Otic capsule dehiscence syndrome: Superior semicircular canal dehiscence syndrome with no radiographically visible dehiscence

P. Ashley Wackym, MD, FACS; Scott J. Wood, PhD; David A. Siker, MD; Dale M. Carter, MD

Abstract
We conducted a prospective longitudinal study of two cohorts of patients who had superior semicircular canal dehiscence syndrome (SSCDS); one group had radiographically confirmed superior canal dehiscence (SCD), and the other exhibited no identified otic capsule dehiscence on imaging (no-iOCD). We compiled data obtained from prospective structured symptomatology interviews; diagnostic studies; three-dimensional, high-resolution, temporal bone computed tomography; and a retrospective case review from our tertiary care referral center. Eleven adults and 1 child with SSCDS were identified, surgically managed, and followed. Six of these patients—1 man and 5 women, aged 29 to 54 years at first surgery (mean: 41.8)—had radiologically confirmed SCD. The other 6 patients—1 man, 4 women, and 1 girl, aged 13 to 51 years (mean: 32.2)—had no-iOCD. The 6 adults with SCD underwent surgery via a middle cranial fossa approach with plugging procedures. The 5 adults and 1 child with no-iOCD underwent round window reinforcement (RWR) surgery. One SCD patient developed no-iOCD 1.5 years after SCD surgery, and she subsequently underwent RWR surgery. Our main outcome measures were patient symptomatology (with video documentation) and the results of diagnostic studies. Other than the character of migraine headaches, there was no difference in preoperative symptomatology between the two groups. Postoperatively, resolution of SSCDS symptoms ultimately occurred in all patients. Both the SCD and the no-iOCD groups experienced a highly significant improvement in postural control following treatment (Wilcoxon signed rank test, p < 0.001). We conclude that the term otic capsule dehiscence syndrome more accurately reflects the clinical syndrome of SSCDS since it includes both superior semicircular canal dehiscence and no-iOCD, as well as posterior and lateral semicircular canal dehiscence, all of which can manifest as SSCDS. We have also included links to videos in which 4 of the SSCDS patients with no-iOCD in this study discussed their symptoms and the results of their surgery; these links are found in the "References" section in citations 12-15. Links to three other videos of interest are contained in citations 10, 11, and 24.

Introduction
In 1998, Minor et al became the first to describe superior semicircular canal dehiscence syndrome (SSCDS). However, a year earlier, Ostrowski et al described 3 cases of this clinical syndrome that were treated with perilymph fistula (PLF) repair; their 3 patients had not undergone high-resolution temporal bone computed tomography (CT) preoperatively. Subsequent to the latter report, the diagnosis of superior canal dehiscence (SCD) was confirmed by CT in 1 of these 3 patients (Timothy C. Hain, MD; personal communication; June 7, 2015). In 2005, Minor described sound- and/or pressure-induced vertigo, oscillopsia, and disequilibrium in a...
review of 65 cases of SSCDS. He reported that 54 patients (83%) had vestibular symptoms elicited by loud sounds, and 44 patients (68%) had pressure-induced (sneezing, coughing, and straining) symptoms. He also described decreased hearing thresholds for bone-conducted sounds (referred to as a pseudoconductive hearing loss or inner ear conductive hearing loss) and lower cervical vestibular evoked myogenic potential (cVEMP) thresholds.

In SSCDS, one of the most disturbing auditory symptoms is autophony, an unpleasant subjective discomfort that occurs while hearing one's own voice during phonation. Affected patients often describe their voice as "echo-like" or "resonant." Some patients with SCD can also hear their eyes move or eyelids blink. Bhutta postulated that patients who hear their eyes move do so via transdural transmission of extraocular muscle contraction.

Zhou et al considered SCD to be "a great otologic mimicker." In their series, they reported autophony and a blocked ear in 94% of patients and a pseudoconductive hearing loss in 86%. Arts et al found electrocochleographic (ECoG) evidence of endolymphatic hydrops in 14 of 15 ears with SCD; all 4 patients who underwent surgical repair experienced a resolution of their endolymphatic hydrops.

Black et al defined PLFs as "defects in the otic capsule or its windows that allow leakage of perilymph from the inner ear perilymphatic space into the middle ear spaces." Of course, before the description of SCD by Minor et al in 1998, it was not known that a defect in the superior canal could allow for leakage of perilymph from the inner ear perilymphatic space into the middle cranial fossa to create a PLF.

The literature contains conflicting reports about the frequency of symptoms and diagnostic test findings in patients with PLF. One illustrative summary that highlights the spectrum of the most common complaints from patients with PLF was published nearly a quarter-century ago by Black et al. We reanalyzed their dataset to determine the percentage of their patients who reported each of the 13 most common complaints; the three most common were disequilibrium, headache, and dizziness (figure 1). Other important clinical symptoms included cognitive dysfunction, nausea, vision disturbance, and subjective as well as objective hearing loss. This range of symptoms is extraordinarily similar to the spectrum of symptoms experienced by patients with SSCDS and vestibular migraine.

In 2009, one of the authors of this article (P.A.W.) began identifying SSCDS patients in his practice who had entirely normal findings on high-resolution temporal bone CT. These patients subsequently underwent round window reinforcement (RWR) surgery, and their symptoms resolved. Some of these patients could also hear their eyes move or eyelids blink. Based on Bhutta’s hypothesis that patients who hear their eyes move do so via transdural transmission of extraocular muscle contraction, these patients might have had an otic capsule defect in an area such as the modiolus that created a third window, just as is the case with SCD.

In this article, we describe our prospective study of 12 SSCDS patients who underwent surgical treatment and who were longitudinally followed for a mean of 3.0 years after their final surgery (range: 2.3 to 3.6 years). Of this group, 6 patients had radiologically confirmed SCD, and 6 had no radiologically identified otic capsule dehiscence (no-iOCD). We also present evidence to support our contention that SSCDS should be renamed otic capsule dehiscence syndrome (OCDS) since its syndromic symptoms and findings on nonradiologic objective test-
Patients and methods

**Patients.** Our study population was made up of 11 adults and 1 child with SSCDS (table 1). Six of these patients—1 man and 5 women, aged 29 to 54 years at first surgery (mean: 41.8)—had radiologically confirmed SCD. The other 6 patients—one man, 4 women, and 1 girl, aged 13 to 51 years (mean: 32.2)—had no-iOCD.

There was no difference in preoperative symptomatology between the two groups other than the character of their migraine headaches (table 2). A history of trauma was common in this series; trauma was reported in 4 of the 6 SCD patients and in 5 of the 6 no-iOCD patients (table 1). None of the SCD patients and 4 of the no-iOCD patients had been assigned a neurobehavioral or psychiatric diagnosis before referral (table 1). Unilateral disease was present in 5 SCD patients and 4 no-iOCD patients.

**Diagnostic testing.** Comprehensive testing was performed pre- and postoperatively with the tuning fork, audiometry, ECoG, cVEMP assessment, vestibular autorotation testing (VAT), moving platform pressure testing, and computerized dynamic posturography.

**Tuning fork testing.** As a screening tool for patients with SSCDS/OCDS symptoms, a low-frequency tuning fork was applied to their knees and elbows, and they were asked if they could hear or feel the vibration in their head. Both 128- and 256-Hz tuning forks were used.

**Audiometry.** Pure-tone audiometry was performed over the frequency ranges of 250 to 8,000 Hz for air conduction and 250 to 3,000 Hz for bone conduction. Testing was performed in a sound-proof booth. Appropriate masking was used for bone conduction and, when needed, for air conduction. Tympanometry was also performed, and acoustic reflexes were tested for ipsilateral and contralateral presentation of tones.

**ECoG.** Preoperative ECoG was performed with gold foil tiptrodes (Etymotic Research; Elk Grove Village, Ill.), which were placed adjacent to the tympanic membrane in the external auditory canal and stabilized on the foam tip of the insert audio transducer. Unfiltered clicks of 100 μsec duration were presented at an intensity of 85 dB nHL. Two replications of averaged responses elicited by 1,500 clicks presented at a rate of 11.7/sec were obtained. Responses were band-pass filtered (20 to 1,500 Hz) and averaged, and the summating potential to action potential (SP/AP) ratio was calculated. An SP/AP ratio of greater than 0.4 was defined as abnormal for purposes of this study, based on commonly used standards for clinical testing.16

**Acoustic cVEMP stimuli and recording techniques.** A commercial auditory evoked potential software system (v. 6.2.1d; Bio-Logic Systems; Mundelein, Ill.) was used for acoustic cVEMP testing. Sound stimuli were delivered monaurally via an intra-auricular transducer with foam earphones (E-A-R Link Insert Earphones; E-A-R Auditory Systems, Indianapolis) as described previously.17

During the recording protocol, patients were seated upright. The skin in the areas of electrode placement was cleansed with alcohol preps prior to electrode placement. The cVEMP measurements were recorded on disposable, self-adhesive, pre-gelled electrodes (Red Dot Ag/AgCl electrodes; 3M Canada; London, Ont.) and lead wires from Bio-Logic. The electrode montage consisted of an active electrode on the top third of the sternocleidomastoid muscle, a reference electrode at the sternoclavicular junction, and a ground electrode on the sternal notch.

During the cVEMP instruction, patients were asked to rotate their head toward the shoulder contralateral to the stimulus, and tilt their head approximately 30° to maximize the contraction of the sternocleidomastoid muscle. The clinician applied the maximum amount of manual resistance that each patient could tolerate while visually confirming the muscle contraction during stimulus delivery.

During the cVEMP measurements, air-conducted stimuli were delivered with a 1,000-Hz, 100-dB-nHL tone burst of positive polarity at a repetition rate of 4.3/sec (a 2 msec rise/fall time and a 2 msec plateau). Evoked myogenic potentials were amplified by 1,000 and band-pass filtered (10 to 1,500 Hz). An average of approximately 80 to 150 sweeps were made per test.

The response parameters were defined as (1) the VEMP p13 potential being the first distinctive trough in the waveform, anticipated to occur at approximately 10 to 14 msec following the stimulus, and (2) the n23 potential being the first distinctive peak in the waveform, occurring at approximately 19 to 23 msec after stimulus onset. Peak-to-peak amplitude was calculated with the Bio-Logic software after peaks were labeled and the amplitude difference between the two peaks was measured. The threshold was defined as the lowest dB SPL at which a p13 and n23 response could be recorded. For reporting purposes, the cVEMP was considered positive when an increase in amplitude and decrease in threshold were observed.

**VAT.** The horizontal and vertical vestibulo-ocular reflexes (VORs) of each patient were tested by the VAT, which is a computerized test based on active head movements over a frequency range from 2 to 6 Hz. At
<table>
<thead>
<tr>
<th>Pt.</th>
<th>Sex</th>
<th>Age at first surgery, yr</th>
<th>Current age, yr*</th>
<th>Diagnosis at initial referral</th>
<th>Trauma</th>
<th>Surgery 1</th>
<th>Surgery 2</th>
<th>Surgery 3</th>
<th>Length of follow-up, yr</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>SCD group</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>F</td>
<td>29.45</td>
<td>32.07</td>
<td>Multiple sclerosis</td>
<td>None</td>
<td>Left SCD</td>
<td>–</td>
<td>–</td>
<td>2.62</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>32.10</td>
<td>34.68</td>
<td>Post-traumatic ELH</td>
<td>Motorcycle accident</td>
<td>Left SCD</td>
<td>–</td>
<td>–</td>
<td>2.58</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>32.18</td>
<td>35.97</td>
<td>ELH, dizziness</td>
<td>None</td>
<td>Right SCD</td>
<td>Left SCD</td>
<td>Right RWR</td>
<td>2.29 (1.5 yr after right SCD)</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>50.33</td>
<td>53.94</td>
<td>Imbalance, falling</td>
<td>Motor vehicle accident</td>
<td>Right SCD</td>
<td>–</td>
<td>–</td>
<td>3.62</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>52.48</td>
<td>55.96</td>
<td>Labyrinthitis</td>
<td>Motor vehicle accident</td>
<td>Left SCD</td>
<td>–</td>
<td>–</td>
<td>3.48</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>54.08</td>
<td>57.70</td>
<td>Migraine, tilting</td>
<td>Motor vehicle accident</td>
<td>Right SCD</td>
<td>–</td>
<td>–</td>
<td>3.62</td>
</tr>
<tr>
<td><strong>No-iOCD group</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>13.15</td>
<td>15.62</td>
<td>Autism as a child, migraine</td>
<td>Motor vehicle accident</td>
<td>Right RWR</td>
<td>–</td>
<td>–</td>
<td>2.47</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>24.73</td>
<td>27.69</td>
<td>Benign ICH; VP shunt ×2</td>
<td>None</td>
<td>Right RWR</td>
<td>–</td>
<td>–</td>
<td>2.97</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>30.03</td>
<td>33.16</td>
<td>Autoimmune inner ear disease</td>
<td>Onset after vaginal delivery of a child</td>
<td>Right RWR</td>
<td>Left RWR</td>
<td>–</td>
<td>2.85</td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>36.65</td>
<td>39.63</td>
<td>Traumatic brain injury</td>
<td>Bicycle accident, TB fx, mandible fx</td>
<td>Left RWR</td>
<td>–</td>
<td>–</td>
<td>2.98</td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>36.78</td>
<td>39.93</td>
<td>Traumatic brain injury</td>
<td>7 concussions, most recent from skiing accident</td>
<td>Right RWR</td>
<td>Left RWR</td>
<td>–</td>
<td>3.03</td>
</tr>
<tr>
<td>12</td>
<td>F</td>
<td>51.63</td>
<td>54.76</td>
<td>Traumatic brain injury</td>
<td>Motor vehicle accident</td>
<td>Left RWR</td>
<td>–</td>
<td>–</td>
<td>3.14</td>
</tr>
</tbody>
</table>

*As of June 1, 2015.

Key: SCD = superior canal dehiscence; ELH = endolymphatic hydrops; RWR = round window reinforcement; ICH = intracranial hypertension; VP = ventriculoperitoneal; TB = temporal bone; fx = fracture.
frequencies higher than 2 Hz, the VORs represent the primary systems for ocular gaze fixation because other ocular movement systems (e.g., smooth pursuit) are minimally effective in this range of frequencies.

For the VAT protocol, patients were seated and fitted with conventional electro-oculographic (EOG) electrodes. Then a lightweight headband was attached to a rotational velocity sensor and an EOG amplifier. Horizontal eye movements were recorded by bilateral electrodes positioned at the outer canthi and by a reference electrode positioned above the bridge of the nose. Vertical eye movements were recorded by electrodes placed above and below one eye. Head velocity was recorded by a calibrated velocity sensor that was fixed to the headband. A computer-generated tone was used as an audible cue to direct the frequency of head motion while the computer program swept the frequencies from 0.5 to 6.5 Hz during the 18-second test epoch. Two instructions were given: (1) “stare at the wall-mounted target” (a 1-cm disk) and (2) “move your head smoothly from side to side in time to the computer-generated tone.”

After a 30-second rest, the same procedure was performed twice more for a total of three evaluations of horizontal head movements, and then it was performed three more times with vertical head movements in a “nose up, nose down” direction. Eye position and head velocity data were amplified and digitized. Data from the first 6 seconds were used for EOG calibration. Gain and phase were computed during the final 12 seconds of the test epoch. In brief, gain is defined as the eye velocity amplitude divided by the head velocity amplitude. Phase is the time lag in degrees of the eye velocity in relation to the head velocity. Asymmetry is the amount of drift of the eye toward one side. All three characteristics are frequency-dependent. An ideal VOR result would be expressed as gain = 1 and phase = 180° with no asymmetry.

An inability of eye velocity to follow head velocity can indicate pathology when gains and phases differ from normal. Eye drifts to the right or left might indicate pathology when they occur systematically toward one side. A VAT result is considered clinically abnormal if two or more means and standard deviations of gain or phase datapoints show error bars that are clearly separable from those of the normal group in one or more of the four plotted graphs: horizontal and vertical, gains and phases.

Asymmetry plots are generated from each patient’s data by determining the ratio of the eye position deviation from the straight-ahead position and the amount of spectral energy at each frequency as a percentage. This is ascertained by Fourier analysis. Asymmetry in VORs suggests that the number of neural impulses per unit of time that contributes to the extraocular muscles is lower on one side, which causes the eye to drift in the orbit to that side during active head movement. Asymmetry suggests the presence of a unilateral lesion, and the direction of the eye drift is toward the side of the lesion.

Moving platform pressure test. Most patients underwent moving platform pressure testing (fistula test) preoperatively as described by Black et al (table 3).5,18

Computerized dynamic posturography. Postural performance was measured in 5 SCD patients (1 patient exceeded the weight limit for the test platform) and all 6 no-iOCD patients on a movable platform before and after surgical intervention. This test was performed on an EquiTest platform (NeuroCom International; Clackamas, Ore.). Patients stood in the center of the platform with their shoes off, their feet shoulder-width apart, and with the medial malleolus aligned with the rotational axis of the support surface and visual surround.

The support surface was made up of a dual forceplate with four force transducers (strain gauges) mounted symmetrically to measure the distribution of vertical forces sampled at 100 Hz. Patients were instructed to maintain an upright stance with their arms folded and their head in a natural upright orientation. Center-of-mass sway angles were derived from the anteroposterior and mediolateral center-of-pressure positions with a low-pass Butterworth filter (2nd order, cutoff frequency at 0.85 Hz), with the center of mass estimated at 55% of the patient’s height.19

During platform testing, sensory organization tests (SOTs) were administered. SOTs pose a set of increasingly challenging conditions to assess a patient’s ability to make effective use of visual, vestibular, and somatosensory information in order to maintain an upright stance. Testing is done under six sensory conditions:

- 1: fixed support surface, eyes open and fixed on a target;
- 2: fixed support, eyes closed;
- 3: fixed support, vision sway-referenced;
- 4: support sway-referenced, eyes open and fixed;
- 5: support sway-referenced, eyes closed; and
- 6: support sway-referenced, vision sway-referenced.20

During some SOTs, the support surface and/or the visual surround was rotated in direct proportion to the patient’s instantaneous anteroposterior sway, which is referred to as sway referencing. Postural sway was measured during 20-second trials; testing included combinations of two somatosensory conditions (fixed-
Table 2. Patient history, symptoms, and physical findings before surgical intervention

<table>
<thead>
<tr>
<th>Pt.</th>
<th>SCD group</th>
<th>No-IOCD group</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>None</td>
<td>Increased headache, nausea, vomiting</td>
</tr>
<tr>
<td>2</td>
<td>Tilting, dizziness,† nausea, headache</td>
<td>Increased headache, nausea, vomiting</td>
</tr>
<tr>
<td>3</td>
<td>Dizziness,† eyes jumping, nausea, cough-induced dizziness</td>
<td>Lightheadedness, waviness, worse cognitive dysfunction</td>
</tr>
<tr>
<td>4</td>
<td>Dizziness,† nausea</td>
<td>Irritation; no dizziness or nausea</td>
</tr>
<tr>
<td>5</td>
<td>Tilting, nausea</td>
<td>Light-headedness, waviness, worse cognitive dysfunction</td>
</tr>
<tr>
<td>6</td>
<td>Increased headache, occasional tilting</td>
<td>Dizziness†</td>
</tr>
<tr>
<td>7</td>
<td>None</td>
<td>Increased headache, nausea, vomiting</td>
</tr>
<tr>
<td>8</td>
<td>Tilting, nausea</td>
<td>Increased headache, and pain</td>
</tr>
<tr>
<td>9</td>
<td>Increased headache, nausea, vomiting</td>
<td>Increased headache, and pain</td>
</tr>
<tr>
<td>10</td>
<td>Dizziness†</td>
<td>Increased headache, and pain</td>
</tr>
<tr>
<td>11</td>
<td>Increased headache and pain</td>
<td>Increased headache and pain</td>
</tr>
<tr>
<td>12</td>
<td>Tilting, nausea</td>
<td>Tilting, nausea</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th></th>
<th>Hearing internal sounds</th>
<th>128- and 256-Hz tuning fork test*</th>
<th>Cognitive dysfunction</th>
<th>Spatial disorientation</th>
<th>Anxiety</th>
<th>Nausea</th>
<th>Migraine</th>
<th>Treated as a vestibular migraine patient</th>
</tr>
</thead>
<tbody>
<tr>
<td>SCD group 1</td>
<td>Heartbeat</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Constant</td>
<td>Frequent ocular migraine, vestibular migraine ×2</td>
<td>Yes</td>
</tr>
<tr>
<td>2</td>
<td>Eyes moving, breathing</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Mild</td>
<td>3 or 4 x per wk, rare vestibular migraine with rotational vertigo, light-sensitive</td>
<td>Yes</td>
</tr>
<tr>
<td>3</td>
<td>Autophony</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Constant</td>
<td>Frequent</td>
<td>No</td>
</tr>
<tr>
<td>4</td>
<td>Eyes moving</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Mild</td>
<td>Frequent, severe, light-sensitive</td>
<td>Yes</td>
</tr>
<tr>
<td>5</td>
<td>Heel strike</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>2x per mo</td>
<td>No</td>
</tr>
<tr>
<td>6</td>
<td>Eyes moving, heartbeat, autophony</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Constant</td>
<td>Daily, light-sensitive</td>
<td>No</td>
</tr>
<tr>
<td>7</td>
<td>Heartbeat, chewing</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Yes, worse on an elevator</td>
<td>24/7, light-sensitive</td>
<td>Yes</td>
</tr>
<tr>
<td>8</td>
<td>Eyes moving, heartbeat, autophony</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Constant</td>
<td>24/7, light-sensitive</td>
<td>Yes</td>
</tr>
<tr>
<td>9</td>
<td>Autophony</td>
<td>Positive</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Constant</td>
<td>24/7, rare ocular migraine, light-sensitive</td>
<td>Yes</td>
</tr>
<tr>
<td>10</td>
<td>Autophony</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Mild</td>
<td>24/7, rare ocular migraine, light-sensitive</td>
<td>Yes</td>
</tr>
<tr>
<td>11</td>
<td>Autophony</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Intermittent</td>
<td>24/7, light-sensitive</td>
<td>Yes</td>
</tr>
<tr>
<td>12</td>
<td>Eyes moving, heel strike</td>
<td>Positive</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Strong</td>
<td>24/7</td>
<td>Yes</td>
</tr>
</tbody>
</table>

* Ability to hear or feel vibration in the head when the tuning fork was applied to the knees and elbows.
† The gravitational receptor asymmetry type of vertigo (e.g., a rocky, wavy, or tilting sensation; a feeling as if on a boat; a feeling of being pushed; or a sense of the floor falling out from beneath oneself).
support and sway-referenced support) and three visual conditions (eyes open, eyes closed, and sway-referenced vision). Three trials of each condition were performed. The anteroposterior peak-to-peak sway angle, \( q \) (in degrees), was used to compute a continuous equilibrium (EQ) score, as follows:

\[
EQ = \left(1 - \left(\frac{q}{12.5}\right)\right) \times \% \text{ trial completed},
\]

where 12.5° was the maximum theoretical peak-to-peak anteroposterior sway and normalized values ranged between 0 and 100. Falls were recorded when patients moved their feet, began to take a step, or raised their arms. In view of the skewed distribution of EQ scores, the nonparametric repeated-measures Wilcoxon signed-rank test was used to compare pre- and postoperative posture performance, and the independent samples Mann-Whitney \( U \) test was used to compare across SCD and no-iOCD groups using Statistical Package for the Social Sciences software (SPSS, v. 22; IBM; Armonk, N.Y.).

**CT of the temporal bone.** Patients underwent temporal bone CT on a helical high-resolution scanner (Somatom Sensation 64-slice scanner; Siemens; Malvern Pa.) with a collimation of \( 12 \times 0.6 \) mm and a reconstruction increment of 0.3 mm. Axial imaging was obtained with reconstructions in sagittal and coronal planes. The images were optimized with a very sharp kernel and a specific window level dedicated to the inner ear (Siemens PLM Software). Next, the axial 0.6-mm raw dataset was loaded onto a viewer (AquariusNET; TeraRecon; Foster City, Calif.) in three-dimensional (3-D) mode. Using the 3-D controls, the left and right superior semicircular canals were manipulated to a “best view in plane” position with the circumference of the canal. The entire bony otic capsule, including the superior semicircular canals, was then evaluated with two different 3-D rendering modes. The first was a gray-scale, minimum-intensity projection mode at 1-mm thickness. The second was a color 3-D volume-rendering mode, also at 1-mm thickness.

The character and size of the dehiscence were measured using the best-view-in-plane images on the workstation. The bone overlying the superior semicircular canal on each side and with each 3-D rendering mode was characterized as either normal, thin, small (SCD ≤2 mm), medium (2 to 4 mm), or large (≥4 mm). For reporting purposes, an image was classified as normal if no dehiscence could be seen in any of the three semicircular canals or anywhere else in the bony otic capsule.

**Magnetic resonance imaging.** Magnetic resonance imaging (Tim Trio 3.0 T MRI; Siemens) was performed in 1 patient, a 32-year-old woman (patient 3) who developed late no-iOCD and a recurrence of her symptoms, to determine if her superior semicircular canals remained plugged. The semicircular canal sequence used to determine if a semicircular canal was patent or plugged was CISS (constructive interference in steady state) 0.6-mm axial acquisitions, which were then evaluated in both 2-D and 3-D volume rendering on the Tera AquariusNET viewer. The 3-D volumes were then evaluated with maximum-intensity projection slabs ranging from 10 to 20 mm. These high-resolution sequences were used to determine whether fluid was present within the superior semicircular canals.

**Technique for SCD surgery.** The same surgical technique was used for all 6 SCD patients. After intravenous administration of 10 mg of dexamethasone and 0.5 g/kg of mannitol, surgery via a traditional middle cranial fossa approach with a craniotomy centered on the zygomatic root and a craniectomy to the skull base was performed. The dura was elevated with an Adson peristomial elevator, and a Fisch retractor was placed, with the retractor tip just past the petrous ridge. With microsurgical techniques, the superior canal was inspected. If the dehiscence was not seen on the superior aspect of the canal, further dural elevation and subsequent use of a Buckingham mirror to identify a dehiscence was completed.

The canal was plugged with temporalis fascia or periosteum. The superior canal was resurfaced with hydroxyapatite bone cement. If the ossicles were in contact with the herniated temporal lobe and dura, Gelfoam was used to fill the middle ear. Gelfoam was also used to fill all of the remaining temporal bone defects.

Acellular hydrated dermis was then trimmed to fit the surface of the temporal bone, and titanium mesh was placed over the defect. Then an additional piece of dermis was used to line all of the exposed dura. If there were any dural defects, the dura was repaired with either a fascia graft or a medial graft fashioned from the hydrated dermis. A single piece of Gelfoam was used to cover all of the exposed dura at the craniotomy/craniectomy site, and then titanium mesh was secured to the skull. Finally, hydroxyapatite bone cement was used to complete the cranioplasty prior to wound closure.

**Technique for RWR surgery.** The basic RWR techniques were similar to those described nearly a quarter century ago. Loose areolar tissue was harvested and minced into 0.25-mm pieces with a #10 Beaver blade. Tisseel, a two-component fibrin sealant, was used to coat the pieces. One component of Tisseel is a sealer protein solution that contains human fibrinogen and...
Table 3. Results of diagnostic studies before surgical intervention

<table>
<thead>
<tr>
<th>Pt.</th>
<th>SCD group</th>
<th>Pseudoconductive hearing loss</th>
<th>Endolympathic hydrops*</th>
<th>cVEMP</th>
<th>VAT gain</th>
<th>Moving platform pressure test</th>
<th>High-resolution temporal bone CT</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SCD group</td>
<td>1</td>
<td>Bilateral</td>
<td>No</td>
<td>Positive, left</td>
<td>Normal</td>
<td>Not performed</td>
<td>Left SCD, right near-SCD</td>
</tr>
<tr>
<td>2</td>
<td>Left</td>
<td>Bilateral</td>
<td>Normal</td>
<td>Normal</td>
<td>Positive, small left</td>
<td>Left SCD</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Bilateral</td>
<td>No</td>
<td>Absent</td>
<td>Normal</td>
<td>Positive, small right</td>
<td>Right channel-like SCD, left near-SCD</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Bilateral</td>
<td>Bilateral</td>
<td>Normal</td>
<td>Normal</td>
<td>Not performed</td>
<td>Right SCD</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Left</td>
<td>No</td>
<td>Normal</td>
<td>Normal</td>
<td>Not performed</td>
<td>Left SCD</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Right</td>
<td>Bilateral</td>
<td>Positive, right</td>
<td>Normal except horizontal ≥4 Hz reduced</td>
<td>Positive, small right</td>
<td>Right SCD</td>
<td></td>
</tr>
<tr>
<td>No-iOCD group</td>
<td>7</td>
<td>Bilateral</td>
<td>No</td>
<td>Positive, right</td>
<td>Normal</td>
<td>Positive, large right</td>
<td>Normal</td>
</tr>
<tr>
<td>8</td>
<td>Right</td>
<td>Bilateral</td>
<td>Decreased amplitude left &gt; right</td>
<td>Normal</td>
<td>Positive, small right</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>Bilateral</td>
<td>No</td>
<td>Normal</td>
<td>Normal</td>
<td>Positive, right</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>Left</td>
<td>No</td>
<td>Absent</td>
<td>Normal</td>
<td>Positive, left</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>Bilateral</td>
<td>No</td>
<td>Positive, bilateral</td>
<td>Aborted (too asymptomatic)</td>
<td>Positive, large bilateral</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>Left</td>
<td>Left</td>
<td>Positive, left</td>
<td>Normal</td>
<td>Positive, left</td>
<td>Normal</td>
<td></td>
</tr>
</tbody>
</table>

* Abnormal SP/AP ratio on electrocochleography.

Key: cVEMP = cervical vestibular evoked myogenic potential (positive indicates increased amplitude and decreased threshold); VAT = vestibular autorotation testing, horizontal and vertical; CT = computed tomography; SCD = semicircular canal dehiscence.
aprotinin, a synthetic fibrinolysis inhibitor that helps prevent premature degradation of the fibrin clot; the other component is a human thrombin solution with calcium chloride. Each of these solutions is prepared and kept isolated in petri dishes into which the minced tissue is divided. An Nd:YAG (532 nm [green wavelength]) laser was used to denude all of the mucosa around the round window niche and around the anterior portion of bone surrounding the annular ligament of the oval window.

After placement of the reinforcement materials, the defocused laser was used to coagulate and denature these materials at the periphery so that greater adherence to the temporal bone could be achieved. The round window was reinforced with the loose areolar tissue coated with the fibrinogen and thrombin solutions. The oval window reinforcement was accomplished with draped grafts around the anterior crus, which were packed into place with Gelfoam. Too much tissue was intentionally placed in the round window niche and also around the stapes because some of it would be resorbed during the healing and connective tissue remodeling phases.

Following reinforcement, the middle ear was filled with Gelfoam, and a tympanomeatal flap was placed into position. Strips of dry Gelfoam were placed across the intact skin and the skin of the tympanomeatal flap, and a small amount of antibiotic ointment was placed over this. Ofloxacin 0.3% otic solution was instilled into the external auditory canal. No additional dressing materials were required.

For patient 3, who had SSCDS and who developed a delayed no-iOCD after SCD plugging, the standard technique for RWR was modified. The bone was drilled off the round window niche with a 0.8-mm diamond bur. Then the perichondrium was thinned with a fascia press and placed directly on the surface of the round window membrane and extended onto the otic capsule.

After the mucosa was denuded with the laser, a 2-mm conchal cartilage graft was harvested with a 2-mm biopsy punch and then split in half and placed on top of the perichondrial graft. Loose areolar tissue was minced into 0.25-mm pieces separated into the two petri dishes that contained either a human fibrinogen solution with aprotinin or a human thrombin solution. The latter was then circumferentially placed like a gasket around the cartilage and onto the perichondrium.

**Ethical considerations.** The procedures followed in this series were performed in accordance with the ethical standards of the responsible committee on human experimentation and with the Helsinki Declaration. Our institutional review board approved the study protocol.

**Results**

Resolution of SSCDS symptoms ultimately occurred in all patients. The mean duration from final surgery to June 1, 2015, ranged from 2.3 to 3.6 years (mean: 3.0) in the SCD group and from 2.5 to 3.1 years (mean: 2.9) in the no-iOCD group (table 1). (Links to videos summarizing the cases of patient 8,12 patient 10,13 patient 11,14 and patient 1215 are available in the “References” section.)

**Diagnostic test findings.** Figures 2 and 3 show the pre- and postoperative cVEMP results in patient 12, a 51-year-old woman with no-iOCD who had an elevated amplitude and decreased threshold. She underwent left RWR surgery on April 12, 2012. On follow-up, she remained asymptomatic more than 3 years after her surgery.

All 12 patients demonstrated a pseudoconductive hearing loss in at least one ear (table 3). Figure 4 shows the preoperative unilateral (left-sided) pseudoconductive hearing loss in patient 12.

High-resolution temporal bone CT in two different 3-D rendering modes showed evidence of SCD in all 6 SCD patients and in none of the no-iOCD patients (figure 5).

Only 1 patient, the 32-year-old woman with bilateral SSCDS (patient 3), underwent MRI with CISS sequences, and it determined that her bilateral superior canals remained plugged (figure 6). She did well for 1.5 years, but after prolonged vomiting, she developed a recurrence of her SSCDS/OCD. She subsequently underwent right RWR surgery with the modified technique, and her symptoms resolved.

**Hearing internal sounds and hearing or feeling a tuning fork applied to the extremities.** Preoperatively, all 12 patients reported hearing internal sounds (table 2). Of note, 3 of the SCD patients and 2 of the no-iOCD patients were able to hear their eyes move. Postoperatively, these sounds ceased in all 12 patients.

Likewise, all 12 patients were able to hear or feel a 128-Hz and 256-Hz tuning fork applied to their knee or elbow preoperatively (table 2).24 (A link to a video showing two representative tuning fork tests is available in reference 24.) This condition also resolved after their surgical procedures were completed.

**Computerized dynamic posturography.** As seen in figure 7, both the SCD and no-iOCD groups experienced highly significant improvement following treatment (Wilcoxon signed-rank test, p < 0.001). The greatest improvement occurred in patients who were most sensitive to the vestibular contributions to postural control (SOT conditions 5 and 6). The difference in pretreatment postural performance between the two groups was not statistically significant (independent samples Mann-Whitney U test). Also, there was no difference
in post-treatment EQ scores.

The no-iOCD group tended to show a more robust improvement in the SOT condition 5 value, although the difference was not statistically significant.

**Discussion**

The most important finding of our study is that SSCDS can occur in both SCD and no-iOCD patients. Therefore, we suggest that the term *superior semicircular canal dehiscence syndrome* should be abandoned and replaced with the term *otic capsule dehiscence syndrome*. Moreover, practitioners should understand that the OCDS designation includes not only patients with SCD and no-iOCD, but those with posterior and lateral semicircular canal dehiscence, as well. All of these conditions can manifest as SSCDS/OCDS.

**What is no-iOCD?** In the words of American surgeon William Stewart Halsted (1852-1922):

“If you put ‘perhaps’ before a statement and the statement turns out to be true, you will get credit for making it; if it turns out to be false, you will not be blamed.”

Clinically, since no-iOCD patients have the same clinical phenotype as SSCDS patients, *perhaps* no-iOCD is really an otic capsule dehiscence in an area such as the modiolus that creates a third window, just as is the case with SCD. If so, these otic capsule defects cannot be visualized with existing CT technology. Reinforcing the round window effectively closes the third window, thereby eliminating or reducing symptoms. Thus, RWR surgery is effective in selected patients not because perilymph leakage from the inner ear into the middle ear has been stopped, but because closing the third window alters the biomechanical properties of the inner ear. If Bhutta’s hypothesis\(^5\) that patients who hear their eyes move do so via transdural transmission of extraocular muscle contraction is correct, it supports the idea that the modiolus is the site of the third window.

![Figure 2](image2.png)

*Figure 2. Patient 12. Image shows the preoperative left and right cVEMP results in the 51-year-old woman with no-iOCD. At the presentation level of 90 dB HL, the amplitudes of the response evoked were 276 μV from her left ear and 107 μV from her right ear.*

![Figure 3](image3.png)

*Figure 3. Patient 12. Image shows the pre- and postoperative cVEMP amplitudes and thresholds in the left ear of the 51-year-old woman with no-iOCD. At the presentation level of 90 dB HL, the amplitudes of the response evoked were 276 μV preoperatively and 66 μV postoperatively. The amplitude of her preoperative threshold was 35 μV at 75 dB HL, and the amplitude of her postoperative threshold was 15 μV at 70 dB HL.*
The use of an RWR technique has been explored in SCD patients who underwent RWR surgery using a variety of materials. Silverstein et al described 22 patients with a confirmed diagnosis of SCD who underwent RWR via a transcanal approach. Six surgeons from four institutions participated in this study. They used various types of tissue, including temporalis fascia, tragal cartilage and perichondrium, fat, loose connective tissue, Gelfoam, and Silastic. A statistically significant alleviation of all symptoms except hearing loss was seen in 19 of the 22 patients who underwent RWR. In contrast, 2 of 3 patients who underwent an alternate treatment, round window niche occlusion, experienced a worsening of symptoms that required revision surgery.

RWR surgery with tissue may reduce the symptoms associated with SCD. Silverstein et al speculated that the reinforcement technique may benefit SCD patients by reducing the third-window effect created by the dehiscence. The terms dizziness, giddiness, and disequilibrium are often used to describe these feelings, but they do not accurately capture the nature of these experiences. As a result of this imprecision, physicians have a poor understanding of the symptoms of otic capsule defects in both SCD and no-iOCD patients. In general, patients with SCD or no-iOCD do not experience rotational vertigo. However, this clinical phenotype can be blurred by vestibular migraine being superimposed on SCD or no-iOCD.

Migraine and the gravitational receptor dysfunction type of vertigo. Vestibular migraine, also referred to migraine-associated dizziness, has become recognized as a distinct clinical entity that affects a high proportion of patients who have vestibular symptoms. It is so common that vestibular migraine should be considered in any patient who presents with dizziness, vertigo, or disequilibrium. A temporal overlap between vestibular symptoms (e.g., vertigo and head-movement intolerance) and migraine symptoms (e.g., headache, photophobia, and phonophobia) is a requisite diagnostic criterion. Findings on physical examination and laboratory testing are usually normal in vestibular migraine, but they can be used to rule out other vestibular disorders with overlapping symptoms such as SSCDS/OCDS, SCD, and...
OTIC CAPSULE DEHISCENCE SYNDROME: SUPERIOR SEMICIRCULAR CANAL DEHISCENCE SYNDROME WITH NO RADIOGRAPHICALLY VISIBLE DEHISCENCE

no-iOCD. The pathophysiology of vestibular migraine is incompletely understood, but it is plausible that it would involve neuroanatomic pathways to and from central vestibular structures, as well as neurochemical modulation via the locus coeruleus and raphe nuclei.

In the absence of controlled trials, treatment options for patients with vestibular migraine largely mirror those for patients with classic migraine. These treatment approaches include prophylactic drug therapy with antiseizure medications (e.g., topiramate, zonisamide), calcium channel blockers (e.g., verapamil), tricyclic antidepressants (e.g., nortriptyline) and, especially for children, beta blockers (e.g., propranolol). (See video in reference 10.)

Anecdotally, approximately one-third of patients with vestibular migraine have endolymphatic hydrops, which is typically bilateral. These patients do not experience autophony or sound-induced dizziness and nausea, but when they have endolymphatic hydrops, they can experience sound sensitivity that borders on a Tullio phenomenon. For this reason, when a high-resolution temporal bone CT with color 3-D volume rendering demonstrates no evidence of SCD, all patients suspected of having no-iOCD should be treated as those having vestibular migraine since medical management, if successful, avoids unnecessary surgery. In our study, 3 of the 6 SCD patients and all 6 of the no-iOCD patients had been treated as vestibular migraine patients before surgical intervention.

Vestibular migraine is an illustration of the overlap among vestibular pathways, migraine circuit triggers, and central mechanisms for premonitory symptom generation. Information transmitted by the peripheral vestibular sensory organs and the vestibular nerve to the medulla and pons is an external trigger within the migraine circuit construct proposed by Ho et al. This model is based on the distribution of the calcitonin gene-related peptide, which has a complex distribution within the vestibular periphery.

Migraine headache is almost always present in patients with the gravitational receptor dysfunction type of vertigo caused by SCD or no-iOCD, but it is infrequent in the rotational receptor dysfunction type of true rotational vertigo. This is an important concept because SCD and no-iOCD can induce classic migraine and its three variants: ocular migraine, hemiplegic migraine, and vestibular migraine. In our study, 2 patients in each group had at least one of these migraine variants; in the SCD group, 1 patient had intermittent ocular migraines and vestibular

Figure 5. Patients 7 through 12 (A through F, respectively). CTs of the right and left superior semicircular canals show no visible SCD (yet they all have otic capsule dehiscence syndrome).
migraines twice and another had intermittent vestibular migraines, while in the no-iOCD group, 2 patients had intermittent ocular migraines. This explains why some patients with SCD or no-iOCD, who normally have only the gravitational receptor dysfunction type of vertigo (disequilibrium) can experience episodes of vestibular migraine and infrequent true rotational vertigo attacks.

It should also be noted that the character of the migraines in our study was different between our two cohorts. All the migraines in the no-iOCD group were characterized as “24/7.” These patients also had a greater degree of light sensitivity, and many of them wore sunglasses during much of their waking day; physicians would also find the lights turned off when entering the examination room.

In our study, as is generally the case in clinical practice, surgical management of SCD and no-iOCD resolves the migraines, although sometimes the migraines persist but with decreased frequency and intensity.11-15

**Autonomic dysfunction.** Autonomic dysfunction occurs in varying degrees in cases of PLF, vestibular migraine, and/or SCD, but in general it is very common. Autonomic dysfunction also occurs in cases of rotational receptor asymmetries. Symptoms include nausea, cold and clammy skin, decreased heart rate, and vomiting. Many investigators have studied the underlying mechanisms and pathways subserving this dysfunction.29-32 In our series, all patients experienced some degree of nausea.

**Cognitive dysfunction.** All patients in our series had experienced cognitive dysfunction before surgery. Cognitive dysfunction is uncommon in the rotational receptor dysfunction types of vertigo (e.g., benign positional vertigo, vestibular neuronitis, and other disorders that produce true rotational vertigo). Patients with SCD or no-iOCD often use the following terms when describing their cognitive function: “fuzzy,” “foggy,” “spacey,” and “out of it.” Their memory and concentration are poor, they have difficulty reading because they perceive that the words are floating on the page, they have trouble finding the right words, and they forget what they wanted to say.

Gurvich et al published an excellent review of the role the vestibular system plays in cognition and psychiatry.33 The two key anatomic regions that provide links between the vestibular system and the neural networks involved in cognitive and emotional processing are the parabrachial nucleus and the hippocampus.29-32 Many of the neuroanatomic regions that are linked to the vestibular system are also implicated in several psychiatric illnesses. The past decade has seen increasing interest in the relationship between the vestibular system and mood, cognition, and psychiatric symptoms. Studies have demonstrated that vestibular stimulation can produce changes in mood, cognition, and psychiatric symptoms.34-36

Patients with SSCDS/OCDS can be assigned a neuropsychologic or psychiatric diagnosis before their vestibular disorder.
is diagnosed. In our series, none of the SCD patients and 4 of the no-iOCD patients had previously been assigned a neuropsychiatric/neurobehavioral diagnosis before referral. All 4 of these no-iOCD patients experienced a resolution of their “psychiatric disorder” after surgical intervention. Unfortunately, the assignment of a neuropsychiatric diagnosis before referral is common in children.11 The hippocampus has been consistently implicated in cognition and models of psychiatric disorders, and there is a large body of evidence supporting vestibular-hippocampal interactions.37-41

We recently completed a study incorporating pre- and postoperative quantitative measurements of cognitive function in a cohort of patients who had one of three conditions: SCD only, no-iOCD only, and both SCD and no-iOCD.28 We studied 17 patients (13 adults and 4 children) with clinical SSCDS/OCDS who were treated surgically. We completed neuropsychology test batteries preoperatively and every 3 months postoperatively for up to 1 year. Tests were conducted with the Beck Depression Inventory (BDI-II), the Delis-Kaplan Executive Function System, the Wide-Range Intelligence Test, and the Wide-Range Assessment of Memory and Learning (WRAML-2), which analyzes four domains: verbal memory, visual memory, attention/concentration, and working memory.

We found a significant decrease in BDI-II scores in all three groups. WRAML-2 analysis showed a statistically significant improvement in visual memory and verbal memory for the no-iOCD-only group and the combined SCD/no-iOCD group. We found no improvement for the SCD-only group. All three groups showed improvement in the attention/concentration domain. On the other hand, no change in working memory was seen in any group, and IQ scores were unchanged. All patients in this study had been diagnosed preoperatively with cognitive dysfunction.

Altered spatial orientation. Patients with vestibular migraine who have SCD and/or no-iOCD often say they have trouble judging distances; they feel detached and separated or not connected when they are around other people, almost as if they are watching a play; or they feel as if they are having an out-of-body experience (in severe gravitational receptor asymmetries).

Clinically, spatial disorientation resolves after surgery, although Baek et al reported that spatial memory deficits following bilateral vestibular loss may be permanent.45 There is also evidence that stimulation of the vestibular system is necessary to maintain normal spatial memory.45 Deroualle and Lopez explored the visual-vestibular interaction in their 2014 review, and they concluded that vestibular signals may be involved in the sensory bases of self-other distinction and mirroring, emotion perception, and perspective-taking.14

Patients with SCD and/or no-iOCD recognize changes in their personality. Smith and Darlington argued that these changes in cognitive and emotional function occur as a result of the role that the ascending vestibular pathways to the limbic system and neocortex play in the sense of spatial orientation.45 They further suggested that a change in the sense of self is responsible for depersonalization and derealization symptoms such as feeling strange, “spaced out,” and not in control of oneself. In our series, preoperative spatial disorientation was nearly universal, as only 1 patient, a no-iOCD patient, did not experience it.

Anxiety. Vestibular disorders can produce anxiety, but the classic sense of impending doom occurs only in patients with the most severe gravitational receptor asymmetries. It is nonetheless quite unnerving because it is a unique type of anxiety, and affected patients characteristically have no insight as why they feel anxious. Much work has been completed in an effort to understand the underlying mechanisms and pathways subserving this dysfunction.29,32,46 In our series, only 3 patients (1 SCD and 2 no-iOCD) experienced this type of anxiety.

Audiometry and electrocochleography. In Minor’s 2005 study of SCD, 70% of patients exhibited a pseudoconductive hearing loss of 10 dB or greater. All 12 patients in our series had a pseudoconductive hearing loss. This finding supports the concept of a third window, regardless of whether it is visible on high-resolution CT.

Homeostasis of the pressure differentials between endolymph and perilymph is maintained as a function of the endolymphatic duct and sac. If this balance between pressure and volume is disrupted, endolymphatic hydrops may result. In a retrospective review of 11 cases of SCD (15 ears), Arts et al found ECoG evidence of endolymphatic hydrops in 14 ears; all 4 patients who had undergone surgical repair experienced a resolution of their hydrops.7 Other models of otic capsule defects, such as animal models with experimental PLF, have demonstrated that endolymphatic hydrops usually resolves within 3 weeks of induction.47,48

Aso and Gibson used intraoperative ECoG to demonstrate abnormal SP/AP ratios in patients with no visible PLF.49 Reinforcement of the round window niche in these patients led to relief of symptoms. In our series, only 3 SCD patients and 2 no-iOCD patients exhibited electrophysiologic evidence of endolymphatic hydrops.
Postclosure endolymphatic hydrops is common after PLF repair.\textsuperscript{90} We have observed this frequently in our postoperative SCD and no-iOCD patients, and it has complicated the recovery of some patients before resolution. A detailed analysis of this situation requires a much larger series and a detailed longitudinal cohort.

**Vestibular evoked myogenic potentials.** Assessment of cervical and oculovestibular evoked myogenic potentials (cVEMPs and oVEMPs, respectively) has emerged as an important test of vestibular (otolithic) function.\textsuperscript{17} While not uniformly observed, significant threshold changes and increased amplitude responses can be seen in patients with SSCDS.\textsuperscript{3,17,22} This is also the case in patients with clinical SSCDS/OCDS whose CT scans are normal. It should be noted that both SCD and no-iOCD patients are particularly bothered by and made more symptomatic by acoustic cVEMP and oVEMP testing.

As shown in figures 2 and 3 (patient 12), the cVEMP amplitude can be elevated and the threshold reduced in patients with no-iOCD, and they can normalize after surgical repair. Our patient 12 had the sound-induced gravitational receptor dysfunction type of vertigo, unremitting migraine headaches, and cognitive dysfunction, and she could hear her eyes move preoperatively, yet her CT findings were normal (figure 5, F). These clinical problems resolved postoperatively after round window and oval window reinforcement with loose areolar tissue.\textsuperscript{15} (See video in reference 15.)

Another group made this observation regarding cVEMPs in PLF nearly a decade ago. Modugno et al reported lowered thresholds in a series of PLF patients who had no radiographic evidence of SCD.\textsuperscript{21}

**Computerized dynamic posturography and moving platform pressure tests.** Patients with SSCDS/OCDS have a high incidence of the gravitational receptor dysfunction type of vertigo, which is often referred to as chronic disequilibrium. While posturography can identify disorders of balance and postural dyscontrol, it cannot distinguish between the various types of otic capsule dehiscence. Black et al noted objective improvement in dynamic posturography after PLF surgery, with 12 of 32 patients having normal tests after PLF repair.\textsuperscript{8}

As shown in figure 7, we found a highly significant improvement in postoperative equilibrium scores for each SOT condition in both the SCD and no-iOCD groups. The greatest improvements were seen in those conditions that are more sensitive to vestibular contributions to posture control (SOT conditions 5 and 6). There was no statistically significant difference in pretreatment postural performance between the two groups.

Black et al pioneered the simultaneous application of pressure to the middle ear and indirectly to the inner ear (i.e., the moving platform pressure test) in patients with PLF during computerized dynamic posturography.\textsuperscript{18} However, their work was completed before SSCDS was recognized. Low-frequency sound application has also been suggested as a useful provocative stimulus during posturography for identifying PLF. However, positive test results have also been seen in Ménière disease, SCD, and non-PLF-related inner ear asymmetric function.\textsuperscript{8}

It is interesting that SCD patients experience mildly positive responses to the moving platform pressure test while patients with no-iOCD have more robust responses. This pattern was observed in our patient 3, who developed delayed no-iOCD after SCD plugging (figure 6).

In conclusion, some patients with SSCDS exhibit no otic capsule dehiscence on imaging. As demonstrated in our prospective series, there were no differences between the SCD and no-iOCD groups in symptoms (other than the character of the migraine headaches) and the results of diagnostic studies other than high-resolution temporal bone CT with color 3-D volume rendering. Closure of the third window resolves symptoms, but successfully treated SCD patients might still develop no-iOCD long after surgical management, which manifests as OCDS. Since OCDS encompasses SCD, no-iOCD, lateral canal dehiscence, and posterior canal dehiscence, we believe that the term **superior semicircular canal dehiscence syndrome** should be abandoned and replaced by the term **otic capsule dehiscence syndrome**.

**References**

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