SVOA Neurology

ISSN: 2753-9180

ScienceVolks Open Access

Research Article

Special Features of Chiari Malformation Type 0

Salim MEZIANI[1], Yacine FELISSI[1]*, Chafik HANDIS[1], Abdelhalim MORSLI[1]

[1]Bab El Oued University Hospital, Department of Neurosurgery.

Dr Youcef El Khatib University of Health Science – Faculty of Medicine, Algiers, Algeria.

***Corresponding Author:** Yacine FELISSI, Mohamed Lamine Debaghine University Hospital, Department of Neurosurgery, Algiers university – Faculty of Medicine, Algiers, Algeria.

https://doi.org/10.58624/SVOANE.2025.06.020

Received: June 14, 2025

Published: July 21, 2025

Citation: MEZIANI S. FELISSI Y. HANDIS C. MORSLI A. Special Features of Chiari Malformation Type 0. *SVOA Neurology* 2025, 6:4, 101-107. doi. 10.58624/SVOANE.2025.06.020

Abstract

Objective: Chiari malformation type 0 (CM-0) is an uncommon condition characterized by syringomyelia and cerebrospinal fluid (CSF) obstruction at the foramen magnum without cerebellar tonsillar descent. This study aims to delineate its clinical, radiological, and surgical aspects to refine diagnostic and therapeutic protocols.

Methods: In a prospective cohort of 80 patients operated for Chiari malformations at Bab El Oued University Hospital between 2017 and 2022, 10 adult cases of CM-0 were identified. All exhibited suboccipital headaches, sensory symptoms, cervicobrachial neuralgia, and syringomyelia. Imaging demonstrated no tonsillar herniation, reduced posterior cranial fossa (PCF) volume (mean 197.4 cm^3 vs. normal 228–244 cm^3), pointed tonsils, and CSF flow blockage. Treatment involved osteo-dural decompression, arachnoid web dissection, and autologous duraplasty.

Results: Headaches resolved postoperatively, with motor improvement reflected by JOA score increase (15 → 16.6). Syringomyelia regressed in length (16.4 → 13.4 vertebral levels) and width (Vaquero index: 0.76 → 0.38). CSF dynamics normalized in 9/10 patients; tonsils changed from pointed to rounded. However, residual symptoms included thermoalgesic dissociation (7/10) and muscle atrophy (9/10). Arachnoid adhesions blocking the foramen of Magendie were observed intraoperatively in all cases.

Conclusion: CM-0 is marked by a small PCF, atypical tonsillar morphology, and obstructive arachnoid pathology. Outcomes resemble those of Chiari type 1, but success hinges on thorough intradural inspection and arachnoid release. Tonsillar configuration and arachnoid anomalies may be pivotal in syrinx formation, affirming CM-0 as a separate clinical entity requiring individualized management.

Keywords: *Chiari Malformation Type 0; Syringomyelia and Cerebrospinal Fluid Obstruction; Posterior Cranial Fossa Volume*

Introduction

Chiari malformation is generally defined as a tonsillar herniation greater than 5 mm below the foramen magnum [1,2]

Although this definition is still widely used, authors have demonstrated that the degree of tonsillar herniation does not influence the severity of clinical signs or the extension of the syringomyelic cavity in Chiari malformation.

Some patients present with syringomyelia with classic clinical features of Chiari malformation without tonsillar herniation. These entities [1-3] have been described by Iskandar and Tubbs et al. as Chiari type 0 [4-7], where the syringomyelia generally resolves after suboccipital decompression. In our work, we will present some specific features of Chiari type 0, observed in 10 adult patients operated on for this malformation.

Materials and Methods

We conducted a prospective study on 80 patients with Chiari type 1 malformation who underwent osteo-dural decompression in the neurosurgery department of Bab El Oued University Hospital, Algiers, Algeria (during the period from December 31, 2017, to June 30, 2022). Among these 80 patients, 10 adults had Chiari type 0.

These 10 patients were female, and their ages ranged from 38 to 50 years, with an average of 44.8 years.

All patients presented with sub occipital headaches aggravated by the Valsalva maneuver. The average JOA score was 15.

Suspended sensory disturbances with thermo-algesic dissociation were found in all ten patients.

Cervicobrachial neuralgia, thenar, and hypothenar muscular atrophy were also observed in all patients.

In addition to the clinical signs noted above, two patients also had static cerebellar syndrome and cranial nerve palsy involvement, including swallowing disorders, vocal cord paralysis, and sleep apnea.

A preoperative cerebrospinal fluid MRI was performed in all patients, revealing the following parameters:

- An absence of tonsillar herniation was noted in all patients.
- The size of the posterior cerebral fossa had an average of 197.4 cm3 (the average size of a PCF in an adult subject is 244.89 cm3 in men and 228.24 cm3 in women[8].
- The shape of the inferior end of the cerebellar tonsils was pointed in all ten patients.
- The absence of a large cistern was noted in all patients.

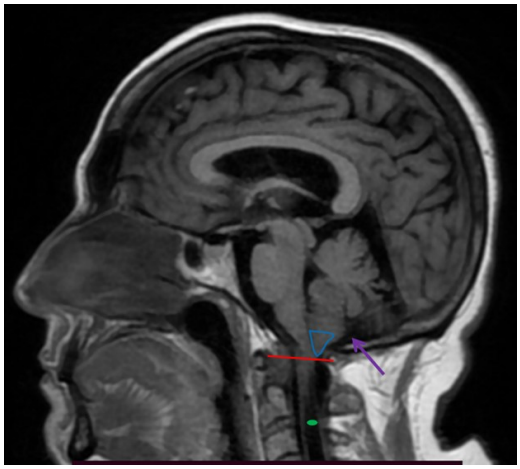

Figure 1. *Shows the absence of tonsillar herniation (tonsil blue triangle, Macrea's line in red), with syringomyelia (green dot), and absence of the large cistern (purple arrow).*

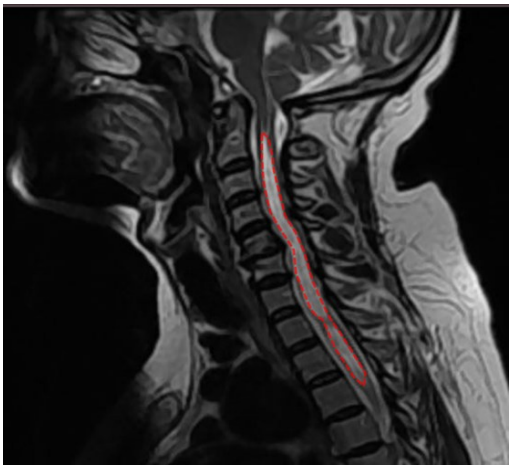

Figure 2. *Syringomyelic cavity surrounded by red dotted lines.*

- A syringomyelic cavity was found in all patients; they have a cervicothoracic location with a longitudinal extension of 16.4 vertebrae on average, and a Vaquero index of 0.76 on average.
- A preoperative MRI was performed in all patients, showing obstruction of CSF flow at the foramen magnum.

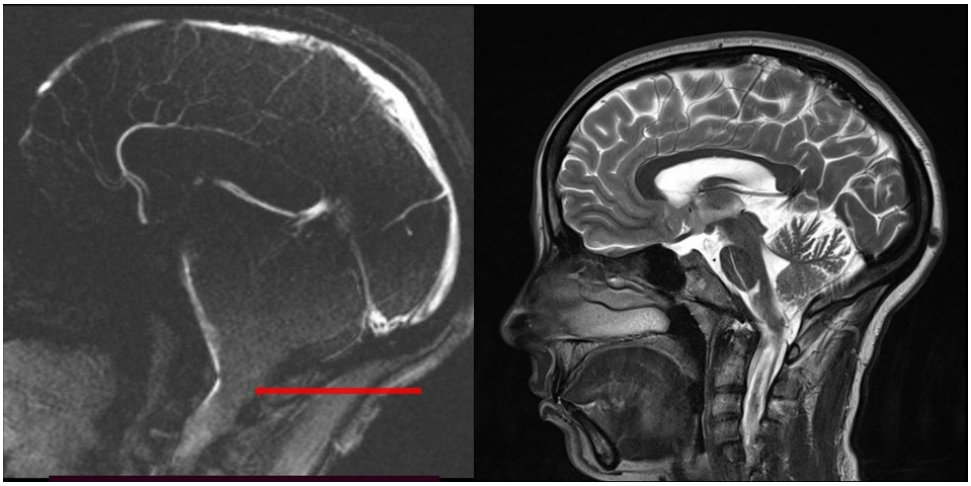

Figure 3. *An MRI cine scan showing obstruction of CSF flow at the foramen magnum. (Red arrow)*

All patients underwent surgery, benefiting from sub occipital osteo-dural decompression.

Ablation of the posterior arch of C1 was systematically performed in all our patients.

The average size of our suboccipital craniectomy was 4 cm.

A Y-shaped opening of the dura mater was performed in all our patients, followed by opening of the arachnoid mater.

- In nine of the ten patients, we found arachnoid adhesions, retaining the cerebellar tonsils at the bulbo-medullary junction.
- In all our patients, we found an arachnoid veil obstructing the foramen of Magendie.

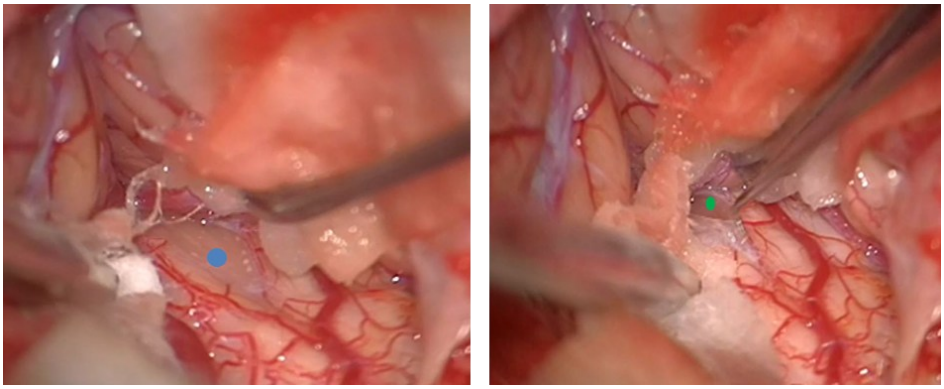

Figure 4. *Intraoperative images, arachnoid veil obstructing the Magendie foramen (blue dot), opening of the arachnoid veil (green dot).*

- Watertight closure of the dura mater was achieved in all our patients using autologous dural plasty harvested from the occipital pericranium.
- During closure of the dura mater, it was left suspended, and the arachnoid membrane was reattached to it to prevent postoperative adhesions.
- Biological glue was used in all patients.

Results

Suboccipital headaches resolved rapidly postoperatively in all our patients.

The postoperative JOA score averaged 16.6.

Suspended sensory disturbances with thermo-algesic dissociation improved in only three patients and stabilized in the rest.

Cervicobrachial neuralgia improved postoperatively in all patients, while thenar and hypothenar muscles atrophy improved in only one patient, with stabilization of this clinical sign in the rest.

Patients with signs of cranial nerve involvement and static cerebellar syndrome improved postoperatively.

A cerebrospinal fluid MRI was performed postoperatively in all patients, revealing the following parameters:

-The shape of the inferior end of the cerebellar tonsils had become rounded after surgery in all our patients.

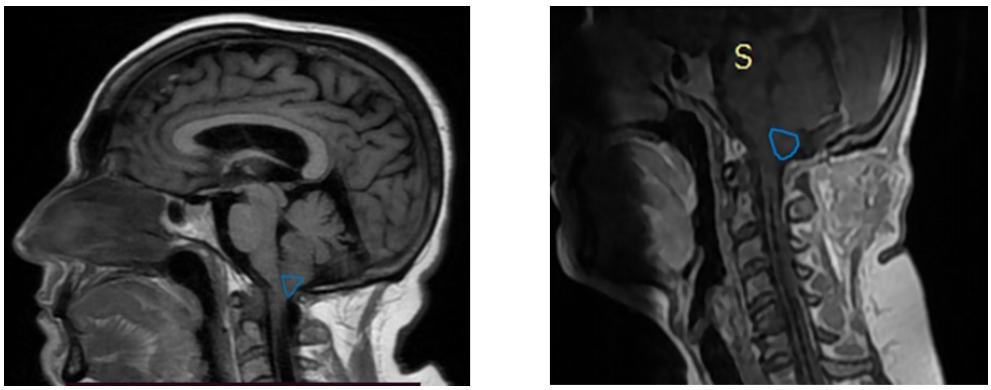

Figure 5. *Left image (lower end of the tonsils pointed preoperatively), right image (lower end of the tonsils rounded postoperatively).*

- The creation of a neo-large cisterna was noted in all patients postoperatively.
- The longitudinal extension of the syringomyelic cavity improved significantly after surgery, going from an average of 16.4 vertebrae preoperatively to 13.4 vertebrae postoperatively.
- The anteroposterior extension of the syringomyelic cavity improved considerably after surgery; from an average Vaquero index of 0.76 preoperatively to a Vaquero index of 0.38 postoperatively.

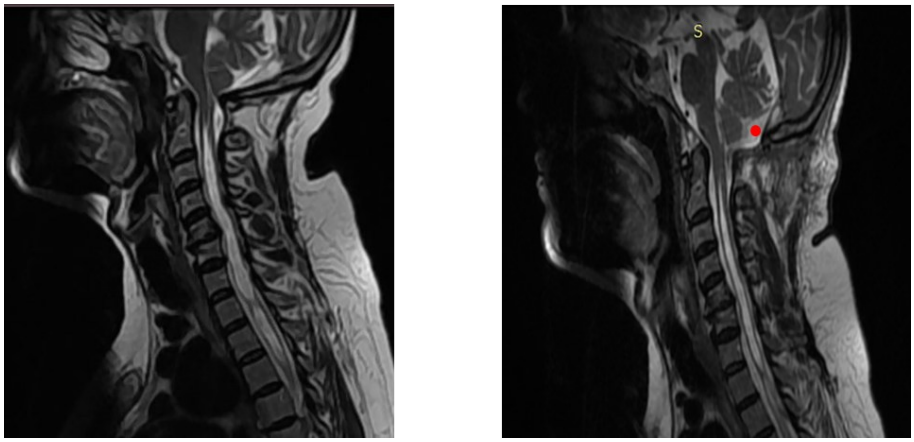

Figure 6. *Image on the left, creation of a neo-large postoperatively (red dot).*

A postoperative MRI was performed in all patients, showing a restoration of CSF flow at the foramen magnum in nine out of ten patients.

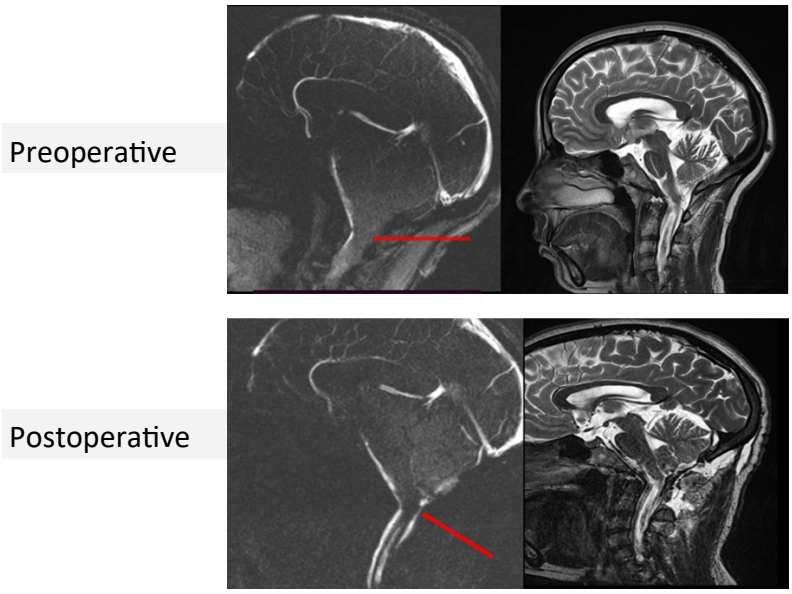

Figure 7. *An MRI cine showing restoration of CSF flow at the foramen magnum (red arrow).*

Discussion

In our series, the rate of Chiari type 0 was 12.5%, in a group of 80 patients with Chiari malformation. Given the rarity of this pathology, there are not many articles in the literature that allow us to define its percentage; in the series by Joshua J. Chern[9], the rate of Chiari type 0 was 3.7%.

These 10 patients were female, ranging in age from 38 to 50 years (average of 44.8 years). In the series by Joshua J. Chern[9] on a pediatric population, 9 were boys, 6 were girls, with an average age of 10.5 years. -The clinical features of our patients were dominated by signs of spinal cord compression due to the syringomyelic cavity, resulting in paresis and thermoalgesic disturbances. Signs due to obstruction of CSF flow at the foramen magnum were secondary, with occipital headaches aggravated by the Valsalva maneuver. The same clinical findings have been reported in the literature.-Cerebrospinal fluid MRIs performed on our patients showed:

The absence of tonsillar ectopia, with the presence of a syringomyelic cavity.

A small posterior cerebral fossa was found in our patients, this was also noted by Joshua J. Chern[9] and Nejat Isik[10]. -a pointed shape of the inferior end of the cerebellar tonsils was observed in our patients, this anatomical peculiarity was described by Tubbs[11] in Chiari type 1 malformations.

Flow MRI was performed in our patients, confirming the obstruction of CSF flow at the occipito-vertebral junction. This MRI was performed in the majority of series, notably in Nejat Isik[10] , Yuan Zhou[12] and Iskandar BJ[2] , except for Joshua J. Chern[9] , where Flow MRI was requested at the beginning of the study, but was often inconclusive postoperatively despite the clinical and radiological improvement of their patients. Therefore, they concluded that the use of flow MRI is no longer useful in these cases. - Each patient underwent posterior cranial fossa decompression with removal of the posterior arch of C-1. The size of the craniectomy was 4 cm in width and height. The same surgical technique was performed in the majority of series, which surgically treat Chiari type 0 malformation in the same manner as Chiari type 1 malformation.

Intradural exploration of the bulbo-medullary junction revealed arachnoid adhesions with a web obstructing the foramen of Magendie. This was noted by Joshua J. Chern[9], suggesting systematic opening of the dura mater in Chiari type 0 malformations, in search of these adhesions and arachnoid web.

All patients underwent duraplasty with autologous pericranium. The majority of authors used the same closure technique.

Clinical signs that improved postoperatively included suboccipital headaches, motor deficit, and cerebellar syndrome. While the clinical signs that did not respond to surgical treatment were thenar and hypothenar amyotrophy and thermoalgesic disturbances, this was noted by Tubbs[11].

We observed in all our patients a regression of the syringomyelic cavity both longitudinally, going from an average of 16.4 vertebrae preoperatively to 13.4 vertebrae postoperatively, and anteroposteriorly, with a Vaquero index on average going from 0.76 preoperatively to 0.38 postoperatively. The same findings were described by Joshua J. Chern[9].

A peculiarity was noted in all our patients, represented by the change in the shape of the lower end of the cerebellar tonsils, which became rounded after surgery; Tubbs[11] noted this. Flow MRI was performed postoperatively in all patients, showing restoration of CSF flow at the foramen magnum in the majority of patients, although flow MRI in Chiari type 0 malformations is not practiced by all authors; indeed, Joshua J. Chern et al[9] used flow MRI early in their studies, but was often inconclusive postoperatively despite clinical and radiological improvement in their patients. Therefore, they concluded that the use of flow MRI is no longer useful in these cases.

Conclusions

Chiari malformation type 0, characterized by syringomyelia and a small posterior fossa without tonsillar herniation, shares clinical and surgical features with type 1. Our findings suggest that a pointed shape of the tonsils and the presence of an arachnoid veil at the foramen of Magendie may contribute to its pathophysiology, warranting systematic intraoperative inspection.

Conflict of Interest

The author declare no conflicts of interest regarding the publication of this case report**.**

Funding

None.

References

1. Tubbs RS, Iskandar BJ, Bartolucci AA, Oakes WJ. A critical analysis of the Chiari 1.5 malformation. *Journal of neurosurgery.* 2004;101(2 Suppl):179-183.
2. Iskandar BJ, Hedlund GL, Grabb PA, Oakes WJ. The resolution of syringohydromyelia without hindbrain herniation after posterior fossa decompression. *Journal of neurosurgery.* 1998;89(2):212-216.
3. Chorobski J, Stępien L. On the Syndrome of Arnold-Chiari: Report of a Case %J Journal of Neurosurgery. 1948;5 (5):495-500.
4. Gardner WJ, Goodall RJ. The surgical treatment of Arnold-Chiari malformation in adults; an explanation of its mechanism and importance of encephalography in diagnosis. *Journal of neurosurgery.* 1950;7(3):199-206.
5. Dockter JL. Sclerotome induction and differentiation. *Current topics in developmental biology.* 2000;48:77-127.
6. Fan CM, Tessier-Lavigne M. Patterning of mammalian somites by surface ectoderm and notochord: evidence for sclerotome induction by a hedgehog homolog. *Cell.* 1994;79(7):1175-1186.
7. Koehler PJ. Chiari's description of cerebellar ectopy (1891). With a summary of Cleland's and Arnold's contributions and some early observations on neural-tube defects. *Journal of neurosurgery.* 1991;75(5):823-826.
8. Greenlee J, Garell PC, Stence N, Menezes AH. Comprehensive approach to Chiari malformation in pediatric patients. *Neurosurgical focus.* 1999;6(6):E6.
9. Chern JJ, Gordon AJ, Mortazavi MM, Tubbs RS, Oakes WJ. Pediatric Chiari malformation Type 0: a 12-year institutional experience. *Journal of neurosurgery Pediatrics.* 2011;8(1):1-5.

10. Isik N, Elmaci I, Kaksi M, Gokben B, Isik N, Celik M. A new entity: Chiari Zero malformation and its surgical method. *Turkish neurosurgery.* 2011;21(2):264-268.

11. Tubbs RS, Beckman J, Naftel RP, et al. Institutional experience with 500 cases of surgically treated pediatric Chiari malformation Type I. *Journal of neurosurgery Pediatrics.* 2011;7(3):248-256.

12. Zhou Y, Wang H, Li N, Lin Y, Zhu L, Cheng H. Chiari 0 malformation with syringomyelia syringobulbia and syrinx cavity in pons. *Interdisciplinary Neurosurgery.* 2016;6:35-37.