Editorial

Chiari malformation Type I

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In their paper, Noudel et al. carefully analyzed the presentation, change in posterior fossa volume (PFV), and outcome of a series of 11 adult patients undergoing posterior fossa (PF) decompression for Chiari malformation Type I (CM-I). They performed an occipital craniectomy and C-1 laminectomy with dural opening and expansion but with meticulous preservation of the arachnoid layer. Their craniectomy was approximately 3 × 3 cm. The authors state that this may be less than optimal in some individuals. They conclude that the clinical response to surgery was significantly influenced by the extent of the PFV expansion and that an ideal bony expansion can be predicted based on preoperative MR imaging findings.

Of the many issues surrounding CM-I surgery, the extent of bony decompression is 1 of the 3 most pertinent issues discussed today. The other 2 issues regard which clinical symptoms will respond to surgery and which anatomical layers should be included in the decompression. The authors propose and validate a technique to measure PFV and then use the technique to assess a response to surgery.

Limitations of the study include the small number of patients analyzed (11), the varying types of their presentation and preoperative imaging findings, and the relatively short duration of follow-up of a chronic disease. The absolute measurements should not be applied to pediatric patients, and the technique of dural opening is especially challenging with the large venous sinuses seen over the PF in small infants and children. A small percentage (5%–10%) of patients with CM-I will have a medullary veil or PF in small infants and children. The authors state that this may be less than optimal in some individuals. They conclude that the clinical response to surgery was significantly influenced by the extent of the PFV expansion and that an ideal bony expansion can be predicted based on preoperative MR imaging findings.

The surgical approach emphasizes the maintenance of the arachnoid to protect the subarachnoid space and the need for a meticulous closure. Soiling of the subarachnoid space with blood or other inflammatory material can be counterproductive to the goal of maintaining free and easy CSF egress from the fourth ventricular outlet.

The authors conclude that a larger bony opening yields a more favorable clinical outcome. The reader is told that no patient suffered from cerebellar “slump” as a consequence of too aggressive of a bony decompression but that this possibility certainly exists, especially when the decompression exceeds 4 × 4 cm. Adding objective measurement of the extent of the bony removal in a CM-I decompression adds significantly to the surgical refinement of this relatively common procedure.

Reference


Response

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We would like to thank Dr. Oakes for the thoughtful comments on the results of our study.

We are especially grateful because he emphasizes that the topic of our study focused on a major question about CM-I surgery, which concerns the identification of the size of the occipital craniectomy leading to the optimal volume for the patient-specific PF. Despite the potential harmful effects of a quantitatively inadequate bony removal, the craniectomy size often remains arbitrarily decided for all patients based on the experience and preference of each neurosurgeon. Too small of a craniectomy may fail to resolve or lead to the recurrence of the primary symptoms, whereas an excessive enlargement may cause cerebellar slump through the craniectomy defect, accounting for an important cause of poor long-term clinical results and requiring additional surgery for PF box reconstruction.

Therefore, performing objective measurements of the extent of the bony removal is likely to provide a significant contribution to the surgical refinements of the technique. The aim of our article was to measure the PFV expansion resulting from the creation of an enlarged cisterna magna when a similar sized and shaped craniectomy was performed, and to search for its correlations with the treatment response to determine the patient-specific PFV expansion, which corresponds to the variation of PFV associated with complete recovery and no complications. The results of the present study showed that the clinical response to treatment was significantly influenced by the amount of PFV increase, allowing us to validate a technique for the accurate measurement of the PFV variations after surgery, which usually concern small variations in CSF volume.

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Above all, performing the same size craniectomy in the 11 patients in our study led us to conclude that the relative PFV increase has wide interindividual variations (1.5%–19.7%) based on the initial PFV. For the same craniectomy size, small PFs harbor a significantly greater PFV increase and a better treatment response, suggesting that the patients who responded partially to surgery would have benefited from greater bone removal, and emphasizing the fact that any approach with a fixed operative technique has limitations. Because of the great variability of the patient-specific findings, the size of the craniectomy should probably be adapted to the PFV of the individual patient.

Our results also confirm that approximate measurements for optimal bone removal in an individual patient could be made on the basis of preoperative MR imaging findings, because in our study, the PFV increase was dependent on the PF size and the basioccipital length. The present study should definitely be considered as a preliminary stage in the perspective of tailoring the amount of bone removal to the patient-specific PF size as measured preoperatively.

We agree with Dr. Oakes about the limitations of our study regarding the small number of patients. We hereby confirm that a more extensive study in which repeatable measures are used in more patients is necessary to support our results and to evaluate further applications; such a study is presently underway in our center. The essential limit when analyzing the correlations between the treatment response and a quantitative parameter such as the PFV increase is the clinical presentation because it still represents the most important prognostic factor. Even when an optimal PFV increase is obtained, final results remain heavily influenced by the presenting signs and the level of disability, a poor preoperative class being associated with a worse postoperative outcome. Because the treatment response may be dampened by the predominance of poor prognostic factors, a careful analysis of the preoperative distribution of the symptoms remains crucial in the management of CM-I.

As stressed by Dr. Oakes, other questions about CM-I surgery deal with the approach to the arachnoid and tonsils, as well as the need for dural opening and grafting. Despite the fact that cerebellar ptosis and CSF-related complications may occur less frequently if the dura is partially thinned without entering the intradural space, we systematically opened the dura mater at the cranio-cervical junction because it has proven to allow a better outcome and an increased rate of syrinx resolution, requiring fewer additional interventions. After the restrictive dura mater has been removed, an effective cisterna magna is able to form as an enlargement of the arachnoid membrane that can expand under the normal pulsating CSF flow, avoiding the need for subarachnoid dissection and tonsillar resection in most cases. Our results confirm that a meticulous extrarachnoidal approach without duraplasty allows for good recovery and the formation of an artificial cisterna magna, without increasing the risk of symptomatic cerebellar ptosis or of pseudomeningocele. Despite the fact that the standardized craniectomy size we performed was not adapted to the patient-specific PFV, the good results we observed support the notion that a large artificial cisterna magna is protective against slump. On the contrary, the potential of arachnoidal scar due to any intraarachnoidal procedure, or the risk of dense meningeal fibrosis due to subsequent inflammatory graft reaction can lead to the obstruction of free CSF flow, increasing the effects of the craniospinal pressure dissociation upon a cerebellum without any bony support. In addition, the dural graft can favor the formation of a compressive pseudomeningocele because the lack of watertightness of dural sutures may allow a unidirectional CSF leak to pass through the sutures, creating a second CSF compartment between the graft and the muscular layer.

However, patients with dense arachnoiditis at the foramen magnum represent a limitation to this approach, and we agree with Dr. Oakes about the fact that the extrarachnoidal technique puts us at risk of missing some arachnoidal adhesions requiring a more aggressive surgery. Since completion of the present study, we have treated 30 patients with the same craniectomy size, but in some cases, especially those patients with marked tonsillar herniation of more than 15 mm and those with severe arachnoiditis, we had to open the arachnoid widely and reduce the volume of the tonsils with bipolar retraction or subpial resection. The results were favorable, with no CSF leak or cerebellar fixation due to fibrosis and with the creation of large retrocerebellar spaces, opening of the cerebellomedullary fissure, and ascension of the hindbrain.

Because CM-I is a dynamic disease, we strongly believe that there is not a single and standardized procedure to treat this process. The craniectomy size and the approach to the arachnoid and tonsils should be decided on a case-by-case basis. The results of our study enhance the need for further measurements in a greater number of patients to validate a reproducible methodology, allowing us to predict the optimal craniectomy size according to the patient-specific PFV.

References

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