Cerebrospinal shunt malfunction: recognition and emergency management

Indwelling shunts to divert CSF flow are essential in treating hydrocephalus. There is a high incidence of shunt malfunction, which accounts for the increasing popularity of endoscopic third ventriculostomy. Failure to recognize and act on symptoms and signs of shunt malfunction may lead to loss of life or to permanent neurological dysfunction. This review provides the basis for assessment and management based on updated clinical knowledge.

The presentation of a patient with a CSF shunt malfunction or infection can pose a significant diagnostic and therapeutic challenge to clinicians in the emergency setting. The ability to recognize and initiate the active management of potential shunt complications is essential given that most emergency departments and acute medical units in the UK do not have access to on-site neurosurgical expertise. Patients with little remaining compensatory reserve may deteriorate suddenly from respiratory arrest, seizure or coning. Shunt blockage may cause blindness and death in the presence of sudden onset and delay in treatment.

This review provides guidance to emergency department staff who are likely to encounter patients attending with shunt-related complications. A literature search was carried out on Medline from 1970 onwards, with no language restrictions. Standard texts on neurosurgery and emergency medicine were consulted. The selected papers were further cross-referenced. One author (AAK) has also drawn on recent experience of working in a neurosurgical unit.

Shunts

Shunts are the mainstay of hydrocephalus treatment. Hydrocephalus is defined as an excessive accumulation of CSF in the ventricular system caused by disturbance of formation, flow or resorption.

The basic shunt structure comprises a short length of proximal ventricular tubing, a subcutaneous one-way valve to control the rate of drainage of CSF (preventing excessive drainage) and prevent back flow, a silicon pumping bulb, an anti-siphoning device, and longer distal tubing (containing valves that prevent back flow). Most shunt components are radio-opaque, ensuring visibility on plain radiographs. Valves are of three types: diaphragm, slit or spring-ball. The valve may be programmable, allowing programmability of opening and closing pressure, and if so the setting is changeable with an external magnet. Some patients may also have a ventricular access device or Ommaya reservoir in the right frontal or right parietal region, which is located proximal to the one-way valve and overlying the burr hole. This allows urgent percutaneous aspiration of CSF in the presence of life-threatening acutely raised intracranial pressure.

Various types of shunt are in clinical use. They are usually named after their draining area and drainage sites. The distal catheter may be placed in the peritoneal or pleural cavities, right atrium, internal jugular vein or gall bladder. The ventriculoperitoneal shunt is the most commonly used shunt at present, as ventriculoatrial shunts are uniquely susceptible to the risks of endocarditis, cardiac tamponade, pulmonary embolism and glomerulonephritis (‘shunt nephritis’), while also not allowing for growth in later life. The ventricular catheter of the shunt is normally inserted into the frontal or occipital horn of the lateral ventricle through the cortical mantle, via a burr hole in the right parieto-occipital region, and the valve is usually located behind the right ear. The tip of the ventricular catheter should lie in the lateral ventricle anterior to the foramen of Monro in order to maximize its distance from the choroid plexus. It demonstrates high attenuation on computed tomography (CT) scanning and appears hypointense on T1 and T2 weighted magnetic resonance imaging (MRI) scanning. The distal catheter is tunnelled distally and subcutaneously down to another incision in the abdomen whereby it is placed in the peritoneal cavity. Most neurosurgeons use medium pressure valves that will drain continuously if the differential pressure is over 10 mmHg.

Ventriculoatrial shunts drain the lateral ventricle through the internal jugular vein and superior vena cava, with the catheter tip placed in the right atrium. This is the shunt of choice when abdominal abnormalities are present, for example if the patient has had extensive abdominal surgery or peritonitis, or is morbidly obese. The ventriculopleural shunt is an alternative if the peritoneal cavity is unavailable. The use of the lumboperitoneal shunt is confined to communicating hydrocephalus

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draining from the lumbar subarachnoid space to the peritoneal cavity. The subdural shunt is usually used for subdural hygroma, draining the subdural space to the peritoneal cavity.

Shunts have become increasingly sophisticated since their introduction in the early 20th century. Problems such as over-shunting or under-shunting have been overcome and modern shunts with programmable valves allow the closing pressure to be altered externally using a special magnetic device. This is sometimes extremely useful in selected shunt patients with intractable headache but it can lead to problems following an inadvertent change of pressure.

Shunt complications

All shunts are prone to malfunction, 30% failing within 6 months of placement, 50% failing by 6 years and 70% failing by 12 years (UK Shunt Registry, unpublished data, 2005). Shunts may also fail unexpectedly after many years without symptoms (Tomlinson and Sugarman, 1995). Shunt malfunction can be the result of obstruction, infection, disconnection at the junction or break, hardware erosion through the skin, over-shunting and subdural haematoma (*Table 1*). Under-drainage is the commonest reason for shunt revision. The overall complication rate from shunts has been reported from 4–30% and the rate of serious complications is 9.4% (Greenberg, 2001). Serious complications include blockage, infection, haemorrhage and permanent neurological deficit (Pople, 2002). Infection occurs in 10% of patients within the first year of placement.

Recognition of shunt complications

Recognition of shunt malfunction involves assessment of the patient as a whole and assessment of the shunt hardware. Shunt evaluation includes ascertaining the reason for initial shunt insertion (e.g. meningitis, tumour, myelomeningocele), documenting the date and reason for the last revision, and the type of valve implanted (programmable/adjustable).

Table 1. Shunt complications that may present to an emergency department

Infection	
Malfunction	Blocked proximal catheter
	Blocked distal catheter
	Valve dysfunction
	Over-shunting
	Disconnection
Invasion of abdominal organs (ventriculoperitoneal shunt)	
Peritonitis, ascites, abdominal pseudocyst (ventriculoperitoneal shunt)	
Pulmonary emboli (ventriculoatrial shunt)	
Glomerulonephritis (ventriculoatrial shunt)	
Pleural effusion (ventriculopleural shunts)	

Adapted from Lennon (2006)

The importance of paying careful attention to the observations of parents, families and carers, particularly if they have had previous experience of shunt block, cannot be overemphasized. Watkins et al (1994) demonstrated that families were at least as accurate as paediatricians in diagnosing shunt block. One should always suspect shunt malfunction in a symptomatic child with a shunt and no alternative convincing explanation for the symptoms.

The history should allow the recognition of raised intracranial pressure and of CNS infection. Acute and florid presentations tend to relate to increasing intracranial pressure or meningitis. Subacute subtle presentations also occur, associated with a progressive insidious rise in intracranial pressure, and may be heralded by neuropsychological, cognitive and behavioural symptoms.

Block is the commonest reported complication in most series. In the largest reported cohort of 1719 patients, 56% experienced at least one episode of shunt block in the 12 years following insertion (Sainte-Rose et al, 1991). The peak ‘danger’ period for blockage is in the first year after insertion with rates as high as 20% recorded in some series (Peacock and Currer, 1984). The commonest site of obstruction is in the proximal catheter. Shunt obstruction may occur proximally in the ventricular catheter because of blockage by choroid plexus, red cells, tumour cells or high protein concentration (protein plug) in CSF, although the latter cause is open to dispute. Blockage of the distal catheter can occur as a result of adhesions within abdominal cavity, especially when associated with low-grade infection.

Presentation of shunt block may be associated with what might be termed ‘classical’ symptoms of raised intracranial pressure, namely headache, vomiting and drowsiness (Lee et al, 1999) or may occasionally be atypical or misleading (Jamjoon and Wilson, 1988). Common classical features include irritability, headache, drowsiness, gait disturbance, sixth nerve palsy, fatigue, failing vision, papilloedema, generalized clumsiness and ataxia. Atypical presentations that have been reported include seizures, abdominal pseudocyst, syringomyelia and hemiparesis (Lee et al, 1999).

The coexistence of headache, vomiting and drowsiness make it very likely that an affected patient has shunt dysfunction; in most cases this will mean acute shunt block. Drowsiness is by far the best predictor of shunt block, while headache and vomiting are less predictive of shunt block (Barnes et al, 2002). The patient may deteriorate suddenly as a result of respiratory arrest, coma, seizure or coning. Shunt blockage may cause blindness and death as a result of dilatation of the third ventricle with compression of optic chiasma or occlusion of posterior cerebral artery by downward transtentorial herniation (Greenberg, 2001). An increase in head circumference, scalp vein distension, persistent bulging of the anterior fontanelle and suture diastasis are seen in infants.

CSF leak around the proximal catheter may indicate either a blocked proximal catheter or disconnection, and may lead to the formation of a fluctuant swelling around the burr hole where the shunt enters the cranium.

Unusual manifestations of shunt malfunction include the Parinaud syndrome in patients with pre-existing intracranial neoplasm, and neck pain, lower cranial nerve palsy and syringomyelia in myelodysplastic patients (Key et al, 1995).

Infection is one of the most devastating complications of shunt placement. Shunt infection usually occurs within a few months of shunt surgery – 70% of infections present within 2 months of surgery. Shunt infection is associated with substantial morbidity, including risk of seizure disorder and decreased intellectual performance.

Approximately 3–12% of patients develop shunt infection (Casey et al, 1997). Shunt infections are commonest shortly after insertion and are usually caused by the patient's own skin organisms, highlighting the need for scrupulous aseptic surgical technique. The most commonly isolated organisms are *Staphylococcus epidermidis* (60–75%), *Staph. aureus* and Gram-negative bacilli. Shunt infection is suggested by fever (usually intermittent and low-grade), rigors, headache, drowsiness, lethargy, anorexia, generalized malaise, irritability, vomiting, peritonism (ventriculoperitoneal shunt), pleurisy (ventriculopleural shunt) and occasionally pain and erythema along the shunt device system. There is an associated high peripheral and CSF white cell count, but the raised peripheral white cell count is of low sensitivity and specificity. CT scan imaging may reveal ventriculitis, with enlarged lateral ventricles demonstrating irregular contrast-enhancing ventricular walls.

Possible complications of over-shunting include intracranial hypotension, slit ventricle and subdural haematoma: 10–15% of long-term shunt patients will develop one of these complications within 6 years of initial shunting. If this occurs, the patient may complain of postural headaches while sitting up, which resolve when lying down (McLone, 2000). Patients may present with nausea, vomiting, lethargy, double vision or up gaze palsy. Sometimes the symptoms may resemble those caused by increased intracranial pressure. The postural variation in symptoms is related to a siphoning effect resulting from a column of CSF in the shunt tube when the patient is erect. Ventricles may be slit like or may be normal in appearance. The shunt pressure may be set too low for an individual patient, leading to over-drainage. Such patients need adjustment of valve pressure either by revision of the shunt or by using programmable valves. The slit ventricle syndrome presents with spontaneous and episodic rise in intracranial pressure, in the presence of a patent shunt system, in association with a small or slit-like ventricular system. The syndrome is associated with a wide range of intracranial pressure, ranging from low to high (Rekate, 1993).

The incidence of subdural haematoma formation following shunt insertion is estimated at 4–23% in adults and 2.8–5.6% in children (Greenberg, 2001). It may be caused by collapse of the brain with tearing of bridging veins. Negative pressure in ventricles because of siphoning may also facilitate the development of subdural haematoma. The collection may be on the same side as the shunt in 32%, on the opposite side in 21% and bilateral in 47% (Greenberg, 2001). They rarely present acutely. While most are clinically not significant, subdural haematoma may be symptomatic in 40% of cases. Symptoms often resemble those of shunt malfunction and these require treatment. Anecdotally, the incidence has been reduced with the use of high-pressure valves.

Abdominal complications may occur on occasions. Bowel perforation, ileus, pseudocyst, ascites or abscess formation are potential problems (Chidambaram and Balasubramaniam, 2000). In rare instances catheter migration has been documented to cause bladder perforation or migration into the scrotal sac resulting in a hydrocoele (Ward et al, 2001).

Initial management

The emergency department management of a patient with shunt malfunction includes resuscitation along standard lines, followed by focused assessment, investigations, emergency treatment and final transfer to neurosurgical care.

Physical examination should include documenting the Glasgow Coma Score, visual acuity and field of vision. Optic fundoscopy should also be performed for papilloedema. A septic screen looking for potential sources of infection is also essential.

The first definitive step involves locating and assessing the shunt hardware. Most reservoirs can be compressed easily and rapidly refill in a few seconds. Silicon pumping bulb incompressibility is usually the result of distal catheter obstruction. If the bulb compresses easily but does not refill, the proximal catheter is blocked. Bulb manipulation requires expertise which may not be readily available in an emergency department, and may precipitate blockage of the CSF pathway by the choroid plexus in the presence of small ventricles. This is because the reservoir capacity, ranging from 20–25 ml, may be drained distally, allowing the choroid plexus to block the ventricular end of the catheter.

In a moribund child, raised intracranial pressure can be relieved by inserting a 25-gauge butterfly needle into the bulb or ventricular access reservoir at an angle of 45° to the skin under aseptic precautions. CSF is drained off until the pressure is 10 cm H_2O. In a stable patient, the

Table 2. Investigations for shunt malfunction

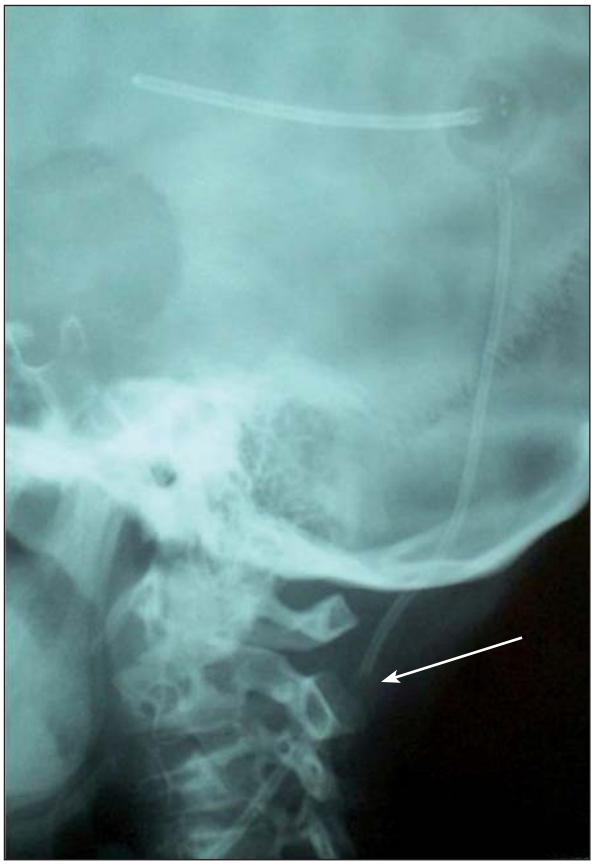
Routine	Full blood count		
	Urea and electrolytes		
	C-reactive protein		
	Blood culture		
	Urinalysis		
Special	Radiological	X-rays shunt series	Lateral skull
			Neck (lateral view)
			Chest (AP)
			Abdomen (AP)
		Computed tomography brain	
	CSF shunt tap		
	Lumbar puncture		
	Intracranial pressure monitoring		

AP = antero-posterior

entire length of the subcutaneous shunt should be inspected and palpated.

Investigations that might be required to evaluate shunt-related complications are listed in *Table 2*. Twenty-five per cent of shunt-infected patients have a white cell count less than $10x10^9$/litre, while a third of patients have a white cell count of more than $20x10^9$/litre. Urea and electrolytes are checked to evaluate renal function. The serum C-reactive protein is almost invariably elevated in the presence of shunt infection. Blood cultures are positive only in one-third of patients. Proteinuria and haematuria may occur with chronic shunt infections.

Figure 1. Broken shunt.

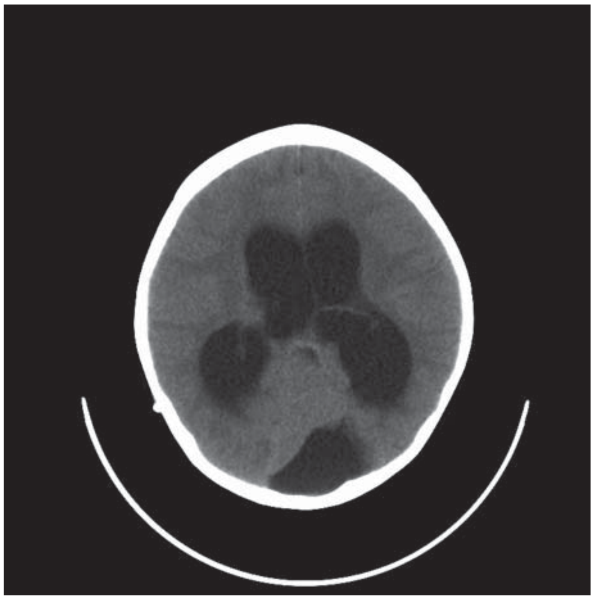


Imaging is an essential adjunct to the clinical evaluation of patients with suspected ventriculoperitoneal shunt malfunctions or complications. The entire course of the shunt is initially examined with plain radiography (shunt series) in the stable patient for the presence of disconnections, kinks, breaks (*Figure 1*), or migration of the shunt tubing that may be confirmed later with other imaging modalities. Cranial CT is the usual preferred imaging mode. It is used to assess the overall size of the ventricular system and to ascertain the presence or absence of peri-ventricular oedema (*Figures 2* and *3*). Comparison with previous CT scans is usually helpful (note patients often retain copies of previous scans).

An increase in hydrocephalus is not always seen in the presence of shunt blockage, and CT scan appearances can widely fluctuate over a relatively short time. Clinical monitoring of the patient is of primary importance. In the neurosurgical unit intracranial pressure monitoring may be required to monitor progress.

Zorc et al (2002) reported on the predictive ability for suspected shunt obstruction on plain radiograph as having a sensitivity of 20% and a negative predictive value of 22%, whereas using a CT scan had a sensitivity of 83% and a negative predictive value of 93%. Combining the two tests gave a sensitivity 83% and negative predictive value of 95%. Obliteration of the perimesencephalic cistern mandates urgent neurosurgical consultation (Johnson et al, 1986). This CT sign was noted in a series

Figure 2. Computed tomography of the head, showing a blocked shunt.



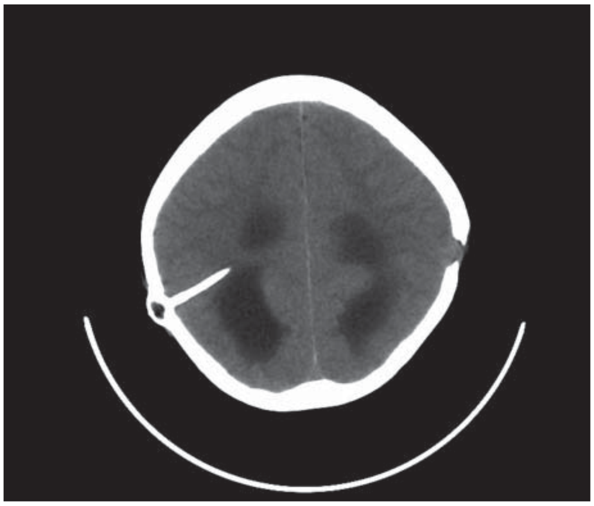


Figure 3. Computed tomography of the head; showing a functioning shunt.

of seven children presenting with life-threatening shunt malfunction. With ventriculitis the lining of the ventricle demonstrates contrast enhancement. Over-drainage is associated with subdural haematoma, post-shunt craniosynostosis and the slit ventricle syndrome. Small ventricles may be non-compliant and do not necessarily imply a normal intracranial pressure. Subdural haematoma in this situation tends to be a space-filling rather than a space-occupying lesion. Cranial ultrasound may be useful in infants, in whom skull sutural fusion has yet to take place.

Abdominal ultrasound may demonstrate a CSF pseudocyst or ascites (Briggs et al, 1984). CSF tap is usually done in neurosurgical units. It is useful for cell count, protein concentration and to exclude residual infection. A protein concentration greater than 4 g/litre may clog up a majority of ventriculoperitoneal shunts. Lumbar puncture is usually not recommended, and may be hazardous in obstructive hydrocephalus with non-functional shunts. CSF white cell is elevated in about 70% of cases with infection, but a positive CSF culture is the gold standard for diagnosis of infection. Bacterial antigen detection and polymerase chain reaction testing may be useful.

Definitive management

With infection, removal of the shunt and placement of a temporary external ventricular drain is indicated. Intensive antibiotic therapy is followed by placement of a new shunt when the infection is cleared.

Morphine derivatives are generally avoided for analgesia: non-steroidal anti-inflammatory agents can be used in the absence of contraindications. Patients should be given empirical antibiotics after taking blood culture, the choice of antibiotics being guided by the hospital antibiotics policy. Most neurosurgical departments use intravenous vancomycin or ceftriaxone because they have good CSF penetration. Urgent neurosurgical consultation is recommended to prevent potential delays in treatment and to prevent devastating complications of shunt malfunction. **BJHM**

The authors are grateful to the neurosurgical unit at Queens Medical Centre, Nottingham, for the radiographic images.
Conflict of interest: none.

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KEY POINTS

- CSF shunts are widely used in treating hydrocephalus.
- Shunts can be associated with morbidity and a risk of death.
- Awareness of potential complications is necessary in emergency care settings.
- Shunt morbidity can be reduced by improvements in design.