Pediatric Chiari malformation Type 0: a 12-year institutional experience

Clinical article

Joshua J. Chern, M.D., Ph.D., Amber J. Gordon, M.D., Martin M. Mortazavi, M.D., R. Shane Tubbs, M.S., P.A.-C., Ph.D., and W. Jerry Oakes, M.D.

Pediatric Neurosurgery, Children's Hospital, Birmingham, Alabama

Object. In 1998 the authors identified 5 patients with syringomyelia and no evidence of Chiari malformation Type I (CM-I). Magnetic resonance imaging of the entire neuraxis ruled out other causes of a syrinx. Ultimately, abnormal CSF flow at the foramen magnum was the suspected cause. The label “Chiari 0” was used to categorize these unique cases with no tonsillar ectopia. All of the patients underwent posterior fossa decompression and duraplasty identical to the technique used to treat patients with CM-I. Significant syrinx and symptom resolution occurred in these patients. Herein, the authors report on a follow-up study of patients with CM-0 who were derived from over 400 operative cases of pediatric CM-I decompression.

Methods. The authors present their 12-year experience with this group of patients.

Results. Fifteen patients (3.7%) were identified. At surgery, many were found to have physical barriers to CSF flow near the foramen magnum. In most of them, the syringomyelia was greatly diminished postoperatively.

Conclusions. The authors stress that this subgroup represents a very small cohort among patients with Chiari malformations. They emphasize that careful patient selection is critical when diagnosing CM-0. Without an obvious CM-I, other etiologies of a spinal syrinx must be conclusively ruled out. Only then can one reasonably expect to ameliorate the clinical course of these patients via posterior fossa decompression. (DOI: 10.3171/2011.4.PEDS10528)

Key Words • posterior fossa • hindbrain hernia • tonsillar ectopia • neurosurgery • pediatrics

Idiopathic cervical syringomyelia can be associated with supratentorial lesions, such as hydrocephalus, craniostenosis, and meningitis. Posterior fossa lesions associated with syringomyelia include tumor, posterior fossa cysts, and various Chiari malformations. One possible mechanism of syrinx formation due to posterior fossa pathology is an alteration of CSF flow at the cranio cervical junction. This notion has been supported by CSF flow studies as well as clinical responses after attempts to alter CSF dynamics via foramen magnum decompression. Possible underlying causes of disrupted CSF flow in the absence of cerebellar tonsillar herniation include a compacted posterior fossa as well as intraoperative findings of veins and arachnoid adhesions at the foramen of Magendie. Because of this hypothesized pathophysiological mechanism, the term “Chiari 0 malformation” was coined, referring to syringomyelia that resolves following posterior fossa decompression and in the absence of tonsillar ectopia.

Herein, we present the salient features of our surgical experience with the CM-0 in children, derived from over a decade of experience with this unique pathological entity.

Methods

Following institutional review board approval, a retrospective analysis of all CM-Is in pediatric patients surgically treated at the Children’s Hospital in Birmingham, Alabama, was performed. Four hundred five patients were surgically treated between 1998 and 2010. In this group, 15 patients fulfilled the criteria for CM-0. All patients had undergone imaging of the head, cranio cervical junction, and spine. Imaging of the head was performed to exclude intracranial pathology, including hydrocephalus. High-resolution MR imaging studies of the posterior fossa were not routinely obtained. Spine MR imaging with contrast enhancement was conducted to examine the extent of syringomyelia, to rule out potential intrins ic spinal cord neoplasm, and to exclude a tethered spinal cord. Flexion and extension radiographs were obtained to

Abbreviations used in this paper: CM-I = Chiari malformation Type I; PICA = posterior inferior cerebellar artery.
rule out craniocervical junction instability. Cerebrospinal fluid flow studies were performed early in this series but not over the last approximate decade because of our experience with false-negative results.

Each patient underwent posterior cranial fossa decompression along with the removal of the posterior arch of C-1. The size of the craniectomy was 2.5 cm in width and in height. Intraoperative ultrasonography was not used in these 15 patients. Intradural exploration of the spinomedullary junction revealed potential arachnoid adhesions that might cover the foramen of Magendie. Occasionally, metal clips were used to attach the arachnoid membrane to the dura mater in an attempt to decrease the likelihood of a posterior fossa CSF hygroma causing acute hydrocephalus. All patients underwent duraplasty with autologous pericranium. Each patient was monitored in the intensive care unit overnight and in general was sent to the ward over the following couple of days.

Results

Patient Presentation

The patient cohort consisted of 9 boys and 6 girls with no history of trauma or infection of the spine (Table 1). Patient ages ranged from 3 to 15 years (mean 10.5 years). The most common presenting symptom was spine-related, including scoliosis (8 patients), limited neck range of motion (1 patient), and torticollis (1 patient). Six patients reported headache and/or neck pain. Four patients reported headaches that were typical of cerebellar ectopia, that is, those that were Valsalva induced, of short duration, and posterior in location. One patient had atypical headache. On examination 1 patient had gait disturbance and was found to have increased tone in the lower extremities. The duration of symptoms varied between 1 month and 3 years. In 1 case, the onset of symptoms occurred after a traumatic event. One patient had a cerebellar hemorrhage at birth, and the dysmorphic posterior fossa contents were suggestive of extensive scar formation. No other inciting events or etiologies were identified in the other cases.

The minimal follow-up in these patients was 1 year. Postoperatively, 10 of the 15 patients experienced significant to complete symptom resolution. One patient had a preoperative scoliosis curve of 80° that continued to progress and eventually required spinal fusion. One patient who had presented with non–Chiari-like headaches continued to experience headaches postoperatively. Three patients who had presented with mild scoliosis (< 35°) demonstrated stable curves after surgery.

Syringomyelia Signs

All 15 patients presented with syringes of various lengths (3–18 levels). All but 1 patient showed marked diminishment in the size of the syrinx. The percent decrease in the syrinx cross-sectional area ranged between 60% and 95%. This decrease was most dramatic in 2 patients who had presented with holocord syringes (Figs. 1–4). In the only exception, the patient had a syrinx extending from the cervical region to the conus with dilation at the conus level. The terminal portion of the syrinx measured 4 mm in diameter and remained unchanged at the 5-year follow-up. In general, the change in syrinx size was first observed between 6 months and 1 year after surgery; however, no routine imaging was typically done prior to 6 months. None of the residual syringes were found to progress with follow-up.

Intraoperative Findings

The posterior arch of C-1 was bifid in 1 patient, and the assimilation of C-1 occurred in 1 patient. One patient had a markedly thickened bone and an upward lipping of the opisthion. Eight patients demonstrated an arachnoid veil occluding the fourth ventricular outlet, which was transected in all patients. In 1 of these 8 patients,
Chiari malformation Type 0

the arachnoid membrane adhesions were extremely dense and necessitated shrinking the cerebellar tonsils with electrical cautery. Interestingly, in 1 case, the PICA formed a loop immediately adjacent to the obex, and it was thought that the redundant PICA loop might have caused intermittent obstruction of the CSF flow at the foramen of Magendie.

Complications of Surgery

No neurological complications occurred as a result of operative intervention in this small cohort. There was neither CSF leakage nor surgical infection. The average hospital stay was 3 days. Postoperative pain was controlled in the majority of patients by using alternating doses of acetaminophen and ibuprofen. No patient required reoperation or additional surgical procedures such as syringo-subarachnoid shunting.

Anatomy of the Posterior Cranial Fossa

We previously suggested that the syringohydromyelia seen in the CM-0 might be due to a compressed posterior fossa and that tonsillar herniation would not be necessary to disrupt normal CSF flow. The most striking finding in the initial group of 5 patients was a caudally displaced brainstem without tonsillar ectopia. We performed similar measurements in the now larger (inclusive) group of 15 patients, and our findings were consistent with those observed in our earlier, smaller study.

At the level of the foramen magnum, the sagittal anteroposterior distance of the spinomedullary junction was 13 mm versus an average of 11 mm, implying caudal descent of the more rotund medulla oblongata. Correspondingly, the tip of the obex was located at or below the level of the foramen magnum in all patients except 2, whereas in a healthy control group, the obex was located 8–17 mm above the level of the foramen magnum. Lastly, at the midsagittal plane, there was an increase in the distance between the basion and the opisthion (average 37.4 mm, compared with 28–33 mm in age-adjusted controls). These observations were consistent with the hypothesis that the contents of the posterior fossa in this group of patients were caudally compressed.

Discussion

While we find it encouraging that with our selection criteria this small group of patients suffering from idiopathic syringohydromyelia seemed to respond well to posterior fossa decompression, we stress once again that CM-0 is a diagnosis made only after other etiologies of spinal syrinx have been conclusively ruled out. As our case number demonstrated, this subgroup is small; CM-0 was found in only 3.7% of our cases. Shunting or fenestrating a symptomatic idiopathic syringohydromyelia continues to be a valid treatment option for this disease entity; in our experience, however, the results have often been disappointing. Therefore, dealing with the primary cause of the syrinx—that is, interruption of CSF flow at
the craniocervical junction—is the logical first step in treatment, just as in patients with CM-I.

**Clinical Characteristics of Patients With CM-0**

We previously described 5 patients with CM-0, and 10 patients were added to this group in the present study. We did not find substantial differences between these 2 patient groups. The predominant symptoms and signs were related to scoliosis and lower-extremity weakness and paresthesias. Among the 8 patients with scoliosis and kyphoscoliosis as the major sign, the curves improved in 4 (completely resolving in 1 case), remained stable in 3, and worsened in only 1. The patient in this last case had a preoperative curve of 80° and was the only person who required spinal fusion following posterior fossa decompression. Complete and partial resolution of lower-extremity symptoms occurred in 3 patients and 1 patient, respectively.

Headache is often a secondary symptom, although more than half of the patients denied having headaches. When a patient does present with headache, it is often Chiari-like (short duration, reproducible with the Valsalva maneuver, and located in the occipital region). For such headaches to occur, the cerebellar tonsils might be intermittently herniating, and this phenomenon is not captured during static MR imaging. This notion may be supported by the fact that all 3 patients who presented with Chiari-like headaches experienced complete resolution of their symptoms postoperatively.

**Lessons Learned From the Past Decade**

As with all surgical procedures, careful patient selection is critical. All of the patients underwent detailed work-ups as described above. The presence of significant syringohydromyelia along with correspondingly credible signs and symptoms largely dictated the decision for surgery. We placed relatively minor importance on nondescript symptoms such as headache; over the years, however, we did expand the indications for posterior fossa decompression in patients, and we were pleased to find satisfactory results among them.

Obtained anthropomorphic measurements suggested that the etiology of CM-0 was CSF flow disturbance, and this hypothesis was further supported by intraoperative observations. While these morphological measurements, which have been confirmed by others, may help to establish a diagnosis, in reality, our decision to operate relies very little on these measurements. It is perhaps even more remarkable then that these patients do share the noted

---

**Fig. 3.** Case 5. Preoperative sagittal MR image demonstrating a CM-0.

**Fig. 4.** Case 5. Preoperative axial MR image showing syrinx resolution, although the resolution was not as dramatic as that seen in the patients featured in Figs. 1 and 2.
Chiari malformation Type 0

anatomical similarities and further support CM-0 as an independent disease entity. Again, the presence of symptomatic syringohydromyelia per se would have prompted our decision to operate with or without the finding of a crowded posterior fossa. Interestingly, there may be some overlap between CM-0 and CM-1.5, as both have shown caudal descent of the brainstem.

Finally, cine MR imaging would seem to be a valuable tool to document the CSF flow pattern before and after surgery in these cases. In our early experience, however, this particular modality was often inconclusive despite clinical and radiologically confirmed improvement. Therefore, the use of cine MR imaging has been abandoned over the last several years.

Conclusions

We stress that this patient subgroup represents a very small cohort within the spectrum of those with Chiari malformations. These patients tend to have caudally displaced brainstems and at surgery are often found to have arachnoid veils occluding the foramen of Magendie. We emphasize that careful patient selection is critical when making the diagnosis of CM-0. Without an obvious CM-I, other etiologies of a spinal syrinx must be conclusively ruled out. Only then can one reasonably expect to ameliorate the clinical course of these patients with posterior fossa decompression.

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: Tubbs. Acquisition of data: Tubbs, Chern, Gordon. Analysis and interpretation of data: Tubbs, Gordon. Drafting the article: Tubbs, Chern. Critically revising the article: all authors. Study supervision: Tubbs, Oakes.

References


Manuscript submitted November 19, 2010. Accepted April 25, 2011.

Address correspondence to: R. Shane Tubbs, Ph.D., Pediatric Neurosurgery, Children’s Hospital, 1600 7th Avenue South, ACC 400, Birmingham, Alabama 35233; email: shane.tubbs@chsys.org.