Indication for Endoscopic Third Ventriculostomy

John Mugamba[1] and Vita Stagno[2]

Key words
- Choroid plexus cauterization
- Endoscopic third ventriculostomy
- Hydrocephalus
- Indications

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
ETV: Endoscopic third ventriculostomy

From the [1]Department of Neurosurgery, CURE Children's Hospital of Uganda, Mbale, Uganda; and the [2]Division of Neurosurgery, Università degli Studi di Napoli Federico II, Naples, Italy

To whom correspondence should be addressed: John Mugamba, M.D. [E-mail: mjohnk2006@yahoo.co.uk]

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BACKGROUND: **Endoscopic third ventriculostomy (ETV) is increasingly prevalent among pediatric neurosurgeons as the initial treatment for hydrocephalus. The combination of ETV and choroid plexus cauterization (ETV/CPC) has improved the success rate among infants with hydrocephalus for whom ETV alone is much less successful. In parts of the developing world where there are economic and human resource constraints, this mode of treatment may be more appropriate than the routine use of shunts, which are prone to failures that require urgent surgical treatment. Here we review indications for the use of ETV or ETV/CPC as the primary treatment for hydrocephalus.**

CONCLUSION: **Primary treatment of hydrocephalus by ETV can avoid shunt-dependence and its complications for many patients. Optimal results depend upon proper patient selection and the use of combined ETV/CPC when treating infants.**

INTRODUCTION

Third ventriculostomy done endoscopically was successfully used by Mixter (40) for treatment of hydrocephalus before the era of valve-regulated shunts. The procedure, however, had setbacks with early high mortality and morbidity and soon it fell out of favor as the shunts with valves gained widespread acceptance (1). Nevertheless, with a continued high rate of shunt malfunction and the rapid development of endoscopic technologies, the rationale for a physiological and anatomical diversion for CSF has gained once again more attention, leading to its reintroduction (35). Since that time, various indications have been proposed for application of endoscopic third ventriculostomy (ETV) in the management of hydrocephalus (9).

The superiority of ETV, if successful, is underlined by being minimally invasive and the CSF absorption being more physiological when the subarachnoid space is patent, and, perhaps most attractive, is the resulting decreased burden of future shunt-related dependency and complications.

The primary and most common indication for ETV is noncommunicating hydrocephalus due to idiopathic or secondary aqueductal stenosis (26, 51, 53). Fairly good success rates after ETV have also been reported for cases of communicating hydrocephalus in idiopathic normal-pressure hydrocephalus (22, 24, 37). Because a growing number of patients with hydrocephalus are considered "possible" candidates for ETV, the reported failure rate for this procedure has consequently increased with time.

A correlation between the state of the cisterns and the cause of hydrocephalus seem to be involved in the failure mechanism, as is highlighted by a higher incidence of failure reported in postinfectious or posthemorrhagic hydrocephalus (18, 69).

Endoscopic third ventriculostomy failure is age-related, being most common in infants. Most failures occur in the first months after surgery, but rare cases of delayed ETV failure have been reported (15, 18). Redo ETV is advisable where the previous ETV procedure has failed (49), particularly when the ETV had been initially successful for a period of time. Recent investigators have reported clinical parameters that help predict the likelihood of ETV success in any particular patient (23, 69, 70).

The aim of this article is to review the main surgical indications for a wide but rational application of ETV in the management of hydrocephalus.

Indications for ETV can be classified as absolute, where ETV should be considered as the first surgical option with a high likelihood of success, or relative, in which the expectation for success is lower and a consensus as to best practice has not been reached.

ACQUIRED AQUEDUCTAL STENOSIS

The occlusion of the aqueduct of Sylvius is one of the more common causes of hydrocephalus. It may result from congenital or acquired conditions. The endoscopic third ventriculostomy is nowadays considered the first and best surgical option for the treatment of hydrocephalus in these patients. The success rate is related to the cause of the occlusion and the age of the patient, as well as the clinical and radiologic characteristics. Among these, the acquired type of aqueductal stenosis has a higher success rate than the congenital form in selected series of patients (34, 35). Increasing patient age, regardless of the underlying cause, seems to favor outcome. This has been related both to the age at onset of hydrocephalus and the age at time of surgery by various authors (3, 35, 48, 61). In adult

patients, ETV is clearly established as the standard of care, and success rates of more than 80% have been reported (26, 31).

ETV IN POSTERIOR FOSSA, THIRD VENTRICLE, AND MIDBRAIN TUMORS

The incidence of hydrocephalus in patients with tumoral lesions located in the posterior fossa is extremely high (20, 53-55, 58). It is usually due to the occlusion of the CSF pathways at the level of Lushka and Magendie's foraminas and/or the compression of the fourth ventricle.

The noncommunicating nature of hydrocephalus highlights, rationally, the effectiveness of ETV in the management of patients with posterior fossa tumors, because it seems to provide a physiological/natural and effective solution to a "transient hydrocephalus" (5, 16). Nevertheless, there is still controversy regarding the timing of the procedure in relation to tumor removal because this alone can reinstate physiological CSF circulation allowing the resolution of hydrocephalus.

In the recent past the application of ETV has been extended also in the treatment of noncommunicating hydrocephalus associated with pineal region, third ventricular, and midbrain tumors (30, 38). Even though recent published results consist of small series of patients, retrospectively analyzed, the endoscopic fenestration of the third ventricular floor can be performed simultaneously with an endoscopic biopsy, leading to both diagnosis of the tumoral lesion and regression of hydrocephalus.

ETV IN CONGENITAL ABNORMALITIES

Endoscopic third ventriculostomy has gained widespread acceptance as an effective alternative to ventricular shunt placement in patients with hydrocephalus in many congenital abnormalities. The cause of hydrocephalus depends on the nature of the anatomic anomaly and can be multifactorial, including obstruction at the level of aqueduct of Sylvius (51), the fourth ventricular foramina (36), the craniocervical junction, or the arachnoid granulations.

The efficacy of endoscopic third ventriculoscopy has been demonstrated for congenital aqueductal stenosis, Dandy-Walker (41), and Chiari malformations (11, 25, 43) with fourth ventricle outlet obstruction (42) and recently has been assessed in the treatment of hydrocephalus in infants with encephalocele (44) and complex faciocraniosynostosis (12). In patients with hydrocephalus associated with Chiari malformation type I and syringomyelia, a reduction in the length and caliber of the syrinx cavity has been noticed following ETV, leading to resolution of the preceding symptoms in 83% of the series (25, 43).

Among the most common causes of congenital hydrocephalus has been that associated with myelomeningocele and Chiari malformation type II (45). Until recently, hydrocephalus was reported to require treatment in 80% to 90% of these children (63, 68). More recently, several authors have reported lower incidences of progressive hydrocephalus requiring treatment in this population, ranging from 52% to 66% (10, 14, 65, 68). A high rate of shunt-related problems, including infections or malfunctions, has disheartened neurosurgeons from performing shunt-placement in favor of endoscopic techniques. Although the anterior displacement of the basilar artery complex and the brainstem are considered limiting factors to access the prepontine cistern, and the third ventricular anatomy can be more complex in infants with Chiari malformation type II, the effectiveness of ETV in combination with bilateral choroid plexus cauterization as primary treatment for hydrocephalus in infants with myelomeningocele has been demonstrated, especially in developing countries (66, 68-70).

Other congenital abnormalities, like colloid cysts (71), arachnoid cysts (13, 62) with midline suprasellar or intraventricular extension, and quadrigeminal cistern cysts are commonly associated with obstructive hydrocephalus. In the same procedure, a CSF diversion through an endoscopic ventriculostomy together with the endoscopic fenestration of the cyst has been found beneficial.

POSTINFECTIOUS HYDROCEPHALUS

A significant number of infections that affect the central nervous system lead to hydrocephalus due to obstruction of the CSF outflow pathways (59, 60). In these cases, hydrocephalus can be caused by impaired CSF absorption or aqueductal or fourth ventricular outlet foramina obstruction. The widespread use of ETV has been already reported by many neurosurgeons in the management of postinfectious hydrocephalus with successful outcomes (28, 64, 67). Recently, the value of endoscopic management has gained attention in cases of chronic hydrocephalus without evidence of outlet obstruction, as in postmeningitis (4, 19, 28, 32, 33) ventricular enlargement (especially purulent and tuberculosis) or in neurocysticercosis (52, 64). In this case, an early extraction of the parasites, together with the ventriculostomy, seem to be effective in treating the accompanying communicating hydrocephalus.

With time, the expansion of indications for ETV has made patient selection difficult, especially in childhood where the effects of age and etiology on outcome have been particularly controversial (3, 48, 56, 61, 66, 69, 70). Although increasing patient age seems to favor outcome, in patients younger than age 2 years with postinfective hydrocephalus, ETV has not been a convincing option. Success rates of 50% or less in this category are reported (3, 35, 61, 63) and the results in the subgroup of patients under 6 months is even poorer (35, 48, 66). In fact, for some authors, it is a relative contraindication (29). However, in the Ugandan experience, ETV for infants younger than age 1 year with postinfectious hydrocephalus was successful in 59%, and successful in 70% of those infants in whom the aqueduct was obstructed (67). Cisternal scarring occurs in many of these postinfectious cases, and this has been demonstrated to more than double the rate of ETV failure (69). The less successful attempts with ETV in infants less than 6 months of age are thought to be due to the insufficient absorptive capacity in the subarachnoid space (26, 29, 48, 57). This has been one explanation for the favorable effect of adding choroid plexus cauterization to the ETV procedure in infants (66).

In developing countries, where the access to care is limited by the lack of neurosurgeons and competent centers, the rationale for performing ETV in patients younger than age 1 year is related to the enormous burden of postinfective hydrocephalus and the need to avoid the enhanced dangers of shunt dependence in this environment (69, 70).

ETV IN POSTHEMORRHAGIC HYDROCEPHALUS

Posthemorrhagic hydrocephalus is one of the common causes of hydrocephalus. It primar-