Factors Affecting Ventriculo-Peritoneal Shunt Failure in Children: A Comprehensive Review

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Abstract

Ventriculoperitoneal shunt (VPS) placement is one of the most commonly performed neurosurgical procedures and is necessary to treat most forms of hydrocephalus in children. These may be more common in developing countries due to poor economic conditions and a dearth of available resources. The shunt malfunction in paediatric patients may be caused by number of reasons, including obstruction, infection, pseudocyst formation and bowel perforation. This paper highlights the various factors affecting the successful outcome of VPS in children.

Keywords: Hydrocephalus, ventriculoperitoneal shunt, children, CSF, pseudocyst.

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INTRODUCTION

A ventriculoperitoneal shunt (VPS) is surgical equipment that relieves pressure on the brain caused due to accumulation of fluid. The VP shunting is a surgical procedure that is mainly done to treat a condition called hydrocephalus (Figure 01). This condition occurs when excess cerebrospinal fluid (CSF) collects in the brain’s ventricles. CSF cushions the brain and protects it from injury inside the skull. The fluid acts as a delivery system for nutrients that brain needs, and it also takes away waste products. Normally, CSF flows through these ventricles to the base of the brain. The fluid then bathes the brain and spinal cord before it’s reabsorbed into the blood. When this normal flow is disrupted, the buildup of fluid can create harmful pressure on the brain’s tissues, which can damage the brain [1]. It is then advised to surgically place VP shunts inside one of the brain’s ventricles to divert fluid away from the brain and restore normal flow and absorption of CSF. Excess fluid can build up around the brain for a number of reasons, including:

1. Overproduction of CSF
2. Poor absorption of CSF by the blood vessels
3. Blockages preventing fluid from flowing throughout the brain

Blockages are the most common cause of hydrocephalus. The cysts, tumours or inflammation in the brain can impede the normal flow of CSF and create an unsafe accumulation. Among the procedures performed for hydrocephalus, cerebrospinal fluid (CSF) diversion with insertion of ventriculoperitoneal (VP) shunt remains the most popular method in pediatric patients [2, 3]. Newer methods for treatment of hydrocephalus have also emerged with the most notable being endoscopic third ventriculostomy (ETV)[4]. ETV is considered to be the preferred treatment for non-communicating hydrocephalus, atleast in older children, especially those with aqueductal stenosis [5, 6]. However, the effectiveness of ETV remains questionable in cases of hydrocephalus secondary to infective etiologies, hydrocephalus associated with meningo(myelo)cele [7] and hydrocephalus in infants [8]. Unfortunately, complications related to VPS placement are also common, and multiple shunt revisions are almost expected throughout a patient’s lifetime. Sometime VPS may become a cause of frustration and anxiety for patients, caregivers, and neurosurgeons alike. The failure rates of VPS have been estimated at approximately 10.9–23.7% within the first year after initial shunt placement [9, 10], with most researchers [11] reporting a significantly large number of shunt revisions and replacements in pediatric patients compared to adults. In one of the study [12] conducted over 64 pediatric patients followed for 15 years, only 15.4% of patients did not require a revision during the period of follow up, and 17.3% of patients required 3 or more revisions. Wu Y et al. [13] Applied the database of the Office of Statewide Health Planning and development to identify 14,355 patients who received a VPS during the time of 1990–2000. The results showed...
the highest rate of shunt malfunction in neonates, followed by children, with adults having the lowest rates of shunt malfunction. The major independent risk factor for VPS complications in children is the type of hydrocephalus, with a significantly more risk linked with congenital and obstructive hydrocephalus compared to non-communicating hydrocephalus. Moreover male gender and low socioeconomic status were associated with an increased risk of VPS complications [14].

Inspite of major advancements in VPS including design of valve, improved sterile techniques prevention of shunt malfunction has failed to demonstrate significant improvement over the past few years in children. The frequency of VPS complications in pediatric patients ranged from 45% to 59 % in different reports [15, 16]. The various factors affecting VPS failure in children can be broadly categories as:

Infections of VP shunt
It is the second most common cause of shunt malfunctioning, reported in approximately 8–14% of pediatric patients who undergo VPS placement [17]. The risk factors for shunt infection include young age, postoperative CSF leak, glove holes during shunt handling [18], African American race [19], public insurance, previous shunt infections and etiology of intraventricular hemorrhage. The highest risk for shunt infections was reported in premature neonates in whom the immune system is not fully developed. The majority of shunt infections occur within the first few weeks to several months after VPS placement [18]. Mostly patients will present with symptoms of infection, such as fever, however, this is not always the case. Patients with shunt malfunciton due to any cause may present with nausea, vomiting, fever and headache although fever is more common in patients with shunt infection. McClinton et al. [20] noted the presence of fever and neutrophil count >10% in the ventricular fluid had a specificity of 99%, a positive predictive value of 94%, and a positive probability of test 92% for predicting shunt infection. The most common causative organisms isolated from infected shunts in order of frequency are *Staph epidermidis, Staph aureus*, and gram negative rods [20, 21]. *Staph epidermidis* and *Staph aureus* are common skin flora, and infection typically occurs during shunt placement due to contamination from the skin. Thus, infections due to these organisms usually present relatively early after shunt placement. Standard therapy for shunt infection typically involves removal of the infected hardware and initiation of intravenous antibiotic therapy. A study [22] by pediatric neurosurgeons in the US showed that the large number of surgeons prefer to remove the infected shunt and place an external ventricular drain, while a smaller number prefer to externalize the infected shunt. Antibiotic impregnated catheters are a new strategy to prevent shunt infections from occurring. Raffa et al. [23] studied the incidence of infections in VP shunt in pediatric patients younger than one year and found that the rate of infection in the patients with antibiotic impregnated shunts was 9% while that in patients with standard shunt catheters it was 34%.

Obstruction of VP shunt
VP Shunt catheter obstruction is the most common cause shunt malfunction. This obstruction can occur in the proximal catheter part, within the valve, or within the distal catheter, but, most of the studies [24, 25] proved that the most common site of obstruction site is the proximal catheter. Basically there are two types of valves used in VPS: programmable and non-programmable. In a study [24] after adjusting for age and etiology of hydrocephalus, it was found that programmable valves were associated with a relative risk reduction of shunt revision of 0.54 and that too in proximal catheter obstruction only. It could be due to the ability of the programmable valves to prevent over-drainage by means of non-invasive adjustments, which would theoretically prevent debris from being pulled into the proximal catheter.

Also the proximal catheter part of VP shunt may become clogged with parenchyma of brain as it is
passed from the cortex into the brain ventricle during its surgical placement. Other theories hold that the proximal catheter may be blocked with pieces of choroid plexus when placed near the foramen of Monroe. Dickerman et al. [25] studied 115 shunt revisions performed at their institution to see if placement of the proximal catheter within the anterior horn of the lateral ventricle, farthest from the choroid plexus would be associated with a lower rate of shunt malfunction at 6 months, but this association was not found, although they did find the lowest rate of shunt malfunction to occur among patients with normal pressure hydrocephalus (NPH). These authors theorized that the patients with NPH had large ventricles, so proximal catheters were less likely to become blocked with choroid plexus.

Another important cause of VP shunt obstruction is the debris, because of blood and proteinaceous fluid may slowly accumulate within and eventually block the thin catheter tubing. This is supported by the findings of few studies [26] that showed that shunt malfunction was slightly more frequent among patients of hydrocephalus with intracranial hemorrhage.

The shunt obstruction diagnosis is mostly done with a combination of CT, shunt series, and shunt tapping or lumbar puncture (LP). The CT is likely to show increased ventricular size and LP may be suggestive of an elevated opening pressure, but this is not always the case [27]. Rocque et al. [28] noted that poor flow of CSF during a shunt tap had a 92% positive predictive value of proximal shunt obstruction.

Abdominal Pseudocyst

Abdominal pseudocyst is a rare complication reported in 1 to 44% of VPS patients [29]. It is often a late complication, with most cases occurring after a year or more of VPS placement or revision. Pseudocysts are formed due to collections of fluid around the tip of the distal catheter part of VPS which are surrounded by a fibrous tissue wall without an epithelium covering. They are believed to arise because of the effect of inflammation and abdominal adhesions that may occur with infections of shunt or due to multiple abdominal surgeries [30]. The most common clinical feature of a VP shunted patient with an abdominal pseudocyst is abdominal pain and distention with a palpable mass, although rarely neurological symptoms may be present. Sometimes, abdominal pseudocysts are hepatic pseudocysts, which can occur when the distal catheter tubing shifts to the surface of the liver and induces chronic irritation and inflammation, leading to right upper quadrant pain worsening after meals which may be accompanied by elevated liver enzymes [31]. The diagnosis of abdominal pseudocysts is made with either ultrasonography or abdominal CT scan. Dabdoub et al. [32] found pseudocyst recurrence in 18.8% of children in whom the distal catheter was repositioned into a non-peritoneal space.

Perforation of Bowel

Bowel perforation by catheter tubing during VPS placement is estimated to occur in 0.1 and 0.6% cases. Most common presentation is protrusion of the tip of the distal catheter through the anus. In children, the majority of cases of bowel perforation occurs by the distal catheter, this could be due to the fact that children are having a relatively thin bowel wall, especially with myelomeningocele in whom innervation of the bowel wall may be insufficient [33].

The stiffness of the catheter tubing is also an important factor which contributes for bowel perforation during VPS. One study [34] compared the stiffness of three common shunt catheters, the Medtronic, Chhabra, and the Codman bactiseal shunt catheters, the results showed that Chhabra shunt catheters are the least stiff among the three; however they do have the highest frictional force.

Bowel perforation by catheter tubing has also occurred due to allergy to silicone. Although it is rare, but patients with such an allergy who receive VPS could develop bowel and abdominal wall perforations and other skin erosions around the distal tip of catheter.

Overdrainage and subdural hematoma

The formation of subdural collections and subdural hematoma can occur after VP shunting procedures as a result of overdrainage of CSF. The subdural hematoma formation incidence is 5% in high pressure hydrocephalus patients. One study [35] found serious over-drainage to be significantly more frequent in NPH patients with an opening pressure of >16 cm of water compared to those with a lower opening pressure. This suggests that patients with NPH and higher ICPs may be more sensitive to increased CSF drainage and thus more likely to develop subdural hematoma. The development of programmable shunt valves has offered a major advantage in treating symptoms of over drainage and subdural hematoma formation without the need for surgery in many cases.

CONCLUSION

The VP Shunt malfunction continues to be a common neurosurgical problem in children with shunted hydrocephalus, often leading to lengthy hospital stays. To prevent morbidity, early detection and proper management are key. VPS obstruction, which most often occurs in the proximal catheter, is the most common cause of VPS malfunction while hardware infection is the second most common. With continued advancements in shunt systems and infection prevention, morbidity resulting from shunt complications may continue to be reduced.
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