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CLINICAL REVIEW

# Contemporary management and recent advances in paediatric hydrocephalus

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The term hydrocephalus relates to an imbalance between the production and absorption of cerebrospinal fluid that may cause an increase in intracranial pressure. Rather than being a single pathological disease, hydrocephalus may be congenital or may arise secondary to intracranial haemorrhage, infection, or tumours.

Although some reports suggest the incidence of paediatric hydrocephalus has declined in many developed countries, others indicate increased rates because of improved survival of premature infants.[1-4] High rates of neonatal infection and neural tube defects in developing countries are linked to higher rates of hydrocephalus than in the developed world. One prospective observational study of hydrocephalus in east Africa estimated more than 6000 new cases a year, and it continues to be a major source of morbidity and mortality.[5]

The management of hydrocephalus was revolutionised in the 1950s with the development of shunts to divert cerebrospinal fluid away from the cranial cavity. This transformed hydrocephalus from a usually fatal condition to a manageable disease. Insertion of a ventriculoperitoneal shunt is now a common procedure in neurosurgical units worldwide.[5] However, patients with implanted ventriculoperitoneal shunts often present with complex and challenging problems that include infection and obstruction of the shunt. Large prospective observational studies have shown a shunt failure rate of about 40% at one year.[6]

We provide an overview of the modern management of paediatric hydrocephalus and an approach to problems that the generalist clinician may encounter. We also discuss future developments in the management of the condition.

## Who gets hydrocephalus?

Hydrocephalus can affect all ages, from infants to elderly people. Infants may have hydrocephalus at birth, caused by a congenital malformation that prevents adequate circulation of cerebrospinal fluid. Acquired hydrocephalus may arise after infection, intracranial haemorrhage, or structural or mass lesions. In this article we discuss paediatric hydrocephalus only, and box 1 lists the common causes of it.

Hydrocephalus may cause pathological changes to brain morphology, microstructure, circulation, biochemistry, metabolism, and maturation.[7] Although treatment does not always reverse the damage, untreated hydrocephalus will lead to progressive neurological decline and eventually death. In essence, once the diagnosis is confirmed a definitive treatment should not be unduly delayed.

## How does hydrocephalus present in children?

Despite the wide variation in aetiology, the clinical presentation of hydrocephalus is remarkably similar in all pathological processes and is mainly related to signs and symptoms of localised or generalised raised intracranial pressure (box 2).

## How is hydrocephalus treated?

Hydrocephalus remains a "surgical" condition in that the definitive treatment involves preventing raised intracranial pressure through diversion of cerebrospinal fluid out of the ventricles of the brain. Most commonly, this is achieved by the implantation of a permanent ventriculoperitoneal shunt—a silicone tube inserted into the lateral ventricle of the brain via a burr hole and tunnelled subcutaneously into the peritoneum (figure). Ventriculoatrial (lateral ventricle to internal jugular vein), ventriculopleural (lateral ventricle to pleural space), and lumboperitoneal (lumbar intradural space to peritoneum) shunts are acceptable alternatives for certain indications, such as when a ventriculoperitoneal shunt has failed or is not possible.

A shunt represents a lifelong commitment and should be carefully considered. Ventriculomegaly alone does not always require insertion of a shunt. Repeat evaluation showing

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Appendix: Types of valve used in cerebrospinal fluid shunt systems (see http://www.bmj.com/content/343/bmj.d4191/suppl/DC1)

**Summary points**

- Hydrocephalus may develop in association with a congenital brain malformation or, more commonly, after a neurological insult such as infection or haemorrhage
- Treatment involves draining cerebrospinal fluid from the lateral ventricles of the brain to another body cavity, usually the peritoneum
- Shunts are mechanical devices that are prone to obstruction, infection, and failure. Delayed recognition of shunt malfunction still causes death
- Endoscopic third ventriculostomy is an effective alternative to shunts in a select group of patients, but the stoma may eventually close and follow-up is needed to avoid fatalities
- Image guided placement of ventricular catheters may reduce the rate of poor shunt placement (further studies are required to evaluate this)
- Catheters impregnated with antibiotics or silver may reduce shunt infection (a large multicentre trial plans to evaluate them)
- Many children and adults can lead normal lives with shunts in situ, including participation in sports. Future prognosis and good health will be largely determined by avoiding complications and detecting shunt malfunction early

**Sources and selection criteria**

The data for this review article were compiled by a combination of Medline search, Pubmed search, a personal archive of references, and consultation with other experts. We reviewed evidence from published randomised controlled studies where available, cohort studies, clinical reviews, and evidence based guidelines.

**Box 1: Common causes of paediatric hydrocephalus**

*Congenital causes*

- Chiari malformation or spina bifida
- Aqueductal stenosis (X linked)
- Dandy-Walker complex
- Congenital arachnoid cysts
- Atresia of foramen of Munro

*Acquired causes*

- Haemorrhage
- Infection
- Traumatic head injury
- Tumour

progression of symptoms, signs, and ventricular size will confirm the need for shunt implantation.

Endoscopic third ventriculostomy has recently been developed, and large prospective multicentre cohort studies have found it to be an effective alternative to ventriculoperitoneal shunting in select cases that should be considered if appropriate.[8] This technique involves inserting a fibreoptic endoscope into the lateral ventricle and advancing it into the third ventricle. Using a balloon catheter, a stoma is created in the floor of the third ventricle, which allows cerebrospinal fluid to drain into the basal cisterns and thus into the subarachnoid space. If the operation is successful it obviates the need for a permanent shunt.

## What problems may clinicians encounter in children with shunts?

Obstruction or infection, and less commonly overdrainage of cerebrospinal fluid, can cause malfunction of the shunt. Long term prospective cohort studies have shown that repeated episodes of malfunction may result in impaired cognitive development and reduced IQ, multiple prolonged hospital admissions, repeat surgery, and increased risk of death.[2 3] The overall failure rate is 50% at two years.[9]

Obstruction most commonly occurs when the proximal ventricular catheter is blocked with choroid plexus or brain parenchyma if the shunt is poorly positioned. This leads to raised intracranial pressure and clinical presentation with signs and symptoms as outlined in box 2.

Large prospective observational studies have reported that infection occurs in 3%–27% of cases after shunt insertion.[4 9] Data from the UK shunt registry report that 15% of shunt revisions are due to infection.[6] Most occur within three months of surgery, and Gram positive organisms are the most common pathogens implicated.[10 11] The incidence of shunt infection is higher in neonates and children less than 1 year old than in other groups.[10 11] Patients with infected shunts may present with signs of raised intracranial pressure, meningism, raised temperature, and raised serum inflammatory markers.

Overdrainage of cerebrospinal fluid via the shunt, which is uncommon, can cause the cerebral mantle to fall away from the overlying dura and may result in tearing of delicate bridging veins and formation of subdural haematomas.[9] Patients may present with “low pressure” headaches, worse on standing than

**Box 2: Symptoms and signs of raised intracranial pressure in newly diagnosed hydrocephalus or shunt malfunction**

*In the infant and young child*

- Irritability
- Impaired level of consciousness
- Vomiting
- Failure to thrive
- Poor feeding
- Developmental delay
- Increasing head circumference crossing centiles
- Poor head control
- Tense anterior fontanelle
- Dilated scalp veins
- "Setting sun" sign (combination of upper eyelid retraction and failure of upgaze)
- Bradycardia
- Apnoeic spells
- Seizures

*In the older child and adolescent*

- Headache
- Vomiting
- Drowsiness or impaired consciousness and coma
- Diplopia
- Worsened seizure control
- Impaired upgaze
- Papilloedema

lying down, or may paradoxically present with focal or generalised signs and symptoms of raised intracranial pressure due to an expanding intracranial haematoma.

Abdominal symptoms secondary to shunt malfunction are relatively rare, but if suspected early referral to a neurosurgeon is warranted.

## How should a clinician manage a patient who seems to have a malfunctioning shunt?

### History and examination

A shunt malfunction is a potentially life threatening emergency. Patients still die from unrecognised shunt failure, and having a high index of suspicion of malfunction in a patient with a shunt who presents with vague symptoms may save lives, especially when a patient presents acutely unwell.

Symptoms and signs of shunt failure are essentially those of raised intracranial pressure (see box 2), but the presenting symptoms may be variable. Headache may be predominant in some cases, and vomiting and drowsiness in others. Children may often present similarly on repeated episodes of shunt block. Listen to the child's parents—if they say that the child is "not right" there is usually something sinister going on. Shunt infection may present with irritability, raised temperature, erythema along the shunt site, and raised intracranial pressure due to suboptimal shunt function.

A patient who is acutely unwell should be managed immediately according to the "ABC" of emergency medicine. The patient should be transported to the nearest emergency department to be assessed, stabilised, and discussed with the regional neurosurgical team.

### Imaging and other investigations

The important question to answer is: "Is the intracranial pressure raised?"

A standard first line investigation for a suspected shunt blockage is urgent cranial computed tomography to estimate ventricular size. If possible, comparison with previous images is useful since a larger ventricle size compared with the previous scan is strongly suggestive of a blocked shunt. Subdural haematomas or collections may be visible with overdrainage. Rapid sequence magnetic resonance imaging, although often less quickly obtainable, provides a radiation-free alternative and may be considered more often in the future.

However, shunt blockage with potentially dangerous raised intracranial pressure may not always result in an appreciable enlargement of ventricular size, and in such cases a clinical history and examination that suggests a blocked shunt should take precedence, with a low threshold for referral to a neurosurgical unit. Ventricular volume can be unreliable, and a child with small ventricles may still have critically raised intracranial pressure. Chronic overdrainage of the ventricular system can result in small ventricles that do not expand when the shunt is blocked even when the pressure is high. This phenomenon is termed the slit ventricle syndrome and is one reason that ventricular volume in itself is not a reliable index of shunt function.

Plain x rays of the shunt tubing (shunt series x rays) are useful to rule out tubing disconnection or fracture leading to shunt malfunction.

Ultrasonography is useful in neonates with an open anterior fontanelle as it allows the diagnosis of ventriculomegaly at the bedside. Serial ultrasound scans can be used to monitor ventricular size, especially in the preterm infant with

intraventricular haemorrhage. If the technical support for ultrasound is not available then urgent cranial computed tomography should be performed if the patient can tolerate transport to the imaging department.

Other investigations, such as for serum C reactive protein and white cell count, may show raised levels in shunt infection. If shunt infection is suspected referral to the regional neurosurgery unit is indicated.

## How can shunt problems be reduced?

### Image guided neuro-navigation for shunt placement

The traditional method of shunt insertion involves a combination of external skull landmarks and interpretation of computed tomography or magnetic resonance imaging. However, a proportion of shunts are poorly placed when inserted using this technique, and these require early revision, exposing the patients to additional risks and potential morbidity.

Stereotactic shunt placement has evolved over the past two decades, and designs that use infrared optical tracking systems or electromagnetic technology to guide shunt placement are now available. A recent multicentre, prospective, cohort study of navigated shunt placement showed a reduced rate of poor shunt placement compared with standard blind shunt placement, which reduced the early shunt revision rate.[12] Whether this will translate into a reduced need for revision shunt surgery in the longer term remains to be seen.

Implantation of the ventricular catheter using an endoscopic technique has been evaluated in a multicentre randomised trial but was not found to be better than non-endoscopic catheter insertion.[13]

### Antibiotic impregnated shunts

The introduction of antibiotic impregnated shunt systems designed to prevent infection (mainly in the early postoperative period, when most infections occur) may represent an advance, but there is no evidence from randomised controlled trials to support their use in practice. Recent large multicentre cohort studies showed that antibiotic impregnated shunt systems reduced infection rates by 1.5%–4%, a potentially important advance.[10] Further studies are under way, and large scale data collection such as that established by the UK hydrocephalus shunt registry will help to elucidate standards for practice.

### Shunt valves

Shunt valves are mechanical devices that regulate cerebrospinal fluid flow. Several types are used in practice (see appendix on bmj.com). However, randomised clinical studies have yet to confirm the superiority of one valve type over another.[9-15] There is a continued focus on developing valve technology in order to regulate cerebrospinal fluid drainage to as close to normal physiological parameters as possible, including the development of intracranial pressure transducers that are remotely readable and can be programmed to drain according to a patient's needs and symptoms.

### The role of neuroendoscopy

Clearly, avoiding shunt insertion is the best way of reducing shunt associated complications. The advent of endoscopic techniques in other areas of surgery, and high resolution cameras combined with improved light sources, has influenced neurosurgical practice.

Endoscopic third ventriculostomy is an alternative to shunt implantation and may be used as a first line treatment for hydrocephalus. Primary endoscopic third ventriculostomy is effective for obstructive triventricular hydrocephalus, which typically arises secondary to aqueduct stenosis, pineal region lesions, posterior fossa tumours, and arachnoid cysts. The technique may also be performed in selected patients to address shunt malfunction (secondary endoscopic third ventriculostomy). Successful endoscopic third ventriculostomy is defined as symptom relief with shunt independence. A large multicentre prospective study demonstrated close to 70% success rates with primary endoscopic third ventriculostomy.[8]

Failure of endoscopic third ventriculostomy occurs if the stoma is closed by formation of a membrane, which usually occurs within the first three months after the procedure. Patients present with recurrent symptoms of raised intracranial pressure. Endoscopic third ventriculostomy has a lower complication rate than ventriculoperitoneal shunts (2%–6% *v* 13.6%), and infection is much less common.[8] Delayed closure can occur, and patients should be followed up as those with ventriculoperitoneal shunts are—there are published case reports of sudden death in patients who have undergone the procedure without any intraoperative complication.[16]

## Follow-up and routine management

The best follow-up paradigm for patients with treated hydrocephalus is not defined. Baseline imaging at three months and one year after shunt insertion at times when the patient is asymptomatic has been recommended based on long term follow-up studies.[9] True shunt independence is rare, and therefore lifelong neurosurgical follow-up with annual review if the patient is stable is a common practice.

On discharge, patients and family physicians can be provided with information leaflets that contain emergency contact details and a list of signs and symptoms indicating potential shunt malfunction. Patients can also be provided with a "shunt alert card" to carry which states the type of shunt the patient has and the symptoms of malfunction. The card emphasises urgent assessment by a neurosurgical unit if the card holder is showing signs of shunt blockage or infection.

There is no evidence to support routine use of prophylactic antibiotics immediately before dental procedures for patients, nor do they require alternative vaccinations or antibiotic prophylaxis protocols.

Common paediatric conditions such as otitis media, urinary tract infection, constipation, or even appendicitis can mimic shunt malfunction. Symptoms of lethargy, headache, and vomiting can be difficult to differentiate from those of shunt malfunction. A systematic approach is required to obtain the correct diagnosis with a low threshold for suspecting shunt malfunction.

During routine or emergency abdominal surgery, paediatric surgeons need not usually be troubled with the shunt and can push it away from the surgical field. If there is intra-abdominal soiling or contamination we advise externalisation of the distal catheter to avoid infection. If the shunt is obviously infected—that is, signs of raised intracranial pressure, meningism, erythematous tracking along shunt tubing—then the neurosurgical team should be contacted to consider complete removal and insertion of a new external ventricular drain system until the infection is cleared and the shunt can be reimplanted. Elective non-infective laparoscopy performed in patients with a ventriculoperitoneal shunt does not require externalisation of the shunt and is probably safe.[17]

Sport related shunt complications are rare and recommendations by neurosurgeons vary.[18] Some neurosurgeons advocate avoiding contact sports whilst others are less restrictive.

In relation to older patients, the presence of a shunt is not a contraindication to pregnancy.[19] Although symptoms akin to shunt malfunction are common during pregnancy they usually resolve with delivery. Labour and delivery should be guided by obstetric evaluation.

## How do we measure the success of a shunt?

The outcome of hydrocephalus will ultimately be determined by the underlying pathological entity that caused it, the treatment selected accordingly, and avoidance of the complications thereof. Historically, successful hydrocephalus treatment was defined as satisfactory ventricular catheter position on computed tomography, no postoperative complications (such as infection or haemorrhage) requiring further surgery, and relief of symptoms of raised intracranial pressure. Such measures are crude, and an absence of shunt failure does not necessarily equate to success.

In a cohort of UK patients with spina bifida followed from birth, patient reported outcomes indicated a poor prognosis for independent living into adulthood.[20 21] More recently small prospective case series have evaluated the role of neuropsychological testing in patients with spina bifida and showed that reading and writing function remain deficient into adulthood and that memory status is positively correlated with functional independence. These tests are time consuming to administer and interpret but provide an indication of the potential for independent living for patients.

Objective outcome measures have been developed by several researchers, primarily aimed at paediatric cohorts and those with spina bifida living into adulthood. These tools range from prospective lifestyle and health assessment questionnaires (patient reported outcomes) to objective measures of physical, social-emotional, and cognitive function (the hydrocephalus outcome questionnaire). The hydrocephalus outcome questionnaire has been validated and can be administered to children older than 5 years who are shunt dependent to measure development and the effects of episodes of shunt malfunction on neurological development and social integration.[2 22]

## Treating hydrocephalus in the developing world

Most cases of hydrocephalus in the developing world are caused by neonatal infection and are theoretically preventable.[5] Hydrocephalus in children is likely to be under-recognised and undertreated in the developing world. The lack of the appropriate medical infrastructure and trained specialists poses considerable challenges in delivering appropriate care.

Neurosurgery for children in resource poor countries is challenging. Two strategies have been adopted: (*a*) increased availability of low cost shunt hardware and follow-up centres, and (*b*) avoiding shunt dependence by means of endoscopic procedures. (see information resources box). Both strategies need to be considered in combination with improvements in public health and maternal and child healthcare, potentially leading to a reduction in infection and neural tube defects.

A large prospective cohort study in Uganda showed that endoscopic third ventriculostomy in combination with choroid plexus coagulation is a valid surgical treatment for childhood hydrocephalus in developing countries, but with lower success rates than in developed nations.[23] Three factors seem to account for the higher failure rate in Uganda: patient prognostic factors, technical variation in the procedure, and more cases aborted intraoperatively. When these factors are taken into consideration, the outcome of endoscopic third ventriculostomy in children in Uganda has been shown to be no different from that of children in developed nations.[23]

## Does fetal screening aid management and prevention of hydrocephalus?

With the advent of prenatal ultrasonography, diagnosis of hydrocephalus in utero has led to attempts at intrauterine fetal surgery. The presumption has been that earlier repair in utero would provide superior outcomes compared with postnatal surgery. However, postnatal treatment is much simpler and is not traumatic to the mother. Procedures performed in utero, such as ventriculoamniotic shunts and serial cephalocenteses, were associated with high morbidity and mortality, and thus in utero treatment of hydrocephalus has largely been abandoned.

However, interest in fetal therapy for hydrocephalus associated with spinal dysraphism has increased recently.[24] A prospective, randomised, multicentre trial compared prenatal repair of myelomeningocele with standard postnatal repair and reported that prenatal surgery reduced the need for shunting and improved motor outcomes at 30 months but was associated with serious maternal and fetal risks.[25] Caution is required in interpreting the potential implications of this study. Although this study suggests potential benefits from intervening early, less traumatic and invasive techniques need to be developed before antenatal surgical repair can be advocated. The long term outcomes associated with intrauterine repair of myelomeningocele remain unclear, and for now patient counselling should reflect that.

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**Ongoing research**

International Infant Hydrocephalus Study—www.iihsstudy.org
International multicentre trial randomising infants with defined aqueductal stenosis and triventricular hydrocephalus to receive either endoscopic third ventriculostomy or CSF shunting. Long term outcome analysis will cover shunt dependence and a more comprehensive analysis of treatment effect and patient outcome (including various aspects of quality of life such as hospitalisation or other sickness time and neurodevelopmental evaluations over the course of 5–7 years). This study was initiated under the aegis of the International Study Group for Neuro-endoscopy and the International Society for Paediatric Neurosurgery.

British Antibiotic and Silver Impregnated Catheters for ventriculoperitoneal Shunts multi-centre randomised controlled trial (BASICS trial)
Funding (£1.72m) is being sought from the National Institute for Health Research for this trial to compare standard silicone, antibiotic impregnated, and silver impregnated catheters in the incidence of shunt infection. The trial will be led by the departments of neurosurgery in Alder Hey Children's Hospital and the Walton Centre for Neurology and Neurosurgery in Liverpool, the lead investigators are Conor Mallucci and Michael Jenkinson.

**Additional educational resources**

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**Information resources for patients and non-specialists**

International Federation for Spina Bifida and Hydrocephalus—www.ifglobal.org/en
Worldwide umbrella organisation for a network of organisations that support people with hydrocephalus and spina bifida

International Program to Advance the Treatment of Hydrocephalus (iPATH)—www.ifglobal.org/ipath/
Program trains and equips neurosurgeons responsible for treating hydrocephalus in developing countries

Society for Research into Hydrocephalus and Spina Bifida (SRHSB)—www.srhsb.org

Association for Spina Bifida and Hydrocephalus (ASBAH)—www.asbah.org
Association of patients with spina bifida and hydrocephalus in the United Kingdom

Hydrocephalus Association—www.hydroassoc.org/
Association of patients with hydrocephalus in the United States

The Hydrocephalus Foundation—www.hydrocephalus.org
A registered US non-profit organisation providing support, educational resources, and networking opportunities for patients and families affected by hydrocephalus

Arbeitsgemeinschaft Spina Bifida und Hydrocephalus (AsbH)—www.asbh.de
Association of patients with spina bifida and hydrocephalus in Germany

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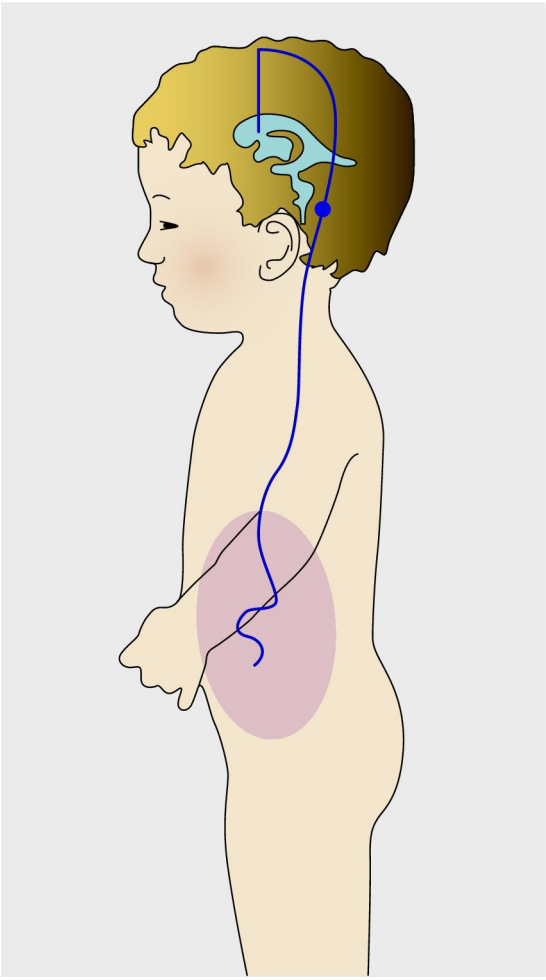
## Figure



Pictorial representation of a shunt system. A shunt consists of three components—a ventricular catheter, a valve, and a distal tube. Many shunts will have a cranial reservoir for “shunt taps,” where a surgeon may sample cerebrospinal fluid and assess patency and intracranial pressure.