Diagnosis and Management of Post Haemorrhagic Ventricular Dilatation (PHVD)

Introduction:
Post Haemorrhagic Ventricular Dilatation (PHVD) is challenging to manage due to the lack of evidence regarding optimal treatment and the threshold and timing of any intervention. A recent review highlights wide variations in the approaches to diagnosis, treatment threshold and mode of intervention between neonatal centres.  

Background:
Intraventricular haemorrhage (IVH) is a well recognised complication of prematurity and PHVD occurs in about 50% of infants with severe IVH (Grade III-IV according to the old classification). Typically, mild or uncomplicated IVH (Grade I-II according to the old classification) is unlikely to lead to PVHD. However, the risk of PHVD increases if the IVH is severe: post Grade III the risk is 78% and post Grade IV 53% respectively. Around half of all neonates that have any IVH will not go on to have ventricular dilatation, 25% will have non progressive dilatation probably secondary to white matter damage and cerebral atrophy. Thus, only 25% of all neonates with any IVH will ultimately have progressive ventricular dilatation requiring treatment. Moreover, in 2/3rds of these cases the ventricular dilatation will spontaneously arrest following treatment (usually serial lumbar punctures), hence only 10% of all cases of IVH will ultimately require CSF shunt insertion.  

PHVD results from progressive accumulation of CSF due to reduced reabsorption and blockage of flow caused by small blood clots and the subsequent chronic arachnoiditis. This leads to ballooning of the ventricles and distortion of the underlying developing brain with eventually rising pressures resulting in progressive periventricular white matter injury.  

Therapeutic interventions aim to reduce the number of VP shunts required and to improve neurodevelopmental outcome. Interventions include repeated lumbar punctures, ventricular taps, ventricular access devices, drug treatment to reduce CSF production, intraventricular fibrinolytic therapy, external ventricular drain, ventriculo-subgaleal shunt, third ventriculostomy and choroid plexus coagulation. However, there is currently no evidence to support any treatment option versus another.  

Diagnosis:
Ventricular dilation post IVH typically develops 10-20 days after the onset of haemorrhage and usually precedes the clinical symptoms of hydrocephalus. Clinical symptoms of raised intracranial pressure can be subtle and include raised/tense fontanelle, separation of head sutures, abnormal tone, irritability, poor feeding, apnoeas, seizures and eventually sun-setting eyes.  

Following an IVH serial cranial ultrasound scans should be performed at least twice weekly to monitor for PHVD. Once ventricular dilatation is noticed the ventricular index (VI) is measured and plotted on the reference chart by Levene (please see the Neonatal Cranial Ultrasound guideline for further information). Observation of the shape of the lateral ventricles e.g. if balloon shaped can suggest hydrocephalus.  

Repeated measurements of the resistive index (RI) (please see the Neonatal Cranial USS guideline) of the anterior cerebral artery may also help in the assessment of raised intracranial pressure. As the ICP increases it decreases the end diastolic velocity, thus causing the RI to increase towards 1.0. Remember that the newborn skull is compliant and so the RI may not always be sensitive for raised intracranial pressure. Raised RI is also not specific for raised ICP; in particular in this context in the presence of a PDA which also may elevate the RI.  

The head circumference (OFC) should be measured and plotted at least every other day. Head circumference growth will be accelerated if the CSF pressure is elevated but be aware that it will often lag behind ventricular enlargement by 1-2 weeks. Normal OFC growth is 1mm/day between 26 and 32 weeks and 0.7mm/day from 33 weeks until term. An increase of >4mm/day over 2 days or if measuring weekly 14mm over 7 days is likely to be excessive.
Measuring the CSF pressure may be helpful, and if > 9mmHg, suggests raised intracranial pressure or hydrocephalus suggesting that CSF should be drained. However, we do not routinely do this on our unit.

Threshold of Intervention:
If a conservative approach is adopted the threshold of intervention for progressive PHVD is reached when an infant presents with rapidly increasing head circumference of >2mm/day and/or displays clinical signs of raised intracranial pressure. Most units also take into account the ventricular index (VI) and initiate treatment once the VI is above the 97th centile +4mm line.

However, some units will intervene earlier at a lower VI measurement of the 97th Centile due to concerns that waiting for the VIs to increase to the 97th centile +4mm level before intervening may be too late. There is some evidence to support this approach (albeit retrospective). With early intervention the rate of shunt insertion is 16% vs 62% and moderate or severe neurodevelopment delay is reduced from 26% to 16% A randomised control trial ELVIS (The Early versus Late Ventricular Intervention Study ELVIS) is currently investigating if earlier intervention improves outcome and will report in due course (currently half way through recruitment).

Initial and Further Management:
Once the intervention threshold is reached, the mainstay of treatment is reducing the CSF pressure through drainage of CSF via lumbar puncture. CSF pressure should be measured and CSF sent for biochemical analysis and microscopy and culture. At least 10ml/kg of CSF should be removed at a rate of 1ml/kg/min. If the patient is very symptomatic or is requiring very frequent LPs, occasionally 20mls/kg of CSF may be withdrawn from the patient. CSF drainage via LP can be a “one off” or repeated serially.

If LPs are unsuccessful in draining CSF, usually due to of non-communication between the spinal canal and the ventricles neonates should be referred to the neurosurgical team for consideration of insertion of a ventricular access device (VAD) (Ommaya or Rickham reservoir). Rarely ventricular taps may be done as a “one off” on the unit, usually as a bridge to surgery for insertion of a CSF reservoir insertion. Ventricular taps should NOT be used as a routine method for CSF drainage as repeated ventricular taps cause intraparenchymal needle track injury.

Once a VAD is in situ this should be used for repeated CSF tapping to control intracranial pressure and head growth, under the supervision of the neonatal team. Each tap should be undertaken using strict aseptic precautions and samples sent for biochemistry and culture. The frequency of tapping the VAD depends on the patient and the clinical situation but usually is once or twice a week. If the patient is requiring CSF tapping more than twice a week, please discuss with the neurosurgical team.

If there is a prolonged need for tapping to maintain normal head growth or persistent excessive head enlargement insertion of a Ventriculoperitoneal shunt should be considered. However, this requires for the patient to weigh more than 2kg, and for the CSF protein to be less than 1.5mg/l.

References:
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