OBJECTIVE: Chiari malformation type I (CM-I) is a craniocervical junction disorder associated with descent of the cerebellar tonsils >5 mm. The prevalence of CM-I is common, including 0.5%—3.5% in the general population, 0.56%—0.77% on magnetic resonance imaging, and 0.62% in anatomic dissection studies. We sought to measure our surgical outcomes related to resolution/improvement of headaches, neurologic outcomes, and syringomyelia compared with reported adult CM-I studies from 2000—2019.

METHODS: From December 2003 to June 2018, the first author (K.I.A.) performed 270° circumferential decompression on adult (>18 years) patients with CM-I. At admission and follow-up, all parameters were numerically evaluated; headaches were self-reported on the visual analog scale, neurologic condition was evaluated using Karnofsky Performance Status and European Myelopathy Score, and syrinx width (if present) was measured on magnetic resonance imaging by grades I—IV. All parameters were analyzed, compared, and statistically tested. We compared results with our previously reported and updated systematic review of operative adult CM-I studies (studies from 2000 to 2019).

RESULTS: In our series, 118/121 (98%) experienced headache improvements and 100% experienced neurologic improvements. Complete syrinx resolution was experienced by 35/43 (81%); 8 (19%) showed significant improvement. In data from reported studies (2000—2019), only 79% experienced headache resolution, 77% improvement of neurologic status, and 74% resolution/improvement of syrinx (mean).

CONCLUSIONS: Our modified 270° circumferential microsurgical foramen magnum decompression for adult CM-I appears to be beneficial in improvement of outcomes, namely in resolution of the syrinx, neurologic symptoms, and headaches. We also confirm the association of body mass index with CM-I. Further studies are needed to confirm our results.

INTRODUCTION

Chiari malformation type I (CM-I) is a craniocervical junction disorder characterized specifically by a descent of the tonsils below the foramen magnum of >5 mm into the spinal canal. The prevalence of CM-I is common and has been reported to be 0.5%—3.5% in the general population, 0.50%—0.77% on magnetic resonance imaging (MRI) studies, and 0.62% in anatomic dissection studies. The age distribution of operatively treated patients with CM-I is approximately even among the adult and pediatric population.

The results of operative treatment for CM-I have been reported to be good over the past 20 years. Improvement and/or resolution of symptoms is generally reported in around 75% of cases, with complete resolution in around 25% of cases. The goal of surgical treatment is to improve quality of life and prevent complications such as hydrocephalus and upper cervical stenosis. However, long-term outcomes and the role of surgical intervention remain a topic of debate.

Key words
- 270° foramen magnum decompression
- Adult
- Body mass index
- Chiari malformation type I
- Operative treatment
- Series

Abbreviations and Acronyms
- BMI: Body mass index
- CMC: Cerebellomedullary cistern
- CM-I: Chiari malformation type I
- CSF: Cerebrospinal fluid
- EMS: European Myelopathy Score
- IQR: Interquartile range
- MRI: Magnetic resonance imaging

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of syringomyelia has been found in 74% of adult populations, of neurologic deficit in 77%, and of headaches in 79%. In operative series of adult CM-I, patients were treated with suboccipital/foramen magnum bony decompression (100%), dural opening (97%), and arachnoid opening and dissection (70%). Occasionally, tonsillar resection has been used (23%). Nevertheless, up to 26% of adult patients do not experience postoperative improvement of syringomyelia, neurologic deficit, and headaches, respectively.1,33

In an attempt to further improve postoperative outcomes, we prospectively adopted a modification of adult CM-I operative treatment by using a circumferential decompression of the foramen magnum of 270°. We present our experiences with this surgical modification in a series of patients. We included details of the technique, patient demographics (including neurologic symptoms, headaches, syrinx presence, and body mass index [BMI, calculated as weight in kilograms divided by the square of height in meters]) as well as outcomes related to these variables. We also compared the variables with the outcomes of reported CM-I adult operative studies from our previously reported and updated systematic review.1

METHODS

This study was approved by the appropriate hospital institutional review board. This is a 14.5-year prospective study, initiated in December 2003 and completed in June 2018. The study included adult patients with CM-I (>18 years old) with or without syringomyelia and with neurologic deficit who were subsequently operated on by the senior author.

Preoperative and Postoperative Analysis and Follow-up of Patients with CM-I

Neuroradiologic Protocol. A thorough preoperative neuroradiologic protocol was designed by an independent neuroradiologic team at our institution, who were not part of this study; only their readings were deemed conclusive. For all patients, readings included anteroposterior and lateral flexion, and extension radiography with particular attention to evaluating atlantoaxial interval and vertical movements of the odontoid process. MRIs of the brain and C-spine, including T1 and T2-weighted axial, coronal, and sagittal cuts across the craniocervical junction were included, as were MRIs of the thoracic spine if the syrinx extended downward. Computed tomography scans of the cervical spine, including thin cuts with sagittal and coronal reconstructions, were performed to evaluate for bone and/or facet abnormalities. Postoperatively, MRI of the C-spine and the anteroposterior, lateral, and flexion-extension lateral C-spine radiography were performed routinely at 2, 6, and 12 months, and then yearly as needed, evaluating the same parameters that were examined preoperatively. More frequent neuroradiologic imaging was performed on a case-by-case basis. CM-I neuroradiologic studies were analyzed and included determinations of the descent of cerebellar tonsils, vertical extent of syringomyelia, and width of the syrinx according to a previously established scale that divides syrinx width into 4 grades: grade I (0%–25%), grade II (26%–50%), grade III (51%–75%), and grade IV (76%–100%).4 The neuroradiologic outcomes of syrinx formation, as well as their resolution or decrease, were precisely recorded based on these criteria and included in our analysis. In addition, we compared and analyzed our syrinx results with other reported adult CM-I operative series from a previously reported systematic review. Particular attention was paid to evaluate and exclude preoperative and postoperative evaluation of Co–C2 instability.

Headaches and Satisfaction Evaluation. Before every preoperative and postoperative examination, patients self-reported the degree of their headaches with a visual analog scale. Patients also self-reported their satisfaction with surgery as “not satisfied” (would not have the surgery again), “satisfied” (most probably would have the surgery again), and “very satisfied” (would definitely have the surgery again).

Neurologic Examination Protocol. Patients were evaluated after reviewing medical history and performing neurologic examination. The severity of neurologic deficit was determined by Karnofsky Performance Status,14 which was evaluated by an independent neurologist. Patients with a Karnofsky Performance Status of 85–100 had a mild neurologic deficit, 65–84 had a moderate deficit, and <65 had a severe neurologic deficit. Furthermore, all patients were evaluated before surgery and at follow-up regarding symptoms of myelopathy using the European Myelopathy Score (EMS).35

BMI

Data concerning a patient’s body weight and BMI were analyzed and used as an indicator of obesity. Using World Health Organization criteria,8 we divided all patients into categories based on their BMI as “underweight” (BMI, <18.5), “normal body weight” (BMI, 18.5–24.9), “overweight” (BMI, 25–29.9), “class I obesity” (BMI, 30–34.9), “class II obesity” (BMI, 35–39.9), and “class III obesity” (BMI, >40). We also analyzed the age of patients, gender, length of stay, presence of postoperative complications, length of follow-up, history of smoking, and presence of diabetes mellitus.

Exclusion Criteria

Patients with complex CM-I, such as basilar invaginations or platybasia, and/or patients who showed any radiologic signs of craniospinal (Co, C1, C2) instability after neuroradiologic expert review that required instrumented stabilization were excluded from this study. In addition, patients with secondary CM-I, such as idiopathic intracrani hypertension, intracranial hypotension and brain sagging, hydrocephalus, Ehlers-Danlos syndrome, or patients <18 years of age, were excluded from the study.

Statistical Methods

Categorical data are represented by absolute and relative frequencies. Numeric data are described by median and interquartile range (IQR) boundaries. The variance of category variables was tested by the χ2 test, as well as by Fisher exact test. The normality of the distribution of numeric variables was tested by Shapiro-Wilk test. The differences of normal distribution of numeric variables between the 2 independent groups were tested by the Student t test and, in cases of a deviation from the normal distribution, by Mann-Whitney U test. The differences of numeric variables between the 2 dependent groups were tested by the Wilcoxon test.
The differences between numeric variables in cases of ≥3 independent groups were tested by the Kruskal-Wallis test. All P values were 2-sided. The level of significance was set at \( \alpha = 0.05 \). Statistical analyses were performed with MedCalc Statistical Software version 18.2.1 (MedCalc Software, Ostend, Belgium) and SPSS version 23 (IBM Corp., Armonk, New York, USA).

Operative Technique

The operation consisted of a 270° circumferential microsurgical decompression of the foramen magnum and opening of the dura and arachnoid membrane (Figure 1), followed by microsurgical decompression, and opening of the foramina of Magendie and Luschka and the bilateral cerebellomedullary cisterns (CMCs). The operation thus re-established cerebrospinal fluid (CSF) flow circumferentially across the craniovertebral subarachnoid spaces.

The patient is first placed in a supine position to harvest a piece of fat from a small abdominal paraumbilical incision for later use. The patient is then placed prone on thoracic-abdominal bolster rolls with the head affixed in a 3-point Mayfield head rest. The posterior suboccipital area is shaved and prepared, and an incision is marked from the external occipital protuberance to the C2 spinous process. After the skin and subcutaneous tissue are incised, an incision is made between the 2 leaflets of the nuchal ligament and paraspinous muscles, which are separated from the squama of the suboccipital bone and C1 lamina with retractors spreading in the laterolateral direction, proximally and distally (Figure 1A). Then, a C1 laminectomy is performed. Using a high-speed drill with a 7-mm coarse round diamond drill bit, an oval sulcus is created in the suboccipital bone with the base toward C1 extending halfway between the rim of the foramen magnum and

![Figure 1. The operative exposure. (A) The exposed suboccipital bone, C1 lamina, and the lower half of the suboccipital ligament is shown and the suboccipital and paraspinous muscles are retracted. The bone is removed along the dashed line. (B) Next, the suboccipital bone flap and C1 lamina are elevated/removed and the suboccipital ligament incised and resected along the dotted line. (C) The posterior 1/5 of the medial aspect of the condyle is drilled to facilitate exposure of both cerebellomedullary cisterns after dural opening. With the dura now exposed, it is incised along the Y-shaped dashed line. (D) The dura and arachnoid membrane are opened. Note the release of arachnoid adhesions, freed entrance to the fourth ventricle, and the Magendie foramen and Luschka foramina/lateral cerebellomedullary cisterns including both vertebral arteries and posterior inferior cerebellar arteries. The cerebellum, brainstem, lower cranial nerves, and craniovertebral subarachnoid spaces have been decompressed and cerebrospinal fluid (CSF) circulation re-established 270°. This fat graft technique was adopted after 3 patients developed pseudomeningocele/CSF leak after surgery and needed reoperation (1 man and 2 women; cases 4, 5, and 11). Since then, we have adopted the fat graft technique as described here in 119 cases and have not experienced another case of pseudomeningocele/CSF leak.36]
the external occipital protuberance (approximately 2 cm or 1 inch). Kerrison rongeurs of 2 mm are used to resect the inner lamina of the bone, followed by elevation of the bone flap.

The posterior medial 1/5 of the occipital condyle is drilled using a 3-mm coarse diamond drill bit using a microsurgical technique. Any bleeding is easily controlled with Gelfoam powder (Pfizer, New York, New York, USA). The exposed suboccipital ligament is then resected (Figure 1B and C).

The dura is opened in the shape of the letter Y and tacked to surrounding soft tissues with 4-0 Nurolon stitches (Ethicon US, LLC, Johnson & Johnson, Bridgewater, New Jersey, USA) (Figures 1C and D and 2A and B). Using a microsurgical technique, the arachnoid membrane is opened in the midline and then in the bilateral CMCs, releasing a gush of CSF from the CMCs and decompressing both Luschka foramina (Figure 2C). Arachnoid adhesions are divided, and bilateral vertebral arteries with the posterior inferior cerebellar artery origin are visualized. This process is enabled with previous drilling of the small parts of posterolateral condyles, which provides an unobstructed view to the CMCs (Figure 3A and B). The entrance in the fourth ventricle is opened by releasing arachnoid adhesions and any arachnoid membrane if present. The tips of the cerebellar tonsils are coagulated to shrivel them. Decompression of craniospinal subarachnoid spaces to about 270° is visualized (Figure 3A and B).

Closure is performed with a triangular piece of bovine pericardium allograft (Durepair [Medtronic, Minneapolis, Minnesota, USA]) precut to fit the allotted space using 4-0 Nurolon running and single sutures. A Valsalva maneuver ensures a watertight closure. The previously harvested abdominal fat is placed cautiously on top of the dura along with fibrin glue to eliminate the dead space created by bone removal and to prevent CSF leak or pseudomeningocele formation. Care is taken to not to overpack the space.

**Follow-Up Protocol.** Follow-up occurs 3 weeks postoperatively for suture removal. Subsequently, the patient is seen 3 months later for MRI study, then in 9 months, and subsequently on a yearly basis, including clinical and neuroradiologic examinations protocols.

**RESULTS**

**Syringomyelia**

A total of 130 patients were operated on during a 14.5-year period (Figure 4). There were no statistically significant differences in the age of the patients in relation to gender or presence of syringomyelia, which was present in 35% of the patients overall.
Figure 4. Imaging from illustrative cases in the series. Note complete resolution of preoperative syrinx in postoperative imaging. Case 10: preoperative (A) and postoperative (B) sagittal T2-weighted spinal magnetic resonance imaging (MRI) of a 41-year-old woman with severe suboccipital headaches and myelopathy. Case 11: preoperative (C) and postoperative (D) sagittal T2-weighted spinal MRI of a 35-year-old woman with myelopathy. Case 63: preoperative (E) and postoperative (F) sagittal T2-weighted spinal MRI of a 33-year-old man with myelopathy. Note preoperative signal change at C2 spinal cord dorsally, which resolved on postoperative MRI. Case 96: preoperative (G) and postoperative (H) sagittal T2-weighted spinal MRI of a 57-year-old man with myelopathy. Note preoperative signal changes in spinal cord at the C1-C2 level, which resolved postoperatively. Case 125: sagittal MRI of a 25-year-old woman with myelopathy. (I–K). T2-weighted sagittal MRI of C-spine (L). Note the de novo CM-I formation after body mass index gain of 10 points over 2 years (body mass index 22–32) T2 and T1 sagittal MRI of C-spine and brain (J and K). Also note postoperative resolution of CM-I on T2-weighted MRI of C-spine (L).
The most common age range was 31–40 years, followed by 21–30 and 41–50 years (Figure 5).

More women had syringomyelia than did men (32 vs. 11), but syrinx was proportionally more common among men than among women (11/18 [61%] vs. 32/112 [29%]), which was statistically significant (Fisher exact test, \( P = 0.02 \)) (Table 2). Patients with syringomyelia were younger, although the difference did not reach statistical significance (Table 2).

The median length of stay was 3 days (IQR, 3–4 days), which was not significantly related to the presence of syrinx. The median time to resolving/improving syrinx was 4 months (IQR, 3–8 months). Longer follow-up time with patients with syrinx was statistically significant (Mann-Whitney \( U \) test, \( P = 0.03 \)) (Figure 4A–D, Table 2).

Syrinx completely resolved in 35 patients (81%) from the subgroup of 43 patients who had syrinx, whereas 8 patients (19%) showed significant improvement. A statistically significant resolution or improvement of syringomyelia was noted post-operatively versus preoperatively (Wilcoxon test, \( P = 0.01 \)) (Table 3; Figures 6–8).

### Headache

Before surgery, headache was reported in 121 patients (93.1%), 36 of whom had syrinx. After surgery, there was improvement in 118 patients (98%), except for 3 (2%), who experienced no change. Self-reported pain reduction of headaches between groups was statistically significant (Wilcoxon test, \( P < 0.001 \)) (Table 2, Figure 9).

### Neurologic Status

Only 6 patients (5%) did not have neurologic deficit before surgery; the remaining patients (n = 124; 95%) had neurologic deficit. Of those patients, 30 (24%) had mild (7 with syrinx), 32 (26%) had moderate (10 with syrinx), and 62 (50%) had severe (26 with syrinx) neurologic deficits.

Of the 43 patients with syrinx, all had neurologic deficits. All patients had complete resolution or significant improvement of their preoperative neurologic deficit. There was no statistically significant difference in the neurologic deficit before surgery between patients with and without syringomyelia (Table 2, Figures 6 and 7). Smoking was reported in 33 patients (25.4%), 9 (20.9%) of whom had syrinx. There is no statistically significant difference related to smoking given the presence of syringomyelia (Table 2, Figure 9).

Mean preoperative EMS was 11.32 (SD, ±3.607). Mean EMS value at follow-up was 16.42 (SD, ±1.419). This improvement in neurologic score was shown to be statistically significant and that there is a correlation between 2 parameters (Figure 9).

### BMI

Twenty-seven of the 130 patients (20.8%) had normal BMI. One patient (0.8%) was underweight, whereas all others were overweight or obese (88%) (Table 2). The vertical range of extension of the syringomyelia cavity was on average 7 vertebrae, ranging from 1 to 18 vertebrae. In 23 of the 43 patients with syrinx (53.5%), the syringomyelia cavity started with the first cervical vertebra (C1), and in 2 (5%) of the examinees, the syringomyelia cavity was...
stretched to the eleventh thoracic vertebra (T11). The syringomyelia cavity started from the second cervical vertebra (C2) in 8 patients (18.6%), from the third cervical vertebra (C3) in 6 patients (14.0%), from the fifth cervical vertebra (C5) in 5 patients (11.6%), and from the seventh cervical vertebra (C7) in 1 patient (14.0%).

Satisfaction with Surgery
For postsurgical patient satisfaction, 97% of respondents were satisfied with the procedure; only 3 respondents were unsatisfied because of lack of headache improvement (Table 3). Obviously, satisfaction rate was directly proportional to resolution/improvement of preoperative headaches and neurologic status.

Comparison with Other Adult CM-I Studies
The duration of our study was 14.5 years. The mean duration of reported adult studies was 8 years (range, 1−38 years, Figure 8). The mean age of adult patients in previously reported studies was 37.8 years (IQR, 36−40 years); in our study, the mean age was 38 years (IQR, 29−44.3 years). In other studies, the median number of respondents was 20 patients per study (IQR, 15−45 patients; range, 1−177); in our study, there were 130 total

Table 2. Distribution of Patients and Presence of Syringomyelia

<table>
<thead>
<tr>
<th>Demographic</th>
<th>Presence of Syringomyelia</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No</td>
<td>Yes</td>
<td>Total</td>
<td>P Value</td>
</tr>
<tr>
<td>Patient sex distribution in the syringomyelia subgroup, n (%)</td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>Male</td>
<td>7 (8)</td>
<td>11 (26)</td>
<td>18 (14)</td>
<td>0.02*</td>
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<tr>
<td>Female</td>
<td>80 (92)</td>
<td>32 (74)</td>
<td>112 (88)</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>87 (100)</td>
<td>43 (100)</td>
<td>130 (100)</td>
<td></td>
</tr>
<tr>
<td>Age of patients (years) relative to syrinx, median (IQR)</td>
<td>37 (28−47)</td>
<td>34 (29−43)</td>
<td>36 (29−44)</td>
<td>0.51</td>
</tr>
<tr>
<td>Length of stay (days), median (IQR)</td>
<td>3 (3−4)</td>
<td>3 (3−4)</td>
<td>3 (3−4)</td>
<td>0.36</td>
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<tr>
<td>Follow-up (months), median (IQR)</td>
<td>42 (17−71)</td>
<td>73 (20−110)</td>
<td>52 (19−85)</td>
<td>0.03</td>
</tr>
<tr>
<td>Time to resolve/improve syrinx (months), median (IQR)</td>
<td>—</td>
<td>4 (3−8)</td>
<td>4 (3−8)</td>
<td></td>
</tr>
<tr>
<td>Headache scores, median (SD)</td>
<td></td>
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</tr>
<tr>
<td>Preoperative</td>
<td>9.7 (0.9)</td>
<td>&lt;0.001†</td>
<td>9.3 (1.8)</td>
<td>&lt;0.001†</td>
</tr>
<tr>
<td>Postoperative</td>
<td>4.1 (2.3)</td>
<td></td>
<td>4.7 (2.4)</td>
<td></td>
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<tr>
<td>Preoperative neurologic deficit, n (%)</td>
<td></td>
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<tr>
<td>Mild</td>
<td>23 (28)</td>
<td>7 (16)</td>
<td>30 (24)</td>
<td>0.19*</td>
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<tr>
<td>Moderate</td>
<td>22 (27)</td>
<td>10 (23)</td>
<td>32 (26)</td>
<td></td>
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<tr>
<td>Severe</td>
<td>36 (45)</td>
<td>26 (61)</td>
<td>62 (50)</td>
<td></td>
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<tr>
<td>Total</td>
<td>81 (100)</td>
<td>43 (100)</td>
<td>124 (100)</td>
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<tr>
<td>Did respondent smoke, n (%)</td>
<td></td>
<td></td>
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<tr>
<td>No</td>
<td>63 (72.4)</td>
<td>34 (79.1)</td>
<td>97 (74.6)</td>
<td>0.52†</td>
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<tr>
<td>Yes</td>
<td>24 (27.6)</td>
<td>9 (20.9)</td>
<td>33 (25.4)</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>87 (100)</td>
<td>43 (100)</td>
<td>130 (100)</td>
<td></td>
</tr>
<tr>
<td>Satisfaction with the outcome of surgery, n (%)</td>
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<tr>
<td>Not satisfied</td>
<td>2 (4)</td>
<td>1 (2)</td>
<td>3 (2)</td>
<td>&gt;0.99*</td>
</tr>
<tr>
<td>Satisfied</td>
<td>16 (18)</td>
<td>8 (19)</td>
<td>24 (19)</td>
<td></td>
</tr>
<tr>
<td>Very satisfied</td>
<td>67 (78)</td>
<td>34 (79)</td>
<td>101 (78)</td>
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<tr>
<td>Total</td>
<td>85 (100)</td>
<td>43 (100)</td>
<td>128 (100)</td>
<td></td>
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</table>

IQR, interquartile range. 
*Fisher exact test. 
†Mann-Whitney U test. 
∥Wilcoxon test. 
§2 test.
patients, which is substantially more patients. There were no statistically significant differences in patient age in our study versus other studies of adult CM-I (Table 3). In the adult CM-I studies, gender is known in 1456 patients (543 males [37%] and 913 females [54%]), whereas our study included female patients (n = 112, 86%) versus male patients (n = 18, 14%); our female:male ratio had a greater margin than published studies.

**Headache as a Preoperative Symptom.** In our 121 patients with headache (93.1%), 118 (98%) experienced improvement, whereas only 3 patients (2%) were unchanged. In other studies of adult patients with CM-I, mean improvement was noted in 79% of patients (Figure 9, top).

**Neurologic Status.** Of the 130 patients in the adult series, mean improvement/resolution was noted in 77%, no change in 20%, and worsening in 3%. In our study, improvement of clinical status in all 130 patients was observed (100%) (Figure 9, middle).

**Syringomyelia.** In other previous studies of adult patients with CM-I, syrinx resolved/improved in 74%, whereas 26% experienced no change. In our study, there was an improvement in all 130 patients (100%) (Figure 9, middle).

**Follow-Up Length.** We observed statistically significantly longer follow-up of patients in our study versus other adult studies (t test, P = 0.002) (Table 2).

**Complications.** There were no operative and perioperative mortalities. Four patients died during follow-up, all of whom were female (case 43, heart attack 4 months after surgery; case 46, complications of gall bladder surgery 6 months after surgery; case 82, heart attack 1 year after surgery; case 90, stroke 2 years after surgery). Three patients developed pseudomeningocele/CSF leak after surgery (1 male, 2 females; cases 4, 5, and 11) and needed reoperation. Since then, we have adopted the fat graft technique as described earlier and have not experienced further cases of pseudomeningocele/CSF leak. Four patients developed superficial wound infection (all female): 3 needed reoperation (debridement, washout, and closure), and 1 was healed only with antibiotics and dressing changes (Table 4).

**DISCUSSION**

CM-I is a well-known clinical entity with a relatively high prevalence reported in 0.24%–3.6% of the population and 0.56%–0.77% based on MRI studies. Symptoms of CM-I can be divided into those related to CSF circulation impairment at the cranio-cervical junction and those related to compression or traction of the brainstem, cerebellum, and cranial nerves (VII–XII). The results of operative treatment of patients over the previous 20 years have been reported to be good, with mean improvement/resolution of syringomyelia in 74% of adult patients with CM-I, improvement/resolution of neurologic deficit in 77%, and improvement of headaches in 79%. However, there is still room for improvement in up to 26% of outcomes across all concerned clinical symptoms.\(^3\,33-37,72\)

This clinical study prospectively hypothesized that a circumferential cranio-cervical decompression of CM-I in adults of 270° achieves re-establishment of CSF circulation between the cranial and spinal subarachnoid compartments and provides decompression of the cerebellum, brainstem, and cranial nerves. This decompression facilitates resolution of preoperative syringomyelia, neurologic deficit, and headaches better than previously reported. The average number of patients in other adult studies is 20.\(^3\,33-35,37,72\) In addition, our study has a longer follow-up (approximately 6 months longer than the mean of other studies; Table 3) and is the only prospective study. Our study period was almost 2 times longer than the average length of other adult studies (15 vs. 8 years).\(^3\,33-35,37,72\)

The goal of our study was to precisely measure, radiologically and clinically, the surgical outcomes regarding resolution/improvement of syringomyelia, neurologic status, and preoperative headaches and to compare those outcomes with our reported systematic review of surgical outcomes of studies of CM-I in adults reported in 1963–2013 and included additional studies from 2014–2019.\(^1\) Also, we investigated the association of BMI in patients with CM-I.
Demographics

In our series, the most common age range was 21–50 years (specifically, 31–40 years) with a mean age of 38 years, which is slightly younger compared with the mean age of 41 years found in the literature (Figure 5). Females were more prevalent than males: 86% versus 14% compared with 57% versus 34% in the literature (with 9% of unknown gender in the literature). The margin of female predominance is larger in our study than in the literature.1-33

Patients without syrinx were more common than patients with syrinx in our series (65% vs. 35%), which was different from in published studies (31% vs. 69%). Syrinx was significantly more common in our male patients than in females; however, this could not be compared because the breakdown of syrinx by gender is not reported in the literature. Our patients with syrinx were also younger compared with other studies, but this difference did not reach statistical significance.1-33

Headache Improvement

Suboccipital headaches that worsen with Valsalva maneuvers (e.g., coughing, sneezing, lifting heavy objects, straining with defecation, or coitus) are the hallmark of CM-I presentation. This is classic tussive, or strain-related, headache of patients with CM-I caused by dural stretching as cranial volume is briefly increased and not simultaneously vented because of CSF flow obstruction caused by the descended tonsils at the foramen magnum.18 This is probably the most common symptom in CM-I, although it is often underreported.1 Headache before surgery has been reported in 93% of our patients, 36 of whom (83.7%) had syrinx. After surgery, there was an improvement in 118 of our patients (98%),

Figure 6. Preoperative (A) and postoperative (B) width of syringomyelia cavity in patients harboring syrinx by grades I–IV, blue, red, green, and purple, respectively. Preoperative neurologic deficit in subjects without (C) and with syringomyelia (D).
with the exception of 3 (2%), who experienced no change. This is clearly an improved outcome compared with the mean 79% headache resolution reported in other reports of adult CM-I. Resolution of headaches and improvement of neurologic deficit closely mirrors patient satisfaction with surgery outcomes, which again has not been previously reported.

**Syringomyelia Improvement**

In other series of adult CM-I, syringomyelia has been reported to resolve or improve on average in 74% of patients. Craniocervical decompression obviously opens cerebrospinal subarachnoid spaces, including egress of CSF from the fourth ventricle, and thus decompression of the syrinx is facilitated. However, 26% of patients still did not achieve syrinx improvement. In our series, all patients (100%) achieved improvement or resolution of syringomyelia. More specifically, of 43 patients with syringomyelia, complete syrinx disappearance was documented in 35 patients (81%), and there was a significant improvement in 8 patients (19%). Four of 5 patients with syrinx had complete resolution and 1 had significant improvement (Figures 6, 7, and 9, bottom). Median time to resolve/improve syrinx was 4 months (IQR, 3–8 months). Therefore, 8 months after surgery, it is unlikely to

![Figure 7. (Top) Comparison of the width of syrinx preoperatively and postoperatively. Note the significant improvement and/or resolution of syrinx preoperatively. (Bottom) The neurologic deficit preoperatively and postoperatively in patients without (left) and with (right) syringomyelia.]
expect any further syrinx improvement. To our knowledge, this observation regarding the timing of syrinx resolution has not been reported previously in CM-I in adults.

Neurologic Improvement
Symptoms of brainstem compression and traction on cranial nerves may be noted together because it may not be possible to distinguish both in every patient. Combined symptoms include nystagmus, hoarseness, speech and swallowing difficulties, and sleep apnea. Balance problems are presumably the result of traction or compression of cerebellar connections. Findings associated with CM-I include nystagmus, impaired gag response, and impaired limb and gait ataxia with resulting impaired balance, and other less common symptoms. Syringobulbia may be encountered in patients with CM-I and may present with lower cranial nerve involvement and, occasionally, with diplopia. Overall, mean neurologic improvement/resolution in other series was noted in 77% of patients, with no change in 20%, and worsening in 3%, which is similar to syringomyelia improvement/resolution values (74%). All our patients (100%) experienced improvement/resolution of symptoms and most presented preoperatively with severe (50%), moderate (26%), and mild (24%) neurologic deficit. Furthermore, we evaluated all patients according to EMS to thoroughly investigate the neurologic impairment and postoperative outcome. Our results have shown a significant improvement after surgery, with mean EMS at follow-up of 16.42. To our knowledge, this is the first study of adult Chiari malformation to use EMS for evaluation of neurologic improvement after surgery. There was no statistically significant difference in severity of preoperative neurologic symptoms between patients with and without syringomyelia.

BMI
This study confirmed a previously reported strong association between increased BMI and adult patients with CM-I with syringomyelia in the literature. Only 1 patient in our current series was underweight (1%) and 27 patients were normal weight (21%), whereas 102 patients (88%) were overweight or obese. In addition, the vertical extent of syrinx was an average of 7 vertebral levels. Again, data that include patient BMI are not available in other CM-I series for comparison. Figure 3 shows this association and confirms previous observations in the literature that increased BMI and CM-I formation are associated. Another important novel observation is the de novo formation of CM-I in a patient after gaining 10 BMI points of weight over 2 years (Figure 4I–K).

Our Operative Technique and Proposed Mechanisms of Improvement
The goals of surgical decompression include relief of brainstem compression and cranial nerve distortion, restoration of the instantaneous venting capability of the cranial CSF compartment, and reduction of the syrinx cavity (when one is present). Enlarging the cisterna magna is based on the premise that CSF pulsations in an enlarged cisterna magna promote ascent of the cerebellar tonsils, as well as aid in the propagation of CSF pulsations into the spinal subarachnoid space. Reducing the size of the cerebellar tonsils contributes to the enlargement of the cisterna magna and initiates ascent of the tonsils. Duraplasty may affect intracranial compliance and seems to be associated with a lower risk of reoperation. As our systematic review suggested, all patients in the adult surgical population with CM-I received bone decompression (100%), almost all received dural opening and dissection (97%), and most received arachnoid opening and dissection (70%).
In an attempt to further improve outcomes of CM-I microsurgical decompression, we adopted a wider, 270° circumferential decompression of the craniocervical junction, which includes C1 laminectomy and suboccipital craniectomy. The craniectomy itself is about 2 cm (approximately 2 cm [1 in]) high to avoid any possibility of cerebellar ptosis. We believe that craniectomy width is more important than height when decompressing craniocervical subarachnoid spaces. Therefore, we include drilling a 1/5 posteromedial aspect of the occipital condyles because craniospinal obstruction of subarachnoid spaces involves a relatively short segment of neuraxis, but frequently also includes blockage of CSF egress from the fourth ventricle via the 3 foramina, obliteration of cisterna magna, and both lateral CMCs. Hence, circumferential, wider decompression is important.

After dural opening and release of any arachnoid adhesions and membranes possibly obstructing the fourth ventricle and/or Magendie foramen, a more aggressive 270° arachnoid dissection (enabled by limited condyle drilling) facilitates a more complete release of arachnoid adhesions obstructing the CSF flow and decompression of the lateral CMCs and Luschka foramina bilaterally. CSF evacuation from the fourth ventricle is therefore widely facilitated. Furthermore, vertical CSF flow is facilitated across the craniospinal junction circumferentially. Coagulating tonsillar tips and dural patch grafting maintains a wide craniocervical decompression simultaneously, which ensures resolution of syringomyelia. This technique modification results in excellent outcomes for headache, neurologic deficit, and syringomyelia resolution/improvement (Figures 1–3).

Importance of Ruling Out Craniocervical Instability

Recently, Goel et al.73 reported their experience with patients with CM-I with complex malformations and craniocervical instability with satisfactory postoperative results with fusion as a predominant treatment option. In our study, we performed extensive preoperative neuroradiologic workup for instability. This study included only patients in whom craniocervical instability was completely ruled out (the radiologic protocol is detailed in the Methods section). Furthermore, we continued to monitor our patients during the lengthy follow-up for several parameters, including instability. None of the patients in the series showed any clinical or radiologic signs of instability during the course of lengthy follow-up, which averaged almost 4.5 years (Table 2).

Limitations of the Study

The limitations of this study include the fact that it is a single-center study by a single neurosurgeon, which may include the
bias of patient selection process, regional patient-specific and demographic-specific bias, and lack of control group.

Although the syrinx size and postoperative improvement, the neurologic examination, and headache severity and improvement were performed by exact means of measurement by an independent expert who was not part of this study, the potential errors cannot be completely excluded.

Although the comparisons between the results of our study and those of the most extensive systematic review of CM-I studies in the literature were between similar clinical parameters and numeric values, the systematic reviews of CM-I literature studies may have results that are negatively biased because they included average (mean) values.

CONCLUSIONS

Based on our results compared with the results of other studies, it indicates that the 270° circumferential microsurgical foramen magnum decompression surgical technique for adult CM-I appears to have a beneficial effect and modest improvement in outcomes, as well as resolutions of the syrinx, neurologic symptoms, and diminishment of headaches. Further substantial research is needed to confirm our results.

CRediT AUTHORSHIP CONTRIBUTION STATEMENT

Kenan I. Arnautovic: Conceptualization, Methodology, Formal analysis, Investigation, Data curation, Writing - original draft. Writing - review & editing, Visualization, Supervision, Project administration. Bawan F. Qaladize: Validation, Investigation, Writing - review & editing. Mirza Pojskic: Validation, Formal analysis, Investigation, Data curation. Writing - original draft, Writing - review & editing, Visualization, Project administration. Bruno Splavski: Validation, Formal analysis, Writing - review & editing. Frederick A. Boop: Validation, Formal analysis, Writing - review & editing.


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