the superior canal.

Hennebert sign similar to those in our patients, although correlation with an abnormality of the superior semicircular canal was not specifically evaluated. Bronstein et al²⁵ reported 3-dimensional eye movement recording in a 55-year-old woman with idiopathic Tullio phenomenon. The slow phases of the evoked eye movements were identical to those observed in our study. Cohen et al²⁶ reported vertical eye movements in response to sound in a 30-year-old man with progressive sensorineural hearing loss. Ostrowski et al²⁷ reported ocular torsion induced by pressure in the external canal in 3 patients. Colebatch et al²⁸ reported click-evoked vestibulocollic reflexes at a lower threshold from the symptomatic ear in a patient who developed the Tullio phenomenon after forceful sneezing.

Many features of the eye movements evoked by sound in patient 1 are similar to those reported in a patient with presumed otolith Tullio phenomenon.²⁹ The patient in the report of Dieterich et al²⁹ was a horn player who was believed to have subluxed his stapes footplate from a playing technique that involved a particularly large increase in posterior nasopharyngeal and middle ear pressure. He developed a sound-induced ocular tilt reaction (skew deviation with ipsilateral hypertropia, ocular torsion with the superior poles of the eyes moving away from the affected ear, and head tilt with ipsilateral ear up). Stimulation of the utricular nerve in cats has been shown to result in a similar profile of eye and head movements.³⁰ Presumed injury of the utriculus from a stapedectomy with a prosthesis extending too deeply into the vestibule resulted in an ocular tilt reaction in the opposite direction.³¹ The sound-evoked ocular tilt reaction in the patient described by Dieterich et al²⁹ was relieved by a procedure that decreased motion of the stapes footplate within the middle ear by insertion of compressed

Silastic foam between the crurae of the stapes. The characteristics of these evoked eye movements are similar to those observed in our patients. At least 2 interpretations are possible—the patient of Dieterich et al²⁹ may also have had a dehiscence of the superior canal or otolith (presumably utriculus) activation occurring as a result of superior canal dehiscence may have contributed to the tonic torsional deviations we observed during administration of tones and/or pressure to the affected ear in our patients.

Diagnosis of the underlying cause of symptoms with avoidance of stimuli that evoke these symptoms was sufficient treatment in 6 of our patients. The disabling disequilibrium that was present in 2 patients may have been caused by persistently abnormal activation of the superior semicircular canal due to the dehiscence. Plugging of the dehiscent canal relieved this symptom and alleviated the Tullio phenomenon and Hennebert sign. In consideration of a surgical approach for plugging the superior semicircular canal, it is important to realize that for the plug to be effective it must be placed between the dehiscence and the ampulla of the superior semicircular canal without leaving any open area of membranous canal between the plug and the ampulla. Sensorineural hearing loss occurred in 1 patient, and the long-term effects of such a plugging procedure remain to be determined.

The cause of superior semicircular canal dehiscence in these patients is not known. The incidence of this finding and its possible occurrence without symptoms remains to be determined. None of the patients in this series had prior otological disease other than sporadic episodes of acute otitis media during childhood. Head trauma and probably changes in middle ear or intracranial pressure with physical exertion preceded the onset of symptoms in 2 patients. Both patients undergo-

ing superior semicircular canal plugging had erosion of bone in the tegmen mastoideum and tympani. This bone erosion was similar to that reported by Gacek³² in patients with arachnoid granulations and cerebrospinal fluid otorrhea. No granulations were noted in the patients in our series, but it is conceivable that such granulations resulted in bone erosion and then resolved. It is also possible that dehiscence of bone overlying the superior semicircular canal may be congenital in some patients.

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