

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

cervical - the upper part of the spine; the neck area

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

cranial nerve - any of the 12 pairs of nerves which originate in the brain as opposed to the spinal cord

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

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

palsy - total paralysis of a body part

respiratory - related to breathing

sleep apnea - condition which involves many episodes of temporary interruption of breathing during sleep

Severe Breathing Problems

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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Acute Respiratory Failure And Sleep-Disordered Breathing In Arnold-Chiari Malformation

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Introduction: One of the more frightening manifestations of Chiari is how it can affect breathing. People with Chiari have been shown to have a higher rate of sleep apnea than normal. Also, although it is not common, there are several case reports of Chiari patients experiencing sudden respiratory failure, meaning they can't breathe. This case discusses someone who experienced both of these problems.

Patient: Thirty-two year old man, who was a long-time smoker, was admitted to intensive care because of acute respiratory failure. Three months earlier he had begun to experience fatigue and headaches. A neurological exam revealed palsy of several cranial nerves, and no gag reflex. His right trapezius muscle and the right side of his tongue were atrophied. MRI revealed a significant Chiari malformation and a long syrinx. Because of his breathing problems, he also underwent sleep testing which revealed severely altered breathing and apnea during sleep.

Treatment: He underwent decompression surgery and his apnea was treated using what is known as positive pressure ventilation.

Outcome: Three months after surgery MRI revealed no compression around his brainstem and the syrinx had shrunk significantly. However, his sleep apnea was still severe and he continues to need to use the ventilation device.

Author's Discussion: The author's believe that the man's severe apnea problems are due to the damage to his cranial nerves, which control the throat area, resulting in actual obstruction of his airways (from weakness) while he sleeps.

Editor's Discussion: Sleep apnea can be either obstructive or central. Obstructive apnea is where part of the throat actually closes off - or obstructs - the airway. Central sleep apnea is due to a problem with the breathing center in the brainstem, which can be compressed by a Chiari malformation. One study showed that people with both Chiari and basilar invagination are likely to suffer from central sleep apnea.

--Rick Labuda

