

### Key Points

1. Complex Chiari cases, which involve additional bony abnormalities have been shown to have poorer outcomes
2. Research from University of Iowa looked at reasons Chiari patients required fusion in the craniocervical junction
3. Identified 4 groups of types of patients: bony abnormalities, anterior decompressions, instability without bony abnormalities, and muscular or ligament weakness
4. The most common reason for fusion was anterior decompression surgery, where the procedure itself causes instability requiring fusion.
5. Overall 92% of the patients experienced symptom improvement
6. Dynamic MRI is critical to identifying stability issues

### Definitions

**anterior** - refers to the front

**assimilation** - in this context, refers to bones that are congenitally fused together

**craniocervical junction (CCJ)** - where the skull and bony spine meet

**dorsal** - towards the back

**dura** - thick, outer covering of the brain and spinal cord

**duraplasty** - surgical technique where the dura is expanded by sewing a patch into it

**dynamic MRI** - MRI of the neck in different positions of flexion and extension

**EDS** - Ehlers Danlos Syndrome, a group of genetic disorders which involve problems with the connective tissue

**extension** - bending the neck backward

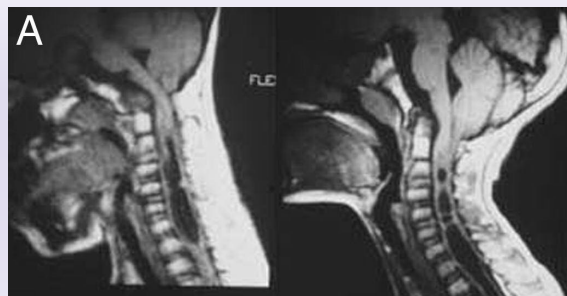
### Cervical Fusion In Chiari Patients

July 31, 2008 -- There is a growing recognition that not all Chiari cases are the same, which can have profound implications for diagnosis and especially treatment. One way to think about Chiari beyond the antiquated definition based on size of herniation has to do with the underlying cause. For example, there is growing evidence that some Chiari cases can be classified as due to small posterior fossas, while others may be related more to tethered cord, and still others may be due to problems with CSF dynamics (such as pseudotumor and hydrocephalus). While we are likely still far away from redefining Chiari along these lines, it is an important advance that surgeons and researchers are thinking along these lines.

A second way to break down Chiari cases is by whether they are simple or complex, where complex cases involve additional abnormalities (such as bony problems of the craniocervical junction) and often require spinal fusion. A recent study from China showed that in a large patients series, so called simple Chiari cases enjoyed a 95% improvement rate with surgery, whereas complex cases had a much lower success rate (74%). Now, a publication from the University of Iowa (Fenoy, Menezes, Fenoy) in the July, 2008 issue of the Journal of Neurosurgery: Spine has taken a deeper look into what constitutes many of what can be classified as complex Chiari cases.

Specifically, the Iowa researchers reviewed their experience over a 10 year period with Chiari and syringomyelia patients who required cervical fusion as part of their treatment. Overall, the group identified 234 such patients, who ranged in age from 2.5 - 86 years; a third of the patients were under 16. Not surprisingly, more than three fourths of the group suffered from head and/or neck pain. In order to identify stability problems in the patients, the doctors used dynamic MRI extensively, which captures images of the neck in different positions. In some cases, this can reveal compression which is not apparent on a standard MRI (Figure 1 below).

**Figure 1: Dynamic MRI Showing Compression In Neck Flexion, but Not Extension**



As stated previously, all the patients underwent fusion to some extent to provide stability to the craniocervical junction. In nearly all the cases, the fusion involved what are called semi-rigid instruments, meaning titanium loops and cables, as opposed to rods and screws. In addition, 119 of the group underwent posterior fossa decompression for Chiari and/or syringomyelia, and 51 underwent both posterior fossa decompression and a transoral decompression to relieve pressure on the brainstem. This type of surgery, which goes through the mouth, always requires some type of fusion for stability.

When the researchers analyzed the patients for why they required fusion, they identified 4 distinct groups (although some patients fit into more than one category):

**Group 1:** Bony abnormalities, such as assimilation, which cause compression

**Group 2:** Previous anterior decompression of the brainstem, where the surgery itself causes instability requiring fusion

**Group 3:** Instability of the CCJ without bony abnormalities

**Group 4:** Instability due to muscle or ligament weakness, such as from EDS or repeated surgeries

The most common reason for fusion was Group 2, representing 44% of the patients (Figure 2). Groups 1 and 2 were about equal at 25% and 26% respectively, followed by group 4 at 14%.

Although, the authors did not analyze outcomes based on this grouping they did report an overall success rate of 92%, based on symptoms improvement.

It is not known how common or uncommon these types of complex Chiari cases are, although that would make for an excellent research study. What this work does make clear is that if there are any questions as to the stability of the craniocervical junction, or if symptoms are related to neck position, that a dynamic MRI is critical to planning a proper course of treatment.

**flexion** - bending forward of the neck

**foramen magnum** - opening at the base of the skull through which the brain connects with the spinal cord

**fusion** - type of surgery intended to increase stability of the spine by joining together different parts

**posterior fossa decompression (PFD)** - surgical procedure for Chiari which creates more space around the cerebellar tonsils

**reducible** - when referring to bone problems, means the problem can be corrected

**transoral** - through the mouth

**ventral** - pertaining to the front or bottom

**cerebellar tonsils** - portion of the cerebellum located at the bottom, so named because of their shape

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

**Chiari malformation I** - 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

**syringomyelia** - condition where a fluid filled cyst forms in the spinal cord

### Source

Fenoy AJ, Menezes AH, Fenoy KA. [Craniocervical junction fusions in patients with hindbrain herniation and syringohydromyelia.](#) J Neurosurg Spine. 2008 Jul;9(1):1-9

**Figure 2: Classification of Patients By Reason For Fusion (234 Total)**

<b>Group 1</b>	Bony abnormalities	25%
<b>Group 2</b>	Anterior decompression	44%
<b>Group 3</b>	No Bony abnormalities	26%
<b>Group 4</b>	Muscular or Ligament Laxity	14%

**Note:** Patients could be assigned to more than one group.

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