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Hydrocephalus and Shunts

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Summary

Hydrocephalus occurs as a result of impaired circulation or absorption of cerebrospinal fluid. Hydrocephalus is not a single disease entity but rather the end result of a wide range of congenital or acquired pathological processes. It is important that this etiological heterogeneity is appreciated when evaluating and treating hydrocephalus. The clinical presentation, the rate of evolution and the long-term prognosis of hydrocephalus will vary according to the age of onset and the nature of the underlying cause. It is important that these issues be borne in mind so that the most appropriate management plan can be formulated. This chapter aims to provide an overview of hydrocephalus, its various causes and the use and complications of the shunt devices to treat it. The role of endoscopy in the treatment of hydrocephalus is discussed elsewhere.

Introduction

Numerous definitions of hydrocephalus have been advanced; common to these is an underlying imbalance between the production of CSF and its absorption. Although overproduction of CSF resulting in hydrocephalus is well recognized in association with choroid plexus papillomas, these are rare tumors and, in practice, hydrocephalus is most commonly the result of some impairment of the circulation or absorption of CSF.

The accumulation of fluid in various intracranial compartments was recognized by Hippocrates (BC 460–377) and Claudius Galen (130–200 AD); however, the first morphological description of hydrocephalus, recognizing the ventricular enlargement and brain destruction is attributed to Andreus Vesalius (1514–64). More complete descriptions subsequently appeared in the works of Jean Louis Petit (1664–1750), in Robert Whytt's "Observations on dropsy of the brain" and in the writings of Giovanni Morgani (1682–1771), who described the clinical features of bulging fontanelle and sutural widening in childhood hydrocephalus. The anatomy of the ventricular system and the CSF pathways began to be understood during this period, facilitated in particular by the studies of Thomas Willis (1621–75). It was he who first proposed the choroid plexus as the site of CSF production and who introduced the concept of absorption into the venous system via what he termed the meningeal "glandules", presumed to be the arachnoid granulations. Franciscus Sylvius (1614–72), Alexander Monro (1733–1817) and Francois Magendie (1783–1855) also made important anatomical contributions but it was not until the more physiological investigations of Key and Retzius (1876) that the modern concept of CSF circulation was established.

During this period, crude attempts at treatment of hydrocephalus by means of repeated

cerebral puncture were employed but at the cost of great morbidity and mortality. Dandy and Blackfan further contributed, developing experimental models of hydrocephalus, and established a means of classification of hydrocephalus, differentiating between the non-communicating (obstructive) and communicating forms and proposing possible treatment strategies. The treatment options they advanced included extirpation of the choroid plexus, removing obstructive pathologies if these could be identified or the creation of conduits to drain the CSF from the intracranial compartment, either internally by the sub-frontal and subtemporal routes or extracranially. Early means of CSF diversion included nephrectomy followed by the plumbing of the ureter into the spinal theca, thus draining CSF to the bladder. Other surgical approaches devised at this time included Torkildsen's procedure of draining the lateral ventricle into the cisterna magna, ventriculocisternostomy in cases of aqueductal obstruction and the first descriptions of endoscopic third ventriculostomy by Mixter – a procedure that has recently undergone a renaissance. Morbidity and failure rates, however, remained high. Removing choroid plexus failed to account for the extra-choroidal production of CSF and early artificial conduits were prone to mechanical malfunction. In the 1950s, synthetic, biologically tolerated polymers, in particular silicone elastomer, became available and thus heralded the onset of the shunt era of hydrocephalus management.

Cerebrospinal Fluid Production and Absorption

Although a small proportion of CSF may be produced from the ependyma and brain parenchyma, the predominant site of CSF production is the choroid plexus, contributing 70–80% of the daily volume. Production occurs by a combination of filtration across the capillary endothelium and active secretion of sodium by the choroidal epithelium [1]. Cerebrospinal fluid production does appear to be reduced in the presence of elevated intracranial pressure and reduced cerebral perfusion pressure; however, the effect is small and production is largely independent of pressure under physiological conditions. By contrast, CSF absorption shows a linear relationship to ICP.

The mechanisms of absorption of CSF have been extensively investigated. Direct absorption by the brain parenchyma, the choroid plexus itself and by lymphatic channels in the region of the cribiform plate have been postulated. It is, however, via the arachnoid villi and granulations that most absorption is presumed to take place. Arachnoid villi are herniations of arachnoidal tissue into the dural venous sinuses. For a long time, two models of CSF absorption were debated. The "closed" concept held that the villi were blind diverticulae and absorption of CSF occurred by a process of seepage across their endothelial covering. The alternative "open" model inferred the presence of channels across the villus, opening and closing in a valve-like manner, permitting the unidirectional flow of CSF. Tripathi and Tripathi [2] attempted to reconcile these opposing views and proposed a transmembrane transport mechanism consisting of vacuoles ferrying CSF across the covering endothelial layer. Interestingly, recent work has focused on the role of the CNS microcirculation in the absorption of CSF – a concept that would have major implications for our understanding of the pathogenesis of hydrocephalus [3]. Whilst the precise mechanism of CSF absorption and the relative contributions of the various absorptive pathways currently remain unclear, our classification and management of hydrocephalus is based on traditional concepts of CSF circulation.

Cerebrospinal Fluid Volume and Composition

Cerebrospinal fluid is produced at a rate of approximately 500 ml per day (0.35 ml/min). The total volume of the CSF varies with age and in the adult is 100–150 ml, of which 15–25 ml is contained within the ventricular system.

Etiology and Pathophysiology of Hydrocephalus

An obstruction at any point along the CSF pathway may result in hydrocephalus. It is usual to distinguish between etiologies that lie within the ventricular system or at the fourth ventricular outflow – obstructive hydrocephalus (non-communicating) – and those that impair circulation through the subarachnoid spaces or

absorption into the venous system – communicating hydrocephalus. Where the etiology is known, it is possible to further divide hydrocephalus into congenital and acquired forms. Examples of the various types of hydrocephalus are shown in Table 24.1, which includes both adult and pediatric hydrocephalus.

Estimating the true incidence of hydrocephalus is complicated by the etiological heterogeneity, the lack of a precise definition of hydrocephalus and also by institutional factors, such as referral patterns and treatment protocols.

Post-hemorrhagic Hydrocephalus

During cerebral development, the germinal matrix is the site of intense cellular proliferation and the source of both the neuronal and glial elements of the cerebral hemispheres. Situated in the periventricular region, between the thalamus and caudate nucleus, the germinal matrix is a large structure in the early developing brain. From the end of the second trimester, it begins to involute – a process that is almost complete by about 34 weeks; therefore, germinal matrix hemorrhage is unusual after this time. The blood vessels of the germinal matrix are irregular structures that have an immature connective tissue architecture; they also lack the auto-regulatory capacity observed in mature cerebral vasculature. Both these factors are thought to contribute to the propensity for vessel rupture in the premature infant.

The incidence of germinal matrix hemorrhage shows an inverse relationship with gestational age. Hemorrhage is detected in 40–45% of premature infants with birth weight less than 1500 g. In neonatal and pediatric practice, approximately 20% of infants who suffer an intraventricular hemorrhage will go on to require a shunt. Clearly, in a number of cases, the condition resolves following conservative management. The population presenting to the neurosurgeon is selection biased and hence the neurosurgical literature quotes greater rates of shunt placement. The risk of progressive hydrocephalus developing is proportional to the grade of hemorrhage.

The majority of intraventricular hemorrhages occur within the first few days after birth. In addition to prematurity, vigorous resuscitation, respiratory distress syndrome, pneumothoraces and seizures are among the factors associated with an increased risk of development of hemorrhage, with pronounced fluctuations in cerebral blood flow being the possible final pathway. Although hemorrhages may occur in the absence of clinical signs, the more extensive lesions may be associated with seizures, bradycardias and apnoeic spells.

Germinal matrix hemorrhage is readily detected on cerebral ultrasound examination and is graded according to the site of the hematoma and the effect upon the ventricular size [4].

Once germinal matrix hemorrhage has been diagnosed, follow-up by serial ultrasound examinations is required, particularly in the

Table 24.1. Types of hydrocephalus.

Obstructive hydrocephalus	Communicating hydrocephalus
Congenital	**Congenital**
Aqueduct stenosis	Arnold Chiari malformation (type II, less commonly type I)
Dandy Walker cyst	Encephaloceles
Benign intracranial cysts (e.g. arachnoid cysts)	Skull base deformity
Vascular malformations (e.g. vein of Galen aneurysms)	
Acquired	**Acquired**
Tumours (e.g. third ventricle, pineal region, posterior fossa)	Infection (intrauterine, e.g. CMV, toxoplasma, post-bacterial meningitis)
Other mass lesions (e.g. giant aneurysms, abscesses)	Haemorrhage (IVH of infancy, sub-arachnoid haemorrhage)
Ventricular scarring	Venous hypertension (e.g. venous sinus thrombosis, arterio–venous shunts)
	Meningeal carcinomatosis
	Oversecretion of CSF (choroid plexus papillomas)

presence of intraventricular extension or ventricular enlargement. The presence of blood and its breakdown products within the CSF may lead to an ependymal reaction, with blockage at the narrower points of the CSF circulation, such as the aqueduct or the outlet foramina of the fourth ventricle with the subsequent development of an obstructive (non-communicating) hydrocephalus. More commonly, the blood products cause an obstruction at the level of the subarachnoid space and arachnoid villi, leading to a communicating hydrocephalus.

Increasing head circumference and progressive ventricular enlargement indicate the need for intervention. A number of therapeutic options are available. The presence of heavy blood staining or excessive amounts of proteinaceous materials and cellular debris in the CSF precludes the early insertion of a shunt. Moreover, in the premature, low-birth-weight infant, the high risk of shunt infection is an additional concern (see above). Temporizing measures may include serial lumbar punctures or ventricular taps via the fontanelle. If repeated ventricular taps are necessary, there is a risk of causing damage to the cerebral mantle, producing areas of porencephaly. This risk can possibly be minimized by the placement of a ventricular catheter and subcutaneous reservoir. CSF can then be aspirated from the reservoir, thus avoiding repeated cerebral puncture. Recent review of the literature has not found evidence to support the use of repeated lumbar puncture or ventricular taps as a means of either reducing death or disability or the need for shunt placement [4a].

Medical measures to control the ventricular dilation are also employed. Acetazolamide, alone or in combination with furosemide, has been used and some have even suggested that such a regime may avoid the need for subsequent shunt placement. Recent evidence, however, has questioned the clinical efficacy of such regimes [5].

Lately, it has been suggested that intraventricular fibrinolytic therapy, instituted soon after the hemorrhage is diagnosed, may prevent the chemical arachnoiditis that develops in response to intraventricular hemorrhage and thus reduce the number of these infants requiring shunt insertion.

Whatever method of treatment employed, it is important that progress is monitored by means of regular clinical evaluation, head circumference measurements and ultrasonography. If progressive hydrocephalus is present, then once the CSF is clear of blood products, a shunt procedure can be performed.

Hydrocephalus and Myelomeningocele

Hydrocephalus complicates open spina bifida in 85–90% of patients. The key to understanding its etiology in this context is the Chiari (type II) malformation. In the fetus with open spina bifida, the meningocele sac acts as a CSF sump. The constant venting of CSF from the developing brain and spinal cord, beyond the time when the caudal neuropore should have closed, removes the distending force that is normally present within the cranial neurocele. This results in a constellation of features, termed the Chiari II malformation [6]. These include disorganization of brainstem topography, a small posterior fossa and, as cerebellar growth exceeds the confines of the small posterior fossa, herniation of the cerebellum through the foramen magnum and up through the incisura. As a consequence, the normal CSF pathways may be compromised at a number of sites, including the cerebral aqueduct, the fourth ventricular outlet and at the perimesencephalic region, resulting in hydrocephalus.

Hydrocephalus may only become apparent following closure of the myelomeningocele. In addition to the usual presenting features of infantile hydrocephalus, features unique to the myelomeningocele patient are bulging of the back wound, occasionally with CSF leakage and signs of brainstem compression due to the Chiari malformation. These latter signs can include stridor, lower cranial nerve palsies and upper-limb weakness and should prompt the search for progressive hydrocephalus. As well as being presenting signs of hydrocephalus in these infants, these signs may similarly herald a shunt malfunction in the older child. It is important to be aware of these atypical modes of presentation of raised intracranial pressure in the myelomeningocele population.

It is doubtful whether any child with myelomeningocele and shunted hydrocephalus can ever be considered to be truly shunt independent. Even in instances where it appears that the shunt is not being used, there are reports of sudden cardio–respiratory arrest attributable to the combination of raised ICP and the Chiari malformation.

Aqueduct Stenosis

As a result of growth of the tectum and tegmentum, the lumen of the neural tube in the region of the mesencephalon becomes narrowed to form the aqueduct of Sylvius. Because of its small caliber, this area of the CSF pathway is vulnerable to obstruction by a number of congenital and acquired pathologies.

Aqueductal stenosis is responsible for approximately 10% of cases of hydrocephalus in childhood. Presentation may, however, occur at any time from birth to adulthood. In congenital forms of aqueduct "stenosis", the aqueduct, rather than being stenosed, is branched or forked into two or more channels. In some instances, the tectum is also deformed and it has been postulated that, here, the primary pathology is a communicating hydrocephalus in which external pressure on the mesencephalic structures has led to obliteration of the aqueduct secondarily.

Scarring or gliosis in the aqueduct following infection or hemorrhage may lead to an acquired aqueduct stenosis. Tumors arising in adjacent structures, such as the tectal plate, the rostral fourth ventricle, posterior thalamus or pineal region, may not be evident on CT but can similarly result in a picture of aqueduct stenosis. MRI is necessary in such cases.

Imaging reveals a tri-ventricular hydrocephalus, with a small or normal-sized fourth ventricle. In addition, MRI will often reveal a trumpeting of the proximal aqueduct and, moreover, will readily identify associated neoplastic lesions that may not be seen on CT scanning. It is for this reason that MRI scanning should be performed routinely in cases of aqueduct stenosis.

Dandy Walker Syndrome

This syndrome comprises agenesis of the cerebellar vermis with cystic dilatation of the fourth ventricle, enlargement of the posterior fossa and hydrocephalus. The hydrocephalus is often absent at the time of birth but is present in 75% of cases by the age of 3 months. Additional brain malformations are present in over half of the cases. Neurodevelopmental delay is reported in up to 70% of cases.

Some controversy exists among neurosurgeons as to the best surgical treatment of this condition. The principal issue is regarding the placement of the proximal catheter: whether this should be placed in the lateral ventricle, the posterior fossa cyst or indeed whether both compartments should be shunted simultaneously. A further option is to shunt the infratentorial compartment in the first instance and then to treat the supratentorial hydrocephalus endoscopically if the ventricles fail to decompress.

Obstructive Hydrocephalus Due to Tumors

Midline tumors – particularly those of the pineal gland and posterior fossa – commonly result in obstructive hydrocephalus and, indeed, this is one of the principal ways in which these conditions present, especially in children. Although preliminary shunting of the hydrocephalus has been advocated, in the majority of cases, the hydrocephalus will resolve following removal of the obstructive pathology. In their series of posterior fossa tumors with hydrocephalus, Kumar et al. reported that a shunt was required in 18.9% of cases [7]. Young age at presentation, incomplete tumor removal and malignant midline tumors were factors increasing the likelihood of shunt requirement.

In situations where more urgent control of the hydrocephalus is required, insertion of an external ventricular drain, a subcutaneous reservoir or an endoscopic third ventriculostomy can be performed, pending definitive surgical treatment of the tumor.

Post-meningitic Hydrocephalus

Hydrocephalus may occur as the result of a range of infectious or inflammatory processes. The effects of chronic inflammation – organization of the inflammatory exudate with scarring or gliosis – can produce obstruction to CSF flow, both within the ventricular system and in the basal cisterns and cortical subarachnoid spaces. Bacterial, parasitic and granulomatous infections are much more likely to lead to hydrocephalus than viral infections. Ventricular enlargement rather than hydrocephalus may occur due to an ex-vacuo phenomenon. This is a result of the severe white matter damage or encephalomalacia that is commonly seen in the

aftermath of bacterial meningitis. The risk of hydrocephalus is greater when treatment has been delayed or is sub-therapeutic.

Purulent infections not uncommonly result in compartmentalization or trapping of parts of the ventricular system (Fig. 24.1). This can make the surgical management of these cases particularly problematic, with the need for multiple ventricular catheters. In some circumstances, neuroendoscopic techniques can be used to fenestrate gliotic septae in an attempt to unify the compartments and thus simplify the shunt configuration.

Hydrocephalus and Venous Hypertension

The role of raised venous pressure as an etiological factor in the development of hydrocephalus has been highlighted in a number of clinical conditions [8], Achondroplasia and syndromic craniosynostosis (e.g. Crouzon and Cloverleaf skull) being the most commonly referred to. The mechanism is thought to be deformity at the skull base, resulting in narrowing of the jugular foramina and impairment of intracranial venous drainage [9]. The raised pressure within the cranial venous sinuses reduces the pressure gradient across the arachnoid villi and granulations impairing the absorption of CSF. In the infant with open sutures and immature myelination, progressive dilatation of the ventricles occurs in these circumstances. By contrast, once the sutures have fused and the cranium is less distensible, a picture more akin to pseudotumor cerebri results.

Hydrocephalus commonly accompanies vein of Galen aneurysms. Again, the mechanism is in part the result raised of venous pressure due to arteriovenous shunting. In this instance, shunting the ventricles may be both harmful and unnecessary, since once the primary anomaly has been treated, the hydrocephalus may come under control.

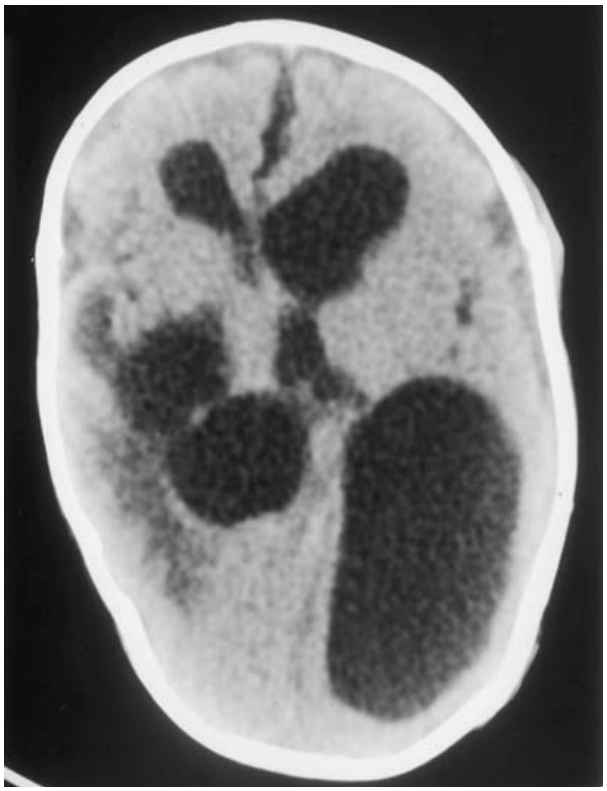

Fig. 24.1. Loculation of the lateral ventricles following bacterial meningitis.

Hydrocephalus Following Subarachnoid Hemorrhage

Hydrocephalus is seen in 10–15% of patients suffering aneurysmal subarachnoid hemorrhage. The incidence is increased where there is intraventricular hemorrhage in addition. Hydrocephalus may appear soon after the initial ictus but must also be considered as a cause of delayed recovery or neurological deterioration later in the illness. Symptomatic hydrocephalus requires intervention, which, in the first instance, will most commonly be by means of external ventricular drainage. There is some concern that the early re-bleeding rate is higher in those patients requiring drainage.

Normal Pressure Hydrocephalus

This condition, most commonly seen in adulthood, is characterized by a clinical picture of gait deterioration, dementia and urinary incontinence in the context of enlarged ventricles on neuroimaging, but relatively normal intracranial pressures. Although in some cases there is a history of subarachnoid hemorrhage, intracranial infection or head trauma, in approximately 60% of cases the etiology remains unknown. The differential diagnosis is wide and the identification of patients that will

benefit from shunting is known to be difficult. A number of investigations have been advocated to aid in the selection of patients suitable for shunt treatment. These include isotope cisternography to assess abnormalities of the CSF pathways, infusion tests to detect increased resistance of CSF outflow and ICP monitoring looking at the frequency of Lundberg B waves which seem to be increased in this condition. Clinical features such as short history, predominance of gait disturbance as a presenting feature and a known etiology are though to portend a favorable response to treatment.

Idiopathic Intracranial Hypertension

This condition has previously been termed benign intracranial hypertension (BIH) or pseudotumor cerebri. The intracranial pressure is raised; however, there are no localising signs, no alteration in level of consciousness, the cerebrospinal fluid composition is normal and there is no evidence of hydrocephalus or other cause of raised ICP on neuroimaging. It is thus a diagnosis of exclusion. Chronic meningitis, venous sinus thrombosis and indeed spinal cord tumors may produce a similar clinical picture. An association with various drugs, metabolic and endocrinological disorders is recognized and this needs to be considered during the clinical evaluation of these patients.

Headache and visual disturbance are the usual clinical features. Papilloedema, optic atrophy and reduced visual fields are often present and permanent visual impairment can result. Close ophthalmological surveillance of these patients is mandatory.

Preservation of vision is the main aim of treatment. Treatment options include medical therapies such as acetazolamide. Drainage of CSF, either intermittently with serial lumbar punctures or continuously by placement of a lumbo–peritoneal shunt (or ventriculo–peritoneal shunt if ventricular size permits), may be required where medical therapy fails. Optic nerve fenestration may lead to visual improvement and may be a useful adjunct in the treatment of this condition. The long-term prognosis is generally good, with spontaneous resolution in a number of cases, though the visual loss may persist.

Arrested Hydrocephalus

Occasionally, hydrocephalus may evolve into a chronic state in which ventricular enlargement persists, yet CSF pressure returns to normal. This situation might be more accurately termed "compensated hydrocephalus". In children who have truly compensated, insertion of a shunt may be detrimental, leading to symptoms of low pressure or chronic subdural formation in addition to the usual complications of shunt devices. On the other hand, it is important not to miss the child whose neurodevelopmental progress is being hindered by the presence of hydrocephalus. If a decision is made not to embark upon surgical treatment, close monitoring is required in order to be sure that there is no disproportionate increase in head growth or progression of ventricular size and to ensure that development proceeds at a satisfactory pace.

Hydrocephalus Versus Ventriculomegaly

The term "hydrocephalus" should be used as a clinical term to imply an active process in which either the circulation or absorption of CSF is impaired (or, more rarely, the production of CSF is excessive), leading to an elevation of intracranial pressure. Increased size of the cerebral ventricles on imaging is more appropriately termed "ventriculomegaly" and does not necessarily equate to hydrocephalus requiring treatment. It is important to emphasize that whilst ventriculomegaly can be readily diagnosed by various imaging modalities, such findings must be interpreted in the context of the clinical symptoms and signs in order to permit a firm diagnosis of active hydrocephalus. In some cases, it is necessary to proceed to invasive measurement of intracranial pressure in order to make this distinction [10].

There are other explanations for ventricular enlargement, for example the ex-vacuo effect of cerebral atrophy associated with aging, head trauma or severe infection. In the child, ventricular enlargement may result from hypoxic ischemic insults. Moreover, in the aftermath of various therapeutic interventions such as steroid therapy, radiation therapy and chemotherapy, ventricular enlargement may

also be seen. In such cases of white matter loss, there is usually concomitant enlargement of the cortical sub-arachnoid spaces and basal cisterns. The periventricular lucency seen in ventricles under pressure is not usually a feature in these circumstances.

A number of structural abnormalities of the brain, such as colpocephaly, holoprosencephaly and agenesis of the corpus callosum, may also be associated with ventricular enlargement and yet do not necessarily require intervention. Again, correlation with the clinical picture is essential.

Clinical Presentation of Hydrocephalus

The presentation of hydrocephalus differs in the case of the neonate and infant compared with the older child or adult.

Prior to closure of the cranial sutures and obliteration of the fontanelles, hydrocephalus results in disproportionate head growth. Thus, over the first 2–3 years of life, measurement of the occipito–frontal circumference and plotting this on a centile chart provides a simple and sensitive test. Wherever possible, sequential measurements (corrected for gestational age) should be obtained in order that the trend of head growth in relation to the centile lines can be demonstrated. Clinical symptoms are often subtle and include general irritability, poor feeding and slow attainment of milestones. In addition to head size, clinical signs include bulging of the fontanelle, separation of the cranial sutures, prominence of scalp veins and sun-setting of the eyes. This latter clinical sign is attributed to pressure on the mid-brain tectum by CSF in the supra-pineal recess. Papilloedema can be difficult to diagnose in the infant and indeed is not uncommonly absent in infantile hydrocephalus and so is an unreliable sign in this context.

In older children and adults, the classical symptom complex of raised intracranial pressure, headache, vomiting and drowsiness is more likely to herald an underlying diagnosis of hydrocephalus. Where hydrocephalus has developed insidiously, cognitive impairment, poor concentration and behavioural changes occur. Visual obscurations and papilloedema are more common than in the younger age group.

In both groups of patients, the presence of bradycardia, hypertension and irregularities in breathing pattern imply critical elevation of intracranial pressure and should be treated promptly.

Investigation of Hydrocephalus

Cranial Ultrasound Scanning (see Chapter 2)

In the neonate, the supratentorial ventricular system can be reliably evaluated using ultrasound. This is the imaging modality of choice in the investigation and monitoring of the infant with an open fontanelle. Hematomas or other ventricular masses responsible for hydrocephalus can also be identified. Ultrasound provides a non-invasive and readily available tool for both diagnostic purposes and, by means of sequential studies, a way of charting changes in ventricular size.

CT and MRI

In order to more fully evaluate the entire ventricular system and investigate the underlying etiology of hydrocephalus, CT or MRI scanning is required. Clearly, there is a range of normal ventricular size and, indeed, ventricular size changes with age, rendering absolute measurements of ventricular dimensions of little use. No single radiological parameter can be relied upon to distinguish hydrocephalus from the other causes of ventricular enlargement mentioned above. Some features, however, are strongly suggestive, particularly when occurring in combination. Enlargement of the temporal horns of the lateral ventricles and enlargement of the third ventricle, commensurate with the enlargement of the rest of the ventricular system, are findings in favor of hydrocephalus. Obliteration of the basal cisterns and effacement of the cortical sulci further support a diagnosis of hydrocephalus. When the ventricles are under pressure, there may be transependymal flux of CSF into the periventricular parenchyma, particularly at the tips of the frontal occipital and temporal horns. This appears as low density on CT scan or a rim of high signal intensity on the T2-weighted MRI scan (Fig. 24.2).

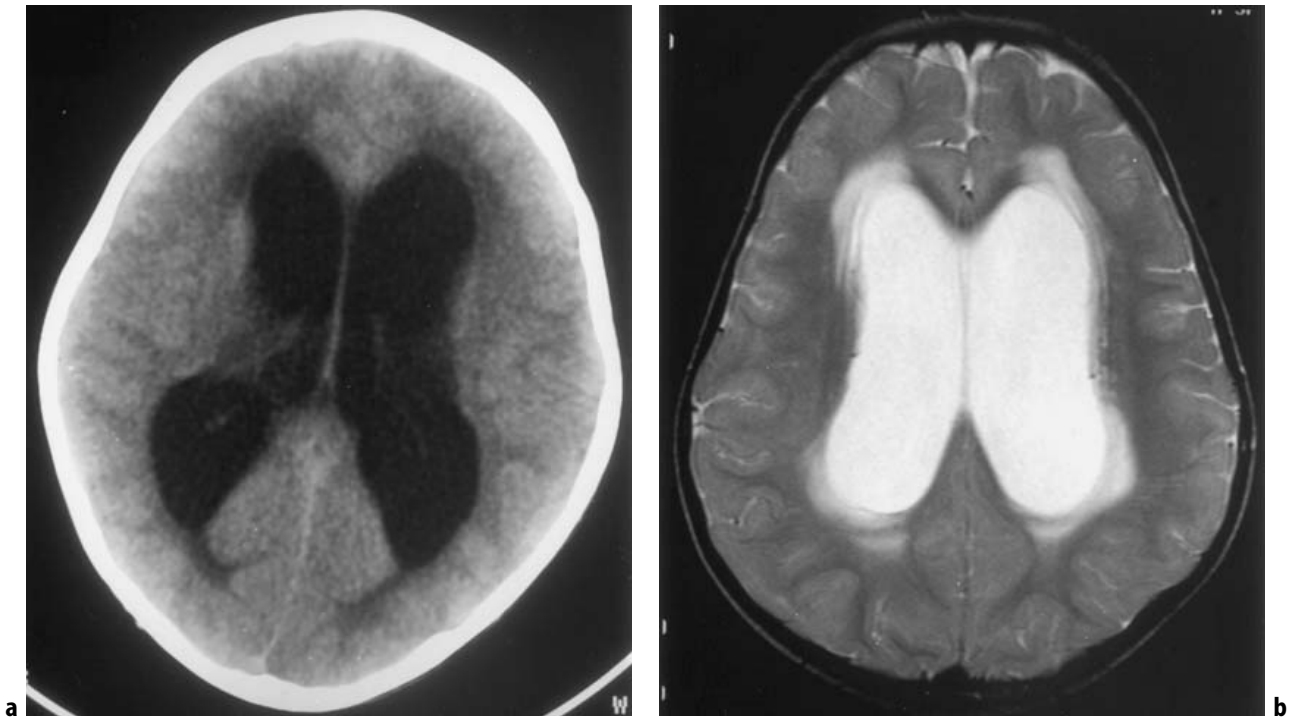

Fig. 24.2. The imaging characteristics of trans-ependymal seepage of CSF in hydrocephalus. On CT, a rim of low density and on MRI, a rim of high-signal intensity on the T2-weighted image.

Treatment of Hydrocephalus

Shunts

Bypassing the site of obstruction to CSF flow or diverting the CSF from the ventricular cavity to a site where it can be more readily absorbed are the basic principles underlying the treatment of hydrocephalus. The ventricular shunt is the mainstay of hydrocephalus treatment and, even in severe hydrocephalus, shunt insertion can have a dramatic effect on the re-expansion of the cortical mantle, particularly in the infant (Fig. 24.3). Neuroendoscopic techniques, in particular third ventriculostomy, also play an important role in the treatment of hydrocephalus and this subject is dealt with in detail elsewhere.

Numerous shunt systems have been devised and marketed, though all have their shortcomings and are prone to similar complications (see below).

The shunt assemblage comprises a proximal catheter, located in the cerebral ventricle, and a distal catheter draining to some alternative site of CSF absorption, most commonly the peritoneal cavity, but drainage to the pleural cavity or right atrium is occasionally employed. Usually, a valve and reservoir are incorporated into the shunt, although the precise configuration is variable.

Proximal Catheter

The most commonly used ventricular catheter comprises a blind-ended silastic tube with a number of side holes adjacent to the catheter tip. Modifications of this basic design will occasionally be encountered, for example flanged catheters. These were designed to protect the side holes from occlusion by tissue at the time of introduction; however, no such advantage has been demonstrated.

A number of devices are available to permit placement of the catheter within the ventricle. A stylet passed down the lumen of the catheter or clipped to its outside is the most simple and widely used. Endoscopes are now available which fit inside specially designed open-ended catheters; these have the advantage of permitting visualization of the ventricular cavity and so aiding optimal placement. Whilst

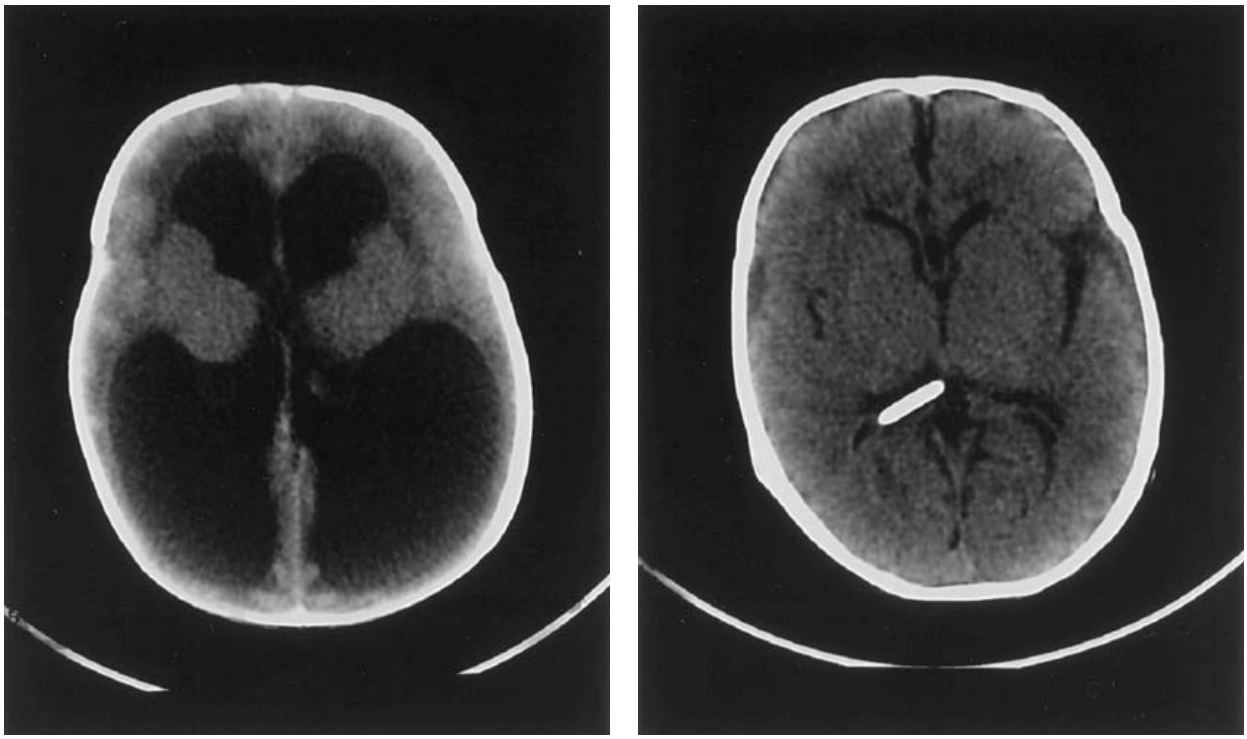

Fig. 24.3. CT scan appearances before (left) and 6 weeks after (right) the insertion of a VP shunt.

this technique may be useful in certain circumstances, it is probably not necessary for routine use.

Valves

A variety of valve designs are currently available (Table 24.2). An array of eponymous names, valve types and manufacturers can lead to great confusion. The table attempts to provide a simple classification of some of the valve designs in common usage but it is by no means exhaustive.

Differential pressure valves are pressure-regulating devices. Pressure regulators maintain a fixed-pressure differential, regardless of flow; this is in contrast to flow regulators that maintain constant flow regardless of pressure. Four types of differential pressure valve are commonly encountered: slit valves, mitre valves, ball and spring and diaphragm valves [11].

For most fixed valves, a low, medium or high-pressure alternative is available. The pressure setting defines the opening or, more commonly, the closing pressure of the valve (the physical characteristics of the valve are the reason for the differences in these two values).

In order to overcome the limitations imposed by fixed-resistance valves, some manufacturers have developed valves whose operating pressure can be varied once the shunt has been implanted. Such devices include the Medos programmable valve (Codman Medos, Le Cocle, Switzerland) and the Sophysa adjustable valve (Sophysa, Orsay, France). An externally applied magnetic field is used to alter the position of an internal rotor and thus vary the pressure setting. The setting is then verified on plain radiograph (Medos) or a compass held over the device (Sophysa).

The pressure gradient across the valve is the difference between the intraventricular pressure and the intra-abdominal pressure in the supine position. In the upright position, the added hydrostatic pressure will increase the differential pressure and so increase the CSF flow through the valve. One of the criticisms of differential pressure valves is that they are subject to this siphoning phenomenon in the upright position.

A number of modifications of shunt design have been devised in order to overcome the problem of siphoning. Anti-siphon or siphon

Table 24.2. Types of CSF shunt valves.

Valve principle	Valve type	Examples
Differential pressure (fixed)	Slit	Codman Holter Codman Denver
	Mitre	Mueller Heyer Schulte in line valve
	Ball and spring	Cordis Hakim valve Codman Medos Hakim valve
	Diaphragm	Mueller Heyer Schulte Pudenz flushing valve PS Medical flow-control valve Codman Accu-flo
	Diaphragm + siphon control device	PS Medical Delta valve
Differential pressure (externally programmable)	Ball and spring mechanism, opening pressure adjusted by externally applied magnetic field.	Codman Medos Programmable Sophysa Adjustable
Flow-control valve	Variable-resistance valve	Cordis Orbis Sigma

control devices (ASDs) can be incorporated in series with the valve. The device, which is placed subcutaneously in series with the valve, houses a mobile membrane that moves in response to a pressure change across it. The outer surface is theoretically at atmospheric pressure. When the pressure within the shunt falls, the membrane moves to occlude the shunt lumen. ASDs are available as separate components to insert below the valve in existing shunts; alternatively they may be incorporated into a valve as in the Delta valve (P.S. Medical Corporation, California, USA) which combines a diaphragm valve and a siphon control device in one.

A different approach to the siphon problem is seen in the Orbis-Sigma valve (Cordis Corporation). By contrast to differential pressure valves, which are pressure regulating, this valve is designed to be a flow-regulating device, permitting a relatively constant flow rate over a wide range of differential pressures.

Unfortunately, at present, there are insufficient data to support many manufacturers' claims of superior function of particular shunt valves. Choice of valve design continues to be, for the most part, a matter of surgical preference, no clear advantages have been consistently demonstrated for any individual model [12].

Distal Catheter

The currently preferred site of drainage is to the peritoneal cavity. Insertion into the peritoneal cavity is either by mini-laparotomy or the use of an abdominal trocar. In childhood, a suitable length of tubing is inserted into the peritoneal cavity in order to compensate for the effects of growth.

Alternative sites include the right atrium, the pleural cavity and the gall bladder. The ventriculoatrial (VA) shunt was the preferred technique prior to the introduction of silastic catheters (previous catheter materials tended to incite a tissue reaction and become occluded); however, the consequences of VA shunt infection, including septicaemia and renal failure, were responsible for significant morbidity and mortality. Furthermore, the positioning of the catheter tip is critical in the atrial shunt (in order to maintain patency) and thus frequent revisions were necessary as the child grew. On occasion, the use of the peritoneal cavity will be precluded, for example following abdominal sepsis or in the presence of extensive post-surgical adhesions, and, in such situations, the atrial site is still used.

Medical Treatment

Medical measures to treat hydrocephalus may be appropriate under certain circumstances. The use of osmotic diuretics such as mannitol is restricted to situations where prompt reduction of intracranial pressure is necessary pending definitive treatment, as in the acute shunt block with neurological deterioration whilst arranging for shunt revision.

Acetazolamide is an inhibitor of the enzyme carbonic anhydrase. The enzyme is present in the choroid plexus epithelium and is necessary for the production of CSF. Acetazolamide is

frequently used to reduce CSF production in cases of benign intracranial hypertension (pseudotumor cerebri). It is occasionally used in the management of neonatal post-hemorrhagic hydrocephalus as a temporising measure whilst the child gains weight and the CSF clears of blood products.

Complications of Shunts

The extensive range of shunt models available and the constant search for new alternatives is unfortunately a reflection of the shortfalls of all shunt systems to date. A complex combination of factors relating to the patient, the surgeon and the shunt device itself are likely to be responsible for the complications that are well known to all who treat hydrocephalus.

Mechanical failure and infection together account for the vast majority of shunt complications. A list of some of the more commonly recognized shunt complications is shown in Table 24.3, some of which are discussed below.

Shunt Blockage

Shunt obstruction is the commonest indication for shunt revision and, in the majority of cases, the cause is blockage of the ventricular catheter. Choroid plexus, brain tissue and cellular debris are frequently responsible for the occlusion. Obstruction of the shunt may, however, occur at any level in the shunt assembly. The risk of mechanical failure is related to the time from shunt surgery, with most of these complications occurring in the first post-operative year [13].

The clinical presentation is usually dominated by signs of raised ICP – headache, vomiting and drowsiness are most common. In the infant population group, enlarging head circumference, tense fontanelle, CSF tracking along the course of the shunt and, rarely, seizures are additional indicators of underlying shunt malfunction. The time course of symptom onset is, however, extremely variable; in some, the onset may be insidious over days or weeks whilst in the more shunt-dependent individuals, rapidly progressive symptoms may develop in the space of a few hours.

Table 24.3. Commonly encountered complications of shunts.

Complication
Infection
Shunt blockage (proximal, valve, distal)
Fracture or disconnection
Migration
Overdrainage
Isolation (trapping) of ventricles
Malposition
Intracranial haemorrhage
Viscus perforation

It has been postulated that the site of insertion of the ventricular catheter has some bearing on the propensity for obstructive complications. The frontal site is preferred by some surgeons, who argue that placement in the frontal horn beyond the foramen of Monro reduces the likelihood of blockage by choroid plexus. This practice does suppose that accurate placement can be attained during blind shunt insertion and that choroid plexus is a major etiological factor in shunt obstructions. Malposition of the ventricular catheter is, however, well recognized and, furthermore, Sekhar has demonstrated that tissues other than choroid plexus, including glial tissue, leptomeninges, chronic inflammatory debris fibrin and thrombus may be responsible for catheter obstruction. In a prospective randomized trial, Bierbrauer et al. [14] failed to show any advantage of frontal placement over the occipital route.

CSF Overdrainage

Despite modifications of valve design including on/off control, anti-siphon devices and, more recently, externally programmable valves, shunts cannot reproduce the balance between CSF production and absorption characteristic of normal physiology. Excessive CSF drainage may result in symptoms of headache, nausea and vomiting, diplopia and a generalized lethargy, sometimes with impairment of school performance. These symptoms can be difficult to distinguish from raised ICP. Resolution of symptoms on lying down is an occasional indicator favoring intracranial hypotension.

The consequences of overdrainage include subdural hematoma formation. This may vary from mild extra-axial collections that are often managed conservatively, to larger symptomatic subdural hematomas that may necessitate intervention. Treatment strategies may include burr hole drainage with or without shunt removal, upgrading the valve to a higher pres-

sure or, on occasion, it is necessary to insert an additional (unvalved) sub-dural shunt; this is sometimes plumbed into the existing shunt below the valve.

In the infant, the lowered intracranial tension following shunt insertion may lead to premature closure of the cranial sutures, producing a secondary craniosynostosis with cranial deformity.

Asymmetrical drainage of the ventricles may also be seen to cause trapping or isolation of part of the ventricular system. It may be difficult to distinguish whether this is a true consequence of the shunt or related to compartmentalization of the ventricles as a result of the original pathology, for example post-meningitic hydrocephalus. Trapping of the fourth ventricle is an example of this process and can be seen after apparently successful treatment of hydrocephalus with a shunt. Isolation of the fourth ventricle may be discovered incidentally or may result in symptoms of raised intracranial pressure or cerebellar disturbance. In symptomatic cases, the fourth ventricle can be drained either by inserting an additional shunt system or by placing a catheter into the fourth ventricle and plumbing this into the existing supratentorial shunt via a T or Y connector. It is important that the connection is made above the valve to ensure that the ventricles are drained at the same pressure.

The Slit Ventricle Syndrome

It is important to distinguish between the radiological "label" of slit ventricles, a not uncommonly seen appearance on post-shunt CT scans where the ventricles are barely recognisable but the patient is free of symptoms, and the less frequently encountered clinical symptom complex that may accompany slit-like ventricles. It has been estimated that only 11% of patients with radiologically confirmed slit ventricles demonstrated the clinical syndrome. The clinical syndrome is usually one of episodic headache, which may be positional, vomiting, occasionally with vague gastrointestinal symptoms and the reservoir, if present, may be slow to refill. The symptoms will frequently have a cyclical pattern, episodes lasting from between a few hours to 2 or 3 days, the individual being quite well in between "attacks".

It is postulated that the condition results from a loss of ventricular wall compliance. The small-volume ventricles intermittently collapse around the catheter, temporarily blocking it. Intracranial pressure has to build up in order to distend the non-compliant ventricles; during this period symptoms will be present. Once the ventricle begins to expand, the catheter can again begin to function and symptoms subside.

The syndrome may be accompanied by either low or high intracranial pressure and differentiating these is frequently difficult on clinical grounds alone. A period of ICP monitoring may be a useful aid in the diagnosis and may guide subsequent treatment [10].

If ICP is low then the therapeutic options include upgrading the valve or insertion of an anti-siphon device. Such maneuvres may be associated with re-expansion of the previously collapsed ventricle.

In the presence of raised ICP, clearly it is essential to establish that the shunt is patent. If this is so and symptoms persist, then subtemporal decompression may afford relief. The removal of bone, usually ipsilateral, to the shunt removes some of the constraint upon ventricular dilation, improving compliance and permitting focal expansion of the ventricular cavity. This has been reported to improve symptoms and reduce the number of subsequent shunt-related problems [15].

Abdominal Complications

Numerous complications have been described in relation to the distal shunt catheter. Viscus perforation can occur either as a complication of the initial insertion or may develop as a result of chronic erosion of the catheter tip through the viscus wall. Perforation of the stomach, the large and small bowel, gall bladder and vagina are all described. The presentation may be obvious, with signs of peritoneal sepsis or occasionally with extrusion of the catheter tip at the anus, umbilicus or vagina. Catheters that have become disconnected and lost in the peritoneal cavity can lead to symptoms and will make it difficult to eradicate infection. Retained catheters should be avoided wherever possible.

Intra-abdominal Fluid Collections

It is rare for ascites to develop as a result of the peritoneal cavity failing to cope with the CSF load, except where there is additional abdominal pathology such as adhesions from previous

sepsis, surgery or ongoing infection. Hydroceles and an increased incidence of inguinal hernia are recognized complications of ventriculoperitoneal shunts in infants.

More common is the formation of localized CSF collections within the peritoneal cavity. Abdominal pain and distension are common symptoms in the presence of such CSF pseudocysts; these can be readily diagnosed on ultrasound examination. The presence of a CSF pseudocyst should always raise the possibility of underlying infection, which has been reported to occur in two-thirds or more of cases [16].

In such cases, management should be aimed at eradication of the infection, with either removal or externalization of the infected shunt. Although some neurosurgeons have recommended conversion to a VA shunt, a history of pseudocyst formation does not necessarily preclude the continued use of the peritoneal cavity as the distal site.

Shunt Infection

Shunt infection is one of the most common complications encountered and one that carries significant morbidity and even mortality. Shunt infections result in prolonged hospitalization, they increase the risk of subsequent shunt malfunction and can lead to physical disability and impaired intellectual development. Reported shunt infection rates in some instances exceed 20%, although a range of 5–15% would be a more realistic figure, examining larger series of pediatric patients. Rates as low as 1% have been achieved in some centers [17].

The subject of shunt infection is a complex one, and beyond the differences between individual neurosurgical units, there are important variations relating to the heterogeneity of the hydrocephalic population. Whilst shunt infection may afflict adults as well as children, it is in the pediatric population that shunt infection rates tend to be greater and the majority of studies have been performed in this group.

Although there are many factors that appear to contribute to shunt infection, it is generally assumed that contamination of the shunt system occurs at or around the time of shunt surgery. Poor surgical technique, excessive handling of the shunt hardware and inadequate operative environment are among some of the general risk factors frequently cited. Specific factors that appear to have particularly strong correlations with shunt infection are postoperative wound infection and CSF leakage; strenuous measures should be taken to avoid these complications.

Within the pediatric age group, patient age also appears to play a significant role. Pople et al. [18] report an incidence of infection of 15.7% in children less than 6 months of age in contrast to a rate of 5.6% in those older than 6 months. Immunological immaturity, different microbiological flora and physical properties of the skin are among the possible factors increasing the risk of shunt infection in the neonate.

The Presentation of Shunt Infection

Ventriculoperitoneal shunt infection most commonly manifests itself soon after an operative procedure – either shunt insertion or revision. Approximately 70% of shunt infections will have presented within 2 months and 80% by 6 months of the surgical procedure [19]. Rarely, shunt infections present later than this, for example following late perforation of a viscus by the distal shunt, incidental abdominal sepsis (e.g. appendicitis) or in association with wound breakdown or erosion of the shunt through the skin.

A high index of suspicion should be maintained in the weeks following surgery. The mode of presentation is a variable constellation of pyrexia and meningism and general irritability in children.

In the case of ventriculoatrial shunts, infection may manifest early on with an acute, overtly septic illness. A more chronic presentation is also well recognized, characterized by prolonged periods of generalized lassitude, sometimes with a low-grade pyrexia or mild anemia. Vague low-back pain, hematuria and hypertension may herald the onset of "shunt nephritis" – an immune complex-mediated nephritis that can result in renal failure.

In suspected shunt infections, CSF examination is needed to confirm the diagnosis and may be obtained by aspiration from the shunt reservoir – a shunt tap. CSF Gram stain and culture are often diagnostic and will aid in selection of appropriate anti-microbial therapy. It is important to appreciate that if the peritoneal end of the shunt is the source of the infection, a negative CSF result may be obtained in the early

stages. Abdominal ultrasound examination, looking for encysted collections of CSF, may be useful in such cases (see above).

Blood cultures are frequently unhelpful in diagnosing VP shunt infection; however, measurement of C-reactive protein (CRP) can be a useful guide, both as part of the initial investigation and as a means of monitoring the effectiveness of treatment.

Organisms Responsible for Shunt Infection

The commensal skin flora is the usual source of pathogens that give rise to shunt infections with the coagulase-negative staphylococci, particularly *S. epidermidis*, the most commonly isolated. *S. aureus* is also well recognized, especially in the context of wound infection or skin breakdown. Enterococci, micrococci and coryneforms account for a significant proportion of the remainder of infecting organisms (Table 24.4).

One of the principal factors which enables coagulase-negative staphylocci to colonize shunt systems is their ability to produce an extracellular slime, which aids adherence of the organisms to the surface of the silicone catheter [20]. This is also one of the main factors responsible for the resilience of these infections to treatment with the shunt in situ.

Treatment of Shunt Infection

Once diagnosed, shunt infection requires prompt and comprehensive treatment with appropriate anti-microbial therapy. Controversy exists, however, as to whether treatment necessitates complete removal of the shunt system or whether the infection can be managed with the shunt in situ. Those who favor treating the infection with the shunt in situ cite the risks of shunt removal, including hemorrhage, from adherent ventricular catheters and the risk of super-added infection associated with temporary external ventricular drainage in support of their policy. The success rates associated with this line of management, however, are poor [21] and the overall morbidity associated with surgical treatment (shunt removal and antibiotic therapy) is lower than with medical therapy alone.

Table 24.4. Organisms responsible for shunt infection.

Organism	Number of cases
Staphylococcus epidermidis	24
Staphylococcus aureus	12
Methicillin-resistant *Staphylococcus aureus*	3
Enterococcus faecalis	2
Escherichia coli	1
Aerococcus viridans	1
Beta-haemolytic Streptococcus group A	1
Pseudomonas putida	1

Data from Great Ormond Street Hospital 1994–97. Courtesy of Dr H. Holzel, Dept of Microbiology (unpublished).

The most common strategy is removal of the shunt and replacement with an external drain for the duration of antibiotic treatment. This permits intrathecal administration of antibiotics if required and serial sampling of the CSF for Gram stain, culture and monitoring of the white cell response. A new shunt is inserted once the CSF is sterilized.

The Role of Antibiotic Prophylaxis in Shunt Surgery

The temporal relationship between time of operation and the occurrence of shunt infection, together with the observation that the commensal skin flora is the commonest source of pathogens, might suggest that antibiotics given at the time of surgery would reduce the incidence of infectious complications. Although there have been numerous studies attempting to demonstrate this, most have failed to reach statistically significant conclusions. One of the principal problems has been enrolling sufficient patients into randomized trials to demonstrate an effect. Two reports have sought to circumvent this problem of type II error using the techniques of metanalysis [22,23]. Both of these reports came out in favor of prophylaxis. Haines and Walters, however, caution that any demonstrable benefit is related to the baseline infection rate; no beneficial effect could be demonstrated when this was less than 5%.

Additional controversy then surrounds the issue of the choice of antibiotic, its route of administration and the duration of prophylaxis. Many antibiotics, including vancomycin, cephalosporins and aminoglycosides, when administered via the intravenous route, fail to achieve significant levels in the CSF, particularly in the absence of inflammation and are thus inappropriate in this respect. Some workers have suggested intraventricular administration

or soaking the shunt components in antibiotic solution prior to insertion.

A more recent technique of antibiotic delivery has been to incorporate antibiotics into the silicone tubing. The antibiotic gradually leaches out from the tubing, providing protection in the early post-operative period. In-vitro studies have demonstrated that shunt colonization was delayed for up to 56 days [24]. Shunts containing antibiotic-impregnated silicone tubing using rifampicin and clindamycin are now available for use; however, the results of clinical trials of such devices are awaited.

Miscellaneous Shunt Complications

Seizures

Since the ventricular catheter must, of necessity, traverse the cerebral cortex, there has been concern as to whether this increases the risk of epilepsy for hydrocephalic patients. Whilst a number of studies have attempted to address this question, drawing meaningful conclusions is compromised by the heterogeneity of cases included and variable definitions of epilepsy. Dan and Wade [25] reported that 9.4% of 180 cases of hydrocephalus of variable etiology developed epilepsy after shunting. These authors cite multiple revisions and use of the frontal route of insertion as particular risk factors. Their methodology and conclusions have, however, attracted criticism. Many authors report the incidence of epilepsy in shunted patients to be much greater than this. In an actuarial analysis of 464 patients, Piatt and Carlson [26] found an incidence of epilepsy (defined by the use of anticonvulsant medication) of 12% at the time of diagnosis rising to 33% at 10 years post-shunting. They emphasize that the underlying cause of the hydrocephalus, in particular post-hemorrhage and post-infection, was a far more important predictor of the risk of epilepsy than any surgical factors, including shunt position, number of revisions or history of shunt infection, none of which reached statistical significance. Reporting cases of congenital hydrocephalus only, Noetzel and Blake also favor patient factors, specifically mental retardation and cerebral malformations, as best indicators of long-term epilepsy in shunted patients [27].

Metastases

Extraneural metastasis of primary CNS tumors is an unusual but well recognized phenomenon. Medulloblastoma is the most common of the neural malignancies reported to spread in this way, although germinoma and astrocytoma are also well described. Lymphatic and hematogenous pathways are the most likely routes of spread; however, whether or not shunt systems provide a potential conduit for tumor spread has given cause for concern. Indeed, the incorporation of filters into shunt systems has been advocated to diminish the risk of this. In an analysis of 160 published cases of extraneural spread of medulloblastoma, Jamjoom et al. [28] suggested that the shunt could be implicated in no more than 11 patients. In a further eight cases of extraneural spread in a series of 415 intracranial tumors, Berger et al. [29] failed to identify any increased risk attributable to the presence of a shunt device.

Hemorrhage Related to Ventricular Catheters

Both at the time of initial insertion and particularly during ventricular catheter removal, hemorrhage may occur. Choroid plexus frequently becomes entwined in the catheter and is easily avulsed, leading to intraventricular hemorrhage. It has been suggested that some degree of intraventricular hemorrhage occurs in almost one-third of ventricular catheter revisions. This complication should always be considered when removing catheters, particularly if they have been in situ for a prolonged period of time. Careful evaluation of the pre-operative CT scan may in fact reveal choroid plexus adjacent to the ventricular catheter, highlighting the possible risk of this complication. When an adherent catheter is encountered, it may be necessary to leave it in situ and pass a new one rather than risk choroid plexus avulsion. Passing a stylet along the lumen of the catheter and cauterizing with diathermy prior to withdrawal may help prevent this complication and aid removal of the catheter. Where bleeding does occur, the catheter should be left to drain until the CSF clears; gentle irrigation to prevent occlusion of the catheter with blood clot can be performed. If the blood fails to clear, an external ventricular drain should be left; the patient needs to be

closely monitored and scanned post-operatively to assess the extent of the hemorrhage.

Silicone Allergy

Allergic reactions to silicone in ventriculoperitoneal shunts have been reported [30], although is it a rare phenomenon. The presentation may comprise fever and malaise, sometimes mimicking shunt infection. Erythema along the path of the shunt or wound breakdown sometimes occurs. The pathological basis of this condition is a foreign-body giant cell reaction with granuloma formation. Treatment necessitates removal of the shunt and replacement with a silicone-free alternative.

The Prognosis of Shunted Hydrocephalus

Numerous variables influence the long-term neurological and cognitive outcome of shunted hydrocephalus. Whilst a number of studies have attempted to elucidate the relative importance of some of these factors, they need to be interpreted with caution. Donders et al. [31] cite sample bias, variable means of assessment of intellectual outcome, inadequate statistical analysis and the failure to account for coexistent medical problems as the main methodological inadequacies of many of these previous studies. Regression analysis of their own series of patients (in whom they attempted to overcome these design shortcomings) highlighted neonatal problems, including anoxia, respiratory distress, CNS infection and early seizures, and also ocular defects (gaze, movement and refraction) as particularly poor prognostic factors for long-term intellectual outcome.

The timing as well as the nature of the cerebral insult also appear to be important determinants of long-term outcome. A prenatal or neonatal cause for hydrocephalus has been associated with a worse outcome when compared with cases in which postnatal onset was identified or where the etiology of the hydrocephalus was unknown [32].

Academic placement may serve as more tangible criterion of outcome than IQ. In a cohort of 155 shunted patients after a follow-up of at least 10 years, Casey et al. [33] found that over half of the children were able to attend a normal school. Forty-one percent, however, required special schooling. Children whose hydrocephalus was the consequence of intraventricular hemorrhage or infection were much more likely to require special schooling.

In this study, more than half (55%) required one or more shunt revisions during the 10-year follow-up period. Furthermore, a mortality rate of 11% for non-tumor-related hydrocephalus was revealed in this study. Precipitous deterioration and sudden death following shunt blockage are well recognized. The adage *Once a shunt, always a shunt* should at least be assumed to be true and serves as a reminder that when the shunted patient presents with new or unexplained symptoms, the possibility of shunt malfunction must be borne in mind and investigated promptly.

Key Points

- *Hydrocephalus is not a single disease entity. Treatment should be formulated based on an understanding of the underlying condition.*
- *Ventricular size is not the sole index by which hydrocephalus and the need for treatment should be evaluated; always correlate with the clinical picture before embarking on surgery.*
- *A wide range of shunt products are available for the treatment of hydrocephalus; it is recommended that the clinician becomes familiar with the use of a particular shunt system and reserves the more novel shunt valves for specific problem cases.*
- *The clinical presentation of shunt malfunction is extremely variable; shunt malfunction should always be considered in the presence of new or unexplained symptoms.*
- *When presented with cases of shunt malfunction, consider whether there is an opportunity to simplify complex shunt systems or perform endoscopic third ventriculostomy.*

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