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ORIGINAL RESEARCH

Reversible Peripheral Vestibulopathy: The Treatment of Superior Canal Dehiscence

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OBJECTIVE: Superior canal dehiscence (SCD) is a recently described disorder that results from absence of bone over the superior semicircular canal. We have reviewed 30 cases of SCD found at our institution and report their presentation, workup, and response to therapy.

STUDY DESIGN: Retrospective chart review of all patients diagnosed with SCD from 1999 to 2004 at the University of Utah.

RESULTS: Thirty patients were identified with SCD. Patients presented with chronic disequilibrium (63%), Tullio's phenomenon (41%), pressure evoked vertigo (44%), hearing loss (30%), and pulsatile tinnitus (7%). ENG performed early in our series revealed abnormal nystagmus with sound presentation, Valsalva, or tympanogram; however, history and CT examination alone was used to identify this condition in most of our patients. Twenty-seven of the 30 patients had some symptoms related to SCD; the other 3 were found to have incidental SCD on CT examination. Of these patients, 14 had severe enough symptoms to warrant operative intervention. All, but one had resolution of their symptoms after completion of intervention.

CONCLUSIONS: Superior canal dehiscence is a highly treatable form of vestibulopathy once recognized. When patients present with typical symptoms, workup with CT is reliable and accurate. Surgical intervention results in reversal of symptoms in most cases with low morbidity.

EBM rating: C-4

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Superior semicircular canal dehiscence syndrome (SCD) is a recently described form of peripheral vestibulopathy in which absence of bone over part of the superior canal leads to significant vestibular symptoms.[1,2] Diagnosis is made by history, examination, and confirmation of missing bone over the superior semicircular canal by high resolution CT. Once recognized, this syndrome can be treated with surgical intervention, which leads to a high rate of symptom resolution and patient satisfaction.

Common symptoms include sound-induced vertigo (Tullio's phenomenon) and pressure/Valsalva-induced vertigo. These symptoms have been historically reported with vestibulopathy caused by Meniere's, Lyme disease, congenital deafness, congenital syphilis, and perilymph fistula.[3-6] With careful workup and a high degree of suspicion, SCD can be differentiated from these other vestibular disorders. Since the senior author (C.S.) began evaluating patients for SCD in 1999, we have regularly identified patients who meet the diagnostic criteria. To date, 30 patients with SCD have been identified at our institution.

In this article, we describe the patient presentation and results of our surgical technique for repair of this disorder in 17 consecutive operations in 14 patients.

MATERIALS AND METHODS

We performed a retrospective chart review of all patients identified at our institution with SCD from January 1999 to July 2004. Patients who presented with symptoms suspicious for SCD, such as Tullio's phenomenon, pressure evoked vertigo, or chronic disequilibrium, underwent CT for further evaluation. The CT performed was a 1 mm per

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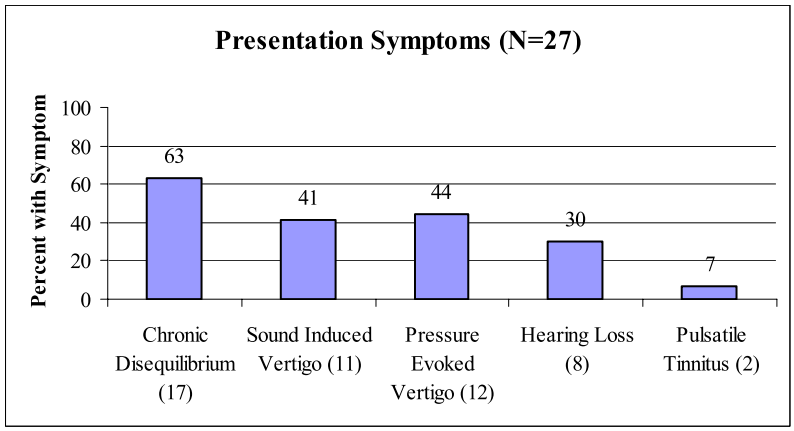

Figure 1 Presentation symptoms by frequency seen in SCD patients.

section, noncontrasted, coronal with a window width/ level setting of 4000/450, a protocol that gives excellent bony detail. We identified 30 patients who met our initial radiographic criteria for SCD, which was absence of bone over the superior semicircular canal on one coronal cut. Three patients had SCD noted on imaging examinations that were performed for other conditions and did not have symptoms felt to be related to SCD. Therefore, these patients were not included in this study.

Historical variables including age, sex, date of presentation, and symptoms at the time of presentation, were noted. Any reports of dizziness with loud sounds or with straining were recorded and the source of such symptoms noted. Physical examination findings and initial audiometric results were noted. Results of vestibular function testing (if performed) were also recorded.

Patients whose symptoms were debilitating were offered surgical intervention. Surgical repair was performed by accessing the dehiscence through a middle fossa approach, identifying the area of dehiscence, and covering the defect with hydroxyapatite bone cement as described by Smullen et al.[7] Early repairs were performed with fascia and CRS bone cement. More recent repairs were performed by placing temporalis fascia over the defect and covering this with "fast setting" hydroxyapatite bone cement (Norian CRS, Synthes Corporation, Paoli, PA). The bone cement was allowed to harden after placement and the temporal lobe retraction released to allow the dura to cover the repair.

Patients were followed for several months after surgery. Those with less than 3 months of follow-up were not included for analysis of response to surgical intervention. Patients were assessed for persistent symptoms, postoperative auditory function including 4 frequency average (0.5, 1, 2, and 3 kHz) of air and bone conduction, speech discrimination, and perioperative complications.

RESULTS

Thirty patients were diagnosed with SCD at our institution over the time period studied. As stated previously, the diagnosis was incidental in 3 patients. These patients were not symptomatic from the dehiscence and therefore not

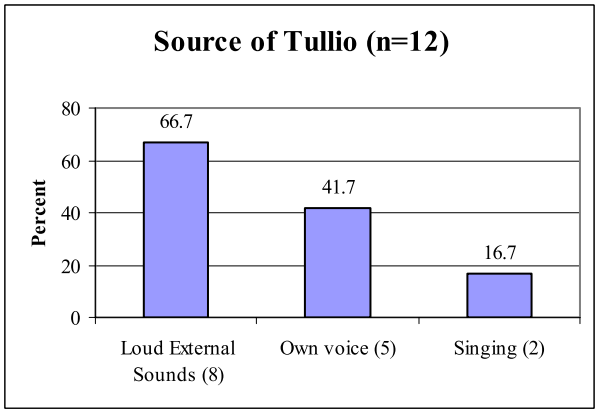

Figure 2 Source of noise-induced vertigo in patients with Tullio's phenomenon.

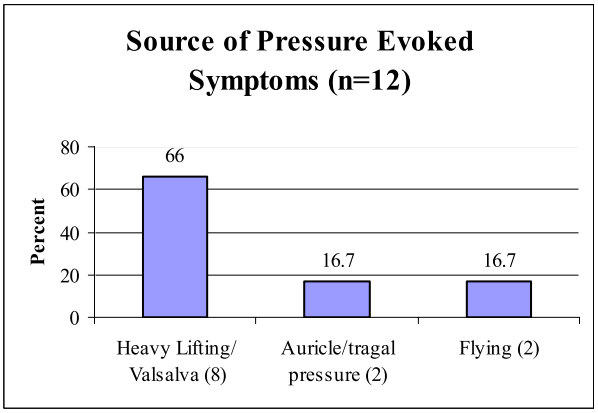

Figure 3 Source of pressure-evoked symptoms by percentage.

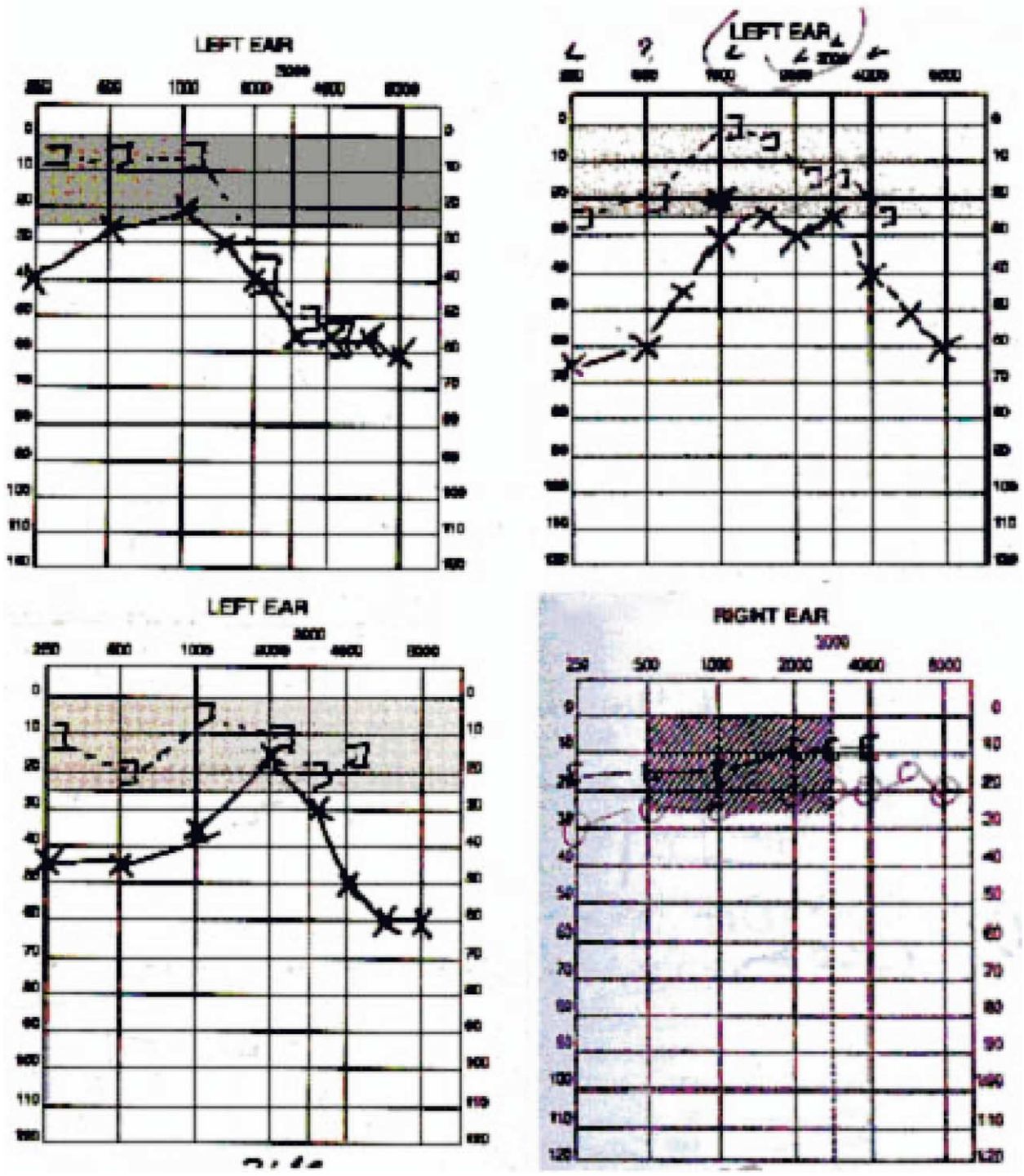

Figure 4 Audiograms from patients diagnosed with SCD on the same ear as a conductive hearing loss was identified. Low frequency air-bone gaps predominate.

included in this study, which left 27 patients to be studied. The average age at the time of diagnosis was 47.5 years. Eighteen were female and 9 were male. Presentation symptoms were diverse and are presented in Fig 1. The most common complaints were constant disequilibrium (63%), Tullio's phenomenon (41%), and vertigo with straining or pressure changes (44%). Hearing loss and pulsatile tinnitus were also common complaints. Hearing loss was seen in 30% of patients; 4 (15%) patients had a conductive hearing loss component attributed to the superior canal dehiscence. The source of the noise and pressure evoked symptoms are shown in Figs 2 and 3.

When conductive hearing loss was seen, it tended to be predominantly in the low frequencies. These audiograms are shown in Fig 4. This is a pattern that has been observed by Minor et al[8] as well in a recent publication. There did not seem

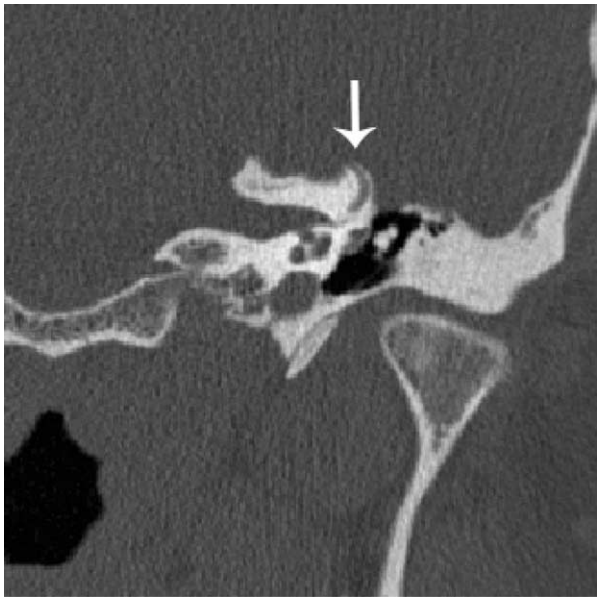

Figure 5 CT shows absence of bone over the superior semicircular canal.

to be the nadir at 2 KHz that is often seen with otosclerosis. Otherwise, there was no distinct pattern recognizable that could help to distinguish the conductive hearing loss in SCD from that seen with other ossicular chain abnormalities.

The diagnosis in each case was confirmed by CT (Fig 5). The SCD was seen on the left side in 14 (52%) patients, on the right side in 4 (15%) patients, and bilateral in 9 (33%) patients. Of the 27 symptomatic patients diagnosed, 14 were symptomatic enough to warrant surgery. Five of these patients were noted to have bilateral dehiscence on preoperative CT. The more symptomatic side was corrected first. Two of these patients had resolution of symptoms after repair of the first side. Later they became symptomatic on the opposite side and subsequently underwent surgery to repair the second side. Another patient with unilateral SCD initially had symptom resolution for 5 months with progressive return of symptoms after that. A repeat CT showed a persistent fistula on the same side and he underwent revision surgical repair. Therefore a total of 17 operations were performed on 14 patients. One patient had less than a month follow-up after surgery at the time of this article preparation and therefore was not included in analysis of treatment results, which leaves 16 operations on 13 patients for symptom control analysis. During the operation, all patients were found to have a fistula on the suspect ear except for one who had a blue line only. This patient had a CT that showed a "dehiscence" on 1 cut only. This is the reason we now insist on 2 consecutive cuts of 1 mm thickness that show dehiscence as our diagnostic criterion.

Of the 13 patients who underwent surgical intervention, 12 had postoperative audiometric data available for review. The average presentation air/bone pure tone averages, air-bone gap, and postoperative hearing results are shown in Table 1. Only 2 patients presented with >10 dB air/bone gap, which decreased but was not eliminated after surgery. One patient had a decrease in sensorineural function (4 frequency average) of 10 dB after surgery. This patient who required revision for persistent dehiscence had a decrease of 19 dB after the second surgery.

The patients' preoperative vestibular symptoms resolved after the surgery in 12 of the 13 patients. As stated above, 1 patient was initially cured but had recurrent symptoms 5 months after surgery that prompted a repeat CT. This revealed a recurrent dehiscence as a result of a shift of the bone cement on the previously operated side, and revision surgery was offered. This was performed and the patient has been symptom-free since. Therefore, only 1 patient had persistent vestibular symptoms after final definitive repair. Interestingly, this was the 1 patient who was noted to have a blue-line and not a true dehiscence of the superior semicircular canal at the time of surgery. She had typical symptoms including vertigo with loud sounds and straining, but she did not have objective physical examination findings such as nystagmus.

DISCUSSION

Superior canal dehiscence is a condition caused by the absence of bone over 1 or both of the superior semicircular canals that leads to dysfunction of the vestibular end organ as a result of altered fluid flow mechanics. The absence of bone allows for dural contact into the inner ear and altered fluid dynamics, the likely explanation for the frequently found Valsalva-associated symptoms. In addition, this "third window" has been theorized to be a cause of "inner ear conductive hearing loss" as well as the Tullio's phenomenon often seen in patients with this disorder.[9,10]

The characteristic presentation of SCD has been described as dizziness in the presence of loud noises or pressure-evoked dizziness from changes in atmospheric pressure, heavy lifting, or straining, the same symptoms that have been historically attributed to perilymphatic fistula. One of these symptoms was present in each patient described in a series of 17 patients by Minor.[1] Most, but not all, of our patients (17 of 26, 65%) presented with at least 1 of these 2 complaints. The most common complaint of

Table 1
Hearing presentation and results for all patients and the intervention subset

	Entire group (N=27, 2 bilateral)
Pre-op Air PTA	26.0 dB
Pre-op Bone PTA	20.9 dB
Pre-op ABG	5.1 dB
Number with ABG >10dB Pre Op	4 (14.3%)

PTA, Pure tone average; *ABG,* air bone gap; *dB,* decibel.

patients in our series was a sensation of constant disequilibrium (15 of 26, 58%). Seven (27%) of the patients who presented with chronic disequilibrium did not have either of the "typical" symptoms of noise or pressure-induced vertigo, which reinforces that a high degree of suspicion for SCD should be maintained for patients with chronic imbalance as well. Only 2 patients in our series did not have a presentation with 1 of these 3 complaints. One had an acute vertigo spell lasting 3 days with a sudden hearing loss and no long-term vestibular symptoms. The other had pulsatile tinnitus on the same side as the dehiscence as their sole complaint.

Pressure-evoked dizziness had a variety of precipitating causes in our study. Although most of the patients with this as a symptom complained of dizziness when they lifted of heavy objects or strained, 2 noted dizziness with flying. Noise-induced dizziness was caused most often by loud impulsive sounds, but in 2 of our patients, the dizziness was evoked with loud singing by the subject. One patient insisted that only certain notes (in the alto range) caused his dizziness.

When we suspect SCD as a possible cause of vestibulopathy, we proceed with high resolution coronal CT of the temporal bone to confirm or refute the diagnosis. All of our patients but 1 had the dehiscence confirmed at the time of surgery. This patient, who appeared to have a dehiscence on 1 cut, at the time of surgery had only a blue line of the superior canal. Because of this false-positive, we now confirm the diagnosis of SCD by the absence of bone over the superior canal on 2 consecutive cuts. In our series, this radiographic criteria would have changed the false-positive rate from 6% to zero.

It should be emphasized that this low false-positive rate is directly attributable to the fact that we are scanning patients with highly suspicious symptoms for SCD. If a larger group of subjects with vague symptoms were screened with CT scans, then false-positives could be found potentially leading to unwarranted surgical exploration. In a recent review, Belden[11] described that a CT scan with 1 mm collimation has a positive predictive value of only 50% in the general population to illustrate this potential pitfall. This is why our patients must have significant symptoms as well as evidence of bone over the superior canal on at least 2 consecutive cuts. If the symptoms are not typical, objective vestibular testing should be performed to look for nystagmus with sound or pressure presentation to verify the CT findings. Alternatively, ultra-high resolution CT scanning (0.5 mm cuts) may also be used to provide better specificity as proposed by Minor.[2]

Recently, we have questioned whether SCD could be accurately diagnosed by MRI as well. This question has arisen out of the fact that a large percentage of the patients we have treated for SCD present with nonspecific disequilibrium. Some of these patients have indications to undergo screening MRI, and it would be convenient for this test to be able to confirm or refute SCD. One recent study suggested that high resolution, T2 weighted, fast spin echo MRI had a sensitivity of 96% and a specificity of 98% for the identification of SCD.[10] We continue to use CT as our modality of choice for SCD evaluation.

Early in the series, we used ENG testing as an adjunct for diagnosis.[12] A total of 7 patients underwent modified ENG testing specifically designed to examine SCD. All of the patients eventually diagnosed with SCD by CT had at least 1 abnormality on ENG, typically abnormal nystagmus with loud sounds, tympanogram, or Valsalva. In our experience, however, we noted that high resolution CT had enough sensitivity and specificity to stand alone the definitive diagnostic test for SCD when typical symptoms were also present. If we suspect, based on history and examination, that the patient's symptoms are multifactorial or atypical, we still use ENG testing to help with the evaluation.

Many of the patients (48%) initially diagnosed at our institution did not require surgery. These patients had mild symptoms or additional diagnosis that compounded their symptoms. For instance, 2 of the nonoperative patients had the diagnosis of benign positional vertigo in addition to SCD during the workup. Although 1 had chronic disequilibrium and the other Tullio's phenomenon in combination with the positional vertigo, they both felt well enough after canalith repositioning that no further treatment was necessary.

Surgery is performed through the middle fossa approach with fascia and hydroxyapatite cement placed over the bony defect (Fig 6). We have had good success with this approach, but there can be technical hurdles. The bone cement we use is a fast setting type (Norian CRS, Synthes Corporation, Paoli, PA) that hardens in 5 minutes and cures readily in a wet environment. This cement is a clear improvement over the slower setting formulation that was used early in our series, which was prone to microfracture and did not set as well in a wet environment. Keeping the hardened cement in position can be difficult, however, as it can slip off the underlying bone when the dura is released. Providing a large surface area of contact between the cement and the middle fossa floor is helpful to prevent displacement. The area covered by the cement is significantly wider than the dehiscence, at least a centimeter in radius from the superior canal. Also, we have found that allowing appropriate time (at least 5 minutes) for hardening and not manipulating the dura after release of retraction to take a "second look," keeps the repair in place.

Surgical repair provides a high success rate (93%). The 1 patient that required revision had displacement of the cement. This has not happened again since we began use of the new cement formulation and the maneuvers described above. The 1 failure in our series was an error of diagnosis rather than a failure of the repair.

Bilateral dehiscence was noted in 9 (33%) of our patients. Of these, 5 went on to surgical repair initially on the side that was felt to be the most symptomatic. Although the patients were counseled that bilateral repair may be needed,

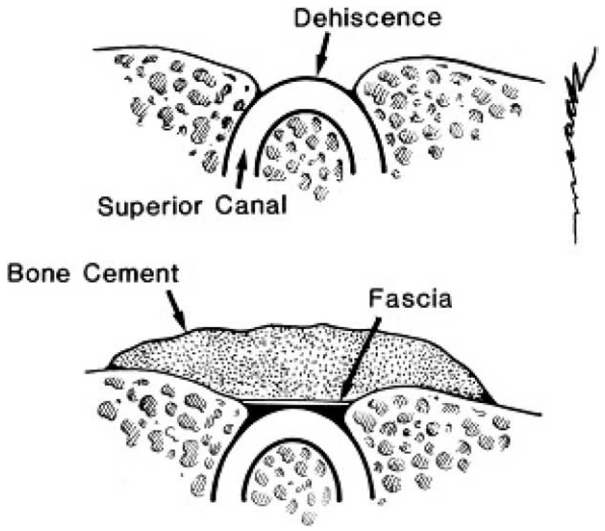

Figure 6 Repair of SCD involves restoration of the bone that separates the superior canal from the intracranial space. This is performed by placing temporalis fascia over the defect and placing bone cement over a broad span of the middle fossa bone over and around the fascia. (Reprinted by permission of C Shelton, MD.)

only 2 (40%) patients had recurrent symptoms that required repair of the opposite side. Both of these patients reported improvement of their symptoms after the initial repair and both are symptom-free after their second surgery. We continue to target treatment to the subjectively most symptomatic side, however, in the hopes that most of the patients with bilateral SCD will not go on to need repair of both sides.

Superior canal dehiscence is a highly treatable vestibulopathy once recognized. Up to 0.7% of individuals may have absence of the bone over the superior semicircular canal on the basis of a recent temporal bone survey by Carey et al.[10] Although it is unlikely that this many individuals have symptomatic disease, it is probable that SCD is under-diagnosed.

Besides imaging patients with classic symptoms of SCD, it is our current practice to image young patients with chronic disequilibrium and those with symptoms that suggest perilymph fistula. We have 3 patients who had pulsatile tinnitus on presentation that resolved after surgical repair in 2. For patients with pulsatile tinnitus with no source identified on the standard workup, we are now performing a CT scan to rule out SCD. The conductive hearing loss seen in SCD can also masquerade as otosclerosis. SCD should be considered in stapedectomy cases with an unchanged postoperative conductive deficit despite uncomplicated surgery. House et al[13] in 1980 described the term "inner ear conductive hearing loss" in reference to 4 patients who seemingly had normal ossicular chains at the time of stapedectomy and no other explanation for their hearing loss. In this article, they note that 3 patients did not have round window reflexes despite normal stapes mobility and states that there could be "some inner ear abnormality not visible" that cause this phenomenon. SCD is 1 possible explanation for a conductive hearing loss seen in the face of normal ossicular mobility.

CONCLUSION

Superior canal dehiscence is a reversible vestibulopathy, with a wide range of presentations, but characteristic imaging findings. The treatment for disabling cases involves closure of the defect with fascia and bone cement. Success of treatment is high, with resolution of symptoms in greater than 90% of patients who require surgical intervention.

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