Use of the ETV Success Score to explain the variation in reported endoscopic third ventriculostomy success rates among published case series of childhood hydrocephalus

Clinical article

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Objective. Published case series of endoscopic third ventriculostomy (ETV) for childhood hydrocephalus have reported widely varying success rates. The authors recently developed and internally validated the ETV Success Score (ETVSS); this is a simplified means of predicting the 6-month success rate of ETV for a child with hydrocephalus, based on age, etiology of hydrocephalus, and presence of a previous shunt. The authors hypothesized that the ETVSS would be able to predict with reasonable accuracy the actual ETV success rate reported among published case series.

Methods. A literature search was performed to identify published pediatric ETV papers that contained enough information with which to calculate an aggregate, mean predicted ETVSS for the cohort. This was then compared with the actual ETV success rate in the cohort. Data were extracted independently in triplicate, including by 2 individuals who were not involved with the development of the ETVSS.

Results. Fifteen papers reporting on 322 patients were included. Interrater reliability was very high in determining the predicted ETVSS (intraclass correlation coefficient 0.99). The predicted ETVSS for each paper agreed strongly with the actual ETV success rate reported in each paper (reliability intraclass correlation coefficient 0.81). There was no significant difference in the magnitude of the predicted ETVSS and the actual ETV success (p = 0.98, paired t-test). In a linear regression model, the predicted ETVSS explained 62% of the variation in actual ETV success. When the entire cohort was combined and analyzed together, the overall mean predicted ETVSS was 57.9%, which was nearly identical to the actual ETV success rate of 59.2%.

Conclusions. The ETVSS closely predicts the actual ETV success rate reported in selected papers published over the last 20 years and explains much of the variation. (DOI: 10.3171/2010.11.PEDS10296)

Key Words • endoscopy • pediatric hydrocephalus • endoscopic third ventriculostomy

Published case series of ETV for childhood hydrocephalus have reported widely varying success rates. A large part of this variation is probably due to patient selection, resulting in different patient prognostic features, such as age and the underlying etiology of the hydrocephalus. These differences, however, have been hard to quantify. Recently, we used logistic regression techniques to develop and internally validate a prediction score that would, with good accuracy, predict the 6-month success rate of ETV, taking into account the patient’s age, the etiology of their hydrocephalus, and presence of a previous shunt. The end product of this was the ETVSS (Table 1), a simple means to predict ETV success, with scores ranging from 0 (meaning virtually no chance of ETV success) to 90 (meaning a roughly 90% chance of ETV success). In a separate publication, we showed, as an example, that the ETVSS could be used to adjust for a patient’s prognostic factors to explain much of the difference in ETV outcome between children treated in the developed world and those treated in sub-Saharan Africa. With the current study, our objective was to determine if the ETVSS could accurately predict the highly variable ETV success rates reported among published case series. In essence, this was a test of the external validity of the ETVSS. If it could predict ETV success among published papers, this would add further confidence in its ability to be applied more broadly in neurosurgical centers around the world.

Abbreviations used in this paper: ETV = endoscopic third ventriculostomy; ETVSS = ETV Success Score; ICC = intraclass correlation coefficient.
TABLE 1: Calculation of the ETVSS*

<table>
<thead>
<tr>
<th>Score</th>
<th>Age</th>
<th>Etiology</th>
<th>Previous Shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>&lt;1 mo</td>
<td>postinfectious</td>
<td>previous shunt</td>
</tr>
<tr>
<td>10</td>
<td>1 mo to &lt;6 mos</td>
<td>myelomeningocele, IVH, nontectal</td>
<td>no previous shunt</td>
</tr>
<tr>
<td></td>
<td></td>
<td>brain tumor</td>
<td></td>
</tr>
<tr>
<td>30</td>
<td>6 mos to &lt;1 yr</td>
<td>aqueductal stenosis, tectal tumor,</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>other</td>
<td></td>
</tr>
<tr>
<td>40</td>
<td>1 yr to &lt;10 yrs</td>
<td></td>
<td></td>
</tr>
<tr>
<td>50</td>
<td>≥10 yrs</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


Methods

Article Selection

A wide search for published ETV case series was performed. This included the following elements: 1) a search of PubMed and Google Scholar in which the search term “endoscopic third ventriculostomy” was used; 2) a cited reference search of relevant papers by using Google Scholar, to identify additional papers that had cited one of the relevant papers; and 3) a search of the bibliographies of all relevant papers. Included papers met the following criteria: 1) were exclusively or primarily pediatric case series published in peer-reviewed journals; 2) reported results of pure ETV (not a stereotactic or open procedure); 3) had enough data published within the paper to be able to calculate an aggregate ETVSS for the sample (for example, the number of patients within each age group had to be provided, not just mean/median age); 4) the data from the patients presented in the paper had not already been used in the development of the ETVSS; and 5) adequate outcome data were available in the publication to determine ETV success at 6 months.

Endoscopic third ventriculostomy failure was generally defined as treatment failure requiring any subsequent surgical procedure for definitive CSF diversion (that is, either shunt or ETV) or death related to hydrocephalus management. However, within each paper, we accepted the individual authors’ definition of ETV failure.

Outcome Measure

The ETVSS (Table 1) ranges from 0 to 90, and the number itself roughly approximates the percentage chance that an ETV will be successful at 6 months. For example, an 8-month-old baby with aqueductal stenosis and no previous shunt would have an ETVSS of (30 + 30 + 10) = 70, or a roughly 70% chance of having a successful ETV without failure at 6 months postprocedure. We have previously demonstrated the internal validity of the ETVSS.13

Data Extraction

For each paper, we calculated the ETVSS based on the number of patients within each category of age, etiology, and previous shunt. We termed this the “predicted ETVSS.” This provided an average ETVSS that represented the proportion of patients within the paper’s cohort who would be predicted to have an ETV that was successful at 6 months. We also calculated the actual proportion of patients within each paper whose ETV was successful at 6 months, as reported by the authors. We termed this the “actual ETV success.”

All of the above data elements were extracted from all papers by each of the 3 authors, working independently and blinded to the others’ responses. Two of the authors (J.R.C. and S.R.B.) had not been involved in the development of the ETVSS and received no extra training beyond reading the published paper describing its development and use.13

Statistical Analysis

 Interrater Reliability. Agreement among the 3 raters in their assessment of predicted ETVSS and actual ETV success was calculated using the ICC derived from a 1-way ANOVA.

   External Validity of the ETVSS. We used the ICC to determine the agreement between predicted ETVSS and actual ETV success. The magnitude of the difference between the predicted ETVSS and the actual ETV success was assessed with the paired t-test. We performed linear regression analysis with actual ETV success as the dependent variable and predicted ETVSS as the sole independent variable. This provided an estimate of the amount of variation in actual ETV success that was explained by the predicted ETVSS (adjusted R^2 value).

   The ICCs are reported as follows: value (95% CI). All analyses were performed using SPSS Advanced Statistics software, version 17.0 (SPSS Inc.).

Results

The literature search identified 15 papers that met our inclusion criteria.1–11,15–18 The papers originated from Germany (4), the US (4), and Australia, Brazil, Canada, Czech Republic, Italy, Korea, and Switzerland (1 paper each). The papers were published in Childs Nervous System (7 papers), Neurosurgery (3 papers), Journal of Neurosurgery: Pediatrics (2 papers), Pediatric Neurosurgery (2 papers), and European Journal of Pediatric Surgery (1 paper) between 1990 and 2010. They reported on a total of 322 patients (a mean of 21.5 patients per study), who represented a broad range of ages, etiologies, and previous shunt status. Table 2 shows the percentage of patients within each ETVSS category.

   The actual ETV success for the 15 papers ranged from 31.3% to 92.3%. The predicted ETVSS for the 15 papers ranged from 41.3% to 85.4%.

   Interrater Reliability

   Among the 3 raters, the interrater reliability was very high for predicted ETVSS (ICC 0.993 [95% CI 0.985–0.998]) and actual ETV success (ICC 0.952 [95% CI 0.891–0.982]).

   External Validity of ETVSS

   There was consistently high agreement between pre-
Predicting success for endoscopic third ventriculostomy

Table 2: Characteristics in 322 patients who underwent ETV

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>% of Patients Included in Study</th>
</tr>
</thead>
<tbody>
<tr>
<td>age</td>
<td></td>
</tr>
<tr>
<td>&lt;1 mo</td>
<td>6.7</td>
</tr>
<tr>
<td>1 mo to &lt;6 mos</td>
<td>40.7</td>
</tr>
<tr>
<td>6 mos to &lt;1 yr</td>
<td>16.0</td>
</tr>
<tr>
<td>1 yr to &lt;10 yrs</td>
<td>19.3</td>
</tr>
<tr>
<td>≥10 yrs</td>
<td>17.0</td>
</tr>
<tr>
<td>etiology</td>
<td></td>
</tr>
<tr>
<td>postinfectious</td>
<td>6.8</td>
</tr>
<tr>
<td>myelomeningocele, IVH, non-tectal brain tumor</td>
<td>31.0</td>
</tr>
<tr>
<td>aqueductal stenosis, tectal tumor, other</td>
<td>62.2</td>
</tr>
<tr>
<td>previous shunt</td>
<td></td>
</tr>
<tr>
<td>previous shunt</td>
<td>15.7</td>
</tr>
<tr>
<td>no previous shunt</td>
<td>84.3</td>
</tr>
</tbody>
</table>

The ETSS did not perfectly predict the actual ETV success, and there are several reasons why there was still residual unexplained variation. The standards defining ETV failure and success were left completely up to the authors of the papers, and might well have varied. There were probably some inaccuracies in extracting data appropriately and accurately calculating the ETSS (which is why we ensured that all data extraction was performed by multiple independent raters). The sample size of the individual papers was often small as well (mean 21.5 patients per paper), so the chance of random variation is greater, which could lead to actual success rates that are much higher or lower than predicted. If one considers the entire cohort of 322 patients, however, the overall predicted ETSS for the sample was within <2% of the actual ETV success rate (57.9% predicted vs 59.2% actual).

Limitations of our study include the fact that we analyzed only a small number of papers from the entire ETV literature. Our selection of papers is biased toward those with smaller sample sizes because of our data extraction needs (the largest paper had 36 patients). This excluded, therefore, many of the very large published series in the literature. The papers with smaller sample sizes in our

Discussion

We have shown that the ETSS explains much of the large variation seen in selected published papers of ETV outcome in children. The current study provides, for the first time, a preliminary assessment of the external validity of the ETSS in a completely independent sample of patients derived from peer-reviewed papers published over the last 20 years from 9 different countries. These papers had actual ETV success rates ranging from about 30% to slightly over 90%. The absolute agreement between the predicted ETSS and the actual ETV success was quite high (>0.75), and the ETSS explained nearly two-thirds of the variation in actual ETV success (R² = 0.62). The corollary to this finding is the suggestion that the performance and outcome of ETV across the diverse settings represented by these papers is very similar. That is, most of the differences in ETV outcome were explained by inherent patient factors (as quantified by the ETSS) rather than extraneous factors.

We have also shown that there is very high interrater reliability (>0.95) in the assessment of the ETSS. Similarly, there was high interrater reliability (ICC > 0.95) in the assessment of actual ETV success for these same papers. The agreement was not 100%, however, and this was related to occasional ambiguities in the published papers. For example, it was sometimes unclear what ETSS category was most appropriate for a child described as “1 month old.” It was left to the rater’s discretion whether this would be categorized as “<1 month old” (and given an ETSS age score of 0) or as “1 to <6 months old” (and given an ETSS age score of 10). Similarly, even though the number of ETV failures and successes was explicitly documented, it was not always clear how many of the failures had occurred 6 months after surgery (since the ETSS is only meant to measure ETV success at 6 months). Despite these ambiguities, however, the agreement among reviewers was extremely high.

The ETSS did not perfectly predict the actual ETV success, and there are several reasons why there was still residual unexplained variation. The standards defining ETV failure and success were left completely up to the authors of the papers, and might well have varied. There were probably some inaccuracies in extracting data appropriately and accurately calculating the ETSS (which is why we ensured that all data extraction was performed by multiple independent raters). The sample size of the individual papers was often small as well (mean 21.5 patients per paper), so the chance of random variation is greater, which could lead to actual success rates that are much higher or lower than predicted. If one considers the entire cohort of 322 patients, however, the overall predicted ETSS for the sample was within <2% of the actual ETV success rate (57.9% predicted vs 59.2% actual).
study were also ones that tended to have more infants, so our sample is biased toward younger patients treated with ETV (more than one-half were younger than 1 year old; see Table 2). As mentioned above, our data extraction was limited by what could be obtained from the published paper, so it might contain inaccuracies. We did not use more traditional statistics to assess predictive properties, such as the C statistic or receiver operating characteristic analysis. Although this would have been preferable, we could not perform these statistical tests because most papers did not provide individual patient-level data and outcomes.

We developed the ETVSS primarily to help inform clinical decision-making. We have, however, used it successfully for research purposes as well. We used the ETVSS in a risk-adjusted analysis to determine the relative success of ETV performed in developed countries versus sub-Saharan Africa. The ETVSS adequately controlled for most patient prognostic factor differences between these 2 populations. We also used the ETVSS to compare outcome of ETV and shunt procedures in a large cohort. This suggested that, after adjusting for patient prognostic factors by using the ETVSS, the chance of delayed ETV failure is perhaps less than the chance of delayed shunt failure. The current study furthers our confidence in the use of the ETVSS for predicting ETV outcome in diverse settings.

Conclusions

The ETVSS closely predicts the actual ETV success reported in selected papers of childhood hydrocephalus published over the last 20 years and explains much of the variation in outcome. This provides further external validation and confidence in the use of the ETVSS in different settings.

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: Kulkarni. Acquisition of data: all authors. Analysis and interpretation of data: Kulkarni, Riva-Cambrin. Drafting the article: Kulkarni. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Statistical analysis: Kulkarni. Study supervision: Kulkarni.

References


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