Symptoms Due To Shunting...Small Posterior Fossa

June 20, 2006 -- Newborns with hydrocephalus - an abnormal accumulation of cerebrospinal fluid in the brain - are shunted to prevent the build-up of CSF from becoming dangerous. Basically, a tube is inserted which drains CSF from the brain to the abdominal cavity (or some other space in the body).

This procedure has been used for quite some time, and while often successful in treating the hydrocephalus, in 1976, researchers Hoffmann and Tucker noted that some children develop Chiari like symptoms some time after this type of shunt is installed. They labeled their finding cephalocranial disproportion, because they believed that shunts which overdrained the CSF caused a mismatch between the brain and skull.

Now, a group from UCLA (Osuagwu et al.), including Dr. Jorge Lazareff who previously published on a Chiari operative technique to reduce trauma to children, has used modern imaging technology to look more closely at the Chiari-shunting phenomenon.

Over the course of their practice, they had observed that some children with a ventriculoperitoneal shunt for hydrocephalus would later develop symptoms suggestive of Chiari. In cases where the symptoms were severe enough to warrant an MRI, they noticed crowding of the posterior fossa - the region in the back of the brain where the cerebellum is located - consistent with what is found in Chiari. Based on their observations, they decided to quantitatively study the anatomy of shunted children who develop Chiari like symptoms and compare them to an age-matched control group. Their findings were recently published in the on-line first section of the journal Child's Nervous System.

To be included in the study, the children had to have developed Chiari type symptoms after having a VP shunt installed for hydrocephalus. Further, there had to be available a pre-shunt CT or MRI, plus a post-shunt MRI. Children were excluded if the pre-shunt imaging showed any signs of posterior fossa crowding or tonsillar herniation. Children were also excluded if a post-shunt MRI showed signs of remaining hydrocephalus.

Based on this criteria, the research team identified 9 children for the study. There were 3 boys and 6 girls, with an average age of 7.3 years. They suffered from a variety of symptoms (see Table 1), with headache, vomiting, and nausea topping the list. From the study group demographics, the team formed a control group of 8 children, matched for age, who received MRI's for other reasons and had no symptoms which could be related to the posterior fossa region.

Next, the researchers examined the post-shunt MRI's and used software to quantitatively measure anatomical features of both the study group and the control group. Similar techniques have been used several time to show that Chiari patients tend to have small posterior fossa's and certain other anatomical features.

When they compared the two groups, they found significant differences in several features (see Table 2). Specifically, in the shunted group, the clivus was shorter, the cerebellar tonsils were herniated, and the posterior fossa volume was smaller. In addition the ratio of the posterior fossa to the rest of the brain was lower for the shunted group.

These results are very similar to what has been found in other research, namely that Chiari patients tend to have specific anatomical abnormalities in the posterior fossa region, which result in an overall smaller skull in the back of the head.

In discussing their results, the authors believe that the theory from the '70's is not quite accurate, in that there is not a global mismatch between the brain and skull, but rather the mismatch is specific to the posterior fossa region, which again is what has been found repeatedly in Chiari patients.

In addition, the authors found no signs of overdrainage of CSF, or evidence of craniosynostosis. They speculate that the insertion of a shunt alters the normal CSF dynamics which recent research has indicated may be critical to proper development. Specifically, the authors state that if the CSF pressure in the skull region is changed due to the shunt, the sutures, or natural spaces between the growing skull plates, are not kept open by this natural pressure and the posterior fossa may not be as large as it should be.

This work not only has clinical significance for this type of patient - it should be noted that three children in the shunt group underwent successful decompression surgery - but it may have broader implications as well which the authors did not address.

It is becoming well established that Chiari involves a small posterior fossa which crowds a normal sized brain in that region, but the question is why? In the medical literature, an as yet to be identified genetic defect is often

Key Points

1. Children with hydrocephalus are often shunted to drain the excess CSF from the brain
2. In the 1970's, it was noted that some children shunted this way developed Chiari like symptoms
3. It was speculated that overdraining CSF led to premature closing of the skull sutures and craniosynostosis
4. This study used MRI to quantitatively measure the skull anatomy of 9 shunted children with Chiari symptoms compared to a control group
5. Found that the shunted group has shorter clivus, tonsillar herniation, and small posterior fossa's
6. These features match other research on Chiari patients versus healthy controls
7. Authors believe that shunting alters the CSF dynamics and pressure resulting in a growth mismatch between the brain and posterior fossa

Definitions

clivus - one of the bones in the skull base region

craniosynostosis - premature closing of the sutures between the skull bones

hydrocephalus - condition where there is an abnormal accumulation of CSF in the brain

nystagmus - involuntary rapid eye movements; usually indicative of a neurological problem

peritoneal - having to do with the abdominal cavity

posterior fossa - region in the back of the skull where the cerebellum is situated

shunt - small, tube-like device which is implanted in order to redirect, or drain, CSF

strabismus - eye misalignment

ventricle - one of several CSF filled spaces in the brain

vp shunt - a shunt used to drain CSF from a ventricle in the brain to the peritoneum

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

cerebellum - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

Chiari malformation I - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

decompression surgery - general term used for any of several surgical techniques employed to create more space around a Chiari malformation and to relieve compression

Source

Osuagwu FC, Lazareff JA, Rahman S, Bash S. Chiari I anatomy after ventriculoperitoneal shunting: posterior fossa volumetric evaluation with MRI. Childs Nerv Syst. 2006 May 30; [Epub ahead of print]

cited as the cause of a small posterior fossa. However, this study which demonstrated an acquired type of Chiari (with the same anatomical features of congenital Chiari) due to altering the CSF dynamics and pressure, suggests that CSF dynamics during important growth phases may also play a critical role.

While it is fun to speculate, much more research will be required to fully understand the possible reasons why the posterior fossa does not develop properly in Chiari patients.

Table 1
Signs & Symptoms Of Study Group (9 Total)

Symptom	# of Patients
Headache	7
Vomiting	9
Nausea	6
Nystagmus	2
Strabismus	2
Hearing Problem	1
Fainting	1
Apnea	1

Table 2
Selected Anatomical Measurements Between Study Group and Control Group

Feature	Control Group	Shunted Group
clivus (mm)	33.1	26.4
tonsillar herniation(mm)	-2.2	8.2
posterior fossa volume (ml)	167.3	137.9
PFV ratio	18.5	13.1

Note: *The differences between the two groups for the above features were all statistically significant; they are also very similar to findings of Chiari patients versus healthy controls; the posterior fossa volume ratio represents the size of the posterior fossa region of the brain compared to the rest of the brain*

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