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REVIEW PAPER

Third ventriculostomy in a single pediatric surgical unit

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Abstract
Purpose Endoscopic third ventriculostomy (ETV) is a successful method of treatment for obstructive hydrocephalus that has become popular over the last 20 years. The purpose of this paper is to study the outcome of infants with obstructive hydrocephalus treated by ETV by a single surgeon and to evaluate the safety, reliability, and efficacy of this treatment.
Methods All data were collected retrospectively. Between July 1999 and June 2005, 14 children underwent an ETV. In one child, a second ETV was performed. The age of the eight female and six male patients at the time of ETV ranged from less than 1 month up to 13 years and 11 months. The indication for an ETV was an obstructive hydrocephalus. Median follow-up period was 5 years and 9 months. The need of a further operation after ETV was defined as a failure of ETV.
Results In six patients, the first ETV was successful. In the remaining eight patients, there was a need for further treatment (ventriculoperitoneal shunt). Although the follow-up shunt failed in one patient, he was successfully treated by a second ETV.
Conclusion Our study suggests that ETV can be successfully done in a small pediatric unit, but with a lower success rate because of small caseload, and therefore, lower experience and routine of the surgeon. Therefore, we propose a centralization of patients to obtain a higher number of cases. We confirm that ETV is a safe, reliable, and efficient method with a better outcome in children than infants.

Keywords Endoscopic third ventriculostomy · Hydrocephalus · Infants · Outcome

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Introduction

Endoscopic third ventriculostomy (ETV) as an alternative to ventriculoperitoneal (vp) shunting has become more important over the last 20 years, as long-term reliability of a vp-shunt is disappointing due to multiple malfunctions, such as mechanical failure and infection [5, 11]. To perform an ETV in obstructive hydrocephalus requires the perforation of the floor of the third ventricle, to create a new orifice that enables a natural outflow of cerebral fluid. The objective of this paper is to study the outcome of infants with obstructive hydrocephalus treated with ETV in a single pediatric unit, where only one surgeon is profound-trained and performing ETVs. Furthermore, this paper is aimed at evaluating the safety, the reliability, and the efficacy of this treatment in such a setting and also to compare the results with other studies with higher number of cases.

Patients and methods

Between July 1999 and June 2005, 14 children with obstructive hydrocephalus underwent an ETV at the pediatric surgical department of the University Children's Hospital Zurich. One child had a second ETV. The children (eight girls, six boys) were aged between less than 1 month

and 13 years 11 months (median: 3 years 4 months). Etiology of hydrocephalus was variable: cyst (n=4), tumor (n=4), Chiari II malformation, and myelomeningocele (n=3), aqueductal stenosis (n=3). Four patients had undergone previous vp-shunt surgery before receiving an ETV. All patients were followed-up with clinical examination and imaging with a median follow up period being 5 years and 9 months. (Table 1) Technically, the intervention was done either with a rigid endoscope (0 degree optic, Storz, Germany) or with a rigid one-way endoscope (0 degree optic, Medtronic, USA). The operation was performed as described in "The manual of endoscopic procedure in neurosurgery" by J.A. Grotenhuis [8].

Results

ETV was successful in six out of 14 patients without the need of further surgical intervention. Four of those six were more than 13 months old at the time of ETV. The other two successfully treated patients were 1 month and 5 months old. In eight patients, ETV was not sufficient, seven children needed a vp-shunt, one child with a remaining shunt requiring revision. Failure of ETV was due to obliterated ETV (n=2), insufficient Cerebrospinal fluid (CSF) absorption (n=4), insufficient flow through the ETV (n=1), and intraventricular bleeding followed by external drainage of CSF (n=1). The failure rate was eight out of ten patients of those younger than 13 months, compared to a zero failure rate in the four children older than 13 months. There was no significant difference in failure rate among the different aetiologies of hydrocephalus. One boy aged 2 months had a second ETV at the age of 3 years and 10 months. During the first ETV, intraventricular bleeding occurred and was treated with an external drainage followed by a vp-shunt insertion. Later, the boy presented with shunt dysfunction and the parents agreed to a second ETV, up to now, this has been successful. No mortality or permanent morbidity followed from the first or second ETV. There were five cases with minimal intraoperative bleeding, resolving spontaneously without interfering with the procedure. In two cases, bleeding was more significant, but not severe, and in one of these two cases, ETV was carried out. In the other case, ETV could not be performed, but in both cases an external drainage was inserted. Postoperative course revealed no complications in five patients. Three patients showed the following temporary symptoms: headache, strabism, and paresis of the oculomotoric nerve. In one case, these symptoms were caused by overdrainage of the remaining shunt. In five other patients, postoperative increase in head circumference, sunset phenomenon, and intensified reflexes suggested that the benefit of ETV was insufficient, or that ETV was unsuccessful. The patient with the abandoned ETV due to intraventricular bleeding displayed no complications in his postoperative course. In the other child with intraventricular bleeding on whom ETV was performed, the drainage blocked and one insertion was put in place. Subsequently, the patient developed epilepsy. Neurological follow-up showed normal findings in six patients. In the other eight patients, all symptoms were known preoperatively, or if noted postoperatively not related to the procedure itself. Thirteen of 14 patients attend normal school education. The remaining patient attends special education for disabled children. Five patients require supportive therapy. Only two patients require a regular intake of medication, namely anticonvulsive medication (Sultiam). In one child, epilepsy is caused by a brain tumor, in the other child, epilepsy presented for the first time after changing his external ventricular drainage as described above. Currently, all successfully treated patients with ETV magnetic resonance imaging (MRI) show a flow void through the floor of the third ventricle, but also four children with failed ETV demonstrate a flow void in MRI.

Discussion

Success rate

Six of 14 patients were successfully treated by their first ETV in our institution, with one patient having successful repeated ETV. MRI results reveal that in 11 children these interventions are technically correct. Literature shows a success rate of 60–90% or higher for carefully selected patients [3, 6, 7, 9, 11, 14, 15]. Our success rate is lower, however, the number of our patients is too small to make a conclusive statement. Furthermore, and importantly, we treated children only, with nine of the 14 patients being younger than 1 year. Another factor explaining our lower success rate compared to the literature might be the fact that a single surgeon operating on a few patients a year may take longer to develop expertise. On the other hand, there has to be considered that there was no third ventriculostomy done in the German-speaking part of Switzerland until 1999. So, one might also argue that this study reflects a learning curve. But as already mentioned above, the number of cases in this study is too small for any conclusive statements.

What influences the failure rate?

It is well-described in the literature that age is an important factor in the failure and long-term reliability of ETV, with reliability being lower in very young children [7, 9, 10].

Table 1 Description of patients

Patient	Sex	Age	Diagnosis causing obstructive hydrocephalus	Procedures (chronically listed)	Complications disturbing the ETV and other severe complications	Outcome
1	Female	3 months	Arachnoid cyst	History of cystoperitoneal shunt History of shunt revision ETV, no flow, obliteration Vp-shunt	None	Regular education, no medication, no neurological problems related to ETV Follow-up 8 years 2 months
2	Male	5 months	Fibrillous astrocytoma	ETV	None	Regular education, no medication, no neurological problems related to ETV Follow-up 7 years 10 months
3	Female	5 months	Thalamic tumor	ETV, normal flow, too low resorption Vp- shunt, shunt dysfunction and revision	None	Regular education with special support, no medication, no neurological problems related to ETV Follow-up 7 years 1 month
4	Female	1 month	Chiari II malformation	ETV	None	Regular education, no medication, no neurological problems related to ETV Follow-up 6 years 10 months
5	Female	1 year 1 month	Chiari II malformation	History of shunt insertion and dysfunction ETV, normal flow, too low resorption Vp-shunt revision	None	Regular education, no medication, no neurological problems Follow-up 6 years 10 months
6	Female	1 month	Membranous aqueductal stenosis	ETV, normal flow, too low resorption Vp-shunt, shunt infection and revision	None	Regular education, no medication, no neurological problems related to ETV Follow-up 6 years 2 months
7	Female	2 months	Blake's pouch cyst	ETV, insufficient flow Vp-shunt, overdrainage due to broken valve	None	Regular education with special support, no medication, no new neurological problems related to ETV Follow-up 6 years 2 months
8	Female	8 months	Retrocerebellar cyst	ETV, no flow, obliteration Extraventricular drainage Vp-shunt, shunt dysfunction, shunt revision	Intraoperative bleeding, extraventricular drainage insertion	Regular education, no medication, no neurological problems Follow-up 5 years 9 months
9	Male	13 years 11	Tectal glioma	History of shunt insertion, shunt	None	Regular education with special support, no medication, no neurological problems related to ETV

Table 1 (continued)

Patient	Sex	Age	Diagnosis causing obstructive hydrocephalus	Procedures (chronically listed)	Complications disturbing the ETV and other severe complications	Outcome
		months		dysfunctions ETV		Follow-up 5 years 7 months
10	Male	1 month	Chiari II malformation	ETV, normal flow, too low resorption	None	Special education, no medication, no neurological problems related to ETV
				Vp-shunt,		Follow-up 5 years 7 months
11	Female	12 years 1 month	Idiopathic aqueductal stenosis	ETV	None	Regular education, no medication, no neurological problems
						Follow-up 5 years
12	Male	2 months	Membrane in the region of the foramina Megendii	Abandoned ETV	Intraoperative bleeding, interruption of ETV, extraventricular drainage insertion	Regular education, no medication, no neurological problems related to ETV
				Extraventricular drainage Vp-shunt, shunt dysfunction Second ETV		Follow-up 4 years 9 months
13	Male	4 years 6 months	Arachnoid cyst	History of cyst fenestration	None	Regular education with special support, no medication, no neurological problems
				History of extraventricular drainage History of Vp-shunt ETV		Follow-up 2 years 6 months
14	Male	13 years 8 months	Pilocystic astrocytoma	ETV	None	Regular education, no medication, no neurological problems related to ETV
						Follow-up 2 years 3 months

Success rate is lower in the first year of life than in older children. A low capacity of CSF resorption and a higher tendency to form new arachnoid membranes are proposed as causes of this age dependency [2, 10]. Although our study group is small, we also recorded a higher failure rate in infants less than 13 months. The fact that four of these patients developed progressive hydrocephalus, despite a patent ETV and three of these showed obstruction (two cases), or insufficient flow through the ETV (one case), seems to confirm this hypothesis. The influence of the etiology of hydrocephalus on the outcome is controversial. It appears that there are no significant differences in outcome as long as ETV is performed in a noncommunicating hydrocephalus. Kadrian et al. found no differences in success and reliability of ETV in particular diagnostic groups although [9]. Feng et al. conclude that ETV is most efficient in obstructive hydrocephalus due to aqueductal stenosis, cysts, and tumors [6]. Our patients show the lowest failure rate in children with hydrocephalus due to brain tumor (zero), however, it has to be considered that in our small study patients with tumors were older than the patients with other etiologies of hydrocephalus. An important influence on the outcome is the operative course [14]. Schröder et al. state that the rate of abandoned procedures ranges from 0 to 26%, mainly due to hemorrhages, anatomical variations, and inability to perform the fenestration of the third ventricle floor. They reported a rate of 0.5% of abandoned procedures. We had one interruption caused by bleeding; no anatomical variations or inability to fenestrate the ventricle floor presented a limiting factor intraoperatively. However, we did find such anomalies in preoperative MRIs in potential ETV patients. The mortality in our study was zero, other studies report a mortality of 0–1% [1, 5, 11, 14, 15]. The main mortality risk factor is perforation of the basilar artery resulting in a massive intraoperative bleeding. Controversy surrounds discussion on the influence of persisting vp-shunts. In one study, all patients with previously inserted shunts failed the ETV procedure, but all were premature babies [2]. It is open to debate whether a vp-shunt is a risk factor for ETV in premature babies or whether prematurity itself is the risk factor. Most other studies conclude that a history of shunt does not influence outcome [3, 7, 9]. Our results match the literature. Despite the small number of patients, we found no lower success rate in children with previous shunt surgery. Failure of ETV generally occurs within the first year after the procedure, and usually after 5 years, no further failures are detected [5–7, 9, 15].Our study concurred with this finding. In two cases, the failure occurred outside of the 12-month period, (at 25 and 46 months), but never later than 5 years postoperatively. In both late cases, the infants were very young at the time of operation (2 and 3 months). Contrary to this, the literature states that failures in ETV occur early after the intervention in very young babies due to lower reliability. We have no explanation for these two late failures in our patients. It is important not to miss late failures as they can be lethal.

As mentioned above, another point to discuss is whether such a small caseload, as in our small series, influences failure rate and outcome. We found a lower success rate as published in the literature, but we had no more severe intraoperative complications having two bleedings with need for extraventricular drainage. Concerning the other intra- and postoperative problems, we were very honest and listed also the least aberrance, as five minimal intraoperative bleedings solving already intraoperatively spontaneously. In the overall outcome, we had no neurological findings related to the ETV itself. The neurological symptoms we found were preexisting due to hydrocephalus or underlying disease. On the other hand, it is a matter of fact that a higher number of patients per year give a surgeon more experience and routine which helps to keep the failure rate small.

Repeated ETV

Siomin et al. have written that a repeated ETV is at least as safe and effective as the first. The selection of patients for a repeated ETV should include the same criteria as recommended for the first [15]. Having followed this guideline, we successfully did a second ETV in one patient. Lack of both an MRI and a trained surgeon being available to perform ETV 7×24 h, 365 days a year, might be another reason we have performed only one repeat ETV, thus far. Furthermore, we carefully assess indication of ETV in all hydrocephalic children before planning a first intervention.

Shunt vs. ETV

Over 8 years we have performed 15 ETV procedures (including two ETVs on one patient). In the same time period, we performed 136 vp-shunts. The main advantages of shunting are that it can be used for all types of hydrocephalus, it is technically straightforward, has been performed in a standardized way over many years, and, additionally, the mortality rate is very low. The benefits of ETV are that all main complications of shunting, such as foreign bodies in the organism, mechanical problems such as disconnection, occlusion, and valve dysfunction can be avoided. Also the risk of infection is significantly lower. Furthermore, ETV restores a kind of physiological liquor circulation without the risk of overdrainage [5]. A shunt often requires more than one intervention, 50% of the inserted shunts fail within 2 years, and 70% within 10 years, as stated by Schroeder et al. in their meta-analysis [14].

Four out of five studies in this meta-analysis were pediatric. De Ribaupierre et al. found that the number of revisions per patient is higher in the group of shunted children: failure rate for ETV at 2, 5, and 10 years was 26, 26, and 30% compared with the failure rate of vp-shunting of 34, 42, and 50% [14]. The reliability of shunting in children, especially in those up to 2 years is lower than in adults [5]. Di Rocco et al. describe a complication rate of 50% in children treated within the first 2 years compared to 30% in those older than 2 years. A higher failure rate of ETV in children is likely to be more acceptable as the complication rate of shunt insertion is also increased. Disadvantage of ETV is a higher risk of intraoperative complications such as injury of the basilar artery or its branches, or damage of nerve structures by the endoscopic instruments. The risk of perforating the basilar artery is lower when the floor of the third ventricle is thin and the structures underneath can be identified. If ETV is performed correctly and the patients are well selected it is a safe, simple, and fast option [6, 14, 15]. It should be considered for all patients, especially children, with obstructive hydrocephalus.

Outcome

In the literature, neurological damage (excluding hypothalamic damage) after ETV is found in 3–4% of cases [5]. Di Rocco et al. state that the actual incidence of neurological complications may be underestimated, as most of these neurological injuries are of short duration, and therefore often unreported. Frequent complications include impairment of short-term memory, other memory dysfunctions, confusion or unconsciousness, postherniation syndrome, aggressive behavior, and disinhibition, impairment of motor skills, hemiparesis, oculomotor palsy, and diabetes insipidus [5]. Epilepsy in children with hydrocephalus is a frequent disorder with an incidence of 15–50% [5, 13]. Epilepsy after ETV caused by multifactorial etiology is rare with an incidence of 0–1% [5]. The endoscope itself is a generally accepted risk factor for epilepsy in ETV. The evidence in literature suggests that the prevalence of seizures in shunted children with a permanent catheter is higher than in the nonshunted patients, and that ETV can actually reduce the risk of epilepsy [5]. In our study, 11 patients showed no neurological findings caused by hydrocephalus or its treatment, while three patients presented with symptoms due to hydrocephalus. In our small sample, we recorded no children with long-term neurological complications related to ETV itself. All long persistent neurological symptoms in our study were caused by the underlying condition or related to procedures other than ETV. Long-term outcomes show that 13 patients are able to attend normal school education. One child with a parenchymal lesion (diagnosed before ETV was done) requires special education, confirming that preexisting parenchymal lesions in neuroimaging are related to impairment in neurological outcome and educational outcome [12]. For the majority of patients, the most important factor in neurological outcome and education are neurological impairments not related to hydrocephalus and its treatment. To our knowledge, no study shows a difference in neurological outcome between ETV and shunts in children. On the other hand, it is well-known that untreated hydrocephalus leads to loss of white and gray matter and to neurological deficits.

Conclusion

Our caseload is too small to allow any significant results or conclusions. Nevertheless, we found according to the literature, that ETV fails more often in very young children but that there are also successfully treated infants. Most ETV fail within the first postoperative year, but late failure can occur up to 5 years postoperatively. Therefore long-term follow-up after ETV is mandatory.

ETV can be done in a single pediatric surgical unit by one well-trained surgeon with comparable results, but the success rate seems to be lower and a higher complication rate can occur. In the German-speaking part of Switzerland, ETV has been carrying out since 1999, so this intervention has a young tradition with not as much as experience as in other areas and countries. Furthermore, the problem in small countries like Switzerland is to have a sufficient large admission area to raise the caseload, and therefore to reach better results, more experience, and more routine. In the German-speaking part of Switzerland, we have two pediatric surgical units where ETV interventions are done and one pediatric surgical unit where only shunts are performed. As an option to ameliorate the performance of ETV, it would be an idea to centralize this treatment in one of these three units with the cooperation of more than one well-trained surgeon, ensuring round-the-clock availability with a high level of safety, efficacy, and reliability of ETV. This option needs a new approach of the Swiss population, who, so far is used to reach an excellent medical standard treatment for children for almost any diagnosis within less than an hour. In such a scenario remains the need to solve the political question of whether we should have a single ETV center or have multiple units delivering local care with lesser experience.

We suggest ETV as a procedure of choice in pediatric obstructive hydrocephalus, especially in children older than 3 months. ETV in case of shunt failure and also repeated ETV in case of failure of the previous ETV should always take into consideration and the same

criteria for indication of ETV should be applied as when treating pediatric obstructive hydrocephalus for the first time.

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