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Case Report

Importance of Preoperative Evaluation in the Diagnosis of Ventriculoperitoneal Shunt Malfunction in an Infant with Hydrocephalus

Paul M. Nowak[1], Irim Salik[2], Jared B. Cooper[3], Michael Jacoby[2], and Samuel M. Barst[2]

ABSTRACT

Hydrocephalus is a commonly encountered condition in the pediatric population, characterized by impaired circulation of cerebrospinal fluid (CSF), or rarely, overproduction. Treatment of hydrocephalus consists of CSF diversion via an endoscopic third ventriculostomy or by the creation of a shunt. We present the case of a 7-month old female with a history of ventriculoperitoneal (VP) shunt placement, who was found to have symptoms of elevated intracranial pressure (ICP) upon presentation for eye exam under anesthesia. The present case details the importance of the anesthesiologist in the identification of VP shunt malfunction in an infant during a preoperative assessment to avoid potential neurologic injury.

From the [1]Department of Anesthesiology, [2]Department of Pediatric Anesthesiology, [3]Department of Neurosurgery, Westchester Medical Center, Pleasantville, NY, USA.

Correspondence to Dr. Paul M. Nowak at paul.nowak@wmchealth.org.

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Hydrocephalus is generally classified into 3 groups: congenital, acquired, or idiopathic, each of which may lead to ventricular enlargement and increased intracranial pressure (ICP) (1). Symptoms of elevated ICP include vomiting, poor feeding, failure to thrive, bradycardia, and apneic spells; older children may complain of headaches, diplopia, and lethargy. Sudden death has also been reported in the literature (1-4). Surgical intervention is indicated and includes shunt placement consisting of a proximal ventricular catheter connected to a valve, which is then connected to a distal catheter that most commonly diverts cerebrospinal fluid (CSF) to the peritoneum (Figure 1, 2). Shunts are regulated by a series of valves which control outflow to help keep CSF within physiologic pressures. Shunts are prone to malfunction; approximately 50% will encounter some kind of problem within the first two years, with the most frequent complication being an obstruction (5). This case recounts the recognition and management of acute VP shunt

obstruction by identifying signs of elevated ICP. In compliance with the Health Insurance Portability and Accountability Act (HIPAA), a written authorization to disclose existing protected health information was obtained.

Case Description

A 7-month-old, 6 kg female was scheduled for an eye exam under anesthesia in preparation for a left corneal transplant. The patient had a history of holoprosencephaly and previously had a VP shunt inserted at 6 weeks of age. Past medical history was notable for an atrial septal defect, a ventricular septal defect, and bilateral corneal opacifications, diagnosed as Peters anomaly. The patient remained in the neonatal intensive care unit 2 months after birth. She had undergone a right corneal transplant 1 month prior to this admission and was scheduled for an exam under anesthesia in preparation for a left corneal transplant. During the preoperative evaluation, the patient's father noted that the patient was less playful and interactive over the last few days, exhibiting poor feeding and multiple episodes of emesis. Physical exam was significant for general inactivity with minimal eye-opening, bulging of the anterior fontanelle, and bradycardia with a heart rate in the 60s. The most recent visit with her neurosurgeon was approximately 1 month ago, where brain magnetic resonance imaging (MRI) demonstrated stable findings.

Suspecting a malfunctioning VP shunt, an emergent neurosurgery consult was requested by the attending pediatric anesthesiologist. A stat computed tomography (CT) scan revealed extensive hydrocephalus with marked dilation of the lateral and 3rd ventricles, thus confirming shunt malfunction (Figure 3). The patient was brought to the operating room (OR) for an emergent shunt revision. Standard ASA monitors were applied, and the patient was induced through a 24-gauge IV with propofol and rocuronium. Intubation was atraumatic and achieved via direct laryngoscopy with a 3.5 cuffed endotracheal tube. Anesthesia was maintained with controlled ventilation delivering sevoflurane at a minimum alveolar concentration of 2. End-tidal carbon dioxide was maintained at about 33-34 mmHg until the patient was spontaneously breathing. Acetaminophen and morphine were administered for

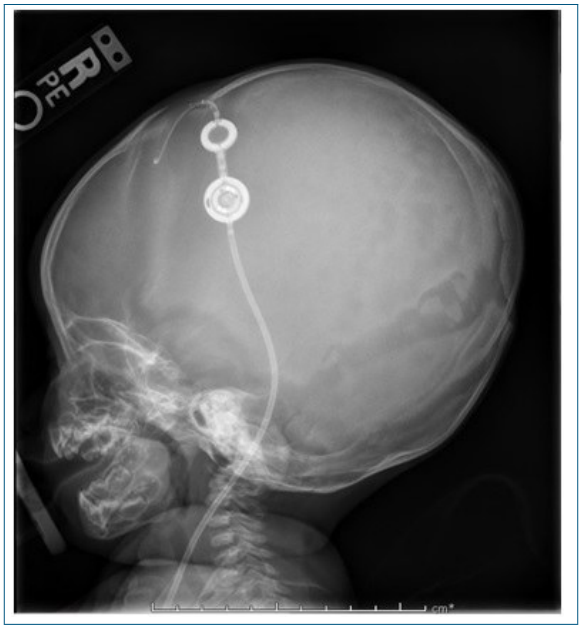

Figure 1. Shuntogram Demonstrating Ventricular Portion of Ventriculoperitoneal (VP) Shunt, Seen Here Terminating in the Right Lateral Ventricle.

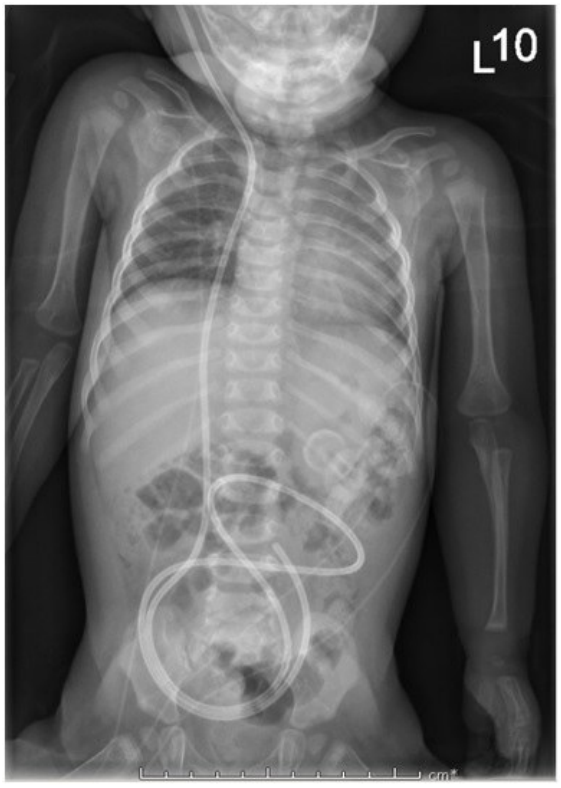

Figure 2. Peritoneal Portion of VP Shunt.

analgesia. The bradycardia experienced during the preoperative period promptly resolved during the case and the patient remained hemodynamically stable throughout the procedure.

Upon interrogation of the VP shunt valve and catheters using manometry, a valve malfunction was diagnosed, prompting replacement. A new ventricular catheter was inserted using neuroendoscopic guidance, and an intraoperative shunt tap confirmed adequate function. At the conclusion of the procedure, the patient was reversed with 0.4 mg of neostigmine and 0.04 mg of glycopyrrolate. Additionally, 4 mcg of dexmedetomidine was administered to facilitate emergence. The patient was extubated and transported to the pediatric intensive care unit in stable condition. A postoperative MRI demonstrated interval improvement in ventricular size. The patient progressed over the course of her admission; repeat brain MRI demonstrated a continued decrease of ventricular size, and the patient was discharged home on postoperative day 6 (Figure 4).

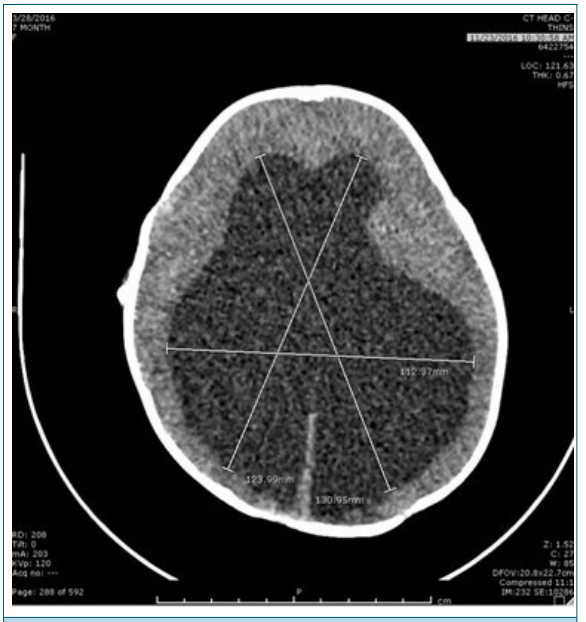

Figure 3. Preoperative Computed Tomography (CT) Scan Demonstrating Marked Dilation of the Lateral Ventricles Secondary to VP Shunt Obstruction.

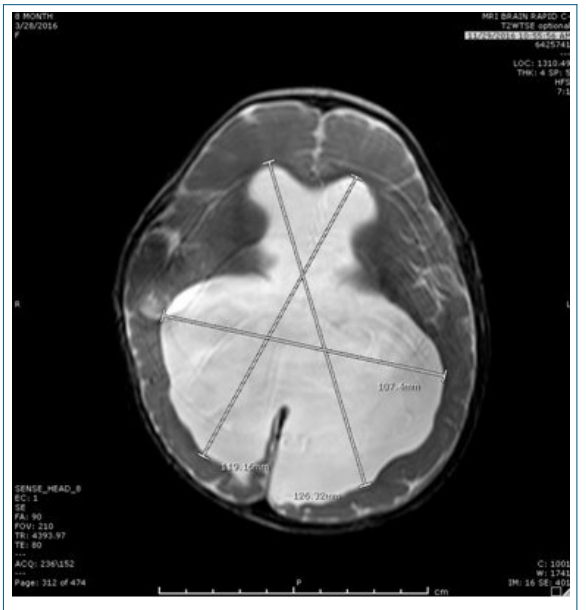

Figure 4. Magnetic Resonance Imaging (MRI) on Postoperative Day 6, the Day of Discharge, Showing Interval Decrease in Ventricle Size.

Discussion

The ventricles of the brain are a communicating network of cavities filled with CSF and are located within the brain parenchyma. The ventricular system is composed of 2 lateral ventricles connected to a single 3rd ventricle via the bilateral foramen of Monroe, the cerebral aqueduct, and the 4th ventricle. CSF is diverted to the spinal column via the foramina of Luschka and Magendie. The choroid plexus, located in the ventricles, produces CSF, which fills the ventricles and subarachnoid space. This process is characterized by constant production and reabsorption (6). VP shunt insertion is often challenging with considerable long-term complication rates. Within the pediatric population, failure rates have been reported to be as high as 38% at 1 year and 48% at 2 years (7). Shunt malfunction most often results from infection or blockage of the shunt that causes it to divert CSF intermittently or not at all. When a blockage occurs, CSF accumulates and can result in symptoms of untreated hydrocephalus. Additional shunt complications can include overdrainage (producing slit-like ventricles or subdural hematoma), underdrainage, multiloculated hydrocephalus, seizures, and abdominal complications including pseudocysts, bowel perforation and hernias (8).

Take Home Messages
• Hydrocephalus is commonly encountered in the pediatric population and is most frequently caused by impaired circulation of CSF.
• Untreated, hydrocephalus may lead to symptoms of elevated ICP; this includes headache, vomiting, lethargy, failure to thrive, bradycardia, and apnea.
• VP shunt insertion is the most common treatment for hydrocephalus despite a high complication rate.
• A thorough preoperative history and physical is essential in a patient with the history of VP shunt placement.

CSF, cerebrospinal fluid; ICP, intracranial pressure; VP, ventriculoperitoneal.

During shunt placement, a catheter is positioned within the foramen of Monroe of the desired ventricle, termed the proximal site, allowing CSF to be diverted through a catheter to the preferred distal site, usually the peritoneal space in a ventriculoperitoneal (VP) shunt. In rare cases, the peritoneal space is inadequate, necessitating the use of an alternative distal site such as the cardiac atrium (ventriculoatrial shunt) or lung pleura (ventriculopleural shunt). Common reasons the peritoneal space is deemed unacceptable include previous intra-abdominal infection and scarring due to prior procedures or intra-abdominal pathology. A retrospective analysis by McGovern et al. found that for normal pressure hydrocephalus, ventriculoatrial shunting appeared to be as safe as ventriculoperitoneal shunting. Although the pleural space may represent an adequate alternative, in children younger than four years of age or in patients with poor pulmonary reserve, the pleura may not provide the necessary CSF absorptive capacity (9).

Two-thirds of catheter obstructions occur at the ventricular end; this is subject to blockage from choroid plexus, brain parenchyma, and infection, while the peritoneal end may become obstructed by omentum and infection (10). At a microscopic level, obstructions occur when holes in the shunt are occluded by granulation tissue, neovascularization, and invasion by the ependymal lining. Another source of obstruction, as demonstrated by this case, is valve failure. It is important to maintain a high index of suspicion for obstruction when caring for a patient with a history of VP shunt. The anesthesiologist was able to play a pivotal role in this patient's care by recognition of a perilous clinical situation through preoperative assessment, and the implementation of a swift surgical intervention in coordination with neurosurgery.

Symptoms of shunt malfunction can develop very rapidly, potentially leading to coma and even death. In infants and toddlers, symptoms include enlargement of the head, a bulging fontanelle, prominent scalp veins, vomiting, lethargy, irritability, downward deviation of eyes, reduced feeding, fever, and swelling or redness along the shunt tract (2,3). Shunt obstruction is confirmed either with brain imaging or fluoroscopically guided injection of iodinated contrast material into the shunt reservoir. The course of the shunt can be carefully examined with plain radiography for breaks, kinks, disconnections, or migration of the shunt tubing. Plain radiography may reveal suture diastasis and increase intracranial ventricle size during episodes of hydrocephalus. Findings related to an acute increase in intracranial pressure caused by shunt malfunction include effacement of cerebral cortical sulci and basal cisterns and transependymal CSF absorption. Chronic hydrocephalus may lead to calvarial thickening, an enlarged and flattened sella turcica, porencephaly, or intracranial cyst formation (8). Ventriculitis and meningitis can be visualized by CT and MRI as enhancement of the ventricular ependymal lining. Imaging analysis is an important adjunct to a strong clinical evaluation of patients with suspected VP shunt malfunction. Additional modalities used for investigation of a potential shunt malfunction include shunt tapping and manometry, valve setting adjustment, headache diaries and placement of an ICP monitoring device.

During the preoperative interview, several questions should be addressed, including when and where the shunt was inserted, the type of shunt, the indication for the shunt, and date of last revision. Common symptoms to inquire about include headache, vomiting, malaise, and irritability (3). On physical exam, bradycardia

and lethargy may be noted. The presence of papilledema is one of the most significant and specific signs (3). However, atypical presentations do occur; thus, if a shunted patient appears unwell, the possibility of an obstructed or malfunctioning shunt must be entertained. Time course is also highly variable, with some patients decompensating rapidly and others experiencing a more insidious onset (3). Regardless, all instances of VP shunt obstruction must be emergently addressed, as delay in care can contribute significantly to morbidity and mortality (2). This case underscores the importance of rapid identification of VP shunt malfunction through a thorough preoperative assessment.

The authors declare no conflicts of interest.

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