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CONGENITAL HYDROCEPHALUS: INNOVATIONS IN THE USE OF ENDOSCOPIC SHUNTING (ETV/CPC) IN NEWBORNS

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Abstract: Introduction: Hydrocephalus, a prevalent neurological condition, is characterized by the abnormal accumulation of cerebrospinal fluid (CSF) in the cerebral ventricles. This accumulation leads to ventricular dilation, damage to brain tissue and the manifestation of various symptoms. Hydrocephalus can be classified clinically into two main categories: congenital hydrocephalus (CH) and acquired hydrocephalus, based on the time of onset. Aim: The aim of this study is to carry out a literature review on neurosurgical methods in the treatment of CH. Methodology: The scientific databases used were: Latin American and Caribbean Health Sciences Literature (LILACS), *Scientific Electronic Library Online* (SCIELO), *National Library of Medicine* (NIH), MEDLINE. The descriptors used in this research were: "Congenital Hydrocephalus", "Endoscopic Shunt", "Shunt". The article was prepared using articles from 2020 to 2025. Results and discussion: Endoscopic third ventriculostomy with choroid plexus cauterization (ETV/CPC) is mainly recommended for cases of obstructive hydrocephalus, where the underlying cause of the condition is clearly identified and the anatomy of the CSF circulatory system is suitable for the procedure. On the other hand, external ventricular drainage involves inserting a catheter into the ventricular system to facilitate the drainage of CSF out of the body. Current treatment also includes ventriculo-peritoneal shunts (VPS) with valves to redirect CSF from the ventricles to the peritoneum. Shunt technology is limited by a number of complications, which include infection after implantation, obstruction of the shunt due to clot formation or obstruction of the catheter by scar tissue or choroid plexus, disconnection and migration of the tube, and excessive or insufficient CSF drainage due to valve malfunction. Conclusion: It can be concluded that the most effective treatment for the pediatric approach is the ETV/CPC me-

thod, since a minimal incision is made and the surgical approach is performed by anatomical landmarks, providing, according to the literature, fewer complications and better long-term effects.

Keywords: Third Endoscopic Ventriculostomy with Choroid Plexus Cauterization; Hydrocephalus, Congenital; Endoscopic Shunt;

INTRODUCTION

Congenital hydrocephalus (CH) is a primary form of hydrocephalus, characterized by the pathological expansion of the cerebral ventricles. Genetic influences are one of the main contributors to the pathogenesis of CH, and epidemiological evidence suggests their involvement in up to 40% of all cases observed globally. CH is often diagnosed during the postnatal period or in the first year of life by the presence of ventriculomegaly, macrocephaly (Duy; Kahle ., 2023)

The global frequency is estimated at around 0.9-1.2 per 1,000 births (Javeed *et al.*, 2023). According to Ellenbogen *et al.* (2020) particularly in developing countries. The mainstay of treatment has long been shunt placement for CSF diversion, but recent years have seen the rise of alternative procedures such as endoscopic third ventriculostomy (ETV pediatric hydrocephalus affects approximately 0.5 to 0.8 per 1,000 births, and is most commonly caused by intraventricular hemorrhage of prematurity, Chiari type II malformation, tumors and infection.

While secondary hydrocephalus is found in cases of obstructive brain tumors, leptomeningeal disease, infection and hemorrhage, these patients usually develop elevated intracranial pressure (ICP) and demonstrate clear obstructive pathology. In addition, all forms of hydrocephalus in children have characteristics such as ventriculomegaly due to excess production of cerebrospinal fluid (CSF), obstruction of the CSF pathways, impaired CSF

flow or an imbalance of production and re-absorption in the subarachnoid granulation (Sakamoto *et al*), 2024)

It is understood that the normal flow of CSF originates from the lateral ventricles, through the foramen of *Monroe*, into the third common ventricle, continues into the fourth ventricle through the cerebral aqueduct and exits through the foramen of *Luschka* and *Mage* into the subarachnoid space. Therefore, hydrocephalus occurs due to flow obstruction and failure of CSF absorption by the normal drainage pathways (Hochstetler; Raskin; Blazer-Yost, 2022) .resulting from a multitude of causes. While the etiologies of hydrocephalus are numerous, many of the acute and chronic symptoms of the condition are shared. These symptoms include disorientation and pain (headaches

Treatments for hydrocephalus mainly include surgical CSF shunting or endoscopic third ventriculostomy with choroid plexus cauterization (ETV/CVC). For multiloculated hydrocephalus, choroid plexus ablation in conjunction with septal fenestration and shunting is still an option for controlling hydrocephalus. Therefore, by eliminating this source of CSF, the neurosurgeon is probably reducing the amount of CSF needed to be reabsorbed by the arachnoid villi, as well as the force applied to the ventricles (Guimarães *et al.*, 2025) . The aim of this article is to review the literature on congenital hydrocephalus and the main neuro-surgical and therapeutic methods used in the management of these patients.

METHODOLOGY

This article is a bibliographic review, with several original articles, which were looked at in scientific databases such as Latin American and Caribbean Literature in Health Sciences (LILACS), *Scientific Electronic Library Online* (SCIELO), *National Library of Medicine* (NIH), MEDLINE. The descriptors used

in this research were: “Congenital Hydrocephalus”, “Endoscopic Shunt”, “Shunt”. The Boolean operators used in this research were AND and OR.

The data was collected between 2020 and 2025, in the last 5 years. the inclusion criteria were studies available in full and free online, the articles used were originals such as literature reviews, randomized and double-blind studies, systematic reviews, their technologies and the impact of their interventions, articles in Portuguese and English were used. while the exclusion criteria for this article were the exclusion of duplicate articles, incomplete works, paid works and articles that were not in English and Portuguese. A total of 59 original articles were found, of which only 23 were selected for this article.

RESULTS AND DISCUSSION

CONGENITAL HYDROCEPHALUS: DEFINITION AND ETIOLOGIES

Hydrocephalus is a prevalent neurological condition characterized by the abnormal accumulation of CSF within the ventricles of the brain. The ventricular system comprises the lateral, third and fourth ventricles, interconnected by foramina. These ventricles are filled with CSF which performs multiple functions, including removing metabolic waste from the brain, maintaining environmental homeostasis and regulating the balance of intracranial pressure. This accumulation leads to ventricular dilation, damage to brain tissue and the manifestation of various symptoms. HC is a condition characterized by the presence of hydrocephalus symptoms in babies during prenatal development. In contrast, acquired hydrocephalus can occur at any time after birth. In this sense, understanding the distinct characteristics and classifications of hydrocephalus is crucial for accurate diagnosis and appropriate treatment planning (Hochstetler;

Raskin; Blazer-yost ., 2022)resulting from a multitude of causes. While the etiologies of hydrocephalus are numerous, many of the acute and chronic symptoms of the condition are shared. These symptoms include disorientation and pain (headaches

The global prevalence rate of HC is approximately one in every five hundred births, with multifaceted predisposing factors in reproduction (Liu *et al.*, 2024). While according to Greenberg *et al.*,(2023)such as global developmental delay and autism spectrum disorder; however, current recommendations are not specific to or inclusive of congenital hydrocephalus (CH HC is present in approximately 1 in every 1,000 live births and is among the most common neurodevelopmental disorders. Therefore, it is an idiopathic disease and is associated with developmental abnormalities, such as genetic mutations linked to ciliopathies, neural tube defects, spina bifida, myelomeningocele, anencephaly, while the acquired form can develop from ischemia or hypoxia, hemorrhage, infection, trauma and brain tumors (Mcknight *et al*).., 2021)

According to Kahle *et al.*,(2024) the secondary or acquired form can be caused by infections or hemorrhages of the central nervous system (CNS), and can be correlated to blockage of CSF pathways, or dysregulation of CSF secretion due to inflammation. Although the clinical causes are unclear, the characteristic pathogenic accumulation of CSF may be associated with brain malformations such as aqueductal stenosis. In addition, HC provides a hypercompliant brain, unable to withstand the pressure exerted by the ventricular CSF, generating passive CSF accumulation and secondary ventricular dilatation.

According to Duy and Kahle(2023) mutations in L1CAM linked to the X chromosome are considered to be the main causes of CH, commonly attributed to obstruction of CSF flow due to stenosis of the cerebral aqueduct,

however, aqueductal stenosis is not always present in all patients. It can also be understood that mutations in related genes lead to various abnormalities in ciliary structures, resulting in ciliary motility disorders and the development of hydrocephalus.

Among other genes associated with the development of HC, one can consider mutations in the DNAH2 and DNAH14 genes, also described in the development of chronic hydrocephalus, which contribute to the structure of motile cilia, have been associated with primary microcephaly, neurodevelopmental disorders and seizures. While the AP1S2 genes, two autosomal recessive MPDZ and CCDC88C have also been reported to cause CH, involved in defects of the aqueduct of Sylvius, growth and movement of the cilia and CNS development. (Sakamoto *et al* .., 2024)

TREATMENT OF CONGENITAL HYDROCEPHALUS

CSF shunting is the main treatment for CH, which can be achieved by means of shunts that drain CSF directly from the high-pressure ventricular cavity into the low-pressure peritoneal or atrial cavity. The shunt method has three basic components: a ventricular catheter placed in the lateral ventricle, a valve that regulates the flow of CSF out of the brain and a distal catheter that ends in a cavity, although the peritoneal cavity as in ventriculo-peritoneal shunt (VPS) is the most common location, ventriculo-atrial shunt (VA) and ventriculo-pleural shunt (VPLS) are also commonly used (Kahle *et al* .., 2024)

On the other hand, external ventricular drainage (EVD) involves inserting a catheter into the ventricular system to facilitate the drainage of CSF out of the body. This procedure is commonly employed for acute transit hydrocephalus and serves as a temporary measure to relieve pressure within the ventricles before definitive treatment is initiated (Javeed *et al* .., 2023)

Neuroendoscopy is another option, with a preference for being more physiological, performed on specifically indicated patients, such as those with obstructive hydrocephalus, and involves inserting an endoscope into the ventricular system with the aim of addressing the primary pathology or altering the CSF mass flow. This technique has several advantages, including minimal tissue damage, rapid recovery and better visibility (Hochstetler; Raskin; Blazer-Yost ., 2022)resulting from a multitude of causes. While the etiologies of hydrocephalus are numerous, many of the acute and chronic symptoms of the condition are shared. These symptoms include disorientation and pain (headaches

Therefore, HC-related treatment seeks to restore CSF circulation and reduce intracranial hypertension. Although PVD is more commonly used, it can cause complications such as infections, surgical revisions and dysfunction of the valvular system. In relation to PVD, neuroendoscopy and laparoscopy for patients with hydrocephalus are safe methods. Neuroendoscopy helps to precisely place the catheter at the desired ventricular end, while laparoscopy allows for minimally invasive and precise insertion of the catheter into the abdominal cavity, and is an important technique for communicating hydrocephalus (Song *et al.* ., 2021)

In this sense, PVD surgery may be indicated for symptomatic hydrocephalus when ETV is contraindicated. Common indications for PVD include spina bifida, congenital hydrocephalus, hydrocephalus due to tumor, hemorrhage, infection or trauma and idiopathic intracranial hypertension (Li; Caird ., 2025)

In addition, according to Javeed *et al.*,(2023) in the first year after the installation of a PVD, the incidence of failure has been reported to be between 11% and 25%. PVD failure is a sequel to multiple complications, which include obstruction that can be complete or

partial, infection of the shunt, the most common being *Staphylococcus epidermidis*, which is part of the skin flora, excessive drainage leading to subdural hematoma (SDH), insufficient drainage, migration of the shunt, peritoneal pseudocysts, intestinal perforation and hernias. In addition, this complication requires revision and replacement, often requiring frequent and lengthy hospital stays.

While the endoscopic shunt or endoscopic third ventriculostomy (ETV) creates a direct low-resistance route for CSF to pass from the third ventricle to the cisterns, this procedure overcomes obstructive hydrocephalus at the level of the third ventricle by creating an ostomy that communicates the third ventricle to the subarachnoid space. ETV is sometimes associated with cauterization of the choroid plexus in an attempt to reduce the volume of CSF produced by the choroid plexus of the lateral ventricles. It is a less invasive surgical procedure, which creates a detour for the CSF, reducing dependence on *shunts* and complications, which is performed by introducing an endoscope in the midline through an introduction observing intraoperative anatomical parts such as the hypothalamic infundibulum, basilar arteries (Guimarães *et al.* ., 2025)

ENDOSCOPIC SHUNT (ETV) IN NEWBORNS

ETV and the PVD technique can improve cognitive function in children with hydrocephalus. According to Dhandapani *et al.*(2021)their differences in cognitive and Quality of Life (QOL 40% of children undergoing PVD and 50% of children undergoing multiple shunt surgery developed cognitive dysfunction, while only 25% of those undergoing ETV showed cognitive deficits. This new route allows the CSF to bypass any obstructed pathways and drain directly from the ventricles. It is recommended mainly for cases of obstructive hydrocephalus, in which the

underlying cause of the condition is clearly identified and the anatomy of the CSF circulatory system is suitable for the procedure.

This technique consists of introducing the endoscope with guide cannula, optics and working channels at the entry point of the skull of less than 1cm, the entry site for surgical realization would be the Kocher point or outer third of the anterior fontanelle in infants, after a 4mm corticectomy, the endoscope is introduced through the parenchyma towards the lateral ventricle and foramen of Monro, the interventricular area must be located, and then the endoscope is directed towards the third ventricle, after detecting the ventricular membrane, a puncture is performed with a Fogarthy type catheter, which is then introduced through a ventriculostomy to the pre-pontic cistern (Diallo *et al*), 2023)

THE combination of ETV with choroid plexus cauterization (CPC) (ETV/CPC) has emerged as an effective treatment for some infants with hydrocephalus, which favours shunt independence, CPC is commonly performed using a flexible endoscope via septostomy and aims to cauterize the choroid plexus as much as possible. ETV, therefore, allows the creation of a pathway for the drainage of CSF flow through the opening of the third ventricle, and according to Islas-Aguilar *et al*, (2024) ETV/CPC avoided reoperation in 66.9% of patients within 6 months.

According to Coulter *et al*, (2021) this technique is best performed in babies over 1 month old, with hydrocephalus secondary to myelomeningocele and aqueductal obstruction. However, failure is more likely in neonates with posthemorrhagic hydrocephalus of prematurity (PHHP), particularly those under 1 month of corrected age and those with a pre-pontine scar, with an incidence of around 10 to 15% of premature newborns (Islas-Aguilar *et al*, 2024) . Diallo *et al*, (2023) reported that hydrocephalus of various etiologies showed an overall procedure success rate of 75%, with

the best success rate in myelomeningocele (87.5%) and the worst in post-infectious disease (50%). ETC/CPC is also effective in hydrocephalus associated with the Dandy-Walker complex (DWC)

In order to perform the ETV technique correctly, it is important to position the trepanation hole 3 cm from the midline using image guidance, assessing a line that extends from the interpeduncular cistern to the foramen of Monro to the skull in the sagittal plane using magnetic resonance imaging (MRI). In this context, trepanation is usually performed on the right side in symmetrical ventricles or on the side of the small ventricle in asymmetrical ventricles to facilitate perforation of the septum pellucidum (Chowhady *et al*,) .2021

According to the aforementioned author, ETV can also be performed for simultaneous biopsy and for pineal or posterior third ventricle tumors, obstructive hydrocephalus with congenital or acquired aqueduct stenosis. While the complication rate can vary from 8 to 15%, intraoperative and early complications include hemorrhage (cortical, ventricular and cisternal vessels), neural injury (fornix, third cranial nerve, hypothalamus and midbrain), subdural collections, pneumoventricle, hyponatremia, seizures, delayed awakening, bradycardia, hypothalamic dysfunction (diabetes insipidus, lack of thirst, amenorrhea) and hyperthermia.

Peralta *et al*, (2023) carried out a study in which they analyzed 11 fetuses with a gestational age of 28.7 weeks on average, percutaneous VTE showed no perioperative complications and was considered viable for human fetuses with progressive ventriculomegaly or severe for mother and fetus, while Rocque *et al*, (2022) described a success rate of 41% in 6 months of performance, and that advanced age is associated with a greater probability of successful VTE.

According to Warf *et al*, (2023) ETV/CPC is a durable and effective treatment for young children, with an estimated 59% of patients

not requiring a postoperative shunt. The rationale behind this approach has traditionally been based on the belief that the choroid plexus produces a large amount of CSF; by eliminating this source of CSF, the surgeon is probably reducing the amount of CSF needed to be reabsorbed by the arachnoid villi as well as the force applied to the ventricles.

On the other hand, ETV alone may be less effective in infants due to higher brain compliance. Regardless, this technique appears to have a higher long-term success rate and a lower infection rate than primary shunt placement, so it should be considered an effective primary treatment option for congenital idiopathic hydrocephalus. Although the success rate is low at 37%, the lower complication rate compared to shunt treatment may justify this procedure in the initial management of hydrocephalus (Izah *et al.*., 2024)

In addition to this benefit, another factor related to PVD is the high incidence of mesencephalic dysfunction during PVD failure, according to Guida *et al.*,(2023) about 37% of neonates who underwent PVD later had global rostral mesencephalic dysfunction syndrome (GRMSD), this condition is reversible

after ETV/CPC, which restores the physiological transtentorial gradient.

Therefore, all patients with SDMRG showed clinical improvement after ETV/CPC, with regression of hyperintensity on T2 MRI of the midbrain in most cases. Two common contraindications for EVT are communicating hydrocephalus and difficulties visualizing the floor of the third ventricle. However, although complication rates were low, the rate of postoperative seizures was quite high (5.1%), with a prevalence of 15% (Coulter *et al.*., 2021)

CONCLUSION

ETV/CPC is a neurosurgical method that provides treatment for CH in the majority of patients under 1 year of age, and for other types of hydrocephalus in childhood, regardless of etiology. ETV has emerged as an effective alternative, and the addition of choroid plexus coagulation (CPC) increases its success, especially in children under six months of age, making it the most indicated and safest treatment, with the lowest risk of complications for children with congenital hydrocephalus.

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