Chiarí malformation Type I is a congenital disorder primarily diagnosed on the basis of caudal displacement of the cerebellar tonsils through the foramen magnum into the upper cervical canal.5,18,34,36,40,53 The exact pathogenesis and natural history of this malformation remain undefined.40 Presentations may include tussive headaches, cerebellar or brainstem dysfunction, or symptoms referable to syringomyelia. Although debate persists regarding the degree of tonsillar ectopia necessary to warrant the diagnosis of CM-I, many clinicians define CM-I as at least 5 mm of tonsillar descent below the foramen magnum.1,4,34,53 Chiarí malformation Type I is often discovered incidentally in asymptomatic patients, and may not progress or warrant intervention.30,57 The authors of numerous reports have demonstrated, however, that acute neurological deterioration can occur in patients with CM-I and acquired Chiarí malformations.5,3,10,11,13,14,29,31,33,43,46,49,56,59,65 Milhorat et al.35 also reported on a case of acute myelopathy secondary to VP shunt malfunction and the onset of acute syringomyelia.

We present our experience in 2 patients with asymptomatic CM-I who experienced quite different manifestations of acute neurological deterioration secondary to VP shunt malfunction. Presumably, VP shunt malfunction created a positive rostral pressure gradient across a stenotic foramen magnum, resulting in tetraparesis from foramen magnum syndrome in 1 patient and acute ataxia and cranial nerve deficits from syringobulbia in the other. Although urgent shunt revisions yielded partial recovery of neurological function in both patients, marked improvement occurred only after posterior fossa decompression. (DOI: 10.3171/2009.4.PEDS0936)

Key Words • Chiari malformation • foramen magnum syndrome • syringobulbia

Abbreviations used in this paper: CM-I = Chiari malformation Type I; CN = cranial nerve; SSEP = somatosensory evoked potential; VP = ventriculoperitoneal.
Acute deterioration in CM-I

Case Reports

Case 1

History and Examination. This 16-year-old boy with cerebral palsy, developmental delays, moderate mental retardation, and shunted hydrocephalus since birth presented to an outside hospital with bifrontal headaches, nausea, and double vision. A radioisotope shunt patency study (shuntogram) was performed and showed evidence of flow from the ventricle into the peritoneal cavity. Over the course of 4 days, mild, progressive weakness developed in all extremities, followed by worsening of headaches and urinary retention requiring placement of a Foley catheter. On arrival at our institution, the child was lethargic, hypertensive (200/100 mm Hg), bradycardic, had bilateral CN VI palsy, and was severely tetraparetic. His motor power was 3/5 (antigravity) in the right upper extremity, and 1/5 (trace movement) in the left upper, left lower, and right lower extremities.

First Operation. Given the signs of acute herniation, an emergency large-volume shunt tap was performed with immediate improvement in mental status and vital signs. The patient’s motor, sphincter, and CN deficits remained stable. He was taken directly to the operating room for a distal VP shunt revision. The patient’s motor examination gradually improved to 4/5 in the right upper extremity, 3/5 in the left upper extremity, and 2/5 in the bilateral lower extremities over the course of few days. He was unable to void on his own postoperatively and a Foley catheter was reinserted.

Magnetic resonance images of the brain and cervical spine were obtained that again demonstrated the previously diagnosed Chiari malformation with low lying tonsils down to the lamina of C-1. Diffusion-positive T2 signal changes were revealed in the lower medulla/dorsal columns of the upper cervical cord consistent with acute infarction, as well as edema of the right cerebellar tonsil (Fig. 1A and B). Although prior imaging studies were not available for viewing, the degree of tonsillar descent (12 mm) was more pronounced compared to the report on the previous MR images that noted 6 mm of tonsillar descent below the foramen magnum. These findings were consistent with suspected herniation and acute foramen magnum syndrome, and probably exacerbated by the child’s congenitally narrow cervical spinal canal.

Second Operation. After the child’s moderate improvement following the shunt revision, the surgical team recommended Chiari decompression to possibly hasten the patient’s recovery, relieve residual compression, and perhaps prevent future deterioration. Given his immediate partial improvement, his family elected to wait for further recovery before another surgery. Magnetic resonance imaging was repeated 2 weeks postoperatively, and demonstrated encephalomalacia in the medulla, atrophy of right cerebellar tonsil, and a slight decrease in the extent of tonsillar descent (Fig. 1C and D). Given his residual compression and the plateau in his neurological recovery, the patient underwent suboccipital craniectomy, C-1 laminectomy, and duraplasty. Intraoperative SSEPs were unreliable but intermittently recordable at the start of the operation. Subsequent to decompression and dural opening, the reproducibility of the SSEP signals dramatically improved.

Postoperative Course. One week after decompression, the patient could ambulate with assistance and void without difficulty. At the 6-week follow-up examination, the patient’s strength and gait had markedly improved. His most prominent deficit was difficulty with joint position sense while ambulating, probably secondary to the infarction in the dorsal columns. He continues to improve with intensive physical therapy.

Case 2

History and Examination. This 14-year-old boy originally presented as an infant with communicating hydrocephalus, and underwent VP shunt insertion at 4 months of age, with a single revision surgery 3 months later. After the initial decrease in size of his ventricular system, serial imaging studies revealed stable ventriculomegaly and CM-I. Of note, no syrinx was present on serial examinations obtained over the course of 8 years, the most recent of which was 6 years prior to the current presentation. He remained well until the day he was admitted after an episode of prolonged vomiting in the setting of a coincident viral illness, and medial deviation of his left eye, weak-
ness of the left side of his face, and hoarseness were noted. On presentation to the emergency room, neurological examination confirmed left CN VI palsy, left peripheral facial paresis (House-Brackman Grade IV), ataxia, and paralysis of the left vocal cord on direct laryngoscopy. Although MR imaging revealed stable ventriculomega-
ly, there was exacerbated descent of his tonsils (8 mm) compared to findings on a prior imaging report that noted only 5 mm of tonsillar descent, cervicothoracic syringomyelia, and contiguous syringobulbia extending into the left posterolateral medulla and pons (Fig. 2A–C).

First Operation. The results of a radioisotope shunt patency study suggested a partial distal shunt malfunction. Intraoperative exploration of the shunt revealed decreased flow across the valve, so the valve was replaced. Postoperatively the patient’s CN VI and VII palsies improved slightly, but the ataxia persisted without improvement.

Second Operation. Given his incomplete recovery, the patient underwent suboccipital craniectomy, C-1 laminectomy, microsurgical fenestration of adhesions, and duraplasty on postoperative Day 6. Notably, dense adhesions were discovered overlying the cerebellar tonsils and foramen of Magendie, possibly the cause of his communicating hydrocephalus.

Postoperative Course. The patient subsequently made a remarkable recovery with near complete resolution of the CN VI and VII palsies (House-Brackman Grade II), swallowing difficulty, hoarseness, and ataxia. Repeated brain and cervical spine MR images obtained 1 week postoperatively demonstrated reestablishment of CSF spaces at the foramen magnum, complete resolution of syringobulbia, and a marked decrease in the size of his cervicothoracic syrinx (Fig. 2D–F).

Discussion

Although CM-I is a congenital malformation, most patients typically experience an insidious disease course and present in adulthood. In a review of > 22,000 patients who underwent MR imaging of the head and cervical spine, Meadows et al. found asymptomatic CM-I to be present in 0.11% of these patients with a mean tonsillar descent 11 mm below the foramen magnum. In a series by Vernooij and colleagues documenting incidental findings on brain MR imaging in 2000 adults, CM-I was identified in 0.9% of patients. Debate exists concerning the degree of tonsillar ectopia required to warrant the diagnosis of CM-I, and controversy remains over indications for surgical intervention. Novegno et al. recently followed a cohort of 22 children with incidentally discovered or minimally symptomatic CM-I and found that 77% remained asymptomatic or improved over a mean follow-up of 5.9 years. Given the imprecise correlation of imaging and clinical findings, and the undefined natural history, establishing precise criteria for intervention—especially in asymptomatic patients—has been fraught with difficulty.

Despite the usually insidious course of CM-I, acute deterioration has been described, and acquired forms of Chiari malformation exist. There have been several reports describing patients with undiagnosed CM-I who
Acute deterioration in CM-I

experienced sudden-onset neurological deficits from syringomyelia.1,2,5,6 Numerous authors have reported acquired tonsillar herniation from spontaneous spinal fluid leaks, lumbar punctures, external lumbar drainage, lumboperitoneal shunts, and missile injuries to the spine.3,10,11,14,20,41,46–49,51,59 All of these causes create a negative pressure gradient caudal to the foramen magnum that can promote tonsillar descent and brainstem compression. Such acquired forms have been observed in patients with occult CM-I, as well as in those without prior posterior fossa abnormalities or tonsillar ectopia. Treatment in these patients involved a combination of blood patching, clamping or removal of lumbar drains or shunts, VP shunt placement, or posterior fossa decompression. Other authors have reported tonsillar descent and neurological deterioration in patients with asymptomatic CM-I after traumatic brain injury and minor trauma.5,13,33,46,58,61 Lee et al.31 reported on a patient with acquired CM-I and an associated syrinx secondary to obstructive hydrocephalus caused by a giant craniopharyngioma. The CM-I resolved with resection of the tumor.

The phenomenon of hydrocephalus or shunt malfunction causing brainstem dysfunction is well-documented in patients with CM-II (Arnold-Chiari malformation) who often have more severe compression of the brainstem and cervicomedullary junction than those with CM-I.8,9,12,27,30,37,39,42,44,45,52,54,56 The authors of numerous reports have described shunt insertion or revision in patients with CM-II leading to resolution of apnea,12,29,52 brainstem and lower CN dysfunction,8,9,39 and improvement in syringomyelia size and its associated deficits.30,37 According to a review of CM-II management by Tubbs et al.,44 assurance of a functioning shunt is mandatory prior to consideration of surgical decompression to avoid the risk of herniation. These authors also assert that progressive syringomyelia in patients with CM-II with shunts should be considered a malfunction until proven otherwise. Similar to CM-I, however, debate also exists concerning the precise indications for posterior fossa decompression in children and infants with CM-II.42,44,45,55 Nevertheless, there is some evidence of progressive neurological deterioration after the onset of brainstem dysfunction,42,44,45,55 and that better outcomes can be achieved with early intervention.25,26,42,44,45,56 The mechanism of deterioration in these examples of patients with CM-I and CM-II appear to be similar—a positive rostral pressure gradient leading to tonsillar descent across a stenotic foramen magnum.

We first described a patient with a preexisting but asymptomatic CM-I, in whom progressive tetraparesis developed, followed by acute tonsillar herniation and brainstem compression caused by VP shunt malfunction. The patient’s neurological deterioration was secondary to a positive rostral pressure gradient across a stenotic foramen magnum at baseline. Although he did improve somewhat after emergency shunt revision, many of his deficits persisted until posterior fossa decompression and duraplasty were performed. Although there was evidence of a small, bilateral medullary infarction indicative of some element of permanence to his deficits, there was exacerbation of his tonsillar descent from baseline, and crowding of the posterior fossa after shunt revision. The significance of the marked improvement in the reproducibility of SSEP signals after decompression is unknown, but may provide further evidence of ongoing compression.38 Nevertheless, the patient had a more dramatic recovery after decompression—indicative of a reversible element to his deficits.

Although debate persists regarding the exact pathogenesis of syringomyelia and syringobulbia, their association with CM-I and stenosis at the foramen magnum is unquestionable.20,22–24,32,41,50,55,60 The incidence of syringomyelia in CM-I varies between 50 and 80%;2,21,36,55 syringobulbia, however, is much less common (3–5.8%).22,36,50,55 While the majority of patients with CM-I and syringomyelia exhibit subacute or chronic symptoms, a few cases of acute neurological deterioration from both syringomyelia and syringobulbia have been described.2,5,6,12,15,62 Milhorat et al.35 reported on a case of acute progressive paraparesis and urinary retention in a patient who had previously undergone treatment for CM-I. The cause of clinical deterioration was VP shunt malfunction and the acute development of a holocord syrinx that had not been present on imaging studies obtained 3 days prior to presentation. This patient’s condition completely resolved after VP shunt revision.

In the second case we presented, given the absence of syringomyelia on serial MR images over an 8-year timespan and the lack of long-tract signs or symptoms, the presence of a new syrinx hints toward an acute or subacute onset of syringomyelia. Given the 6-year interval between the last imaging study that had demonstrated the absence of a syrinx, however, we cannot determine whether the syrinx had been present and progressing during that time, or whether the partial VP shunt malfunction resulted in de novo syrinx formation caused by an increased intracranial pressure gradient. The patient’s acute-onset brainstem symptoms were probably secondary to the acute development of syringobulbia from increased intraspinal pressure caused by vomiting and Valsalva maneuvers that forced the syrinx to dissect in a rostral direction into the brainstem. This theory is consistent with the patient’s acute neurological deterioration and the subtle finding of intraparenchymal signal changes on T2-weighted and FLAIR images around the cavity of the bulbar syrinx, a finding absent around the syrinx at the levels of the cervicothoracic spinal cord and caudal medulla (Fig. 2C). This may be similar to the transseptal flow observed around the lateral ventricles in patients with acute hydrocephalus.

The cases we have presented here underscore the need for careful evaluation in patients with Chiari malformation and symptoms of shunt malfunction. One must have a high index of suspicion for a potential shunt malfunction, and proceed with aggressive evaluation and timely treatment. In both patients we have reported on in the present study, persistent compression was suspected given their limited recovery after VP shunt revision, and the results of imaging; taken together, these findings underscored the need for surgical correction of the CM-I, even though correction of the inciting event (VP shunt malfunction) had already taken place.

Conclusions

We reported on 2 quite different and dramatic cases
of acute neurological deterioration caused by shunt malfunction in patients with previously asymptomatic CM-I. Given the devastating neurological sequelae of compression at the foramen magnum, surgeons must have a low threshold for diagnostic and therapeutic interventions in the setting of suspected shunt malfunction and Chiari malformation. In selected cases, suboccipital decompression may be instrumental to recovery, and provide prophylaxis against future insults.

Disclaimer

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

References

37. Milhorat TH, Johnson WD, Miller JI, Bergland RM, Hollen-
Acute deterioration in CM-I


Address correspondence to: Robert E. Elliott, M.D., Department of Pediatric Neurosurgery, New York University, Langone Medical Center, 317 East 34th Street, Suite 1002, New York, New York 10016. email: robert.elliott@nyumc.org.