CASE SERIES

10.2478/AMB-2026-0046

SURGICAL MANAGEMENT OF CHIARI MALFORMATION TYPE I IN THE PEDIATRIC PATIENTS WITH SYRINGOMYELIA AND RELATED TO KLIPPEL-FEIL SYNDROME: A CASE SERIES AND LITERATURE REVIEW

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Abstract. ***Background:*** *Chiari type I malformation (CM-1) is a birth defect where the back part of the skull does not develop properly, leading to the formation of a small and shallow space at the back of the head. The aim of this case series is to determine the causes and pathologies associated with Chiari type 1 malformation and the possible management and treatment, as well as the best outcomes that aid in the postoperative evolution of CM-1 decompression.* ***Case Presentation:*** *A 13-year-old patient presented to the emergency department with severe, frequent, and daily headaches of five years' duration. An 8-year-old girl presented to our clinic with severe headaches of one year's duration. A 9-year-old boy presented to the clinic with weeks of severe headaches and numbness in the fingers of his right hand. MRI was performed, which diagnosed Arnold-Chiari syndrome type 1, fusion of the C1-C2 vertebrae, and triphalangeal fingers of the hand, which are associated with the Klippel-Feil syndrome (KFS).* ***Conclusion:*** *CM-1 is associated with syringomyelia, which is more prominent in Case 2, whereas Case 3 is associated with Klippel–Feil syndrome; therefore, karyotyping and Sanger sequencing of the GDF6 gene were recommended.*

Key words: *Chiari malformation type 1, posterior fossa decompression, foramen magnum, Klippel-Feil syndrome, syringomegaly*

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Received: *19 August 2025;* ***Revised/Accepted:*** *08 January 2026*

INTRODUCTION

The Chiari type I malformation is a congenital disorder characterized by embryonic underdevelopment of the occipital bone that can result in a pathology of the narrow and shallow posterior cranial fossa. Its accumulation at the posterior level causes the tonsils to descend into the foramen magnum, compressing the anatomical structures. This malformation may manifest with syringomyelia, as well as hydrocephalus, craniosynostosis, scoliosis, or a tethered spinal cord [1]. The CM-1 was described or reported by Hans Chiari in the 1890s. The classification arose from autopsies of children; he identified three distinct deformities: Chiari types 1, 2, and 3. Later, in 1895, a Chiari type 4 phenotype was identified [2]. The Klippel-Feil sequence is often associated with Chiari type 1 and sleep-disordered breathing. Klippel-Feil is a congenital condition that often presents with congenital cervical fusion or reduced cervical flexion and posterior lower hair implantation. Using a polysomnogram two months postoperatively for decompression of Chiari type 1 malformation shows severe sleep apnea, even though the patient is usually asymptomatic [3]. The differential diagnosis is divided into Chiari type 1, non-myelomeningocele, and Chiari type II with isolated myelomeningocele, without a posterior cranial fossa abnormality [4]. The standard treatment for Chiari type I malformation is decompression of the posterior fossa or foramen magnum. There are existing techniques for extradural decompression, such as intra-arachnoid tonsillar resection. The extra-arachnoid approach is more traditional, including suboccipital craniotomy with duroplasty and the use of a dural patch in duroplasty [5].

The aim is to decompress the posterior cranial fossa to reverse the symptoms of the Chiari type 1 malformation, such as headache with paresthesia of the upper limbs, due to compression of the syrinx with the presence of syringomegaly and the symptoms associated with Klippel-Feil syndrome, by decompressive trepanation and laminectomy of the craniovertebral junction.

CASE PRESENTATION

CASE 1

A 13-year-old patient was brought to the emergency room with severe, frequent daily headaches that, according to her family, began 5 years ago. Therefore, it was decided to admit her to our center, the Morozovskaya Children's Hospital, Moscow. She had a past medical history of measles. The physical examination revealed the following measurements: height: 158 cm; body weight: 40 kg; temperature: 36.6 °C; BMI: 16 kg/m²; body surface area: 1.32 m²; Glasgow Coma Scale (GCS) score: 15. An MRI revealed a decrease in the amygdala of more than 6 mm to the foramen magnum. A brain CT scan confirmed the diagnosis of an Arnold-Chiari malformation, type I. Description of surgery: The patient was in the prone position. A linear skin incision along the midline in the occipital region was made, followed by resection of the lower third of the occipital bone squama and the posterior arch of the C1 vertebra. Marked dural pulsation was observed. Therefore, it was decided to limit the bone decompression. Soft tissue and skin sutures were applied. During the surgical procedure, a craniovertebral junction decompression was per-

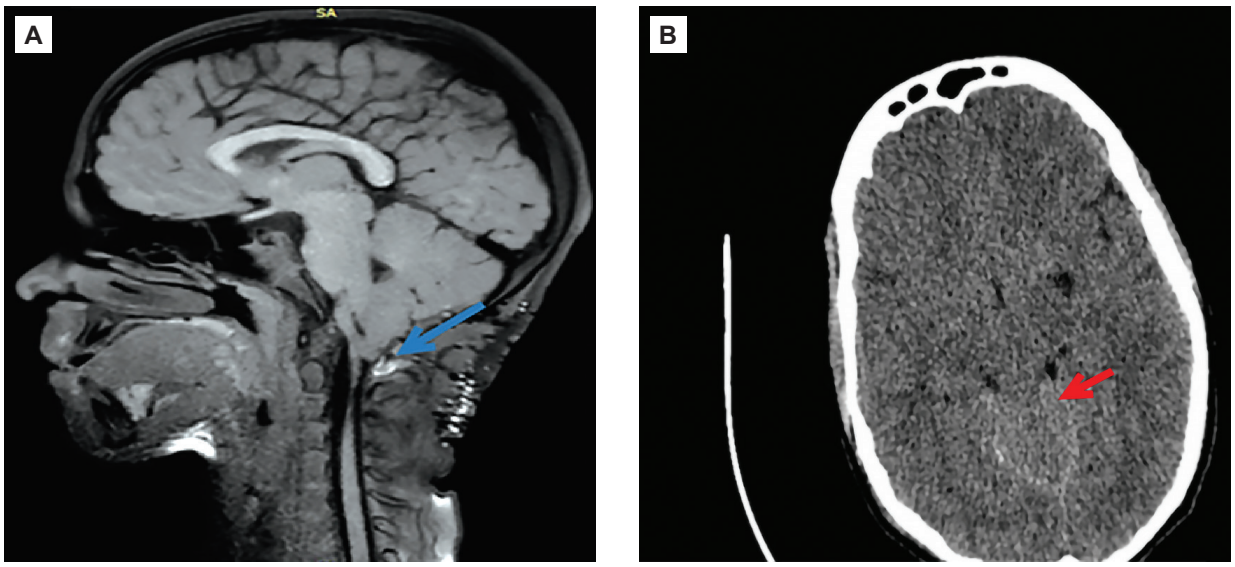

Fig. 1. a) A sagittal T1-weighted MRI image is shown. The tonsils are shown to be drooping. Note the blue arrow. **b)** Axial CT image shows compression of the posterior cranial fossa structures due to the descent of the cerebellar tonsils

formed, with a hypodense lesion at the level of the lateral basal ganglia on the left side. During the postoperative period, conservative treatment was based on 2 g of ceftriaxone IV once a day for 5 days. During follow-up in the ICU, the inpatient treatment phase was completed, and the patient was discharged under the supervision of a pediatrician, neurologist, and ophthalmologist with recommendations. An MRI of the brain and entire spinal cord was performed after 6 months without relevant findings.

CASE 2

An 8-year-old girl came for a consultation with severe headaches that had been occurring for a year. She reported that her headache had been increasing in frequency to 2-3 times per week, was located in the occipital region, was worse in the morning, and was of medium intensity, lasting 15-20 minutes with spontaneous relief at night. She presented no complaints at that moment. During the physical examination, her height was 140 cm, body weight was 25 kg, tempera-

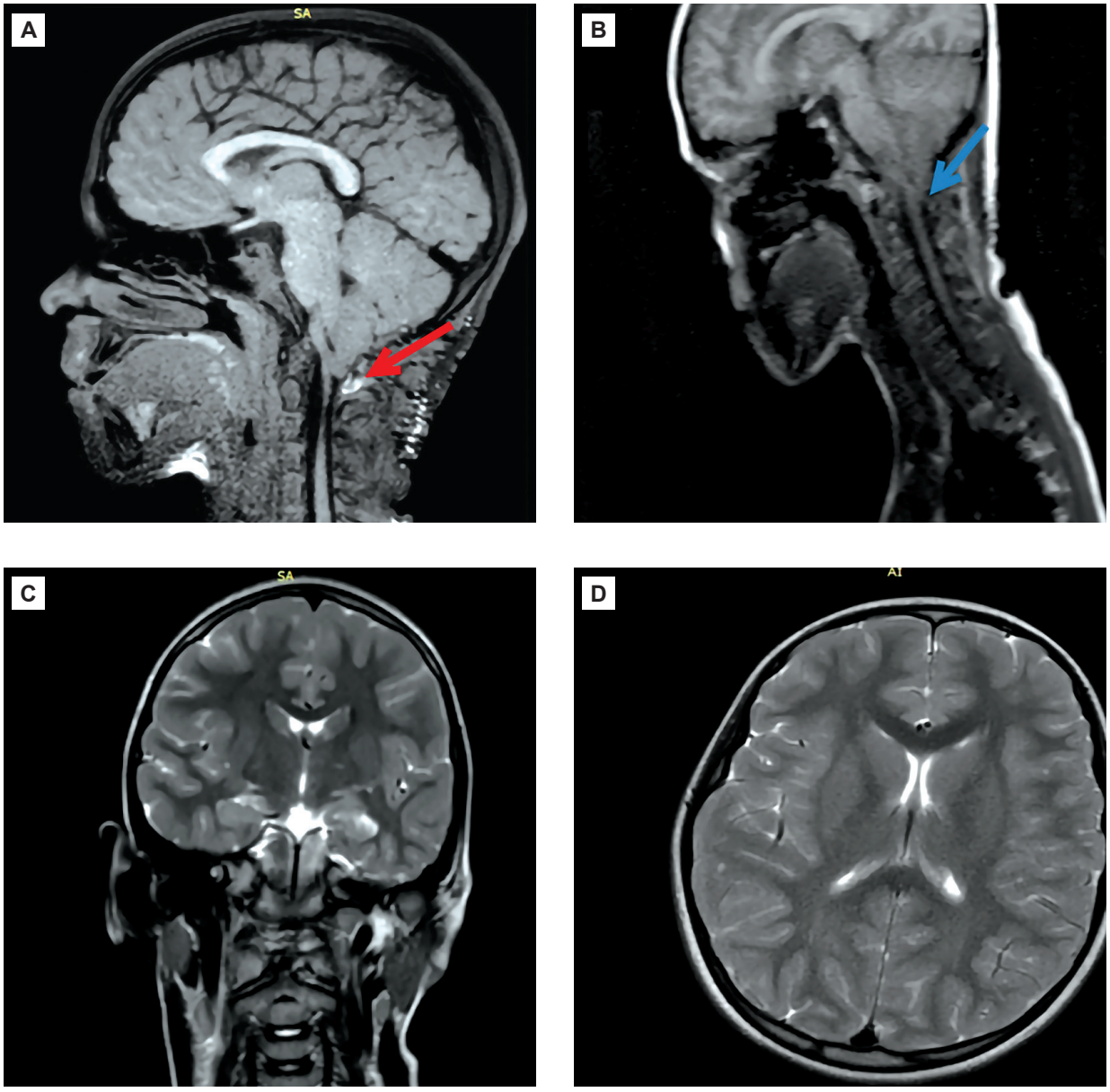

Fig. 2. a) Preoperative sagittal image of a brain and cervical spine MRI: decompression of the craniovertebral junction. A descent of the cerebellar tonsils below the foramen magnum. See the red arrow. **b)** Images of the sub- and supratentorial structures using a series of FLAIR, DWI, SWI, and T1- and T2-weighted brain CT scans. See the blue arrow. **c)** Coronal image. A postoperative defect of the posterior arch of the C1 vertebra and squamous cell occipital bone is observed, as well as soft tissue edema in the occipital region. **d)** The cerebellar tonsils and medulla oblongata prolapse into the foramen magnum to the level of the middle third of the C2 vertebra. The midline structures are not displaced, and the cerebral ventricles are normal in shape and size

ture was 36.6 °C, BMI was 12.8 kg/m², and body surface area was 0.99 m². There were no concomitant symptoms. The initial decision was to prescribe nootropic treatment. An outpatient MRI scan of the brain was performed, which diagnosed decompression of the craniovertebral junction. The magnetic resonance imaging showed a Chiari 1.5 malformation, also known as the bulbar variant of Chiari malformation. Therefore, the decision was made to admit the patient to the Morozovskaya Children's Hospital in Moscow. Three days after preoperative preparation, the surgical procedure of decompression through the craniovertebral joint was performed. Description of the surgery: In the prone position, a linear incision was made in the cervico-occipital region, the occipital bone and first cervical vertebra were skeletonized, and the posterior arch of the first vertebra and the occipital squama were resected with forceps. The dura mater was observed to be non-pulsatile. Under microscopic control, the dura mater was opened, and after opening, a large cistern was visualized at the level of C2, with a clear pulsation. An aponeurosis implant was sutured into the incision, and the patch area was reinforced with Tachocomb.

Because of the seriousness of the surgical intervention, the patient was transferred to the intensive care unit (ICU) several hours later for postoperative recovery and observation. Two days later, another MRI of the brain and cervical spine was performed: images of the sub- and supratentorial structures were obtained through a series of FLAIR, DWI, SWI, T1-, and T2-weighted brain CT scans. A postoperative defect of the posterior arch of the C1 vertebra and the squama of the occipital bone was observed, as well as soft tissue edema in the occipital region. Conservative treatment was recommended 3 days later. During follow-up, the patient's recovery was remarkable, and they were discharged. The MRI scan is shown in Figure 2.

CASE 3

A 9-year-old boy was brought to the clinic with weeks of severe headaches and numbness in the fingers of his right hand. While performing his physical examination, the following measurements were observed: height: 140 cm; body weight: 30 kg; temperature: 36.7 °C; BMI: 15.3 kg/m²; body surface area: 1.08 m². Two days later, an MRI was performed that diagnosed Arnold-Chiari syndrome type 1, fusion of the C1-C2 vertebrae, and triphalangeal fingers of the right hand. Differential diagnosis was made with Klippel-Feil syndrome among the chromosomal abnormalities, as well as with monogenic syndromes, which are characterized by triphalangeal fingers of the hand: preaxial polydactyly type 2 and triphalangeal thumb syndrome with polysyndactyly. We performed a brain computed tomography scan two days after the CT scan. The cerebellar tonsils protruded into the cervicothoracic syringomyelia, so the patient was admitted for surgery. Postoperative gas accumulation, a small amount of cerebrospinal fluid, and fragments of bone density in soft tissues were observed. In the surgical procedure, a trepanation with decompressive laminectomy of the craniovertebral junction was performed, which consisted of follow-up on the third and fourth postoperative days in the intensive care unit (ICU); the patient showed a total improvement and was discharged without complications. The scan results are shown in Figure 3.

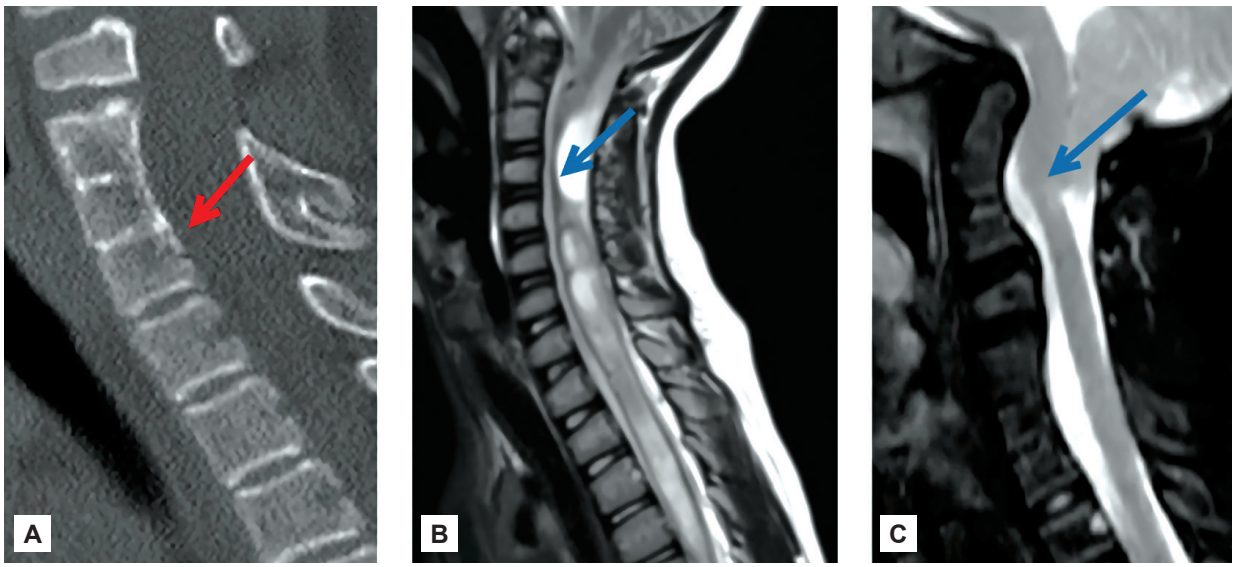

Fig. 3. a) A T1-weighted MRI is shown. **b)** A T2-weighted image shows descent of the cerebellar tonsils, with syringomyelic cavities, syrinx fluid, and associated strong suspicion of Klippel-Feil syndrome. **c)** The herniated cerebellar tonsils show compression of the cervical spinal cord, with syringomyelia

DISCUSSION

Craniovertebral junction decompression

The patients were placed face down on the operating table, with the head rigidly fixed with a Mayfield clamp and tilted forward. The skin was repeatedly treated with an antiseptic solution. A linear incision was made along the midline in the cervico-occipital region, separating the soft tissues. A flap measuring approximately 2 x 1 cm of fascia was excised from the left half of the wound and temporarily removed from the wound. The occipital bone and one or two cervical vertebrae were skeletonized. A variant anatomy of the atlanto-occipital region was observed, with dense adhesions on the first and second cervical vertebrae, as well as an extremely dense connective tissue bridge between the occipital bone and the first cervical vertebra. The axis of the cervical spine formed an acute angle with the occipital squama, making laminectomy difficult. An electric burr was used to create a burr hole in the occipital squama. Subsequently, a suboccipital burr was performed with forceps, visualizing the dura mater. A C1-C2 laminectomy was performed with forceps, revealing relaxation of the brain beneath the dura. The dura was opened with a saber-shaped incision; the brain exhibited moderate pulsation, and dense arachnoid adhesions were observed. The adhesions were dissected, cerebrospinal fluid began to flow, and a firm, pronounced cerebral pulsation was observed.

After a posterior cranial fossa approach for decompression of Arnold-Chiari malformation, the most common or frequent complications involve pseudomeningocele, meningitis, hydrocephalus or cervical instability. MC-1, if not corrected in time, may cause bilateral vocal cord paralysis due to obstruction of the medulla oblongata as well as the cerebellum through the foramen magnum. This pressure or traction on the vagus nerve can be relieved after suboccipital decompression surgery or C1 laminectomy. Therefore, during this procedure, extensive extraction can cause cranial nerve injuries [8].

Hemohydrodynamic interactions in Chiari syndrome due to venous drainage and regulation of intracranial volume changes

The alternative drainage pathways, such as the cerebral venous collection cistern factors related to cerebral arterial blood flow; if it increases, the venous drainage pathways will sync. This venous factor will also have a negative correlation with cervical CSF volume in patients with Chiari syndrome and, therefore, with the syrinx. This correlation will not be present in patients without a syringe. Therefore, there will be a partial decrease in CSF. Cervical volume will also not be compatible with CSF and conventional volume. Therefore, parameters should be used to compensate for the lack of CSF in the compliance function in the venous factor, allowing its capacity to increase in intracranial vascular volume changes during the same systole [9].

Surgical Innovations in Chiari Malformation Treatment

MISS spinal techniques treat spinal cord injuries and related lesions. Once the injury is reduced, the muscles, bones, and tissues can lead to immediate recovery with reduced postoperative pain and, therefore, shorter hospital stays compared to open surgery [10, 11, 12]. Several studies have supported the potential of these techniques for Chiari type 1 malformations. One of the current trends is the endoscopic technique for the management of Chiari type 1 malformations [13]. This approach also helps reduce postoperative pain. Also, the use of an exoscope as an alternative to a microscope allows for superior and unrestricted visualization of images [14]. This will be helpful for the surgeon, especially in procedures involving the occipital region [15].

Chiari type 1 malformation may be pathophysiologically associated with abnormalities of the craniocervical junction, both cranial and spinal cord anchoring, as well as intraspinal hypotension, cranial construction, and intracranial hypertension, whether or not accompanied by hydrocephalus [6, 14]. However, patients with Klippel-Feil syndrome often present with atlantoaxial instability; a cervical spinal cord injury may also develop. In these patients, hypermobility may present with spinal cord injury but not with neurological signs. It may also be associated with juvenile rheumatoid arthritis and fibrodysplasia ossificans progressiva, as well as ankylosing spondylitis [7, 15].

CONCLUSION

This study is a case series of Chiari malformations. As with all clinical manifestations, the main symptom in all three cases was headache, which persisted for months or years. Case 1: During the surgical procedure, a decompression of the craniovertebral junction was performed, revealing a hypodense lesion in the left lateral basal ganglia. Postoperatively, conservative treatment consisted of 2 g of ceftriaxone IV once daily for 5 days. Theoretically, due to the compression of anatomical structures in the posterior cranial fossa and the hypodense lesions of the left lateral basal ganglia, which could also be attributed to the headaches and, above all, to inflammation of these structures, administering a blood-brain barrier protectant during those days yielded good postoperative

results. The patients presented with syringomyelia, especially case 2, which is a manifestation of Chiari type 1 syndrome. A postoperative defect of the posterior arch of the C1 vertebra and the squamous part of the occipital bone was observed, as well as soft tissue edema in the occipital region. Conservative treatment was recommended 3 days later. Case 3, on the other hand, is associated with Klippel-Feil syndrome. Therefore, postoperative gas accumulation, a small amount of cerebrospinal fluid, and bone density fragments in soft tissues were observed. Postoperative treatment and follow-up in the intensive care unit were necessary; the blood-brain barrier recovered during conservative treatment. Karyotyping for GDF6 gene analysis by Sanger sequencing was recommended. For more reliable confirmation of this syndrome, despite its phenotypic visibility, Chiari malformation type 1 and its variants present characteristic clinical features, and posterior cranial fossa decompression remains the preferred treatment option. This helps relieve headaches and, in some cases, intracranial pressure.

Conflicts of Interest: *There is no conflict of interest*

Ethical approval statement: *The ethical guidelines outlined in Good Clinical Practice in the Declaration of Helsinki of the World Medical Association were strictly followed, ensuring that each patient provided informed consent before participating in the research.*

The study was carried out according to the latest revision of the Helsinki Declaration regarding medical research involving human subjects. Morozoskaya Children's City Clinical Hospital, Moscow, Russia. No. Reference L035'00115-77/00096790, 103, February 2, 2015.

Use of artificial intelligence: *No generative AI or AI-assisted technologies were used during the writing process of this manuscript.*

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