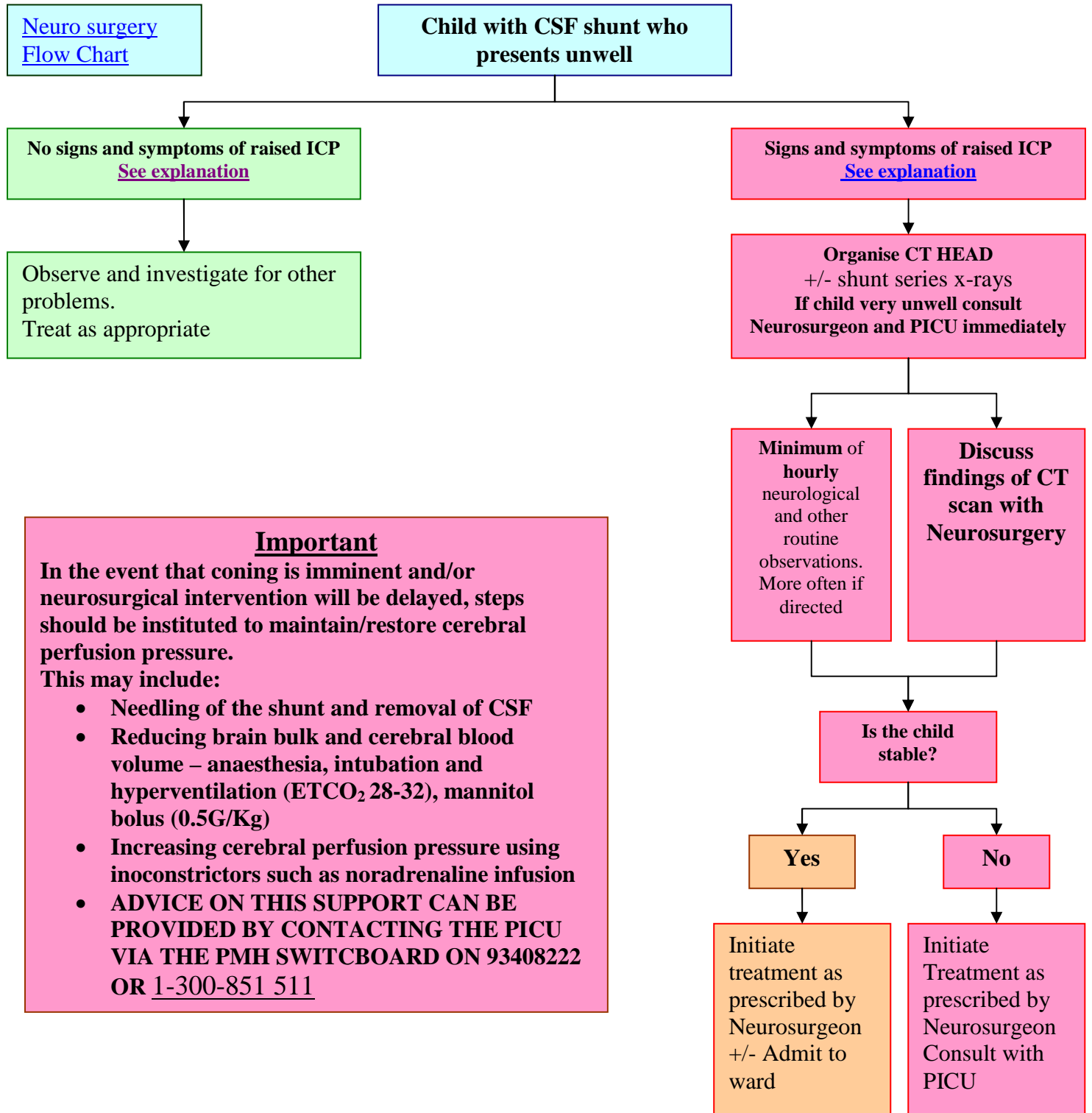


Hydrocephalus and CSF Shunt Blockages

Emergency Department Protocol



Hydrocephalus is **NOT** a single disease entity. It is either due to subnormal CSF re-absorption, obstruction along the flow pathways or very rarely increased production.

It may be:

Congenital (e.g. myelomeningocele, Dandy Walker syndrome, stenosis of the aqueduct of Sylvius)

Or

Acquired (e.g. post meningitis, post hemorrhagic, obstruction due to tumour.)

Treatment options include

- Insertion of a CSF diversion shunt (the main treatment modality)

Or

- Endoscopic third ventriculostomy (internal diversion via perforating the floor of the third ventricle allowing CSF to flow from the 3rd ventricle directly to the cortical subarachnoid space)

BOTH shunt systems are prone to blockages.

Early and prompt detection of shunt blockage is vital as delay can lead to markedly raised intracranial pressure, coning and death.

Presentation

Parents **may** know what is usual for their child in the event of a blockage. The importance of paying careful attention to the observations of the child's parents, and other carers, particularly if they have had experience of shunt blockage in the past, cannot be overemphasised.

History and symptoms may be **variable**:

- Headache
- Drowsiness
- Vomiting
- Irritability

Less commonly:

- seizures,
- abdominal problem (tenderness, distension or peritonism),
- fever,
- lethargy.

Intermittent shunt dysfunction/blockage may lead to a more protracted time course with chronic headaches, cognitive decline or developmental standstill.

Two prospective studies of 104 admissions showed that drowsiness was by far the best, BUT not a definite clinical predictor of a shunt blockage. Headache, vomiting, irritability and duration of symptoms were less predictive as to whether the child's shunt was actually blocked.

The presence of pyrexia may suggest the shunt is malfunctioning because it is infected OR an alternative diagnosis.

The presence of drowsiness, headache and vomiting together make it very likely that the patient has shunt dysfunction.

In a shunted child, presenting unwell, shunt blockage must be a differential and not excluded until proven otherwise – **EARLY CONSULTATION WITH THE NEUROSURGICAL TEAM IS ADVISED.**

Impaired or falling Glasgow Coma Scale, bradycardia, papilloedema OR sun setting eyes all imply dangerously elevated intracranial pressure. This constitutes a neurosurgical emergency.

IMMEDIATE REFERRAL TO THE NEUROSURGICAL TEAM IS MANDATORY.

Call the on-call Neurosurgery team at PMH on 9340 8222. If, in the event of such an emergency and there is a delay on this phone number, you can also contact the on-call neurosurgery registrar at SCGH 9346 3333 for advice.

Examination of shunt system

Shunt evaluation is diagnostically unreliable. Any abnormal shunt findings should be discussed with the neurosurgery team.

In a child with an open fontanelle, this should be soft and pulsatile. Fluid tracking along subcutaneous shunt tubing may also indicate shunt blockage.

Investigations that aid in diagnosis are

- Brain CT – to detect result of shunt malfunction: Enlarged ventricles may imply shunt obstruction

A scan showing obviously dilated ventricles **when compared with previous scans** is a definite indication of shunt blockage/hydrocephalus

A “normal” CT scan **without a previous scan DOES NOT** reliably exclude the diagnosis of a shunt blockage/hydrocephalus. The child may have **slit ventricles**. In this circumstance, as the child is to the right of the compliance curve, a small increase in volume (subtle/ minimal change in ventricle size) will result in a large increase in ICP.

- Plain X-ray/ Shunt series – may demonstrate a disconnection
- CSF Sampling – this should only to be done by the neurosurgical team or after consultation with neurosurgery.
- FBC and CRP may help in elucidating infection/ shunt dysfunction.

Management

The child should be observed with hourly **neurological** and **Glasgow Coma Scale** assessments until definite diagnosis is made. Hourly blood pressure and pulse observations should also be recorded.

If blocked or infected, the shunt will require revision/removal – URGENT CONSULTATION WITH THE NEUROSURGICAL TEAM IS REQUIRED.

In the event that coning is imminent and/or neurosurgical intervention will be delayed, steps should be instituted to maintain/restore cerebral perfusion pressure.

This may include:

- **Needling of the shunt and removal of CSF**
- **Reducing brain bulk and cerebral blood volume – anaesthesia, intubation and hyperventilation (ETCO₂ 28-32), mannitol bolus (0.5G/Kg)**
- **Increasing cerebral perfusion pressure using vasoconstrictors such as noradrenaline infusion**

- **ADVICE ON THIS SUPPORT CAN BE PROVIDED BY CONTACTING THE PICU VIA THE PMH SWITCHBOARD ON 93408222 OR 1-300-851 511**

References:

D Thompson. Shunt Blockage – Clinical guidelines for Great Ormond Street Hospital for Children, NHS Trust. http://www.ich.ucl.ac.uk/clinical_information/clinical_guidelines/cm_g_guideline_00006.

Watkins L, Hayward R, Andar U, Harkness W. The diagnosis of blocked cerebrospinal fluid shunts: a prospective study of referral to a paediatric neurosurgical unit. *Childs Nerv Syst* 1994; 10:87-90

Barnes NP, Jones SJ, Hayward RD, Harkness WJ, Thompson D. Ventricularperitoneal shunt block: what are the best predictive clinical indicators? *Arch Dis Child* 2002; 87:198-201.

