Suboccipital decompression during posterior cranial vault remodeling for selected cases of Chiari malformations associated with craniosynostosis

Clinical article

WILLIAM W. SCOTT, M.D.,1 JEFFREY A. FEARON, M.D.,2 DALE M. SWIFT, M.D.,3 AND DAVID J. SACCO, M.D.3

1Department of Neurosurgery, University of Texas Southwestern Medical Center; 2Department of Plastic and Craniofacial Surgery, Medical City Hospital; and 3Department of Pediatric Neurosurgery, Children’s Medical Center, Dallas, Texas

Object. The optimal management of Chiari malformations in the setting of craniosynostosis is not well established. In this report the authors describe their outcomes with the combined technique of simultaneous suboccipital decompression (SOD) during posterior cranial vault remodeling (PCVR).

Methods. A retrospective review was performed of all patients undergoing PCVR and simultaneous SOD. Demographic data, diagnosis, imaging studies, operative intervention, and clinical follow-up were evaluated.

Results. Thirty-four patients were identified as having undergone a simultaneous PCVR/SOD for Chiari malformation associated with craniosynostosis. Eighty-eight percent of these patients had syndromic, multisutural craniosynostosis, and the remaining patients had unilateral lambdoid craniosynostosis. There were no postoperative complications as a direct result from this combined procedure. Two patients required a subsequent direct approach for decompression of the Chiari malformation. The interval between these subsequent surgeries was 3 years and 19 months.

Conclusions. Chiari malformations are commonly associated with syndromic, complex craniosynostosis and isolated lambdoid craniosynostosis. In appropriately selected patients, a combined posterior cranial vault enlargement and SOD of the foramen magnum is associated with a low complication rate and appears to be an effective procedure.

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KEY WORDS • Chiari malformation • suboccipital decompression • posterior cranial vault remodeling • craniosynostosis

Chiari malformations are commonly associated with both syndromic and nonsyndromic craniosynostosis, including multisutural and isolated lambdoid craniosynostoses.1–3,5–7,9,10 In this population of patients, the Chiari malformation is thought to be acquired secondary to abnormal skull development as a result of premature suture fusion(s).2,3,11 These fusions result in diminished cranial volumes, with small posterior fossae predisposing this population to cerebellar tonsillar herniation and its potential consequences.12,13,17,18 These findings distinguish these patients from those in the general population with Chiari malformation Type I.

The optimal management of Chiari malformation in the setting of craniosynostosis is not well established. In addition, experience gained from treating and observing patients with Chiari malformation in the general population may not be relevant given the fundamental differences in skull growth between patients with craniosynostosis and those without. Beginning in 1995, we began selectively treating patients with craniosynostosis-associated Chiari malformation with posterior cranial vault enlargements and in-continuity SOD of the foramen magnum. In select cases, in which substantial transosseous compensatory occipital venous drainage was encountered, a variation of this combined procedure was performed that permitted preservation of this important collateral venous outflow. The purpose of this report is to describe this unusual surgical approach and to examine our outcomes with this combined technique.

Methods

After obtaining institutional review board approval, we performed a retrospective review of all patients undergoing PCVR and simultaneous SOD. These operations were performed by a single craniofacial surgeon (J.A.F.) and 1 of 2 pediatric neurosurgeons (D.M.S. or D.J.S.). Demographic data, diagnosis, imaging studies, operative intervention, and clinical follow-up were evaluated.

Surgical Technique

Patients were placed prone on a horseshoe headrest. A coronal incision was used in all cases. The scalp was
reflected to the level of the superior nuchal line, and then a subperiosteal dissection was performed with elevation of the musculature to the level of the foramen magnum. Importantly, abnormal venous drainage is common in this patient population and is often noted exiting the central occipital bone at or above the level of the confluence of sinuses (Fig. 1). Sometimes, this can be clearly appreciated on standard preoperative MRI (Fig. 2). Very early in our experience, this finding would preclude us from attempting a combined PCVR/SOD. However, with experience, we sought to preserve these anomalously enlarged veins by leaving them in a small “island” of surrounding bone (Fig. 3), maintaining the scalp drainage. In conjunction with the preparatory craniectomy for PCVR, the anomalous exiting venous drainage is identified, and the bone through which it penetrates is preserved, effectively creating this island of bone. Once this venous channel has been preserved, the subperiosteal dissection then proceeds inferiorly around both sides of this island of bone to fully appreciate and subsequently remove the suboccipital bone up to the lateral margins of the foramen magnum.

This exposure also assists in cases of lambdoid synostosis, where the typical lambdoid correction entails a lower osteotomy that is less than 2 cm away from the posterior aspect of the foramen. Cervical laminectomies were performed in select cases; however, the dura matter was left intact in all cases in an effort to minimize overall operative risk. A PCVR was performed, based on the presenting skull configuration, to significantly enlarge the overall cranial volume to include the posterior fossa (Fig. 4). Postoperative care was similar to that for other craniosynostosis patients. Follow-up MRI was performed to monitor the position of the cerebellar tonsils (Fig. 5).

Results

Thirty-four patients were identified as having undergone a simultaneous PCVR/SOD for Chiari malformation associated with craniosynostosis. The operative dates spanned from 1995 to 2011. The mean age of these patients at the time of surgery was 23.6 months (range 10 months to 9 years). There were 16 males and 19 females (46% and 54%, respectively). The mean follow-up was 3.8 years (6 months to 14 years).

The majority of these 34 patients (88%) had syndromic multisutural craniosynostosis including 9 with Pfeiffer syndrome, 8 with Apert syndrome, 6 with Mercedes-Benz pattern complex craniosynostosis, 5 with Crouzon syndrome, 1 with Shprintzen-Goldberg syndrome, and 1 with an unusual 6p duplication (with a Pfeiffer-like phenotype). The remaining 4 patients (12%) had unilateral lambdoid craniosynostosis.

Twenty-one patients (62%) underwent prior cranial vault procedures. In 13 of these patients (38%) a previous anterior cranial vault remodeling had been performed, and in 5 (15%) a previous PCVR was performed. In 3 cases (9%) both a previous anterior cranial vault remodeling and PCVR had been performed.

Hydrocephalus was present in 17 patients (50%). Of these, 15 underwent prior surgical treatment for hydrocephalus. At the time of the PCVR/SOD, 10 patients had a ventriculoperitoneal shunt in place and 5 patients had a functional endoscopic third ventriculostomy. In the 2 patients who did not undergo preoperative treatment of their hydrocephalus, the PCVR/SOD alone has been adequate treatment and no further intervention has been necessary.

A C-1 laminectomy was performed in 8 cases (24%). The rationale for performing a C-1 laminectomy varied but included syringomyelia, tonsillar herniation beyond the C-1 ring and radiographic spinal cord compression, and myelomalacia. A C-2 laminectomy was performed in 1 case. The inconsistent inclusion of cervical laminectomies does not allow for conclusions regarding its effectiveness, but it does show that it is possible with this exposure.

Syringomyelia was present in 7 patients. Five of these patients had postoperative images available for review. In all 5 cases, the syrinx was found to be significantly smaller postoperatively. In addition to these 7 patients with syringomyelia and the 2 patients with untreated hydrocephalus, 5 other patients had symptoms that were thought to be related to the Chiari malformation. This included 4 patients with central sleep apnea and 1 patient with progressive swallowing difficulties.

Twenty patients underwent a PCVR/SOD without obvious clinical symptoms referable to the Chiari malforma-
tion. This was a subset of patients in whom a PCVR was indicated for poor skull growth and intracranial volume concerns, in combination with a Chiari malformation noted on preoperative imaging. In 19 of these cases, the Chiari malformation was radiographically progressive compared with prior images. Postoperative imaging was available for review in 71% of these cases. Improvement in the posterior fossa size was noted in all cases. The Chiari malformation resolved in 35%, was improved in 35%, and was stable radiographically in 30%.

There were no postoperative complications that appeared to be the direct result of this new procedure. However, 2 patients (6%) required a subsequent direct approach for decompression of the Chiari malformation. The rationale for repeat surgery was persistent tonsillar herniation resulting in worsening central sleep apnea in one patient with Crouzon syndrome and the development of a new syrinx in another patient with a unilateral lambdoid craniosynostosis. The interval between surgeries was 3 years and 19 months, respectively. In these subsequent surgeries, a midline suboccipital incision was made, the prior suboccipital craniectomy was exposed and slightly widened, a C-1 laminectomy was performed, and a duraplasty was completed.

**Discussion**

The association between craniosynostosis and Chiari malformation is well recognized and can be found in both syndromic and nonsyndromic patients. In 1972 Saldino et al. described the association between Chiari malformation and craniofacial synostosis. In 1998 Cinalli et al. mentioned a simultaneous SOD and occipital remodeling; yet to this day, significant controversy exists as to the best management of this clinical entity. Some centers have argued that the Chiari malformation should be addressed only if the patients are symptomatic; on the other hand, other groups have recommended decompression of the posterior fossa for all cases of Chiari malformation associated with craniosynostosis regardless of symptoms.

Most believe that Chiari malformation in the setting of craniosynostosis is an acquired defect, occur-
Suboccipital decompression during cranial vault remodeling

ring sometime during the postnatal period as a result of premature fusion of the cranial sutures. Historically, studies noted a higher incidence of Chiari malformation associated with Crouzon syndrome (in upward of 70% of cases), followed by oxycephaly and Pfeiffer syndrome. More recently, certain subtypes of Pfeiffer syndrome have been shown to have the highest incidence, approaching 100% in these cases. An association of Chiari malformation with Apert syndrome has also been noted in 29% of cases. Consistent with these observations, 88% of our patients had syndromic multisuture craniosynostosis.

Complicating the management of Chiari malformation in this population is the frequent coexistence of hydrocephalus and venous hypertension. This, combined with the abnormal skull anatomy, differentiates this population of patients with Chiari malformation from the patients with Chiari I malformation in the general population. In a recent review of 383 consecutive patients treated for craniosynostosis over a 15-year period, Strahle et al. noted Chiari malformations in 29 patients. Seventeen of these cases were diagnosed before craniosynostosis repair, and 12 cases were diagnosed after, highlighting the dynamic nature of Chiari malformation in patients with craniosynostosis. Of the 17 patients with Chiari malformation diagnosed prior to craniosynostosis repair, 10 underwent SOD either concurrently (5 patients), prior to craniosynostosis repair (2 patients), or after repair (3 patients). During follow-up, 12 patients were eventually diagnosed with a Chiari malformation, including 7 patients with an associated syrinx. Six of these patients ultimately underwent a Chiari decompression at a mean of 3.4 years after the craniosynostosis repair. The follow-up of the patients diagnosed with a Chiari malformation, but who were not treated, is not given. Summarizing these data, in their series, the Chiari malformation was surgically treated in 16 of the 29 patients: 5 during a combined procedure and 11 at a separate operative setting. It was their observation, and has been that of others, that it is possible for the radiographic appearance of the Chiari malformation to improve after craniosynostosis repair alone. Interestingly, this has been our experience as well. However, possibly given the high percentage of children with syndromic and multisutural craniosynostosis whom we have encountered, we have not found this to be durable over time.

How effective have these Chiari decompressions been, especially when found in conjunction with the craniosynostoses? Sacco and Scott examined reoperations for Chiari malformation in a 14-year retrospective review. Sixteen patients (16%) were undergoing reoperations for Chiari I malformation. However, of the population of patients with Chiari I malformation, 9 patients also had craniosynostosis. When this group alone was examined, 5 (55%) required reoperations. The authors opined that the population of patients with Chiari malformation and craniosynostosis may have a different natural history and may require a different approach from that used in Chiari malformation in the general population.

Given the rarity of Chiari malformation in children with syndromic synostosis, it would be extremely difficult to demonstrate when posterior decompression is sufficient treatment versus when it is not sufficient. It is likely that this is largely dependent on at least 2 factors: how far inferiorly the tonsils have descended and how tight the foramen magnum is (that is, how much skull base growth inhibition is present). We presume one would appreciate that those children with tight Chiari malformations are not going to appreciate sufficient decompression with an isolated posterior expansion. In this current series, we were motivated to reduce the total number of operations that children with craniosynostosis and Chiari malformation must undergo. With this in mind, beginning in 1995, we initiated a combined posterior skull enlargement procedure, with an in-continuity SOD of the foramen magnum. As this was an evolving technique, our indications slowly changed. We began by utilizing this approach for those with severe or symptomatic Chiari malformation who also required calvarial enlargement. However, once we realized how facile this additional step was, we began treating children whom we believed were at significant risk for developing symptoms. Our current review of 34 cases suggests that this combined procedure can be performed safely.

In this challenging subgroup of patients, a complex venous anatomy is often encountered (with large transossosseous venous channels occurring in the region of the confluence of sinuses), particularly among the more severe subtypes of Apert and Pfeiffer syndromes. Previous reports have attributed a perioperative death to intraoperative ligation of enlarged posterior transossosseous venous drainage. Along the same lines, separate reports have been published, recommending avoiding surgical intervention because of the perceived risk to the venous system. In an effort to maximize venous drainage, we developed a technique to preserve these outflow tracts by leaving an island of surrounding bone through which anomalous venous channels course, while still decompressing the foramen magnum with this same surgical exposure. It has been our experience that this anatomy is predictable and may be adequately imaged with standard preoperative MRI. Thus far, we have yet to encounter a large transosseous channel inferior to the region of the confluence of sinuses.

This retrospective review suggests that a combined posterior cranial vault enlargement with an in-continuity SOD of the foramen magnum can be effective at addressing presenting pathology, including progressive tonsillar herniation, syringomyelia, central sleep apnea, and hydrocephalus. In addition, it appears that this combined procedure is not associated with a significantly higher complication rate. Given that one of our surgical goals is to minimize the total number of operative interventions in these complex patients, we have performed this combined procedure on select patients who require a PCVR and who also have a Chiari malformation, whether it is overtly symptomatic or not. It is our contention that this adds very little to the risk of the procedure and may decrease the need for subsequent surgical interventions. How many operations are being saved by combining these procedures is a good question that is obviously difficult to ascertain with a retrospective study. However, there have been previous reports that demonstrate that the Chiari malformations associated with the syndromic synostoses (and the complex synostoses) are typically progressive. Our experience in treating a fairly significant number of
patients suggests that a high percentage of children require subsequent decompression. In terms of recurrence, 2 patients (6%) went on to develop a progressive Chiari malformation, with both clinical and radiographic consequences. These 2 patients underwent secondary direct decompressions with a duraplasty at a mean of 3.6 years after the initial procedure, a rate that we believe compares favorably with the known natural history of Chiari malformation in this patient population.2,3,6

After our review, we remain enthusiastic about a combined posterior cranial vault enlargement and SOD of the foramen magnum. However, we recognize that our retrospective study design does not permit comparisons to 2-stage procedures (vault enlargement and subsequent Chiari decompression). It is also important to note that although we have observed patients in this series for up to 9 years postoperatively, our average length of follow-up of just less than 4 years is still relatively short, so the incidence for subsequent secondary decompressions might be underrepresented. We believe that for these complicated patients, the presence of an anomalous transccipital cerebral-scalp drainage system is not a definitive contraindication to a posterior decompression, as a surgical preservation of this system is possible. Obviously, each surgeon will need to balance the individual risks and benefits of this combined procedure for their patients.

Conclusions

Chiari malformations are commonly associated with syndromic, complex craniosynostosis and isolated lambdoid craniosynostosis. We have found that in a selected group of patients with craniosynostosis and Chiari malformation, a combined posterior cranial vault enlargement and SOD of the foramen magnum (including the preservation of compensatory transccipital venous drainage) is associated with a low complication rate and appears to be an effective procedure. Use of this technique may decrease the total number of surgical interventions required in these complex cases.

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 to the study and manuscript preparation include the following. Conception and design: Scott, Sacco. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: Scott. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Scott. Administrative/technical/material support: Fearon, Swift, Sacco. Study supervision: Sacco.

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