Cerebrospinal Fluid Shunt Problems in Pediatric Patients

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As more and more pediatric-aged patients survive what used to be considered untreatable diseases, pediatricians need to be more familiar with some of the technological advances made in caring for these children. The placement of cerebrospinal fluid (CSF) shunts is currently the most commonly performed neurosurgical procedure, with an estimated 18,000 placed each year. These shunts are placed to allow the diversion of CSF into another area of the body outside the brain. Unfortunately, placement of these shunts is accompanied by a high rate of complications, which include malfunction, obstruction, infection, malposition, and migration. This article discusses the indications, complications, recognition, and management issues concerning these shunts for the practicing pediatrician.

CEREBROSPINAL FLUID CIRCULATION

Cerebrospinal fluid is a clear aqueous solution which is an ultrafiltrate of plasma that bathes the brain and spinal cord. The low specific gravity of the CSF in relation to the brain reduces the mass effect of the brain to a minimal amount and thus serves as a protective cushion to prevent the brain's full weight from producing traction on nerve roots, blood vessels, and delicate membranes. The CSF also provides the chemically appropriate environment necessary for neurotransmission and the removal of metabolic products. Approximately 70% of CSF is produced by the choroid plexus located in the lateral, third, and fourth ventricles. The remainder is formed in extrachoroidal sites such as the ventricular ependyma, Sylvian aqueduct, subarachnoid pial surface, and brain and spinal cord parenchyma. The rate of formation is approximately 0.35 to 0.40 mL/minute or 500
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tumors is an extremely rare cause of hydrocephalus. Hydrocephalus is usually caused by an obstruction of CSF flow between its point of production within the ventricles and its point of absorption in the arachnoid villi. Hydrocephalus has been described as communicating and noncommunicating. Communicating implies that the obstruction lies in the region of the basal cisterns, arachnoid villi, or subarachnoid spaces, which blocks the resorption of CSF. The ventricles are patent and freely communicate with each other. Noncommunicating implies that the blockage is proximal to the foramina of the fourth ventricle (Lushka and Magendie). CSF thus accumulates in the lateral and third ventricles because it can no longer drain by way of the usual pathway (Figure 1).

The earliest descriptions of hydrocephalus date back to the accounts of Hippocrates and Galen. However, it was not until the 20th century that extracranial diversion of CSF became a possibility. To quote McLaurin, it can be said that "the history of the evolution of ventricular shunting for hydrocephalus is largely a history to prevent the complications of shunting." There are a variety of conditions that may cause hydrocephalus. These include brain tumors, congenital malformations (ie, Arnold-Chiari malformations, Dandy-Walker cysts, aqueductal stenosis, Vein of Galen arteriovenous malformations), and inflammatory processes (infections, hemorrhage). Aqueductal stenosis is a condition where the long, thin aqueduct of Sylvius (Figure 1) is blocked. This may be from a tumor, infection, or a central nervous system (CNS) hemorrhage. Arnold-Chiari is a condition where part of the cerebellum and fourth ventricle extend down through the foramen magnum and cause obstruction of CSF flow by compressing the fourth ventricle. Infections such as bacterial meningitis may cause blockage of the resorption of CSF and lead to a communicating hydrocephalus. Approximately 30% of all infants with periventricular-intraventricular hemorrhage will develop ventriculomegaly and 20% to 50% of these patients will require a CSF shunt. Head trauma may cause an intracranial hemorrhage, which will lead to obstruction of CSF flow and produce hydrocephalus. Central nervous system tumors in children commonly occur in the posterior fossa and may cause hydrocephalus (See the article entitled "Pediatric Brain Tumors" by Valentino, Conway, Shiminski-Maher, and Siffert on


to 600 mL/day. The turnover time for CSF is approximately 5 to 7 hours. The actual intraventricular volumes are approximately 40 to 60 mL in infants, 60 to 100 mL in young children, 80 to 120 mL in older children, and 100 to 160 mL in adults. CSF moves from the point of production through the CSF pathways by way of several mechanisms: (1) via a pressure gradient that exists between the site of formation (15 mm H2O) and the site of resorption at the superior sagittal sinus (9 mm H2O), (2) cilia on the ependymal cells, (3) vascular pulsations, and (4) respiratory variations. After its formation in the lateral ventricles, CSF passes through the paired intraventricular foramina of Monro into the midline third ventricle, then through the aqueduct of Sylvius and into the fourth ventricle, and ultimately into the subarachnoid space by way of one of two pathways. These include an egress of CSF through the foramen of Magendie or the paired foramina of Lushka of the fourth ventricle. The CSF then bathes the brain and spinal cord and is ultimately resorbed on the superior surface of the brain by the arachnoid villi that drain into the superior sagittal sinus (Figure 1).

HYDROCEPHALUS

Hydrocephalus occurs when there is an imbalance in the production and absorption of CSF in the brain. The overproduction of CSF by choroidplexus
The most important caveat to remember is that in any patient with a CSF shunt in place, the shunt is the source of the problem until proven otherwise.

pp. 579-587). Infants with a Vein of Galen malformation have a large arteriovenous shunt. This leads to the shunting of blood from the high pressure arterial side to the low pressure venous side, ultimately leading to an elevation of the cerebral venous pressure that will cause a decrease in the resorption of CSF into the cerebral venous system. Dandy-Walker cysts are congenital cysts that occlude the fourth ventricle and produce noncommunicating obstructive hydrocephalus. The most effective treatment for hydrocephalus is placement of a CSF shunt; however, these devices are not without complications.

CEREBROSPINAL FLUID SHUNT SYSTEMS

The pediatrician should be familiar with three types of major complication of CSF shunts: mechanical failure of the shunt device, infection, and functional failure from either over- or underdrainage. A general knowledge of the basic shunt system is important for the physician who cares for a child with a shunt. The system usually consists of three basic parts: a radiopaque silastic ventricular catheter, which is placed into the ventricle; a one-way valve; and a distal portion that is made of radiopaque silastic tubing (Figure 2). There are a large number of shunt systems available and each neurosurgeon has his or her own preference. You should be familiar with the systems in use in your area. Make a note in your records of the type of shunt and valve the patient has. The patient and their parents should be familiar with this information as well.

The tip of the ventricular catheter has multiple small perforations to allow CSF to drain freely. This portion of the shunt connects to a unidirectional valve that allows movement of CSF out of the brain and the CSF passes through the valve and ultimately into the distal portion of the shunt. Each valve has a different opening pressure. The pressures are classified as (1) a low-pressure valve that needs 2 to 4 cm H2O to open, (2) a medium pressure valve that requires 4 to 6 cm H2O to open, and (3) a high-pressure valve that requires 8 to 10 cm H2O to open. The advantage of these different valves is to allow the neurosurgeon a choice in determining the drainage pattern of the CSF through the shunt. Each patient with hydrocephalus is different, and this is borne in mind when a particular shunt system is chosen. The ultimate goal is to have a system that prevents over- and underdrainage. Some systems have reservoirs that allow a source from which one can access CSF measure intracranial pressure (ICP), and evaluate and possibly decompress a malfunctioning shunt. The third component of the shunt system is the distal tubing, which is longer than the proximal portion and usually tracked subcutaneously down into the peritoneum. Ventriculoperitoneal shunts are the most effective and dependable shunts for children with hydrocephalus. However, if the peritoneal cavity cannot be used because of complications such as infection, adhesions, or peritoneal malabsorption, other sites must be considered. These include the cardiac atria, pleural space, bladder, ureter, stomach, fallopian tube, and gallbladder.

VENTRICULOATRIAL SHUNTS

Following the introduction of polyethylene tubing in the late 1950s, ventriculoatrial (VA) shunts became the standard treatment for hydrocephalus. These were placed from the cerebral ventricle into the internal jugular vein and right atria. Data accrued over subsequent years demonstrated that serious and life-threatening complications are more frequent in children with VA compared with ventriculoperitoneal shunts. These complications have included pulmonary embolism secondary to thrombus formation, endocarditis, cor pulmonale, cardiac arrhythmias, septic
TABLE

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<thead>
<tr>
<th>Symptoms of Shunt Malfunction</th>
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<tbody>
<tr>
<td><strong>Infants</strong></td>
</tr>
<tr>
<td>Enlargement of the baby's head</td>
</tr>
<tr>
<td>Fontanel is full and tense when the infant is upright and quiet</td>
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<td>Prominent scalp veins</td>
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<td>Swelling or redness along the shunt tract</td>
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<td>Fever</td>
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<td>Vomiting</td>
</tr>
<tr>
<td>Irritability</td>
</tr>
<tr>
<td>Sleepiness</td>
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<tr>
<td>Downward deviation of the eyes</td>
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<td>Seizures</td>
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emboli, migration of the catheter into the coronary sinus, and shunt nephritis.

VENTRICULOPLEURAL SHUNTS

The pleural space is yet another option for the distal catheter of the shunt system. Potential problems arise from the fact that pressure in the pleural space is subatmospheric during inspiration and thus excessive CSF drainage may occur. Pleural effusions, proximal shunt obstruction, subdural hemorrhage, and symptomatic intracranial hypotension may occur. Other considerations include coexisting pulmonary disease, respiratory reserve, and derangements of ventilatory control, particularly in patients with kyphoscoliosis and Chiari malformations. A pleural effusion may place these patients into severe respiratory failure.

LUMBOPERITONEAL SHUNTS

The advantages of LP shunts include the lack of need for cerebral cannulation, the possibility of using regional rather than general anesthesia, freedom from proximal obstruction by choroid plexus and brain parenchyma, low infection rate, and the ability to place the shunt in the presence of slit ventricles. However, in addition to the usual complications of shunting (eg, obstruction, infection, overdrainage, and underdrainage), these shunts have unique complications, including arachnoiditis resulting in hyperlordosis, back pain, root involvement, scoliosis, and acquired tonsillar herniation.

SHUNT MALFUNCTIONS

Shunt malfunction is the most common complication of CSF shunts. The risk for shunt failure is greatest in the first months following placement. The mean survival time for a ventriculoperitoneal shunt is 5 years, and approximately 80% of patients will require a revision by 10 years. Malfunctions occur as the result of obstruction (proximal and distal), infection, over- and underdrainage, mechanical malfunction, and from shunt migration. The most common signs and symptoms include headache, vomiting, nausea, altered mental status, lethargy, and a general feeling of malaise. Many of these findings are age related (Table). The most important caveat to remember is that in any patient with a CSF shunt in place, the shunt is the source of the problem until proven otherwise. A parent of a child with a shunt who states that their child is not acting or looking right is often correct in making the diagnosis of shunt malfunction.

OBSTRUCTION

Obstruction is the most common cause of shunt malfunction. Shunt obstruction may be life-threatening and it must recognized and treated immediately. The obstruction may occur at either the proximal end (the portion of the shunt in the ventricle) at the interposed valve or at the distal end (usually in the peritoneum). Proximal obstruction occurs much more frequently than distal obstruction. It is usually caused by the proximal tip of the catheter becoming occluded with choroid plexus,ependymal cells, glial tissue, brain debris, fibrin, or blood, or if the tip migrates into the brain parenchyma. The valve between the proximal and distal tubing may also become blocked with brain debris or tissue colonization. Distal obstruction may result from kinking of the tubing, disconnection of the shunt tubing, migration of the catheter outside of the peritoneum, intra-abdominal infection, pseudocyst formation, or being clogged with debris.
Obstruction may also be associated with an infection in almost one third of cases.

INFECTION

Shunt infections are a major cause of morbidity and mortality. The incidence of infection is between 5% and 8% in different series. Infection may involve the shunt equipment, the wound, the CSF, or at the proximal site where the shunt drains. Risk factors for infection include the age of the patient, condition of the skin, presence of other sites of intercurrent infection, type of operation (the highest rate is seen in operations for previously infected shunts), a shunt ending requiring revision (higher rate for ventricular portion of the catheter), and postoperative wound dehiscence or scalp infection. Most shunt infections (70%) present within the first 2 months following surgery, and that number increases to almost 90% by 6 months following shunt insertion or revision.

The organisms most frequently implicated in shunt infections are staphylococci. Approximately 40% of shunt infections are caused by Staphylococcus epidermidis and 20% by Staphylococcus aureus. Other organisms include streptococci, enterococci, gram-negative rods, and yeast. The pathogenesis of shunt infections includes, in addition to colonization, possible retrograde and hematogenous spread of organisms. Because the majority of infections occur in the first 2 months following surgery, the consensus is that there is endogenous spread of organisms located on the patient's or operating room staff's skin at the time of the surgery. The surgical wound had been found to be colonized in 58 of 100 cases before closing. Thirty-two of these patients had the identical organism found in their nose, ear, or scalp preoperatively, and the remainder were presumed infected by aerial contamination.

After insertion, the shunt becomes coated with a glycoproteinaceous film with material produced in the serum. This serves as a potential binding site for bacteria. These bacteria reproduce and produce a biofilm on the shunt surface, which will ultimately lead to a shunt infection. The fact that the bacteria are embedded in this biofilm makes it very difficult to eradicate an infection. Bacteria that are deeply embedded are not eradicated by systemic antibiotics alone. They continue to reproduce and will ultimately lead to another infection. S. epidermidis is able to produce a slime (glyocalyx). This allows it to adhere to the shunt material and evade the body's immune defenses. This slime explains why it is such a common cause of shunt infections. Electronmicrographic studies have shown that the seemingly smooth surface of the shunt actually is filled with irregularities in the form of pits and lumps that may offer "shelters" for the bacteria. There is also a relative lack of phagocytosis in the CNS. This is due to the fact that the leukocytes do not adhere well to the shunt material, and that pols recognize the shunt material itself as foreign, causing them to discharge their antibacterial enzymes (myeloperoxidase) against the shunt and rendering them ineffective against invading microorganisms.

Clinical signs depend on the site of the infection. Wound infections manifest with fever, reddening of the incision or shunt tract, and, as they progress, discharge of pus from the incision. Patients with meningitis or ventriculitis usually have fever, headache, irritability, and neck stiffness or nuchal rigidity. Peritonitis is less common, and these patients present with fever, anorexia, vomiting, and abdominal tenderness. The clinical symptoms also depend on the infecting organism; S. epidermidis-infected patients look rather well. Their complaints may be similar to those seen in patients with shunt obstruction (Table). A thorough history and physical examination must be performed to exclude other possible infections (otitis media, pharyngitis, gastroenteritis, urinary tract infection, viral syndrome).

The evaluation of a patient with a suspected shunt infection, in addition to the history and physical examination, should include a complete blood count, blood cultures (if appropriate), and a specimen of CSF. Other markers of infection and inflammation have been studied, like C-reactive protein, but they are not specific or sensitive exclusively for shunt infections. Following consultation with the neurosurgeon, a shunt tap should be done. The area of the shunt site is prepped in a sterile manner with betadine and alcohol. A 23-gauge butterfly needle is then inserted into the valve and an opening pressure obtained. If no fluid is obtained, this implies a proximal shunt obstruction. The CSF should be sent to the laboratory for cell count, culture, Gram's stain, glucose, and protein. Controversy exists as to the exact number of white blood cells that may be seen in a patient with a shunt infection, but in infected patients, the mean number of white blood cells was 79 cells/mm³. Eosinophilia has also been associated with shunt infection as eosinophils are not usually present in the CSF.

The pediatrician and neurosurgeon should discuss the main treatment options for CSF shunt infections: (1) should the shunt hardware be removed, (2) does the patient need an intermittent external ventricular drain (EVD), and (3) should intraventricular antibiotics be given. The combination of CSF shunt removal, interval antibiotic treatment, and an EVD carries the highest shunt infection cure rate and the
lowest mortality and is the treatment of choice.\textsuperscript{10,11} After appropriate cultures have been obtained, intravenous antibiotics should be administered. A good starting regimen includes vancomycin for staphylococcus coverage. A third-generation cephalosporin or aminoglycoside can be added if gram-negative organisms are suspected. Patients with gram-negative infections appear much sicker; the source of these organisms is usually the gastrointestinal tract. The antibiotic treatment may be modified as the culture results become available. Intraventricular antibiotics should be considered in patients with CSF cultures that are positive for organisms after 3 days of intravenous antibiotics. Reinternalization of the shunt may be performed after three subsequent negative CSF culture results have been demonstrated.

**SLIT VENTRICLE SYNDROME**

Slit ventricles are a complication of a functioning shunt. In fact, the shunt works too well and there is overdrainage of CSF (Figure 3). Risk factors for developing slit ventricles include patient's age at time of shunt implantation, existence of brain atrophy, prior shunting, and brain compliance and biomechanical properties of the skull. Slit ventricle syndrome develops in only a small number of patients who have slit ventricles. It is due to CSF overdrainage with a growing brain that fills the intracranial space. The intracranial space is comprised of the brain, blood, and CSF, covered by the inelastic dura and encased in a bony skull. The Kellie-Monro hypothesis states that if one of these components changes, one or both of the other compartments must change to maintain equilibrium in the cranial space (See “Accidental Head Injury” by Zuckerman and Conway on pp. 621-632). The growing brain does not allow for the presence of much CSF, and thus these patients become very sensitive to any changes that may affect brain fluid content, cerebral blood flow, and the rate of CSF drainage through the shunt. Symptoms are similar to those of increased ICP. Mechanisms that may explain the symptoms include intermittent obstruction, low pressure syndrome, and vasomotor instability.

In patients with intermittent obstruction, the slit ventricles collapse around the catheter, and the ventricular walls obstruct the catheter. The ventricles then slowly fill with CSF, the ICP rises, and the ventricles open a small amount, allowing the shunt to work again. These patients usually have headaches and nausea, become lethargic, but lie down for an hour or 2 and then feel fine. Patients with low pressure syndrome are sensitive to changes in position. They overdrain when they are in the upright position. Symptoms usually include an intense headache. When the patient lies down (horizontally), the overdrainage is treated. Vasomotor instability implies that any changes in cerebral blood volume will produce symptoms. Migraines may do this by causing

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*Figure 3. (A) CT demonstrating normal ventricular system. (B) CT demonstrating hydrocephalus and ventricular dilatation. (C) CT demonstrating slit ventricles.*
cerebral vasodilatation, resulting in an increase in cerebral blood volume. Management of patients with slit ventricle syndrome includes observation, avoiding shunts if possible, avoiding overdrainage, and migraine therapy. Only a small number of patients need surgical revision of the shunt.12

OTHER COMPLICATIONS

Other complications that may occur in patients with shunts include disconnections and fractures (Figures 4 and 5). As patients grow, the shunt must be free to move under the skin. Fixation at any point will create tension in the material, which may then fracture or disconnect. Patients with dislocated or fractured shunts may have a fibrous tract that formed around the shunt tubing and allows the CSF to flow through the gap between the broken catheters. Abdominal complications have included migration into the scrotum and inguinal canal, small bowel obstruction, intussusception, abdominal pseudo-cysts, volvulus, and perforation of the colon13 (Figure 6). It must be recalled that shunt placement is a blind procedure and the catheter may be inadvertently misplaced. Subcutaneous fluid collections usually result from shunt obstructions, large ventricles, large dural openings, and overdredsection of the subcutaneous tissue to tunnel the catheter, particularly in young infants with loose skin. Overdrainage of CSF may cause the brain to shrink away from the dura and lead to a tearing of the bridging veins producing a subdural hematoma. Subdural collections may also be found following a surgical procedure or following head trauma.

EVALUATION AND MANAGEMENT OF PATIENTS WITH MALFUNCTIONING SHUNTS

Patients who present with possible shunt obstruction may show signs of increased ICP such as headache, nausea, vomiting, lethargy, and papilledema. Other presentations may be more vague and include blurred vision, back or neck pain, gait disturbances, and personality changes. The pediatrician should involve the neurosurgeon early in the evaluation process. Young infants may demonstrate an enlarging head, poor head control, full bulging fontanel, engorgement of head veins, Macwesn's sign (cracked pot sound heard on percussing over the dilated ventricles), sixth cranial nerve palsy, hypertensive reflexes, irregular respirations with apnea, and the "setting sun sign" (upward gaze palsy). Seizures
have also been noted, but their predictive value for a shunt malfunction is questionable because many children with shunts have underlying CNS disease which may predispose them to seizures. One should evaluate these patients for not only shunt malfunction, but anticonvulsant levels as well. Patients who present with seizures as a sign of shunt malfunction usually have several other clinical signs such as fever, headache, vomiting, and respiratory compromise.14 The evaluation of these patients begins with a complete history and physical examination. An important part of the examination is to palpate over the shunt apparatus and to determine, if possible, the type of shunt and if a valve is present. Once the valve is located, its patency is assessed. An easily depressed valve suggests there is distal patency, and a rapid refill of the chamber implies that there is proximal patency as well. Studies have shown that the sensitivity of this examination was only 18% to 20% for shunt obstruction and the value of a negative pumping test (indicating patency) was only 61% to 82%, thus making this an unreliable sign of shunt malfunction.15

The second portion of the evaluation is to perform an imaging study that includes a "shunt series or survey." This is a lateral radiograph of the skull and a radiograph that includes the neck, thorax, and abdomen. The purpose of this series is to ascertain the location and connections of the shunt apparatus (Figures 4 and 5). A head CT is usually also done. However, it is imperative to have a prior study to evaluate the size of the patient's ventricles (Figure 3). Ventriculomegaly may persist in some patients despite a functioning shunt, and, conversely, some patients have shunt malfunctions and high ICP despite small or unchanged ventricles on CT. The pediatrician must remember that a negative CT does not rule out a shunt obstruction.

A head sonogram may be useful in a younger patient. Again, however, one needs an earlier study to compare it to. Studies that evaluate "flow through the shunt" may be falsely negative in patients with partially obstructed shunts because the development of raised ICP is related to an increase in outflow resistance and not necessarily the complete shutoff of the outflow.16 The force used to inject the contrast material may be enough to clear out the obstructing plug.

After all of the above have been completed and the case discussed with a neurosurgeon, an attempt to aspirate fluid from the shunt must be done. It must be fluid from the shunt that is used. It cannot be taken in a sterile fashion as described earlier in this article. An elevated opening pressure or the inability to withdraw CSF implies a shunt obstruction. CSF should be sent for cell count, culture, Gram's stain, and a glucose and protein assessment. Once shunt obstruction has been demonstrated, shunt revision is necessary.

If the child is clinically stable and has only mild signs of obstruction, the neurosurgeon may opt to wait 6 to 12 hours to rehydrate the patient before going to the operating room. These children should be closely monitored in a pediatric intensive care unit if one is available as they may decompensate quickly. Other medical therapies could include the administration of mannitol, furosemide, and/or acetazolamide. Obviously, if the child becomes more obtunded or comatose, has focal neurologic findings, posturing, or autonomic instability (hypertension and bradycardia), the patient should be resuscitated following the ABCs (airway, breathing, circulation) and the revision performed immediately.

CONCLUSIONS

As more and more children survive previously life-threatening CNS diseases, the number of shunt-dependent children will increase. The major complications of these shunts include obstruction and infection. The signs and symptoms of obstruction range from mild (headache and nausea) to life-threatening signs of intracranial hypertension. Fever may only be present in half of those patients with shunt infections. The most important concept to remember is that in any patient with a CSF shunt, the shunt is the problem until proven otherwise. These children are difficult to evaluate and we need to work closely with our neurosurgical colleagues to allow the best possible outcomes for these children.

Addendum

For anyone interested in obtaining more information on hydrocephalus and ventriculoperitoneal shunts, please contact the "Hydrocephalus Association" located at 870 Market Street, Suite 955, San Francisco, CA 94102.

REFERENCES