



External validation of the ETV success score in 313 pediatric patients: a Brazilian single-center study

Leopoldo Mandic Ferreira Furtado¹ · José Aloysio da Costa Val Filho¹ · Eustaquio Claret dos Santos Júnior¹

Received: 9 June 2020 / Revised: 23 November 2020 / Accepted: 10 December 2020
© Springer-Verlag GmbH Germany, part of Springer Nature 2021

Abstract

The endoscopic third ventriculostomy (ETV) success score (ETVSS) was developed to predict the success rate of ETV at 6 months. In this study, the authors assessed the performance of this score for > 6 months, i.e., at 12 months, and provided external validation in Brazilian children. All children undergoing first ETV (without choroid plexus cauterization) at a Brazilian single institution for > 20 years were included in the study. The ETVSS was retrospectively calculated for each patient and compared with the actual success of the procedure observed at 6 and 12 months after the procedure. A total of 313 eligible children underwent initial ETV, 34.18% of whom had undergone shunt placement before ETV. The most common etiologies were aqueductal stenosis (45%) and non-tectal brain tumors (20.8%). ETV was successful at 6 months in 229 patients (73.16%) compared with the 61.3% predicted by the ETVSS. The overall actual success rate observed at 1 year after ETV was 65.1% (204 patients). The area under the receiver operating characteristic curve was 0.660 at 6 months and 0.668 at 1 year, which suggested a tendency for the ETVSS to underestimate the actual success rate of ETV at both timepoints. The ETVSS showed good success prediction in accordance with the actual ETV success rate and proved to be useful during the decision-making process of ETV.

Keywords Third ventriculostomy · Validity · Neuroendoscopy · Follow-up · Hydrocephalus · Pediatric neurosurgery

Introduction

Endoscopic third ventriculostomy (ETV) has been considered an important procedure in the treatment of occlusive hydrocephalus (mainly aqueductal stenosis) in the last decades [1–4]. However, the selection of this procedure for hydrocephalus remains controversial because of other factors, such as infection, hemorrhage, and myelomeningocele [5–10]. Because of its lower success rate, it is difficult for young neurosurgeons to decide when to perform ETV.

The ETV success score (ETVSS) was developed by Kulkarni et al. [11] to predict the clinical response after ETV based on age, previous shunt surgery, and cause of hydrocephalus in pediatric populations. This score has been validated in several studies across North America, Europe, and the UK [12–20]. Nevertheless, no validation of the ETVSS has been

reported in Latin America. Furthermore, the ETVSS aims to predict the ETV success rate at 6 months; thus, studies addressing its benefit at 1 year are limited [13, 17].

This study aimed to assess the ETVSS in a Brazilian pediatric population to evaluate its performance at 6 months and 1 year after ETV and provide external validation.

Materials and methods

Patients and eligibility criteria

After obtaining approval from the ethics committee, we conducted a retrospective observational study of all consecutive pediatric cases of ETV at a single institution in Minas Gerais, Brazil, between January 1996 and December 2016, through a review of medical records. The STROBE guidelines were followed in this study to provide more high level of evidence in observational studies. The primary ETV procedure was indicated for the combination of clinical symptoms of hydrocephalus and radiological evidence of lateral and third ventricle dilation. All patients who did not undergo at least 1 year of follow-up and children who had undergone choroid plexus

✉ Leopoldo Mandic Ferreira Furtado
lmandicster@gmail.com

¹ Department of Pediatric Neurosurgery, Vila da Serra Hospital, Alameda Oscar Niemeyer, 499, Nova Lima, Minas Gerais 34000-000, Brazil

cauterization and redo ETV or secondary ETV were excluded from this study. Demographic data, etiology of hydrocephalus, and postoperative complications were considered.

The criteria of age and etiology were based on those established by the ETVSS model. Age was stratified into five groups (< 1 month, 1 month completed to 6 months, 6 months completed to 1 year, 1 year completed to 10 years, and > 10 years). Seven groups were considered based on the etiology, i.e., post-central nervous system infection, myelomeningocele, intraventricular hemorrhage, non-tectal brain tumors, tectal brain tumors, and other causes (aneurysm of the vein of Galen and Dandy–Walker syndrome was considered as other causes).

Endoscopic technique

All surgeries were performed under general anesthesia with the patients on a dorsal decubitus position with their heads stabilized in a neutral position on a horseshoe-shaped headrest. A U-shaped skin surgical incision was used. In most procedures, the burr hole was placed based on computed tomography or magnetic resonance imaging findings. The entry point was usually located behind the hairline in the mid-pupillary line. If the anterior fontanelle was still open, the lateral border was carefully dissected, and osteoplastic minicraniotomy was performed with the bone fixed using 3–0 silk surgical suture, according to a previous publication of the senior author (J.A.C.V.F) [21]. The dura was opened without coagulation, and cortex mantle perforation was performed with diathermia before lateral ventricular cannulation using a peel-away sheath. A rigid neuroendoscope (Aesculap or Karl Storz) was used in all cases and advanced into the lateral ventricle, aiming at the foramen of Monro. The main third ventricular landmarks were identified, such as the mammillary bodies and tuber cinereum. Endoscopic ventriculostomy was performed between the mammillary bodies and infundibulum. The tuber cinereum was carefully punctured using a 4-French Fogarty catheter. The stoma was enlarged by inflating the balloon to achieve adequate fenestration. The Lilliequist membrane was opened in all cases to optimize the flow of the cerebrospinal fluid (CSF) through the subarachnoid space. If hemorrhage occurred, irrigation was performed using normal saline solution at body temperature.

Validity of the ETVSS

The ETVSS was retrospectively applied to each patient using the following criteria: age, hydrocephalus etiology, and previous shunt, in accordance with the original article published by Kulkarni et al. [11] (Table 1). The score ranged from 0 to 90 and estimated the probability that an ETV was successful at 6 months. We compared our overall 6-month and 12-month ETV success rates with the mean predicted 6-month success

rate based on the ETVSS. A receiver operating characteristic (ROC) curve was built to test the ability of the ETVSS to discriminate the success from the failure of the technique at both timepoints. An ETVSS > 0.7 was usually considered satisfactory as a clinical prediction rule. Furthermore, we identified the cutoff point of the ETVSS regarding the improved prediction of the actual success of ETV. The covariates of age, etiology, and previous shunt were independently analyzed to estimate the value of each parameter in the prediction of ETV success.

ETV success criteria

After discharge from the hospital, postoperative assessment was performed in the private office of the senior neurosurgeon of this study, and the data and those obtained from the hospital charts were recorded. All patients considered for this study have returned in a quarterly regimen for at least 1 year of follow-up. The data obtained from the patient charts in the hospital were analyzed and collected in the private office by this study author and the resident of neurosurgery. The ETV success was defined as a resolution of the symptoms of hydrocephalus, such as amelioration of bulging anterior fontanelle, control of head circumference, and reversion of ocular impairment signs and improvement in neurodevelopmental delay, in children aged < 2 years, and improvement in severe symptoms of intracranial hypertension, such as impairment of consciousness, vomiting, and headache in older children.

In our institution, usually we did not consider the persistence of ventriculomegaly as a criterion for ETV failure, and clinical aspects were adopted as the main criteria. Furthermore, the diagnosis of ETV failure is based on persistence of symptoms corroborated by worsening of ventricular dilation. In the present study, brain imaging findings were not reassessed, and the information used was based on the charts. Therein, ETV failure was considered when there was a subsequent need for redo ETV or shunt implantation based on clinical and radiological evidence of the persistence or progression of hydrocephalus.

Statistical analysis

SPSS version 20 (IBM, Armonk, NY, USA), Minitab 16 Statistical Software (Minitab, PA, USA), and Microsoft Excel 2010 (Microsoft Corporation, Redmond, WA, USA) were used in the data analyses. A confidence level of 95% was considered.

Student's *t* test was used to compare the mean ETVSS between the patients who exhibited success and those who had ETV failure at both timepoints.

The two-proportion equality test (chi-squared test) was used to compare the success rates among the covariates of

Table 1 ETV success score

| ETVSS | | | |
|-------|-----------------------|---|-------------------|
| Score | Age | Etiology | Previous shunt |
| 0 | < 1 month | Postinfectious | Previous shunt |
| 10 | 1 month to < 6 months | | No previous shunt |
| 20 | | Myelomeningocele, intraventricular hemorrhage, non-tectal brain tumor | |
| 30 | 6 months to < 1 year | Aqueductal stenosis, tectal tumor, other etiology | |
| 40 | 1 year to < 10 years | | |
| 50 | ≥ 10 years | | |

The ETVSS was calculated as the age score + the etiology score+ the previous shunt score. Reprinted with permission from Kulkarni Av et al.: J Neurosurg Pediatr 6:310–315, 2010

age, etiology, and previous shunt. Differences were considered significant at a P value < 0.05.

Results

Patient characteristics

During the study period, the senior neurosurgeon (J.A.C.V.F) performed the majority of ETV procedures at our institution. After the application of the exclusion criteria, 313 pediatric patients were considered for the analysis (Fig. 1). There were 179 boys and 134 girls ($P < 0.001$). The most common etiologies were aqueductal stenosis (45%), non-tectal brain tumors (20.8%), and myelomeningocele (15.3%). Previous shunt was observed in 34.18% of children (Table 2).

Moreover, 65 children (40 boys and 15 girls) were classified as having non-tectal brain tumors of the following types: pilocytic astrocytoma of the cerebellum (19 patients), medulloblastomas (14), ependymomas of the fourth ventricle (9), diffuse glioma of the pons (12), exophytic brainstem glioma (6), hemangioblastoma (1), ependymoblastoma (1), glioma of the cerebellum (1), cerebellar teratoma (1), and ganglioglioma of the fourth ventricle (1).

Effectiveness of the ETV

The overall ETV success rate was 73.2% after 6 months and 65.2% at 1 year. In patients with obstructive hydrocephalus secondary to aqueductal stenosis, the ETV success rate was 83.6% in the first 6 months and 76.6% at 1 year. The covariates of age, etiology, and previous shunt placement were significant predictors of the success of ETV in the first 6 months after the procedure (Tables 3, 4, and 5).

In the follow-up period, 109 ETV procedures were considered failures, with most of them detected in the first 6 months postoperatively (77.06%). In the non-tectal brain tumor category (65), all tumors were located in the posterior fossa, and

ETV preceded tumor resection, with all 22 failures occurring in the first 6 months postoperatively. The ETV success rate for pilocytic astrocytomas of the cerebellum was 84.2%, while it was 50% for medulloblastomas, 55.6% for ependymomas, and 66.7% for diffuse gliomas of the pons. All six cases of exophytic brainstem glioma achieved success. Other

Table 2 Characteristics of the 313 pediatric patients who underwent primary ETV between 1996 and 2016

| Age at ETV | <i>n</i> (%) |
|-----------------------------|--------------|
| ≤ 1 month | 25 (8) |
| > 1 to < 6 months | 54 (17.3) |
| 6 months to < 1 year | 54 (17.3) |
| > 1 to 2 years | 19 (6.1) |
| > 2 to 10 years | 98 (31.3) |
| ≥ 10 years | 63 (20.1) |
| Etiology of hydrocephalus | |
| Aqueductal stenosis | 141 (45) |
| Non-tectal brain tumor | 65 (20.8) |
| Myelomeningocele | 48 (15.3) |
| Other | 18 (5.8) |
| Intraventricular hemorrhage | 17 (5.4) |
| Tectal tumor | 13 (4.2) |
| Postinfectious | 11 (3.5) |
| Previous shunt | 107 (34.18) |
| ETVSS | |
| 10 | 9 (2.9) |
| 20 | 5 (1.6) |
| 30 | 20 (6.4) |
| 40 | 37 (11.8) |
| 50 | 53 (17) |
| 60 | 36 (11.5) |
| 70 | 63 (20.1) |
| 80 | 67 (21.4) |
| 90 | 23 (7.3) |

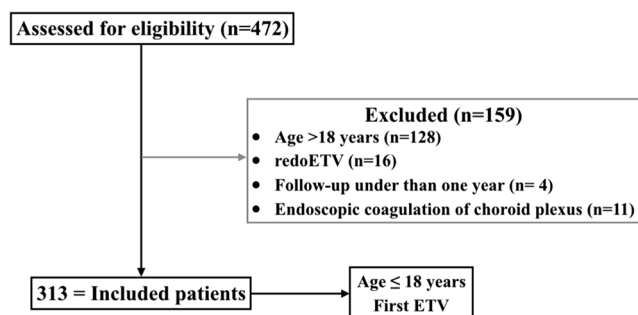


Fig. 1 Flow diagram of the selection process that was used to identify patients who underwent ETV for inclusion in this study

pathological samples, such as ependymoblastoma, glioma, and teratoma of the cerebellum, showed ETV failure. Complications developed in 5.8% (18) of patients and included subdural hemorrhage (3 patients), CSF leakage (3), hyponatremia (2), meningitis (2), transient ophthalmoparesis (2), transient hemiparesis (1), brain hemorrhage (1), long-standing fever (1), and severe intraoperative hemorrhage (3). One patient in this series died due to complication of a hemorrhage that developed 1 week after the procedure.

External validation of the ETVSS

The majority of the probability predicted by the ETVSS was between 50% and 70%, followed by > 80%, whereas a lower ETVSS was detected in a minority of cases (Table 6).

Overall, we observed a discrepancy between the actual success rate and that predicted by the ETVSS, with a tendency toward underestimation of the actual success of the procedure at the 6-month and even 1-year timepoint.

The ETVSS exhibited a moderate accuracy for predicting the actual ETV success according to the ROC curve analysis, with an area under the curve of 0.660 at 6 months and 0.668 at 1 year after ETV (both, $P < 0.05$) (Fig. 2). From the ROC curve, we obtained the best cutoff points to predict the success of ETV, i.e., 0.45 at 6 months (sensitivity, 83.8%; specificity, 40.5%) and 0.65 at 1 year (sensitivity, 57.8%; specificity, 67.9%).

Table 3 ETV success at 6 months according to ETVSS age group

| Age | ETV | | <i>P</i> value |
|--------------------|----------------------|----------------------|----------------|
| | Success <i>n</i> (%) | Failure <i>n</i> (%) | |
| < 1 month | 14 (56) | 11 (44) | < 0.001 |
| 1 to 6 months | 38 (70.4) | 16 (29.6) | |
| 6 months to 1 year | 38 (70.4) | 16 (29.6) | |
| 1 year to 10 years | 85 (72.6) | 32 (27.4) | |
| > 10 years | 54 (85.7) | 9 (14.2) | |
| Sum | 229 (73.2) | 84 (26.8) | |

Table 4 Success of ETV at 6 months according to etiology

| Etiology | ETV | | <i>P</i> value |
|-----------------------------|----------------------|----------------------|----------------|
| | Success <i>n</i> (%) | Failure <i>n</i> (%) | |
| Aqueductal stenosis | 118 (83.7) | 23 (16.3) | < 0.001 |
| Non-tectal brain tumor | 43 (66.2) | 22 (33.8) | |
| Myelomeningocele | 26 (54.2) | 22 (45.8) | |
| Other etiology | 15 (83.3) | 3 (16.7) | |
| Intraventricular hemorrhage | 8 (47) | 9 (53) | |
| Tectal tumor | 11 (84.6) | 2 (15.4) | |
| Postinfectious | 8 (72.7) | 3 (27.3) | |
| Sum | 229 (73.2) | 84 (26.8) | |

The application of Student's *t* test to compare the mean ETVSS between patients with failed and successful ETV at both timepoints yielded a cutoff ETVSS of 50% at 6 months and 70% at 1 year. At both timepoints, the difference between success and failure was significant ($P < 0.001$) (Table 7).

Discussion

In this study, we selected a large group of children who underwent ETV, which represents an experience of a Brazilian single institution as a way to better define the accuracy of ETVSS and help in decision-making of the pediatric neurosurgeon to estimate the probability of success of ETV for the first time in Latin America and provide external validation of this scoring system. The overall analysis of ETV success observed in this present study, which depicted the overall ETV success rate of 73.2% and lower morbidity, undoubtedly corroborated several previous studies that reported the effectiveness and safety of the neuroendoscopic technique [16, 19, 20, 22–27].

With the increasing application of neuroendoscopy as an important option for hydrocephalus treatment in the 1980s, many studies that aimed to evaluate the relationship of children's age and effectiveness of the ETV and results demonstrating great heterogeneity have been published [5, 26, 28]. Zaben et al. [27] performed a meta-analysis to study the effectiveness of ETV in extremely young children and after applying the inclusion criteria in 629 studies published between 1990 and 2018 at this regard, included 19 eligible

Table 5 ETV success at 6 months according to previous shunt

| | Success <i>n</i> (%) | Failure <i>n</i> (%) | <i>P</i> value |
|----------------|----------------------|----------------------|----------------|
| Previous shunt | 67 (28) | 40 (45) | 0.002 |
| Sum | 229 (73.2) | 84 (26.8) | |

Table 6 Success of ETV based on the ETVSS

| Variable | Number of cases (%) | Six-month success rate |
|-------------------------|---------------------|------------------------|
| ETVSS (<i>n</i> = 313) | | |
| < 40% | 71 (22.7) | 52.1% |
| 50–70% | 152 (48.6) | 77% |
| > 80% | 90 (28.7) | 83.3% |

studies, and found that the success rate ranges from 0% to as high as 83%, with a mean of 53%, and presented a wide range of follow-up of 1 week to 106 months, with a mean of 27 months. One of the causes attributed to such discrepancy was the different success criteria. The higher ETV failure rate in this population reported in some studies was explained by Shizuo Oi and Concezio Di Rocco using a novel evolutionary theory in which they described a minor pathway as a main CSF circulation in children aged < 6 months in which the CSF is absorbed by ependyma, choroid plexus, and neuronal cells instead of arachnoid villi, which is not influenced by ETV [29]. However, the success was not properly explained in the current literature. Our study reported an ETV success rate of 70% in children aged < 1 year and 56% in children aged < 1 month, which could be considered high and with a 12 months of follow-up according to the purpose of the study, which was to define the ETVSS accuracy in this timepoint. In spite of this impressive success rate initially encouraging us to perform this approach in children with extremely low age, caution must be taken before the indication of neuroendoscopy in this population. Technically, the closer structures in the third ventricle floor carry an additional risk to damage the basilar artery and oculomotor nerve during ostomy. Additionally, these patients have increased risk of developing complications, such as hypothermia and acid-basic disturbances during the procedure, mainly at < 1 month. Herein, the controversies regarding the indication of neuroendoscopic approach for extremely low-aged children remain, and further and more strengthened studies with prospective design, longer follow-up, and more

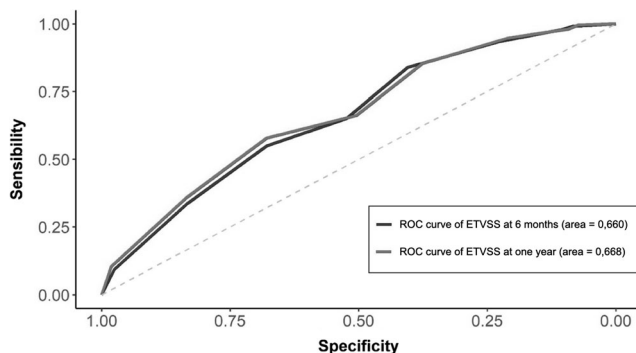


Fig. 2 ROC curve of the ETVSS predicted the actual success at 6 months and 1 year after ETV. The area under the curve was 0.660 at 6 months and 0.668 at 1 year

Table 7 Mean ETVSS in patients exhibiting successful and failed ETV

| | Success | | Failure | |
|------------|---------|-------|---------|-------|
| | Mean | SD | Mean | SD |
| Six months | 0.637 | 0.181 | 0.517 | 0.216 |
| One year | 0.648 | 0.179 | 0.525 | 0.209 |

The values were converted to facilitate the calculation. For example: 0.637 means 60% ETVSS

accurate measurements will be needed to better define the long-standing ETV effectiveness in this population.

The covariates of etiology of hydrocephalus and previous shunt placement were also correlated with the overall ETV success rate. This confirms the hypothesis proposed by Kulkarni et al. [11] that differences in patient age, cause of hydrocephalus, and previous shunt status are responsible for variances in the initial ETV success, as reported in several studies [1, 22, 30–32]. Aqueductal stenosis and tectal tumors presented the highest ETV success rate in these previous studies. Otherwise, we observed an unexpected success rate in postinfectious patients, suggesting that this etiology does not completely exclude the indication of ETV, which could present other benefits, such as cleaning of the ventricles; moreover, if the Liliequist membrane is adequately opened and the naked basilar sign is achieved, this may be an independent intraoperative success variable [23].

The management of hydrocephalus related to posterior fossa tumors is controversial and can be conducted by shunt placement, tumor resection plus external ventricular drainage, tumor resection alone, or ETV before surgery. In the present study, all patients underwent ETV 3 days before tumor resection. The rationale was the achievement of better conditions during tumor resection, treatment of intracranial hypertension due to hydrocephalus, and avoidance of shunt placement. In this context, shunt freedom was achieved in 66% of cases. However, some series have reported success rates of ETV before posterior fossa tumor resection < 70% [22, 33] and a similar ETV failure rate [34]. A discrepancy was observed in the literature, with a tendency toward reporting higher success rates in more recent series [24, 35–38]. One explanation for this observation may be the acute onset of hydrocephalus in tumor cases, according to Oertel et al. [24], who hypothesized the lower probability of membrane formation, as observed in chronic cases. Conversely, other authors have advocated that factors such as midline position of tumors and subtotal tumor resection are associated with shunt dependence of the posterior fossa tumors [33]. In the present series, we observed a higher rate of success in the resection of benign tumors, such as pilocytic astrocytoma (84.2%), compared with malignant tumors, such as medulloblastomas (50%) and ependymomas (66.7%). We speculate that malignant tumors and those that

are more difficult to resect, together with the need of neoadjuvant therapies, affect CSF absorption and contribute to ETV failure. The present results confirmed that ETV was an efficient treatment before posterior fossa tumor resection.

Postoperatively, it is common to observe at least some degree of ventriculomegaly, which should not be considered as ETV failure. Although ventriculometric data, such as Evans' index and frontal and occipital horn ratio, could not be used in this study, the most significant variable for predicting ETV success was clinical improvement in patients. Several studies have demonstrated a lack of differences in neurocognitive performance between patients who underwent shunt or neuroendoscopic treatment based on ventricular volumetry [39, 40].

The ETVSS underestimated the overall success rate of ETV at both time points, and the ROC curve analysis demonstrated an insufficient accuracy level. However, we considered the cutoff value of 50% to aid the decision-making process of ETV after 6 and 12 months, because the score maintained its accuracy and the area under the ROC curve at both time points was highly similar (Fig. 2). Nevertheless, the accuracy could decrease if a longer follow-up period is considered. Breimer et al. [13] evaluated the ETVSS at 6 and 36 months after ETV and demonstrated higher accuracy in the first 6 months than that in other studies, with an area under the ROC curve of 0.82, which decreased to 0.73 at the end of 36 months after ETV (Table 8). Our impression was that the ETVSS combined with the major clinical variables helped neurosurgeons reach a decision. However, this contention warrants further evidence of the external validity of the ETVSS. Similarly, Labidi et al. performed an external validation of the ETVSS by enrolling a mixed population of 168 patients and using an ROC curve analysis to test the accuracy of the ETVSS, with an area under the curve of 0.61 [19], and this finding was corroborated by Foley et al. [15], who demonstrated the same accuracy of the ETVSS in another external validation study conducted on 112 pediatric population of Ireland (Table 8).

Naftel et al. conducted external validation of the ETVSS using a sample of 151 consecutive pediatric cases. These authors

reported a success rate of 68.4% at 6 months after ETV. They also reported a similar rate of complications of 9.3% and highlighted the fact that the ETVSS overestimated the ETV success in patients with a success prediction < 70% [20].

The reasons for this discrepancy are unclear. However, one possible explanation is the unexpected success observed in patients who had previous central nervous system infections. This population is normally underscored by the ETVSS and can exhibit variations in ventricular injury after infections. Perhaps, it would be more adequate to not score cases of multiloculated hydrocephalus.

Another explanation could be the different proportions observed in the present study for an ETVSS < 40%, as we detected a larger number of patients with a lower ETVSS than did the prospective study performed by Kulkarni et al.: 71 (22.6%) children had a success prediction rate of 40% and actual ETV success rate of 52.1% compared with 14 (4.2%) children with a success prediction rate of 21.4% in the previous study [23]. Furthermore, in the present study, the high ETVSS presented with a low failure rate of 16.7% compared with other studies [11, 30]. Gianaris et al. considered that the ETV success rate is higher in patients with acute increase in intracranial pressure [2].

Among the ETVSS variables, age < 1 month, hemorrhage related to prematurity, and previous shunt exhibited the strongest associations with ETV failure. However, neurosurgeons should evaluate the advantages and disadvantages of the procedure before reaching a decision regarding shunt placement.

Our study clearly had some limitations, as it was a retrospective study, which could be related to imprecise data and miscalculation, and lacked blinded reviewers in the assessment of predictive variables. Moreover, the absence of outcome measurements, such as neurocognitive data, may have introduced a bias. Regarding the high ETV success rate observed in extremely young children, we strongly recommend further studies with prospective design and longer follow-up. Nevertheless, this study corroborated the usefulness of the ETVSS in clinical practice.

Table 8 Accuracy of the ETVSS in predicting the actual ETV success among studies

| Authors and year | City and state or country | Study design | Sample size (<i>n</i>) | Area under ROC curve | | |
|----------------------|---------------------------|--------------|--------------------------|----------------------|-----------|-----------|
| | | | | 6 months | 12 months | 36 months |
| Kulkarni et al. [11] | Multicentric study* | P OS | 618 | 0.68 | NR | NR |
| Naftel et al. [20] | Alabama, EUA | R OS | 151 | 0.74 | NR | NR |
| Breimer et al. [13] | Groningen, Netherlands | R OS | 104 | 0.82 | NR | 0,73 |
| Labidi et al. [19] | Québec, Canada | R OS | 168 | 0.61 | NR | NR |
| Foley et al. [15] | Dublin, Ireland | R OS | 112 | 0.61 | NR | NR |
| Current study 2020 | Nova Lima, Brazil | R OS | 313 | 0.66 | 0.67 | NR |

12 pediatric institutions in Canada, Israel, and the UK. *P*, prospective; *OS*, observational study; *NR*, not recorded

The ETVSS is a valid score that allowed adequate prediction of ETV success rate even after 6 months of neuroendoscopic approach in a large Brazilian pediatric population and could be considered as a valuable tool for neurosurgery decision-making.

Authors' contributions Conceptualization: Leopoldo Mandic Ferreira Furtado; methodology: Leopoldo Mandic Ferreira Furtado; formal analysis and investigation: Leopoldo Mandic Ferreira Furtado and José Aloysio da Costa Val Filho; writing—original draft preparation: Leopoldo Mandic Ferreira Furtado; writing—review and editing: Leopoldo Mandic Ferreira Furtado, José Aloysio da Costa Val Filho, and Eustaquio Claret dos Santos Júnior; supervision: José Aloysio da Costa Val Filho.

Data availability The data that support the findings of this study are available in Mendeley (data.mendeley.com) with the identifier <https://doi.org/10.17632/3g9y568hhw.1>.

Compliance with ethical standards

Conflict of interest The authors declare that they have no competing interests.

Ethics approval This study was approved by the ethical board with protocol number CAAE: 31084920.7.0000.5125.

Consent to participate Not applicable.

Consent for publication Not applicable.

Code availability Not applicable.

References

- Drake JM, Canadian Pediatric Neurosurgery Study G (2007) Endoscopic third ventriculostomy in pediatric patients: the Canadian experience. *Neurosurgery* 60:881–886; discussion 881–886. <https://doi.org/10.1227/01.NEU.0000255420.78431.E7>
- Gianaris TJ, Nazar R, Middlebrook E, Gonda DD, Jea A, Fulkerson DH (2017) Failure of ETV in patients with the highest ETV success scores. *J Neurosurg Pediatr* 20:225–231. <https://doi.org/10.3171/2016.7.PEDS1655>
- Kulkarni AV, Sgouros S, Constantini S, Investigators I (2016) International Infant Hydrocephalus Study: initial results of a prospective, multicenter comparison of endoscopic third ventriculostomy (ETV) and shunt for infant hydrocephalus. *Childs Nerv Syst* 32:1039–1048. <https://doi.org/10.1007/s00381-016-3095-1>
- Warf BC (2005) Comparison of endoscopic third ventriculostomy alone and combined with choroid plexus cauterization in infants younger than 1 year of age: a prospective study in 550 African children. *J Neurosurg* 103:475–481. <https://doi.org/10.3171/ped.2005.103.6.0475>
- Costa Val JA, Scaldaferrri PM, Furtado LM, de Souza BG (2012) Third ventriculostomy in infants younger than 1 year old. *Childs Nerv Syst* 28:1233–1235. <https://doi.org/10.1007/s00381-012-1740-x>
- Elgamal EA, El-Dawlatly AA, Murshid WR, El-Watidy SM, Jamjoom ZA (2011) Endoscopic third ventriculostomy for hydrocephalus in children younger than 1 year of age. *Childs Nerv Syst* 27:111–116. <https://doi.org/10.1007/s00381-010-1254-3>
- Lam S, Harris D, Rocque BG, Ham SA (2014) Pediatric endoscopic third ventriculostomy: a population-based study. *J Neurosurg Pediatr* 14:455–464. <https://doi.org/10.3171/2014.8.PEDS13680>
- Tamburrini G, Frassanito P, Iakovaki K, Pignotti F, Rendeli C, Murolo D, Di Rocco C (2013) Myelomeningocele: the management of the associated hydrocephalus. *Childs Nerv Syst* 29:1569–1579. <https://doi.org/10.1007/s00381-013-2179-4>
- Teo C, Jones R (1996) Management of hydrocephalus by endoscopic third ventriculostomy in patients with myelomeningocele. *Pediatr Neurosurg* 25:57–63; discussion 63. <https://doi.org/10.1159/000121098>
- Tuli S, Drake J, Lamberti-Pasculli M (2003) Long-term outcome of hydrocephalus management in myelomeningoceles. *Childs Nerv Syst* 19:286–291. <https://doi.org/10.1007/s00381-003-0759-4>
- Kulkarni AV, Drake JM, Mallucci CL, Sgouros S, Roth J, Constantini S, Canadian Pediatric Neurosurgery Study G (2009) Endoscopic third ventriculostomy in the treatment of childhood hydrocephalus. *J Pediatr* 155:254–259 e251. <https://doi.org/10.1016/j.jpeds.2009.02.048>
- Azimi P, Mohammadi HR (2014) Predicting endoscopic third ventriculostomy success in childhood hydrocephalus: an artificial neural network analysis. *J Neurosurg Pediatr* 13:426–432. <https://doi.org/10.3171/2013.12.PEDS13423>
- Breimer GE, Sival DA, Brusse-Keizer MG, Hoving EW (2013) An external validation of the ETVSS for both short-term and long-term predictive adequacy in 104 pediatric patients. *Childs Nerv Syst* 29:1305–1311. <https://doi.org/10.1007/s00381-013-2122-8>
- Durnford AJ, Kirkham FJ, Mathad N, Sparrow OC (2011) Endoscopic third ventriculostomy in the treatment of childhood hydrocephalus: validation of a success score that predicts long-term outcome. *J Neurosurg Pediatr* 8:489–493. <https://doi.org/10.3171/2011.8.PEDS1166>
- Foley RW, Ndoro S, Crimmins D, Caird J (2017) Is the endoscopic third ventriculostomy success score an appropriate tool to inform clinical decision-making? *Br J Neurosurg* 31:314–319. <https://doi.org/10.1080/02688697.2016.1229744>
- Furlanetti LL, Santos MV, de Oliveira RS (2012) The success of endoscopic third ventriculostomy in children: analysis of prognostic factors. *Pediatr Neurosurg* 48:352–359. <https://doi.org/10.1159/000353619>
- Garcia LG, Lopez BR, Botella GI, Paez MD, da Rosa SP, Rius F, Sanchez MA (2012) Endoscopic third ventriculostomy success score (ETVSS) predicting success in a series of 50 pediatric patients. Are the outcomes of our patients predictable? *Childs Nerv Syst* 28:1157–1162. <https://doi.org/10.1007/s00381-012-1836-3>
- Kulkarni AV, Riva-Cambrin J, Browd SR (2011) Use of the ETV success score to explain the variation in reported endoscopic third ventriculostomy success rates among published case series of childhood hydrocephalus. *J Neurosurg Pediatr* 7:143–146. <https://doi.org/10.3171/2010.11.PEDS10296>
- Labidi M, Lavoie P, Lapointe G, Obaid S, Weil AG, Bojanowski MW, Turmel A (2015) Predicting success of endoscopic third ventriculostomy: validation of the ETV success score in a mixed population of adult and pediatric patients. *J Neurosurg* 123:1447–1455. <https://doi.org/10.3171/2014.12.JNS141240>
- Naftel RP, Reed GT, Kulkarni AV, Wellons JC (2011) Evaluating the Children's Hospital of Alabama endoscopic third ventriculostomy experience using the endoscopic third ventriculostomy success score: an external validation study. *J Neurosurg Pediatr* 8:494–501. <https://doi.org/10.3171/2011.8.PEDS1145>
- Costa Val JA (2009) Minicraniotomy for endoscopic third ventriculostomy in babies: technical note with a 7-year-segment

- analysis. *Childs Nerv Syst* 25:357–359. <https://doi.org/10.1007/s00381-008-0748-8>
22. Brockmeyer D, Abtin K, Carey L, Walker ML (1998) Endoscopic third ventriculostomy: an outcome analysis. *Pediatr Neurosurg* 28: 236–240. <https://doi.org/10.1159/000028657>
 23. Kulkarni AV, Riva-Cambria J, Holubkov R, Browd SR, Cochrane DD, Drake JM, Limbrick DD, Rozzelle CJ, Simon TD, Tamber MS, Wellons JC 3rd, Whitehead WE, Kestle JR, Hydrocephalus Clinical Research N (2016) Endoscopic third ventriculostomy in children: prospective, multicenter results from the Hydrocephalus Clinical Research Network. *J Neurosurg Pediatr* 18:423–429. <https://doi.org/10.3171/2016.4.PEDS163>
 24. Oertel JM, Baldauf J, Schroeder HW, Gaab MR (2009) Endoscopic options in children: experience with 134 procedures. *J Neurosurg Pediatr* 3:81–89. <https://doi.org/10.3171/2008.11.PEDS0887>
 25. Stovell MG, Zakaria R, Ellenbogen JR, Gallagher MJ, Jenkinson MD, Hayhurst C, Mallucci CL (2016) Long-term follow-up of endoscopic third ventriculostomy performed in the pediatric population. *J Neurosurg Pediatr* 17:734–738. <https://doi.org/10.3171/2015.11.PEDS15212>
 26. Yadav YR, Jaiswal S, Adam N, Basoor A, Jain G (2006) Endoscopic third ventriculostomy in infants. *Neurol India* 54: 161–163
 27. Zaben M, Manivannan S, Sharouf F, Hammad A, Patel C, Bhatti I, Leach P (2020) The efficacy of endoscopic third ventriculostomy in children 1 year of age or younger: a systematic review and meta-analysis. *Eur J Paediatr Neurol* 26:7–14. <https://doi.org/10.1016/j.ejpn.2020.02.011>
 28. Baldauf J, Oertel J, Gaab MR, Schroeder HW (2007) Endoscopic third ventriculostomy in children younger than 2 years of age. *Childs Nerv Syst* 23:623–626. <https://doi.org/10.1007/s00381-007-0335-4>
 29. Oi S, Di Rocco C (2006) Proposal of “evolution theory in cerebrospinal fluid dynamics” and minor pathway hydrocephalus in developing immature brain. *Childs Nerv Syst* 22:662–669. <https://doi.org/10.1007/s00381-005-0020-4>
 30. Ogiwara H, Dipatri AJ Jr, Alden TD, Bowman RM, Tomita T (2010) Endoscopic third ventriculostomy for obstructive hydrocephalus in children younger than 6 months of age. *Childs Nerv Syst* 26:343–347. <https://doi.org/10.1007/s00381-009-1019-z>
 31. Sacko O, Boetto S, Lauwers-Cances V, Dupuy M, Roux FE (2010) Endoscopic third ventriculostomy: outcome analysis in 368 procedures. *J Neurosurg Pediatr* 5:68–74. <https://doi.org/10.3171/2009.8.PEDS08108>
 32. Wellons JC 3rd, Tubbs RS, Banks JT, Grabb B, Blount JP, Oakes WJ, Grabb PA (2002) Long-term control of hydrocephalus via endoscopic third ventriculostomy in children with tectal plate gliomas. *Neurosurgery* 51:63–67; discussion 67–68. <https://doi.org/10.1097/00006123-200207000-00010>
 33. Ray P, Jallo GI, Kim RY, Kim BS, Wilson S, Kothbauer K, Abbott R (2005) Endoscopic third ventriculostomy for tumor-related hydrocephalus in a pediatric population. *Neurosurg Focus* 19:E8. <https://doi.org/10.3171/foc.2005.19.6.9>
 34. Sherrod BA, Iyer RR, Kestle JRW (2020) Endoscopic third ventriculostomy for pediatric tumor-associated hydrocephalus. *Neurosurg Focus* 48:E5. <https://doi.org/10.3171/2019.10.FOCUS19725>
 35. Bhatia R, Tahir M, Chandler CL (2009) The management of hydrocephalus in children with posterior fossa tumours: the role of pre-resectional endoscopic third ventriculostomy. *Pediatr Neurosurg* 45:186–191. <https://doi.org/10.1159/000222668>
 36. El-Ghandour NM (2011) Endoscopic third ventriculostomy versus ventriculoperitoneal shunt in the treatment of obstructive hydrocephalus due to posterior fossa tumors in children. *Childs Nerv Syst* 27:117–126. <https://doi.org/10.1007/s00381-010-1263-2>
 37. Ruggiero C, Cinalli G, Spennato P, Aliberti F, Cianciulli E, Trischitta V, Maggi G (2004) Endoscopic third ventriculostomy in the treatment of hydrocephalus in posterior fossa tumors in children. *Childs Nerv Syst* 20:828–833. <https://doi.org/10.1007/s00381-004-0938-y>
 38. Sainte-Rose C, Cinalli G, Roux FE, Maixner R, Chumas PD, Mansour M, Carpentier A, Bourgeois M, Zerah M, Pierre-Kahn A, Renier D (2001) Management of hydrocephalus in pediatric patients with posterior fossa tumors: the role of endoscopic third ventriculostomy. *J Neurosurg* 95:791–797. <https://doi.org/10.3171/jns.2001.95.5.0791>
 39. Kulkarni AV, Donnelly R, Mabbott DJ, Widjaja E (2015) Relationship between ventricular size, white matter injury, and neurocognition in children with stable, treated hydrocephalus. *J Neurosurg Pediatr* 16:267–274. <https://doi.org/10.3171/2015.1.PEDS14597>
 40. Warf B, Ondoma S, Kulkarni A, Donnelly R, Ampeire M, Akona J, Kabachelor CR, Mulondo R, Nsubuga BK (2009) Neurocognitive outcome and ventricular volume in children with myelomeningocele treated for hydrocephalus in Uganda. *J Neurosurg Pediatr* 4:564–570. <https://doi.org/10.3171/2009.7.PEDS09136>

Publisher's note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.