Limitations of Neuroendoscopic Treatment for Pediatric Hydrocephalus and Considerations from Future Perspectives

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Abstract

Neuroendoscopy has become common in the field of pediatric neurosurgery. As an alternative procedure to cerebrospinal fluid shunt, endoscopic third ventriculostomy has been the routine surgical treatment for obstructive hydrocephalus. However, the indication is still debatable in infantile periods. The predictors of late failure and how to manage are still unknown. Recently, the remarkable results of endoscopic choroid plexus coagulation in combination with third ventriculostomy, reported from experiences in Africa, present puzzling complexity. The current data on the role of neuroendoscopic surgery for pediatric hydrocephalus is reported with discussion of its limitations and future perspectives, in this review.

Key words: hydrocephalus, neuroendoscopy, cerebrospinal fluid, pediatric, shunt

Introduction

The cerebrospinal fluid (CSF) shunt has long been the classic treatment for pediatric hydrocephalus. It can resolve nearly all forms of hydrocephalus, regardless of the etiology. It is a simple procedure that can aid in having a relatively normal life. However, its failure rate is significant. The reported rate of shunt malfunction in the first year of placement is approximately 30%, and about 10% per year thereafter. The risk of infection is between 5% and 10%. In addition, the high rates of shunt complication such as slit ventricle syndrome are unacceptable by current standards. Children with shunts are dependent on surgical maintenance throughout their lives.1–8) Therefore, the advent of neuroendoscopy was received with enthusiasm. Endoscopic third ventriculostomy (ETV) has been a routine surgical practice for the past two decades and provides an alternative to the CSF shunt. It is a straightforward procedure for diversion of the CSF and does not require placing devices in children’s bodies. The utility and safety of ETV have been proven for obstructive hydrocephalus that occur secondary to aqueductal stenosis. However, for other indications, it is necessary to examine the anatomy and etiology, as well as the patient's age. During follow-up after ETV, late failure can occur and may lead to rapid deterioration. The aim of this report is to review the current data on the use of neuroendoscopy in children and to discuss the limitations and future perspectives regarding this procedure.

Patient Selection and ETV in Infants

ETV has two main purposes: to restore CSF communication between the ventricle and subarachnoid space and to reduce transmantle pulsatile stress by increasing compliance of the ventricular wall.9) Since the 1990s, patients with aqueductal stenosis have been considered ideal candidate for ETV.10,11) ETV quietly developed into a common procedure, without any prospective randomized trials comparing its efficacy to that of the CSF shunt. In spite of strict patient selection, the overall ETV success rate does not exceed 80%.12–16) However, as neuroendoscopic technology has evolved and pediatric neurosurgeons have gained experience with the procedure, the indications for ETV have broadened. For example, patients with hydrocephalus who had a previously failed shunt have become ETV candidates. The success rate of ETV for shunt malfunction is notable, around 80%.17–20) In recent years, the use of ETV to treat many pathological conditions has been debated. These include myelomeningocele, Chiari type 1 malformation, Dandy-Walker malformation, and previous meningitis or hemorrhage.14,19,21,22)
In infants, the benefits conferred by ETV may be considerable due to the high complication rate of CSF shunts. However, there are two concerns with the use of ETV in infants. The first is safety of the procedure, including the possibility of damage to the ventricular and cisternal structures during surgery, CSF leakage, or infection soon after surgery and late closure of the stoma with rapid deterioration. The second concern is that an infant successfully treated with ETV may be transformed from active hydrocephalus to an arrested type. These infants often have larger ventricles than children treated with CSF shunts. No study has attempted to correlate the larger ventricle size with any measurements of psychomotor development. According to reports from a multitude of international studies, the shunt independence rate ranges from 25% to 89%. Two-thirds of the reports suggested that success rate is dependent on age of the infant at the time of ETV. To increase ETV success rates, greater accuracy of appropriate patients would be advised. However, because of the high rate of shunt failure and complication, ETV is sometimes preferred as a first-line treatment. Further, the range of what is defined as failure in this age group is very wide. A multicenter prospective randomized study on infants up to 2 years of age with no flow at the level of the aqueduct, named the International Infant Hydrocephalus Study (IIHS) is now under way. This study focuses primarily on the neurodevelopmental outcomes associated with different treatment paradigms at 5 years, and includes a comprehensive assessment of relevant risks and benefits.

Failed ETV and Its Management

ETV failure is a possible event. Although most failures from ETV occur in the early period, within a few days to 2 weeks following the procedure, late failure after many months may lead to rapid deterioration and even sudden death. A rapid increase in intracranial pressure caused by late obstruction of the stoma is typically regarded as the mechanism of failure. Early failure is attributed to the incorrect surgical technique or different criteria in the selection of patients. However, the predictors of late failure are still unknown. Therefore, patients who have undergone successful ETV should be followed on an ongoing basis. Neurosurgeons should encourage patients and their parents to return as soon as possible if any adverse symptoms develop, because these may have severe consequences. Setting the CSF reservoir concurrently with ETV is one option for the treatment of emergencies, even though it means implantation of a foreign material. In Japan, a follow-up magnetic resonance imaging (MRI) examination including sagittal T2-weighted images, cine-MRI, or constructive interference in steady state (CISS) is often scheduled to detect the CSF flow across the stoma. However, there is currently no evidence regarding whether the patients with no flow on MRI following ETV may be at a greater risk to develop clinical symptoms. For these patients, a repeat ETV may be performed immediately rather than close observation. In the literature, repeat ETV has a good success rate. Therefore, this is one option for patients with a failed ETV, and it provides a means of even avoiding the CSF shunt.

ETV vs. Aqueductoplasty

Endoscopic aqueductoplasty (EAP) is a means to restore the physiological CSF dynamics. This procedure provides an alternative to ETV, because it avoids the risk of severe arterial bleeding. EAP has been performed in cases with membranous or short segmental occlusion of the sylvian aqueduct. However, the long-term results of EAP have not been as successful as one would expect. EAP has been shown to fail frequently. Schroeder et al. reported a re-closure rate of 50%, and proposed that one contributing factor to re-closure could be lower aqueductal CSF flow through the stoma than that following ETV. In addition, aqueductoplasty is generally considered a riskier procedure due to the higher risk of injuring midbrain structures. It may lead to neurological deficits such as oculomotor or trochlear nerve palsy, Parinaud’s syndrome, and periaqueductal syndrome. Therefore, ETV, which has higher long-term success rates and less risk, would be a better alternative for membranous, short segmental, and even tumor-related occlusion of the aqueduct. However, the condition of isolated fourth ventricle (IFV) is an exception. Almost all patients with IFV have a medical history of hydrocephalus within first year of life, mostly post-infectious or post-hemorrhagic. Further, following shunt placement, they often experience complicated overdrainage with aqueductal stenosis. Aqueductoplasty could be a means of establishing CSF communication to the formerly isolated ventricular compartment; however, a stent is mandatory to keep this pathway open. EAP with a stent could be one choice in the endoscopic treatment of IFV.

ETV vs. CSF Shunt in Children

As described above, ETV has been widely applied for pediatric hydrocephalus as an alternative to the CSF shunt mainly in an attempt to avoid foreign body implantation and to better simulate physiological
CSF dynamics. However, the results of CSF shunt have rarely been compared to ETV. According to the pediatric study by de Ribaupierre et al., the failure rate of ETV was 26% and that of ventriculoperitoneal (VP) shunt was 42% at 5 years follow-up. In a review of the literature, the same trend was seen in other studies. Kulkarni et al. reported that the relative risk of ETV failure is initially higher than that for the CSF shunt. However, the risk became progressively lower at approximately 3 months following the procedure, and was approximately half the risk of shunt failure at 2 years. The decrease in ventricle size is usually smaller and happens more slowly after ETV compared to shunt. Pediatric neurosurgeons are sometimes concerned about the relationship between these neuroimaging changes and neuropsychological outcomes. Hirsch reviewed the postoperative intelligence quotient (IQ) was not significantly different in 70 patients with shunts vs. 44 who underwent percutaneous third ventriculostomy. Other reports also have found similar results. Recently, Kulkarni et al. reported that at 1 year following surgery, the quality of life and IQ scores were not significantly different between an ETV and a VP shunt group.

**Challenging Procedure of ETV + CPC**

There has been a resurgence in the technique of choroid plexus coagulation (CPC) in combination with ETV ever since this procedure was performed for African children with hydrocephalus of various etiologies in 2005. Warf and colleagues highlighted that hydrocephalus with shunt dependency is inadvisable in developing countries because of limited access to medical centers in the event of shunt malfunction. In 2005, Warf and colleagues reported the results of a combined ETV + CPC trial performed mainly in infants with post-infectious hydrocephalus and in those with myelomeningocele. A total of 266 patients underwent ETV + CPC, whereas ETV alone was performed in 284 patients. The results demonstrated that the ETV + CPC increased the success rate in infants from 47% to 66%. In 2008, they reported the long-term results of ETV + CPC in hydrocephalus with myelomeningocele. The intention-to-treat analysis showed a shunt independence rate of 76% in 338 infants, which was higher than the results of ETV alone reported in the literature. Further analyses regarding ETV + CPC performed in patients with encephalocele, Dandy-Walker complex, and congenital aqueductal stenosis have been conducted using their database. The success rates achieved were between 70% and 80%. However, all pediatric neurosurgeons should be cautious about the meaning of “success.” In these reports, success usually referred to controlling raised intracranial pressure and avoiding an extra-cranial CSF shunt. Cognitive function was not the main outcome index. In addition, all these results have been reported from Africa. In the coming years, the challenge will be to see whether these results from Africa can be extrapolated to developed countries. The main advantage of ETV + CPC was avoidance of a shunt in patients who were difficult to follow-up. In Africa, geographic and socioeconomic constraints reasons for poor follow-up. The low possibility of cognitive salvage or the high rate of shunt complications, such as holoprosencephaly or hydroanencephaly, may be further reasons in developed nations. Technically, Warf and colleagues used a steerable endoscope via a single burr hole, sometimes in combination with septostomy, and coagulated the bilateral choroid plexus from the foramen of Monro to the entrance of the temporal horn. This procedure appeared to be safe in some cases of severely dilated ventricles. For Japanese neuroendoscopic surgeons, the flexible endoscope is more familiar than it is in other European nations. Nevertheless, almost all pediatric neurosurgeons still believe that the use of ETV + CPC in pediatric hydrocephalus needs further evaluation.

**Conclusion**

It is difficult to determine the best strategy for CSF diversion in pediatric hydrocephalus. ETV is one of the alternative procedures to the CSF shunt in conditions of obstructive hydrocephalus such as aqueductal stenosis. However, the preferred indications in infants have not been fully determined. The mechanisms and predictors of late ETV failure that might cause rapid deterioration are not still known. Recently, other clinical dilemmas, such as the use of ETV + CPC have arisen. Definition of the optimal indications, perfection of endoscopic techniques, developing strategies for follow-up, and considering socioeconomic constraints are required from pediatric neurosurgeons when selecting a surgical strategy in pediatric hydrocephalus.

**Conflicts of Interest Disclosure**

The authors declare that they have no conflicts of interest.

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