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OUTCOMES OF MICROSURGICAL TREATMENT OF CHIARI TYPE I MALFORMATION USING A DIFFERENTIATED INTRAOPERATIVE STRATEGY FOR DETERMINING THE EXTENT OF INTERVENTION

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Abstract
Despite the presence of numerous publications and analytical reviews in the international literature, a universally accepted algorithm for the surgical treatment of Chiari malformation type I (CM-I) does not currently exist. In the domestic literature, the number of publications devoted to the surgical management of CM-I remains limited, which, in our opinion, highlights the relevance of this study.

Keywords: Chiari malformation, diagnosis, surgical treatment.

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Introduction

The widespread use of magnetic resonance imaging (MRI) for evaluating various neurological symptoms has led to an increased number of diagnoses of Chiari malformation type I (CM-I), especially in children, as the prevalence of low-lying cerebellar tonsils is higher in the pediatric population than in adults [1]. In most cases, CM-I is an incidental finding, unrelated to the original reason for performing the MRI. Nevertheless, such patients are often referred to a neurosurgeon for consultation. CM-I is characterized by the descent of the cerebellar tonsils more than 5 mm below the level of the foramen magnum. A key difference from Chiari malformation type II (Arnold–Chiari syndrome) in children is the absence of associated spinal cord malformations (e.g., myelomeningocele), as well as the absence of caudal displacement of the brainstem and the fourth ventricle into the cervicomedullary junction [2].According to the literature, MRI signs of CM-I are found in 1% to 3% of children in the general population, and 30% to 50% of these patients also present with syringomyelia of the cervical spinal cord [3–5]. However, only one-third of patients exhibit clinical symptoms, and no more than 15% require surgical intervention [6]. The clinical manifestations of CM-I include Valsalva-induced headaches in the occipital and cervical regions, as well as dysphagia, respiratory dysfunction, dysesthesias, pyramidal insufficiency, cerebellar symptoms (such as ataxia and nystagmus), and other neurological signs [7]. Surgical indications are typically based on persistent and progressive clinical symptoms of CM-I [8]. The surgical techniques described in the literature range from extradural bony decompression to resection of the herniated cerebellar tonsils and stenting of the foramen of Magendie in cases of severe or recurrent syringomyelia [9]. Surgical decision-making becomes particularly complex in the presence of additional anomalies (such as basilar impression) or instability at the craniovertebral junction, which are observed in approximately 15% of patients [10].

Despite the abundance of international publications and reviews on this topic, there is still no universally accepted surgical treatment algorithm for pediatric CM-I. In many cases, the extent of the surgical intervention is determined by the operating surgeon based on personal experience. In the domestic literature, there

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are significantly fewer publications addressing surgical treatment of CM-I in children, which, in our view, underlines the relevance of the present study.

Materials and Methods

We conducted an analysis of the diagnostic and surgical treatment outcomes of 43 pediatric patients, aged from 1 to 17 years (mean age: 8.48 years), diagnosed with Chiari malformation type I (CM-I). Patient history and clinical examination were used to assess the presence of general cerebral symptoms (headache, nausea, vomiting), bulbar dysfunctions (respiratory, swallowing, and phonation disturbances), conduction disorders (sensory disturbances, pyramidal insufficiency), visual impairments (reduced visual acuity, diplopia), cerebellar symptoms (ataxia, nystagmus), and autonomic symptoms (fatigue, lethargy, hyperhidrosis, etc.). MRI scans were used to determine the degree of cerebellar tonsil ectopia and the presence of syringomyelia. Additional diagnostic methods (e.g., evoked potentials, Doppler ultrasonography) were performed in some patients, but these were considered auxiliary and not decisive in determining the treatment strategy, and therefore were not analyzed separately in this study. Patients with craniovertebral junction anomalies associated with instability or ventral brainstem compression were excluded from the study. Surgical treatment was performed in patients with clinical symptoms and was carried out using a standard technique. The surgical procedure included resection of the edge of the foramen magnum and adjacent portions of the occipital bone, laminectomy of the C1 vertebra, and dissection of adhesions between the outer layer of the dura mater and adjacent tissues. The decision to extend the scope of surgical intervention was made intraoperatively based on visually assessed signs of adequate extradural decompression (e.g., clearly transmitted pulsations of the dura mater) or intraoperative ultrasonographic findings, the use of which is also described in the literature [11]. If signs of persistent compression of the subarachnoid spaces were identified, dural opening and duraplasty were performed using a graft from the periosteum or muscle fascia.

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Results

In the early postoperative period, respiratory dysfunctions regressed in all patients. Headache and vomiting resolved in 92% of cases, general cerebral and autonomic symptoms in 94%, visual disturbances in 80%, dysphagia in 60%, and sensory deficits in 39% of children. Pyramidal insufficiency regressed in 32% of cases, and cerebellar symptoms in 28%. No worsening of neurological symptoms was observed following surgery. Postoperative complications were noted in two patients who underwent duraplasty: one developed a pseudomeningocele, and another developed a cerebrospinal fluid (CSF) leak with subsequent meningitis (2 out of 12 patients — 16.6%). No complications were observed in the group that underwent extradural decompression. Long-term follow-up data were available for 33 patients (76.7% of the cohort), including 9 patients (75%) from the duraplasty group and 24 patients (77%) from the extradural decompression group. None of the patients experienced respiratory disorders during follow-up. No statistically significant difference was found in clinical outcomes (regression of symptoms) between patients who underwent surgery with or without duraplasty.Two children from the extradural decompression group underwent reoperation due to recurrence of general cerebral symptoms. In both cases, symptoms resolved following the second surgery, which involved enlargement of the craniectomy window without dural opening.

Discussion

Chiari malformation type I (CM-I) is increasingly considered an acquired condition with a multifactorial etiology, including disturbances in cerebrospinal fluid (CSF) dynamics and venous outflow, skull developmental anomalies, craniovertebral junction instability, as well as metabolic and endocrine disorders. CM-I is rarely diagnosed in newborns and typically is not accompanied by structural changes in the cerebellum or brainstem. Moreover, numerous reports have documented cases of cerebellar tonsil ectopia reversal following treatment of the underlying pathology. Nevertheless, a reduction in posterior cranial fossa volume associated with CM-I may lead to the development of clinical symptoms even with minimal tonsillar herniation [12]. The disease most commonly

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manifests in older children and is characterized by a polymorphic clinical presentation. Among the most frequently reported symptoms are paroxysmal headaches (triggered by straining, coughing, etc.), dysphagia, respiratory dysfunction, autonomic disturbances (e.g., fatigue, weakness, somnolence), sensory and motor deficits (e.g., dysesthesias, pyramidal insufficiency), dizziness, gait disturbances, and others [13]. Ataxia and nystagmus may also be observed. In some cases, CM-I may be associated with delayed speech development and behavioral disorders. Given the wide spectrum of symptoms, misinterpretation is common. For instance, migraine-like headaches, behavioral abnormalities, seizures, and scoliosis should be cautiously associated with CM-I [14, 15]. One of the most characteristic clinical features of CM-I in children is respiratory dysfunction, which may affect up to 70% of patients with confirmed diagnosis. The cause is thought to be related to either direct compression of the respiratory center or paresis of the vocal cords and pharyngeal muscles [16]. In our cohort, the majority of patients were aged 3 to 12 years, with general cerebral symptoms predominating, and clinical signs of respiratory dysfunction were observed in 37% of cases. Surgical treatment of CM-I is generally recommended only for symptomatic patients, even when syringomyelia is present [13, 17, 18]. Reports of postoperative mortality and a relatively high rate of complications (up to 20%) [19] emphasize the importance of a differentiated approach when deciding whether to expand the surgical intervention. It is known that the risk of complications is higher when the dura mater is opened [20]. However, in pediatric patients, the high elasticity of tissues and ongoing cranial development may allow for correction of craniocephalic disproportion through extradural decompression alone. While this approach was previously believed to be less effective, more recent studies have not confirmed this concern [20, 21]. Nevertheless, some surgeons continue to favor duraplasty in all cases to prevent reoperation and promote faster resolution of syringomyelia [21]. In our study, extradural decompression proved effective, particularly for symptoms of brainstem dysfunction. Respiratory symptoms fully regressed in all patients. Conduction disturbances and cerebellar signs showed gradual improvement. In the long-term follow-up period, positive dynamics were also noted in speech development.

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Dural opening and duraplasty were performed in 12 patients (28%) who showed persistent subarachnoid space compression intraoperatively. Our analysis indicated that patients requiring duraplasty more frequently presented preoperatively with signs of hypertensive syndrome (headache, vomiting), visual disturbances, and respiratory dysfunction. However, these associations were not statistically significant. The need for duraplasty did not correlate with patient age, the extent of tonsillar descent, or the presence of syringomyelia. Long-term outcomes revealed no clinical advantage of duraplasty over extradural decompression in terms of symptom regression. We also found no significant difference in the frequency of duraplasty between patients with and without syringomyelia, although cyst sizes were not specifically tracked over time. Literature data show that even after duraplasty, syrinx cavities reduce in size or resolve in just over 50% of cases, remain stable in about one-third, and may increase in others [15]. According to previous studies, headaches, nausea, or vomiting may persist in 35% of patients after surgery, and sensorimotor deficits in 45% [15]. In some cases, symptoms may even worsen despite treatment. Only 38% of patients with predominant coordination disorders experience regression of these symptoms postoperatively [24]. In our series, long-term follow-up showed persistent headache in 21% of patients, conduction and cerebellar symptoms in 12%, and vomiting and bulbar symptoms in 6%. Among the 9 patients who underwent duraplasty, headache persisted in 3, and vomiting in 2. However, sensory, visual, and bulbar symptoms fully regressed in this group. The recurrence of headache, vomiting, and other symptoms in some patients, as well as characteristic complications such as pseudomeningocele and CSF leakage in the duraplasty group, may indirectly suggest persistent CSF circulation disturbances or other underlying issues requiring further evaluation and management. These findings may reduce the specificity of our proposed intraoperative decision-making algorithm for determining the extent of intervention. The need for reoperation in two patients from the extradural decompression group—often considered a drawback of this technique—was likely due to ossification of the bony defect [18].

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Conclusion

The outcomes of surgical treatment for Chiari malformation undoubtedly depend on thorough preoperative evaluation and careful selection of candidates for surgery. In our study, headache was the least specific clinical manifestation of CM-I, whereas respiratory dysfunction was the most specific symptom. The surgical approaches commonly compared in the literature—CM-I correction with and without dural opening—should, in our view, be considered as sequential stages of the same operative procedure. The final decision regarding the extent of the intervention should reasonably be based on intraoperative assessment of the adequacy of extradural decompression. According to our data, none of the preoperative parameters evaluated showed a statistically significant association with the decision to perform duraplasty, likely due to individual patient variability.

REFERENCES

1. Abel F, Tahir MZ. Role of sleep study in children with Chiari malformation and sleep disordered breathing. Child's Nervous System. 2019;35(10):1763-1768. https://doi.org/10.1007/s00381-019-04302-0
2. Aitken LA, Lindan CE, Sidney S, Gupta N, Barkovich AJ, Sorel M, Wu YW. Chiari type I malformation in a pediatric population. Pediatric Neurol- ogy. 2009;40(6):449-454.https://doi.org/10.1016/j.pediatrneurol.2009.01.003
3. Broussolle T, Beuriat PA, Szathmari A, Rousselle C, Di Rocco F, Mot- tolese C. Management of Chiari type I malformation: a retrospective analysis of a series of 91 children treated surgically. Acta Neurochirurgica. 2021;163(11):3065-3073.
4. Carey M, Fuell W, Harkey T, Albert GW. Natural history of Chiari I malformation in children: a retrospective analysis. Child's Nervous System. 2021;37(4):1185-1190.
5. Chae JK, Greenfield JP. Revision Chiari Surgery in Young Children: Predictors and Outcomes. Pediatric Neurosurgery. 2021;56(6):529-537. https://doi.org/10.1159/000519201
6. Chai Z, Xue X, Fan H,Sun L, Cai H, Ma Y, Ma C, Zhou R. Efficacy of Pos-

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terior Fossa Decompression with Duraplasty for Patients with Chiari Malformation Type I: A Systematic Review and Meta-Analysis. World Neurosurgery. 2018;113:357-365.e1.

7. Chatrath A, Marino A, Taylor D, Elsarrag M, Soldozy S, Jane JA Jr. Chi- ari I malformation in children-the natural history. Child's Nervous System. 2019;35(10):1793-1799.
8. 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. https://doi.org/10.3171/2011.4.PEDS10528
9. Davidson L, Phan TN, Myseros JS, Magge SN, Oluigbo C, Sanchez CE, Keating RF.. Long-term outcomes for children with an incidentally discovered Chiari malformation type 1: what is the clinical significance?. Child's Nervous System. 2021;37(4):1191-1197.
10. Fons K, Jnah AJ. Arnold-Chiari Malformation: Core Concepts. Neonatal Network. 2021;40(5):313-320. https://doi.org/10.1891/11-T-704
11. Furtado SV, Thakar S, Hegde AS. Correlation of functional outcome and natural history with clinicoradiological factors in surgically managed pediatric Chiari I malformation. Neurosurgery. 2011;68(2):319-328. https://doi.org/10.1227/NEU.0b013e31820206e5
12. Jiang E, Sha S, Yuan X, Zhu W, Jiang J, Ni H, Liu Z, Qiu Y, Zhu Z. Comparison of Clinical and Radiographic Outcomes for Posterior Fossa Decompression with and without Duraplasty for Treatment of Pediatric Chiari I Malformation: A Prospective Study. World Neurosurgery. 2018;110:465-472. https://doi.org/10.1016/j.wneu.2017.11.007
13. Klekamp J. Chiari I malformation with and without basilar invagination: a comparative study. Neurosurgical Focus. 2015;38(4):E12. https://doi.org/10.3171/2015.1.FOCUS14783
14. Massimi L, Frassanito P, Bianchi F, Tamburrini G, Caldarelli M. Bony decompression vs duraplasty for Chiari I malformation: does the eternal dilemma matter? Child's Nervous System. 2019;35(10):1827-1838. https://doi.org/10.1007/s00381-019-04218-9

Modern American Journal of Medical and Health Sciences
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Website: usajournals.org

15. McGirt MJ, Attenello FJ, Datoo G, Gathinji M, Atiba A, Weingart JD, Carson B, Jallo GI.Intraoperative ultrasonography as a guide to patient selec- tion for duraplasty after suboccipital decompression in children with Chiari malformation Type I. Journal of Neurosurgery: Pediatrics. 2008;2(1):52-57. https://doi.org/10.3171/PED/2008/2/7/052
16. Mikulis DJ, Diaz O, Egglin TK, Sanchez R. Variance of the position of the cerebellar tonsils with age: preliminary report. *Radiology*. 1992;183(3):725-728. https://doi.org/10.1148/radiology.183.3.1584927
17. Riordan CP, Scott RM. Fourth ventricle stent placement for treatment of recurrent syringomyelia in patients with type I Chiari malformations. Journal of Neurosurgery: Pediatrics. 2018;23(2):164-170. https://doi.org/10.3171/2018.7.PEDS18312
18. Sanakoeva AV, Korshunov AE, Kadyrov ShU, Khukhlaeva EA, Kushel' IuV. Posterior decompression of the craniovertebral junction in syringomy- elia combined with Chiari-1 malformation in children. Burdenko's Journal of Neurosurgery. 2017;81(3):48-57. (In Russ.). https://doi.org/10.17116/neiro201781348-56
19. Singhal A, Bowen-Roberts T, Steinbok P, Cochrane D, Byrne AT, Kerr JM. Natural history of untreated syringomyelia in pediatric patients. Neurosurgical Focus. 2011;31(6):E13.
20. Strahle J, Muraszko KM, Kapurch J, Bapuraj JR, Garton HJ, Maher CO. Chiari malformation Type I and syrinx in children undergoing magnetic resonance imaging. Journal of Neurosurgery: Pediatrics. 2011;8(2):205-213. https://doi.org/10.3171/2011.5.PEDS1121
21. Strahle J, Smith BW, Martinez M, Bapuraj JR, Muraszko KM, Garton HJ, Maher CO. The association between Chiari malformation Type I, spinal syrinx, and scoliosis. Journal of Neurosurgery: Pediatrics. 2015;15(6):607-611. https://doi.org/10.3171/2014.11.PEDS14135