

## Definitions

**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 which surrounds, and protects, the brain and spinal cord

**Chiari malformation** - 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** - common term for any of several variations of a surgical procedure to alleviate a Chiari malformation

**growth hormone** - created by the pituitary, this hormone controls growth of the body

**laminectomy** - surgical removal of part (the bony arch) of one or more vertebrae

**magnetic resonance imaging (MRI)** - diagnostic test which uses a large magnet to create images of internal body parts

**posterior fossa** - depression on the inside of the back of the skull, near the base, where the cerebellum is normally situated

## Chiari And Growth Hormone Deficiency

Case Studies is a feature designed to highlight interesting patient cases reported in the research. Given the lack of knowledge about CM/SM, much of the published research comes in the form of case studies - doctors describing one or two patients they have seen and treated - as opposed to rigorous scientific studies. While this type of publication doesn't advance the scientific cause as much, it does give us a window into some of the issues surrounding CM/SM, including lasting side effects and related conditions. And hopefully, some of our readers will say, "Hey, that's just like me!" and know they are not alone in what they are going through.

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### Chiari I Malformation And Idiopathic Growth Hormone Deficiency In Siblings

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**Introduction:** There is mounting evidence that Chiari is actually due the bony underdevelopment of the posterior fossa region of the skull, resulting in not enough room for the normally sized brain. However, it is not clear why this underdevelopment occurs in the first place. This case study discusses three brothers whose unique presentations may offer us some clues.

**Patient 1:** A 7 year old boy with no signs or symptoms of Chiari other than mild scoliosis. However, he was only in the 10th percentile of size for his age and he was diagnosed with growth hormone deficiency. MRI's revealed a Chiari malformation and a syrinx. He was surgically decompressed and his syrinx had resolved by a six-month follow-up. Three years later, the boy's scoliosis is stable, he has no neurological symptoms, and takes growth hormone replacement.

**Patient 2:** Patient 1's 4 year old brother was also diagnosed with growth hormone deficiency. He had no Chiari related symptoms, but an MRI did reveal a Chiari malformation with no syrinx. He is being monitored with yearly MRI's which to date have shown no change in his condition.

**Patient 3:** Patient 1 and 2's 2 year old brother. This brother is healthy with no signs of growth hormone deficiency. Because his parents were concerned, an MRI was done, but there were no signs of Chiari, a syrinx, or scoliosis. He received no treatments.

**Author's Discussion:** The authors have previously reported a study which showed that a small group of children with growth hormone deficiency had small posterior fossa features similar to those found in Chiari patients. They hypothesize that growth hormone deficiency may lead to Chiari through the underdevelopment of the posterior fossa region and believe that this case strengthens their theory. They also point out that in cases of growth hormone deficiency and Chiari it would be interesting to see if hormone replacement therapy leads to improvement of the Chiari - meaning the tonsils actually ascend - and surgery can be avoided.

**Editor's Discussion:** Case studies are often of limited value in gaining a deeper understanding of Chiari; however this study is an exception. By presenting the case of 3 brothers, 2 with growth hormone deficiency and Chiari, and one with neither, they add to their own body of evidence that growth hormone deficiency may be a cause of Chiari. If validated, this would be an extremely interesting and important finding as it may provide an opening for a non-surgical treatment in some cases.

--Rick Labuda

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