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Intradural pathology and pathophysiology associated with Chiari I malformation in children and adults with and without syringomyelia

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OBJECTIVE The pathophysiology underlying tonsillar herniation and CSF obstruction in Chiari malformation Type I (CM-I) is unclear, and the cause of CM-I–associated syringomyelia is not well understood. A better understanding of this pathophysiology is important for an improved treatment strategy. Therefore, the authors sought to identify, characterize, and examine the intradural pathology and CSF flow pathophysiology in the posterior fossa and at the level of the foramen magnum that occurs in the setting of CM-I. They determined the incidence of these intradural findings and assessed differences across age, with the degree of tonsillar herniation, and in the presence and absence of syringomyelia.

METHODS A prospective database initiated in March 2003 recorded all intraoperative findings during surgical treatment of children and adults with CM-I with or without syringomyelia. A total of 389 surgeries for CM-I were performed in 379 patients between March 2003 and June 2016. A total of 109 surgeries were performed in 109 patients with CM-I (without osseoligamentous abnormalities) in whom both a posterior fossa extradural and intradural decompression with duraplasty was performed (first-time intradural procedures). Using a surgical microscope, intradural pathology and obstruction of CSF channels were identified and assessed. Student t-tests and Fisher's exact tests compared groups in a series of univariate analyses, followed by multivariate logistic regression.

RESULTS The following intradural pathological entities were observed (prevalence noted in parentheses). These include those that did not obstruct CSF flow channels: opacified arachnoid (33.0%), thickened arachnoid (3.7%), ischemic and gliotic tonsils (40.4%), tonsillar cysts (0.9%), and inferior descent of the fourth ventricle and cervicomedullary junction (CMJ) (78.0%). The following intradural pathological entities were observed to obstruct CSF flow channels: medialized tonsils (100%), tonsil overlying and obstructing the foramen of Magendie (21.1%), intertonsillar and tonsil to CMJ arachnoid adhesions (85.3%), vermian posterior inferior cerebellar artery branches obstructing the foramen of Magendie (43.1%), and arachnoid veils or webs obstructing or occluding the foramen of Magendie (52.3%). Arachnoid veils varied in type and were observed in 59.5% of patients with CM-I who had syringomyelia, which was significantly greater than the 33.3% of patients with CM-I without syringomyelia who had an arachnoid veil ($p = 0.018$). The presence of CM-I with an arachnoid veil had 3.22 times the odds ($p = 0.013$, 95% CI 1.29–8.07, by multivariate logistic regression) of being associated with syringomyelia, adjusting for tonsillar herniation. The inferior descent of the fourth ventricle and CMJ occurred with a greater degree of tonsillar herniation ($p < 0.001$) and correlated with a cervicomedullary kink or buckle on preoperative MRI.

CONCLUSIONS Intradural pathology associated with CM-I with or without syringomyelia exists in many forms, is more prevalent than previously recognized in patients of all ages, and may play a role in the pathophysiology of CM-I tonsillar herniation. Arachnoid veils appear to partially obstruct CSF flow, are significantly more prevalent in cases of CM-I with syringomyelia, and therefore may play a role in the pathophysiology of CM-I–associated syringomyelia.

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KEY WORDS Chiari malformation Type I; tonsillar herniation; arachnoid veil; arachnoid web; arachnoid adhesions; opaque arachnoid; ischemic tonsils; gliotic tonsils; syringomyelia; cerebrospinal fluid; craniovertebral junction

ABBREVIATIONS CM-I = Chiari malformation Type I; CMJ = cervicomedullary junction; PICA = posterior inferior cerebellar artery.

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Chiari malformation Type I (CM-I)[2,6–8] is a condition characterized by inferior displacement of the cerebellar tonsils through the foramen magnum, is associated with syringomyelia, and has been well described in the literature, with various presentations and a constellation of signs and symptoms.[23,24,38] There are extensive case series of both pediatric[38] and adult patients in which presentation, nonoperative or operative management, surgical procedure, and outcome analysis are described, with widely varying results.[24]

A limitation to better outcomes and optimum surgical management of CM-I is the fact that the pathophysiology of tonsillar herniation and CSF obstruction in CM-I is unclear and the cause of CM-I–associated syringomyelia is not well understood.[36] Evidence suggests that a combination of posterior fossa extradural[1,3–5,9,11,13,21,24,25,27,28,32,33,36,37,40] and intradural pathology[41] play a role in the tonsillar herniation of CM-I, CSF obstruction at the foramen magnum, and CM-I–associated syringomyelia.[29]

Multiple studies have described the extradural pathology in CM-I, which occurs in development and results in a smaller posterior fossa—a finding that is believed to result in the tonsillar herniation and CSF obstruction seen in CM-I.[1,3–5,9,11,13,21,24,25,27,28,32,33,36,37,40] However, other studies have found a normal-sized posterior fossa in patients with CM-I,[32,36,40] suggesting that other factors play a role in the pathophysiology of tonsillar herniation. Few studies have described or analyzed intradural pathology in CM-I. Therefore, we sought to characterize the intradural pathology and associated CSF flow pathophysiology in the posterior fossa and at the level of the foramen magnum that occurs in the setting of CM-I. We reviewed all intradural pathology and CSF flow pathophysiology encountered in first-time posterior intradural decompression and duraplasty for the surgical treatment of CM-I from a prospective database spanning 2003 to 2016. We determined the incidence of these intradural findings and assessed differences across age, with degree of tonsillar herniation, and in the presence and absence of syringomyelia.

Methods

Patient Population

The senior author (A.H.M.) initiated a prospective database of all craniovertebral junction abnormalities,[10] including CM-I,[23] in 1978. In March 2003, we modified the database to incorporate and record all intraoperative findings during the surgical treatment of children and adults with CM-I with and without syringomyelia, with the focus on better understanding the pathophysiology of this disorder. A total of 389 surgeries for CM-I were performed in 379 patients between March 2003 and June 2016 (Fig. 1). A total of 109 primary posterior procedures were performed in 109 patients in whom both a posterior fossa extradural and intradural decompression with duraplasty (extra-intradural) was performed (first-time intradural procedures). All were without craniovertebral junction abnormalities. All surgeries were performed by the senior author between March 2003 and June 2016, and by the first author (B.J.D.) and senior author between July 2015 and June 2016. The University of Iowa's institutional review board approved this study.

Patient Selection: CM-I Treatment Algorithm

The first and senior authors used a tailored treatment strategy for each patient with CM-I with and without syringomyelia. All patients underwent preoperative evaluation and, if deemed symptomatic, a posterior decompression was offered. Symptomatic patients included those who presented with Valsalva maneuver–induced headaches, bulbar signs and symptoms, cerebellar signs and symptoms, cranial nerve deficits, or spinal cord signs and symptoms. Intraoperative ultrasound was used to determine sufficient subarachnoid space and CSF flow around the tonsils. We used ultrasound throughout the different stages of the extradural operation to assess for effective decompression. The presence of CSF behind the tonsils or tonsillar ascent were criteria for effective extradural decompression. In patients with CM-I without syringomyelia, we used intraoperative ultrasonography for determination of adequate decompression and the need for intradural exploration, decompression, and duraplasty. In most patients with CM-I and syringomyelia, we performed a posterior extra-intradural decompression due to data suggesting that patients with syringomyelia have a higher prevalence of intradural pathology.

Overall Surgical Approach: Posterior Extra-Intradural Decompression With Cervical Fascia Duraplasty

All posterior extra-intradural decompressions for CM-I with or without syringomyelia were performed similarly. A posterior occipital cervical incision was made and a subperiosteal dissection was performed to expose the occiput, foramen magnum, and C-1 lamina. A high-speed electric drill and rongeurs were used to remove approximately 25–30 mm of the posterior foramen magnum. The occiput was removed superiorly by approximately 30 mm to the inferior nuchal line. The posterior 25–30 mm of the superior two-thirds of the C-1 lamina was removed, and the occipital-cervical epidural compressive band was removed. Ultrasound was performed to assess the adequacy of the extradural decompression. In these 109 cases, the microscope was then used for opening of the dura mater, intradural aspects of the surgery, and cervical fascia duraplasty.

Use of Microscope for Intradural Aspects of Procedure and for Recording Intradural Findings

Using a surgical microscope under high-power magnification, the posterior fossa dura was opened starting at the superolateral quadrant on the right, and proceeding down to the midline at the foramen magnum, and then along the dorsal axis of the cervical canal toward the lamina of C-1 in a curvilinear or hockey stick–like fashion. Care was taken to leave the arachnoid intact during dural incision. Stay sutures held apart the exposure. Hemostasis in the venous sinuses was secured with titanium clips.

On completion of the dural opening, the arachnoid was inspected and divided in the midline, held apart temporarily, and then attached to the dural edges with titanium

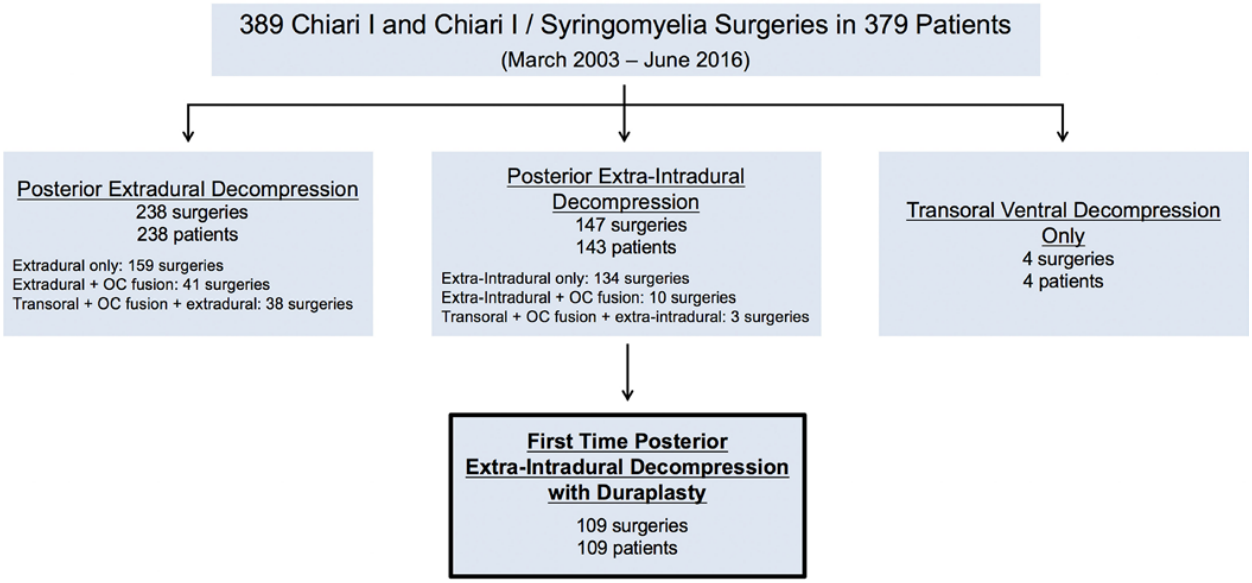

FIG. 1. Flow diagram illustrating the total number of CM-I surgeries and patients with CM-I, and the breakdown per type of surgery from March 2003 to June 2016 in our prospective database. A total of 109 primary posterior procedures were performed in 109 patients in whom both a posterior fossa extradural and intradural decompression with duraplasty (extra-intradural) were performed (first-time intradural procedures). All patients were without craniovertebral junction abnormalities. OC = occipital. Figure is available in color online only.

clips. Arachnoid adhesions were lysed and the foramen of Magendie (opening of the fourth ventricle) was explored, and any adhesions and arachnoid veils were cauterized and lysed.

Intradural pathology and CSF flow pathophysiology were noted and recorded. Prior to the initiation of this study, the senior author had identified and reported on arachnoid adhesions, posterior inferior cerebellar artery (PICA) branches obstructing the foramen of Magendie, and tonsils occupying the outlet to the fourth ventricle in patients with CM-I.[23] Further study of intraoperative findings in CM-I led to recording the following intradural pathology in this prospective study: arachnoid opacification, thickened arachnoid, arachnoid beading, ischemic and gliotic tonsils, inferior descent of the fourth ventricle and cervicomedullary junction (CMJ), tonsillar cysts, medialized cerebellar tonsils, cerebellar tonsil overlying the foramen of Magendie, intertonsillar and tonsil to CMJ arachnoid adhesions, vermian PICA branches obstructing the foramen of Magendie, and arachnoid veils obstructing the foramen of Magendie. Pathological entities that appeared to obstruct CSF channels and CSF flow through the foramen of Magendie and the foramen magnum were noted and recorded.

Cerebellar tonsillar reduction was performed using low-power bipolar piarachnoid electric coagulation of the cerebellar tonsils such that the tonsils ascended upward and outward. This opened the roof of the fourth ventricle and exposed the egress of the fourth ventricle. The integrity of the pia mater was maintained to avoid potential adhesion, scarring, and recurrence. Modified cisterna magna reconstruction was performed with placement of a patulous cervical fascia dural graft.

Statistical Analysis

Descriptive statistics were calculated, and Student t-tests (continuous variables) and Fisher's exact tests (dichotomous variables) were used to compare various groups in a series of univariate analyses. The following groups were compared: pediatric CM-I group (< 18 years old) with the adult CM-I group (≥ 18 years old); and CM-I no syringomyelia group with the CM-I syringomyelia group. Tonsillar herniation was compared in the presence and absence of each intradural pathological entity. To assess for intradural pathology associated with syringomyelia, the following were initially examined in a series of univariate analyses, followed by a multivariable regression analysis based on a stepwise approach, including univariable analyses with $p < 0.20$ and $p < 0.05$ as the cutoff for inclusion in the final adjusted model: patient age, sex, tonsillar herniation, patient age < 18 or ≥ 18 years old, and the presence or absence of syringomyelia, cervicomedullary kink or buckle, arachnoid opacification, thickened arachnoid, arachnoid beading, ischemic and gliotic tonsils, tonsillar cysts, fourth ventricular and CMJ descent, medialized tonsils, tonsil overlying the foramen of Magendie, intertonsillar and tonsil to CMJ arachnoid adhesions, vermian branch of PICA obstructing the foramen of Magendie, and arachnoid veil or web obstructing the foramen of Magendie. Statistical analyses were conducted with the help of the SAS 9.2 (SAS Institute, Inc.) and LogXact 8 (Cytel, Inc.) software.

Results

Demographics and Preoperative Radiographic Characteristics

A total of 109 patients underwent primary posterior

fossa extra-intradural procedures (Fig. 1). The mean age of the patients was 17.7 years (range 2–61 years) (Table 1). Sixty-eight patients (62.4%) were pediatric (< 18 years old) and 41 patients (37.6%) were adults (≥ 18 years old). A total of 70 female and 39 male patients underwent surgery. The mean tonsillar herniation was 15.0 mm (range 4.5–26.4 mm). Syringomyelia occurred in 79 patients (72.5%). Cervicomedullary compression with cervicomedullary buckle was observed in 85 patients (78.0%). No hydrocephalus was observed in any of these 109 patients preoperatively.

Arachnoid Opacification, Thickened Arachnoid, and Arachnoid Beading

On opening the dura, the arachnoid was kept intact. The arachnoid was opaque in 33.0% (36/109) of patients (Table 1). The degree of opacification varied from patient to patient (Figs. 2–5). In some cases the arachnoid was completely opaque (Fig. 4C), whereas in others it was somewhat semitranslucent (Figs. 2C and 3C) and therefore less opaque. In some cases the arachnoid had peculiar strands within it, for which the etiology was unclear. The arachnoid was vascularized in some cases. In 3.7% (4/109) of patients, the arachnoid was thickened. In 0.9% (1/109) of patients, the arachnoid was as thick as the dura. Very tiny beading of the arachnoid was found on the surface of the cerebellar tonsils in 2.8% (3/109) of patients. Arachnoid opacification was significantly different across age, degree of tonsillar herniation, and presence or absence of syrinx (Tables 2–4).

Ischemic and Gliotic Tonsils

Ischemic and gliotic tonsils were identified as having a pale or whitish color compared to the cerebellum, most often noted at the tonsillar tips (Fig. 2). The tonsils were inspected in all patients and 40.4% (44/109) of patients were found to have ischemic and gliotic tonsils (Table 1). In 12.8% (14/109) of patients, only the right tonsil was ischemic and gliotic; in 9.2% (10/109) of patients only the left tonsil was ischemic and gliotic; and in 18.3% (20/109) of patients, both tonsils were found to be ischemic and gliotic. Patients with ischemic and gliotic tonsils had a significantly greater degree of tonsillar herniation than patients without ischemic and gliotic tonsils (18.2 mm vs 12.8 mm, $p < 0.001$, Table 3). There was no significant difference between patients with and without syringomyelia (Table 4).

Tonsillar Cysts

A tonsillar cyst was observed in 0.9% (1/109) of patients (Table 1). In the 1 patient, the tonsillar cyst was evident on the preoperative MR image. Intraoperatively the tonsillar cyst was noted to exert mass effect on the CMJ, and because of this mass effect the brainstem was rotated away from the cyst (Fig. 5).

Inferior Descent of the Fourth Ventricle and CMJ

The fourth ventricle and/or the CMJ were observed to be displaced inferiorly below the foramen magnum in 78.0% (85/109) of patients (Table 1). The inferior displacement of the CMJ correlated with the cervicomedullary buckle seen on the preoperative MRI in these 85 patients (Fig. 5). There was no difference across age or based on the presence or absence of a syrinx (Tables 2 and 4). Patients with inferior descent of the fourth ventricle and CMJ had a significantly greater degree of tonsillar herniation than patients without descent of the fourth ventricle and CMJ (15.9 mm vs 11.7 mm, $p < 0.001$, Table 3).

Medialized Tonsils

The cerebellar tonsils were medially approximated and crowded the foramen of Magendie, limiting CSF egress

TABLE 1. Intradural pathology associated with CM-I with and without syringomyelia in 109 patients of all ages

Characteristic	Value
Demographics & preop variables	
Total patients	109 (100)
Age in yrs	17.7 (2–61)
Male sex	39 (35.8)
Tonsillar herniation in mm	15.0 (4.5–26.4)
Age <18 yrs; pediatric group	68 (62.4)
Syringomyelia	79 (72.5)
Cervicomedullary buckle	85 (78.0)
Nonobstructive intradural pathology	
Arachnoid opacification	36 (33.0)
Thickened arachnoid	4 (3.7)
Arachnoid beading	3 (2.8)
Ischemic & gliotic tonsils	44 (40.4)
Rt	14 (12.8)
Lt	10 (9.2)
Both	20 (18.3)
Tonsillar cysts	1 (0.9)
4th ventricular & CMJ descent	85 (78.0)
Obstructive intradural pathology	
Medialized tonsils	109 (100.0)
Tonsil overlying foramen of Magendie	23 (21.1)
Rt	16 (14.7)
Lt	7 (6.4)
Intertonsillar & tonsil to CMJ arachnoid adhesions	93 (85.3)
Vermian branch of PICA obstructing foramen of Magendie	47 (43.1)
Rt	14 (12.8)
Lt	10 (9.2)
Both	23 (21.1)
Arachnoid veil obstructing foramen of Magendie	57 (52.3)
Complete	11 (10.1)
Perforated	31 (28.4)
Inferior two-thirds	7 (6.4)
Inferior one-third	8 (7.3)

Values are expressed as the number of patients (%) or as the mean (range), unless otherwise indicated.

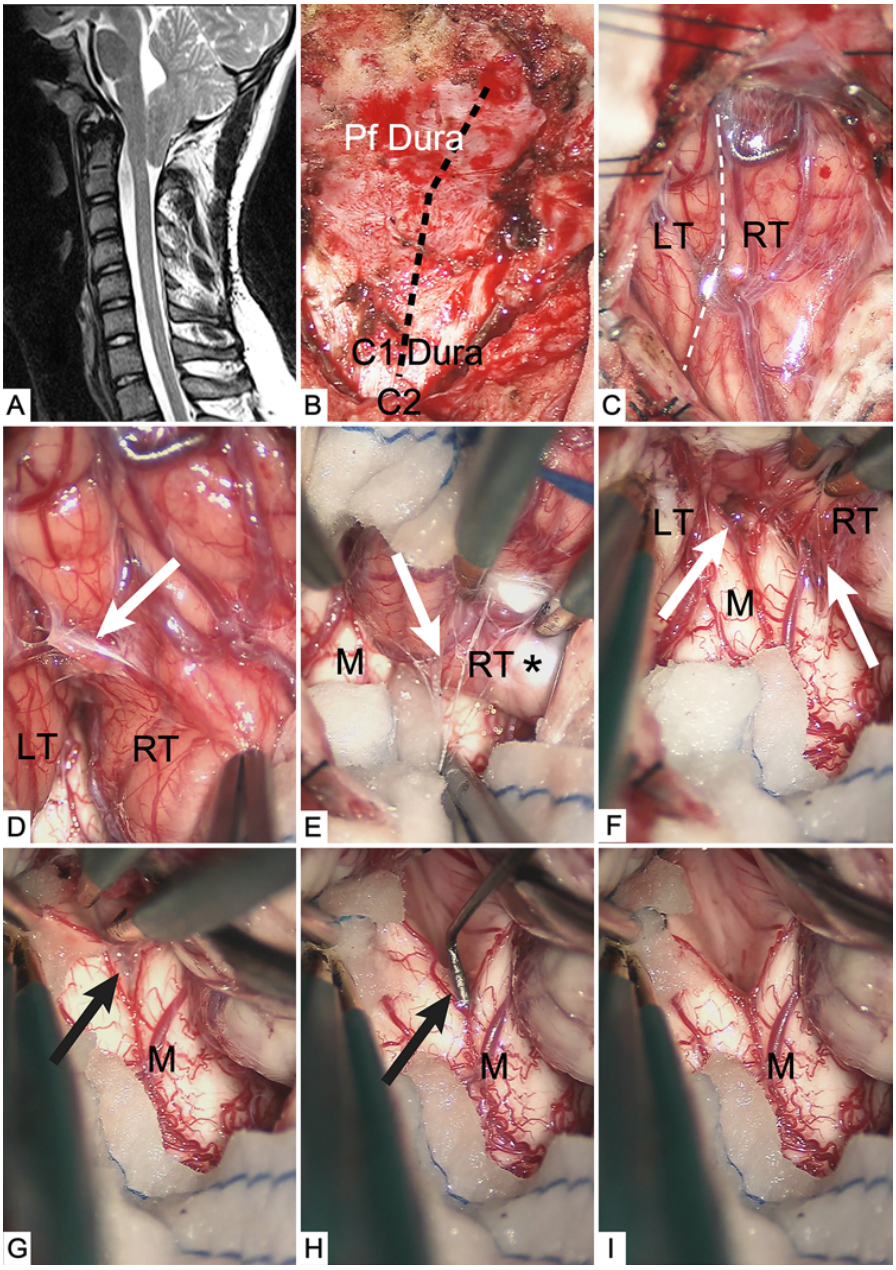

FIG. 2. An 18-year-old man with CM-I without syringomyelia. Intraoperative illustration of tonsillar herniation, right tonsil overlying the cisterna magna and occluding the foramen of Magendie, ischemic and gliotic tonsils, thick arachnoid adhesions, fine arachnoid adhesions, adhesions surrounding the foramen of Magendie, and inferior one-third arachnoid veil overlying the foramen of Magendie. **A:** Preoperative sagittal T2-weighted MRI study demonstrating CM-I with tonsillar herniation and brainstem compression. **B:** Suboccipital craniectomy was performed superiorly to the inferior nuchal line and approximately 30 mm in diameter, with foramen magnum decompression and C-1 laminectomy. The *dotted line* illustrates the proposed dural opening. **C:** On opening the dura, the arachnoid was semitranslucent, with opaque strands present. The right tonsil descended to below the lamina of C-1 and completely filled the cisterna magna and covered the foramen of Magendie. *Dotted line* delineates the right tonsil from the left tonsil. **D:** Separation of the tonsils revealed thick intertonsillar arachnoid adhesions (*arrow*). **E:** After pulling up the right tonsil, the tonsillar tip was noted to be ischemic and gliotic (*asterisk*). Fine arachnoid adhesions (*arrow*) were attached to the tonsil and medulla. **F:** Fine arachnoid adhesions (*arrows*) surrounding the foramen of Magendie prevented complete opening of the foramen. **G:** Bipolar spreading to open up the foramen of Magendie. The inferior one-third of the foramen of Magendie was covered by an arachnoid veil (*arrow*). **H:** Opening of the arachnoid veil (*arrow*). **I:** Completely opened foramen of Magendie. LT = left tonsil; M = medulla; Pf = posterior fossa; RT = right tonsil. Figure is available in color online only.

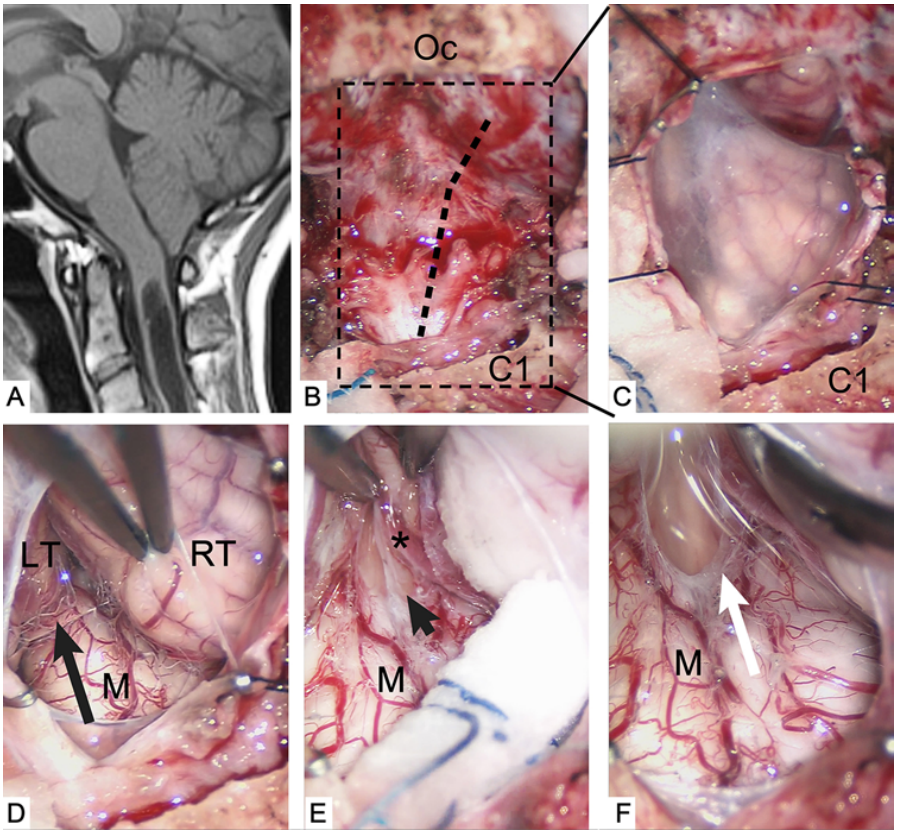

FIG. 3. A 32-year-old woman with CM-I and syringomyelia. Intraoperative illustration of opaque arachnoid, medialized tonsils, fine arachnoid adhesions, PICA vermian branch obstructing the foramen of Magendie, and inferior two-thirds arachnoid veil overlying the foramen of Magendie. **A:** Preoperative sagittal T1-weighted MRI study demonstrating CM-I with tonsillar herniation, cervicomedullary buckle below foramen magnum, and syringomyelia. **B:** Suboccipital craniectomy was performed superiorly to the inferior nuchal line and approximately 30 mm in diameter, with foramen magnum decompression and superior two-thirds C-1 laminectomy. The *dotted line* illustrates proposed dural opening. **C:** On opening the dura, the arachnoid was visibly opaque but still semitranslucent. **D:** Separation of the medialized tonsils revealed intertonsillar and tonsil to medulla fine arachnoid adhesions (*arrow*). **E:** After exposing the foramen of Magendie, the right PICA vermian branch (*asterisk*) was found obstructing the foramen of Magendie. Additionally, an inferior two-thirds arachnoid veil (*arrowhead*) over the foramen of Magendie was found partially occluding CSF outflow. **F:** The arachnoid veil was lysed and excised, and the edges were coagulated to prevent recurrence. This allowed complete opening of the foramen of Magendie (*arrow*). Oc = occiput. Figure is available in color online only.

out of the fourth ventricle in some fashion in all 109 patients (Table 1). The opening to and visualization of the fourth ventricle was only observed by displacing the tonsils laterally and superiorly with retractors (Figs. 3–6). In some cases, the cerebellar tonsils descended medially but also inferiorly in the left lateral gutter, compressing the lower cranial nerves—the 9th, 10th, and 11th.

Tonsil Overlying the Foramen of Magendie

As discussed above, the cerebellar tonsils were medially approximated in all cases. However, in 21.1% (23/109) of patients, the tonsils were asymmetrically medialized such that one of them filled the cisterna magna and overlaid and obstructed the fourth ventricle and foramen of Magendie, thus limiting CSF egress out of the fourth ventricle (Table 1, Fig. 2). There was no difference across age, degree of tonsillar herniation, or presence or absence of syrinx (Tables 2–4).

Intertonsillar and Tonsil to CMJ Arachnoid Adhesions

Arachnoid adhesions occurred in 85.3% (93/109) of patients (Table 1). Adhesions were present in varying degrees in each patient (Figs. 2–5). In some cases the adhesions were fine, thin, and limited in number (Figs. 2E, 2F, 3D, and 6D), and in others the adhesions were thick and quite prevalent (Fig. 2D). Some of the thick adhesions were more consistent with scarlike tissue. The location of adhesions also varied between patients. Adhesions were present between the tonsils as well as between the tonsils and posterior CMJ and over the foramen of Magendie. Because of these adhesions, the tonsils adhered to the posterior aspect of the CMJ and to each other. In fact, the arachnoid adhesions were so severe in some cases that CSF egress out of the foramen of Magendie was completely occluded by adhesions between the tonsils, medulla, and the roof of the fourth ventricle. There was no difference across age, degree of tonsillar herniation, or presence or absence of syrinx (Tables 2–4).

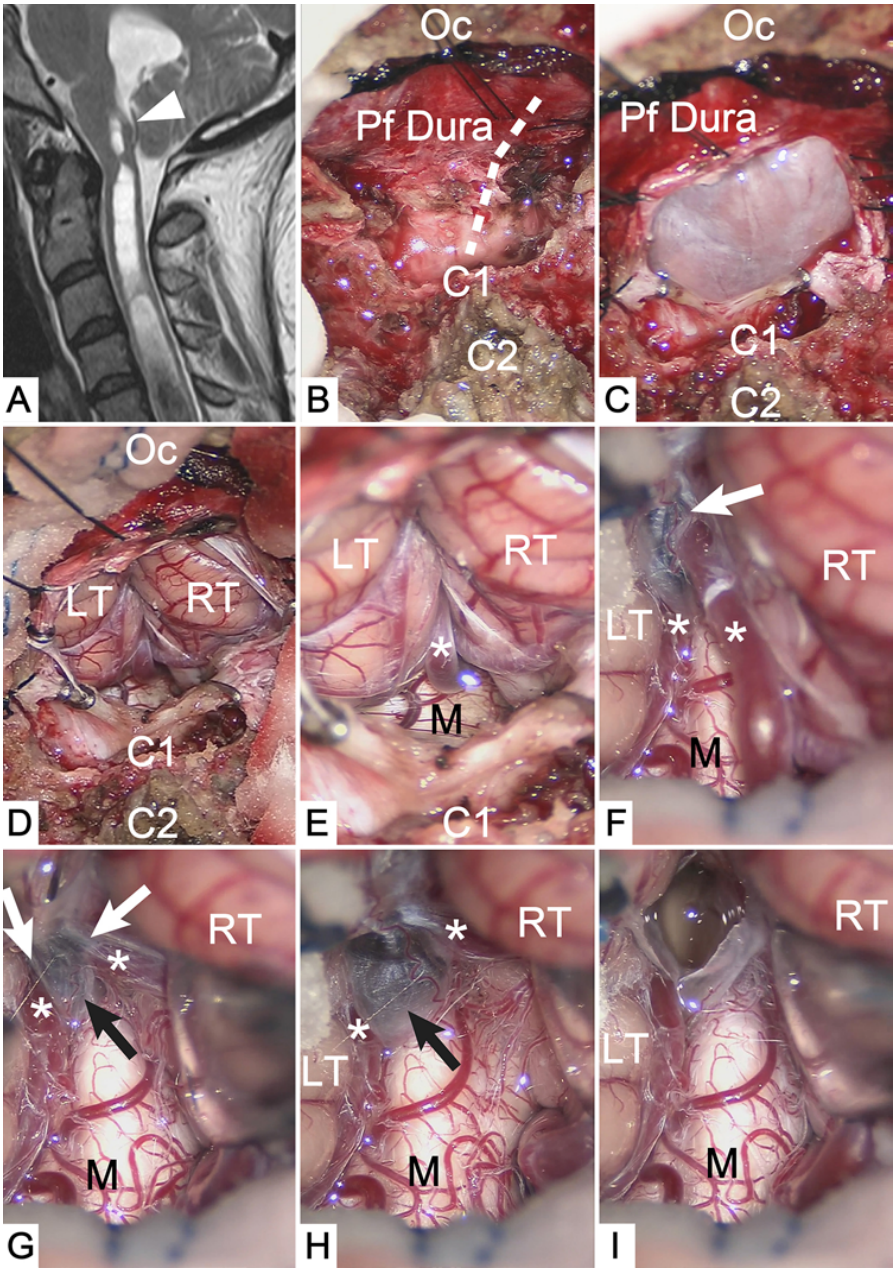

FIG. 4. A 40-year-old woman with CM-I, syringobulbia, and syringomyelia. Intraoperative illustration of opaque arachnoid, medialized tonsils, bilateral PICA vermian branches obstructing the foramen of Magendie, and perforated (almost complete) arachnoid veil overlying the foramen of Magendie. **A:** Preoperative sagittal T2-weighted MRI study demonstrating CM-I with tonsillar herniation, syringomyelia, and syringobulbia. A PICA vermian branch (*arrowhead*) was observed on MRI to be obstructing the foramen of Magendie. **B:** Suboccipital craniectomy was performed superiorly to the inferior nuchal line and approximately 30 mm in diameter, with foramen magnum decompression and superior two-thirds C-1 laminectomy. The *dotted line* illustrates the proposed dural opening. **C:** On opening the dura, the arachnoid was completely opaque and was thickened when incised in the midline. **D:** After incising the arachnoid in the midline, the medialized tonsils were observed. **E–H:** Bilateral PICA vermian branches (*asterisks*) were visualized entering (*white arrows*) and obstructing the foramen of Magendie, which was occluded by a partial arachnoid veil (*black arrows*). The arachnoid veil was partially vascularized by small branches from the PICA vermian branches. **I:** Opening of the arachnoid veil completely opened the foramen of Magendie. Figure is available in color online only.

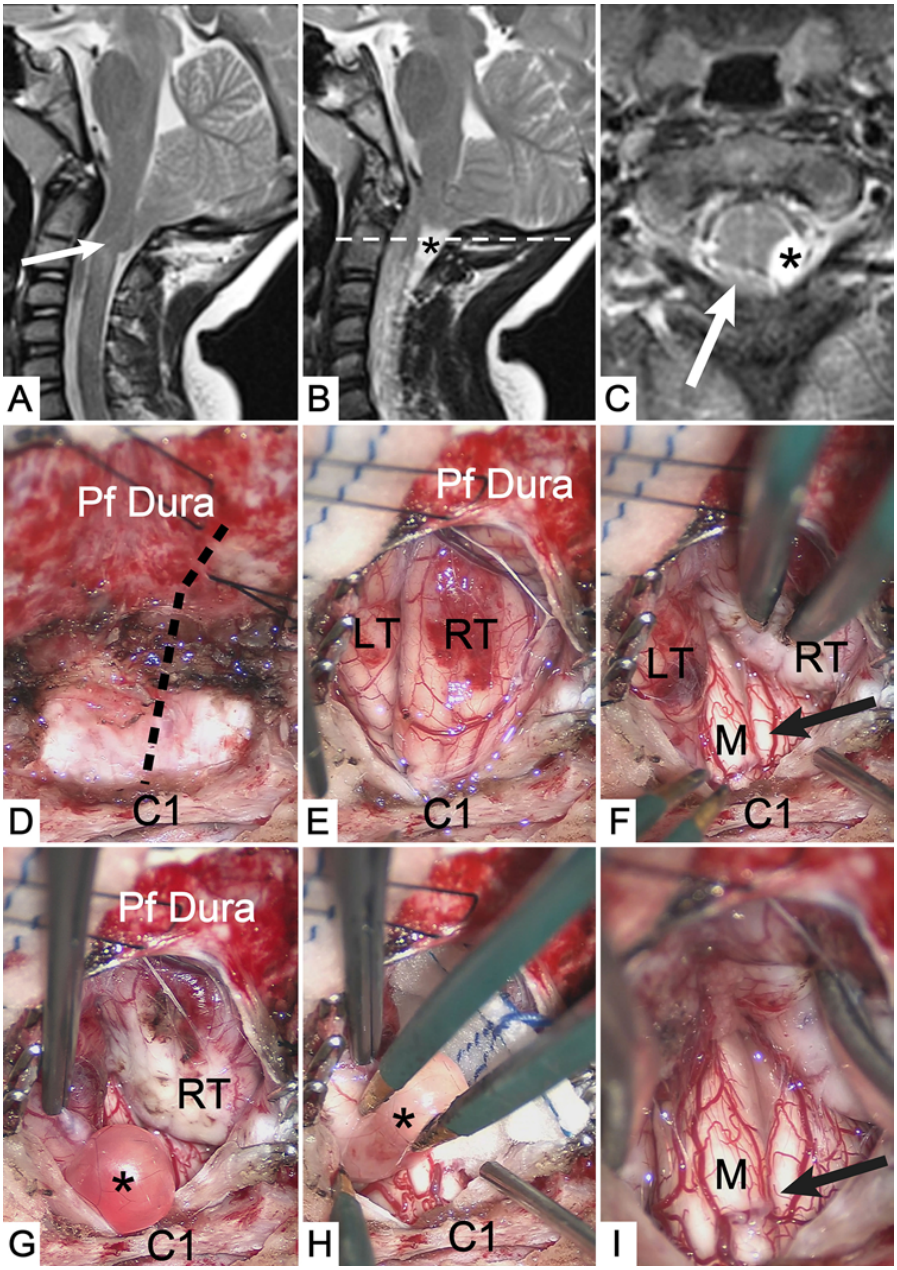

FIG. 5. A 6-year-old girl with CM-I with left tonsillar cyst and without syringomyelia. Intraoperative illustration of right tonsil obstructing the foramen of Magendie, left tonsillar cyst, inferior descent of fourth ventricle, and inferior descent of the CMJ, which corresponds with the cervicomedullary buckle seen on MRI. **A:** Preoperative midline sagittal T2-weighted MRI study demonstrating CM-I with tonsillar herniation and cervicomedullary buckle (*arrow*). **B:** Preoperative parasagittal T2-weighted MRI study demonstrating tonsillar herniation with tonsillar cyst (*asterisk*). **C:** Preoperative axial T2-weighted MRI view through the slice denoted by the *dotted line* in panel B, demonstrating tonsillar herniation with tonsillar cyst (*asterisk*) causing mass effect on the medulla. Notice that the right tonsil (*arrow*) has descended across the midline and is overlying the foramen of Magendie and exerting mass effect on the medulla. **D:** Suboccipital craniectomy was performed superiorly to the inferior nuchal line and approximately 30 mm in diameter, with foramen magnum decompression and superior two-thirds C-1 laminectomy. The *dotted line* illustrates the proposed dural opening. **E:** On opening the dura, the arachnoid was translucent and the right tonsil was noted to be filling most of the cisterna magna and obstructing the foramen of Magendie. **F:** With retraction and low-power bipolar reduction of the right tonsil, the foramen of Magendie was exposed and found to be free of adhesions and veils and was noted to be inferiorly descended to the level of the foramen magnum. Just inferior to the foramen of Magendie, a dorsal cervicomedullary enlargement (*arrow*) was observed corresponding with the cervicomedullary buckle seen in panel A. **G–I:** After pulling up the left tonsil, the tonsillar cyst (*asterisks*) was observed. The cyst had mass effect on the medulla and rotated the medulla to the right. Figure is available in color online only.

TABLE 2. Intradural pathology associated with CM-I in 109 pediatric and adult patients

Characteristic	Value		p Value
	Age <18, n = 68	Age ≥18, n = 41	
Demographic & preop variables			
Age in yrs	10.3 (2–17)	29.9 (18–61)	**<0.001**
Male sex	27 (39.7)	12 (29.3)	0.308
Tonsillar herniation in mm	15.5 (4.5–24.7)	14.2 (5–26.4)	0.197
Syringomyelia	52 (76.5)	27 (65.9)	0.271
Cervicomedullary buckle	55 (80.9)	30 (73.2)	0.179
Nonobstructive intradural pathology			
Arachnoid opacification	13 (19.1)	23 (56.1)	**<0.001**
Thickened arachnoid	2 (2.9)	2 (4.9)	0.246
Arachnoid beading	0 (0)	3 (7.3)	0.012
Ischemic & gliotic tonsils	30 (44.1)	14 (34.1)	**0.029**
Rt	9 (13.2)	5 (12.2)	NA
Lt	9 (13.2)	1 (2.4)	NA
Both	12 (17.6)	8 (19.5)	NA
4th ventricular & CMJ descent	55 (80.9)	30 (73.2)	0.179
Tonsillar cysts	1 (1.5)	0 (0)	NA
Obstructive intradural pathology			
Medial tonsils	68 (100)	41 (100)	NA
Tonsils overlying foramen of Magendie	15 (22.1)	8 (19.5)	0.380
Rt	9 (13.2)	7 (17.1)	NA
Lt	6 (8.8)	1 (2.4)	NA
Intertonsillar & tonsil to CMJ arachnoid adhesions	59 (86.8)	34 (82.9)	0.343
Vermian branch of PICA obstructing foramen of Magendie	27 (39.7)	20 (48.8)	**0.028**
Rt	10 (14.7)	4 (9.8)	NA
Lt	4 (5.9)	6 (14.6)	NA
Both	13 (19.1)	10 (24.4)	NA
Arachnoid veil obstructing foramen of Magendie	36 (52.9)	21 (51.2)	0.759
Complete	11 (16.2)	0 (0)	NA
Perforated	15 (22.1)	16 (39.0)	NA
Inferior two-thirds	4 (5.9)	3 (7.3)	NA
Inferior one-third	6 (8.8)	2 (4.9)	NA

NA = not applicable.
Values are expressed as the number of patients (%) or as the mean (range), unless otherwise indicated. Student t-tests for continuous variables and Fisher's exact tests for dichotomous variables were used to compare patients with CM-I in children (< 18 years old) and adults (≥ 18 years old). Boldface type indicates statistical significance.

The PICA Vermian Branches Obstructing the Foramen of Magendie

One or both PICA vermian branches were sandwiched between the descended tonsils and the posterior CMJ and were found to reside within the fourth ventricle and to obstruct the foramen of Magendie in 43.1% (47/109) of patients (Table 1). Once the medialized tonsils or the tonsil overlying the fourth ventricle were displaced laterally to allow inspection of the foramen of Magendie, the vermian branches of the PICA were found in these 47 patients. These branches were medially approximated and intertwined with arachnoid adhesions and were found to obstruct the foramen of Magendie (Figs. 3 and 4). Often, the PICA vermian branches were attached to the medial aspect of the cerebellar tonsils, and with tonsillar reduction outward and upward, the vessel was freed from obstruction. There was a significant difference across age (Table 2) and degree of tonsillar herniation (Table 3), but not with the presence or absence of syrinx (Table 4).

Arachnoid Veil or Web Obstructing Foramen of Magendie

Arachnoid veils or webs are thin arachnoid membranes that either completely occluded or partially occluded the foramen of Magendie and were found in 52.3% (57/109) of patients (Table 1, Video 1).

VIDEO 1. Clip is an operative illustration of the opening of an arachnoid veil completely occluding the foramen of Magendie and CSF flow in a 6-year-old girl with CM-I and syringomyelia (see Fig. 6). Prior to opening the veil, no pulsatile CSF is observed to flow through the foramen of Magendie. With lysis of the veil, CSF is observed to flow well into the cisterna magna from the foramen of Magendie—and free pulsatile CSF flow is then observed through

TABLE 3. Tonsillar herniation in CM-I stratified by the presence or absence of each intradural pathology

Intradural Pathology	Present or Absent	Mean Tonsillar Herniation in mm (range)	p Value
Arachnoid opacification	+	13.6 (4.5–26.4)	**0.049**
	−	15.7 (5.0–24.0)	
Thickened arachnoid	+	13.5 (7.1–24.0)	0.562
	−	15.1 (4.5–26.4)	
Arachnoid beading	+	16.0 (5.0–23.0)	0.747
	−	15.0 (4.5–26.4)	
Ischemic & gliotic tonsils	+	18.2 (8.0–26.4)	**<0.001**
	−	12.8 (4.5–22.5)	
4th ventricular & CMJ descent	+	15.9 (4.5–26.4)	**<0.001**
	−	11.7 (5.0–21.0)	
Tonsils overlying foramen of Magendie	+	14.7 (6.3–24.0)	0.193
	−	16.3 (4.5–26.4)	
Intertonsillar & tonsil to CMJ arachnoid adhesions	+	15.0 (4.5–26.4)	0.820
	−	15.3 (6.3–22.0)	
Vermian branch of PICA obstructing foramen of Magendie	+	13.7 (6.3–24.7)	**0.019**
	−	16.0 (4.5–26.4)	
Arachnoid veil obstructing foramen of Magendie	+	15.1 (4.5–24.7)	0.872
	−	14.9 (5.0–26.4)	

+ = present; − = absent.
Student t-tests for continuous variables and Fisher's exact tests for dichotomous variables were used to compare the group without syringomyelia with the group with syringomyelia. Boldface type indicates statistical significance.

the foramen magnum. Copyright Brian J. Dlouhy. Published with permission. Click here to view.

Different types of arachnoid veils were observed. These included veils that completely occluded the foramen of Magendie (Fig. 6), were partially perforated (Fig. 4), or covered the inferior two-thirds or inferior one-third portion of the foramen of Magendie (Fig. 3). Arachnoid veils were observed in 59.5% (47/79) of patients with CM-I who also had syringomyelia, which was significantly greater than the 33.3% (10/30) of patients with CM-I without syringomyelia who had an arachnoid veil (p = 0.018, Table 4). Logistic regression results showed that an arachnoid veil was associated with increased risk of syringomyelia in adjusted analyses. The presence of CM-I with an arachnoid veil had 3.22 times the odds (p = 0.013, 95% CI 1.29–8.07, by multivariate logistic regression) of having syringomyelia, adjusting for tonsillar herniation (Table 5). There was no difference in arachnoid veils across age or degree of tonsillar herniation (Tables 2 and 3).

Complete veils occurred in 12.7% (10/79) of patients with CM-I who also had syringomyelia but in only 3.3% (1/30) of patients with CM-I without syringomyelia. These complete veils had no perforations and completely prevented CSF flow through the foramen of Magendie—CSF ballooned the arachnoid veil outward. Once the veil was punctured, immediate egress of CSF was observed. In most cases, these complete arachnoid veils were vascularized, with small vessels within them. Once opened, the fourth ventricle CSF flow was visualized as well as the choroid plexus.

In perforated arachnoid veils, tears or perforations were found within the veil, which partially allowed CSF to flow through the foramen of Magendie. They had the appearance of webs. These occurred in 28.4% (31/109) of patients. The inferior two-thirds arachnoid veils occluded the inferior two-thirds of the foramen of Magendie. This occurred in 6.4% (7/109) of patients. The inferior one-third arachnoid veils occluded the inferior one-third of the foramen of Magendie. This occurred in 7.3% (8/109) of patients.

Intradural Pathology Obstructing and Not Obstructing Intraoperative CSF Flow

We observed intraoperatively that intradural pathology was either obstructive or not obstructive to CSF flow channels through the foramen of Magendie and foramen magnum. Pathology not obstructive to CSF channels and flow included the following: arachnoid opacification, thickened arachnoid, arachnoid beading, ischemic and gliotic tonsils, tonsillar cysts, and the inferior descent of the fourth ventricle and CMJ.

Pathology that was obstructive to CSF channels and flow included the following: medialized cerebellar tonsils, cerebellar tonsils overlying the fourth ventricle and foramen of Magendie, intertonsillar and tonsil to CMJ arachnoid adhesions, vermian PICA branches obstructing the foramen of Magendie, and arachnoid veils obstructing or occluding the foramen of Magendie.

Discussion

Key Results and Interpretation

This is the first study to identify, characterize, and ex-

TABLE 4. Intradural pathology associated with CM-I stratified by the presence or absence of syringomyelia

Characteristic	Value		p Value
	CM-I w/o Syringomyelia, n = 30	CM-I w/ Syringomyelia, n = 79	
Demographic & preop variables			
Age in yrs	19.0 (3–42)	17.2 (2–61)	0.493
Male sex	9 (30.0)	30 (38.0)	0.507
Tonsillar herniation in mm	17.0 (8–26.4)	14.3 (4.5–23)	**0.014**
Cervicomedullary buckle	24 (80.0)	61 (77.2)	1.000
Nonobstructive intradural pathology			
Arachnoid opacification	15 (50.0)	21 (26.6)	**0.024**
Thickened arachnoid	3 (10.0)	1 (1.3)	0.063
Arachnoid beading	2 (6.7)	1 (1.3)	0.183
Ischemic & gliotic tonsils	15 (50.0)	29 (36.7)	0.275
Rt	4 (13.3)	10 (12.7)	NA
Lt	3 (10.0)	7 (8.9)	NA
Both	8 (26.7)	12 (15.2)	NA
4th ventricular & CMJ descent	24 (80.0)	61 (77.2)	1.000
Tonsillar cysts	1 (3.3)	0 (0)	NA
Obstructive intradural pathology			
Medial tonsils	30 (100)	79 (100)	NA
Tonsils overlying foramen of Magendie	5 (16.7)	18 (22.8)	0.604
Rt	5 (16.7)	11 (13.9)	NA
Lt	0 (0)	7 (8.9)	NA
Intertonsillar & tonsil to CMJ arachnoid adhesions	27 (90.0)	66 (83.5)	0.549
Vermian branch of PICA obstructing foramen of Magendie	15 (50.0)	32 (40.5)	0.394
Rt	3 (10.0)	11 (13.9)	NA
Lt	5 (16.7)	5 (6.3)	NA
Both	7 (23.3)	16 (20.3)	NA
Arachnoid veil obstructing foramen of Magendie	10 (33.3)	47 (59.5)	**0.018**
Complete	1 (3.3)	10 (12.7)	NA
Perforated	8 (26.7)	23 (29.1)	NA
Inferior two-thirds	1 (3.3)	6 (7.6)	NA
Inferior one-third	0 (0)	8 (10.1)	NA

Values are expressed as the number of patients (%) or as the mean (range), unless otherwise indicated. Student t-tests for continuous variables and Fisher's exact tests for dichotomous variables were used to compare the group without syringomyelia with the group with syringomyelia. Boldface type indicates statistical significance.

amine intradural pathology in patients of all ages (children and adults) who had CM-I with and without syringomyelia. In this prospective study, we found that intradural pathology exists in many forms and is much more prevalent than previously recognized in patients of all ages. Interestingly, a subset of this pathology obstructed intraoperative CSF flow channels at the level of the foramen of Magendie and the foramen magnum, and therefore may play a role in the pathophysiology of both CM-I tonsillar herniation and CM-I–associated syringomyelia. Further supporting this, almost all of the intradural pathological findings were present in patients from the very young to the very old, suggesting that much of this pathology is not acquired from long-standing tonsillar herniation but is inherent to the pathophysiology of both CM-I tonsillar herniation and syringomyelia. An analysis of intradural pathology that obstructed CSF flow intraoperatively revealed that arachnoid veils were significantly more prevalent in patients who had CM-I with syringomyelia compared with those who had CM-I without syringomyelia.

Cerebellar Tonsil CSF Obstruction: Medialized Tonsils and Tonsil Overlying the Foramen of Magendie

The cerebellar tonsils in CM-I appeared to obstruct CSF flow channels in all patients. The tonsils were inferior in descent and medially approximated in all cases. However, in a subset of patients (21.1%), tonsillar medialization was asymmetrical, and one cerebellar tonsil occupied most of the cisterna magna and overlaid the fourth ventricle, obstructing CSF flow through the foramen of Magendie and the foramen magnum. In many cases, the cerebellar tonsil needed to be lifted superiorly and laterally to allow better CSF flow intraoperatively. The prevalence of this finding in this cohort is similar to the prevalence (26% of patients with cerebellar tonsillar occupation and obstruction of the foramen of Magendie

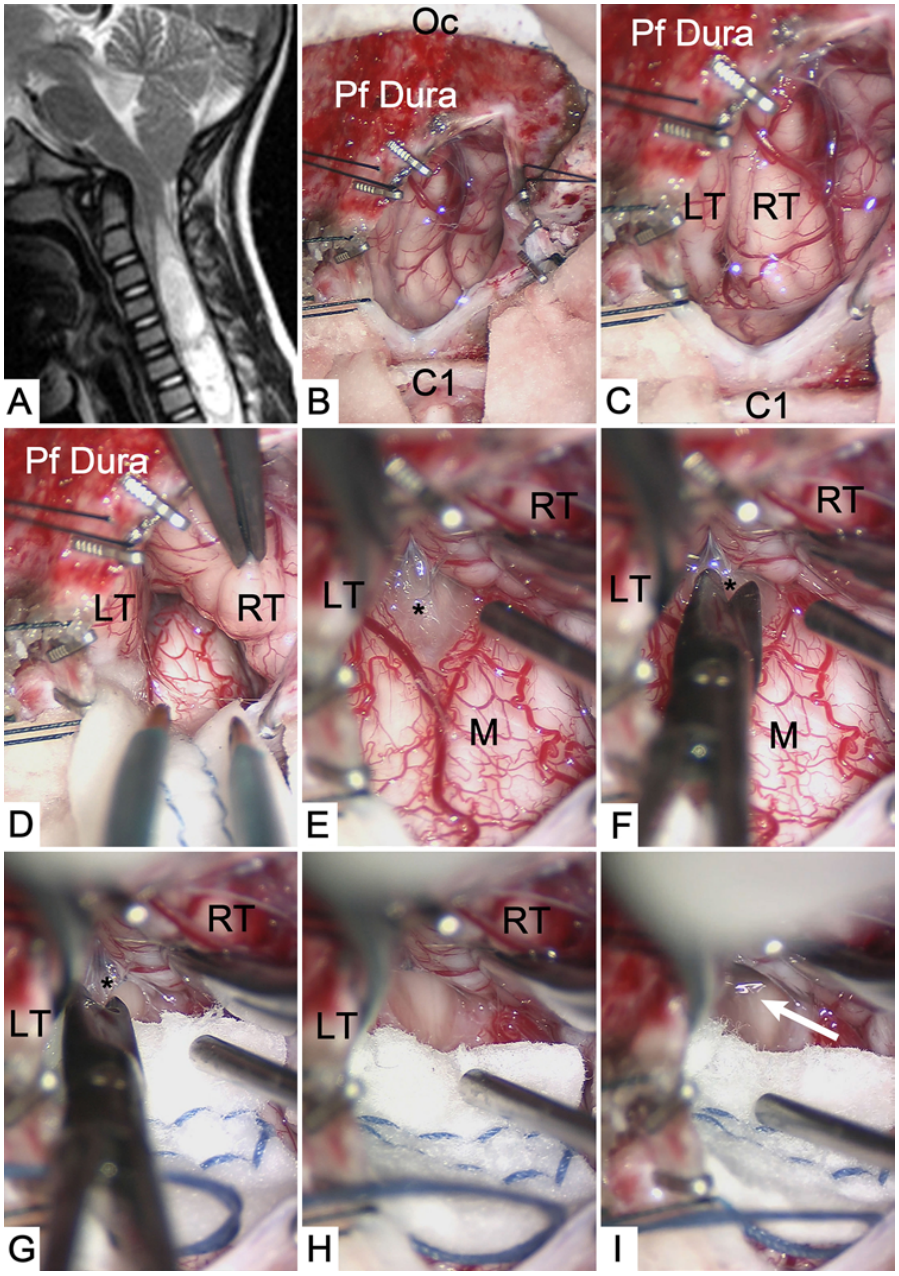

FIG. 6. A 6-year-old girl with CM-I and syringomyelia. Intraoperative illustration of medialized tonsils and complete arachnoid veil occluding the foramen of Magendie. **A:** Preoperative sagittal T2-weighted MRI study demonstrating CM-I with tonsillar herniation and syringomyelia. **B and C:** Suboccipital craniectomy performed superiorly to the inferior nuchal line and approximately 30 mm in diameter, with foramen magnum decompression and superior two-thirds C-1 laminectomy with a curvilinear dural opening. On opening the dura, the arachnoid was almost completely translucent and tonsils were observed to be medialized. The arachnoid was thickened when incised in the midline. **D:** Separation of the medialized tonsils revealed fine intertonsillar arachnoid adhesions. **E:** With inspection of the foramen of Magendie, a complete veil (*asterisk*) was observed. No CSF was flowing through the foramen of Magendie. **F and G:** The arachnoid veil (*asterisk*) was lysed and cut out, and the edges were cauterized to prevent recurrence. **H and I:** Complete opening of the foramen of Magendie with CSF noted to flow through it after opening, noted by the glistening appearance of CSF (panel I, *arrow*). Figure is available in color online only.

TABLE 5. Obstructive intradural pathology and risk analysis for syringomyelia performed using logistic regression analysis

Intradural Pathology	Total No. of Patients	Syringomyelia, n = 79	
		No. (%)	Adjusted OR (95% CI)
Tonsillar herniation in mm, continuous			**0.89 (0.81–0.98)**
Arachnoid veil			
No	52	32 (61.5)	1.0
Yes	57	47 (82.5)	**3.22 (1.29–8.07)**

Adjusted odds ratios were determined using multivariate logistic regression analysis. Boldface type indicates statistical significance.

and foramen magnum) in the previous cohort of patients treated by the senior author.[23]

The PICA Vermian Branches Obstructing the Foramen of Magendie

In the present cohort, we found that one or both PICA vermian branches were sandwiched between the tonsils and obstructed the foramen of Magendie in 43.1% of patients. Similarly, in a previous report by the senior author,[23] PICA branches were found to be sandwiched between the cerebellar tonsils in 25% of patients with CM-I. We suspect that the slightly greater prevalence in the current cohort is due to our being more aware of this pathological finding intraoperatively. The PICA vermian branches[19] are often attached to arachnoid adhesions and one of the cerebellar tonsils. With lysis of arachnoid adhesions and piarachnoid coagulation of the tonsils, the vessel can be pulled away from the foramen of Magendie, thereby opening and improving CSF flow. The vermian PICA branches can be observed to enter the foramen of Magendie on preoperative MRI, and this can be useful to assess whether this plays a role in the patient's CM-I.

Arachnoid Veils or Webs Obstructing and Occluding the Foramen of Magendie

We found that arachnoid veils of all types were more common than previously reported and were significantly more common in patients who had CM-I with syringomyelia than in those without. A veil (all types, from occluding only one-third of the foramen of Magendie to complete occlusion) was present in 33.3% of patients with CM-I alone and in 59.5% of patients who had CM-I with syringomyelia, suggesting that arachnoid veils may play a role in the pathogenesis of syringomyelia.

Tubbs et al.[41] found that the foramen of Magendie was obstructed by an arachnoid veil or web in 12.5% (10/80) of patients who had CM-I and syringomyelia. Remarkably similar to this report, a veil completely occluding the foramen of Magendie was found in 12.7% of our patients with CM-I and syringomyelia. Saez et al.[31] reported that the foramen of Magendie was obstructed and wholly occluded by an arachnoid veil in 27% of patients. It is unclear if these other reports also found partial and incomplete veils in a similar prevalence to our cohort, because they did not note these types of veils in their reports. The etiology of arachnoid veils is unknown. Tubbs et al. suggested that these veils were remnants of the rhombic lip, as described by Gardner and Goodall,[12] and therefore congenital. However, other acquired causes such as infection, inflammation, and trauma may be possible.

Veils appeared to either partially or completely obstruct flow of CSF through the foramen of Magendie. The complete occlusion was evident when the veil would billow in and out with CSF pulsations but would not allow CSF to flow through the foramen of Magendie. With opening of these veils, first by puncturing and then excising it, and cauterizing the edges to prevent recurrence, we found CSF to then flow freely through the foramen of Magendie (Video 1).

Intertonsillar and Tonsil to CMJ Arachnoid Adhesions

Arachnoid adhesions have been reported and are well known to exist in the posterior fossa in patients with CM-I.[17] We observed that 85.3% of patients with CM-I had adhesions, which existed in 2 different types. Arachnoid adhesions were thin and fine, and in other cases much thicker—almost scarlike. Adhesions were found between tonsils, between the tonsils and CMJ, and in some cases surrounded the foramen of Magendie. We found no difference in adhesions between patients with and without syringomyelia. The cause of arachnoid adhesions in CM-I is unknown, but reports have found an association between such adhesions and birth trauma.[26] Others have shown an association between birth trauma and CM-I with syringomyelia.[15] Klekamp[17] found an association with the degree of arachnoidopathy and syringomyelia. This arachnoidopathy included arachnoid adhesions that occluded the foramen of Magendie.

Other Intradural Pathology Not Obstructive to CSF Flow

The arachnoid was found to be opaque and thickened in patients with CM-I. As with the cause of arachnoid veils, webs, and adhesions, the cause of opacification of the arachnoid is unknown. However, as suggested previously, inflammation, infection, or trauma may play a role. Other findings included the inferior descent of the fourth ventricle and CMJ. The medullary buckle[34] seen on preoperative MRI correlated with the inferior descent of the CMJ intraoperatively.

We found ischemic and gliotic tonsils in 40.4% of patients. Ischemic and gliotic tonsils are believed to result from the inferior descent and impaction of the cerebellar tonsils through the foramen magnum and into the cervical canal by the pulsating passage of CSF.[18,30,39] The ischemia and gliosis is probably a combination of both physical impaction and loss of blood supply. The systolic impaction of the tonsils into the canal further exacerbates CSF obstruction, and tonsillar CSF pulsations continue to push the tonsils further into the canal.

Only a few reports in the literature have documented the presence of tonsillar cysts[35,39]—cysts at the tip of the cerebellar tonsils—in patients with CM-I. In a report by Tubbs et al.,[39] 6% (4/67) of patients with CM-I were found to have tonsillar cysts, and these authors suggested that cerebellar tonsillar ischemia led to tonsillar cysts. We found a tonsillar cyst in only 1 patient (0.9%), a 6-year-old

girl with CM-I. Tonsillar cysts do not appear to be a common finding.

Pathophysiology of CM-I: Extradural and Intradural Causes of Tonsillar Herniation

Evidence suggests that some patients with CM-I have a congenital or developmental hypoplastic posterior fossa and reduced posterior fossa volume that is associated with underdevelopment of the occipital bone, low-lying tentorium, or thickened or elevated occipital bone.[1,3–5,9,11,13,21,24,25,27,28,32,33,36,37,40] It is postulated that this small posterior fossa cannot hold the volume of the cerebellum, thus leading to tonsillar herniation through the foramen magnum and to CSF obstruction. How the constriction of the cerebellum in development results in downward tonsillar herniation has been illustrated in our findings. Medialized tonsils, obstruction of the foramen of Magendie by a cerebellar tonsil, and the inferior descent of the tonsils could be a direct result of the congenital or developmental posterior fossa hypoplasia. However, other studies have not shown a reduction in posterior fossa volume in patients with CM-I,[32,36,40] indicating that tonsillar herniation may occur through other mechanisms.

The widespread prevalence of intradural pathology that is obstructive to CSF flow in patients with CM-I suggests that this pathology may also play a role in tonsillar herniation. Arachnoid veils, PICA vermian branches, arachnoid adhesions, medialized tonsils, and fourth ventricular tonsils obstruct free pulsatile CSF flow through the foramen of Magendie and foramen magnum. This obstruction can lead to an abnormal craniospinal pressure gradient[42,44,45] and systolic CSF flow or pulsations that impact the cerebellum and cerebellar tonsils.[14,29] It is this effect of CSF flow or pulsations on the cerebellum that may result in downward herniation of the tonsils through the foramen magnum. The intradural restriction or obstruction of CSF flow through the foramen of Magendie and foramen magnum by the abundance of intradural pathology in patients with CM-I provides an additional mechanism in this complex pathophysiology and may help explain why some patients do not have a reduction in posterior fossa volume.

Pathophysiology of CM-I With Syringomyelia: Extradural and Intradural Causes of Syringomyelia

Previous studies have suggested that syringomyelia in the setting of CM-I occurs from obstructed CSF flow at the level of the foramen magnum due to tonsillar herniation. According to Gardner and Goodall[12] and Williams,[43] albeit through different proposed mechanisms, this obstruction results in CSF entering the central canal and development of a spinal syrinx. However, according to Oldfield et al.,[29] obstruction at the foramen magnum creates a pistonlike effect of the tonsils on spinal CSF, which creates increased spinal subarachnoid pressure and increased subarachnoid pulse pressure, driving CSF into the spinal cord. Regardless of the cause of syrinx formation, all theories suggest that syringomyelia results from obstructed CSF flow at the level of the foramen magnum. However, most patients with CM-I have tonsillar obstruction, and therefore it is unclear why some have an associated syrinx and others do not.

Our findings suggest that intradural pathology disrupts and restricts CSF flow at the level of the foramen magnum. In a multivariate analysis, we found that arachnoid veils that obstruct the foramen of Magendie were significantly more common in patients with CM-I who have syringomyelia than in those with CM-I but without syringomyelia, suggesting that arachnoid veils may play a role in the pathophysiology of CM-I–associated syringomyelia. Lack of proper flow through the foramen of Magendie disrupts the normal CSF flow through the fourth ventricle into the cisterna magna and through the foramen magnum. Obstruction at the level of the foramen of Magendie and at the foramen magnum from both intradural and extradural pathology is consistent with the proposed theories on the pathogenesis of CM-I–associated syringomyelia by Gardner and Goodall,[12] Williams,[43] and Oldfield et al.[29] Obstruction of the foramen of Magendie by arachnoid veils or other intradural pathology could potentially force CSF through the central canal according to Gardner and Goodall, and Williams. On the contrary and in agreement with the theory proposed by Oldfield et al., obstruction of the foramen of Magendie by an arachnoid veil or other intradural pathology obstructs CSF flow out of the fourth ventricle, which could lead to downward herniation and the pistonlike effect of the tonsils on spinal CSF, creating increased spinal subarachnoid pressure and increased subarachnoid pulse pressure, potentially leading to syrinx formation.

Clinical Importance of These Findings

Successful treatment of CM-I occurs when normal pulsatile CSF flow through the outlet of the fourth ventricle and across the foramen magnum is restored and brainstem and upper cervical spinal cord compression is relieved.[13] There is enough evidence to suggest that any mechanism that resolves the obstruction of normal CSF pathways and decompresses the brainstem will result in proper treatment. The surgical treatment of CM-I in patients with and without syringomyelia varies from posterior fossa extradural decompression only[16] to posterior fossa extra-intradural decompressions[20,22,23] and their variations, as well as anterior decompressions.[21]

The widely varying results in the surgical treatment of CM-I are multifactorial but reflect the fundamental lack of understanding of the pathophysiology of CM-I tonsillar herniation and CM-I–associated syringomyelia. The heterogeneity of CM-I, in which both extradural and intradural pathology play a role in the pathophysiology, varies among patients. Because of this, a universal single best surgical treatment for all cases of CM-I, either with or without syringomyelia, is unlikely to be found. Precision medicine relies on a better understanding of the pathophysiology of a disease, and only through this will we be able to distinguish which patients should be treated and how they should be treated, in a tailored treatment strategy.

Our data suggest that intradural pathology may play a role in the pathophysiology of tonsillar herniation and syringomyelia. Predicting which patients require both a posterior fossa extradural and intradural decompression with duraplasty to achieve proper CSF flow will be a challenge for future studies. However, our data suggest that arachnoid veils are more common in patients with CM-I and

syringomyelia, implying that these patients may do better with both a posterior fossa extradural and intradural decompression and duraplasty.

Limitations of the Study

Although this study was prospective in nature, it was not a randomized study. Therefore, there is selection bias for patients with CM-I who underwent intradural exploration and duraplasty. During this study, most patients with CM-I who had syringomyelia underwent both posterior fossa extradural and intradural decompression. However, patients with CM-I without syringomyelia underwent an intradural decompression and duraplasty if the foramen magnum still appeared obstructed on intraoperative ultrasound after the posterior fossa extradural decompression. Because of this, the latter cohort may be more severe in terms of obstruction at the level of the foramen magnum. Therefore, the intradural pathology observed in the CM-I cohort without syringomyelia may be of higher prevalence than the whole population of CM-I without syringomyelia. Furthermore, some variables reported here are categorical and therefore depend on the judgment of the operating surgeon in distinguishing "yes" versus "no" in reporting these findings. There is no way to provide a more continuous variable for some of these findings. However, the data presented here had an internal control because a single surgeon provided the intradural findings and reports for all patients.

Conclusions

Intradural pathology associated with CM-I with or without syringomyelia exists in many forms, is much more prevalent than previously recognized in patients of all ages, and may play a role in the pathophysiology of CM-I tonsillar herniation. Arachnoid veils may play a greater role in the pathogenesis of CM-I–associated syringomyelia than other intradural CSF obstructive pathology. Arachnoid veils appear to partially obstruct CSF flow, are significantly more prevalent in cases of CM-I with syringomyelia, and therefore may play a role in the pathophysiology of CM-I–associated syringomyelia.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Dlouhy, Menezes. Acquisition of data: Dlouhy, Menezes. Analysis and interpretation of data: Dlouhy, Menezes. Drafting the article: Dlouhy. Critically revising the article: Dlouhy, Menezes. Reviewed submitted version of manuscript: Dlouhy, Menezes. Approved the final version of the manuscript on behalf of all authors: Dlouhy. Statistical analysis: Dlouhy, Dawson. Administrative/technical/material support: Dlouhy, Menezes. Study supervision: Dlouhy, Menezes.

Supplemental Information

Videos

Video 1. https://vimeo.com/226722608.

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