Hydrocephalus

Description

An increase in the volume of cerebrospinal fluid (CSF) occupying the cerebral ventricles secondary to either impaired absorption or, less commonly, increased production of fluid.

Terms Used When Describing Hydrocephalus

- **Communicating** This implies communication between the ventricles and the subarachnoid space. Usually due to ↑CSF production, occasionally due to ↓absorption or ↓drainage
- **Non-communicating** CSF flow obstructed within the ventricles or between the ventricles and the subarachnoid space.
- **Normal pressure hydrocephalus** The CSF pressure remains normal or is only intermittently raised. Failure to reabsorb the CSF is compensated by reduced production. The condition may be congenital (+myelomeningocoele or –myelomeningocoele [infantile hydrocephalus]) or acquired.
- **Hydrocephalus ex vacuo** This is seen in conditions associated with cerebral atrophy and shrinkage, such as Alzheimer's Disease and Pick's Disease. The ventricles expand and there is no increase in CSF pressure.

Epidemiology

Congenital hydrocephalus prevalence 82 per 100,000 live births
Acquired hydrocephalus incidence is unknown.

Causes

Congenital causes in infants and children

- Stenosis of the aqueduct of Sylvius
- Dandy-Walker malformation/Arnold-Chiari malformation type 1 and type 2
- Agenesis of the foramen of Monro
- Congenital toxoplasmosis
- Bickers-Adams syndrome (stenosis of the aqueduct of Sylvius, severe mental retardation, and in 50% by an adduction-flexion deformity of the thumb)

Acquired causes in infants and children

- Mass lesions (20% of all cases in children, e.g. medulloblastoma, astrocytoma)
- Intraventricular haemorrhage (e.g.prematurity, head injury, or rupture of an AVM)
- Infections - meningitis, cysticercosis in some areas
- ↑Venous sinus pressure - related to achondroplasia, craniostenoses, venous thrombosis
- Iatrogenic - e.g. hypervitaminosis A
- Idiopathic

Causes of hydrocephalus in adults

- Subarachnoid haemorrhage
- Idiopathic (one third of cases)
- Head injury
- Tumours
- Iatrogenic - posterior fossa surgery
- Congenital aqueductal stenosis (may not be symptomatic until adulthood)
- Meningitis, especially bacterial
- Normal pressure hydrocephalus
- All causes of hydrocephalus described in infants and children

Presentation

Features in Infants

- Poor feeding, irritability, reduced activity, vomiting
- Rapid increase in head circumstance or head circumference ≥98th centile
- Dysjunction of sutures, dilated scalp veins, tense fontanelle
- Setting-sun sign
- Increased limb tone

Features in Older Children and Adults

- Papilloedema
- Failure of upward gaze
- Macewen sign (a "cracked pot" sound on percussing the head)
- Unsteady gait due to spasticity in the legs
- Large head (although the sutures are closed, the skull still enlarges due to chronic ↑ICP)
- Unilateral or bilateral sixth nerve palsy secondary to increased ICP

Other Features Specific to Adults

- Cognitive deterioration
- Neck pain
- Nausea and vomiting
- Blurred and double vision
- Incontinence

Differential Diagnosis

- Brainstem gliomas
- Causes of sudden visual loss
- Childhood migraine variants
- Craniopharyngioma
- Frontal lobe epilepsy
- Frontal lobe syndromes
- Frontal and temporal lobe dementia
- Glioblastoma multiforme
- Intracranial epidural abscess or bleed
- Meningioma
- Mental retardation
- Oligodendroglioma
- Pituitary tumours
- Primary CNS lymphoma
- Pseudotumor cerebri
- Subdural empyema or haematoma

Investigations

Skull X-ray

Neuroradiology - CT or MRI

Generalised ventricular dilation suggests communicating hydrocephalus. Dilation of the lateral & 3rd ventricles suggests an aqueduct stenosis, if 4th ventricle normal;or a post. fossa mass if not.

Ultrasound - through the ant fontanelle used in infants.

Management

Drugs

Temporizing measure: Frusemide and acetazolamide ↓secretion of CSF. Isosorbide ↑absorption.

Surgery

Treat the underlying cause The cure rate is 80% if a tumour can be identified.

Repeated lumbar punctures is only valid in communicating hydrocephalus.

Insertion of a shunt is common e.g. ventriculoperitoneal shunt.

Other surgical procedures Choroid plexectomy, choroid plexus coagulation, and endoscopic cerebral aquaductoplasty. Endoscopic fenestration of the 3rd ventricle floor in non-communic

Prognosis

Untreated congenital hydrocephalus is often fatal within the first four years of life. The prognosis in other conditions depends on the underlying cause.