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# Multimodal Evaluation Of Cerebrospinal Fluid (csf) Dynamics Following Extradural Decompression For Chiari I Malformation

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**Multimodal evaluation of cerebrospinal fluid (CSF) dynamics following extradural  
decompression for Chiari I malformation**

A Thesis Submitted to the  
Yale School of Medicine  
in Partial Fulfillment of the Requirements for the  
Degree of Doctor of Medicine

By  
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2015

Keywords: Chiari I malformation, extradural decompression, cine flow MRI

Running head: CSF dynamics following extradural Chiari I decompression

## **Abstract**

Chiari I malformation is a hindbrain abnormality characterized by tonsillar herniation  $\geq 5$  mm below the level of the foramen magnum. Extradural decompression is a minimally invasive technique for treating Chiari I that involves a suboccipital craniectomy with removal of the outer dural layer. Other techniques that involve opening of the dura, arachnoid or even manipulation of the cerebellar tonsils have since been the mainstay of treatment, but are associated with a higher complication rate and longer hospital stays. While there is no agreement on which surgical method is optimal, mounting evidence demonstrates that extradural decompression effectively treats clinical symptoms with a minimal reoperation rate. Many of the symptoms associated with Chiari I are now understood to be related to obstructed cerebrospinal fluid (CSF) flow. Therefore, one goal of a successful decompression is improved CSF dynamics. Phase-contrast cine flow MRI provides insight into CSF dynamics before and after surgical decompression. This study describes a surgical series of 18 patients with Chiari I malformation undergoing extradural decompression, for which clinical improvement was correlated with radiologic changes. All 18 patients presented with symptomatic Chiari I malformation, confirmed on imaging to have tonsillar herniation  $\geq 5$  mm. Two patients had associated syringomyelia. All patients underwent suboccipital decompression and C1-laminectomy with dural splitting. Patients were categorized as having complete, partial, or no resolution of their symptoms. Posterior fossa area (PFA), cisterna magna area (CMA), and tonsillar herniation were assessed on T2-weighted magnetic resonance imaging (MRI). Improvement in CSF flow was evaluated with phase-contrast cine flow MRI. All patients received standard pre- and post-operative MRIs, 8 (44.4%) patients had pre- and

post-operative phase-contrast cine, while the rest had only post-operative cine. Patients who had a complete resolution of their symptoms also had a greater relative increase in cisterna magna area compared with those with only partial improvement ( $p = 0.022$ ). In addition, those with complete improvement had smaller pre-operative cisterna magna area compared with those who had either partial (0.020) or no (0.025) improvement. Ten (91%) of the 11 patients with improved flow also had improvement in their symptoms. There was one post-operative complication of dysphagia and dysphonia. None of the patients have required a second operation. Extradural decompression has the potential to be the first-line treatment for Chiari I malformation, but has since been lacking an objective measure by which to assess surgical success, as well as the need for reoperation. In our study, an increase in the CSF spaces and improved CSF dynamics and were associated with greater resolution of clinical symptoms. Including cine imaging as part of routine pre- and post-operative evaluation can help identify which patients are most likely to benefit from surgery, as well as determine a satisfactory decompression.

## **Acknowledgements**

I would like to thank my thesis advisor Dr. Michael Diluna, in addition to all of the members of my thesis committee, Dr. Murat Gunel, and Dr. Charles Duncan, in addition to the neurosurgery department for their support. In particular, the work would not have been possible without the help of Dr. Ryan Grant, who assisted with the radiographic analysis of the data and editing of the final publication. I would also like to thank Michael Hajek for his help editing the final manuscript. A version of this manuscript has been accepted for publication in the Journal of Neurosurgery: Spine.

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## **Introduction**

### *History of Chiari malformations*

Described most famously by Hans Chiari in the 1890's, Chiari malformations include a number of hindbrain malformations that range in severity<sup>1</sup>. In a series of over 40 post-mortem hindbrain anomalies, Chiari observed elongated portions of the cerebellum, a hypoplastic or absent fourth ventricle, and occasionally a flattened medulla<sup>2</sup>. He ultimately concluded that the observed anatomical findings were a result of chronic congenital hydrocephalus, which in turn caused a downward displacement of the brainstem and cerebellum with subsequent widening of the spinal canal<sup>2</sup>. Chiari also commented on changes within the spinal cord related to the presence of a lumbar myelomeningocele<sup>2</sup>. A number of other individuals had also reported on various hindbrain malformations prior to Chiari's description, though none to such an extent or in such great detail as Chiari<sup>3</sup>. For example, as early as 1641, Nicolaes Tulp wrote about a myelodysplastic patient with possible hindbrain herniation<sup>3</sup>. In 1881, Theodor Langhans gave his account of a patient with tonsillar herniation, hypothesizing that the obstruction of CSF and blood flow caused subsequent syringomyelia<sup>3</sup>.

The Chiari classification system is still commonly used to describe hindbrain abnormalities<sup>1</sup>. Chiari types I-III are graded based on increasing amounts of herniation through the foramen magnum. In type I, there is herniation of just the cerebellar tonsils. Type II Chiari malformations is classified by a more pronounced herniation, including the vermis and brainstem, while type III malformations include an encephalocele of the hindbrain structures. Type IV, on the other hand, represents cerebellar hypoplasia<sup>1</sup>. Since

Chiari's initial classification, other variants have been described and are included in this system. Chiari type 0 corresponds to a syringomyelia (defined as damage to the spinal cord caused by a fluid filled pocket) that resolves with decompression, whereas type 1.5 describes tonsillar herniation with an additionally caudally displaced brainstem<sup>1</sup>.

#### *Clinical presentation of Chiari I malformation*

Chiari I malformation is an anatomical abnormality characterized by cerebellar tonsillar herniation of  $\geq 5$  mm below the level of the foramen magnum<sup>1,4,5</sup>. Classically, patients can present with headache, long tract signs (such as loss of sensation, position sense, weakness, spasticity, or incontinence), bulbar symptoms (such as difficulty swallowing, dysarthria, or other dysfunction of the lower cranial nerves), or other neurologic impairment. A number of other anatomic and radiologic findings have been found in association with Chiari type I. For example, basilar skull and craniocervical junction abnormalities are often found in association with Chiari I, where patients may also have smaller, more shallow posterior fossas, along with smooth cerebellar tonsils resulting from compression<sup>1</sup>. While most patients with type I do not have hydrocephalus, many have a decreased cisterna magna area<sup>1</sup>, thus leading to a diminished space for CSF flow. Finally, patients with Chiari type I may also have syringes, defined as a fluid filled cavity within the spinal cord, most commonly in the lower cervical or upper thoracic spine<sup>1</sup>.

The incidence of Chiari type I is thought to be 0.5-3.5%<sup>6</sup>, though some studies may underestimate the actual number, because many individuals with this anatomical anomaly are asymptomatic<sup>7</sup> and therefore go undetected. The most common complaint for patients



with Chiari I is localized occipital headache/pain which can be exacerbated by Valsalva maneuvers<sup>1</sup>. Symptoms are thought to derive from impaired dynamic CSF flow<sup>5,6,8</sup> or direct compression of local anatomy<sup>9,10</sup>. Compression of the brainstem and lower cranial nerves can cause dysphagia, sleep apnea, gagging, and other symptoms. Chiari type I has been reported in association with a syrinx in approximately 65% of patients<sup>6</sup>. As such, patients may also have a distribution of symptoms related to the location of a syrinx<sup>1</sup>, such as a loss of sensation or pain in a dermatomal distribution. The presence of a syrinx in these patients is additionally associated with a concurrent spinal deformity; patients with syringes > 6 mm had a greater risk of having scoliosis<sup>11,12</sup>.

### *Pathophysiology*

Chiari I malformations develop under one of two circumstances: the acquired type occurs in relation to trauma or other conditions that increase intracranial pressure, whereas the etiology of the congenital type is not yet fully understood.<sup>1</sup> Chiari's characterization of these malformations as a result of increased pressure is the basis for the hydrodynamic theory of their formation, which was later expanded upon by W. James Gardner, Bernard Williams, and Edward Oldfield<sup>1</sup>. In a sense, Chiari and others were correct in their characterization of the pathophysiology of this malformation as an obstruction of normal CSF dynamics<sup>7</sup>. Since then, a number of other proposals have emerged to explain the origins of this condition. In the "cranial constriction" mechanism, underdeveloped occipital bones are thought to confine hindbrain structures in a posterior fossa that is too small<sup>6</sup>. The age of onset can vary by patient<sup>13</sup>, though the condition tends to have a female predominance<sup>6</sup>. A variety of physiologic parameters, including the geometry and

compliance of surrounding anatomy, as well as the velocity, pressure, and resistance to CSF flow, are recognized as key factors in the pathophysiology of Chiari type I<sup>7,14</sup>. According to the Monro-Kellie doctrine, which states that the sum of all volumes within the cranial vault (namely blood, CSF, and brain) must remain constant, expansion of the blood vessels during systole displaces approximately 1.5 mL of CSF from the cranial vault, thereby causing oscillations in CSF flow in the spinal canal<sup>15</sup>. These fluctuations, in turn, cause pressure waves across adjacent tissue and pressure gradients across channels in the system<sup>15</sup>. The systolic pressure waves are further complicated by the anatomy of the subarachnoid space<sup>15</sup>. In addition, CSF flow can vary based on age, as well as physiologic measures such as blood pressure<sup>15</sup>. In fact, the Valsalva maneuver has been shown to transiently lower CSF velocity and raise CSF pressure<sup>15</sup>. The current understanding states that the abnormal morphology of the hindbrain causes an interruption of CSF pulsations at the foramen magnum<sup>7</sup>. The increased resistance at this region leads to an increased pressure gradient, which in turn exacerbates the compression of neural structures<sup>7</sup>. In a numerical simulation of CSF flow based on MRI, a patient with Chiari I was found to have 1.5 times greater peak pressure than the healthy control<sup>7</sup>. Patients with Chiari I were also found to have more turbulent and increased CSF flow<sup>15,16</sup>.

It is less clear how the abnormal fluid dynamics in Chiari I leads to syrinx formation, but one theory suggests that it simply a result of CSF obstruction in the spinal pathway<sup>17</sup>. However, the presence of syringomyelia is not typically associated with hydrocephalus, nor additional arachnoid adhesions<sup>18</sup>. This differs markedly from experimental models of

syringomyelia in which hydrocephalus, spinal cord injury, or arachnoiditis incite the pathology<sup>18</sup>. Nevertheless, no animal model exists for this disease process<sup>18</sup>. Numerous clinicopathological and radiological studies have been conducted to better understand the CSF origin of syrinx<sup>18</sup>. Some evidence also suggests that tapering of the spinal canal and its subsequent effect on CSF pressures may be involved<sup>7,15</sup>. Yet, pressure studies do not support a simple pressure gradient in the pathophysiology of syrinx formation, as the degree of CSF blockage can vary across patients<sup>18</sup>. An alternative theory suggests that there is decreased CSF space compliance (volume change per pressure change) in Chiari I caused by tonsillar herniation<sup>18</sup>. In turn, this reduces the ability of the spinal veins to reabsorb CSF causing a build-up of CSF in the central canal<sup>18</sup>. Syrinx progression may be caused by increased pressure in the cervical subarachnoid space that, in turn, compresses the spinal cord and propagates pressure waves through the syrinx with each cardiac pulsation<sup>19</sup>.

In the search for a genetic explanation, whole genome screening revealed regions on chromosomes 15q21.1–22.3 and 9q21.33-33.1 to be associated with Chiari malformations<sup>1</sup>. Whole genome expression analysis of dura from pediatric patients brought forth several candidate genes, including ones involved in the dorso-ventral axis pathway.<sup>13</sup> The clinical and likely etiologic heterogeneity of the disease makes it especially difficult to identify a singular genetic cause. Given that these malformations do not follow a monogenic inheritance pattern<sup>1</sup>, they may emerge from multiple entities causing the same anomaly.

*Review of surgical techniques*

The mainstay of treatment for Chiari I malformation is surgery, with the goals to decompress hindbrain structures and restore CSF flow<sup>1,20</sup>. A comparison of patients treated operatively vs. non-operatively demonstrated that 94.5% of patients who underwent surgery saw improvement in their symptoms compared to 47.1% of those treated conservatively<sup>21</sup>. In conjunction with other pathology, such as a tumor or craniosynostosis (defined as the premature fusion of the fibrous sutures in an infant's skull), the surgical treatment may be more extensive<sup>1</sup>. Many individuals meet the radiologic criteria for Chiari type I; therefore, determining which associated symptoms will likely benefit from surgery poses an additional challenge<sup>20</sup>. Asymptomatic children with incidentally diagnosed Chiari I (based on tonsillar herniation) showed a favorable outcome when followed conservatively over an average of 5.8 years: 3 out of 16 later developed symptoms and only 2 were subsequently treated for resultant hydrocephalus<sup>22</sup>. A small proportion of children may have spontaneous improvement or worsening of their condition (assessed as the appearance of symptoms, change in syrinx, or change in flow), though the degree of tonsillar herniation did not differ from those whose condition remained stable<sup>23</sup>. Indications for treatment include the presence of a syrinx, as well as debilitating symptoms that impede the patient's quality of life<sup>1</sup>. In a study of pediatric patients with syringes, the majority remained stable in size through non-operative treatment, though 2 out of 17 increased in size<sup>24</sup>. It is therefore important that patients with syringes receive regular re-imaging and re-evaluation of their symptoms. The majority of patients undergoing decompressive surgery for Chiari I malformation note post-operative improvement in their quality of life<sup>25</sup>. Headaches exacerbated by Valsalva

maneuvers are thought by some to be an accurate predictor of whether patients are likely to benefit from surgery<sup>21,26</sup>. Decompressive surgery was more likely to resolve pain than either sensory or motor deficits<sup>11</sup>. Furthermore, there is some indication that operating closer to the first presentation of symptoms gives a greater chance of postoperative improvement, especially in the presence of a syrinx<sup>20</sup>. This suggests that early detection and implication of Chiari I as the cause of a patient's symptoms allows for a greater improvement after surgery. Objective measures to help predict a patient's improvement will correspondingly help to select patients who are good candidates for surgery.

The first documented surgical decompression for hindbrain malformations included accounts by Wilder Penfield, Donald Coburn, and Cornelis Joachimus Van Houwenige Gradftdijk, who sought to treat obstructive hydrocephalus in a patient with tonsillar herniation and myelomeningocele by decompressing the posterior fossa, as well as removing "redundant" cerebellar tissue<sup>3</sup>. In 1950, W. James Gardner and Robert Goodall published the most widely known series of these first decompressions, in which they described 17 patients with Chiari malformation<sup>3,27</sup>. For most of these aforementioned patients, symptoms began during adulthood. Patients presented most commonly with weakness or numbness in at least one extremity. Other reported symptoms included headache, pain, tinnitus, hearing loss, imbalance, double vision, difficulty breathing, dysphagia, hoarseness, and incontinence<sup>27</sup>. Many of the patients also had associated bony anomalies such as platybasia (a flattening of the skull based caused by an abnormal relationship between the occiput and cervical spine) and Klippel-Feil syndrome (an abnormal fusion of two or more cervical vertebrae)<sup>27</sup>. In addition to decompressing the

posterior fossa, the procedure also involved removal of the arch of C1, incision of the dura, which was left open, as well as efforts to reopen the foramen of Magendie. Thirteen of the seventeen patients had an explicit improvement in their symptoms whereas 3 had worsening of their quadriparesis<sup>27</sup>. Gardner additionally demonstrated in a later series that such decompressive treatment could also be used to resolve syringomyelia<sup>3</sup>. Following their report, posterior fossa decompressions became the standard for treating symptomatic hindbrain herniation<sup>3</sup>. Between 1965 and 2013, over 145 operative series of Chiari type I patients were published in the literature<sup>6</sup>.

While surgical techniques to widen the posterior fossa, decompress neural structures, and improve CSF flow have been widely described<sup>10</sup>, Chiari I malformation has no single definitive treatment<sup>4,8,28</sup>. The classic method of decompression includes a suboccipital craniectomy and C1 laminectomy, either with or without dural opening and subsequent autograft or allograft duroplasty. Numerous additional surgical techniques have been described in the literature<sup>29</sup>, including opening and dissection of the arachnoid<sup>6</sup>, manipulation or resection of the cerebellar tonsils, as well as stenting or shunting of the fourth ventricle and syrinx to improve CSF dynamics<sup>30</sup>. Most reported cases involve opening of the dura (92%), with a smaller majority also involving opening of the arachnoid (66%)<sup>6</sup>, suggesting that this level of invasiveness is still common practice. A small proportion of cases in which the arachnoid was opened also involved tonsillar resection or syrinx shunting<sup>6</sup>. A higher proportion of the purely adult series (97.3%) than the pediatric series (81%) describe opening the dura as part of the posterior decompression<sup>6</sup>. Postoperative evaluation of neurologic improvement has typically been

a subjective evaluation, with the majority of patients (75%) reporting at least some improvement in their symptoms<sup>6,31</sup>. Patients with associated hydromyelia (a dilation of the central canal) may have lower rates of improvement and have a worse outcome following surgery<sup>31</sup>. The reported rates of residual syrinx following decompression range from 0 to 22% (average 6.7%) and even with radiographic improvement may be associated with lasting signs of spinal cord injury<sup>32</sup>. One group reports re-exploration of the decompression, lysis of adhesions, and duroplasty as an effective treatment for recurrent syringomyelia<sup>33</sup>. Alternatively, Alzate *et al.* report using a syringosubarachnoid shunt in patients with syringomyelia, with a complete disappearance in 15 cases, a decrease in size in 17, and no change in size in only 2<sup>11</sup>.

These surgical techniques are not without complications, which are often related to penetrating the dura or bleeding<sup>34</sup>. In addition, the entrance of blood, muscle, or other debris in the CSF space can cause arachnoiditis and subsequent obstruction of flow<sup>20</sup>. Across all of the reported case series from 1965-2013, the median complication rate was 4.5%<sup>6</sup>. Other complications include re-closure of the foramen and hydrocephalus secondary to the formation of a posterior fossa hygroma (a CSF-filled sack)<sup>1</sup>.

In Chiari type I malformation, symptoms can recur even with adequate surgical intervention of any kind<sup>8,20,26</sup>, and therefore the surgeon must ensure sufficient execution of surgical technique to minimize potential treatment failure. First described in 1993 by Isu *et al.*, extradural Chiari I decompression – that is, the removal of only the outer layer of dura – is an alternative surgical technique that avoids the risks of CSF leak and

vascular injury, while allowing decompression of the expandable inner dural layer<sup>4,28</sup>. In this dural splitting technique, there must be two identifiable dural leaflets, with an extensive outer leaflet resection performed in order to allow relaxation of the inner dural layer<sup>8</sup>. A retrospective comparison of patients undergoing extradural decompression with those undergoing duroplasty demonstrated a similar rate of symptom improvement (73% vs. 87%), with more patients (11 vs. 23) overall undergoing duroplasty<sup>31</sup>. While duroplasty more reliably led to an improvement in hydromyelia, opening the dura was also associated with more surgical complications, including CSF leak and infection. For certain patients, a less invasive procedure that can still allow symptom improvement might be preferable in light of the lower risk of operative complications. In their experience of 80 patients undergoing decompression with dural opening and 16 patients undergoing bony decompression only, Hayhurst *et al.* found greater improvement in headaches in patients with dural opening<sup>20</sup>. However 74% of those who had extradural decompression had at least some improvement in their headaches. The authors comment that the optimal procedure for restoring CSF dynamics might vary by patient. 72% patients for whom the dura was opened additionally had cauterization of the cerebellar tonsils<sup>20</sup>. Patients who had undergone dural opening also had a higher complication rate of 30%, compared to a complication rate of 12.5% in the extradural group<sup>20</sup>. A randomized study performed in 27 Turkish patients demonstrated similar rates of clinical improvement when the dura was opened with duroplasty (73.3%) compared to when the dura was left intact (83.3%)<sup>35</sup>. Three patients in the duroplasty group additionally had complications of CSF fistulas<sup>35</sup>. Yeh *et al.* similarly found greater complications, as well



as more cases of post-operative headache, pain, and nausea in patients undergoing duroplasty compared to those undergoing bony decompression alone<sup>36</sup>.

### *Informing the treatment paradigm*

Thus far, MRI has been the mainstay for diagnosing Chiari type I, as well as assessing post-operative outcomes<sup>7</sup>; however, standard MRI gives only a static measure of the anatomical distortion and radiologic findings that may not adequately correlate with clinical symptoms<sup>7</sup>. Some studies suggest that patients with symptomatic Chiari malformations may also have smaller posterior fossa volumes<sup>7</sup>. Heiss *et al.* showed that patients with Chiari had decreased anteroposterior diameters of the CSF spaces near the foramen magnum<sup>19</sup>. One group compared the anatomical and physiologic parameters of individuals with and without Chiari I malformation (based on the definition of tonsillar herniation > 5 mm) using MRI<sup>37</sup>. They further subdivided those with Chiari I into those with typical and atypical symptoms<sup>37</sup>. Individuals with Chiari I had shorter clival and supraocciput length, greater crowding of the hindbrain in the posterior fossa, smaller fourth ventricular volume, cord displacement, and intracranial pressure<sup>37</sup>. These differences were even more significantly different in the “typical” Chiari sub-group<sup>37</sup>. Another group also showed underdevelopment of the posterior fossa structures in Chiari I, and further developed a probability model to better diagnosis Chiari I, beyond the classical definition<sup>38</sup>. The authors argue that Chiari I should be defined along the basis of occipital bone undergrowth, rather than purely tonsillar herniation, which can be caused by other conditions in the absence of posterior fossa crowding<sup>38</sup>. Posterior fossa volume measurements can even be automated from MRI’s to facilitate the ease of obtaining this

informative measure<sup>39</sup>. More recently, one group developed a “Chiari severity index,” an integration of clinical symptoms and pre-operative radiologic features to prognosticate the likelihood of improvement from surgery<sup>40</sup>. Headache and myelopathic symptoms were the most indicative clinical features, whereas the presence of a syrinx > 6 mm was the most significant radiologic predictor<sup>40</sup>. Yet given that a significant portion of the underlying pathology may be secondary to CSF flow obstruction<sup>41</sup>, a more dynamic measure of the pathophysiology of Chiari I could more effectively inform treatment decisions.

A number of different imaging and other techniques have emerged to evaluate CSF flow dynamics in better detail<sup>7</sup>. Phase-contrast cine flow MR imaging captures the CSF flow by comparing the signals from stationary and moving nuclei in two data sets<sup>42</sup>. Stationary nuclei have identical phases (giving a net phase of zero), whereas moving nuclei move from one field to another (giving a net phase value and therefore, a signal)<sup>42</sup>. By subtracting the two data sets, only moving nuclei are seen on the final image<sup>42</sup>. The grayscale intensity of the image is related to the CSF velocity, with rostral and caudal flow seen as shades of black and white, respectively<sup>42</sup>. Importantly, cine-flow MRI provides an important tool to assess CSF flow as it can be used *in vivo*. Multiple studies have additionally demonstrated abnormal CSF flow velocities in patients with Chiari I compared with healthy controls, and in turn, an improvement in these abnormally high velocities following decompressive surgery<sup>7,17</sup>. Similar findings were found in a prospective study of patients undergoing radionuclide cisternography as a measure of CSF flow dynamics<sup>43</sup>. Patients who had abnormal pre-operative flow were

correspondingly the most debilitated, and thus had greatest clinical improvement following surgery<sup>43</sup>. Cine findings have even been correlated with symptomatology; occipital headaches were associated with abnormal CSF flow on cine, whereas frontal or generalized headaches did not have an association with flow<sup>44</sup>. Symptomatic patients have simultaneous bidirectional flow whereas asymptomatic patients do not have this problem<sup>15</sup>. Cine findings have been less informative in the etiology of syrinx. One study found that patients who developed a syrinx had a longer period caudal flow in the ventral subarachnoid space<sup>18,45</sup>. Another study used cine-flow MRI to compare patients with Chiari I with healthy controls and found that those with the disease had abnormalities in CSF flow at the craniocervical junction<sup>46</sup>. Namely, they had heterogeneous flow at the craniocervical junction, anterolateral flow jets, and flow vortex formation – abnormalities which were more pronounced in patients with concurrent syringomyelia<sup>46</sup>. Patients additionally had higher peak flow velocities at the craniocervical junction, and maximum velocities that occurred later in the cardiac cycle<sup>46</sup>. In a study of 130 patients with Chiari I malformation defined by tonsillar herniation, McGirt *et al.* found that 81% had decreased flow on pre-operative cine flow MRI<sup>47</sup>. Of the patients with abnormal pre-operative flow, 91% experienced post-operative improvements in flow<sup>47</sup>. Those without improvements on cine were twice as likely to have no change in their symptoms<sup>47</sup>. Additionally, those with “normal” pre-operative flow were 3.4 times more likely to have symptom recurrence following surgery<sup>47</sup>. Another study compared CSF velocities and pressures in symptomatic patients with Chiari I before and after surgery, as well as relative to healthy controls<sup>17</sup>. Aqueductal and cervical spinal CSF velocities were higher in patients with Chiari I than healthy controls<sup>17</sup>. These abnormally high velocities

decreased following decompression, but remained significantly higher than the CSF velocities of patients without Chiari I<sup>17</sup>. The authors maintain that pre-operative velocities may help predict which patients are likely to experience symptom improvement<sup>17</sup>. These findings were corroborated by Heiss *et al.* who found increased velocities but decreased CSF flow at the foramen magnum of patients with Chiari I<sup>19</sup>. In addition, in those with the disease, transmission of pressure across the foramen magnum was impeded, spinal CSF compliance was decreased, and subarachnoid and pulse pressure were both increased in the cervical cord<sup>19</sup>. Finally, syrinx fluid was found to flow in opposite directions during different phases of the cardiac cycle<sup>19</sup>. All these measures normalized following surgery, and in particular, fluid velocity and flow within the syrinx decreased<sup>19</sup>. With this, phase-contrast cine flow MRI not only can contribute to the pre-operative evaluation, but can also corroborate whether a satisfactory decompression has occurred. Cine flow can help identify patients in whom hindbrain pathology is directly responsible for their symptoms<sup>47</sup>.

### **Purpose**

The purpose of this study was to both shed light on extradural decompression as a potential first line surgical treatment for Chiari I malformation and to introduce objective measures to assess surgical success. In addition, the work was intended to contribute to a decision paradigm to prospectively determine which patients are most likely to benefit from surgery, as well as which surgical techniques would be optimal for which patients. Cine flow imaging to evaluate CSF flow will become increasingly useful to assess

surgical outcomes, especially for the multimodal evaluation of a successful extradural decompression.

### **Hypothesis**

I present a retrospective series of 18 patients with Chiari I malformation undergoing extradural decompression, for whom I assessed posterior fossa decompression as well as CSF dynamics using both T2-weighted and phase-contrast cine flow MRI. I proposed that patients who experienced clinical improvement would also have corresponding radiologic changes. Furthermore, I predicted that the degree of the MRI findings would vary across different levels of clinical improvement.

### **Specific aims**

In symptomatic patients with Chiari I malformation:

- 1) To assess clinical improvement and changes in functional status (as determined by a scale used by Noudel *et al.*<sup>48</sup> (Table 2)) following extradural decompression
- 2) To assess radiologic improvement in tonsillar herniation, posterior fossa area, cisterna magna area, and CSF flow following extradural decompression
- 3) To compare radiologic improvement across patients with varying levels of neurologic improvement

### **Patients and Methods**

I collected all of the retrospective clinical data, including pre- and post-operative symptoms, determined the functional grades and the change in clinical improvement

based on clinic notes, calculated the absolute and relative differences in posterior fossa area (PFA, mm<sup>2</sup>), cisterna magna area (CMA, mm<sup>2</sup>), and tonsillar herniation, calculated the change in functional grade, and analyzed the radiologic data according to the degree of clinical improvement. Another investigator, blinded to the clinical outcomes, collected the PFA, CMA, and tonsillar herniation measurements directly from the imaging as well as determined whether there was a change in CSF flow between pre- and post-operative imaging.

This study received IRB approval to collect, store, and analyze retrospective data obtained from clinic notes and radiographic reports.

#### *Patient population*

Eighteen patients with Chiari I malformation were treated surgically by the senior author (MLD). Age at admission ranged from 10 to 50 years old (mean = 28.8). Three of the eighteen patients (16.7%) were male, and fifteen (83.3%) were female (Table 1).

Table 1. Clinical summary

M = male, F = female; Y = yes, N = no

\*Comparison of pre- and post-operative T-2 weighted signal; confirmed with post-operative Cine

	Age	Sex	Headache	L'hermitte's sign	Hoffman's sign Left-N Right-Y	Syrinx	Functional Assessment		Clinical improvement	Post-operative complications
							Pre-op	Post-op		
1)	28	F	Y	N	Left-N Right-Y	N	IV	II	P+	N
2)	32	F	Y	N	N	N	I	I	N	N
3)	21	M	Y	N	N	N	II	I	N	N
4)	17	F	Y	N	N	N	II	I	P++	N
5)	34	F	Y	N	N	N	III	I	P++	N
6)	39	F	Y	N	Y	N	III	I	P++	Dysphagia, hypophonia
7)	42	F	Y	N	N	N	I	I	P	N
8)	50	F	Y	N	N	N	I	0	C	N
9)	14	F	Y	N	N	N	I	I	P+	N
10)	44	F	Y	Y	Y	N	IV	I	P+	N
11)	29	M	Y	N	N	N	I	I	P++	N
12)	10	F	Y	N	N	Y	II	I	P++	N
13)	33	F	Y	N	N	N	II	I	P++	N
14)	30	F	Y	N	N	N	II	I	P+	N
15)	15	F	Y	N	N	N	II	I	P	N
16)	19	F	Y	N	N	N	II	0	C	N
17)	42	F	Y	N	N	Y	II	I	P	N
18)	20	M	Y	N	N	N	II	I	P++	N

### *Neurologic assessment*

Symptoms and clinical exams were obtained from a retrospective review of the surgeon's clinic notes. Functional grade was determined using a grading scale previously reported in the literature<sup>48</sup>.

Table 2. Functional grading system from *Noudel et al., 2011*.

<b>Grade</b>	<b>Clinical examination</b>	<b>Functional Assessment</b>
0	Normal	Asymptomatic or complete recovery
I	Normal	Slight impairment
II	Objective signs	Slight impairment
III	Objective signs	Personal & socioprofessional disturbances
IV	Disabled	Persistence of autonomy
V	Disabled	Loss of autonomy

Patients received an initial post-operative follow-up visit in the clinic between 5-40 days (mean = 16.27), with continual follow-up at several month intervals thereafter. Clinical improvement was determined by a clinician who only assessed clinical symptoms and was blinded to the radiologic results. Patients were determined to have improved symptoms if it was clearly documented in any post-operative clinic notes. Patients were categorized as having no, partial, or complete resolution of their symptoms based on their last documented visit.

#### *Radiologic studies*

The diagnosis of Chiari I malformation was made using sagittal T1-weighted MRI brain studies, and was defined as tonsillar herniation of at least 5 mm below the level of the foramen magnum. The change in tonsillar herniation was calculated as the post-operative tonsillar herniation minus the pre-operative value. PFA and CMA were measured using T2-weighted imaging at the mid-sagittal plane. The method for determining the PFA and CMA was based on previously reported methods of measuring posterior fossa<sup>48</sup> and cisterna magna<sup>9</sup> volumes; the contents of the posterior fossa were measured as the neural structures along with the surrounding subarachnoid space, while excluding the thickness



of the skull<sup>48</sup>. The CMA was measured as the height of the cisterna magna multiplied by the depth of the cisterna magna perpendicular to the occipital dura divided by two (which assumes that the area of the cisterna magna is approximately a triangle)<sup>9</sup>. The increase in PFA was calculated as the post-operative PFA subtracted by the pre-operative PFA, whereas the relative PFA increase was calculated as the PFA increase divided by the pre-operative PFA. The relative CMA increase was calculated as CMA increase divided by the pre-operative CMA. Narrowing of the CSF spaces anterior and posterior to the spinal cord was assessed on T2-weighted imaging. Post-operative improvement in CSF flow was demonstrated by comparing T2-signal on pre- and post-operative images and by looking for differences in signaling in the posterior fossa and around the spinal cord. Phase-contrast cine flow imaging was compared pre- and post-operatively, when available, and assessed on all patients post-operatively to confirm adequate CSF flow. Post-operative MRI with cine was obtained approximately three months after surgery.

### *Surgical Technique*

All 18 patients underwent extradural surgical Chiari I decompression in the prone position. A standard suboccipital craniectomy was performed in order to ensure a wide decompression of the cerebellar hemispheres, brainstem, and midline structures. For all patients, a C1 laminectomy was also performed to decompress the cervical spinal cord. The atlanto-occipital ligament was divided and the underlying outer dura leaflet incised and reflected radially, while maintaining the integrity of the inner leaflet of the dura. The patients were then closed in a standard multi-layer fashion, similar to intradural procedures to guard against any potential CSF leak not noted intraoperatively.

### *Statistical analysis*

A univariate analysis was used to compare groups, and  $p < 0.05$  was considered significant. Microsoft Excel 2011 was used to organize the data in a de-identified encrypted file. Statistical analyses were performed using a TI-87 graphing calculator.

## **Results**

### *Demographics, Presenting Ailments, and Pre-operative Radiographic Assessment*

As stated earlier, 3 male (16.7 %) and 15 female (83.3%) patients were analyzed, with the average age being 28.8 (range 10 – 50) years. All 18 patients had headache as a presenting symptom, with subtypes including Valsalva/exertional, cluster, migraine, and tension. Headache locations were frontal, temporal, and occipital. Notably, many patients initially had multiple headache types. Additional reported symptoms included upper and lower extremity pain and/or numbness, neck pain, dizziness and vertigo, imbalance, visual symptoms, photosensitivity/phobia, nausea, fatigue, difficulty breathing, as well as compressive symptoms such as dysphagia and hiccups (Table 3). Twelve patients reported exacerbation of their symptoms with coughing, sneezing, exertion, or other Valsalva-like maneuvers. Only three patients had positive Hoffman's and/or L'Hermitte's signs. Other neurologic findings included downbeating nystagmus ( $n = 4$ ), hypophonia ( $n = 2$ ), and diminished sensation in the extremities ( $n = 1$ ). Four patients had pain and restriction with neck movement. Patients' functional grades ranged from a normal clinical exam with slight impairments (I) to disabled but with the persistence of autonomy (IV).

Table 3. Pre-operative symptoms

Symptom	Number of patients
Headache (all types)	18
Valsalva exacerbation of symptoms	12
Upper and/or lower extremity pain, numbness, or paresthesias	9
Neck pain	12
Compressive symptoms	
Dysphagia	5
Hiccups	1
Nausea	5
Dizziness/Vertigo	10
Imbalance	4
Photosensitivity/phobia	2
Visual symptoms	2
Fatigue	1
Difficulty breathing	1

Posterior fossa area ranged from 2489.57 to 3576.52 mm<sup>2</sup> (mean = 2997.78), which likely overlapped with the normal range (as this parameter can vary by age and gender)<sup>49</sup>. Notably, 10 patients (55.6%) had no measurable cisterna magna, with areas ranging from 0 to 15.53 mm<sup>2</sup> (mean = 4.95). Cerebellar tonsillar herniation ranged from 4.5 to 20.99 mm (mean = 9.86). Two patients had syringes associated with their condition (11.1%) (Table 1).

#### *Neurological outcomes*

Two of the 18 patients (11.1%) reported a complete resolution of their symptoms, 14 (77.8%) reported partial resolution, and 2 reported no improvement in their symptoms. Two patients had a recurrence of some symptoms that had initially improved. The latency to begin seeing an improvement in symptoms ranged from 7 days to 9 months. The

change in functional grade following treatment ranged from no difference to an improvement of 3 grades. Patients' post-operative functional grades ranged from 0 to II.

Sixteen patients (88.9%) noted some improvement in their headaches. Six patients had complete resolution of their headaches, five had decreased frequency, two were able to control their headaches with over the counter analgesics, and four (including one of the patients with decreased headache frequency) had resolution of some, but not all, headache subtypes. Refractory headache types included migraine (n = 2), exertional (n = 1), and temporally located headaches (n = 1). Other symptoms that were alleviated post-operatively included upper and lower extremity sensory changes (n = 7), neck pain (n = 4), dizziness/vertigo (n = 5), visual symptoms (n = 1), and dysphagia (n = 2) (Table 4).

Table 4. Post-operative improvement in symptoms

Symptom	Number of patients
Headache (all types)	16
Upper and/or lower extremity pain, numbness, or paresthesias	7
Neck pain	4
Dizziness/vertigo	5
Visual symptoms	1
Dysphagia	2

Two patients did not report any improvements in symptoms within 1 year of follow-up. Patients who did not report immediate clinical improvement continued to experience headaches (n = 2), neck pain/stiffness (n = 2), dizziness (n = 1) and photophobia (n = 1). No patient has had a worsening of symptomatology following surgery.

### *Radiologic Outcomes*

Seventeen patients (94.4%) had an increase in posterior fossa area following decompression. Post-operative posterior fossa area ranged from 2746.23 to 3572.89 mm<sup>2</sup> (mean = 3193.68) with an average increase of 209.31 mm<sup>2</sup> compared to pre-operative areas (Table 5). All 18 patients had an increase in cisterna magna area following decompression, with an average change of 41.84 mm<sup>2</sup>. Post-operative cisterna magna area ranged from 10.60 to 119.56 mm<sup>2</sup> (mean = 33.90). Following decompression, tonsillar herniation ranged from 4 to 18.67 mm (mean = 8.015). Sixteen patients saw an improvement in tonsillar herniation, with one patient experiencing no difference and one with a 0.3 mm worsening of herniation. Of the two patients with syringes, neither demonstrated resolution on 3-month post-operative imaging.

Pre-operative T2-weighted imaging demonstrated restricted signal anteriorly, posteriorly, or circumferentially around the cerebellar tonsils and spinal cord in 14 of 18 (77.8%) patients. Cine flow imaging was obtained pre- and post-operatively for 8 patients (44.4%) and only post-operatively for 10 patients (55.6%). Eleven (61.1%) patients demonstrated a post-operative improvement in CSF flow around the foramen magnum and/or cerebellar tonsils (Figures 1, 2). Four patients (22.2%) did not show a change in CSF dynamics following treatment, but had adequate flow on both pre- and post-operative imaging. Three patients (16.7%) continued to have restricted flow after decompression.

Table 5. Summary of radiologic and clinical treatment responses

M = male, F = female, PFA = posterior fossa area, CMA = cisterna magna area, Y = yes, N = no; N = no clinical improvement, P = partial, + = slight improvement, ++ = good improvement, C = complete

Age	Sex	Posterior fossa area (mm <sup>2</sup> )		PFA increase (mm <sup>2</sup> )	Relative PFA increase [Postop-Preop]		Cisterna magna area (mm <sup>2</sup> )		CMA increase (mm <sup>2</sup> )	Relative CMA increase [Postop-Preop]		Tonsillar herniation (mm)		Change in tonsillar herniation (mm)	Change in CSF dynamics	Functional assessment		Clinical improvement
		Preop	Postop		Preop	Postop	Preop	Postop		Preop	Postop	Preop	Postop					
1)	28	F	3256.11	3313.33	57.22	0.018	3.77	13.89	10.12	0.729	9.8	8.9	0.9	Y	IV	II	P+	
2)	32	F	2807.75	3064.55	256.8	0.091	9.19	63.13	53.94	0.854	4.5	4	0.5	N	I	I	N	
3)	21	M	3576.52	3564.79	-11.73	-0.003	9.95	51.29	41.34	0.806	11.8	8	3.8	Y	II	I	N	
4)	17	F	3347.82	3572.89	225.07	0.067	0	53.2	53.2	1.000	10.78	8.17	2.61	N	II	I	P++	
5)	34	F	3498.78	3559.37	60.59	0.017	6.26	40.91	34.65	0.847	10	9.71	0.29	Y	III	I	P++	
6)	39	F	2918.87	3194.07	275.2	0.094	0	42.6	42.6	1.000	8.13	8	0.13	Y	III	I	P++	
7)	42	F	2523.1	2866.31	343.21	0.136	0	12.6	12.6	1.000	20.99	18.67	2.32	Y	I	I	P	
8)	50	F	2847.72	3074.21	226.49	0.080	0	14.97	14.97	1.000	17.75	15.84	1.91	Y	I	0	C	
9)	14	F	2489.57	3015.68	526.11	0.211	0	10.6	10.6	1.000	11.58	11.88	-0.3	Y	I	I	P+	
10)	44	F	2581.62	2772.73	191.11	0.074	15.53	29.69	14.16	0.477	13.34	13	0.34	N	IV	I	P+	
11)	29	M	3127.76	3132.52	4.76	0.001	9.71	39.99	30.28	0.757	5.23	4.6	0.63	Y	I	I	P++	
12)	10	F	2919.87	3169.25	249.38	0.085	0	67.33	67.33	1.000	6	4	2	N*	II	I	P++	
13)	33	F	2883.07	3264.16	381.09	0.132	6.95	70.4	63.45	0.901	5	5	0	Y	II	I	P++	
14)	30	F	2538.6	2746.23	207.63	0.082	5.74	14.44	8.70	0.602	9	5	4	Y	II	I	P+	
15)	15	F	3391.44	3546.79	155.35	0.046	0	119.56	119.56	1.000	7	4	3	N*	II	I	P	
16)	19	F	2844.41	3040.92	196.51	0.069	0	67.8	67.80	1.000	6.5	4.5	2	N*	II	0	C	
17)	42	F	2868.91	2997.54	128.63	0.045	0	33.39	33.39	1.000	10	5	5	Y	II	I	P	
18)	20	M	3237.19	3531.29	294.10	0.091	0	74.4	74.40	1.000	10	6	4	N*	II	I	P++	

### *Neurological Outcome and Radiographic Correlation*

We assessed the patients' radiologic changes relative to clinical improvement and change in functional grade. There was no statistically significant difference in the relative change in posterior fossa area between patients with no, partial, and complete improvement in their symptoms ( $p > 0.05$ ). Patients with complete resolution of their symptoms had a significantly greater relative increase in their cisterna magna area following surgery than those with partial symptomatic improvement ( $p = 0.022$ ) (Figure 3). Patients with a complete resolution of their symptoms had significantly smaller cisterna magna areas pre-operatively compared with those who had no ( $p = 0.025$ ) or partial ( $p = 0.020$ ) improvement (Figure 4). Pre-operative posterior fossa area was not significantly different between any of the groups.

Ten (91%) of the 11 patients with improved CSF dynamics also had either partial or complete resolution of their symptoms. All four of the patients with adequate but unchanged CSF flow experienced at least a partial improvement in their symptoms. Two (66.7%) of the three patients who continued to have restricted flow post-operatively experienced a partial improvement, with one showing no change in their clinical symptoms.

There were no significant effects of the degree of improvement in functional grade on radiologic changes. One patient experienced a change of 3 grades (from Grade IV to

Grade I); because she was the only patient to experience this substantial an improvement, she was excluded from the between-group comparisons.

#### *Surgical complications*

There were no cases of CSF leak or post-operative infection, including meningitis. One patient developed dysphagia and hypophonia post-operatively that was thought to be due to traumatic injury to her vocal cords during intubation. None of the patients, including those without clear symptom improvement, have since sought reoperation.

### **Discussion**

Chiari I malformation is a common neurological ailment that often presents with headache and signs of brainstem compression, which is thought to be secondary to an impairment in dynamic CSF flow. Patients with Chiari type I malformations demonstrate a thickened dural band at the craniovertebral junction with histological evidence of hyalinosis, calcification, and/or ossification<sup>50</sup>. To restore CSF dynamics, surgical decompression is sought, with the mainstay of therapy being a posterior intradural decompression. In this case series, we demonstrate that an extradural decompression may be appropriate for increasing the area of important CSF spaces such that there is an improvement in CSF dynamics, and consequently, neurological symptoms.

#### *Extradural Chiari decompression is a viable and reproducibly effective procedure*

Extradural decompression, while the most minimally invasive of the types of surgical decompression, nonetheless has been shown to be an effective technique for achieving symptomatic relief<sup>34,51,52</sup>. Theoretically, the effectiveness of the dural splitting technique



depends on relieving the outer dural layer compression of the cord, and the expandability of the newly decompressed inner dural layer. When assessing the biomechanical properties of entire versus split dura mater, the former appears to be more fragile under stress, while the latter allows a greater volume of deformation at a lower stress level<sup>53</sup>. In the case series by Isu *et al.*, all seven patients demonstrated Chiari I malformation<sup>28</sup> and subsequently underwent a suboccipital craniectomy and either a C1-C2 or C1 laminectomy, followed by removal of the outer dural layer. The authors used intraoperative ultrasonography to confirm satisfactory decompression, as visualized by the pulsation of the cerebellar tonsils after removal of the outer dural layer. Six of the seven patients (86%) experienced improvement in their neurologic symptoms. Remarkably, Isu *et al.* showed that within several months, all seven patients had a reduction in their syrinx size. Lee *et al.* similarly found that there was no difference in the rate of syrinx improvement or resolution between those undergoing intradural and extradural decompression<sup>54</sup>. This is congruent with our series in which 88.9% patients experienced either partial or complete improvement in their symptoms. Interestingly, Munshi *et al.* found that in the three patients for whom there was improvement in the hydromyelia, there was also an increase in posterior fossa volume<sup>31</sup>. While neither of the patients in our series had resolution of their syringes, both had increased posterior fossa area. Three months follow-up may have been too short a time to see a noticeable difference in the size of the syringes, which may eventually decrease on subsequent imaging.

Chauvet *et al.* reported a series of eleven patients treated surgically with extradural decompression<sup>4</sup>, who initially presented with various symptoms, including headache and/or neck pain secondary to brainstem compression and/or syringomyelia. All patients experienced some improvement in symptoms, with six of the eleven (54.5%) patients having complete resolution. There were no fluid leaks or collections in the post-operative site, and no cases of aseptic meningitis. Post-operative MRI showed that 10 of the 11 (91%) patients experienced extensive decompression with reestablishment of the cisterna magna in four patients, which is consistent with our findings. In our series, all 18 patients experienced an increase in cisterna magna area following extradural decompression. Furthermore, we found that patients with a complete resolution of their symptoms had a significantly greater increase in relative cisterna magna area compared to those with partial resolution of their symptoms. This difference did not hold true for the absolute increase in cisterna magna area. In fact, patients with complete symptom relief had significantly smaller pre-operative cisterna magna area compared to those with patients with either partial or no symptom improvement. Our results suggest that if symptom relief is desired, adequate restoration of CSF spaces should be the goal of satisfactory decompression.

*Extradural Chiari decompression is safer and more cost effective*

When treating Chiari I malformation without syringomyelia, some authors recommend posterior fossa decompression with extradural durotomy, proceeding only with intradural exploration when adequate CSF flow cannot be achieved<sup>55</sup>. In extradural decompression, some suggest using intraoperative ultrasonography to determine adequate CSF flow<sup>36,55</sup>,

whereas with intradural techniques, it is difficult to assess CSF flow intraoperatively<sup>28</sup>. While there is no consensus on which surgical technique is more effective<sup>8,30,34,52</sup>, an external Chiari decompression via dural-splitting avoids the complications of a dural-penetrating craniectomy and laminectomy<sup>4,28</sup>, where there is the potential for CSF leak, subsequent pseudomeningocele, as well as meningitis<sup>8,28</sup>. The need for potential reoperation given inadequate symptom resolution is balanced by the lower complication rate<sup>18,34</sup>. However, others argue that the complication rate using an intradural opening with duroplasty is near negligible<sup>30</sup>. In a comparison of duroplasty with intradural manipulation and dural splitting, extradural decompression led to faster operating times, shorter hospital stays, and therefore lower total hospital costs while yielding comparable clinical results<sup>8,10</sup>. More specifically, dural splitting for Chiari I without syringomyelia was deemed to be safer and more cost effective, not including the cost savings from fewer overall complications<sup>10</sup>.

*Pre-operative assessment and prediction of surgical success – a multimodal approach*

In their original series Gardner and Goodall used encephalography to visualize the obstructive hydrocephalus, which they used to inform their decision to operate<sup>27</sup>. Since then, various techniques to assess the severity of compression, tonsillar herniation and flow obstruction have been described to both predict and determine operative success<sup>35,43</sup>. In their series of 11 patients undergoing intradural decompression without duroplasty, Noudel et al. measured the increase in posterior fossa volume (PFV) and correlated it with clinical outcomes<sup>48</sup>. They found that the degree of PFV increase positively correlated with symptomatic improvement ( $p = 0.014$ ). There was a greater increase in

PFV with a smaller starting volume, and that an increase of at least 15% was needed for a complete recovery. The authors further suggest that an optimal PFV increase could be predicted to improve patient symptoms. We did not find a correlation between posterior fossa area and symptom responsiveness, however this may be related to the lack of dural opening in our series. Rather than just visualizing the compression of hindbrain structures, high resolution MRI may prove additionally useful by showing arachnoid adhesions and other obstructions that may benefit from dural opening. In addition, this approach may show the pathophysiologic effects on surrounding tissue.

While traditional imaging techniques may be employed to assess the degree of tonsillar herniation and thus identify the Chiari I malformation, they may not be sufficient when evaluating whether or not to operate<sup>18,41</sup>. After all, a “ball-valve” mechanism exacerbated by Valsalva-like maneuvers may be responsible for a dynamic obstruction of CSF flow and subsequent headache symptoms<sup>18,41</sup>. Dynamic imaging studies therefore provide additional utility in assessing the degree of impairment. In addition to MRI, some describe using ultrasonography with color Doppler imaging to demonstrate anatomical compression, as well as impaction of CSF flow<sup>30</sup>.

Phase-contrast cine MRI is an alternative study with even more clinical utility given that a flow-sensitive pulse demonstrates the amplitude and direction of CSF and blood flow relative to anatomical structures<sup>41</sup>. Cine flow imaging may have the greatest utility in an asymptomatic or minimally symptomatic patient with an incidental finding of tonsillar herniation, when surgical treatment based only on abnormal anatomy has uncertain

benefit<sup>41</sup>. While traditional MR imaging is unable to capture Valsalva-related changes in CSF flow, cine flow can measure the more dynamic fluctuations relevant to patients with CSF obstruction<sup>56</sup>. Although we were only able to obtain pre-operative cine flow images for 8 of 18 (44.4%) patients in this study, multiple studies have demonstrated that cine flow MRI has additional value for evaluating which patients are most likely to benefit from surgical intervention. Patients with Chiari malformation demonstrate abnormal CSF flow dynamics on cine compared to healthy controls<sup>7,57</sup>, and cine parameters can be used to identify those with the disease<sup>58</sup>. Ventureyra *et al.*, showed that symptomatology correlated well with abnormal cine flow imaging<sup>5</sup>. Asymptomatic patients almost invariably had normal cine flow studies and did best without surgical treatment<sup>5</sup>. Conversely, symptomatic patients treated with suboccipital decompression with duroplasty had improvement of symptoms, as well as on post-operative imaging<sup>5</sup>. Patients with abnormal CSF flow on pre-operative cine are the most likely to have maximal benefit from surgical treatment, as these patients are less likely to have symptom recurrence<sup>26</sup>. In patients with Chiari type I undergoing decompressive surgery, computational modeling of CSF flow dynamics demonstrates lower post-operative CSF velocities and pressures<sup>14</sup>. In our study, of the 14 patients with restricted flow pre-operatively, 12 (85.7%) had either a partial or complete improvement in their symptoms. Nevertheless, of the 11 patients with improved flow, 10 reported at least a partial improvement in their symptoms. This suggests that restoration of CSF dynamics may be nonetheless beneficial. One limitation of cine flow MRI is that it only measures dependent flow, as the patient is lying down during the study. It is therefore possible that some patients may falsely show adequate flow that would otherwise be compromised in

the standing position. Furthermore, CSF flow dynamics will certainly vary based on other physiologic parameters; therefore, establishing what is “normal” will require an analysis that incorporates a diverse set of healthy controls. Subarachnoid CSF velocities were obstructed to varying degrees in Chiari I and have even been reported to be normal in 19-33% of pediatric patients with Chiari I<sup>18</sup>. Furthermore, some studies have suggested lower systolic velocities, whereas others have reported higher systolic velocities in patient cohorts, and none have demonstrated the entry-point of CSF flow into a syrinx<sup>18</sup>. Therefore, while cine-flow MRI is a tool with a lot of potential to inform our understanding of the pathophysiology of Chiari I, there is much work to be done.

## **Conclusion**

Extradural decompression provides the benefits of surgical decompression while avoiding the complications of intradural techniques. The dural-splitting surgical technique has the potential to be the first line treatment for Chiari I malformation. Our data suggest that surgical enlargement of CSF spaces along with either adequate or improved CSF dynamics allows for the greatest chance of success following extradural decompression. Patients with either absent or severely reduced cisterna magna areas, in addition to restricted flow, may be the most likely to benefit from a basic decompression. In cases where patients remain symptomatic despite the lack of radiologic findings, a more invasive technique might better address intradural pathology. Nevertheless, further studies with larger cohorts, ideally randomized to the two techniques (intradural versus extradural) would corroborate this study and hopefully bring the extradural Chiari decompression technique into the mainstream spotlight.

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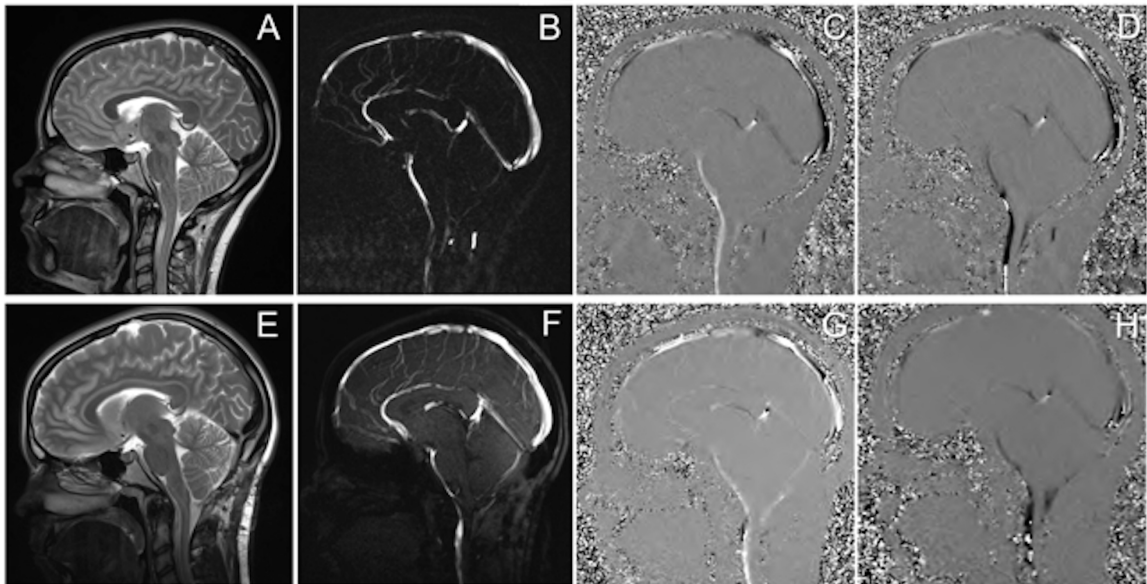


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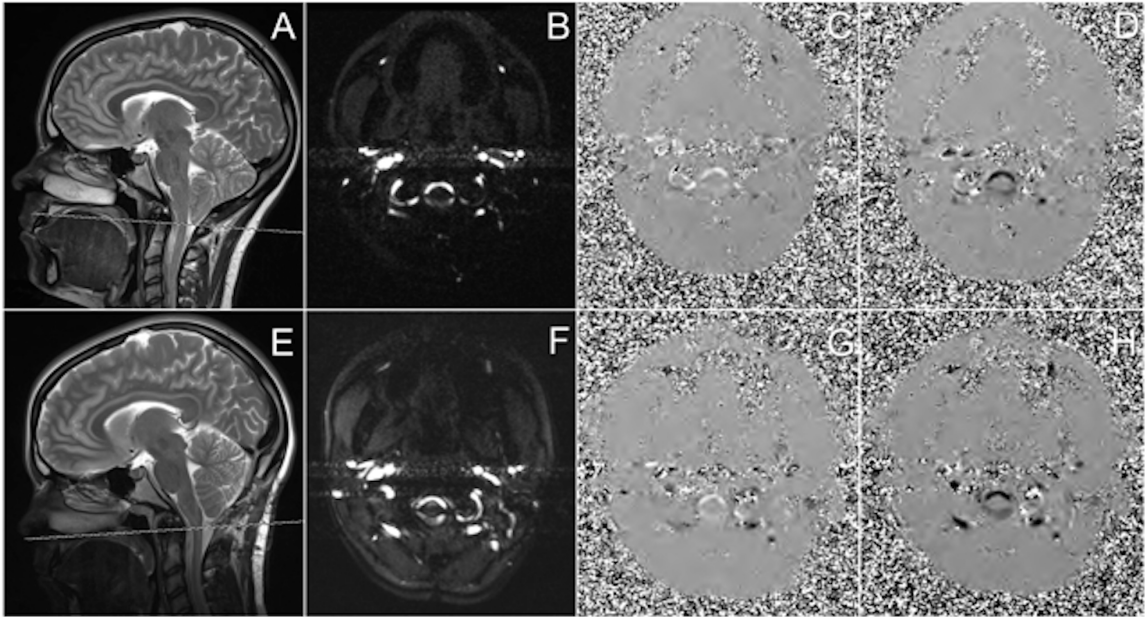
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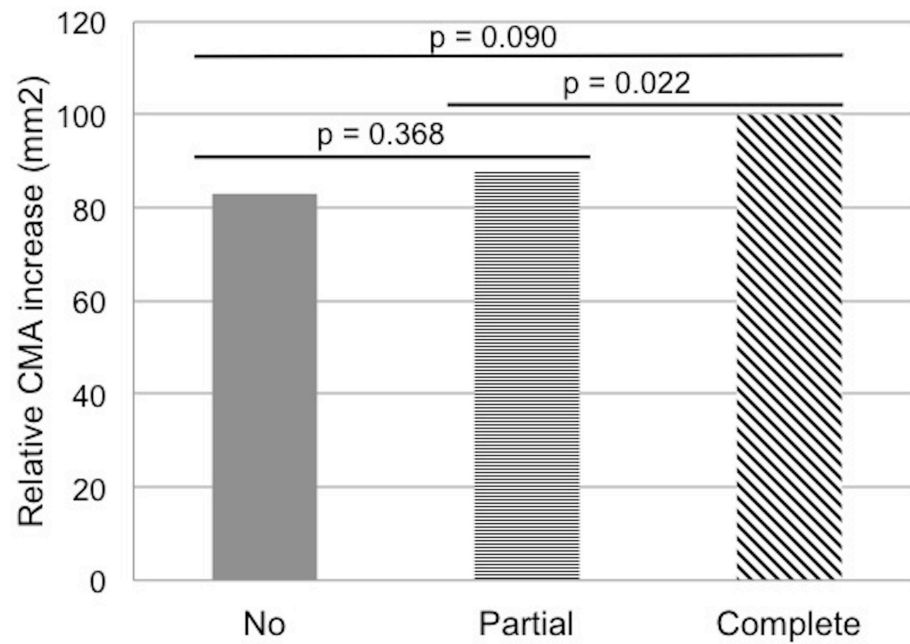
## Figures



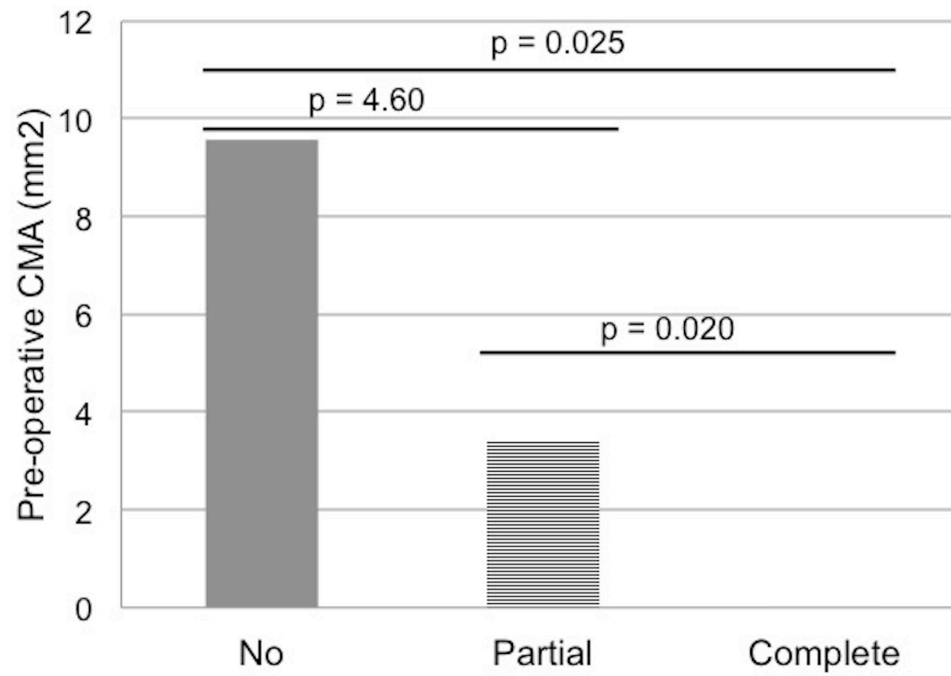
**Figure 1.** Pre- (A-D) and post-operative (E-H) sagittal MRI in a representative patient with clinical and radiologic improvement; T2-weighted (A, E), composite cine (B, F), phase-in cine (systole) (C, G), and phase-out cine (diastole) (D, H). Post-operative T2-weighted (E) imaging demonstrates some improvement in tonsillar herniation. Post-operative composite (F), phase-in (G) and phase-out (H) cine demonstrate robust flow at the cervicomedullary junction and around the tonsils as well as improved biphasic flow. Flow in the cerebral aqueduct is minimally improved and a blush of flow is now noted in the 4th ventricle



**Figure 2.** Pre- (A) and post-operative (E) T2-weighted sagittal images indicate the level of pre- (B-D) and post-operative (F-H) axial cine flow MRI in a representative patient with clinical and radiologic improvement; Composite cine (B, F), phase-in cine (systole) (C, G), and phase-out cine (diastole) (D, H)



**Figure 3.** Relative increase in CMA (mm<sup>2</sup>) across clinical improvement levels



**Figure 4.** Pre-operative CMA (mm<sup>2</sup>) across clinical improvement levels