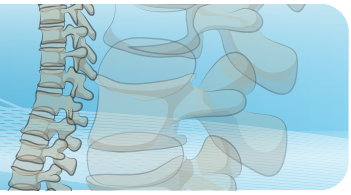


Hydrocephalus

Hydrocephalus is present in more than 80% of children with spina bifida. In these children, the hydrocephalus is caused by the Arnold Chiari Malformation. This is due to the hindbrain herniating through the foramen magnum and into the upper cervical canal, causing the normal flow of Cerebrospinal Fluid (CSF) from the 4th ventricle into the spinal canal to be obstructed, thus resulting in overfilling of the ventricles. Most often signs of hydrocephalus become more noticeable after the lesion is closed and drains are removed, as there is no longer an outlet for the CSF to leak (McLone and Dias 2003). Approx 80-90% of children with spina bifida and hydrocephalus will require a ventriculo-peritoneal shunt (usually 1-2 weeks following back closure) to help reduce intracranial pressures (Shaer 1997, Wagner et al 2002, Rintoul et al 2002).

Johnson et al (2006) proposed that dilated ventricles during early brain development, even if hydrocephalus is no longer active, may alter or injure corticospinal pathways and thus lead to weakness or spasticity, further exacerbating fine and gross motor deficits. They found that unrecognised increases in intracranial pressure (ICP) associated with chronic ventricular dilation, can have a negative impact on cognitive and neurodevelopmental outcome.

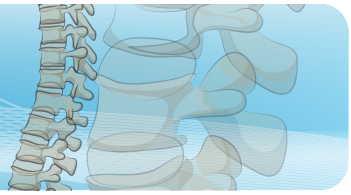
A study by Mataro et al (2000) highlighted the importance of monitoring ICP in patients with assumed arrested hydrocephalus, because only a few of these patients will have truly arrested hydrocephalus. Furthermore, children who have more shunt revisions and infections of the CNS, such as meningitis and shunt infections, have been found to have greater learning difficulties and poorer intellectual functioning.

Hydrocephalus is diagnosed and monitored using.

- Progressive head circumference measurements
- Assessing for papilledema (swelling of the optic disc due to raised intracranial pressure)
- Ultrasound (before closure of sutures)
- Computerized axial tomography (CT) scan and
- Magnetic resonance imaging (MRI)

Hydrocephalus is generally managed by the insertion of a vetriculo-peritoneal (VP) shunt, which drains CSF away from the lateral ventricles (Stein and Schut 1979). It is now nearly 50 years since the first effective shunt was used, and this method of management has been generally available for more than 40 years. With the development of ventriculoscopic techniques (allowing the ventricles to be visualised), procedures such as third ventriculostomy to improve CSF flow, are sometimes being used. With the use of this and other techniques, it may be possible to avoid inserting shunts to manage hydrocephalus in some patients, and thus reduce the risk of shunt complications, however, evidence to support this remains inconclusive, particularly in the long term.

Rintoul et al (2002) showed that the incidence of shunting for hydrocephalus is associated with the level of the lesion, with a significantly lower rate of shunting for patients with better functional levels. They showed that related to functional level, 97% of thoracic, 87% of lumbar and 37% of sacral level MMC patients were shunted. In relation to bony level, 100% thoracic, 88% lumbar and 68% sacral level lesions required shunting.



VP shunting has a relatively high complication rate with Rintoul et al (2002) reporting that 55% of patients with a VP shunt required revision of the shunt within the first year of life. Longer term follow up studies have reported a failure rate of greater than 60% (Talamonti et al 2007) with the majority of those requiring 2 or more revisions (Talamonti et al 2007, Bowman et al 2001). Shunt obstruction is the main cause of failure, followed by infection (Talamonti et al 2007, Caldarelli et al 1996). Occasionally, a ventriculo-pleural or a ventriculo-atrial shunt will be used if a serious infection within the peritoneum prevents placing of the shunt in this area.

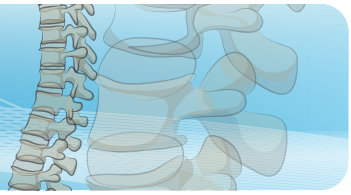
The high incidence of shunt failure in children with hydrocephalus further highlights the need for close monitoring of neurological signs by the physiotherapist.

Signs and Symptoms

Signs and symptoms of hydrocephalus or more specifically shunt failure can be overt or may be very subtle. The table below illustrates some of the more common and identifiable signs and symptoms of shunt failure. These need to be investigated carefully to aid in differential diagnosis of shunt failure from other reasons for neurological deterioration.

Table: Signs and Symptoms of increased intracranial pressure/shunt failure

Age	Signs of shunt failure
Newborn to 18 months	Tense or bulging fontanelle Increased head circumference Sunset eyes Irritability Poor feeding, vomiting Drowsiness Swelling along shunt tract Separation of sutures Prominent scalp veins Papilledema Seizures
Older Child or Adult	Irritability Personality Change Headache Nausea and vomiting Seizures Swelling along shunt tract Decreased coordination, balance Deterioration in school performance Pain in site repair Blurred vision Papilledema



Spasticity, scoliosis, reduced vital signs and opisthotonus may develop if the increased pressure isn't relieved.

The child with MMC and hydrocephalus may develop neurological deterioration due to shunt dysfunction. This deterioration may be gradual and subtle or more rapid and obvious. The physiotherapist should always be alert to potential signs of shunt dysfunction to assist in the prompt diagnosis and management and thus prevent permanent neurological damage.

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