ARTÍCULO ORIGINAL

Gac Méd Caracas 2023;131(4):829-841
DOI: 10.47307/GMC.2023.131.4.6

Differentiated diagnostic and surgical algorithm for isolated and combined Chiari malformation Type I

Diagnóstico diferencial y algoritmo quirúrgico para malformación de Chiari Tipo I aislada y combinada

Buranbek Diusheev[1*], Akylbek Akmataliev[2], Chyngyz Turdugulov[3], Nurlan Kachiev[4]

SUMMARY

Introduction: *Arnold-Chiari malformation (ACM) type I is a craniovertebral junction pathology with ectopy of the cerebellar tonsils through the great occipital foramen with compression of the brain stem and upper cervical spinal cord. Until now, there have been more than 20 types of surgical interventions for the Arnold-Chiari malformation type.* **Objective:** *The main purpose of this research is the decompression of craniovertebral stem structures and occipital foramen, and the search for effective methods to repair cerebrospinal fluid (CSF) circulation.* **Methods:** *The complex examination and surgical treatment at the Neurosurgery Department No. 1 of the National Hospital of the Ministry of Health of the Kyrgyz Republic (Bishkek, Kyrgyzstan) passed 201 patients (aged 16-74) with symptomatic ACM I from 2008 to 2020.* **Results:** *Regression of preoperative clinical symptomatology was detected in 117 cases (58 %), significant reduction of hypertension and vestibulo-ataxic disturbances in 76 cases (38 %), and stabilization of the process with preservation of focal damages in 8 cases (4 %). In 10 (5 %) cases, there was syringomyelia progression, for which syringosubarachnoid shunting was carried out in the second stage. In the presence of a tight filum terminale syndrome detected in 4 (2 %) patients, the first step was excision of the filum terminale followed by regression of clinical manifestations. The most effective results of surgical interventions were obtained when the pathogenetic approach was followed, in particular, the cerebrospinal fluid flow repair during craniocervical decompression; a regression of both the underlying disease and concomitant pathologies was achieved.*

Keywords: *Cerebrospinal fluid, syringomyelia, hydrocephalus, craniocervical decompression, craniovertebral junction.*

DOI: https://doi.org/10.47307/GMC.2023.131.4.6

ORCID: 0000-0001-7417-9419[1]
ORCID: 0000-0002-7337-2601[2]
ORCID: 0009-0008-1704-3660[3]
ORCID: 0000-0003-4117-478X[4]

Department of Neurosurgery. National Hospital. 720040, 1 Togolok Moldo Str., Bishkek, Kyrgyzstan

*Corresponding autor: Buranbek Diusheev. E-mail: buranbekdiusheev@yahoo.com

Recibido: 21 de julio 2023
Aceptado: 1 de septiembre 2023

RESUMEN

Introducción*: La malformación de Arnold-Chiari tipo I es una patología de la unión craneovertebral con ectopía de las amígdalas cerebelosas a través del gran agujero occipital con compresión del tronco encefálico y la médula espinal cervical superior. Hasta el momento, existen más de 20 tipos de intervenciones quirúrgicas en malformaciones tipo Arnold-Chiari.* **Objetivo:** *El objetivo principal de esta investigación es la descompresión de las estructuras del tallo*

craneovertebral y el agujero occipital, y la búsqueda de métodos efectivos para reparar la circulación del líquido cefalorraquídeo (LCR). **Métodos:** *El examen complejo y el tratamiento quirúrgico en el Departamento de Neurocirugía No. 1 del Hospital Nacional del Ministerio de Salud de la República Kirguisa (Bishkek, Kirguistán) pasaron 201 pacientes (de 16 a 74 años) con ACM I sintomático de 2008 a 2020.* **Resultados:** *Se detectó regresión de la sintomatología clínica preoperatoria en 117 casos (58 %), reducción significativa de la hipertensión y alteraciones vestibuloatáxicas en 76 casos (38 %), estabilización del proceso con preservación de daños focales en 8 casos (4 %). En 10 (5 %) casos hubo progresión de la siringomielia, por lo que se realizó derivación siringosubaracnoidea en la segunda etapa. Ante la presencia de un síndrome de filum terminale apretado detectado en 4 (2 %) pacientes, el primer paso fue la escisión del filum terminale seguido de la regresión de las manifestaciones clínicas. Los resultados más efectivos de las intervenciones quirúrgicas se obtuvieron cuando se siguió el enfoque patogénico, en particular, la reparación del flujo de líquido cefalorraquídeo durante la descompresión craneocervical; se logró una regresión tanto de la enfermedad de base como de las patologías concomitantes.*

Palabrasclave*:Líquidocefalorraquídeo,siringomielia, hidrocefalia, descompresión craneocervical, Unión craneovertebral.*

INTRODUCTION

Arnold-Chiari malformation (ACM) type I stands as a distinctive and intricate craniovertebral junction (CVJ) pathology that engenders a significant impact on neurological health (1). This malformation manifests through the displacement of the cerebellar tonsils, vital components of the brain responsible for balance and coordination, downward through the great occipital foramen (2). As researchers continue to delve into the nuances of ACM type I, insights into the underlying pathophysiology and the dynamic relationship between the brain, spinal cord, and surrounding structures are continually being refined, paving the way for improved diagnostic techniques, treatment strategies, and ultimately, enhanced patient outcomes (3,4).

The history of the study of ACM morphology and its corresponding symptomatology is well known (5,6). Meanwhile, this malformation aetiology and pathogenesis remain controversial; however, all discussions are based on the disembryogenetic theory (7,8). Neuroimaging advanced techniques have dramatically increased the number of patients with this pathology in neurosurgical institutions. Pre- and postoperative brain magnetic-resonance (MRI) capabilities using different modules with the assessment of cerebrospinal fluid (CSF) circulation and hemodynamics have significantly expanded the range of surgical interventions in ACM (9).

The foundation for ACM development is rooted in disturbances during fetal cerebellar formation. Genetic factors play a pivotal role in embryogenesis, particularly genes associated with paraxial mesoderm differentiation, such as *GDF7*, *GDF3*, *ALDH1A2*, *FLT1*, and *CDX1* (10). The inadequate development of the occipital bone derived from the paraxial mesoderm leads to the improper formation of the small posterior cranial fossa, contributing to ACM. However, the precise genetic underpinnings remain to be fully elucidated, likely involving multiple gene mutations working in a complex interplay (11). Embryonic asphyxia and respiratory infections further contribute as triggers for malformation. The pathophysiology involves several mechanisms, including elevated intracranial pressure, ventricular fluid accumulation, skull constriction and hindbrain herniation, spinal canal pressure reduction, spinal cord compression, and abnormal atlantoaxial joint mobility, often linked to connective tissue anomalies.

Surgical interventions for ACM I primarily aim to decompress craniovertebral structures and restore CSF circulation. The decompression of the great occipital foramen, often combined with duraplasty, is a common approach to reestablish cerebrospinal fluid flow through the foramen magnum (12). Although the first decompression surgery dates back to 1930 by Dutch surgeon Graftdijk (1), the diverse morphological variations in the CVJ region result in a polymorphic array of clinical manifestations, especially in conjunction with conditions like syringomyelia, hydrocephalus, and fixed terminal filum (13). Consequently, a standardized algorithm for surgical intervention methods in ACM I remains elusive. Despite the proposal of over 20 surgical

approaches, a consensus is lacking regarding the optimal timing, sequencing, and evaluation criteria for multistage interventions (14).

This paper aims to identify and establish the most effective set of criteria for choosing appropriate diagnostic and treatment methods. The primary objectives of this study revolve around overcoming key challenges to achieve its overarching purpose. These objectives encompass the comprehensive collection and analysis of patient data, both in isolated cases of ACM I and when coexisting with other anomalies. Additionally, the study assessed patients' conditions before surgical interventions, monitor their statuses following these procedures, and meaningfully interpret the outcomes.

MATERIALS AND METHODS

The results of the complex examination and surgical treatment of 201 patients with symptomatic ACM I treated at the Neurosurgery Department No. 1 of the National Hospital of the Ministry of Health of the Kyrgyz Republic (Bishkek, Kyrgyzstan) from 2008 to 2020, aged 16-74 years were analysed (Figure 1).

All patients had magnetic resonance (MR) imaging of the brain and CVJ, sometimes to the lumbosacral region, to eliminate syringomyelia and tethering syndrome associated with ACM I. The degree of cerebellar ectopia into the subarachnoid space of the spinal cord was

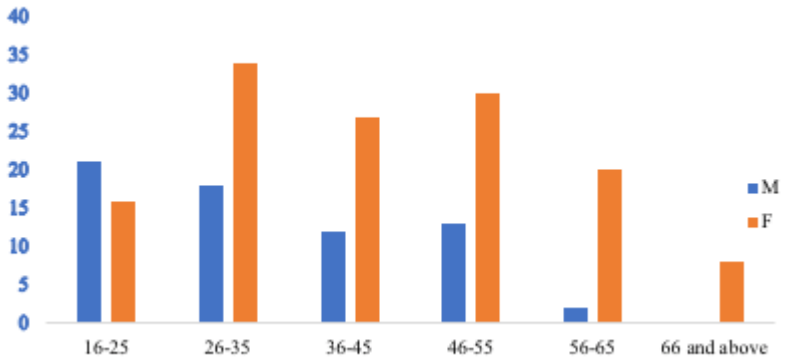

Figure 1. Distribution by gender and age of patients with symptomatic ACM I.

determined by the Chamberlain line (the line connecting the edge of the hard palate and opisthion) and the eligibility criteria for patients was a descent of more than 5 mm. In the last 3 years, phase-contrast cardiac synchronized MR trials to study CSF circulation (studied in 20 patients) and MR angiography were performed. To clarify bone changes, the patients underwent computed tomography with topometry.

All patients in this group took the examination algorithm (Figure 2).

Upon completion of the algorithm for examination of patients with ACM I, a detailed diagnosis of ACM I was made, which determined the tactics of surgical treatment of the patient. Excel spreadsheets were used to process the data. The work was performed in compliance with all ethical standards and under conditions of non-disclosure. Before the start, the patients were informed about the goals of the work, its plan, and the manipulations foreseen for the performance. All methods have not been introduced for the first time but were used earlier in other institutions.

DIFFERENTIATED DIAGNOSTIC AND SURGICAL ALGORITHM

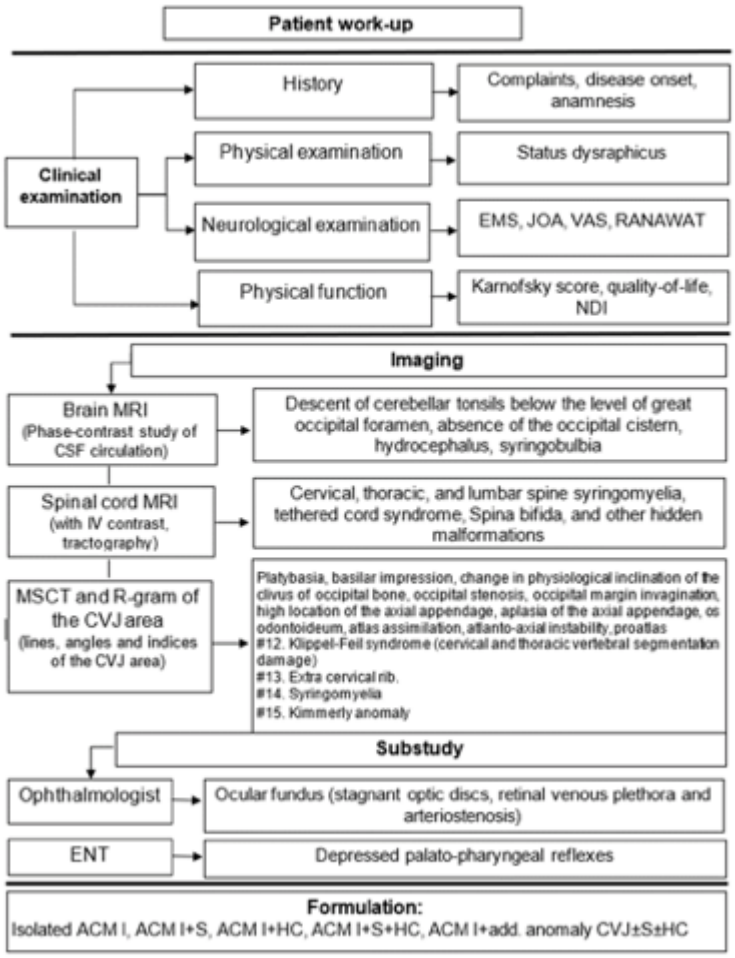

Figure 2. Algorithm of complex examination of a patient with ACM type I
Note: HC – hydrocephalus, S – syringomyelia). ACM I – Arnold-Chiari I type, S – syringomyelia, HC – hydrocephalus, CVJ – Cranio-vertebral junction.

RESULTS

During the objective examination, in more than half of the patients – 113 (56 %), different signs of dysraphic status were found: kyphoscoliosis, gothic palate, different position of auricles and shoulder blades, short neck, keeled and funnel chest, flatfoot, facial asymmetry. Comorbid bone structure anomalies of the craniovertebral area were found in this group of patients (Table 1).

Status dysraphicus and common comorbid bone anomaly in the craniovertebral region are

Table 1

The incidence of associated ACM I craniovertebral junction anomalies

Bone anomaly of the cranio-vertebral region Anomaly	Quantity	%
Platybasia (flattening of the skull base)	18	8.9
Basilar impression	3	1.5
Elevation of the odontoid process	1	0.5
Kimmerly anomaly	8	4

quite common in the ACM I group. No correlation was found between the cerebellar tonsil ectopia below the Chamberlain line on brain sagittal MR imaging and the above-mentioned changes. In almost all patients, the disease manifested with pain in the cervico-occipital region, intensifying with coughing, with progression, accompanied by nausea and vomiting. Then vestibulo-attachment disorders with obnubilations associated with congestive changes on the ocular fundus joined. As the disease progressed, bulbar disorders with dysarthria, dysphagia, and dysphonia were defined. Extreme manifestations of motor disorders were di-tetraparesis with sensory impairment, more often dissociated. The highly variable clinical symptoms in the patients were summarized in Table 2.

Table 2

ACM I clinical manifestations in a group of patients. Distinguished syndromically by symptoms

Syndromes	Manifestations
Hypertension syndrome – 193 (96%)	Pain in the cervico-occipital region, intensifying with coughing, diffuse bursting headaches, nausea, vomiting; photo and noise phobia at the headache maximum
Vestibulo-ataxic (cerebellar) – 169 (84%)	Dizziness, shaky walk
Focal syndromes: 1) Movement abnormalities – 48 (24%)	Paresis, changes in reflexes.
2) Sensory abnormalities – 32 (16%)	Impaired sensibility.
3) Pelvic abnormalities – 12 (6%)	Disorder of diuresis and defecation,
4) Bulbar abnormalities – 16 (8%)	Dysarthria, dysphagia, dysphonia.

The dependence of ACM I clinical manifestations on cerebellar tonsil ectopia through the great occipital foramen is shown in Figure 3.

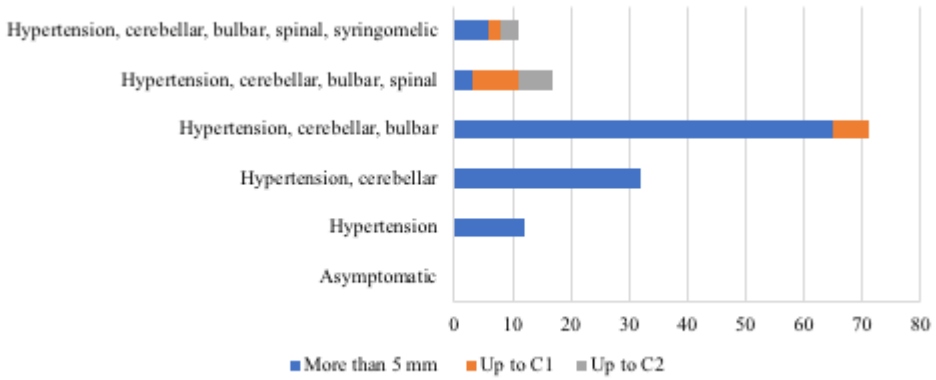

Figure 3. ACM I clinical manifestations depend on the degree of cerebellar amygdala prolapse below the level of the great occipital foramen.

All surgeries were performed in the Concorde-type supine position, with conservative resection of the posterior margin of the greater occipital foramen (3×3 cm) and laminectomy of the C1 vertebra, or otherwise craniocervical decompression (CCD) without duroplasty. The scope of surgical intervention in the clinic has undergone a certain development with experience. Initial CCD without duroplasty is performed extremely rarely, the area of decompression of the occipital bone is markedly reduced. The optimal size is considered to be 3×3 cm, to prevent prolapse of the cerebellar tissue into the CVJ area and prevent craniocervical instability. It should be noted that isolated CCD without duroplasty often leads to dynamic changes directly in the subarachnoid space up to the elimination of the cerebellar tonsils' ectopy with their spontaneous "return" to the vacated cisterna cerebellomodularis. However, the above-described scope of surgical intervention was often insufficient (Figure 4).

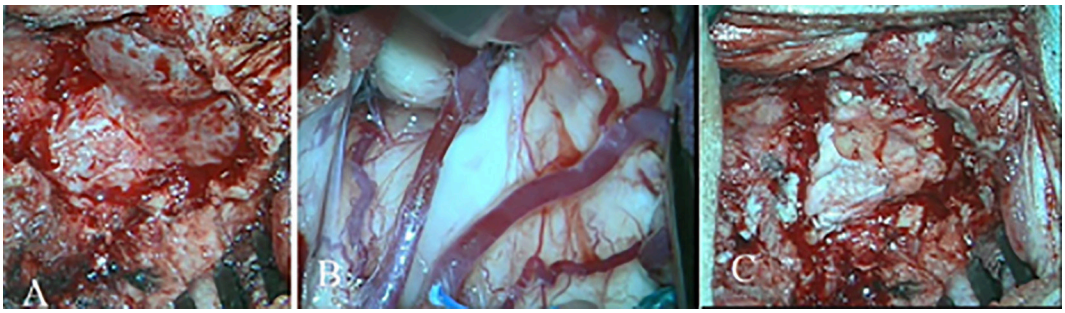

Figure 4. The main stages of surgery.
Note: A – Bone decompression with conservative resection of the occipital bone and C1 vertebral arch. B – Revision of the posterior cranial fossa with the main anatomical structures' visualization, adhesiolysis, and repair of CSF flow; C – Dura mater plastic repair by autograft from the fasciae latae.

After the duramater incision, the arachnoid membrane was examined. In the absence of visible changes in the arachnoid membrane and visualization of the structures involved, the surgery was completed by dura mater plasty, in 70 cases (35 %). Thus, the further scope of surgical intervention changes accordingly. In the case of severe adhesions, the arachnoid meninges were dissected and meningoradiculomyelolysis was performed throughout with the repair of CSF flow. Magendie's foramen was inspected for patency in 109 cases (54 %). If there was a strangulation sulcus on the descended cerebellar tonsils below the C1 vertebral arch, their subpial resection with enlargement of Magendie's foramen was performed. Then the surgery was also completed with dura mater plasty of the fasciae latae. The edges of the dissected arachnoid meninges were sutured to the dura mater to prevent scarring and adhesion. Most often, the duramater was sutured after plasty to tighten it to the surrounding bone structures to prevent adhesions and early formation of the cisterna cerebellomodularis. No stents in Magendie's foramen or artificial sheaths were used. Relatively small accesses, up to 6-7 cm in length, and careful layer-by-layer suturing of muscles and subcutaneous fat made it possible to achieve adequate sealing and prevent liquorrhea. On the day following the intervention, a lumbar puncture was performed to sanitize the cerebrospinal fluid (CSF).

Afterward, the patients had control MRI examinations in 3-6 months. In the presence of superior cervical syringomyelia with severe neurological symptoms, the above surgery was supplemented with syringostomy at one time. Follow-up studies were conducted from 6 months to 12 years. According to the obtained data, complete regression of preoperative clinical symptomatology was detected in 117 cases (58 %), significant reduction of hypertension and vestibulo-ataxic disturbances in 76 cases (38 %), stabilization of the process with preservation of focal disturbances in 8 cases (4 %). There was no postoperative mortality (Figure 5).

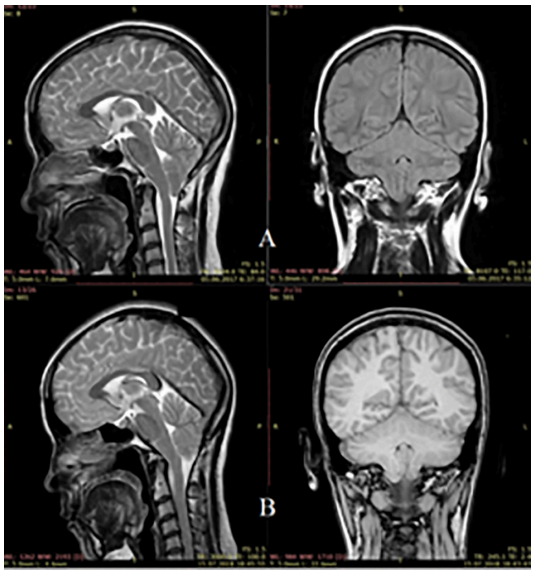

Figure 5. Brain MRI (T1-2 mode in the sagittal and frontal planes) of a patient with isolated Arnold-Chiari malformation type I before and after CCD with revision of the posterior cranial fossa and duroplasty.
Note: A – The descent of the cerebellar tonsils below the level of the foramen magnum by 11 mm; B – Formation of the occipital cistern, liquor flow restored.

When ACM I was combined with spinal cord syringomyelia, 80 cases (40 %), assumed to be the first stage of CVJ surgery, it resulted in a spontaneous reduction of syringomyelia severity, and the second stage of surgery, syringostomy obviated in 9 (4.5 %) patients (Figure 6).

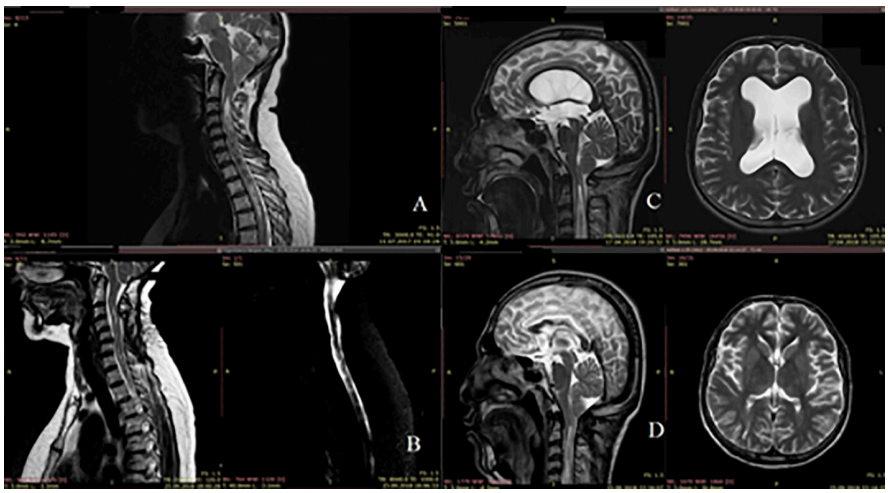

Figure 6. Brain and cervical spine MRI, T2 mode.
Note: A – Expansion of the central canal of the spinal cord and descent of the tonsils of the cerebellum by 8 mm before surgery: B – the result of syringomyelitic cyst regression after craniocervical decompression, the result after 6 months; C – Total expansion of the ventricles of the brain due to a block at the level of the foramen of Mogendie before surgery; D – positive dynamics in the form of syringomyelitic cyst reduction after ventriculo-peritoneal shunting with concomitant hydrocephalus, the result after 3 months.

A total of 59 (29.4 %) patients had reduced syringomyelia in ACM I+S. In 10 cases (5 %), progression of syringomyelia with increasing motor, sensory, and pelvic disorders was noted in the control MRI study from 1 to 3 months, for which syringo-subarachnoid shunting was performed as the second stage, provided there was no block at the Majandi level (based on the phase-contrast MRI study). In these patients, regression of syringomyelitic cysts with the repair of neurological functions was noted in all of them at 3 months of follow-up.

The next associated ACM I pathology was hydrocephalus in 10 cases (5 %), which spontaneously regressed in half of the patients. Five patients underwent VP shunting (Figure 6), and all patients achieved hydrocephalus regression. In the presence of a tight filum terminale syndrome detected in 4 patients (2 %), the first step was excision of the filum terminale followed by regression of clinical manifestations. A number of patients had complications such as lycvoria in 3 cases (1.5 %), and in 2 patients (1 %) secondary bacterial meningitis developed in the early postoperative period. These complications were eliminated during an inpatient period.

Overall, the impact of comorbidities such as hypertension, diabetes mellitus, and other chronic diseases on the course of Arnold-Chiari malformation type 1 (ACM I) and treatment outcomes represents a significant area of research. Chronic conditions like hypertension and diabetes can impact overall health and potentially complicate treatment and recovery processes. Hypertension, for instance, may exacerbate symptoms related to cerebrovascular function and increase the risk of complications during surgery. Diabetes can affect wound healing, immune responses, and overall recovery after surgical interventions. Moreover, these comorbidities might influence the response to surgical treatments, as well as the overall effectiveness of decompression and symptom alleviation.

Also, it is important to note that thorough rehabilitation after surgical interventions for Arnold-Chiari malformation type I (ACM I) is essential to enhance the effectiveness of long-term treatment results. Postoperative recovery entails physical therapy to improve mobility, muscle strength, and posture, addressing discomfort and muscle atrophy. Neurological rehabilitation targets deficits in coordination, sensation, and muscle function, aiding neuromuscular re-education and functional independence. Pain management techniques alleviate immediate discomfort while contributing to long-term pain reduction. Addressing potential cerebrospinal fluid-related complications and hydrocephalus management through education and close monitoring is integral. Additionally, psychological support and coping strategies enhance patients' psychosocial well-being, collectively ensuring that rehabilitation plays a pivotal role in minimizing complications, promoting recovery, and elevating the overall quality of life for ACM I patients.

DISCUSSION

Despite a better understanding of the pathogenetic mechanisms, significant improvement in diagnostic methods, a large number of clinical studies performed, and accumulated experience, the management of patients with Chiari malformation type I remains unresolved. The main works carried out in different countries recognize the inexpediency of surgical treatment in asymptomatic ACM I (15). Patients with brainstem and cranial nerve dysfunction, as well as hydro syringomyelia associated with ACM I, require surgical treatment (16). The wide range of surgical interventions in ACM I demonstrates the versatility of morphological changes accompanying this pathology. The previously prevailing CCD with duroplasty still is the basic surgical approach, which is applicable in most patients with ACM I and is performed by most neurosurgeons (17,18). The findings of these papers agree with the findings of this article regarding the importance of surgical intervention in specific cases of CM1. However, the author's study focuses on comorbid craniovertebral anomalies and details postoperative outcomes, while the second study recognizes the global consensus on surgical strategies and the popularity of MCC.

Preoperative detailing of morphological changes in ACM I determines the basic strategy in planning surgery and the tactics of intervention are determined directly during surgery (19). The

changes detected during surgery in the subdural and subarachnoid spaces of the CVJ determine its final volume. Many authors suggest not to dissect the arachnoid membrane if there are no obstacles at the level of Magendie's foramen, being sure of good CSF permeability for the prevention of subsequent postoperative adhesions (20). Most authors recommend duroplasty to form a cisterna cerebellomedullaris with artificial sheaths, which, in their opinion, simplifies the surgery and is a reliable way to prevent scarring and adhesions. For this purpose, duroplasty with autotissue from the fasciae latae was applied, fixing its dome to the periosteum of the occipital bone (21). Previous studies and the current study agree on the symptoms of Chiari malformation type 1, such as hypertension, movement disorders, and cranial nerve damage, with the severity of symptoms correlating with cerebellar amygdala prolapse. However, the papers differ in the description of postoperative outcomes, treatment of complications, and surgical techniques.

The latter measure with a proper microsurgical technique without bleeding allows satisfactory clinical results. In addition, the use of autologous tissue reduces the risk of liquorrhea due to better edge sealing during suturing and early healing. The use of a proper microsurgical technique indicates that the surgery is being performed with high magnification and precision using specialized instruments. Microsurgery allows the surgeon to work on delicate structures with minimal tissue damage, reducing the risk of bleeding. This is crucial in surgeries involving sensitive areas like the brain or spinal cord. The choice of this technique is also justified by the fact that it leads to satisfactory clinical outcomes. This means that the technique has proven to be effective in achieving the desired results for patients.

The connection between ACM I and syringomyelia is well-known. However, the treatment strategy for syringomyelia in these patients also remains controversial (22). Many neurosurgeons prefer a pathogenetic approach, suggesting that the repair of CSF flow eliminates CSF dynamic abnormalities in the CVJ and leads to syringomyelia regression. This principle causes the disappearance or reduction of syringomyelia up to 90 % (23). In the absence of results from the first stage of intervention and the rapid progression of syringomyelia, many authors recommend syringo-subarachnoid shunting. The results obtained in the present study confirm the feasibility of this algorithm (24). However, there is also the opposite tactic of patient management, when syringostomy and even syringo-peritoneal shunting is the first-line therapy in the case of extensive and large syringomyelic cavities. This may be considered a less rational strategy, and it could be used only in exceptional cases of the "avalanche" phenomenon of spinal and bulbar disorders (25). All the sources, including the author's work, underline the complex challenges in managing ACM I patients and highlight ongoing debates within the medical community regarding optimal treatment pathways.

The work of Liang et al., also proved the effectiveness of surgical correction with the inclusion of duroplasty in the case of Arnold-Chiari type I malformation (26). During surgery, compression of the tonsils, cerebellar hemisphere, and bone tissue was detected in most patients. Decompression of the large occipital foramen and craniocervical department was performed in parallel with the removal of the atlanto-occipital membrane. At the same time, patients underwent a resection of the arachnoid membrane of the cerebellar-medullary fissure. Afterward, an artificial dura mater implant was installed. The last two manipulations were introduced to expand the posterior fossa without resection of the cerebellar tonsils and shunting. The operation was performed for 21 patients with subsequent monitoring of their condition for 6-36 months after the intervention. In 15 cases, syringomyelia was recorded, but in all these cases there was a significant relief of the symptom. Among the complications, osteocompression of the cerebellar hemisphere was observed in 12 patients; in 18 cases – the appearance of a thickened tissue ring, which was fixed between the cerebellar hemisphere and the arachnoid membrane. No fatalities have been recorded. Thus, artificial duroplasty in this case became an alternative to shunting. Comparatively, both studies underscore the complexity of ACM I and the diverse symptomatology associated with the condition. They highlight the importance of tailored surgical approaches to address various manifestations.

Marković et al. described a case of atypical Arnold-Chiari type I malformation without syringomyelia (27). The patient suffered from movement coordination disorder for 6 years. During the last 7 months, the fettle has worsened and mild quadriaparesis has been added to the symptomatology with a dominant mobility disorder of the lower limbs. The damage to the cranial nerves has been diagnosed immediately before the operation. During the surgical intervention, resection of the posterior arch of the first cervical vertebra and C2 laminectomy, microsurgical displacement of the tonsils, and an artificial dura mater implant were performed. The result of the intervention was a significant improvement in the patient's condition. In children, only 15 % of Arnold-Chiari type I malformation manifests itself as symptoms. In such cases, surgical intervention is recommended for dysfunction of cranial nerves and nerves of the brain stem, loss of sensitivity, syringomyelia, and occipital pain, provided with radiological detection of malformation. Targeted surgical intervention, as in the case of adult patients, also includes decompression by removing the occipital bone, the posterior margin of the foramen magnum, and a part of the posterior arch of C1. In specific cases, it is possible to remove part of the cervical vertebrae, and cerebellar tonsils, and use Dural plastic surgery. Usually, the operation is completed within 2-3 hours. The rare complications after surgery are represented by spasms of the neck muscles, which are eliminated by muscle relaxants; development of infection, and liquorrhea (28-30). Although these studies focus on ACM I and surgical interventions, they differ in patient populations, symptom correlations, outcomes, and focus. Together, these studies make valuable contributions to understanding the complexities and nuances of treating ACM I given the different patient profiles and manifestations.

While surgical resection remains the primary treatment for solid tumor malignancies, it is well established that surgery alone is often inadequate for achieving long-term cures. In cancers such as breast, colon, and lung, relapse rates remain high even after apparently complete surgical removal of the primary tumor (31,32). The high risk of recurrence is attributable to micrometastases and residual localized disease not eliminated by surgery. Therefore, careful post-surgical monitoring and consideration of adjuvant therapies are critical for preventing progression. Administration of systemic chemotherapy or radiation as adjuvant therapy plays a key role in eradicating microscopic residual disease and improving outcomes. Additionally, there is increasing recognition that lifestyle factors including diet, physical activity, smoking cessation, and stress reduction can favorably impact cancer recurrence risks (33-35). Such lifestyle changes may act by inhibiting microscopic tumor growth, reducing inflammation, and improving overall health.

Detecting relapse at the earliest possible point is also crucial, as a disease that is caught early is more likely to be responsive to salvage therapy. Intensive radiographic and clinical follow-up in the post-surgical setting allows prompt identification of progression while therapeutic options are still viable. Though surveillance protocols vary by cancer type, regular cross-sectional imaging and biomarker testing generally facilitate the earliest possible diagnosis of recurrent disease. Management of surgically-treated malignancies does not end with the operation itself. Only through diligent long-term monitoring, selective administration of adjuvant therapy, promotion of lifestyle changes, and a multidisciplinary commitment can the risk of relapse be minimized.

Surgical intervention in ACM is associated with a wide range of intraoperative and postoperative complications. Adherence to anatomical access with cutting along the white line of the muscular complex of the cervico-occipital area, delicate periosteal dissection of the CVJ bone structures with careful haemostasis, and the same delicate suturing thereafter allow for avoiding traditional postoperative complications (36,37). A microsurgical technique with subdural and subarachnoid "dry" space is a condition for the prevention of vascular and neural damage. Postoperative development of liquorrhea is the most common complication in these patients, often associated with inadequate sealing of the dura mater (38). Careful closure of the dura mater is important to prevent postoperative liquorrhea.

The research gap addressed by this study involves a comprehensive understanding of the

diagnostic and treatment processes for patients with Arnold-Chiari malformation type 1 (ACM I) and associated conditions. While previous studies have examined symptoms and surgical outcomes, this study contributes by highlighting the prevalence of comorbid abnormalities in craniovertebral bone morphology, particularly platybasia, which was common among patients.

CONCLUSIONS

In the diagnostic process, patients with Chiari malformation type 1 were found to have comorbid abnormalities in the morphology of the bones of the craniovertebral region, among which platybasia was the most common, but they were not associated with the degree of ectopy of the cerebellar tonsils. The hypertensive syndrome was found in 96 % of patients, motility disorders in 48 %, and cranial nerve damage in 8 %. These symptoms were closely correlated with the degree of ectopy and were most clearly expressed in patients with prolapse of more than 5 mm. 201 patients underwent surgery. If syringomyelia was detected, the operation was supplemented with syringostomy, which made it possible to eliminate the symptom of syringomyelia in 5 % of patients, however, in 29 % of cases after I+S, relapse was observed. As a result of the operation, preoperative symptoms completely disappeared in 58 % of patients; in 38 % there was an alleviation of vestibulo-atactic abnormalities and hypertension syndrome; stabilization of the process with preservation of focal damages was noted in 4 % of cases. Patients with progressive postoperative syringomyelia and hydrocephalus (5 %) underwent shunting, as a result of which the symptoms subsided after 3 months. In 2 % of cases after surgeon treatment, a tight filum terminale syndrome has been developed, which was eliminated by excision of the filum terminale.

Enhancing care for Arnold-Chiari malformation type 1 involves leveraging high-resolution MRI and computational modelling for accurate diagnosis, alongside tailored surgical strategies based on herniation severity and associated conditions. Intraoperative tools like ultrasound and neurological monitoring aid precise decompression. Optimization of surgical techniques such as suboccipital craniectomy and duraplasty, and exploration of adjuvant therapies like CSF shunting, contribute to improved outcomes. Standardized metrics, follow-up procedures, and data analysis through patient registries further refine treatment approaches.

The significance of this study lies in its potential to revolutionize the approach to diagnosing and treating patients with ACM I, ultimately improving their quality of life and healthcare outcomes. The key limitations of this study on surgical treatment for Arnold-Chiari malformation type 1 include the retrospective observational design without a control group, modest sample size from a single center, lack of long-term postoperative follow-up, incomplete evaluation of factors underlying clinical outcomes, limited use of advanced imaging modalities for surgical planning, no assessment of adjuvant therapies or quality of life impact, and no analysis of costs versus benefits. The main future research priorities for improving surgical management of Arnold-Chiari malformation should focus on prospective multicenter randomized controlled trials incorporating long-term follow-up, advanced diagnostic imaging, analysis of prognostic factors, cost-effectiveness, quality of life impacts, adjuvant therapies, and basic science models to better define optimal treatment protocols for restoration of normal anatomy and physiology.

REFERENCES

1. Fric R, Eide PK. Chiari type 1 – A malformation or a syndrome? A critical review. Acta Neirochir (Wien). 2020;162(7):1513-1525.
2. Chu W, Chen X, Xue X. Treatment of symptomatic Chiari I malformation by "all-factors-surgery": A report of 194 cases. Eur Spine J. 2021;30:1615-1622.
3. Gallo P, Copley PC, McAllister S, Kaliaperumal C. The impact of neurosurgical technique on the short- and long-term outcomes of adult patients with Chiari I malformation. Clin Neurol Neurosurg. 2021;200:106380.
4. Nash J, Cheng JS, Meyer GA, Remler BF. Chiari type I malformation: Overview of diagnosis and treatment. Wisconsin Medical J. 2002;101(8):35-40.
5. Schijman E. History, anatomic forms, and pathogenesis of Chiari I malformations. Child's Nervous System. 2004;20(5):323-328.

6. Strahle J, Muraszko KM, Kapurch J, Bapuraj JR, Garton HJL, Maher CO. Natural history of Chiari malformation Type I following decision for conservative treatment: Clinical article. J Neurosurg. 2011;8(2):214-221.
7. Joseph N, Rajderkar D, Baker D, Prabhakaran S. Arnold-Chiari malformation: An uncommon etiology for a brief resolved, unexplained event in an infant. J Clin Sleep Med. 2020;16(11):1983-1984.
8. Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y. Pathogenesis of Chiari malformation: A morphometric study of the posterior cranial fossa. J Neurosurg. 1997;86(1):40-47.
9. Sakushima K, Tsuboi S, Yabe I, Hida K, Terae S, Uehara R. Nationwide survey on the epidemiology of syringomyelia in Japan. J Neurological Sciences. 2012;313(1-2):147-152.
10. Urbizu A, Garrett ME, Soldano K, Drechsel O, Loth D, Marcé-Grau A, et al. Rare functional genetic variants in *COL7A1*, *COL6A5*, *COL1A2* and *COL5A2* frequently occur in Chiari Malformation Type 1. PLoS One. 2021;16(5):e0251289.
11. Barpujari A, Kiley A, Ross JA, Veznedaroglu E. A systematic review of non-opioid pain management in Chiari Malformation (Type 1) patients: Current evidence and novel therapeutic opportunities. J Clin Med. 2023;12(9):3064.
12. Rosenblum JS, Pomeraniec IJ, Heiss JD. Chiari Malformation (update on diagnosis and treatment). Neurol Clin. 2022;40(2):297-307.
13. Labula R, Talla Nwotchouang BS, Inrahimy A, Allen PA, Oshinski JN, Klinge P, et al. A new hypothesis for the pathophysiology of symptomatic adult Chiari malformation Type I. Med Hypotheses. 2022;158:110740.
14. Alluhaybi AA, Abdulqader SB, Alanazi T, Altuhayni K, Albanyan A. Chiari I malformation with neurogenic hypertension after suboccipital decompression. Child's Nervous System. 2021;37(2):659-663.
15. Zhang Y, Zhang N, Qiu H, Zhou J, Li P, Ren M. An efficacy analysis of posterior fossa decompression techniques in the treatment of Chiari malformation with associated syringomyelia. J Clin Neuroscience. 2011;18(10):1346-1349.
16. Valentini L, Visintini S, Saletti V, Chiapparini L, Estienne M, Solero, CL. Treatment for Chiari 1 malformation (CIM): Analysis of a pediatric surgical series. Neurological Sciences. 2011;3:321-324.
17. Yundt KD, Park TS, Tantuwaya VS, Kaufman BA. Posterior fossa decompression without duraplasty in infants and young children for treatment of Chiari malformation and achondroplasia. Pediatric Neurosurg. 1996;25(5):221-226.
18. Sindou M, Chávez-Machuca J, Hashish H. Craniocervical decompression for Chiari type I-malformation, adding extreme lateral Foramen Magnum opening and expansile duroplasty with arachnoid preservation. Technique and long-term functional results in 44 consecutive adult cases – Comparison with literature data. Acta Neurochirurgica. 2022;144(10):1005-1019.
19. Sindou M, Gimbert E. Decompression for Chiari type I-malformation (with or without syringomyelia) by extreme lateral foramen magnum opening and expansile duraplasty with arachnoid preservation: Comparison with other technical modalities (literature review). Advances and Technical Standards in Neurosurgery. 2009;34:85-110.
20. Zhang ZQ, Chen YQ, Chen YA, Wu X, Wang YB, Li XG. Chiari I malformation associated with syringomyelia: A retrospective study of 316 surgically treated patients. Spinal Cord. 2007;46(5):358-363.
21. Vega A, Quintana F, Berciano J. Basichondrocranium anomalies in adult Chiari type I malformation: A morphometric study. J Neurological Sciences. 1990;99(2-3):137-145.
22. Eule JM, Erickson MA, O'Brien MF, Handler M. Chiari I malformation associated with syringomyelia and scoliosis: A twenty-year review of surgical and nonsurgical treatment in a pediatric population. Spine. 2022;27(13):1451-1455.
23. Aghakhani N, Parker F, David P, Morar S. Long-term follow-up of Chiari-related syringomyelia in adults. Neurosurgery. 2009;64(2):308-315.
24. Weprin BE, Iskandar BJ, Hedlund GL, Grabb PA, Oakes WJ. The resolution of syringohydromyelia without hindbrain herniation after posterior fossa decompression. J Neurosurg. 1998;89(2):212-216.
25. Zérah M. Syringomyelia in children. Neurochirurgie. 1999;45(1):37-57.
26. Liang CJ, Dong QJ, Xing YH, Shan M, Wen LX, Qiang ZY. Posterior fossa decompression combined with resection of the cerebellomedullary fissure membrane and expansile duraplasty: A radical and rational surgical treatment for Arnold-Chiari type I malformation. Cell Biochemistry and Biophysics. 2014;70(3):1817-1821.
27. Marković M, Berisavac I, Bojović V, Kostić B, Dulejić V. Surgical treatment of Arnold-Chiari malformation type I in an adult patient. Vojnosanitetski Pregled. 2008;65(8):648-652.
28. Albert GW. Chiari malformation in children. Pediatric Clinics of North America. 2021;68(4):783-792.
29. Tursynova A, Omarov B, Sakhipov A, Tukenova N. Brain Stroke Lesion Segmentation Using Computed Tomography Images based on Modified U-Net Model

with ResNet Blocks. International J Online and Biomedical Engineering. 2022;18(13):97-112.

30. Belenichev I, Burlaka B, Bukhtiyarova N, Aliyeva O, Makyeyeva L, Bak P. The effect of intranasal administration of an IL-1b antagonist (rail) on the state of the nitroxydergic system of the brain during modeling of acute cerebrovascular accident. Azerbaijan Pharmaceut Pharmacoth Jl. 2022;22(1):78-85.
31. Slivkina N, Abduldayeva A, Tardjibayeva S, Doszhanova G, Kuanyshbayeva G. The health of the population, according to prenosological diagnostics. Georgian Medical News. 2020;(303):188-193.
32. Nuradilova D, Kaliyeva L, Vaitkiene D, Kalimoldayeva S, Issenova S. Urogenital mixed infections in reproductive aged women with pelvic inflammatory disease. Georgian Medical News. 2021;(312):114-118.
33. Sagandykova NS, Fakhradiyev IR, Sajjala SR, Taukeleva SA, Shemetova DE, Saliev TM, et al. Patient-specific CFD simulation of aerodynamics for nasal pathology: A combined computational and experimental study. Computer Methods in Biomechanics and Biomedical Engineering: Imaging and Visualization. 2021;9(5):470-479.
34. Nazarchuk O, Dmyrtriiev D, Babina Y, Faustova M, Burkot V. Research of the activity of local anesthetics and antiseptics regarding clinical isolates of *Acinetobacter baumannii* as pathogens of postoperative infectious complications. Acta Biomedica. 2022;93(1):e2022003.
35. Latka K, Kolodziej W, Pawlak K, Sobolewski T, Rajski R, Chowaniec J, et al. Fully Endoscopic Spine Separation Surgery in Metastatic Disease—Case Series, Technical Notes, and Preliminary Findings. Medicina (Lithuania). 2023;59(5):993.
36. Sakaguchi T, Tanaka M, Sake N, Latka K, Fujiwara Y, Arataki S, et al. The Most Significant Factor Affecting Gait and Postural Balance in Patients' Activities of Daily Living Following Corrective Surgery for Deformity of the Adult Spine. Medicina (Lithuania). 2022;58(8):1118.
37. Navruzov SN, Polatova DS, Geldieva MS, Nurieva EI. Possibilities of study of the main cytokines of the immune system in patients with osteogenic sarcoma. Voprosy Onkologii. 2013;59(5):599-602.
38. Sahalevych AI, Sergiychuk RV, Ozhohin VV, Khrapchuk AYu, Dubovyi YO, Frolov OS. The Modified Procedure of Totally Tubeless PNL. International J Biol Biomed Engineer. 2022;16:82-89.