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STATEMENT OF FINANCIAL DISCLOSURE

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AHC Media

Cerebrospinal Fluid Shunt Emergencies

Cerebrospinal fluid (CSF) shunt failures account for more than 15,000 pediatric hospital admissions per year,[1] and have an estimated mortality rate of 1-2%.[2] The prompt recognition and treatment of shunt failure in the emergency department (ED) is of the utmost importance to limit morbidity and mortality.

— Ann M. Dietrich, MD, Editor

CSF shunts are mechanical conduits that drain CSF from the ventricular system to a distal site of absorption. They are used as a way to decrease elevated intracranial pressure (ICP) as a result of poor absorption and/or overproduction of CSF within the ventricular system.[3,4]

Hydrocephalus is one of the most common present-day reasons for pediatric shunt placement.[3,5,6] Hydrocephalus refers to enlargement of the ventricles and can be a result of infection, hemorrhage, neoplasm, spinal dysraphism, trauma, or a congenital etiology.[4,6] An estimated 125,000 U.S. children have shunt-dependent hydrocephalus,[1] with an estimated prevalence of 1-1.5%.[4,5]

Shunt failure is common; 40% of shunts fail within the first year[2,3,7-10] and 56-80% by 10 years.[2,11,12] Between 8-10% of shunts eventually become infected.[3,7,8,13] Shunt malfunction can lead to an acute increase in ICP, which may be a life-threatening emergency, making early detection and treatment critical to reducing morbidity and mortality.

CSF Shunts

Cerebrospinal fluid shunts drain CSF from the ventricular system to a site of absorption. Shunts are made up of four components: the proximal catheter, the reservoir, the valve, and the distal catheter. The proximal catheter originates in the lateral ventricle. The catheter exits the intra-cranium through a burr hole and connects to a reservoir in the overlying subcutaneous tissue. The reservoir allows access for CSF sampling and pressure monitoring. From the reservoir, a one-way valve controls flow into the distal catheter. There are five broad categories of valves: differential pressure, flow regulated, anti-siphon controlled, programmable, and gravitational. Each category represents the continued effort to improve the effects of over-drainage associated with hydrostatic pressure changes and reduce the need for surgical intervention when altering valve settings (*see Table 1*). A distal catheter is attached to this valve, which is then tunneled subcutaneously into another body cavity where CSF can easily be reabsorbed.[11,14,15] The peritoneum is the most common site for distal catheter termination — when in such a location, the shunt is called a ventriculoperitoneal (VP) shunt. When

EXECUTIVE SUMMARY

- A high index of suspicion is needed to diagnose shunt malfunction; patients may present with non-specific signs and symptoms.
- Shunt failure is most common the first couple months after shunt placement; previous shunt revision and infection are known predictors of shunt failure.
- To assess for shunt function current image studies must be compared with prior exams.
- Quick-brain MRI is preferred over CT.
- Delayed recognition can lead to significant morbidity and mortality.

the peritoneum is a non-viable option, ventriculoatrial, lumbarperitoneal, ventriculopleural, and ventriculogallbladder are alternative possibilities.[2,8,16] Numerous types and brands of CSF shunts exist, each with their own advantages and disadvantages.[14,17] Knowing the type of shunt a patient has is critical to patient management.

Signs and Symptoms of Shunt Failure

The signs and symptoms of shunt failure are vast and non-specific. Often, they mimic common childhood illnesses such as gastroenteritis, otitis media, appendicitis, migraines, and viral illnesses.[3,4,18] Having a high index of suspicion for shunt failure is critical to diagnosis.

Clinical presentation varies according to age, duration of malfunction, and etiology of failure (infectious vs non-infectious).[7,14] (*See Table 2.*) The most predictive factors of failure are bulging fontanel, fluid collection along the shunt, depressed level of consciousness, irritability, abdominal pain, nausea and vomiting, abnormal shunt pump test, accelerated head growth, and headache. Predictors more specific for an infectious etiology include: purulent drainage, skin erosion, meningismus, erythema, peritonitis, abdominal pain, CSF leakage, and irritability.[3] Notably, although fever is strongly associated with shunt infection, it is not a requirement to make the diagnosis.[3,14] Time since initial surgery and previous history of infection are other important factors to consider.[19] To date, it is unclear whether seizures have any predictability in regards to shunt failure.[2,18]

When there is concern for repeat failure, knowing how a patient previously presented with shunt failure can be helpful.[14] Younger age at time of insertion and short time interval since prior surgical revision are important predictors of repeated shunt failure.[8]

It is important to note that while the presence of certain signs and symptoms increases the likelihood of shunt failure, the same cannot be said for their absence. Thus, an astute physician must remain wary, as the lack

Table 1. Shunt Valve Classifications[7,14,17,41-43]

Type of Valve Can have a combination of the valves below	Description	Brand Examples
Differential Pressure	Allows flow through the valve when the pressure gradient between the proximal and distal catheter reaches a set point. Valves are posture independent.	- Medtronic Fixed Differential Pressure - Aesculap Fixed Differential Pressure - Codman Fixed Differential Pressure
Flow Regulated	Maintains constant CSF flow through the valve.	- Orbis-Sigma
Anti-Siphon	Reduces the siphoning, or over-drainage effect, that occurs in the upright position due to hydrostatic pressure differences.	- Delta (Medtronic PS Medical)
Programmable	Allows valve pressure settings to be altered as an outpatient without the need for surgical intervention.	- Strata (Medtronic PS Medical) - Codman-Hakim Programmable Shunt (Codman) - Certas Valve (Codman)
Gravitational	Posture dependent valves that change resistance based on a patient's posture in efforts to overcome siphoning.	- ProGAV (Aesculap) - Paedi-Gav (Aesculap)

For more information on types of shunts, please visit the ISPN (international Society for Pediatric Neurosurgery) webpage at http://www.ispneurosurgery.org/ and view the shunt guide section.

Table 2. Signs and Symptoms of Shunt Failure

Infectious and Non-infectious

- Abdominal mass[8]
- Abdominal pain[3]
- Accelerated head growth[3]
- Apnea[40]
- Ascites[8]
- Ataxia[14]
- Bradycardia[40]
- Bulging fontanel[3]
- Decreased level of consciousness (mental status changes)[3,18]
- Dilated/sluggish pupils[40]
- Fever[3]
- Fluid collection along the shunt[3]
- Focal deficits[11]
- Headache[3]
- Irritability[3]
- Lethargy[2]
- Nausea/vomiting[3,18]
- Papilledema[11]
- Purulent drainage[3]
- Seizures[2,18]
- Shunt site swelling[2]
- Shunt site erythema[18]
- Signs and symptoms of increased intracranial pressure[8]
- Skin erosion[3]
- Splaying of cranial sutures[8]

of any signs and symptoms does little to rule out the possibility of shunt failure.[3,18]

Mechanisms of Shunt Failure — Non-Infectious Causes of Shunt Failure

Mechanical Failure

Fracture: Catheter fracture occurs due to the biomechanical forces of patient growth and catheter degradation from host reactions.[8] Scar tissue surrounding the catheter provides a temporary conduit between catheter fragments, often causing a delay in symptomatology and detection.[11] The neck is the most common region for fracture development.[14,20]

Disconnection: Catheter disconnection occurs due to surgical error or material defect. Disconnection is seen shortly after shunt surgery and typically located at the valve level.[8,11,20]

Migration: Migration occurs when the catheter (proximal or distal) moves from its original position to a location that inhibits proper drainage. This can occur due to improper length (both too short and too long), fracture, disconnection, tethering, or perforation.[8,11,21] Evidence of catheter migration and subsequent erosion/perforation into viscera can be found in almost any body cavity, including the ventricular, intrathoracic, abdominal, and pelvic cavities.[11]

Misplacement: Misplacement occurs when the catheter tip (either proximal or distal) is inappropriately positioned during surgery, making for poor CSF flow. Misplaced shunts are typically noted shortly after surgery, but may also have a delayed presentation.[8]

Obstruction

Obstruction is the most common cause of shunt malfunction, making up 56-83% of shunt failures.[2,9,15,21] The two most common places for obstruction to occur are at the proximal catheter tip and at the shunt valve. The proximal catheter tip can be obstructed due to ingrowth of the choroid plexus; the shunt valve can be blocked by blood and debris.[8,11,14] Distal catheter obstruction is less common and occurs due to adhesions, scarring, migration, tube kinking (most commonly at the level of the valve connection or abdominal entry site), catheter twisting, or distal catheter obstruction (internal obstruction from inflammatory debris and tissue build-up or external obstruction from pseuduocyst formation).[8,14,21]

Over-drainage

Over-drainage occurs when a functioning shunt drains more CSF than appropriate. Acute over-drainage results in extra-axial fluid collections or subdural hematomas. Chronic over-drainage results in slit ventricle syndrome (SVS), the precise definition of which remains to be determined. Generally speaking, SVS refers to symptomatic small ventricles — small ventricles in association with intermittent symptoms suggestive of shunt malfunction (increased intracranial hypertension), typically associated with postural changes. Although small ventricles occur in 50% of shunted children, SVS only has a reported incidence of < 2%. Nonetheless, SVS accounts for a disproportionate number of shunt revisions in pediatric neurosurgery. The etiology of SVS is theorized to be due to chronic over-drainage during the period of rapid brain growth, leading to poor brain parenchymal compliance and obstruction of the catheter by the collapsed ventricular system.[11,14,22]

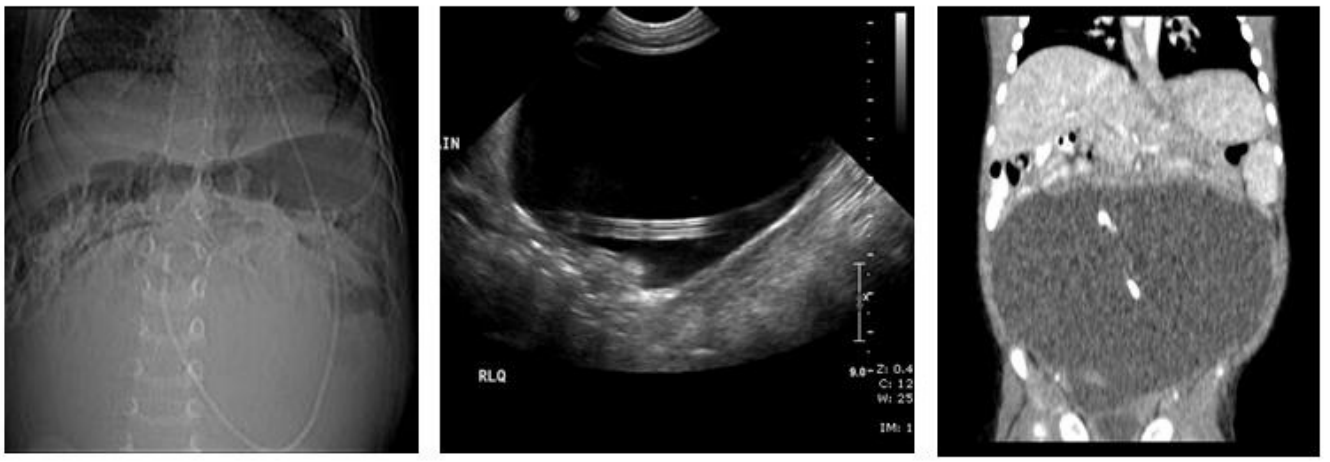
Ventricular Loculations

Ventricular loculations are non-communicating pockets of CSF within the ventricular system. They form gradually and are either congenitally acquired or develop after an episode of ventriculitis or hemorrhage. Shunt malfunction occurs due to a shunt's inability to access and drain all the CSF so that a loculated compartment, separated from the shunt, continues to increase in size and creates symptoms of increased intracranial pressure.[11,22] A special type of loculation, known as isolated or "trapped" fourth ventricle, occurs when the fourth ventricle is unable to communicate with both the third ventricle above and basal cisterns below. In the face of a functioning shunt, CSF is diverted away from the Sylvian aqueduct promoting its closure. If there is poor CSF absorption or fourth ventricular obstruction, CSF will accumulate in the fourth ventricle producing signs and symptoms of increased ICP. This phenomenon typically occurs in patients with a history of shunt revisions and post-infectious hydrocephalus.[4,22]

Abdominal Complications

Ascites: A rare complication that causes shunt malfunction in patients with comorbid heart, liver, or kidney disease. The ascites fluid increases pressure within the peritoneal cavity, which changes the CSF intracranial-peritoneal pressure differential and eventually leads to shunt malfunction.[22]

Figure 1. Example of a CSFoma (Pseudocyst) with KUB (left), Abodominal Ultrasound (middle), and Coronal CT Image (right) of the Same Child



Images provided by Achint K. Singh, MD, University of Texas Health Sciences Center San Antonio

Constipation: Severe constipation can increase intra-abdominal pressure and thereby alter the CSF intracranial-peritoneal pressure differential, eventually leading to shunt malfunction.[5]

Inguinal Hernia: Increased intra-abdominal pressure following VP shunt placement increases the likelihood of subsequent inguinal hernia development, especially in neonates and males.[21,23]

Perforation: May occur acutely (intra-operatively) or chronically via erosion of the shunt through an abdominal or pelvic viscus.[11,22] Chronic perforations typically present indolently with tubing emerging from the urethra or anus.[22] Perforations into the bladder, vagina, diaphragm, intestine, gallbladder, and bronchial tree have all been documented.[21]

Pseudocyst: Pseudocyts are loculated intra-abdominal fluid collections centered around the terminal portion of the peritoneal catheter (*see Figure 1*).[11,22] They are caused by adhesions due to infection, prior abdominal surgery, reactions to CSF proteins and shunt material, or migration of the omentum over the catheter tip.[11,21] Pseudocysts present as abdominal masses (with or without pain), signs of abdominal obstruction, and/or neurological symptoms,[22] and occurrence is 0.7-10%.[21]

Mechanisms of Shunt Failure — Infectious Causes of Shunt Failure

Approximately 8-10% of shunts become infected.[8] Depending on the microbial source, shunt infection and subsequent failure can present both acutely or indolently. A high index of suspicion is needed in all patients presenting to the ED with nonspecific signs and symptoms, with or without fever, as the consequences of a missed/delayed diagnosis are severe.[13,14,21,24] Most shunt infections occur within the first 4-6 months after placement and are caused by skin-colonizing organisms present at the time of surgery.[13,20,25] *Staphylococcus* species account for up to 90% of infections,[25] with coagulase negative staphylococci (CONS) alone accounting for 75%, due to its unique ability to create a biofilm (biofilms allow CONS to escape both antimicrobials and the host immune system).[13] When present, Gram-negative organisms represent ICU colonization, neonatal pathogens, or abdominal pathology. Should cultures contain Enterobacteriaceae or anaerobes or be polymicrobial, bowel perforation by the peritoneal catheter should be a concern.[13,21] Other infectious organisms that can be seen include *Proprionibacterium* species, *Corynebacterium*, *Haemophilius influenza*, and fungi.[25]

Known risk factors for infection include time since placement, age at insertion, history of prior infection, and history of revision (*see Table 3*).

Mechanisms of Shunt Failure — Special Shunts

When placement of the distal shunt catheter into the peritoneum is contraindicated, alternative locations are used. Notably, these locations are less common and associated with a higher rate of failure.[11]

Ventriculoatrial Shunts

Due to their risk profile, ventriculatrial (VA) shunts are not as common as they once were. The atrium is accessed via the facial, subclavian, or internal jugular vein, making length critical to proper VA shunt functioning. Catheters that are too long allow for catheter migration into the right atrium, through a patent foramen ovale, into the pericardial space, or through the septum; if too short, migration with growth up the SVC may produce poor drainage.[11] Other complications include bacterial endocartidits, pulmonary embolism, pulmonary hypertension, cor pulmonale, cardiac arrhythmias, cardiac tamponade, venous thrombosis, tricuspid valve pathology, infection, and shunt nephritis.[2,8,11] Shunt nephritis is the result of activation of the complement

Table 3. Known Risk Factors for Shunt Infection

- < 4-6 months since CSF shunt placement[13,19,20,25]
- Shunt placement under 1 year of age[13,19]
- Previous shunt revisions[13,19,44]
- Previous shunt infections[19,44]
- Hydrocephalus due to intraventricular hemorrhage[13]
- Complex initial shunt[13]
- Ventriculoatrial shunt[13]

Table 4. Pumping the Shunt

Single Reservoir

- Apply pressure to reservoir
- Resistance to compression is concerning for distal malfunction
- No refill is concerning for proximal malfunction

Double Reservoir

- Apply and maintain pressure to proximal reservoir
- Press distal reservoir. If resistance to compression, concern for distal malfunction
- Release proximal reservoir. Delayed refill is concerning for proximal malfunction

Adapted from Greenberg[4] and Magita[10]

cascade due to chronic infection and subsequent glomerular immune complex deposition. Associated symptoms include hypertension, nephrotic syndrome, hematuria, fever, anemia, hepatosplenomegaly, and non-thrombocytopenic purpura. Unlike VP shunts, due to the bloodstream location of VA catheters, when shunt infection is present, blood cultures are often positive. Likewise, if bacteremia from a non-catheter-related etiology occurs, colonization of the shunt subsequently ensues. Most VA complications are late occurrences.[8,11]

Ventriculopleural Shunts

Ventriculopleural shunts are used as a temporary measure, as the risks include hydrothorax, pneumothorax, fibrothorax, pleural emphyema, pleural effusion, chest pain, pneumonia, respiratory failure, and bronchial perforation.[2,11,14]

Lumboperitoneal Shunts

Lumboperitoneal shunts are associated with tonsillar herniation and arachnoiditis.[26]

Diagnostic Testing

Currently, there is no consensus guideline to aid physicians in the diagnostic workup of shunt failure. All imaging modalities have their unique advantages and disadvantages. Clinical judgment must be used in determining what type(s) of imaging modality to use.[27]

Shunt Pump Test

Pumping the shunt is a compression maneuver clinicians can perform to test for both proximal and distal shunt obstruction. How to perform the test depends on whether the shunt has a single or double reservoir (*see Table 4*). In general, resistance to reservoir compression is concerning for distal obstruction, whereas delayed refill with decompression is concerning for proximal obstruction.

Although previously thought to be helpful in the diagnosis of shunt malfunction, its utility is not as valuable as once believed.[14] The reported sensitivity and specificity for detecting obstruction is 11-20% and 63-99%, respectively,[10] with the specificity declining with increasing age.[3] The positive predictive value ranges from 12% to 86% and the negative predictive value from 65% to 93%.[8,10] Thus, a negative shunt pump test cannot rule out shunt obstruction,[3,18] nor does a positive test in an asymptomatic individual necessitate a workup.[28] Newer studies have even suggested that the test may actually cause obstruction by drawing the choroid plexus into the shunt.[14] Thus, if the test is to be performed, it may only be used as an adjunct to clinical decision making, not to definitively rule in or out shunt obstruction.[10]

Shunt Series

Shunt series refers to the collection of radiographs that view the entire course of a shunt. It includes frontal and lateral views of the head and neck and frontal views of the chest and abdomen. It is used to evaluate shunt continuity (disconnections, fractures, catheter migrations) rather than shunt patency.[11,14,21] Some valves and connectors are radiolucent while others are radiopaque. Normally, radiolucent material can be mistaken for shunt disconnection/fracture. Knowing what type of shunt the patient has can help distinguish this.[11,14] Comparison to prior imaging is helpful.

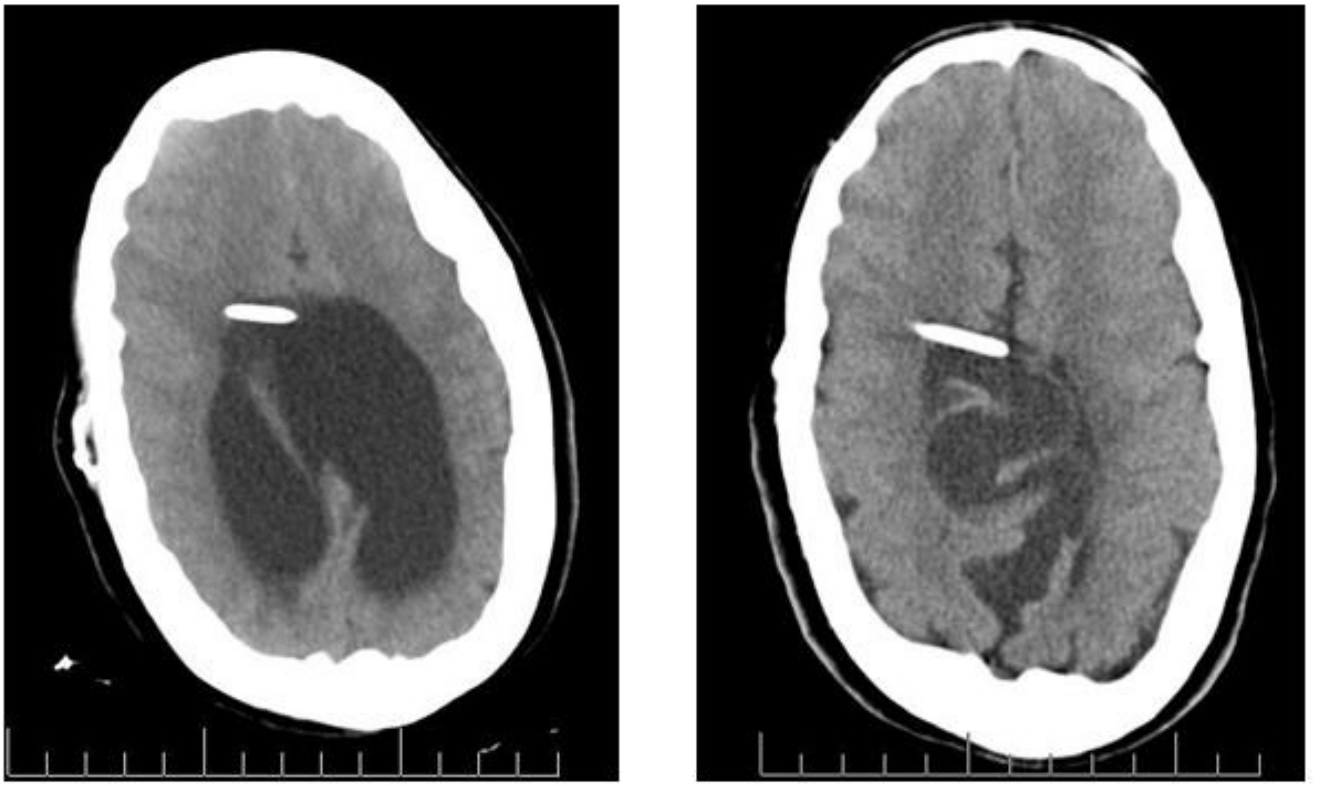
Controversy surrounds the use of the shunt series in cases of possible dysfunction. Some studies suggest obtaining it in all cases of suspected shunt malfunction to identify the presence of shunt discontinuity; MRI or CT will only show ventricular size and morphology changes.[29] Other studies suggest its use only when there is concern for a mechanical cause of shunt failure due to the low sensitivity of the test.[20,27,30] Clinical judgment is advised as well as developing a protocol with your local neurosurgeon to determine in which instances a shunt series should be performed.

Quick-Brain MRI/CT

Quick-brain MRI and CT are imaging modalities used to provide information on ventricular morphology, shunt location, and shunt integrity to aid in detection of shunt failure. Shunt failure can present with enlarged,

Figure 2. 16-year-old Female with VP Shunt Malfunction who Presented with Headache and Vomiting

CT image on left was done in the ED showing enlargement of both lateral ventricles as compared to her previous CT performed when asymptomatic (right).



Images provided by Daniel J. Dire, MD, University of Texas Health Sciences Center San Antonio

normal, or small-sized ventricles.[14,21,31] Although ventricular enlargement is indicative of shunt failure,[8,14,31] shunt malfunction occurs in 16-24% of patients with no ventricular changes on imaging.[2] Comparison with prior imaging, if available, is imperative (*see Figure 2*).[8,14,31] Comparison of ventricular morphology both from baseline imaging and/or prior imaging from past obstruction can guide a clinician's judgment; changes are often time consistent and predictable, but not always.[31] Secondary signs of obstruction include blurring of ventricular margins due to transependymal flow of CSF, peri-shunt edema, and subgaleal fluid collections.[11,14] Infection can occasionally be detected by visualization of debris within the ventricles and/or leptomeningeal/ependymal enhancement. Care must be taken when interpreting pachymeningeal enhancement as this can be seen for months postoperatively.[11,24] As an imaging modality, CT has good specificity but poor sensitivity (54-83%)[1] and negative predictive value for predicting shunt obstruction;[27] therefore, clinical findings in correlation with imaging are needed to make decisions regarding shunt management.

Quick-brain MRI (also known as rapid-brain MRI, rapid-sequence MRI, fast-sequence MRI, single-shot fast-spin echo) is quickly replacing CT as the diagnostic imaging modality of choice in the workup of shunt malfunction.[1,11,14,32-34] Quick-brain MRI works by taking 1-minute images of the brain in three separate planes, thus reducing the need for sedation and eliminating radiation exposure without altering the test characteristics for detecting shunt failure.[1] Decreased radiation exposure is a significant benefit for patients with CSF shunts who receive an average of 2.6 head scans per year.[12,32,34] Quick-brain is as sensitive as CT in detecting shunt malfunction with comparable sedation needs[1,14,33] and minimal increase in time to complete the scan.[1]

Quick brain MRI is not without its limitations. Contraindications include pacemakers, defibrillators, and cochlear implants.[1] Blood, air, and implanted devices can be difficult to visualize with quick-brain MRI and make CT a reasonable alternative when specifically trying to visualize a hemorrhage, pneumocephalus, or when contrast material is needed.[32,33] Care must be taken with programmable shunt valves, as these may be adjusted by the MRI magnet. Prior to MRI imaging, the radiologist should confirm that the valve is resistant to reprograming at the magnetic field to be used. Post scan, it is good practice to check the valve to ensure that the settings have not changed. More

Table 5. Steps to Performing a CSF Shunt Procedure[4,10,15,40]

- Place the patient in recumbent position.
- Identify the reservoir.
- Prep the area for a sterile procedure (shave, clean with isopropyl alcohol, prep with Betadine, drape area, use sterile gloves and sterile technique). Local anesthesia is usually not necessary, but topical anesthetic creams may be useful prior to the procedure in young children.
- Enter the reservoir perpendicularly with a 25 gauge needle (preferably non-coring needle) until a popped is felt.
- Do not advance too far so as to puncture the back wall of the reservoir.
- Adjust needle until flow returns.
- Spontaneous flow indicates some proximal catheter function. If no spontaneous flow, may gently aspirate CSF with syringe. The inability to aspirate CSF may indicate proximal occlusion. The ability to easily aspirate CSF may indicate ventricular pressure is near zero.
- Measure opening pressure with a manometer (e.g., from a lumbar puncture kit) and making sure to level the manometer at the level of the ear (which is the external marker for the level of the cerebral ventricles).
- Following opening pressure, if a distal reservoir is present, occlude and re-measure pressure. A rise in pressure indicates some function of the valve and distal shunt.
- Collect fluid for studies (culture and sensitivity, glucose, protein, cell count).
- If a shunt is being tapped secondary to cerebral herniation, remove fluid until pressure is < 20 cm H_2O.

recent valves have been made that are resistant to change at the 3T level.[11]

Shuntogram

Shuntogram is a radionucleotide shunt study used to evaluate shunt patency and velocity. Shunt obstruction is seen when the radionucleotide injected into the shunt reservoir fails to flow throughout the length of the catheter.[11,14] This test is especially useful when there is concern for obstruction but no change in ventricular size.[14] Notably, CT combined with shuntogram increases the sensitivity compared to CT alone.[27]

Ultrasound

Ultrasound is a new and emerging imaging modality being used as an adjunct in the evaluation of shunt malfunction. It is cheap, fast, easily accessible, and has the added benefit of no radiation exposure.[35] Reports have been made about its use in detection of abdominal shunt-related pathology (pseudocyst, abscess),[14] shunt discontinuity,[36,37] and aiding the physician in accessing the shunt reservoir.[36,37] Some studies have looked at shunt flow patterns and cerebral blood flow with Doppler ultrasound in attempts to extrapolate information regarding shunt function.[36-39] Other studies have looked at using ultrasound to measure optic nerve sheath diameter as a predictor of increased ICP; however, these studies revealed that this test is neither sensitive nor specific enough to be used as a screening modality in the evaluation of shunt malfunction.[35] Work continues to be done in the field of ultrasonography in hopes to further its use as an imaging modality in the workup of shunt malfunction.

Shunt Management

When there is concern for CSF shunt malfunction, always consult a neurosurgeon to assist in shunt evaluation. A neurosurgeon can tap the shunt for both therapeutic and diagnostic purposes, remove/repair the shunt, place a temporary extraventricular draining device, or perform a third ventriculostomy.[4,40]

Non-infectious

Any child that presents with a potential shunt dysfunction should be placed on a cardiac monitor and have frequent neuro checks performed. If the shunt is found to be dysfunctional, the child should be managed, preferably, at the site where the shunt was placed. An emergent transfer should be arranged in consultation with the accepting neurosurgeon. If a patient comes into the ED with concern for cerebral herniation and impending death from increased ICP (fixed and dilated pupils, presence of Cushing's triad, unresponsiveness) and transfer to a higher level of care or a neurosurgeon is not immediately available, the ED physician may need to tap the shunt to emergently decrease the ICP.

Prior to performing a procedure, the physician must know what kind of shunt the patient has and where to tap for each individual shunt type. The physician needs to be aware that there are significant risks associated with accessing the shunt, including infection, CSF leak, damage to the shunt, and local hematoma.[40] The steps to perform a procedure are listed in Table 5 (*see Figure 3*).

Infectious

There are no universal guidelines for management of CSF shunt infections.[13] Only a neurosurgeon should tap the shunt reservoir and obtain CSF cultures.[11,13] This may be delayed until patient is transferred to a higher level of care or in conjunction with a neurosurgeon.

To increase detection of shunt pathogens, aerobic, anaerobic, and fungal cultures should be obtained and extended incubation times should be requested. Prolonging incubation time will lead to higher detection rates of fastidious organisms and microorganisms in patients pre-treated with antibiotics. New and emerging is the use of polymerase chain reaction as a diagnostic tool in detecting shunt pathogens. Current studies show that many negative cultures have tested positive with polymerase chain reaction.[13]

Empiric coverage should be directed toward common shunt pathogens and have the ability to penetrate the central nervous system.[13] For

Figure 3. Example Tapping a Shunt Reservoir with a 25 Gauge Butterfly Needle

To obtain pressures, attach a manometer (not shown) to the distal end of the butterfly needle catheter. Additional photographs of a shunt tap can found at http://emedicine.medscape.com/article/81058-overview#a7.

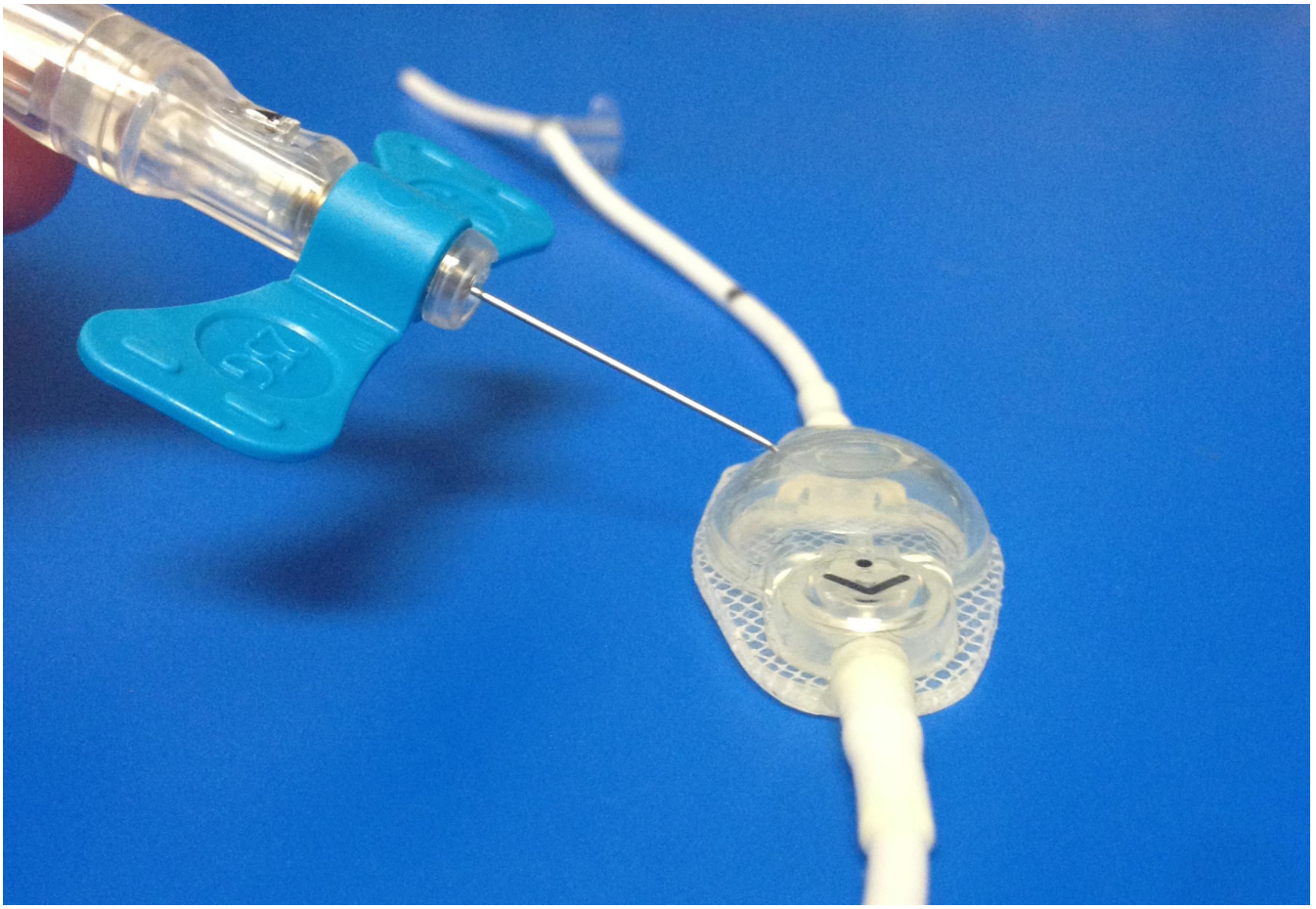


Image provided by Whitney M. Wroe, MD, University of Texas Health Sciences Center San Antonio

Gram-positive coverage, vancomycin is typically used due to the emergence of methicillin-resistant *Staphylococcus*. Cefepime, ceftazidime, or meropenem are appropriate for Gram-negative coverage as they include coverage of *Pseudomonas aeruginosa*.[13] When infection is present, a neurosurgeon will typically do one of the following:

- Remove the shunt, place a temporary external ventricular drain, and give IV antibiotics.[13,24]
- Replace the shunt and give IV antibiotics.[13,24]
- Serial ventricular taps and give IV antibiotics without surgical revision.[13] Notably, there are lower success rates when hardware is kept in place.[24]

The duration of antibiotics varies, and the use of intraventricular antibiotics is controversial.[13]

Summary

Shunt failure is common. Symptoms are often non-specific and require a high index of suspicion for diagnosis. There is no guideline or consensus on diagnostic imaging, as each imaging modality has its advantages and disadvantages. Diagnosis of shunt malfunction ultimately is a clinical one, relying on the combination of symptoms, physical exam, and imaging results. Missed diagnosis can result in significant morbidity and mortality.

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CME Questions

1. When imaging the brain in the evaluation of CSF shunt malfunction, why is quick-brain MRI preferred over CT?
   a. There is easier access to MRI.
   b. There is better visualization of brain with MRI.
   c. MRI is a faster imaging technique.
   d. There is no radiation exposure with MRI.
2. A patient with a VP shunt presents to the ED with a headache. Which of the following is most indicative of shunt malfunction?
   a. Imaging that shows enlarged ventricles in the ED and 4 months ago when outpatient imaging was obtained for annual screening
   b. Imaging shows small ventricles in the ED and 4 months ago when outpatient imaging was obtained for annual screening
   c. Imaging shows enlarged ventricles in the ED and normal sized ventricles 4 months ago when outpatient imaging was obtained for annual screening
   d. Imaging shows normal sized ventricles today and enlarged ventricles during a prior episode of obstruction 4 months ago
3. Slit ventricle syndrome could be present in which of the following cases?
   a. Patient with small ventricles on imaging and headache that occurs when changing from supine to standing position
   b. Patient with small ventricles and no symptoms
   c. Patient with normal sized ventricles and persistent headache
   d. Patient with history of small ventricles and enlarged ventricles on current imaging
4. Who is most likely to have a shunt infection?
   a. An 18-year-old with a shunt placed 1 year ago
   b. A 6-month-old with a shunt placed at 5 months of age
   c. A 15-year-old with a shunt placed at 1 month of age
   d. A 5-year-old with shunt placed 4 months prior
5. Which of the following are contraindications to rapid MRI?
   a. Pacemakers, defibrillators, and cochlear implants
   b. CSF shunt and pacemakers
   c. Gastrostomy tubes and cochlear implants
   d. Mechanical heart valves, pacemakers, and defibrillators

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6. Both before and after obtaining a quick-brain MRI, the physician must do which of the following?
   a. Perform the shunt pump test
   b. Obtain a shunt series
   c. Call the shunt manufacturer for approval and then report on outcome of test
   d. Check the shunt valve setting

7. What is the imaging modality of choice to detect shunt continuity and shunt patency respectively?
   a. Shunt series, shuntogram
   b. Quick-brain MRI, shunt series
   c. Ultrasound, shuntogram
   d. Shunt series, CT

8. Where are the two most common sites for shunt obstruction to occur?
   a. Distal catheter tip and valve
   b. Proximal catheter tip and valve
   c. Distal catheter tip and at the level of the neck
   d. At the level of the neck and proximal catheter tip

9. What is the most common cause of shunt malfunction?
   a. Obstruction
   b. Overdrainage
   c. Pseuduocyst
   d. Perforation

10. Most shunt infections are caused by which of the following?
    a. Skin colonizing organisms present at the time of surgery
    b. Fungus
    c. Rare anaerobes
    d. Seeding from bacteremia

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PEDIATRIC EMERGENCY MEDICINE **REPORTS**

Practical, Evidence-Based Reviews in Pediatric Emergency Care

Cerebrospinal Fluid Shunt Emergencies

Shunt Valve Classifications[7,14,17,41-43]

Type of Valve Can have a combination of the valves below	Description	Brand Examples
Differential Pressure	Allows flow through the valve when the pressure gradient between the proximal and distal catheter reaches a set point. Valves are posture independent.	- Medtronic Fixed Differential Pressure - Aesculap Fixed Differential Pressure - Codman Fixed Differential Pressure
Flow Regulated	Maintains constant CSF flow through the valve.	- Orbis-Sigma
Anti-Siphon	Reduces the siphoning, or over-drainage effect, that occurs in the upright position due to hydrostatic pressure differences.	- Delta (Medtronic PS Medical)
Programmable	Allows valve pressure settings to be altered as an outpatient without the need for surgical intervention.	- Strata (Medtronic PS Medical) - Codman-Hakim Programmable Shunt (Codman) - Certas Valve (Codman)
Gravitational	Posture dependent valves that change resistance based on a patient's posture in efforts to overcome siphoning.	- ProGAV (Aesculap) - Paedi-Gav (Aesculap)

For more information on types of shunts, please visit the ISPN (international Society for Pediatric Neurosurgery) webpage at http://www.ispneurosurgery.org/ and view the shunt guide section.

Known Risk Factors for Shunt Infection

- < 4-6 months since CSF shunt placement[13,19,20,25]
- Shunt placement under 1 year of age[13,19]
- Previous shunt revisions[13,19,44]
- Previous shunt infections[19,44]
- Hydrocephalus due to intraventricular hemorrhage[13]
- Complex initial shunt[13]
- Ventriculoatrial shunt[13]

Pumping the Shunt

Single Reservoir
- Apply pressure to reservoir
- Resistance to compression is concerning for distal malfunction
- No refill is concerning for proximal malfunction

Double Reservoir
- Apply and maintain pressure to proximal reservoir
- Press distal reservoir. If resistance to compression, concern for distal malfunction
- Release proximal reservoir. Delayed refill is concerning for proximal malfunction

Adapted from Greenberg[4] and Magita[10]

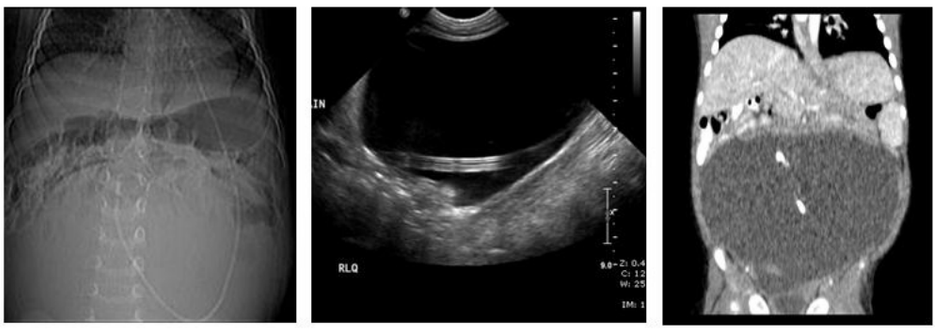
Steps to Performing a CSF Shunt Procedure[4,10,15,40]

- Place the patient in recumbent position.
- Identify the reservoir.
- Prep the area for a sterile procedure (shave, clean with isopropyl alcohol, prep with Betadine, drape area, use sterile gloves and sterile technique). Local anesthesia is usually not necessary, but topical anesthetic creams may be useful prior to the procedure in young children.
- Enter the reservoir perpendicularly with a 25 gauge needle (preferably non-coring needle) until a popped is felt.
- Do not advance too far so as to puncture the back wall of the reservoir.
- Adjust needle until flow returns.
- Spontaneous flow indicates some proximal catheter function. If no spontaneous flow, may gently aspirate CSF with syringe. The inability to aspirate CSF may indicate proximal occlusion. The ability to easily aspirate CSF may indicate ventricular pressure is near zero.
- Measure opening pressure with a manometer (e.g., from a lumbar puncture kit) and making sure to level the manometer at the level of the ear (which is the external marker for the level of the cerebral ventricles).
- Following opening pressure, if a distal reservoir is present, occlude and re-measure pressure. A rise in pressure indicates some function of the valve and distal shunt.
- Collect fluid for studies (culture and sensitivity, glucose, protein, cell count).
- If a shunt is being tapped secondary to cerebral herniation, remove fluid until pressure is < 20 cm H_2O.

Signs and Symptoms of Shunt Failure

Infectious and Non-infectious
- Abdominal mass[8]
- Abdominal pain[3]
- Accelerated head growth[3]
- Apnea[40]
- Ascites[8]
- Ataxia[14]
- Bradycardia[40]
- Bulging fontanel[3]
- Decreased level of consciousness (mental status changes)[3,18]
- Dilated/sluggish pupils[40]
- Fever[3]
- Fluid collection along the shunt[3]
- Focal deficits[11]
- Headache[3]
- Irritability[3]
- Lethargy[2]
- Nausea/vomiting[3,18]
- Papilledema[11]
- Purulent drainage[3]
- Seizures[2,18]
- Shunt site swelling[2]
- Shunt site erythema[18]
- Signs and symptoms of increased intracranial pressure[8]
- Skin erosion[3]
- Splaying of cranial sutures[8]

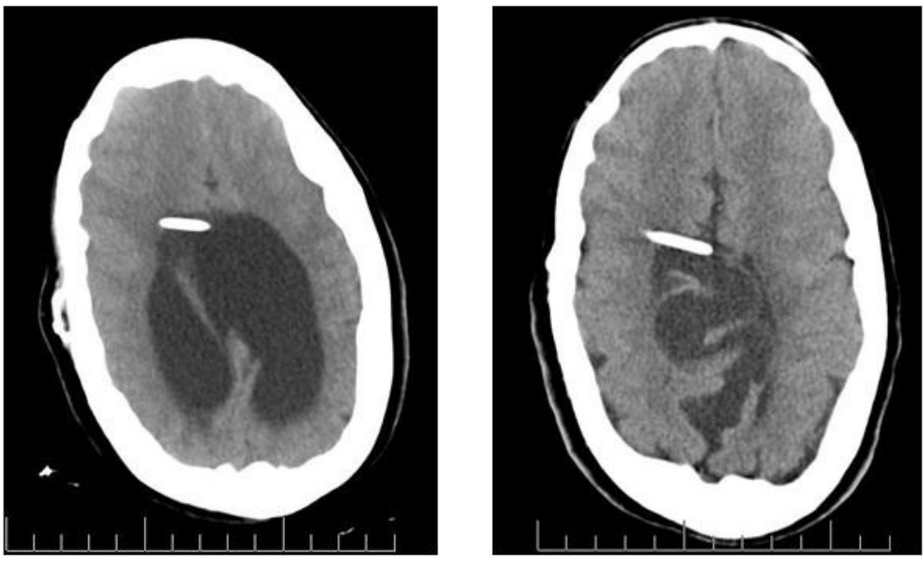
Example of a CSFoma (Pseudocyst) with KUB (left), Abodominal Ultrasound (middle), and Coronal CT Image (right) of the Same Child



Images provided by Achint K. Singh, MD, University of Texas Health Sciences Center San Antonio

16-year-old Female with VP Shunt Malfunction who Presented with Headache and Vomiting

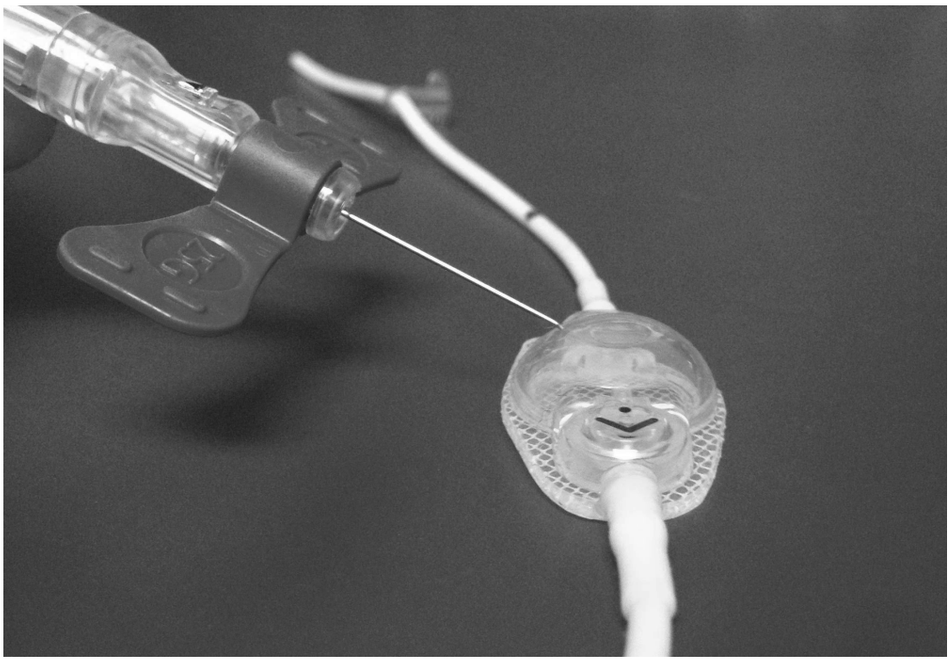
CT image on left was done in the ED showing enlargement of both lateral ventricles as compared to her previous CT performed when asymptomatic (right).



Images provided by Daniel J. Dire, MD, University of Texas Health Sciences Center San Antonio

Example Tapping a Shunt Reservoir with a 25 Gauge Butterfly Needle

To obtain pressures, attach a manometer (not shown) to the distal end of the butterfly needle catheter. Additional photographs of a shunt tap can found at http://emedicine.medscape.com/article/81058-overview#a7.



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