Spina Bifida Hydrocephalus Ireland

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Hydrocephalus & Headaches

It is not uncommon for people with Hydrocephalus to experience headaches. This information will discuss headaches and Hydrocephalus in an attempt to give a better understanding of the issues.

Individuals with Hydrocephalus shunted or not, are frequently troubled by headaches. A diagnosis of the cause of headaches can be difficult and complicated, and, as with the management of any chronic pain, it requires tremendous patience on the part of the person with Hydrocephalus and the GP or Consultant.

Possible Causes of Headaches

Dr. Harold L. Rekate, Chief of Paediatric Neurosurgery at the Barrow Neurological Institute in Phoenix, Arizona, suggests five different reasons for headaches in a person with Hydrocephalus:

- Intermittent proximal shunt obstruction. This is often referred to as the classic 'slit ventricle syndrome.' Frequently the headaches last from 10-90 minutes and resolves on their own. Often occurring in the late afternoon, they can happen at any time. The headache can be severe, and may be associated with vomiting, photophobia (aversion to light), and it can resemble, to some extent, a migraine.
- Small ventricles when the shunt fails and the ventricles can't grow to accommodate cerebro spinal fluid (CSF). This is more severe in people whose headaches come and stay; are present mostly in the morning, and can be associated with double vision. Their headaches are usually progressively more severe.
- Intermittent failure of the shunt can produce a variety of headaches. The length of time that failure occurs is indeterminate and not predictable.
- Extremely low shunt pressure can cause headaches that are similar to spinal headaches. In these cases, headache complaints are minimal when the patient is lying down but become more severe when the patient sits up or stands.
- Migraine, a common affliction, can also occur in a person with Hydrocephalus. Often there is a positive family history. Varying degrees of neurological dysfunction, headaches, vomiting, difficulty with vision and impairment of consciousness (including stupor) have been documented. Migraine attacks in shunted children and adolescents can create a disconcerting clinical situation for the patient, the family and the physician.

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Dr. Gordon McComb, Head of Neurosurgery at Children's Hospital in Los Angeles, identifies similar reasons for headaches, but he narrowed them down to just three causes: migraine, shunt failure and low pressure.

Mechanics

Brain volume, blood volume and CSF volume determine intracranial pressure (ICP). If one of these goes up, pressure in the brain will rise unless one of the others compensates by decreasing. In Hydrocephalus, this balance is distorted and an unnatural condition takes place. Under normal conditions, a person should have one ounce of spinal fluid in the ventricles and about four ounces of spinal fluid around the outside of the brain. When all the components of the brain are functioning, without a shunt, the brain has the ability to be elastic. That is, an increase in volume means an increase in pressure (and vice versa), the brain is compliant, or able to adapt. When a shunt is in place, the brain's ability to compensate for things like coughing, or straining at stool, disappears.

Additionally, when we enter REM sleep, about 80-90 minutes after we fall asleep, plateau waves and high intracranial pressure (CIP) develop. In people with shunts who undergo ICP monitoring overnight, those changes are dramatic. The things that cause pressure to go up create huge changes in the ICP of people who have been shunted. Under REM sleep, the normal rise of the ICP pressure is absent and is accentuated by the fact that there is no compliance. That is, there is no extra spinal fluid in the brain to be able to be displaced.
The placing of a shunt creates an unnatural situation. The brain fills the intracranial space while the shunt drains essentially all of the available CSF from the ventricles. The result is a large brain in a fixed (NOT elastic) solid skull, with very little room for changes in intracranial pressure. If changes in cerebral blood flow occur, resulting in increased blood volume in the intracranial space, then increased intracranial pressure will result, possible causing a headache.

Headaches can be related to the altered pressures inside the skull once the shunt is place. They can occur when the intracranial pressure is too high, and also when the intracranial pressure is too low. And what's too high for one person can be too low for another.

Treatment

Children and adults with Hydrocephalus have headaches, just like everyone else. It is the frequency and severity, suggests Dr. McComb, which determines the possible relationship between Hydrocephalus and headaches. "If the headaches are getting progressively

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worse, many times it's (due to) an intermittent malfunction of the shunt. We change the proximal end of the shunt and that's it. (The upper end of the shunt is the proximal end, and the bottom end is the distal.) "Just because a CT scan does not show enlarged ventricles does not mean that the shunt is not working. In a given percentage of cases, the ventricles are going to remain the exact same size whether the pressure is normal or elevated," adds Dr. Rekate.

With slit ventricle syndrome, argues Dr. McComb, small ventricle size is not the problem. The pro-blem occurs if the shunt clogs and the ventricles don't dilate (get larger). If this happens repeatedly, and headaches accelerate, it often means that there is a blockage of the shunt that builds up and then releases, and builds up and releases. Again, replacing the proximal ventricular catheter, the part of the shunt that is plugged, often takes care of the problem, and relieves the headache.

One of the things that both Drs. Rekate and McComb recommend is sketching out a plan of action. This involves a patient-doctor conference where a time line and course of action are established. "We'll set up parameters and if these parameters are exceeded, then we'll go ahead and fix the shunt. So, if the headaches occur at such and such a frequency and continue getting more frequent, the criteria have been put into place to change the shunt." Both physicians suggest that it is very important to put all of the various pieces together to get a perspective, as well as to individualize the care of each and every patient.

Drs. Fred Epstein and Rick Abbott, of Beth Israel Hospital in New York City, and Jeffrey H. Wisoff, of New York University Medical Center, suggest that difficulties in the diagnosis and treatment of headaches arise when there have been no changes in ventricular size and headache symptoms are of a more chronic, non-progressive nature. This can be caused by intracranial hypotension (low) or intracranial hypertension (high). They and their col-leagues recommend Intracranial Pressure Monitoring (ICP) for severe, persistent cases where CT or MRI demonstrates no enlargement of the ventricular system. ICP monitoring involves hospitalizing the patient for a day or two, inserting the monitoring device and continually measuring the pressure inside the brain. The patient is alert and active so that pressures can be recorded in relation to body position and activity. If the pressure changes can be correlated with the patient's symptomatology, the shunt can then be revised with either a higher or lower pressure valve. Dr. McComb suggests that another way to test shunt function is to inject a tracer into the shunt and do a flow study.

The cause of many headaches can be related to the altered pressures and functions inside the head once a shunt has been placed. "Unfortunately," say Dr. Rekate, "The perfect

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valve is one you don't have to put in." Today, most of the valves are pressure differential valves which react to the pressure above versus the pressure below. The valve can't tell the difference between 300 and 100 because the pressure differential is the same. Research continues on flow regulated valves as well as shunts that are programmable.

Dr. Jack Walker, past President of the American Society of Paediatric Neurosurgeons, suggest that in some cases when intracranial compliance is extremely low, treating patients with migraine therapy will often produce an improvement because of the stability provided to the intracranial vasculture by such medication. "Dilation of cerebral vessels and increased blood flow may not occur as often due to the vasoactive drugs and stabilization of the patient's intracranial blood flow. Relief of the symptoms may occur even though the symptoms were not caused by a true migraine mechanism."

The criteria for establishing the diagnosis of migraine includes "Recurrent headaches with symptom-free intervals, as well as nausea, vomiting, abdominal pain, hemicrania, throbbing pulsating pain, complete relief after rest, aura and a family history of migraine," states Drs. Hector James and Thomas Nowak of Children's Hospital of San Diego. When a protocol of shunt testing scans, and ICP monitoring, and perhaps shunt revision with a valve change have failed to alleviate the condition, management with medications for migraine may be indicated. Propranolol, Periaction and Inderal are just a few of the medications prescribed for migraine control.

Conclusion

Hydrocephalus is not a disease; it is the brain reacting to a blockage. Placement of a shunting device is currently the most common way to control this blockage. However, as noted above, shunting creates an unnatural condition. While a number of people shunted for Hydrocephalus have few or no problems with their shunts, the limited statistics that are available suggest that more than 50% will need some kind of revision. Because headaches can be an indication of malfunction or obstruction, establishing a mutually respecting relationship with your neurosurgeon and your medical team, is the best way to insure continued, comprehensive care.

The occurrence of headaches in children and adults with Hydrocephalus, especially if recurring, can be a complicated problem that requires tremendous patience thorough medical attention and an agreed up plan of action.

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