# CASE FOUR

**Short case number: 3_12_04**
**Category: Children & Young People**
**Discipline: Paediatrics Surgery**
**Setting: Hospital ward**
**Topic: Large Heads_hydrocephalaus**

## Case

**You are the intern covering the wards for the evening. Nine month old Mia Samuels is 2 days post operative following the insertion of a ventrico-peritoneal shunt because of hydrocephalus. You are asked to see her because she is unsettled and has vomited twice.**

## Questions

1. You realise that you can't quite recall what hydrocephalus is, so decide to quickly 'look it up' before you go to see her. Outline the anatomy of brain and ventricles and the physiology of CSF production and flow, explaining the abnormalities that occur in hydrocephalus.
2. On arrival on the ward, you review Mia's medical records and note that she was referred to the neurosurgeon because her head circumference has increased beyond the 97$^{th}$centile. What are the key clinical features that need to be assessed in the child with possible hydrocephalus? How does the clinical presentation differ in older children?
3. What investigations are often used to assess hydrocephalus and why?
4. You are assessing Mia, when her mother returns to the ward. She asks you if there is a problem with the shunt. Outline the anatomy of a ventrico-peritoneal and ventrico-atrial shunt and explain the complications that can occur and how these present clinically.

**Suggested reading:**


- Flett, P., & Russo, R. (2012) Neural tube defects, large heads and hydrocephalus. In South, M. & Isaacs, D. (Eds) *Practical Paediatrics (pp613 – 624).*Edinburgh: Churchill Livingstone/ Elsevier.

## ANSWERS

1. There are 4 ventricles within the brain; 2 lateral ventricles, third ventricle and fourth ventricle. The l*ateral ventricles are two curved shap*ed c[av]ities located within the cerebrum, separated by a thin medial partition (septum pellucidum).
   They protect the brain from trauma and provide a pathway for the circulation of CSF. The third ventricle is a narrow cavity located between the two hemispheres of the diencephalon and serves as a route for transportation of CSF from the lateral ventricles to the fourth ventricle. The fourth ventricle is a diamond shaped cavity located behind the pons and medulla oblongata, it extends into the medulla oblongata and becomes continuous with the central cord of the spinal cord. It protects the brain from trauma.
   The choroid plexus is a cluster of specialized capillaries enclosed by specialized cells that line the brain ventricles, it is located behind the pons and medulla oblongata, produces cerebrospinal fluid and provides a barrier between blood and CSF.

   CSF cushions delicate neural structures, supports the brain and transports nutrients, chemical messengers and waste products.
   Normal CSF production is 0.20 – 0.35 ml/min, most CSF is produced in the choroid plexus with a small amount derived from the 'weeping' (or transmission) of tissue fluid by the brain into the ventricles. Normal route of CSF from production to clearance is the following: From the choroid plexus, the CSF flows to the lateral ventricle, then to the interventricular foramen of Monro, the third ventricle, the cerebral aqueduct of Sylvius, the fourth ventricle, the 2 lateral foramina of Luschka and 1 medial foramen of Magendie, the subarachnoid space, the arachnoid granulations, the dural sinus, and finally into the venous drainage.

   Hydrocephalus refers to a group of conditions characterized by:
   • an increase in cerebrospinal fluid (CSF) volume
   • ventricular dilatation
   • elevation of intraventicular pressure

   Hydrocephalus occurs when there is an imbalance between the formation and absorption of CSF. Impaired absorption is almost always due to some degree of obstruction along the CSF pathways. If the passage of CSF is obstructed within the ventricular system, the resultant hydrocephalus is labelled *non-communicating,* while if obstruction exists in the surface pathways, the hydrocephalus is described as being *communicating.*

   **Non-communicating**

   **Aqueduct stenosis or atresia**
   • Commonest site of intraventricular obstruction in infants with congenital hydrocephalus
   • May occur as an isolated anomaly or be associated with myelomeningocoele and the Arnold-Chiari malformation
   Sporadic
   Familial
   • inherited as a sex-linked trait, features include a short flexed thumb, mental retardation and other cerebral abnormalities.

   **Obstruction at the fourth ventricle**

   - Dandy-Walker syndrome
   - Cystic dilatation of the fourth ventricle, with cerebellar hypoplasia, other structural brain
     anomalies may also occur.
   - Associated with atresia of the exit foramina of the fourth ventricle

- Hydrocephalus may be present at birth or may develop subsequently
- Diagnosis is suggested in typical cases by the shape of the skull and the presence of cerebellar signs
- Arachnoiditis

**Obstruction due to intracranial mass lesions**

- Should always be considered in any child where head enlargement develops in late infancy
- or childhood
- Neoplasms, cysts
- Childhood tumours usually arise in the posterior cranial fossa and include medulloblastoma, astrocytoma and ependymoma
- Intracranial pressure develops early, because of their close proximity to the fourth ventricle
- Ataxia, incoordination, nystagmus and papilloedema are suggestive of the diagnosis
- Differential diagnosis includes craniopharyngioma, gliomas, pineolomas and arachnoid
- cysts
- Haematoma
- Galenic vein aneurysm

**Ventricular inflammations (rare)**

**Communicating**

**Arnold-Chiari malformation**

With myelomeningocoele (Type 2)

Without myelomeningocoele (Type1)

- Consists of:
    - downward displacement and elongation of the hind brain herniation of the medulla, cerebellar vermis and inferior part of the fourth ventricle into the upper cervical canal
- CSF flow is impaired, usually within the subarachnoid space
- Hydrocephalus usually develops in early infancy
- Frequently associated with cranium bifidum, myelomeningocoele and hydromyelia

**Encephalocoele**

**Meningeal adhesions**

Post inflammatory

Post haemorrhagic

- May be secondary to neonatal meningitis (post inflammatory adhesions), or intraventricular or subarachnoid haemorrhage
- Hydrocephalus is common, and is usually communicating
- Neurological deficit, developmental delay and seizures are usually the result of the infective process, but the hydrocephalus, if not relieved, will aggravate the brain injury.

**Choroid plexus papilloma**

- A rare cause of hydrocephalus
- Hydrocephalus is produced by excessive fluid secreted by the tumour, sometimes with obstruction to CSF flow
- Recurrent haemorrhage from the tumour may play a role
- Total excision of the tumour usually leads to a resolution of the hydrocephalic process

2. Clinical features of hydrocephalus are influenced by the following:
   - patient's age
   - cause
   - rapidity of onset
   - location of obstruction
   - duration

   Symptoms in infants
   - poor feeding
   - irritability
   - reduced activity
   - vomiting

   Symptoms in children
   - slowing of mental capacity
   - headaches (initially in the morning) that are more significant than in infants because of skull rigidity
   - neck pain suggesting tonsillar herniation
   - vomiting, more significant in the morning
   - blurred vision: this is a consequence of papilloedema and later of optic atrophy
   - double vision: this is related to unilateral or bilateral sixth nerve palsy
   - stunted growth and sexual maturation from third ventricle dilatation: this can lead to obesity and to precocious puberty or delayed onset of puberty
   - difficulty in walking secondary to spacticity: this affects the lower limbs preferentially because the periventricular pyramidal tract is stretched by the hydrocephalus
   - drowsiness

Classically, hydrocephalus is recognized by a progressive increase in occipitofrontal head circumference out of proportion to other bodily dimensions. A single head circumference measurement that greatly exceeds the 97$^{th}$ percentile strongly suggests the existence of hydrocephalus. Where head enlargement is equivocal, and neurological abnormality is absent, serial head measurements will often indicate the need for further diagnostic studies. It must be emphasized that, once enlargement of the skull is clinically obvious, the ventricles are already grossly dilated and the cerebral cortex is thinned.

Clinical signs that frequently precede obvious enlargement of the head include:
- a large and bulging fontanelle
- thinning of the bones of the calvarium
- widening of the coronal, sagittal and lambdoidal sutures

With advancing hydrocephalus:
- the scalp thins and becomes shiny and pale
- there is upward retraction of the eyelids
- the eyes are fixed in a downward gaze (the 'setting sun' sign)
- hair appears sparse
- superficial scalp veins become distended
- the brow overhangs the small, triangular face

The shape of the skull should be noted. A large protruding occiput is typical of a Dandy-Walker cyst, while an asymmetrical head may be due to unilateral obstruction at the foramen of Munro. In addition, auscultation for cranial bruit should be performed over the eyeballs and over the calvarium.

In children other signs include
- failure of upward gaze: this is due to pressure on the tectal plate through the supraspineal recess. The limitation of upward gaze is of supranuclear origin. When the pressure is severe, other elements of the dorsal midbrain syndrome (i.e. Parinaud syndrome) may be observed, such as light-near dissociation, convergence-retraction nystagmus, and eyelid retraction (Collier sign)
- Macewen sign: a'cracked pot' sound is noted on percussion of the head.

In the older child with 'arrested' hydrocephalus, it is important to evaluate the mental and psychological status. These children are frequently talkative, jovial and euphoric ('cocktail party syndrome') but their capacity for concentration, language comprehension and abstract thinking is often lacking.

3. In all cases of hydrocephalus investigations are required to confirm the diagnosis, determine the extent of the disorder and if possible define the aetiology. Investigations are also of assistance in deciding the need or otherwise for active treatment and also as a means of assessing the success or otherwise of treatment. The plain skull X-ray may be a useful initial investigation.

*Ultrasound*

The widespread use of ultrasound scanning has in recent times greatly facilitated the assessment of infants with suspected hydrocephalus. Real-time ultrasound imaging through the open fontanelle provides a clear demonstration of the ventricles and may define other structural anomalies. This non-invasive risk-free investigation can be undertaken with little or no sedation and can be repeated as often as required. Ultrasound examination during pregnancy can indicate whether the foetus has hydrocephalus.

*Computed tomography*

In the older child, and occasionally in infants where more detail is required, computed tomography (CT) scanning is the investigation of choice. This technique provides excellent detail of the intracranial anatomy and the images may be enhanced by the injection of contrast material.

*Magnetic resonance imaging*

MRI is rarely undertaken as a primary investigation but may be of value in defining the cause of the condition. It can evaluate for Chiari malformation or cerebellar or periaqueductal tumours. It affords better imaging of the posterior fossa than CT.

CT/MRI criteria for acute hydrocephalus include the following:

· size of both temporal horns is greater than 2mm, clearly visible. In the absence of hydrocephalus, the temporal horns should be barely visible.
· ratio of the largest width of the frontal horns to maximal biparietal diameter (i.e. Evans ratio) is greater than 30% in hydrocephalus
· transependymal exudate is translated on images as periventricular hypoattenuation (CT) or hyperintensity (MRI T2-weighted)
· ballooning of frontal horns of lateral ventricles and third ventricle (i.e.., 'Mickey Mouse' ventricles) may indicate aqueductal obstruction.
· upward bowing of the corpus callosum on sagittal MRI suggests acute Hydrocephalus

CT/MRI criteria for chronic hydrocephalus include the following:

· temporal horns may be less prominent than in acute hydrocephalus
· third ventricle may herniate into the sella turcica
· sella turcica may be eroded
· macrocrania (i.e., occipitofrontal circumference >98$^{th}$ percentile) may be Present
· corpus callosum may be atrophied (best appreciated on sagittal MRI)

4. The indications for treatment are based on a clear understanding of the natural history of the disorder. Three patterns may be described:
   • the process continues, followed by neurological deterioration
   • the process progresses to a point, then stabilizes ('compensated hydrocephalus')
   • the process is temporary

In the majority of patients, the ventricles will continue to enlarge and the overlying brain will become stretched, compressed and thinned.
The definitive treatment of hydrocephalus is a surgical procedure. The usual method of treatment is by a shunt that diverts the CSF to some other site in the body.

*Ventriculoperitoneal shunt.* This is the operation performed most frequently in paediatric patients with hydrocephalus. A Silastic catheter is placed in a lateral ventricle through a burr hole and the other end of the tube is passed subcutaneously to the abdomen and then placed in the peritoneal cavity. A valve is interposed and an adequate length of tube is placed in the peritoneal cavity to allow for growth. The peritoneum absorbs CSF effectively.

*Ventriculoatrial shunt*. In this procedure the lower end of the shunt is passed via a neck vein to the right atrium. The catheter is so designed so that CSF can pass from the catheter tip but blood cannot flow back into the lumen. The turbulent blood flow in the atrium prevents thrombus formation around the catheter. This operation is not undertaken often in childhood as maintenance may involve the lengthening of the atrial catheter on several occasions.
A ventriculopleural shunt is considered second line. It is used if other shunt types are contraindicated.

*Complications of ventricular shunts*.
The operation is generally well tolerated with infrequent early difficulties.
Common complications include meningitis, ventriculitis, and shunt obstruction.
The most common presentation of a child with a blocked shunt is that of a vague illness. Irritability and vomiting are frequent and headache may be present. The symptoms are very similar to those of many childhood illnesses and difficulties are often experienced in trying to decide whether the symptoms are a consequence of shunt malfunction or an unrelated illness. Definite signs of raised intracranial pressure, if present, are of great assistance but are often not ascertained readily. Palpation of the shunt mechanism may also frequently be inconclusive.
The treatment of shunt obstruction is usually a simple procedure and involves the replacement of the defective component. However, a small number of patients suffer from repeated episodes of obstruction and management can be difficult and may involve many variations of shunt equipment and surgical technique.