Pitfalls in the treatment of hydrocephalus

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Abstract
This article aims to describe the causes and types of hydrocephalus, as well as explain why treatment options may vary from case to case. Potential complications of treatment are also discussed.

What is hydrocephalus?

Hydrocephalus is a condition which develops when there is an abnormality in either the production or the removal of cerebrospinal fluid (CSF) from the ventricles within the brain and the spaces that surround the brain. The resulting build-up of fluid within these spaces, usually in the ventricles, causes an increase in intracranial pressure which can cause a variety of symptoms. The majority of patients with hydrocephalus have problems with absorption of the fluid rather than over-production. CSF is made by the choroid plexus, a rich network of blood vessels mostly located in the lateral ventricles of the brain and to a smaller extent within the fourth ventricle. Once secreted into the ventricles, the fluid then travels through small channels, including one particularly important channel known as the cerebral aqueduct, before exiting the ventricular system and entering the space around the outside of the brain. The CSF bathes the brain before coming up to the midline surfaces over the top of the brain where it is re-absorbed by a primarily passive process into the venous sinuses, notably the superior sagittal sinus, and then re-incorporated into the bloodstream.

The location of the difficulty in communication between the site of production and the site of absorption distinguishes the two main types of hydrocephalus known as communicating and non-communicating (or obstructive) hydrocephalus.

In communicating hydrocephalus, there is free-flow of CSF to the venous sinuses as well as through the subsequent connection to the spinal canal, where the CSF also bathes the spinal cord. The abnormality lies in delayed or reduced absorption at the venous sinus level. Common causes for communicating hydrocephalus include subarachnoid haemorrhage and meningitis.

In obstructive hydrocephalus there is a physical barrier to the fluid getting to the venous sinuses and the spinal column, leading to a build-up of fluid within the ventricular system. Common causes for obstructive hydrocephalus include tumours, particularly those that obstruct the cerebral aqueduct, and haemorrhages that lead to a clot being lodged somewhere within the ventricular system (also commonly at the aqueduct or within the cerebellum region).

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Symptoms and signs of hydrocephalus

Patients with hydrocephalus generally present with symptoms of raised intracranial pressure, and may arise in conjunction with symptoms caused by the underlying cause of the hydrocephalus. In broad terms, the hydrocephalus itself causes different sets of symptoms and clinical findings depending on the age of the patient.

In babies where the skull has not yet fused, hydrocephalus tends to present with vomiting, drowsiness, listlessness and irritability. If the history is more chronic, i.e. over several weeks, then an increase in head circumference may also be noticed by the parents. Occasionally parents may also comment that the child is unable to look upwards (known as 'sun-setting'); their neurological development may cease or they may even lose developmental milestones.

In older children and adults the primary symptom will be headaches associated with loss of appetite and vomiting, increased sleepiness, lack of concentration or confusion. These symptoms can progress to blurred vision, and the visual deterioration may be severe enough to leave the patient blind. Eventually there may be decreased conscious level, coma and death. Although these last consequences are relatively unusual outside of a very acute setting in developed countries, every year there are deaths or major disabilities as a consequence of undiagnosed or inadequately treated hydrocephalus in the UK.

Some neurological symptoms, such as cranial nerve palsies (often leading to double vision) or seizures can occur in any age group.

Investigation

The main investigation needed to make the diagnosis is imaging. In the vast majority of cases, this will consist of a computerized tomography (CT) scan (Figure 1). This is a quick (15–60 seconds) and relatively easily obtainable test. Most hospitals have access to 24-hour CT, and those that do not usually have agreements with local larger units to transfer patients for a scan. There is a small radiation dose with each scan, so it must still be ordered when felt necessary, not as a 'routine'.

In patients who have a more chronic history, are being considered for third ventriculostomy (Figure 2), or being investigated for an underlying cause of hydrocephalus (such as a tumour), a magnetic resonance image (MRI) scan may be obtained. This requires more time (up to 45 minutes),

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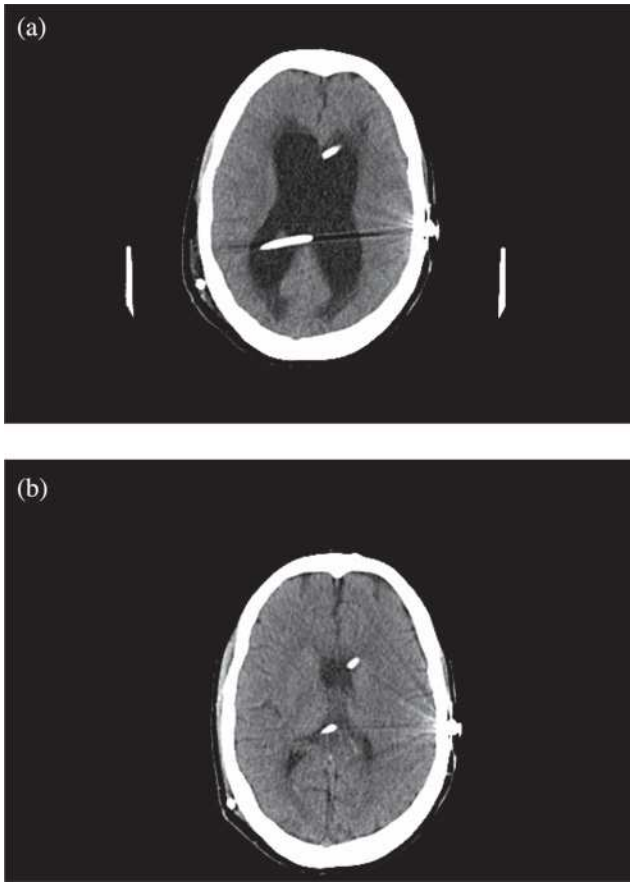

Figure 1 Axial CT scans of a young woman with multiple shunts *in situ*. She presented with a blocked shunt (Figure 1a). Note the presence of dilated ventricles as compared to the post-revision scan (Figure 1b)

and is not easy to obtain due to the relative lack of scanners. Even in major neurosurgical units, MRI may not be available 24 hours a day. However, it is radiation-free, something that is important if the patient has had a large number of CT scans, or for example, is pregnant.

In babies, an ultrasound is a very quick and safe method of imaging the ventricles. It relies on a bone-free area of the scalp, known as the anterior fontanelle, being present as in most babies up to the age of 18 months. The quality is dependent on the skill of the operator, and often is difficult to compare exactly with previous scans, but nonetheless, is a very useful test as it can be repeated whenever needed as it is radiation-free.

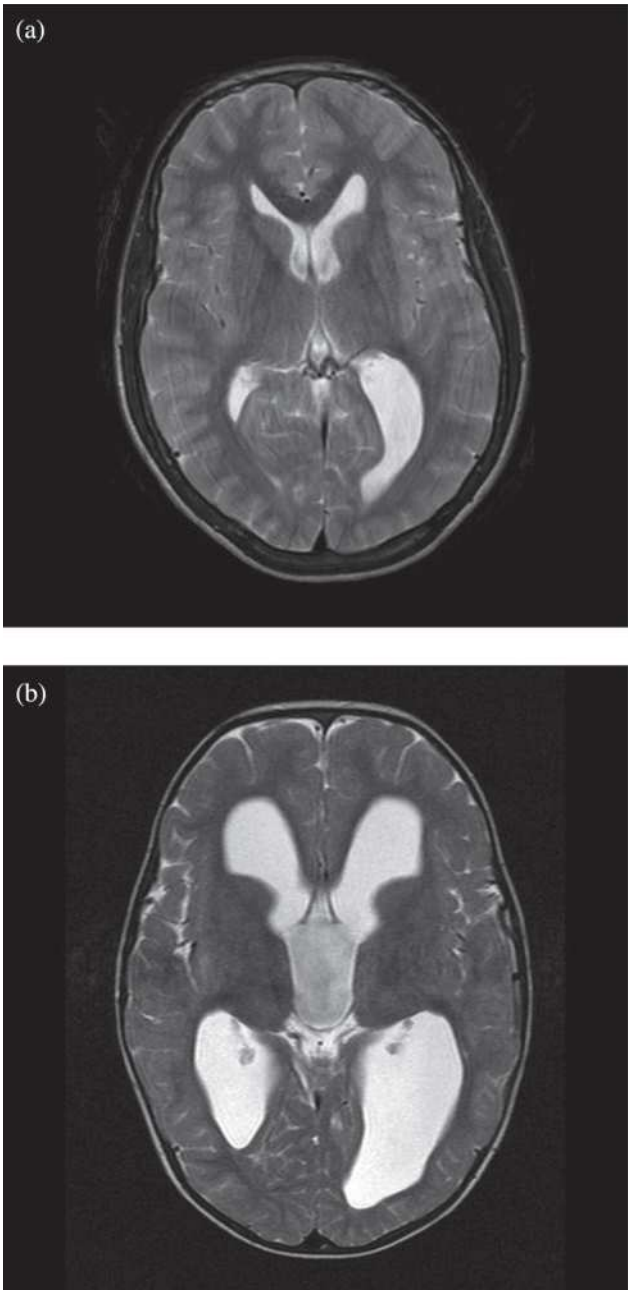

Figure 2 Scans of a patient who was scanned to exclude hydrocephalus secondary to a previously diagnosed subarachnoid haemorrhage (Figure 2a, negative for hydrocephalus). Some months later, he presented with symptoms of raised intracranial pressure, and had the scan (Figure 2b) confirming hydrocephalus that required treatment

Treatment of hydrocephalus

The treatment of hydrocephalus is by way of immediate management if presenting as an emergency followed by a long-term treatment plan. Treatment for the acute condition depends on distinguishing obstructive from communicating hydrocephalus.

Communicating hydrocephalus is treated relatively easily by performing a lumbar puncture, which can be carried out by almost any physician within a hospital setting. A needle is passed between the laminae (bones) of the lowest (lumbar) part of the spine and into the CSF space lying behind, which communicates with the ventricles. Fluid can then be drained from the spine via the needle, thereby lowering the intracranial pressure within the ventricles. However, in cases of obstructive hydrocephalus, there is not free-flow of fluid from the ventricles to the lumbar CSF spaces and, therefore, a lumbar puncture

cannot be performed safely. Instead, direct drainage from the ventricles is necessary, usually by insertion of an external ventricular drain, a tube passed through the brain into the ventricular spaces and connected to an external drainage bag which collects the excess CSF (Figure 3).

If a lumbar puncture is performed in the presence of obstructive hydrocephalus, then there is the potential for a life-threatening complication known as coning. If the pressure within the spinal canal is lowered by removal of CSF via the lumbar route, then the differential pressure between the intracranial compartment and the spinal compartment will actually rise. This can force brain tissue down into the craniocervical junction, and compress this very important part of the brain known as the medulla. Since it contains heart rate, breathing and other vital centres, its dysfunction following the compression (the coning effect) can rapidly cause life-threatening deterioration in a patient.

Although there are some clinical signs that distinguish between communicating and obstructive hydrocephalus, these are difficult to elicit and clinicians who are not neurological specialists would not be expected to note the difference (indeed there may be often no way of distinguishing without a scan). Therefore, when patients with a presumed diagnosis of hydrocephalus first present, a scan should be carried out both to confirm the diagnosis and that it is safe to carry out a lumbar puncture. In very extreme cases where a patient is losing consciousness without accessibility to a neurosurgeon or to a CT scan, it would be acceptable to perform a lumbar puncture without a scan, as a life-saving measure, especially if it is likely that meningitis is the cause of the hydrocephalus. This situation will be very rare in the UK, because of the widespread availability of both neurosurgeons and CT scans, and most NHS Trusts have guidelines available for performance of lumbar punctures in the setting of suspected meningitis.

Once the acute situation has been managed, consideration can then be given to the need for long-term drainage. Where meningitis or subarachnoid haemorrhage is the underlying cause of the hydrocephalus, it may be that intermittent drainage of the fluid is sufficient, either by repeated lumbar punctures or by the external ventricular drainage system. Once the infection or the blood load is cleared, spontaneous CSF re-absorption is possible and, therefore, no long-term CSF diversion procedure may be necessary.

Figure 3 A baby treated with an external ventricular drain. By raising or lowering the relative height of the external drainage collector, the intracranial pressure can be varied (siphon effect)

However, many patients will require one of two long-term drainage procedures: shunting or third ventriculostomy. The choice of procedure rests with the cause of the hydrocephalus.

Shunting

A shunt is a plastic tube which is used to divert CSF from the ventricles or from the lumbar CSF spaces to one of three destinations from where it is spontaneously re-absorbed. The most common destination is the peritoneum which lines the abdominal cavity. The next most common site is the pleural space around the lungs, and the least popular site is into a vein in the neck which eventually drains into the atrium of the heart. Generally, the abdomen is tried first as it is the safest for the patient. The other two destinations are generally only used as a second/third choice as they have a slightly higher complication rate unless performed by those surgeons more experienced in these procedures. The procedure of inserting both ventricular and lumbar shunts carries risks of complications including infection and blockage.

The infection rate varies from unit to unit, but as a rough estimate between 8–13% of shunts will become infected mostly within six months of implantation. The recent introduction of new types of catheters with impregnated antibiotics is likely to reduce this rate, although the long-term figures are not yet available.

Roughly 50% of ventriculoperitoneal shunts need to be revised within 5 years due to obstruction, and the rate is higher in lumbar shunts. Shunts can block as a result of protein debris within the ventricular catheter, the delicate valve mechanism that controls flow, or as a result of the omentum of the abdomen wrapping around the distal tip. The blockage of a shunt by omentum is a 'natural' reaction by the body and does not imply negligent insertion.

Other complications that may occur as a result of the shunt procedure include injury to the brain, intracerebral haemorrhage, epilepsy, trauma to the neck and chest wall (when tunnelling the catheter under the skin to its destination), and injury to internal organs such as the lungs, bowel, gall bladder and bladder, depending on the location of the drainage point. If the shunt has been directed into the atrium, injury to the cardiac cycle (including arrest), incompetence of cardiac valve function, renal complications (nephritis) and septicaemia have all been reported.

Third ventriculostomy

Because of the high risk of complications with insertion of a shunt, there has been an increase in use of third ventriculostomy. The procedure involves the use of an endoscope (a fibre optic telescope), inserted into the ventricles, to

form a hole through the floor of the third ventricle. This creates a communication between the third ventricle and the subarachnoid space, allowing the fluid pathway to bypass the cerebral aqueduct and fourth ventricle. This method, therefore, best works when the cause of the hydrocephalus is an obstruction after the third ventricle (e.g. a cerebellar tumour). This technique avoids the need for any implantation, thus avoiding the infection risk. The rate of re-obstruction appears to be around only 1% year, appreciably lower than that for shunt blockage. Risks during the operation, however, are higher, and include injury to brain, seizures, memory problems (due to injury to the fornix), pituitary gland damage, and injury to the basilar artery, which has a high chance of being fatal. A smaller bleed may require insertion of an external drain to clear the blood.

A third ventriculostomy procedure is not always appropriate, for example, there may be anatomical reasons during an endoscopy why ventriculostomy may not be safe, and a shunt may be chosen instead. In most modern units, with careful selection, around 60–70% of patients with hydrocephalus are considered for ventriculostomy, with a 70–90% success rate. If the CSF cannot be absorbed, then a shunt can always be placed as a last resort. Occasionally, a patient with a longstanding shunt in who is admitted with a blockage may receive a ventriculostomy in preference to a replacement of the shunt (Figure 4).

Long-term complications

Once a patient has been treated for hydrocephalus with either of these two long-term methods, there is a possibility of infection or failure at any time. As mentioned above, infection after a shunt tends to be within the first six months. Patients may show systemic signs of infection such as a raised temperature, redness or swelling along the shunt line, abdominal pain, or frank neurological deterioration if the infection spreads to the ventricles (ventriculitis). Infection after ventriculostomy may present after a CSF leak from the wound or *de novo*. The symptoms tend to be head-related, such as headache and confusion, as well as temperatures.

Blockage of shunts may occur acutely or over a longer period. If a patient is highly dependent on their shunt, their symptoms will be more acute and severe, and may include worsening headache, nausea and vomiting, drowsiness, confusion, seizures and visual disturbance.

Where the obstruction takes place over a longer period of time, the same symptoms may occur in a more insidious manner, but there may be more significant mental deterioration, and children often show a drop in learning ability.

Similar symptoms may occur after an obstructed ventriculostomy. This is important for doctors to recognize, as the absence of a shunt has led to false reassurance that 'it can't be hydrocephalus or you'd have had a shunt put in' when a patient re-presents with symptoms.

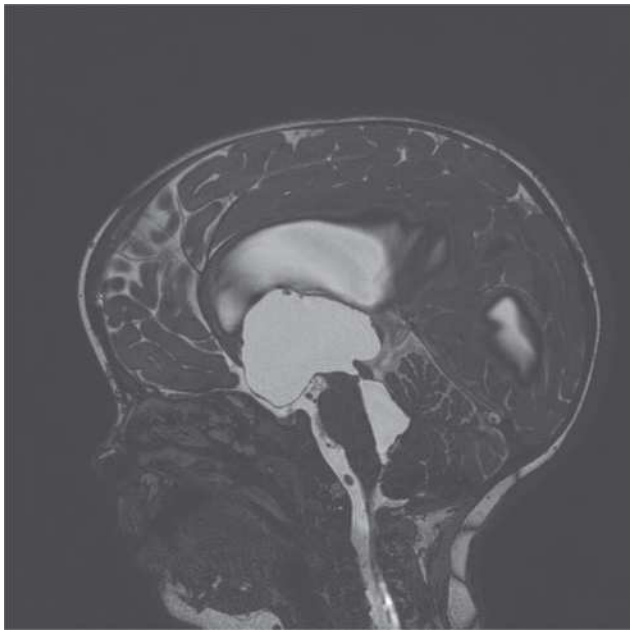

Figure 4 Preoperative scan of a patient treated with a third ventriculostomy, showing the approach and target for the procedure (arrow). Note the cause of the hydrocephalus is obstruction of the aqueduct (circled)

Treatment of long-term complications

Any patient presenting with symptoms of post-operative problems needs a thorough assessment. This involves taking a standard history and carrying out an examination, in particular looking at neurological aspects. Investigation will usually involve a scan; in an acute setting, a CT scan will suffice as it is quick, easy to obtain, low risk, and provides the necessary information in almost all cases. The scan may show gross ventricular dilation with other suggestive findings such as periventricular oedema (swelling around the ventricles), which confirms the diagnosis. However, some scans may not be so clear-cut and comparison with old scans is imperative. In a few cases (usually where there is post-infective or haemorrhagic hydrocephalus or other longstanding pathologies such as spina bifida), there may be no change in the ventricle size, even if the pressure is rising. This is because the walls of the ventricles do not have their usual elastic nature, which normally allows them to dilate to accommodate the increased fluid. Thus a small change in volume may lead to a dramatic increase in pressure. The condition is sometimes known as stiff ventricle syndrome due to the inability for the ventricles to change in size. If they are fixed at a very small size, they are also sometimes known as slit ventricle syndrome. These cases cannot always be diagnosed with scans, and the diagnosis depends on clinical acumen. In cases of doubt, intracranial pressure monitoring may need to be performed (Figure 5). There may, however, be sufficient suspicion to warrant direct surgical exploration of such patients. Neurosurgical referral is mandatory if raised intracranial pressure is suspected in these cases.

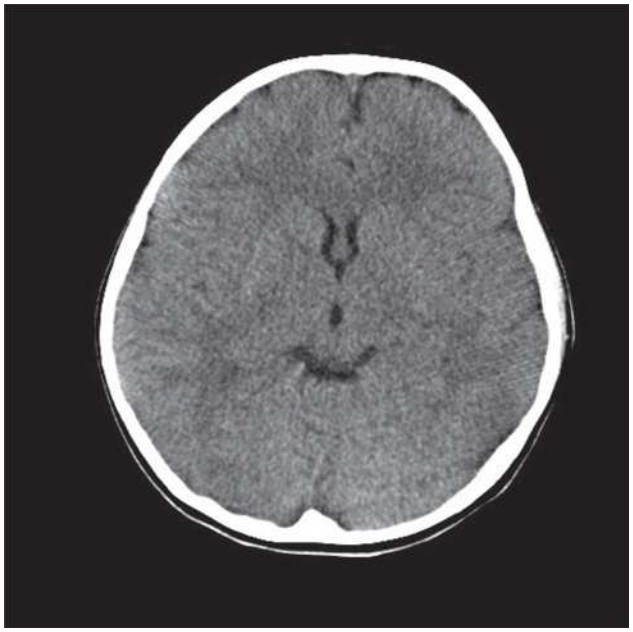

Figure 5 A child presenting with symptoms and signs of raised intracranial pressure. The scan shows small, normal sized ventricles, but ICP monitoring confirmed grossly raised pressures, and this child had a computer-guided insertion of a shunt, to good effect

When to operate?

The timescale for exploring a suspected blocked shunt varies. In the presence of lowered conscious level or other major problems, the surgery will clearly need to go ahead as soon as possible. If the patient is reasonably well, without deteriorating vision or other symptoms, then they may be put on the next available operating list, and it would be unusual for a patient with a blocked shunt to be monitored for much longer than this, unless their medical condition is well-known. In a few cases there may be a chance that the symptoms will resolve with careful observation, but this requires the treating physician to know that particular patient's 'hydrocephalus habits' very well, and for them to be ready to operate if deterioration occurs.

Summary

The symptoms and signs of raised intracranial pressure caused by hydrocephalus should be recognized by all physicians. Investigation and treatment regimens are well-established. In the case of 'treated' hydrocephalus, the spectre of recurrent raised intracranial pressure should also be remembered. In most cases the answer is clear and a good history, examination, CT scans and other investigations can rule out raised intracranial pressure. However, there are no tools outside of intracranial pressure monitoring to absolutely exclude recurrent raised pressure. In those cases with a convincing history and high index of clinical suspicion neurosurgical referral is mandatory regardless of the imaging findings and may justify open surgical exploration to make the final diagnosis, rather than risk the potentially fatal consequences of failing to treat raised intracranial pressure.

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