

### **Title and Author**

Title: Chiari Malformation with Associated Syringomyelia

Author: Kara Laue, RT(R)

E-mail: klaue@unmc.edu

Phone: 308-630-1140

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Supervisor/Education Coordinator: Adam Stevens, MA, RT(R)(CT)(MR)

Author E-mail: klaue@unmc.edu

Affiliation: University of Nebraska Medical Center School of Allied Health

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### **Introduction and Patient History**

Chiari Malformations (CM) are characterized by cerebellar tonsils that descend through the foramen magnum and into the cervical spinal canal. The extruding cerebellar tissue then causes obstruction of the cerebral spinal fluid (CSF), resulting in increased pressure and pulsatility of the CSF flow. It has been postulated that the pulsatile flow can cause the cerebellar tonsils to act like pistons capable of driving fluid from the spinal canal into the spinal cord itself. This results in a condition called hydrosyringomyelia, with the resulting fluid filled cavities being individually referred to as syrinx. This presentation will demonstrate the characteristic Magnetic Resonance Imaging (MRI) appearance of CM and associated hydrosyringomyelia.

This patient is a 33 year old female with the only known symptom of a headache and some vertigo (dizziness). She has not had any prior studies to the date of this MRI exam that demonstrated her condition.

### **Method**

MRI Cervical Spine Without and With Contrast  
Philips 1.5 T Intera with the NV-16 Neurovascular 16-Channel Coil

Seven imaging sequences were performed on the patient's cervical spine to look for etiology. Multihance (gadobenate dimeglumine) Gadolinium was administered for the contrast images.

Protocol	TR/TE/TI	FOV	Slice Thickness	Bandwidth	Acquisition Matrix	NSA
Survey/ Locator	26/4.4/NA	FH- 300 mm RL- 300 mm AP- 32 mm	3:1	240 HZ	240X200	2
T2-W Turbo Spin Echo (TSE)	325/120/NA	FH- 220 mm RL- 43 mm AP- 220 mm	3:1	450 HZ	276X201	8
T1-W TSE	375/7.8/NA	FH- 220 mm RL- 43 mm AP- 220 mm	3:0.6	211 HZ	244X228	6

<b>STIR</b>	3947/70/120	FH- 230 mm RL- 53 mm AP- 230 mm	3:1	192.9 HZ	256X182	4
<b>T2-W TSE</b>	3946/135/NA	FH- 135 mm RL- 150 mm AP- 150 mm	3:1	287.6 HZ	168X118	6
<b>T2-W Fast Field Echo (FFE)</b>	727/9.2/NA	FH- 127 mm RL- 160 mm AP- 160 mm	3:1	108.8 HZ	140X160	3
<b>T1-W TSE Post Contrast</b>	375/7.8/NA	FH- 220 mm RL- 43 mm AP- 220 mm	3:1	213.8 HZ	276X244	6
<b>T1-W Spin Echo (SE) Post Contrast</b>	463/8.0/NA	FH- 127 mm RL- 150 mm AP- 150 mm	3:1	304.5 HZ	168X130	4

### Findings and Discussion

According to the radiologist report dictated by Dr. Scott Anderson, the cerebellar tonsil extends five millimeters down through the foramen magnum, exemplifying a mild Type I CM. A large syrinx in the cervical spine is noted which begins at the craniocervical junction and extends down to T2-3 level of the thoracic spine. The greatest axial dimension measures 1.4 x 1.2 centimeters at the level of C7.

People with Type I CM are often unaware of the condition because they are asymptomatic. Other types of CM experience symptoms such as numbness in their extremities, posterior neck pain, balance problems, vertigo, vision problems, dysphasia, tinnitus or buzzing in the ears, vomiting, and headaches. For infants, irritability when eating, excessive drooling or dysphasia can be indications of a CM.

### Conclusion, Treatment, and Prognosis

This particular patient was recommended to another hospital for further evaluation and treatment, so the events after this MRI are unknown.

No medical treatment is known for patients with syringomyelia, however, there is a variety of surgical treatments that have been proposed for syringomyelia. Several types of surgeries can be performed either in the cervical spine or the lower portion of the skull, the occipital portion, shunt placement, and percutaneous needling which is advocated as a possible mode of therapy; however, rapid refilling of the hydromyelic cavity from the ventricular system follows aspiration of fluid at the time of surgery.

Medications may be used to ease certain symptoms caused by CM, but in no way cures CM. If a CM is asymptomatic, presents no symptoms, no surgery or treatment should be performed. When an individual presents symptoms with their CM or is causing a syringomyelia, surgery is the only treatment available to correct this condition and somewhat prevent damage to the central nervous system, although more than one surgery is usually required. With surgery, most individuals notice a reduction in their symptoms and prolonged periods of relative stability.

One surgery is a posterior fossa decompression, which is performed on only adults with CM. This creates more room for the cerebellum and alleviates pressure on the spinal column. CM may also be treated surgically with only local decompression of the overlying bones, decompression of the bones and release of the dura, which is a thick membrane covering the brain and spinal cord, or decompression of the bone and dura and some degree of cerebellar tissue

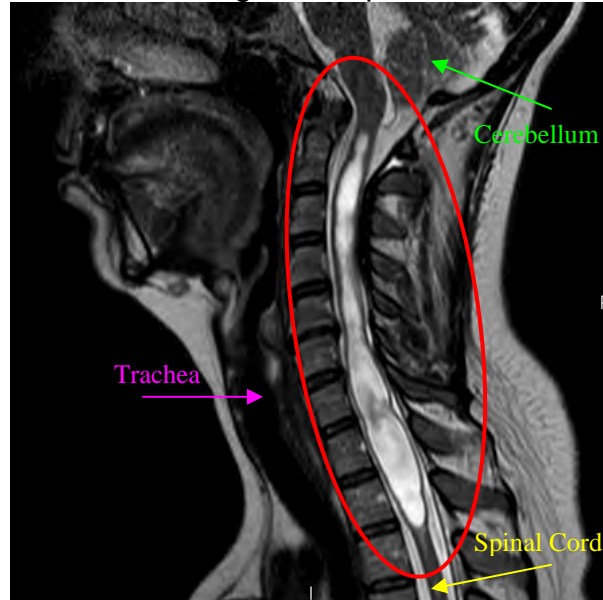
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resection, or removal of tissue. Decompression involves removing the back of the foramen magnum and often the back of the first few cervical vertebrae to the point where the cerebellar tonsils end. More space is provided for the brainstem, spinal cord, and descended cerebellar components. In an individual who has Type II CM, the surgery is treated similarly, only the tissues in the spinal canal are decompressed and the back of the skull is left alone. The goal for this surgery includes optimal decompression of nerve tissue and reconstruction of normal CSF flow around and behind the cerebellum.

The benefits of surgery should always be weighed carefully against its risks. Although some patients experience a reduction in their symptoms, there are no guarantees that surgery will reduce symptoms. Nerve damage can not be reversed, and as stated before, more than one surgery may be required. Patient education is important, especially knowing to avoid high-impact exercise like running and jumping, which can cause neck instability. Prognosis depends on the underlying cause of CM and the syringomyelia, the magnitude of neurological dysfunction, and the location and extension of the syrinx. Individuals who present with moderate or severe neurological deficits fare much worse than those with mild deficits. Mortality rates are lower in this modern world with surgical interventions and better treatment of complications that come with surgery, i.e. pulmonary embolism, or blood clots in the lungs.

#### Images

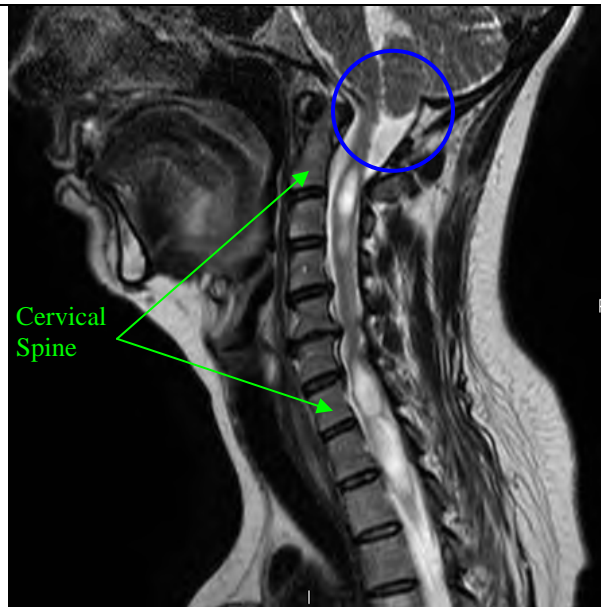
T2 Weighted Turbo Spin Echo  
Sagittal C-Spine



Red Circle: Large Spinal Cord Syrinx

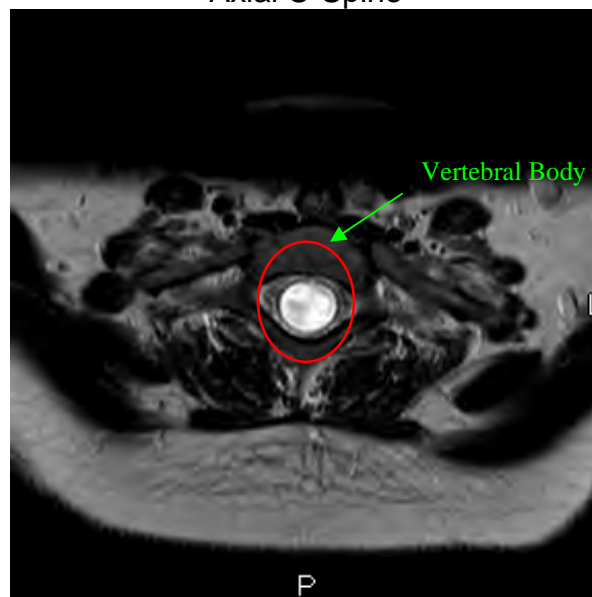
T2 Weighted Turbo Spin Echo  
Sagittal C-Spine

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Blue Circle: Type I Chiari Malformation

T2-Weighted TSE  
Axial C-Spine



Red Circle: Axial Cut of Syrinx in the Cervical Spine

### References

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