USING DIFFERENT DURAPLASTY VARIANTS IN THE TREATMENT OF PATIENTS WITH CHIARI MALFORMATION TYPE I

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The emergence of modern neuroimaging techniques has allowed for effective identification of patients with Chiari malformation. It is now possible to create a more accurate picture of this anomaly. Different surgical treatments can now be developed but this has led to standardization issues and lack of uniform recommendations for treatment of patients with Chiari malformation. A study conducted by the authors found that surgery involving the opening of the dura mater and subsequent plastic reconstruction by autoplastic graft was the optimal surgery option (with respect to volume of surgical treatment of such patients).

Keywords: Chiari malformation, Arnold–Chiari malformation, dura mater, aseptic meningitis

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ОПЫТ ИСПОЛЬЗОВАНИЯ РАЗЛИЧНЫХ ВАРИАНТОВ ПЛАСТИКИ ТВЕРДОЙ МОЗГОВОЙ ОБОЛОЧКИ В ЛЕЧЕНИИ БОЛЬНЫХ С АНОМАЛИЕЙ (МАЛЬФОРМАЦИЕЙ) КИАРИ I ТИПА

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Появление современных методов нейровизуализации позволило эффективно выявлять пациентов с аномалией Киари, сформировать более точную картину аномалии. Возникла возможность развития различных методов хирургического лечения, что в настоящее время приводит к проблемам стандартизации, а также отсутствию единых рекомендаций в лечении пациентов с мальформацией Киари. По результатам проведенного авторами исследования, оптимальной по объему хирургического лечения подобных пациентов была операция со вскрытием твердой мозговой оболочки и последующей ее пластикой аутотрансплантатом.

Ключевые слова: мальформация Киари, аномалия Арнольда–Киари, твердая мозговая оболочка, асептический менингит

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Chiari malformation is often a congenital defect, characterized by displacement of the cerebellar tonsils below the level of the foramen magnum. This leads to chronic craniovertebral junction compression, blocks the normal flow of cerebrospinal fluid (CSF) and disrupts normal blood circulation in this area (fig. 1) [1, 2].

There are currently 6 types of Chiari malformations (table) [1, 3–6]. The most common are types I and II malformations: type I is observed in 3.3–8.2 cases per 1,000 people, while type II is found in 1–2 cases per 1,000 people [1, 7].

Surgery is the main method of treatment when indicated (syringomyelia and progression of clinical symptoms).

In 1932, Van Houweninge Graftidijk reported the first attempts at surgical correction of this deformity. He tried to restore normal CSF flow at the level of the deformity by resecting the tongue of redundant tissue or by resecting the bone over the posterior surface of the malformation and incising the underlying dura. The treatment did not produce positive results [8–10]. In 1938, Penfield and Coburn described a surgical technique involving resection of the lower part of the cerebellar tonsils at this anomaly [11]. With the emergence of the Gardner and Williams concepts, surgical treatment of syringomyelia (fig. 2) and Chiari malformation became theoretically justified. The name Gardner (or "basic technique")

Types of Chiari malformations

Types	Diagnostic criteria
I	Displacement of the cerebellar tonsils below the level of the foramen magnum by more than 5 mm or presence of spinal cord syringomyelia with displacement of the cerebellar tonsils below the level of the foramen magnum by more than 2 mm
II	The cerebellar tonsils, cerebellar vermis, fourth ventricle and the medulla oblongata shift downward through the spinal canal
III	The contents of the posterior cranial fossa descend into the meningeal sac located in the occipital bone defect
IV	Cerebellar hypoplasia without shift. This Chiari malformation type is not accompanied by herniation of the CNS structures, so it is often not included in conventional classification
0	Cerebellar tonsils fill the entire cisterna magna but do not go beyond it. It is characterized by "overflowing" of the posterior cranial fossa
1,5	Cerebellar tonsillar dystopia, small stretching of the fourth ventricle and brain stem (possible slight flattening or crimping) by minimal changes from oral spinal cord sections. It is a transitional (boundary) type between types I and II, which combines features of both types of deformities, or embryologically incomplete Type II without concomitant myelodysplasia

was given to the surgery procedure described by them for correction of syringomyelia in Chiari malformation in 1965. This surgery technique is used in various modifications to this day. The operation involves suboccipital resection trepanation, anterior cervical laminectomy, dissection of arachnoid adhesions, readjustment of the foramen of Magendie and closure of the entrance to the central canal of the spinal cord with a muscle slice. The surgery involves alignment of the CSF flow pressure at the level of the craniovertebral junction [12, 13].

Currently, Gardner's surgery — in its classical form — is performed rarely. The proposed modifications for this surgery are performed in two ways. The first type is resection trepanation of the posterior cranial fossa, dissection of the dura mater and arachnoid mater with or without resection of the cerebellar tonsils with or without cisterna magna reconstruction. The second type is resection trepanation of the posterior cranial fossa with the opening of the dura mater and manipulations on it [14–17].

There are also other surgery types: Endoscopic third ventriculostomy (ETV), transoral decompression and craniovertebral decompression with occipitospondylodesis [5].

Performance of ETV as a first stage in the treatment of patients with Chiari malformation type I (CM-I) and concomitant hydrocephalus is currently recognized as the gold standard, displacing the previously used shunt system. According to researchers [6], the procedure is up to 95 % effective. There is a small group of CM patients suffering directly from intracranial hypertension symptoms for whom surgical treatment can be limited to only ETV [6]. However, craniovertebral junction compression is subsequently required for most patients with Chiari malformation and concomitant hydrocephalus.

Transoral decompression in CM patients should be used in cases of severe anterior compression and basilar invagination. Most scientists believe that given the injury rate of this method, it is reasonable to use the standard craniovertebral decompression with a one-time stabilizing surgery as the first stage of surgical treatment for patients with a combination of anterior and posterior compressions [18].

One-time performance of craniovertebral decompression and stabilizing surgery is indicated in a relatively small group of patients with CM-I, atlantoaxial dislocation and high risk of cervical spine instability revealed at the pre-surgical examination stage. The provoking factors for spinal instability in patients with CM-I are disruption of neck muscle innervation amidst syringomyelic cysts in the upper cervical level, muscular fibrosis repeating their compression and stretching and improper surgical wound closure [18, 19].

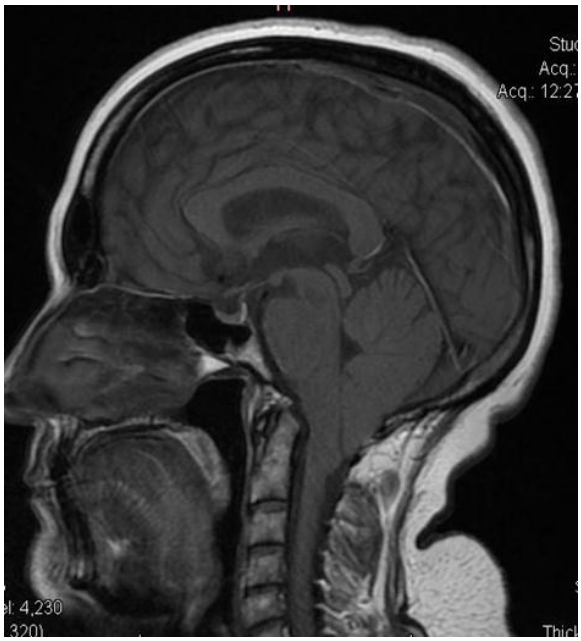

Fig. 1. A T1-weighted sagittal MRI scan, from a patient with Chiari malformation, cerebellar tonsillar prolapse

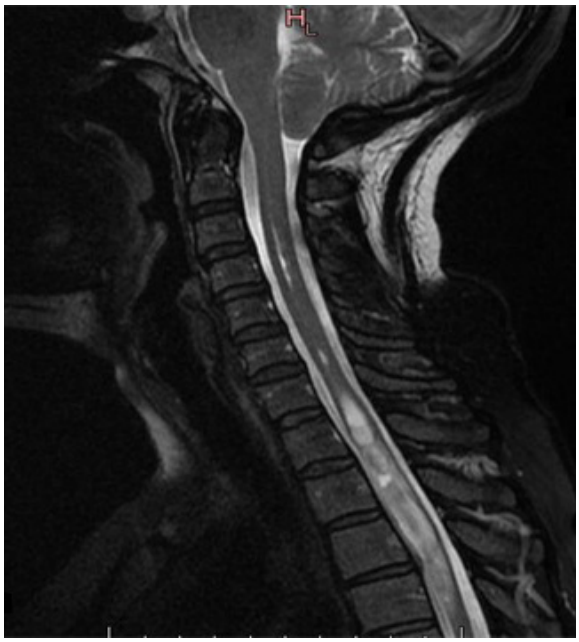

Fig. 2. A T1-weighted sagittal MRI scan. Syringomyelia — disorder often associated with Chiari malformation

The purpose of the surgery is to decompress the craniovertebral junction in order to free the flow of CSF and blood circulation at this level by easing CSF outflow from the foramen of Magendie and cisterna magna to the spinal subarachnoid space.

There are a number of post-op complications in duraplasty. In our study, we decided to focus on two main complications – aseptic meningitis and pseudomeningocele.

Aseptic meningitis is an inflammatory response by the meninges. It differs from purulent postoperative meningitis by the absence of pyogenic pathogen diagnosed through microscopic examination. This syndrome has a characteristic clinical picture (pyrexia, meningeal symptom, inflammatory changes in the blood and cerebrospinal fluid), which is not significantly different from that of purulent postoperative meningitis. Aseptic meningitis syndrome is accompanied by immunologic phenomena of transient postoperative reactive inflammation associated with lymphocyte activation [20].

Pseudomeningocele is a pathological extradural accumulation of cerebrospinal fluid in the soft tissues, communicating with the subdural space through the dura mater defect (Fig. 3).

There are also a number of other post-operative complications: accumulation of lamellar subdural hygroma in the cerebellar hemisphere, pneumocephalus and arachnoiditis. The patient's sitting position during surgery also leads to such intraoperative complications as air embolism.

Research objective: To determine the role of duraplasty in Chiari malformation surgeries.

Tasks:

– to ascertain the incidence of various clinical symptoms in Chiari malformations;

– to assess the incidence of post-op complications depending on the surgical technique;

– to assess the response to surgical treatment under various duraplasty techniques.

METHODS

The study included 34 patients with Chiari malformations. They were treated at the neurosurgical department of Interregional Clinical and Diagnostic Center in Kazan from 2010 to 2014.

Clinical method, preoperative brain imaging, direct intraoperative imaging and retrospective analysis were all deployed for the study.

All the patients underwent neurological examination in the preoperative and early postoperative periods. The basic neurological functions were assessed.

Preoperative imaging consisted of brain MRI (capturing the craniovertebral junction) and spine MRI along the length depending on the syringomyelia level. MRI scan was performed using the Signa HDxt 1.5T magnetic resonance scanner (General Electric, USA) with at least 1.5 T magnetic field intensity in the axial, sagittal and coronal projections, as well as in hydrography regime to assess the CSF flow dynamics in the craniovertebral junction.

The main criteria for Chiari malformation diagnosis were neuroimaging data (fig. 4) [21–23] and clinical examination based on identification of specific clinical syndromes (cerebellar, hypertension-hydrocephalic, syringomyelic, radicular, bulbar and vertebrobasilar insufficiency syndromes [24]). In most CM-I cases, MRI revealed descent of the cerebellar tonsils below the McRae line, syringomyelia and brain stem compression. In Chiari Malformation type II (CM-II), descent of the cerebellar tonsils, Z-shaped deformation of the medulla oblongata, quadrigeminal adhesion (beak-shaped bend in the quadrigemina), medulla elongation and low brain attachment were also detected. In CM-II cases, MRI revealed hydrocephalus, syringomyelia in the craniovertebral junction, isolated fourth ventricle, cerebellar-medullary compression, and agenesis/ dysgenesis of the corpus callosum [5].

The average age of the patients was 45 years (from 18 to 69 years). The ratio between number of men and female was 1 : 3.2 respectively. It is worth noting that the most common surgical procedure was performed in patients aged 50–60 years. Neurological symptom progression was the indication for surgery.

The surgery procedures were carried out in a sitting position. The soft tissue was dissected according to Naffziger-Towne method (Babchin's modification). In the next step, the lower parts of the squamous part of occipital bone were resected, C1 laminectomy and sometimes C2 was carried out. All the patients were divided into two groups based on the extent of further intervention.

The first group included 8 (23.5 %) patients, whose dura mater was not opened. In this case, distinct pulsation of dura mater could be seen.

The second group had 26 (76.5 %) patients, whose dura mater was opened with a Y-shaped incision. In most cases, the dura mater turned out to be thickened and it did not pulse. Venous sinuses expanded quite often. After dissection of the dura mater, the level of descent of the cerebellar tonsils and subarachnoid space was examined and assessed. In the case of pronounced adhesions, the subarachnoid space was readjusted with release from adhesions of vessels, medulla and cerebellar tonsils. Next, duraplasty was performed to restore the integrity of the dura mater. The second group was divided into two subgroups depending on the type of duraplasty performed:

– 2A: 14 (41.2 %) patients — the dura mater was opened followed by allograft duraplasty surgery. Artificial dura mater DURAFORM (Codman Neuro, USA) was used as the allograft.

– 2B: 12 (35.3 %) patients — the dura mater was opened

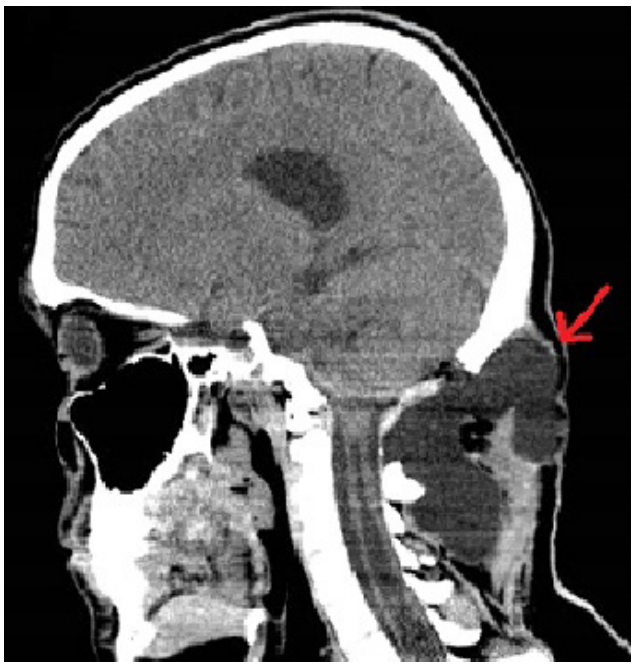

Fig. 3. A sagittal CT scan. Pseudomeningocele (marked by arrow) — one of the post-op complications from Chiari malformation surgery

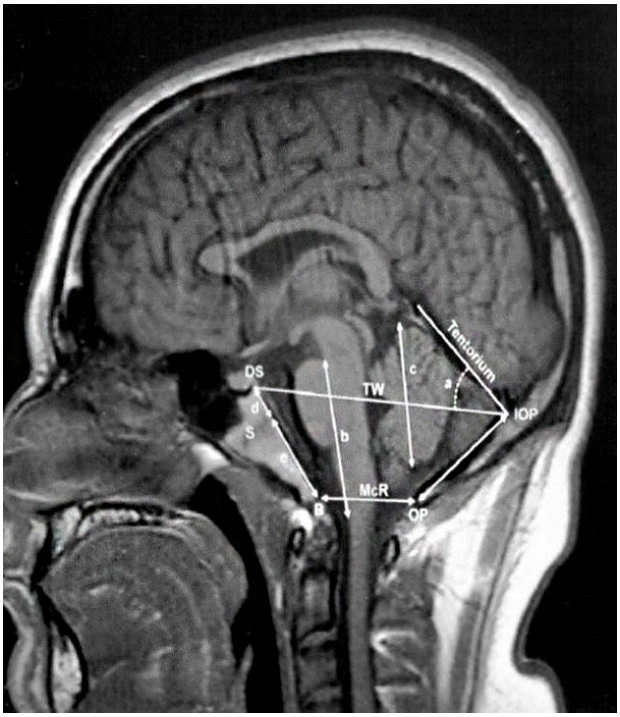

Fig. 4. Reference points of the posterior fossa used in diagnosis of Chiari malformation (Sekula et al., 2005 [21])

d + e — length of clivus; S — sphenooccipital synchondrosis; d — length of basisphenoid between the top of the dorsum sellae and the sphenooccipital synchondrosis of the clivus; e — length of the basiocciput between the synchondrosis and the basion; b — length of the hindbrain between the midbrain-pons junction and the medullocervical junction; a — angle of the cerebellar tentorium to Twining's line; c — length of cerebellar hemispheres; DS — top of the dorsum sellae; IOP — internal occipital protuberance; OP — opisthion; IOP to OP — length of supraocciput; B — basion; TW — Twining's line; McR (B to OP) — McRae's line.

followed by allograft duraplasty surgery. A previously marked occipitalis muscle aponeurosis served as the allograft. On the access point, the soft tissues were dissected up to the occipitalis muscle aponeurosis. The aponeurosis was cut off as a 3 × 5 cm flap, and leaned back to the side. The base of the flap was directed towards the occipitalis muscle. Thus, the aponeurosis flap remained on the pedicle via which it was supplied with necessary nutrients till the end of the operation. Afterwards, dissection was carried out linearly like in other groups. At the duraplasty stage, the pedicle was cut off, and the flap was sutured to the edges of the opened dura mater. So the cisterna magna was increased, which resulted in more craniovertebral junction decompression.

Resection of the cerebellar tonsils was not performed in any of the patients.

Response to surgery in the early postoperative period was analyzed. During comparison, neurological regression, incidence of postoperative complications resulting from duraplasty inefficiency, and dependence on duraplasty technique were first factored in.

Statistical data analysis was performed using Microsoft Office Excel 2010.

RESULTS

Analysis of the medical records of patients with Chiari malformation revealed the incidence of neurological syndromes identified during preoperative clinical examination (fig. 5). Cerebellar syndrome was detected in majority of the cases (88.2 %). Bulbar syndrome and vertebrobasilar insufficiency syndrome were the least observed (in less than half of the patients).

Clinical method and postoperative brain imaging were used in analyzing the response to surgical treatment. Neurological examination took into account regression of the characteristic clinical symptom, whose presence criterion included the following: no complaints from the patient, sensitivity improvement under the corresponding dermatitis and increase in strength in the limbs, reduction in cerebellar symptoms and signs of intracranial hypertension.

Response to treatment was assessed in the early and late post-op periods. After full treatment, all the patients showed a positive response to treatment.

Hypertension-hydrocephalic syndrome regressed faster than others. Within the first days after surgery, characteristic neurological symptoms of intracranial hypertension disappeared in the patients. Within 3–4 months, there was complete or partial disappearance of bulbar, cerebellar and syringomyelic syndromes. Restoration of sensitivity and muscle strength was observed. MRI scans showed that syringomyelic cysts disappeared within 6 months to 1 year.

The following results were obtained (fig. 6).

In the group of patients who underwent craniovertebral junction decompression followed by duraplasty, neurological syndromes regressed in 21 (80.8 %) out of the 26 patients within the first 20 days of surgery: in the first subgroup with allograft duraplasty surgery — in 11 (79 %) out of 14 patients; in the second subgroup with allograft duraplasty surgery — in 10 (83 %) out of 12 patients.

A time limit of 20 days was chosen arbitrarily according to the results obtained from data study.

In the group of patients who underwent craniovertebral junction decompression transition without opening of the dura mater, regression of neurological syndromes in 75 % of cases occurred no earlier than one month from the date of the surgery. In 2 (25 %) out of 8 patients, the main syndromes regressed within the first 20 days.

Post-op complications associated with inefficiency of duraplasty — aseptic meningitis and pseudomeningocele — were found only in patients who underwent craniovertebral junction decompression with the opening of dura mater and subsequent allograft duraplasty surgery (42.9 %). Moreover, of all the patients from this subgroup (n = 14), aseptic meningitis was observed in 5 (35.7 %) patients in the postoperative period, and pseudomeningocele in 1 (7.1 %) patient.

In the subgroup of patients who underwent craniovertebral junction decompression with the opening of dura mater and subsequent allograft duraplasty surgery, the above-mentioned post-operative complications were not observed.

DISCUSSION

Identifying an efficient surgical correction method and standardizing this method is an important task since surgery is the primary treatment for patients with this deformity. However, the decision on extent of surgery in some cases cannot be standardized, and any addition to the main surgery procedure should be determined intraoperatively. The main goal of surgery is to decompress the craniovertebral junction and restore normal flow of cerebrospinal fluid in the area.

In a number of cases, bone decompression is enough to achieve the surgery target. This is evidenced by a clear pulsation of the dura mater, but the sole criterion cannot provide the required reliability. Therefore, we believe that somatosensory evoked potentials should be used intraoperatively. This method

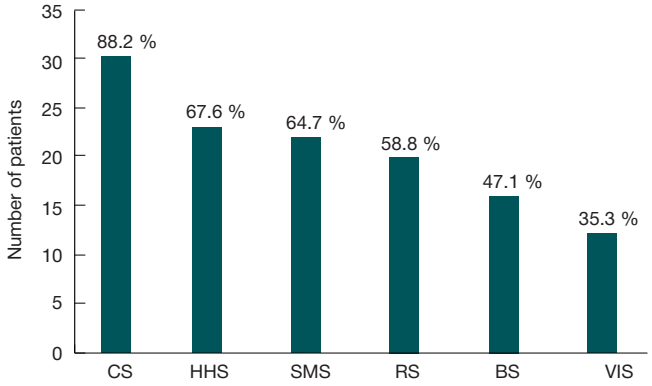

Fig. 5. Incidence of syndromes in Chiari malformationMC — мозжечковый синдром

CS — cerebellar syndrome; HHS — hypertension-hydrocephalic syndrome; SMS — syringomyelic syndrome; RS — radicular syndrome; BS — bulbar syndrome; VIS — vertebrobasilar insufficiency syndrome.

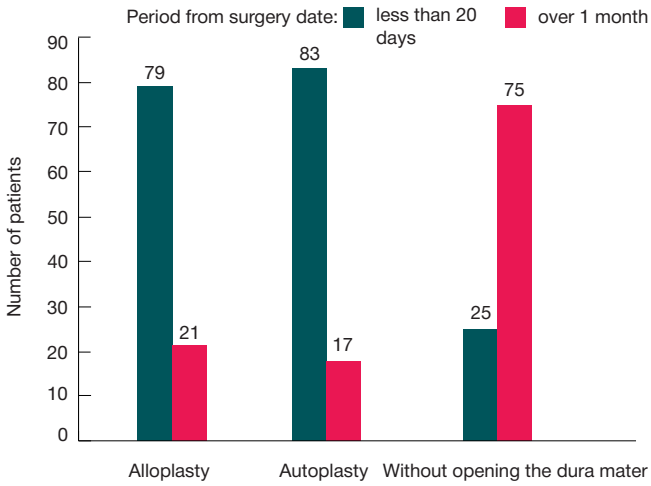

Fig. 6. Duration of neurological syndrome regression under different types of surgery

gives fairly accurate information on possible post-operative regression. The evidence of this includes intraoperative results obtained by us, namely the positive dynamics in 2 out of 8 patients who did not undergo opening of the dura mater. These 2 patients from the first group showed the fastest regression of neurological symptoms.

Our study found that surgery with the opening of the dura mater and subsequent allograft duraplasty surgery is the most optimal extent of surgical treatment of type 1 Chiari malformation. This finding is consistent with the data from world literature [25–27].

CONCLUSIONS

Based on the outcome of surgical treatment of patients with Chiari malformation type I, it can be concluded that craniovertebral junction decompression with the opening of the dura mater and subsequent sealing with autograft is an effective method for surgical treatment of patients with this deformity. This method reduces the risk of post-op complications. The relative majority of patients with the most rapid regression of neurological symptoms serve as a proof that this method is effective.

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