Clinico-otoneurological assessment of the early postoperative outcome in surgical treatment of Chiari malformation and syringomyelia

Levchenyuk Sofia¹, Derkach Yuriy²

¹Doctor-otolaryngologist, otoneurology group, State Institution "Romodanov Neurosurgery Institute, the National Academy of Medical Sciences of Ukraine", Kyiv, Ukraine

²Candidate of medical sciences, doctor-neurological surgeon, Department of spinal cord pathology, "Romodanov Neurosurgery Institute, the National Academy of Medical Sciences of Ukraine", Kyiv, Ukraine

Address for correspondence:
Levchenyuk Sofia
E-mail: mdslevchenyuk@gmail.com

Abstract: although the pathology of Chiari malformation is extensively studied, many questions have not yet been solved. In particular, the clinical manifestations of this pathology have not been studied in detail entailing the lack of the unified approach for the optimal complex of the diagnostic workup and the selection of the optimal surgical intervention. These questions remain the point of discussion in the scientific literature. The present study is based on the retrospective analysis of the diagnostic findings and treatment outcomes of 59 patients – 24 males (40.7 %) and 35 females (59.3 %) – treated in the Department of spinal cord pathology of the State Institution "Romodanov Neurosurgery Institute, the National Academy of Medical Sciences of Ukraine" in 2010-2020. The age of the patients ranged from 16 to 50 years (36.9 ± 3.7 on the average). All patients were examined according to the conventional standards allowing for the assessment of the pre- and postoperative disease course. The following surgical interventions were provided: bone decompression for the inferior parts of the cerebellum at the upper cervical level; the recovery of the CSF flow at the region of craniovertebral junction; the expansion of the space in the fossa cranii posterior and/or craniovertebral junction. The data were processed and analyzed with the aid of STATISTICA 13 software. The quantitative variables were compared by non-parametric methods. The aim of the study was to analyze the preoperative and early postoperative clinico-otoneurological status in patients with Chiari malformation and syringomyelia. The clinical aspects of the disease were evaluated. Most clinical signs and symptoms in patients with Chiari malformation were associated with otolaryngological disturbances (hearing impairment, tinnitus, hoarseness, vertigo, etc.) and cerebellar signs (ataxia, nystagmus). Taking into account non-specific signs and symptoms of the disease, the otoneurological evaluation in these patients is undoubtedly of high importance for the preoperative differential diagnosis as well as the postoperative assessment of treatment outcome. The further study of the problem will allow us to timely diagnose Chiari malformation, to predict the course of the disease and the risk of the possible complications providing the clinicians with important information for decision-making on the optimal treatment strategy. The clinical and instrumental otoneurological examination is an important component of the systemic assessment of the clinical-and-functional status of patients with syringomyelia caused by Chiari malformation that allows for objectifying the dynamics of the otoneurological aspects of the disease in the course of the treatment.

Keywords: Arnold-Chiari Malformation, syringomyelia, headache, hearing, vertigo, tinnitus.
Introduction

Chiari malformation (CM) is an impaired development of the central nervous system representing the heterogeneous group of the diseases varying by their clinical-and-anatomical features. CM is characterized by a downward displacement of cerebellar tonsils or vermis (and sometimes the brain stem and the fourth ventricle) through the foramen magnum (FM). CM can be associated with hydrocephalus, spina bifida, syringomyelia, curvature of the spine (kyphosis and scoliosis) and tethered cord syndrome (Yang, C., Deng, X., Yang, J., Xu, Y., 2019). Prior to the widespread adoption of MRI, CM was considered as a rare pathology. In particular, this was due to the polymorphism of the neurological symptoms and the difficulties with the instrumental diagnosing. At present, the incidence of this pathology is 3.3-8.2 per 100 thousands persons (Soleman, J., Roth, J., Constantini, S., 2019).

The clinical pattern of the disease depends largely on the age at the onset, the concomitant pathology and the presence of syringomyelia. Although the CM pathology is extensively studied, many questions have not yet been solved. In particular, the clinical manifestations of this pathology have not been studied in detail entailing the lack of the unified approach for the optimal complex of the diagnostic workup and the selection of the optimal surgical intervention. These questions remain the point of discussion in the scientific literature. (Lin, W. et al., 2018; Jiang E. et al., 2018; Khalsa, S. et al., 2018). These factors indicate the expediency of the present study.

Aim

The aim of the study was to analyze the preoperative and early postoperative clinico-otoneurological status in patients with CM and syringomyelia.

Materials and methods

The study is based on the retrospective analysis of the diagnostic findings and treatment outcomes of 59 patients with their disease codified as Q07.0 according to the International Disease Classification 10 (WHO 2007). The patients were treated in the Department of spinal cord pathology of the State Institution "Romodanov Neurosurgery Institute, the National Academy of Medical Sciences of Ukraine" in 2010-2020.

The standard demographic data of the patients (age, sex) as well the information of case history (duration of the illness, time and sequence of the symptoms, the features of the prehospital disease course) were collected prior to the surgery.

The general clinical, laboratory and clinico-neurological examinations were provided with the aim of the assessment of the peculiar features of the disease course.

The complex clinical-and-instrumental otoneurological examination and the assessment of the neurological status was performed prior to the surgery and within 4-10 days following surgery (depending on the severity of state and the consciousness level of the patient). This examination comprised the assessment of the complaints, the standard otolaryngological inspection (anterior and posterior rhinoscopy, otoscopy, oropharyngoscopy), the qualitative assessment of the innervations by the cranial nerves (CN) (І, V , VIІ, VIII, IX, X, XI, XII), and the assessment of the function of auditory and vestibular analyzers.

For the audiometry, the serial audiometer MA-51 (Germany) was used in compliance with the requirements of the standards.

MRI of the brain with the extension to the craniovertebral site of C2 vertebra was obtained on 1.5 T high-field MRI unit. The MRI protocol comprised the sequences with contrast enhancement using intravenous paramagnetic contrast agent. Based on MRI findings, the volumes of the posterior cranial fossa (PCF) and the sryngomyelia cysts were measured and the extent of the dislocation of the cerebellar tonsils through FM was assessed. These volumes were evaluated based on the analysis of the MR tomograms of PCF, cervical and thoracic divisions of the backbone.

The surgical treatment was aimed at PCF decompression. The following surgical interventions were provided: bone decompression for the inferior parts of the cerebellum at the upper cervical level; the recovery of the CSF flow at the region of craniovertebral junction; the expansion of the space in the PCF and/or craniovertebral junction. Under general anesthesia, a piece of the occipital bone was removed with the back of the cervical vertebrae 1 and/or 2 to the point where the cerebellar tonsils end. The great occipital foramen
was widened relieving the compression of the brainstem, the spinal cord, and cerebellar tonsils.

All patients were discharged in satisfactory condition for the outpatient follow-up at their places of residence.

The data were processed with the aid of STATISTICA 13 (StatSoft Inc., USA, license No. JPZ8041382130ARCN10-J) and SPSS 17.0 (IBM, USA). Taking into account a small sample size, non-parametric methods were used for comparing the quantitative data. The data are presented as median and interquartile interval (Me (Q25; Q75)). The differences between independent groups were compared by Mann–Whitney test. The strengths of association between variables was assessed by Spearman's correlation. The results were considered statistically significant at p < 0.05.

All patients provided the informed consent in written for the participation in the study according to the Declaration of Helsinki by the World Medical Association on the ethical principles for medical research involving human subjects (1964–2008), Directive 2001/20/EC on the conduct of clinical trials on human subjects, and the Order by the Ministry of Health of Ukraine No. 690 of 23.09.2009 with the amendments.

Results and Discussion

The group under study comprised 59 patients. Among them 48 (81.4%) patients had syringomyelia syndrome; 11 (18.6%) patients were without syringomyelia. There were 24 (40.7%) male patients and 35 (59.3%) female patients. The age of the patients ranged from 16 to 50 years (on average 36.9 ± 3.7 years). The mean age of male and female patients did not differ significantly (p > 0.05).

The clinical pattern of CM and syringomyelia was characterized by the symptoms of the damage of central and peripheral nervous system, the dysfunction of the visceral organs, vegetative-trophic, vegetative-vascular, and muscular-fascial disorders. Such variety of the symptoms could be explained by the damage of spinal cord that provides the connection between the central nervous system and effector organs. Furthermore, the radices of the spinal cord are directly connected with specific body areas. This pathology is also associated with the impairment of the exteroceptive, interoceptive, and proprioceptive functions. Table 1 demonstrates the pre- and postoperative distribution of the clinical symptoms in the patients under study.

### Table 1. The pre- and postoperative frequency of clinical symptoms

<table>
<thead>
<tr>
<th>Clinical symptom</th>
<th>Preoperative</th>
<th>Postoperative</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>abs. number</td>
<td>%</td>
</tr>
<tr>
<td>Headache in cervico-occipital area</td>
<td>48</td>
<td>81.4</td>
</tr>
<tr>
<td>Unilateral tinnitus aurium</td>
<td>6</td>
<td>10.2</td>
</tr>
<tr>
<td>Bilateral tinnitus aurium</td>
<td>11</td>
<td>18.6</td>
</tr>
<tr>
<td>Bilateral asymmetrical sensorineural hearing loss</td>
<td>5</td>
<td>8.5</td>
</tr>
<tr>
<td>Bilateral symmetrical sensorineural hearing loss</td>
<td>12</td>
<td>20.3</td>
</tr>
<tr>
<td>Unilateral sensorineural hearing loss</td>
<td>4</td>
<td>6.8</td>
</tr>
<tr>
<td>Objective (rotary) vertigo</td>
<td>3</td>
<td>5.1</td>
</tr>
<tr>
<td>Subjective (nonrotary) vertigo</td>
<td>26</td>
<td>44.1</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>28</td>
<td>47.5</td>
</tr>
<tr>
<td>Facial hypesthesia (paresthesia)</td>
<td>14</td>
<td>23.7</td>
</tr>
<tr>
<td>Dysfunction of caudal CNs</td>
<td>13</td>
<td>22</td>
</tr>
<tr>
<td>Statocoordinatory impairment</td>
<td>49</td>
<td>83.1</td>
</tr>
<tr>
<td>Conductive sensory disorders</td>
<td>36</td>
<td>61</td>
</tr>
</tbody>
</table>

Ukrainian scientific medical youth journal, 2023, Issue 3 (141)
http://mmj.nmuofficial.com
Preoperatively, 48 (81.4 %) patients complained of the headache in cervico-occipital area of varying intensity. The headache became more intensive after coughing, laughing, or straining (Valsalva maneuver). Such headache was caused by the compression and the involvement of dura mater and upper cervical spinal radices to the adhesive process. Only 11 (18.6 %) patients did not complain of the headache (even episodically). Hypertensive-hydrocephalic syndrome was diagnosed in 9 (15.3 %) patients in combination with the internal hydrocephaly confirmed by MRI visualization.

The impaired coordination ranks the second among the symptoms in patients with CM. In our group, the impaired coordination was diagnosed in 49 (83.1 %) patients. These impairments were not severe manifesting predominantly as staggering as well as Romberg position and intentional tremor in coordination tests. No severe manifestations of the cerebellar impairment such as the scanning speech or the decreased muscle tone due to cerebellar dysfunction were observed. Among subjective disorders in CM, the subjective vertigo is considered as the classic. Such complaints were found in 26 (44.1 %) patients. The isolated attacks of the objective (rotary) vertigo were evident in 3 (5.1 %) patients.

Nystagmus is the most frequent eye movement deficit in CM. The patients with Chiari malformation present with various types of nystagmus; each type has unique pathophysiology and central correlate. The orientation of the eye-in-orbit determines the intensity of nystagmus (Ghasia, F. F., Gulati, D., Westbrook, E. L., & Shaikh, A. G., 2014). In 28 (47.5 %) patients, the central positional nystagmus without latent period and with duration exceeding 1 minute and the alternate direction was diagnosed. More often, the nystagmus in patients was vertical and stable and characterized by the downward direction in the null position. This nystagmus intensified when the eyes displaced eccentrically or with the inclined head. Sometimes the rotatory component was present with combination of the visual and vestibular ataxia with near fall forward or backward as well as the deficit of the vertical slow consecutive movements. We believe that such symptoms are due to the compression of the stem structures and the inflow blood vessels and/or the impairment of the central vestibular-ocular reflex. The similar point of view is also argued elsewhere.

Gaze-evoked nystagmus, a common sign of cerebellar dysfunction, could then be interpreted as a disorder in a neural network that mathematically integrates a velocity (move) command into a position (holding) command (Shadmehr R., 2017). We also observed the association between the manifestations of nystagmus with the functional load: inclination of the head, Valsalva maneuver, etc.

The sensorineural hearing loss of varying grades were diagnosed in 21 (35.6 %) patients < unilateral – in 4 (6.8 %) patients, bilateral symmetrical – in 12 (20.3 %) patients, and bilateral asymmetrical – in 5 (8.5 %) patients. No cases of the conductive and mixed hearing loss were revealed. In all cases, the audiometric curves were of the descending type. The unilateral tinnitus aurium (on the side of the lesion) was evident in 6 (10.2 %) patients, the bilateral – in 11 (18.6 %) patients.

No cases of the facial nerve damage were found. Facial paresthesia was observed in 14 (23.7 %) patients due to the damage of the substantia gelatinosa of the superior cervical spinal segments and the sensory nucleus of the spinal trigeminal tract. In 13 (22 %) patients, the damage of the caudal CNs (IX, X, XI, XII) was diagnosed. In these patients, dysphagia was diagnosed more often, sometimes in combination with dysphonia and dysarthria. The taste was not impaired. The conductive sensory disorders were more pronounced when large syringomyelia cysts were present – in 36 (61 %) patients. The impairments of the pelvic organs were not evident.

The early postoperative clinical-and-otoneurological findings are presented in Table 1. The frequency of headache, statocoordinatory and vestibular-ocular-motor impairments dropped drastically as compared to preoperative data. The regression of the headache was evident in 29 (49.2 %) patients with accompanying decrease in its intensity. The complete regression of the statocoordinatory impairments was registered in 30 (50.8 %) patients. The improvement in the category of conduction sensation disorders was
also evident. While the number of patients with the impaired CN function remains practically the same, the grade of the impairments decreased.

The morphometric indices of the PCF volumes and the sizes of siringomyelia cysts calculated based on MRI data are given in Table 2. The preoperative extent of dislocation of the cerebellar tonsils ranged from 1.0 mm to 29.0 mm (mean 12.1 mm, the standard deviation 6.29). After the surgery, the cerebellar tonsils were located above the level of the great occipital foramen in all patients.

According to the literature (De Vlieger, J., Dejaegher, J., & Van Calenbergh, F., 2019) the morphometric indices that characterize the base of the skull and the axial dimensions of the brainstem and cerebellum in patients with CM point to the decreased size of the corresponding bone structures and the disproportion between the volume of the neural structures and the capacity of the PCF to fit them in.

We have analyzed the correlation between the PCF volume, the extent of the descent of cerebellar tonsils, the duration of the disease, the volume of the siringomyelia cyst and the severity of statocoordinatory and conductive sensory disorders, the headache and the impairment of CN function prior to the surgery and after surgery.

The impairment of the caudal CN function correlated with PCF volume (the smaller PCF – the more pronounced bulbar syndrome: $R = 0.62$) and with the preoperative extent of the descent of cerebellar tonsils ($R = –0.65$). Correlation with other neurological impairments was less strong.

Table 2. Pre- and postoperative morphometric indices of the PCF volumes and the sizes of siringomyelia cysts

<table>
<thead>
<tr>
<th>Index</th>
<th>PCF volumes (cm³)</th>
<th>Cyst volume (cm³)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>range</td>
<td>mean</td>
</tr>
<tr>
<td>Prior to surgery</td>
<td>110-218</td>
<td>157.95</td>
</tr>
<tr>
<td>A week after surgery</td>
<td>125-228</td>
<td>170.960</td>
</tr>
</tbody>
</table>

Correlation with other neurological impairments was less strong.

The correlation was also shown between the headache intensity and the decreased PCF volume as well as larger extent of the descent of cerebellar tonsils.

The motor impairments correlate neither with PCF volume nor with extent of the descent of cerebellar tonsils. Instead, they correlated with the volume of the siringomyelia cyst ($R = 0.89$) and less strongly with other neurological impairments. The conductive impairments of the surface sensitivity correlated only with the volume of the siringomyelia cyst ($R = –0.83$).

The small sample size in our study seems to limit the statistical capabilities. As a result, we could not be able to assess all factors affecting the outcome of the surgical treatment. In future studies, we are going to extend the follow-up period and to assess the clinical-and-otoneurological status depending of the patient's age, the time of symptoms onset, the pattern and the extent of the dysfunctions. The present study demonstrated that most of the clinical signs and symptoms in patients with CM are associated with otolaryngological disturbances (hearing impairment, tinnitus, hoarseness, vertigo, etc.) and cerebellar signs (ataxia, nystagmus). Taking into account non-specific signs and symptoms of the disease, the otoneurological evaluation in these patients is undoubtedly of high importance for the preoperative differential diagnosis as well as the postoperative assessment of treatment outcome. The further study of the problem will allow us to timely diagnose CM, to predict the course of the disease and the risk of the possible complications providing the clinicians with important information for decision-making on the optimal treatment strategy.
Conclusions
1. Most of the clinical signs and symptoms in patients with CM are associated with otolaryngological disturbances (hearing impairment, tinnitus, hoarseness, vertigo, etc.).
2. Clinical-and-instrumental otoneurological examination is an important component of the systemic assessment of the clinical-and-functional status of patients with CV and syringomyelia that allows for objectifying the dynamics of the otoneurological aspects of the disease in the course of the treatment.
3. The aim of the surgical intervention in CM should be aimed at the decompression of the inferior parts of the cerebellum, the recovery of the CSF flow at the region of craniovertebral junction, and the expansion of the space in the fossa cranii posterior and/or craniovertebral junction.

Acknowledgment
The authors acknowledge the Academician of the NAMS of Ukraine Yevhenii Heorhiiovych Pedachenko for his longstanding support and supervising.

Financing
This study has not been supported by the external funding.

Conflict of interests
The authors have no conflict of interests to declare.

Consent to publication
All authors read the text of the manuscript and agreed with its publication.

ORCID ID and author contribution
(A, B, D, E, F) Sofia Levchenyuk 0000-0003-0830-1169 (A, B, C, D, E, F) Yurii Derkach
A – Research concept and design, B – Collection and/or assembly of data, C – Data analysis and interpretation, D – Writing the article, E – Critical revision of the article, F – Final approval of the article

REFERENCES
Клініко-отоневрологічне оцінювання результатів хірургічного лікування мальформації Кіарі та сирингомієлії в ранньому післяоперативному періоді

Левченюк Софія1, Деркач Юрій2

1Лікар-отоларинголог групи отоневрології державної установи «Інститут нейрохірургії ім. акад. А.П. Ромоданова Національної академії медичних наук України»
2Канд. мед. наук, лікар-нейрохірург відділення патології спинного мозку державної установи «Інститут нейрохірургії ім. акад. А.П. Ромоданова Національної академії медичних наук України»

Address for correspondence:
Levchenyuk Sofia
E-mail: mdslevchenyuk@gmail.com

Анотація: попри тривале вивчення патології мальформації Кіарі, багато питань й дотепер залишаються невирішеними, зокрема недостатня вивченість клінічних проявів даної патології та відсутність єдного підходу до оптимального комплексу діагностичних заходів і вибір оптимального хірургічного втручання залишається предметом дискусій у науковій літературі. Дослідження ґрунтується на ретропроспективному аналізі результатів обстеження та лікування 59 пацієнтів, які знаходились на лікуванні у відділенні патології спинного мозку державної установи «Інститут нейрохірургії ім. акад. А.П. Ромоданова НАМН України» з 2010 по 2020 рр. З них чоловіків було 24 (40,7%), жінок – 35 (59,3%). Середній вік пацієнтів складав від 16 до 50 років (в середньому 36,9±3,7 років). Всі пацієнти обстежені відповідно існуючим стандартам, з метою оцінки особливостей перебігу захворювання до- та після операції. Застосовано методи хірургічного лікування: кісткова декомпресія нижніх відділів мозочка, верхньошийного рівня; відновлення лікворотоку в області краніовертебрального переходу; збільшення загального обсягу задньої черепної ямки та або КВП. Обробку та аналіз даних проводили за допомогою програми STATISTICA 13, порівняння за кількісними ознаками здійснювали з застосуванням непараметричних методів. Метою дослідження був аналіз клініко-отоневрологічного статусу пацієнтів з МК та сирингомієлією по до оперативного втручання та в ранньому післяоперативному періоді. Визначалась особливості клінічної картини, більшість клінічних ознак і симптомів МК, пов’язані із отоларингологічними порушеннями (порушення слуху, шум у вухах, запаморочення тощо); з мозочковими порушеннями (ригідність, ністагм). Через неспецифічні ознаки і симптоми захворювання, клініко-інструментальне отоневрологічне дослідження, важливо виявити для диференційнії діагностики на доопераційному етапі та післяоперативних результатів хірургічного лікування. Подальше вивчення цієї проблеми зумовить можливість своєчасно діагностувати МК і прогнозувати подальший перебіг захворювання, а також досягнути вивчення фізіологічних ускладнень, отримати важливу інформацію для вибору оптимальної тактики лікування. Клініко-інструментальне отоневрологічне дослідження, важлива складова, системного оцінювання клініко-функціонального статусу пацієнта з сирингомієлією викликає значний інтерес, союз правовий в процесі лікування.

Ключові слова: Мальформація Кіарі, головний біль, слух, запаморочення, шум у вухах.