CLINICAL ARTICLE

Use of Endoscopic Third Ventriculostomy (ETV) Success Score to Predict the Outcome of Endoscopic Third Ventriculostomy in Obstructive Hydrocephalus

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ABSTRACT

Object: *In February 2011 Abhaya V. Kulkarni et al. reported endoscopic third ventriculostomy success score in order to predict the outcome of obstructive hydrocephalus. After ETV the object of the present study was to evaluate this predictive value in our own setup, prospectively.*

Materials and Methods: *From April 2011-November 2012, 110 endoscopic third ventriculostomy procedures were performed for obstructive hydrocephalus of different etiologies at the department of pediatric neurosurgery, the Children's Hospital Lahore. All of these cases were more than 6 months old and the data was analyzed by the senior author.*

Results: *A total number of 110 patients were operated between April 2011 – November 2012. The patients' ages ranged from 6 months – 13 years. No child below 6 months was included in this study. Children were stratified into 4 age groups: 6 months – 1 year (Group – 1), 1 year – 2 years (Group – 2), 2 years – 10 years (Group – 3) and more than 10 years (Group – 4). The score was calculated for each patient before surgery according to ETVSS and at the end of 6months the success or failure of the ETV was determined by clinical, radiological measures. Out of 110 patients, only 80 were available at the completion of 6months period after surgery. The ETV was successful in 56 patients (70%). Patients below 1 year achieved lowest success. Of the ten patients with a high probability of ETV success, eight (80%) were successfully treated.*

Conclusion: *The results show that EVT success can be predicted very well by ETVSS and it should help in establishing surgical selection criteria in order to obtain high success rate. It can also help in preparing the patients and their families to the expected outcome.*

Key Words: *Paediatric Neurosurgery, ETV, ETVSS, Obstructive Hydrocephalus.*

Abbreviations used in this paper: *ETV = endoscopic third ventriculostomy; ETVSS = endoscopic third ventriculostomy scoring system; CSF = cerebrospinal fluid; CPC = choroid plexus cauterization, MMC = Myelomeningocele, IVH = Intraventricular Haemorrhage.*

INTRODUCTION

In the early days of neuroendocoscopy, it was thought that an ideal case for Endoscopic Third Ventriculostomy is a child more than 6 months old, with moderate degree of obstructive hydrocephalus of congenital origin. However, the paediatric neurosurgeons throughout the globe continued to operate upon the children of any age with variable etiology and getting widely variable results. In June 2010, Kulkarni AV et al. developed and internally validated the ETV Success Score (ETVSS).[1] The predictive range is 0 – 90. This is simplified means of predicting the 6 – month success rate of ETV for a child with hydrocephalus, based on age, etiology of the hydrocephalus and presence of a previous shunt. Presently, endoscopic third ventriculostomy has become an established treatment for obstructive hydrocephalus. However, the controversy exists over which persons are appropriate candidates for the procedure because selection of candidates for ETV affects its success. Adequate management of hydrocephalus is important and should aim at creating the best circumstances for cerebral development. Obstructive Hydrocephalus with an apparently normal CSF resorptive system is a classical indication of third ventriculostomy.[6] The apparent advantage of ETV over CSF shunting is the more definitive and more physiological solution to excessive CSF in the brain. In all ETVSS strata, the risk of ETV failure becomes progressively lower, compared with the risk of shunt failure, with increasing time from the surgery. In the best ETV candidates (those with ETVSS of $\geq$ 80), however, the risk of ETV failure is lower than that of shunt insertion right after surgery.[5]

ETV is considered conceptually preferable to VP shunt placement due to the avoidance of lifelong dependency and associated complications such as infections and dysfunctions.[9]

MATERIAL AND METHODS

From April 2011 – November 2012, 110 endoscopic third ventriculostomy procedures were performed for obstructive hydrocephalus of different etiologies at the department of pediatric neurosurgery, the Children's Hospital Lahore. All of these cases were more than 6 months old and the data was analyzed by the senior author.

RESULTS

Out of the 110 patients who were operated, 30 were not available at 6 months follow-up because five patients were those in whom ETV was done and later were operated for posterior fossa tumors and could not survive in the follow-up period. One case of Dandy Walker syndrome and twelve patients with myelomaningocele and co-morbid factors also succumbed. Rest of the 18 patients did not turn up for follow-up and only 80 patients were available for 6 months follow-up. Keeping in view the previous experience of poor outcome of ETV in children less than 6 months of age , such patients were not included in the study. The patients who showed clinical improvement and did not require any other CSF drainage procedure in next 6 months were declared cured by ETV. In order to keep the intracranial CSF production low, we routinely used Acetazolamide for 6 weeks (post-operatively) in all these patients. Some of these patients also underwent ventricular tapping according to the need. These patients were followed up over next 6months clinically, by serial cranial ultrasound, CT Scan and at times MRI. The early post-operative MRI changes in successful ETV, we looked for, were straightening of lamina terminalis and disappearance of sagging of floor of 3rd ventricle. ETV failure was defined as treatment failure requiring any subsequent definitive CSF diversion procedure like shunt or ETV and death related to hydrocephalus management. For all these endoscopic third ventriculostomy procedures, KARL Storz Rigid Neuroendoscope was used along with Fogarty Catheter no.[5] The data of available 80 patients was analyzed.

The etiology of the obstructive hydrocephalus has a profound effect on the outcome of ETV. The etiology pattern in our study was as follows:

The presence of a shunt in a patient undergoing

Table 1: *Age Groups of 80 Patients who Underwent 6 months follow-up after Endoscopic Third Ventriculostomy.*

Age	Number
Group – 1 (6 months – 1 year)	37
Group – 2 (1 year – 2 years)	9
Group – 3 (2 years – 10 years)	24
Group – 4 (More than 10 years)	10

Table 2: *Etiology of Obstructive Hydrecaphalus.*

Etiology of Obstructive Hydrocephalus	Number
Post-Infectious	5
Myelomeningocele MMC, IVH, Non-Tectal Brain Tumor	29 + 5 + 7
Aqueductal stenosis, Tectal Tumor, Others	26 + 3 + 5

IVH: Intraventricular haemorrhage

The presence of a shunt in a patient undergoing ETV has a negative impact on the outcome and preoperative status of our patients was as follows:

Table 3: *Shunt Surgery (Previously).*

Previous Shunt	No previous shunt
13	67

The predictive range of ETVSS is between 0 – 90.

For example, a 6 year old child with tectal brain tumor without previous shunt will have an ETVSS of 40 + 30 + 10: 80 which means roughly 80% chance of having successful outcome of ETV.

In Group – 1, ages range from 6 – 12 months and mean age was 8.8 months. The average score on ETVSS was 59.18 and actual success was 56.75%.

While in Group-2, ages ranged from 12 months – 2 years and mean age was 14.3months. The predicted score in this group was 66.4 while the actual success was 66.6%.

In Group – 3, ages range from 2 – 10 years and mean age was 6.8 years. The predicted score for this group was 70.4 and the actual score was 76.19%.

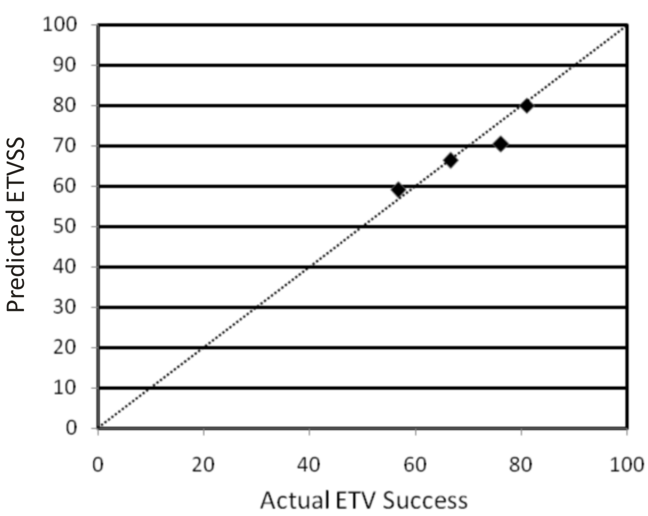
Table 4: *Calculation of ETVSS (Kulkarni AV et al, 2010).*

Score	Age	Etiology	Prior Shunt
0	< 1 month	Post-infectious	Previous shunt
10	1 month or < 6 months		No previous shunt
20		Myelomeningocele, IVH, non-tectal brain tumor	
30	6 months to < 1 year	Aqueductal stenosis	
40	1 year to < 10 years		
50	≥ 10 years		

IVH: Intraventricular haemorrhage   Predictive Range of ETVSS 0 – 90

Table 5: *Group Wise Success of ETV.*

Group	Age Range	Mean Age	Predicted Score	Actual Success Score
Group 1	6 – 12 months	8.8 months	58.18%	56.75%
Group 2	1 – 2 years	14.3 months	66.4%	66.6%
Group 3	2 – 10 years	6 – 8 years	70.4%	76.19%
Group 4	10 – 13 years	12 years	81%	80%



Graph 1: *Actual ETV Success.*

While in Group – 4, ages range from 10years-13years and mean age was 12 years. The predicted value of ETVSS for this group was 81 while actual ETV success was 80% (Table 5 and Graph 1).

There was no death directly attributable to the ETV procedure and in all those cases where the ETV failed, shunt procedures were done. None of these ETV failure cases were subjected to another ETV procedure though there are reports of success of re-ETV procedure from 50 – 90%.

The dashed line predicts the theoretical match between predicted ETVSS and actual ETVSS in four groups.

The complications were managed successfully and there was no procedure related death.

Table 6: *Complications in 80 Patients Undergoing ETV.*

Complication	No. of Patients
Infection	2
CSF leak	3
Per-operative Hemorrhage	1
Cranial Nerve Injury	0
Death	0

DISCUSSION

Reported ETV success varies among studies from 30 – 90%.[2] This variance in success is less likely a result of surgeon ability or operative technique than a function of patient risk factor.[9] Kulkarni et al. analyzed the results from 15 published ETV studies and each study reported different level of success. The in-depth analysis demonstrated that the ETVSS model accounted for the variance among these patients. In our own series of endoscopic third ventriculostomies since 1999, more than 1200 cases of obstructive hydrocephalus have been operated. There is an impression that patients with gross hydrocephalus and post-tuberculous hydrocephalus have very poor outcome maybe because of the involvement of arachnoid granulations. Further studies are needed to determine the effect of gross hydrocephalus and especially post-tuberculous hydrocephalus on the predicted value of ETVSS. Although the role of ETV appears to be expanding, but the challenge to better define the outcome of ETV in comparison with standard shunting, remains there[1]. It has been shown by Abhaya V. Kulkarni that ETVSS predicts not just short-term ETV outcome but also long-term ETV outcome.[1] He has shown that 3 years' success rates for high and moderate ETVSS groups were 72% and 52% respectively. Suggesting that, early ETV success leads to long term success as well. It is important to recognize the limitation of this analysis. In the best ETV candidates (those with ETVSS $\geq$ 80), however, the risk of ETV failure is lower than that of shunt insertion right after surgery. While for less than ideal ETV candidates (ETVSS $\leq$ 70) the risk of ETV failure is higher than that of shunt insertion and only becomes lower after 3 – 6 months of surgery. Benjamin et al. in his study has shown that combined ETV-Choroid plexus cauterization (ETV – CPC) is significantly superior to ETV alone for infants younger than 1 year of age with congenital auqeductal stenosis.[4] Our study does not include children under 6 months of age presenting with obstructive hydrocephalus during this period. Mansoor Foroughi et al. has described that assessment of third ventricular floor and lamina terminalis morphology is useful in predicting clinical success of ETV and in the follow-up in 96% cases.[8] Andrew J. Durnford from UK and L. Fani from Netherlands have also validated the ETVSS in their studies.[3,6]

Generally, authors have used various terms to describe the patients undergoing ETV e.g. primary, secondary, repeat and salvage ETV. Primary ETV is the term which is used for the patients undergoing ETV as first CSF – diversion procedure. The term Secondary ETV is used when the patient has previously been shunted. Repeat ETV is the term which is synonymous with re-do ETV. The term Salvage ETV represents an important subgroup of patients undergoing ETV for whom likelihood of success is low.[9] This is a group of patients who does not typically meet our pre-operative criteria for ETV but whose malignant frequency of shunt failure or poor options for distal absorption lead to the ETV attempt. Salvage ETV may have up to 25% success rate but this is very important because these are the patients who are saved from their malignant cycle of shunt revisions. Change in ventricular size is not believed to be immediate, it may take months to years after surgery. Sacko et al. noted that 59% of patients with successful ETV had no change in ventricular size at 3 – month of follow-up.[10] Romeo A. et al. have found that majority of the changes occur between 3 months – 1 year post-operatively (Romeo A., Naftel R., Reed G., et al. Endoscopic Third Ventriculostomy for tectal plate Gliomas: long-term outcomes and ventricular size. Presented at the AANS / CNS Section on Pediatric Neurological Surgery Meeting, Cleveland, Ohio Dec, 2010). Kulkarni et al. noted that for a change in ventricular size to be plainly, subjectively apparent, a 15% change in the frontal and occipital ratio must be present.[9] An increase in ventricular size at first post-operative imaging was associated with a significantly increased risk of ETV failure compared with ventricles that decreased in size or remained stable.

The location of extra-ventricular, intra-cisternal blockage is theoretically and practically important. With improved imaging of the cisterns and CSF flow within them, it is conceivable that third ventriculostomy through lamina terminalis or the pineal recess could offer other options to address hydrocephalus due to cisternal blockage at the tentorial incisura.[9]

Triantafyllos Bouras and Spyros Sgouros analyzed 34 series of ETV for complications and concluded that ETV can be regarded as a low-complication procedure with an overall complication rate of 8.5%, morbidity rate of 2.4%, mortality rate of 0.21% and delayed "sudden death" rate of 0.07%.[11]

CONCLUSION

The study shows that in a very large majority of patients it is reliable to apply the predictive value of ETVSS and it can help us in choosing the proper patients who have good chance of getting rid of this disabling disease of hydrocephalus. This prediction becomes very valuable in a country like ours where there is acute shortage of neurosurgeons and ETVSS will help them to decide whether to go for shunt or ETV.

DISCLOSURE

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper. Author contributions to the study and manuscript preparation include the following: Conception and design: Malik Muhammad Nadeem. Drafting the article: all authors. Statistical analysis: Uzair. Study Supervision: Nadeem.

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