Definitions

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

central canal - center of the spinal cord

communicating syringomyelia - type of syringomyelia where CSF can flow from the 4th ventricle to the syrinx in the central canal

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

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

foramen magnum - opening at the base of the skull, through which the spinal cord passes

hydrocephalus - condition involving an abnormal build-up of CSF in the brain and enlargement of the ventricles

intractable hiccups - hiccups which last more than 24 hours

lesion - injury

shunt - tube like device used to divert - or drain - CSF and improve flow

syringobulbia - condition similar to syringomyelia, where a syrinx forms in the brainstem area

syringomyelia (SM) - neurological condition where a

Hiccups; Rapid Onset Syringomyelia

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: Hiccups & Syringomyelia

Reported In: Neurosurgery (Case Report). January, 2004.
Doctors: Dr. Seti; et al. Dept. of Neurosurgery, Hokkaido University, Japan
Patient:

- 27-yr old male with progressive numbness on right side of body
- Developed intractable hiccups over a period of 2 weeks
- Neural exam showed some weakness and loss of sensation
- MRI revealed Chiari 1 malformation, a cervical syrinx, and syringobulbia (syrinx in the brainstem area)
- Underwent standard decompression surgery
- Symptoms improved, including hiccups
- Post-op MRI's revealed the syringobulbia improved, but the cervical syrinx did not
- No hiccups at 6 month follow-up

Observations:

- The cause of most intractable hiccups is unknown
- Hiccup control center is thought to be in/near the brainstem
- Some reports attribute intractable hiccups to brainstem lesions
- Treatments include drugs, electrical stimulation of nerves, and home remedies, but are not very effective
- Authors believe this case of hiccups was caused by the syrinx in the brainstem

Ed Note: *This case illustrates how varied the symptoms associated with Chiari and syringomyelia can be. While most patients share one or two common symptoms, a lot of patients also have one or two strange symptoms depending on the specifics of their anatomy and neurological involvement. Doctors do not always spend much time in trying to decipher symptoms, even though it can be important validation for a patient.*

CASE 2: Rapid Onset Syringomyelia

Reported In: Neurosurgery (Case Report). November, 2003
Doctors: Dr. Milhorat et al.; The Chiari Institute, North Shore Long Island Jewish Health System, New York
Patient:

- 29 year old female
- Prior surgery - decompression and shunt - for Chiari, syringomyelia, and hydrocephalus
- Developed worsening headaches and leg weakness over 8 days
- MRI revealed hydrocephalus and a large syrinx
- MRI 3 days earlier had shown NO hydrocephalus or syrinx
- Underwent emergency surgery for a failed shunt

neurological condition where a fluid filled cyst forms in the spinal cord

syrinx - fluid filled cyst in the spinal cord

ventricle - CSF filled space in the brain

vertebra - segment of the spinal column

- Symptoms improved
- MRI 3 weeks after surgery showed no hydrocephalus or syrinx

Observations:

- Ventricles can enlarge - or dilate - very quickly when there is an obstruction to CSF flow
- In this case, because CSF could flow from the 4th ventricle to the central canal, a syrinx also developed rapidly
- This is an example of communicating syringomyelia - there is a path from the 4th ventricle to the central canal
- Usually with age, the central canal closes off and CSF can not flow from the 4th ventricle into it

Ed Note: *This case illustrates how understanding CSF flow dynamics is critical to understanding syrinx formation. With a flow obstruction, a syrinx developed in only 3 days. This case also shows how each person's unique anatomy affects whether and where a syrinx will form.*