HERNOV, et al., provide an innovative and provocative treatment for a group of shunt-treated patients that all neurosurgeons have difficulty treating—those with so-called “slit-ventricle syndrome.” In their series, the complication rate is virtually zero and shunts were able to be explanted in all patients. The study is retrospective, involves a small group of 15 patients, and the follow-up period is relatively brief—1 year or less in 40% of the patients. The proposed treatment also raises a significant issue, which is that many of their patients appear to have been asymptomatic at the time of the staged procedure involving endoscopic third ventriculostomy (ETV) and shunt removal.

In the neurosurgical literature and in the minds of neurosurgeons there are many interpretations and descriptions of slit-ventricle syndrome. As in this series, the patients with this condition usually have undergone shunt placement, suffer from headaches, and have small ventricles. As the authors discuss comprehensively, however, within this clinical constellation exists a variety of shunt problems, including blocked shunts, shunt overdrainage, and headache apparently unrelated to shunt function. Although the authors use the definition of slit-ventricle syndrome proposed by Rekate—which, in addition to headaches lasting 10 to 30 minutes, includes a slow-filling shunt reservoir—this does not always allow one to distinguish among different types of shunt problems. The pathophysiology of slit-ventricle syndrome is, as demonstrated in Fig. 1 of this article, highly theoretical and has been validated only in part and only in a few patients. Often intracranial pressure (ICP) monitoring, shunt-flow studies, and even operative exploration are required to determine the shunt problem. Two of the patients in this series experienced moderate headache and normal ICP at the time of operation; it is possible that these patients could have been suffering from headaches unrelated to shunt function.

Surgical Technique

The authors’ surgical technique is quite innovative. A small-diameter (2.5-mm) fiberscope with a working channel is used to pass along the ventricular catheter into the slit ventricle and remove the ventricular catheter under direct vision. The ICP is measured through the ventricular catheter following disconnection from the valve. Next, an infusion study is performed to estimate ventricular compliance with the aid of an external ventricular drainage catheter whose outlet is set to 10 cm H₂O. If, as determined by subjective assessment, the ventricles expand enough to allow access to the third ventricle, compliance is assumed to be adequate and ETV is performed. Otherwise, the ventricular catheter is revised and a Codman-Hakim programmable valve, usually set to an opening pressure of 20 cm H₂O, is inserted.

In patients treated by the two-stage method, the larger-diameter scope was used electively at a mean of 16 months later to perform ETV in the ventricles that had expanded as a result of the programmable shunt. The ETV is performed with a 4.8-mm scope, and an opening in the floor of the third ventricle is made midway between the infundibulum and mammillary bodies by using a No. 4 French Fogarty catheter; care is taken to fenestrate all membranes below the level of the third ventricular floor.

The advantage of this particular technique of ETV is that it allows expansion of the ventricles to be performed immediately or on an outpatient basis by using an indwelling shunt, whereas traditional procedures involve externalizing the shunt and expanding the ventricles by performing external ventricular drainage in the hospital.¹ The patients in this study remained in the hospital for a prolonged period of time (2–3 weeks) for observation, however, following ETV.

The third ventricular floor in patients with small ventricles is often fairly thick, and there is a concern about injuring either hypothalamic structures, and or vascular and neural structures below the floor. In Fig. 3 the floor of the third ventricle appears partially translucent; moreover, the absence of serious complications in this report attests to the authors’ excellent surgical technique and indicates that such concerns about potential injury may be overemphasized.

Eleven of the 14 patients were treated using the two-stage procedure at a mean of 16 months after the ventricular catheter and valve revision. Although this schedule was apparently part of a planned two-stage treatment, the
patients appeared to be asymptomatic at the time of the second surgery with the exception of a few patients with blocked shunts. The authors previously advocated the use of the fiberscope and Codman-Hakim programmable valve for slit-ventricle syndrome in 20 patients, 11 of whom are discussed in this report. It is not clear why the authors have changed their philosophy and now believe the added ETV is a better procedure. Moreover, eight of the patients in the previous report also appear to have undergone surgery when they were not asymptomatic due to the authors’ fear that trouble might occur if shunt revisions were neglected.

Subecting complex, shunt-treated patients to yet another surgical procedure when they are well—particularly a procedure with the significant risks of an ETV—is of debatable value. Many patients who were well 1 year after the initial procedure might question the wisdom of undergoing another operation to prevent future problems. The authors’ excellent results may justify this approach in the short term; however, their follow up is limited. Authors of other series with longer follow up report that the long-term success rate of ETV may be closer to 75% in those shunt-treated patients with aqueductal obstruction. Although the authors state in the conclusion that “absence of a shunt results in the absence of all shunt-associated complications,” rapid deterioration and death have occurred, albeit rarely, in patients who have undergone ETV long after the procedure and in at least one case following shunt removal.

The authors have made an important contribution to the growing literature on the use of ETV for patients with failed shunts due to a variety of causes. This report pertains to those patients with slit ventricles, a particularly challenging group. These patients require extensive investigations to determine the causes of their symptoms and, if endoscopic treatment is contemplated, a surgery very skilled in this modality. Prolonged follow up and careful risk–benefit analysis will be required to determine if treatments such as the one proposed by Chernov, et al., are durable and if “ETV is an optimal primary treatment modality for hydrocephalus, regardless of its cause,” as stated in this paper.

References

Response: We are very thankful to Dr. James Drake for his thoughtful comments. He clearly underlined several pitfalls of our study, including its retrospective nature, small number of cases, and insufficient duration of follow up. We agree that some individuals with slitlike ventricles due to shunt treatment, but without slit-ventricle syndrome, could be enrolled. Should those patients be treated prophylactically or not? The majority of those in the neurosurgical community will say that they should not undergo prophylactic treatment. It should not be forgotten, however, that 40 to 68% of these patients become symptomatic a mean of 20 months after the neuroimaging diagnosis. Moreover, at the time of shunt obstruction 77% of patients rapidly deteriorate into lethargy and show associated narrowing of the ambient cistern on computerized tomography scanning. In addition, the neuropsychological effects of chronic intracranial hypotension, while not clear yet, may be harmful. Therefore, it is our policy to propose active treatment to all patients with slitlike ventricles due to shunt treatment, but the reasons for such an approach are definitely questionable.

What is not questionable is that patients with slit-ventricle syndrome should be treated. The problem is how does one treat these patients? As a result of the heterogeneity caused by incorporation of the several different conditions with the same name, the complex pathophysiology of the complication cannot be explained by any simplified scheme. Choice of the optimal management strategy may be based on ICP monitoring, shunt-flow studies, and operative exploration; however, the rate of recurrences is high if the shunt is preserved. Therefore, we proposed removal of the shunt and ETV. No significant perioperative complications were encountered. We did not observe unusual thickening of the third ventricular floor; however, it should be noted that in the majority of patients ETV was performed after a long period of ventricular enlargement resulting from implantation of the Codman-Hakim programmable valve. The latter by itself showed effectiveness in the management of slit-ventricle syndrome and frequently resulted in complete resolution of symptoms. Should those patients undergo the additional risk of an ETV? Let us look at the problem from another angle. Should those patients be left with a life-long risk of shunt complications when there is a real opportunity for the elimination? There is no doubt that ETV may be accompanied by late deterioration, which necessitates detailed information from the family and careful follow up. Two thirds of our patients exhibited some degree of persistent ventricular dilatation, and we could not be sure that some of those patients will not need implantation of a shunt later on. Nevertheless, based on the long-term follow-up examinations the complication rate of ETV is definitely lower compared with that for shunts.

Based on present and previous experience, we wish to emphasize that neurofiberscopic interventions can be successfully performed in patients who do not suffer from hydrocephalus. In the present series it was possible to perform primary ETV and shunt elimination in four patients with slit-ventricle syndrome. It appears that it is not the presence of ventriculomegaly but rather sufficient preservation of brain compliance that is the key factor in safe intraventricular neuroendoscopic manipulations. The risks and benefits of such procedures, however, particularly for management of slit-ventricle syndrome, need further prospective evaluation. In this way, the importance of highly
developed surgical skills and the availability of adequate neurofiberscopic devices should not be neglected.

References


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