Results of Surgical Treatment of Syringomyelia Associated with Chiari 1 Malformation. An Analysis of 125 Cases

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The rate of Chiari malformation (CM) ranges from 3 to 8 per 100,000 population. In 62—80% of cases, CM is accompanied by the development of syringomyelia (SM) at various levels. The clinical picture in these patients is a combination of CM and SM manifestations. However, SM symptoms often prevail, which creates some problems in the disease diagnosis and in the choice of optimal treatment. Objective. On the basis of our own experience of surgical interventions, we aimed to clarify the indications for surgical treatment of SM associated with CM and to define the optimal extent of surgery and criteria for evaluation of treatment outcomes. Material and methods. Two hundred twenty five patients with a combination of syringomyelia and Chiari 1 malformation were examined in the period from 2011 to February 2015. Of them, 125 patients were operated on. The mean age of the operated patients was 36.8 years. The mean time from the appearance of the first signs of the disease to surgery was 75.8±2 months. All operations were performed by a single surgeon. All operations were carried out with the patient in the semi-sitting (89.6%) or prone (10.4%) position. The operation included sparing suboccipital craniectomy, C1 arch resection, restoration of cerebrospinal fluid (CSF) circulation along the posterior surface of the cerebellum, and reconstruction of the dura mater (DM) in the craniovertebral junction region. Results. Exploration of the arachnoid mater of the cisterna magna after dura opening revealed no arachnopathy in 78 (62.4%) patients (Chiari 0 malformation according to Klekamp). Type 1 arachnopathy (by Klekamp) was detected in 31 patients (24.8%), and type 2 arachnopathy was observed in 16 (12.8%) cases. The condition of 109 (88%) patients was evaluated one year after the surgery. Sixty one (56%) patients had partial or complete regression of the preoperative neurological symptoms. The disease stopped progressing in 44 patients (40%). The disease was worsened in 4 (3.7%) patients. No recurrence of a CSF circulation disturbance at the craniovertebral level was observed during follow-up. Early postoperative complications occurred in 4 (3.2%) patients: wound CSF leakage in 1 (0.8%) patient, acute epidural hematoma in 1 (0.8%) patient, and aseptic meningitis in 2 (1.6%) patients. Temporary deteriorations in the condition (headache worsening, meteosensitivity) were detected in 11 (8.9%) patients. The symptoms regressed by the end of the 1st postoperative month. There were no deaths. Conclusions. The indication for surgery in patients with a combination of CM and SM is the presence of neurological symptoms associated with syringomyelia and their progression as well as headache caused by herniation of the cerebellar tonsils that significantly deteriorates the patient’s quality of life. The main criteria for evaluating the treatment efficacy include stabilization of the clinical symptoms and/or an improvement in the patient condition. Suboccipital craniectomy followed by DM reconstruction and restoration of CSF circulation in the craniovertebral region is an effective treatment for syringomyelia associated with Chiari 1 malformation. Keywords: syringomyelia, Chiari malformation, CSF circulation, suboccipital decompressive craniectomy.

Chiari malformation (CM) and syringomyelia (SM) are two diseases that occur independently of each other. However, they often occur in combination, which may fundamentally change the treatment strategy. According to various authors, the CM rate ranges from 3 to 8 per 100,000 population; in 62—80% of cases, CM is accompanied by the development of SM at various levels of the spinal cord. The clinical picture in these patients is a combination of CM and SM manifestations. However, the SM symptoms often prevail, which creates some problems in the disease diagnosis and in the choice of optimal treatment.

To date, various approaches for treating these diseases have been reported in the literature that are based on theories explaining causes of the SM development and progression. However, none of the theories can fully explain all clinical and pathophysiological aspects of this condition. Therefore, further investigation of the pathogenesis of cyst formation in the spinal cord is required. The most popular and widespread theory of development of CM-associated SM is a theory by W. Gardner (1950) that is based on the assumption that obstructed CSF outflow from the cisterna magna into the spinal subarachnoid space leads to hydrodynamic shocks of the CSF systolic wave from the IVth ventricle to the central canal walls, which results in the canal expansion and formation of a syringomyelic cavity. On this basis, restoration of CSF circulation in the craniovertebral junction region is the main goal of surgical treatment. However, there are various unresolved problems in this field: what are the symptoms for choosing a surgical technique; what treatment outcomes should be considered satisfactory; how to treat patients with CM, SM, and basilar impression, etc.

On the basis of our own experience of surgical treatment of these patients, we tried to define the optimal extent of surgery and criteria for assessing treatment outcomes and to answer some arisen questions.

Material and Methods

A total of 225 patients with a combination of syringomyelia (SM) and Chiari malformation type 1 (CM1) were examined in the period from 2011 to February 2015. Of them, 125 patients (52 males and 73 females) were operated on. The indication for surgery in all patients was the presence of neurological symptoms associated with SM, their progression, and headache caused by cerebellar tonsillar herniation that significantly deteriorated the patients’ quality of life. The presence of CM and SM in the absence of clinical manifestations of the diseases was not an indication for surgery, regardless of the depth of
cerebellar tonsillar herniation as well as the size and localization of a syringomyelic cyst. These patients received supportive conservative treatment and were followed-up (100 patients in our study).

The mean age of operated patients was 56±8 years; the mean time from the appearance of the first signs of the disease to surgery was 75±82 months. A thorough neurological examination of the patients was carried out before surgery; postoperative examinations were performed every 6—12 months. The maximum follow-up duration was 3.8 years; the minimum duration was 6 months (median, 1.4 years). The disease manifested as dissociative disorders of sensation in 65% of patients, cervico-occipital pain in 37.1% of cases, pyramidal symptoms in 8.9% of cases, and hypertension symptoms in 1.6% of patients. Most patients had all of the listed symptoms at the time of hospitalization. All patients underwent preoperative MRI of the brain and entire spinal cord; further examinations were performed every 6—12 months. Some patients with a concomitant craniovertebral region anomaly (basilar impression, platybasia, C1 assimilation, etc.) underwent computed tomography (CT).

Preoperative phase-contrast MRI with cardiac synchronization was conducted in 23 (18.4%) patients. This examination was needed to confirm a block of CSF flow in the craniovertebral region. The examination was especially indicated for patients who had previously undergone decompression in order to decide whether the repeated procedure was advisable.

All operations were performed by a single surgeon with the patient in the semi-sitting (89.6%) or prone (10.4%) position. A 4—5 cm skin incision in the occipital region was followed by suboccipital craniectomy of up to 3 cm in diameter and C1 laminectomy (Fig. 1).

The dura mater was opened with a Y-shaped incision. The arachnoid mater of the cisterna magna was maximally preserved during DM opening. Then, DM was inspected for adhesions. If arachnopathy (type 1 and 2, by Klekamp [11]) signs were detected, the cisterna magna was opened, adhesions between the arachnoid mater and the cerebellum, medulla oblongata, and spinal cord were dissected, CSF circulation along the posterior surface of the cerebellum was restored, and the foramen of Magendie was inspected (Fig. 2).

In patients with pronounced arachnoid scars, scar dissection was limited to the midline to provide a connection between the intracranial subarachnoid space and the spinal cord space. Adhesions on the lateral surface of the medulla oblongata and spinal cord were not dissected to avoid damage to the structures or small blood vessels (Fig. 3).

If the cerebellar tonsils descended to the C2 level and below, the tonsils were subpially resected. At this stage, some patients who had isolated dilation of the fourth ventricle were implanted with a shunt going from the IVth ventricle through the foramen of Magendie (most often obliterated by adhesions in this situation) to the subarachnoid space of the cisterna magna. Then, reconstruction of the dura mater in the craniovertebral junction region was performed. For this purpose, we used artificial dural substitutes (Gore, DuraForm, SeamDura, DuraPair, etc.) in all patients. This material was used to reduce the risk of recurrence of CSF circulation disturbances due to postoperative adhesions between the dura mater and the cerebellar surface. Some patients with an appropriate thickness of the occipital bone underwent reconstruction of a defect with a titanium implant. This reduced the adhesion process outside the dura mater and eliminated compression of the arachnoid space by swollen muscles (Fig. 4).

Particular attention was paid to careful muscle suturing. A small, 4—5 cm, incision, not reaching the external occipital protuberance, allowed for complete wound sealing with a muscular layer.

In the early postoperative period, most patients underwent brain MRI to rule out hemorrhagic and ischemic foci. Further examinations were carried out after 4—6 months, 6—12 months, and then annually. The clinical symptoms and surgical treatment outcomes were evaluated in the same period. Because

Figure 1. The extent of adequate bone resection for Chiari 1 malformation. A CT scan of the skull bones and subsequent 3D reconstruction.
progression of clinical disease symptoms was the indication for surgery, treatment outcomes were considered as satisfactory in the case of stabilization or improvement of the patient’s condition.

**Results**

Chiari 1 malformation is often associated with other malformations of the craniovertebral region. In our series, apart from the descended cerebellar tonsils, 56 (44.8%) patients had hyperostosis of the occipital bone, platybasia, basilar impression, C1 assimilation, and a Klippel–Feil anomaly.

Exploration of the arachnoid mater of the cisterna magna after dura opening revealed no arachnopathy in 78 (62.4%) patients (Chiari 0 malformation according to Klekamp); thus, no opening of the arachnoid mater was required. Type 1 arachnopathy (by Klekamp) was detected in 31 patients (24.8%), and type 2 arachnopathy was observed in 16 (12.8%) cases. These patients underwent dissection of adhesions and restoration of CSF circulation along the posterior surface of the cerebellum and spinal cord. In 12 (9.6%) cases, isolated dilation of the IVth ventricle required forth ventriculosubarachnoid shunting. The cerebellar tonsils were resected in 6 (4.8%) patients if the tonsils descended to the C2 level and below and significantly impeded CSF circulation.

Two (1.6%) patients with concomitant hydrocephalus underwent, apart from suboccipital decompression, one-stage ventriculoatrial shunting and the major surgery.

Two (1.6%) patients with concomitant basilar impression underwent one-stage transnasal endoscopic resection of the dens, suboccipital craniectomy, C2 arch resection, and occipitospondylosis.

Control MRI 4 months later revealed that SM resolved in 19 (15.3%) patients, decreased in 89 (71.8%) cases, and remained unchanged in 16 (12.9%) patients (Fig. 5 and 6).

The condition of 109 (88%) patients was evaluated one year after the surgery (77 patients were present, while the condition of the remaining 32 patients was evaluated remotely). A total of 61 (56%) patients had partial or complete regression of the preoperative neurological symptoms. The disease stopped progressing in 44 (40%) patients. The disease was worsened in 4 (3.7%) patients only. Cardiac-synchronized phase-contrast MRI of the craniovertebral junction region was performed in these patients 4–12 months after suboccipital decompression to evaluate CSF circulation. No signs of a CSF flow block were detected. Therefore, the patients underwent

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**Figure 3. An intraoperative image.**

Adhesiotomy (type 2 arachnopathy) and detachment of DM from the brain along its posterior surface (indicated by red arrows) were performed. The foramen of Magendie is opened, a ventriculocisternal shunt is placed through the foramen into the IVth ventricle cavity (blue arrow).
syringosubarachnoid shunting for stabilization of neurological symptoms (Fig. 7).

No recurrence of CSF circulation disturbances at the craniovertebral level was observed in the operated patients during follow-up.

Early postoperative complications occurred in 4 (3.2%) patients: 1 (0.8%) patient had wound CSF leakage; 1 (0.8%) patient had an acute epidural hematoma; and 2 (1.6%) patients had aseptic meningitis (DM in both patients was reconstructed with a dural substitute of the same brand). Temporary deteriorations (headache worsening, meteosensitivity) were observed in 11 (8.9%) patients. These symptoms regressed by the end of the 1st postoperative month.

Pseudo-meningocele signs, which were not accompanied by any neurological symptoms and cosmetic disturbances and did not require changes in the treatment approach, were detected during control MRI of the brain in 3 (2.4%) patients. There were no deaths.

Discussion

Given the affordability of MRI in our country, diagnostics of CM and SM is not problematic. However, these conditions are often overdiagnosed. This is due to the fact that, to date, the value of a clinically significant displacement of the cerebellar tonsils is still disputed. According to most experts, a 5 mm or larger displacement in adult patients is considered as pathology, whereas this displacement may be physiological in children due to cerebellum growth. A disturbance of CSF circulation at the craniovertebral level can also be caused by a bone abnormality in the absence of a cerebellum displacement (Chiari malformation type 0, first described by B. Iskandar in 1998 [4―6]). In these controversial cases, phase-contrast MRI and assessment of CSF circulation in this region are recommended [7].

Given the diversity of disease clinical manifestations, it is important to identify the main symptoms and assess their dynamics. According to many authors, the presence of CM and pronounced SM in the absence of clinical symptoms and signs of disease progression is not an indication for surgery.

Despite the fact that suboccipital decompression is widely used for treating CM1, there is no generalized algorithm of this manipulation. Surgery proposed by W. Gardner included extensive craniectomy of the posterior fossa, opening of the IVth ventricle, and plugging the obex with a piece of muscle; the dura mater was not closed. The author reported five fatalities after 74 operations. Similar rates of postoperative mortality and patient condition deterioration have had other surgeons to use less invasive procedures, such as preservation of the intact arachnoid mater after dura opening, incision of only the outer dura layer, or bone decompression alone. In our opinion, the optimal and most accepted surgical technique is that described above. However, the decision on the extent of surgery is made by the neurosurgeon for each particular case based on the disease pathogenesis in a particular patient. Availability of MRI scans enables analyzing disease, allowing for all anatomical peculiarities, and planning surgery. The complexity of decision making is related primarily to the rarity of these diseases.

We used only artificial dural substitutes for DM reconstruction. In our opinion and according to J. Klecamp,
this reduces the risk of recurrence of CSF circulation disturbances in the craniovertebral region due to adhesions.

According to the modern concepts, drainage of a syringomyelic cyst is a temporary symptomatic procedure. This operation may cause secondary fixation of the spinal cord and SM, and, therefore, it should be used as rarely as possible. In our series of observations, this procedure was required only in 4 patients with SM progression. A decision on syrinx shunting should be made no earlier than 4 months after suboccipital decompression. In the case of normalization of CSF circulation at the craniovertebral level, this time is enough to evaluate clinical symptoms that are the main criterion for the assessment of treatment results. Despite adequately performed decompression, a syringomyelic cyst may not change or reduce for this period.

The disease rarity and, as a result, low awareness of neurologists and neurosurgeons on its pathogenesis, treatment options, and possible outcomes lead to the situation where some patients are not operated on for years and, finally, become profoundly disabled. On the other hand, we often face consequences of undue surgeries performed in the absence of indications for them or surgeries performed incompletely. As a result, some doctors conclude inoperability or meaninglessness of surgical treatment. As our study shows, identification of the indications for surgery and appropriate surgical treatment provide good outcomes with the minimum complication rate and zero mortality in more than 90% of patients.

**Conclusion**

A set of surgical procedures, including suboccipital craniectomy, C1 arch resection, and subsequent DM reconstruction and restoration of CSF circulation in the craniovertebral region, which are performed on time and according to indications, is an effective approach for treatment of SM associated with CM1.

The indications for surgery include the presence of SM-associated neurological symptoms, their progression, and/or headache caused by herniation of the cerebellar tonsils and significantly deteriorating the patients’ quality of life.

The essential extent of surgery includes economic resection of the occipital squama (up to 3 cm), C1 arch resection, exploration of the subarachnoid space of the cisterna magna, and dissection of arachnoid adhesions, if present, for restoration of CSF circulation along the posterior surface of the cerebellum, followed by expansive DM reconstruction using artificial dural substitutes in the craniovertebral junction region.

The main criteria of the treatment efficacy in patients with combined pathology, CM1 and SM, include stabilization of the clinical symptoms and improvement of the patient condition.

**Authors declare no conflict of interest.**
The article presents a clinical study of outcomes of surgical treatment in 125 patients with a combination of Chiari malformation and syringomyelia performed in the period between 2011 and 2015. The indication for surgery included neurological symptoms associated with syringomyelia, their progression, and also headache resulting from cerebellar tonsillar herniation and significantly affecting the patient’s quality of life. The authors performed a detailed analysis of the clinical and MRI semiotics of this group of patients and described a surgical technique that typically involved suboccipital craniectomy of the foramen magnum (about 3 cm in diameter) and C1 laminectomy, a Y-shaped incision of the dura mater with subsequent exploration of the arachnoid mater and evaluation of arachnopathy severity (according to Klekamp), which was the basis for deciding on opening the cisterna magna. If the cerebellar tonsils descended to the C2 level and below, subpial resection of the tonsils was performed. All patients underwent DM reconstruction in the craniovertebral junction region using artificial dural substitutes (Gore, DuraForm, SeamDura, DuraPair, etc.). Some patients were also implanted with syringosubarachnoid and ventriculosubarachnoid shunts. The condition of 109 (88%) patients was evaluated one year after the surgery. The disease stopped progressing in 44 (40%) patients; 61 (56%) patients had a partial or complete regression of the preoperative neurologic symptoms. The disease was progressing only in 4 (3.7%) patients. Control MRI 4 months later revealed that syringomyelia disappeared in 19 (15.3%) patients, decreased in 89 (71.8%) cases, and remained unchanged in 16 (12.9%) patients. The authors also evaluated the rate of postoperative complications, which was 3.2%. The authors conclude that suboccipital craniectomy, C1 arch resection, and subsequent DM reconstruction and restoration of CSF circulation in the craniovertebral region, performed on time and according to the indication, to be an effective treatment for syringomyelia associated with Chiari malformation type 1.

In our view, this work is a valuable and topical study since it presents a detailed description of the data obtained in a large, for domestic literature, group of 125 patients. Also, high quality of acquisition of the epidemiological and clinical data should be mentioned.

A significant achievement of the study is unification of the indications for surgery, surgical technique, and approach for postoperative data analysis. Thus, the authors demonstrated the efficacy of surgical treatment in a rather homogeneous group of patients. The technical aspect of performed interventions does not raise any questions since the intervention is based on the classic approach to surgery for Chiari malformation, which stresses the efficacy and relative safety of the approach. The clinical and magnetic resonance results of surgery 4 and 12
months after operations and a low rate of complications indicate high expertise and surgical skills of the authors.

Thus, most questions to the article are not methodological but rather conceptual and are related to the ambiguity of approaches to surgical treatment for Chiari malformation.

For example, the authors performed reconstruction of the dura mater in all patients, without exception. This is the rule of thumb approach, but if it is pathogenetically reasonable in all cases? In our practice, we routinely use ultrasound to decide whether dura reconstruction is required. We believe that a noninvasive examination of volume relationships in the craniovertebral region as well as of CSF circulation and the blood flow velocity in the straight sinus using transcranial dopplerography is a highly effective and safe method to identify preoperatively a leading pathophysiological syndrome (compression, CSF circulation, or mixed type disturbance). Identification of this syndrome in the clinical diagnosis provides the surgeon with information about a possible surgical approach: DM reconstruction using a dural autograft (local aponeurotic flap) is suitable in the case of obstructed CSF circulation and mixed disturbances, while bone decompression is essential in the case of compression disorders. Intraoperative ultrasound allows ensuring the adequacy of decompression and, if necessary, modifying an intervention approach. Probably, evaluation of this parameter would enrich the study with practical information.

The use of shunt surgery in patients with syringomyelia is associated with some questions. According to the monograph “Syringomyelia” by Jörg Klekamp, an acknowledged leader in the treatment of CSF circulation disorders, the failure rate of syringosubarachnoid and ventriculosubarachnoid shunting in patients with syringomyelia and arachnopathy is 92—100%; at the same time, Klekamp says that the technique can be used in particular cases (apparently, the authors were guided by this note). It would be interesting to give more attention to the efficacy/inefficacy of shunting in the present study.

Finally, the surgical technique used by the authors is also debatable. For instance, in our practice, we often use endoscopic techniques, including flexible endoscopy of the subarachnoid space (thecaloscopy), endoscopic cyst fenestration, and an endoscopic portal approach to the foramen magnum in some cases. Undoubtedly, the choice of a surgical technique is the exclusive prerogative of the surgeon, and this is why the uniform surgical approach used in the study is both an advantage and disadvantage of the work.

Thus, we believe that the article “Results of surgical treatment of syringomyelia associated with Chiari 1 malformation. An analysis of 125 cases” to be topical and highly informative work. The issues that it raises are absolutely reasonable and reflect the complexity and diversity of common pathologies, such as Chiari malformation, syringomyelia, and arachnopathy. Discussions in the neurosurgical society induced by similar studies should encourage us for generalization of the available data and continuous analysis of surgery outcomes.

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