Chiari malformation associated with craniosynostosis

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Object. Chiari malformation (CM) Type I is frequently associated with craniosynostosis. Optimal management of CM in patients with craniosynostosis is not well-established. The goal of this study was to report on a series of pediatric patients with both craniosynostosis and CM and discuss their management.

Methods. The authors searched the medical records of 383 consecutive patients treated for craniosynostosis at a single institution over a 15-year period to identify those with CM. They recorded demographic data as well as surgical treatment and outcomes for these patients. When MR imaging was performed, cerebellar tonsillar descent was recorded and any other associated findings, such as hydrocephalus or spinal syringes, were noted.

Results. A total of 29 patients with both CM and craniosynostosis were identified. Of these cases, 28% had associated occipital venous abnormalities, 45% were syndromic, and 52% also had hydrocephalus. Chiari malformation was more likely to be present in those patients with isolated lambdoid synostosis (55%), multisuture synostosis (35%), and pansynostosis (80%), compared with patients with coronal synostosis (6%) or sagittal synostosis (3%). All patients underwent surgical repair of craniosynostosis: 16 had craniosynostosis repair as well as CM decompression, and 13 patients did not undergo CM decompression. Of the 7 patients in whom craniosynostosis repair alone was performed, 5 had decreased tonsillar ectopia postoperatively and 5 had improved CSF flow studies postoperatively. Both patients with a spinal syrinx had imaging-documented syrinx regression after craniosynostosis repair. In 12 patients in whom CM was diagnosed after primary craniosynostosis repair, 5 had multiple cranial vault expansions and evidence of elevated intracranial pressure. In 5 cases, de novo CM development was documented following craniosynostosis repair at a mean of 3.5 years after surgery.

Conclusions. Chiari malformation is frequently seen in patients with both multi- and single-suture lambdoid craniosynostosis. Chiari malformation, and even a spinal cord syrinx, will occasionally resolve following craniofacial repair. De novo development of CM after craniosynostosis repair is not unusual.

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KEY WORDS • Chiari malformation • craniosynostosis • surgical treatment

THE association between CM and craniosynostosis has been recognized for several decades.36 Chiari malformation occurs in patients with both syndromic and nonsyndromic forms of craniosynostosis.5,7,17,18,41,42 Up to 70% of individuals with Crouzon syndrome and 50%–82% of those with Pfeiffer syndrome have associated CM.5,6,7 There are also several reports of CM with nonsyndromic synostosis involving the sagittal, coronal, and even the metopic sutures.17,25,41 Cerebellar tonsillar ectopia in patients with craniosynostosis is thought to arise from disproportionately slow growth or the small size of the posterior fossa in many cases.5 Hydrocephalus, venous hypertension, and associated congenital brain anomalies have also been proposed as possibly important factors leading to CM development in these patients.5,34,39,60 The optimal management of CM in the setting of craniosynostosis is not well established. Some surgeons advocate simultaneous surgical correction of craniosynostosis and CM, and others suggest that CM should only be treated if it is symptomatic or associated with a syrinx.5,7 We describe our experience with a group of children with synostoses of one or more cranial sutures and CM, and we propose a management strategy for these complex cases.

Methods

We performed a retrospective single-center review of all patients under 18 years of age who were treated for craniosynostosis between 1994 and 2009. The University of Michigan institutional review board granted approval for the study. For this analysis, craniosynostosis was defined as clinical or radiological evidence of premature fusion of at least one cranial suture. Chiari malformation was defined as cerebellar tonsillar descent greater than or equal to 5 mm below the foramen magnum.23,28 In each
case, we recorded demographic information, characteristics of the craniosynostosis including suture location, and the number of sutures involved. When pre- or postoperative imaging studies were performed (Table 1), we recorded characteristics of the CM including a measurement of tonsillar descent below the foramen magnum, CSF flow analysis data on cine MR imaging, and any associated findings on imaging including venous anomalies and hydrocephalus. Cerebrospinal fluid flow data were recorded for all patients in whom dedicated CSF flow sequences were shown on MR imaging. At our institution, changes in signal intensity on sagittal phase-contrast CSF flow studies are observed in the CSF spaces anteriorly and posteriorly at the level of the cervicomедullary junction. The alternating bright and dark signals seen in the CSF spaces in the cine mode are diminished or absent when there is abnormal flow. On the axial and sagittal phase-contrast images, any change in the signal intensity of the cerebellar tonsils in the cine mode suggests tonsillar pulsations. Cerebrospinal fluid flow was categorized as abnormal if flow was decreased anteriorly or posteriorly at the foramen magnum or decreased at the foramen magnum with abnormal tonsillar pulsations based on the initial report of the radiologist. Surgical results were recorded for those patients who underwent craniosynostosis repair with or without CM decompression.

## Results

A total of 383 individual patients underwent surgical craniosynostosis repair over the specified time interval. Of these patients, 183 children were evaluated for isolated sagittal synostosis; 80 for isolated coronal synostosis; 71 for isolated metopic synostosis; 9 for isolated lambdoid synostosis; and 40 for multisuture synostosis craniosynostosis. Forty-six patients (12%) were diagnosed with a craniofacial dys syndrome. Specifically, 16 had Crouzon syndrome, 9 had Pfeiffer syndrome, 9 had Saethre-Chotzen syndrome, 2 had craniofacial dys dyssynostosis, and 1 had Norman-Roberts syndrome. Chiari malformation was diagnosed in 29 (8%) of the patients treated for craniosynostosis (Table 2, Fig. 1). Children with single-suture lambdoid synostosis (p < 0.001) or multisuture craniosynostosis (p < 0.001) were much more likely to have associated CM than all other patients with craniosynostosis (Fig. 2). Of the 9 individuals with isolated lambdoid synostosis, 5 (56%) were diagnosed with CM (Figs. 3 and 4) in contrast to no patients (0%) of those 71 with isolated metopic synostosis, 5 (3%) of the 183 with isolated sagittal synostosis, and 5 (6%) of the 80 with isolated coronal synostosis. Of the 40 patients with multisuture synostosis, 5 had pansynostosis defined as premature closure of all sutures (metopic, coronal, sagittal, and lambdoid), and 4 (80%) of these patients were diagnosed with CM. In patients with multisuture synostosis, 14 (35%) of 40 had CM compared with 15 (4%) of the 343 with single-suture involvement (p < 0.001). Including 5 patients with single-suture lambdoid craniosynostosis as well as 12 patients with lambdoid suture involvement in multisuture craniosynostosis, lambdoid synostosis was found in 17 patients (59%) with CM. Multisuture craniosynostosis without lambdoid suture involvement was not significantly more likely to be associated with CM (Table 2) than single-suture disease (p = 0.3). Of the patients with CM, 15 (52%) had associated hydrocephalus. Of the 29 patients with craniosynostosis and CM, 10 (34%) also had syringomyelia and 8 (28%) had associated abnormalities of cerebral venous drainage.

Each of the 29 patients underwent surgical correction for their craniosynostosis. The mean age at time of craniosynostosis repair was 1.8 years (range 2 months to 9 years). Of the patients who underwent surgical craniosynostosis repair over the specified time interval, 40 had CM compared with 15 (4%) of the 343 with single-suture involvement (p < 0.001). Including 5 patients with single-suture lambdoid craniosynostosis as well as 12 patients with lambdoid suture involvement in multisuture craniosynostosis, lambdoid synostosis was found in 17 patients (59%) with CM. Multisuture craniosynostosis without lambdoid suture involvement was not significantly more likely to be associated with CM (Table 2) than single-suture disease (p = 0.3). Of the patients with CM, 15 (52%) had associated hydrocephalus. Of the 29 patients with craniosynostosis and CM, 10 (34%) also had syringomyelia and 8 (28%) had associated abnormalities of cerebral venous drainage.

### Table 1: Preoperative and postoperative imaging in patients with craniosynostosis according to suture involvement

<table>
<thead>
<tr>
<th>Craniosynostosis Type</th>
<th>No. of Patients</th>
<th>No. of Preop Studies (%)</th>
<th>No. of Postop Studies (%)</th>
<th>Mean MRI Follow-Up in Yrs (range)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>CT</td>
<td>MRI</td>
<td>CT</td>
</tr>
<tr>
<td>sagittal</td>
<td>183</td>
<td>161 (88)</td>
<td>12 (7)</td>
<td>49 (27)</td>
</tr>
<tr>
<td>coronal</td>
<td>80</td>
<td>80 (100)</td>
<td>24 (30)</td>
<td>43 (54)</td>
</tr>
<tr>
<td>lambdoid</td>
<td>9</td>
<td>9 (100)</td>
<td>7 (78)</td>
<td>2 (22)</td>
</tr>
<tr>
<td>metopic</td>
<td>71</td>
<td>71 (100)</td>
<td>6 (8)</td>
<td>14 (20)</td>
</tr>
<tr>
<td>multisuture</td>
<td>40</td>
<td>40 (100)</td>
<td>20 (50)</td>
<td>29 (73)</td>
</tr>
<tr>
<td>w/o lambdoid involvement</td>
<td>21</td>
<td>21 (100)</td>
<td>14 (67)</td>
<td>18 (86)</td>
</tr>
<tr>
<td>w/o lambdoid involvement</td>
<td>19</td>
<td>19 (100)</td>
<td>6 (32)</td>
<td>11 (58)</td>
</tr>
</tbody>
</table>

### Table 2: Incidence of CM according to suture involvement

<table>
<thead>
<tr>
<th>Craniosynostosis Type</th>
<th>Total No. of Patients</th>
<th>No. of Patients w/ CM (%)</th>
<th>p Value†</th>
</tr>
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<tr>
<td>single-sutures</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>sagittal</td>
<td>183</td>
<td>5 (2.8)</td>
<td>NS</td>
</tr>
<tr>
<td>coronal</td>
<td>80</td>
<td>5 (6.3)</td>
<td>NS</td>
</tr>
<tr>
<td>metopic</td>
<td>71</td>
<td>0 (0)</td>
<td>0.05‡</td>
</tr>
<tr>
<td>lambdoid</td>
<td>9</td>
<td>5 (55.6)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>total</td>
<td>343</td>
<td>14 (4.4)</td>
<td></td>
</tr>
<tr>
<td>multiple sutures</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>w/o lambdoid involvement</td>
<td>21</td>
<td>12 (57.1)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>w/o lambdoid involvement</td>
<td>19</td>
<td>2 (10.5)</td>
<td>&lt;0.01‡</td>
</tr>
<tr>
<td>total</td>
<td>40</td>
<td>14 (35)</td>
<td></td>
</tr>
</tbody>
</table>

*NS = not significant.
† Single-suture cases compared with other single-suture cases and multiple-suture cases compared with other multiple-suture cases.
‡ Negatively correlated with CM.
Chiari anomaly and craniosynostosis

repair, 17 were diagnosed with CM before undergoing surgical correction for craniosynostosis and 12 were diagnosed with CM following craniosynostosis repair.

Chiari Malformation Diagnosed Prior to Craniosynostosis Repair

Of the 17 patients diagnosed with CM prior to craniosynostosis repair, 10 underwent CM decompression. Five of these children were treated with simultaneous craniosynostosis repair and CM decompression. In each case, calvarial exposure of the posterior fossa was accomplished by reflecting the scalp posteriorly from a single bicoronal incision and performing a suboccipital craniectomy. In each case, the posterior arch of C-1 was never removed and the dura was never opened. In 3 patients, an initial craniosynostosis repair was followed by CM decompression, and in 2 CM decompression was performed prior to craniosynostosis repair. When CM decompression was carried out, either before or after craniosynostosis repair, the CM decompression was performed via a standard midline posterior scalp incision from the inion to the upper cervical spine, and a bony decompression was conducted. The dura mater was never opened. Seven patients diagnosed with CM prior to craniosynostosis repair did not undergo CM decompression at any time during the study period (Table 3). Of these, 6 had a decrease in tonsillar ectopia and 5 had improved CSF flow studies following craniosynostosis repair alone. The 2 patients in this group with syringes prior to craniosynostosis repair both had decreased syrinx width following craniosynostosis repair.

Chiari Malformation Diagnosed After Craniosynostosis Repair

Following craniosynostosis repair, 12 patients were diagnosed with CM. The mean age at time of CM diagnosis was 5.6 years (range 0–16 years) with a mean interval from the initial surgery of 3.7 years (range 197–2780 days). All of these patients underwent both cine MR imaging and spine MR imaging. At the time that CM was diagnosed, cine MR imaging revealed abnormal CSF flow in 10 (83%) of the 12 patients and spine MR imaging demonstrated a syrinx in 7 patients (58%). Five patients (42%) required more than 1 CVR procedure and 7 (58%) had increased ICP found after placement of an invasive ICP monitoring device. Six of these patients (50%) had tonsillar descent over 10 mm and CSF flow studies with restricted anterior and posterior flow at the foramen magnum. These patients ultimately underwent posterior fossa decompression at a mean interval of 3.4 years following the initial craniosynostosis repair. Of the 12 patients diagnosed with CM following craniosynostosis repair, 5 had at least 1 prior MR imaging study that demonstrated normal tonsillar height, thus indicating de novo CM development. The mean interval between the initial repair and CM diagnosis in these patients was 3.5 years. Four of the patients with documented
de novo CM development had a history of hydrocephalus and VP shunt placement in the interim between craniosynostosis repair and CM diagnosis.

Discussion

An association between CM and craniosynostosis has been recognized for many years.36 Chiari malformation is especially likely to be found in those with multisuture or syndromic craniosynostosis. Cinalli et al.3,4 examined 95 patients with syndromic craniosynostosis and found CM in 70% of those with Crouzon syndrome, 75% of those with oxycephaly, 50% of those with Pfeiffer syndrome, and 100% of those with the Kleeblattschädel deformity.26,43 Chiari malformation was found in only 1.9% of patients with Apert syndrome in that series.4 Importantly, that group used cerebellar tonsillar descent of less than 2 mm below the basion-opisthion line as the diagnostic criterion, perhaps leading to a larger number of diagnosed cases of CM. Other reports, however, have confirmed the frequent association of CM with craniofacial syndromes.7,9,33 Francis et al.9 found an associated CM in 5 of 10 patients with Crouzon syndrome. Fearon and Rhodes7 found that 84% of the 28 patients they treated for Pfeiffer syndrome had associated CM. Half of the children with Pfeiffer syndrome underwent placement of a VP shunt for hydrocephalus, and CM was diagnosed in every patient in whom a VP shunt was placed. Because CM has been found so frequently in children with Pfeiffer syndrome, Fearon and Rhodes have advocated routine screening of these children with MR imaging.
Chiari anomaly and craniosynostosis

Many cases of single-suture nonsyndromic craniosynostosis associated with CM have also been reported.2,17,25,41 Most of these cases involve the lambdoid suture, but CM has been reported even in conjunction with sagittal, metopic, and unilateral coronal synostosis.2,17,25,41 Leikola et al.17 reported on a series of 124 patients with single-suture craniosynostosis and, on imaging, found that 7 (5.6%) had CM. Sgouros et al.38 showed that cranial base growth is altered in both multi- and single-suture craniosynostosis, even in cases that do not exhibit premature fusion of a cranial base synchondrosis. Tubbs et al.41 found that 30% of those with metopic ridges had an associated CM, and they postulated that this may be the result of reduced anterior fossa volume. In our series, 15 (52%) of 29 patients with CM and craniosynostosis had single-suture disease, including syndromic and nonsyndromic cases. Five of these cases involved the lambdoid suture alone. We found CM in over half of all cases of isolated lambdoid synostosis. Chiari malformation was also found in isolated sagittal and coronal synostosis in 5 patients each when syndromic cases were included. Three patients with single-suture, nonsyndromic sagittal craniosynostosis and no patients with single-suture, nonsyndromic coronal craniosynostosis had associated CM. Because these sutures are involved much more often than the lambdoid suture, CM is significantly less associated with single-suture disease in these locations.

On the basis of the previously reported series and our own experience, we believe it is clear that syndromic and multisuture synostosis is frequently associated with CM. Furthermore, the frequent association of lambdoid synostosis with CM is well established.4 Lambdoid suture involvement is predictive of CM formation even in patients with multisuture or syndromic forms of craniosynostosis.4,43 Cinalli et al.4 have postulated that CM is much more likely in patients with Crouzon syndrome than those with Apert syndrome, because the former is associated with significantly earlier closure of the sagittal and lambdoid sutures.

The frequency of CM in various types of craniosynostosis can guide the practitioner in the decision to obtain an MR imaging study to screen for CM. Cinalli et al.5 found that approximately one-third of their patients who had CM associated with craniofacial disorders were either symptomatic or had a spinal cord syrinx. They advocated screening for CM in all patients with complex or syndromic craniosynostosis. We generally screen patients with syndromic craniosynostosis and patients with lambdoid synostosis with brain MR imaging prior to surgical correction of the craniosynostosis. We do not routinely screen asymptomatic individuals with single-suture craniosynostosis at locations other than the lambdoid suture.

Although some brain malformations associated with craniofacial disorders may be the primary result of a common genetic abnormality, most now believe that CM associated with craniosynostosis is acquired postnatally as a result of abnormal skull development.27 Chiari malformation has been associated with underdevelopment of the occipital bone and a small posterior fossa,21,22,24 as well as with primary bone disorders that affect the posterior fossa skull such as osteopetrosis and fibrous dysplasia.28 Craniofacial disorders that lead to a small or deformed posterior fossa may result in crowding of posterior fossa contents and tonsillar descent in affected patients.3 Most cases of CM associated with craniosynostosis are found after the prematurely fused suture has resulted in a significant skull deformity.4 This assertion is supported by several reported cases of de novo CM formation follow-

![Fig. 4. Imaging studies obtained in a 6-month-old girl presenting for evaluation of severe plagiocephaly.](image)

A: Reconstructed CT scan confirming synostosis of the right lambdoid suture. Inset: Two-dimensional depiction of the "viewing angle" that is depicted in the larger 3D image. B: Sagittal MR image demonstrating cerebellar tonsillar descent in a pegged configuration consistent with a diagnosis of CM. The patient underwent posterior cranial vault reshaping including barrel-stave osteotomies and removal of suboccipital bone at the posterior rim of the foramen magnum. The dura was not opened and posterior arch of the first cervical vertebra was not removed. C: Two-year postoperative MR image demonstrating no evidence of a CM.
ing diagnosis of a craniofacial syndrome. Hopkins and Haines\textsuperscript{14} reported on a case of Seckel syndrome in a patient in whom rapid CM development was demonstrated on serial imaging. Ranger et al.\textsuperscript{26} reported on a patient with Pfeiffer syndrome whose initial brain MR imaging findings were normal; 2 months after craniofacial reconstruction and VP shunt placement, however, MR imaging showed the development of CM, providing more evidence for an acquired pathogenesis. In patients with Crouzon syndrome, premature closure of the lambdoid suture is associated with an increased risk of CM.\textsuperscript{4,11,20} Apert syndrome, however, which typically presents with a normal or larger than normal posterior fossa, is not associated with CM.\textsuperscript{32}

Hydrocephalus is frequently associated with craniofacial anomalies and may occasionally play a role in CM pathogenesis in these patients.\textsuperscript{4,9,10,43} Children with craniofacial disorders as well as hydrocephalus are more likely to have a CM than children with a craniofacial disorder in the absence of hydrocephalus.\textsuperscript{4,9,34} Hydrocephalus has also been reported to develop after repair of craniosynostosis, with subsequent development of tonsillar herniation.\textsuperscript{9,43} In our own series, 4 of the 5 patients with de novo CM on serial imaging developed hydrocephalus after craniosynostosis repair and were treated with VP shunt placement prior to the CM diagnosis. Our own results, combined with those of prior reports, suggest that hydrocephalus plays a role in the pathogenesis of CM in some cases.\textsuperscript{4,9,10,34,43} We recommend routine screening for CM in patients with both craniosynostosis and hydrocephalus.

Craniofacial anomalies may be associated with venous outflow impairment that may lead to venous hypertension and increased ICP.\textsuperscript{1,9,12,13,28,31,33,39,40} Some surgeons suggest that cerebellar tonsillar herniation may result from increased venous turgor.\textsuperscript{9,28} Venous abnormalities are more often associated with complex and syndromic forms of craniosynostosis.\textsuperscript{33,39} Abnormal venous anatomy must be accounted for when considering surgical treatment of CM associated with complex craniofacial syndromes. For this reason, we screen all craniosynostosis and CM patients with preoperative CT venography to identify any transosseous venous channels. Furthermore, opening the dura for CM decompression in these cases may increase the risk of hemorrhage due to abnormal venous sinuses. In our experience, opening the dura has not been necessary for the treatment of CM in these patients. In some cases, preoperative detection of major venous collaterals at the site of a proposed CM decompression may suggest that even bone removal at the foramen magnum is not safe and should not be performed.\textsuperscript{1,37}

In general, most surgeons agree that CM should not be treated unless it is symptomatic or, in some cases, associated with a spinal syrinx. It is possible that the indications for surgical treatment of CM may be different for individuals with associated craniosynostosis. In some cases, CM decompression can be done simply at the time of a planned craniosynostosis repair. In this retrospective analysis, it is impossible to accurately describe all of the factors that went into each surgical decision. In general, we approach patients with both lambdoid synostosis and CM with an eye to expanding the posterior vault with over-correction. When a significant CM is seen on preoperative imaging, we often will include a posterior fossa decompression in the operation to repair the craniosynostosis. To the limits of this retrospective analysis, we do not believe that CSF flow was ever used as an important criterion for performing a posterior fossa decompression. Several groups recommend posterior fossa expansion surgery as the treatment of choice for all cases of CM identified prior to craniosynostosis correction, even in the absence of symptoms.\textsuperscript{5,38,44} It is possible that, in many instances, the CM will resolve or improve following craniosynostosis repair (Table 3). Di Rocco and Velardi\textsuperscript{6} reported on a single case in which a supratentorial cranial expansion resulted in resolution of an acquired CM. Given the very young age of patients undergoing craniosynostosis repair, it is important to consider that any bony decompression may be less durable compared with CM decompression performed later.\textsuperscript{5} For this reason, Fearon and Rhodes\textsuperscript{3} have advocated delaying posterior remodeling procedures in patients with Pfeiffer syndrome until at least 13 months of age. For patients with CM diagnosed before craniosynostosis repair, we reserve CM decompression for patients who exhibit symptoms of CM or a spinal cord syrinx af-

### Table 3: Effect of CVR without posterior fossa decompression on preoperatively diagnosed CM*  

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Sutures</th>
<th>Tonsillar Location†</th>
<th>Pre-CVR CSF Flow‡</th>
<th>Syrinx</th>
<th>Post-CVR Tonsillar Location†</th>
<th>CSF Flow‡</th>
<th>Syrinx</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>coronal</td>
<td>C-1</td>
<td>decreased ant &amp; pst</td>
<td>no</td>
<td>stable</td>
<td>improved</td>
<td>no</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>multiple</td>
<td>C-2</td>
<td>decreased pst</td>
<td>no</td>
<td>improved</td>
<td>improved</td>
<td>no</td>
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<td>3</td>
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<td>C-2</td>
<td>decreased ant</td>
<td>no</td>
<td>improved</td>
<td>improved</td>
<td>no</td>
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<tr>
<td>5</td>
<td>M</td>
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<td>C-2</td>
<td>decreased ant &amp; pst</td>
<td>yes</td>
<td>improved</td>
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<td>improved</td>
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<td>C-2</td>
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<td>improved</td>
<td>improved</td>
<td>no</td>
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</tbody>
</table>

* ant = anterior; pst = posterior.
† Tonsillar location is designated by vertebral level of tonsillar descent.
‡ Cerebrospinal fluid flow at the foramen magnum is described as decreased anterior, posterior, or demonstration of abnormal tonsillar motion.
Chiari malformation and craniosynostosis
ter an extended interval following surgical correction of the skull deformity. Exceptions may be made if the CM is symptomatic or associated with a spinal syrinx. The presence of posterior fossa transosseous venous collaterals, however, would lead us to reconsider any posterior fossa decompression.

In cases associated with hydrocephalus or intracranial hypertension, it may be necessary to treat the hydrocephalus or relieve the ICP to treat the CM. Pouratian et al. have reported on 2 cases of acquired CM associated with craniosynostosis and hydrocephalus that were initially treated with CSF diversion. Others have also reported on CM development after treatment of hydrocephalus. The role of endoscopic third ventriculostomy is not clear. Fearon et al. reported on a small series of patients with craniofacial dysostosis, CM, and hydrocephalus; patients who did not require surgical management of the CM underwent endoscopic third ventriculostomy, in contrast to those treated with a VP shunt for their hydrocephalus. In most cases, we prefer to treat the hydrocephalus with a shunt procedure prior to treating CM.

There are several limitations to our study. In this retrospective analysis, the treating physicians may have recommended a different treatment for patients depending on the perceived severity of the craniosynostosis or the CM. Patients with symptoms, a spinal syrinx, or a greater degree of tonsillar cerebellar descent were more likely to be surgically treated. Therefore, any attempt to compare outcomes of patients by treatment pattern will reflect this selection bias. Furthermore, MR imaging was not performed in all patients presenting with craniosynostosis. Our selective use of MR imaging in patients with craniosynostosis has resulted in a sampling bias that must be considered in any analysis of these results. Sampling bias also needs to be considered when pondering the role of hydrocephalus in these patients, because individuals with hydrocephalus were more likely to undergo repeated imaging. Although we reported on a large number of patients with craniosynostosis, CM presentations and treatments were diverse in this group of patients. This diversity resulted in relatively small sample sizes for the analysis of individual treatment strategies. Finally, none of the patients in this series were treated using minimally invasive strip craniectomy techniques that are increasingly used for treatment of craniosynostosis. It is possible that these techniques will result in different rates of occurrence and different treatment outcomes for CM.

Conclusions

Chiari malformation is seen frequently in patients with syndromic, multisuture and single-suture synostosis of the lambdoid suture. In some cases, the CM and craniosynostosis present concurrently. In other patients, we have noted the development of CM after the surgical treatment of the craniosynostosis. In patients presenting with both CM and craniosynostosis, we recommend treating the craniosynostosis first, with simultaneous posterior fossa expansion only if there are neurological symptoms or spinal syringes. When CM decompression is not carried out at the time of craniosynostosis repair, it should be considered if neurological symptoms or a spinal syrinx persist after the initial surgery. Neurosurgical follow-up is particularly necessary for patients with multisuture, syndromic, or lambdoid craniosynostosis, as well as for those patients requiring treatment of hydrocephalus.

Disclosure

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

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References


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