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Original Research Article

Endoscopic third ventriculostomy for hydrocephalus: a study of thirty cases

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ABSTRACT

Background: Raised intracranial pressure because of abnormal accumulation of CSF with in the ventricles of brain is a potentially life-threatening condition. Ventriculoperitoneal shunt remains mainstay of treatment even today. However, shunt failure is not uncommon. Hence endoscopic procedures are now gaining popularity. The outcome of ETV with respect to age ,etiology and long-term outcome needs further elucidation.
Methods: Thirty patients with hydrocephalus who underwent endoscopic treatment were analysed retrospectively with respect to age, diagnosis, surgical outcome, treatment failure and complications.
Results: Total of 28 patients successfully underwent ETV and with an overall success rate of 78%. Moreover, the success rate of ETV was found to be 100% in the 0-1 and 80% in 1-2 years age groups. 2 patients required re operation in the form of VP shunt.
Conclusions: ETV is a safe and effective surgical modality with comparable outcome in paediatric and adult populations.

Keywords: CSF diversion, ETV, Fenestration, Hydrocephalus

INTRODUCTION

Raised intracranial pressure because of abnormal accumulation of CSF within the ventricles of brain is a potentially life-threatening condition and commonly seen in neurosurgical patients. It is classically described as communicating or non-communicating hydrocephalus, based on the presence or absence of CSF outflow obstruction.[1]

Ventriculoperitoneal shunt is easily available, in-expensive and remains mainstay of treatment even today. However, shunt failure is not uncommon and revisions may not give outcomes. Hence endoscopic procedures are now gaining, popularity as an alternative to VP shunt.[2] These include endoscopic third ventriculostomy, endoscopic aqueductoplasty, and endoscopic aqueductal stenting.

Clinical and radiological criteria are used for defining success of ETV. Clinical criteria include resolution of the signs of raised ICP i.e., improvement in the level of consciousness, resolution of occular movement abnormalities, resolution of headaches, reduction in the head circumference and decrease in the fontanelle tension (in infants). Radiological criteria includes reduction in size of the third ventricle (i.e., by 25%) on three months follow up and A 15% reduction in size of the third ventricle within 1 month is considered as a reliable indicator of favourable outcome.[3,4] Usually the extent of ventricular size reduction postoperatively is inversely proportional to the duration and magnitude of symptoms preoperatively conversely reduction in ventricular size

post ETV may or may not indicate a successful procedure as it has been found to reduce marginally in both settings.[5]

The aim and objective of the present endeavor was to study and compare clinical outcome of patients undergoing endoscopic third ventriculostomy for hydrocephalus.

METHODS

Authors present our institutional experience of endoscopic management of hydrocephalus through the present study. All patients were followed up for a minimum period of 6 months to study the efficacy, complications of the procedures

Inclusion criteria

- All patients diagnosed to have a hydrocephalus were included.

Exclusion criteria

- Patients in active stage of meningitis and or ventriculitis.
- Patients with intraventricular hemorrhage
- Patients not consenting to undergo endoscopic treatment.

A total of 30 patients of hydrocephalus were managed between January 2017 and August 2018. All thirty patients who underwent endoscopic treatment for hydrocephalus were analysed. Of these twenty eight patients underwent endoscopic third ventriculostomy. One patient with complex multiseptate hydrocephalus underwent fenestration with shunting. The third ventricular floor anatomy was unfavourable in this patient and hence ETV was not attempted. In another patient who was previously shunted and had presented with shunt dysfunction, with an omaya insitu, ETV was attempted but the procedure was abandoned.

The patients were assessed both clinically and radiologically to ascertain the effect of the endoscopic treatment. The resolution of clinical signs and symptoms were considered for determination of success/ failure of the treatment offered.

RESULTS

This study includes 30 patients. Six months follow up data of 29 patients were available and 1 patient is lost to follow up. Three patients died during Follow up. ETV was performed in 28 patients, which included Five previously shunted patients who presented with shunt dysfunction. ETV was not attempted in view of complex anatomy in one patient. While ETV was attempted but abandoned due to distorted anatomy in 1 patient. Favourable results were obtained in 23 patients. Overall results were unfavorable in 5 patients. 2 failed ETV required insertion of shunt for CSF diversion and remaining 3 died.

The study participants were grouped into 5 groups. Infants made only 16.6% of the study group while 63.3% patients were younger than 30 years. The youngest patient was 4 months age and the oldest patient in this study was 55 years old. The male female ratio was 1:1 in this study comprising 30 consecutive patients treated by Endoscopic procedure for various etiology (Table 1).

Table 1: Age-wise distribution of study participants.

Age (in years)	Number of patients	Percentage (%)
Less than 1	5	16.66%
1-15	14	46.66%
16-30	5	16.66%
31-45	3	10%
46–60	3	10%
Total	30	100%

Table 2: Distribution of study participants based on diagnosis.

Etiology of hydrocephalus	No. of patients	Percentage (%)
Aqueductal stenosis	8	26.66
Meningitis	6	20
Congenital hydrocephalus with MMC	1	3.33
Dandy Walker malformation	2	6.66
Space occupying lesion	13	43.33
Lateral ventricular cyst	1	
Third ventricular cyst	1	
Pineal gland tumour	2	
Posterior fossa tumour	9	
a. Cerebello pontine angle Tumours	4	
b. Meningioma	1	
c. Ependymoma	2	
d. Medulloblastoma	2	
Total	30	100

Of the 30 consecutive patients admitted over the period of 1 year, 43% patients were diagnosed with obstructive hydrocephalus due to a space occupying lesion. Other than tumours Aqueductal stenosis was the single most common cause 26% (Table 2).

ETV + Fenestration was done for 4 patients, a 6-month child with lateral ventricle cyst, two patients of age 1 year with dandy walker cyst and complex hydrocephalus each, and a 50 year old patient with a third ventricular cyst. All four patients showed resolution of presenting complaints. ETV with EVD was done in a 40 year old patient with a

CP angle tumour and hydrocephalus in view of intraoperative bleed. ETV with EVD with Biopsy was done for patients with posterior third ventricular Tumours. The biopsy and the ETV were done through a single burr. Fenestration with shunting was done for 6-month-old patient with a complex hydrocephalus. In an 8-year-old patient who had developed meningitis and ventriculitis due to shunt infection was initially managed was taken up for ETV after resolution of infection for persistent symptoms of raised intracranial pressure (Table 3).

Table 3: Distribution of study participants based on procedure.

Procedure	No. of patients	Percentage (%)
ETV	21	70%
ETV+fenestration	4	13.33%
ETV+ EVD	1	3.33%
ETV+EVD+biopsy	2	6.66%
Fenestration+VP shunt	1	3.33%
Septostomy	1	3.33%
Total	30	100%

ETV = Endoscopic third Ventriculostomy, EVD = external ventricular drain.

However, ETV could not be done due to the post ventriculitis septations and thick third ventricular floor. Hence a septostomy with Revision of the VP shunt was done.

The only intraoperative complication was intraventricular hemorrhage witnessed in 3 out of 30 patients. There were no complications while introduction of the ports into the ventricle. In two cases the hemorrhage occurred while taking biopsy from the tumour and in only one case we witnessed a hemorrhage with creation of the stoma, which was easily manageable by irrigation and tamponade, an external ventricular drain was inserted in all these patients.

Twenty two patients responded well and remained symptom free over the first 6 months follow period. In our study ETV was done in 8 patients with aqueductal stenosis. All improved and remained symptom free during the follow period except one patient who was lost to follow up after 7th postoperative day. In 13 patients with hydrocephalus due to obstruction by SOL, 2 patients developed symptoms of raised ICT within 3 months after initial improvement following ETV. In both these patients a VP shunt was done (Table 4).

In patients less than 1 year success was 100%. The procedure was successful in 1 patient with aqueductal stenosis, 1 patient with lateral ventricle cyst, 1 patient with meningitis and 1 patient with MMC. One patient was lost to follow up. The success of ETV in children between the age group of 1-2 years was also 80%. 2 patients with aqueductal stenosis, 1 patient with dandy walker malformation and 1 with meningitis had good outcome. Adverse outcome was met in patient with tubercular meningitis who died on the same day after surgery (Table 5).

Table 4: Association between procedure and outcome.

Procedure	No. of patients	Re solution after ETV	Re-ope rate	Death	LAMA
ETV	21	18	1	1	1
ETV+ fenestration	4	4	0	0	0
ETV+ EVD	1	0	0	1	0
ETV+ED+ biopsy	2	0	1	1	0
Total	28	22	2	3	1
Percentage (%)	100	78	7.14	10.7	3.5

Note :1) One of the patients had complicated hydrocephalus in whom fenestration of multiple cyst was done followed by shunt placement, ETV was not attempted due to complex distorted anatomy; 2) In one patient ETV was attempted but could not be done due to unfavourable anatomy. In this patient septostomy with omaya placement was done.

Table 5: Association between age and outcome.

Age	Total	Resolution	Death	LAMA
Less than 1 year	5	4	0	1
1-2 years	5	4	1	0

DISCUSSION

Endoscopic third ventriculostomy is the well-established treatment of hydrocephalus and is used routinely.

Success rate of ETV and age

It is believed that the clinical response of adult patients to ventriculostomy differs from the pediatric population, because of the duration of hydrocephalus, the differences in CSF circulatory dynamics and reduced elasticity of the brain.[3]

There are controversies regarding the success of ETV in infants. There is difference of opinion regarding, whether ETV should be done in patients younger than 6 months age2. poor results are reported in neonates and in infants, younger than 2 months. Shim KW et al suggested that simultaneous endoscopic third ventriculostomy and ventriculo peritoneal shunt should be performed in infantile hydrocephalus due to poor results of ETV alone.[6,7]

Kulkarni AV et al, reported the relative higher risk of initial failure in ETV, than shunt in children.[8] The relative

risk becomes progressively lower for ETV after about 3 months. Patient could experience a long-term treatment survival advantage after an early high-risk period of ETV failure as compared to shunt. They observed that it might take several years, however, to realize this benefit.

The success rate of ETV in 21 patients less than two years of age was analyzed by Baldauf et al.[9] It was found in this study that the success rate of ETV in children less than two years of age suffering from obstructive hydrocephalus depends on age and etiology with an overall success rate of 43%. In infants, ETV was successful in 37.5% of cases. On analyzing ETV done for 41 hydrocephalus patients younger than two years, Sufianov et al.[10] observed that ETV was successful in 71.4% of children between one to two years and in 75.0% of children less than one year. He et al, have reported sixteen successful ETV procedures done out of seventeen attempted cases of infantile hydrocephalus of varied etiology.[11] A retrospective analysis by Jernigan et al, of 5,416 infants with hydrocephalus who underwent CSF diversion either in the form of shunting or ETV observed a failure rate of 64% after ETV, higher than the 40% failure rate seen post shunting.[3] This rate was even more pronounced if ETV was done within 3 months of birth.

The radiological criteria are not reliable and cannot be used as a sole entity in determining successful outcome of ETV thus clinical correlation is more important.[3]

In present study, authors have used only clinical criteria as indicator of success. Although radiological signs were included in the study, the success was not determined by using them. In our study 5 patients of hydrocephalus were less than 1 year old amongst 1 had aqueductal stenosis, 1 had lateral ventricle cyst, 1 had postmeningitis HCP and 1 patient with HCP with MMC. Overall success rate was 100% with the exception of one patient who was lost to follow up after one week.

The success of ETV in children between the age group of 1-2 years was also 80%. 2 patients with aqueductal stenosis, 1 patient with dandy walker malformation and 1 with meningitis had good outcome. Adverse outcome was met in patient with tubercular meningitis who died on the same day after surgery.

Outcome of ETV in relation to the underlying etiology

Results of ETV are better in congenital hydrocephalus due to aqueductal stenosis as compared to post hemorrhagic or post infective hydrocephalus.

Teo, based on a review of different series of patients treated with ETV, assumes effectiveness of the treatment in 50% to 90% in selected cases of aqueductal stenosis in adults.[12] According to him, patients with primary or secondary aqueductal stenosis, with third ventricle hydrocephalus, with an oval-shape third ventricle, over 2 years of age, previously shunted, who have never been a subject to radiotherapy, with a thin translucent floor of the third ventricle have a good prognosis. Patients who do not meet these criteria become shunt-dependent in 50% of cases Gangemi et al, evaluated the effectiveness of ventriculostomy in treatment of obstructive hydrocephalus twice, based on multicenter studies in Italy.[13,14] In this study involving 140 patients of mixed age, under observation for 6-12 years, a good outcome was reported in 87.1% of cases regardless of patients' age and etiology of hydrocephalus.

In present study ETV has been shown to have 100% success rate in patients of aqueductal stenosis in the short duration of follow up for 6 months.

The patients of aqueductal stenosis included in the study were between the age group of 4 months - 16 years. In most of the patients the width of the foramen Monroe was adequate, the floor of the third ventricle was thinned out and pulsating with the arterial pulsations. Visualization of the perforating branches of the basilar artery along with opening up of the second membrane was satisfactory. In one of the patients, there was difficulty in reaching to the floor of third ventricle due to large head and inadequate length of the scope assembly and the floor was thick and nontranslucent hence the stoma created could not be adequately dilated and successful opening of the lilenquist membrane was not visualized but CSF flow was established through the stoma. This patient is doing well on 6 months follow up.

In our experience secondary HCP due to meningitis (pyogenic or tubercular) are technically difficult for endoscopic third ventriculostomy with higher chances of failure, especially in acute stage of disease due to an inflamed, thick and opaque third ventricle floor.[15] Due to poor results in an acute phase of tubercular meningitis, shunt insertion could be advised to start with, and the ETV is performed when shunt malfunction occurs. Clinical and radiological recovery could be very slow, especially in poor grade.[16] ETV helps to divert the CSF to areas which were inaccessible and clears exudates from the areas which had impaired absorption, thus helping to improve drug delivery. ETV has been shown to have a success rate between 60-85% in most series published.[15]

In post meningitis hydrocephalus, 75% success rate has been observed in our study. Out of 6 patients between the age group of 6months to 42 years, ETV was abondened due to difficult anatomy in one patient. In this patient septostomy with omaya placement was done, while ETV was not attempted in another patient in view of complex anatomy who underwent endoscopic fenestration of multiple cyst.

On 4 out of 6 patients ETV was performed, 3 showed overall clinical improvement after primary ETV. There was 1 death in a patient of post tubercular meningitis due to sepsis. This patient was previously shunted twice.

ETV can be used in the management of hydrocephalus associated with myelomeningocele. Narrow antero-posterior length of the third ventricular floor and narrow prepontine cistern are not infrequently observed in such cases, which may pose problems during ETV. Pre-operative evaluation and intra-operative inspection of these findings are very important in successful performance of ETV.[17]

ETV in spina bifida patients more than 6 months of age after shunt failure has been shown to have a good long term success (approximately 80%).[18] ETV in spina bifida has shown good short term success (100%) in our study.

ETV in dandy walker malformation can be effective means to achieve reduction in hydrocephalus and is recommended line of management.[19] Endoscopic third ventriculostomy alone, ETV with aqueductal stent placement and ETV with fenestration of the occluding membrane could be performed effectively and safely depending on existing pathology in DWS.[19]

In studied group the success rate was 100%. In one out of 2 patients, who were diagnosed with dandy walker malformation, a large periventricular cyst was present which was fenestrated into the lateral ventricle.

The long term success in HCP due to Tumour is usually difficult to assess as in most cases ETV is done for palliation. Endoscopic interventions help in diagnosis by biopsy as well as CSGF diversion for relief of raised ICP. Tissue diagnosis could be obtained in 83%-100% cases.[20] Endoscopic biopsy and ETV through a single or dual port was found to be safe and effective, and it avoids second surgery. Shono et al, found it to be safe and claimed that the risk of tumor dissemination due to the neuro-endoscopic procedures was minimal when the appropriate chemotherapy and radiotherapy were provided postoperatively.[21]

ETV should be considered as an alternative procedure to VP shunt in controlling severe hydrocephalus, related to posterior fossa tumors while patients await their definite tumor excision.[22,23]

Present study included 13 patients of hydrocephalus secondary to a space occupying lesion, of whom 2 were diagnosed of having an arachnoid cyst, 2 of pineal gland tumour, and 9 from posterior fossa tumour. The age group included was between 6 months to 55 years. ETV had a success rate of 80%. There were 2 mortalities in this subset of patients.

For benign lateral and third ventricle arachnoid cyst, ETV+cyst fenestration was done. Both patients had a good outcome.

Two patients of pineal gland tumour underwent ETV+ EVD+biopsy. Intraoperative hemorrhage was present in both. In 1 of the 2 cases did not show clinical or radiological evidence of improvement and was shunted while the other patient died 10 days after surgery despite a functional EVD.

In 9 patients with tumour of posterior fossa, ventriculo-stomy was the temporary alternative. Improvement of clinical condition was observed in 8 (88%) patients. Among these 8 patients, ETV was used as a primary procedure in 7 (77%) patients and as a secondary procedure in 1 patient for shunt malfunction. One death was reported in this group.

In present study involving 30 patients, between the age group of 2 months to 55 years, observed for a period of 6 months, clinical improvement was reported in 83% (25) of the cases, radiological improvement in (22) 73% and (6) 20% showed no radiological improvement while in (2) 7% radiological imaging could not be done. 6% (3 of 25) of the patients who were treated successfully, demonstrated no change in ventricular diameter. In contrast among the patients who responded unsuccessfully to primary procedure (5 of 30) and ultimately required secondary shunt insertion or died, none demonstrated radiological improvement. 78% success was observed when ETV was used as a primary procedure in hydrocephalus of various etiologies with mixed age group.

In patients with a distorted and unfavourable ventricular floor anatomy, septostomy with fenestration of septae with VP shunt is best palliation for hydrocephalus.

In present study, there were 5 previously shunted patients who presented with shunt malfunction in mixed age group. 4 (80%) patients underwent ETV, 3 (60%) had good outcome thus 60% success rate was seen for secondary ETV for a single procedure. Authors have used ETV as compliment to previous shunt. Shunt was left in situ. ETV was done as an additional procedure.

Varying reports in the literature cite an ETV success rate of approximately 65-70% in cases of shunt malfunction.[24] Woodworth et al, see ETV as a good complement to shunt treatment in cases of complications and necessary revision.[25] When performing a secondary procedure, some studies recommend removal or blockage of the malfunctioning shunt. This is shown to increase the longevity of the procedure, Endoscopic procedures like septostomy, fenestration have also shown good results.

Complications

Authors encountered bleeding as the only procedural complications in 3 cases (10%). One patient developed subdural hygroma immiediately post procedure which resolved spontaneously. There were no wound complications, meningitis.

Over all complication rate after ETV is about 2% - 15%, but the permanent complications are few and include

fever, bleeding, hemiparesis, gaze palsy, memory disorders, altered concousness, diabetes insipedus, weight gain and precocious puberty. Intraoperative neural injury, such as thalamic, forniceal, hypothalamic and midbrain injuries are observed. Intraoperative bradycardia and hemorrhages including fatal hemorrhage due to basilar artery rupture are also reported. Attempts to perforate the ventricular floor can lead to bleeding, especially in hydrocephalus following an infection and hemorrhage.

In 1 patient authors could not decisively visualize the stoma adequacy in view of the large head of the patient. The scope assembly could not be progressed to the stoma site. Possibly this issue can be circumvented by extra length scope and sheath assembly.

The patents with pineal gland space occupying lesion underwent ETV followed by biopsy. In both these cases bleeding was from the biopsy site and was significantly brisk and was controlled by bipolar cauterization and irrigation. Although the bleeding was controlled both these patients had unfavorable outcome, one required a VP shunt while the other patient whose general condition was poor preoperatively and post operatively died on postoperative day 7, despite a functional EVD.

Post procedure complications

Early complications reported in literature namely Central nervous system infections, fever, CSF leak and post-operative intracranial hematomas, Diabetes insipidus, weight gain, precocious puberty and abnormal prolactin levels were not seen in our study. Two patients developed seizures while Two patients required VP shunt for failure of ETV. As our study has a short follow up of 6 months, we cannot comment of delayed complications.

In present study, mortality rate was 10% (3 of 30). One patient of ETV with biopsy with post third ventricle space occupying lesion was complicated by intraoperative bleed from the biopsy site and stoma site. His general condition was poor preoperatively and postoperatively, died on postoperative day 7 despite a functional EVD.

One patient of ETV with CP angle space occupying lesion was complicated by bleeding from ventriculostomy site. The bleeding had abated on table. Patient had functional EVD. This patient withstood the procedure well and was awake alert till postoperative day 5 when she went into status eplepticus and could not be revieved despite all measures.

One patient with post traumatic HCP who had undergone a VP shunt followed by shunt revision. He subsequently developed ventriculitis and meningitis and was controlled by exteriorisation of the shunt and antibiotics. He was taken up for ETV and had uneventful EVD but developed high grade fever followed by tonic posturing on postoperative day 0.

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