Pediatric Endoscopic Third Ventriculostomy: A Narrative Review of Current Indications, Techniques and Complications

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Abstract

Hydrocephalus is a disorder in which excess cerebrospinal fluid (CSF) collects in the brain. Possible managements for hydrocephalus contain CSF deviation through ventriculoperitoneal shunt assignment and endoscopic third ventriculostomy. An endoscopic third ventriculostomy has been in trend for the past two decades, as a tool in the field of the neurosurgery, for the management of hydrocephalus. Its utility has been confirmed consistently in congenital and acquired aqueductal stenosis, although the outcomes in communicating hydrocephalus and hydrocephalus secondary to other etiologies have not been impressive. It is a relatively harmless technique with the appropriate selection of patients with a low rate of enduring morbidity. Further studies in child should focus on the predictive factors that are specific to the older population. A combination of clinical, radiological, and intraoperative findings may be necessary to plan a clinical prediction rule suitable to this group of patients. The purpose of this review is to describe the current indications, management outcomes, techniques and complications of this method.

Keywords: Pediatric, Endoscopic Third Ventriculostomy, Hydrocephalus

1. Context

Hydrocephalus is a disorder in which excess fluid collects in the brain. The fluid is actually cerebrospinal fluid (CSF); a clear fluid nearby the brain and spinal cord (1). Pathophysiologically, hydrocephalus is observed as an inequity in the formation and absorption of cerebrospinal fluid (CSF) to an adequate magnitude creating accumulation of fluid leading to a promotion of intracranial pressure. Compensatory alterations particularly in very young and very old matters might happen that can decrease usual CSF pressure to a normal range (2).

The frequency of hereditary and childhood hydrocephalus in the advanced nations has been projected to variety from 0.5 to 0.8 per 1000 living deliveries (3, 4). Hydrocephalus characterizes a great health care load in the US, by ventriculoperitoneal (VP) shunting and its related hospital burdens accumulation nearly $2 billion yearly in children 0 - 18 years. The load of hydrocephalus is informed to be upper in developing countries (5, 6). This review purposes to describe the present indications, managing outcomes, technique and complications of endoscopic third ventriculostomy in pediatric patients with hydrocephalus.

2. Evidence Acquisition

We searched PubMed and Ovid databases using the key words, including pediatric, endoscopic third ventriculostomy, and hydrocephalus. Exclusion criteria were as follows: 1) articles published in any language other than English, and 2) patients older than 18 years at the time of surgery. Finally, we reviewed 66 abstracts dating from 1995 to 2015.

3. Results

3.1. Clinical Landscapes

3.1.1. 0 - 2 Years

Before 2 years, the head expands extremely because the cranial sutures are exposed. This enlargement is virtually invariably the presenting sign (7, 8).

1) Shape of Head: An abnormal head form might propose the diagnosis. Occipital prominence is perceived in Dandy walker malformation. An unreasonably large forehead is common by aqueductal stenosis. The distended cranial magnitude is quite evident and helpful in diagnosis.

2) Anterior Fontanelle: Usually the anterior fontanelle is small, depressed in a relaxed sitting patient but with hydrocephalus, it is enlarged and full even once the infant is silent and upright.
3) Percussion Note: The percussion note of a typical infant skull is of a “Crack pot”, though with hydrocephalus it starts similar to a watermelon. This sound is predominantly striking when examiner places his ear contrary to the infant’s skull while percussing.

4) Scalp Veins: The scalp veins are typically protuberant, mainly in crying infants.

5) The Eyes: As the hydrocephalus developments, the eyes are displaced downward by pressure on the thinned orbital roofs. This movement of eyes sources the sclera to be visible above the iris, called as the “setting sun” sign (9).

6) Cranial Nerves: Optic atrophy is a communal finding in progressive hydrocephalus owing to the compression of the optic chiasma and the optic nerve by dilated anterior third ventricle and increased intracranial pressure (ICP). Abducent nerve paresis secondary to stretching is common (9).

7) Growth Retardation: In hydrocephalic children, growth failure and delayed neurological growth are common. Head and trunk switch is principally affected (9).

3.1.2. More Than 2 to 12 Years

Children who have previous (infantile) unrecognized, advanced hydrocephalus by normal or retarded neurological development may be typically diagnosed after an incidental head injury leading to fast deterioration of neurological functions. They usually have somewhat enlarged heads with optic atrophy or papilloedema, abnormal hypothalamic functions and spastic lower limbs (9). Performance intelligence quotient (IQ) is inferior than verbal IQ and learning difficulties are common (9).

3.2. The Classification Being Used Currently Is as Follows

1) Noncommunicating or obstructive hydrocephalus in which flow of CSF from the ventricles to subarachnoid space is blocked. Therefore, there is no communication among the ventricular system and the subarachnoid space. The commonest reason of this category is aqueduct blockage.

2) Communicating or nonobstructive hydrocephalus in which flow is not obstructed, nonetheless CSF is incompetently reabsorbed in the subarachnoid space. So, there is communication among the ventricular system and the subarachnoid space. The commonest cause of this group is postinfective and posthemorrhagic hydrocephalus. This category might also be subclassified into congenital and acquired (10, 11).

3.3. Diagnostic Tools

3.3.1. Ultrasonography

It is a noninvasive technique used only in patients in whom the anterior fontanelle is open.

3.3.2. Computed Tomography

CT scan superseded other invasive surveys like ventriculography and pneumoencephalography. It has a main role in precise valuation of ventricular size, extracerebral spaces and site of obstruction.

3.3.3. Magnetic Resonance Imaging (MRI)

This is also a noninvasive investigation (12). It may also be used in antenatal judgment of hydrocephalus.

3.4. NonSurgical Managing

Drugs such as isosorbide which produce hyperosmotic diuresis and those such as acetazolamide which diminution the secretion of CSF may be treated as temporary organization of clinical situation. Their brief action and side effects prevent their lengthy use in the definitive treatment of hydrocephalus. Wrapping of the infant’s head was started as a treatment of hydrocephalus, the object being to force CSF into alternate absorptive paths (13).

3.5. Surgical Managing

The surgical procedures used in modern times for release of hydrocephalus have been exceptionally varied. Following procedures were tried before the definitive treatment the hydrocephalus evolved (13).

Processes planned to decrease CSF formation: 1) excision of choriod plexus, and 2) cauterization of choriod plexus.

Processes for decompression of ventricles: 1) repeated ventricular puncture, 2) open ventricular drainage, and 3) closed ventricular drainage.

3.5.1. Indications for Shunt Surgery

The indication of surgical intervention are associated to intracranial hypertension, neurological dysfunction, the existence or absence of pathological lesion, evidence and degree of ventricular dilatation, and the nature and the location of obstruction. Some surgeons insert the shunt in any patient with big ventricles whereas others limit it to those with potentially reversible deficit or progressive deterioration. Progressive ventriculomegaly on CT scan joint with observation of developmental discrepancies in infants and intellectual and motor disability in older children are a few criteria (14).

Recent techniques included: 1) ventriculo peritoneal shunting, 2) ventriculo-atrial shunting, and 3) third ventriculostomy.

Ventriculoperitoneal and ventriculoatrial shunts have played a chief part in the effective managing of patients by hydrocephalus. Insertion of a shunt can be associated with
a long-time risk of infection and despite technological advancements; there is a risk of shunt impediment as well as overdrainage that can result in momentous morbidity, demanding recurrent shunt revisions (15-18). The reported rate of shunt malfunction in the first year of placement is 30%, and afterward it is around 10% per year. The accumulative risk of infection is approximately 20% per patient, with most centers recording rates between 5% and 10% (15, 19).

As an alternative to a shunt, an endoscopic third ventriculostomy (ETV) has been in routine neurosurgical training for the past two decades, with long-term consequences being stated in the current writings. To perform an ETV, the third ventricular floor is fenestrated between the mamillary bodies and infundibular recess in the midline, to connect the ventricular system through the basal cisterns (15-17).

An endoscope was first used in neurosurgery in the early 1930s when an ureteroscope was used to congeal the choroid plexus of ventricle. Subsequently, by Dandy’s classification of hydrocephalus into communicating and non-communicating categories and further modifications of it, bypass processes to overcome obstruction of the cerebrospinal fluid (CSF) pathways have been established (19).

3.5.2. Endoscopic Third Ventriculostomy Indications

Defining the paramount candidates for ETV has been hard, by means of incompatible reports on who are the finest candidates, predominantly with respects to the consequence of age and etiology. Information have showed that indications for ETV is a meaning of age, independent of age, a role of etiology, or a function of together age and etiology (16, 20-27).

The developers of third ventriculostomy such as Bergland, Hoffman, Patterson and, Hirsh suggested that the technique must first be kept for patients by obtrusive or Noncommunicating hydrocephalus, in whom the usual mechanism of CSF absorption still be existent at the arachnoid granulations (28-30). Really, the writings presented that ETV is more actual if the hydrocephalus is due to aqueductal stenosis or compression of aqueduct or fourth ventricle by means of tumors. Reasons of hydrocephalus such as intraventricular hemorrhage, subarachnoid hemorrhage, and meningitis have been used by some to dismiss patients from ETV. However, these have newly become relative contraindications in opinion of the decrease of the morbidity and mortality of ETV (30-35).

The endoscopic third ventriculostomy has been tried with respectable victory rates reaching up to > 90% in patients by posterior fossa tumors and brain stem gliomas with hydrocephalus (35-37). The ETV has been of confirmed rate in patients with posterior fossa tumors, who have determined hydrocephalus after tumor resection (36). Mega cisterna magna, Dandy-Walker malformations, retrocerebellar cysts and hydrocephalus secondary to fourth ventricular outlet obstruction have all been treated effectively with ETV, with or without aqueductal stenting (38-40).

Warf ET al. (40) have described the consequences in a large amount of pediatric patients with postinfectious hydrocephalus. The achievement rates in these patients are inferior to in patients with hydrocephalus, owing to aqueductal stenosis of noninfectious causes. In their practice, the victory rates in children elder than one year of age were 81% in patients with postinfectious hydrocephalus and 91% in nonpostinfectious hydrocephalus. The success of ETV in infants through postinfectious hydrocephalus was significantly fewer at 59% (40).

Hydrocephalus related by myelomeningocele could be of the obstructive or communicating type. These patients need treatment in infancy, but the ETV does not offer as respectable outcomes as in older patients, with failure rates of up to 50% being described in patients with myelomeningocele (5, 41). However, in a current series, Warf et al, have confirmed good consequences in up to 78% of infants with myelomeningocele related hydrocephalus treated with ETV and choroid plexus cauterization (39-41). Endoscopic third ventriculostomy remnants a practicable choice in the managing of hydrocephalus in patients with Chiari I malformation and syringomyelia, with shunt freedom being reached in up to 94% of the patients and resolve of the syrinx being detected in a major number of patients (5, 6, 42). In children with Chiari II malformation, the reason of hydrocephalus is improbable to have an obstructive etiology and ETV has been planned to play a very restricted role in this suggestion (6).

Endoscopic third ventriculostomy is an active decision in patients with hydrocephalus who existing with shunt dysfunction due to underdrainage or overdrainage. The total rate of shunt malfunction in patients shunted through infancy is informed to be 48%, 52%, and 63% at one, two, and five years, with 20% of the patients having more than three revisions in five years (43). The ETV in patients with shunt dysfunction was efficacious in around 80% of the patients (16, 33, 39). Patients with aqueductal stenosis had the best consequence; however, postinfectious and myelomeningocele patients had a deprived result.

Infants presenting by hydrocephalus are not suitable applicants for ETV as they have not well-established absorptive surfaces in the subarachnoid spaces and have an open anterior fontanelle, with a lax skull (6, 44). Yet, the ETV has been tried by numerous writers in infants with hydrocephalus, due to variable etiologies. Og iwara et al. (45-48) reported a global success rate of 34.8% in infants fewer than six months of age. Even as success rates have been acceptable in lonely aqueductal stenosis, there is a high
rate of failure with other etiologies as well as in preterm infants.

3.5.3. Surgical Anatomy and Technique

Awareness of the ventricular anatomy is vital in performing an ETV. A burr hole is prepared on a point at the frontal area that will plan a straightforward line connecting the foramen of Monro and the tuber cinereum, the portion of the floor of the third ventricle to be punctured. Stereotactic navigation is frequently working to choose the entry point and define the route of the endoscope (47, 48). The foramen of Monro is usually the first structure visualized after entry of the endoscope into the frontal horn of the lateral ventricle. The typical interpretation of the foramen of Monro is showed in Figure 1 (49).

The choroid plexus of the lateral ventricle schemes frontward to the foramen, and the septal vein is situated anteromedially beside the septum pellucidum. It joints the thalamostriate vein, positioned posterolaterally, at the posterior rim of the foramen of Monro to arrange the internal cerebral vein. The scope is formerly forward-thinking through the foramen of Monro. The endoscopic vision of the floor of the third ventricle has shown in Figure 2 (49).

The paired mammillary bodies are visible on the posterior boundary of the floor of the third ventricle (47, 48). The goal of fenestration is the tuber cinereum, the zone among the mammillary bodies and the infundibulum of the pituitary gland. It is important to remind that the basal artery and the oculomotor nerves are located right below the tuber cinereum. A probe is then approved through the working channel of the endoscope. When the tip of the probe seems in view of the endoscope, it is directed to the tuber cinereum wherever a fenestration is complete. This has to be ended judiciously in order to escape bleeding from the basal artery or injury to the oculomotor nerve. The hole is then inflamed by a Fogarty balloon. (Figure 2) presented the fenestration being complete by the probe. When hemostasis is attained, the endoscope is withdrawn mildly (47, 48, 50).

3.6. Complications of Endoscopic Third Ventriculostomy

In an assessment of complications subsequent ETV in a large series of patients, long-lasting morbidity was 2.38% and permanent neurological problems happened in 1.44%. The global complication rate was 8.5%. The additional complications connected to ETV involved intraoperative hemorrhage from the, choroid plexus ependymal veins or basilar artery (3.7%), and enduring precocious puberty, weight gain, and diabetes insipidus. Quick postoperative mortality owing to sepsis and hemorrhage was 0.21% (27). Complication rates of about 8% to 9% were informed in additional series (47, 48, 50). Intraoperative complications comprised intraventricular hemorrhage, cardiovascular fluctuations, such as, bradycardia during fenestration and inflation of the balloon of the Fogarty catheter, and damage to the hypothalamus and fornix (27). Inside the 30 days subsequent ETV, the complications stated included a CSF leak, subdural fluid collection and ventriculitis (27, 51, 52). The risk of complication was higher with recurrence ETV processes and in patients by previous shunts (53, 54).

Misplacement of the fenestration in the third ventricular floor usually accounts for most of the intraoperative hemorrhagic and cranial complications. This can be avoided by recognizing the mammillary bodies, foramen of Monro, dorsum sellae, infundibular recess and staying in the midline (2, 52-54).

Bradycardia has been described in up to 41% of the cases throughout fenestration of the third ventricular floor and the mechanisms hypothesized comprise motivation of the preoptic area, at which time there might be related hypotension. Stimulus of the posterior hypothalamus sources tachycardia with hypertension (52, 55, 56). Use of normal saline as a substitute of Ringer's Lactate for irrigation can decrease the risk of hyperkalemia-induced bradycardia.

Subdural hygromas as well as chronic subdural hematomas have been informed subsequent ETV, attributable to the sudden extreme discharge of CSF or a big cortical puncture letting way out of CSF into the subdural space (57, 58).
4. Conclusions

Amongst some common disorders like iron deficiency anemia and febrile seizure (39) and some rare disorder like lumbar discopathy and traumatic spine disorders (60,61), hydrocephalus still remains a troublesome problem in child. Endoscopic third ventriculostomy has been found to be a real alternative approach to shunt placement in obstructive hydrocephalus secondary to aqueductal stenosis, with good long-time results. Use of ETV in other situations should be sensible, with an appropriate review of the patient’s ventricular anatomy and etiology of the hydrocephalus. Eventually, the management of hydrocephalus needs to be modified with both the options of ETV and shunt being appropriate in individual situations. Further studies in child should focus on the predictive factors that are specific to the older population. A combination of clinical, radiological, and intraoperative findings may be necessary to plan a clinical prediction rule suitable to this group of patients.

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