lenges of inherited mild bleeding disorders: a bait for poorly explored clinical and basic research. J. Thromb Yaemost. 2019; Feb.; 17(2): 257 - 270. Doi: 10.1111/jth. 14363.

10. Sabih A., Babiker H.M. Von Willebrand desease. Stat Pearls Publishing; 2021. Jan. PMID: 29083708

11. Katneni U.K. [et al.]. Von Willebrand factor

/ ADAMTS-13 interactions at birth: implications for thrombosis in the neonatal period. J Thromb Haemost. 2019; 17(3): 429 - 440. Doi: 10.1111/ ith. 14374.

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## INTRADURAL LIPOMA IN A NEWBORN

Intradural lipoma (spinal cord lipoma) is a rare benign tumor in the spinal cord consisting of white fatty tissue. The article presents a clinical case of intradural lipoma in a newborn.

Keywords: newborn, intradural lipoma, spinal cord, tethered spinal cord syndrome, skin appendage, surgical treatment.

Introduction. Intradural lipoma is a rare benign tumor of white fatty tissue inside the spinal cord. This formation of dysembryogenetic genesis is located in the lumbar-sacral region, originating from the conus of the spinal cord. The clinical picture of intradural lipoma includes: rudimentary appendages (tail), hypertrichosis, subcutaneous formation in the lum-

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bar region of a soft-elastic consistency. Intradural lipomas limit the mobility of the spinal cord, which is called tethered spinal cord syndrome [5;6].

Tethered spinal cord syndrome (TSCS) is a complex of pathological symptoms that is caused by tension of the spinal cord due to fixation of its caudal part [1]. Received widespread publicity in 1976, neurologist Hoffman published observations of typical symptoms in 31 patients. In children, the true incidence is unknown. According to some data, 0.8-1.4 cases per 1000 live births [9;11]. Risk factors include folic acid deficiency in the first trimester of pregnancy. TSCS is manifested by impaired sensitivity and motor function of the lower extremities, dysfunction of the pelvic organs, and skin symptoms in the lumbar region [8]. Surgical treatment consists of eliminating fixation factors: removal of pathological tissue (lipomas, dermal sinus), excision of the pathologically altered filament terminale. The earlier the surgery is performed, the lower the patient's risk of developing severe neurological deficits in the future.

Clinical case. Below is a clinical case of a newborn with a congenital malformation of the musculoskeletal system and tethered spinal cord syndrome at the State Institution of the Republic of Sakha (Yakutia) Republican Hospital No. 1 - National Center of Medicine named after M.E. Nikolaev, Perinatal Center.

The child was born to a 26-year-old woman from the first pregnancy, which occurred in the 1st trimester цшер grade 1 anemia (she took Maltofer), and in the 2nd trimester with shortness of breath. She was observed by a cardiologist with cardiac arrhythmias - frequent single extrasystole with episodes of bigeminy, trigeminy, quadrigeminy. Rare single supraventricular extrasystole, no treatment was carried out. Taking into account the Rh negative blood type - blood group AB(IV) Rh(-), Rh immunization was carried out at 30 weeks. Prenatal ultrasound diagnostics carried out within the screening period without any special features. The boy was born from the first spontaneous labor at 39.1 weeks in a cephalic presentation, after medical preparation of the birth canal with mifepristone. The condition of the child at birth is satisfactory, the Apgar score is 8/10 points. Physical parameters correspond to gestational age: birth weight 3300 g, body length 54 cm, head circumference 34 cm, chest circumference 33 cm. Among the features of the course of labor is a long anhydrous period of 12 hours, the waters are light. From the first hours of life, the child is breastfed. During the initial examination on the mother's abdomen in the delivery room, special attention was drawn to a soft tissue formation (dimensions 1.5x1.5 cm) in the sacral region, from the top of which a "tail" up to 2.0 cm long extends.

From the medical history of the child's mother it is known: she has been registered with the antenatal clinic since the early stages of pregnancy, and has regular visits. Denies bad habits, injuries, surgeries, blood transfusions. Gynecological diseases - ovarian cyst, cystectomy in 2022. Common diseases - chicken pox, rubella, acute respiratory infections, chronic bronchitis. The epidemiological history is calm. In a registered marriage, the husband is 25 years old, healthy, not burdened by heredity.

At the age of 3 days of life, a newborn boy was transferred from the neonatal department to the pathology department of newborns and premature infants for further examination and treatment with a diagnosis of: Q 79.9 Congenital malformation of the musculoskeletal system, unspecified.

At the time of examination at the Department of Pathology of Newborns and Premature Children, the boy's weight was 2980g (weight loss 9%). The condition is moderate according to the disease. On examination he is active, has an emotional cry, and opens his eyes. The physique is proportional. Breastfed. Muscle tone is physiological, reflexes are evoked, d = s. In particular, support reflexes, automatic walking, plantar reflex are symmetrical, physiological. The head is round in shape. Large fontanelle 1.0x1.0 cm. The seams are tight. Skin with icterus zone 3-4 according to Kramer, elements of toxic erythema throughout the body. According to bilitest 50/40 units. Continuous phototherapy was prescribed. The umbilical remnant is in a bracket, the periumbilical area is without signs of inflammation. Auscultatory breathing is puerile, there are no wheezes. BH 56 per minute. Heart sounds are clear and rhythmic. Heart rate 140 per minute. The abdomen is soft and accessible to palpation. The liver and spleen are not enlarged. Urinates on his own in his diaper. There was no stool during the examination. External genitalia according to the male type, testicles in the scrotum. No dysfunction of the pelvic organs was noted during the observation period.

Locally in the sacral region there is a soft tissue formation measuring 1.5x1.5 cm, from the top of which there is a "tail" up to 2.0 cm long (Fig. 1).

The newborn was consulted by specialists in the Perinatal Center of the "Republican Hospital No. 1-National Center of Medicine named after M.E. Nikolaev" of the Ministry of Health of the Republic of Sakha (Yakutia) in Yakutsk.

The child was examined by a surgeon (1st day of life): formation of the sacral region. An MRI examination was prescribed to exclude a connection with the spinal canal, with a subsequent decision on surgical treatment. Surgeon (10th day of life): stigmas of dysembryogenesis, skin growth in the coccygeal region. - Planned excision of the formation under local anesthesia is recommended.

Consulted by a neurosurgeon: intradural lipoma at level S 4.5. Fixed spinal cord. Skin formation in the gluteal region on the right. It is recommended that MRI of the lumbar spine and sacral spine be performed once a year.

Paraclinically: In the general blood test on the 4th day, moderate reticulocytosis is noted, other indicators for the entire observation period are within the age norm. In a biochemical blood test,



Fig. 1. Third day of life

hyperbilirubinemia is noted from the 4th day of life due to the indirect fraction of bilirubin. Indicators of the acid-base state of the blood are within the References values. The general urine analysis was unremarkable.

Ultrasound examination of the brain, abdominal organs, cervical spine - without pathologies. According to ultrasound, the thymus gland shows a moderate increase due to its width. Echocardiography is unremarkable. Electrocardiogramsinus rhythm with heart rate 155 per minute. Electrical axis of the heart sharply to the right. Block of the posterior branch of the bundle branch. Violation of intraventricular conduction. The potentials of the right ventricle are increased.

Conclusion of magnetic resonance imaging of the lumbosacral spine and spinal cord: at level S 4.5, an intradural lipoma measuring 5x6x11 mm is determined. The spinal cord is fixed to the lipoma. In the soft tissues of the gluteal region on the right, a soft tissue structure measuring 18x4 mm is detected on the skin.

Surgical intervention was performed on the 11th day of life (Fig. 2): in the department of pediatric surgery, under local anesthesia with novocaine 0.5% - 0.2 ml, the skin appendage of the coccygeal region was cut off with an electric knife, with the application of 1 suture. After 10 days, the suture is removed, the postoperative area is without any features.

Histology of the skin appendage: skin with underlying fibro-fatty tissue.

The child was discharged on the 14th day (Fig. 3). With a weight of 3608g. Condition is satisfactory, breastfeeding. The skin is subicteric, with regression. Postoperative area without inflammation.

Based on clinical and laboratory manifestations and magnetic resonance imaging data, the main clinical diagnosis was made: Q 06.8 Tethered spinal cord syndrome: intradural lipoma at the level of S4, S5. Fixed spinal cord. Congenital cutaneous appendage of the coccygeal region. Associated: P59.0 Neonatal jaundice of newborns.



Fig. 2. 3rd day after surgical removal of the skin appendage

Conclusion: Carrying out prenatal ultrasound diagnostics with an expert-class device would make it possible to suspect this pathology in utero, and refer the pregnant woman for additional examination - MRI of the fetus, as well as psychologically prepare the mother for childbirth with such a pathology in the child. This clinical case draws the attention of neonatal and pediatric doctors to the presence of a syndrome such as tethered spinal cord syndrome. Manifested by skin symptoms in the lumbar region; impaired pain and tactile sensitivity of the lower extremities; violation of pelvic functions, which can occur at any age [8]. In this patient, the skin appendage of the sacral region, which did not communicate with the spinal cord canal, was surgically

Spinal lipomas are often part of a complex of congenital anomalies; therefore, additional neuroimaging of the brain and spinal cord is required to exclude craniospinal dysraphism.

Outpatient follow-up for intradural lipomas after surgical treatment: observation by a neurologist, pediatrician, urologist, orthopedist, ophthalmologist. MRI control three months after surgery. If there are no signs of relapse of spinal cord tethering, MRI is indicated annually for up to three years.

## References

- 1. Bataeva E.P., Zeleneva A.YU., Kalinina L.R. Klinicheskij sluchaj nejrogennoj disfunkcii mochevogo puzyrya u rebenka na fone lipomy spinnogo mozga [A clinical case of neurogenic bladder dysfunction in a child with spinal cord lipoma]. Dal'nevostoch. medicin. zh-l [Far Eastern Medical Journal. 2016; 4: 101-103 (In Russ.).]
- 2. Zyabrov A.A. Sindrom fiksirovannogo spinnogo mozga (klinika, diagnostika, hirurgicheskaya korrekciya, blizhajshie i otdalennye rezul'taty) v detskom vozraste: avtoref. diss... kand.med. nauk [Fixed spinal cord syndrome (clinic, diagnosis, surgical correction, immediate and long-term results) in childhood: PhD abstract dissertation. St. Petersburg. 2012; 26 (In Russ.).]
- 3. Klinicheskie rekomendacii RF 2013-2017 (Rossiya) "Diagnostika i lechenie sindroma fiksirovannogo spinnogo mozga u detej" [Clinical recommendations of the Russian Federation

2013-2017 (Russia) 'Diagnosis and treatment of fixed spinal cord syndrome in children'. St. Petersburg, 2015:18 (In Russ.).]

4. Voronov V.G., Syrchin E.F., Zyabrov A.A. [et al.] Sindrom fiksirovannogo spinnogo mozga: sovremennye predstavleniya obetiologii i patogeneze, klinicheskoj kartine, diagnostike i lechenii (obzor nauchnyh publikacij) [Fixed spinal cord syndrome: modern ideas about the etiology and pathogenesis, clinical picture, diagnosis and treatment (review of scientific publications)]. Nejrohirurgiya i nevrologiya detskogo vozrasta [Neurosurgery and neurology of childhood. 2011; 2 (28): 53-65 (In Russ.).]

5. Khachatryan V.A., Sysoev K.V. Ob aktual'nyh problemah patogeneza, diagnostiki i lecheniya sindroma fiksirovannogo spinnogo mozga (analiticheskij obzor) [On the actual problems of pathogenesis, diagnosis and treatment of fixed spinal cord syndrome (analytical review)]. Nejrohirurgiya i nevrologiya detskogo vozrasta [Neurosurgery and neurology of childhood. 2014; 3:76-87 (In Russ.).]

6. Agrabawi HE. Incidence of neural tube defect among neonates at King Hussein Medical Center Jordan. Eastern Mediterranean health journal, 2005, Jul;11(4):819-23.

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## **POSITIVE CONTRIBUTION** OF TIMELY DIAGNOSIS AND CORRECTION OF ADRENAL DYSFUNCTION REQUIRING EXTRACORPOREAL MEMBRANE **OXYGENATION IN PATIENTS** WITH SEVERE PNEUMONIA IN THE EARLY **POSTPARTUM PERIOD**

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Introduction. The main diagnoses leading to the use of ECMO in pregnant women and the postpartum period are acute respiratory failure (62.7%). Adrenal dysfunction due to critical illness (CAD) often determines the severity of the patient's condition and the outcome of their illness. Aim. Timely detection and adequate correction of CAD in women in labor using ECMO.

Materials and methods. A patient after delivery with community-acquired severe bilateral polysegmental pneumonia who required the use of ECMO. Results. Against the background of combined treatment with norepinephrine and hydrocortisone, early stabilization of hemodynamics and septic complications was achieved.

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Conclusions. CAD is characterized by vascular insufficiency requiring the use of vasopressors. Timely and adequate correction of adrenal dysfunction during the development of a critical condition makes it possible to reduce the severity of the intensive care patient's condition and improve the outcome.

Keywords: critical illness, hydrocortisone, cortisol, extracorporeal membrane oxygenation, adrenal glands, adrenocorticotropic hormone, hypothyroidism, thyroxine.

Introduction. Respiratory distress syndrome (RDS), associated with lung damage from the H1N1 influenza virus-RDS, can develop rapid and almost total lung damage [2]. Due to the longterm restoration of gas exchange in the lungs with H1N1 - RDS, the risk of the need for veno-venous extracorporeal membrane oxygenation (VV ECMO) increases. The combination of primary viral and secondary inflammatory lesions of the lungs causes the development of a combination of viral-bacterial sepsis with multiple organ dysfunction (MOD), one of the components of which may be adrenal dysfunction caused by critical illness (CAD) [11]. CAD determines the severity of the patient's condition and the outcome of their disease [4, 6]. Timely and adequate correction of CAD often improves the outcome of the disease in intensive care patients [5, 6, 7]. However,

the problem of adrenal dysfunction (AD) remains outside the scope of intensive care directions implemented by the treating team of the intensive care unit. The presented clinical observation illustrates for the first time the importance of timely detection and adequate correction of CAD in a postpartum woman with severe community-acquired pneumonia that required VV ECMO.

Results and discussion. Patient L., 37 years old, 28 weeks of pregnancy, on the 15th day of illness with a diagnosis of "Acute purulent right-sided otitis media" was hospitalized in the ENT department of the regional hospital. On the third day of hospitalization, during treatment, the severity of the condition worsened: cough with light sputum, shortness of breath, weakness. On examination, breathing is spontaneous, respiratory rate (RR) is 37 times per minute, O2