Modern Trends in the Treatment of Hydrocephalus in Infants

Thesis

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Abstract

The aim of this study is to review the literature and recent publications regarding the endoscopic third ventriculostomy as a treatment of hydrocephalus in infants.

Treatment of hydrocephalus in infants is one of the most common clinical problems in neurosurgical practice. Placement of a ventriculoperitoneal (VP) shunt is still the standard for the surgical management of the disease; yet shunt infection and shunt failure are common problems.

Endoscopic third ventriculostomy (ETV) presents an alternative to shunt insertion. According to published data from several groups, infants younger than 1 year of age have a higher failure rate for ETVs compared with that of older children.

The study was done prospectively on 16 cases of obstructive hydrocephalus in infants younger than 6 months age to assess the success rate of ETV as a primary treatment for hydrocephalus in such age group, in cases of evident failure a VP shunt was applied.

Despite eliminating the factors suggested as causes of failure of ETV in infants below 6 months such as the etiology regarding the communicating type, thickness of the third ventricular floor, history of previous intracranial hemorrhage or CNS infection, still the success rate did not exceed 12.5%.

Key words: Congenital hydrocephalus, obstructive hydrocephalus, endoscopic third ventriculostomy, pediatric neurosurgery.

Contents

List of Abbreviations

VP	Ventriculo-peritoneal
ETV	Endoscopic Third Ventriculostomy
CSF	Cerebrospinal Fluid
ICP	Intracranial Pressure
CT	Computed Tomography
MRI	Magnetic Resonance Imaging
SSAS	Spinal Subarachnoid Space
CSAS	Cortical Subarachnoid Space
NPH	Normal Pressure Hydrocephalus
TID	Three times daily (Ter in die)
QID	Four times daily (Quater in die)
CNS	Central Nervous System

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Introduction and aim of the study

Treatment of hydrocephalus in infants is one of the most common clinical problems in neurosurgical practice. Placement of a ventriculoperitoneal (VP) shunt is still the standard for the surgical management of the disease; yet shunt infection and shunt failure are common problems. Potential risks and complications exist with shunt surgery especially among infants during their 1st year of life.[1,2,3,4]

Endoscopic third ventriculostomy (ETV) presents an alternative to shunt insertion. According to published data from several groups, infants younger than 1 year of age have a higher failure rate for ETVs compared with that of older children.[5,6,7,8] Discussed possible explanations include the plasticity of the developing brain which may lead to occlusion of the ventriculostomy site and the open cranial sutures which may cause failure of the pressure gradient across the ventriculostomy. The conclusion has been drawn that patients younger than 1 year of age are not candidates for ETV and should undergo placement of a VP shunt instead.[65]

We discuss our experience regarding the question of whether the success of ETVs is determined by the age of the patient or by the cause of his or her hydrocephalus.

In this work we were assessing the outcome of ETV in infants below 6 months in a definite etiological entity which is obstructive hydrocephalus which is theoretically the most indicated type for ETV.

Review of Literature

In hydrocephalus, an excess of cerebrospinal fluid (CSF) accumulates within the ventricular system of the brain, and intracranial pressure (ICP) increases as a result. The condition is not a disease but results from various conditions that affect the fetus, infant, and child. Nor is it an “all-or-none” phenomenon. Some children can be managed with observation if the hydrocephalus is mild,[66] and some cases may resolve without treatment.[67]

Adequate treatment for hydrocephalus did not exist before the valve-regulated shunt was developed in the early 1950s. Before then, most children who were born with or acquired hydrocephalus died, and the condition of the survivors was poor. At that time, the diagnosis of hydrocephalus depended on angiography or injection of air into the ventricular system; a significant percentage of patients now seen by neurosurgeons would not even have been diagnosed. They also would not have been captured in natural history studies of patients with hydrocephalus from that era. The natural history of children with stable, moderately enlarged ventricles, with large heads is still unknown.

In 1973, the first computed tomography scanner was installed in the United States. About the same time, real-time cranial ultrasonography began to be performed routinely in neonatal intensive care units. The diagnosis of hydrocephalus no longer depended on invasive and dangerous tests, and milder forms of hydrocephalus were discovered. In the mid-1980s, the development of the magnetic resonance imaging (MRI) allowed neurosurgeons not only to diagnose hydrocephalus but also to determine the actual site of obstruction to the flow of CSF that caused it.

Consequently, the ability to diagnose the underlying condition leading to hydrocephalus has improved, and new treatment paradigms have been developed to manage many of these infants and children.

- **PATHOPHYSIOLOGY:**

To understand the pathophysiology of hydrocephalus, one must first understand the normal dynamics of CSF flow, the interaction of CSF flow with the viscoelastic properties of the brain, and how both relate to cerebral blood flow. CSF is produced at a rate of about 0.33 ml/minute by two distinct processes. First, CSF is produced by an energy-requiring process performed by the choroid plexus in the lateral, third, and fourth ventricles. This process depends on the enzyme carbonic anhydrase and can be blocked by the carbonic anhydrase inhibitor acetazolamide (Diamox). Originally, the choroid plexus were thought to be the sole source of CSF production, but CSF production returns to normal after the choroid plexus have been removed. The remainder of the CSF is produced as a by-product of cerebral and white matter metabolism. The brain and spinal cord extracellular fluid flows into the ventricular system and central canal of the spinal cord by bulk flow. In normal circumstances, choroidal CSF production constitutes 50% to 80% of the total CSF production.

After its production, the ventricular CSF flows through a series of narrowings from one compartment to the next. The compartments begin with the lateral ventricles through the foramina of Monro to the third ventricle. From there it flows through the aqueduct of Sylvius into the fourth ventricle. From there it exits the outlet foramina of the fourth ventricle (Luschka and Magendie) and flows into the cisterna magna, where it mixes with the CSF from the spinal subarachnoid space (SSAS).

Finally, the CSF flows through the cortical subarachnoid space (CSAS) to be absorbed through specialized organs, the arachnoid villi, into the sagittal sinus.

The energy for the circulation of CSF is generated by the pumping of arterial blood into the choroid plexus by the left ventricle of the heart. A parallel circuit involves cerebral blood flow, the total of which is about 1 L/minute, in contrast to the 0.33 ml/minute associated with CSF. Arterial blood is pumped into the choroid plexuses and the parenchymal arterioles at the mean arterial pressure. In both locations, the mean arterial pressure immediately drops to the level of the ICP in the ventricles and the brain parenchyma. The channels between the compartments within both the CSF and the flowing blood pathways can be thought of as resistors in a circuit. Each channel can also be obstructed by some pathological processes that lead to abnormal CSF and ICP dynamics.

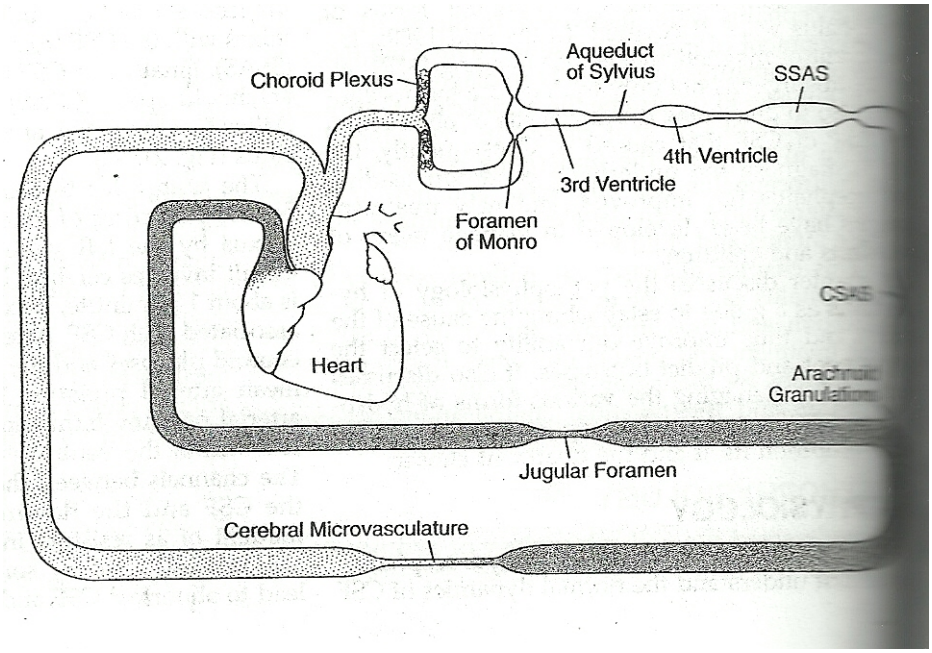

Fig. 1: Schematic diagram of the circulation of CSF as a hydraulic circuit.[9]

Depending on which point along the circuit an obstruction occurs, the differential diagnosis of the cause of the hydrocephalus, the treatment options, and the probable outcome of treatment vary.[68] It is not always possible to determine the point of obstruction at presentation without subjecting a patient to unjustifiable risk or discomfort. However, with MRI, particularly with CSF flow studies, considerable information can be obtained about the physics of the CSF system in an individual patient. Occasionally, a radiographic dye must be injected into the CSF pathways to determine the actual point of obstruction. Further, CSF may be obstructed at more than one point along the pathway. In particular, Chiari II malformations can cause as many as four different sites of obstruction to CSF flow. Severe meningitis can also cause multiple sites of obstruction.

- **CLASSIFICATION BASED ON SITE OF CSF FLOW OBSTRUCTION:**

In the earliest studies of the pathophysiology of hydrocephalus, Dandy and Blackfan[9] classified hydrocephalus into two types: communicating and non-communicating. This classification was based on the injection of a supravital dye into the lateral ventricle and its collection in the SSAS after a spinal tap. Because shunting device were unavailable then, they attempted several operations to treat obstructive hydrocephalus, including an open third ventriculostomy, originally sacrificing an optic nerve and removing the choroid plexuses of the lateral ventricles, which they believed were the source of all the CSF production. Probably owing to the extreme nature of hydrocephalus in the era when transillumination was

the standard diagnostic test, the mortality associated with these operations was high, and the success rate was extremely low.

In 1960, Ransohoff and coworkers recognized the basically obstructive nature of all hydrocephalus and proposed a new classification using the terms intraventricular obstructive (non-communicating) and extraventricular obstructive (communicating) hydrocephalus. Obstruction at the foramen of Monro, aqueduct of Sylvius, and outlet foramina of the fourth ventricle leads to intraventricular obstructive hydrocephalus, and obstruction at the basal cisterns (CSAS to SSAS), arachnoid villi, or venous outflow creates extraventricular obstructive hydrocephalus. Using the imaging modalities now available to neurosurgeons, the classification of the hydrocephalus should reflect the actual anatomic site of obstruction, because each site is associated with different causes and clinical syndromes.

- **Obstruction of One Foramen of Monro:**

Like most other forms of hydrocephalus, unilateral monoventricular hydrocephalus can be congenital or acquired. The congenital absence or constriction of the foramen of Monro is a rare birth defect. Even when severe, it is often missed in infancy because the child is perceived to be normal. The condition is often diagnosed when a disproportionately rapid increase in head circumference occurs. At this point, neuroimaging studies usually reveal a markedly enlarged lateral ventricle on the side of the occlusion. Uniquely in this condition, the calvarium on the side of the enlarged ventricle is greatly expanded, and the falx encases the normal hemisphere far from the midline. Careful neurological assessment often

reveals increased tone of the contralateral upper and lower extremities, a finding that can become more obvious later in life.

Acquired obstruction of the foramen of Monro is caused by mass lesions in the region or by inflammatory processes (bacterial ventriculitis). When obstruction of the foramen of Monro is due to ventriculitis, it is usually part of global compartmentalization of the ventricular system involving multiple sites of obstruction. Very complex treatment strategies are required. Tumors in this location are most common in children include craniopharyngiomas, opticohypothalamic astrocytomas, choroid plexus papillomas, and germinomas. In a few older children and adults, colloid cysts are important causes of obstruction at this location. In such cases, the foraminal obstruction is usually bilateral, yielding the picture of biventricular hydrocephalus.

Probably the most common tumors leading to univentricular obstruction of the foramen of Monro are subependymal giant cell astrocytomas. Although these rare tumors may be the presenting sign of this condition, they are found only in the context of tuberous sclerosis. The tumors arise in the region of the foramen of Monro and are very small and asymptomatic in these patients. Like so many of the tumors associated with the phakomatoses, their natural history is unpredictable. They can, however, grow and cause severe symptoms very rapidly.

Treatment options at presentation: At diagnosis, patients with congenital absence of one foramen of Monro usually have severe unilateral dilatation of one ventricle and a very thin ipsilateral cortical mantle. The pathophysiology of this condition is clear. Because the ipsilateral brain has been chronically subjected to severe thinning, it is unlikely that the cerebral

mantle will be reconstituted significantly on that side by simple perforation of the septum pellucidum. Most neurosurgeons would elect to shunt the affected ventricle, with or without performing the endoscopic septum pellucidotomy at the same time. This approach gives the cortical mantle the best chance to reconstitute significantly and maintains the option of enabling the patient to do without the shunt when it fails.

When a tumor obstructs the foramina of Monro, removal or debulking of the lesion unblocks the points of obstruction, and no shunt is needed after surgical treatment of the tumor itself. Treating hydrocephalus by unblocking the foramen is an important strategy in the case of subependymal giant cell astrocytomas. Uniquely in this case, it is difficult to maintain a working shunt. The tumor tends to block the ventricular catheter, and its protein content upstream from the point of obstruction is very high. Gross total resection of the tumor is probably essential because it is so difficult to maintain a working shunt in this context.

- **Obstruction of the Aqueduct of Sylvius:**

The cause of constriction or occlusion of the aqueduct of Sylvius can be either congenital or acquired. After hydrocephalus associated with spina bifida, the congenital form is the second most common diagnosis of hydrocephalus in utero. In this context, it is usually an isolated and unexpected finding. It can occur in families as a sex-linked condition found only in boys. It is also found in many animal models of hydrocephalus. Pathologically, the usual postmortem description is of a “forked” aqueduct, analogous to attempting to create a tunnel by beginning to dig through the mountain from either end by missing in the center. In both animal models and human studies, hydrocephalus itself has been found to cause further