Sinus pericranii is a vascular malformation composed of extracranial veins and a prominent venous varix connected to an intracranial venous sinus via emissary veins. Although rare traumatic associations have been reported, most suggest a congenital etiology for this malformation, given its association with developmental venous anomalies and other vascular lesions. Sinus pericranii was first described by Hecker in 1845 and elaborated by Stromeyer in 1850. Of this curiosity Stromeyer wrote that it “…consists of a blood bag on the skull, which stands in connection with the veins of the diploe and through these with the sinuses of the brain….”

Due to the rarity of sinus pericranii, its pathogenesis and natural history remain unclear. Thus, the optimal management strategy for this lesion is also unknown. Symptomatic sinus pericranii or those resulting in a cosmetic deformity are strongly considered for surgical obliteration. Reports of an association with hemorrhage, sinus thrombosis, intracranial hypertension, and infection have led some to aggressively treat asymptomatic sinus pericranii. Decision making is complicated by difficulty in determining which sinus pericranii are safe for surgical intervention.

The angioarchitectural classification of sinus pericranii into dominant or accessory varieties is an attempt to facilitate making a determination about which lesions may be safely obliterated. The late Pierre Lasjaunias and co-workers defined dominant lesions as those where the main stream of contrast material flow uses the sinus pericranii to drain the brain, bypassing the usual venous outlets. In contrast, accessory lesions were defined as those in which only a small part of the venous outflow occurs through the extradiploic vessels. This qualitative angiographic designation has become the main factor in determining whether a sinus pericranii can be treated. It is believed that dominant sinus pericranii must be preserved while accessory sinus pericranii are candidates for surgical intervention.

Although we have found the aforementioned classification scheme helpful for surgical decision making, we have developed a protocol for additional angiographic interrogation of sinus pericranii that may further ensure the safety of intervention. The lesion compression technique we describe here enables one to make an assessment of the adequacy of intracranial venous outflow during temporary functional absence of a sinus pericranii. Additional assessment allows the surgeon or interventional specialist to proceed confidently with obliteration of a sinus pericranii deemed redundant within the intracranial circulation.
Presentation and Noninvasive Imaging

A 7 month-old, healthy female infant presented electively with a pulsatile, right parasagittal scalp mass (Video 1).

**VIDEO 1.** Clip showing pulsatile scalp mass. Copyright Jason A. Ellis. Published with permission. Click here to view.

As per the parents’ assessment, modest growth over several months was noted. Doppler ultrasonography demonstrated spectral waveforms within the scalp concerning for a vascular malformation (Fig. 1A). Subsequent MRI showed a small tangle of prominent flow voids in the region of interest, with 1 vessel traversing the parietal calvaria and draining into the superior sagittal sinus (Fig. 1B and C).

Angiographic Technique

Although a diagnosis of sinus pericranii was made noninvasively and the typically benign nature of the condition was described, the parents wished to proceed with intervention if deemed safe. Thus, catheter cerebral angiography was performed to more fully assess the angioarchitecture of the sinus pericranii and the associated venous drainage pattern of the brain.

Given the patient’s young age, the procedure was performed under general endotracheal anesthesia to ensure high-resolution imaging without motion artifacts and to allow for direct transfer to the operating suite for excision if deemed appropriate. Following sterile preparation and draping, a 4-Fr arterial sheath was placed in the right common femoral artery using a modified Seldinger technique with a micropuncture needle. A 4-Fr hockey-stick catheter was passed over a Bentzon guidewire to sequentially obtain standard 6-vessel arteriography, including the bilateral common and internal carotid arteries as well as the bilateral vertebral arteries. The sinus pericranii was noted to opacify at the expected interval in the venous phase without arteriovenous shunting. This occurred in conjunction and connection with the superior sagittal sinus draining to the scalp veins (Fig. 2A and B). The sinus pericranii drained only a small portion of the intracranial venous outflow and was, by definition, accessory.\(^23\)

A pressure dressing was subsequently applied to the scalp directly above the sinus pericranii. Repeat arteriography with external compression in place demonstrated normal arteriovenous transit opacification of the intracranial vessels, but no opacification of the extracranial scalp vessels through the sinus pericranii (Fig. 2C and D). The intracranial venous drainage pattern, including that of the superior sagittal sinus, was normal, suggesting that the sinus pericranii may be safely excised.

Surgical Excision

The patient was transported to the operating theater while under the same general anesthesia used to perform catheter angiography. Positioning was prone with the head
on a cerebellar headrest (Fig. 3). After usual prepping and draping, a slightly parasagittal incision was made near the vertex slightly to the right of midline with care taken not to incise the underlying venous anomaly. Using bipolar and sharp dissection, numerous scalp veins as well as a single feeder to the sinus pericranii from the sagittal sinus was identified, cauterized, and ligated. The subgaleal plane was swept circumferentially down to the level of the bone, with stripping of the periosteum a distance of 3–4 cm radially. The pericranium was reduced with cautery to the edge of the skull defect through which the sagittal sinus communicated. At this point, the pulsations in the scalp from the malformation were completely absent. Final inspection, putting the patient in the Trendelenburg position, and executing a Valsalva maneuver indicated no evidence of residual sinus pericranii or abnormal scalp feeders/tributaries. The patient tolerated the procedure well and was discharged home neurologically intact on the first postoperative day.

**Discussion**

Although more than 100 cases of sinus pericranii have been reported in the literature, its relative rarity has precluded standardization of diagnosis, classification, and management. Not surprisingly, the majority of reported cases are unclassified with respect to dominance. Most reports suggest that sinus pericranii are clinically benign lesions that predominantly raise cosmetic concerns. However, rare reports of hemorrhage (spontaneous and traumatic), infection, air embolism, intracranial hypertension, and sinus thrombosis have led some practitioners and patients to pursue aggressive intervention. Accurate clinical and radiographic assessment of sinus pericranii is therefore essential prior to intervening on this probably benign lesion.

Févre and Modec proposed the first classification system for sinus pericranii in 1936. Their scheme included descriptions of sinus pericranii as: 1) closed systems arising from and draining into intracranial sinuses; 2) draining systems that act as collaterals for intracranial flow; and 3) extracranial lesions draining into intracranial sinuses. Gandolfo and colleagues proposed a more clinically useful categorization, explaining that dominant sinus pericranii are untreatable because they serve as a major venous outflow channel to the intracranial compartment, whereas accessory sinus pericranii are highly treatable because only a small portion of the intracranial venous outflow traverses them. Classification of sinus pericranii as dominant or accessory has arguably been the most important criterion for determining whether this anomaly may be safely obliterated. In our opinion, the compression technique we describe for functionally, but reversibly, obliterating sinus pericranii provides additional assurance that intracranial venous outflow will not be compromised by resection or embolization. Discordant pre- and post-compression venous phase angiography favors conservative management. Although not performed in our case, use of awake neurological testing and/or evoked potential monitoring may provide additional assurances.

It is notable that various compression maneuvers in patients with sinus pericranii have been previously described. However, we are unaware of these techniques being used in the manner we describe to determine therapeutic options. Volkman in 1950 used direct compression to differentiate true sinus pericranii from pseudo–sinus pericranii (hemangioma, cavernoma, etc.). Kaido et al. described compression of the jugular veins during cerebral angiography, but this maneuver provided little information for clinical decision making. Madsen and colleagues employed compression of a cutis aplasia lesion associated with a sinus pericranii during CT venography to assess intracranial drainage patterns.

**Conclusions**

Sinus pericranii are benign congenital vascular malformations associated with a rare incidence of complications. Treatment via endovascular or open surgical methods should be attempted only after angiographic confirmation that the lesion is accessory. We believe that direct sinus pericranii compression during catheter angiography is a safe way to determine if intracranial venous outflow may be normal after lesion treatment. Validation of this technique in additional patients will be necessary to confirm this finding.
References


Disclosure
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Ellis. Acquisition of data: Ellis, Mejia Munne. Analysis and interpretation of data: Ellis, Feldstein, Meyers. Drafting the article: Ellis, Mejia Munne. Critically revising the article: Ellis, Feldstein, Meyers. Reviewed submitted version of manuscript: Ellis, Feldstein, Meyers. Approved the final version of the manuscript on behalf of all authors: Ellis.

Supplemental Information
Video

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