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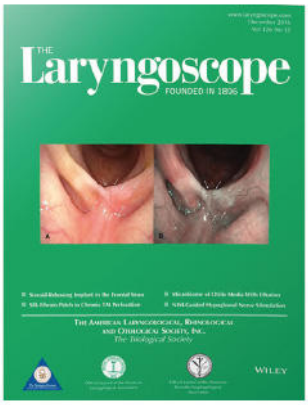


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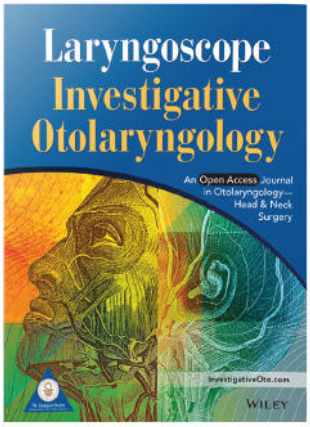


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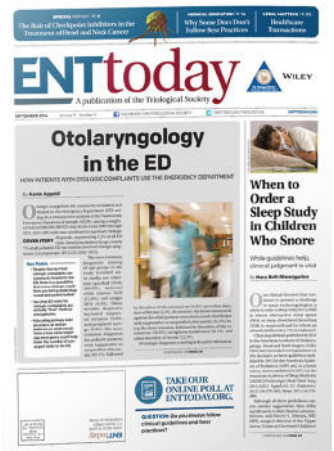


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# Evaluation and Treatment of Pulsatile Tinnitus Associated With Sigmoid Sinus Wall Anomalies

David J. Eisenman, MD; Prashant Raghavan, MD; Ronna Hertzano, PhD, MD; Robert Morales, MD

**Objective:** Describe clinical and radiographic features of sigmoid sinus wall anomalies (SSWA) associated with pulsatile tinnitus (PT) and determine factors predictive of response to surgery.

**Methods:** Preoperative diagnostic imaging and treatment response were reviewed after surgical repair of 40 ears among 38 consecutive patients presenting with PT associated with SSWA who underwent transtemporal sinus wall reconstruction.

**Results:** Twenty-three ears had isolated sigmoid sinus dehiscence, and 17 had diverticulum. The rates of transverse sinus stenosis (TSS) and empty sella, 66% and 32% respectively, were significantly higher than in historical controls ($P = 0.02$ and 0.001). Thirty-six out of 40 subjects (90%) had complete resolution of their PT following surgery, including all those with a diverticulum. For subjects with dehiscence alone without diverticulum, a favorable response to surgery was strongly associated with the presence of TSS ($P = 0.01$) and empty sella ($P = 0.02$).

**Conclusion:** Sigmoid sinus diverticulum and dehiscence are a clinically important cause of PT. Women of childbearing age with an elevated body mass index (BMI) are commonly affected, and there is a high rate of associated TSS and empty sella. Transtemporal sinus wall reconstruction has a high rate of success in appropriately selected patients. Patients with isolated sinus wall dehiscence without diverticulum, TSS, or empty sella are less likely to respond to transtemporal sinus wall reconstruction. These data imply a multifactorial cause of PT in at least some patients with SSWA.

**Key Words:** Pulsatile tinnitus, sigmoid sinus diverticulum and dehiscence, idiopathic intracranial hypertension, transverse sinus stenosis.

**Level of Evidence:** 4

***Laryngoscope*, 00:000–000, 2018**

## INTRODUCTION

Pulsatile tinnitus (PT) is the abnormal self-perception of an internally generated vascular somatosound. Although the Hippocratic authors (c. 500–400 BCE) reportedly described a phenomenon of "auto-auscultation of the blood stream,"[1(p.288)] one of the earliest detailed case reports of PT in the classical literature is found in the Babylonian Talmud (Gittin 56b, c. 500 CE). In a clinical vignette mixed with both plausible and fanciful elements, the Talmud describes the illness and death of the Roman emperor Titus at the age of 42 in the year 81 CE. Titus suffered from a rhythmic, drum-like noise in his head. He observed serendipitously that the banging of a blacksmith's hammer masked the sound, after which he hired such a workman to sit before him, banging continuously to mask his self-perception of the somatosound. This provided transient relief for 30 days, after which the masking was no longer effective. He ultimately succumbed to the illness, and the Talmud reports that postmortem examination revealed an intracranial mass with the appearance of a small bird. Roder and Murison, who both felt that this story provided the most reliable diagnostic information on Titus' enigmatic death at a young age, accurately identify some of the relevant clinical data in the anecdote and suggest that it is evidence that the emperor died from complications related to a cerebellopontine angle tumor. However, they both likely misidentify the presumed histopathology as an acoustic neuroma or meningioma rather than as the far more likely jugular foramen paraganglioma with both intracranial and middle ear involvement, a well-known neoplastic cause of PT (with the segment connecting the larger and smaller portions of the lesion perhaps resembling the neck of a bird, connecting the smaller head with the larger body).[2,3(pp.68–9)]

Atkinson distinguishes an "intrinsic" versus "extrinsic" murmur as causes of tinnitus, the latter arising from a "phonetic" or "vibratory" source, whereas the former is a purely "subjective impression without a sound (!) basis."[4] Fowler had previously termed the two contrasting types "vibratory" and "non-vibratory,"[5] but Atkinson rejected that in favor of his own classification. Engstrom and Graf attribute the "first account of a

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TABLE I.
Etiologies of Pulsatile Tinnitus Classified by Mechanism

| Abnormal Sound Production | Abnormal Sound Perception | Combinations | Unknown Mechanism |
|---|---|---|---|
| Acquired dural vascular lesions (malformations and fistulae) [8] | Third mobile window syndromes[12] | Superior petrosal sinus-superior semicircular canal fistula (Fig. 1) | Migraine[13] |
| Temporal bone and cerebellopontine angle vascular neoplasms[9] | Conductive hearing loss[7] | Cochlear-carotid fistula (Fig. 2) | Meniere's disease |
| Fibromuscular dysplasia[14] | Eustachian tube dysfunction[7] | Dehiscent tegmen tympani with dura or cephalocele in contact with ossicles Perigeniculate neoplasm with cochlear erosion | Idiopathic intracranial hypertension[15] |
| Carotid artery stenosis or dissection[16] | Patulous Eustachian tube[17] | | |
| Aberrant carotid artery[16] and other middle ear vascular anomalies[18] | | | |
| Sigmoid sinus wall anomalies[7,10,19,20] Transverse sinus stenosis[21,22] | | | |

stethoscopical murmur ... auscultated over the cranium" to Fischer in 1833, although a specific reference is not provided.[1] These authors also introduce the term "objective" tinnitus, which corresponds to Atkinson's "extrinsic" category, and contrast it with "subjective," which corresponds to his "intrinsic." They define objective tinnitus to mean that "the murmur in a persons (sic) ear *under favourable conditions* [emphasis added] can be heard by an examinator."[1]

The first use of the term *pulsatile tinnitus* was in 1977; Holgate et al. referred specifically to the self-perception of a vascular somatosound.[6] Because there are nonvascular causes of pulsatile or rhythmic tinnitus, this should more properly be referred to as *pulse-synchronous tinnitus*, a term that the above authors also use in the same article. However, the term *pulsatile tinnitus* has become so ensconced in clinical practice and the literature that it now has a specific code in the new International Classification of Diseases-10th edition (H93.AX). Thus, this article will retain the parlance of pulsatile tinnitus to describe the self-perception of vascular somatosounds.

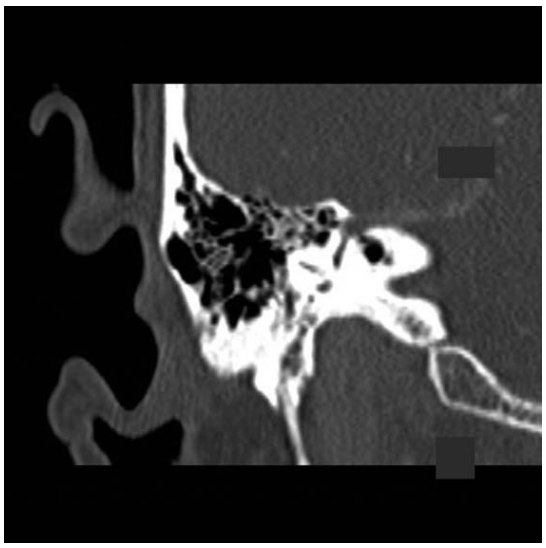

Fig. 1. Coronal computed tomography demonstrating superior petrosal sinus-superior semicircular canal fistula in the right temporal bone, with aberrant vein of Rosenthal entering the sinus adjacent to the fistula.

Pulsatile tinnitus is a form of objective tinnitus. It contrasts with the more common subjective tinnitus, which is typically a central phenomenon often associated with sensorineural hearing loss and noise exposure. Objective tinnitus arises from a mechanical sound source, although not necessarily a pathologic one, and is found more commonly in patients with normal sensory hearing. Pulsatile tinnitus can be caused by abnormal sound perception of a normal internal stimulus, such as in third mobile window syndromes,[7] or by abnormal sound production, such as with acquired dural vascular lesions[8] or temporal bone paraganglioma.[9] Venous etiologies of PT have been described and identified with increasing frequency.[7,10] A common nonpulse-synchronous cause of objective tinnitus is the rhythmic clicking or fluttering associated with middle ear or palatal myoclonus.[11] A more comprehensive list of potential causes of PT is found in Table I.

Sigmoid sinus wall anomalies (SSWA) are an often unrecognized yet potentially treatable venous etiology of PT.[7,10,19,20,23] These include both sigmoid sinus diverticulum and sigmoid sinus dehiscence[19,20] (Fig. 3A–B). These lesions are an uncommon incidental finding in

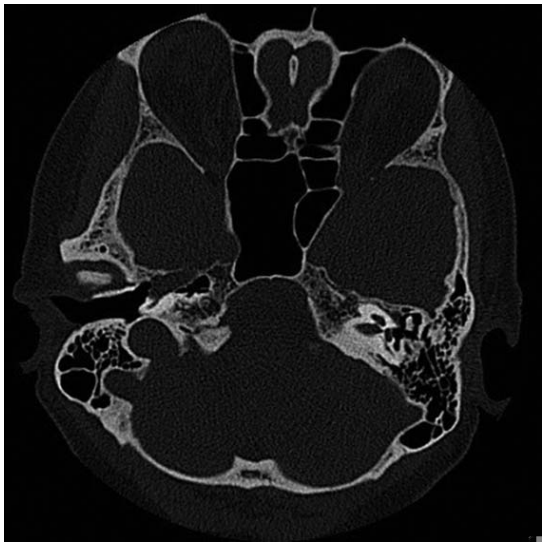

Fig. 2. Axial computed tomography demonstrating a cochlear–carotid fistula of the right temporal bone.

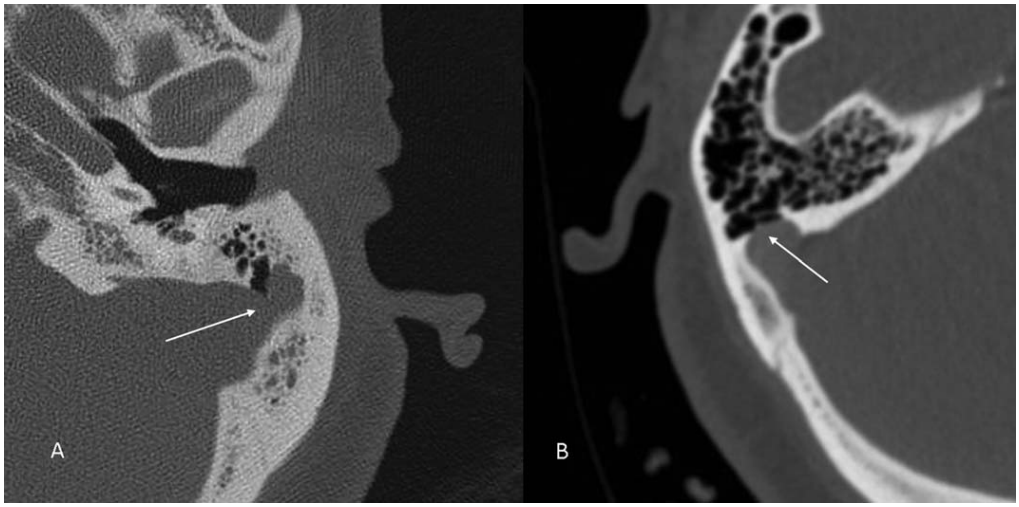


Fig. 3. (A, B) Typical Examples of Sigmoid Sinus Wall Anomalies. (A) Left pedunculated sigmoid sinus diverticulum with narrow neck (axial CT). (B) Right sigmoid sinus dehiscence (axial CT).

temporal bone computed tomographic (CT) scans performed for reasons other than PT[24] but are the most commonly identified radiographic abnormalities in CT scans performed for the evaluation of PT.[7,10,24] The pathophysiology of the anatomic development of SSWA and the genesis of the pathological sound remain uncertain. Epidemiologically, PT due to SSWA is far more common in women than in men,[20,25] and the majority of affected women have an elevated body mass index (BMI), with a mean of 35.5 $kg/m^2$ in one large series.[25] These findings, coupled with the results of another study demonstrating elevated lumbar spinal opening pressures in patients undergoing surgery for SSWA,[26] suggest a pathophysiologic relationship to idiopathic intracranial hypertension (IIH), an established cause of PT affecting the same demographic, although also by means not yet explained.[27] The increased prevalence of transverse sinus stenosis (TSS) and other radiographic findings associated with IIH in patients with SSWA also support this association.[10,28] Despite all of this evidence, the association between SSWA, TSS, and IIH is still not understood. Elucidation of this complex issue is further complicated by the fact that the nature of the relationship between IIH and TSS is itself poorly understood. It is clear that there is an increased prevalence of transverse sinus stenosis in patients with IIH.[29,30] One study demonstrated a 93% rate of bilateral stenosis in subjects with IIH as compared to a rate of 6.8% in normal controls.[30] Although other studies have suggested a higher rate of TSS in normal controls,[31] the rate of *bilateral* TSS is clearly much lower in normal controls than in the IIH population. The cause of this high prevalence of stenosis in patients with IIH is uncertain. Two theories have been advanced.[32] The first postulates that stenosis of the intracranial venous outflow in one or both transverse sinuses leads to elevated intracranial pressure (i.e., TSS causes IIH). This theory is supported by the fact that stenting of the transverse sinuses in patients with IIH often decreases lumbar opening pressures and relieves symptoms of IIH, including PT.[33] The second theory postulates that transverse sinus stenosis is caused by extraluminal compression due to elevated intracranial pressure (i.e., IIH causes TSS). This theory is supported by the observed reversal of stenosis, with re-establishment of venous outflow following lumbar puncture with removal of cerebrospinal fluid.[34] It is possible that both theories are pathophysiologically significant, and that a vicious cycle in which each factor exacerbates and perpetuates the other leads to the final phenotype.

Pulsatile tinnitus itself has also been associated with TSS, independent of the presence of elevated intracranial pressure, and there are some data in the literature supporting the efficacy of transverse sinus stenting in this population.[21,22] However, interpretation of these data is complicated by the fact that these studies did not assess for the presence of possible associated SSWA, despite the fact that studies have demonstrated a high prevalence of SSWA in this patient population.

Identification of a sigmoid sinus diverticulum as a cause of PT, along with effective treatment via an endoluminal approach, was first described by Houdart et al. in 2000.[35] Subsequent additional case reports followed shortly thereafter.[36,37] Following these reports, successful surgical correction via a transtemporal, extraluminal approach was introduced in the neurotologic literature in 2007,[38] with another report in the neurosurgical literature shortly thereafter.[39] Sigmoid sinus wall dehiscence without diverticulum as a cause of PT, as well as its surgical repair with a standardized transtemporal approach, were described in a larger case series in 2011,[19] with subsequent smaller series again following shortly thereafter.[26,40] In light of the presumed common pathophysiology of sigmoid sinus diverticulum and dehiscence, similar patient demographics, and similar successful approaches to the evaluation and treatment, the constellation of sigmoid sinus abnormalities encountered in clinical practice has come to be known as

*sigmoid sinus wall anomalies*.[25] This appellation accounts for the possibility that other related anomalies may yet be described and that a definitive classification scheme of the subtypes is still lacking.

Despite the success of both endoluminal and transtemporal approaches to treatment, not all patients with PT and SSWA are cured by surgery. [25,41] This finding implies that, excluding technical failures, the radiographic finding of a SSWA alone in at least some patients does not suffice to account for the PT. In the absence of an objective means of demonstrating that PT in a specific patient results from a radiographically identified anomaly, additional criteria are needed to appropriately select candidates for surgery and minimize failures. The strong association between SSWA and IIH suggests the possibility that TSS, which is also more highly prevalent in patients with SSWA,[41] may also play a role in the production of PT in this pathologic process as well. The uncertainty about the cause-and-effect relationship between IIH and TSS has led to the suggestion that there is a self-perpetuating vicious cycle, with the two interacting to produce the end clinical result.[32] A similar hypothesis can be posited for a relationship between TSS and SSWA, that is, that the two interact in varying degrees to produce symptomatic PT. This hypothesis, if correct, would suggest that sigmoid sinus wall anomalies are the missing link explaining the connection between IIH and PT. If correct, this pathophysiology also suggests that treatment of PT in this population might be accomplished by addressing either pathologic entity if both are necessary but not sufficient to cause the clinical symptom.

This study reports the clinical findings and results of a consecutive series of 40 patients with SSWA treated surgically for symptomatic relief of ipsilateral PT. The goal of this study is to document the typical presenting clinical findings, including features associated with IIH, and to describe factors associated with surgical success or failure as measured by postoperative elimination of the subjective index symptom. The data suggest that not only are SSWA strongly associated with TSS, but the presence of TSS is also associated with surgical success. Knowledge of such factors may improve surgical outcomes, lead to a better understanding of the pathophysiology of SSWA, and potentially aid in the identification of alternative treatment options.

## MATERIALS AND METHODS

### *Study Design and Radiologic Analysis*

A retrospective review of data from 40 transtemporal sinus wall reconstructions for radiographically identified SSWA associated with PT was undertaken. All procedures were performed in a single center— all but one performed by the senior surgeon. There were 36 patients with unilateral defects and two with bilateral defects repaired. All patients had adequate preoperative temporal bone CT imaging available to establish a diagnosis. Many of the scans were performed at outside facilities and thus varied in technique and quality. Any other available imaging (e.g., invasive or noninvasive vascular studies) was also reviewed. All the studies were reviewed independently in a blinded fashion by two fellowship-trained neuroradiologists, each with more than 10 years of clinical experience. The radiologists were aware that all patients had undergone surgery for PT but were blinded to the clinical diagnosis (diverticulum or dehiscence), laterality, and surgical outcome. For patients who had bilateral surgery (n = 2), both sides were considered separately. For purposes of the statistical analysis, any patient with intraoperative confirmation of a sinus wall dehiscence or diverticulum was included in the study group. Dehiscence was defined as the absence of the normal layer of bone overlying the sigmoid sinus on at least three consecutive 0.6 mm cuts on the axial CT, with preservation of the normal sigmoid contour and no focal outpouching or irregularity. If mastoid air cells were in direct contact with the soft-tissue sinus wall, then the sinus was defined as dehiscent at that level. Diverticulum was defined as a focal outpouching of the sigmoid sinus protruding into the mastoid air cell system or mastoid cortex, with irregularity of the contour of the surface of the sinus. All patients with diverticulum had dehiscence of overlying bone as well. Transverse sinus stenosis was defined as any focal area of prominent caliber change in the vessel comprising at least a 50% decrease in diameter, although these were typically a slit-like appearance. Dominance of a sinus was defined as greater than 150% of the diameter of the smaller side. Empty and partially empty sella were defined by standard radiographic criteria.

### *Surgical Technique*

The surgical technique was like one previously described.[3] Cortical mastoidectomy is performed, and the sigmoid sinus is identified and skeletonized. The anomalous area is fully decompressed, along with a small circumferential area of normal sinus wall. A diverticulum, if present, is reduced with bipolar cautery. If rupture occurs, bleeding is controlled with topical application of an absorbable, oxidized cellular polymer (Surgicel Nu-Knit, Ethicon LLC, Somerville, NJ). The sigmoid sinus, along with adjacent dura if necessary, is elevated from the posterior petrous face. A soft tissue graft of temporalis fascia or dermal replacement matrix (AlloDerm, LifeCell Corp., Bridgewater, NJ) is insinuated deep to the petrous bone in the epidural space. The bony defect is reconstructed with synthetic bone cement (Hydroset, Stryker Corp., Kalamazoo, MI). A layer of autologous bone paté can be placed overlying the cement to protect it from fracture during closing and to promote fibrovascular ingrowth. In appropriate circumstances, when there is a sufficiently robust surrounding sigmoid sinus wall, the soft tissue graft can be secured with epidural sutures to obliterate dead space and discourage formation of an extramural collection that could compress the vessel. The suture ends are left long and tied (or tied down a second time) onto the surface of the cured bone cement over a roll of Surgicel Nu-Knit (Ethicon LLC) to securing all three layers of the reconstruction together (Fig. 4 A–E).

### *Data Analysis and Compliance*

Outcomes were dichotomously classified as either a treatment success or failure (responders vs. nonresponders) based on subjective resolution of the presenting symptom at least 1 month postoperatively. All subjects deemed nonresponders had persistence of their pulsatile tinnitus, and all responders had complete resolution at the 1-month follow-up period. Previous studies have documented durability of short-term results over a longer period of observation.[19]

Descriptive and comparative statistics were calculated using Microsoft Excel 12.3.6 (Microsoft Corp., Redmond, WA) and SPSS 22 (IBM Corp., Armonk, NY). Contingency tables were analyzed using Fischer exact test, or the chi-square test when the number of values sufficed—both with two tails. This study was approved by the University of Maryland School of

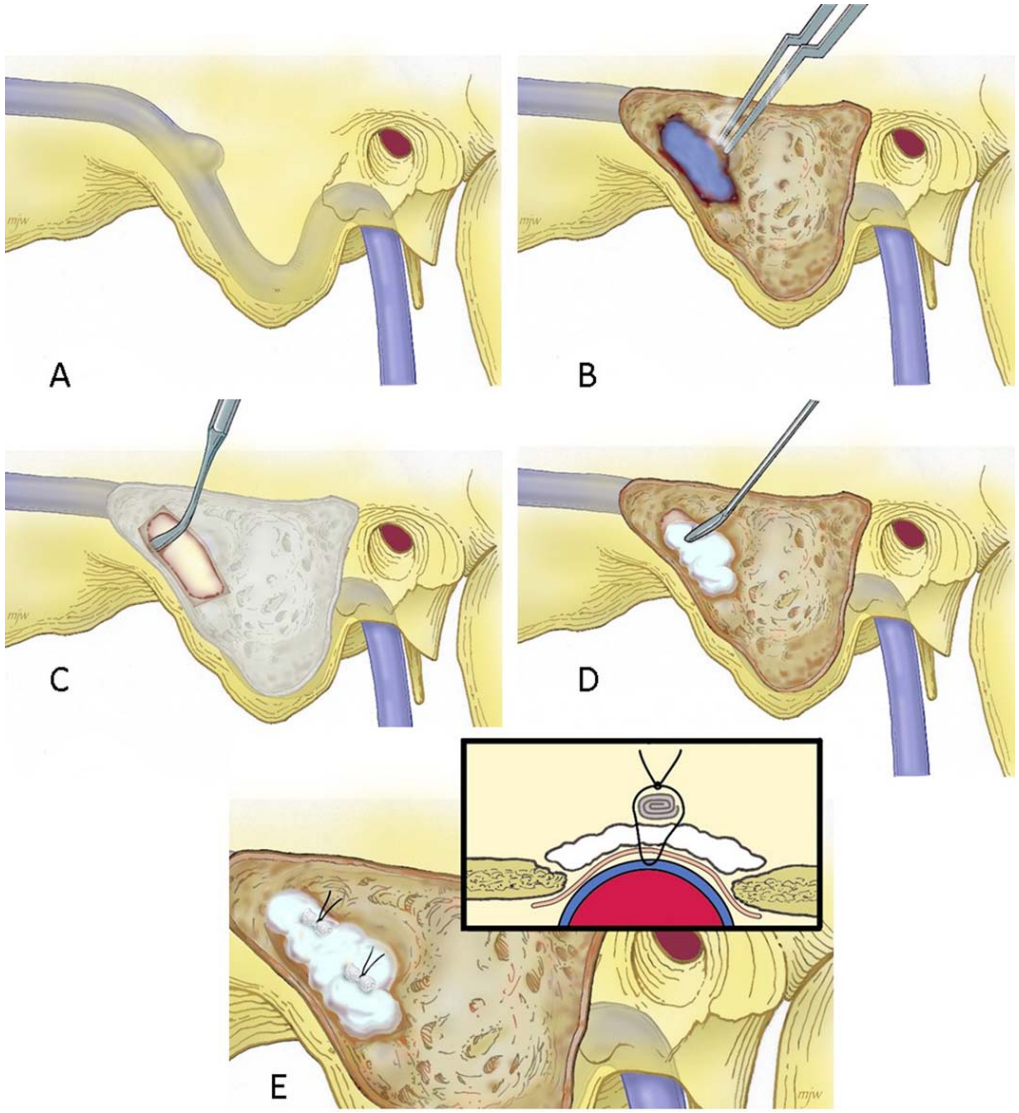


Fig. 4. Surgical technique of Sinus Wall Reconstruction. (A) Preoperative schematic of a sessile diverticulum. (B) Skeletonization and decompression of sigmoid sinus after mastoidectomy, with reduction of the diverticulum using bipolar cautery. (C) Placement of soft tissue graft. (D) Application of bone cement. (E) Optional epidural suture(s) through partial thickness of sigmoid sinus wall.

Medicine Institutional Review Board for Human Subjects Research (protocol HP-00059407). The authors have no conflicts of interest.

## RESULTS

The complete data set documenting clinical findings (including patient characteristics and associated findings) and surgical outcomes is found in Table II.

### *Clinical Findings*

Descriptive statistics for the cohort are presented in Table III. All but four subjects were female: cases 2, 5–13 (subject with bilateral operations), 34, and 36. Age, symptom duration and BMI were not significantly different for subjects with diverticulum and dehiscence (independent two-tailed $t$ test, $P = 0.22$, 0.08, and 0.6, respectively), although the relatively low $P$ value for symptom duration does not exclude the possibility of a falsely negative result due to insufficient power. The difference in laterality of pathology between subjects with diverticulum and dehiscence was not statistically significant (chi-square, $P = 0.33$).

The transverse dimension of dehiscence ranged from 2.5 mm to 11.25 mm, although the largest was an outlier in subject 36 whose surgery was not successful. Excluding that patient, the largest transverse dimension was 8 mm. The median dehiscence size was 4.1 mm on the right and 4.2 mm on the left, with an interquartile range of 3.3 mm to 5.3 mm on the right and 3.7 mm to 4.5 mm on the left. Measurement of diverticula is somewhat arbitrary because they may be multilobulated, with individual lesions having different dimensions at different levels. The smallest diverticulum operated on protruded from the level of the sigmoid sinus wall 3 mm and the largest 10 mm.

TABLE II.
Patient Dta

| Case | Age | Gender | Duration | BMI | Side | Diagnosis | R TSS | L TSS | Empty Sella | Surgical Outcome |
|---|---|---|---|---|---|---|---|---|---|---|
| 1 | 58 | Female | 4 | 52 | right | diverticulum | NA | NA | NA | success |
| 2 | 44 | Male | NA | 29 | right | dehiscence | yes | yes | yes | success |
| 3 | 53 | Female | 10 | 28.4 | left | diverticulum | yes | yes | no | success |
| 4 | 21 | Female | NA | 29.1 | right | dehiscence | no | no | no | success |
| 5 | 15 | Male | 1.5 | 27.4 | right | dehiscence | NA | NA | no | success |
| 6 | 36 | Female | 0.2 | 31.6 | right | dehiscence | yes | yes | no | success |
| 7 | 51 | Female | 10 | 39.2 | right | diverticulum | no | no | yes | success |
| 8 | 47 | Female | 0.5 | 31 | right | diverticulum | yes | no | yes | success |
| 9 | 70 | Female | 1.5 | 29.4 | right | diverticulum | no | no | no | success |
| 10 | 43 | Female | NA | 32 | right | dehiscence | yes | no | yes | success |
| 11 | 57 | Female | 0.7 | 31.8 | right | diverticulum | no | no | no | success |
| 12 | 57 | Female | 1 | 48 | right | diverticulum | no | no | no | success |
| 13* | 17 | Male | 0.2 | 34.2 | left | dehiscence | NA | NA | no | success |
| 14 | 35 | Female | 3 | 17 | right | diverticulum | NA | NA | NA | success |
| 15 | 26 | Female | 0.5 | 43.7 | left | dehiscence | NA | NA | yes | success |
| 16 | 28 | Female | NA | 43.4 | right | diverticulum | NA | NA | NA | success |
| 17 | 37 | Female | 7 | 54.2 | left | dehiscence | NA | no | NA | success |
| 18 | 27 | Female | NA | 22 | right | dehiscence | NA | NA | no | success |
| 19 | 31 | Female | 0.25 | 28.3 | right | diverticulum | yes | yes | no | success |
| 20 | 62 | Female | 0.5 | 22 | left | dehiscence | no | no | no | failure |
| 21 | 56 | Female | 1 | 36.3 | right | dehiscence | no | NA | yes | success |
| 22† | 31 | Female | 0.5 | 28.3 | left | dehiscence | yes | yes | no | success |
| 23 | 27 | Female | 2 | 35 | right | dehiscence | NA | no | no | success |
| 24 | 41 | Female | 3 | 42 | right | dehiscence | no | yes | yes | success |
| 25 | 38 | Female | 1 | 40.6 | left | dehiscence | NA | yes | no | success |
| 26 | 23 | Female | 0.75 | 27.2 | right | dehiscence | yes | yes | no | success |
| 27 | 36 | Female | 1.5 | 44.4 | right | diverticulum | NA | no | no | success |
| 28 | 27 | Female | 1.75 | 35.4 | left | diverticulum | yes | no | no | success |
| 29 | 32 | Female | 1.5 | 30.9 | left | dehiscence | no | yes | yes | success |
| 30 | 57 | Female | 0.75 | 38.1 | left | dehiscence | NA | no | no | failure |
| 31 | 59 | Female | 0.5 | 40.2 | left | dehiscence | no | no | no | failure |
| 32 | 40 | Female | 1.75 | 36.1 | left | diverticulum | NA | NA | yes | success |
| 33 | 38 | Female | 0.5 | 48.7 | right | dehiscence | NA | NA | NA | success |
| 34 | 41 | Male | 9 | 26.52 | right | diverticulum | NA | NA | no | success |
| 35 | 65 | Female | 5 | 44.04 | right | diverticulum | yes | NA | NA | success |
| 36 | 62 | Male | 6 | 23.4 | left | dehiscence | no | no | no | failure |
| 37 | 25 | Female | 2 | 33.8 | right | dehiscence | yes | NA | yes | success |
| 38 | 55 | Female | 2 | 41.4 | left | diverticulum | no | NA | yes | success |
| 39 | 30 | Female | 0.25 | 37.9 | right | diverticulum | NA | NA | no | success |
| 40 | 33 | Female | 1.5 | 49.59 | right | dehiscence | NA | NA | no | success |

*Second side of case 5.
†Second side of case 19.
BMI = body mass index; L TSS = left transverse sinus stenosis; R TSS = right transverse sinus stenosis.

The vast majority of subjects had dominance of their transverse-sigmoid system on the side of their symptoms (n = 24) or codominance of the systems (n = 11). Cases 3, 13, and 19 had contralateral dominance: two with diverticulum and one with dehiscence. Case 36, one of the four surgical nonresponders, also had contralateral dominance. Subject 18's preoperative imaging was inadequate for accurate assessment.

Data on TSS are presented in Table IV. There were no significant differences in the prevalence of TSS between the cases with diverticulum and dehiscence. For comparison, a large study of consecutive patients undergoing CT angiography (CTA) demonstrated a prevalence of unilateral TSS of 33% in an unselected population.[31] The difference in prevalence of unilateral TSS between our study population and these historical

TABLE III.
Population Descriptive Statistics

| | Gender | | Age (median years (IQR))* | Side* | | Duration of Symptoms (median years (IQR))* | Body-Mass Index (median kg/m$^2$ (IQR))† |
|---|---|---|---|---|---|---|---|
| | Female | Male | | Right | Left | | |
| Diverticulum | 16 | 0 | 47 (35–57) | 13 | 4 | 1.75 (0.925–4.25) | 36.1 (29.4–43.4) |
| Dehiscence | 19 | 5 | 36 (26.5–43.5) | 13 | 10 | 1 (0.5–1.75) | 33.8 (28.65–40.4) |
| Total | 35 | 5 | 38 | 26 | 14 | 1.5 (0.5–2.5) | 34.6 (28.85–41.55) |

*Difference between diverticulum and dehiscence for age, duration of symptoms, and BMI not significant. Two-tailed $t$ test, $P = 0.22$, 0.08, and 0.6, respectively.
†Difference in prevalence of diverticulum and dehiscence based on laterality of pathology not significant, chi-square, $P = 0.33$.
BMI = body mass index; IQR = interquartile range.

controls is statistically significant (chi-square, $P = 0.02$). Six of 18 subjects with available data (33%) had bilateral TSS: two of the subjects with diverticulum and four with dehiscence. Of the cases with TSS contralateral to their symptomatic side, six had bilateral TSS and three had isolated contralateral TSS. Of those six, one was a subject (case 19–22) who underwent bilateral surgery. For comparison, the above cited study demonstrated a prevalence of bilateral TSS of 5% in its population.[31] The difference in prevalence of bilateral TSS between the present cohort and these historical controls is statistically significant (chi-square, $P = 0.0001$). The BMI does not differ for subjects with and without TSS (independent two-tailed $t$ test, $P = 0.14$).

Thirty-four cases had adequate imaging to assess the contents of the sella turcica. Eleven (32%) of these cases had empty or partially empty sellae, leaving 23 (68%) without this radiographic finding. Six cases did not have adequate imaging to assess the sella turcica. As compared to a consecutive cohort of patients with IIH studied elsewhere,[42] this rate of empty sella was significantly lower (32% vs. 66%, chi-square, $P = 0.007$), although it is significantly higher than the rate found in their controls (0%, Fisher exact test, $P = 0.001$).

### *Surgical Outcomes*

Transmastoid sinus wall reconstruction resolved the PT in 36 out of 40 patients (responders) for a 90% success rate. Surgery was successful in 100% of patients with a diagnosis of diverticulum (17 of 17), and in 83% of patients with dehiscence alone (19 of 23). This difference is not statistically significant (Fischer exact test, $P = 0.15$). All the failures (nonresponders) occurred in patients with unilateral left-sided dehiscence (Figs. 5–7). Age, symptom duration, and BMI did not differ between the responders and nonresponders (independent, two-tailed $t$ test; $P = 0.09$, 0.90, and 0.60, respectively).

For the four nonresponders, the blinded neuroradiologists both agreed with the clinical diagnosis in two (subjects 30 and 36), but they disagreed on the diagnosis in the other two (subjects 20 and 31), with one agreeing with the clinical diagnosis of dehiscence and the other observing either a thin layer of intact bone or noncontiguous areas of dehiscence. Intraoperatively, the two latter subjects had noncontiguous foci of dehiscence within a region of extremely thin bone. Because the presence of any dehiscence was confirmed intraoperatively, these subjects were included in the final analysis, despite the fact that their radiographic findings were subtler and potentially ambiguous. For comparison, representative images from CT scans of four subjects with dehiscence who responded to surgery are shown in Figure 8.

Because these results demonstrate that the radiographic and intraoperative finding of dehiscence is itself not 100% predictive of surgical success, and there may be cases of dehiscence in which the preoperative imaging is equivocal, other variables associated with outcome were sought. Patients with SSWA have features found in cohorts of patients with IIH[25,28]; therefore, the presence or absence of such features was assessed for its impact on surgical outcome. All subjects with diverticulum were responders; thus, those cases are not included in this analysis. Among cases with dehiscence alone and sufficient available data, there was a strong association between the absence of TSS and the persistence of PT following surgery (Fisher exact test, $P = 0.01$) (Table V).

Similarly, none of the nonresponders had an empty sella even though the prevalence of empty sella for the entire cohort was higher than that for historical controls. For the subset of subjects with dehiscence alone,

TABLE IV.
Prevalence of Transverse Sinus Stenosis in Cases With Available Data.

| | Ipsilateral TSS* | Contralateral TSS* | Any TSS* | No TSS* | Bilateral TSS* |
|---|---|---|---|---|---|
| Total | 11 of 23 (48%) | 9 of 24 (37.5%) | 14 of 22 (64%) | 8 of 22 (69%) | 6 of 18 (33%) |
| Diverticulum | 4 of 9 (45%) | 3 of 10 (30%) | 5 of 9 (56%) | 4 of 9 (45%) | 2 of 8 (25%) |
| Dehiscence | 7 of 14 (50%) | 6 of 14 (43%) | 9 of 13 (69%) | 4 of 13 (31%) | 4 of 10 (40%) |

*Differences between cases with diverticulum and dehiscence not significant for any of the columns. Fischer exact test, $P = 1.0$, 0.7, 0.7, 0.7, and 0.6, respectively.
TSS = transverse sinus stenosis.

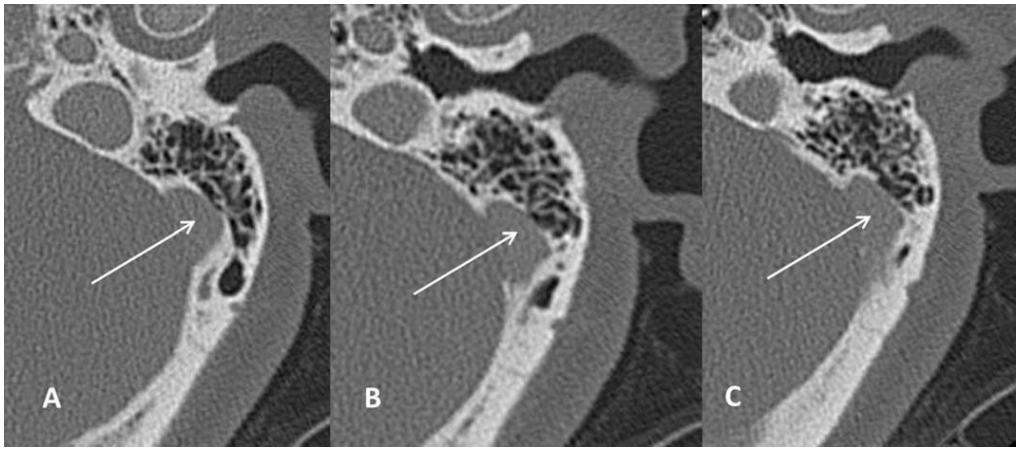


Fig. 5. (A–C) Left-sided dehiscence, nonresponder, case 30, three axial images.

the presence of an empty sella was also associated with surgical response (Fisher exact test, $P = 0.02$) (Table VI). It should be noted that there was only one responder (subject 4) who had neither TSS nor empty sella (refer to Discussion section for caveat regarding this patient).

## DISCUSSION

### *Sigmoid Sinus Wall Anomalies and Pulsatile Tinnitus: A Proposed Pathophysiology*

Sigmoid sinus wall dehiscence and diverticulum are relatively recently recognized entities that have been shown to be a cause of PT. Numerous pieces of data, including demographic and radiographic, as outlined in the Introduction section of this article, suggest an association with idiopathic intracranial hypertension. The results of the present study further support this by demonstrating a high rate of TSS and empty sella in the cohort, findings that are also common in the IIH population. Furthermore, a statistically significant absence of TSS and empty sella in patients with sigmoid sinus dehiscence whose tinnitus did not respond to sinus wall reconstruction suggests that a proximal stenosis, elevated intracranial pressure, or both, may play a pathophysiologic role not only in development of the anomaly but also in production of the sound for patients with dehiscence alone. This is suggested by the fact that none of the nonresponders in the present cohort had TSS or empty sella, implying that dehiscence alone in the absence of one of these associated findings may be necessary but not sufficient to cause PT; however, it leaves unexplained the cause of PT in the nonresponders.

The nearly constant location of SSWA immediately distal to the transverse-sigmoid junction suggests that these may evolve from mechanical perturbations of flow proximally in the transverse sinus. The high prevalence of TSS, either ipsilateral or contralateral, suggests that elevated flow velocity rates from a proximal ipsilateral stenosis, or elevated flow volume rates in a dominant sinus with a contralateral stenosis, could be responsible for progressive focal erosion of the bony sinus wall, leading to dehiscence and potentially the ultimate formation and enlargement of a diverticulum (Fig. 9A–B). Once the pathology has been established, the anomalous flow

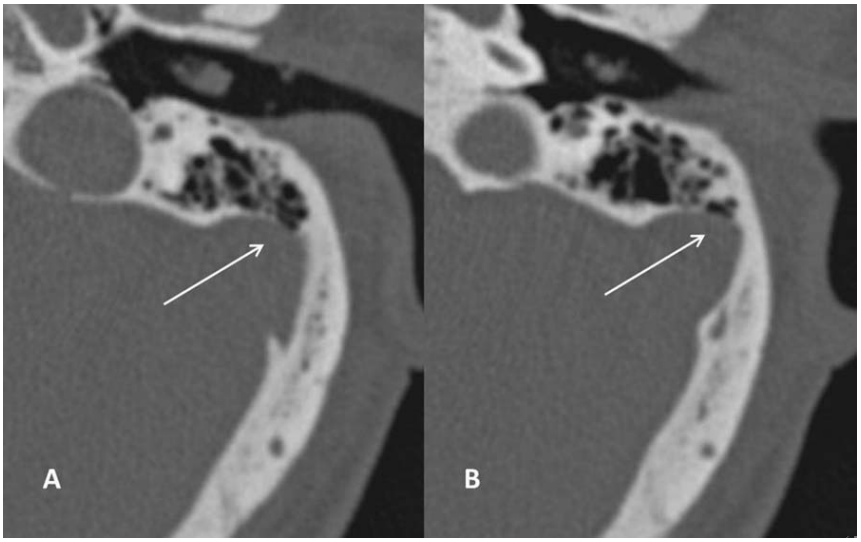


Fig. 6. (A, B) Left-sided dehiscence, nonresponder, case 31, two nonconsecutive axial images.

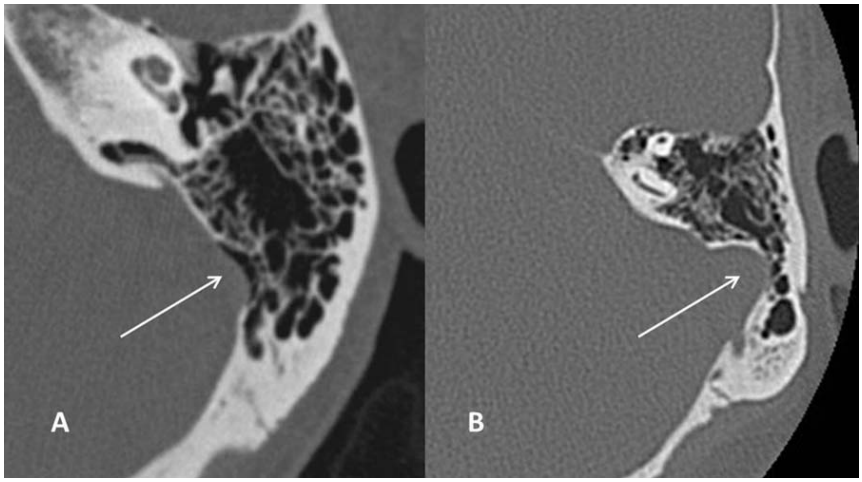


Fig. 7. (A, B) Left-sided dehiscence, nonresponders. (A) Case 36. (B) Case 20.

causes vibration of the dehiscent and/or thinned sinus wall, or turbulence in a diverticulum, and becomes audible. It is still unknown if turbulent flow is required for sound production in both processes. It is likely that there is turbulent flow in diverticula. However, it is possible that sound production from dehiscent sinus walls comes not from turbulence but rather from vibration of the dehiscent and usually thinned vessel wall, resonating like the reed of a wind instrument.

This proposed pathophysiologic mechanism suggests a delicate balance of different factors may play a role in creating the perceived sound. These factors could include degree and length of stenosis, size of dehiscence, viscosity and other flow characteristics of the blood affecting its Reynolds number, outflow resistance due to jugular vein stenosis or elevated central venous pressure (e.g., from an elevated BMI), degree of thinning of the vessel wall, or intracranial extramural venous pressure such as

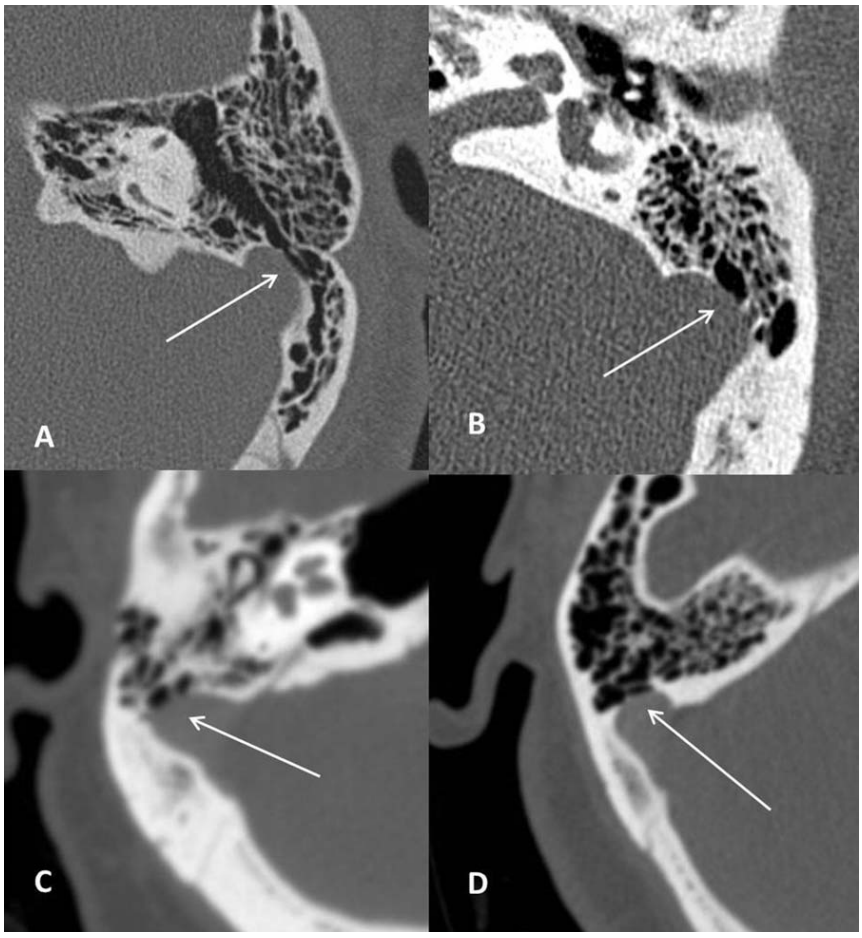


Fig. 8. Representative preoperative images from subjects with successful repair of sigmoid sinus dehiscence without diverticulum (A) subject 15, (B) subject 29, (C) subject 6, (D) subject 33.

TABLE V.
Contingency Table for Rate of TSS in Responders and Nonresponders With Available Data*

| | Responders | Nonresponders | Total |
|---|---|---|---|
| Any TSS | 9 (69%) | 0 (0%) | 9 (69%) |
| No TSS | 1 (8%) | 3 (23%) | 4 (31%) |
| Total | 10 (77%) | 3 (23%) | 13 (100%) |

*Subjects with dehiscence alone.
TSS = transverse sinus stenosis.

TABLE VI.
Contingency Table for Rate of Empty Sella in Responders and Nonresponders With Available Data.*

| | Responders | Nonresponders | Total |
|---|---|---|---|
| Radiographic: empty sella | 7 (33.3%) | 0 (0%) | 7 (33.3%) |
| Normal: sella turcica | 10 (47.7%) | 4 (19%) | 14 (66.7%) |
| Total | 17 (81%) | 4 (19%) | 21 (100%) |

*Subjects with dehiscence alone.

in patients with intracranial hypertension. It is possible that the relative weight of each of the multiple factors in the balance could differ from patient to patient but still result in the symptom of PT. This theory also suggests the possibility that, in at least some patients with dehiscence alone, treatment of the abnormal sound production might be accomplished by addressing one of the other dominant factors, if present, rather than by repairing the dehiscence itself. As mentioned earlier, these conclusions hold only for sigmoid sinus dehiscence because all subjects with a diverticulum responded to surgery regardless of the associated findings. It is certainly feasible that once a diverticulum has formed, the turbulent flow in the pouch alone would suffice to produce an audible PT, even in the absence of other contributing factors. If this multifactorial theory is correct, it could also explain the clinical observation that patients can have mild, intermittent recurrences of a less intense PT after successful surgery if they gain weight or develop IIH[25] because the relative impact of those factors would have increased. It should also be noted that this pathophysiologic mechanism does not explain why a SSWA sometimes develops ipsilateral to a TSS and sometimes contralateral to it. This could be due to underlying anatomic factors, such as relative outflow dominance, skull curvature, location or type of stenosis, or other unidentified factors, and requires further study.

The cause of surgical failure in the four subjects in this series is uncertain. The SSWA may have been an incidental finding and not the cause of their PT, or it may have been a minor portion of a multifactorial etiology, carrying insufficient weight as compared to other unidentified factors to be necessary or sufficient. Case 31 also had a high-riding left jugular bulb, with a diverticulum dehiscent into the vestibular aqueduct and thin bone over the ipsilateral superior semicircular canal, although no dehiscence—either or both of which may have been causing PT. Dong et al. found multiple vascular anomalies to be the rule in the 242 patients who they imaged with CT for unilateral PT, and they suggested that in some cases remediation of more than one anomaly would be required for cure.[10] To the best of our knowledge, no alternative etiology for PT has been identified in the other three nonresponders to date. Digital subtraction angiography and neuro-ophthalmologic evaluation were recommended for those who had not had those done preoperatively, but not all patients complied with these recommendations. A technical surgical failure can also not be excluded. These four subjects were imaged subsequently to assess the accuracy and integrity of the repair, and no technical errors could be identified.

Two nonresponders were noted intraoperatively to have a very thin layer of bone, with one or more small central foci of noncontiguous dehiscence. They were treated similarly to others with complete decompression of the affected area. A similar finding was described by Geng et al. and was also associated with poor surgical response in their series.[41] The other two nonresponders had confluent dehiscence typical of that seen in the responders.

There was only one responder for whom complete data were available and who had a unilateral dehiscence without either TSS or empty sella (case 4). The patient

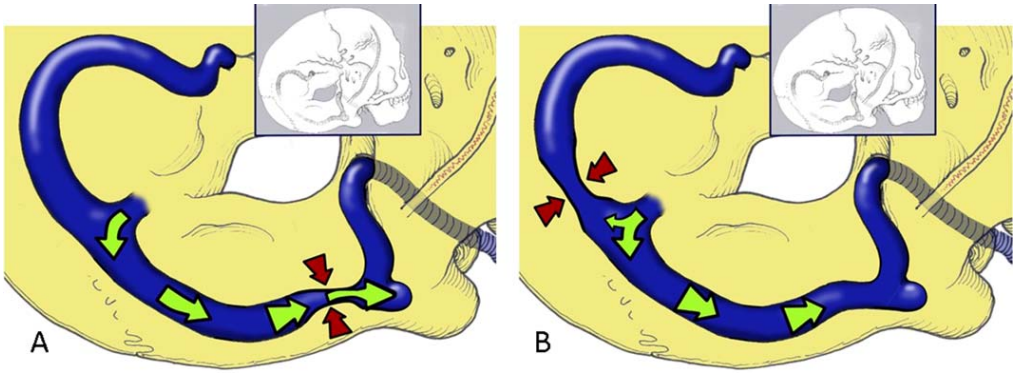


Fig. 9. Proposed pathophysiologic development of SSWA. (A) Ipsilateral transverse sinus stenosis with resultant increase in flow velocity exerting increased pressure on sinus wall. (B) Contralateral transverse sinus stenosis with resultant increase in ipsilateral flow volume.

was treated early in the series for a right-sided dehiscence with an audible bruit when examined with a Toynbee tube in the external auditory canal. The patient developed postoperative visual loss and was found to have elevated intracranial pressure, despite the absence of sinus thrombosis or stenosis, and no radiographic evidence of optic nerve sheath edema. The etiology of the patient's postoperative complication was never elucidated, but it was strongly suspected that the patient in fact may have had undiagnosed visual loss preoperatively. Perhaps some perioperative phenomenon pushed the patient over a threshold from subclinical IIH to manifest with an acute symptomatic presentation. If correct, it suggests that although the patient did not have venous outflow stenosis typical in patients with IIH, the patient may have had undetected IIH preoperatively. If that is indeed the case, there were no responders in the series with unilateral dehiscence without signs or symptoms of IIH. Caution is advised in selecting patients for surgery who have unilateral dehiscence without TSS or empty sella because these data suggest a much higher likelihood of surgical failure. Even if sigmoid sinus diverticulum is a later stage development following dehiscence, the same restrictions may not apply to treatment of diverticulum because a diverticulum, once established, probably results in turbulent flow sufficient to cause PT even in the absence of other factors. This suggestion is supported by the observed 100% success rate of surgery in patients with diverticulum.

### *Diagnostic Challenges*

There are two challenges in definitive diagnosis of PT due to SSWA: 1) identifying the abnormal radiographic finding, and 2) determining that the patient's perceived PT is due to the abnormality.

The former relates to a broader discussion about the optimal imaging technique to employ for the chief complaint of PT. Mattox and Hudgins have proposed a diagnostic algorithm to choose the proper test and pinpoint the most likely etiology.[7] Based on that decision, they recommend appropriately tailored diagnostic imaging based on the most likely diagnosis. Although this is a rational approach, it is one that may overly depend on the thoroughness and reliability of the history, physical exam, and the clinician's diagnostic acumen. Reardon and Raghavan have employed a modified CT angiography technique for imaging of venous etiologies of tinnitus.[43] This study protocol sets the image windowing as for a standard temporal bone protocol, but delays image acquisition as compared to a normal CTA protocol to ensure adequate venous filling. This technique is highly reliable for the detection of SSWA, dural venous stenoses, third mobile window pathology, middle ear and jugular foramen vascular lesions and neoplasms, otosclerosis, and other causes of conductive hearing loss. It will also identify most of the soft signs of an acquired dural vascular lesion,[44] with a possible exception of early venous filling because the injection timing is delayed. Asymmetric venous filling can still be identified. This is an efficient approach to initial diagnostic imaging evaluation. It should be noted that many clinicians rely on MR angiography, sometimes with traditional high-resolution postcontrast MR imaging as their initial study of choice. Although 3D inversion recovery images might identify major dural venous stenoses, this approach will overlook most sigmoid sinus diverticula, all sigmoid sinus dehiscence, and most third-window pathologies and otosclerosis, and is not as sensitive as CTA for acquired dural vascular lesions.[44] Proper choice of study is only the first step; careful interpretation of the images is critical, being mindful of the wide range of potential pathology that may be encountered. Sigmoid sinus wall anomalies, even some very obvious ones, are frequently overlooked, and many radiologists are not sensitive to the subtle signs of the other common pathologies. The treating otolaryngologist will need to review carefully those cases initially reported as negative. In addition, none of these studies will identify a cervical carotid bruit from stenosis or dissection. Careful clinical examination should identify such etiologies, supplemented by carotid duplex studies or CTA of the neck, if concern persists.

### *Therapeutic Decisions*

Treatment of PT suspected to be due to a SSWA should begin with a thorough evaluation to look for other potential causes, including vascular and nonvascular. In the absence of another obvious cause of the symptom, all patients should be evaluated by a neuro-ophthalmologist to exclude IIH, even those without elevated BMI. If feasible, consideration should be given to advising an initial attempt at meaningful weight loss. However, anecdotal experience suggests a low likelihood of success with this approach alone.

A variety of techniques for treatment of PT due to SSWA have been proposed. These include both endoluminal[35–37,45–47] and transmastoid/extraluminal approaches.[19,38,40,48] The former has the potential advantage of being able to simultaneously address an associated TSS and confirm adequate postprocedural vessel patency. It has the disadvantage of requiring long-term anticoagulation, as well as potential complications related to the placement of a stent or coiling material. Broad-necked diverticula cannot be excluded from the circulation without simultaneous placement of a stent to prevent migration of the coiling or other obliterative material. Endoluminal treatment has only been described for sigmoid sinus diverticulum, not isolated dehiscence. Although it is conceivable that stenting could treat PT due to dehiscence by stiffening the soft tissue sinus wall, this latter pathology lends itself more readily to transtemporal surgical repair.

The transtemporal techniques described include soft tissue grafts of fascia, dermal replacement tissues, or muscle, placed either deep to the posterior petrous face in the epidural space or directly onto the sinus wall superficial to the bone. Securing the soft tissue graft can be accomplished with bone cement, bone dust, suture, mini-plate, or a combination of these. Some authors have recommended deliberate compression of the sinus wall with more rigid grafts such as cartilage.[49]

Transtemporal approaches have the advantage of direct, binocular visualization and confirmation of the pathology, which is particularly important in the case of isolated dehiscence. Long-term anticoagulation is not required, and depending on the technique and materials employed, complication rates should be very low. It is unknown what role each layer of reconstruction plays in the success of surgery, and it is possible that there is redundancy in a multi-layered repair.

The greatest risks of transtemporal sinus wall reconstruction, aside from the potential for persisting PT, relate to the disruption of venous outflow and elevation of intracranial pressure. Extraluminal compression and intraluminal thrombosis have both been described.[19,50] Because the latter is typically a more concerning problem, a modest regimen of perioperative coagulation may be beneficial, using 325 mg of aspirin and 75 mg of clopidogrel daily starting the evening after surgery. Currently, clopidogrel is continued for 2 weeks after surgery and aspirin for 4 weeks. Although it is possible this could increase the risk of extraluminal compression from an epidural hematoma, that problem is less likely to be symptomatic and more likely to resolve uneventfully. Postoperative imaging in the absence of concerning symptoms is not required, although a contrast-enhanced CT of the temporal bones to assess the reconstruction and sinus patency may be considered. Typical postoperative imaging findings have been reported.[50] Cases of asymptomatic extraluminal compression may be managed expectantly without complications.

Clinical suspicion for elevated intracranial pressure should be high if the patient complains of persisting or worsening headache more than 1 week after surgery. Such a complaint should trigger imaging and urgent neuro-ophthalmologic evaluation. Intraluminal thrombosis should be treated with more aggressive anticoagulation, in consultation with a vascular neurologist, if not otherwise contraindicated. Extraluminal compression could be potentially treated with angioplasty, if clinically indicated. Lumbar puncture should be considered and could be both diagnostic and therapeutic. Persistently elevated pressure despite medical therapy might require a neurointerventional procedure or subarachnoid shunting.

### *Study Limitations*

There are a number of limitations to this study. The retrospective nature of the data collection, coupled with a recent rapid growth in knowledge about SSWA, resulted in numerous absent data points, particularly from subjects evaluated and treated earlier in the series. Although the significance of this data, despite the limitation of power, suggests a robust result, future studies with prospective these data collection are needed. Data analysis was also hampered to a certain extent by the small number of surgical nonresponders. Logistic regression would have been an optimal statistical approach for a priori, hypothesis-driven analysis of this type of data set, which was attempted. However, because there were too few events (surgical failures) and multiple interdependent variables, the model suffered from multicollinearity and was not viable.

### *Future Directions*

These data provide strong support for an association between SSWA and TSS and may provide the link, or at least a link explaining the known association of PT and IIH. There is still much to learn about these relationships. Among other unknowns discussed above, more specific information about the role of TSS is needed. Do flow aberrations from a TSS create SSWAs, produce PT in the presence of an existing SSWA, or both? What degree, type, and location of TSS are required for whatever role they play? Are the effects of a focal arachnoid granulation similar to those of a tapered extraluminal stenosis? What are the relative roles of other variables, such as outflow pressure and viscosity, in the production of the final phenotype? Reliable bench and/or computer models that faithfully reproduce the in vivo physiology would allow for manipulation of individual variables and would help elucidate the relative weight of and interaction between the different variables. Precise knowledge of that information would help determine optimal treatment protocols, which may differ from patient to patient.

## CONCLUSION

Sigmoid sinus diverticulum and dehiscence are important causes of pulsatile tinnitus that often are overlooked in this patient population. Most, although not all, affected patients are overweight women in their childbearing years. The pathology typically affects a dominant or codominant sigmoid sinus and is strongly associated with transverse sinus stenosis and empty sella. The stenosis may be ipsilateral or contralateral to the SSWA. Patients with PT and a SSWA should be screened for idiopathic intracranial hypertension, and medical treatment should be initiated if this is identified. Both endoluminal and transtemporal approaches to treatment have been described and have met with high rates of success. Intervention for patients with sigmoid sinus dehiscence without an associated transverse sinus stenosis or empty sella should not be undertaken without a clear understanding in the informed consent process that failure to achieve symptom relief is highest in this subpopulation


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