Syringomyelia

To the Editor: We read with great interest the recent article by Heiss and colleagues (Heiss JD, Suffredini G, Smith R, et al: Pathophysiology of persistent syringomyelia after decompressive craniocebral surgery. Clinical article. J Neurosurg Spine 13:729–742, December, 2010).5 The authors conducted a prospective study in 16 patients with Chiari malformation Type I (CM-I) who had persistent syringomyelia despite previous craniocebral decompression. The authors found partial blockage of CSF pathways at the craniovertebral junction with abnormally elevated CSF pressure and pulse pressure in the cervical subarachnoid space. During surgery, ultrasonographic observation demonstrated pulsatile movement of the cerebellar tonsils. On the postoperative MR imaging study, the CSF pathway had been restored with normalization of the cervical subarachnoid space. The size of the syrinx decreased in 15 of the 16 patients. Based on these findings, the authors concluded that persistent blockage of the CSF pathways at the foramen magnum resulted in increased pulsation of the cerebellar tonsils, which created elevated CSF pressure waves, which in turn affected the external surface of the spinal cord to force CSF into the spinal cord through Virchow-Robin spaces.

We wholeheartedly agree with the authors that persistent syringomyelia after decompressive craniocebral surgery is often caused by inappropriate decompression of the CSF pathways, recurrence of its blockage, or arachnoid adhesion either previously unnoticed or newly formed, resulting in insufficient normalization of the CSF dynamics. The condition should be treated with additional surgery if the pathology is evident on imaging studies. However, we have serious concern about the authors’ conclusion regarding the pathophysiology of syringomyelia from the data presented in this article. The authors’ hypothesis is that the pulsatile movement of the tonsils creates elevated pressure waves in the subarachnoid space, which in turn force the CSF into the spinal cord through Virchow-Robin spaces. We understand that the evidence supporting this hypotheses is that 1) the pressure of the cervical subarachnoid space as well as its pulse pressure were elevated preoperatively and were subsequently normalized after surgery; and 2) pulsatile movement of the tonsils was observed on intraoperative ultrasonographic images. Yet, we believe that these pieces of evidence have rather weak linkage to their aforementioned hypothesis.

First of all, the subarachnoid pressure is constantly lower than the syrinx pressure, which is necessary for the syrinx to keep its distended status.3 This is directly proven by the pressure measurement described in the authors’ previous publication.4 The fact that the authors observed increased subarachnoid pressure in patients with CM-I does not necessarily mean that this pressure difference is thereby simply reduced. The pressure difference may be unchanged or become greater resulting in the syrinx pressure becoming much higher than the subarachnoid pressure, producing a larger pressure gradient compared with the normal status. Actually, in our mathematical model of spinal CSF dynamics,1,2 the absolute cervical subarachnoid pressure increased when we decreased the capacitance of the cisterna magna, simulating the CM-I (data not shown). However, at the same time, the pressure difference between the syrinx and the subarachnoid pressure was also increased, compared with the normal status, with the syrinx pressure becoming much higher than the subarachnoid pressure.1 In that situation, we will have to imagine the CSF entering the syrinx through the spinal cord against the higher pressure gradient impeding such entrance. In the first place, the authors’ hypothesis that the CSF enters the syrinx against the impeding pressure gradient (higher syrinx pressure against lower subarachnoid pressure) seems to us somewhat difficult to concede. We are afraid that the data presented in this article do not support the authors’ conclusion on the pathophysiology of syringomyelia in CM-I.

Second, the authors’ intraoperative findings showed 10 cases of intradural adhesion. We know that the arachnoid adhesion itself can block the normal CSF dynamics and become the cause of syringomyelia, not necessarily accompanied by any piston-like movement of any object.2,3 Although the authors state, “The cerebellar tonsils pulsed actively, descending during cardiac systole and ascending during diastole,” they did not specify whether they observed this phenomenon in all of the patients or only in some, or whether they observed this phenomenon in the patients with arachnoid adhesions. They also did not describe whether the intensity of that movement varied in individual cases. This information is the most important piece in the
authors’ argument leading to their aforementioned hypothesis. Lack of this information critically undermines the logic of this article.

We highly esteem the authors’ effort and their presentation of well-studied data that are of high quality. However, we are afraid that the presented data do not necessarily support the authors’ conclusion.

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Disclosure

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References


RESPONSE: We thank Drs. Chang, Tsuchiya, and Matsui for reading and commenting on our article and for their interest in our work. Their letter begins with a statement of concordance with our clinical management of patients with persistent syringomyelia after craniocervical decompression of CM-I. The rest of their letter 1) attempts to refute the mechanism of persistence of syringomyelia that we proposed in this and previous manuscripts;4,5 and 2) supports their hypothesis of syrinx pathogenesis.2 We are pleased to respond to their comments.

The usefulness of a scientific theory largely depends on its ability to explain observations. We wish to correct a misstatement in their letter about one of our previous observations. Their third paragraph states incorrectly that in one of our previous papers we reported that the syrinx pressure was greater than the cervical subarachnoid pressure. In contrast, we specifically wrote in that article, “Syrinx pressure was identical to cervical subarachnoid pressure (syrinx 15 ± 5.8 mm Hg; cervical 15.1 ± 4.7 mm Hg; correlation coefficient 0.997).” The remainder of this paragraph of their letter refers to their mathematical modeling, which is based on the supposition, which was unsupported by the data in our paper, that syrinx pressure exceeds subarachnoid pressure in patients. Furthermore, it should be acknowledged that their hypothesis is derived from an analysis based on a simple electric circuit, rather than a biological model, and which has no clinical or laboratory data supporting it.

The next paragraph criticizes the brevity of our description of cerebellar tonsillar pulsation observed by intraoperative ultrasonography. This criticism is easily rectified. All patients in our study on the pathophysiology of persistent syringomyelia had CM-I, so their cerebellar tonsils extended through the foramen magnum. The CSF spaces at the foramen magnum were narrow, similar to those in patients in our previous study who had not previously undergone surgery.1 Arachnoiditis was limited to the foramen magnum region and was dorsal in location, allowing it to be relieved by a dorsal approach in all but one of the patients. Tonsillar motion was similar in amplitude (2–3 mm) to patients who had not previously undergone craniocervical decompression. In the one patient with extensive arachnoiditis in the craniocervical area that could not be relieved, tonsillar motion was restricted dorsally but persisted in the medulla and anterior part of the tonsil. In all cases, brain expansion during systole appeared to result in a piston motion on the CSF at the foramen magnum and in the upper cervical spinal canal. As noted, this motion on a subarachnoid space with reduced compliance produced enlarged CSF pressure waves in the cervical subarachnoid space. Craniocervical decompression that effectively opened the obstructed CSF spaces at the foramen magnum increased CSF flow at the foramen magnum, reduced craniocervical compliance and cervical pressure, and led to syrinx resolution. Finally, the authors seem to have the notion that the degree of visible movement of the tonsils is linked to their having a piston-like effect on the spinal subarachnoid space. That is not necessarily so, since the physical movement of the tonsils to impart an exaggerated pulse pressure in the spinal subarachnoid space is related to the degree of altered compliance in that space. Patients with much-reduced compliance may have limited tonsillar motion associated with high pulse pressures in the spinal subarachnoid space. Thus, our observations in the study of pathophysiology associated with persistent or recurrent syringomyelia are entirely consistent with our hypothesis.3,5

The hypothesis for syrinx pathogenesis espoused by the authors’ letter is based on observations in one patient with syringomyelia, observations that are contrary to those that have been reported by other authors.1 Their case report proposes that syrinx enlargement results from a reduction in cervical subarachnoid pressure and that syrinx reduction results from an increase in cervical subarachnoid pressure. The subject of their paper was not a patient with CM-I and syringomyelia, as in our study, but a 60-year-old with “a history of multiple surgeries to treat her syringomyelia associated with adhesive arachnoiditis.” In their case report, MR imaging demonstrated a distended syrinx extending from the T-6 segment to the conus and a narrow syrinx proceeding from the T-6 to the C-1 spinal segment, consistent with a syrinx originating from arachnoiditis in the lower thoracic spine. The authors performed the following 3 surgical interventions: 1) a cervical subarachnoid-peritoneal shunt using a medium pressure valve, which was followed by enlargement of the cervical extension of the thoracic syrinx seen on
MR imaging 2 months later, “although her preoperative symptoms were subjectively relieved;” 2) foramen magnum decompression and lumbar theca exploration, which “transiently reduced the size of syrinx,” although reexpansion of the cervical part of the syrinx occurred 7 months later, associated with stable neurological status and the use of a walker; and 3) revision of the cervical subarachnoid-peritoneal shunt by replacing the medium-pressure valve with a programmable valve set at a higher pressure. This procedure resulted in improvement in lower-extremity spasticity, but “lower-extremity weakness kept her wheelchair bound.” The diameter of the cervical extension of the syrinx was smaller when evaluated by cervical spine MR imaging 2 months after surgery. The authors assume that the cervical portion of the syrinx became smaller because the cervical subarachnoid pressure increased after replacing the previous medium-pressure valve with a programmable valve set at a higher pressure. At no time in this patient’s evaluation or treatment was subarachnoid or syrinx pressure measured. The cumulative effect of the 3 surgical procedures was that the patient’s paraparesis advanced and the cervical portion of the syrinx returned to the diameter it had before these 3 surgical treatments.

The most likely explanation of the findings of the case report is that revision of the shunt by placing a programmable valve resulted in improvement in CSF drainage, despite the authors’ contention that the medium-pressure “shunt system was functioning normally.” We base this assumption on 5 published reports that have demonstrated reduction in syrinx size and stable or improved neurological function after thecal shunting. The most recent of these articles reported on 7 patients with thecoperitoneal shunting rostral to a subarachnoid block. Six of the 7 patients improved symptomatically and the other patient remained stable. Of the 6 patients available for imaging follow-up, all had reduction in syrinx size. In summary, the observation made in the case report that syrinx enlargement was caused by a decrease in cervical subarachnoid pressure and that syrinx reduction resulted from an increase in cervical subarachnoid pressure is refuted by many published reports. Possible explanations for their case report yielding an observation that is discordant with other published reports are that 1) replacement of the valve improved shunt function or 2) spontaneous drainage of the cervical portion of the syrinx might have occurred in the 2 months between placement of the higher-pressure valve and an MR imaging study that documented reduction in the diameter of the cervical portion of the syrinx.

Obstruction of the free pulsatile movement of CSF in the subarachnoid space appears to be a critical etiological element in syringomyelia pathogenesis. This observation supports certain surgical treatments, such as in our current and previous articles, treatments that directly relieve the obstruction, to successfully address the abnormal pathophysiology and treat syringomyelia.

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