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INTRODUCTION

Superior semicircular canal dehiscence (SSCD) syndrome was first described by Minor et al. in 1998.\(^1\,2\) It was identified in a cohort of patients with vestibular and auditory symptoms induced by pressure and sound changes in the middle ear. Anatomically, SSCD is characterized by the thinning, or complete absence, of bone overlying the superior semicircular canal, resulting in a third-window effect that renders the inner ear more susceptible to pressure changes in the middle ear.\(^3\)

Symptoms of SSCD syndrome include vertigo and disequilibrium in response to loud sounds (Tullio phenomenon) and in response to bearing down. Affected patients often complain of autophony and hearing their own footsteps, heartbeats, and eyeball movements.\(^3\) Audiometric evaluation often reveals a low-frequency conductive hearing loss with a 5 to 10 decibel (dB) air–bone gap.\(^4\) In some cases, bone curve thresholds less than 0 dB can be seen, often referred to as a supranormal, “negative,” threshold. The presence of a nonphysiologic third window diverts hydroacoustic flow to the site of dehiscence in the bony superior canal rather than the cochlea. This generates conductive hearing loss, hyperacusis, and vertigo. Additionally, it has been proposed that the hypersensitivity of the inner ear to intracranial pressure also contributes to hearing loss, vertigo, and pulsatile tinnitus.\(^3\)

The diagnosis of SSCD is confirmed by reduced vestibular-evoked myogenic potential (VEMP) testing as well as high-resolution computed tomography (CT) scans of the temporal bone to visualize the defect.\(^5\,6\)

The clinical incidence of SSCD is unknown. Cadaveric studies of more than 1 thousand temporal bones showed an incidence of 0.7%, with an additional 1.3% showing very thin temporal bone layers (< 1.5 mm).\(^7\,8\) The etiology of this condition is also incompletely understood but is thought to arise from a disruption in the ossification of the bony labyrinth overlying the superior canal.\(^7\)
Nonsurgical management is recommended for mildly symptomatic or asymptomatic patients. For those with bothersome symptoms, surgery is preferred. Several methods have been reported to help visualize and repair the bony defect. These include the middle fossa craniotomy and the transmastoid approach. The middle fossa craniotomy approach carries a higher theoretical risk of seizures from temporal lobe retraction. The transmastoid approach has a lower theoretical risk of cerebrospinal fluid (CSF) leaks and facial nerve injury.

Defects can be resurfaced using cartilage, fascia, bone graft, or hydroxyapatite cement. Alternatively, dehiscent canals can be plugged using bone pâte or other materials. Although plugging has been suggested to achieve better symptomatic control, it has been suggested to carry a higher risk of hearing loss and vestibular damage when compared to resurfacing.

Traditionally, the middle fossa craniotomy approach has been used for resurfacing, whereas the transmastoid approach has been used for plugging. Other techniques such as transtympanic reinforcement of the round window have been reported, with mixed long-term success rates. Few studies have examined the success of resurfacing in the treatment of SSCD; these have shown mixed results. In a recent meta-analysis by Giorgetti et al., no significant differences in surgical success, defined as complete or partial disappearance of symptoms, was found between resurfacing or plugging. However, a meta-analysis by Vlastarakos et al. determined that resurfacing had a success rate of 50% as compared with 97% for plugging; here, success was defined as symptom resolution. Follow-up in this study was mostly between 3 and 6 months. Most recently, Powell et al. reported on their experience with transmastoid resurfacing with bone pâte and fibrin sealant. Here, success was defined by an improvement in symptoms and quality of life, and the authors found significant improvements in 6 of 9 symptoms; most patients reported that the surgery improved their quality of life.

We previously reported on the transmastoid resurfacing technique, with the added benefit of direct visualization of the dehiscence using a Buckingham mirror. This technique combines the benefits of a transmastoid approach and resurfacing. In our previous study, symptom resolution was reported in all four patients, with an improvement in hearing seen in three of four patients (75%) based on up to 4 years of postoperative follow-up. This study reports our growing experience with this surgical technique, describing postoperative symptom changes and patient satisfaction as well as hearing changes, need for re-operation, and complications.

**MATERIALS AND METHODS**

Ethics approval was obtained from our regional research ethics board at our institution (Dalhousie University, file 2014–103). We generated a database of all patients who underwent SSCD resurfacing via the transmastoid approach at our institution between 2005 and 2014. We included patients who were 18 years or older and whose diagnosis of SSCD was confirmed by VEMP testing and high-resolution temporal bone CT scans. We excluded patients with previous surgery for SSCD on the ipsilateral ear and those with no follow-up. We did not evaluate patients for a history of migraine or intracranial hypertension. All patients were operated on and followed by the senior author (M.B.). Surgical complications and follow-up were determined with a retrospective review of operative notes.

To evaluate pre- and postoperative symptom quality and overall patient satisfaction, we used a cross-sectional outcomes recall survey. Questionnaires were mailed to all patients (Supporting Appendix S1). Only patients who responded to the questionnaire were included in the study. Patients were asked to report their symptomatic ear(s) and which ear(s) were operated on. Those who had surgery on both ears were asked to report the outcomes of their surgeries separately. As such, we treated surgeries to the left and right ears for the same patient as independent events.

In the surveys, we asked patients to report the change in the severity of their symptoms after surgery (voice echoing, hearing their own heartbeat, hearing their eyeballs move, dizziness, lightheadedness, dizziness with loud sounds, dizziness with straining, hearing their own footsteps, tinnitus) on a scale of −5 to +5. Negative values implied worsening symptoms postoperatively; 0 depicted no change; and positive values depicted symptomatic improvement. We later decided to exclude dizziness and lightheadedness from any analysis due to its highly subjective and quite variable patient experience. Postoperative symptoms included in the questionnaire, with the exception of dizziness and lightheadedness, were assumed to lateralize to the operated ear. To avoid pseudolateralization of nonlateralizing symptoms, in our analysis of patients with a bilateral surgical history we only considered a single, most recent set of dizziness with loud sounds and dizziness with straining. Patients who had a re-operation that was not a transmastoid resurfacing procedure were excluded from this analysis. Next, patients were asked to report their overall satisfaction with the surgery on a subjective scale of −10 to +10. Negative values implied decreasing satisfaction, and positive values implied increasing satisfaction. Similarly, patients whose revision surgery was not a transmastoid resurfacing procedure were excluded from this analysis. To compare all subjective symptom scores among patients, values were plotted against time since surgery. We tested for relationships between these symptom scores and time since surgery using first-order linear models.

**TABLE I.**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number of patients (ears)</td>
<td>10 (12)</td>
</tr>
<tr>
<td>Right ears (%)</td>
<td>6 (50%)</td>
</tr>
<tr>
<td>Left ears (%)</td>
<td>6 (50%)</td>
</tr>
<tr>
<td>Average age, years (± SD)</td>
<td>52 (± 11 y)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>1/10 (10%)</td>
</tr>
<tr>
<td>Female</td>
<td>9/10 (90%)</td>
</tr>
<tr>
<td>Average follow-up, years (± SD)</td>
<td>4.7 years (± 3.0 y)</td>
</tr>
<tr>
<td>Average surgery to survey time, years (± SD)</td>
<td>4 years (± 2.8 y)</td>
</tr>
<tr>
<td>Complications</td>
<td></td>
</tr>
<tr>
<td>CSF leak</td>
<td>4/12 (33%)</td>
</tr>
<tr>
<td>Vestibular hypofunction</td>
<td>1/12 (8%)</td>
</tr>
<tr>
<td>Otitis externa</td>
<td>1/12 (8%)</td>
</tr>
<tr>
<td>Re-operation performed</td>
<td>4/12 (33%)</td>
</tr>
<tr>
<td>Average time to symptom recurrence, months (± SD)</td>
<td>4.3 months (± 4.1 SD)</td>
</tr>
</tbody>
</table>

CSF = cerebrospinal fluid; SD = standard deviation.
The effect of surgery on hearing was determined by comparing pre- and postoperative audiograms. The most recent audiogram conducted before the first transmastoid resurfacing surgery was used, as well as the most recent audiogram conducted after the surgery (4–6 weeks postoperatively). Average postoperative changes in air and bone curves were assessed by nonlinear, locally weighted regression curves. All 12 ears were included in this analysis.

We considered surgical failure to have occurred if the patients received a re-operation. Revision surgery was offered to patients with residual and bothersome symptoms after primary surgery once. Open circles indicate patients who received a second operation. Patients who received a second operation that was not a transmastoid resurfacing are excluded.

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operation. Kaplan-Meier curves were created to estimate the expected probability of surgical success. All data were tabulated and summarized using the Statistical Package for Social Sciences version 24.0 (SPSS, IBM Corp., Armonk, NY). Graphing, linear and nonlinear models, and survival analysis were prepared in R 3.4.3 (R Core Team, 2017).

RESULTS

Nineteen patients with SSCD underwent surgery through the transmastoid resurfacing approach at our institution between 2005 and 2014. A total of 10 patients (53%) responded to the questionnaire (12 ears in total) and were included in the study. Six of twelve ears were right ears (50%). Nine of 10 patients (90%) were female, and on average the patients were followed for a period of 4.7 years (± 3 years standard deviation [SD]) after surgery. Two out of 10 patients had bilateral symptoms. The average patient age was 52 years (± 11 years SD). The average time between surgery and the receipt of our survey was 4 years (± 2.8 years SD).

The average time from surgery to symptom recurrence was reported at 4.3 months (± 4.1 months SD) (Table I). Eleven of 12 ears (92%) were resurfaced using cartilage overlay and pericondrium/fascia underlay; seven of 11 ears (58%) were supplemented with Tisseel (Baxter Healthcare, Wayne, PA); and four of 11 (36%) were supplemented with bone pâté. Bone pâté and pericondrium alone were used in one of 12 (8%) ears.

There was a very broad spectrum of postoperative symptom scores. Not all symptoms scores were completed by all patients. Scores reflected patient impressions after a second revision surgery, if applicable. After excluding patients with a re-operation that was not transmastoid resurfacing (n = 3) and after excluding nonlateralizing symptom data (dizziness, lightheadedness) from those remaining patients with a bilateral surgical history (n = 1), using a scale of −5 to +5, patients reported an overall improvement in voice echoing (+3.6 [95% CI: 1.9, 5.4]), hearing their own heartbeat (+2.7 [95% CI: 0.7, 4.7]), hearing eyeball movement (+3.7 [95% CI: 2.3, 5.1]), dizziness with straining (+4.2 [95% CI: 2.5, 5.8]), hearing own footsteps (+3.1 [95% CI: 1.1, 5.2]), and tinnitus (+3.0 [95% CI: 1.1, 4.9]) after surgery (Fig. 1). Dizziness with loud sounds was not different after surgery (symptom score + 2.4 [95% CI: −0.6, 5.5]). There was no significant linear relationship between any of the eight symptoms scores and time since surgery (P > 0.05), as well as no significant difference in scores among patients who required re-operation and patients who did not (P > 0.05). On a scale of −10 to +10, patients reported an overall positive satisfaction with the surgery (+6.8 [95% CI: 2.0, 11.5]) (Fig. 1).

The effect of surgery on hearing was evaluated using locally weighted regressions of the difference between pre- and postoperative air and bone curves (Fig. 2) (n = 12). The trendlines showed a monotonic decline in air and bone conduction at frequencies above 2500 Hz (Fig. 2), although this change was small, below 4000 Hz. There was substantial interpatient variability (Fig. 2; gray lines).

Surgical failure, defined by re-operation, occurred in four of 12 ears (33%). Re-operations occurred at 11.9, 22.4, 31.3, and 32.1 months, respectively. Of the four patients who were re-operated on, two of four (50%) underwent transmastoid plugging of the superior semicircular canal; one of these patients received a second revision surgery (67 months after the initial surgery). One of four (25%) underwent round window plugging, and one of four (25%) underwent a second transmastoid resurfacing surgery. Postoperatively, the patients who underwent a resurfacing/plugging of the superior canal reporting complete resolution of their symptoms. The patient with the round window plugging reported a significant improvement in symptoms but not complete resolution.

Second-order linear regressions comparing the air and bone conduction threshold curves of patients who required a reoperation compared to those who did not showed no significant differences (P = 0.67 and 0.97 for the air and bone-difference curves, respectively). This can also be visualized by comparing solid (not failed) versus dashed (failed) gray lines of the raw data in Figure 2.

Follow-up times based on chart review ranged from 9.6 to 124.4 months per patient. Correcting for variable...
lengths of follow-up, estimated probability of surgical success (avoiding re-operation) was determined using Kaplan-Meier Survival Analysis (Fig. 3) \((n = 12\) ears). At about 36 months after surgery, expected surgical success was 0.571 (95% CI: 1, 0.326). Data are available for up to 124 months after surgery, with no failures (re-operations) after 32.1 months.

Complications were seen in six of 12 (50%) ears operated on. Five (42%) procedures resulted in disequilibrium postoperatively, lasting on average 10 days. The most common complication was an intraoperative CSF leak, seen in four of 12 ears (33%). All CSF leaks were identified intraoperatively and repaired. In one of four (25%) of these cases, persistent symptoms were identified on follow-up that required a craniotomy approach for repair. There was no difference in age between those who had a CSF leak (52 ± 11 years SD) compared to those who did not (52 ± 12 years SD). Other complications included vestibular hypofunction and otitis externa (1 of 12 ears for each [8%]) (Table I).

**DISCUSSION**

This study reports on our patients’ experience and the clinical outcomes of transmastoid resurfacing using a cartilage overlay technique in treating superior canal dehiscence syndrome. We assessed the patient-oriented outcomes of symptom severity and perceived satisfaction using questionnaires, which is similar to the approach used by Powell et al.\(^{13}\) Overall, the surgery was deemed satisfactory by most patients’ subjective scoring (Fig. 1). Furthermore, patients reported an overall improvement in voice echoing, hearing their own heartbeat, hearing eyeball movements, dizziness with straining, hearing own footsteps, and tinnitus. However, there was a subset of patients (4 of 12 ears) who required re-operation. Their questionnaire results were collected well after both the initial surgery and the re-operation, and hence may reflect satisfaction after revision surgery.

Using nonlinear regression curves, we demonstrated an increase in air and bone curve thresholds (indicative of a decline in hearing acuity) at frequencies greater than 2.5 kHz postoperatively. These findings are similar to ones reported by Powell et al.\(^{13}\) Middle cranial fossa plugging and transmastoid resurfacing approaches have both been linked to mild high-frequency sensorineural hearing loss postoperatively.\(^{15,16}\) This might be iatrogenic, related to the surgery itself; however, the exact cause remains unknown.

In our cohort of patients, 33% of ears required re-operation, which by our definition constituted surgical failure. Based on Kaplan-Meier Analysis, which accounts for patients lost to medium- and long-term follow-up, we estimate a 57.1% probability of surgical success (avoiding re-operation) at 36 months (95% CI: 100%, 32.6%) (Fig. 3), with data up to 120 months suggesting stability at this point. These data are important with respect to patient counseling and expectation management. The definition of surgical success and failure varies greatly in the literature. In our study, we defined surgical failure as the need for revision surgery, which was offered to patients with persistent and bothersome symptoms. Overall, the patients in our study reported an improvement in the severity of their symptoms after surgery.

There is no standard definition for resurfacing. The use of several materials including bone pâte, perichondrium, cartilage, fascia, cortical bone, and bone substitutes such as hydroxyapatite has been described.\(^{3}\) We have previously reported on using a construct composed of a fascia underlay and tragal cartilage overlay, held together by Tisseel (Baxter Healthcare).\(^{11}\) Most of the patients included in this study underwent resurfacing with a cartilage overlay and an underlay of pericondrium or fascia, supplemented with bone pâte or Tisseel (Baxter Healthcare).

The low success rate for resurfacing might be explained by the lack of long-term stability provided by the grafts. For instance, in two of four (50%) patients who failed surgery in this series, return of symptoms was preceded by upper respiratory tract infection symptoms characterized by coughing, sneezing, and vomiting, which may have destabilized the repair. Alternatively, the graft may have been placed inadequately during surgery. Sharon et al. reported on a large cohort of patients who underwent revision surgery for SSCD. In most cases, they found that the material used during the primary surgery to resurface or plug the canal was present but not entirely covering the dehiscence.\(^{17}\) We visualized the position of the primary repair in three of four patients during revision surgery. Indeed, in two of three patients (67%), the repair construct had migrated from its original position. In one of three patients (33%), the repair was in its original position but appeared to have resorbed. As such, we believe that graft migration and resorption are likely contributing to the lack of long-term symptom resolution seen after our procedure.

Several meta-analyses have examined the surgical complications related to SSCD treatment. No significant differences in outcomes or complications were found between resurfacing and plugging, whether through a transmastoid or middle cranial fossa approach.\(^{15,14}\) Vestibular hypofunction was only seen in one of 12 (8%) ears in our study. This was diagnosed postoperatively through a history of prolonged vertigo as well as a positive head thrust and Fukuda stepping test toward the operated ear. None of our patients underwent vestibular testing postoperatively. Although it is hypothesized that plugging is more likely to result in postoperative vestibular hypofunction and sensorineural hearing loss, we did encounter these complications in our cohort.

CSF leak was seen in four of 12 ears (33%). Xie et al. recently reported on over 240 cases repaired through middle fossa and transmastoid approaches. No dural tears were encountered via the transmastoid approach.\(^{18}\) In our cohort, all CSF leaks occurred during dural elevation. Additionally, in all those patients the tegmen was noted to be low and/or dehiscent around the superior semicircular canal. There was no difference in age between those who had a CSF leak and those who did not. The rate of CSF leak in our study is much higher than previously described in the literature. It is important to acknowledge that this as a potential complication, which can help in operative planning and perioperative patient counseling.

Our sample size was a significant limitation. Only 10 of 19 eligible patients responded to the questionnaires.
Our Research ethics board mandated that only a response to the questionnaire implied consent to participate in the study; therefore, we were unable to include the remaining patients in the cohort. Because this is an uncommon surgery, future work is best done through multicenter case series. Our use of postoperative questionnaires, which facilitated quick and inexpensive data collection, unfortunately also introduces the possibility of recall bias; patients more remote from a surgery or those who received a re-operation may have recalled their symptom burden differently. We attempted to address some of these limitations by excluding data was appropriate from patients with bilateral disease or certain re-operations. We also tested for evidence of time dependence of symptoms scores using linear modeling; here, there were no such patterns. There are no validated questionnaires specific to SSCD with regard to symptoms. Additionally, reporting on the outcomes of various treatments for SSCD in the literature is highly variable. To further mitigate such risks, future studies could utilize validated questionnaires such as the Autophony Index or the Dizziness Handicap Index to report on specific symptomatic outcomes.

CONCLUSION

Here, we add to the body of evidence on the surgical management of SSCD. Transmastoid resurfacing approach using an underlay graft technique carried a long-term success rate of 57.1% but with high patient satisfaction, although sometimes after re-operation with the same technique. Importantly, we report a high intraoperative CSF leak rate, which in all cases was encountered during elevation of the dura mater. The findings of this study have enabled us to better counsel patients on the outcomes and complications of this procedure, allowing for better informed decision making and expectation management. Offering resurfacing as the primary approach should involve a detailed discussion on the possible need for a plugging procedure in the event of surgical failure. Alternatively, canal plugging as the primary surgical approach for SSCD is a viable option.

BIBLIOGRAPHY