Hydrocephalus

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Kai Arnell and Tomas Wester

Hydrocephalus is defined as an excessive amount of cerebrospinal fluid (CSF) under increased pressure in the ventricular system. The incidence is 0.4–2.5 per 1000 live births. CSF is mainly produced by the choroid plexus of the ventricular system. It is circulating through the foramen of Monro to the third ventricle and then through the aqueduct of Sylvius to the fourth ventricle. The CSF enters the subarachnoid space through the foramina Luschka and Magendie. It is absorbed into the blood stream by the arachnoid villi on the surface of the brain. The CSF production is age-dependent. Neonates produce approximately 25 mL/24 h, and adults produce approximately 700 mL/24 h. The intraventricular pressure at rest varies between 0 and 10 cm H_2O.

K. Arnell
Department of Women's and Children's Health, Uppsala University, Uppsala, Sweden
e-mail: Kai.Arnell@surgsci.uu.se

T. Wester (✉)
Unit of Pediatric Surgery, Karolinska University Hospital, Karolinska Institutet, Stockholm, Sweden
e-mail: tomas.wester@karolinska.se

49.1 Causes

The cause of hydrocephalus can be obstruction to the flow of CSF (noncommunicating hydrocephalus), overproduction of CSF, or failure of its re-absorption (communicating hydrocephalus). Pathological processes that cause hydrocephalus are congenital malformations, neoplasms, bacterial meningitis, prenatal infections (toxoplasmosis, listeriosis, or cytomegalovirus infection), subarachnoid or intraventricular haemorrhage, and overproduction of CSF by choroid plexus papillomas. The three most important malformations that cause congenital hydrocephalus due to obstruction of the CSF flow are aqueductal stenosis, Arnold-Chiari II malformation, and Dandy-Walker malformation. Aqueductal stenosis accounts for about 15% of cases of hydrocephalus and occurs in several anatomical forms. Myelomeningocele, when combined with Arnold-Chiari II malformation, is associated with hydrocephalus in about 90% of cases. This malformation leads to CSF flow obstruction due to herniation of the medulla oblongata, cerebellar tonsils, and vermis through the enlarged foramen magnum and compression of the fourth ventricle. The Dandy-Walker malformation is a developmental defect of the foramina of Luschka and Magendie, with absence of the cerebellar vermis, a distended fourth ventricle, and an enlarged posterior fossa. Hydrocephalus can be an isolated finding but it can also be part of various malformation syndromes, such as Meckel-Gruber, Walker-Warburg, and X-linked hydrocephalus.

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49.2 Clinical Features

An important clinical feature of hydrocephalus in neonates and infants is enlargement of the head circumference, which can easily be overlooked if the head circumference is not measured repeatedly. The shape of the head is often abnormal. The anterior fontanel is large and bulging. Cranial suture separation and distended scalp veins are common. The" sunset sign" may be present. Vomiting, behavioural changes, drowsiness, and headache are common symptoms in infants and young children with hydrocephalus. Other symptoms are failure to thrive, irritability, delayed motor and psychosocial development, and mental retardation.

49.3 Diagnosis and Monitoring

Ultrasound, CT scans, and MRI distinguish hydrocephalus from other conditions. Hydrocephalus is frequently diagnosed with antenatal ultrasound. Ultrasound is also the method of choice for serial examinations of premature babies to detect intracranial haemorrhages and secondary hydrocephalus. CT and particularly MRI are useful to show the detailed anatomy and reveal the cause of hydrocephalus as well as associated abnormalities or neoplasms. The investigation of the patient with hydrocephalus also includes a TORCH screen and chromosomal analysis if a prenatal infection or a chromosomal abnormality is suspected. The patient should undergo an eye examination, and associated anomalies such as a cardiovascular malformation should be excluded.

Mild hydrocephalus, caused by intracranial haemorrhage, is sometimes monitored by repeated measurement of the skull circumference, ultrasound scans, and clinical signs of progressive hydrocephalus; it may finally resolve without treatment. On the other hand, in cases with progressive ventricular dilatation, rapidly increasing skull circumference, and clinical features of increased intracranial pressure, the condition requires treatment by surgical diversion of CSF.

49.4 Surgical Treatment

Ventriculoperitoneal shunts are the primary option for surgical diversion of CSF in infants and children. Ventriculoatrial shunts are used only under certain circumstances because of the risk of serious sequelae such as thrombosis, pulmonary embolism, and septicaemia. Many different shunt systems are available. Systems with a programmable valve are preferred.

Endoscopic third ventriculotomy has become an option particularly for treatment of aqueductal stenosis. The technique eliminates the need for a shunt and thereby the complications of shunting. There appear to be a correlation between success rate and age, with poorer outcome in infants less than 6 months of age, although conflicting data have been reported.

49.4.1 Preoperative Routines

The preoperative routines are important to minimize the risk of *Staphylococcus epidermidis* shunt infections. The patient should be carefully prepared before the operation. Cloxacillin is administered as prophylaxis at induction of general anaesthesia.

The child is placed in a supine position on the operating table with the head turned to the left if the shunt is to be placed on the right side (Fig. 49.1). The right shoulder and neck are elevated to stretch the skin and avoid creases that make the subcutaneous tunneling of the abdominal catheter difficult. The scalp should be shaved and well prepared. The placement of the incision depends upon whether a frontal or a parieto-occipital approach is selected for ventriculoperitoneal shunting.

Fig. 49.1 Patient positioning for placement of a ventriculoperitoneal shunt

49.4.2 Frontal Approach

A frontal approach has several advantages. Less brain is traversed than with the posterior approach, and the distance to the ventricle is shorter, which makes cannulation of very small ventricles easier. The disadvantage of a frontal approach is that a longer subgaleal tunnel is required and an extra occipital incision is needed for tunneling. The sagittal and coronal sutures are marked, as well as the place of entrance of the ventricular catheter, which should be located 2 cm from the midline and 2 cm anterior to the coronal sutures. In neonates and infants with massive hydrocephalus, the catheter will be placed in the fontanel. In most cases, the ventricular catheter will be inserted at the edge of the fontanel or through the frontal bone (Fig. 49.2). It is important to avoid having any part of the shunt system lying under the wound. Prepare the child and drape it in a sterile fashion. The entire operation field should be covered with a plastic sheet to avoid contact between the shunt system and the skin. The skin is very thin in neonates, particularly in preterm babies, and the blood supply is vulnerable, so infiltration with xylocaine and adrenaline is not used. Bipolar diathermy is preferred for haemostasis. The flap is carefully dissected from the underlying galea and a subcutaneous stay suture is put in the flap. When entering at the edge of the fontanel, the galea is opened and a circular hole is nipped in the bone edge. In older children with bone under the flap, the periosteum is removed. A burr hole is made. The bone is very thin in neonates and infants, and it is important to be careful. The dura is coagulated using a fine-pointed forceps, and a small opening is made. Ventricular catheters may be either straight or prebent, but prebent catheters are recommended for infants and children. From a CT scan, the optimal length of the ventricular catheter can be estimated. The ventricular catheter is introduced through the small opening to get a snug fit and avoid leakage of CSF. The direction of the ventricular catheter is chosen so that the tip will be located in the anterior horn of the right ventricle. The catheter is passed through the brain perpendicular to the skull, aiming at the inner canthus of the ipsilateral eye.

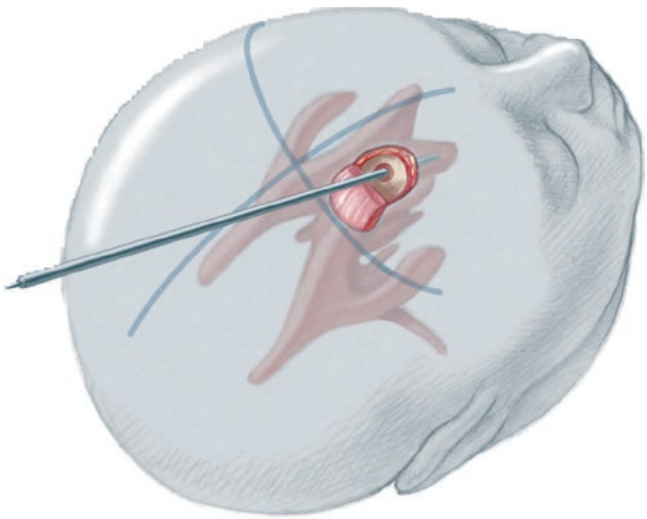

Fig. 49.2 Location of the ventricular catheter using a frontal approach

49.4.3 Parieto-Occipital Approach

In a parieto-occipital approach, the scalp incision is semicircular, lying well behind the posterior parietal eminence. Care must be taken to avoid having any part of the shunt system located under the wound, which would predispose for breakdown of the wound and infection. The burr hole is placed 3 cm above and behind the ear. The catheter is passed parallel to the sagittal suture, aiming at the glabella (Fig. 49.3). The length of the ventricular catheter should allow the tip to extend 1 cm anterior to the coronal suture in order for the catheter to be placed in the anterior horn.

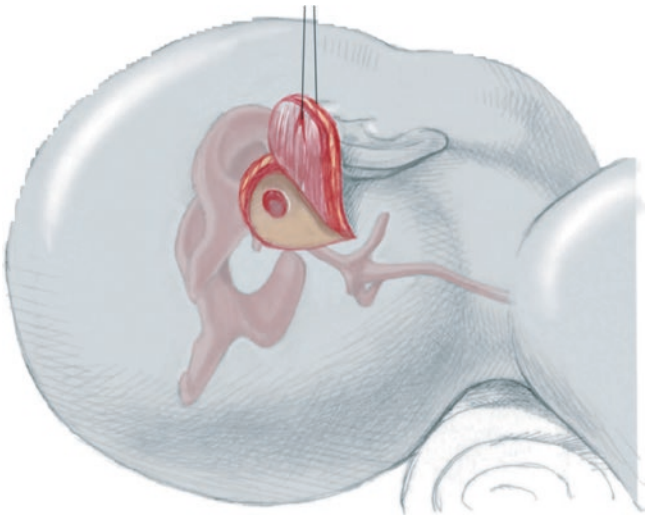

Fig. 49.3 Location of the ventricular catheter using a parieto-occipital approach

49.4.4 Completion of Shunt Placement

The abdominal incision is made 2 cm below the costal margin, over the rectus abdominis muscle (Fig. 49.4). The muscle fibres are split longitudinally and the peritoneal cavity is opened.

The tunneling device is pushed from the scalp incision subcutaneously to the abdominal incision (Fig. 49.5). The passage over the clavicle is controlled to avoid skin contusion or perforation. The stylet is withdrawn and the peritoneal end of the catheter is pushed through the tunneling device.

The ventricular catheter is introduced into the lateral ventricle in the direction of the glabella and the stylet is withdrawn. The valve is pressed to test the function. CSF should be seen dripping distally. The scalp incision is closed with absorbable subcuticular sutures. The system is tested once again before the abdominal catheter is inserted through the small opening into the peritoneal cavity (Fig. 49.6). At least 35–40 cm of the catheter can be placed in the abdomen of a full-term baby. The peritoneum is closed in a water-tight manner, as are the layers of the abdominal wall. Absorbable sutures are used for the skin closure.

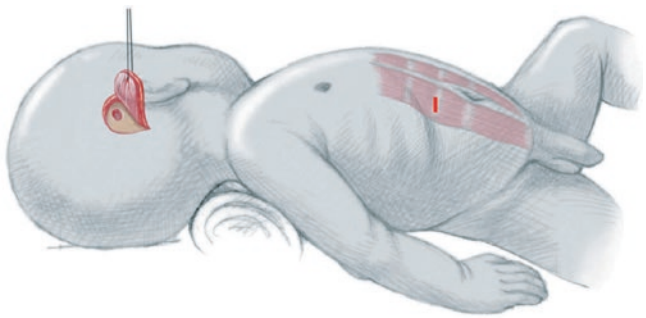

Fig. 49.4 The abdominal incision

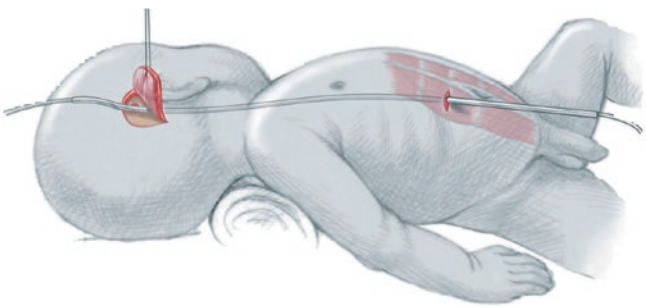

Fig. 49.5 Use of the tunneling device

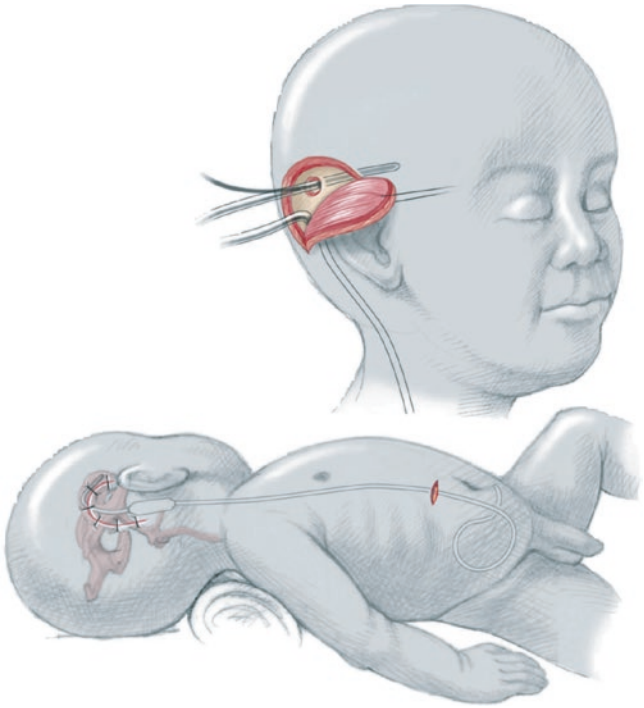

Fig. 49.6 Insertion of the catheter into the peritoneal cavity

49.5 Complications

Shunt complications are relatively frequent, owing to infections, catheter obstruction, improper placement of the catheters, and short or disconnected catheters. Despite careful preparation and antibiotic prophylaxis, there is a risk of perioperative infections with *Staphylococcus epidermidis*, *Staphylococcus aureus*, or *Proprionebacterium* species. Neonates and infants are more sensitive to infections and can also have haematogenous infections with origin in the urinary tract or respiratory tract. The incidence of infections has been reported to range from 10% to 15%. The infections are treated by removal of the infected shunt system, external ventricular drainage, and use of intravenous and intraventricular antibiotics before insertion of a new system.

Technical advances have led to the development of shunt systems that are much more sophisticated than those initially produced 50 years ago. The programmable valves can be noninvasively adjusted to a suitable pressure, giving the brain optimal conditions for development. In spite of the introduction of programmable valves, however, problems due to overdrainage are encountered, resulting in slit ventricles. An antisiphon device can be inserted between the valve and the abdominal catheter to reduce this problem. There are also valves available that include an antisiphon mechanism.

The incidence of reoperations due to shunt malfunction used to be very high. Today, the incidence has decreased because of the awareness of strict preoperative and perioperative routines, antibiotic prophylaxis, and the use of longer catheters.

49.6 Outcome

The long-term outcome of children with hydrocephalus depends on many factors, such as the severity and duration of the ventricular dilatation, the aetiology of the hydrocephalus, associated intracranial malformations, and whether severe infections have occurred. It is important to follow these patients and monitor their neurologic and psychosocial function as well as their growth and onset of puberty.

Suggested Reading

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