Key Points

1. Most Chiari cases are thought to be congenital; however Chiari can be acquired due to tumors, shunts, etc.
2. Case study showing that a 3 year old girl developed Chiari due to a fatty filum terminale
3. It is thought that a thick or fatty filum is too tight and causes a downward pulling on the spine
4. Downward traction was actually proposed as a cause of Chiari inb 1938, but has not been a focus until recently
5. Adds to the idea that the filum plays a role in at least some Chiari cases
6. Need to investigate the elastic properties of the filum in Chiari patients

Definitions

congenital - present at birth; something you are born with

conus medullaris - cone shaped area at the lower end of the spinal cord

filum terminale - thread like structure that connects the lower end of the spinal cord to the bony spinal column

Klippel-Feil - disorder which involves abnormal fusion of 2 or more vertebrae

spinal cord - bundle of nerve fibers that runs from the base of the brain all the way down the back, through the bony spine

tethered cord syndrome (TCS) - condition where the spinal cord is improperly attached, or tethered, to the spine

traction - applying a pulling type force or tension

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

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

MRI Documents Acquired Chiari Due To Fatty Filum

November 20, 2006 -- Chiari is commonly thought of as a congenital condition, meaning that people are born with it, however the medical literature has clearly shown that Chiari can be acquired as well.

Ed. Note: *I have always found it interesting that Chiari is considered congenital, yet symptoms may not occur until later in life and as a result of some type of trauma. In one sense aren't the symptoms of Chiari then acquired in some cases?*

Tumors, arachnoid cysts, and certain types of shunts have all been shown to lead to acquired Chiari, and now a report out of the University of Washington has presented MRI evidence that Chiari can be acquired due to a fatty filum terminale.

The case study, published in the October, 2006 issue of the Journal of Neurosurgery: Pediatrics, involves a three year old girl and highlights the growing awareness of the role that the filum terminale may play in Chiari. The filum terminale is a thin, thread like structure which essentially anchors the bottom of the spinal cord to the bones of the spinal cord.

As discussed elsewhere in this issue, a filum which is unusually thick or fatty may lose its natural eslasticity and pull down on the spinal cord, effectively placing it in traction. When this force is enough to pull the end of the spinal cord below the L2 vertebra it is considered to be tethered cord. This type of downward traction can cause symptoms such as bladder and bowel dysfunction and leg pain and weakness. A tight filum is treated by cutting it and releasing the tension.

Recently, some experts have begun to screen Chiari patients for tethered cord and are sectioning the filum in addition to (or instead of) decompression surgery. Also recently, a Spanish neurosurgeon, Royo-Salvador, proposed that Chiari and scoliosis are both caused by tight filums exerting downward pressure on the spine. This makes some sense theoretically as a healthy, elastic filum is critical to the natural embryological growth upward of the spinal cord. In fact, the same concept - that downward traction could cause tonsillar herniation - was actually proposed in 1938 (Penfield & Coburn) but went out of favor because animal studies failed to demonstrate that it was true.

In the Washington case, the little girl was seen for balance problems, Klippel-Feil anomalies, and progressive scoliosis which had increased from 12 degrees to 25 degrees. An MRI showed multiple fusions of her vertebrae, a Chiari 1 malformation, and a fatty filum (a filum with fat in it looks different on MRI). Interestingly, when the doctors pulled an MRI that had been done when she was born, they clearly saw no indication of a Chiari malformation. From this, they concluded that the pulling of the fatty filum led to acquired Chiari in her case.

Even though she did not have any leg pain or bladder problems, the surgeons decided to section her filum. During the surgery, they noted that the filum was indeed under a lot of tension and sprang back when it was cut. Fifteen months after her surgery, the girl's balance problems had resolved and her scoliosis had stabilized.

This case adds to the growing recognition that a tight filum may play a role in some Chiari cases and highlights the need to somehow quantify the elastic properties of the filum in Chiari patients.

Related C&S News Articles:

Minimal Tethered Cord Shows Abnormal Anatomy

Controversy Surrounds Occult Tethered Cord Syndrome

New Surgery Proposed For Chiari

The Many Faces Of Chiari

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

Source

Source: Abel et al. Acquired Chiari Malformation Type 1 Associated With A Fatty Terminal Filum. J Neurosurg 2006 Nov;105(4 Suppl Pediatrics):329-32.

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