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Review

Evolution of surgical interventions for hydrocephalus: patient preferences and the need for proper information

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ABSTRACT: The treatment of hydrocephalus has undergone remarkable transformation since it was first documented over two thousand years ago. Currently, the focus of hydrocephalus research is on minimally invasive techniques of treatment. This article reviews the evolution of hydrocephalus therapy, and examines current attitudes towards modern methods. We relied on journal publications, as well as literature on hydrocephalus obtained from the Internet (Google, Yahoo and PUBMED search) making use of the following search terms: "hydrocephalus: history; treatment; complications", "cerebrospinal fluid shunt", "endoscopic third ventriculostomy: indications of; complications of; advantages; disadvantages; successes; failure". Numerous medical and surgical approaches have been adopted in the treatment of hydrocephalus in the past. However, the breakthrough that ushered in the modern era of hydrocephalus treatment was the introduction of valve-regulated systems in the middle of the last century. Endoscopic third ventriculostomy has evolved to become an alternative to traditional shunts. Cerebrospinal fluid shunt procedures are very effective in the treatment of hydrocephalus and have radically transformed the outcome of the disorder. However, they have a number of limitations. The alternative to shunts, endoscopic third ventriculostomy, is relatively safe, effective and durable. It does not, however, succeed in every patient; and also has some potentially devastating complications. Preoperative counseling is imperative to ensure that patients are properly guided.

KEYWORDS: Cerebrospinal fluid shunt; Complications; Endoscopic third ventriculostomy; Hydrocephalus; Indications; Patient selection

INTRODUCTION

Hydrocephalus is a clinical entity characterised by excess accumulation of cerebrospinal fluid (CSF) in the ventricles and subarachnoid pathways, usually as a result of disturbance in its formation, flow or absorption. The term "hydrocephalus" itself is derived from the Greek words *hydro* (water) and *cephali* (head)[1].

Hippocrates (5th century B.C.), widely regarded as the father of medicine, is credited to be the first to document hydrocephalus, the treatment of which he reportedly attempted by ventricular punctures[2,3]. Since then, treatment of the condition has undergone remarkable transformation, partly because of the increased understanding of the anatomy and physiology of the ventricular system and CSF pathways, recent technological advances with accompanying improved instrumentation, and the discovery of biocompatible materials in the last century. Thus, many of the erstwhile methods of treatment which were either unsuccessful or fraught with complications have been replaced by modern management techniques.

An increasing number of neurosurgeons are keying into the new treatments. Similarly, with the increased volume of information available to patients, an increasing number of them now also specifically request for some of the modern

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methods as against traditional means of CSF diversion.

This paper examines some of the major landmarks in the evolution of hydrocephalus therapy, and highlights the role of patient selection and appropriate training of practitioners of modern methods in ensuring optimal results and prevention of some of the potentially devastating complications.

EVOLUTION OF INTERVENTIONS FOR HYDROCEPHALUS

Cases of hydrocephalus were described by Galen (130 – 200 A.D.), who thought that the disorder was a result of extra-axial accumulation of water rather than enlargement of the ventricles – an impression that led to countless errors in diagnosis and treatment[4,5].

Early and medieval Arabian physicians also described the condition. Prominent among these was Abul-Qasim Al-Zahrawi who, in a 30-volume treatise on medicine, dealt with various aspects of neurosurgery, including the diagnosis and treatment of hydrocephalus. He was the first to describe in detail the evacuation of superficial intracranial fluid in hydrocephalic children[4,6].

Vesalius (1514 – 1564), an Italian physician, clarified several aspects of the anatomical and pathological characteristics of hydrocephalus, pointing out that in one of his patients, "the 'water' had not collected between the brain and its surrounding membrane, but within the ventricles of the brain". He also thought that the CSF was a vaporous substance, the "spiritus animalis," which was produced in the ventricles and provided energy and motion to all parts of the body[6-8].

Thomas Willis in 1664, suggested that the choroid plexus was responsible for the production of CSF, contrary to the popular opinion at the time, which also held that the ventricles contained a vapor during life and which, after death, condensed and gravitated to the spaces in and around the brain and spinal cord. Robert Whytt, in the mid 18th century was, however, the first to depict hydrocephalus as a disease. He described several cases which were associated with tuberculous meningitis which he ascribed as cause of the condition[7].

With the advancements in the understanding of the physiology and anatomy of the ventricular system and cerebrospinal fluid in the 19th century[8], the stage was set for the entrance of modern shunting techniques. In 1825, Magendie described the circulation of CSF within the brain (which was later named the "third circulation" by Cushing), and identified the midline foramen of the fourth ventricle. This was followed, three and half decades later, by the description of two additional lateral outlet foramina by Luschka. The detailed description of the meninges, the subarachnoid spaces (including the basal cisterns), the ventricles and the arachnoid villi by Key and Retzius in their anatomical atlas was another major milestone[7].

Treatments

Before the late 19th century, treatment of hydrocephalus was more conservative than active intervention[9]. The early attempts in treatment were mostly unsuccessful owing to poor understanding of the pathophysiology of the condition. Cures were thought to be rare and treatments fraught with complications.

Some of the therapeutic measures that were applied include: intraventricular injection of iodine, head wrapping, carotid artery ligation and phlebotomy. Other reported attempts at medical cures included the use of thyroid extract, dyes, diuretics and purgatives such as rhubarb, jalap and calomel. Most of these failed and it became obvious that the hydrocephalus was primarily a disease best treated surgically, although it was without definitive cure[10].

Reports on many of the early surgical interventions in the literature were neither clear nor could they be substantiated by sufficient evidence[4]. In 1891, Quincke described lumbar puncture as an effective treatment for hydrocephalus[8,10] while Keen was credited with the first description of continuous ventricular drainage[10,11].

What may be regarded as one of the important forerunners of modern day shunts was credited to Mikulicz who in 1893 attempted the first permanent shunt by draining CSF from the lateral ventricle to the subarachnoid, subdural and subgaleal spaces using gold tubes and cat-gut strands[4,10,12].

In 1908, the "Balkenstich Method", a procedure involving perforation of the corpus callosum with drainage of CSF into the subdural spaces was introduced by the German surgeons, Anton and von Bramann. The procedure, however, remained unpopular because of the low cure rates and high mortality associated with it[13].

Other approaches that were attempted by different workers include: drainage of the CSF from the temporal horn of the lateral ventricle into the cheek fat pad, and drainage through the roof of the orbit (ventriculo-orbitotomy). But neither of these met with significant success[10].

Payr, also in 1908, introduced shunting of CSF from the ventricle into the sagittal sinus and jugular veins using vein grafts[14]; and in the same year, Kausch used a rubber conduit to drain the lateral ventricle into the peritoneal cavity. Unfortunately, this did not initially attract much interest[9,10,15]. Heile, at about this time, attempted to divert CSF from the spinal canal to the peritoneal cavity by various techniques including suturing of the gut serosa to the dura mater and the use of conduits like

veins and latex tubes. He was also credited with the first attempt to divert CSF to the urinary system[16]. Matson and his colleagues also reported diverting CSF to the urinary system from the ventricles and lumbar subarachnoid space; but their technique required the performance of nephrectomy, and was associated with several complications like fluid and electrolyte disorders and infections[12,17,18]. Cushing also attempted to divert CSF from the spinal subarachnoid space to the peritoneal cavity or retroperitoneum with silver cannulae passed through apertures in the L-4 vertebral body[8,10].

Several other workers attempted to bypass obstructions within the ventricular system by various methods. Dandy was however, credited with the introduction of third ventriculostomy to bypass aqueductal stenosis[10,12,19] - the technique being later refined by Stookey and Scarff [7,10,12,20].

Torkildsen, diverted CSF by passing a shunt from the lateral ventricle to the cisterna magna (ventriculocisternostomy). Even though this technique was initially successful, its acceptance was limited by the postoperative morbidity which was also considerable[21].

Other workers also attempted diversion of CSF to different sites such as the heart, jugular vein, pleural cavity, thoracic duct, gallbladder, ileum, fallopian tube and salivary ducts[10]. Over time, however, the right atrium and peritoneal cavity became the choice destinations for CSF diversion.

However, the breakthrough that ushered in the modern era of hydrocephalus surgery was the introduction of valves and flow-regulated shunt systems, and biocompatible synthetic materials in the middle of the 20th century[10]. This development enhanced the safe and reliable diversion of CSF devoid of many of the complications of unregulated CSF drainage that preceded it[10,12,22,23].

Since then, there have been remarkable improvements both in the surgical techniques and in the properties of the shunt devices themselves. Numerous systems are currently available; each with some modification designed to improve performance, enhance safety or prevent known complications. More recently, devices with programmable valves for fine-tuning CSF flow rates were also introduced[9].

Before the era of modern shunting techniques and more sophisticated hardware however, children with hydrocephalus generally had a poor prognosis, and majority of them were not offered surgical intervention. Among untreated cases, only about 20% reached adulthood, with the survivors having a 50% chance of living with permanent brain damage. Currently, most children with hydrocephalus reach adulthood with proper shunt management; and studies show that over 50% of these are educable[1].

The effectiveness of shunts is high[24]; and shunting procedures have thus become standard treatment for most types of hydrocephalus, having radically transformed the outcome of the disorder.

In spite of the impressive advances in shunt techniques, hardware properties, diagnostics and follow-up however, shunts still have inherent tendencies for complications, such as malfunction and infection[25,26]; with more than 10% of ventriculoperitoneal shunts having to be revised at some point for various reasons (such as growth of the child, mechanical obstruction of the device, infection, etc)[27]. In addition, the reported long-term shunt-related mortality rate of shunt-treated patients remains high (2.9% to 12.4% at 10 years follow-up)[28].

An often ignored aspect in the follow-up of shunt-treated patients is the psychological burden of having to live with a foreign body (both on the patients themselves and on their families). Clinical data show that this could be quite profound. Eghwrudjakpor and Essien reported the case of a near disastrous alteration in family psychodynamics resulting from the placement of a ventriculoperitoneal shunt in one of their hydrocephalic patients[29,30].

With the limitations of shunts in the treatment of hydrocephalus becoming more obvious, it became necessary to seek plausible alternative methods of treatment of the condition. Currently, the focus of hydrocephalus research is on, among others, minimally invasive techniques of treatment of the condition[2].

ENDOSCOPIC THIRD VENTRICULOSTOMY

One of the significant advances in the modern treatment of hydrocephalus has been the evolution of endoscopy. Its re-entry in neurosurgery in the 1980s and 1990s[31] was partly prompted by the need for precise placement of ventricular catheters. With technological advancements and the miniaturisation of equipment accompanying it, it is now possible to carry out actions at the level of the floor of the third ventricle. This has brought about a renewal of interest in endoscopic third ventriculostomy (ETV) as an alternative treatment for obstructive hydrocephalus[25,26,29.32].

Endoscopic third ventriculostomy, by allowing drainage of cerebrospinal fluid from the ventricles directly into the subarachnoid space[27], re-establishes a physiological route of CSF dynamics. It is not, however, an entirely novel concept. At the beginning of the 20th century, attempts were made to introduce scopes into the ventricular system and remove the choroid plexus[2].

Before the advent of modern valve-regulated shunt systems, third ventriculostomy through open craniotomy (which was described by Dandy in 1922[33]), was the main treatment modality for hydrocephalus. The procedure, however, fell out of

favour because of the high morbidity and mortality associated with it.

Endoscopic third ventriculostomy (ETV) was performed by William Mixter in 1923 with the aid of a urethroscope which he used to examine and perform the procedure in a child with obstructive hydrocephalus[34,35]. Twenty years later, McNickle introduced a percutaneous method of performing the procedure. This resulted in a reduction in the rate of complications while bringing about significant improvement in the success rate[36]. Other developments closely followed with the introduction of the leucotome, stereotactic frame, etc[37].

Currently, ETV is much more refined and is relatively common practice; and has become the treatment of choice in patients (especially adults) with acquired or late-onset occlusive hydrocephalus in many tertiary care neurosurgical centres[4,24].

Indications for endoscopic third ventriculostomy

ETV was initially performed as a bypass procedure for the treatment of hydrocephalus in patients with aqueductal stenosis (AS)[38]; and currently, AS is still one of the major indications for it. Nevertheless, several workers think that ETV should not be used to treat AS because the subarachnoid space in many of these patients is not sufficiently developed[34,39].

The list of reported indications for the procedure includes a wide range of unrelated disorders, all of which have hydrocephalus as the common denominator. They include Dandy-Walker malformation, idiopathic stenosis of the fourth ventricular outlet foramina of Magendie and Luschka, pineal and tectal tumours, hydrocephalus due to intraventricular haematoma or that due to the mass effect caused by intraventricular tumours, posterior fossa tumours; and suprasellar arachnoid cysts. ETV may also have a role in the management of hydrocephalus associated with Chiari malformation, long standing overt ventriculomegaly (LOVA), persistent shunt infection, and intraventricular haemorrhage (IVH) that are resistant to regular shunting techniques. ETV is also thought to be effective in many cases of "slit-ventricle" syndrome as a means of getting the patients to become shunt-independent. In the latter case, it is first necessary to first exteriorise the shunt in order to control CSF drainage until an adequate working diameter of the ventricles is achieved[28,38,40-52].

Patient selection

Despite the resurgence of interest in ETV, not every hydrocephalic patient benefits from the procedure; with wide variation in the success rates being reported by different series. This variation is thought to be partly due to differences in the criteria used for selecting patients[26] and differences in technique. Patient selection thus has a very important role in determining the overall success rate of the procedure. At present, however, there are no common criteria for patient selection[24]; but clinical evidence shows that certain features increase the probability of success (which is defined by shunt independence)[27,38]. These include:

1. The presence of obstructive hydrocephalus. ETV has the highest chance of being successful in patients with pure obstruction of CSF flow within the ventricular system. Preoperative Magnetic Resonance Imaging (MRI) is particularly useful in this respect because it can clearly demonstrate any existing blockade. ETV has also been performed in patients with non-obstructive hydrocephalus. The long-term effectiveness of this indication is however not known.
2. Age of patient above one year. There are few studies which indicate the effectiveness of ETV in infants. Clinical data, however, suggest that the overall success rate in newborn infants is not as high as in older age groups[53]. It is thought that this may be because the child's brain is still developing and the pressure generated within it during this period is no enough to keep the third ventriculostomy patent.
3. Onset of hydrocephalus should be relatively recent. The chances of success appear to be highest among adult patients in this group.
4. There should be no intracranial infection or subarachnoid haemorrhage.
5. The ventricles should be dilated. This would afford easier access of the instruments to the floor of the third ventricle.
6. Ventricular anatomy should be normal.

It is opined that the decision to perform ETV should preferably be taken jointly with the patient or his family after proper counseling[27]. It is important that they be made to understand the limitations of the procedure; and be prepared to have a regular shunt inserted should the ETV fail.

Advantages of endoscopic third ventriculostomy

Endoscopic third ventriculostomy has several advantages over traditional shunts. These have contributed to a large extent to its appeal among neurosurgeons, patients and their families alike.

1. In the first place, the procedure, when successful, eliminates the dependence on mechanical shunt devices with all their limitations; while normalising CSF dynamics in patients with obstructive hydrocephalus. With the absence of foreign object implanted in the body, the risk of infection is

significantly reduced[29]; and there is no risk of tissue reaction.
2. The duration of surgery is comparatively shorter; which means that the patient is subjected to less anaesthesia.
3. The success rate is high among appropriately selected patients.
4. Fewer incisions are needed to perform an ETV.
5. The procedure itself is elegant, relatively low-risk and its performance is comparatively simple.
6. The problem of CSF over-drainage (with the attendant complications such as slit ventricle syndrome) is completely eliminated.
7. The long term complication rate of ETV is less compared to regular shunts. Revisions are also rare. However, some patients need shunt placement to improve despite a patent ventriculostomy[24].
8. Revision of ETV is relatively infrequent.

Complications of endoscopic third ventriculostomy

Even though endoscopic third ventriculostomy has been greatly refined, especially in recent years, the procedure is not without complications. Though they are much fewer, some of the reported complications are quite serious.

In a series of 339 paediatric patients who underwent ETV for hydrocephalus, Beems and Grotenhuis reported a complication rate of 7.7%[45], while Amini and Schmidt reported a complication rate of 14% in their series of 36 adult patients, even though none of the complications in the latter resulted in any permanent sequelae[38].

One of the most serious complications of ETV is injury to the Basilar Artery complex or its perforating vessels, which can result in catastrophic haemorrhage or brainstem infarction[38]. Others include bleeding at the level of the choroid plexus, ventricular wall, ventriculostomy, or interpeduncular cistern. Intraparenchymal or subdural hemorrhage and infection are less common complications[38,54,55].

Some patients who have undergone ETV fail to improve despite a patent ventriculostomy; and many of these need to have regular shunting devices inserted[24]. In some of these cases, a repeat ETV may be indicated. Such repeat procedures are usually effective and safe. But they have also sometimes been associated with complications such as convulsions, pulmonary oedema, raised intracranial pressure and cardiac arrhythmia[51].

Success rates

The reported success rate of ETV varies from 50 to 94%. The success of ETV appears to depend on several factors, e.g. the technique used, the experience of the endoscopic neurosurgeon and, very importantly, the type of patients selected for the procedure[26,37,38,45,56,57]. The chances of success appear to be highest among adult patients with recent-onset aqueductal stenosis (such as those with posterior fossa metastatic disease). The scoring system (the ETV success score), has been found to be very useful in predicting the probability of success of the procedure in the treatment of hydrocephalus

Failure of endoscopic third ventriculostomy

Despite the appeal and attractiveness of ETV, it is not always successful. It is essential that patients or their relatives be made aware of this fact before it is performed. The failure rate is thought to be highest in the first 2 to 3 months following the procedure. Clinical data show that if ETV does not fail during the first few months, the chances of it failing subsequently are considerably reduced[27,58], However, failures have also been reported years later.

In their retrospective analysis carried out to determine the success or failure of endoscopic third ventriculostomy in 170 patients who underwent it as a primary procedure and 63 patients who underwent ETV as a secondary procedure for shunt malfunction, O'Brien et al reported that among the patients in whom the procedure failed in both groups, more than 95% were evident within 1 month of the procedure[58].

One of the commonest causes of failure of ETV is the presence of Liliequist's membrane which may obstruct the outflow of cerebrospinal fluid from the fenestration made in the floor of the third ventricle[59]. In their series of 36 adult patients who underwent ETV, Amini et al found that 22% in whom the procedure was initially successful later had closure of the fenestration with recurrence of symptoms at a mean interval of 3.75 years[38].

A number of other factors predispose to failure of ETV probably by obliterating the cerebrospinal fluid (CSF) pathways. Some of the better known ones include intraventricular haemorrhage, tuberculous meningitis, repeated shunt infections and Chiari I malformation.

Patient follow-up post ETV

Thus, in patients who undergo this treatment, long-term periodic follow-up review should be carried out so as to detect problems early. This is particularly important during the first few months, because studies show that the chance of blockage of the fenestration is most likely at this time. Failure to recognise and promptly treat this complication could be disastrous[38]. It is important that patients and their families are properly informed that the procedure is not a cure for

hydrocephalus. They should also be made aware of the possibility of shunt insertion should the need arise.

THE FUTURE OF ETV

Due to the attractiveness and elegance of ETV, more neurosurgeons now perform the procedure and the number is expected to grow. In view of the obvious advantages, more and more patients and their families now also specifically request for it as an alternative to traditional shunts. The number of such requests is likely to escalate with increase in the amount of information available to patients. It is of utmost importance, therefore, that neurosurgeons be properly informed so as to give appropriate counseling to patients to enable them make informed choices.

Even though the techniques of endoscopic third ventriculostomy have continued to improve, neurosurgeon should also always bear in mind that the seemingly simple procedure holds the potential for very serious complications. Thus, there is need for appropriate training of endoscopic neurosurgeons in order to prevent avoidable complications, and ensure the best results in operated cases. It is imperative, therefore, that surgeons continue to report their experience with the complications of ETV so that the procedure can continue to be made as safe as possible[60].

CONCLUSION

The ultimate goal in the treatment of patients with hydrocephalus is to get them to be shunt-free[28]. Endoscopic third ventriculostomy offers this chance.

The procedure affords a safe, effective and durable method of treatment for hydrocephalus, especially in older children and adults (including those with chronic compensated hydrocephalus and brainstem tumours)[38]. The overall outcome depends on the cause of the disorder.

It is important that preoperatively, there should be adequate counselling of patient and family about all aspects of the procedure, including risks, successes, possibility of failure and the long-term complications.

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