Choroid plexus coagulation for pediatric hydrocephalus: review of historical aspects and rebirth

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No conflicts of interest to declare.

Rev. Chil. Neurocirugía 42: 15-18, 2016

Abstract

This study aims to review historical aspects and rebirth of the endoscopic choroid plexus coagulation (CPC) for pediatric hydrocephalus. The first CPC procedure goes back to early 1930s. After the development of other treatment methods and the understanding of CSF dynamics, the application of CPC dramatically decreased by 1970s. In 2000s, there was a rebirth of CPC in combination with endoscopic third ventriculostomy (ETV), and remains one of the options for the treatment of pediatric hydrocephalus in selected cases. CPC might provide a temporary reduction in CSF production to allow the further development of CSF absorption in infant, and adding to ETV for infants with communicating hydrocephalus may increase the shunt independent rate thus avoiding the consequence of late complication related to the shunt device. This is important for patients who are difficult to be followed up, due to geographical and/or socioeconomic difficulties. And also adding CPC to ETV for obstructive hydrocephalus in infants younger than 1 year of age may also increase the successful rate. Furthermore, CPC may be an option for cases with high chance of shunt complication such as multiloculated hydrocephalus, extreme hydrocephalus and hydranencephaly. In comparison with the traditional treatment of CSF shunting, the role of CPC needs to be further evaluated in particular concerning the neurocognitive development.

Key words: Pediatric hydrocephalus, Choroid plexus coagulation, Endoscopic third ventriculostomy.

Introduction

Pediatric hydrocephalus is one of the most frequently diagnosed diseases in pediatric neurosurgical practice. Approximately 60% of the total cases of hydrocephalus in children are congenital or acquired in childhood. It´s a typical surgical disease, and If left untreated, most cases are lethal. Traditionally, shunts are the main treatment since 1940s. However, sometimes shunt dependency in children with hydrocephalus is a terrible problem, and is more dangerous in emerging countries than developed countries because of the difficulties that prevent access to prompt neurosurgical intervention, in cases of infection or shunt malfunction[8,22,23].

In this scenario, mainly in obstructive hydrocephalus, neuroendoscopic techniques such as endoscopic third ventriculostomy (ETV) become vitally important in the management of these patients. ETV is successful in about 80% of children older than 1 year of age, regardless of the cause of obstructive hydrocephalus. For infants younger than 1 year of age, isolated ETV remains controversial, probably because of a deficiency in CSF absorption. Benjamin C. Warf, at CURE Children´s Hospital of Uganda, hypothesized that the addition of choroid plexus coagulation (CPC) at the time of the ETV, to reduce the rate of CSF production, might improve the outcome[22]. This pioneer neuroendoscopic procedure of the early 20th century, abandoned due to limited technology and complications, has been and adapted current neurosurgery.

Historical aspects

Faivre, in 1854 and Luschka, in 1855 were the first researchers to suggest that the choroid plexus is the source of CSF[3,29]. Cushing supported this hypothesis through intraoperative observations[4]. Extrachoroidal fluid production was suggested by Weed in 1914, by animal studies[29]. Dandy, in 1918, demonstrated in an animal study that unilateral hydrocephalus was produced when the 4th ventricle was blocked together with the access through the foramen of Monro to the contralateral plexectomized lateral ventricle[5,30]. Furthermore, in the same year, he also demonstrated that CSF is produced by the choroid plexus in an animal study. Based on this result, he performed choroid plexus extirpation in four infants with communicating hydrocephalus by open surgery. In this series, one infant with moderate hydrocephalus and myelomenigocele was well on 10 months follow-up, and the other three infants with severe hydrocephalus died until 4 weeks after the operation[5,30].

After, in 1932, Dandy also used a rigid Kelly cystoscope to inspect the lateral ventricles in two hydrocephalic children[6]. Cauterization of the choroid plexus was attempted in one case. He further detailed the technique of endoscopic coagulation of the choroid plexus in 1938[7,30]. CPC was first described by Putnam, in 1934[30]. In the subsequent years, besides CPC, other surgical treatments of hydrocephalus were introduced including endoscopic third ventriculostomy (ETV) and extrathecal CSF shunts. In a review, from 1934 to 1957, there were 95 cases of CPC. The mortality rate was 15%, while the mean success rate was 60% with an average follow-up period of 8 years. On the other hand, there were 1087 cases of various kinds of CSF shunt, including 230 ventriculoperitoneal shunts (VPS). The mean mortality rate was 10%, and the mean initial success rate was 60% with an average follow-up time of 2 years. The result of the reviews showed a shift from CPC to shunts, perhaps due to poor and limited technology. However, the late complication rate for shunt was 57%[17,30]. Scarff, in 1970, published the first large series of CPC cases, his own series of 39 children treated during 23-year period, with 67% of success[18]. Milhorat, in 1974[13], reported his series of 12 cases underwent choroid plexectomy. Among 11 survivors, 8 (72%) failed and required further shunt[13]. After this report, that CPC in rhesus monkeys reduced CSF production by only 40%, it declined in favor of the shunts[30].

Literature concerning neuroendoscopy, from the 1980s to 2004, showed success rate of CPC was between 30 and 52%[9,10,16]. In small series, two out of three cases were successful[1,14,30]. Griffith, in 1986, gave a detailed account of endoscopic intracranial neurosurgery, through a report of the results of 71 CPC patients with or without shunt, from 1972 to 1982[9]. The selection criteria were infants with hydrocephalus who had progressively enlarging head circumference with ventricles grossly dilated and absent superficial CSF space on CT scan. Behavioral changes were also considered. In his series, 30% were not shunt dependent. The success rate was 54, 58, and 22% for myelomenigocele, communicating, and obstructive hydrocephalus groups, respectively. The same author, in 1990[10], further reported the results of 32 childhood hydrocephalus cases treated by CPC between 1985 and 1988 with CT scan examination. Eighteen cases were below the age of 6 months. Patient selection was the same as in his previous report. In addition, all cases showed marked ventricular dilatation on preoperative CT scan. Different to his previous series, he added postoperative perfusion of the ventricular system with artificial CSF to clear the post-coagulation blood and protein released to the CSF. The average follow-up time ranged 1 to 4 years. Fifty-two percent of the patients were shunt independent. Those required shunting was all in an interval of $\leq$12 weeks except one. Among the successful group, most of the patients showed a head circumference similar to the preoperative size[30].

Pople and Ettles, in 1995[16], reviewed the results of CPC in 116 children with hydrocephalus operated from 1973 to 1993[15,16]. The mean age was 2 years and the overall hydrocephalus control rate was 49.5%. Among communicating hydrocephalic children with slow to moderate rate of increase in head circumference, the long-term control rate was 64%. On the other hand, only 35 % achieved long-term control without CSF shunts in patients presented with tense fontanels and rapidly progressed hydrocephalus and the authors suggested that the main indication for CPC was mildly progressive communicating hydrocephalus in infants. In these patients, it seemed that the balance between production and absorption of CSF could be restored by only a small reduction in outflow from the choroid plexus of the lateral ventricle. In contrast, CPC was not recommended for rapidly progressive hydrocephalus with acutely raised intracranial pressure[15,16,30]. In fact, until this time, these first experiences were quite controversial, perhaps because of technological limitations[8].

Rebirth

In the late 1990s to early 2000s, due to advancement in neurosurgical technology, the mortality rate in isolated CPC has decreased, but the key issue for its decline in clinical practice is its efficacy[30]. At this time, Warf´s Uganda series, for the first time investigated the benefic effect of ETV associated to CPC, again arousing interest in this technique. It was concluded that ETV/CPC procedure is superior to ETV alone in infants younger than 1 year of age, particularly among those with nonpostinfectious hydrocephalus and myelomeningocele, but longer follow-up with neurocognitive assessment is necessary[22].

Dr. Warf published their results of ETV/CPC for children in Africa in 2005, majority of them were infants[22]. The long-term outcome and neurocognitive outcome were reported in 2008[24] and 2009[21], respectively. They highlighted that shunt dependency in children with hydrocephalus is more dangerous in developing countries than in developed countries because of the limits to access competent centers in the event of shunt malfunction or infection[22]. Between 2001 and 2004, ETVs were performed as the initial treatment in 550 patients presented with hydrocephalus. After evaluating the initial results, it was decided to perform bilateral lateral ventricle CPC including the temporal horn using flexible endoscope from a single approach in addition to the ETV to assess the benefit. Warf and Campbell, in 2008[24], reported the long-term result of ETV/CPC for East African infants with hydrocephalus related to myelomeningocele. Among the 338 infants whose myelomeningocele was repaired prior to 6 months of age, 258 patients (66%) who had been followed up for > 6 months required treatment for hydrocephalus. There were 93 cases (mean age, 3 months) who had completed ETV/CPC with >1 month follow-up. They have achieved a successful (shunt independent) rate of 76%. This successful rate was higher in ETV/CPC cases than those by ETV alone for infant age 6 months or younger with hydrocephalus in association with myelomeningocele as reported in the literature[11,20]. Still, in 2009, Warf et al.[21] reported the neurocognitive outcome and ventricular volume in children with myelomeningocele treated for hydrocephalus in Uganda. The modified

Bayley Scales of Infant Development (BSID-III) and the frontal / occipital horn ratio (FOR) were used to compare three groups of patients with myelomeningocele. For the modified BSID-III, there was no statistically significant difference between treatment groups of VPS and ETV/CPC. For the ventricular size, the mean FOR were 0.7, 0.65 and 0.62 for the VPS, the ETV/CPC and the not required-treatment groups respectively without statistically significant difference in between. The authors suggested that future work was needed to compare outcomes by using a larger control group of children treated primarily with VPS shunt placement. Still, the same group have applies ETV/CPC in encephalocele, with the successful rate was 85%[27], and in obstructive hydrocephalus due to aqueductal stenosis, with success rate of 81.9% in patients received ETV/CPC[28].

Dandy-Walker complex is another condition treatable by ETV/CPC, according Warf´s Africa series[26]. Inside also Uganda scenario it was the larger series of this disease treated by neuroendoscopy, and this technique should be strongly considered as the primary management in place of the traditional standard of creating shunt dependence. For this disease, the success rate was 74% for Dandy-Walker malformation, 73% for Dandy-Walker variant, and 100% for mega cisterna magna. Eighty-eight percent of the cases were younger than 12 months and 95% had an open aqueduct at the time of ETV/CPC. None required posterior fossa shunting in a mean follow-up of 24.2 months[26]. From the same African series by Dr. Warf, the use of the ETV/CPC for communicating hydrocephalus is a viable option[23]. It was significantly more successful than ETV alone in treating congenital idiopathic hydrocephalus of infancy. In this study with sixty-four infants (mean/median age, 6.1/5.0 months), sixteen consecutive patients were treated by ETV alone, and the subsequent 48 by ETV/CPC (mean/median follow-up 34.4/36.0 months). ETV was successful in 20% and ETV/CPC in 72.4% at 4 years ($p < 0.0002$, logrank test; $p = 0.0006$, Gehan-Breslow-Wilcoxin; hazard ratio 6.9, 95% CI 2.5-19.3. It was assumed that the primary effect of ETV, as a pulsation absorber, and of CPC, as a pulsation reducer, may be to reduce the net force of intraventricular pulsations that produce ventricular expansion. On the other hand, ETV alone may be less successful for infants because of greater brain compliance. Regardless, this technique appears to have a higher long-term success rate and a lower infection rate than primary shunt placement and should be considered an effective primary treatment option for congenital idiopatic hydrocephalus For multiloculated hydrocephalus, ablation of the choroid plexus in conjunction with septal fenestration and shunting was an option to control the hydrocephalus. Zuccaro and Ramos[31] reviewed their series of 93 cases with multiloculated hydrocephalus. Choroid plexectomy / fulguration was performed in 14 cases (eight by endoscopy and six by craniotomy). The authors concluded that each patient must be studied individually, because variable success rate[30] Initial experience with ETV/CPC for post-hemorrhagic hydrocephalus of prematurity has revealed the importance of prepontine cistern status and the predictive value of FIESTA MRI imaging[25]. In hydrocephalus in premature infants with IVH and hydrocephalus ETV/CPC is a safe initial procedure for, obviating the need for a shunt in selected patients. Even though the success rate is low (37%), the lower rate of complications in comparison with shunt treatment may justify this procedure in the initial management of hydrocephalus. As several of the studied factors have shown influence on the outcome, patient selection based on these observations might increase the success rate[2]. Besides ETV/CPC combined technique, nowadays there are new indications for isolated CPC, such as in extreme hydrocephalus and hydranencephaly[12,14,19]. Avoidance of a CSF shunt is desirable in these conditions, since the thinness and fragility of their scalp, besides the common presence of infected scalp ulcers at the parietal bosses. Morota and Fujiyama, in 2004[14], described the technique of unilateral transparietal approach for bilateral CPC using a flexible neuroendoscope for three infants with IVH related hydrocephalus. Two of them were shunt independent. The authors suggested that the characteristics of favorable candidates for CPC with severe advanced hydrocephalus like hydroanencephalic hydrocephalus, slow progressive hydrocephalus and lack of or thinned out septum pellucidum to make the bilateral endoscopic access possible. Malheiros et al., in 2010[12], with a series of seventeen patients, have completed CPC in 9 patients; the procedure successfully controlled excessive head circumference and signs of increased intracranial pressure in 8 of these patients (88.8%). One endoscopic procedure in a hydranencephalic child failed after 7 months, resulting in VPS placement. Thus, of the 10 patients randomized to CPC, 8 were treated successfully by CPC (80%), and 2 went on to have a VPS. There were no complications related to this method of treatment. So, the authors concluded that CPC is an acceptable alternative to VPS for treatment of hydranencephaly and near hydranencephaly, because It is a single, definitive, safe, effective, and economical treatment that may avoid the complications of shunting. In another recent study[19], in severe congenital hydrocephalus and hydranencephaly, CPC stabilizes macrocephaly in approximately 40% of infants with and can be considered as an alternative to VPS placement. Patients were followed from 30 to 608 days (median of 120 days). Of the 30 evaluable patients, CPC was considered to be successful in 13 (43.3%), including 8 of 20 patients with severe hydrocephalus and 5 of 10 with hydranencephaly. Failure of CPC was evident from increased head circumference in 14 (82%) of 17 patients and from CSF leakage in 3. Of the 17 failures, 13 occurred within 3 months of surgery. Six patients died: 3 whose CPC procedures were failures, 2 whose CPC was successful, and 1 postoperatively. Of the 17 in whom CPC failed, 10 subsequently underwent VPS insertion This African study concluded that isolated CPC stabilizes macrocephaly and can be considered as an alternative to shunt placement[19].

Conclusion

CPC remains one of the options for the surgical treatment of pediatric hydrocephalus mainly in communicating hydrocephalus, and this procedure might provide a temporary reduction in CSF production to allow the further development of CSF absorption. Adding CPC to ETV for infants with hydrocephalus of various non postinfectious causes may increase independency related to the shunt device. This result is very important for patients who are difficult to be followed up, due to geographical and socioeconomic constrains, as in developing countries. CPC may also be an option for cases with high chance of shunt complication such as hydranencephaly and as an adjunct therapeutic measure for complex cases of hydrocephalus such as multiloculated hydrocephalus. Unfortunately, it´s not known about the long-term effects of coagulating about 80% of a child´s choroid plexus and leaving larger ventricles, associated or not to ETV, specially in terms of neurocognitive aspects. However, it is a promising way to improve outcome of hydrocephalus, reducing shunt dependency, mainly in emerging countries. More studies with larger series are necessary to define the evident benefits of this technique.

Recibido: 15 de diciembre de 2015
Aceptado: 12 de febrero de 2016

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