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Ventriculoperitoneal Shunt Infections in Patients with Hydrocephalus

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Ventriculoperitoneal (VP) shunt infections can be detrimental to the pediatric patient with hydrocephalus and his or her family. Although there are many causes of hydrocephalus in the pediatric patient, the standard treatment of care is surgical placement of a VP shunt. Current treatment modalities when a ventriculoperitoneal shunt infection has occurred and the differences between the causes of late and early shunt infections are reviewed. Most importantly, the signs and symptoms that the families of the child undergoing a VP shunt must know during the postoperative period, and the tools they can use to help combat and prevent VP shunt infections are discussed.

Infections of cerebrospinal fluid (CSF) shunts in pediatric patients are associated with significant mortality and morbidity (Baird, O'Conner, & Pittman, 1999; Barnes, Jones, Hayward, Harkness, & Thompson, 2002; Vinchon, Lemaitre, Vallee, & Dhellemmes, 2002). Shunt infections also are the cause of frequent stressful hospitalizations for many children and their families.

Case Study

Imagine Samantha. She has hydrocephalus and has just been readmitted to an acute care facility because her ventriculoperitoneal shunt (VP shunt) has become infected again and has to be replaced. She is only seven years old, but she already has had nine major neurological operations because of shunt malfunctions and infections. She's a bright, beautiful girl who would rather be playing with her toys, playing dress up, painting, or drawing. However, over the next several days, she will need to have her VP shunt externalized from her cranium to drain off her infected CSF. Daily cultures of her CSF will be drawn to

check the status of her infection. She will need a combination of caustic intravenous antibiotics to eradicate the bacteria responsible for her infection. She's going to miss school, miss playing with her friends, and be surrounded by strangers for weeks. After her infection is clear, she has more surgery to which she can look forward. Perhaps this time the shunt won't become reinfected. The grim truth is that it most likely will. Samantha could be any pediatric patient admitted with a VP shunt infection. Sadly, many children with hydrocephalus are readmitted again and again due to repeated shunt infections and shunt malfunctions.

What is Hydrocephalus?

Hydrocephalus is derived from a Greek word literally meaning "watery head." Hydrocephalus is a condition not a disease. Although there are numerous causes, most are due to impaired CSF absorption, CSF adsorption, or alternate CSF absorptive pathways. In many cases, the cause remains unknown. Regardless of the cause, the result is an abnormal or excessive accumulation of CSF around the brain. Table 1 summarizes the risk factors associated with hydrocephalus. According to the Hydrocephalus Foundation (2004), hydrocephalus is fairly common in children and accounts for approximately 1.5 per 1,000 live births. However, because no national registry or database for hydrocephalus exists, it is difficult to establish the prevalence and incidence. The National

Institute of Neurological Disorders and Stroke (NINDS) (2004) estimates that hydrocephalus affects one in every 500 children.

Hydrocephalus can be congenital or acquired. Congenital hydrocephalus occurs during fetal development and can be the result of complex interaction of genetic and environmental factors. Hydrocephalus is associated with infections during the prenatal period, especially toxoplasmosis, cytomegalovirus, and rubella. It is also associated with hemorrhage or internal bleeding particularly when compounded with prematurity. Bleeding, infection, trauma, tumors, vascular problems, and structural anomalies have been strongly associated with the development of hydrocephalus (NINDS, 2004). Hydrocephalus is also associated with numerous pediatric diseases, especially spina bifida. Acquired hydrocephalus in children can occur at birth or anytime during the child's life. Some of the more common causes of acquired hydrocephalus in the pediatric patient are infection, meningitis, tumor, or head injury.

Certain medical conditions are highly associated with hydrocephalus, including aqueductal stenosis, neural tube defects, myelomeningocele, tumors, arachnoid cysts, and Dandy-Walker syndrome. The most common type of congenital hydrocephalus is aqueductal obstruction or stenosis. The stenosis results from a narrowing or blockage from the aqueduct, or it may be caused by infection, hemorrhage, or a tumor. This blockage causes the CSF to accumulate upstream producing hydrocephalus. Spina bifida, or neural tube defects (NTD), refers to the condition in which the vertebrae, muscles, and ligaments that support the spinal cord are impaired, not the spinal cord. A myelomeningocele is an open NTD that is highly associated with widespread abnormalities of the central nervous system, including Chiari II malformation and hydrocephalus that occur in 90% of NTDs (Fudge, 2002). In Chiari II malforma-

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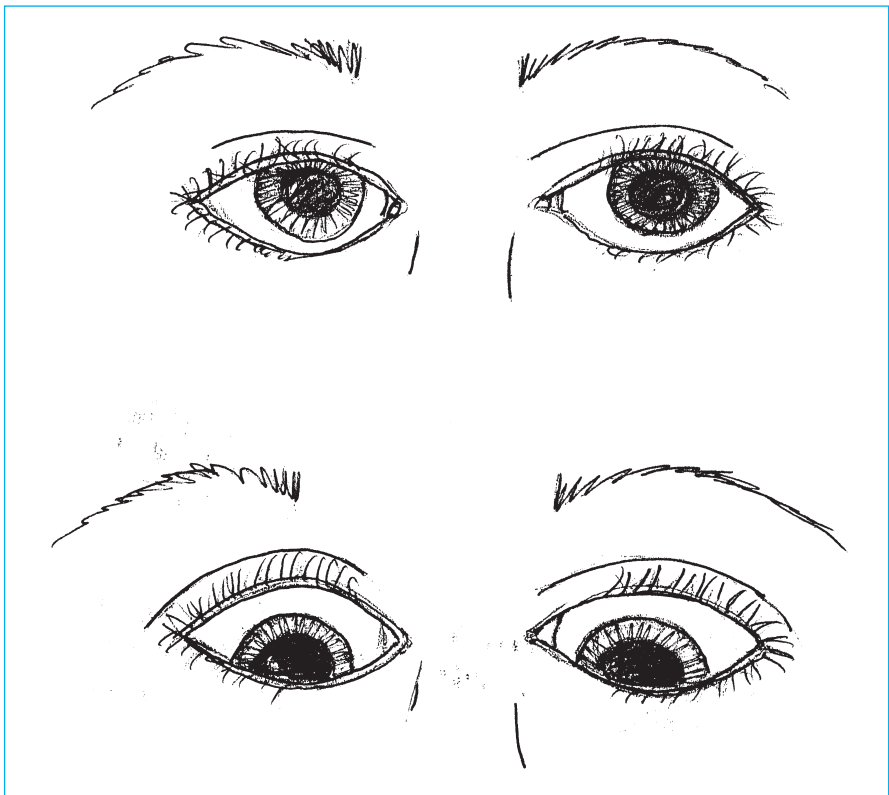
The CE Posttest
can be found
on pages 468-469.

Table 1. Risk Factors Associated with Hydrocephalus

- Idiopathic
- Meningitis
- Traumatic head injuries
- Brain tumors
- Complications associated with prematurity and fetal exposure
 - Intraventricular hemorrhage
 - Subarachnoid hemorrhage
 - Infections during fetal development (toxoplasmosis, CMV, rubella)
- Genetic inheritance, structural anomalies, and neural tube defects
 - Aqueductal stenosis
 - Spina bifida
 - Chiari II malformations
 - Myelomeningocele

Sources: Fudge, 2002; NINDS, 2004.

Figure 1. Drawing of Normal Eyes Verses “Sunsetting” Eyes



tions, the flow out of the fourth ventricle is blocked. In children, brain tumors are most commonly found in the back of the brain (Fudge, 2002). The tumor fills or compresses the fourth ventricle as it grows, which then blocks the CSF flow out of the fourth ventricle. Arachnoid cysts can occur anywhere in the brain; however, in children they are usually found in the back of the brain near the third ventricle. These cysts have entrapped fluid that may block the CSF pathways. In Dandy-Walker syndrome,

the fourth ventricle is enlarged because of partial or total closure of its outlets. Children with this syndrome also typically have a portion of the cerebellum that fails to develop. The syndrome also can be associated with abnormal, or lack of, development of other parts of the brain. Most children with this particular syndrome require two shunts, one placed in the lateral ventricle and one placed in the fourth ventricle (Fudge, 2002; Neurology Channel, 2004).

Types of Hydrocephalus

There are two main types of hydrocephalus that affect pediatric patients: noncommunicating and communicating. Noncommunicating hydrocephalus is basically of an obstructive nature. The CSF pathways within the ventricles become blocked and obstructed. Although this blockage can occur anywhere in the ventricular system, it usually lies either within the narrow passageways connecting the ventricles or where the CSF exits the fourth ventricle into the subarachnoid space. The small opening of the fourth ventricle fails to develop or develops immaturely, which also can lead to an obstruction in the outflow of CSF. In communicating hydrocephalus, the CSF moves freely around the ventricles, but it is blocked after it attempts to exit the ventricles. The CSF also becomes impeded as it passes over the brain. Although the absorption sites are blocked, the CSF moves freely throughout the ventricles, and the ventricles “communicate” with each other (Fudge, 2002; NINDS, 2004).

Symptoms of Hydrocephalus

Symptoms of hydrocephalus vary with age, disease progression, and individual differences in tolerance to CSF (NINDS, 2004). Infants and toddlers with hydrocephalus are most often identified during well child exams because of the infant’s rapidly enlarging head. The infant may have a problem holding his or her head up. The skin over the scalp appears shiny and thin with prominent scalp veins. Infants also may present with signs of increased intracranial pressure such as a bulging anterior fontanel, splitting sutures, and a phenomenon known as Macewan’s crackpot, the sound that is heard with head percussion. Infants may present with early signs such as vomiting, sleepiness, and irritability, and late signs involving failure to thrive, delay or loss of developmental milestones, seizures, and eyes that deviate downward called “sunsetting” eyes (see Figure 1). In contrast, infants less than 2 weeks of age often exhibit no symptoms (NINDS, 2004). Table 2 summarizes the more common signs and symptoms of hydrocephalus found during early infancy.

In late infancy and with older children whose skull bones have fused, symptoms may include nausea, irritability, drowsiness, chronic headaches, urinary incontinence, sunsetting eyes, seizures, and vision disturbances. Vision problems may include papilledema, blurred vision, and

Table 2. Signs and Symptoms of Hydrocephalus in Early Infancy

- Asymptomatic
- Sleepiness
- Rapidly enlarging head
- Persistent vomiting
- Failure to thrive
- Irritability
- Delay or loss of developmental milestones
- Downward deviation of eyes, "sunseting" eyes
- Seizures

Sources: Fudge, 2002; NINDS, 2004.

diplopia. Initially, these children may present with problems concerning balance and poor motor coordination. Some children with hydrocephalus present with delays in walking or speech and have learning disabilities throughout the learning years of childhood. They may also exhibit changes in personality or cognition, including memory loss (NINDS, 2004). These signs and symptoms of hydrocephalus in late infancy and childhood are summarized in Table 3. It is also important to remember that these symptoms account for the most typical ways hydrocephalus manifests itself and that symptoms can vary from individual to individual.

When left untreated, the CSF circulating around the brain accumulates, leading to an increased level of pressure inside the cranium vault that can result in substantial brain damage if allowed to accumulate. However, with treatment, the excessive accumulation of CSF can be diverted to prevent brain injury.

Treatment Standards for Hydrocephalus

The most common and effective treatment of hydrocephalus involves a surgical placement of a shunt by a neurosurgeon. Shunts were first introduced in the 1950s specifically for the treatment of hydrocephalus (Schreffler, Schreffler, & Wittler, 2002). In acute care facilities in the U.S., hydrocephalus accounts for 70,000 hospital admissions (McGuirt et al., 2003). There are over 25,000 shunt operations performed each year in the United States, and of these, 18,000 are operations for initial shunt placement (Hydrocephalus Foundation, 2004). Because of this effective surgical procedure, morbidity and mortality

rates in this population have fallen (McGuirt et al., 2003; Tuli et al. 2000). The shunt allows the excess and obstructed CSF to be drained from around the brain. This modified drainage of CSF prevents the dangerous accumulation of CSF, the continual rise in intracranial pressure, and subsequently, brain injury.

Shunts typically transfer and drain the CSF from the lateral ventricles of the brain and empty the CSF into the peritoneal cavity. Less commonly and more risky, the shunt can be placed to drain the CSF into the right atrium of the heart, ureter, pleura, fallopian tube, or gall bladder (Hydrocephalus Association, 2003). Generally, placement of a VP shunt is preferred because it has fewer risks than alternative placements and is easier to surgically perform. Physicians generally choose other sites only if the usual VP site cannot be used (for example, if there is excessive scarring in the peritoneum from multiple shunt or abdominal surgeries), or if another site is required for a particular child's circumstances. The shunts are made of silastic tubing and are approximately 1/8 inch in diameter. VP shunts are placed subcutaneously in the fatty portion just below the skin. A small hole is surgically placed in the cranium, and the ventricular end of the shunt is gently placed in the lateral ventricle. The distal end is then passed to the abdominal cavity through a small opening in the perineum. Shunt systems come with a variety of options such as programmable valves, precision valves, anti-siphon devices, and antimicrobial impregnated catheters, but all have similar functions (Hydrocephalus Association, 2003). As with any foreign object placed in the body, infection is potentially a dangerous life-long complication.

Endoscopic Third Ventriculostomy

Endoscopic third ventriculostomy (ETV) is a procedure in which a small opening is made in the thinned floor of the third ventricle. This allows movement of the CSF out of the obstructed ventricle system. This opening then allows for a free flow of the CSF into the basal cisterns for absorption. ETV is not a cure for hydrocephalus and is not the appropriate choice of treatment for many types of hydrocephalus (Hydrocephalus Association, 2004). The concept of ETV is not new, but because of the improvements in endoscopic equipment and the ability of a MRI to visualize brain anatomy, there

Table 3. Signs and Symptoms of Hydrocephalus in Late Infancy and Childhood

- Nausea
- Chronic headaches
- Irritability
- Blurred vision
- Papilledema
- Diplopia
- Drowsiness
- Delays with walking or speech
- Urinary incontinence
- "Sunsetting" eyes
- Changes in personality or cognition including memory loss

Sources: Fudge, 2002; NINDS, 2004.

is new enthusiasm for this procedure. However, there are many limitations. According to Fudge (2002), one limitation is that many neurosurgeons will not perform the procedure on children below the age of two years. The five-year patency rates of ETV range from 50 to 80%. The initial complications rate of ETV is much higher than that of shunt placement; these complications are also higher than the associated risks of shunts malfunction. Even when successful, children with ETV need to have periodic neurosurgical evaluations. ETV primarily is used as an alternative to shunting with obstructive types of hydrocephalus in older children. Research is ongoing to determine the benefits of ETV in children with communicating and/or non-communicating hydrocephalus.

Treatment Standards for Shunt Infections and Obstructions

VP shunts can malfunction for a variety of reasons. Malfunction can be due to obstruction, equipment error, or infection. Even with the progressive improvement in the treatment of hydrocephalus patients, many pediatric patients require numerous shunt revisions due to infection or malfunction. Shunt infections are generally diagnosed by the combination of symptoms the child has and a positive CSF (shunt) culture. The initial CSF specimen is usually obtained from a shunt tap. The neurosurgeon or specially educated nurse usually performs the shunt tap. The shunt tap also can give valuable clues to the neurosurgeon on the viability of the shunt.

According to Fudge (2002), infection is usually caused by the child's own bacterial organisms and not acquired from exposure to other children or adults who are ill. The majority of these shunt infections occur within several months of the initial operation. Only about 10% of shunt infections occur more than one year after the last shunt operation, with most occurring within eight months of the initial shunt surgery or revision (Baird, O'Conner, & Pittman, 1999). The primary bacteria culprit for shunt infections is *Staphylococcus epidermidis* (Fudge, 2002). Isaacman, Poirier, Hegenbarth, Lillis, and Scarfone (2003) report that shunt infections typically result from contamination at the time of surgery, and that 80% of infection is present in the first 6 months after placement.

McGuirt et al. (2003) examined 301 patients who had undergone VP shunt revisions because of primary shunt failures. The ratio of primary shunt insertion to subsequent surgical revisions was 1:3.6. In a subgroup of pediatric patients, the study found more than 12 revisions were required over each patient's lifetime. Although this surgery is considered clean, the infection risk is extremely high. In many instances, the failure rate after one year was 40% (McGuirt et al., 1999).

Risk Factors for Shunt Malfunctions and Shunt Infections

According to Tuli et al. (2000), identifying the specific risk factors that make a patient more prone to develop a shunt malfunction or infection has been poorly researched. Their study examined 1,183 shunt failures in 839 pediatric patients with hydrocephalus. Results showed that the risk factors most closely associated with repeat shunt surgeries were the patient's age at the time of their initial shunt surgery, the cause of the patient's hydrocephalus, and the time from the previous shunt surgery. Being very young at the time of initial surgery placed children at a higher risk for developing subsequent shunt failures requiring additional surgery. Additionally, repeated episodes of shunt failure within six months was 1.5 times higher than those occurring after six months.

Treatment for Shunt Infections

The most appropriate treatment when a shunt infection does occur is controversial. According to Schreffler et al. (2002), in the 40 years since

shunts were first introduced, only one randomized trial with the goal of validating the most effective treatment has been published. Schreffler and colleagues' research examined the three most common methods of treatment for shunt infections and found that the most effective method is to immediately remove the infected shunt, place an external ventricular drain, and administer IV antibiotics until the CSF sterility is obtained. After CSF sterility is maintained for several days, a new shunt can then be safely placed. Research is underway to determine if removal of the total shunt is necessary or if removal of the distal or proximal portion is sufficient. McGuirt et al. (2003) examined reutilization of the distal and proximal aspects of an infected shunt and concluded that repeated shunt failures were greater when the distal catheter was reused. An antimicrobial impregnated shunt catheter is in use; however, to date, no published data or clinical trials confirm a decrease in infection rates.

Causes of Early and Late Infections

Controversy also surrounds the criteria for determining what constitutes late versus an early infection. Baird et al. (1999) examined early shunt infections; these occurred within a year of surgery and specific causative organisms were identified. The most common organisms cultured in early shunt infections are *S. epidermidis*, *S. aureus*, *diphtheroids*, and *E. coli*. The skin is the portal of entry for the majority of early VP infections, and most early shunt infections are believed to be the result of skin flora introduced during surgery (Isaacman et al., 2003). Meticulous operative technique is thought to be the best strategy to prevent shunt contamination.

In late shunt infections, after one year of shunt placement, the incidence of infection is less understood, and the various sources of infection have been poorly studied. Vinchon et al. (2002) examined 1,793 pediatric patients who developed shunt infections more than one year after surgery. In many cases, it was found that the origin of the infection was unclear and the relationship of the previous surgical experience unrelated. However, many patients with late shunt infections were found to have peritonitis. Often, the etiology is related to peritonitis and most associated with an infected, inflamed, or ruptured appendix. Other sources of infection included hematogenous,

bowel perforation, and direct inoculation. The direct inoculation was an isolated case that was due to the child having a head injury, which damaged the skin overlaying the shunt. There have been documented cases of children developing a shunt infection secondary to acquiring an ear infection, adolescent acne, or tonsillitis (Vinchon et al., 2002). Despite the cause, late infections remain an ongoing, life-long risk to any patient with a shunt placement.

Why Are Shunt Infections So Critical?

Pediatric patients with VP shunts are an especially vulnerable population. Children are developing and growing not only physically but also psychosocially. As with placement of any foreign body, patients that have had a shunt placed are always at an increased risk for developing an infection. The placement of the shunt makes the infection more likely to involve the CSF fluid and cause brain injuries. Because of the constant threat of CSF infections, patients undergoing shunt surgery and their families need to know how to maintain optimal health and nutrition. Although over half the patients with hydrocephalus are at a high developmental level of functioning, others are neurologically compromised or developmentally delayed. These developmentally challenged children are at an even greater risk of having undetected shunt problems because they may have difficulty communicating their distress. Parents need to be made aware that maintaining optimal overall health is important, and given the proper tools and resources to help their child achieve it. Nutritional support, physical therapy, and occupational therapy are resources that the parent will need, and access should be made available to them.

Preventing Shunt Infections

Preventing a postoperative shunt infection in the hydrocephalus pediatric patient is critical. Common sense dictates that stringent hand washing and the use of universal precautions must be enforced to help prevent health care-acquired infections, especially in the immediate postoperative period. Parents should be educated on the importance of this practice when they or others are in direct contact with their child. Standard practice does not indicate that special immunization schedules are needed for shunted children. Unless they are in the immediate postoperative phase,

these children are not at increased risk during flu season or after dental work just because of their shunt placement.

Although prior research indicates that most infections occur due to surgical technique, no new research indicates that early shunt infections are due to any other cause. However, it is prudent for the bedside nurse to maintain aseptic techniques when performing incision care and to also reinforce the importance of maintaining this aseptic technique to the parents. Parents also need to be educated on proper site care management, the correct way to cleanse the wound sites, and what signs and symptoms to look for postoperatively.

Treatment regimen will vary depending upon the standing protocol for the institution and surgeon preference. To date, research has not identified which postoperative wound care and or site care techniques are most effective in preventing infection in a patient with a shunt placement. Incision care uses different methods and products to cleanse the incision depending on the institution where the surgery was performed and the surgeon's preference. Techniques using betadine, antibiotic shampoos, hydrogen peroxide, antibiotic ointments, and plain soap and water have been used; however, to date, none of them have been researched adequately for their efficacy.

Signs and Symptoms of Shunt Infections

Most importantly, families need to understand what to do when their child demonstrates signs or symptoms of a shunt malfunction or infection. The parents and caregivers of children with hydrocephalus are the first line of defense against brain injury, especially when their children cannot verbally express themselves. First, parents need to understand the importance of timely treatment, and should be instructed to contact their child's neurosurgeon immediately if symptoms occur. If the child exhibits signs and symptoms when the office is closed, they should go to the nearest emergency room indicating that the child has had a shunt placed and clearly outlining the signs or symptoms the child is exhibiting. The importance of prompt medical intervention cannot be stressed enough to parents. According to Barnes et al. (2002), death, blindness, or major neurological insult can occur with any delay in seeking proper treatment. ReKate (1991) states that prompt treatment after a shunt revision is even more

Table 4. Signs and Symptoms of Shunt Malfunction/Infection

- Any drainage or odor from the incision sites
- Any tenderness or bogginess at incision sites
- Fever
- Headaches
- Nausea or vomiting
- Abdominal pain
- Irritability
- Sleepiness or drowsiness
- Changes in school performance
- Any change in level of consciousness
- "Sunsetting" eyes

Sources: Barnes et al., 2002; Fudge, 2002; Isaacman et al., 2003; and ReKate, 1991.

Table 5. (Example) Parent and Caregiver Discharge Teaching Tool

1. Date and Type of shunt placed _____
2. Cleanse incisions as per order (example):
 - i. Wash entire head with antibacterial shampoo every day.
 - ii. Cleanse incision with providone iodine three times a day, days 1-13.
 - iii. Starting on the 14th day, cleanse with 1/2 strength hydrogen peroxide three times a day. This begins on day MM/DD/YYYY.
3. Follow up with (Neurosurgeon) Dr. _____ in 2 weeks, call XXX-XXXX for appointment.
4. Report **immediately** any of the following to your child's doctor or go to the nearest emergency room:
 - Any drainage or odor from the incision sites
 - Any tenderness or bogginess at incision sites
 - Fever (greater than 100.8°/orally)
 - Headaches
 - Nausea or vomiting
 - Abdominal pain
 - Irritability
 - Sleepiness or drowsiness
 - Changes in school performance
 - Any change in level of consciousness
 - Downward deviation of eyes – "sunsetting" eyes

critical than before the first shunt surgery. The patient may have only hours between the onset of symptoms, coma, and then death. Parents also need to give this important information to their child's teachers and other caregivers.

Sometimes, differentiating between a shunt infection or a shunt malfunction is difficult because shunt malfunctions can mimic the signs and symptoms of a shunt infection (Barnes et al., 2002). However, this distinction should not be an issue for parents because they do not need to diagnose their child. They need to be educated and empowered with the information to report when something is wrong with the shunt's performance. Parents should be instruct-

ed to immediately report any ominous signs and symptoms. Barnes et al. (2002) found that a shunted patient that presents with any one of these three cardinal signs – headache, vomiting, or drowsiness – needs immediate evaluation. Presentation of any of these symptoms strongly suggests that a shunt malfunction has occurred. According to Isaacman et al (2003), children with VP shunt malfunction present to the emergency department with a myriad of symptoms. If a child has had a previous shunt malfunction, the parent will often state, "He/she is acting just like the last time the shunt malfunctioned." Barnes et al. (2002) found that health care providers should especially listen to parents and primary

care givers of patients with hydrocephalus. The families were found to be at least as accurate as the primary care physicians in diagnosing a shunt malfunction.

According to Fudge (2002), shunt infection frequently results in fever and may occur alone or in conjunction with a shunt obstruction. Infection also may present with reddening or swelling along the shunt tract. However, Fudge reported a high incidence of a clinical presentation of abdominal pain in patients with an infected VP shunt; yet, there was no report of fever. In older, verbal children, the initial complaint of a shunt malfunction often is a progressively worsening headache (Rekate, 1991). Parents and teachers should suspect shunt malfunction and/or infection if the child complains of headaches, sleeping in school, personality changes, or loss of any previously acquired developmental skills (Rekate, 1991). Parents, teachers, and staff should be educated to report any of these signs and symptoms immediately. Table 4 outlines the signs and symptoms indicating that a shunt malfunction or infection may have occurred.

What to Do if a Shunt Infection Or Malfunction Occurs

Patients do not have to exhibit all of these signs; any one of them could constitute a shunt malfunction and/or infection. If any of these symptoms occur, the patient needs to be examined immediately by their pediatric neurosurgeon. Many pediatric neurosurgeons perform shunt taps so they can better assess the shunt viability, check pressures, and assess for infection.

Taking care of hydrocephalus patients with a ventriculoperitoneal shunt can be challenging, rewarding, and fulfilling. Frequent admissions to hospitals because of shunt infections and/or malfunctions also can make delivering care frustrating. By properly educating the family, the bedside nurse is in a unique position to have an impact on patients' and families' lives. The nurse needs to empower and educate the family of the importance of aseptic techniques when taking care of their child's surgical site. The nurse also needs to stress the importance to the family that their child should maintain optimal health with proper nutrition and exercise. Most importantly, the nurse needs to supply the families with life-saving information of the signs and symptoms of a shunt malfunction

and/or infection. Families and patients must recognize the importance of acting quickly if any of these signs occur.

Conclusion

Nurses need to standardize and perfect the techniques of site care, postoperative care, and education at the patient's bedside. Table 5 is an example of a shunt discharge tool for parents and caregivers. It states what time the shunt was placed and the date of insertion, and also provides written instructions for families for follow up care, as well as warning signs and symptoms that need immediate intervention. When families recognize the symptoms of shunt problems and respond promptly, a better outcome for the life of their child is achieved.

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