Neurosurgical Forum LETTERS TO THE EDITOR

Shunt dysfunction and constipation

TO THE EDITOR: We appreciated the article by Morais et al.² describing a 6-year-old shunt-treated spina bifida patient in the emergency department, presenting with mixed symptoms of shunt malfunction and abdominal pathology (Morais BA, Cardeal DD, Andrade FG, et al: Reversible ventriculoperitoneal shunt dysfunction and chronic constipation: case report. *J Neurosurg Pediatr* 22:147–150, August 2018). The authors demonstrate concomitant ventricular dilatation and abdominal constipation, which resolved with nonsurgical management. The discussion included the possibility that intracranial hypertension could cause bowel dysfunction. At our institution, we frequently observe the clinical phenomenon of transient shunt malfunction related to constipation. Here, we provide 3 recent cases, which expand the scope of the problem.

The first case involved a 16-year-old boy with neonatal posthemorrhagic hydrocephalus, ventriculoperitoneal (VP) shunt surgery, epilepsy, and history of chronic constipation, who presented to the emergency department with refractory vomiting and headache over a 24-hour period. MRI revealed ventricular enlargement, and an abdominal radiograph showed increasing colonic burden of stool (Fig. 1). A shunt tap was performed with brisk flow, low opening pressure, and good distal runoff. The patient was hospitalized for medical management. Over the next day, his symptoms and radiographic changes remitted (Fig. 1).

In our second case, a 16-year-old boy with spina bifida, cervicothoracic syrinx, and shunted hydrocephalus, the patient presented to the clinic for routine follow-up, with no symptoms of shunt malfunction. However, the patient's mother noted very hard stools and decreased frequency of bowel movements. MRI of the brain and spine was remarkable for simultaneous increase in his ventricular size as well as caliber of his cervical syrinx (Fig. 1). Given these findings, he was scheduled for an elective shunt revision if imaging findings did not improve after outpatient bowel regimen. Follow-up imaging at 2 weeks revealed his ventricular size had returned to baseline (Fig. 1), and no surgical intervention was pursued.

Our third recent case was a 13-year-old boy with history of congenital hydrocephalus that had been shunted as an infant. He presented to the clinic for annual follow-up without symptoms of shunt malfunction. Abdominal radiographs revealed increasing stool burden. MRI revealed increased ventriculomegaly (not pictured). He was started

FIG. 1. The first patient's asymptomatic baseline imaging is shown (A and D), along with evidence of ventricular enlargement and increased stool burden (B and E), with resolution after medical management of constipation (C and F). The second patient's asymptomatic baseline imaging is also shown (G and J). Ventricular enlargement along with increased caliber of cervical syrinx was seen (H and K), that resolved with aggressive bowel regimen (I).

on an aggressive bowel regimen and follow-up imaging showed return to his baseline.

As the aforementioned cases illustrate, constipation-related shunt malfunction can occur within multiple different etiologies of hydrocephalus, even with a reassuring shunt tap, and could also change syring flow dynamics. It is thought that shunt malfunction can be related to multiple conditions that increase intraabdominal pressure, such as pregnancy and obesity.^{1,4} Sustained increases in intracranial pressure have even been observed in tapped shunts of patients undergoing laparoscopic procedures, which resolve after the procedure, implying that the increased abdominal pressure could cause relative shunt malfunction.³ We agree with Morais et al. and have also found constipation to be an important cause for reversible shunt malfunction.² These cases highlight the need for clinicians to include a thorough evaluation of constipation in the routine workup for VP shunt patients, as this can reduce the need for surgery.

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Response

We thank Dr. Northam and colleagues for their interest and comments regarding our paper. Chronic constipation commonly affects neurological patients (e.g., dysraphism and cerebral palsy patients) and therefore coexists with

hydrocephalus. Our paper's goal was specifically to highlight this condition as one reversible cause of VP shunt malfunction.

The first literature report that proposed constipation as a possible factor in VP shunt malfunction was Bragg et al. in 1994.¹ However, since then, there are few well-documented cases.^{2–4} We believe this clinical phenomenon is often forgotten and even misdiagnosed, leading to potential unnecessary surgeries for children.

The authors contribute here with 3 cases of constipation-related shunt malfunction. We would like to add some comments. First, the 3 cases involved children who were 13 years of age or older, individuals who are more prone to present with specific signs or complaints of associated obstipation. We should keep in mind that this phenomenon can be even more common in younger children due to their vulnerability to increased abdominal pressure and that their symptoms tend to be more unspecific. Therefore, we recommend an active clinicoradiological investigation.

Second, we found it interesting that two of the described cases were asymptomatic; however, a routine follow-up revealed a ventricular dilatation had reverted with constipation treatment. This suggests that this phenomenon is likely to be more frequent in a day-by-day reality than expected.

Additionally, the authors' proposition of constipation-related shunt malfunction also changing the syring flow dynamics is really interesting and should be further investigated.

In summary, we recommend that constipation should be treated and properly addressed in the scenario of shunt malfunction, if possible, prior to new surgical procedures. We were happy to notice that this issue has been recognized and further considered when daily dealing with shunt malfunction.

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Shunt occlusion by ventricular material

TO THE EDITOR: I read with interest the article by Kraemer et al.² (Kraemer MR, Koueik J, Rebsamen S, et al: Overdrainage-related ependymal bands: a postulated cause of proximal shunt obstruction. *J Neurosurg Pediatr* 22:567–577, November 2018). The authors present observations from a retrospective series of ventricular shunt revisions performed between 2008 and 2015, identifying ependymal bands as the cause of proximal catheter obstruction in the majority of cases. They have labeled as “ependymal bands” the tissue that has been drawn into the openings of the ventricular catheter. They hypothesize that these bands form because of siphoning and overdrainage, and they note that the bands are seen in patients with small ventricles who undergo shunt revision.

As a former trainee of the senior author, my attention was first drawn to this phenomenon during residency. Since that time, I have performed 89 ventriculoperitoneal shunt revisions in my own practice, in nearly all of which I have used endoscopy for placement of the new ventricular catheter. Although I have not systematically collected the data necessary to make a scientific claim, it is my observation that in essentially all cases of ventricular catheter obstruction, ependymal bands are present. Furthermore, these bands are present regardless of the size of the ventricles or the appearance of the preoperative imaging. If one can find the spot in the ventricle where the previous catheter tip was located, almost invariably there are the bands. Furthermore, if the ventricles are dilated, one can sometimes find rows of bands corresponding to 2 rows of catheter holes on one ventricular wall, and then find another row on the opposite ventricular wall, several millimeters away, indicating that this was the surface in contact with the catheter when the ventricle was small. In short, I am a firm believer in ependymal bands.

However, I would challenge the notion that this phenomenon is a consequence of overdrainage or siphoning. Ventricular shunts function by allowing CSF to flow out of the ventricle. All modern shunts include a 1-way valve preventing flow in the other direction. Therefore, to some degree, ventricular tissue will be drawn into every shunt. This is not something that can be mitigated by preventing overdrainage. Rather, it is the fundamental way that shunts work, and therefore will always play a role in shunt failure. I would hypothesize that, in fact, this is the major reason that shunts fail. Our treatment for hydrocephalus is primarily based on the bulk-flow model of CSF physiology: too much CSF in the brain; CSF must be removed. Increasing evidence suggests that the true underlying problem in hydrocephalus is alteration in brain compliance and abnormal CSF pulsatility, along with problems with absorption.^{3,4} As we continue to treat this complex problem with a simple drain, we will continue to face high rates of shunt failure.

I would also challenge the naming of the obstructive tissue as “ependymal bands.” Whereas historically neurosurgeons have posited that choroid plexus is the offending

tissue in shunt obstruction, it is now recognized that this is not always (or even often) the case. However, analyses of the material occluding shunt catheters has not revealed ependymal tissue either.¹ Rather, a mixture of glial, inflammatory, and reactive tissue has been described. Furthermore, while the cases labeled in the present manuscript as “severe” may resemble bands when viewed from outside the catheter, most often the appearance of the obstructing tissue is most similar to nipples. Therefore, for lack of a better term, I would propose to call this phenomenon “ventricular nipples.”

For the foreseeable future, we are clearly stuck with shunts. Therefore, research into limiting the formation of ventricular nipples, whether by mechanical or biological modifications, should be highly encouraged. The authors are to be commended for this excellent description of a previously unreported but ubiquitous finding.

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Response

We always appreciate Dr. Rocque’s comments, whether verbal or in print, as they are typically thought-provoking and colorful. Ependymal bands do indeed seem ubiquitous, and they do develop in ventricles that collapse when shunts are functional and may or may not dilate when shunts obstruct. Yet, we diverge with Dr. Rocque on the notion that regardless of valve mechanism, CSF drainage through a shunt system will always draw in tissue, making failure unavoidable. Decidedly, the force required to pull water into a conduit is different from one that pulls soft tissue. For instance, when drinking through a straw, one has to apply greater suction to drink a thick milk shake versus pure water. Furthermore, according to Bernoulli’s principle, faster flow in itself creates a suctioning effect. We believe that there are finer points of fluid flow that need to

be explored and understood in order to build a shunt system that would maintain effectiveness over the long term.

The cellular composition of ependymal bands has been intriguing to us as well. The histology literature describes debris that had broken off from the ependyma and settled in the ventricular catheter. In our experience, this points to a severe and chronic process, starting early on with mounds of ependymal tissue, which with continued long-term suction via gravitational forces become progressively stiffer, presumably secondary to inflammation, gliosis, and cellular damage, thus eventually losing ependymal architecture. Something has to drive the inflammatory changes. The data suggest that this is most likely a process of suction.

Finally, we appreciate the vividness of Dr. Rocque's proposed nomenclature, but think that it may prove uncomfortable to use when explaining the phenomenon to colleagues, students, and patients. Still, the exact terminology used to describe the ependymal protrusions is not

nearly as important as understanding the paradigm of drainage-driven tissue distortion and shunt obstruction, and the consequence of such understanding on present-day decision-making and future valve design.

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