**Objective:** To characterize (1) the impairment and recovery of functional balance and (2) the extent of vestibular dysfunction and physiological compensation following superior canal dehiscence syndrome (SCDS) surgical repair.

**Design:** Prospective study.

**Setting:** Tertiary referral center.

**Participants:** Thirty patients diagnosed as having SCDS.

**Interventions:** Surgical plugging and resurfacing of SCDS.

**Main Outcome Measures:** Balance measures were assessed in 3 separate groups, each with 10 different patients: presurgery, postoperative short-term (<1 week), and postoperative long-term (≥6 weeks). Vestibular compensation and function, including qualitative head impulse tests (HITs) in all canal planes and audiometric measures, were assessed in a subgroup of 10 patients in both the postoperative short-term and long-term phases.

**Results:** Balance measures were significantly impaired immediately but not 6 weeks after SCDS repair. All patients demonstrated deficient vestibulo-ocular reflexes for HITs in the plane of the superior canal following surgical repair. Unexpectedly, spontaneous or post–head-shaking nystagmus beat ipsilesionally in most patients, whereas contrabeating nystagmus was noted only in patients with complete canal paresis (ie, positive HITs in all canal planes). There were no significant deviations in subjective visual vertical following surgical repair (P = .37). The degree of audiometric air-bone gap normalized 6 weeks after surgery.

**Conclusions:** All patients undergoing SCDS repair should undergo a postoperative fall risk assessment. Nystagmus direction (spontaneous and post–head-shaking) seems to be a good indicator of the degree of peripheral vestibular system involvement and central compensation. These measures correlate well with the HIT.


SUPERIOR CANAL DEHISCENCE syndrome (SCDS) was originally described in 1998 and has since been well documented as a disorder characterized by vertigo and oscillopsia in response to loud sounds (Tullio phenomenon) or Valsalva maneuvers. Patients report other symptoms, including pulsatile tinnitus, conductive hyperacusis, and chronic disequilibrium. One of the most common, and often bothersome, symptoms reported in SCDS is that of autophony, or the increased loudness perception of one’s own voice. Clinically, patients with SCDS typically demonstrate a myriad of auditory and vestibular signs, including conductive hearing loss, enlarged vestibular evoked myogenic potential (VEMP) amplitudes with abnormally low thresholds, and eye movements in the plane of the superior canal in response to loud acoustic stimuli or Valsalva. A diagnosis of SCDS is confirmed with high-resolution multislice computed tomography (CT).

For patients whose symptoms are debilitating, the current treatment is that of surgical canal plugging or resurfacing via the middle cranial fossa or transmastoid approach. During canal plugging, fascia strips and bone chips are placed into the open lumen of the superior canal, which effectively eliminates energy transfer between the labyrinth and the cranial cavity, thus resolving symptoms. Surgical plugging of the dehiscent superior canal has been demonstrated to improve hearing thresholds, normalize VEMP amplitudes and thresholds, and relieve...
sound- and pressure-induced vertigo. Functionally, individuals with SCDS exhibit considerable improvement in their perception of dizziness following surgical repair as well as improvement in their perception of autophony.

Postoperatively, reduced superior canal vestibulo-ocular reflex (VOR) gain in response to head impulses has been demonstrated following canal plugging. However, a small percentage of patients also demonstrate global vestibular deficits with additional reductions in both posterior and horizontal canal VOR. The likelihood of global vestibular dysfunction has been found to be related to the size of the dehiscence. Agrawal et al report that for each increase in dehiscence length of 1 mm, the odds of postoperative vestibular hypofunction increase 2.6-fold.

Although a variety of studies have elucidated the effectiveness of SCDS repair (normalization of hearing, VEMP, sound- and pressure-induced symptoms, and autophony), to our knowledge, no studies have characterized the recovery of functional balance following surgery. Therefore, the purpose of this research study was to characterize (1) the impairment and recovery of functional balance and (2) the extent of vestibular dysfunction and subsequent physiological compensation following SCDS surgical repair.

### METHODS

#### STUDY POPULATION

Thirty patients diagnosed as having SCDS (mean age, 46 years [range, 28-61 years]) participated in the study (see Table for a breakdown of patient groups). Of the 30 patients diagnosed as having SCDS, 20 underwent surgical canal plugging plus resurfacing through the middle cranial fossa approach while the other 10 patients declined surgical intervention and were put in the “presurgery” group. A diagnosis of SCDS was based on patient case history; visualization of a dehiscent superior canal on high-resolution CT with multiplanar reconstructions of the superior canal as well as clinical assessments, including cervical and ocular VEMP testing; bedside assessment consisting of head impulse testing in each canal plane and examination for Tullio phenomenon and Hennebert sign; and air- and bone-conduction audiometry. Seven patients were diagnosed as having left SCDS, 7 patients as having right SCDS, and 16 patients as having bilateral SCDS. Of those diagnosed as having bilateral SCDS, symptoms of autophony, pulsatile tinnitus, conductive hyperacusis, and the presence of either a Tullio phenomenon and/or Hennebert sign were referred primarily to the left ear in 8 patients, to the right ear in 6 patients, and referred equally to both ears in 2 patients. All patients gave informed consent for testing through a protocol approved by the institutional review board at the Johns Hopkins University School of Medicine, Baltimore, Maryland.

#### SURGICAL TECHNIQUE

The surgical techniques for plugging the dehiscent superior canal have been described previously. Image guidance was used to confirm the location of the dehiscent canal. The size of the dehiscence was measured by a small scale, in millimeters, which was laid next to the dehiscence. With the use of magnification, dehiscence length and width were measured. The canal was then plugged with fascia and bone pate, filling the perilymphatic space and thus compressing the membranous canal without disruption. Once plugged, a layer of Hydroset Bone Cement (Stryker Corp) several millimeters thick was placed over the repair and allowed to set until palpably firm. Finally, a fascia graft was overlaid and sealed in place with fibrin glue.

#### MEASURES OF BALANCE

In total, 30 patients were diagnosed as having SCDS (Table). Data from all patients were not obtained at all time frames; therefore, for appropriate statistical analysis patients were divided into 3 separate groups of 10 individuals each: presurgery, postoperative short-term (<1 week), and postoperative long-term (≥6 weeks). All balance measures were completed to assess each patient’s fall risk. All patients participated in the following measures of balance.

#### Modified Clinical Test of Sensory Integration on Balance

The Modified Clinical Test of Sensory Integration on Balance (mCTSiB) was completed as a bedside examination for postural control. Patients were instructed to remove their shoes and stand erect with their arms crossed at their chest and their feet together. While timed, patients were asked to maintain balance for 30 seconds during the following 4 trials: (1) eyes open on firm support, (2) eyes closed on firm support, (3) eyes open on high-density foam, and (4) eyes closed on high-density foam. Minimal sway was allowed; however, timing was stopped in the event of a fall, reaching out, opening of the eyes, or taking a step.
For the Dynamic Gait Index (DGI), patients were scored on a 24-point scale assessing 8 aspects of gait. For each aspect, a 4-point scoring system was used, with a score of 3 indicating normal ability and a score of 0 indicating severe impairment. An overall maximum score of 24 was possible, with a score of 19 or less indicating increased fall risk.

Five Times Sit to Stand Test
During the Five Times Sit to Stand Test (FTSST), each patient was instructed to sit in a standard chair with arms crossed over the chest. Timing was started on the word “go.” Patients were to fully stand up and sit down 5 times. Timing stopped when patients fully sat down the fifth time.

Gait Speed
Gait speed was measured over a distance of approximately 6 m. Patients were instructed to walk at a comfortable pace from approximately 1.5 m before the start line until 1.5 m after the finish line. Gait speed was then calculated by dividing the distance (6 m) by time (seconds).

MEASURES OF VESTIBULAR AND AUDITORY COMPENSATION
Ten patients, seen for Measures of Balance, were also followed in both the postoperative short-term phase (<1 week) and in the postoperative long-term phase (>6 weeks) to document vestibular and auditory compensation (Table). Vestibular compensation measures were only completed in patients postoperatively to characterize physiologic compensation following surgical repair of SCDS, with the exception of audiometric testing, which was measured prior to surgery. The following vestibular compensation and function measures were obtained in this group.

Subjective Visual Vertical
The Subjective Visual Vertical (SVV) was assessed at the bedside, using a simple method developed by Zwergal et al. The participant was instructed to put his or her head within the opening of a large bucket, so that peripheral vision was obstructed (Figure 1A). The participant instructed the examiner to rotate the bucket left or right until the line was perceived as truly vertical. The offset of the line was determined by a plumb line and protractor calibrated with the gravitational vertical.

Head Impulse Test in Each Canal Plane
For the Head Impulse Test (HIT), patients were instructed to look at the examiner's nose while their head was quickly moved in the plane of the canal. Head impulses were randomized and completed numerous times in each canal plane. The presence of a refixation saccade indicated a positive HIT result.

Spontaneous Nystagmus and Post–Head-Shaking Nystagmus
First, visual fixation was removed via infrared video goggles (In View, MicroMedical Technologies). The eyes were observed for the presence of spontaneous and gaze-evoked nystagmus. The head was then shaken at a frequency of 2 Hz for 20 cycles before suddenly stopping, and the eyes were then observed for the presence of nystagmus. Nystagmus direction was recorded.

Audiometric Testing
Air conduction and bone conduction thresholds were obtained in response to octave frequencies from 250 to 8000 Hz. For bone-conduction audiology, the audiometer is calibrated for negative bone conduction thresholds; therefore, an extended bone conduction audiogram was obtained.

STATISTICAL ANALYSIS
A between-groups, 1-way analysis of variance (ANOVA) was used to examine the effects of SCDS surgical repair on balance measures. The independent variable was group (presurgery, 10 patients), postoperative short-term (10 patients), and postoperative long-term (10 patients). The dependent variables were individual balance measures (DGI, mCTSIB, gait speed, and FTSST). Post hoc analyses were completed using Tukey's honest significant difference.
For the 10 patients seen at the postoperative short-term phase and again at the postoperative long-term phase, we used a repeated measures ANOVA and correlation analysis to examine the progression of vestibular compensation and recovery of audiometric thresholds. The dependent variables were SVV, HIT results, nystagmus tests, and degree of air-bone gap.

RESULTS

PATIENTS

There was no significant difference in patient age between the presurgery group (mean age, 45.5 years [range, 33-57 years]), postoperative short-term group (mean age, 44.7 years [range, 28-61 years]), or postoperative long-term group (mean age, 49.5 years [range, 39-61 years]) \((F_{2,29}=0.80; P=.46)\). In the postoperative short-term group, surgical plugging of the dehiscent superior canal occurred in the left ear for 4 patients and in the right ear for 6 patients with a mean dehiscence area of 3.58 mm\(^2\) (range, 1.7-7.0 mm\(^2\)). In the postoperative short-term group, surgical plugging of the dehiscent superior canal occurred in the left ear for 6 patients and in the right ear for 4 patients with a mean dehiscence area of 3.13 mm\(^2\) (range, 1.25-6.0 mm\(^2\)). There was no significant difference in dehiscence area between the postoperative short-term and long-term groups \((F_{1,16}=0.30; P=.59)\).

MEASURES OF BALANCE

mCTSIB Scores

Mean mCTSIB scores are shown in Figure 2. There were no significant differences in mCTSIB score between groups for the first 3 conditions; however, significant differences in mCTSIB performance were noted between groups for condition 4 (eyes closed, stance on high-density foam) \((F_{2,27}=3.67; MSE=147.27; P=.04)\). Patients in the postoperative short-term group (mean, 8.7 seconds) demonstrated significantly reduced mCTSIB times compared with the presurgery group (mean, 22.21 seconds). While not statistically significant \((P=.98)\), the postoperative long-term group (mean 20.5 seconds) did improve to similar performance with the presurgery group (Figure 2).

DGI Scores

Mean DGI scores for each group are shown in Figure 3, with a line drawn horizontally at 19, denoting the cutoff for significantly increased fall risk.\(^{16}\) There were significant differences in mean DGI scores between groups \((F_{2,27}=10.75; MSE=6.86; P<.001)\). Patients in the postoperative short-term group (mean DGI, 18.2) demonstrated significantly worse DGI scores compared with both the presurgery (mean DGI, 22.8) and postoperative long-term groups (mean DGI, 23). No patients in the presurgery or postoperative long-term group obtained an abnormal score on the DGI \((\leq 19)\).

FTSST and Gait Speed Scores

There were no significant differences in FTSST between groups \((F_{2,26}=1.82; MSE=12.53; P=.18)\). Likewise, there were no significant differences in gait speed between groups \((F_{2,25}=0.13; MSE=0.05; P=.88)\).

Next, we examined the relationship between gait speed, DGI score, and FTSST between groups. As shown in Figure 4, gait speed was positively correlated with DGI scores \((r=0.86; P=.003)\) and negatively correlated with FTSST \((r=-0.72; P=.03)\), in the postoperative short-term group. Lower DGI scores and increased FTSST scores were both associated with slower gait speed, indicating their similar relationship to increased fall risk.

MEASURES OF VESTIBULAR COMPENSATION

Vestibular compensation measures were completed in 10 patients (5 men and 5 women; mean age, 44.8 years [range, 28-54 years]) at both the postoperative short-
term and long-term phases. Five patients had a right SCD repair, and 5 had a left SCD repair. The mean time between surgery and evaluation date in the postoperative short-term phase was 4.7 days (range, 2-10 days) and was 58.1 days (range, 39-106 days) in the postoperative long-term phase. The mean dehiscence area, measured surgically, was 3.19 mm$^2$ (range, 1.75-5.20 mm$^2$) in 8 patients. Two patients had “blue-lined” bone (1 on the right and 1 on the left) where the canal wall was intact but transparent and thin, and therefore it was surgically opened and plugged.

**SVV Deviations**

Subjective visual vertical (SVV) deviations were all normalized as right-sided deviations (absolute value of the SVV). There were no significant differences in SVV deviation between the postoperative short-term and long-term phases ($F_{1,9}=0.91; \text{MSE}=2.38; P=.37$). Only 2 of 10 patients offset the SVV outside the normal range (0±2.3°) in the postoperative short-term phase and 1 of 10 did so in the postoperative long-term phase.

**HIT Results**

In the postoperative short-term phase, all patients (100%) exhibited a positive HIT result in the plane of the surgically operated superior canal, which remained consistent in the postoperative long-term phase. In addition, 4 patients (40%) had positive HIT results in the ipsilateral posterior canal, and 2 patients (20%) had positive HIT results in all canal planes ipsilateral to the operated ear. No patients demonstrated positive HIT results on the contralateral side. Abnormal HIT findings normalized in the horizontal canal in 1 patient by the postoperative long-term phase. One patient initially demonstrated positive HIT results in the superior and posterior canals but had complications with aural fullness and fluctuant hearing loss following SC repair and then demonstrated positive HIT results in all canal planes by the postoperative long-term evaluation.

**Spontaneous Nystagmus and Post–Head-Shaking Nystagmus**

In the postoperative short-term phase, spontaneous nystagmus was noted in 6 of 10 patients (60%), and post–head-shaking nystagmus was noted in 8 of 10 patients (80%). Unexpectedly, the spontaneous and/or post–head-shaking nystagmus was ipsilesional, with the fast component beating toward the repaired side (ie, left superior canal repair elicited left-beating spontaneous and/or left-beating post–head-shaking nystagmus) in 7 of 10 patients (70%). There was neither spontaneous nor post–head-shaking nystagmus in 1 of 10 patients (10%). In the remaining 2 patients (20%), the spontaneous and/or post–head-shaking nystagmus was contralesional, with its fast component beating away from the repaired side. Interestingly, the 2 patients with contralesional nystagmus were the same patients who demonstrated positive HIT results in all canal planes ipsilateral to the operated ear. Similarly, the patient described as having complications of aural fullness and fluctuant hearing loss whose HIT progressed from 2 canals to 3 initially presented with ipsilesional-beating nystagmus and then developed contralesional-nystagmus by the postoperative long-term evaluation, following this same pattern.

**Dehiscence Size**

To determine if dehiscence size had an effect on vestibular compensation measures, dehiscence size (area, length, and width) was correlated with all vestibular compensation measures (SVV offset, HIT, and presence of nystagmus). A significant positive correlation was found between SVV offset in the postoperative short-term stage and dehiscence area ($r=0.85; P=.003$); however, there were no other significant relationships between dehiscence size and vestibular compensation measures. As well, individuals with blue-lined bone, in whom the thin canal wall was surgically opened and plugged, did not evidence any postoperative findings greater than their peers.
Audiometric Testing

There were no significant mean differences in degree of air-bone gap among the presurgery and postoperative short-term and long-term phases; however, as shown in Figure 5, the degree of air-bone gap was largest in the low frequencies and demonstrated improvement in degree across all frequencies by the postoperative long-term phase. In the postoperative short-term phase, the number of canals exhibiting a positive HIT result was significantly correlated with the degree of air-bone gap at 1 kHz ($r=0.75$; $P=.19$), 2 kHz ($r=0.71$; $P=.03$), and 4 kHz ($r=0.74$; $P=.02$). There was no significant relationship at 250 Hz ($r=-0.14$; $P=.70$), 500 Hz ($r=-0.43$; $P=.21$), 1 kHz ($r=-0.53$; $P=.11$), 2 kHz ($r=-0.62$; $P=.06$), and 4 kHz ($r=-0.13$; $P=.71$) between degree of air-bone gap and head impulse in the postoperative long-term phase and hence audiometric thresholds had normalized.

Previous reports have demonstrated that surgical plugging of the dehiscent superior canal is advantageous in the treatment for SCDS. Postoperatively, patients report improvement in perceptions of imbalance and autophony and resolution of dizziness evoked by loud sound and/or pressure. Clinically, improved hearing thresholds and normalized VEMP amplitude and thresholds have also been documented. However, plugging of the superior canal causes demonstrable immediate and long-term deficits in VOR function, specifically in the superior canal, and studies to date have not reported on the overall functional implications of this VOR deficit on balance.

MEASURES OF BALANCE

Our data suggest that both static (mCTSIB, condition 4) and dynamic (DGI) balance are significantly impaired acutely after SCDS repair; however, both outcome measures recovered by 6 weeks. While gait speed and the FTSSST results did not reveal differences among the presurgery and postoperative short-term and long-term groups, these outcome parameters were strongly correlated with DGI, providing further evidence that these patients are at increased fall risk acutely following surgery. Although it was not the intent of this study to evaluate the effectiveness of vestibular rehabilitation therapy, our data suggest that vestibular rehabilitation would be useful during the early postoperative recovery period. Accordingly, each patient did undergo an inpatient and outpatient physical therapy evaluation with training in gaze and gait stability exercises. At 6 weeks, the outcome measures used had normalized, which may have been related to the physical therapy intervention. Further studies regarding long-term implications and the effects of physical therapy on outcome parameters are planned.

MEASURES OF VESTIBULAR COMPENSATION

We did not find that the clinical test of SVV was sensitive for identifying vestibular dysfunction following SCDS repair. This may reflect the fact that SVV is a measure of otolith function and that function may be well preserved following SC plugging. This finding is supported by the significantly lower, but still present, VEMP responses in most patients following SC plugging.

All patients presented with positive HIT results in the plane of the SC directly following surgical repair, and this finding remained present more than 6 weeks following surgical repair. This is in agreement with other reports. Following SC repair, global unilateral loss of semicircular canal function has been estimated to occur more frequently in the postoperative short-term phase (38% of patients) and improve by the postoperative long-term phase (10%-11% of patients). This is consistent with the rate of 20% (2 of 10 patients) in the current study. Agrawal et al reported resolution of the positive horizontal HIT findings from the immediate postoperative period to 6 weeks postoperatively in 10 of 14 patients. In this study, HIT results remained unchanged from the postoperative short-term evaluation to the postoperative long-term evaluation in 8 of 10 patients. In the remaining 2 patients, 1 patient demonstrated resolution of the horizontal HIT findings from the postoperative short-term evaluation to the postoperative long-term evaluation. However, another patient demonstrated progression of vestibular involvement, which was coupled with aural fullness and fluctuant hearing loss following SC repair. The likelihood of global vestibular loss has been reported to increase with increased dehiscence size; however, findings from the current study do not corroborate this because there was no significant relationship between dehiscence size and number of canals with a positive head impulse sign ($r=-0.14$; $P=.74$) in the postoperative short-term time frame. The likely explanation for this disparity is the small sample size in the current experiment (10 patients) compared with that of Agrawal et al (42 patients).

Physiologically, the mechanism predisposing or causing vestibular dysfunction in adjoining canals is not yet understood. When involvement extends into the posterior canal, it has been suggested that the canal...
plug may have extended into the common crus. However, global vestibular dysfunction following superior canal plugging may be the result of puncturing the membranous canal during canal plugging, overpressurizing the system by compacting the plug materials in the face of the incompressible labyrinthine fluids, or chemical labyrinthitis that selectively affects the vestibular system. Regardless of the degree of vestibular involvement following SCDS repair, the degree of hearing loss and balance function was found to normalize by 6 weeks. Likewise, individuals with blue-lined bone did not evidence any postoperative findings greater than their peers. In fact, both patients had positive HIT results in the superior canal only, indicating that opening the canal wall did not result in any greater peripheral vestibular damage.

The presence of spontaneous and/or post–head-shaking nystagmus is considered an indication of incomplete static (spontaneous) and dynamic (post–head-shaking) central compensation. Most patients (80%) evidenced spontaneous and/or post–head-shaking nystagmus directly following surgical repair. The direction of nystagmus seems to be a sensitive indicator regarding the extent of peripheral vestibular system damage following surgical repair. The only 2 patients who had evidence of global reduction in VOR function (ie, positive HIT findings in the plane of all canals ipsilateral to the surgery side) also had contralateral nystagmus, indicating that the direction of spontaneous and head-shaking nystagmus postoperatively may be an indication of the extent of peripheral vestibular system damage ($r = 0.79; P = .007$). The remaining patients demonstrated ipsilesional, or irritative, nystagmus. One patient, with left superior canal repair, presented acutely with positive HIT results in both the left superior and left posterior canals in addition to having an irritative left-beating spontaneous nystagmus. However, at the 6-week follow-up, each canal HIT result was positive on the left side, and a paretic, right-beating post–head-shaking nystagmus was found, consistent with ipsilesional vestibular hypofunction.

The finding of 70% of patients with either spontaneous or post–head-shaking nystagmus that was directed irritatively (ie, toward the operated side) was surprising. Irritative nystagmus is considered to occur with increased excitability of vestibular sensory cells and has been recorded on injecting potassium chloride through the guinea pig tympanic membrane. One potential mechanism for the irritative nystagmus in these patients may be the change in ion concentration of the surrounding perilymph. It is possible that small defects created in the membranous canal cause an exchange of potassium between perilymph and endolymph, which in turn leads to the irritative nystagmus acutely. For larger tears, a more permanent hypofunction may be the result, accounting for the small number of individuals demonstrating paralytic nystagmus with concomitant vestibular system paresis.

In conclusion, it is recommended that all patients undergoing SCDS repair undergo a postoperative fall risk assessment to include DGI, gait speed, FTSSST, and mCTSIB. In addition, vestibular rehabilitation is likely useful during the early postoperative recovery period. The assessment of nystagmus direction (spontaneous and post–head-shaking) seems to be a good indicator of the degree of peripheral vestibular system involvement and central compensation. These measures correlate well with the HIT.

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