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Clinical-Neurophysiological Aspects Of The Chiari Malformation Type 1

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Abstract :The work presents the results of the clinical and neurophysiological examination of 207 patients with Chiari malformation of type 1. The subjective and objective neurological symptoms of Chiari malformation type 1 were analyzed in detail, depending on the degree of ectopy of cerebellum tonsils according to MRI data. Neurophysiological criteria for clinical syndromes of Chiari malformation 1 have been identified in the study of evoked potentials (BAEP, SSEP, ENMG). The Clinical Neurophysiological Scale (CNPHS) has been proposed for objective assessment of the degree of neurological deficit and choice of conservative or surgical tactics.
Keywords: ectopy of tonsils, brainstem Auditory Evoked Potentials (BAEP), median nerve somatosensory evoked potentials (SSEP), Clinical Neurophysiological Scale (CNPHS)

1. INTRODUCTION

The wide development of modern methods of neuroimaging gave an impulse to increase the detection of central nervous system anomalies (3,5,6). Nowadays, the "gold standard" for Chiari malformation diagnosis is MRI research, which allows identifying craniovertebral transition pathology, in particular accurately, different types of Chiari malformations, often combined with the basilar impression, with the pathology of liquor spaces and structural bone disorders of the atlantooccipital articulation (1,4,5,6). Neurological disorders in craniovertebral junction malformation are extremely diverse and consist of both brainstem compression and liquorodynamic disorders.

Neurological manifestations in Chiari malformation type 1 have been described in sufficient detail in the literature. Many authors distinguish from 3 to 6 variants of clinical syndromes (3,5). The analysis of neurological disorders dynamics does not always provide objective information. Still, a more informative functional quantitative scale of neurological symptomatology assessment, proposed by Egorov and co-author, 2002 (3), determines the dynamics of neurological disorders in the pre- and post-operative period in CM1 type.

Unfortunately, there is still no unified system for assessing the degree of functional deficit in this pathology. At the same time, the high proportion of subjective neurological disorders makes it difficult to determine the degree of compensation for the disease (2,5). The question - to observe or to operate? and if to operate, what volume of surgical treatment to choose? - is extremely important in the modern practice of neurologists and neurosurgeons. Application of modern neurophysiological methods of diagnostics allows us to objectively

define the level of pathological disorders and the degree of functional involvement of these or those anatomical structures. Comparisons of subjective, objective disorders in CM1 type and neurophysiological disorders according to BAEP, SSEP and ENMG data facilitate the choice of conservative and surgical tactics. However, we have not found such results in the literature, which was the reason for our research.

2. MATERIAL AND METHODS

207 patients with Chiari malformation 1 were examined according to MRI data, outpatient and inpatient treatment at the Republican Specialized Scientific-Practical Medical Center of Neurosurgery of the Ministry of Health of RU. The standard in determining the degree of cerebellar almond prolapse in cases of Chiari malformation was the Chamberlain line running from the hard palate to Opistion (a point located in the center of the rear edge of the FOM) (2,8,9). We considered acceptable the displacement of cerebellum tonsils beyond the Chamberlain line to be 5 mm. In our studies, we used the Chamberlain line to guide the anatomical anomalies of the craniovertebral transition and the degree of ectopy of cerebellar tonsils.

We analyzed clinical symptoms in 207 patients with Chiari malformation type 1 (CM 1 type) according to MRI study. Among them, 73 men and 134 women aged 14 to 62 years. All patients were divided into groups according to the degree of displacement of cerebellar tonsils below the Chamberlain line. The first group consisted of 17 patients with ectopy of 0-5 mm, and the second group consisted of 75 patients with ectopy of 6-10 mm, the third group consisted of 84 patients with ectopy of 11-15 mm, in the fourth group 31 patients had the displacement of more than 15 mm according to MRI scan.

All patients were examined using a multimodal neurophysiological protocol that includes acoustic stem evoked potentials (BAEP), somatosensory evoked potentials (SSEPs) and motor evoked potentials (ENMG) (7). The research was conducted on a 4-channel complex “Synapsis” (Neurotech, Russia) with computer data processing.

The standard vertex-mastoid removal (M1-Cz, M2-Cz) was used for the BAEP, the stimulation was performed through headphones with 0.1 ms biurally with 20 Hz feed frequency and 70 dB sound. In SSEP, the withdrawal electrodes were installed using the standard C4-Fz method - when n.medianus S C3-Fz was stimulated - when n.medianus D was stimulated. Stimulation was performed by electrical pulses in the projection of the median nerve at the wrist level with a current of 15-20 mA, frequency 2 Hz.

We performed the default stimulation EMG for the lingual-throat and accessory nerves with the setting of the recording electrodes according to the muscle innervation presented below. If necessary, we supplemented the nerves under study based on the clinical syndrome.

Cranial nerve-innervation of the muscles
III, IV, VI - Extraocular muscles
V - Masseter, temporalis
VII -Frontalis, orbicularis oculus, orbicularis
oris, mentalis, others
IX - Stylopharyngeus
X - Pharyngeal and laryngeal muscles
XI - Sternocleidomastoid, trapezius
XII – Tongue

3. RESULTS AND DISCUSSION

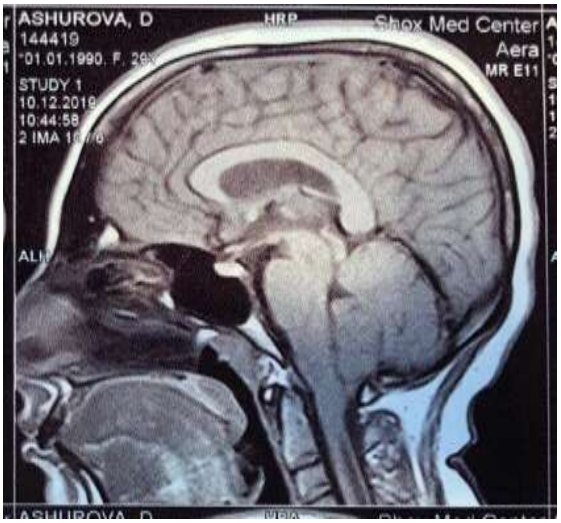
All the examined patients had objective and subjective symptoms consisting of a combination of cerebellar, stem and liquorodynamic disorders. Table 1 suggests the distribution of the main subjective clinical symptoms by groups in patients with CM1 type depending on the degree of ectopy of cerebellar tonsils below the Chamberlain line according to MRI data.

Table 1. Subjective clinical symptoms of CM1 type in comparison with the degree of cerebellar tonsils prolapse

Tonsil ectopy, mm	**Ⅰ gr. 0-5 mm (n=17)**		**Ⅱ gr. 6-10 mm (n=75)**		**Ⅲ gr. 11-15 mm (n=84)**		**Ⅳ gr. More than 15 mm (n=31)**	
General symptoms	number	**%**	number	**%**	number	**%**	number	**%**
Headache -neck and occipital -dotted -retroorbital	 13 - 3	 76.5 - 17.6	 52 14 9	 69.3 18.7 12.0	 58 12 10	 69.0 14.3 11.9	 29 - 2	 93.5 - 6.5
Coordination and vestibular symptoms								
giddiness	12	70.6	47	62.3	62	73.8	16	51.6
Nausea	8	47.0	14	18.7	18	21.4	9	29.0
Tinnitus	4	23.5	21	28.0	35	41.7	10	32.3
Ophthalmologic symptoms								
blurred vision	8	47.0	22	29.3	18	21.4	11	35.5
Double	-	-	4	5.3	3	3.6	-	-
Bulbar symptoms								
throat lump feeling	2	11.8	12	16.0	25	29.8	14	45.2
swallowing difficulties	-	-	4	5.3	14	16.7	8	25.8
Snorting	-	-	9	12.0	11	13.1	7	22.6
Sensitive disorders								
numb face	1	5.9	7	9.3	5	5.95	2	6.45
tongue numbness	-	-	3	4	8	9.5	4	12.9
numbness in the extremities	6	35.3	38	50.7	61	72.6	27	87.0
body numbness	-	-	17	22.7	36	42.9	25	80.6
facial pain	2	11.8	7	9.3	10	11.9	-	-
pain in the limbs	4	23.5	67	89.3	70	83.3	18	21.4
diffuse pain throughout the body	2	11.8	53	70.7	32	38.0	20	64.5
Motor disorders								
upper limb weakness	-	-	14	18.7	27	32.1	18	58.0
lower limb weakness	-	-	16	21.3	24	28.6	14	45.2

gait instability	1	5.9	8	10.7	17	20.2	14	45.2
Vegetative disorders								
shortness of breath	8	47.0	35	46.7	45	53.8	9	29.0
cold feeling in the limbs	10	58.8	22	29.3	39	46.4	12	38.7

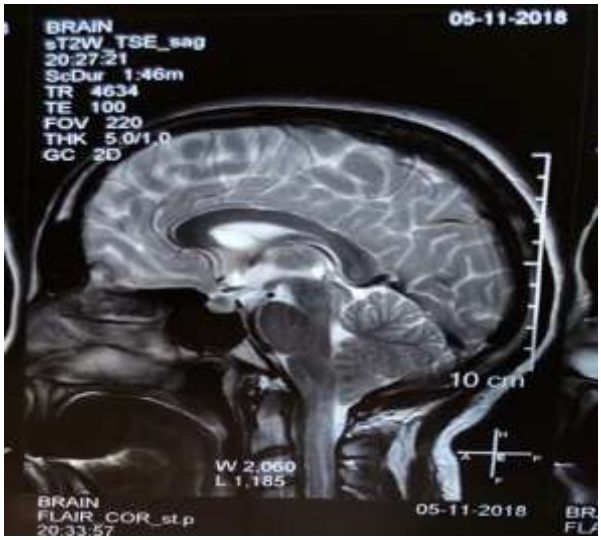
The I group included 17 patients with a small degree of ectopy of cerebellar tonsils below the Chamberlain as per MRI. Fig.1 shows the patient's MRI image with a 4 mm displacement of cerebellar tonsils.



Pic. 1. Example MRI of patient A., with a 4 mm almond prolapse on 2 sides below the Chamberlain level.

As it can be seen from the presented table in the group of patients with 0-5 mm low tonsils prolapse, the main subjective clinical symptoms were general cerebral and vegetative disorders. The majority of 13 (76.5%) patients of this group were disturbed by the headache of cervical-occipital localization related to the head position. Complaints of dizziness were present in 12 (70.6%) patients of I group and often, in 47% of cases, were accompanied by nausea. Subjective eye symptoms in the form of blurred and blurred vision were observed in 8 (47%) patients. Numbness and pain in the extremities dominated in 6 (35.3%) and 4 (23.5%) patients of I group, respectively. Patients of this group practically made no complaints about the motor sphere. Vegetative disorders were characteristic of clinical manifestations in the group with small ectopy of tonsils. The feeling of air shortage was complained about by 8 (47%) patients, and coldness of limbs disturbed 10 (58.8%) persons in the I group. Thus, the group of patients with ectopic tonsils of the cerebellum was dominated by subjective symptoms of headache, dizziness, numbness and pain in limbs, as well as feelings of air shortage and coldness in limbs.

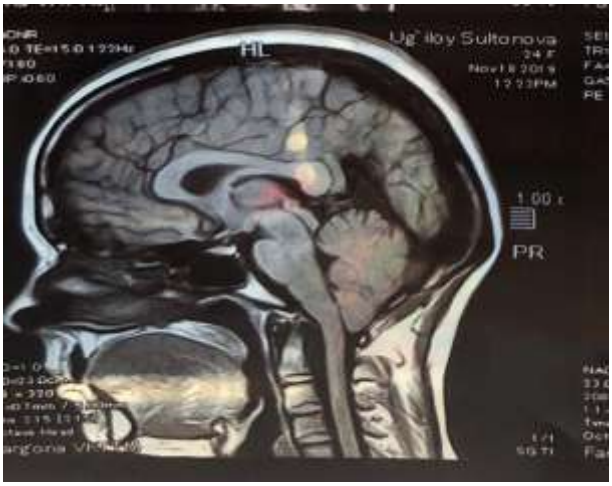
The II group included 75 patients with moderate depression of cerebellar tonsils 5-10mm below the Chamberlain level according to MRI data. Fig.2. suggests an MRI image of patient B, with ectopy of cerebellum tonsils by 7 mm at CM1 type.



Pic.2. Example of MRI of the brain of patient B. with the displacement of cerebellar tonsils 7mm below the Chamberlain line from 2 sides for type 1 AK.

As it follows from Table 1., in the structure of subjective clinical symptoms in patients of the Ⅱ group headache of cervical-occipital localization prevailed in 52 (69.3%) patients, parietal and retroorbital pain occurred in 18.7% and 12.0% of cases respectively. In 47 (62.3%) patients of the Ⅱ group, systemic dizziness was observed, with tinnitus disturbing 21 (28.0%) patients with ectopy of 5-10mm, nausea symptoms were much rarer in 14 (18.7%) subjects examined. Subjective eye disorders in the form of blurred vision were observed in one third of the patients (29.3%) in the Ⅱ group. Sense of lumpiness in the throat, hypertension and difficulty in swallowing were noted in no more than 12 (16.0%) patients with moderate tonsil lowering by MRI. Attention is drawn to the predominance of sensitive disorders in the Ⅱ group. Numbness and pain in extremities were disturbed by 67 (89.3%) patients in this group, complaints about diffuse pain in the whole body were made by 53 (70.7%) subjects. Motor disorders were much rarer in comparison with those in the sensitive group. Weakness in the upper extremities was subjectively noted by 14 (18.7%) patients, while weakness in the lower extremities was disturbed by 16 (21.3%) subjects of the II group. 8 (10.7%) of patients in this group complained about a feeling of instability while walking. Vegetative disturbances in the form of air shortage and coldness of the limbs were observed in 35 (46.7%) and 22 (29.3%) patients of this group, respectively. Considering the above mentioned, we can say that in CM1 type patients with moderate ectopia of cerebellum tonsils 5-10mm according to MRI data headache, different localizations in combination with vestibular disorders in the form of dizziness and tinnitus prevail in the structure of subjective clinical symptoms. Sensitive disorders in the form of numbness and pain in the extremities, as well as diffuse pain throughout the body, are of the greatest importance. Vegetative disorders in patients in the II group occurred in less than half of the observations and were not of primary importance.

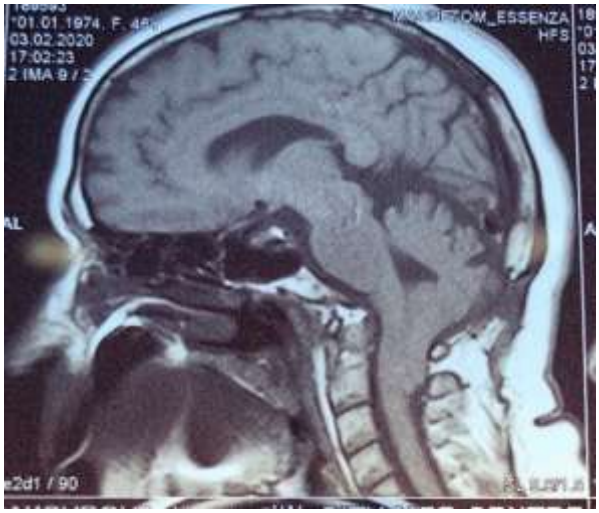
Further, we analyzed subjective clinical symptoms in 84 patients of Ⅲ group with significant displacement of cerebellar tonsils 11-15mm below Chamberlain line according to MRI study. A picture of the brain MRI of patient C., which belongs to this group of patients, is presented in Pic.3.



Pic.3. Example MRI of patient C. with type 1 Chiari malformation with 14mm cerebellum ectopy of tonsils on the right and 12mm on the left.

In Ⅲ group of patients' headache structure was dominated by cervical-occipital localization headache in 58 (69.0%) patients. Dark and retroorbital pain was observed in almost the same number of cases and was 14.3% and 11.9%, respectively. Patients with pronounced ectopia of cerebellum tonsils were disturbed by systemic dizziness in 62 (73.8%) patients, tinnitus was observed in 35 (41.7%) patients of this group, nausea accompanied 18 (21.4%) cases. Complaints about blurred vision were present in 18 (21.4%) of the surveyed Ⅲ groups. Bulbar symptoms were noted in the form of a coma feeling in the throat and difficulties in swallowing in 25 (29.8%) and 14 (16.7%) patients with a pronounced displacement of cerebellar tonsils at type CM1, respectively. Numbness in the extremities prevailed in 61 (72.6%) patients with sensory disorders. Complaints of extremity pain, a burning character in 70 (83.3%) subjects of this group require attention. Numbness and diffuse pains in the whole body were significantly rarer in 36 (42.9%) and 32 (38.0%) patients in the Ⅲ group. In the structure of subjective motor disorders, 27 (32.1%) patients of this group had weakness in the upper extremities. In comparison, weakness in the lower extremities and subjective gait instability was present in 28.6% of cases. Vegetative disorders disturbed almost half of the patients in the Ⅲ group. The feeling of air shortage was complained about by 45 (53.8%) subjects, while the coldness of limbs was noted by 39 (46.4%) patients. In general, subjective clinical symptoms in the group of patients with pronounced ectopia of cerebellum tonsils at CM1 type consisted of the headache of different localization, vestibular, coordination disorders. Dissociated sensory disturbances were a characteristic feature of this group, while motor disorders were observed to a lesser extent despite pronounced displacement of cerebellar tonsils.

The Ⅳ group included 31 patients with type CM1 with an acute displacement of cerebellar tonsils exceeding 15 mm according to MRI study. An example of the MRI scan of patient Z from this group is presented in Pic.4.



Pic.4. MRI of patient Z., 34 years old with craniovertebral transition anomaly with the displacement of cerebellar tonsils below Chamberlain line up to 24 mm from 2 sides.

As it follows from Table 1., above, in subjective clinical symptoms in Ⅳ patients the headache of cervical-occipital localization prevailed in 29 (93.5%) patients, which in almost all subjects were connected with head and body positions, which, in our opinion, was caused by cervical myofascial disorders. Complaints of dizziness and tinnitus were present in 16 (51.6%) and 10 (32.3%) patients of this group, respectively. A variety of complaints of difficulty in swallowing, lumpy throat and hypertension were reported in 45.2%-22.6% of cases. The range of subjective sensory disorders was very wide - the patients complained of numbness in the tongue, the individuals in 12.9% of cases, and numbness in extremities and body was noted by 27 (87.0%) and 25 (80.6%) patients in Ⅳ group. Diffuse pains in the whole body were reported in 20 (64.5%) cases. One of the important subjective symptoms in this group were motor disorders in the form of upper extremity weakness prevalence in 18 (58.0%) patients and lower extremity weakness in 14 (45.2%) patients of this group. 12 (38.7%) patients of Ⅳ group had a cold feeling in their extremities. Summarizing the above mentioned, in the structure of subjective clinical symptoms in patients of CM1 type with the sharply defined displacement of cerebellum tonsils over 15 mm, sensitive and motor disorders, expressed in upper limbs, prevailed. Bulbar symptoms were equally combined with vestibular disorders and had moderate severity.

Thus, subjective clinical symptoms in patients with CM1 type had different degrees of severity and were directly correlated with the degree of ectopy of cerebellar tonsils according to MRI research. Minimal ectopy was dominated by focal, vestibular and vegetative disorders, while moderate tonsil prolapse was accompanied in most cases by vestibular and sensitive disorders. Subjective combination of focal, bulbar and sensory disorders was revealed in patients with pronounced ectopia of cerebellum tonsils in CM1 type. In the coarse prolapse of cerebellar tonsils, over 15 mm MRI data revealed significant sensitive and motor disorders against the background of moderate bulbar symptomatology.

We analyzed the spectrum of objective neurological disorders in 207 patients with CM1 type depending on the degree of ectopy of cerebellar tonsils according to MRI data, the results of which are presented in Table 2.

Table 2. Objective clinical symptoms of CM1 in comparison with the degree of amygdala cerebellum prolapse.

	Ⅰ gr. 0-5 mm	**Ⅱ gr. 6-10 mm**	**Ⅲ gr. 11-15 mm**	**Ⅳ gr. more than 15**

	(n=17)		**(n=75)**		**(n=84)**		**mm (n=31)**	
Focal and vestibular symptoms	**number**	**%**	**number**	**%**	**number**	**%**	**number**	**%**
nystagmus -spontaneous setup	-	-	2	2.67	5	5.95	-	-
-horizontal	7	41.1	18	24.0	20	23.8	3	9.68
-rotator	1	5.88	10	13.3	8	9.52	4	12.9
-vertical	-	-	5	6.67	12	14.3	6	19.4
- “hitting down.”	2	11.8	27	36.0	38	45.2	14	45.2
- “kicking up.”	-	-	1	1.33	3	3.57	-	-
static ataxy	4	23.5	32	42.7	37	44.0	8	25.8
dynamic ataxy	6	35.3	38	50.7	40	47.6	11	35.5
Symptoms of cranial nerve damage								
Diplopia	-	-	1	1.33	5	5.95	2	6.45
face hypoesthesia	3	17.6	12	16.0	14	16.7	4	12.9
dysphagia	-	-	4	5.33	20	23.8	11	35.5
dysarthria	-	-	2	2.67	12	14.3	4	12.9
dygevsia	-	-	2	2.67	12	14.3	4	12.9
palate paresis	-	-	4	5.33	21	25.0	10	32.3
tongue paresis	-	-	2	2.67	15	17.9	6	19.4
neck muscle paresis	-	-	-	-	4	4.76	6	19.4
Reflexive sphere								
decrease or absence of corneal reflex	-	-	8	10.7	12	14.3	2	6.45
decrease or absence of the throat reflex	-	-	4	5.33	18	21.4	10	32.3
decrease or absence of BR,TR	-	-	-	-	22	26.2	18	58.0
decrease or absence of PR, AR	-	-	-	-	7	8.33	6	19.4
decrease or absence of abdominal reflexes	-	-	-	-	16	19.0	12	38.7
BR,TR increase	-	-	4	5.33	14	16.7	10	32.3
PR increase	2	11.8	8	10.7	28	33.3	10	32.3
pathological reflexes	-	-	2	2.67	14	16.7	10	32.3
	-	-	-	-	8	9.52	10	32.3
	-	-	2	2.67	16	19.0	8	25.8

Sensitive disorders								
radicular hypoesthesia	-	-	-	-	14	16.7	1	3.26
dissociated	-	-	10	13.3	22	26.2	6	19.4
segmental "jackets"	-	-	7	9.33	16	19.0	12	38.7
dissociated	-	-	-	-	4	4.76	-	-
segmented into "semi-jackets"	-	-	-	-	8	9.52	4	12.9
dissociated	-	-	5	6.67	10	11.9	7	22.6
Motor disorders								
sluggish upper monoparesis	-	-	5	6.67	18	21.4	4	12.9
flaccid top paraparesis	-	-	12	16.0	20	23.8	12	38.7
sluggish lower monoparesis	-	-	-	-	-	-	-	-
flaccid lower paraparesis	-	-	-	-	5	5.95	4	12.9
spastic lower paraparesis	-	-	9	12.0	12	14.3	-	-
spastic tetraparesis	-	-	-	-	8	9.52	7	22.6
spastic hemiparesis	-	-	-	-	7	8.33	-	-
upper limb hypotrophy	-	-	10	13.3	20	23.8	17	54.8
lower limb hypotrophy	-	-	-	-	4	4.76	8	25.8
fibrillations in -upper limbs -lower limbs -the body	 - -	 - -	 2 -	 2.67 -	 12 -	 14.3 -	 7 2	 22.6 6.45
	-	-	-	-	5	5.95	5	16.1
Vegetative disorders								
heart rhythm disorder	10	58.8	22	29.3	18	21.4	14	45.2
respiratory disorder	7	41.2	18	24.0	15	17.9	12	38.7
hyperhidrosis	12	70.6	31	41.3	27	32.1	8	25.8
acrocyanosis	5	29.4	16	21.3	4	4.76	8	25.8
dermographism	8	47.1	7	9.33	12	14.3	4	12.9
Pelvic disorders								
in terms of delay	-	-	-	-	8	9.52	7	22.6
incontinent	-	-	5	6.67	4	4.76	6	19.4

In the first group of patients with minimal ectopia of tonsils, up to 5 mm below the Chamberlain line in objective examination in 7 (41.1%) cases, the horizontal nystagmus was noted, with the eyeballs aside by 30 degrees from the central position (setting). Rotary nystagmus was observed only in a single case, while the “beating down” nystagmus was a pathognomonic symptom of Chiari malformation in 2(11.8%) of patients in this group. In 4 (23.5%) I patients of the group unsteadiness in the Romberg position was observed, intentionality and failure at palcenosis and heel knee test were revealed in 6 (35.3%) cases.

Hyposthesia of the half of the face was determined in only 3(17.6%) of the subjects of this group from the symptoms of cranial nerve damage. Other disorders of cranial nerve function, particularly caudal nerve function, such as dysphagia, dysarthria, tongue and soft palate muscle paresis, were not observed in patients with a small cerebellar tonsil displacement. In the study of the reflex sphere in Ⅰ patients, no malformation was detected. Cervical and pharyngeal reflexes were preserved, tendon reflexes from upper and lower limbs were induced normally, pathological signs were absent. Objective sensory disorders in the form of dissociated pain and temperature sensitivity disorder in patients of Ⅰ group were not determined. Complex disorders of the sensitive sphere by the alternative type were not detected. Motor disorders of mono-, para-, tetraparesis type were not identified, muscle trophicity was preserved, pathological muscle twitches (fibrillation and fasciculosities) were not detected. The diversity of vegetative disorders prevalent in patients with ectopic tonsils up to 5 mm requires attention.

Thus, cardiac rhythm disturbance was observed in 10 (58.8%) patients of this group, tachypnea disorders and night apnea elements were observed in 7 (41.2%) subjects. Palm hyperhidrosis was detected in an objective study in 12 (70.6%) cases, dermographism was noted in 8 (47.1%) observations. No pelvic disorders were noted in this group of patients with CM1 type.

Thus, in the structure of objective neurological disorders in patients of the first group of CM1 type with the displacement of cerebellum tonsils up to 5 mm coordination and vestibular disorders with clear manifestations of vegetative dysfunction prevailed. Objective sensory and motor disorders were not detected, although, in the above analysis, the specific weight of subjective sensory disturbances was rather high. Subjective symptoms were prevalent over objective neurological data in this group of patients.

Further, we studied neurological symptoms in 75 patients of the Ⅱ group with CM1 type with the displacement of cerebellum tonsils 6-10mm below the Chamberlain line. An objective analysis of vestibular and focal disorders revealed the presence of spontaneous nystagmus in only 2 (2.67%) patients in this group. In comparison, the setting nystagmus was found in 18 (24%) subjects, and the “kicking down” nystagmus characteristic of the Chiari malformation was observed in 27 (36.0%) patients of the Ⅱ group. Static ataxia phenomena were noted in 32 (42.7%) observations, and signs of progressive ataxia were determined in practical cases in half of 38 (50.7%) patients of this group. Objective dysfunctions of cranial nerves were presented in a rather diverse way. Soft palate dysphagia and paresis were determined in 4 (5.33%) patients of the II group, dysarthria and dysgeusia in 2 (2.67%) cases. The largest number of 12 (16.0%) patients revealed hypoesthesia in half of the face by zones of Zelder. Disorders in the reflex sphere were revealed in 8 (10.7%) cases of corneal reflex absence, a decrease in the pharyngeal reflex was noted in 4 (5.33%) patients of this group. In the study of tendon reflexes in 8 (10.7%) patients, a symmetrical increase of the knee reflex was revealed, and in 4 (5.33%), both biceps and triceps reflexes were increased. Pathological signs in the form of symptoms of oral automatism and positive foot reflexes of Babinsky were observed in only 2 (2.67%) patients in this group. In the structure of objective sensory disorders, segmental dissociated disorders of pain and temperature sensitivity by the type of “jacket” were noted in 10 (13.3%) patients of the Ⅱ group, and the symptom of “half jacket” was revealed in 7 (9.33%) cases. Alternating sensitiveness disorders in 5 (6.67%) patients of the examined group according to the Wallenberg-Zakharchenko syndrome type with sensitivity disorder on the face. According to the segmental type on one side and hemianesthesia on the opposite side, according to the conductive type, indicating possible compression of stem structures by displaced amygdala cerebellum in CM1 type, require special attention. Motor impairment was detected in 12 (16.0%) patients in the form of

sluggish upper paraparesis, while sluggish upper monoparesis was observed in only 5 (6.67%) cases. Sluggish lower paraparesis was not determined in this group, whereas spastic lower paraparesis prevailed in 9 (12.0%) patients of the Ⅱ group. Slimming and hypotrophy of the upper limb muscles was observed in 10(13.3%) patients and was more often asymmetrical due to different degrees of pathological involvement of the segmental apparatus. In single cases in 2 patients (2.67%), there were observed fibrillations in the upper extremities indicating the dysfunction of anterior spinal structures at the level of the cervical section. Objective dysfunctions of the vegetative sphere were found in a rather large number of observations. Thus, the disturbance of heart rhythm was registered in 22 (29.3%) patients, and the change of breathing frequency in the form of tachypnea was registered in 18 (24.0%) patients of this group. Hyperhidrosis and acrocyanosis were registered in 31 (41.3%) and 16 (21.3%) cases, respectively. Pelvic disorders in the form of urinary incontinence were observed in 5 (6.67%) patients of the II group, which in our opinion could be due to fixation of the spinal cord at the lumbosacral level, which is often combined with Chiari anomaly. Thus, in the group of patients with ectopy of cerebellar tonsils 6-10mm below the Chamberlain line, objective neurological symptomatology consisted of vestibular-coordination disorders, sensitive disorders of dissociated segmental type, peripheral pauses in upper limbs, and moderate vegetative dysfunction. We analyzed objective neurological symptoms in 84 patients of Ⅲ group with ectopy of cerebellum tonsils 11-15mm below the Chamberlain line level. In the structure of vestibular disorders, there were revealed nystagmus of different natures: spontaneous nystagmus was observed in 5 (5.95%) patients, setting horizontal nystagmus prevailed in 20 (23.8%) patients of this group, rotator nystagmus was observed in 12 (14.3%) cases. In almost half of the examined 38 (45.2%) nystagmus, “beating down” was observed, which is a defining sign of CM1 type. The phenomena of static ataxia in the form of instability in the Romberg posture were recorded in 37 (44.0%) cases, and progressive ataxia with the difficulty of performing finger and heel knee tests was noted in 40 (47.6%) patients of Ⅲ group. The symptoms of cranial nerve lesions were also widely represented in this group. Thus, diplopia and restriction of eyeball motility to the side were found in 5 (5.95%) patients, half-face hypoesthesia was found in 14 (16.7%) cases. The most pronounced symptoms of dysfunction of the caudal nerve group were dysphagia in 20 (23.8%) patients and dysarthria and dysgeusia in 12 (14.3%) subjects of this group. Muscle paralysis of the soft palate was pronounced in almost a quarter of the Ⅲ group patients in 21 (25%) cases. Disorders of the reflex sphere were observed in almost all patients of this group. A decrease in corneal reflex was observed in 12 (14.3%) subjects of the examined group. In comparison, the absence of the throat reflex was observed in 18 (21.4%) patients of Ⅲ group, which indicated a significant impairment of functions of the oblong brain structures in CM1 type. Phenomena of hyporeflexion in the form of tendon reflexes decrease in upper and lower limbs were found in 22 (26.2%) and 7 (8.33%) patients, respectively. The increase of knee reflexes in 28 (33.3%) patients of this group in combination with pathological foot signs was noticed in 16 (19.0%) patients, which was an objective symptom of pyramidal pathway insufficiency in CM1 type. The structure of objective sensory disorders was dominated by segmental dissociated disorders by the type of “jacket” in 22 (26.2%) cases, dissociated segmental disorders by the type of “half jacket” were observed in 16 (19.0%) patients examined. Alternating sensitive disorders of Babinskiy-Najott and Wallenenberg-Zakharchenko syndrome type were present in 10 (11.9%) patients of this group, which indicated the pathology of stem structures on the border of the bridge valley and elongated brain.Conductive hypoesthesia was observed in 8 (9.52%) subjects and was mainly due to the presence of syringomyelitis in the spinal cord. The prevalence of sluggish upper paraparesis was observed in 20 (23.8%) patients, while sluggish monoparesis

was observed in 18 (21.4%) cases. Spastic motor disorders in the form of lower paraparesis and tetraparesis were present in 12 (14.3%) and 8 (9.52%) patients in this group, respectively. In the majority of cases 23.8% of muscular trophicity evaluation was accompanied by upper limbs muscular hypotrophy, more often in distal sections bilaterally. Pathological fibrillation in the shoulder belt muscles was noted in 12 (14.3%) patients, whereas fibrillation in the lower limbs in Ⅲ group was absent.

Vegetative disorders in the form of cardiac and respiratory rhythm disorders, increased sweating to varying degrees were detected up to 32.1% of cases in this group. Pelvic dysfunction by type of delay was twice as prevalent over incontinence type dysfunction (8 and 4 patients), which indicated the predominance of conductive disorders from craniovertebral transition level or cervical spinal cord level. It can be assumed that physical, neurological symptoms with pronounced ectopy of cerebellar tonsils 11-15 mm below the Chamberlain line consist of symptoms of caudal group lesions, dissociated segmental sensory disorders, in more severe cases with the formation of intermittent syndromes. Motor disorders in Ⅲ group have both segmental and conductive character, due to conduction disturbance both at the level of the medulla oblongata (pyramid crossover) and the presence of syringomyelic cavities (the effect of which we will consider in subsequent chapters of our study).

We conducted a symptomological analysis in 31 patients Ⅳ group with ectopy of cerebellar tonsils over 15mm below the Chamberlain line. The predominance of "kicking down" nystagmus was observed in 14(45.2%) patients of this group, while horizontal and rotator nystagmus were much rarer in 9.68% and 12.9% of cases respectively. Static ataxia was observed in 8(25.8%) subjects and progressive ataxia was observed in 11(35.5%) subjects. The dominant symptoms of caudal nerve lesions such as dysphagia and soft palate paresis were observed in 11(35.5%) subjects, while dysarthria and dysgeusia were observed in 12.9% of cases. Paralysis of the tongue and neck muscles was observed in 6 patients (19.4%), which also indicated lesions of the lower stem structures.

In the structure of reflex disorders, the absence of the throat reflex was 32.3%, while the corneal reflex was not caused in only 6.45% of Ⅳ patients. The tendinous reflexes in the upper limbs were sharply reduced in 18(58.0%) patients. The hyporeflexion in the lower limbs was observed in 6(19.4%) of the subjects. The biceps, triceps and knee reflexes were equally increased in 32.3% of cases. Pathological signs in the form of symptoms of oral automatism (palm chin and trunk reflex) were caused in 10(32.3%) of Ⅳ patients, which may be due to lymphatic dysfunction and encephalopathic development. Sensitive disorders in the form of segmental dissociated disorders of the "jacket" type were detected in 6(19.4%) patients of this group, and the "half jacket" symptom was detected twice as often in 12(38.7%) subjects. Alternating sensitivity disorders of the Wallenberg-Zakharchenko syndrome type also prevailed over conductive disorders, which confirmed the predominant lesion of stem structures with a sharply pronounced displacement of cerebellar tonsils. Motor disorders were represented by sluggish paraparesis in upper limbs (38.7%), often asymmetrical with triple superiority over paraparesis in lower limbs (12.9%). Spastic tetraparesis was determined in 7(22.6%) of Ⅳ patients, while spastic lower paraparesis was not detected at all. The presence of upper limb muscle hypotrophy in half of the patients in this group is noteworthy.

In contrast, muscle fibrillation in the shoulder belt was noted in 7(22.6%) of the examined subjects. Common fibrillation in the lower limbs and torso was observed in 2 cases in patients of Ⅳ group with a severe common form of syringomyelia against the background of CM1 type. A relatively equal distribution of objective vegetative symptoms was observed

in 25.8-38.7% of patients in this group. Pelvic dysfunction was manifested in 7 (22.6%) and 6 (19.4%) types of delay and incontinence, respectively.

Hence, with the coarse displacement of cerebellum tonsils more than 15mm below the Chamberlain line according to MRI data in objective neurological status dysfunction of cranial nerves at the level of the medulla oblongata, segmental motor disorders at the level of a cervical section with predominant involvement of anterior spinal cord structures as well as dissociated sensory disorders at the segmental type prevailed. The above disorders found in Ⅳ patients are, in our opinion, mostly due to the formation of extensive syringomyelic cavities, especially at the level of the upper cervical spinal cord.

Having analyzed the abovementioned complex of subjective and objective neurological symptoms in patients with Chiari abnormality of the 1st type, we singled out 4 types of clinical syndromes - cerebellar, bulbar-barrel, pyramidal, syringomyelic, which were most clearly formed in the groups of patients. The distribution of these syndromes in CM1 patients depending on the degree of displacement of cerebellar tonsils below the Chamberlain line is proposed in Table 3.

Table 3: Neurological syndromes in Chiari abnormality type 1 depending on the degree of ectopy of cerebellar tonsils.

	Ⅰ group. 0-mm (n=17)		**Ⅱ group. 6-10 mm (n=75)**		**Ⅲ group. 11-15 mm (n=84)**		**Ⅳ group. More than 15 mm (n=31)**	
syndromes	**number**	**%**	**number**	**%**	**number**	**%**	**number**	**%**
cerebellum	10	58.8	39	52.0	12	14.3	2	6.45
Bulbar	-	-	10	13.3	20	23.8	9	29.0
pyramid	-	-	5	6.67	21	25.0	1	3.23
Syringho-	-	-	18	24.0	31	36.9	19	61.3
myelitis	7	41.2	3	4.0	-	-	-	-

As can be seen from the presented data, cerebellar syndrome with clinical symptoms of dizziness, tinnitus, various types of nystagmus, static and dynamic ataxia was the most frequently encountered in patients of Ⅰ and Ⅱ group with a lowered almond to 6-10mm and accounted for more than half of the observations (58.8% and 52.0% respectively). The representation of cerebellar syndrome in patients of Ⅲ and Ⅳ groups was significantly less. Indeed, in the group with pronounced ectopy of 11-15 mm tonsils, the cerebellar syndrome was observed in 12 (14.3%) patients, while in the group with ectopy of more than 15 mm only in 2 subjects were observed. Such a low number of patients in Ⅲ and Ⅳ group with cerebellar syndrome manifestations, in our opinion, is connected with the cerebellar pathway intact with a pronounced displacement of cerebellar tonsils. Bulbar-barrow syndrome with clinical symptoms of dysphagia, dysarthria, soft palate paresis, tongue muscles were most frequently observed in the Ⅳ group in 29.0% of cases. In comparison, in the Ⅲ group, it was observed in 20(23.8%) patients. The number of patients with the bulbar syndrome in the Ⅱ group was much smaller in comparison with Ⅲ and Ⅳ groups and was only 13.3%, while in the Ⅰ group, no such patients were observed at all.

The predominance of the bulbar-bulb syndrome in groups of patients with the pronounced displacement of cerebellar tonsils was expected since it is in coarse cerebellar ectopy that the structures of the elongated brain are compromised. Clinical manifestations of pyramid syndrome in the form of spastic pares and increased tendon reflexes were typical for

Ⅲ group of patients with ectopy of 11-15 mm tonsils and made up 25%, whereas in Ⅱ and Ⅳ groups similar patients were noted only in 6.67% and 3.23% respectively; in Ⅰ group of patients with the pyramid, the syndrome was not observed. We attribute the dominance of pyramid syndrome in Ⅲ group to concomitant lychemical disorders in the brain, development of hypertension-hydrocephalic syndrome, and conduction disorder at the level of pyramid pathway intersection. Syringomyelic syndrome with different types of sensitive segmental dissociated disorders was most widely represented in Ⅳ group of patients with tonsil prolapse more than 15mm, made more than half of observations - 19(61.3%) in this group. In Ⅱ and Ⅲ the group of patients with syringomyelitis symptoms was twice less in 18(24.0%) and 31(36.9%) respectively, which was due to rarer formation of syringomyelitis cavities in the spinal cord in patients of these groups. In Ⅰ there was no syringomyelitis in the cervical or thoracic spinal cord.

It should be noted that we couldn't single out a certain clinical syndrome in 10 of our patients with CM1 type; 7 patients of Ⅰ group and 3 patients of II group, showed meager subjective complaints without objective violations, cerebellar tonsil prolapse in this category of patients was a "find" in MRI study.

4. CONCLUSIONS

1. Subjective and objective clinical symptoms in patients with CM 1 type had different degrees of severity and directly correlated with the degree of ectopy of cerebellar tonsils according to MRI study.
2. The structure of clinical syndromes in the case of CM 1 type includes cerebellar, bulbar-barrel, pyramid and syringomyelic syndromes, the formation of which is caused not only by the degree of prolapse of cerebellar tonsils but also by liquorodynamic disorders at the craniovertebral transition level.

3) The multimodal neurophysiological study using BAEP, SSEP and ENMG allows to objectify the functional state of stem structures in CM 1 type.

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