Management of the patient with a ventriculoperitoneal shunt presenting with headache

Introduction

CSF shunts have been the definitive treatment for patients requiring CSF diversion (e.g. hydrocephalus, syrinx and pseudomeningocoele) for several decades. CSF may be shunted from intracranial or spinal CSF spaces to various drainage sites, including the peritoneum, cardiac atria and pleura. The most commonly encountered shunt in neurosurgery is the ventriculoperitoneal system, which typically comprises three parts: the proximal tubing (ventricular catheter), the valve (with or without an antisyphon device) and the distal tubing (running from the head to the peritoneum).

Over one third of ventriculoperitoneal shunts will fail within 1 year of insertion (Drake and Saint-Rose, 1995; Wu et al, 2007). The consequences of untreated shunt failure are potentially fatal; prompt diagnosis and timely intervention are vital in preventing long-term morbidity or mortality in these patients.

Patients with previous obstructive hydrocephalus may have undergone an endoscopic third ventriculostomy as an alternative to shunt insertion. These may also fail and if they do, the clinical presentation and workup shares much in common with that described below for patients with ventriculoperitoneal shunts.

Shunt failure

Shunt malfunction can be categorized by aetiology into infective, mechanical and functional failure (Chumas et al, 2001).

Infective

Quoted infection rates for ventriculoperitoneal shunts vary but it is likely that the overall rate is 5–15% (Kulkarni et al, 2001; Wu et al, 2007). About 75% of shunt infections occur within 1 month of insertion and 90% within 1 year (Samandouras, 2010). While there are many possible causative microorganisms, the most commonly encountered are the patient's own bacterial skin flora, frequently either *Staphylococcus epidermidis* or *Staph. aureus*. Where shunts have distal insertion within the peritoneum, it is important to consider enteric organisms such as *Escherichia coli* and *Klebsiella* as potential causes of infection. Antimicrobial prophylaxis may be undertaken as a preventative step in patients with shunts in situ undergoing certain procedures such as laparoscopic surgery. Contrary to popular belief, there is no evidence to support prophylaxis for dental procedures (Acs and Cozzi, 1992; Lockhart et al, 2007).

Mechanical

Mechanical causes account for at least half of all ventriculoperitoneal shunt failures (Drake and Saint-Rose, 1995). The most common reason for mechanical failure is obstruction of the shunt tubing or valve. The most common site for occlusion is the ventricular catheter, as a result of infiltration by choroid plexus. A second risk factor for shunt blockage is where there is a substantially elevated CSF blood load (as may be seen in the early stages of post-haemorrhagic hydrocephalus) which may clot within the shunt's lumen. Other mechanical causes of failure include disconnection, fractures or migration of the tubing.

Functional

Functional failure describes a situation where the shunt system is either over- or under-draining (over-drainage is more common). Over-draining ventriculoperitoneal shunts siphon excessive CSF volumes from the ventricle to the abdomen, the consequences of which include subdural haematoma and the so-called 'low-pressure state' (described later). Patients whose shunt is under-draining present with signs and symptoms of raised intracranial pressure, although the deterioration may be sub-acute as some CSF is still draining through the shunt. Patients with functional shunt malfunction may require shunt valve replacement to adjust their drainage, or in certain cases a programmable valve may be adjusted using a magnetic device. Programmable valves are potentially susceptible to magnetic fields and may need reprogramming following a magnetic resonance imaging scan (see individual manufacturer's website for details).

History

All neurosurgical units should have an open door policy for patients with ventriculoperitoneal shunts whereby they can (and should) directly contact the centre in question, rather than a GP or local accident and emergency department.

In patients presenting with headache the history is key – while these patients experience 'benign' headaches exactly as would any other person, they are also at increased risk of more sinister causes for their symptoms. Patients who are well enough will often know a great deal about their previous history. If the patient is unable to give a full history because of infancy, disability or drowsiness, a history from the next of kin (often a parent) is vital – ignore this at your (and more importantly the patient's) peril.

A malfunctioning shunt is a neurosurgical emergency – fast the patient from the outset, rather than waiting until the history, examination and investigations have all been completed.

Always take a focussed shunt history to obtain details of the patient's previous history, asking:

- When was the original shunt inserted and why?
- Have there been any revisions? If so what are the details?
- What is the current shunt configuration? Patients often know exact details of the type of shunt and valve that they have. Many patients will have multiple

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shunts and fragments from previous operations, so it is important to know which shunt is believed to be the functioning system.

- When, where and by whom was the current shunt inserted?
- Perhaps the most pertinent question to ask of patients who have undergone previous malfunctions is 'When you had a problem before, did it present in this manner?'

One technique for taking a headache history in these patients is to ask about key symptom groups that tend to lend themselves to one diagnosis or another (*Table 1*).

Consider alternate causes of headache such as subarachnoid haemorrhage, cerebral ischaemic events, migraines or medication headaches.

Finally, remember to undertake a thorough review of systems, especially in paediatric patients. Children with viral infections may present as potentially malfunctioning shunts, it has been suggested that there is decreased CSF absorption in the unwell child. Clarify the sequence of symptoms and explore other potential diagnoses such as gastroenteritis, viral infections, constipation, ear, nose and throat problems.

Examination

Patients with shunt malfunction have the potential to deteriorate rapidly. If the patient might be too drowsy to protect his/her airway, request urgent anaesthetic assistance immediately and support the airway until a definitive airway can be provided.

With the exception of the aforementioned scenario, begin with general inspection. Remember that you are essentially looking for evidence of hydrocephalus or systemic infection. Does the patient appear drowsy? If so, shunt malfunction is likely.

More formal examination should include but not be confined to a full neurological examination. Patients tend to present with decreased consciousness and other signs of raised intracranial pressure, rather than with lateralizing signs. Fundoscopy should always be performed (although normal fundi do not rule out hydrocephalus or raised intracranial pressure). In patients presenting with decreased Glasgow coma scale who are thought to be in status epilepticus, it is important to carefully examine to ensure that they are not in fact displaying decerebrate posturing as a result of raised intracranial pressure.

In paediatric patients, neurological examination should include measuring and plotting a head circumference and palpating the fontanelle in those young enough to do so (usually those under the age of 12 months).

The shunt should be examined along the whole tract from head to abdomen to look for any areas of erythema (suggestive of infection) or fluid collection. Pay particular attention to the surgical sites, usually a curvilinear incision behind the ear and a subcostal abdominal scar. When examining the shunt valve try to locate the CSF reservoir (if present); this should depress and refill briskly. If the reservoir depresses but does not refill (or is slow to do so) this suggests a blockage proximal to the shunt valve. If the reservoir is firm and does not depress easily, this suggests a problem with the shunt valve or the tubing distal to it. Even in experienced hands, this is an unreliable sign as 40% of obstructed shunts will depress and refill normally (Aghababian, 2011).

Abdominal examination should be performed, not only to palpate for any CSF collection or tenderness but also to assess for other causes of symptoms such as nausea and vomiting (if applicable) and for any evidence of abdominal complications of ventriculoperitoneal shunt insertion, including peritonitis. If there is the suspicion of an abdominal CSF collection (typically caused either by a migrated or mispositioned peritoneal catheter tip or by malabsorption of CSF in viral illnesses) an ultrasound of the abdomen should be requested.

Where a diagnosis of an infected shunt is considered, it is important to search for other potential sources of sepsis.

Table 1. Potentially shunt-related presentations

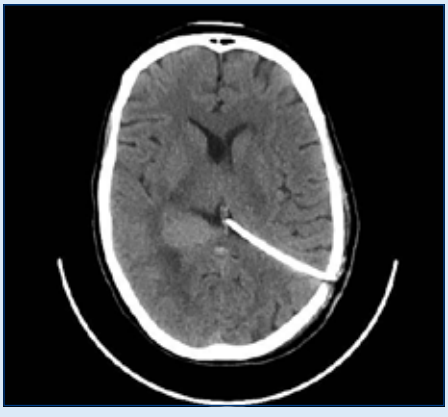
Raised intracranial pressure	Constant 'pressure' headache, worse on lying or bending, worse on waking
	Nausea +/- vomiting
	Visual disturbances (e.g. diplopia and problems with upward gaze)
	Drowsiness +/- confusion
	Balance problems
	Seizures (new or of increased frequency)
Infection	Progressive headache
	Fevers, chills or rigors
	Meningism, photophobia
	Seizures (new or of increased frequency)
	Urinary tract infection
Low-pressure symptoms	Nausea +/- vomiting
	Headache worse on sitting and standing, eases on lying down
	Signs and symptoms of subdural haematomas; limb weakness or decreased or fluctuating Glasgow coma scale (which may occur as a consequence of overdrainage)

Investigation

If there is clinical suspicion of shunt malfunction, appropriate investigations should be obtained without delay.

An unenhanced computed tomography scan of the head (*Figure 1*) can be performed rapidly in most hospitals and can

Figure 1. A computed tomography scan of the head showing a left-sided ventricular catheter sitting within the left lateral ventricle. The ventricles appear well decompressed.



be used to assess for evidence of hydrocephalus, confirm shunt catheter position and exclude several other causes of headache. When assessing the computed tomography, it is important to compare the images with previous scans to ascertain if there has been any change (remember to check whether previous scans were performed when the patient was well or unwell). Some patients with shunts may have copies of a 'well scan' with which any new imaging can be compared; failing this, the local neurosurgery centre may have images on their own system. Do not be falsely reassured by an apparently normal computed tomography in the context of someone who clinically appears to have shunt malfunction.

When a computed tomography scan is requested, so too should be shunt series X-rays (*Figures 2–4*). This consists of X-rays of the shunt tubing as it passes from cranium to abdomen, i.e. skull (anteroposterior and lateral) and anteroposterior: neck, chest and abdomen. When viewing the shunt series, the shunt should be traced from one end to the other, looking for any disconnections or kinks. Common sites for disconnection are where one component meets another, e.g. the connection between proximal catheter and valve or valve and distal tubing. A second reason for shunt malfunction that can be diagnosed on shunt series is migration of the distal tubing out of the peritoneal cavity and into the subcutaneous tissue, as seen on lateral abdominal films.

Ventriculoperitoneal shunt patients presenting acutely need urine dipstick and urgent bloods sending for full blood count, clotting screen, urea and electrolytes, group and save and inflammatory markers plus blood cultures if infection is suspected. Do not wait until a decision is made to take a patient to theatre before preparing them for surgery; send the bloods off as early as possible.

Where discussion with a tertiary neurosurgery centre is anticipated, remember to ask for any imaging (current and previous) to be sent to the department in question (often these can be electronically linked from one trust's system to another).

Sampling CSF from the shunt system (or tapping the shunt) enables CSF to be analysed for evidence of infection, as well as allowing measurement of the CSF pressure. In extreme presentations of acute hydrocephalus, larger volumes of CSF may be aspirated from the shunt as a life-saving procedure to decompress the ventricles before transfer to a neurosurgical centre. A shunt tap should never be performed without prior discussion with the local on-call neurosurgical registrar, the rationale being that a shunt tap could introduce infection into what may actually be a sterile system (*Table 2* gives practical details of how to perform a shunt tap).

Figure 2. A lateral skull X-ray showing the ventricular catheter, valve (circled) and distal tubing in continuity; there is no evidence of disconnection.

Figure 3. A chest X-ray showing the shunt tubing running vertically, to the left of the mediastinum.

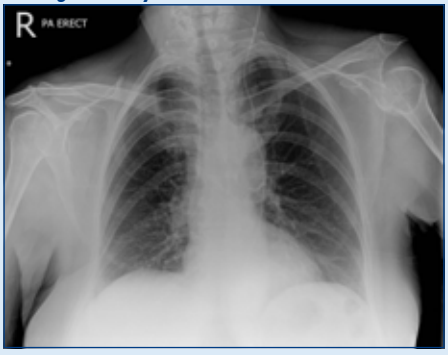


Figure 4. An abdominal X-ray showing that the tubing is sitting uncoiled within the peritoneal cavity.

Table 2. How to tap a shunt

Palpate the shunt reservoir	
Wash hands and prepare equipment	Clean trolley with a sterile field (such as a dressing pack)
	Sterile gloves
	Alcohol-free povidone-iodine solution
	25G (orange) 'butterfly' needle
	Manometer
	5 ml syringe
Clean the skin using the iodine solution and allow to dry	
Insert the butterfly needle into the shunt reservoir	
Connect the manometer to the butterfly and note the pressure of the CSF within the shunt system by holding the manometer level with the reservoir	
If CSF does not drain spontaneously, try lowering the height of the manometer to encourage drainage before checking CSF pressure	
Slowly aspirate 2 ml of CSF and send for analysis (Gram stain, microscopy, culture and sensitivity)	
Withdraw the needle and re-clean the skin	
A dressing is not usually required	

Management

Pending urgent transfer to a neurosurgical unit, the management of these patients is largely supportive, giving analgesia, antiemetics and anticonvulsants where appropriate and observing for any fluctuations in Glasgow coma scale, providing airway support if needed.

The definitive management of the malfunctioning shunt will be performed by the local neurosurgical centre and will vary depending on the underlying cause and local practice: mechanical failure is typically treated with surgical exploration and replacement of all or part of the shunt; antibiotics alone are not usually sufficient to treat shunt infection and surgical removal and substitution with an external ventricular drain (this allows continued CSF drainage and for the administration of intrathecal antibiotics if required) until the infection has been fully treated is almost always required; functional failure typically mandates a valve replacement or adjustment although in certain circumstances neurosurgeons may decide to first surgically insert an intracranial pressure monitor as a diagnostic adjunct.

As mentioned previously, in the rare scenario of the patient presenting in extremis, the neurosurgeon may ask the referring clinician to perform a therapeutic shunt tap to temporarily decompress the ventricles as a life-saving intervention. Similarly, if a patient has a communicating hydrocephalus, it is possible to perform a lumbar puncture to buy time by draining CSF.

Conclusions

Shunt malfunction is common. Prompt diagnosis and intervention are crucial for patients whose shunts have failed. While there are numerous causes for headache in these patients (as with any patient), the patients (or their parents) will often have a great deal of prior experience and will, themselves often know if something is wrong with the shunt. **BJHM**

Conflict of interest: none.

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KEY POINTS

- Always consider shunt malfunction in patients presenting with headache.
- If considering shunt malfunction, remember to keep the patient fasted until this has been excluded.
- Investigate early whenever shunt malfunction is suspected.
- Discuss potential cases with neurosurgery promptly; many patients with shunts will be well known to the department.
- Never tap a shunt without checking with your local neurosurgery centre first.
- Trust the patient and/or the family – if they say that the shunt is not working, they are usually right.

TOP TIPS

- History is key. Where applicable ask: 'Is this how it felt when your shunt failed last time?' If the answer is 'Yes', the shunt will almost certainly have failed again.
- Always look at the patient's fundi – even if you are a relative novice, you will quickly become comfortable with this technique if you practice.
- When reviewing a shunt series, trace the shunt tubing carefully from top to bottom. It may be necessary to adjust the contrast settings of your monitor to adequately visualise this. Pay particular attention to the connections within the system, i.e. either side of the valve.
- When evaluating a computed tomography scan of the head, insist on comparing with a previous scan if at all possible.
- Remember that a seemingly 'normal' scan does not guarantee that the shunt is working.