Operative nuances of endoscopic third ventriculostomy for pediatric obstructive hydrocephalus

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ABSTRACT

Background: Pediatric obstructive hydrocephalus is a relatively prevalent condition that may cause significant social and financial burdens. Current standard management is ventricular shunting with implantation of various ventriculoperitoneal (VP) shunt devices. This procedure is costly and has high complication rate. Endoscopic third ventriculostomy (ETV) is available as an option and considered to be superior to VP shunt.

Methods: All of the pediatric obstructive hydrocephalus cases admitted to Sanglah General Hospital between 2005 until 2016 that were treated by ETV were described.

Results: In 184 cases of ETV procedure in Sanglah General Hospital for pediatric obstructive hydrocephalus we have 92.6% of success rate.

The complications are quite low, one patient (0.5%) died because of bleeding; two patients (1%) suffered wound infection, one person (0.05%) had meningitis and Cerebrospinal Fluid (CSF) leakage on three patients (1.6%), ETV cannot be done in eight patients (4.3%) due to the unidentifiable third ventricle.

Conclusion: ETV is a simple yet effective procedure for pediatric obstructive hydrocephalus. Knowledge about intraventricular anatomic variations is very important in order to provide high success rate and low risk of complication.

INTRODUCTION

The incidence of congenital hydrocephalus is around 2 cases per 1000 live births. There are 812 cases of congenital hydrocephalus in Bali and Nusa Tenggara from 1992-2006 with an average of 4 patients per month. Most of the patients (80%) are categorized as obstructive (non-communicating) hydrocephalus due to the blockade on CSF flow. Without proper treatment, most patients will either die or severely disabled.

Implantation of ventricular shunt systems such as VP shunt and ventriculoatrial shunt is the conventional therapy for hydrocephalus. It has several possible complications such as over or underdrainage, shunt malfunctions, and infections. Complications could not entirely be prevented. Endoscopic Third Ventriculostomy (ETV) showed its superiority compared to ventricular shunting, by avoiding shunt-related complications. It is conducted by creating CSF diversion through basal cistern and subarachnoid spaces thus bypassing the cerebral aqueduct. ETV procedure is cost less and has high success rate and low risk of complication.

This article was based on our personal experiences in treating 184 hydrocephalus patients using ETV.

HISTORY OF VENTRICULOSTOMY

Ventriculoscopy was first introduced by Walter E. Dandy in 1900 for the therapy of hydrocephalus. He is the first person using the first plexectomy in order to perform endoscopy in patients with communicating hydrocephalus. He also developed the basic sub-frontal approach to open the base of the third ventricle. This procedure was accompanied by high mortality and morbidity rate.

In 1923, William Mixter worked on ETV using urethroscope. Tracy J. Putnam modified so that it met requirements as ventriculoscope which could be used to cauterize the choroid plexus in children with hydrocephalus. However; valve regulated shunt system was then discovered and considered simpler. This new technique was deemed a huge success, causing ETV techniques did not progress for the next 30 years.

In 1947, H.F. McNickle introduced percutaneous ETV technique which lowered the number of complications and increase the success rate. The 1970s leukotome was added to widen the perforation at the base of the third ventricle without...
resulting in trauma to the surrounding structures. Development of manufacturing technology and fiber optic lenses allowed the development of small neuroendoscope with arrange-possible edges and excellent optical resolution. As a result, ETV became the first-choice procedure for treating obstructive hydrocephalus with the success rate of 50-94%.19

Applications of currently developed ETV techniques are very broad nowadays. They includes obstructive hydrocephalus due to aqueduct stenosis, hydrocephalus with myelomeningocele, hydrocephalus associated with Dandy-Walker malformation, communicating hydrocephalus, normal pressure hydrocephalus, hydrocephalus secondary from intracranial hematoma, secondary hydrocephalus due to posterior fossa tumors, hydrocephalus after shunt malfunctions, hydrocephalus associated with fasiocraniosonostosis, hydrocephalus in patients with tuberculous meningitis and combination of the Choroid Plexus Cauterization (CPC) with ETV.7,9

Patients candidate
Clinical and radiological investigations are important in influencing the success rate of ETV.4 The investigations will show right and left lateral ventricle widening. The sagittal slices are needed to confirm that there is enough distance between the clivus and basilar artery (AB) from the base of the third ventricle so that the endoscope can enter safely into the third ventricle, allowing it to move safely without risking injury to the lateral wall of the third ventricle.

Age, etiology of hydrocephalus, previous installation of VP shunt and meningitis are the relative contraindication for ETV.14 It is possible to perform ETV on small or slit-like ventricular system caused by the previous installation of a VP shunt. Consideration should take on infants less than six months, the slit-like ventricle form, thin cerebral mantle and communicating hydrocephalus because of the low success rate.19,17

Endoscopic Anatomy of the Ventricles
Beign accustomed to the anatomy of the ventricle system is crucial.2,4,7,8 Foramen of Monro is the most often first seen structure. It is a pair of foramen that connects the lateral ventricle with the third ventricle. The head of the caudate nucleus is located on the lateral side, while septum pellucidum is located on the medial side. If followed on top of choroid plexus, it will lead to the foramen of Monro. Vein of septum pellucidum is positioned at the anteromedial, delivering venous blood to thalamostriate vein located on the posterolateral side of posterior edge foramen of Monro, then to the internal cerebral veins that run on tela choroidea of the third ventricle. A pair of Fornix shaped like the letter C is an efferent-output bundle that connects the hippocampus with mammillary bodies.4

When the endoscope enters the third ventricle, there is a landmark, lateral wall that is consist of the two third of the anterior thalamus and hypothalamus related to the gray matter at the base of the third ventricle. The edges of the left and right lateral third ventricles connect to intermediate mass by a band of gray matter. The posterior edge consists of the pineal body, habenular commissural, posterior commissural and cerebral aqueduct. Cerebral aqueduct is a channel connecting the third and fourth ventricles. The fourth ventricle is approximately 15 mm in length and 1 mm in diameter.

The base of the third ventricle is formed primarily by the hypothalamus. There are some recess and prominence for the landmark: optic recess, chiasma opticum, infundibulum, tuber cinereum, and mammillary bodies. In the majority of cases, the base of the third ventricle (tuber cinereum) is thin and translucent. The location of interpeduncular fossa (pons cistern) is below the base of the third ventricle. CSF streamed from the third ventricle to the interpeduncular fossa with endoscopy guidance.4 Variations in the anatomy of ventricles are found in more than one-third of the cases. Variations of the thickness of the floor and its position are the most common.4 Etus et al. (2016) reported

Figure 1 Anatomic variations and steps during ETV (Maliawan & Wisnu Wardhana, 2017)
intraventricular anatomical variations that includes thick and prominent intermediate mass, narrow tuber cinereum, hollow or thick floor of the third ventricle, small anterior chamber of the third ventricle, existence of interhypothalamic adhesions, vascular floor of the third ventricle, and existence of adhesions or bridges between mammillary bodies.

Instruments
There are various devices for ETV procedure. We emphasize for using one type or brand of the instrument only until the operators are comfortable with those tools. The tools consist of rigid rod lens endoscope with a diameter of 4.2 mm, a disposable endoscope with 13 cm rigid shaft and 2.1 mm working channel, 30 degree optic lens, control panel (contains a video camera, an optical coupler, a xenon light source, video recorder, and printer), 2 French ventricular cannula, Bugbee wire, monopolar cautery, No. 4 Fogarty balloon catheter. Warm ringer used for irrigation during the procedure.

The Procedure
The success of the ETV depends primarily on recognition and understanding of ventricular system anatomy and its variations on hydrocephalus state. After general anesthesia, the patient is placed in the supine position with the head about neutral position laying on the doughnut-shaped pillow. The head is then elevated 30 degrees, and position of the burr hole should be at the highest point to prevent excessive drainage of CSF, anatomical distortion and air influx during the procedure.

A single 6-10 mm burr hole is placed 3 cm from the midline (about the mid pupillary line), 1 cm anterior to the coronal suture (Kocher point) where the normal lateral ventricle, foramen of Monro projected. Dura mater is opened curvilinearly followed by insertions of 14 French peel-away catheters for cannulation of the lateral ventricle. The insertion is directed, on anterior to posterior projection, towards the medial canthus of the ipsilateral eye and, on lateral projection, towards the external acoustic meatus. Endoscopy was directed into lateral ventricle with gentle traction, without causing trauma to the brain. The average distance from dura mater to the foramen of Monro is 6 cm in adults. Some neurosurgeons choose the flexible endoscopy to maximize maneuverability, but we prefer rigid endoscope.

Next step is the identification of foramen of Monro. Subsequently, the endoscope is directed to enter the third ventricle. The base of the third ventricle is approximately 9 cm from the dura mater. The length is highly variable depending on age and severity of hydrocephalus. Next is the identification of mammillary bodies and the infundibulum. Sometimes basilar artery (BA) can be seen clearly through the thin and translucent base of the ventricle.

At this point, the neurosurgeon should perform fenestration using Bugbee wire without or with monopolar electrocaugulation in front of the BA at the base of the third ventricle, between the mammillary bodies and the infundibulum. After that, the Bugbee wire is removed, followed by insertion of no. 4 Fogarty balloon catheter into the stoma. The balloon is inflated very gradually using 0.2-0.4 ml of normal saline to widen the stoma. Balloon dilation will expand the stoma to achieve about 5 mm in diameter. Incorrect balloon placement during inflation can lead to uncontrolled bleeding. The thick Liliequist membrane may prevent the flow of CSF even after fenestration of the base of third ventricle. Ensure there is no obstructing arachnoid membrane. Do not be too aggressive in exploring the prepontine cistern because it could damage the perforating artery or its branches. Identify the BA, stalk hypophysis sometimes visible.

Undulation (flappy movement) from the edges of the stoma indicates adequate CSF flow. There is one case of redo ETV in Sanglah General Hospital. The stoma was intact, but the Liliequist membrane was not opened. After conducting further fenestration, the patient did not need any shunt. Fenestration of the base of ventricles may use the endoscope itself, laser, or monopolar and bipolar cauterization. Once the endoscope removed, foaming gel is used
to cover brain parenchyma corticotomy continued with galea closure using absorbable suture, and the skin with nonabsorbable interrupted suture or skin staples.

If there is significant intraventricular hemorrhage, placement the EVD (external ventricular drainage) maintained for 1-2 days is mandatory. The shunt that was previously implanted is removed, and clinical observation must be performed during 3-5 days. If there are still signs of increased intracranial pressure, consider performing a lumbar puncture. Withdraw approximately 10-20 cc of CSF. It can be repeated two times for two weeks. If there is no clinical improvement, ETV is considered failed, and the patient is classified as shunt-dependent.

**Postoperative care**

The patient should be observed in intensive care unit for at least one day. The patient can usually be discharged on the second or third day after the procedure. Head CT scan performed during early post-ETV period showed that ventricle size does not decrease significantly in the most of the patients, compared with shunting procedure.

When discharged, the patient should be free from the pain of the head without major fontanelle bulging. Patients who had previously undergone VP shunting sometimes takes up to 10 days to adjust to the new CSF absorption system. Sometimes external ventricular drain (EVD) is needed. Ideally after two months, MRI or CT scan may show the narrowing of the ventricular system and the disappearance of periventricular edema. Cine-phase contrast MRI may reveal the patency of the stoma directly, so it could be useful as an indicator of favorable outcome following the third ventriculostomy.

An elastic bandage is arranged on the head so that the shape of the head is not flat. If regularly adjusted, it may help shrinking head circumference significantly. Treatment failure is considered if after two weeks the symptoms of high intracranial pressure persist and after two times lumbar puncture performed.

**Outcome and Predictors**

The success rate of third ventriculostomy procedure is 71-85% on obstructive hydrocephalus cases. Gorayeb et al. (2004) demonstrated lower success rate (64%) in children under one-year-old. Higher success rates already reported for patients with stenosis of the aqueduct. Secondary ETV proved to be a viable option in the management of shunt malfunction with a success rate of 66% which is comparable to primary ETV. Endoscopic third ventriculostomy success score (ETVSS) can be used to predict the outcome of the procedure.

The ETVSS ranged from 0 to 90. The sum of the score roughly shows the success rate of ETV after six months postsurgery. Patients are classified based on high ETVSS (80-90), moderate ETVSS (50-70) and low ETV (score under 40). Analysis of 15 articles regarding ETV showed success rate ranged from 31.3%-92.3%. Incorporation of these data with predicted ETVSS revealed that the result is significant with success score is between 41.3% to 85.4%.

**Possible Complications**

Potential complications of ETV includes leakage of CSF from the location of the incision, injury to fornix, thalamus, hypothalamus or basilar artery (causing Intraventricular bleeding), obstruction of a previous patent stoma, upward brain herniation, and infection. The mortality rate of ETV ranged 1.1-1.8%. Most significant complications of ETV are only transient. The rate of major complications varies around 7% of the cases with only one-sixth of them leading to permanent neurological damages. The third ventriculostomy-related complications usually occurs during the initial 3-6 months after procedure, comparable with lifetime risk of shunt-related complications.

In 184 cases of ETV in Sanglah General Hospital for obstructive hydrocephalus, we experienced very low complications rate (Table 1). ETV procedure cannot be done in eight patients (4.3%) due to unidentifiable third ventricle. Intraventricular bleeding usually occurs in the first 100 cases (related to operator’s experiences and learning curve). Postoperative complications are known to be minimal in experienced hands.
Shunting procedure carries a higher risk of postoperative infection compared with ETV. There are significant lower amount expression of CSF proinflammatory markers (IL-1β, IL-6, and Neural Growth Factor/NGF) on seven days after ETV compared with VP shunt on children with obstructive hydrocephalus.16

CONCLUSION

ETV should be considered as the first option on all obstructive hydrocephalus and most of the nonobstructive hydrocephalus to avoid shunt-related complications. Correct patient selection and precise intraventricular orientation are very important for the preoperative planning and during the procedure. In our experiences, ETV has a small percentage of failures and complications. This procedure can be the first choice treatment for pediatric obstructive hydrocephalus.

REFERENCES