Post haemorrhagic hydrocephalus

**Introduction**

Post haemorrhagic hydrocephalus (PHH) is defined as progressive ventriculomegaly caused by disturbances in cerebrospinal fluid flow or absorption following intraventricular haemorrhage.

True hydrocephalus must be distinguished from:

- Transient ventriculomegaly: this is common following IVH but resolves completely within 4 weeks.\(^1\)
- Static ventriculomegaly: this occurs secondarily to the loss of cerebral tissue either because of the destruction, or the failure of development, of cerebral white matter.\(^2\)

**Incidence and risk factors:**

The incidence is probably declining.

Fernell et al 1994 \(^3\) report in an epidemiological study of infantile hydrocephalus that 17/1000 (1.7%) infants of <32 weeks gestation developed post haemorrhagic hydrocephalus. The incidence of post haemorrhagic hydrocephalus requiring shunt insertion in infants <32 weeks gestation in John Spence Nursery in the last five years (1994-1998) was 6/750 (0.8%).

**Natural history**

The natural history of hydrocephalus can usually be considered as acute or sub acute.

- Acute hydrocephalus develops very rapidly after IVH and is probably a direct consequence of blood clots blocking CSF flow.
- Sub acute hydrocephalus develops more insidiously and may not be apparent for several weeks after IVH. This is thought to be the consequence of an obliterative arachnoiditis in the posterior fossa reducing CSF resorption.

**Consequences:**

The incidence of major neuro-developmental handicap in this group is high. A 30 month follow up study reports multiple handicap rates of between 50% - -70% with the higher rates in those infants with associated parenchymal damage.\(^9\)

**Diagnosis:**

Diagnosis of PHH is by clinical assessment and ultrasound evaluation. Particular attention should be paid to those infants who have sustained an IVH.

**Clinical evaluation**

Possible clinical signs are:

- Head circumference crossing centile lines or enlarging at over 1.5 cm per week
- A tense anterior fontanelle
- Separation of the cranial sutures
- Symptoms of rising intracranial pressure (apnoea, vomiting, abnormal posture)
Ultrasound evaluation

The role of ultrasound:

- Early identification of evolving hydrocephalus
- Confirmation of suspected hydrocephalus
- Monitoring the progression of hydrocephalus
- Differential diagnosis of transient ventriculomegaly and static ventriculomegaly

Infants with hydrocephalus should be monitored as follows:

1. Serial occipital frontal head circumference measurements

All babies in the nursery should have weekly head circumference measurements plotted on growth charts appropriate for their gestational age. Plotted head circumference measurements deviating upwards away from their initial centile or increasing at >1.5cm per week should alert the clinician to the possibility of rapidly evolving hydrocephalus. In such situations when hydrocephalus is confirmed by cranial ultrasound measurements should be increased to twice weekly.

2. Ultrasound monitoring and measurement of the ventricular index

All infants with hydrocephalus should have weekly cranial ultrasounds.

Measurement of the ventricular index (distance from the falx to the lateral most aspect of the lateral ventricle) can be made by cranial ultrasound and compared with gestationally appropriate standards (see graph). At present there is no evidence that such measurements provide useful prognostic information with regard to the need for shunt insertion. However, these measurements are being employed in research protocols and may be useful in identifying the optimal time for shunt insertion in the future.

Ventricular index should be recorded and plotted against Levene’s standards in any infant being monitored for hydrocephalus.
Research observations with potential clinical roles

Doppler studies

Decreasing diastolic velocities and an increased pulsatility index have been reported on cerebral artery Doppler waveforms in infants with hydrocephalus. However the methodology used in various studies has varied considerably and there is not yet any convincing evidence that these studies give any more useful information than plotting head circumferences on a chart.

Sensory evoked potentials

Sensory evoked potentials are altered or abolished in the hydrocephalic rat. Their clinical role in intracranial monitoring in adults has been assessed, but as of yet their role in monitoring the hydrocephalic infant has not been defined.

Interventions:

The only intervention currently undertaken in the John Spence Nursery is the surgical insertion of a ventricular reservoir or a ventriculoperitoneal shunt. The decision to intervene is made by a paediatric neurosurgeon and is based on clinical and ultrasound evaluation.

Other interventions have been described; a brief summary follows

Repeated ventricular or lumbar taps

The effect of repeated ventricular or lumbar taps compared with conservative management of infants with post haemorrhagic hydrocephalus have been studied in four controlled trials. Meta-analysis of these studies gives pooled odds ratios for each of shunt placement, death, disability and multiple disability very close to 1.0. This intervention also appears to be associated with an increased risk of CSF infection. Therefore early repeated CSF tapping cannot be recommended for neonates at risk of, or actually developing, post haemorrhagic hydrocephalus.

Post haemorrhagic intraventricular streptokinase

Multiple blood clots in the CSF causing blockage of the channels of CSF resorption are thought to be the initial cause of post haemorrhagic ventricular dilatation. Therefore it has been hypothesised that lysis of clots by intraventricular injections of thrombolytic agents may reduce the development of hydrocephalus. There has only been one randomised trial of intraventricular fibrinolytic therapy which showed no significant difference in the numbers of infants dying or requiring a peritoneal shunt. The study was small and treatment commenced at a median postnatal age of 12 days so larger study with earlier intervention may be warranted. The data at present do not support the use of thrombolytic agents.

Drug treatment

Acetazolamide and frusemide are widely used in the treatment of post haemorrhagic hydrocephalus and are thought to reduce the production of CSF. A recent international randomised controlled trial assessing their role in the management of post haemorrhagic hydrocephalus showed no benefit in terms of mortality, shunt insertion or disability at 1 year. Preliminary results actually suggest that the use of frusemide and acetazolamide may be associated with higher rates of shunt insertion and neurological morbidity. Their use cannot be recommended.

Ventriculostomy

The role of ventriculostomy in the management of post haemorrhagic hydrocephalus has not been defined.
### Key Points

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<tr>
<th>Key Points</th>
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<td>The incidence of post haemorrhagic hydrocephalus is declining</td>
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<td>The incidence of associated neurodevelopmental handicap is high</td>
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<td>Infants with developing hydrocephalus should have weekly cranial ultrasounds and ventricular index measurements</td>
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<td>Repeated post haemorrhagic ventricular or lumbar taps are not beneficial</td>
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<td>Post haemorrhagic intraventricular streptokinase is not beneficial</td>
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<td>Drug treatment of post haemorrhagic hydrocephalus with frusemide or acetazolamide is not beneficial</td>
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### References


2. Leviton A, Gilles F Ventriclemegaly, delayed myelination, white matter hypoplasia and "periventricular leukomalacia": How are they related ? 1996 *Paeditr Neurol* 15:127


4. Levene MI Measurement of the growth of the lateral ventricle in preterm infants with real time ultrasound *Arch Dis Child* 1981. 56:900-904


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