


Shunt Devices for Neurointensivists: Complications and Management

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Abstract Cerebrospinal fluid diversion has become the mainstay treatment in hydrocephalus for over 50 years. As the number of patients with ventricular shunt systems increases, neurointensivists are becoming the first-line physicians for many of these patients. When symptoms of a shunt malfunction are suspected and access to a neurosurgeon is limited or delayed, workup and temporizing measures must be initiated. The article highlights the functional nuances, complications, and management of current programmable shunt valves and their MRI sensitivity.

Keywords Shunt · Neurointensivists · Valves · Complications

Introduction

Impaired reabsorption or excessive cerebrospinal fluid (CSF) production from a variety of underlying etiologies results in hydrocephalus and subsequent intracranial pressure elevations [1, 2]. Eventually, 75,000 patients require shunting device placement in North America annually [3–5]. Tolerability has improved with the introduction of biocompatible products designed for long-term implantation and regulated shunt valves. Nevertheless, malfunction is still a frequent clinical problem. The most common

causes of shunt malfunction are often diagnosed with a thorough history, examination, and radiographical correlates. Thereby, neurointensivists are becoming increasingly involved with shunt malfunction management, in the critical care unit in university centers, the emergency room, and wards in community centers with neurosurgical services. Notably, there is a paucity of ventricular shunt literature directed to neurointensivists [6]. Therefore, we will review shunt device features and illustrate complications and their management from the neurointensivist's perspective.

Shunt Components

Most ventricular shunts involve a ventricular catheter, a valve, and a distal catheter. The catheters are visible on both X-ray and CT scans (Figs. 1, 2). The ventricular catheter is the most proximal part of the shunt, traveling from the ventricle to the valve. Its obstruction owing to choroid plexus material, gliotic tissue ingrowth, elevated protein content in the CSF, or infection represents the most common cause of shunt failure [3, 7]. These patients may develop a progressive high-pressure headache, inattention, visual deficits, and eventually an impaired mental status leading to coma.

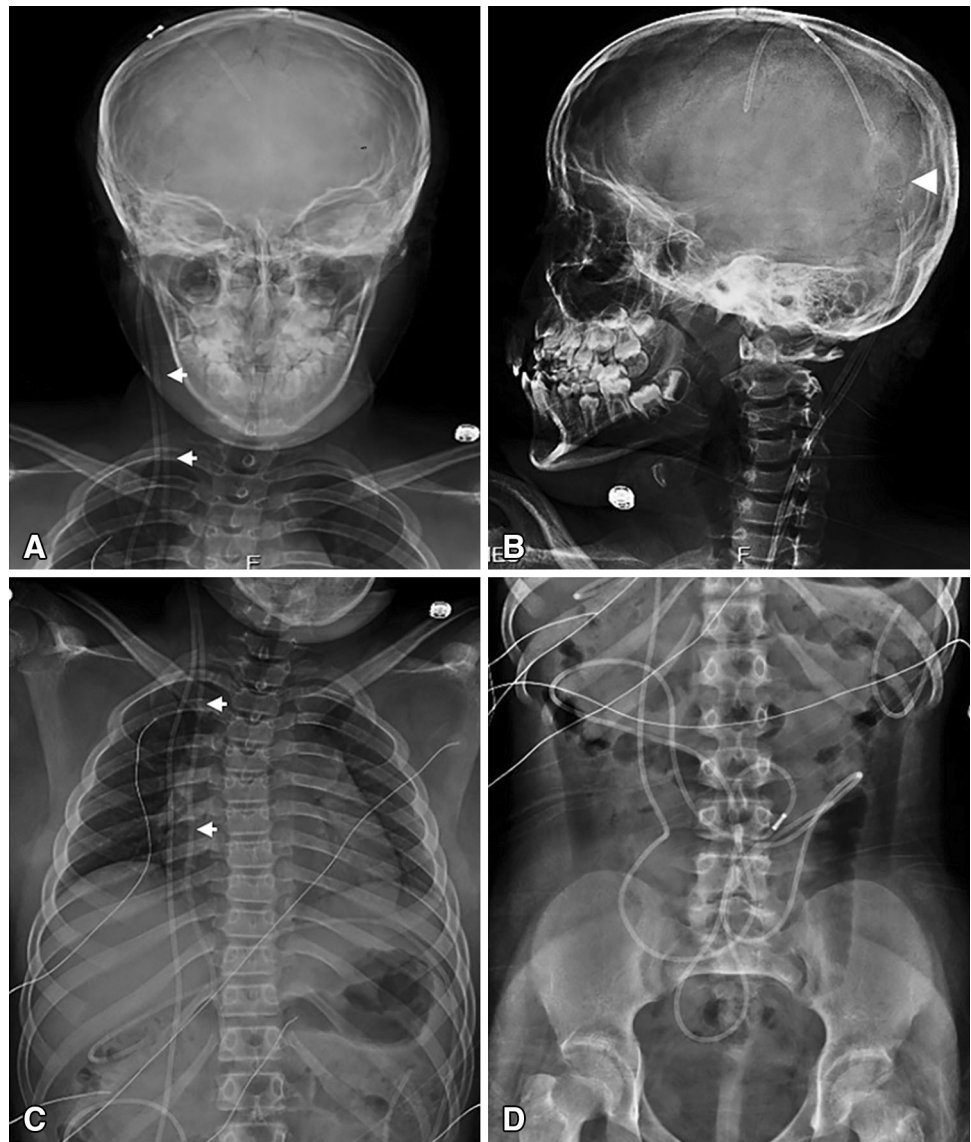
Valves are designed to prevent complications of excessive drainage such as low-pressure headache, intracranial hemorrhage, or subdural hematoma [7–11]. Fixed-pressure and flow-regulated valves allow CSF drainage above a predetermined intracranial pressure threshold or set resistance (Tables 1, 2) [10–12]. Importantly, fixed-pressure and flow-regulated valves do not allow for target range modifications without revision surgery. Conversely, programmable valves allow external fine adjustments in the

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Fig. 1 Plain radiographs of a shunt series. **a, b** AP and lateral cranial X-rays. Skulls films evaluate the continuity of the proximal system and evaluate the valve as referenced with the *bold triangle*. *White arrows* also show retained old distal catheter system in this patient, **c, d** AP X-ray of the chest and abdomen is commonly used to trace the distal tubing into the peritoneum in cases of a ventriculoperitoneal shunt



intracranial pressure thresholds for drainage. A magnetically driven calibration tool placed directly over the skin overlaying the valve allows clinicians to instantly change the threshold for drainage. In the event that a patient presented with low-pressure symptoms such as positional headaches, or conversely, with constant high-pressure headaches, trials of small adjustments can be made in the outpatient setting. Programmable valves can interact with magnetic fields. Thereby, MRI scans could modify the shunt settings [8], unless the patient carries MRI compatible valves and a 3-T or lower magnetic field MRI is involved (Table 3).

Typically valve settings are confirmed via skull X-ray (Fig. 2). Occasionally, programmer devices allow for transcutaneous valve readings. If a skull X-ray is required, a comparison with the manufacture's diagram will allow correct interpretation and reading of the setting (Fig. 2).

Clinical history and the increasingly available previous medical records may provide prior imaging that may ascertain reported recent valve setting changes.

The distal shunt tubing is tunneled subcutaneously posterior to the pinna and connects the valve to the anatomical CSF reabsorption site (from most common to rarest: peritoneum, atrium, pleura, gall bladder, azygos vein, and ureter) [13–15]. Distal locations for CSF diversion present with unique risk profiles (Table 4).

Ventricular Access Device

Some proximal catheters will be connected to a ventricular access device such as an Ommaya or Rickham reservoir. These devices allow percutaneous access to the ventricular system. Two clinical indications for these reservoirs include newborn post-hemorrhagic hydrocephalus and

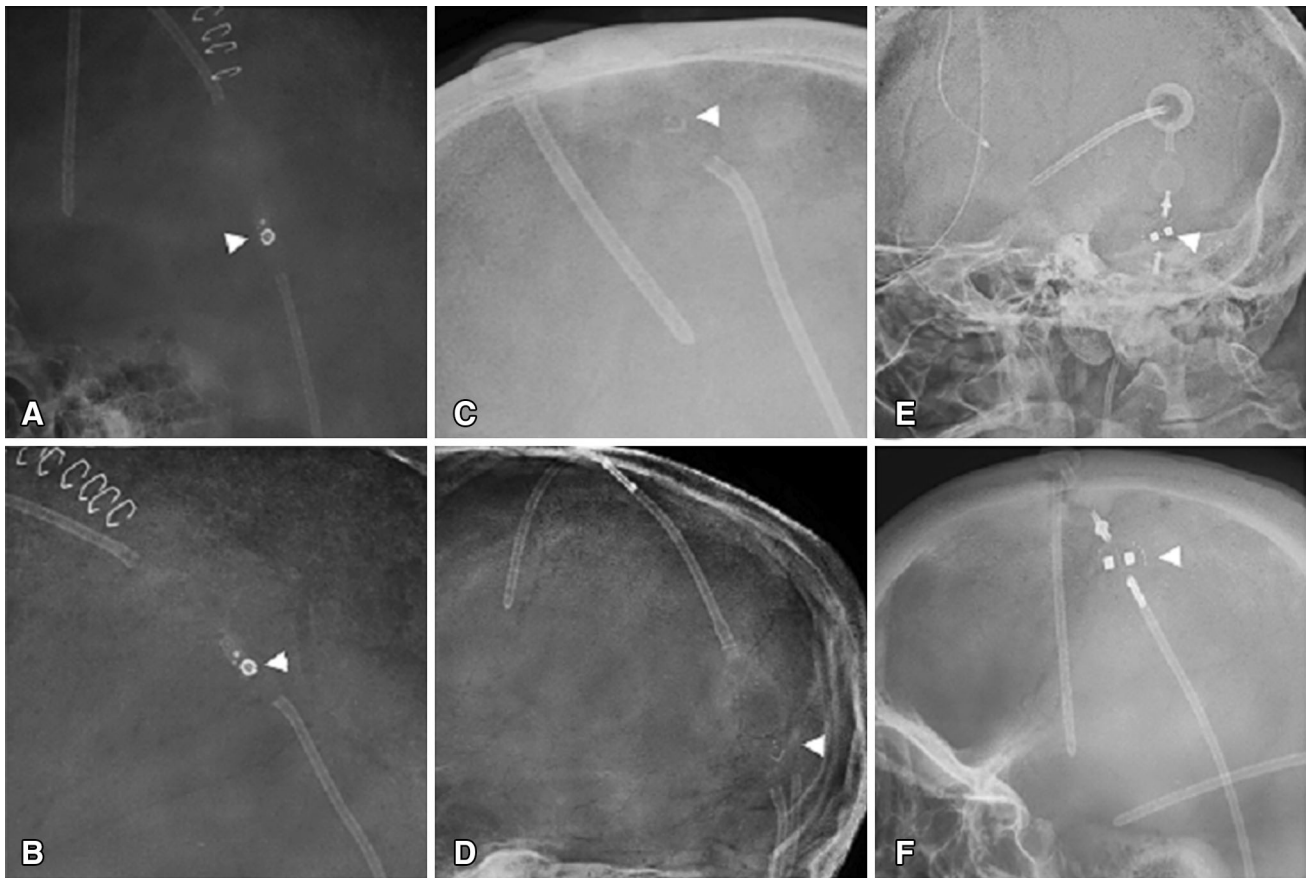


Fig. 2 Radiological identification of different valve types. **a, b** Codman Hakim[®] set at 40 and 120 (4 and 12 cm H₂O), respectively, **c, d** Medtronic[®] low and medium fixed-pressure valves (3–4.5 and

8.5–10.5 cm H₂O), respectively, **e, f** Sophysa[®] programmable valves set at 14 and 17 cm H₂O, respectively

conditions requiring intrathecal drug delivery [16]. Ventricular access devices may be accessed via a small-gauge butterfly needle inserted perpendicular to the skin at the apex of the dome of the reservoir in aseptic conditions similar to those required for a lumbar puncture.

Diagnostic WorkUp for Shunt Malfunctions

A majority of shunt malfunctions may be diagnosed radiographically. Thus, a suspected shunt malfunction assessment includes a radiological shunt series and imaging of the brain (Table 3) [17]. Radiographs help identify valve settings, shunt continuity, position and give a programmable valve's settings, which should be verified in case of recent manipulation (Fig. 1) [18]. A non-contrast brain CT or a fast-spin T2-weighted MRI should also be performed to evaluate for evidence of hydrocephalus development [17, 19, 20].

Shunt puncture may be indicated in the setting of concerning symptomatology in the absence of

radiographic findings. This approach allows direct testing of the proximal catheter. Besides, failure to obtain CSF is diagnostic of a shunt failure. Notably, individuals with a clinical suspicion of pseudotumor cerebri or normal-pressure hydrocephalus, may benefit from a radionuclide tracer injection to evaluate shunt patency [17, 20, 21]. Shunt taps can be performed safely and with minimal risk of introducing an iatrogenic infection by utilizing standard sterile technique to prepare the site over the reservoir, and insert a 23-gauge butterfly needle into the reservoir [22]. Intracranial pressure should be measured at the level of the foramen of Monro (i.e., tragus), and CSF fluid sent for analysis. Values greater than the valve setting suggest a shunt malfunction in the appropriate clinical setting (Table 3). Lumbar puncture might also be helpful. However, clinicians must realize that reservoir pumping is typically ineffective because 40% of obstructed shunts will show normal refill [22–24]. This approach might be diagnostic and therapeutic in severe cases, or if a delay in accessing neurosurgical care occurred [23, 25, 26].

Table 1 Programmable pressure valves

Programmable pressure valves	Strata	Medtronic
		Magnetic programmable ball and spring delta valve
	Codman-Hakim	Can be manipulated by MRI
		Codman (Johnson and Johnson)
		Magnetic programmable ball and spring valve
		One centimeter H ₂ O incremental settings
	Sophy	Can be manipulated by MRI
		Sophysa
		Silicone-coated chamber
		Ball cone with variable pressure set to spring
	Polaris	One centimeter H ₂ O incremental settings
		Can be manipulated by MRI
		Sophysa
		Silicone-coated chamber
	ProGav	Magnetic field self-locking safety feature
		One centimeter H ₂ O incremental settings
		MRI compatible
		Aesculap

MRI Magnetic resonance imaging

Table 2 Non-programmable valves

Fixed-pressure valves	Hakim	Codman (Johnson and Johnson)
		Built-in siphon guard
		Zero–five centimeters H ₂ O outflow resistance
	PS Medical	Medtronic
		Four different fixed-pressure increments
		Injectable proximal reservoir
Chhabra	Very low cost	
	Gravitational three-ball mechanically regulated system for incremental drainage	
Flow-regulated valves	Delta	Medtronic
		Elastomer diaphragm regulate
	Orbit-Sigma	Five different flow levels
		Integra
		Three-stage variable resistance mechanism
		MRI compatible

MRI Magnetic resonance imaging

Shunt Complications Common to Any Site

All neurointensivists are familiar with acute hydrocephalus and high-pressure symptoms ranging from characteristic headaches to a Cushing's response. However, other circumstances might be more challenging from the diagnostic perspective. A thorough medical history is crucial since relatives and patients with recurrent shunt malfunctions will faithfully describe how the shunt fails.

Over-Drainage and the “Siphon Effect”

Over-drainage represents a significant long-term ventriculo-peritoneal (VP) shunt complication. It may occur in patients with impaired ventricular compliance (i.e., chronic normal-pressure hydrocephalus). In these circumstances, VP malfunction manifests with a clinical picture suggestive of low intracranial pressures with headaches, nausea, and vomiting. Frequently, acute or chronic subdural hematoma

Table 3 Diagnostic imaging and tests to evaluate a shunt malfunction

Plain radiographs	
AP + lateral skull film	Visualization of proximal catheter, continuity of tubing, and valve including settings
AP chest and abdominal film ^a	Visualization of continuity and localization of distal catheter system
Cranial imaging	
Axial CT or rapid T2 sequence MR imaging of the brain	Assessment of the ventricular system and proximal catheter placement
Dynamic imaging	
Radionuclide tracer studies	Proximally injected tracer can be tracked moving through a functioning shunt in cases where chronic malfunction is suspected without other signs on imaging
Invasive procedures	
Shunt tap	Direct access to proximal catheter system, which can confirm flow with spontaneous flow
Lumbar puncture	In cases of known communicating hydrocephalus, elevated opening pressure can correlate with a shunt malfunction

^a Abdominal film is only necessary for peritoneal shunts

Table 4 Common symptoms and causes of shunt malfunctions

	Ventriculoperitoneal	Ventriculoatrial	Ventriculopleural
Unique signs and symptoms	Abdominal pain, swelling at incision site, low-grade fevers, decreased appetite, and nausea/vomiting	Septicemia, swelling, or pain of unilateral upper extremity from DVT formation, low-grade fevers	Shortness of breath, pleuritic chest pain, fevers
Proximal obstruction or infection	Proximal obstruction or infection can be caused by bacteria, debris, inflammation, or poor wound healing in all shunt systems		
Distal obstruction	Mechanical Peritoneal fat, colon, debris	Mechanical Thrombus, occlusion	Mechanical Debris, pleural scarring
	Relative Chronic constipation causes increased abdominal pressure Ascites Carcinomatosis	Relative SVC syndrome, right-sided heart failure	Relative Pleural effusion, scarring
Infection	Spontaneous peritonitis Liver failure Cancer patients	Bacteremia Hemodialysis patients Sepsis Endocarditis	Pleural empyema
	Abdominal infections Cholecystitis Diverticulitis Appendicitis Abdominal surgery		
Malabsorption	Pseudocyst formation Infection Malabsorption from scar tissue	Shunt malfunction symptoms, no radiographic findings	Pleural effusion or empyema formation
Disconnection or technical error	Distal tubing migration or disconnection in peritoneal cavity	Shunt fracture with migration in the SVC or right side of heart	Migration of distal tubing into pleural space
	Preperitoneal shunt tubing placement	Kinking or malpositioned distal catheter tubing at venous access site	Malpositioning or kinking of pleural tubing

DVT Deep venous thrombosis; *SVC* Superior vena cava

from chronic over-drainage develops, particularly in normal-pressure hydrocephalus patients [4, 24]. Valve adjustment or shunt removal might be required if collections occurred. The fluid collection may require drainage via a separate catheter or burr hole in select cases [3, 4, 25].

The “siphon effect” occurs in patients when a large change in the pressure gradient within the distal cavity occurs. For instance, following sustained recumbence or performing Valsalva maneuvers, the sharp change in the abdominal compartment will draw fluid from the distal shunt tubing into the abdomen. Under these circumstances, rapid CSF shunting leads to immediate onset of low-pressure symptoms follows. Although antisiphon systems are installed in many valves, not all devices have them (Tables 1, 2). When siphoning occurs, the amount of fluid allowed to drain off is proportional to the setting of the valve, with high-pressure valves allowing less volume drainage than low-pressure valves. This is a challenging diagnosis to make in the absence of radiographic evidence of over-drainage and is most often diagnosed based on a detailed history.

Slit-Ventricle Syndrome

Slit-ventricle syndrome (SVS) is a radiographic diagnosis of slit (minute) ventricles. The etiology arises typically from three different CSF system abnormalities. The first is with a non-compliant ventricular system where no changes in size are seen in response to intracranial pressure changes. Thus, the neuroimaging remains stable despite potential elevations in intracranial pressure. This most often arises in pediatric populations following chronic shunting. Diverse pathophysiological mechanisms have been proposed, including long-term CSF over-drainage brain tissue expansion with ependymal changes leading to rigidity of the ventricular wall, and underproduction of CSF. Other mechanisms such as capillary resorption delay, which may lead to intraventricular and extraventricular pressure dissociation may contribute to SVS.

More commonly, SVS is seen in cases of idiopathic intracranial hypertension or pseudotumor cerebri. This disease is one of the chronic long-standing elevated intracranial pressures. The underlying pathophysiology is of great debate. However, many authors believe it is related to sinovenous stenosis or increased arterial inflow with poor venous drainage of the brain leading to elevated intracranial pressure and SVS on imaging [26]. SVS can also present from chronic over-drainage with low-pressure signs and symptoms as previously discussed. Workup for these patients may include shunt tap to ensure flow and typically opening pressure measurements through a lumbar puncture. Treatment often depends on the etiology of the SVS and may include successful removal of the shunt

following clamping protocol, endoscopic third ventriculostomy with or without shunt removal, or lumboperitoneal shunt for subarachnoid space drainage to prevent proximal obstruction in anatomically small lateral ventricles [27].

Shunt Malfunction

Varying study definitions explain the wide range of shunt dysfunction prevalence (8–64%) [3, 7, 8, 21, 22, 28, 29]. However, there is consistency that approximately 50% of shunt failures occur within a year of its implantation [7, 29]. Shunt malfunction symptoms depend on its etiology and the type of shunt and valve placed (Tables 1, 2, 4) [3, 4, 21, 30].

Shunt Obstruction

Shunt obstruction is the most common cause of failure. Clinicians should always search for abdominal tenderness. When present, an emergent abdominal ultrasound is indicated to rule out pseudocyst.

Proximal occlusions are typically ensuing in the acute phase. Conversely, distal catheter obstruction or fracture occurs in the period following the initial 2 years after placement (Fig. 3) [14, 28]. In the latter case, surgical exploration is often necessary for confirmation.

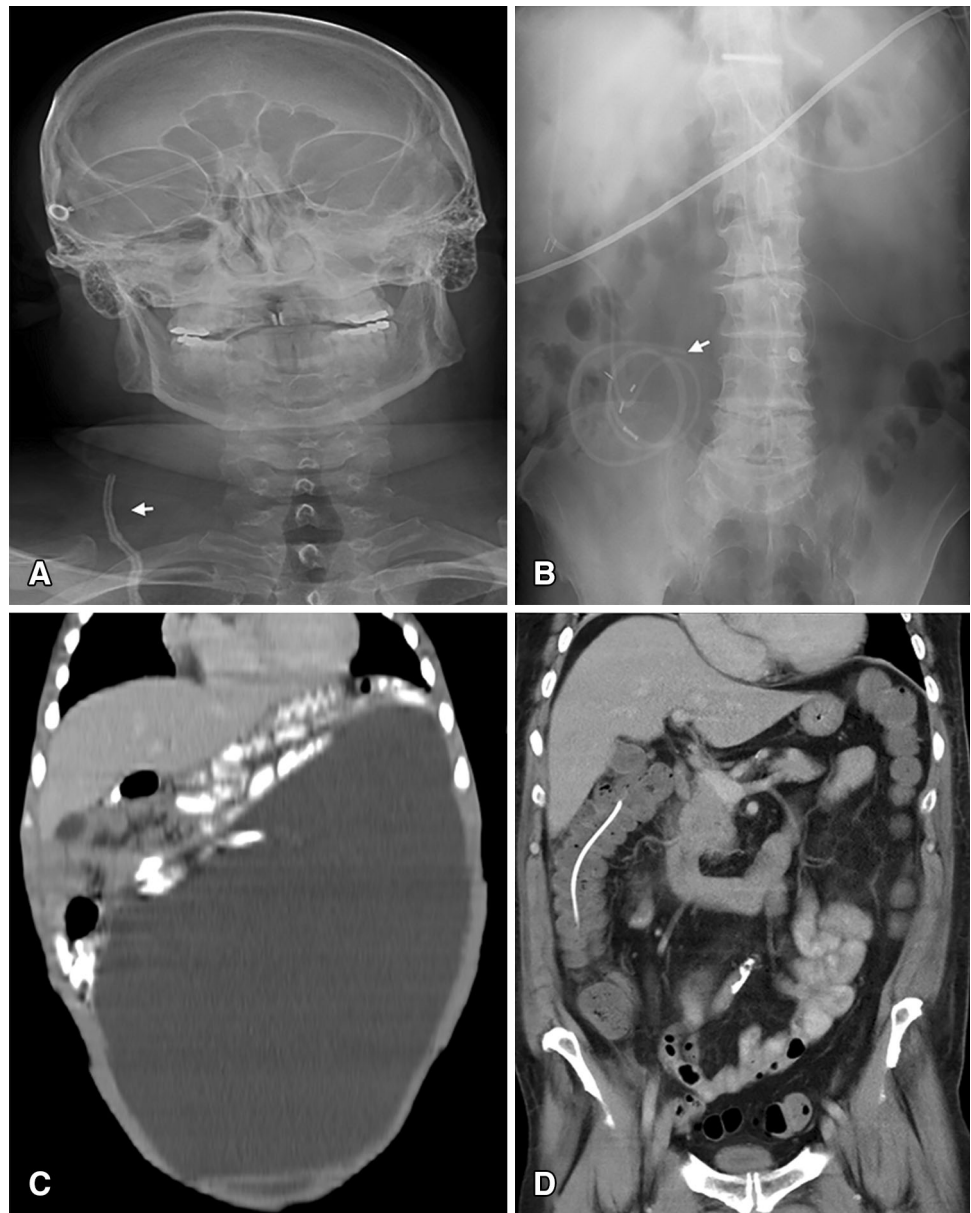
Technical Error and Disconnections

Individuals with a shunt placed decades ago might initially present with insidiously raising intracranial pressures. It may manifest as “arrested hydrocephalus” with radiographical ventriculomegaly, yet not clinically acute hydrocephalus. In these instances, the hardware is typically fractured or occluded. Indeed, disconnection of ≥ 2 components secondary to a procedural error or suture breakage may occur. Fluid accumulation or drainage from the wound near the site of disconnection would be evident and malposition confirmed by a shunt series [28]. Conversely, scar tissue can fixate the tubing and subsequently stretch and fracture the catheter, which may migrate distally in the peritoneum, rectum, or urinary bladder [1, 2, 4, 31].

Infection

The incidence of shunt infection varies among series (1.6–17%) owing to the lack of diagnostic guidelines in adults [23, 32–36], although tentative criteria have been proposed in pediatric populations [33, 37, 38]. Nearly 85% of the infections occur during shunt placement or in the perioperative period [32, 39, 40]. Most frequently, these occur from direct surgical wound contamination by

Fig. 3 Radiological diagnosis of typical shunt device complications. Plain radiographs of skull and abdomen. **a** Shunt fracture and distal migration of tubing are seen in a patient presenting with symptoms of a shunt malfunction, **b** preperitoneal tubing is visualized coiled in the abdominal wall in a patient presenting with a painful and tense abdominal incision site, **c** patient presented with intractable abdominal pain, CT imaging of the abdomen revealed massive abdominal pseudocyst, **d** patient presenting with signs of meningitis and CT revealed migration of shunt tubing into the ascending colon



skin flora (i.e., *Staphylococcus epidermidis* or *aureus*, *Corynebacterium*, and *Propionibacterium acnes*) [33, 36, 41]. Conversely, infections with a peritoneal origin due to gram-negative agents (i.e., *Pseudomonas aeruginosa*, *Serratia marcescens*, *E. Coli*, *Stenotrophomonas*) or *Candida albicans* often occur in the subacute or chronic period [33, 41–45]. When gram-negative infection occurs, a gastrointestinal source, such as migration of catheter into the bowel, ought to be ruled out with a CT of the abdomen.

Unlike meningitides, shunt infections may display a vague clinical picture with headache, fever, and malaise. Empirical broad-spectrum antibiotics should be initiated once CSF cultures have been drawn [37, 46]. Other

laboratory investigations should include complete blood count and inflammatory markers including erythrocyte sedimentation rate and C-reactive protein. These values are relevant in the diagnostic workup and to trend the therapeutic response. Complications of infection such as a ventriculitis involve high mortality rates (30–40%) and may warrant intraventricular antibiotics [37, 46]. Local infections manifest with different symptoms depending on the distal site of draining. For example, peritoneal shunts might present with abdominal pain, swelling at incision site, low-grade fevers, decreased appetite, and nausea/vomiting. Ventriculoatrial shunts may present with septicemia, swelling, or pain of unilateral upper extremity from DVT formation, low-grade fevers.

In the event of bacteremia, the hardware should be rapidly removed. A sterile extraventricular drainage device would be indicated until the infection is fully treated with antibiotics. Pleural shunts might declare with shortness of breath, pleuritic chest pain, fevers.

A Cochrane review including seventeen randomized control trials examined the value of systemic antibiotics and catheter-impregnated antibiotics as primary prophylaxis for intraventricular catheter infection in an aggregate of pediatric and adult populations. Both perioperative systemic antibiotics and antibiotic-impregnated catheters decreased infections with a number needed-to-treat of 20 and 11, respectively [47]. The data were insufficient to address all-cause mortality. Although encouraging, these data may need to be interpreted with caution as the studies were not analyzed with intention-to-treat, but per-protocol, which may bias the results. In addition, a recent meta-analysis of 36 non-randomized studies including 16,796 procedures suggested that antimicrobial-impregnated and coated shunt catheters may reduce risk of skin flora infections [47]. Ultimately, were an infection confirmed, shunt removal and replacement with a temporary drainage system or partial externalization would follow at the neurosurgeon's discretion for there are no definitive guidelines [37].

Peritoneal Shunt Complications and Considerations

Pseudocyst may develop as a consequence of a sterile inflammatory response to high protein CSF content or shunting system components, as well as owing to a subacute infection [48]. Typically, a history of abdominal surgeries, several shunt revisions, and more rarely allergies to silicone, ethylene oxide, or latex herald the possibility of a pseudocyst [49–51].

Patients will often present with acute hydrocephalus symptoms accompanied by abdominal pain, discomfort, or poor PO intake. Pseudocysts are diagnosed on abdominal ultrasound or CT imaging (Fig. 3). Management includes externalization of the catheter from the abdomen. CSF from the shunt and pseudocyst, and the distal catheter should be cultured. Shunt re-internalization will follow once an infection is treated or ruled out [49–51]. In the setting of recurrent pseudocyst formation or the diagnosis of an intra-abdominal infection, the shunt is typically converted to a ventricular–atrial (VA) shunt. In the event that free air is found on radiographs or distal catheter is identified protruding from the anus, hollow viscus perforation must be considered. Antibiotic therapy should be started immediately, and the intra-abdominal shunt should be externalized or removed [52].

Clinical Pearls

Three specific subgroups of patients with shunted hydrocephalus that require further discussion are patients with Chiari malformation, NPH, and baclofen pumps with shunts.

Chiari Malformation

Untreated Chiari malformations with a shunt malfunction may present with worsening Chiari symptoms owing to progressive downward herniation and subsequent intracranial pressure elevations. Careful physical examination, diagnostic imaging, and shunt tap will evidence shunt malfunction. Treatment may involve shunt revision and Chiari decompression [53].

Baclofen Pump and Shunts

With regard to baclofen pump carriers, patients under consideration for shunt placement must have their shunts assessed for functionality prior and after placement to ensure medication will be cleared properly. Shunt dependency can lead to baclofen toxicity if underdrainage or obstruction is present [54].

Shunt Management in Normal-Pressure Hydrocephalus

Shunt complications in NPH represent a clinical challenge. Symptoms may include those of over-drainage, which include low-pressure headaches and presence of subdural fluid collections. Symptoms of shunt failure most often manifest as an exacerbation of NPH symptoms. The workup includes a careful examination, brain CT, shunt series, and shunt tap if needed. If these studies are unrevealing and over-draining is suspected, ligation or removal of the shunt may be indicated. Conversely, if underdrainage and malfunction were suspected, a lumbar puncture with high volume drainage would aid in diagnosis. The issue is further complicated in that often times NPH patients require minimal changes in the programming of the valve. For instance, a change in the setting of a Codman-Hakim valve from 70 to 50 may improve an NPH patient who presents with symptoms of shunt malfunction.

Seizure

Finally, breakthrough seizures may occur within 48 h following shunt placement. Indeed, cortical irritation secondary to the procedure may account for the seizures.

However, shunt malfunction must be ruled out for it may manifest with seizure activity.

Choice of Shunt Site Placement

Alternatives to VP shunts must be considered when distal complications have occurred or peritoneal placement is contraindicated (i.e., from elevated intra-abdominal pressures, scarring and prior infections, history of major abdominal surgeries, or malignancies). Placement options include ventriculoatrial, ventriculogallbladder, ventriculopleural, and ventriculoazygous shunts. There are no guidelines to support efficacy for an alternate distal site for implantation [55]. However, many neurosurgeons advocate for VA shunts as a second choice since long-term durability is similar in VA and VP shunts.

VentriculoAtrial Shunt Complications and Considerations

VA shunts pose a unique set of potential challenges for patients. Distal obstruction from thrombus formation or scarring and stenosis of the superior vena cava may occur [56]. Thus, swelling of the upper extremity should raise the suspicion for thrombus or fibrosis in this vessel. Duplex ultrasound and vein mapping of both upper extremities should be performed [57]. There is no supportive evidence for primary prophylaxis for thrombus formation with anticoagulation or antiplatelet medication in the setting of atrial shunts [58]. Indeed, if a patient developed pulmonary hypertension, pulmonary thromboembolism must be ruled out. If confirmed, oral anticoagulation would be required.

Besides thrombus formation, VA shunts when infected may induce bacteremia. In patients with positive blood cultures, the catheter must be removed to treat the infection and avoid endocarditis or septic emboli. Other serious complications include distal catheter migration into the right ventricle, which must be retrieved endovascularly [59]. Despite the aforementioned possibilities, recent retrospective series reported reassuring safety data in VA shunts as compared to VP shunt in adults with normal-pressure hydrocephalus [57]. VA recipients had no cardiopulmonary complications after almost four years of average follow-up. Nevertheless, prospective randomized trials are lacking.

Pleural Complications and Considerations

Ventriculopleural shunts also present with unique complications including pleural empyema, effusions, respiratory failure, and catheter migration into the pleural space

[60, 61]. Patients with this shunt often present with dyspnea or respiratory failure when fluid accumulates. Once identified, thoracentesis and/or chest tube placement should be performed if the patient is symptomatic. Prompt externalization and attempted aspiration of the pleural collection can also be attempted [60, 61]. If the fluid is concerning for an infection, thoracic surgery should be involved as endoscopic procedures may be required to treat the infection.

Rare Distal Site Alternatives, Complications, and Considerations

The gall bladder, azygos vein, and sternum are all rare sites that can be considered for distal implantation. All have proven efficacy in small case reports [62–64]. Ventricular–gallbladder shunts develop infection or obstruction in around 10% of patients [62]. Azygos vein shunts are technically difficult and have similar risk profiles to VA shunts [63]. Sternal shunts are rare with the unique risk of osteomyelitis to the sternum [64].

Future Directions

Less invasive methodologies for rapid shunt malfunction diagnosis are in constant development. For example, FDA-approved subcutaneous thermal dilution technology (e.g., ShuntCheck III®) cools the upstream CSF via transcutaneous refrigeration of the shunt catheter and enables downstream temperature measurements. A change in temperature is considered verification that there is flow in the shunt system. In addition, constantly evolving information technology in handheld devices and smartphones (e.g., HydroAssist™) allows patients to record a detailed history of shunt settings and clinical correlates are critical for accurate diagnosis.

Conclusions

Shunt systems are complex and vary widely. A basic understanding of shunt design, symptoms of malfunction, unique presentations from distal CSF reabsorption sites, and diagnostic tests is essential for neurointensivists. Were a shunt malfunction suspected in a setting where access to a neurosurgeon is limited or delayed, the workup and temporizing measures described in this manuscript should be immediately initiated, while neurosurgical consultation is sought emergently.

Compliance with Ethical Standards

Conflict of interest None of the authors have a conflict of interest.

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