Evaluation of the ETV Success Score to Improve Decision Making on the Management of Hydrocephalus Secondary to Pediatric Posterior Fossa Tumors

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Significance:
This study is significant as it assesses the utility of the ETV Success Score in guiding clinical decisions regarding the management of hydrocephalus secondary to pediatric posterior fossa tumors, potentially offering a valuable tool to optimize patient care and outcomes.

Abstract

Introduction: Children with posterior fossa tumors (PFT) often have hydrocephalus, which can be treated via endoscopic third ventriculostomy (ETV) before the tumor is removed, thereby improving surgical outcomes. Additionally, a scoring system that forecasts ETV success can help with this procedure's decision-making. The purpose of the research is to assess the effectiveness of electrotherapy (ETV) in treating PFT-related hydrocephalus and the predictive validity of the ETV effectiveness score.

Materials and Methods: A retrospective analysis was conducted on patients aged < 18 years who received ETV prior to PFT resection between 2022 and 2023. For every patient, the ETV success score was acquired retroactively, and its ability to forecast success contrasted with the actual success was noted.

Results: In the first six months after ETV, a worldwide success rate of 61.7% was observed in 40 surgical patients. For patients who were reviewed a year after surgery, this percentage decreased to 52.8%. The histological level of the tumor and the age of the victim did not correlate with the recovery rate. At both time periods, the ETV effectiveness score showed adequate accuracy (AUC = 0.671 at six months, AUC = 0.649 at one year).

Conclusion: In summary, ETV has been shown to be a safe treatment that may effectively cure hydrocephalus in individuals with PFT. In addition, the ETV efficacy score can be a valuable tool for predicting the likelihood of successful endoscopic surgery.

Introduction

Pediatric patients frequently encounter posterior fossa tumors (PFT), particularly those age < two. Hydrocephalus is a common occurrence in patients with PFT, resulting from blockage of the caudal cerebrospinal fluid drainage pathways, including the aqueduct of Sylvius, and the median and lateral apertures of the fourth ventricle (the Magendie and Luschka foramens, respectively).[1-6] Approximately 80% of patients with PFT develop hydrocephalus. Studies suggest that despite tumor excision being the primary therapeutic approach, approximately 16%-33% of hydrocephalus cases persist post-surgery. This highlights the complexity of managing hydrocephalus secondary to PFT and underscores the importance of comprehensive evaluation and treatment strategies to address this condition effectively in pediatric patients.[7-11]

The standard treatment for hydrocephalus, endoscopic third ventriculostomy (ETV), optimizes conditions for posterior fossa tumor (PFT) resection and enables hardware-free restoration of cerebrospinal fluid flow.[12-14] ETV is favored over tumor removal, external ventricular drainage, or shunt device implantation in several studies. Yet, conflicting evidence persists regarding the reliability and safety of ETV as the primary intervention for pre-PFT hydrocephalus.[15-17] Despite these uncertainties, ETV remains a promising therapeutic option, underscoring the importance of further research to elucidate its efficacy and safety profile in managing hydrocephalus associated with posterior fossa tumors.

For all etiologies of hydrocephalus, the Endoscopic Third Ventriculostomy Success Score (ETVSS) serves as a validated tool to predict procedure outcomes within the initial 6-12 months post-ETV.[18] To assess the utility of ETVSS in clinical decision-making for individuals with these tumors, we aimed to evaluate its predictive ability specifically in the context of PFT.

Materials and methods

Eligibility Criteria

In this study, following approval from the regional ethics committee and validation through the Pakistani government website, a protocol number (#) was assigned. Eligible participants were individuals under 18 years of age who had undergone electrothermal venting (ETV) prior to microsurgical resection of posterior fossa tumors (PFT) and had not undergone shunt placement as their primary treatment for hydrocephalus. Participants were recruited between 2022 and 2023. All surgeries were performed by the same group of pediatric neurosurgeons at a single pediatric neurosurgery center. The study adhered to the STROBE criteria for reporting. (19)

To evaluate the efficacy of endoscopic third ventriculostomy (ETV) specifically for hydrocephalus secondary to posterior fossa tumors, individuals with tumors in the pineal region, mesencephalictectum, or supratentorial region were excluded from the analysis. Moreover, patients who lacked a minimum six-month follow-up period were not included in the study. These stringent inclusion criteria ensured that the study focused exclusively on the effectiveness of ETV in managing hydrocephalus associated with posterior fossa tumors, thereby providing valuable insights into its therapeutic potential for this specific patient population.

Data Collection

Data were retrieved from the electronic medical records of hospitals and private clinics between February and April 2023. The neurosurgical team

compiled a database containing information on all the patients treated between 2022 and 2023.

The study focused on several key factors: (1) demographics, (2) anatomical structures affected by the tumor, (3) tumor histology, and (4) treatment outcomes. Additionally, ETV Success Scores (ETVSS) were calculated considering patient age, the presence of a PFT causing hydrocephalus, and prior shunt placement. Since all patients had the same ETVSS score due to the etiology of hydrocephalus (PFT) and the absence of prior shunt placement, age was the only variable potentially affecting ETVSS outcomes.

Resolution of intracranial hypertension-related symptoms serves as a primary indicator of endoscopic third ventriculostomy (ETV) efficacy. Treatment failure was defined as the persistence of symptoms, such as nausea, headache, and cognitive impairment, or the occurrence of cerebrospinal fluid leakage. To evaluate the effectiveness of ETV, the study compared actual versus anticipated procedure success using Endoscopic Third Ventriculostomy Success Score (ETVSS) scores at six months and one-year post-surgery for each participant. This comprehensive assessment allowed for a thorough evaluation of the efficacy of ETV in managing hydrocephalus associated with posterior fossa tumors, thereby providing valuable insights for clinical decision-making and patient care.

Given the study duration and evolving surgical techniques, such as the adoption of peel-away sheaths for ventricular puncture, the potential impact of the learning curve was considered. This factor acknowledges the need to account for variations in surgical proficiency over time, and the incorporation of novel approaches into practice. By acknowledging the learning curve, this study aimed to ensure accurate interpretation of results and effectively assess the efficacy and safety of emerging techniques in the management of hydrocephalus associated with posterior fossa tumors.

The patients were positioned supine with their head neutrally aligned in a horseshoe headrest and then rotated into a prone position for surgery. Imaging modalities such as MRI or CT scans guided the selection of the entry site and informed predictions regarding neuroendoscopic difficulty based on anatomical landmarks such as the dimensions of the Monroe foramen, proximity between the tuber cinereum and basilar tip, and cortical mantle width. Kocher's maneuver facilitated access to the frontal horn of the lateral ventricle, ensuring precise and effective navigation during the surgical procedure. This meticulous approach to patient positioning and surgical planning optimizes the success of neuroendoscopic intervention for hydrocephalus management.

Preventive measures to mitigate cerebrospinal fluid (CSF) leakage and infection include preoperative hair removal, creating a small arch-shaped incision with anterior convection, and using a No. 11 scalpel for coagulation and dural exposure after burr hole creation. A peel-away sheath was then used for ventricular puncture to reduce the risk of upward herniation. Fenestration of the tuber cinereum and Liliequist membrane was performed using a neuroendoscope, followed by sealing of the burr hole with bone fragments embedded in fibrin glue and geofoam. Skin closure was achieved using separate sutures to ensure meticulous wound management and minimize postoperative complications.

Statistical Analysis

Data analysis was performed using Microsoft Excel 2010 (Microsoft Corporation, Redmond, WA, USA) and SPSS version 25 (IBM Corp., Armonk, NY, USA). A significance level of $P < 0.05$ denoted statistical significance for observed differences.

Furthermore, to assess the discriminative ability of ETVSS in predicting positive and negative outcomes at the 6- and 12-month intervals, a receiver operating characteristic (ROC) curve was generated. Traditionally, a clinical prediction rule is considered adequate if the ETVSS exceeds 0.7. In this context, we identified the optimal ETVSS cut-off value to improve prognostic accuracy.

Results

Forty pediatric patients who underwent endoscopic third ventriculostomy (ETV) prior to posterior fossa tumor (PFT) resection were included in this study. The sample comprised 27 subjects (67.5%, $p = 0.926$) with a mean age of 6.3 years (SD = 3.62).

Within the cohort, 15 individuals (55.5%) were diagnosed with high-grade tumors. Specifically, the diagnoses included five cases of medulloblastoma, four cases of diffuse pontineglioma, two cases of grade III ependymoma, and one case of teratoma. Additionally, there were six cases of pilocyticastrocytomas, four cases of grade II ependymomas, one case of ganglioglioma of the fourth ventricle, one case of cerebellar hemangioblastoma, and one case of grade II glioma of the fourth ventricle. This diverse array of tumor types underscores the complexity of hydrocephalus management in the context of posterior fossa tumors.

The success rate of endoscopic third ventriculostomy (ETV) was notably higher (55.6%) in patients with low-grade tumors than in those with high-grade tumors (44.5%, $p = 0.046$). Anatomically, tumors predominantly affected the cerebellar parenchyma in 18 patients (45%), the floor of the fourth ventricle in 13 patients (32.5%), and the brainstem in 7 patients (17.5%). However, this distribution did not significantly influence the success rate of ETV ($P = 0.638$). These findings underscore the importance of tumor grade in predicting ETV outcomes and highlight the various anatomical locations of posterior fossa tumors.

The duration of the study did not significantly influence the success rate of ETV ($P = 0.409$). Table 1 illustrates the effect of age on ETV efficacy. No

intraoperative deaths were reported during neuroendoscopic procedures.

Table-1: Effect of age on ETV efficacy

Age	N	Success (%)	P value
<03 YEARS	5	5 (12.5)	0.084
>03–10 YEARS	21	21 (52.5)	
;>10 YEARS	14	14 (35)	

During the study, one patient experienced intraoperative bleeding post-tuber cinereumostomy, leading to ETV failure due to hydrocephalus from a diffuse pontineglioma. Subsequently, the patient underwent shunt placement and was discharged without any complications. At six months post-ETV, the success rate was 61.7%, which decreased to 52.8% after one year. The receiver operating characteristic (ROC) curves in Figures 3 and 4 depict the predictive capability of the Endoscopic Third Ventriculostomy Success Score (ETVSS) at these time intervals.

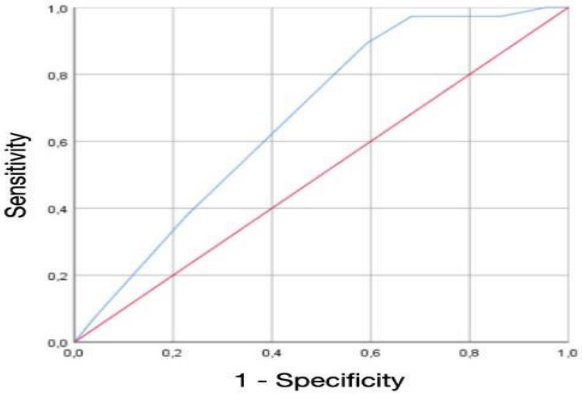

Figure-1: The efficacy of the endoscopic third ventriculostomy scoring system (ETVSS) in forecasting ETV efficacy over the first six months following surgery is shown by the receiver operating characteristic (ROC) curve, which has an area under the curve of 0.671 (p = 0.029).

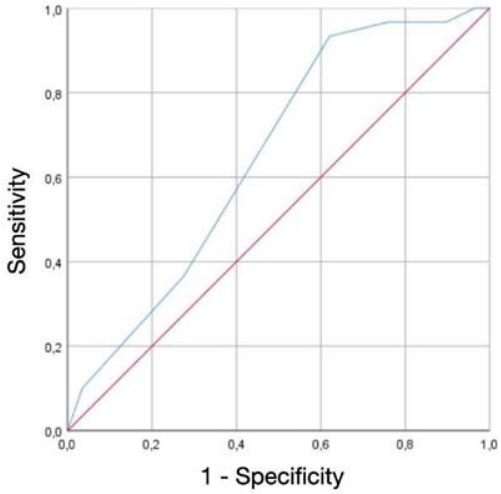

Figure-2: A year following the ETV process, the ROC curve shows the precision of the ETVSS, with an area underneath the graph of 0.649 (p = 0.050).

Discussion

In our single-institutional study, endoscopic third ventriculostomy (ETV) was chosen as the primary treatment modality for hydrocephalus prior to posterior fossa tumor (PFT) resection. This strategic approach facilitated enhanced surgical planning, thereby alleviating intracranial pressure and diminishing the risk of intra-operative hemorrhage.[19] Consequently, it optimized the conditions for tumor resection while reducing the likelihood of postoperative hydrocephalus occurrence. Initially, approximately six out of ten patients achieved successful outcomes with ETV, with approximately half maintaining success at the one-year follow-up mark. Similarly, Srinivasan et al.[20] conducted a comparative study of ETV versus non-ETV groups, revealing a lower shunt dependency rate in the former (22%) than in the latter (32%). Despite limited literature support, our study highlights the advantages of ETV preceding tumor resection, particularly when considering the Endoscopic Third Ventriculostomy Success Score (ETVSS) in clinical decision-making processes (see Table 2).

Table-2

Study	Elbeltagy et al.[14]	El-Ghandour[27]	Azab Et al.[12]	Srinivasan Et al.[20]	Furtado et al.
Year	2010	2011	2013	2020	2023
N	40	32	17	31	59
Failure Rate	37.5%	6.2%	11.8%	21%	37.3%

Technical challenges, post-tumor excision complications, and changes in cerebrospinal fluid (CSF) dynamics during oncologic therapy can contribute to endoscopic third ventriculostomy (ETV) failure. Surgical obstacles may involve hemorrhage, ostomy occlusion, and difficulty fenestrating the Liliequist membrane.[21] Additionally, CSF absorption may be hindered by central nervous system infections post-surgery, potentially causing the "snow globe effect," where tissue debris obstructs the ostomy in the tuber cinereum. These factors underscore the complexity of ETV procedures and highlight the importance of meticulous surgical technique, postoperative management, and infection prevention strategies to enhance the success rates of ETV for hydrocephalus management.[22]

Pediatric neurosurgeons must remain vigilant regarding the risks inherent in endoscopic procedures such as intraoperative tumor bleeding and upper diencephalic herniation due to pressure imbalances.

Consequently, endoscopic third ventriculostomy (ETV) should be performed exclusively by proficient practitioners who ensure comprehensive cerebrospinal fluid (CSF) drainage and precise endoscopic navigation.[23] Although this study did not document upward herniations, irrigation was employed to manage intraoperative hemorrhage effectively. In contrast, the shunt alternative introduces additional risks, including equipment-related complications, such as infections and malfunctions, as well as potential tumor dissemination into the peritoneum, tumor hemorrhage, and herniation induced by over-drainage. Pediatric neurosurgeons must carefully weigh these considerations when determining the most appropriate treatment approach for hydrocephalus management.[24]

In a comprehensive meta-analysis conducted by Dewan et al. (8), which compared the efficacy of endoscopic third ventriculostomy (ETV) versus shunt placement in managing hydrocephalus secondary to posterior fossa tumors (PFT), it was found that the ETV group exhibited a lower incidence of complications (17%) compared to the shunt group (31%, $p = 0.012$).[25] Complications observed included infections, shunt malposition, extra-axial hemorrhages, and cranial nerve palsies. Additionally, Riva-Cambrin et al. (26) proposed a robust scoring system designed to evaluate the risk of persistent hydrocephalus following tumor removal. This scoring system incorporates key factors such as papilledema, histopathological type, presence of cerebral metastases, and age at PFT diagnosis, thereby facilitating risk assessment and informing medical decision-making processes.

Despite the Canadian Preoperative Prediction Rule for Hydrocephalus highlighting age < two years as a key predictor for post-tumor surgery hydrocephalus, our study did not identify a statistically significant variance in hydrocephalus incidence among age groups ($p = 0.084$). Similarly, there was no notable difference ($p = 0.047$) in the likelihood of hydrocephalus among tumors with distinct histology. Notably, the Endoscopic Third Ventriculostomy Success Score (ETVSS) exhibited superior accuracy in predicting the effectiveness of ETV in addressing hydrocephalus associated with posterior fossa tumors (PFT). These findings underscore the potential value of ETVSS in optimizing treatment strategies for PFT-related hydrocephalus.[27]

The predictive capacity of the Endoscopic Third Ventriculostomy Success Score (ETVSS) is paramount in guiding decision-making, given that the primary benefit of ETV is hydrocephalus management without equipment dependency or risk of peritoneal dispersion. However, the retrospective design of our study presents a key limitation, potentially introducing bias due to treatment duration variability. Nonetheless, data sourced from a single institution, where the principal neurosurgeon conducted most surgeries, mitigated the bias arising from diverse surgical techniques. This enhances the evaluation of the results, underscoring the study's reliability despite its retrospective nature.

Conclusion

Therefore, pediatric neurosurgeons must meticulously weigh the benefits and drawbacks of hydrocephalus arising from patient-positioning therapy. Although addressing both conditions concurrently through tumor excision without prior intervention may seem ideal, intracranial hypertension increases the risk of intraoperative hemorrhage. Thus, a thorough assessment of the risks and benefits is essential to ensure the safest and most effective treatment approach for pediatric patients with hydrocephalus associated with posterior fossa tumors.

Tumor resection is a viable and effective elective procedure once hydrocephalus is successfully managed, with skilled practitioners being able to perform endoscopic third ventriculostomy (ETV). Additionally, the Endoscopic Third Ventriculostomy Success Score (ETVSS) exhibits notable predictive accuracy, aiding informed decision-making regarding treatment strategies. This underscores the importance of effectively managing hydrocephalus to optimize the success of subsequent tumor resection procedures, highlighting the role of ETVSS in guiding clinical decisions and ensuring favorable outcomes in patients with posterior fossa tumors.

Thus, despite the inherent risks of endoscopic third ventriculostomy (ETV), it offers fewer complications and superior surgical conditions compared with tumor resection without preoperative intervention. However, prospective controlled studies are required to further support this claim. Such investigations would provide valuable insights into the efficacy and safety of ETV as a therapeutic option for hydrocephalus associated with posterior fossa tumors. With robust evidence from well-designed studies, healthcare providers can make informed decisions regarding the optimal treatment approach for patients, ensuring the best possible outcomes while minimizing risks.

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