

Slit Ventricle Syndrome

One of a series of fact sheets produced by the Family Support Service

Slit ventricle syndrome is a combination of symptoms which, for example, can appear in patients with a functioning shunt and in whom the brain has lost part of its elasticity. The symptoms consist of headaches, vomiting, drowsiness etc. Infuriatingly enough, these symptoms resemble those of shunt malfunction. The appearance of the symptoms is very cyclical, often with the regularity of a clock; for example, the patient is well for three weeks, then violently ill and sleepy for 24 hours and then well again. There is more often than not no cause for the symptoms, although a minor viral illness can produce them. Often the patient is taken to hospital and imaging is done which shows the ventricles to be small, or even unchanged, compared to previous imaging. To increase the frustration the symptoms usually disappear spontaneously.

WHAT ACTUALLY HAPPENS?

In ordinary circumstances, the shunt drains the cerebrospinal fluid (CSF) from the ventricles to a cavity in the body. In certain patients, this causes the ventricles to collapse. The ventricle closes on the ventricular catheter that drains the ventricles and blocks off the outflow of CSF. That's when the symptoms appear. The pressure in the brain rises very quickly and the patient becomes ill. However, as the brain has lost some of its elasticity due to the initial disease that caused the hydrocephalus the ventricles do not blow up rapidly and the symptoms can persist. After a while the ventricles get slightly bigger and the normal CSF drainage resumes. All these changes in shape and size may not be easy to detect and often the illness goes unexplained. Slit ventricles are small ventricles, sometimes so small that they are rarely visible on CT scan (Computer Tomography), or MRI Scan (Magnetic Resonance Imaging). Slit ventricles can occur after severe head injury or viral infection of the brain. In both conditions, the brain becomes so swollen that the fluid is pushed out of the ventricles. Slit ventricles can appear after cerebrospinal fluid diversion-shunts for example. One of the criteria for a happily functioning shunt is decompression of the ventricular system when compared to the previous CT scan or MRI. Inherent in any shunt system is the change in the pressure in the skull and brain. In some patients, this can lead to a siphon effect, much as when you siphon petrol from a car. What happens next

all depends on the resilience of the brain. At a very young age, the brain is very watery and will easily change shape. The ventricles will usually become smaller, sometimes up to a point that they become slit-like. This effect is unpredictable and very little can be done about it, although insertion of an anti-siphon device may help. It is important to know that not all small or slit ventricles cause symptoms. What proportion of patients with small ventricles develop slit ventricle syndrome is not really known. We know that the patients at risk of developing slit ventricle syndrome are those who were shunted at an early age and where a low drainage pressure shunt system was used. This does not imply inadequate treatment. It just means the effect of a shunt can be unpredictable.

WHAT CAN BE DONE?

This is a difficult question. The critical point is to make sure that the shunt is working properly. This means making sure there is no intermittent shunt blockage due to malfunction of the system. Often this can be done by measuring the pressure in the skull (intracranial pressure monitoring-ICP) and with extra imaging (CT scan or MRI). Once satisfied with the functioning of the shunt, there are several options available. One option is volume expansion procedures. This means that since the ventricles are small, the ventricle, and so the catheter inside the brain, can be given some extra space by removing a piece of bone on the side of the skull. This is called a subtemporal decompression. This simple procedure carries very little risk and often resolves the problem. Other volume expansion procedures consist of moving larger portions of the skull to make the total volume bigger, as for example in forehead enlargement. This type of procedure is more elaborate and is suitable for very young children. Tampering with the shunt system is usually a bad idea, at least if satisfied with its functioning.

A number of patients who have slit ventricle syndrome have been treated with endoscopic third ventriculostomy. This was only possible because these patients presented with shunt failure and large ventricles. Therefore slit ventricle syndrome does not rule out endoscopic treatment but ventricles must be large enough to allow access.

If you have any questions or would like further information, please do not hesitate to contact the Family Support Workers at:

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