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CLINICAL CASE

DOI 10.25789/YMJ.2025.89.26

UDC 116.851

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CLINICAL CASE OF RHEUMATIC CHOREA WITH CARDIAC INVOLVEMENT IN A 13-YEAR-OLD CHILD IN THE REPUBLIC OF SAKHA (YAKUTIA)

During the last two decades, the prevalence of acute rheumatic fever has significantly decreased to isolated cases nationwide. In this article, a clinical case of rheumatic chorea, with choreic hyperkinesia syndrome, in a 13-year-old child with cardiac involvement is described. Modern concepts of therapy of rheumatic chorea are presented.

Keywords: chorea, streptococcus, fever, myocarditis, children, Sakha, Yakutia.

For citation: Argunova V.M., Burtseva T.E., Sleptsova P.A., Afonskaya M.V., Egorova V.B., Ammosova A.M., Kostik I.A., Kostik M.M., Evseeva S.A. Clinical case of rheumatic chorea with cardiac involvement in a 13-year-old child in the Republic of Sakha (Yakutia). *Yakut Medical Journal.* 2025; 89(1): 107-110. <https://doi.org/10.25789/YMJ.2025.89.26>

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Introduction. Rheumatic chorea (syn. Sydenham's chorea, minor chorea) is a post-streptococcal immune-mediated inflammatory neuropsychiatric movement disorder. The disease has been known since the Middle Ages and was called “St. Vitus' dance” at the beginning of observation. In 1686 Thomas Sydenham described “a kind of convulsion which affects boys and girls from the tenth year of life to puberty”. Sydenham's chorea, is a great diagnostic criterion for acute rheumatic fever (ARF). It debuts between 5 and 15 years of age, with a peak at 8-9 years of age [12], girls are more commonly affected [4,5,9].

Rheumatic chorea is an inflammatory postinfectious lesion of the CNS that occurs 4-8 weeks after a streptococcal infection, predominantly pharyngitis. Typically, bilateral involvement is noted, but approximately 20% of patients have hemichorea. The disease is a consequence of an autoimmune reaction following infection with “group A β -hemolytic streptococcus” (GABHS). The phenomenon of molecular mimicry is thought to underlie the pathogenesis of the disease. After frequent macroorganism contacts with GABHS, predisposed individuals develop

autoreactive lymphocytes and antibodies directed against epitopes of group A β -hemolytic streptococcus that cross-react with human cells [8]. This cross-reactive immune reaction explains the heterogeneity of the symptoms of ARF, which is usually manifested by skin lesions (subcutaneous nodules, erythema annulare), joint pain, fever, and cardiac involvement (myocarditis, transient atrio-ventricular block, heart valve endocarditis).

The clinical picture is composed of choreic hyperkinesia, muscle hypotonia (up to imitation of paralysis), disorders of statics and coordination, vascular dystonia, emotional lability, and psychiatric disorders. The duration of the attack is 3-6 months, residual phenomena can be observed for a year.

The Jones criteria [3] are used to diagnose acute rheumatic fever (Table 1).

Differential diagnosis. The most difficult cases to diagnose are cases of isolated minor chorea. The circle of differential diagnosis includes PANDAS, obsessive-compulsive syndrome, transient tics, autoimmune encephalitis, systemic autoimmune diseases (systemic lupus erythematosus, antiphospholipid syndrome), primary angiitis of the central

nervous system, Wilson-Konowalow disease [1].

The methods of treatment can be divided into three main groups: antibiotic therapy, symptomatic therapy and immunomodulatory therapy. Antibiotic therapy and antibiotic prophylaxis is currently the most studied method of treatment. Penicillin antibiotics are most commonly used; in case of penicillin intolerance, macrolides are used. Immunomodulatory therapy is used in severe and refractory cases, as well as in patients with severe side effects from symptomatic therapy [13,14] to shorten the duration of the disease and prevent complications [6,7]. There are different regimens for the use of glucocorticosteroids. For example, Fusco et al, suggested intravenous methylprednisolone for 5 days (25 mg/kg per day) followed by oral therapy with deflazacortin for 3 months (0.9 mg/kg per day) [2]. Paz et al. applied prednisolone (2 mg/kg/day, maximum dose 60 mg/day) [10]. Intravenous immunoglobulin and plasmapheresis are used as second-line therapy [11,12]. Anti-epileptic drugs, tranquilizers, and neuroleptics are used to control hyperkinesia [15] (Fig. 1)).

To provide a clinical case of rheumatic chorea with cardiac involvement in a 13 year old child is described below as a demonstration. The child, 13 years old, of Sakha nationality, was admitted on October 9, 2024 to the Reception and Diagnostic Department of the Pediatric Center of the Pediatric Center of the Republican Hospital No. 1-NCoM named after M.E. Nikolaev with complaints of: involuntary movement of the limbs, which the child cannot control, unable to hold objects (a spoon, a cup), slow response to speech, irritability, staring, speech disorders (speech is unintelligible), rapid fatigue, pain in the heart area, poor sleep, excessive tearfulness. Not only the writing abilities were disturbed in the girl, the child could not sit, stand. In the last 3 days bulbar disorders appeared - water was pouring out of the mouth, she could not swallow, did not eat, almost did not drink.

From the medical history: according to data reported by her mother, the girl has been suffering from frequent purulent sore throats since the age of 2 years, the last one was at the end of August 2024 with outpatient treatment. Since 10.09.2024 arthralgias appeared in the area of the right hand, legs, knee joints. After 4 days, swelling of the wrist joint appeared, in connection with which they applied to the district hospital. On 16.09.2024 she was examined by a neurologist, neurological disorders were excluded, reactive arthri-

tis was diagnosed, nimesulide, pentoxifylline, amoxiclav were prescribed. Despite the therapy, arthralgias persisted, cough was added. From 30.09.2024 to 07.10.2024 with the diagnosis of acute bronchitis the girl received inpatient treatment in the children's department of the district hospital, where periodic uncontrolled, chaotic movements of the upper and lower limbs more on the right side were observed. According to the girl's statements, the compulsive movements occurred by themselves. Antibacterial (ceftriaxone), antiviral therapy, expectorant medications were carried out. The joint syndrome was eliminated. Despite the persistence of severe neurological symptoms and psychoemotional instability, the girl was discharged home.

After discharge from the hospital, the mother noticed that the child's hyperkinesias became more frequent and intensified. Due to the worsening of the condition, the parents applied to the district hospital again. On 08.10.2024, the girl was examined by a district neurologist and re-hospitalized to the children's department of the district hospital with a preliminary diagnosis of minor chorea. The pediatrician consulted the rheumatologist of the cardio-rheumatology department of the PC, who recommended urgent hospitalization in the department. She was urgently delivered by air ambulance to the cardio-rheumatology department of the PC.

Past medical history: Child from the 1st pregnancy, without pathology. Delivery was on time, independent. Birth weight was 3270 g, body length was 52 cm. Past diseases were acute respiratory viral infections, acute respiratory infections, chickenpox. Since 2 years purulent sore throats were up to 5 times a year, in 2024 they were almost monthly. Hereditary factor: mother is 34 years old, healthy, father is 37 years old, healthy. On the father's line: rheumatism in a native aunt. Allergologic anamnesis: food allergy - plum, apple, nectarine - of Quincke's edema type.

On admission: Height of 151 cm, weight of 47 kg, HR of 18 per min, saturation of 98%, HR of 108 beats per min, BP of 99/52 mm Hg. The condition was severe. The well-being was disturbed due to chaotic movements of the limbs, which the child did not control. Consciousness was clear, position in bed: constantly on the move. On examination she responded adequately, calmly. Not speaking, because the language "did not obey". Appetite was reduced, she was not able to eat. Sleep was restless. In the neurologic status there were choreic storm, bulbar

disorders. The physique was normosthenic. The musculoskeletal system was visually unchanged. Skin covers were flesh-colored, moderately moist, clean. Turgor of tissues was preserved. Cyanosis was absent. No edema. The mucous membranes were pink, moist, clean. The conjunctiva of the eyes was pale pink. Tears were present. The pharynx was not hyperemic. Palatine tonsils without plaque, scarred. Subcutaneous fatty tissue was moderately expressed, evenly distributed. Peripheral lymph nodes were not enlarged. Nasal breathing was free, without discharge. Auscultatory respiration was even, vesicular, no wheezing in all fields. Percutaneously the borders of the heart were not changed. Auscultating heart tones were clear, rhythmic, there were no coarse noises. The tongue was clean, moist. The abdomen was not enlarged, soft on palpation, accessible to deep palpation, painless. The liver on the edge of the rib arch. Spleen was not enlarged. The stool was formed. The area of the kidney projection was not externally changed, the symptom of effluage was negative on both sides. Urination was free, painless. Urine was light-colored, transparent. There were no meningeal signs, focal neurological symptoms.

Neurological status: Consciousness was clear. During examination involuntary movements of eyeballs: rotation, involuntary sprawling movements of arms, legs, twitching of shoulders. Hyperkinesia increased with verticalization. Frequently changing postures. Pupils of rounded shape, equal. Photoreaction was lively. Eye slits were equal. The volume of eyeball movements was full, there was no nystagmus. The face was symmetrical. Tongue tip straight. (-) ROA. Muscle tone in the limbs was diffusely reduced. No paresis. Tendon reflexes from the limbs were alive, D=S. No pathologic or meningeal signs. Sensitivity was not disturbed. In Romberg's test she staggered. Ticotic hyperkinesia was widespread - chorea.

Paraclinically:

In the general blood test there was leukocytosis and acceleration of COE up to 29.00 mm/h. Biochemical blood test from 09.10.2024: hyperproteinemia, hypoalbuminemia, C-Reactive protein - 1.18 mg/l (0.00 - 10.00). Antistreptolysin-O level - 1522.20 IU/mL (0.00 - 200.00) on 30.09.2024 (Table 2).

ECG from 09.10.24: sinus rhythm.

Echocardiography from 10.10.24: AV insufficiency of 1 degree, MV 1-2 degrees, TV 1 degree. Ectopic attachment of MV chords. Heart cavities are not dilated. EF 69%. Splitting of pericardial sheets. Echocardiography dated 28.10.2024: AV

Table 1

The modified Jones criteria for the diagnosis of rheumatic fever

A. Evidence of prior GABHS infection of the pharynx for all patient groups	
Primary ARF	2 major criteria or 1 major plus 2 minor criteria
Recurrent ARF (with a history of verified ARF or existing chronic rheumatic heart disease)	2 major criteria or 1 major plus 2 minor criteria or 3 minor criteria
B. Major criteria	
<i>Low risk populations</i>	<i>Moderate or high risk populations</i>
Carditis clinical and/or subclinical	
Arthritis	
Polyarthritits	Monoarthritis or polyarthritits Polyarthralgia
Chorea	
Ring-shaped erythema	
Rheumatic nodules	
C. Minor criteria	
<i>Low-risk populations</i>	<i>Moderate or high risk populations</i>
Polyarthralgia	Monoarthralgia
Fever ($\geq 38,5^{\circ}\text{C}$)	Fever ($\geq 38,0^{\circ}\text{C}$)
Erythrocyte sedimentation rate (ESR) $\geq 60\text{mm/h}$ and/or C-reactive protein $\geq 30\text{ mg/l}$	ESR $\geq 30\text{ mm/h}$ and/or C-reactive protein $\geq 30\text{ mg/l}$
Age-adjusted PR interval prolongation on ECG (if carditis is a major criterion)	

insufficiency of 1-2 degree. Regurgitation on PA valve of 1 degree, on TV of 1 degree. Additional trabeculae in the LV cavity. Heart cavities are not dilated. Ejection fraction 71%.

MRI of the brain from 09.10.24: Topographic position of anatomical structures of cranio-vertebral transition is not disturbed. The middle structures of the brain are not displaced. In the frontal and parietal lobes on both sides subcortically, periventricularly there are foci of gliosis up to 3 mm in size. No changes in signal intensity from internal capsules, basal ganglia, thalamus were noted. The corpus callosum is of normal thickness and signal intensity. Brainstem and cerebellum have usual configuration and signal intensity. In diffusion mode at $b=1000$ no zones of acute ischemia or edema in brain structures were noted. The ventricular system is not dilated. Lateral ventricles are symmetrical. III and IV ventricles along the midline. There is no periventricular edema. Subarachnoidal convexital, cisternal spaces are not dilated. The pituitary gland is of normal size, its contours are smooth, clear, its structure is not changed. Pituitary pedicle along the midline. Chiasma and suprasellar cistern without features. Internal auditory canals are symmetrical, normal width on both

sides. No pathologic formations were found in the area of the pontine cerebellar cisterns. Thickening of the mucosa of the maxillary sinuses on both sides, cells of the lattice labyrinth. The structure of orbits is not changed.

The cervical spine and spinal cord were imaged on a series of tomograms. Physiologic lordosis was preserved. The shape and structure of the cervical vertebral bodies were unchanged, and the relationship between the C1 and C2 vertebrae was intact. No pathologic changes in the signal intensity from the bone marrow of the vertebral bodies and paravertebral soft tissues were detected. The

signal intensity from the intervertebral discs was not changed. The intervertebral discs did not protrude beyond the dorsal surface of the vertebral bodies. The nerve roots were intact. The spinal cord was located in the center of the spinal canal, had normal thickness and signal intensity. After contrasting the foci of pathologic accumulation of paramagnetic in the substance and in the brain membranes were not revealed. Conclusion: MR signs of residual encephalopathy. MR signs of bilateral maxillary sinusitis, ethmoiditis.

Ultrasound of abdominal cavity organs from 10.10.2024 without pathology.

Electroencephalography with activating tests from 10.10.24g: Bioelectrical activity of the brain is formed according to age. No pathologic, epileptiform activity was revealed.

Holter ECG monitoring from 15.10.24y: Conclusion: during the time of ECG monitoring the main rhythm was registered - sinus rhythm, episodes of supraventricular rhythm driver migration to unstable atrial rhythm mainly in the night hours. Episodes of marked and moderately - marked arrhythmias mainly in the night hours.

On the basis of complaints, anamnesis, clinical data, laboratory and instrumental investigations a clinical diagnosis was made: Main disease: Rheumatic chorea with heart involvement (I02.0): severe chorea. Carditis with mitral, aortic, tricuspidal valve lesions, mitral insufficiency of the 1-2 degree, aortic insufficiency of the 1st degree, tricuspidal insufficiency of the 1st degree. Arthritis. The III degree of activity. Complication: CHF of the 0-1 degree. FC 1. Associated diseases: chronic decompensated tonsillitis (J 35.0). Acute catarrhal sinusitis, acute catarrhal ethmoiditis (recovery) (J 01.8).

The child was examined by an otorhinolaryngologist on 09.10.2024: Acute catarrhal rhinosinusitis. Hypertrophy of the palatine tonsils of 1-2 degree.

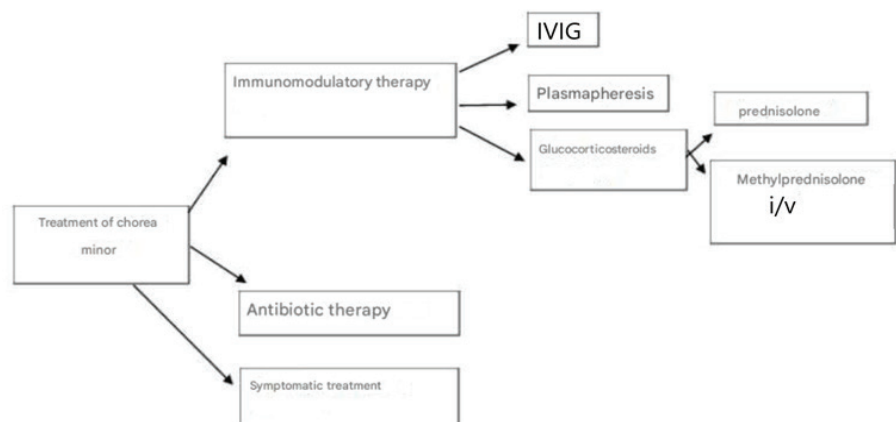


Table 2

Dynamics of laboratory indicators

Laboratory data	09.10.24	14.10.24	21.10.24	28.10.24	05.11.24
Erythrocytes x1012/l	4.92	5.23	5.28	5.09	4.97
Hemoglobin, g/l	120	126	130	127	126
Platelets x109/l	336	323	349	329	344
Leukocytes x109/l	5.85	9.38	12.32	8.38	9.21
COE, mm/hour	29.0	14.0	8.0	16.0	20.0
C-Reactive protein, mg/l	1.18				
Anti-streptolysin O	585.2				

In the dynamics the child was repeatedly consulted by a neurologist.

18.10.24. Consciousness is clear. During examination, involuntary movements of eyeballs: rotating, involuntary movements of hands in distal parts, twitching of shoulders. Pupils of rounded shape, equal. Photoreaction is alive. Eye slits are equal. The volume of eyeball movements is full, there is no nystagmus. The face is symmetrical. Tongue tip straight. (-) reflexes of oral automatism. Muscle tone in the limbs is diffusely reduced. No paresis. Tendon reflexes from the extremities are evoked, D=S. There are no pathologic and meningeal signs. Sensitivity is not disturbed. She is stable in the Romberg test. Finger-to-nose test is performed satisfactorily. Walking around the ward, gait is disturbed, with atactic component.

21.10.2024. In the dynamics of the neurological status during the examination involuntary movements in the hands, grimaces, gait with atactic component.

29.10.2024: Significant improvement in the neurological status, there are practically no involuntary movements. Hypotonia of muscles persists. There is staggering in the Romberg pose. 06.11.2024: according to neurological examination coordinator tests performs satisfactorily. Kernig's symptom is negative. She is stable in the Romberg pose.

The child received antibacterial (Ampicillin-sulbactam 1.5 g 3 times a day intravenously, followed by prophylaxis with bicillin-5, immunosuppressive therapy with glucocorticosteroids (methylprednisolone 10 mg/kg per injection 500

mg #3, then per oral - prednisolone 1 mg/kg/day), carbamazepine (Finlepsin) 100mg*2 (5 mg/kg) in hospital with positive dynamics.

Walking well on discharge. Consciousness is clear. During examination, exophthalmos is not noticeable. No hyperkinesia. Pupils are rounded, equal. Photoreaction is lively. Eye slits are equal. The volume of eyeball movements is full, nystagmus is preserved. The face is symmetrical. Tongue tip slightly to the right. Muscle tone in the limbs is reduced diffusely. No paresis. Tendon reflexes from the limbs are alive, D=S. No pathologic and meningeal signs. Sensitivity is not disturbed. Performance of coordinator tests, but with errors.

She was discharged with improvement under the supervision of a pediatrician, neurologist. Recommended: to continue therapy with carbamazepine, prednisolone, aspirin, bicillin. Re-hospitalization in 3 months.

Conclusion: rheumatic chorea is currently a very rare condition with which pediatric practitioners are not well acquainted. Rheumatic chorea is a neurologic disease that can cause involuntary movements in children and rarely in adults. The presence of cardiovascular involvement allows the diagnosis of rheumatic chorea and ARF, but cardiac involvement in the first few months of the disease may be nonmanifest and manifest only as instrumental findings. It is necessary to include cardiologic examination and search for streptococcal infection in all patients with hyperkinetic syndromes. Rational antibiotic therapy for infection caused by

GABHS may reduce the likelihood of developing ARF.

The authors declare no conflict of interest.

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