ORIGINAL ARTICLE

Dysfunction of endoscopic third ventriculostomy in children

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Abstract

Introduction: Neuroendoscopy is particularly useful as an adjunct in the treatment of hydrocephalus. CSF physiology differs among neonates, children and adults leading to different and conflicting results in the treatment of hydrocephalus using neuroendoscopy. The aim of our study was to review the literature regarding to analyze predictive clinical and imaging models available and discuss specific aspects of the endoscopic approach to hydrocephalus in infants
Methods: Review of the medical literature to analyze predictive clinical and imaging models available and discuss specific aspects of the endoscopic approach to hydrocephalus children. Data of our series of ETV were analyzed. The patients were categorized in 3 groups: Group A (< 6 months of age), Group B (from 6 months to 1 year-old) and group C (>1 year-old).

Results: Group A - 12 patients , group B - 17 patients and group C - 85 patients. The etiology of hydrocephalus was tumors in 33 (29 %), aqueduct stenosis in 33 (29 %), cerebral malformations in 24 (21 %). The ETVSS in the low, moderate and high ETVSS groups was respectively 40%, 70.9% and 92.6%, the actual success rate: 58%, 65% and 86%. The complication rates in groups A, B and C were 33 %, 24 %, and 8 %, respectively (p=0.022).
Conclusion: Endoscopic third ventriculostomy provides very good results for a number of indications in children. Every effort should be made to optimize the selection of surgical candidates on the basis of their clinical features.

Key words: Neuroendoscopy, Endoscopic third ventriculostomy; Newborn; Obstructive Hydrocephalus; ventriculoperitoneal shunt

Introduction

The use of an endoscope to treat hydrocephalus is a standard technique having its origin in the early 20th century when Sir Walter Dandy began treating hydrocephalus by cauterizing or endoscopically removing the choroid plexus (1). After that, several ventriculostomy techniques were developed. In the past two decades, the introduction of new instruments such as rod lenses, Hopkins optics and high-resolution cameras has led to a significant increase in the number of neuroendoscopic procedures (2,3).

Neuroendoscopy is particularly useful as an adjunct in the treatment of hydrocephalus. It is an attractive method because it is simple, durable and eliminates the need for lifelong implanted hardware (3,4).

Although the introduction and remarkable advances in endoscopic techniques and shunt

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hardware have allowed for more effective management of hydrocephalus, consistent successful treatment of this pathology remains one of the greatest challenges in pediatric neurosurgery (5,6). Until the end of the last century just a few long-term follow-up series of ETV in patients under 1 year had been published (7–10). Controversy exists regarding whether children under 1 year of age have a higher risk of failure than older children, accompanied by an increased risk of complications and possible need for a second intervention (i.e. ETV or shunt) (11). Published case series concerning the endoscopic management of hydrocephalus in children have reported widely success rate, ranging from less than 30% to up to 100% (12–25). However, there is an extensive and conflicting literature concerning the role of patient's age and etiology of the hydrocephalus in the success rate (SR) of endoscopic procedures. The multiplicity of different outcomes that have been published brings up the heterogeneity in conducting the studies and analyzing the results (20,26,27). A better understanding of the physiopathology of cerebrospinal fluid (CSF) flow and hydrocephalus in infants has led to a redefinition of the success criteria following ETV (25,28,29).

Recently, some authors have tried to establish clinical, radiological and surgical models that could predict the likelihood of failure of the endoscopic procedure, in order to allow an optimal selection of possible ETV candidates.

Based on our own experience during the past 10 years and a review of the current literature, we raise important issues that could play a statistically significant role on the success and therefore failure rates of ETV in the treatment of hydrocephalus in young infants. We also bring some questions into perspective regarding its future management.

Material and Methods

We reviewed the medical literature, as well as our own data, in order to analyze predictive clinical and imaging models available and discuss specific aspects of the endoscopic approach to hydrocephalus in infants (30).

Data of our series of 114 ETV procedures in pediatric patients were analyzed with Fisher's exact test or chi-square test to determine whether each factor was correlated with the success of the endoscopic procedure for categorical data (SPSS version 17; SPSS, Inc., Chicago, IL). Cumulative survival rates were estimated using Kaplan-Meier methods. Log-rank test and Cox proportional hazard analysis with 95% confidence interval (CI) were used in order to access the association of the following factors with ETV failure: (1) patient's age at surgery, (2) underlying pathology, (3) birth conditions, (4) presence of previous shunt and (5) postoperative complications.

ETVSS was also retrospectively applied in our series in order to compare the predicted with our actual long-term SR. ETVSS was then calculated for each patient using data prior to the endoscopic surgery. Reoperations and cases with a follow-up of less than one year were excluded for this analysis. Kaplan-Meier survival curves as well as hazard ratios with CI 95% were used to compare the groups.

The length of follow-up was chosen as a time scale for statistical analysis. P-value < 0.05 was deemed significant. The ratios of children requiring permanent postoperative shunts or further surgical interventions in the different subgroups were compared.

The length of hospitalization and the learning curve related to the endoscopic approach were also analyzed.

Clinical indications and prognosis factors

Historically, endoscopic third ventriculostomy (ETV) always seemed to be a promising technique and can be considered nowadays a standard procedure for obstructive hydrocephalus (4,31). Lower success rates have been observed for the treatment of hydrocephalus related to other underlying pathologies, such as post-infectious, post-hemorrhagic and malformations of the central nervous system (i.e. myelomeningocele) (32). Herein, current predictive clinical, radiological and surgical models for ETV outcome are summarized. The main factors that have been discussed in the literature are the role of the patient's age group, underlying pathology, previous shunt surgery, prematurity, imaging findings and surgical technique.

ETVSS – A predictive clinical model

Kulkarni et al. (2009) evaluated the influence of the patient's age at surgery, cause of hydrocephalus and the presence of previous shunt as clinical factors to create "the ETV Success Score" (ETVSS) (33). The Score was first tested by the same group and has been externally validated by many other centers worldwide (34,35).

The score is based on patient's age at surgery, etiology of hydrocephalus and presence of previous shunt. Patients are then supposed to have low (≤40), moderate (50-70) or high (≥80) probability of success after ETV. The score ranges from 0 to 90 and approximates to the percentage chance that an ETV will be successful at 6 months, as previously described elsewhere (16). In a retrospective review of 151 consecutive pediatric patients who underwent ETV, Naftel et al. (2011) assessed patients' clinical features, radiological and operative findings, complications, and compared ETV actual success with those predicted by the ETVSS model. The C-statistic was 0,74 (95% CI

0.65-0.83) and suggested that ETVSS is accurate in discriminating failure from success. The actual success rate at 6 months follow-up was 68% compared to the predicted SR of 76.5% +- 12.5% (SD) (35). García et al. (2012) published similar results in a series of 50 children with a mean follow-up of 33.92 months (ranged from 6-92) (36). For those patients who underwent successful ETV, the mean ETVSS was 71.03 (95% CI, 66.23-75.84), and in those for whom the ETV was not successful, the mean ETVSS was 60 (95% CI, 53.09-66.90). They stated that the score may help surgeons to establish improved selection criteria and also advise patients and families about expected outcomes. Although the score was originally developed to predict 6-month outcome, the authors and other groups have recently demonstrated its application in anticipating ETV long-term outcomes (34–36). Durnford et al. (2011) retrospectively analyzed 181 consecutive cases of ETV performed in children and concluded that the ETVSS accurately predicted outcome at 36 months. In that series, the low, medium and high chance of success strata had mean predicted probabilities of success of 82%, 63% and 36%, and actual SR of 76%, 66% and 42%, respectively (34). Once again, the mean probability of success was significantly higher in those with a successful ETV than in those with a failed ETV, (p=0.001).

ETV Operative Technique

The surgical techniques are well established and were developed based on a detailed understanding of third ventricular anatomy, surgical trajectories and improved technology. Several methods have been proposed and been described in detail elsewhere (37,38). Indeed, surgical techniques vary depending on teams' preferences and availability of more sophisticated equipment. In our series, operations were done using a rigid GAAB Karl Storz® neuroendoscopic system equipped with a No. 8 French diameter Hopkins rod lens system with a 0˚ optic, a No. 3 French working channel and an irrigation channel produced by Storz® (Tuttlingen, Germany). All procedures are performed free hand after the induction of general anesthesia.

Postoperative follow-up

Routine postoperative outpatient follow-up appointments were scheduled within one week, and then one month, three months and every six months. Success was defined by the following criteria: when no further intervention was required to treat hydrocephalus and the absence of signs or symptoms of raised intracranial pressure (30,39).

Results

The following data summarize the results from our complete series of 114 consecutive ETVs performed as a retrospective, unblended study, concerning short-term results and long-term success rates of the endoscopic treatment of hydrocephalus in children (30).

Clinical Results

A total of 114 patients were studied. There were 49 male patients (43 %) and 65 female patients (57 %) ranging in age from 11 days to 18 years (mean age, 6.17 ± 1.02 years). These patients fell into three groups. In Group A the 12 patients (mean age 2.75 ± 0.89 months) were < 6 months of age, in-group B the 17 patients were between 6 months and one year of age (mean age 7.82 ± 1.12 months) and in-group C the 85 patients were older than one year (mean age 8.28 ± 1.03 years).

The etiology of hydrocephalus was as follows: tumors in 33 (29 %), aqueduct stenosis (AS) in 33 (29 %), cerebral malformations in 24 (21 %), cystic malformations in 6 (5.3 %), intraventricular hemorrhage in 5 (4 %), meningitis or ventriculitis in 3 (2.6 %) and other etiologies in 10 (9 %) patients.

In all cases ETV was performed as a single and straightforward procedure and the main goal of the endoscopic intervention was to restore the cerebrospinal fluid (CSF) flow pathways. Thirteen patients (11.4%) were preterm. Twenty-nine procedures (25%) were performed in patients under one year of age.

The overall SR for CSF circulation restoration was 80% (91 patients).

Table 1 summarizes clinical features of the patients, SR and complication rate (CR) of ETV in this series according to the CSF restoration.

Age group and SR

According to the age group, we observed 58 % (7/12) SR in group A; 65 % (11/17) in group B, and 86 % (73/85) in group C (p=0.02). The long-term outcome also varied with age.

Table 2 represents a bivariate logistic regression of risk factors associated with ETV failures for different group of patients.

Patients of group A and B had respectively a 4.3-fold (95% CI 1.1-15.9, p=0.02) and a 3.3-fold (95% CI 1.0-10.6, p=0.04) increased risk of ETV failure compared to patients of group C.

Table 1 - Cerebrospinal fluid circulation restoration procedures in 114 patients; SR and CR calculated according to the number of patients and number of procedures

Factor	N (%) Out of 114 patients	SR	p Value SR	CR	p Value CR
Sex			p=0.431		p=0.516
Male	49 (43%)	81.6% (40/49)		12.2% (6/49)	
Female	65 (57%)	78.5% (51/65)		13.8% (9/65)	
Age			p=0.02		p=0.02
<6 months	12 (10.5%)	58% (7/12)		33.3% (4/12)	
6months	17 (15%)	65% (11/17)		23.5% (4/17)	
1year	85 (74.5%)	86% (73/85)		8.2% (7/85)	
>1year					
Birth Conditions			p=0.006		p=0.001
Term	49 (43%)	83.7% (41/49)		6.1% (3/49)	
Pre-term	13 (11.4)	46.2% (6/13)		53.8% (7/13)	
Previous Shunt			p=0.023		p=0.286
Yes	21 (18.4%)	61.9% (13/21)		19% (4/21)	
No	93 (81.6%)	83.9% (78/93)		11.8% (11/93)	
Underlying Pathology			p=0.001		p=0.017
Tumors	33 (28.9%)	90% (30/33)		9.1% (3/33)	
AS	33 (28.9%)	88% (29/33)		6.1% (2/33)	
Brain Malformations	24 (21.1%)	79.2% (19/24)		20.8% (5/24)	
Cystic	6 (5.3%)	83.3% (5/6)		0	
Lesions	5 (4.4%)	60% (3/5)		20% (1/5)	
Hemorrhage	3 (2.6%)	0		33% (1/3)	
CSF infection					
Learning Curve			p=0.017		p=0.009
First period	24 (21%)	62.5% (15/24)		29.2% (7/24)	
Late period	90 (79%)	84.4% (76/90)		8.9% (8/90)	
ETVSS			p=0.001		p=0.033
High	54 (47.3%)	92.6% (50/54)		5.6% (3/54)	
Moderate	55 (48.2%)	70.9% (39/55)		20% (11/55)	
Low	5 (4.3%)	40% (2/5)		20% (1/5)	

Values represent number of patients (%) unless stated otherwise; SR= success rate; CR= complication rate; CSF= cerebrospinal fluid; AS= aqueductal stenosis

Hydrocephalus etiology and SR

In Group A the etiology of hydrocephalus was related to AS in 5/12 (42 %) cases. Complex cystic lesions, arachnoid cysts and brain or spinal malformations (dysraphism, Dandy Walker, Chiari malformation) were each observed in 3 cases (25 %). Out of the 17 patients in group B, malformations were found in 6 (35 %), AS in 4 (24 %), hemorrhage in 2 cases (12 %), and infection in one case. In group C an obvious predominance of pure obstructive hydrocephalus [i.e. posterior fossa tumors and AS] was observed in comparison to the other groups [Group A, B and C: 41 % (5/12), 24 % (4/17), and 67 % (57/85) respectively], (p=0.001).

The overall analysis according to etiology showed a success rate of 90 % (30/33) in hydrocephalus associated to posterior fossa tumors, 88 % (29/33) in AS and 83 % (5/6) in cystic lesions. Lower success rates were observed in cases of myelomeningocele, intraventricular hemorrhage and ventriculitis (p=0.001).

Table 2 - Bivariate logistic regression, adjusted HRs, 95% CIs, and p value for risk factors associated with ETV failure

Factor	Bivariate Analysis		
	HR	CI 95%	p Value
Sex			
Female	1		
Male	0.8	0.3-2.0	p=0.67
Age			
>1 year	1		
6 months- 1year	3.3	1.0-10.6	p=0.04
<6 months	4.3	1.1-15.9	p=0.02
Birth Conditions			
Pre term	6.4	1.7-24.1	p=0.006
Previous Shunt	3.2	1.1-9.0	p=0.02
Underlying Pathology			
AS	1		
Tumors	0.7	0.1-3.5	p=0.69
Brain Malformations			
Cystic Lesions			
Hemorrhage	1.9	0.4-8.0	p=0.37
CSF infection	1.4	0.1-15.7	p=0.76
	4.8	0.6-38.3	p=0.13
	7.2	1.4-36.6	p=0.01
Complication	13.2	3.8-44.8	p=0.000
Hemorrhage	13.2	2.1-79.6	p=0.005
Infection	6.6	0.3-112.3	p=0.19
CSF leak	13.2	1.1-156.4	p=0.04
Learning Curve			
SecondPeriod	1		
First period	3.2	1.1-8.8	p=0.021
ETVSS			
High	1		
Moderate	5.1	1.5-16.5	p=0.006
Low	18.7	2.3-146.9	p=0.005

HR= Hazard ratio; CI= confidence interval; AS= aqueductal stenosis; CSF= cerebrospinal fluid

Poorer outcomes were more frequent in premature infants. Preterm birth was associated to a 6.4-fold increased risk of ETV failure when compared to their full-term counterparts (95% CI 1,7-24,1; p=0.006).

Among patients with AS there was no statistical difference between the age groups: group A 60 % (3/5), group B 75 % (3/4) and group C 92 % (22/24) (p=0.104).

The mean interval between ETV and failure was 4 months (ranging from 15 days to 9 months).

Re-ETV was performed due to failure of the first attempt in two patients. Twenty-one out of 114 (18 %) ETVs were done in children that had been previously shunted. Previous shunt-surgery was strongly correlated with failure of the endoscopic procedure.

The mean follow-up period was 65 months (ranging from 10 months to 9 years). A Kaplan–Meier analysis illustrates that the proportion of functioning ETVs became stable at rates of 58 to 86% after the 1st postoperative year in different age groups (Log Rank (Mantel-Cox); Chi-Square= 7.639, p = 0.006).

ETV Success Score

In the present series ETVSS was retrospectively applied in order to evaluate its suitability for long-term (>12 months) SR prediction. The predicted chance of SR in the low, moderate and high ETVSS groups was respectively 40%, 70.9% and 92.6%. Those values are close to the actual following SRs for groups A, B and C: 58%, 65% and 86%. The low and moderate ETVSS groups were found to show respectively an 18.7 and 5.1-fold increased risk of failure of the endoscopic procedure than the high ETVSS group (CI 95% 2.3-146.9, p=0.005 and CI 95% 1.5-16.5, p=0.006).

For the group in which ETV was successful, the mean ETVSS was 74.8, variance 14.35 (95% CI 69.37-78.22). The mean ETVSS in the group that did not show a favorable outcome was 63.04, variance 14.74 (95% CI 59.67-68.1). Therefore, ETVSS accurately predicted the overall long-term success rates in our population.

Learning curve and complication rate (CR)

The overall CR in this series was 13 % (15/114). They included: intraventricular hemorrhage in 6 patients, infection in 5 [i.e. meningitis (2), ventriculitis (3)], CSF leakage in 3 and transient dysphasia in one case. The mortality rate was 1,7 %. One patient developed severe ventriculitis, and another patient died due to respiratory complications in the postoperative period. Two patients (1.8 %) developed epilepsy and needed further treatment after the endoscopic procedure. The complication rates in groups A, B and C were 33 %, 24 %, and 8 %, respectively (p=0.022).

Analyzing the outcome in two different periods of time (between 2000-2004 and 2005-2010), an improvement in the success rate of the neuroendoscopic procedures can be clearly seen [i.e. from 63 % (15/24) to 84 % (76/90)] (p=0.017). We observed a significant CR reduction in the same period (29 % and 9 %, p=0.009).

The overall length of hospitalization was less than 3 days in 68 % and less than 4 days in 90% of the patients.

Discussion

Over the last few years, intracranial neuroendoscopy, particularly ETV, has found its place in pediatric neurosurgery. Current experience throughout the world shows that this treatment is a good alternative to shunts in many cases of brain disease, and particularly in obstructive hydrocephalus. ETV is considered to be a simple, fast, and safe procedure in children (3,8,14,20,40,41).

In this single-center experience of 116 consecutive ETV procedures performed in children, the overall SR to restore CSF circulation was 80 %, and the absolute complication rate was 13 %. These data are in accordance with those reported in the literature (26,31,41–43).

Despite technological advances in the last century concerning the management of hydrocephalus, the issue about timing of ETV and its effectiveness in younger patients is still controversial (4,7,8,16,20,21,25,26,44).

Also, there is no consensus regarding the appropriate age of patients to be treated with ETV. In most series younger children were not included. Thus, among the authors who have defended ETV, some suggest that it should be attempted only in children older than one year (9,17). The concept of communicating versus obstructive hydrocephalus has been challenged. Controversial issues reside mainly on the fact that the CSF hydrodynamics changes during development and therefore are not identical to adults in the neonatal and infantile periods (25,28,29). The lower SR after ETV in younger children may be explained by the poor CSF reabsorption capacity of newborns due to immaturity of the arachnoid granulations. In addition, the anterior fontanelle is widely opened and the sutures are splayed in infants, contributing to the maintenance of a low intracranial pressure (9,10,13,29).

On the other hand, some authors advocated that ETV has the same long-term results in children younger than 6 months and older children; thus, patient age should no longer be considered a contraindication to using the technique (45–47) and, in the case of delayed failure (usually secondary to

obstruction of the stoma), this can often be managed by repeating the procedure (44–46).

The literature review showed that SR in children under 6 months of age ranged from 32% to 44.9 %, and in children older than one-year age it ranged from 56 % to 71 % (31,33–35,40,42).

Univariate and multivariate analysis showed in our series that both the etiology of hydrocephalus and patient´s age were relevant factors predicting success. These results are in accordance with other authors (8,14,20,26,36,46–48).

Indeed, an overall increased risk of ETV failure in children younger than 1 year was observed, but if stratified by age and underlying pathology, logistic regression showed that intraventricular hemorrhage, myelomeningocele and previous CSF or shunt infection were also strongly associated with failure of the ETV. On the other hand, no statistical difference on outcome was observed among patients with AS, regarding to age group (younger or older than 1 year) (p=0.104). Some authors also reported similar results analyzing outcome and underlying pathology (7,8,10,49). Likewise, presence of previous shunt, preterm birth and postoperative complications [i.e. infection, intraventricular hemorrhage and CSF leakage] are important factors contributing to poor ETV outcome.

Kulkarni et al. proposed the ETVSS aiming to predict the 6 months follow-up outcome of patients submitted to ETV (16). Later on, ETVSS has also been proven to be applicable in order to predict long-term outcome (34–36,50).

In our series the overall predicted and actual long-term SR were clearly correlated (respectively 74.8% and 79.8%, p<0.001). Moreover, the Cox proportional hazard ratio analysis undoubtedly indicated an increased relative risk of ETV failure for the “low” and “moderate” ETVSS groups. Both Kaplan-Meier survival curves explicit a similar proportion of functioning ETVs after the 1st postoperative year in different groups. Even though the Log-rank analysis showed poor cumulative survival for both “Group A” (<6months) and “low ETVSS”, the latter predicted even worse outcome (respectively 58% and 40%). Furthermore, the relative risk predicting ETV failure was much higher when considering associated factors by means of the ETVSS (HH for the “low ETVSS”: 18.7, CI 95% 2.3-146.9, p = 0.005) than just the patient’s age group (HH for “Group A”: 4.3, 95% CI 1.1-15.9, p = 0.02). These data strengthen the importance of several different factors other than age on ETV outcome.

We are surely in agreement with accurately selecting patients in whom ETV is more likely to be successful. In this regard, the ETVSS seems to be easily applicable and could also prepare patients and their families for expected outcomes. Nevertheless, factors that do not count for ETVSS, such as prematurity and postoperative complications, showed high hazard ratios and may also be taken in consideration when predicting ETV failure.

Costs

Garton et al. (2002) in a retrospective analysis of 28 children who underwent ETV failed to prove cost-effectiveness of this therapy when compared to shunt placement (51). In that series the authors showed a 54% ETV success rate over a median follow-up of 35 months. However, as reported in the same paper the short period of observation after surgery clearly represents only a small window of time in the life of those patients. If late failures after ETV are much rarer than CSF shunts, longer follow-up would presumably improve the cost-effectiveness ratio of ETV. Other than that, the reported overall success rate of 54%, even for a pediatric cohort, is much lower than what has been recently published in the literature, and probably this might have increased the costs of patients treated with neuroendoscopy. Other authors are in agreement that in a long-term analysis, when length of hospitalization, operating room time, need for reoperation, readmission and postoperative complications are taken into account, ETV seems to be more advantageous than shunting; nevertheless, an accurate patient selection is mandatory (52).

Surgical Complications and Learning Curve

As previously reported in the medical literature, another factor that influenced the outcome in this series was surgical experience (36,43). Patients operated on during the first period of this series (2000-2004) had a 3.2-fold (95% CI 1.1-8.8, p=0.021) increased risk of ETV failure in comparison to those treated more recently.

We also observed a remarkable reduction of the complication events after ETV procedures in two different periods analyzed (from 29 % to 9 %, p=0.009). According to the literature review the overall CR ranged in individual series from 2 % to 44.9 % (2,31,33–35,40,43). Recently, Bouras & Sgouros published an extensive review of complications associated with ETV (53). Their analysis included 2985 ETVs performed in 2884 patients and they concluded that ETV can be regarded as a low-complication procedure, with an overall complication rate of 8.5 %, permanent morbidity rate of 2.4 %, mortality rate of 0.21 %, and delayed “sudden death” rate of 0.07 %.

What to do when ETV fails?

Recently many authors agree that re-ETV should be considered in carefully selected cases of failed ETV, prior to VP shunt insertion (13,25,49,54,55). Usually the mechanism of failure is closure of the stoma due to local inflammatory reaction and its incidence is also related to the underlying pathology. Wagner and Koch (2005) observed three different patterns of endoscopic findings by repeating ETVs in

cases that had initially failed: 1) occlusion of the ventricular stoma; 2) narrowing of the stoma; 3) patent stoma with newly formed arachnoid membranes in the basal cisterns below the floor of the third ventricle (13). They concluded that concerning ETV failure, in cases of obstructive hydrocephalus, the formation of new arachnoid membranes or scars are more relevant factors than poor CSF absorption. They hypothesized that infants have a higher tendency to form new membranes than older children, so this may explain the higher ETV failure rates in patients younger than 1 year. However, the review of the current literature shows that the success rate of re-ETV can be high, ranging from 13 to 90%, even in children under 2 years old. As well as in primary ETV, patients with previous CNS infection, hemorrhage or foreign body within the ventricle have poorer outcomes after the second attempt (56). Reclosure of CSF pathways occurs not only at the floor of the third ventricle but also at a lower level in the basal cisterns, even though the stoma itself might remain patent (13). The mean interval between the first attempt of ETV and the re-ETV ranged in the literature from 6 days to 36 months. Finally, authors have agreed that a T2 fast spin echo or two-dimensional phase contrast MRI are both indispensable for an accurate evaluation of ETV failure. Then, patients in whom no flow through the ventriculostomy can be demonstrated should be submitted to endoscopic exploration. A repeat ETV is indicated in patients with a closed stoma. The others might require insertion of a cerebrospinal fluid shunt (45). Although re-ETV has been shown to be a plausible alternative with encouraging results in the management of unsuccessful ETV cases, the main cause of poor outcome in infants remains an open question and more investigation is needed.

In the present series reoperations due to failure of the first attempt of endoscopic procedure were observed in only two cases, both procedures were successful, and the patients required no further intervention. These results are similar to other series (44).

Therefore, despite the fact that some patients suffering from reocclusion of the ventriculostomy might have to undergo shunting, several authors consider well worth trying a repeat ETV in selected cases (17,44,45,57).

Conclusions

Endoscopic third ventriculostomy provides very good results for a number of indications in children. Tumor-related CSF circulation problems and AS seem to be particularly well suited for ETV regardless of patient's age. Intraventricular hemorrhage, previous CNS infection, myelomeningocele, prematurity and the presence of previous shunt have been associated with a high failure rate in all age groups, particularly in infants under 6 months of age. Complication rate reduction was associated with accumulated surgical experience. The ETVSS was suitable to predict long-term outcome in our series, but further refinement and prospective validation of this model may be required. Every effort should be made to optimize the selection of surgical candidates on the basis of their clinical features.

Conflict of interest

The authors declare that they have no conflict of interest related to this article.

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