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Hundred Pediatric Cases Treated for Chiari Type II Malformation with Hydrocephalus and Myelomeningocele

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Abstract

Background Chiari malformation type II (CM-II) may not always present as an asymptomatic disorder but prove to be difficult in managing. This is especially true for neonates who show the worst prognosis. There is confounding data over whether shunting or craniocervical junction (CVJ) decompression should be employed. This retrospective analysis summarizes the results of 100 patients diagnosed and treated for CM-II along with hydrocephalus and myelomeningocele.

Methods We reviewed all the children who were diagnosed and surgically treated for CM-II at the Moscow Regional Hospital. Surgical timing was decided on the clinical conditions of each patient. Urgent surgery in the more compromised patients (usually infants) and elective surgery for patients with less severe conditions was performed. All patients first underwent CVJ decompression.

Results The retrospective review yielded 100 patients operated on for CM-II with concomitant hydrocephalus and myelomeningocele. The average herniation was 11.2 ± 5.1 mm. However, herniation level did not correlate with clinical findings. Concomitant syringomyelia was observed in 60% of patients. More severe spinal deformity was observed in patients with widespread syringomyelia ($p = 0.04$). In children of the younger age group, cerebellar symptoms and bulbar disorders were more frequently observed ($p = 0.03$), and cephalic syndrome was noted much less frequently ($p = 0.005$). The severity of scoliotic deformity correlated with the prevalence of syringomyelia ($p = 0.03$). Satisfactory results were significantly more often observed in patients of the older age group ($p = 0.02$). Patients with unsatisfactory results at the time of treatment were significantly younger ($p = 0.02$).

Conclusion If CM-II is asymptomatic, then no specific treatment is prescribed. If the patient develops pain in the occiput and neck, then pain relievers are prescribed. If a

Keywords
- Chiari II malformation
- craniocervical junction decompression
- myelomeningocele
- spina bifida
- syringomyelia

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patient has neurological disorders or concomitant syringomyelia, hydrocephalus or myelomeningocele, surgical intervention is indicated. The operation is also performed if the pain syndrome cannot be overcome within the framework of conservative therapy.

Introduction

Chiari malformation type II (CM-II) is the extrusion of the cerebellar tonsils and vermis and the fourth ventricle through foramen magnum, resulting in the compression of the medulla oblongata and the spinal cord. It is almost always present with spinal myelomeningocele (MMC). Among other things, CM-II leads to a violation of the outflow of cerebrospinal fluid (CSF) resulting in obstructive hydrocephalus. CM-II is the most common Chiari malformation that presents itself during childhood and has an incidence of 1/1000 births.[1]

A congenital malformation, Chiari syndrome, was first described by the German pathologist Hans Chiari in 1890. Julius Arnold was studying the displacement of the tonsils from the cerebellum and caudal brain into the foramen magnum at the same time. This anomaly occurs with a frequency of 1 to 3:1000 newborns. Children born with MMC in 95% of cases have a malformation of the posterior cranial fossa (CM-II) as an associated pathology.[2]

Morphological manifestations of this pathology in the fetus can be detected during an ultrasonography (USG) and predetermine further prognosis. Very often, the detection of CM-II becomes the reason for termination of pregnancy.

The formation of the defect is based on a combination of pathological changes. The spinal cord in its lower part with fixation to the posterior wall of the spinal canal is unable to move cephalic during fetal growth. As a result, the medulla oblongata and partly the cerebellum enter through a large opening into the lumen of the spinal canal. With difficulty of normal CSF flow, hydrocephalus develops. At the same time, the medulla oblongata is severely deformed, the cerebellum is compressed and delayed in development. The process often progresses during the intrauterine and postnatal periods. Depending on the type of hernia, there are three types of Chiari syndrome.[3] This study focuses on CM-II in the newborn and pediatric cases. This type of pathology is characterized by a complex of brain deformities, an elongated shape of the dysplastic (almond-shaped) cerebellum and brain stem, which are embedded (wedge-shaped) in the large foramen. On the side of the lumbar spinal cord, there is a pathological development in the form of spina bifida—meningocele or MMC. Pathology of the brain is accompanied by dysgenesis of the corpus callosum, absence of a transparent septum, obstructive hydrocephalus, polymicrogyria, porencephaly, schizencephaly, and fenestration of the falx of the brain. Changes in the hindbrain in type II Arnold-Chiari malformation include more pronounced elongation and caudal displacement of the structures of the posterior cranial fossa into the spinal canal. In addition to these changes, changes in the vault and base of the skull are often observed. The differential diagnosis is with a CM-I (no MMC) and an isolated MMC without anomaly of the posterior cranial fossa.[4]

The clinic is mainly associated with the compression mechanism of damage to the nervous structures and impaired hemolytic dynamics. In this case, intracranial hypertension and hydrocephalus develop. CM-II anomaly has clinical manifestations that become noticeable from the first minutes of a child's life (spina bifida).[5] Children with this pathology are born with a hydrocephalic skull shape. Hydrocephalus interferes with normal development, newborns suffer from respiratory, palpitation, and swallowing disorders (bulbar disorders). Often the disease is accompanied by convulsive seizures. Children develop nystagmus, apnea, stridor, paresis of the vocal cords, dysphagia with regurgitation, and impaired tone in the limbs. The severity of neurological symptoms primarily depends on the severity of CSF flow disorders, and not on the degree of cerebellar tonsil ectopia.[6]

Although there is no clear algorithm for the treatment and surgical steps of CM-II, most commonly accepted approach is primarily allowing for sufficient CSF drainage by a craniovertebral junction (CVJ) decompression and addressing MMC.[2] CM-II presents itself in the newborn with more severe symptoms than type I. Paresis of the vocal cords, impaired tone in the extremities, and apnea and dysphagia with regurgitation are common clinical findings.[7]

Although clinical findings may be subtle, some life-threatening situations require immediate intervention to prevent morbidity and mortality. The relevance of this study is based on the demonstration of the key factors in the development of the problem and the lack of proper research in this direction. Although this anomaly has already been studied sufficiently, there is no single approach to address all the issues related with CM-II, which necessitates the study of the presented anomaly. Despite the advances in neonatal imaging, CM-II is usually diagnosed clinically; thus, a clear understanding of the pathologies and the treatment approaches should be familiar to those who work close with this entity. In this study, we report our 20-year experience with the surgical management of 100 pediatric cases diagnosed and treated for CM-II along with hydrocephalus and MMC.

Materials and Methods

This retrospective analysis was carried out on 100 patients aged between 0 and 17 years operated on for CM-II associated with hydrocephalus and MMC at the Moscow Regional

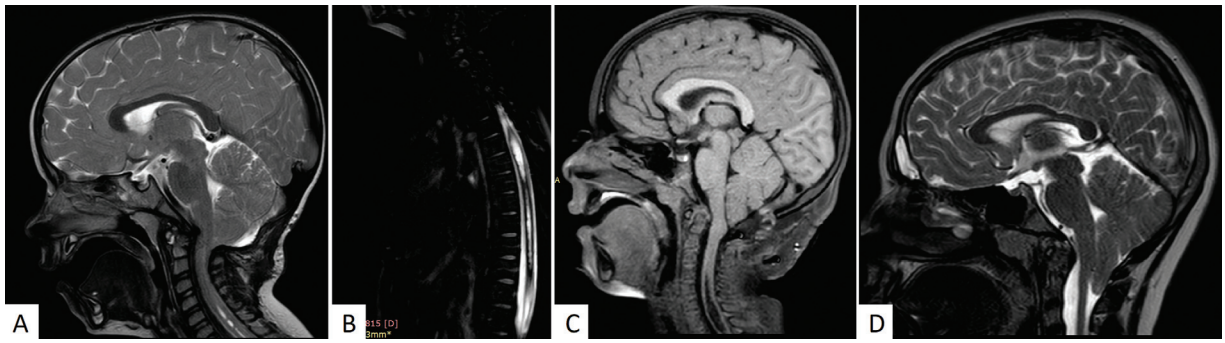

Fig. 1 (**A**) A 3-year-old boy with downward placement of cerebellar tonsils and brain stem. (**B**) Dramatic syringomyelia extending from C6 to lower thoracic levels in the same patients. Concomitant scoliosis was also present. (**C**) A 5-year-old girl diagnosed for Chiari malformation type II with cervical syringomyelia who underwent craniocervical junction decompression. (**D**) A 13-year-old boy with a severe herniation of cerebellar tonsils and brain stem. The patient did not show any major symptoms in the first decade of his life. This may have been due to a rather large foramen magnum and spinal canal at the C1-2 level.

Hospital. All patients underwent clinical, neuroimaging and laboratory studies. Magnetic resonance imaging (MRI) of the brain and the spinal cord was the main method of neuroimaging of CM-II malformation associated with hydrocephalus and MMC in children.

The examination of the patients included an assessment of the neurological status and examination of the MRI data of the brain and spinal cord before and after the operation (►**Fig. 1**).

Operations were performed by the neurosurgical department under general anesthesia. After fixation with the Mayfield skull clamp in the prone position a linear midline incision of the occipitocervical region was made. The muscles were dissected from the midline. A wide suboccipital and C1 posterior arc decompression was performed. Under the microscope, dura was opened linearly. There usually were a large number of dense adhesions between the arachnoid and inner membrane of the dura mater. This usually resulted in arachnoid fenestration. After a high flow of CSF, normal tonsil pulsation was observed. The fascia was then used for expansive duraplasty.

The analysis of the results of surgical interventions was carried out on the basis of protocols of operations and intraoperative observations. We analyzed the relationship between the immediate and long-term results of operations with the volume of the operation and preoperative parameters, including gender, age, clinical manifestations, and degree of dystopia of the cerebellar tonsils, the presence, and prevalence of syringomyelia.

Results

A total of 100 patients were operated on for CM-II from March 2002 till May 2022 (►**Table 1**). Patients underwent a comprehensive neurosurgical examination, which included general somatic, clinical neurological, neuropsychological, examination by an otoneurologist (35) and an ophthalmologist (24). When analyzing the history of 100 patients, pathological conditions of the perinatal period (intrauterine hypoxia, rapid delivery) were detected in 68 (68%) patients and birth trauma in 19 (19%) patients. Another 27 (27%) cases had a history of repeated craniocerebral trauma in childhood and adolescence with the site of application of the traumatic effect in the occipital region. Examination of the entire length of the spinal cord was carried out in 77% of cases. According to MRI data, the degree of herniation of the cerebellar tonsils below the McRae line averaged 11.2 ± 5.1 mm. The average degree of tonsil dystopia in patients with syringomyelia was 11.6 ± 4.6 mm and 10.8 ± 5.7 mm in patients without syringomyelia. It was found that the degree of tonsil herniation did not correlate with the severity of clinical manifestations and did not affect the development of syringomyelia or its prevalence ($p > 0.05$). Being a retrospective study, exact timing of the operations was unable to be obtained. Based on the available data, surgery was performed after diagnosis and correct correlation of findings with symptoms (►**Table 1**).

Concomitant syringomyelia was observed in 60% of patients. Depending on the localization, syringomyelia at the cervical level was detected in 30%, 40% in cervicothoracic, 10% in thoracic, and holocord in 20%. The prevalence of syringomyelia over five vertebral levels was 70%. The diameter of syringomyelia was determined in 60% of patients. Scoliotic spinal deformity of more than 10 Cobb degrees at the time of hospitalization was diagnosed in 45% of patients and was observed only in patients with concomitant syringomyelia. More severe spinal deformity was observed in patients with widespread syringomyelia (cervicothoracic and holocord, $p = 0.04$).

Table 1 Distribution of patients by sex and age

Age groups	Male	Female	Total
0–5 years	22	21	43
6–10 years	5	12	17
11–13 years	10	8	18
14–17 years	10	12	22
Total	47	53	100

Table 2 Clinical manifestations of patients associated with brain stem and cranial nerve dysfunction

Neurological syndrome, symptom	Number of patients
Reflex cough, choking	72
Syringomyelic syndrome	60
Pyramidal signs	58
Intermittent apnea	54
Neurogenic dysphagia	46
Behavioral syndromes	45
Nystagmus (especially downward)	37
Vegetovascular dystonia	36
Stridor	35
False Croup syndrome	35
Hypertension–hydrocephalic syndrome	28
Weak or no cry	27
Paraparesis of the upper limbs	25
Monoparesis of an upper limb	23
Cerebellar syndrome	23
Tetraparesis	22
Paresis of the facial muscles	21
Opisthotonus	21
Aspiration	11

Note: Patients showed a combination of symptoms therefore the total number does not correspond to 100.

Clinical manifestations of CM-II associated with brain stem dysfunction and the lower group of cranial nerves (CNs) are presented in ►**Table 2**. Severe obstructive hydrocephalus was more frequent in children aged between 0 and 5 years and 6 and 10 years (68%, 61%) compared with older age groups (27%, 23%).

Patients with tethered cord syndrome presented with dermatological findings, lower extremity neurological findings, spinal deformities, and pelvic floor dysfunction findings. A special group consisted of newborns ($n = 19$), in whom manifestations of CM-II differed significantly from the older children. The neonates were more likely to develop rapid neurological deterioration within hours. The brain stem and cranial nerve dysfunctions were as follows: neurogenic dysphagia, postfeeding cyanosis, nasal regurgitation, prolonged feeding time, salivary accumulation, and decreased/absent gag reflex. Apnea due to impaired respiratory stimulation was found in 12 newborns and 13 newborns had inspiratory stridor. Laryngoscope examination revealed paralysis of the abductor and rarely adductor vocal cords. All newborns had weakness in the upper limbs, downward nystagmus, a weak cry, and weakness of the facial muscles.

Ventriculomegaly without periventricular edema and signs of CSF occlusion, according to preoperative MRI, was observed in 15% of patients. In patients of the younger age group (0–5 years), ventriculomegaly was observed statistically significantly more often ($p = 0.02$), and syringomyelia was observed much less frequently ($p = 0.04$). The analysis of groups of patients with and without syringomyelia was carried out (►**Table 3**).

The clinical manifestations of the disease in different age groups were different. In children of the younger age group, cerebellar symptoms and bulbar disorders were more frequently observed ($p = 0.03$), and cephalic syndrome was noted much less frequently ($p = 0.005$). The clinical manifestations of the disease in patients with and without syringomyelia were dominated by the cephalic syndrome. Headaches were more common in children in the older age group (6–17 years; $p = 0.01$).

In patients with concomitant syringomyelia, symptoms of myelopathy were observed in 40% of patients and more often (29%) manifested as progressive scoliosis. In other cases (31%), motor, sensory disorders, a combination of motor and sensory disorders, sympathetic disorders (Horner's syndrome), and radicular pain were observed.

Severe ataxia and moderate bulbar symptoms were observed in children under 7 years of age statistically more often ($p < 0.05$). The main complaints from parents were

Table 3 Clinical symptoms of CM-II patients in accordance with syringomyelia

Clinical symptom	Syringomyelia absent *n* (%)	Syringomyelia present *n* (%)	Total (%)
Scoliosis	2 (5)	30 (40)	32 (32)
Bulbar disorders	7 (15)	9 (11)	16 (16)
Forced head position	–	8 (10)	8 (8)
Cephalic syndrome	30 (80)	40 (55)	70 (70)
Sensory deficits	2 (5)	18 (25)	20 (20)
Motor deficits	2 (5)	19 (27)	21 (21)
Dizziness	7 (15)	8 (10)	15 (15)
Cerebellar symptoms	9 (11)	25 (35)	34 (34)
Convergent strabismus	2 (5)	7 (15)	9 (9)

Abbreviation: CM-II, Chiari malformation type II.
Note: Patients showed a combination of symptoms therefore the total number does not correspond to 100.

Table 4 The emotional and behavioral symptoms before surgery

Emotional–behavioral disorders	0–5 years[a]	6–10 years[a]	11–13 years[a]	14–17 years[a]	Overall
Emotional lability	26	8	4	9	47%
Conduct violations	6	1	1	2	10%
Neuroticization	22	8	1	20	51%
Psychopathological symptoms	7	0	0	1	8%
Obsessive-compulsive traits	19	6	4	7	36%

[a]Values are given in number of patients.

increased fatigue, memory loss, capriciousness, and frequent irritability of children. The first symptoms of the disease were cluster headache (87%), coordination disorders (78%), and oculomotor disorders (24%). The early psychomotor development of the patients did not stray from the average age limits; most of the children were adaptive and attended school. An in-depth diagnosis of emotional and behavioral patterns was carried out (►**Table 4**).

According to the scales for diagnosing the emotional–behavioral sphere, 46% of children had emotional instability and 41% had a high level of neuropsychic stress (irritability, high anxiety, an abundance of fears, autonomic dysfunction, decreased appetite). Attention was drawn to the frequency and severity of obsessive–compulsive behavioral traits. In young children, persistent onychophagia and chronic lip biting were frequently observed (44%). Almost one-third of the patients in the 11 to 17 years of age group had compulsive-obsessive features in the form of obsessive rituals, vague fears, and early awakenings with a feeling of severe anxiety. At the same time, parents noted the occurrence of these features some time before the onset of the first symptoms of the disease. Moderate behavioral disorders were observed more often in patients under 7 years of age (13%). Cases of pathopsychological symptoms (nightmares, aggressiveness) were noted in the younger group (16%). In addition, a 16-year-old patient had a psychotic disorder in the form of delusions of persecution and visual hallucinations.

Progressive scoliosis was observed only in patients with syringomyelia. Although the literature describes cases of scoliosis in patients without syringomyelia due to the presence of increased cerebrospinal pressure in the central canal, in the present study, the severity of scoliotic deformity correlated with the prevalence of syringomyelia ($p = 0.03$).

In the postoperative period, clinical improvement was noted in 80% of patients, stabilization in 15%, and regression of syringomyelia was noted in 5% of cases. Complete or partial regression of the cephalic syndrome after surgery was observed in 63% of patients ($p = 0.003$), symptoms of trunk lesions and cranial nerves in 15%, cerebellar symptoms in 13%, and symptoms of myelopathy in 9%.

Satisfactory results were significantly more often observed in patients of the older age group ($p = 0.02$). Patients with unsatisfactory results at the time of treatment were significantly younger ($p = 0.02$). In this group, cerebellar symptoms were more often detected ($p = 0.02$). After the operation, 26 patients developed cerebellar mutism syndrome, 22 patients developed cerebellar cognitive-affective syndrome without loss of speech, and 52 children had no complex symptoms.

When comparing preoperative clinical and neuropsychological parameters in the selected groups, there were no significant differences in the groups in terms of gender and age. However, there was a trend toward the greatest vulnerability of children aged between 0 and 5 years to the occurrence of neuropsychological syndromes.

When evaluating the individual results of surgical treatment of CM-II associated with hydrocephalus and MMC in children, their relationship with the method of posterior decompression of the CVJ was well established. Our results were significantly better in patients who underwent duraplasty ($p = 0.01$), and worse in the group of patients with extradural decompression (EDD; $p = 0.03$).

In patients without syringomyelia, clinical results were statistically significantly better with extra-arachnoid duraplasty (EAD; $p = 0.04$). Intra-arachnoid dissection and duraplasty (IAD) (with or without shunting) were accompanied by a higher frequency of positive MRI dynamics ($p = 0.0003$). After shunting, regression of syringomyelia (partial or complete) was observed in all cases ($p = 0.02$). Children of the younger age group more often had an unsatisfactory result in the postoperative period ($p = 0.02$). Postoperative complications without syringomyelia were more often observed in patients of the younger age group ($p = 0.04$). The reoperation rate was 13.6%. When analyzing the methods of posterior decompression of the CVJ, it was found that EDD was associated with a greater need for repeated operations ($p = 0.008$).

Discussion

Depending on the quality of pregnancy monitoring and the volume of medical research, CM can be diagnosed even in the womb or sometime after birth. A number of authors believe that the cause of the pathology is the small size of the posterior cranial fossa, and other authors are inclined to believe that the anomaly develops against the background of an increased size of the brain. The development of type 2 malformation may be accompanied by other dysplasias of the nervous system. The difficulty of diagnosing this disease at an early age is because the clinical picture may not immediately warrant a suspicion. With the development of the disease, the brain stem is affected and may cause a

compression of cranial nerve nuclei. This threatens normal vision and hearing, the processes of swallowing, and coordinated movement.[8]

McRae line is clinically useful in measuring the level of cerebellar herniation; however, this itself is not a major prognostic factor. The degree of herniation should not be overestimated for the progression of the disease or syrinx cavity as evidenced by previous studies[9] and this study. There was no statistically significant relationship between the severity of symptoms and syringomyelia with level of cerebellar herniation. However, the presence of syringomyelia was found to be well correlated with the incidence of spinal deformity. The syrinx diameter is suggestive of deformity and scoliosis.[10] This should be taken into consideration as 60% of the patients presented with syringomyelia. On the other hand, the direct relationship between level of tonsillar herniation without syringomyelia to spinal deformity needs further evaluation.

The relationship between hydrocephalus and CM-II is well established. Earlier it was thought that the increased intracranial pressure by hydrocephalus caused the tonsillar herniation. However, it was later proven that the herniation itself plugged the foramen magnum causing a disturbance in the flow of CSF.[11] Younger children in this series had more severe hydrocephalus requiring urgent intervention. This may be because more severe hydrocephalus presenting with debilitating neurological findings is diagnosed at a younger age and treated earlier. This was observed in newborns as well. Newborns diagnosed with CM-II had a more rapid neurological deterioration, which further emphasized the importance of fetal diagnosis.

Our cases hide a wide range of neurological manifestations due to CM-II. Frequent choking, syringomyelic syndrome, pyramidal signs, intermittent apnea, and neurogenic dysphagia were the most commonly encountered findings. This is not unusual of CM-II as the tonsillar herniation causes a compression of the brain stem leading to numerous clinical findings. Although these findings are suggestive and expected for CM-II, almost half of the patients had various emotional and behavioral findings. Although psychiatric evaluation of patients with CM-I has been discussed within the literature,[12] evaluation of nonmotor behaviors caused by cerebellar dysfunction in patients with CM-II is lacking. Although cerebellar pathologies are well known for coordination and movement deficits, its relationship with cognitive impairment, language disorder, executive function impairment, visuospatial deficits, regressive personality, emotional lability, and dramatic mood swings are still being investigated.[13] It is important for families and physicians to recognize the co-existence of psychological disorders that may be caused by the disease itself or developmental disturbance due to prolonged hospitalization and treatment. The early detection of cerebellar cognitive and emotional pathologies would result in an earlier therapeutic intervention and restore normal emotional and cognitive functioning.

According to the results of our study, in patients with a syrinx cavity diameter of more than 6 mm, a 70% improvement in the patients with myelopathic symptoms and an 80% improvement in the group of patients without myelopathic symptoms were observed. Based on the results obtained, we formulated criteria for the effectiveness of surgical treatment of CM-II in children:

1. Dynamics of clinical symptoms of the disease after surgery (for patients without syringomyelia).
2. Dynamics of neuroimaging data according to the results of control MRI: the appearance of subarachnoid liquor spaces in the area of the craniovertebral junction, the large occipital cistern; reduction in the length (on sagittal slices) and diameter (on axial slices) of syringomyelia (in patients with concomitant syringomyelia).

Treatment focuses on decompression of the tonsillars concomitantly brain stem and restoration of a normal CSF flow. Surgery still remains the most effective method in achieving these goals and alleviating symptoms. Patients in this study benefited greatly from CVJ decompression. However, it should be noted that patients who underwent EDD had a higher rate of reoperation. Extra-arachnoid duraplasty and IAD resulted in clinically better results that were statistically significant. Bony decompression alone does not allow for sufficient decompression in the subdural area as the overlying dura is usually thicker and fibrotic. Thus, opening of the dura with a widening duraplasty and fenestration of arachnoid for CSF diversion should be strongly considered.

In the presence of concomitant diseases, there are indications for surgical intervention even in the early years of life. When deciding on the appointment of an operation, the presence or absence of anomalies of the bony structures, the presence or absence of hydrocephalus, and the condition of the hernial sac are taken into account. Identification of these pathologies is a must for diagnosis and surgical intervention.

To reduce the risks and prevent anomalies, prenatal diagnostic tools are vital. Prenatal diagnosis should be carried out in a timely manner using USG at 18 to 20 weeks of gestation focusing on the posterior fossa. At the end of the first trimester, USG allows for the identification of the hemispheres and large cisterns and more significantly in this case, changes in the shape and size of the cerebellum. Intrauterine cerebellar pathologies are almost always indicative of some type of CM. In 70 to 85% of cases, CM is accompanied by hydrocephalus or spinal hernia. Therefore, the detection of signs of pathology in the fetus should be the basis for excluding defects in the development of the spine.[14] USG is able to diagnose intracranial pathologies as well. A "banana"-shaped cerebellum (obliterated cisterna magna) with the frontal bones indented as in a "lemon" shape is almost always indicative for CM-II along with spina bifida. This should be supported by a study of the spine and the ventricles as well before drawing a conclusion. Ventricle shapes may be dynamic and change as the disease progresses. USG should be performed both in the horizontal and sagittal plane but may be difficult due to obesity or patient incompliance. In suspected cases, MRI may be employed starting from the 20th week of pregnancy.[15]

Based on the analysis of clinical and neuroimaging data of patients, the following indications for surgical treatment in children with CM-II associated with hydrocephalus and MMC were formulated:

1. "Classic" pain in the craniovertebral region, aggravated or provoked by Valsalva maneuver (with straining),
2. Persistent headaches with the ineffectiveness of conservative treatment,
3. Neurological symptoms of the level of the cervicomedullary junction,
4. The presence of syringomyelia and symptoms of myelopathy,
5. The progression of syringomyelia,
6. Syringomyelia at the cervical level and symptoms of the disease; widespread syringomyelia at the cervicothoracic level or throughout the spinal cord with a diameter of more than 6 mm (more than half the diameter of the spinal cord) in the absence of symptoms of the disease.

The presence of widespread syringomyelia with a diameter of more than 6 mm (more than half the diameter of the spinal cord) in combination with symptoms of myelopathy worsens the outcome of the disease.

Conclusion

If CM-II is asymptomatic, then no specific treatment is prescribed. If the patient develops simple pain in the occiput and neck not aggravated by Valsalva maneuver, then pain relievers are prescribed. If a patient has neurological disorders, surgical intervention is indicated. The operation is also performed if the pain syndrome cannot be overcome within the framework of conservative therapy. Indications for surgical treatment are disorders of the cranial nerves and disorders of the sensitivity of the extremities and paresis.

CM-II associated with hydrocephalus and MMC in children is a severe pathology of the central nervous system. Despite the fact that the disease is infrequent, it is this type of disease that develops in newborns during pregnancy and in the first years of life. The only definite treatment is surgery. For the purpose of timely detection of diagnostics, it is recommended to conduct ultrasound with scanning of the brain and examination of the spine in several planes, as well as conduct MRI. The comparative analysis of the results of surgical treatment of CM-II, associated with hydrocephalus and MMC in children, carried out in the study, the revealed features of the clinical and neuroimaging picture of the disease, made it possible to clarify the definition criteria and indications for surgical treatment and to optimize the choice of the posterior decompression volume to improve the immediate and long-term results of treatment. In order to prevent and correct neuropsychological complications, further retrospective clinical and neuropsychological studies are needed in pediatric patients with CM-II associated with hydrocephalus and MMC.

Ethical Approval
The ethical principles outlined by the Helsinki Declaration have been followed.

Conflict of Interest
None declared.

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