

Key Points

1. Up to 30% of children born with spina bifida also have Chiari
2. Lipomyelomeningocele is related to spina bifida and earlier research has noted that some patients have both LMMC and Chiari
3. Group from UAB reviewed their medical database to identify LMMC patients
4. There were 54 such patients and of those, 7 (13%) had herniations greater than 5mm
5. However, the posterior fossa volume of the LMCC patients was not significantly different from healthy controls
6. Some people think that downward traction on the spinal cord can cause Chiari; however not all research supports this idea
7. The exact link between LMCC and Chiari is not known

Definitions

conus medullaris - the lower end of the spinal cord; when the cord is tethered, the conus sits lower relative to the bony vertebra

filum terminale - small thread of tissue at the bottom of the spinal cord; if abnormal can result in TCS

laminectomy - surgical technique where part of one or more vertebra are removed

lipomyelomeningocele - birth defect where a lump of fatty tissue protrudes from the spinal canal through the spinal column

lumbar - the lower back area

myelomeningocele - spina bifida; birth defect where the spinal cord nerves and membranes protrude through the bony spine

TCS - Tethered Cord Syndrome; loose name for a spectrum of problems that all result in abnormal traction, or tension on

Chiari Linked To Lipomyelomeningocele

March 31, 2007 -- Up to thirty percent of children born with spina bifida (myelomeningocele) are also born with a Chiari malformation. Now, a report from researchers at the University of Alabama-Birmingham suggests that an unusually high percentage of children born with a related birth defect, lipomyelomeningocele, may also have Chiari malformations.

Like spina bifida, lipomyelomeningocele (LMMC) is a birth defect involving the neural tube. However, in LMMC a lump of fatty tissue is situated partially in the spinal cord itself and protrudes through the bony part of the spine to create a bulge under the skin. LMMCs are often located in the lumbar region, are usually visible, and sometimes are accompanied by an unusual hairy patch of skin over the defect.

Since the spinal cord is tethered to the fatty tissue, LMMC is one of the causes of Tethered Cord Syndrome and can cause progressive nerve damage if not treated. In general, LMMC is diagnosed in children, but as [Surgical Outcomes For Adult Tethered Cord Surgery](#) highlights, it can remain undetected in some people until they are adults. LMMC is treated surgically, which can involve a laminectomy, freeing the spinal cord from the fatty tissue and closing the defect where the fatty tissue entered the spinal cord.

Over the years, the medical literature has noted a possible association between LMMC and Chiari, but nothing definitive had emerged. However, in the March, 2007 issue of the *Journal of Neurosurgery: Pediatrics*, the UAB team presents strong evidence of some type of connection between the two conditions.

Specifically, the researchers reviewed the medical database at the Children's Hospital in Birmingham to identify patients treated for LMMC between 1992-2005. From there, they looked for patients who had had images taken both in the lumbar region for the LMMC and at the cervical level from which they could identify Chiari malformations. In total, they found 54 LMMC patients, ranging in age from 1 day to 15 years, for which about half had both sets of images available.

Using the MRIs, the team identified 7 patients with tonsillar herniations of greater than 5mm, which is how they defined Chiari for this study. This finding represents 13% of the overall 54 LMMC patients. Despite the fact that the average herniation for the group was a sizeable 10mm, only two of the 7 Chiari patients had been treated for Chiari related symptoms or syringomyelia.

To further explore the link between LMMC and Chiari, the researchers used the MRIs of the Chiari patients to measure their posterior fossa volume. Recall that one of the leading theories on the underlying cause of Chiari is that an undergrowth of the skull in the posterior fossa region results in crowding and herniation of the brain contents. In this case however, the posterior fossa volume of the study group was not significantly different from a group of age matched, healthy controls (see Table 1), and only one patient had a significantly smaller posterior fossa region.

To determine whether the downward traction put on the spinal cord by the tethering effect of the fatty tissue played a role in the herniation of the cerebellar tonsils, the scientists also correlated the positional level of the conus medullaris to the size of the cerebellar herniation. The conus medullaris is at the bottom of the spinal cord, and tethered cord is classically defined as a low lying conus relative to the bony vertebra, because the abnormal tension pulls the spinal cord down. However, in this group there was no relationship between the level of the conus and the amount of tonsillar herniation.

Given that the overall rate of Chiari in the general population is almost assuredly 1% or less, the authors are confident that the fact that 13% of their LMMC patients also had Chiari is not due to chance. However, the actual link between Chiari and LMMC remains unclear.

The idea that downward traction at the bottom of the spine could cause Chiari was actually first introduced many years ago. Recently, this concept has been revived as Tethered Cord Syndrome has received more attention and one surgeon published a report advocating sectioning the filum terminale to treat Chiari.

However, research in this area is mixed. A recent case study seemed to show clear MRI evidence that a fatty filum terminale, which puts the spinal cord under pressure, actually led to the development of a Chiari malformation in a child. On the other hand, several studies have shown that placing the bottom of the spinal cord in traction does not effect the cerebellar tonsils near the top of the spine.

Lipomyelomeningocele as a birth defect is thought to occur between the third and fifth weeks of development, and it could be that a related problem occurs at the same time which results in Chiari. However, the most likely candidate for such a problem, namely a small posterior fossa, was also not supported by the data in this study.

the spinal cord

traction - a pulling force

vertebra - individual bony segments of the spine

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

Source

Source: Tubbs R, Cuong J, Rice W, Loukas M, Naftel R, Holcombe M, Oakes W. Critical Analysis of the Chiari 1 Malformation Found In Children With Lipomyelomeningocele. J Neurosurg Ped. 2007 Mar;106:196-200.

Thus, while this research presents compelling evidence of a link between LMMC and Chiari, the nature of that link remains a mystery.

Table 1
Posterior Fossa Volume (cm³) of Lipomyelomeningocele Patients vs Healthy Controls

Age Range	Patients	Controls
0 - 5 yrs	141	145
6-10 yrs	144	151
11-15 yrs	153	159

Note: No significant differences were found on average between the PFV volumes of the two groups; only one patient had a small PFV

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