See the corresponding article in this issue, pp 729–742.

J Neurosurg Spine 13:727–728, 2010

Editorial

Chiari malformation Type I

Mark M. Souweidane, M.D.

Neurological Surgery and Pediatrics, Weill-Cornell Medical College, New York, New York

The treatment of symptomatic patients with Chiari malformation Type I (CM-I) has incited an exhaustive range of opinions regarding an optimal treatment strategy. What is universally agreed upon is the need for adequate osseous decompression. Beyond that technical component of surgery, great variance and occasional vehemence is expressed in opinions pertaining to optimal treatment. Amidst this backdrop is the expectation that with treatment, approximately 90% of patients will enjoy symptomatic and radiographically demonstrated improvement with negligible morbidity.[3,6] In the rare event of incomplete resolution, the neurosurgeon has a variety of treatment alternatives at his or her disposal that are dependent on the primary procedure and the postoperative radiographic images. These maneuvers include repeat osseous decompression, tonsillar resection, detethering, fourth ventricular fenestration, ventricular CSF diversion, syrinx shunting, and pseudomeningocele repair. Selecting the best approach can be vexing.

Dr. Heiss and colleagues[2] have offered an elegant and detailed assessment of patients requiring a secondary procedure for symptomatic CM-I. Subarachnoid space dimensions, MR imaging–based CSF velocity measurements, morphological assessments, subarachnoid pressure recordings, and compliance calculations were compared before and after secondary surgical treatment. Additionally, and somewhat surprisingly, these measurements were compared with those in healthy volunteers. In their paper the authors confirm what has long been suspected by most neurosurgeons: inadequate osseous decompression is the preeminent cause of failure of primary surgery. Their excellent clinical results refute any doubt regarding the benefit of reexploration and highlight the relative merits of their approach as compared with syrinx shunting. This information is welcomed and certainly provides confidence to the surgeon faced with the "failed Chiari patient." Their study offers important insights pertaining to not only the secondary procedure but also the primary procedure—points that deserve further commentary.

Secondary Decompression

What remains nebulous in the interpretation of the work by Dr. Heiss and coauthors[2] is what technical aspect of the secondary procedure was most useful. While it is true that each patient underwent further osseous decompression, many also had simultaneous procedures aimed at reestablishing a patent subarachnoid space (that is, release of adhesions, detethering, pseudomeningocele repair, and so forth). If osseous decompression was the sole factor responsible for improvement, could one infer from the results that an extradural procedure alone would adequately address the problem? This conclusion is unlikely and the authors justifiably used a comprehensive surgical approach with admirable results. Thus, since one cannot conclude with confidence that additional bony removal was the sole source of success, it would be prudent to mimic the authors' approach incorporating intradural exploration and duraplasty given their excellent results.

Primary Decompression

The current study also provides some valuable observations that pertain to the adequacy of primary procedures for patients with CM-I. Because the study included patients who had relapsed as well as those having persistent symptoms, a case could be made that the initial surgery was inadequate in only a subset of patients. Osseous regrowth, pseudomeningocele formation, and intradural adhesions have all been implicated as possible causes for symptom relapse following a seemingly successful primary procedure. However, in this series the duration between primary and secondary procedures was relatively short, exceeding 5 years in only 1 patient. Further, the youngest patient was 12 years of age, making osseous regrowth an unlikely explanation for failure. These features of a relatively short interval and a patient age beyond skeletal maturity both favor the impression that the primary procedure might have been enhanced. This conclusion raises a vital question of what maneuvers may have been altered at the time of the primary procedure to reduce the potential of delayed failure. Currently at the forefront of this topic is the importance of dural opening. It is interesting to note that 3 of the patients in the current report had dural sparing procedures as initial treatment. Undeniably, the potential for a secondary procedure increased when the dura mater was not opened as part of the initial surgery.[1,4] This "less invasive" modification has its genesis in the intent to maintain a clear subarachnoid space and reduce the potential for CSF-related complications (that is, hydrocephalus, CSF fistulae, and bacterial meningitides). However, it has been shown that the actual risk of performing duraplasty is negligible in experienced hands, and treatment options must be carefully chosen to maximize patient outcome.[3,5,6] Another important point that is underscored in the current paper is the value of a thorough osseous decompression. The lateral limits of the foramen magnum and spinal canal should always serve as the anatomical reference in a lateral

dimension during decompression. Any abbreviation of this wide osseous decompression risks a poor patient outcome. The authors relied on autologous duraplasty and wide bone removal for impressive outcomes. In all likelihood, if both of these maneuvers were done at the time of initial surgery, there may have been fewer patients requiring a second procedure.

In all probability, the surgical variability in treating patients with CM-I is at times born out of need. Given patient variability (that is, age, associated conditions, anatomy, syrinx, and so forth), it is likely that there exists no single unifying procedure for every symptomatic patient with a CM-I. It is anticipated that with forthcoming comparative clinical studies that rely on objective measurements and standardized outcome scales, using methodologies similar to some of those utilized in the current study, treatment could be tailored in the hopes of reducing the need for secondary procedures.

References

1. Durham SR, Fjeld-Olenec K: Comparison of posterior fossa decompression with and without duraplasty for the surgical treatment of Chiari malformation Type I in pediatric patients: a meta-analysis. **J Neurosurg Pediatr 2:**42–49, 2008
2. Heiss JD, Suffredini G, Smith R, DeVroom HL, Patronas NJ, Butman JA, et al: Pathophysiology of persistent syringomyelia after decompressive craniocervical surgery. Clinical article. **J Neurosurg Spine 13:**729–742, 2010
3. Hoffman CE, Souweidane MM: Cerebrospinal fluid-related complications with autologous duraplasty and arachnoid sparing in type I Chiari malformation. **Neurosurgery 62:**156–161, 2008
4. Mutchnick IS, Janjua RM, Moeller K, Moriarty TM: Decompression of Chiari malformation with and without duraplasty: morbidity versus recurrence. **J Neurosurg Pediatr 5:**474–478, 2010
5. Oakes WJ: Chiari malformation Type I. **J Neurosurg 106:** 185–186, 2007
6. Tubbs RS, McGirt MJ, Oakes WJ: Surgical experience in 130 pediatric patients with Chiari I malformations. **J Neurosurg 99:**291–296, 2003

Response

John D. Heiss, M.D.,[1] and Edward H. Oldfield, M.D.[2]

[1]Surgical Neurology Branch, National Institute of Neurological Disorders and Stroke, National Institutes of Health, Bethesda, Maryland; and [2]Department of Neurosurgery, University of Virginia Health System, Charlottesville, Virginia

We appreciate Dr. Souweidane's review of our article and his comments regarding the controversy surrounding the method of craniocervical decompression that best treats CM-I and syringomyelia. We concur that neurosurgeons generally agree about the need for adequate bony decompression in the primary treatment of CM-I and syringomyelia and often differ about other aspects of the procedure, such as the advisability of opening the dura, arachnoid, and fourth ventricle and of reducing the size of the cerebellar tonsils. It is our opinion that the effectiveness of surgery is most dependent on the ability of the procedure to permanently eliminate the blockage to normal CSF flow at the foramen magnum, which initiates the mechanism of syringomyelia.[1,2] It has been our practice in patients without previous surgery to perform an adequate bony decompression, open the dura, and preserve the arachnoid of the cisterna magna. In most cases the arachnoid of the cisterna magna will bow posteriorly after opening the dura, and observation through the translucent arachnoid will reveal whether or not significant adhesions are present within the subarachnoid space. In the rare case in which subarachnoid scarring or an abnormal band of arachnoid is present, the arachnoid membrane is opened and intraarachnoidal adhesions are cut to allow free pulsatile CSF flow across the subarachnoid space. In our study of patients who previously underwent surgical treatment, adhesions between the dura and arachnoid made it impossible for us to open the dura and preserve the arachnoid membrane, resulting in entrance into the subarachnoid space in the reoperated patients. Our study indicates that a strategy of reexploration of the previous craniocervical decompression in patients with failed CM-I and syringomyelia surgery will usually reform the dorsal CSF pathway at the foramen magnum, permanently reduce the size of the syrinx, and arrest neurological progression from syringomyelia. On the other hand, the occasional patient in whom arachnoiditis extends beyond the craniocervical junction will not be helped by this approach because the surgery will not remove the block to the free flow of CSF during the cardiac cycle nor resolve syringomyelia. In our article we mention other potential reasons for the failure of craniocervical decompression, including hydrocephalus, basilar invagination, instability, hypermobility, and prominent retroflexion of the dens. We agree with Dr. Souweidane that the treatment for CM-I should be modified as required to adequately address associated conditions.

The presence of a healthy control group proved to be essential to recognizing the magnitude of differences in CSF flow and cervical pulse pressure between patients and healthy volunteers. Healthy volunteers were available and willing to serve as participants in this institutional review board–approved clinical study. Potential risks were minimized by the skills of the neuroradiologists experienced in the research techniques; actual effects were limited to an occasional spinal headache.

The clinical, radiographic, and physiological findings in this study are consistent with the theory previously proposed for the mechanism of syringomyelia.[1,2] Failed surgery is signaled by the continued progression of myelopathy, distention of the spinal cord by the syrinx, and obstructed CSF movement at the foramen magnum during the cardiac cycle. Successful primary or secondary surgery reverses this pathophysiological mechanism and results in syrinx reduction and the arrest of neurological progression. *(DOI: 10.3171/2010.5.SPINE10213)*

References

1. Heiss JD, Patronas N, DeVroom HL, Shawker T, Ennis R, Kammerer W, et al: Elucidating the pathophysiology of syringomyelia. **J Neurosurg 91:**553–562, 1999
2. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ: Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. **J Neurosurg 80:**3–15, 1994