https://doi.org/10.48047/AFJBS.4.3.2022.130-139

African Journal of Biological Sciences

Journal homepage: http://www.afjbs.com

Research Paper Open Access

Role of Surgery in Chiari Type I Malformation

Yasser Ahmed Abdalraheem*, M.D; Adham Rabie Abd alaziem, M.D; Hany Mohailaba, M.D

Neurosurgery Department, Faculty of Medicine, Beni-Suef University, Beni-Suef 62511, Egypt

***Corresponding author:** Yasser Ahmed Abdalraheem,

E-mail: yasserneuro2020@gmail.com

Volume 4, Issue 3, Nov 2022

Received: 15 Sep 2022

Accepted: 16 Oct 2022

Published: 18 Nov 2022

doi: 10.48047/AFJBS.4.3.2022.130-139

Abstract:

Background: The foramen magnum lowers the cerebellar tonsils in Chiari type 1 malformation (CM-1). Patients may be asymptomatic or have mild to severe symptoms. **Objective**: To determine whether surgery is effective in treating Chiari malformation type I symptoms. **Methodology**: From March 2019 to March 2022, the Beni-Suef University Hospitals research team retrospectively studied 10 CM-1 patients who received surgical decompression. All patients underwent preoperative neurological, ophthalmological, and radiographic tests. All patients got craniocervical junction MRI and brain CT. Microsurgical procedures included suboccipital craniectomy (2 cm x 2 cm), C1 laminectomy, Y-shaped dural incision, cerebellar tonsil excision, and augmentative duroplasty with fascia lata graft. After 24 hours, all patients had CT brain scans. We scheduled post-op MRIs for three and six months. **Results**: The average age of the patients was 29.2 years, with 60% being female. All patients (100%) developed Valsalva-aggravated occipital headache, neck pain in 8 patients, and dizziness and tiredness in 7 individuals. There are eight instances of Chiari malformation type I associated with syringomyelia. After surgery, 5 patients (50%) had good results, 3 (30%) had medium outcomes, and 2 (20%) had poor outcomes. A follow-up MRI six months after surgery showed a moderate to large decrease in syringomyelia syrinx size in four patients, a slight reduction in two and no change in size compared to pre-operative measures in two individuals. All procedures worked. For chronic wound collection, only one case necessitated a ventriculoperitoneal shunt. **Conclusion**: Patients' needs and symptom intensity should guide the customization of CM-1 therapy. In symptomatic patients, decompression of the posterior fossa bone, cautious durotomy, tonsil reduction, and duroplasty with fascia lata graft all improve clinical and radiological outcomes. Closing the dura watertight prevents cerebrospinal fluid leaks and wound infection.

Keywords: Chiari type I malformation, Suboccipital craniectomy, Duroplasty

1. Introduction:

Chiari type I malformation (CM-1) is characterized by the displacement of the cerebellar tonsils downwards beneath the foramen magnum and congenital hypoplastic posterior fossa. Despite the emergence of various theories, the exact cause of CM-1 remains unknown [1-2]. Nishikawa et al. [3] studied 50 healthy people and 30 people with CM-I. They found that the posterior fossa wasn't developing properly in the CM-I patients, which pushed on the cerebellum and brainstem and let them protrude through the foramen magnum[3]. Furthermore, hydrocephalus, posterior fossa tumors, lumbar-peritoneal shunts, and posterior fossa vascular abnormalities are the causes of acquired CM-1 [4,5]. Patients with CM-1 may have no symptoms and require monitoring rather than surgery [6].

Occipital headaches, the most common type, account for almost all cases (98%). When you cough, laugh, or bend over, you might trigger a vagus nerve, which can make your headache worse. Other common symptoms include diplopia, vertigo, tinnitus, otalgia, pain in the neck and shoulders, weakness in the upper limbs, paresthesia, decreased fine motor skills, dysphagia, sialorrhea, sleep apnea, and intermittent nausea and vomiting. The weakening of the intrinsic hand muscles and the loss of sensation in the upper limbs that looks like a cape could be signs of syringomyelia, which happens to 45–75% of children with CM-1. About a quarter of people with CM-1 also have scoliosis [7, 9].

People may have symptoms at any age; however, it often doesn't appear until the thirties, even though it's a congenital defect. The gold standard for diagnosing CM-1 is cranio-cervical magnetic resonance imaging (MRI). Tonsillar herniation of 3–25 mm or more may indicate CM-1. Traditional medical practice dictates that patients with persistent symptoms and tonsillar herniation located below the foramen magnum as shown by MRI have the option of surgical decompression. With or without duroplasty, surgical decompression is the standard treatment for CM-1 [10, 11, 12].

The underlying cause of CM-1 may sometimes be treated through surgery, such as taking out tumors from the posterior fossa or putting in a ventriculoperitoneal shunt to treat hydrocephalus [13]. This study's primary goal is to assess the value of the surgical management in the improvement of the symptoms & signs of patients with chiari malformation type I.

2. Material and Methods:

2.1. Study design and participants:

This is study was done retrospectively; it included 10 cases with CM-1 that were surgically managed between March 2019 and March 2022, at the neurosurgery department Beni-Suef university hospitals.

Inclusion criteria:

Cases with confirmed CM-1 via MRI scan of craniocervical junction from any age or sex with persistent symptoms and had surgery were involved in the study.

Exclusion criteria:

A symptomatic patients, cases with mild stationary symptoms that had no surgery, or patients those were unfit for surgery due to their medical conditions.

All patients in the review were subjected to:

2.2. Clinical assessment:

• Collecting a patient's medical history, this should include any chronic diseases, medications, or health issues such as high blood pressure, diabetes, heart disease, or diabetes mellitus. The mode of onset, length, and development of his complaint were also part of the present history. Headaches, vomiting, eye issues (loss of vision, blurred vision), sphincter dysfunction, heavy feeling, tingling, and numbness were the particular symptoms that patients were asked about. Pulse, blood pressure, respiration rate, temperature, evaluation of the thorax and abdomen, evaluation of the genitourinary system, and study of the skeletal system are all parts of a thorough examination.

• Comprehensive eye and neurological examination, including testing of visual acuity, motor and sensory systems, and cognitive function.

2.3. Investigations:

- Radiological: MRI brain to exclude presence of any primary causes as hydrocephalus or posterior fossa tumors, MRI cranio-cervical junction to confirm the diagnosis of CM-1 .
- Laboratory: Preoperative routine investigations i.e. CBC, bleeding profile, renal function, liver function, random blood sugar. Also, blood grouping and cross matching.

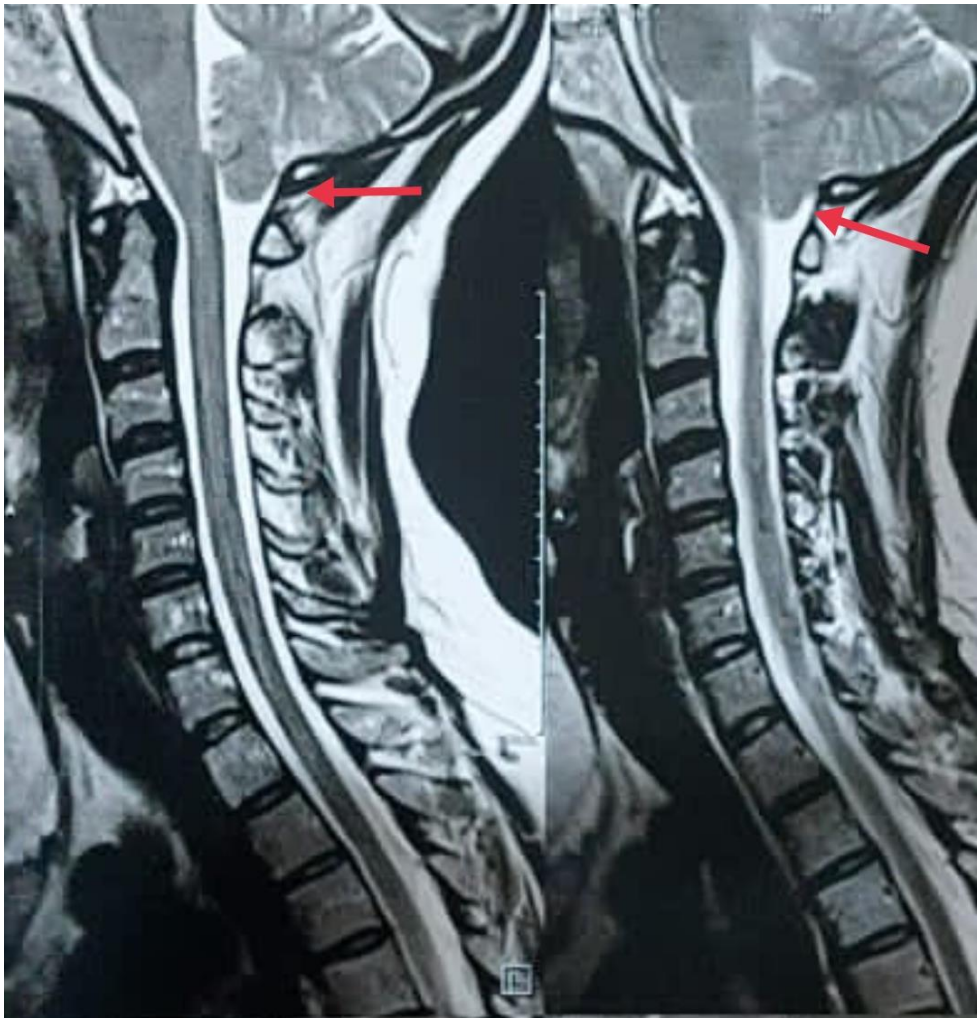

Figure (1): Cerebellar tonsils herniation through foramen magnum.

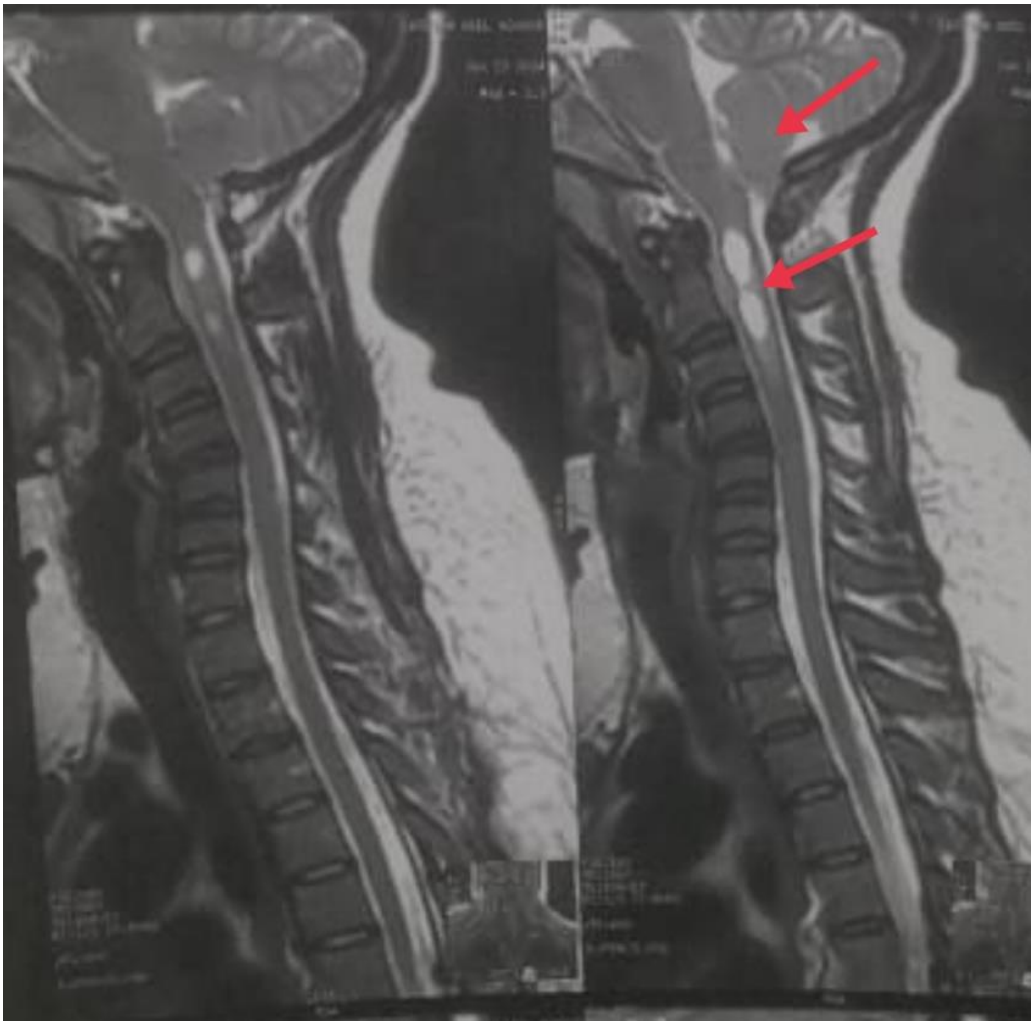

Figure (2): Shows cerebellar tonsils herniation through foramen magnum with upper cervical syringomyelia

2.4. Surgical intervention:

Surgical intervention was used to treat all cases in this research. Duroplasty with a fascia lata graft was performed in conjunction with posterior fossa decompression in all patients. A suboccipital craniectomy of about 2cm x 2cm, C1 laminectomy, Y-shaped dural incision, and reduction of the cerebellar tonsils were regularly executed using microsurgical techniques. The tips of the cerebellar tonsils were excised using a subpial approach, either by coagulation or a combination of both methods, depending on the necessary extent to achieve patency and full decompression of the fourth ventricle. Dural augmentation with fascia lata grafts was performed in all instances **(14)**.

2.5. Postoperative management:

Immediate postoperative data, including motor strength and degree of awareness, was evaluated in all patients. Standard intensive care units stay after surgery for about 24 hours to provide careful patient monitoring. Eight hours after surgery, the first CT scan of the brain was performed as per protocol. Typically, 5 to 15 days after surgery, we observed patients as outpatients. After three to six months, clinicians looked for signs of symptom improvement

or worsening, and radiologists used MRI and CT scans to look for ventriculomegaly and measure the size of the syrinx, if present.

2.6. Clinical outcome evaluation:

The clinical outcome was evaluated as follows: (I) Excellent: patients exhibiting significant improvement with no residual symptoms resulting in any disability; (II) Fair: patients with residual symptoms causing mild disability in daily activities; (III) Poor: patients showing no improvement whatsoever.

2.7. Ethical consideration:

No written informed consent was provided due to the retrospective nature of the study. Approval was obtained from the department council of neurosurgery Beni-Suef University.

2.8. Statistical analysis:

The collected data was coded to improve data processing and then entered twice into Microsoft Access. The data was analyzed using SPSS Inc.'s SPSS software version 22 on Windows 7. Basic descriptive analysis of qualitative data utilizing numerical values and percentages, with arithmetic means used to quantify central tendency and standard deviations to analyze the dispersion of quantitative parametric data.

3. Results:

Out of the 10 patients, 6 patients (60%) were females while 4 patients (40%) were males. Our mean age in the study was 29.2 (with minimum age was 19 years while the maximum age of present study cases was 50 years). Seventy-five percent of the patients were aged between 20 and 40 years.

All patients (100%) had the characteristic occipital headache intensified by Valsalva exercises. All patients exhibited one or more of the following symptoms, listed in descending order of prevalence: neck pain in 8 patients, dizziness and fatigue in 7 patients, numbness and paresthesia of the extremities in 6 patients, weakness of fine motor skills in 4 patients, cerebellar dysfunction (ataxia) in 2 patients, and sphincteric manifestations in 1 patient (Table 1).

Table (1): Showing the clinical presentation.

Clinical presentation	Number of patients	Percentage %
Headache	10	100
Neck pain	8	80
Dizziness& fatigue	7	70
Extremities numbness & parathesia	6	60
Weakness of fine movements	4	40
Cerebellar dysfunction (ataxia)	2	20
Sphincteric dysfunction	1	10

In our research, 5 patients (50%) exhibited excellent outcomes, 3 patients (30%) shown good outcomes, and 2 patients (20%) had bad outcomes (Table 2).

Table (2): Showing the clinical outcome.

Clinical outcome	Number
excellent	5
Fair	3
Poor	2

Cervical syringomyelia was present in 8 patient (80%) pre-operative. 6 months after surgery follow up MRI CS was done for patients with syringomylia that revealed, moderate to marked reduction in the syrinx size in 4 patients, mild reduction in 2 patients, and the same syrinx size as pre-operative in 2 patients. All surgeries passed safely. 2 patients had post-operative wound collection that were managed primary with repeated aspiration and close follow up, and one of them needed ventriculo-perotineal shunt (VPS) insertion.

4. Discussion:

It's possible that CM-1 will have a negative effect on quality of life and cause problematic symptoms that need surgical intervention. However, there is still a paucity of understanding among scientists on how the preoperative and intraoperative conditions of CM-1 patients affect the outcomes of their operations. This study's primary goal is to assess the value of the surgical management in the improvement of the symptoms & signs of patients with chiari malformation type I.

This study focuses on ten individuals diagnosed with CM-1 abnormalities who underwent surgery at Beni-Suef University Hospital's neurosurgery department between 2019 and 2022. Our study's female-to-male ratio of 1.5:1 is in agreement with **Alzate et al.'s** [15] results and suggests that there is a preponderance of females because 60% of the patients were female. The age at presentation ranged from 19 to 50 years, with a mean of 29.2 years. According to **Alzate et al.** [15]**,** most instances occurred throughout adolescence.

Our research showed that Valsalva maneuvers aggravated occipital headaches in every single subject. Following is a list of symptoms that all patients displayed, ranked from most common to least: 8 patients reported neck pain, 7 reported dizziness and fatigue, 6 reported numbness and paresthesia of the extremities, 4 reported weakness in fine motor skills, 2 reported cerebellar dysfunction (ataxia), and 1 patient reported sphincteric manifestations. All patients had headaches, while 89% expressed pain in the neck, 83% claimed dizziness and fatigue, and 72% felt numbness; these results are in good agreement with **Kamal's study** [16]. Consistent with previous research, this suggests that the most common symptom in CM-1 patients is an occipital headache made worse by Valsalva movements [7, 8].

The only certain way to diagnose CM-1 is via MRI of the craniocervical junction. Tonsillar herniations of 3–25 mm or more may indicate CM-1. Additionally, we need to determine if syringomyelia is concurrently present. During our examination, we identified 8 cases with preoperative concurrent syringomyelia. To ensure that no intracranial abnormalities, such as hydrocephalus, were present, all of our study participants had CT scans of the brain prior to surgery. The gold standard for finding people who should have surgical decompression is a mix of MRI, medical history, and a physical assessment. It is

common practice to offer surgical decompression to patients who display chronic symptoms with tonsillar fall beyond the foramen magnum, as seen on MRI.

All patients undergoing posterior fossa decompression in this study underwent duroplasty using a fascia lata graft. Five patients reported a substantial improvement, three a moderate improvement, and two a poor result throughout the post-operative follow-up period. In a similar vein, 84% of patients in **Mueller et al.** [17] reported a considerable improvement in their condition after surgery. Eight patients, or 80%, had cervical syringomyelia before surgery. Four patients had a moderate to large reduction in syrinx size, two had a modest reduction, and two showed no change in size compared to pre-operative measures during a follow-up MRI six months after surgery for syringomyelia.

All surgical procedures proceeded smoothly. A VPS insertion was necessary for one patient, and regular suctioning and careful monitoring mostly managed the postoperative wound collections in the other two patients. There were no deaths among our patients. These results align with **Kamal's study** [16]**.**

Currently, we only suggest surgical intervention for symptomatic patients in CM-I situations, as the condition's natural evolution appears to be relatively harmless in asymptomatic persons. Early surgical intervention tends to yield better functional results for patients. There are a variety of surgical procedures available for these patients. Surgeries including the suboccipital craniectomy, C1 (atlas) laminectomy, and augmentative duraplasty seem to be the most often performed procedures. In some cases, strong adhesions at the interface between the skull and the neck may make it impossible to separate the arachnoid layer, coagulate, and remove the cerebellar tonsils. For patients suffering from symptomatic syringomyelia due to Chiari Malformation Type I, our main recommendation is posterior fossa decompression in conjunction with augmentative duroplasty.

5. Conclusions:

Chiari type I malformation is a neurological condition defined by the fall of the cerebellar tonsils into the spinal canal, which may cause headaches, neck pain, balance problems, and a variety of other neurological symptoms. The severity of the symptoms and the patient's specific needs should guide the management. We advise caution and frequent follow-up for asymptomatic cases or those with mild symptoms that are manageable with simple medical treatment. For symptomatic patients, we may recommend surgical decompression to relieve pressure on the brain and spinal cord.

Conflict of Interests: No conflict of interests is declared.

Fund: non-fundable.

6. References:

1. Massimi L, Novegno F, di Rocco C. Chiari type Imalformation in children. Adv Tech Stand Neurosurg. 2011; 37:143-211.
2. McVige JW, Leonardo J. Neuroimaging and theclinical manifestations of Chiari malformationtype I (CMI). Curr Pain Headache Rep. 2015; 19:18.
3. Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y. Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. Journal of Neurosurgery. 1997; 86: 40–47.
4. Vega A, Quintana F, Berciano J. Basichondrocranium anomalies in adult Chiari type I malformation: a morphometric study. Journal of the Neurological Sciences. 1990; 99: 137–145.
5. Stovner LJ, Bergan U, Nilsen G, Sjaastad O. Posterior cranial fossa dimensions in the Chiari I malformation: relation to pathogenesis and clinical presentation. Neuroradiology, 1993; 35: 113–118.
6. Meadows J, Kraut M, Guarnieri M, et al. Asymptomatic Chiari Type I malformations identified on magnetic resonance imaging. J Neurosurg. 2000; 92: 920-66.
7. Mueller DM, Oro JJ. Prospective analysis of presenting symptoms among 265 patients with radiographic evidence of Chiari malformation type I with or without syringomyelia. J Am Acad Nurse Pract. 2004; 16:134-138.
8. Fischbein R, Saling JR, Marty P, Kropp D, Meeker J, Amerine J, et al. Patient-reported Chiari malformation type I symptoms and diagnostic experiences: a report from the national Conquer Chiari Patient Registry database. Neurol Sci. 2015; 36:1617-1624.
9. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, et al. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. Neurosurgery. 1999; 44:1005-1017.
10. Chavez A, Roguski M, Killeen A, Heilman C, Hwang S. Comparison of operative and nonoperative outcomes based on surgical selection criteria for patients with Chiari I malformations. J Clin Neurosci. 2014; 21:2201-2206.
11. Hankinson T, Tubbs RS, Wellons JC. Duraplasty or not? An evidence-based review of the pediatric Chiari I malformation. Childs Nerv Syst. 2011; 27: 35-40.
12. Lee A, Yarbrough CK, Greenberg JK, Barber J, Limbrick DD, Smyth MD. Comparison of posterior fossa decompression with or without duraplasty in children with type I Chiari malformation. Childs Nerv Syst. 2014; 30:1419-1424.
13. Akar E, Kara S, Akdemir H, et al. 3D structural complexity analysis of cerebellum in Chiari malformation type I. Med Biol Eng Comput. 2017; 55:2169-82.
14. Alden TD, Ojemann JG, Park TS. Surgical treatment of Chiari I malformation: indications and approaches. Neurosurgical Focus. 2001; 11(1): 1-5.
15. Alzate JC, Kothbauer KF, Jallo GI, Epstein FJ. Treatment of Chiari type I malformation in patients with and without syringomyelia: a consecutive series of 66 cases. Neurosurg Focus. 2001; 11 (1): 1-9.

16. Kamal HM. Posterior Fossa Decompression with Microscopic Fenestration of Foramen of Magendi and Duroplasty in Treatment of Adult Chiari Type 1 Malformation: Preliminary Results in 18 Consecutive Patients. Egyptian Journal of Neurosurgery. 2009; 24: 63-78.
17. Mueller D, Oro JJ. Prospective analysis of self-perceived quality of life before and after posterior fossa decompression in 112 patients with Chiari malformation with or without syringomyelia. Neurosurg Focus. 2005; 18: 2.