Hydrocephalus

Ahmed K Toma

Abstract

Hydrocephalus is a term applied to conditions where there is disturbance in cerebrospinal fluid (CSF) dynamics. These are often, but not necessarily, associated with enlarged cerebral ventricles and increased intracranial pressure. Hydrocephalus could be idiopathic or secondary to congenital or acquired conditions. Clinical presentation depends on the age of presentation, with paediatric hydrocephalus presenting with increased head circumference or features of raised intracranial pressure. In the elderly, normal pressure hydrocephalus is a specific entity that presents with a classical triad of gait, cognitive and continence impairment. Urgent neurosurgical management of acute hydrocephalus is often required. The main method of treatment is surgical, mostly by inserting a CSF drainage shunt. These are catheters that divert CSF to a body cavity, with an embedded valve mechanism to control drainage. Unfortunately shunt complications are common. Recognizing a shunt complication and early referral to a neurosurgical unit is important to prevent serious consequences to the patient.

Keywords hydrocephalus; shunt; CSF

Definition

Hydrocephalus is generally known as a condition caused by cerebrospinal fluid (CSF) accumulation within the brain resulting in enlargement of the cerebral ventricles. In infants with open fontanelles, this is commonly associated with increased head circumference, while in older children and adults it is likely to cause raised intracranial pressure. The term (hydrocephalus) is a modern Latin adaptation from Greek *hudrokephalon*, from *hudro-* 'water' and *kephalē* 'head'.[1]

However, there are certain conditions that are considered as hydrocephalus, where the ventricles are small or normal in size (like slit ventricles syndrome), or where the ventricles are large but in the presence of low or normal intracranial pressure (like normal pressure hydrocephalus). Therefore, a generally accepted definition of hydrocephalus is as follows.

> '*A spectrum of conditions where there is a disturbance in cerebrospinal fluid dynamics*'.[2]

In this article, we summarize the pathogenesis, clinical presentation and management of the common hydrocephalic syndromes.

Pathogenesis

CSF is a clear fluid that surrounds the brain and spinal cords and fills the cerebral ventricles. The cerebral ventricles are four interconnected cavities of the brain, lined by ependyma. CSF is mainly produced by active secretion at the choroid plexuses of the cerebral ventricles.[3]

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CSF circulates from the two lateral ventricles, through the interventricular foramen of Monro to the third ventricle, then the cerebral aqueduct of Sylvius, to the fourth ventricle. From the fourth ventricle, CSF passes to the subarachnoid space around the brain and spinal cord through the foramen of Magendie in the midline and the two foramina of Luschka laterally. The CSF circulation comprises not only a directed flow of CSF, but in addition a pulsatile to and fro movement. The CSF circulation around blood vessels penetrating from the subarachnoid space into the Virchow Robin spaces. We are starting to understand that important physiological functions, for example the regeneration of the brain during sleep, may depend on CSF circulation.[4]

CSF is formed at a rate of about 0.5 ml/minute. The daily volume of CSF produced in adult humans is about 500 ml. The total CSF space in young adults is about 150 ml, that is, the CSF is totally replaced about four times each day. Only about 25% of CSF volume lies within the ventricles. The rest resides in the cranial and spinal subarachnoid space.[3]

There has been a gradual paradigm shift in understanding the site of CSF absorption. Previously, it was thought that CSF is absorbed into the blood by the arachnoid villi in the dural venous sinuses. Currently accepted theory is that CSF is cleared from the CNS by bulk flow along sleeves of the subarachnoid space surrounding cranial nerves that enter the nose and eyes, as well as spinal nerves.[3,5]

The relationship between the volume of intracranial components and intracranial pressure (ICP) is important in the pathophysiology of many hydrocephalic syndromes. The Monro-Kellie doctrine states that the skull is a closed bony box with constant volume. An increase in volume of one of the cranial constituents, or the presence of a mass lesion (tumour or haematoma), would result in raised intracranial pressure.

$$\text{v.intracranial (constant)} = \text{v.brain} + \text{v.CSF} + \text{v.blood} + \text{v.mass lesion}$$

A physiological buffer exists, where some compensation is possible as CSF and blood move into the spinal canal and extracranial vasculature respectively. Beyond this point, ICP rises

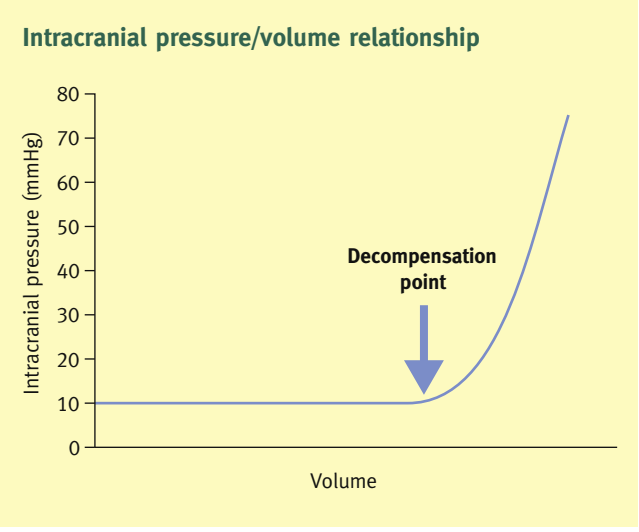

Figure 1

dramatically (Figure 1). Increased ICP can lead to one of the life-threatening herniation syndromes ('coning').[6,7]

Classification

There are different classification schemes based on different criteria. The most widely used classification is based on the CSF dynamics, where hydrocephalus is classified as obstructive (non-communicating) or non-obstructive (communicating hydrocephalus) depending on the presence of blockage of the major CSF circulation pathway. Hydrocephalus syndromes are also classified based on the aetiology as congenital, acquired or idiopathic. Hydrocephalus is also commonly classified as neonatal, infantaile, paediatric or adult hydrocephalus. Adult hydrocephalus is often subclassified as high or normal (low) hydrocephalus.[2,8]

Aetiology

The underlying causes of hydrocephalus syndromes can be broadly classified into congenital, acquired and idiopathic categories. Congenital hydrocephalus usually presents in neonatal period. Chiari malformation (Type 2) is commonly associated with myelomeningocele. The Dandy–Walker complex is a specific entity with hydrocephalus caused by atresia of foramina of lushka and magendi, with agenisis of cerebellar vermis and large posterior fossa cyst. Aqueductal stenosis usually present in childhood. However, first presentation could be delayed until adulthood. The classical presentation is that of triventricular hydrocephalus: enlarged third and lateral ventricles with a small fourth ventricle. The underlying cause is either forking, septum, true stenosis, or gliosis of the aqueduct. An X-linked recessive gene is a rare cause of aqueduct stenosis.

Acquired causes of hydrocephalus include haemorrhage, infection or tumours. In premature infants, bleeding into the germinal matrix commonly produces intraventricular haemorrhage. This is often complicated by hydrocephalus. In adults, communicating hydrocephalus is a common complication of subarachnoid haemorrhage. Post-meningitis hydrocephalus is seen post-pyogenic or post-tuberculous meningitis.

Mass lesions including tumours blocking CSF pathways results in obstructive hydrocephalus. In children, the posterior fossa tumours ependymoma, astrocytoma or medulloblastoma usually first present with raised ICP caused by hydrocephalus. A colloid cyst of the third ventricle results in intermittent hydrocephalus or acute hydrocephalus. Pineal tumours cause aqueduct stenosis and triventricular hydrocephalus.

Choroid plexus papilloma or carcinoma are rare entities that produce hydrocephalus by excessive CSF production.

Normal pressure hydrocephalus is a disease of the elderly population. The underlying cause is not fully understood. The currently accepted theory is that of pulse-induced encephalopathy, in which the ventricles are enlarged because of the chronically transmitted hammering effect of arterial pulse caused by arteriosclerosis.[8–10]

Epidemiology

The incidence of paediatric hydrocephalus is less than 1 in 1000 live births. In developed countries, there has been a decline in hydrocephalus caused by congenital malformation or infection. On the other hand, the incidence of post-haemorrhagic hydrocephalus in prematurely born infants has increased with improving survival rates. In developing countries, neonatal infection and myelomeningocele are linked to higher rates of hydrocephalus compared with the developed world.[8]

Population-based studies estimate the prevalence of normal pressure hydrocephalus in the elderly population to be 1.4–2.9%, and the incidence has been estimated to be 5.5/100,000/year. Several studies have estimated the prevalence of normal pressure hydrocephalus in the dementia population to be between 1.6% and 5.4%.[11]

Diagnosis

The clinical presentation of hydrocephalus varies depending on the aetiology and age. In neonates, hydrocephalus often presents with an enlarged head and craniofacial disproportion. Other features include: irritability, vomiting, apnoeic spells, failure to thrive, and poor feeding. On examination, the scalp is often thin and glistening with dilated veins. Fontanelles are tense and bulging with diastasis of the sutures. The 'setting sun' sign is a combination of upper eyelid retraction and upgaze failure.

In older children with closed skulls and adults, hydrocephalus presents with features of raised ICP. Headache is classically worse in the morning. (This is due to relative hypercapnoea during sleep. Hypercapnoea causes increased ICP.) Vomiting will often relieve the headache as a result of hyperventilation clearing CO_2 and hence improving ICP. Diplopia is caused by six nerve palsy. The six cranial nerve (abducent), is a thin long nerve that can be distorted by raised ICP. Signs include papilloedema and sixth cranial nerve palsy. If untreated, impaired consciousness will follow. Further rise in ICP will result in brain herniation (coning). Impending coning is associated with hypertension, bradycardia and irregular breathing. The patient will have decerebrate posture and start to have pupillary changes. Without urgent intervention, irreversible coning and death will follow within minutes.[8–10]

The classical presentation of normal pressure hydrocephalus is the triad of gait impairment, dementia and incontinence.

Impairment of gait is the most readily recognized feature of idiopathic normal pressure hydrocephalus. It has been variably described as apractic, bradykinetic, glue-footed, magnetic, parkinsonian, short-stepped, and shuffling. Weakness of the legs is not usually evident on neurological examination.

Increased frequency and urgency without actual urinary incontinence may be seen in early stages of the disorder.

Progression to frank urinary incontinence usually occurs with disease progression.

The principal cognitive symptoms seen in idiopathic normal pressure hydrocephalus are suggestive of a subcortical process, mainly involving frontal lobe functions, such as attention, psychomotor speed, verbal fluency and executive functions. Recognition memory is relatively preserved compared with recall. In patients with severe idiopathic normal pressure hydrocephalus, cognitive impairment, the impairment of attention, psychomotor speed, literal fluency, and executive function is disproportionately severe, whereas the impairment of memory and orientation is disproportionately mild compared with Alzheimer's disease.[12]

Investigations

In infants *ultrasonography* can be used to image the brain through the open anterior fontanelles. It can detect haemorrhage

