doi: 10.17116/engneiro201579653-60

Cerebrovenous Orthostatic Reactivity in Craniovertebral Junction Pathology (Chiari Malformation)

V.A. SHAKHNOVICH, E.V. MITROFANOVA, V.N. SHIMANSKIY, N.A. KONOVALOV, A.N. SHKARUBO

Burdenko Neurosurgical Institute, Moscow, Russia

Objective. Chiari malformation is characterized by herniation of the cerebellar tonsils into the foramen magnum, which leads to disturbance of CSF circulation through the craniovertebral junction. Orthostatic stress, which makes CSF flow through the craniovertebral junction, is an adequate method to detect these disorders. It is accompanied by changes in intracranial pressure, affecting the cerebrovenous orthostatic reactivity (CVOR), which is non-invasively assessed in patients with Chiari malformation. **Material and Methods.** The study involved 35 patients with Chiari malformation (26 patients with Chiari I and 9 patients with Chiari II malformation) aged 4 to 58 years. Hydrocephalus and syringomyelia was diagnosed in 4 and 6 examined patients, respectively. Transcranial Doppler sonography was used to record the venous blood flow in the straight sinus of the brain while changing body position on the tilt table from +90° to –30°. **Results.** There is significant CVOR abnormality in most patients with Chiari malformation (more than 90%), which is characterized by either increased CVOR (sometimes 5—6-fold compared to the upper normal level (considerable hyperreactivity) or complete absence of any changes during orthostatic load (nonreactivity). Before surgical treatment, CVOR of patients with Chiari malformation is often characterized by nonreactivity, as well as a moderate or significant hyperreactivity. After surgical treatment (foramen magnum decompression), patients with Chiari malformation demonstrate significant normalization of the craniovertebral volume ratios and CVOR is often characterized by normoreactivity (in 63%) or, more rarely, moderate hyperreactivity. The venous flow velocity in the straight sinus of the brain in patients with Chiari malformation can be increased before the surgery and is normalized after surgery. Conclusions. The high incidence of disturbance of CVOR (over 90%) was revealed in patients with Chiari malformation. After surgical treatment, CVOR was completely normalized in more than half of these patients (63%).

Keywords: *venous brain circulation, craniovertebral junction, Chiari malformation.*

Abbreviations

ICP — intracranial pressure;
CVVRs — craniovertebral volume ratios;
CVJ — craniovertebral junction;
CM — Chiari malformation;
VBFV — venous blood flow velocity;
TCD — transcranial Dopper (sonography);
CVOR — cerebrovenous orthostatic reactivity.

The craniovertеberal junction (CVJ) consists of two upper cervical vertebrae and the adjacent regions of the skull base. The group of appreciably common types of CVJ pathologies includes Chiari malformation (CM): descent of the cerebellar tonsils into the foramen magnum causing compression of the medulla oblongata. In the norm, the cerebellar tonsils lie above the foramen magnum. In patients with CM, cerebellar tonsils descend to the level of the first or sometimes the second cervical vertebrae, thus blocking the CSF flow. Hence, CM is characterized by a disproportion between the volume of the contents of the posterior cranial fossa and its size. As a result, the cerebellar tonsils are displaced below (⩾5 mm) the level of the magnum foramen (Chiari I). The greater displacement of the cerebellar tonsils and the low cerebellar vermis to the second cervical vertebra or below, as well as caudal displacement of the lower brain stem into the magnum foramen, is typical of the stronger pronounced CM (Chiari II). The occurrence of CM can be as high as 560 per 100,000 people [1]. Herniation of the cerebellar tonsils into the foramen magnum may disturb cerebrospinal fluid (CSF) circulation through the CVJ. Pathology of the CVJ may alter the CSF flow dynamics and the conditions of venous outflow from the cranial cavity. Phase-contrast MRI is currently widely used to assess the dynamics of CSF flow through the CVJ in patients with CM [2—4]. This method allows one to assess the changes in the CSF flow velocity through the CVJ and makes it possible to detect if it is blocked [4]. When analyzing the results of these studies, N. Chaffer et al. [4] mentioned that the data were controversial and there was a chance of errors. N. Alperin et al. [5] performed phase-contrast MRI studies and revealed statistically significant changes in the CSF flow velocity through the CVJ neither in patients with CM nor in the control group consisting of healthy individuals. The same authors used the modified MRI to quantify the changes in viscoelasticity of the cerebrospinal system. The method demonstrated that the pathological changes in the CVJ in patients with CM result in a statistically reliable decrease

© Group of authors, 2015

e-mail: vsnevrologiya@gmail.com

in compliance of the cerebrospinal system and, therefore, increase its elastance [5]. Meanwhile, the opposite effect (significantly increased brain compliance and reduced elastance) is the main result of surgical decompression of CVJ in patients with CM. These changes can be caused by normalization of the CSF flow through the CVJ in patients with CM. Meanwhile, the changes in the degree to which the brain structures are displaced are much smaller after surgical decompression of the foramen magnum compared to the increase in compliance of the cerebrospinal system [5]. Hence, the nonfocal factors (changes in the viscoelastic properties of the cerebrospinal system) are much more specific for CM than the local changes in the CSF flow velocity through the CVJ. CM is known to be an inherited disorder. However, the clinical manifestations of CM tend to occur in adulthood. N. Alperin et al. [5] believe that such a late manifestation of the clinical signs of CM is because brain elastance gradually increases with age [6]. A non-invasive ultrasound method that allows one to assess the viscoelasticity of the cerebrospinal system, as well as venous brain circulation and the CSF dynamics, has been developed at the N.N. Burdenko Neurosurgical Institute [7, 8].

The aim of the present study was to use this method to investigate the viscoelastic properties of the cerebrospinal system, CSF dynamics, and cerebral venous flow in patients with CM.

Clinical material

The study involved 35 patients with CM (Chiari I – 26 patients, Chiari II – 9 patients) aged 4 to 58 years. Four of the examined patients had hydrocephalus; six patients had syringomyelia. The most frequent complaints included neck pain, severe headache that was made worse by sneezing or coughing; swallowing disturbance; sensory and motor disorders, such as numbness of the limbs and ataxia. The average size of herniation of cerebellar tonsils into the foramen magnum was 8.2±3.1 mm. No correlation between the degree of herniation into the foramen magnum and the severity of neurological symptoms was found.

The procedure of our own studies

According to the Monro—Kellie doctrine [9, 10], the major intracranial volumes of the cerebral tissue, blood, and CSF are normal. One of these volumes can be changed only if the volumes of other components of intracranial contents are altered correspondingly. The normal volumes of the cerebral tissue is 1100—1300 cm^3; CSF, 130—150 cm^3; and blood, 60—80 cm^3 [11]. Stability of the cerebral hemodynamics is ensured by functioning of the complex, multi-component cerebral blood flow regulation system [12—14]. In addition to quantification of each intracranial volume (cerebral tissue, blood, and CSF), the total characteristics of the craniovertebral volume ratio (CVVR) that is determined as the tightness of the entire cerebrospinal system [15—17] is also crucial. For this purpose, the dependence between the additional volume in the CSF space and the resulting changes in intracranial pressure (ICP) is studied. Sterile normal saline injected intralumbarly is usually used as the additional solution; the changes in ICP can be assessed simultaneously [15—18]. Unfortunately, the invasiveness of this method significantly limits its clinical use. The changes in CVVRs occur under natural physiological conditions. When the vertical body position is changed to the antiorthostatic position, CSF partially migrates from the spinal CSF space into the intracranial CSF space. This natural, non-invasive, intracranial infusion of CSF through the CVJ increases the intracranial volume of CSF and ICP. Under these conditions, bridging veins running in the subarachnoid space are compressed in a cuff-like manner, which is accompanied by weaker venous outflow through the deep cerebral veins and the straight sinus [19]. Hence, it is no wonder that there is a good correlation between the ICP measured using a catheter inserted into the ventricular system and blood flow in the straight sinus recorded by Transcranial Dopper (TCD) sonography [20], which allows one to non-invasively assess ICP. ICP can be measured non-invasively by varying the patient's body position on a tilt table [21]. Transcranial Doppler registration of venous blood flow in the straight sinus under orthostatic stress is an adequate method for assessing the CVVRs and allows one to assess the hemodynamic status [8, 22]. We studied the changes in the velocity of systolic venous blood flow in the straight sinus and the amplitudes of venous blood flow pulse vibrations under orthostatic load both in healthy volunteers **(Fig. 1)** and in patients with pathology. To non-invasively study venous blood flow velocity (VBFV) in the straight sinus by TCD, we placed an ultrasound sensor on the cranial surface near the external occipital protuberance. The straight sinus was located at a depth of 55 mm. CVOR was studied in 45 healthy individuals aged 2 to 82 years. **Fig. 1** shows the orthostatic profile of venous circulation in a healthy woman.

Fig. 1 demonstrates that VBFV in the straight sinus remains stable in a wide range of changes in body position on the tilt table (from +90° to 0°). Meanwhile, VBFV in the straight sinus gradually increases in the range of angles between 0° and –30°. The reason for these changes in venous blood flow may lie behind postural fluctuations in intracranial pressure that is based on redistribution of CSF within the cerebral and spinal CSF spaces. When using a similar functional load (changes in body position on the tilt table within the range from +90° to –30°) in 7 patients operated on for brain tumors, with their ventricular pressure continuously measured using implanted telemetric sensors, we have also revealed two areas where ventricular pressure was stable and gradually increased [21]. The stable pressure area was typically located in the range from +25°, +35°, up to +90°, while the increasing pressure area corresponded to the range of

orthostatic load from –25° to +30°; the dependence between the body tilt angle and an increase in ventricular pressure approached the linear one. Both the experimental [23] and clinical [15] studies showed the presence of two areas corresponding to stable and increasing ICP as the additional intracranial volume was increased. The stable pressure area characterizes the intracranial spatial compensation (the reserve capacity), while the increasing pressure zone characterizes elastic compensation (elastance gradient).

In our studies, systolic flow velocity in the straight sinus was registered by TCD sonography as the body position on the tilt table was gradually changed from +90° to –30°. The dependence between systolic flow velocity and body position is characterized by two straight lines: 1) the spatial compensation area, where systolic flow velocity is stable and 2) the elastance compensation area, where systolic flow velocity gradually increases. The tilt angle of this line characterizes the elastance of the craniovertebral contents, while the intercept point of these two lines characterizes its reserve capacity. VBFV is stable in the spatial compensation area and characterizes the state of cerebral venous circulation. As a result of our studies, we detected the group of patients with VBFV in the straight sinus remaining stable (nonreactivity) as their body position on the tilt table was varied within the entire range, from the orthostatic position (+90°) to antiorthostatic position (–30°). Meanwhile, in other patients, VBFV in the straight sinus increased to different extents as their body position on the tilt table was changed in the same range of angles. Taking these data into account, the term "cerebrovenous orthostatic reactivity" is the most adequate term characterizing changes in venous blood flow under orthostatic functional load. To quantify CVOR, the degree to which VBFV in the straight sinus increased (cm/s) is divided by the entire range of variation of body position on the tilt table (degrees) within which VBFV in the straight sinus was increasing. The major quantitative characteristics of CVVR in healthy individuals were appreciably stable, while the range of their fluctuations was reported in the clinical protocol **(Table). Table** lists the results of studying the cerebral venous circulation, the CSF dynamics, and craniovertebral volume ratios in healthy individuals.

The data listed in **Table** demonstrates that the reserve capacity of CVVR in healthy individuals ranges between 0° and +15°; the velocity of systolic venous flow in the spatial compensation area (VBFV) varies between 14 and 28 cm/s; the amplitude of pulse vibrations of the venous flow in the same zone (AMP) in the norm ranges from 6 to 8 cm/s; and cerebrovenous orthostatic reactivity (CVOR) ranges from 0.15 to 0.35 cm/s/°. The CSF dynamics is assessed according to the time required for the velocity of venous blood flow to become stable again as the tilt table position is changed from the horizontal to the vertical one (no more than 5 s in the norm).

All the results were processed statistically. The method was found to be highly reproducible when comparing the results of repeated studies for the same person. The data listed in Table demonstrate that CVOR is stable and varies within a small range in healthy individuals.

Meanwhile, when the craniovertebral volume ratios were varied to different extents, the CVOR significantly differed from the norm: it could both increase (sometimes 5—6-fold compared to the upper limit of the norm — significant hyperreactivity) and substantially decrease (up to complete absence of any changes in venous blood flow in the straight sinus under orthostatic load — nonreactivity). Different patients had different degrees of changes in CVOR (nonreactivity, hyporeactivity, normoreactivity, moderate or significant hyperreactivity).

The quantitative characteristics of changes in cerebrovenous orthostatic reactivity (CVOR cm/s/°) are represented as follows.

Fig. 2 shows different types of cerebrovenous orthostatic reactivity (CVOR).

One of the variants of CVOR is nonreactivity (upper curve in **Fig. 2**), which is characterized by no changes in VBFV in the straight sinus within the entire range of body positions on the tilt table (from –30° to +90°).

Results

Among the examined 35 patients with CM, only 3 (9%) patients had no changes in CVOR before the surgery. In the remaining 32 cases, various types of CVOR disorders were preoperatively observed (nonreactivity, moderate or significant hyperreactivity). Meanwhile, CVOR was completely normalized after surgical management in more than half cases (63%), while the remaining patients had moderate cerebrovenous orthostatic hyperreactivity.

Cerebrovenous orthostatic nonreactivity was preoperatively observed in 14 patients with CM. The same patients had increased VBFV in the straight sinus.

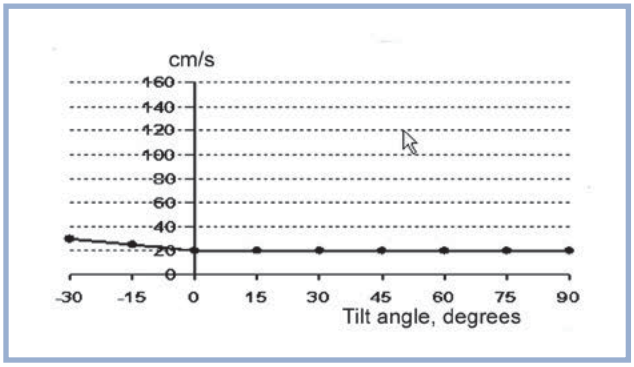


Fig. 1. **Y axis shows the orthostatic profile of venous cerebral circulation in a healthy 34-year-old woman (estimation based on systolic flow velocity). X axis shows variation of body position on a tilt table.**

The results of examination of a 37-year-old woman with CM can be used as an example of cerebrovenous orthostatic nonreactivity accompanied by increased VBFV and normalized CVOR after surgical decompression of the foramen magnum. Six years ago after a traumatic brain injury (falling from her own height and hitting the back of her head while remaining conscious) the patient started to have head and neck pain, which later was gradually aggravated. One year after the pain syndrome had emerged, MRI showed CM (invagination of cerebellar tonsils into the foramen magnus 8 mm below the Chamberlain—Chiari II line). The patient permanently received conservative therapy; however, neck pain was gradually aggravated, especially after another head injury one year later (a downward shock into the occipital region). A year ago the patient started to feel occasional numbness in the left hand. One of the headache attacks was accompanied by severe vertigo and loss of consciousness. Repeated MRI exam showed the more significant downward displacement of the cerebellar tonsils by 13 mm below the Chamberlain line. Neurological examination showed pain in the occipital area, mild symptoms affecting the brain stem and cerebellum presented as gaze-evoked nystagmus on lateral gaze, instability when performing Romberg's test, and transient hypoesthesia in the C5—C7 units on the left side.

The patient was subjected to C1 laminectomy and craniovertebral junction decompression. The early postoperative period was uncomplicated; regression of neurological symptoms included alleviation of neck pain and headache, elimination of sensory disorders and brain stem–cerebellar symptoms.

Fig. 3 shows the results of examining the patient.

Before surgery, the patient had cerebrovenous orthostatic nonreactivity, which can be attributed to blockage of CSF flow through the craniovertebral junction. In addition, venous flow velocity in the straight sinus was preoperatively increased (45 cm/s) due to increased ICP. After the surgery, venous flow in the straight sinus and CVOR were normalized (venous flow velocity decreased to 20 cm/s; CVOR corresponded to normoreactivity – 0.33 cm/s/°), which can indicate that the ICP had normalized and the normal CSF flow through the craniovertebral junction had recovered.

Patients with CM typically had cerebrovenous orthostatic hyperreactivity before surgery (18 cases). After surgical management, the CVOR in this group of patients decreased to normoreactivity.

Assessment of the preoperative distribution of CVOR most frequently showed cerebrovenous orthostatic nonreactivity and moderate hyperreactivity. Meanwhile, normoreactivity and significant hyperreactivity were detected much less often. In the same group of patients, cerebrovenous orthostatic hyporeactivity was not detected at all before the surgery (**Fig. 4**, left-hand side).

The CVVR values in patients with CM significantly changed after surgical management: cerebrovenous orthostatic normoreactivity and, less frequently, moderate hyperreactivity were detected in most patients (**Fig. 4**, right-hand side). Patients after surgery showed neither cerebrovenous orthostatic nonreactivity nor significant hyperreactivity, although these conditions took place before surgery. Cerebrovenous orthostatic nonreactivity in patients with CM was accompanied by the increased mean SFV values in the straight sinus reaching –32.6 cm/s (normal value, –24.3 cm/s); the difference between the venous flow velocity in patients with CM and the normal value was statistically significant ($p<0.004$). Surgical management resulted in a statistically significant decrease in the SFV in the straight sinus ($p=0.003$).

Discussion

The CVJ plays a crucial role in interaction between the CSF systems in the brain and spinal cord. CVJ pathologies may change this interaction and cause imbalance in the CSF dynamics and cerebral venous circulation. Investigation of these disorders contributes to elucidating the degree to which the CVJ is obstructed, which may have diagnostic value when determining the indications for surgical management and for evaluating its effectiveness.

It has been found that a change in the body position of a healthy individual causes significant variations in ICP: it decreases in the orthostatic (vertical) position and increases in the horizontal or antiorthostatic (head below horizontal) position [21]. Migration of CSF through the CVJ can be one of the reasons for postural fluctuations in ICP when changing body position. Gravitational effects under the same orthostatic load may cause migration of CSF through the CVJ in different directions: in the caudal direction in orthostatic position

Cerebral venous circulation, CSF dynamics, and craniovertebral volume ratios. Examination protocol

Parameter	Norm
Systolic flow velocity (SFV_1) in the straight sinus in horizontal position, cm/s	14—28
Pulse amplitude (AMP_1) in the straight sinus in horizontal position, cm/s	6—8
Systolic blood flow (SFV_2) in the straight sinus in the spatial compensation area, cm/s	14—28
Pulse amplitude (AMP_2) in the straight sinus in the spatial compensation area, cm/s	6—8
CSF dynamics	No more than 5 s
Cerebrovenous orthostatic reactivity (CVOR), cm/s/°	0.15–0.35
Reserve capacity, °	0 (+15)

and in the oral direction in antiorthostatic position. Hence, applying an orthostatic load is an adequate method for assessing passability of the CVJ as it detects blockage of the CVJ and different degrees of its stenosis. The role of gravitational effects on the CVVR values and cerebral venous circulation is especially noticeable in a zero-gravity environment during space flights [25]. Different conditions of venous blood outflow from the cranial cavity may also play a role for postural fluctuations in ICP. In horizontal position, venous blood flows out of the skull mostly via the jugular veins, while in vertical position it flows via the vertebral plexus veins [25, 26, 28]. The volume flow rate of venous blood via the internal jugular veins in vertical position is 70 mL/min, while increasing tenfold to reach 700 mL/min in horizontal position [26]. Compliance of the cerebrovascular system increases significantly (2.8-fold) in the vertical position; hence, its elastance decreases [27]. Meanwhile, ICP and cerebral elastance depend exponentially on the volume of intracranial contents [18]. The intracranial volume of blood and CSF decreases in the vertical position, thus reducing ICP [28]. Pathology of the CVJ may change the CSF dynamics and conditions of venous blood outflow from the cranial cavity.

We have detected to main types of changes in CVOR in patients with CM: nonreactivity and hyperreactivity

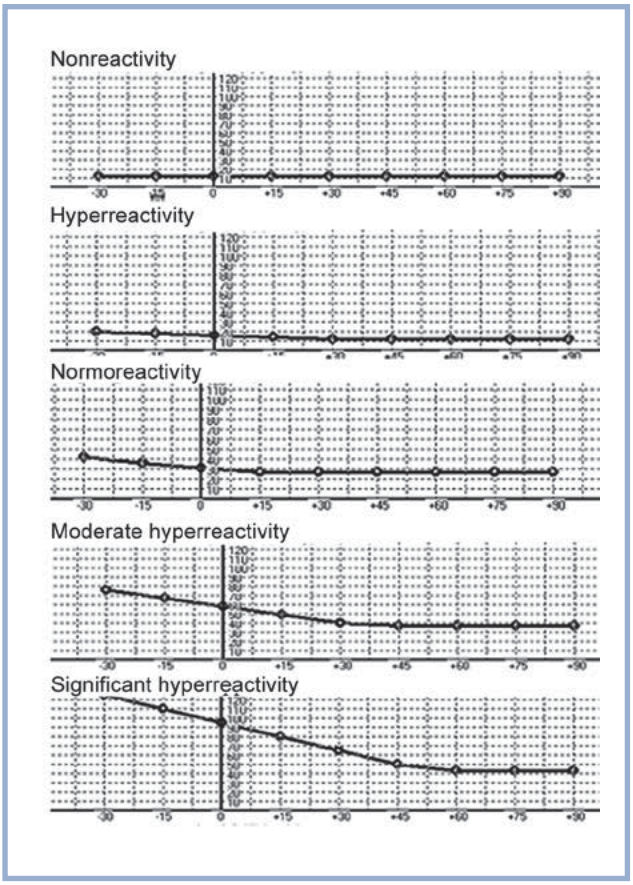


Fig. 2. **Different degrees of cerebrovenous orthostatic reactivity (CVOR).**

(either moderate or significant). Cerebrovenous orthostatic nonreactivity in pathology of the CVJ is most likely to be caused by blockage of CSF flow between the subarachnoid space of the brain and spinal cord. Disintegration of the cerebral and spinal CSF space seems to be the main reason for syringomyelia in patients with Chiari malformation [28—30]. Furthermore, herniation of cerebellar tonsils into the foramen magnum causes compression of the internal vertebral venous plexus located extradurally. It disturbs redistribution of venous blood outflow from the skull under orthostatic load, which may also contribute to development of cerebrovenous orthostatic nonreactivity. In a healthy person under antiorthostatic conditions, CSF partially migrates from the spinal CSF space into the cerebral one, resulting in an increase in ICP. The strong correlation between ICP and SFV in the CVJ [20] may be the reason for increasing SFV in the CVJ under antiorthostatic conditions. The absence of an increase in SFV in the CVJ under antiorthostatic conditions and development of cerebrovenous orthostatic nonreactivity may attest to blockage of the CVJ and disintegration of the CSF space in the brain and spinal cord in patients with CM. Registration of ICP under orthostatic load using ICP sensors located extradurally is an adequate method for assessing the passability of the CVJ. The use of this method in patients with CM made it possible to detect the statistically significant weakening of postural fluctuations in ICP, which attested to disruption of passability of the CVJ [31]. However, the invasiveness of this method substantially limits its clinical use. Not only cerebrovenous orthostatic nonreactivity was detected in patients with CM but also hyperreactivity, which was most likely caused by obstruction of CSF flow through the CVJ, resulting in increased ICP and elastance of CVVR. Increased elastance (decreased compliance of the brain) in patients with CM was also detected by N. Alperin et al. [5] who used modified MRI. After surgical management (decompression of the foramen magnum), the examined patients had significantly normalized CVVR values, while CVOR was typically characterized by normoreactivity or, less frequently, by moderate hyperreactivity. No cases of cerebrovenous orthostatic nonreactivity that is typical of blockage of the CVJ were detected after surgical management of patients with CM.

Surgical decompression of the CVJ resulted in either partial or complete normalization of CVOR after nonreactivity detected before the surgery. In patients who preoperatively had cerebrovenous orthostatic hyperreactivity, the degree of hyperreactivity always decreased (usually to normal values) after surgical management. According to the data obtained by N. Alperin et al. [5] using modified MRI, a significant increase in compliance of the brain (and, therefore, a decrease in its elastance) is the main effect of surgical decompression of the CVJ in patients with CM. These changes can result from normalization of CSF flow

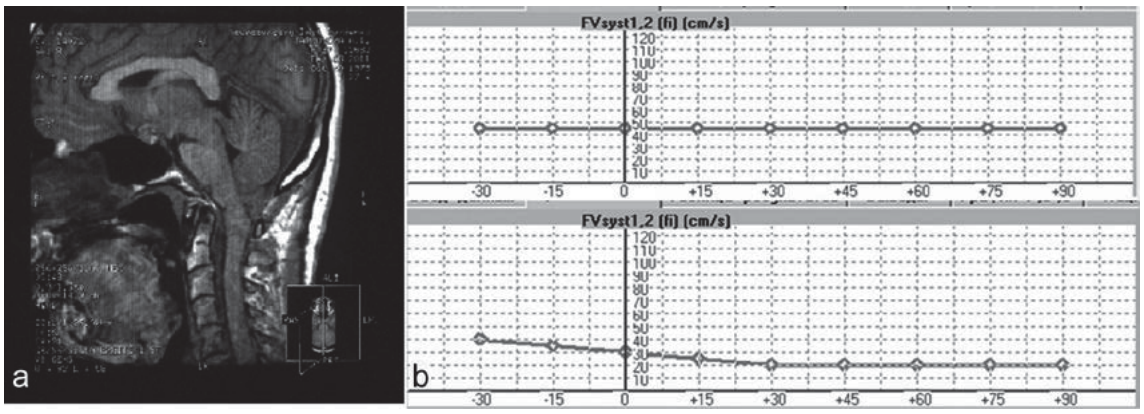


Fig. 3. **Results of examining the 37-year-old patient with Chiari malformation.**

a — Preoperative MRI scan of the brain shows that cerebellar tonsils displaced downward, into the foramen magnus; b — study of cerebrovenous orthostatic reactivity (right top corner — before surgery; right bottom corner — after surgery).

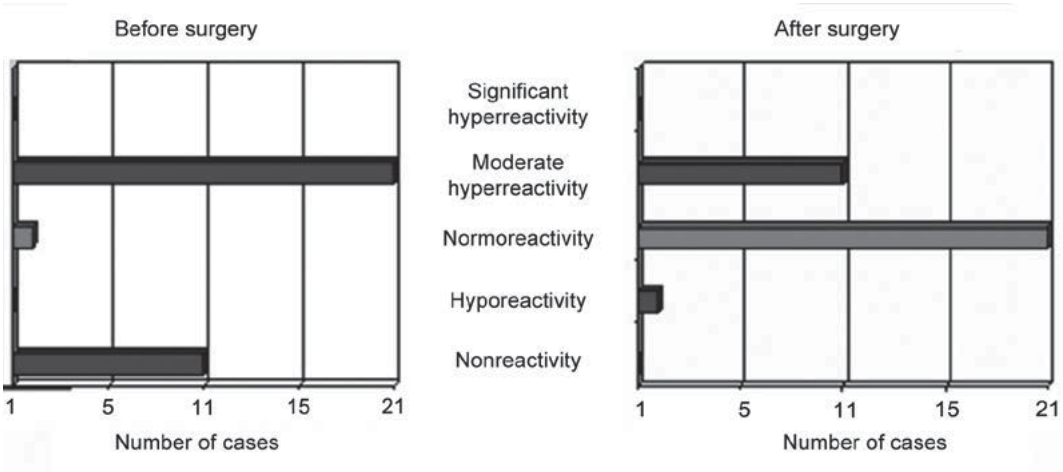


Fig. 4. **Occurrence of different degrees of changes in cerebrovenous orthostatic reactivity (CVOR) in patients with Chiari malformation before (left-hand side) and after (right-hand side) surgery.**

Y axis represents the different degrees of changes in CVOR (nonreactivity, hyporeactivity, normoreactivity, moderate hyperreactivity, and significant hyperreactivity). X axis shows the number of cases.

through the CVJ. In patients with CM, changes in the degree of displacement of cerebral structures and CSF flow velocity through the CVJ after surgical decompression are much less pronounced that the increase in compliance of the craniovertebral contents.

The main result of the studies is that the high occurrence of CVOR disorders (over 90%) in patients with CM has been detected. Meanwhile, more than half of these patients (63%) show total normalization of CVOR after surgical management, while the remaining patients show a statistically significant decrease in the degree of pathology. Surgical management completely eliminates cerebrovenous orthostatic nonreactivity (being indicative of blockage of the CVJ) and statistically reliably reduces the degree of hyperreactivity that is typical of CVJ stenosis. These data demonstrate that CVOR has a high diagnostic significance for assessing the passability of the CVJ.

The preoperative changes in CVOR (nonreactivity, hyperreactivity) and its normalization after surgical management was observed not only in patients with CM but also in individuals having tumors in the CVJ [32] and invagination of the odontoid process of the C2 vertebra [33], which may indicate that they depend on disturbance of CSF dynamics and hemodynamics in the CVJ rather than on the features of different clinical entities. Blockage of the CVJ is characterized by cerebrovenous orthostatic nonreactivity, which was usually combined with statistically significant increase in SFV in the CVJ (up to 80 cm/s) compared to that in the control group of healthy

volunteers; SFV in the CVJ is typically normalized after surgical management. Taking into account the strong correlation between SFV in the CVJ and ICP [20], we can suggest that the twofold increase in the mean SFV values compared to the norm may be caused by intracranial hypertension.

Cerebrovenous orthostatic nonreactivity was also observed in patients with idiopathic normal pressure hydrocephalus [22]; however, it was characterized by the significantly reduced SFV in the CVJ (up to 10 cm/s) for this clinical entity. Cerebrovenous orthostatic nonreactivity in patients with idiopathic normal pressure hydrocephalus is probably caused by cerebral atrophy.

Hence, examination of the patients with CM has revealed strong disturbance of CVOR that was normalized either partially or completely after surgical treatment. It is reasonable to conduct ultrasonography of cerebral venous circulation pre-, intra-, and postoperatively.

Conclusions

1. Patients with craniovertebral junction pathology (Chiari malformation) are likely to exhibit cerebrovenous orthostatic nonreactivity and moderate or significant hyperreactivity before surgical management.

2. After surgical treatment (decompression of the foramen magnum), patients with Chiari malformation exhibit significant normalization of craniovertebral volume ratios; cerebrovenous orthostatic reactivity is typically characterized by normoreactivity or, less frequently, by moderate hyperreactivity.

3. Venous flow velocity in the straight sinus in patients with craniovertebral junction pathology (Chiari malformation) may be increased before surgery and is normalized after surgical treatment.

Authors declare no conflict of interest.

REFERENCES

1. Elster AD, Chen VYM. Chiari I malformations clinical and radiologic reappraisal. *Radiology*. 1992;183:347-353.
   doi: 10.1148/radiology.183.2.1561334.
2. Reutov A. *Diagnostic principles and surgical treatment of patients with Chiari I malformation [dissertation]*. Moscow; 2012. (In Russ.).
3. Reutov A, Kornienko V, Arutyunov N, Fadeeva L. Features of CSF flow on the upper cervical level in the norm and in patients with Chiari I malformation. *Vestnik rentgenologii i radiologii*. 2009;4:37-42. (In Russ.).
4. Shaffer N, Martin B, Loth F. Cerebrospinal fluid hydrodynamics in type I Chiari malformation. *Neurological Research*. 2011;l33:3:1-15.
   doi: 10.1179/016164111X12962202723805.
5. Alperin N, Sivaramakrishnan A, Lichtor T. Magnetic resonance imaging – based mearsuments of cerebrospinal fluid and blood flow as indicators of intracranial complains in patients with Chiari malformation. *J Neurosurgery*. 2005;103:46-52.
   doi:10.3171/jns.2005.103.1.0046.
6. Kiening KL, Schoening WN, Lanksch WR. Intracranial compliance as a bed-side monitoring technique in severely head-injured patients. *Acta Neurochirurg*. Suppl 2002;81:177-180.
   doi:10.1007/2F978-3-7091-6738-0_46.
7. Shakhnovich V, Galushkina A. Features of cerebral venous circulation in patients with benign intracranial hypertension. *Angiologiya i sosudistaya khirurgiya*. 1998;4(1):65-71. (In Russ.).
8. Shakhnovich AR, Shakhnovich VA, GalushkinaAA. Non-invasive assessment of the elastance (E) and reserve capacity of the intracranial content (RCIC) via FV measurement in the straight sinus (SS) by TCD during body tilting test (BTT). *J Neuroimaging*. 1999;9:141-149.
   doi: S0929-8266(97)90130-0.
9. Monro A. *Observations on the structure and function of the nervous system*. Creech and Johnson, Edinburgh, 1783.
10. Kellie G. An account of the appearances observed in the dissection of two of three individuals presumed to have perished in the storm of the 3D, and whose bodies were discovered in the vicinity of Leth on the morning of the 4th, November 1824, with some reflections on the pathology of the brain. Trans, Med.-Chir. Soc. Edindburgh 1824;1:84-169.
11. Popovic D, Khoo M, Lee S. Noninvasive monitoring of intracranial pressure. *Recent patents on biomedical engineering*. 2009;2:165-169.
    doi: LiveAll1.
12. Shakhnovich V. *Cerebral Ischemia. Neurosonology*. Moscow; 2002:305. (In Russ.).
13. Shakhnovich V. *Ischemic Stroke*. Moscow; 2006:308. (In Russ.).
14. Shakhnovich V. *Modern capabilities of neurosonology in neurosurgery*. In: Dreval ON, eds. Neurosurgery: Handbook for Physicians. Moscow; 2013:524-556. (In Russ.).
15. Shevchikovskiy E, Shakhnovich A, Konovalov A, Tomas D, Korsak-Slivka I, Dubova S, Kunitskiy A, Slivka S, Milovanova L, Ditko P. Computer-assisted intensive monitoring of patients in a neurosurgical clinic. *Voprosy neirokhirurgii*. 1980;3:7-16. (In Russ.).
16. Szewczykowski J. A method of estimating intracranial decompensation in man. J *Neurosurg*. 1976;45:155-158.
    doi:10.3171/jns.1976.45.2.0155.
17. Szewczykowski J. A fast method of estimating the elastance of the intracranial system. *J Neurosurg*. 1977;47:19-26.
    doi:10.3171/jns.1977.47.1.0019.
18. Marmarou A, Shulman K, La Morgese J. Compartmental analysis of compliance and outflow resistance of the cerebrospinal fluid system. *J Neurosurg*. 1975;43:523-534.
    doi: 10.3171/jns.1975.43.5.0523.
19. Valdueza JM, Schultz M, Harms L. Venous transcranial doppler ultrasound monitoring in Acute Dural Sinus Thrombosis. *Stroke*. 1995;26:7:1196-1199.
    doi:10.1161/01.str.26.7.1196/
20. Schoser BG, Riemenschneider N, Hansen HC. The impact of raised intracranial pressure on cerebral venous hemodynamics: a prospective venous transcranial Doppler ultrasonography study. *J Neurosurg*. 1999;91:744-749.
    doi: 10.3171/jns.1999.91.5.0744.
21. Chapman P, Cosman ER, Arnold MA. The relationship between ventricular fluid pressure and body position in normal subjects and subjects with shunts: a telemetric study. *Neurosurgery*. 1990;26(2):181-189.
    doi: 10.1227/00006123-199002000-00001.
22. Shakhnovich A, Shakhnovich V. Cerebrovenous orthostatic reactivity (CVOR) in patients with hydrocephalus and intracranial hypertension. *Neirokhirurgiya i nevrologiya detskogo vozrasta*. 2013;3:11-25. (In Russ.).
23. Langfitt TW, Weinstein JD, Kassel NF. Cerebral vasomotor paralysis produced by intracranial hypertension. *Neurology*. 1965;15:21:622-641.
    doi: 10.1212/wnl.15.7.622.
24. Israilyan L, Shimanskiy V, Otamanov D, Poshataev V, Lubnin A. Intraoperative positioning of neurosurgical patients (sitting or lying). *Anesteziologiya i reanimatologiya*. 2013;4:18-25. (In Russ.).
25. Galushkina AA, Shakhnovich VA, Myasnicov VI, Stepanova SI. The influence of space flight (SF) on cerebral arterial inflow (CAI) and cerebral venous outflow (CVO). *European journal of ultrasound*. 1997 May;5:Suppl. 1:52.
    doi: S0929-8266(97)90124-5.
26. Valdueza JM, von Munster T, Hoffman O, et al. Postural dependency of the cerebral venous outflow. *Lancet*. 2000;355:200-201.
    doi: S0140673699048047.

27. Alperin N, Lee SH, Sivaramakrishnan A, et al. Quantifying the Effect of Posture on Intracranial Physiology in Humans by MRI Flow Studies. *Journal of Magnetic Resonance Imaging*. 2005;22:591-596.
doi: 10.1002/jmri.20427.
28. Gisolf J, Lieshout J, Haurden H. et al. Human cerebral venous outflow pathway depends on posture and central venous pressure. *J Physiol*. 2004;569:1:317-327.
doi: 10.1113/jphysiol.2004.070409.
29. Klekamp J. The pathophysiology of Syringomyelia- Hystorical overview and current concept. *Acta Neurochirurg*. 2002;144:649-664.
doi: 10.1007/978-3-642-56023-1_2.
30. Levine DN. The pathogenesis of syringomyelia associated with lesions of foramen magnum: a critical review of existing theories and a proposal of a new hypothesis. *J of Neurol Sci*. 2004;220:3-21.
doi: S0022510X04000607.
31. Poca MA, Sahuquillo J, Topczewski T, et al. Posture-induced Changes in Intracranial Pressure: A Comparative Study in Patients with and without a Cerebrospinal Fluid Block at the Craniovertebral Junction. *Neurosurgery*. 2006;58:5899-5906.
doi: 10.1227/01.neu.0000209915.16235.6d.
32. Mitrofanova E, Shakhnovich V, Shkarubo A, Andreev D. Cerebrovenous orthostatic reactivity in patients with craniovertebral junction tumors. *Rossiyskiy neirokhirurgicheskiy zhurnal im. prof. A.L. Polenova*. 2014;6(special issue):158. (In Russ.).
33. Mitrofanova E, Shakhnovich V, Shkarubo A, Andreev D, Gromov I. Venous brain circulation and CSF dynamics in craniovertebral junction pathology (invagination of the invagination of the odontoid process of the C2 vertebra). *Rossiyskiy neirokhirurgicheskiy zhurnal im. prof. A.L. Polenova*. 2014;6(special issue):253-255. (In Russ.).
34. Gushcha A, Shakhnovich A, Kashcheev A, Arestov S, Abuzaid S. A novel minimally invasive method for surgical treatment of the Arnold—Chiari malformation: The experimental clinical study. *Rossiyskiy neirokhirurgicheskiy zhurnal im. prof. A.L. Polenova*. 2010;2(4):28-29. (In Russ.).

Commentary

This paper focuses on the topical problem, studying the volume ratios between the craniovertebral junction structures and the features of the CSF dynamics and hemodynamics in this area in patients with Chiari I and II malformation during the preoperative and postoperative periods.

Chiari malformation is a common disorder that requires neurosurgical management in most cases. Nevertheless, the data on deep pathophysiological mechanisms of clinico-neurological disorders in this malformation have not been thoroughly studied yet. These factors are responsible for the scientific and practical value of the observations reported in this paper.

The study involved 35 patients with Chiari malformation. The venous blood flow in the straight sinus was registered in patients lying on the tilt table in the position varied between +90° and −30° (the original procedure was designed by the authors). It was found that either moderately or significantly increased cerebrovenous orthostatic reactivity (CVOR) (hyperreactivity) or no changes under orthostatic load (nonreactivity) was typical of patients with Chiari malformation in the preoperative period. Surgical management (decompression of the craniovertebral junction and dura mater reconstruction) normalizes CVOR, which is characterized by normoreactivity (63% of cases). Operative treatment is also found to normalize blood flow velocity in the straight sinus.

The material discussed and the conclusions drawn by the authors allow one not only to objectify the outcomes of neurosurgical treatment but also to reveal the key pathophysiological mechanism that is responsible for the type of pathology in patients with Chiari malformation. We believe that adding non-invasive ultrasound methods for assessing the condition of the craniovertebral junction to the algorithm of preoperative examination is reasonable and will improve early and delayed surgery outcomes. It will also allow one to specify the indications for surgery, which is particularly topical for Chiari malformation not accompanied by hydrocephalus or syringomyelia.

It should be mentioned that blood flow velocity in the straight sinus can be measured both intra- and perioperatively. There are Russian and foreign studies showing the relationship between the changes in these parameters revealed intraoperatively and the strategy of surgical intervention (decompression of the craniovertebral junction only or its combination with dura mater reconstruction). These studies can reasonably continue the work described in this paper.

A.O. Gushcha (Moscow, Russia)