Case Studies | 3.03

Definitions

**aura** - abrupt neurological change - usually visual - which precedes a migraine headache

**basilar invagination** - condition where the odontoid - part of the second cervical vertebra - is abnormally high and crowds the lower brainstem

**brainstem** - lowest part of the brain, connects with the spinal cord

**cerebrospinal fluid (CSF)** - clear liquid in 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 traction** - sustained stretching of the neck using an outside force

**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

**craniocervical junction** - area where the skull meets the spine

**dura** - thick outer layer covering the brain and spinal cord

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

**indomethacin** - anti-inflammatory drug (not a steroid)

**Intracranial Pressure (ICP)** - pressure of CSF inside the skull area

**migraine** - severe, recurring headache often accompanied by nausea and vomiting

**propranolol** - drug used to treat migraine headaches, considered

Traction and a Cough Headache

Case Studies is a feature designed to highlight interesting patient cases reported in the research. Given the lack of knowledge about CMSM, 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 CMSM, 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: Resolution of SM and Basilar Invagination with Traction**

**Reported In:** Journal of Neurosurgery: Spine (Case illustration); April 2003

**Doctors:** Joseph and Rajshekhar, Christian Medical College and Hospital; Vellore, India

**Patient:**
- 37 year old male, Chiari I, syringomyelia and basilar invagination; verified by MRI
- Symptomatic for 5 years
- Numbness, weakness, and stiffness in limbs, unable to walk unaided
- Underwent 4 weeks of intermittent cervical traction
- Symptoms improved and was able to walk by himself
- MRI after traction showed reduction of basilar invagination, less crowding of the cerebellar tonsils, and significant reduction of the syrinx
- Surgery was deferred and patient was stable 9 months later

**Observations:**
- Association between Chiari I and basilar invagination is well recognized
- Spontaneous resolution of CMSM is not common (see Can syrinxes resolve on their own?); however improvement of basilar invagination with traction has been documented
- Authors speculate that traction created more space in the craniocervical junction and allowed the cerebellar tonsils to ascend to a more normal position; this in turn led to improved CSF flow
- Nine month follow-up is much too short to determine if the improvement is permanent or just temporary

**Ed Note:** This case clearly illustrates our fundamental lack of knowledge regarding the exact causes of syrinx formation and even under what conditions a Chiari malformation exists. We need to keep in mind this is only one case; while I don’t have the documentation, I clearly remember hearing about cases where traction made Chiari symptoms much, much worse. As a side note, many people with Chiari have associated anomalies in the same area such as basilar invagination; however the surgical correction for basilar invagination can be more traumatic than a standard decompression and may involve going in through the mouth - including breaking the jaw - and fusion of vertebra.

**CASE 2: Chiari Headache Responsive To Medicine**

**Reported In:** Headache (Brief Communications); April, 2003

**Doctors:** Gabriella Buzzi, MD, PhD et al.; IRCCS Santa Lucia Foundation; Rome, Italy

**Patient:**
- 54 year old female
- Short-lasting headaches - for 10 years - from coughing, sneezing, bending over, and lifting weights
- Passing numbness in arms when lifting weights
- Headaches went away when triggering activity was stopped

**Ed Note:** This case clearly illustrates our fundamental lack of knowledge regarding the exact causes of syrinx formation and even under what conditions a Chiari malformation exists. We need to keep in mind this is only one case; while I don’t have the documentation, I clearly remember hearing about cases where traction made Chiari symptoms much, much worse. As a side note, many people with Chiari have associated anomalies in the same area such as basilar invagination; however the surgical correction for basilar invagination can be more traumatic than a standard decompression and may involve going in through the mouth - including breaking the jaw - and fusion of vertebra.

**Patient:**
- 54 year old female
- Short-lasting headaches - for 10 years - from coughing, sneezing, bending over, and lifting weights
- Headaches went away when triggering activity was stopped
- Passing numbness in arms when lifting weights
- Also suffered from migraines without aura 6 times per month; treated with aspirin
migraine headaches, considered an anti-anxiety drug

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

syrinx - fluid filled cyst in the spinal cord

thoracic - having to do with the middle part of the spine in the chest area

vertebra - segment of the spinal column

- MRI revealed Chiari and a syrinx from C7 to T7
- Surgery was recommended, but patient wanted to postpone
- Propranolol was prescribed; 20mg 3 times per day
- After 3 months, migraines were not as severe and the exertional headaches were greatly improved with sneezing now being the only trigger
- Patient continued to improve for 3 months but then experienced recurrence of the Chiari headaches
- Propranolol was tapered to 40mg/day; indomethacin was prescribed - 25mg 3 times per day
- Chiari headaches disappeared within 48 hours
- On current medicine, patient experiences an average of 2 migraines per month, but no secondary (Chiari) headaches

Observations:
- Exertional headache is very common with Chiari malformation and may include headaches during sex
- The actual mechanism of exertional headache is not completely understood (see What causes the dreaded Chiari cough headache?), but it may involve a pressure spike or inflammation of the dura
- Indomethacin may work by reducing blood flow to the brain and thus lowering intracranial pressure; or it may block inflammation of the dura

Ed Note: The exact relationship between intracranial pressure, Chiari, syringomyelia and headaches is unknown; does Chiari cause elevated pressure or does elevated pressure "cause" Chiari?. Given the variation among people one can assume that people create and absorb CSF at different rates and if the system is out of balance, higher pressure may result. This type of imbalance may respond better to medicine than surgery (see also Dr. Bejjani’s research Treatment options after failed surgery.)