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EC NEUROLOGY
Review Article

Endoscopic Third Ventriculostomy and Cauterization of the Choroid Plexus with Rigid Endoscope: Revision and Technical Note

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Received: July 20, 2020; **Published:** August 10, 2020

Abstract

Background: The treatment of hydrocephalus in childhood continues to be one of the great challenges of modern paediatric neurosurgery. The different forms and technologies of shunt derivations while saving children also mean another disease, often as lethal as the underlying disease. The alternative of endoscopic third-ventriculo cisternostomy (ETV) has not shown satisfactory results in some cases of hydrocephalus, such as in children under 6 months of age, myelomeningocele's patients, post-infection, among others. The addition of choroid plexus coagulation (CPC) seems to open a new perspective in the endoscopic treatment of hydrocephalus.

Objective: In this article, the author intends to share his personal experience in the treatment of hydrocephalus by ETV and CPC using a rigid endoscope. Technical aspects before, during and after surgery will be presented and commented on.

Methods: The author summarizes the principles that led to the indication of coagulation of the choroid plexus by endoscopic approach and presents a brief bibliographic review discussing the advantages and disadvantages of the method, results obtained and complications. The operative technique is described and commented.

Results: Although there is still much discussion about the effectiveness of choroid plexus coagulation in the endoscopic treatment of hydrocephalus, the author has obtained some good results especially in shunt dysfunctions, in hydranencephaly/extreme hydrocephalus and in some cases of hydrocephalus in the first year of life.

Conclusion: Despite some controversies, the international literature and the author's experience have shown that the addition of choroid plexus coagulation to the third endoscopic ventriculostomy can improve results in selected cases of hydrocephalus even with the use of rigid endoscopes.

Keywords: *Hydrocephalus; Endoscopy; Choroid Plexus; Coagulation*

Introduction

The treatment of hydrocephalus in childhood continues to be one of the great challenges of modern paediatric neurosurgery, especially as it is a complex and variable disease related to etiological and pathophysiological aspects. Despite technological advances, hydrocephalus occupies between 40 - 60% of the neurosurgical agenda and about 50% of admissions are for treatment's complications. Many years have gone by since the development of the first valved shunt derivations and we still do not achieve efficiency in their functioning and infection rates at tolerable levels. International literature still points to revision rates of 1.6 - 3/patient, infectious complications of 0.5 to 35%, valvular dysfunction between 38 - 70%, secondary subdural collections to hyper-drainage in 10% and long-term mortality between 5 - 40%. We can affirm that the different forms and technologies of shunt derivations, while saving children also mean another disease, often as lethal as the underlying disease. Infectious and mechanical complications increase treatment costs and morbidity and mortality [1].

In the last 30 years, with the appearance of new endoscopic technologies and advances in video systems, the use of neuroendoscopy gave hope that an internal derivation could resolve the hydrocephalus without the implant of shunt derivations. However, after some years of experience, international literature has shown good results in general in around 40% of cases. Considering the aetiology of hydrocephalus, those associated with the presence of suprasellar cysts or the stenosis of aqueduct reach success rates higher than 70%, on the contrary in cases associated with myelomeningocele and post-infectious hydrocephalus the success rate is down 50% and 30%, respectively. In secondary hydrocephalus after intraventricular haemorrhages, especially in the newborn premature infants, due to rupture of the germinative matrix, less than 40% are resolved endoscopically. In those patients with extensive brain malformations, the ETV has failed in its entirety. If we consider the age group of patients, the majority of publications show poor results in children under 2 years of age, especially below 6 months. Despite so many discouraging data, there is a consensus that early mortality associated with ETV is around 0.5 - 1%, well below that referred to implant valvulated bypasses [1].

The physiology and pathophysiology of cerebrospinal fluid (CSF) dynamics are still not fully understood. We are aware of the existence of a circulation that goes from the choroidal arterial network to the main cerebral venous sinuses. Choroidal plexus are responsible for 60 - 80% of the production of CSF, the rest being produced by ventricular ependyma. There are indications that the pathological increase in intracranial pressure can interfere in CSF production, as well as very low blood pressure levels can stimulate this production. The reabsorption of the CSF depends on even more complex mechanisms including the differentials between the CSF pressure and the venous pressure. Pacchioni granulations are known valvular structures that open whenever the CSF pressure is higher than the central venous pressure. There are also other alternative ways of reabsorbing the CSF such as the ventricular ependyma, the root spaces of the spinal cord and the lamina cribrosa. The CSF pressure is not constant and varies with the pulse and with the respiratory rate. A very important aspect to be considered is that Pacchioni granulations do not exist in the first years of life, which leads us to conclude that the CSF absorption in the infant occurs through the alternative route, also called the minor CSF route. This characteristic may be one of the explanations for the failure of ETV in patients under 2 years of age [2].

Since 1918, the coagulation of the choroid plexus (CPC) was already known as a surgical technique aimed at the treatment of hydrocephalus. Dandy, Putman, among others, used the CPC, but the results were catastrophic which caused the technique to be abandoned. In the 70s, the development and improvement of endoscopic, as well as the navigation within the CSF space were becoming more secure. At first it seemed that the endoscopic approach would replace the implant of ventricle-peritoneal shunts, which arose after 1940. But the failures in hydrocephalus persisted in patients younger than 2 years of age, in those associated with infections, haemorrhages and malformations of the central nervous system. The CPC by the endoscopic route was again considered in order to decrease the production of CSF. Pople and Griffith published a series of endoscopic CPC cases at the end of 1993 with only 33% good results, but with many complications [3].

Benjamin Warf published in 2005 on his experience in the African continent, comparing series where they were made just to ETV with series where it was associated with CPC. The results were significantly better in patients undergoing ETV and CPC, with 65% of good results in children under 6 months of age and 81% in children under 1 year. The existence of infectious complications was 1% and mortality in patients undergoing ETV/CPC was 1/3 of that observed in those who underwent just ETV [4]. In 2013 he published another comparative study between ETV and ETV/CPC, this time in patients with idiopathic congenital hydrocephalus, observing good results in 20% and 72.4% respectively [5].

Despite other published studies not having repeated the good results achieved by Benjamin Warf, ETV/CPC usually appears with some advantages in relation to the ETV individually. Zandian., *et al*. published a meta-analysis in 2014, comparing ETV with ETV/CPC involving 10 studies. The studies involved patients with hydrocephalus associated with haemorrhages, infections, Dandy-Walker malformation and neural tube defects. Success rates were higher for ETV/CPC in all cases, especially in patients older than 2 years of age (84%) [6]. Chamiraju (2014), in her series of 27 premature patients with post-haemorrhagic hydrocephalus, ETV/CPC was a success in 37% of the cases [7]. Weil., *et al*. (2015) published the result of a cohort involving 85 patients with less than 2 years of life. More than half of the patients were hydrocephalus after intraventricular haemorrhage, 14% had stenosis of the aqueduct and 8% associated myelomeningocele. In all cases, a rigid endoscope was used and ETV/CPC was performed. The results were considered successful at 42%, 38% and 37% in a follow up of 6 months, 12 months and 24 months respectively. It was registered 7% of complications and a mortality of 3.6% [8]. Pedrosa., *et al*. published in 2017 a series of 42 patients with extreme hydrocephalus or hydranencephaly treated with endoscopic CPC. They considered the procedure effective at 70.6%, which is in accordance with the experience of Da Cunha, published in 2018 (See table 1, a series of 13 patients with extreme hydrocephalus, in which 4 only CPC was performed and in 2 cases septostomy was necessary due to multiloculated hydrocephalus) [1,8].

Age	Gender	Procedure
12* y	F	ETV + CPC
05y	F	CPC
09* y	F	ETV + CPC
15* y	M	ETV + Septostomy + CPC
11* y	M	CPC
1y	F	CPC
2y	F	ETV + CPC
1y	F	ETV + Septostomy + CPC
7y	M	ETV + CPC
3y	F	ETV + CPC
4y	F	CPC
7y	M	CPC
16* y	F	ETV + CPC

Table 1: *Extreme hydrocephalus cases. 100% success rate.*
(*) Shunted before.

There are some controversies in the use of rigid neuroendoscope. Some authors argue that the flexible endoscope is indispensable for an efficient cauterization of the choroid plexus. In most published reports, rigid neuroendoscopes were used and, Fallah., et al. (2017) analysing a series of 84 cases of ETV/CPC, in which the cauterization was subtotal in 58 (69%) patients and partial in 26 (31%) and found no statistically significant differences in the results [10].

Pre-operative image acquisition

The choice of the treatment method is an important step and requires the etiologic identification of hydrocephalus and a good investigation by images. Although many of our patients, for economic reasons have access only to computed tomography or even only transfontanellar ultrasound, magnetic resonance imaging is the best alternative to visualize the structures of the central nervous system. The FIESTA sequences can provide important information about the third ventricle floor, cerebral aqueduct and the presence of fibrosis and adhesions in the pre-pontine cistern. Magnetic resonance imaging can be an important tool in assessing prognosis, risks and planning the surgical route.

Operating technique

The indispensable equipment for the realization of ETV/CPC includes a neuroendoscope, whether rigid or flexible; tweezers and scissors for endoscopic use; high definition video equipment; Forgarty catheter N° 3 or N° 4; and equipment for coagulation, monopolar, bipolar or LASER. Among other equipment that can be of great help, we include trans-operative ultrasonography and neuronavigation.

As previously mentioned, many services do not have flexible endoscopes, but according to many reports already published, the same results can be achieved with rigid endoscopes. For better management and greater efficiency, the authors recommend short and thinner rigid endoscopes in newborns and infants. In macrocranias and extreme hydrocephalus, longer endoscopes are recommended so that it can reach any extension of the choroid plexus.

The use of bipolar is more efficient than the monopolar, paying close attention to the intensity of the coagulation so as not to cut the artery of the choroid plexus and thus cause bleeding. Another concern is with the propagation of heat and the heating of CSF during the coagulation process. Continuous irrigation of the ventricles must be assured with control of the return temperature. In the author's experience, contact LASER allows a more efficient coagulation, with less heat and detritus productions.

Neuronavigation is an important resource in multiseptate hydrocephalus and more central nervous system malformations, but with the disadvantage of not offering images in real-time. Neuronavigation can be of great help in the planning of the approach and neuro endoscopic path. Transoperatory ultrasonography allows real-time images but requires specific training for the use of this technology.

The patient should be positioned in dorsal decubitus with a slight anterior flexion of the head and elevation of the operating table. This can help minimize the formation of pneumocephalus. The head should be well fixed to avoid any movement during endoscopy. Asepsy and preparation of the operative field. Incision of the skin 1,5 - 2 cm from the midline, at the level of the coronary suture. In the newborn trepanation is not necessary, a careful cross-cut or straight incision of the dura mater can be made. In older children, trepanation, haemostasis with bone wax is necessary before dura incision. After coagulation of the dura mater and adjacent cortex, the endoscope should be introduced slowly and carefully, guided by the neuronavigator and the video system. Introduction of the endoscope in the lateral ventricle. Identification of the choroid plexus. The ETV should be performed first. Following the choroid plexus in the postero-anterior direction we find the foramen of Monro. We cross the foramen of Monro and we are in the third ventricle. Between the mamillary bodies, superiorly, with the aid of a Forgaty catheter number 4, we gently puncture the floor of the third ventricle, filling the balloon to enlargement of the orifice. Then we must make sure that the Lilliquist's membrane is open. The choroid plexus coagulation must be performed in the craniocaudal direction, with the care of coagulating the nourishing artery that accompanies the choroid plexus (Figure 2a-2c). The coagulation should be as extensive as possible, without injury to adjacent brain structures. The bilateral approach is facilitated in cases of single ventricle, absence of septum or fenestrated septum. Having a normal septum, a septotomy may be performed or the contralateral ventricle may be approached through a new trephination. At the end, after the endoscope is removed, be careful with the hermetic closing of the dura-mater to avoid CSF fistula. If there is bleeding, the implantation of an external drainage catheter in the ventricle is recommended. The patient should be observed in an intensive care unit for 48 - 72 hours. The author considers as a surgical success the cases that evolve with intracranial hypertension control without the need of implantation of a shunt (Figure 1).

Advantages, disadvantages and complications

Many reports in the international literature and the experience of the authors show that endoscopic CPC improves the results of ETV, naturally restoring the CSF circulation and minimizing the effects of the poorly absorbed syndrome in some hydrocephalus cases of hy-

Figure 1: *Flow Void through the third ventricle floor, pos ETV.*

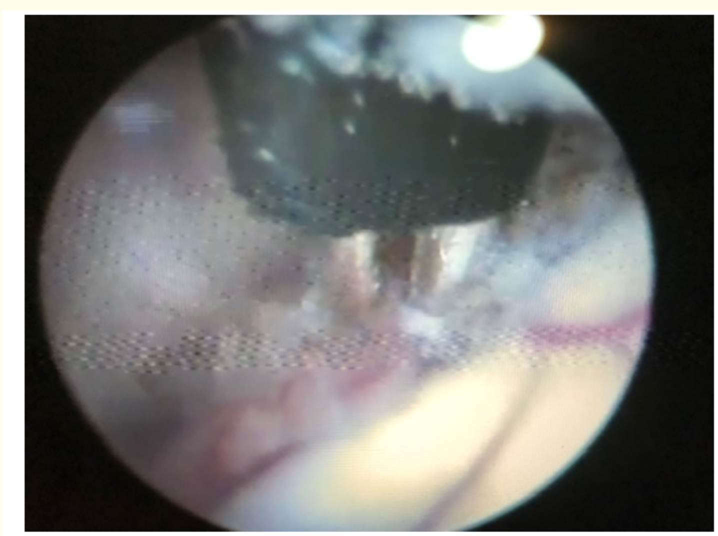

Figure 2a: *Coagulation of the choroid plexus with a bipolar forceps.*

drocephalus. In extreme hydrocephalus and hydranencephalies, there are reports that only with endoscopic CPC was it possible to control hypertensive hydrocephalus without the need for shunt derivative implants [1,8].

However, performing CPC presents some problems.

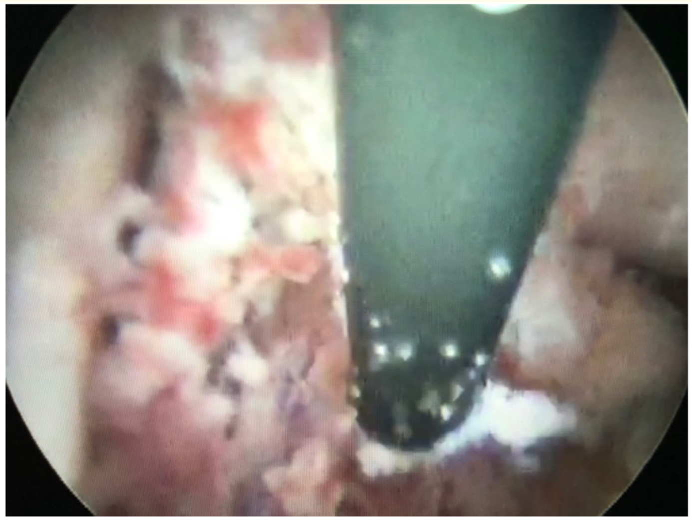

Figure 2b: *Continuation of coagulation of the choroid plexus with a bipolar forceps.*

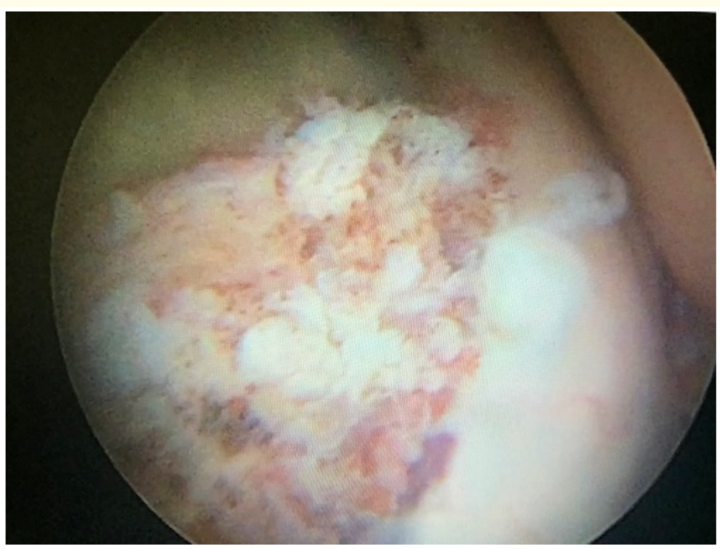

Figure 2c: *Coagulated left choroid plexus.*

The surgical time is longer since it is important to access the largest possible extension of the choroid plexus on both sides. This may mean in many cases performing a septostomy or even bilateral trepanation. The use of flexible endoscopes allows an extensive approach to the choroid plexus. However, few services have this type of equipment and trained neurosurgeons for its use.

A greater occurrence has been observed of seizures and transient endocrinological disturbances in patients undergoing CPC. One of the hypotheses for these occurrences would be the heat released during coagulation. The use of LASER instead of monopolar or bipolar can minimize the thermal effect of coagulation on neighboring tissues. Possible late complications of CPC in newborn, especially premature babies deserve observation through new studies.

In addition to the difficulties referred to above, there are complications inherent to ETV such as small bleeds, subdural collections, pneumocephalus, CSF leakage, infections, and severe perforation of the basilar artery. As of late complications, we can mention the enclosure of cisternostomy and the formation of new membranes in the cisternal space. Although rare, we have to remember of a serious complication later, sudden death due to transtentorial herniation associated with arrested hydrocephalus. ETV with or without CPC requires pressure monitoring, particularly in acute hydrocephalus. The monitoring can be done through a ventricular catheter connected to a closed external collector (external ventricular drainage). The drain must initially remain closed for monitoring only. The author recommends monitoring intracranial pressure for 48 - 72 hours before hospital discharge.

In table 2, the author presents 45 infants undergoing neuroendocopic treatment. Patients were divided into 2 groups, only ETV (18) and ETV + CPC (27). The second group included a patient from the first group in which a second approach was performed with the performance of CPC. There was no significant difference between the two groups.

ETV 18 patients Age: 3 - 10 months Results: 39% Failed x 61% Good results	**ETV + CPC** 27 + 1(*) Age: 2 - 11 months Results: 39% Failed x 61% Good results (*) Patient initially submitted to ETV, later treated with ETV + CPC.

Table 2: *45 patients.*

Table 3 shows a series of 13 patients with myelomeningocele and hydrocephalus initially treated with shunts. The patients returned with clinical and radiographic signs of shunt dysfunction. In all cases, the shunts were removed and ETV + CPC was performed. The approach was considered a success in 85% of cases.

Age	Gender	Complications	Results
10y	F	Infection	EVD/Shunt
09y	F		Success
05y	F		Success
07y	M		Success
16y	M		Success
10y	M		Success
13y	F	CSF Fistula	Shunt
17y	F		Success
18y	F		Success
12y	F		Success
14y	F		Success
04y	M		Success
02y	F		Success

Table 3: *ETV + CPC performed on 13 patients with myelomeningocele and hydrocephalus who returned with signs of shunt dysfunction (February 2014 - October 2016).*

Conclusion

Despite the existence of some controversies and the need for further investigation on the indications and long-term effects, the international literature and the author's experience have shown that the addition of choroid plexus coagulation to the third-ventricle cisternostomy can improve the treatment of hydrocephalus, even with the use of a rigid endoscope.

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Volume 12 Issue 9 September 2020