CHIARI MALFORMATIONS

What is a Chiari malformation?

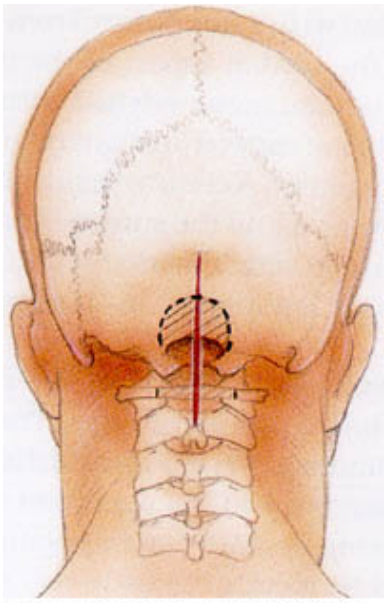
The Chiari malformation or Arnold Chiari involves crowding of the cerebellum at the base of the skull, where it meets the brainstem. This crowding can lead to blockage of spinal fluid flowing around the brain, and a syrinx or cyst in the spinal cord tissue can develop (syringomyelia). There are several subtypes of Chiari.

How is it diagnosed?

Usually an MRI of the brain and spine is required to diagnose a Chiari malformation. Flow studies can be added to assess the circulation of spinal fluid around the brain and brainstem. Xrays of the neck may be requested to assess stability of the spinal column (*see examples below).*

What are treatment options?

Surgery may be recommended if symptoms develop related to the malformation. These can be a certain type of severe headache and neck pain, sleep apnea, balance and coordination problems, dizziness, numbness and tingling in the extremities. The goal is to alleviate symptoms, restore spinal fluid flow around the brain and brainstem and allow resolution of the syrinx (cyst) in the spinal cord. A tailored surgical approach to your child's symptoms and imaging findings will be offered.



Operative exposure of a Chiari I malformation.

SYRINGOMYELIA

How is Syringomyelia diagnosed?

MRI (Magnetic Resonance Imaging) is the leading diagnostic tool used in determining if there is a syrinx. The MR imager takes pictures of body structures, such as the brain and spinal cord, in vivid detail. MRI Testing will show the syrinx in the spine or any other conditions, such as the presence of a tumor. MRI is safe, painless, and informative.

Is there any treatment?

Surgery is usually recommended for syringomyelia patients. Recurrence of syringomyelia after surgery may make additional operations necessary; these may not be completely successful over the long term.

In some patients it may be necessary to drain the syrinx, which can be accomplished using a catheter, drainage tubes, and valves.

What are the different forms of Syringomyelia?

Generally, there are two forms of Syringomyelia. The disorder may be related to a congenital abnormality of the brain called Arnold Chiari malformation. A syrinx may then develop in the cervical region of the spinal cord. Some people with this form of the disorder also have hydrocephalus (water on the brain), a condition in which CSF accumulates in the skull, or arachnoiditis, in which a covering of the spinal cord is inflamed.

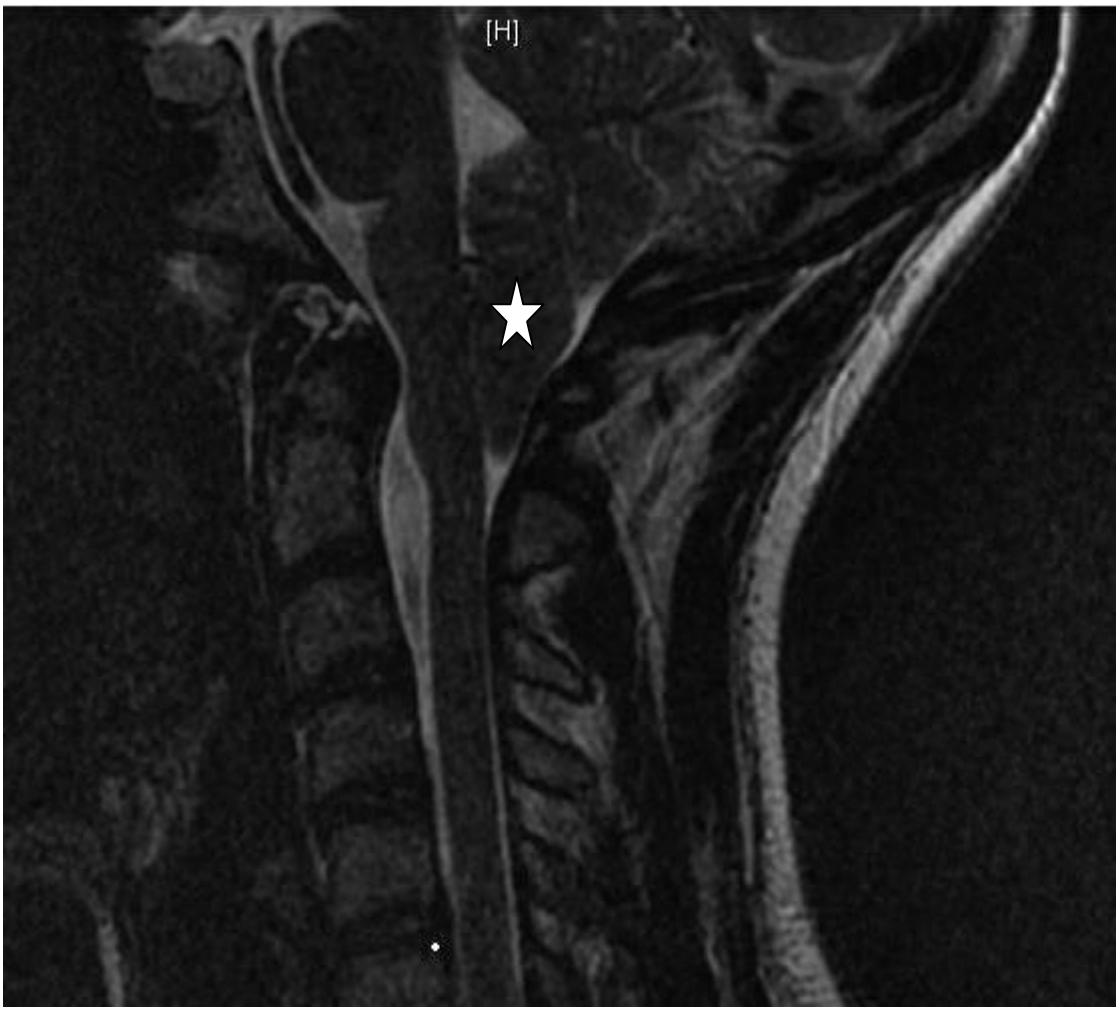
The second major form of syringomyelia occurs as a complication of trauma, meningitis, hemmorrhage or tumor. Here, the cyst or syrinx develops in a segment of the spinal cord damaged by one or more of these conditions.

What is the prognosis?

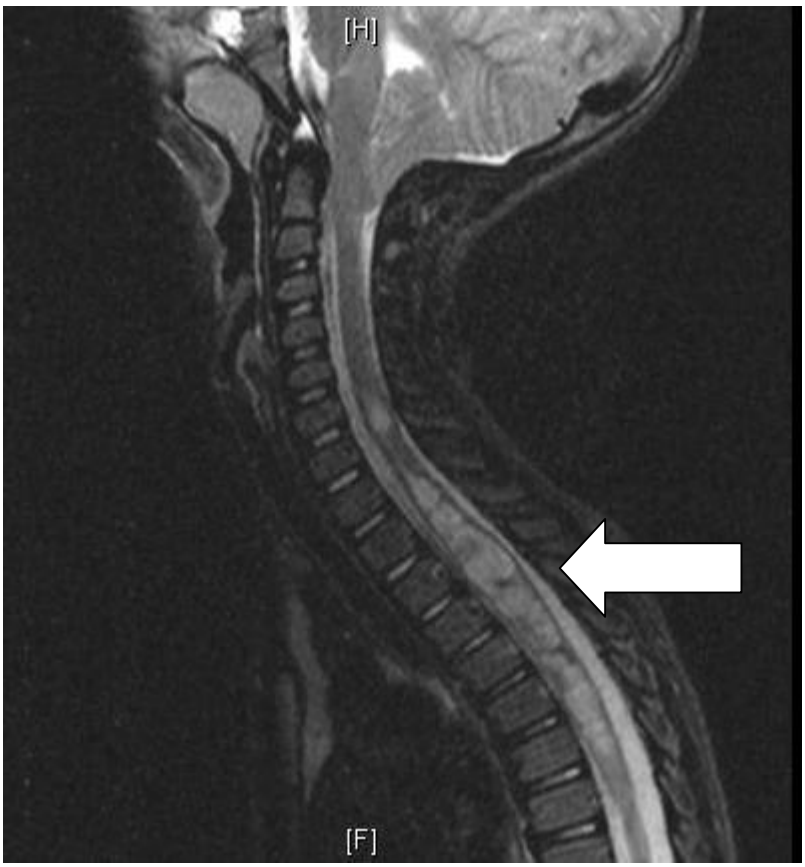
Symptoms usually begin between the ages of 25 and 40 and may worsen with straining or any activity that causes cerebrospinal fluid pressure to fluctuate. Some patients, however, may have long periods of stability. Surgery results in stabilization or modest improvement in symptoms for most patients. Delay in treatment may result in irreversible spinal cord injury.

The condition may lie dormant and undetected for months or years until a symptom or variety of symptoms become bothersome enough to warrant medical attention. Many people with 8M are not diagnosed until mid-life.

A number of medical conditions can cause an obstruction in the normal flow of cerebrospinal fluid (CSF), redirecting it to the spinal cord itself. This results in the formation of a syrinx (cyst that fills with CSF). Pressure differences along the spine cause the fluid to move within the cyst. It is believed that this continual movement of fluid results in cyst growth and further damage to the spinal cord and connecting nerves.



Chiari malformation (star on the cerebellum pushing down into the opening in the skull)



Chiari with syringomyelia (syrinx) – see arrow pointing to the syrinx