Definitions

brainstem - the lowest part of the brain which connects with the spinal cord and controls automatic functions such as breathing and swallowing

cardiology - branch of medicine which deals with the heart

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

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

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

cluster headache - a painful, recurring headache

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

hypertension - high blood pressure

idiopathic - due to unknown causes

syringomyelia (SM) - neurological condition where a fluid filled cyst forms in the spinal cord

syrinx - fluid filled cyst in the spinal cord

Chronic Hypertension; Cluster Headache

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.

CASE 1: Cessation Of Chronic Hypertension After Posterior Fossa Decompression In A Child With Chiari Malformation

Reported In: Journal of Neurosurgery (Pediatrics). February, 2004.
Doctors: Tubbs, Wellons, Blount, Oakes, Grabb; University of Alabama & Children's Hospital - Birmingham, Alabama
Patient:

- 16-yr old boy with headaches (in the back of his head), dizziness, and clumsiness
- MRI revealed a significant Chiari I malformation with no syrinx
- Since age 10, the boy had suffered from chronic, idiopathic hypertension (high blood pressure)
- Diet and exercise had not helped, he was taking medication
- Cardiological tests were normal
- Patient underwent decompression surgery
- 2 months later, all symptoms had resolved, including the hypertension
- He no longer has to take blood pressure medication

Observations:

- Chiari has not been thought of as a cause of chronic hypertension, but it has been noted in association with some cases
- One theory is that compression of the blood supply in the brainstem can cause high blood pressure
- Hypertension treatments that involve decompression of the brain stem have had mixed results

Ed Note: *This case highlights two things: first the connection between the blood and CSF system in the brain - as the heart beats, blood flows into the brain and forces CSF out into the spinal cord - and second, that Chiari exhibits a near endless number of possible symptoms.*

CASE 2: Cluster-Like Headache: Association With Cervical Syringomyelia And Arnold-Chiari Malformation

Reported In: Cephalalgia February, 2004
Doctors: Seijo-Martinez et al.; Pontevedra Hospital, Spain
Patient:

- 36 year old female
- Recurrent headaches for past 6 months
- Occur several times each day at irregular times and last for 20-30 minutes; never at night
- Excruciating pain in the right front part of her head
- Could cause a headache by bending over
- MRI revealed a large cervical syrinx and a minor Chiari malformation
- Decompression surgery resolved the headaches

Observations:

- By the International Headache Society criteria, this patient suffered from cluster headaches, but her case was not typical
- Cluster headaches are usually regular
- Headache associated with head position is a sign of Chiari, but most Chiari headaches are in the back of the head and are aggravated by coughing and straining (not in this case)
- Her malformation was mild, but her syrinx was large
- Authors believe that at times when her syrinx expanded, it irritated a nerve which caused her headaches

Ed Note: *This case is interesting because even though she had headaches, they weren't the classic Chiari type headaches. Also interesting is that her malformation was small, she didn't seem to have symptoms associated directly from it, and yet she had a large syrinx. This is further evidence that using length of herniation as a definition for Chiari is not very useful and we have a long way to go to understand how and why syrinxes form.*