

Chapter 36

Treatment of Hydrocephalus: Mere Survival is Not Enough

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“Hardly any other pathological condition has been accorded more determined attention on the part of the medical profession...than has hydrocephalus. And in hardly a single other condition have cures been so illusive or so often wrecked on purely mechanical obstacles.”

- Leo Davidoff, 1929 [8]

Seventy-five years after Leo Davidhoff made this statement, treatment failure remains the rule, not the exception, in the management of hydrocephalus. Hydrocephalus is the one condition we deal with in neurosurgery where we have made little progress since the development of the shunt valve. We are still using the same basic shunt designs and mechanisms that we have used through all of these years. Shunts definitely save lives. We try to control hydrocephalus, not cure it. There is no question that the ability to place intracranial shunts has benefited many thousands of patients since their development approximately 50 years ago. Our focus, however, in the management of hydrocephalus seems to be on the valve control of cerebrospinal fluid (CSF) flow and less on the correction on the underlying pathophysiology of hydrocephalus.

Hydrocephalus is a disease process that has been with mankind throughout recorded history. It is among the earliest diseases documented in literature (2, 18, 19). Hippocrates, the father of medicine, is thought to be the first physician to actually attempt to treat hydrocephalus. After Hippocrates, there have been numerous references regarding attempts at management of “water on the brain.” Along with trauma, hydrocephalus is one of the oldest documented neurosurgical conditions.

Hydrocephalus has a long history of stimulating research into basic anatomy and physiology. Attempts to control this very difficult and progressive problem led to much of the early scientific interest in anatomy and physiology. By the beginning of the 20th century, the basic understanding of hydrocephalus was expanding rapidly. Although the concept of bypassing or removing obstruction to CSF flow, or draining CSF into other body cavities, was understood by the late 19th century (13, 17, 22), the ability to accomplish this in a safe and effective surgical procedure was not achieved until the mid-20th century. Choroid plexectomy, fenestration of the corpus callosum, and open fenestration of the lamina terminalis were successful in many cases, but the resulting morbidity and mortality was felt to be unacceptable (7, 8, 27). Successful diversionary attempts were limited to lumbar subarachnoid to ureter catheter systems.

The development of valve-regulated shunts in 1952 was a landmark event in the surgical management of hydrocephalus. Nulsen developed a valve regulated shunt system based on a spring and ball mechanism while working with an engineer whose son had hydrocephalus (21). Shortly thereafter, Pudenz et al. (23) developed a one-way slit valve that was successful in the management of hydrocephalus. Since that time, many varieties of shunt valves and shunt parts have been brought forth. Although thousands of lives have been saved through shunt technology, it is also true that the overall design of shunt valves has changed little through the past 50 years.

To understand why mere survival of hydrocephalus is not enough, we must look at the common problems found in

long-term survivors of shunted hydrocephalus. The typical patient with shunted hydrocephalus faces a future of uncertainty because of the one underlying fact that the shunt must continue to function to survive. There is no doubt that the successful treatment of hydrocephalus allows the brain to develop to its fullest potential. If the cause of the hydrocephalus is related to a congenital malformation of the brain, infection, tumor, trauma, etc., the prognosis is most often related to the underlying condition. If, however, the hydrocephalus occurs in an otherwise normal brain, then relief of the intracranial pressure before permanent neurological damage occurs will usually allow full development of brain function. Assuming, of course, that the shunt continues to function appropriately.

Complications of shunting are numerous and too varied to discuss in full at this time. Progress is being made in the management of shunt infection, tolerance to foreign material, and the recognition and treatment of the cognitive outcomes associated with hydrocephalus. It is the management of recurrent malfunction that remains a serious problem for all too many shunted patients, especially those shunted early in childhood. The major contributor to recurrent malfunction is the development of very small or slit-like ventricles and chronic shunt overdrainage. Although overdrainage seems to occur in most patients shunted in childhood, it is the development of craniocerebral disproportion that is the primary underlying pathophysiology.

PATHOPHYSIOLOGY OF SHUNT OVERDRAINAGE

It is well documented that shunts tend to over drain CSF over time (1, 4, 9–11, 15, 20, 24–26, 28). None of the particular valve designs that have been used clinically has eliminated the phenomenon of chronic overdrainage. Some designs seem to allow overdrainage at a slower rate, but all of them eventually allow the ventricles to become smaller and smaller, thus setting up the potential for craniocerebral disproportion and the development of the slit ventricle syndrome (SVS). The definition of SVS has been characterized in numerous ways. It is probably best stated as repeated, intermittent symptoms of shunt malfunction in a patient who seems otherwise healthy. It is characterized by headaches, varying degree of lethargy, and may or may not be associated with nausea or vomiting.

There is rapid brain growth during the first 2 years of life, especially the first 12 months. During this period of time, the brain is grows rapidly, and in the child with a functioning shunt, CSF is being drained from the ventricular system. The ventricles are becoming progressively smaller as the CSF is drained. Small amounts of CSF are pushed out of the cranium through the shunt system with each pulsatile beat of the heart. Eventually, we have a cranium that is filled with brain parenchyma, blood, meninges, and vasculature, but only a small amount of CSF remains (Fig. 36.1). This leads to loss of normal intracranial compensatory mechanisms and the potential for the development of SVS. It has also been documented that, in children younger than 18 years, more than 95% of initial CSF shunt placements are in children younger than 1 year. This leads to a situation where the development of cerebrocranial disproportion seems inevitable in so many children.

Slit ventricle syndrome is a phenomenon that seems to occur only in children shunted early in life. Adults do not seem to get SVS. It is true that adults can develop small ventricles, and they can have symptoms suggestive of over drainage, such as low pressure headaches; however, the full syndrome characterized by a very tight intracranial space and disproportion between the cranium and brain is typically seen only in those children shunted early in life.

If one looks at the head circumference chart for a child shunted in infancy, you note that the occipitofrontal circumference (OFC) is typically above the 95th percentile when the child is shunted. Over time, however, there is

decreased growth of the OFC because of the continued overdrainage of CSF. By the time the childhood years are reached, the head size is often on the small side (Fig. 36.2). This reflects the fact that CSF is lost at an excessive and nonphysiological rate. The normal pulsatile mechanisms are not present to assist in cranial expansion. The resulting cranial growth is purely related to brain growth, hence the development of a tight intracranial space and the loss of normal intracranial compensatory mechanisms.

The incidence of SVS has varied. Walker and Fried (28) found small or slit-like ventricles in 64% of all shunted patients in their study. Nine percent of the patients maintained normal ventricular size, 9% had slightly enlarged ventricles, and 18% had moderate to marked enlarged ventricles. The incidence of surgical intervention for symptomatic SVS was 11% of this population of shunted patients. Therefore, the incidence of SVS requiring intervention was relatively small in this study.

TREATMENT OF SLIT VENTRICLE SYNDROME

The modalities available to treat SVS are many. The fact that there are many options available and yet the problem remains a difficult one is a testament to how ineffective our treatment can be. Although any one option may be effective for a given patient, it remains a bit of a roll of the dice in any given clinical situation. A few of the options available are listed in Table 36.1.

In the landmark shunt design trial, we learned that approximately 40% of shunts will have failed by the end of the first year, and only 50% will still be working after 2 years. Other studies only serve to confirm that shunts fail at an alarming rate.

I believe that the prevention of shunt overdrainage and the subsequent development of slit ventricles is where we must focus our attention for the near future if we are to significantly improve the long-term outcome of children shunted in infancy. Because adults apparently do not develop brain and cranial disproportion (SVS), and because most children shunted in infancy do develop this condition, one has to wonder at what age the shunt could be placed without the expectation of the development of SVS. Because approximately 80% of the brain growth is present by the end of 2 years of life, would placing shunts in patients older than 2 years old have the same risk of the development of slit ventricles as those children shunted who are younger than 12 months? If we shunted children at age 5, 10, or 15, at what point along the growth curve could we expect to see long-term effects mimic more the adult population than that of children?

The prevention of slit ventricles must be directed to the underlying cause, and that treatment must begin early in life. Prevention is the only likely treatment for the near future. If we can, we should delay the development of small ventricles as long as possible. Is this the place for programmable valves? It could be, but we don't have that data. If we were to shunt infants with programmable valves and then periodically assess the size of their ventricles and turn the pressure in the valve up as the ventricles become smaller maybe we could delay the eventual development of slit ventricles for an extended time. It is apparent from other studies that the development of slit ventricles in the typical infant shunted early in life takes approximately one year (16). If we could delay that until 2, 3, or 4 years of age, would we see a different pattern in the long-term outcome of children shunted for hydrocephalus?

OTHER OPTIONS FOR MANAGING HYDROCEPHALUS

Neuroendoscopy

Endoscopic third ventriculostomy (ETV) has become a relatively routine procedure that can be performed in the management of both communicating and obstructive hydrocephalus (3, 6, 12, 14). The proper selection of patients, however, still remains unclear. The best candidates for treatment seem to be those patients who develop an obstructive form of hydrocephalus later in life. Their success is presumably based upon the fact that they have had normal CSF pathways. Removing the obstruction seems to restore flow to these normal pathways. Obstructive hydrocephalus secondary to congenital aqueduct stenosis is not quite so successfully treated. There is still some debate as to whether these children are best treated with ETV in infancy or shunted first and considered for ETV at a later time. There is increasing evidence that ETV can be effective in communicating hydrocephalus, perhaps caused by bypassing obstruction to CSF flow in the basal cisterns and CSF flow pathways around the cranial base. However, it seems certain that patient selection criteria will continue to improve. ETV will remain an important part of the armamentarium of the neurosurgeon to treat obstructive hydrocephalus.

Intrauterine Shunting

Advances in fetal medicine have led to the resurgence of the potential for treating hydrocephalus in an intrauterine environment. Although this has been tried with limited success, it seems certain that advances in maternal and fetal care and appropriate patient selection are leading us again toward the probability of intervening at an earlier stage (5). A current multi-center randomized trial for intrauterine treatment of myelomeningocele is underway. Data from this trial regarding hydrocephalus, as well as the safety of operating on the fetus, will undoubtedly affect our decisions in the near future regarding the intrauterine treatment of fetal hydrocephalus.

Smart Shunts

We must continue to look for other options for children shunted with hydrocephalus. The future will bring other options related to the control of CSF production and absorption. Perhaps different valve designs will be more effective in long-term treatment and eventually the development of smart shunts. Smart shunts will be able to react to appropriate intracranial physiology and will drain or pump CSF in response to changes in intracranial dynamics. These shunts will not drain on a continuous basis, but will more likely drain or pump CSF as required to help control intracranial dynamics.

Basic Research

The need for ongoing research in hydrocephalus has never been greater. We are stuck in the management of hydrocephalus with shunts, and this is an imperfect system. Our understanding of hydrocephalus is changing. We are learning more of what we do not know. We do not know with certainty the site of absorption of CSF. There is increasing evidence that the arachnoid villi are not the primary site of CSF absorption. We have a poor understanding of why the ventricles dilate, even in communicating hydrocephalus. We do not understand the differences between obstructive hydrocephalus, communicating hydrocephalus, normal pressure hydrocephalus, and low-pressure hydrocephalus. Much knowledge is required to help us have a better understanding of this disease and hopefully lead

us to better therapies, both medically and surgically. Mere survival is not enough. We must look to survival with full potential and the best quality of life.

TABLE 36.1. Possible treatments for slit-ventricle syndrome

Observation

Anti-migrainous therapy

Shunt revision (ventricular catheter, valve, or both; flow control device)

Intracranial pressure monitoring and external ventricular drainage

Subtemporal decompression

Cranial morcellation

Endoscopic third ventriculostomy

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FIG. 36.1 Noncontrast computed tomography scan showing the typical findings of slit ventricles. Note the complete collapse of the ventricles around the shunt catheter and the loss of basal cisterns and subarachnoid pathways. There is no room for increased intracranial volume, thus the loss of normal compensatory mechanisms.

FIG. 36.2 A typical OFC growth chart showing declining percentiles of the OFC in a shunted patient. Many patients end up on the small side of the curve because of chronic overdrainage of CSF.