Table 3 shows the results of the genealogical research method, which show the manifestations of dystonic syndrome in parents and siblings of patients with SCA type 1. Patients 6 and 7 are siblings. However, the father did not experience symptoms of cervical dystonia, who died at the age of 59 years. It is worth noting that in patient No. 7, cervical dystonia was diagnosed before the symptoms of ataxia. In the remaining patients, no hereditary form of SCA type 1 with dystonia was observed in relatives. According to genealogical inheritance, the mutation in the *ATXN1* gene was transmitted through the maternal line in 5 cases. On the paternal side in 4 cases. There were more natives from the Abyisky and Tattinsky districts, which are geographically located at a distance of 1800 km from each other. Abyisky ulus is located in the tundra arctic zone, and Tattinsky in the central zone of the Republic of Sakha (Yakutia). Botulinum therapy was performed in 3 patients, with a positive effect in 2 cases. The rest were not carried out due to the severity of the condition and the patients' disagreement with the procedure.

Conclusion. Thus, the study showed that in hereditary SCA type 1, manifestations of muscular dystonia may occur in the form of its focal form - cervical dystonia. Is this a manifestation of clinical polymorphism or is it a combination of two neurodegenerative diseases, when the olivopontocerebellar neurodegenerative process is a trigger for the development of further neurodegeneration in a certain area of the extrapyramidal system? The answer to this question can be provided by further molecular genetic studies, namely whole-genome sequencing. Considering the positive effect of botulinum therapy for cervical dystonia in practical medicine, this treatment method may become promising in the management of patients with SCA type 1 and improve their quality of life.

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A CLINICAL CASE OF THE EFFECTIVE USE OF VALVE BRONCHOBLOCATION IN THE COMPLEX TREATMENT OF A PATIENT WITH CASEOUS PNEUMONIA WITH MULTIDRUG RESISTANCE OF THE PATHOGEN TUBERCULOSIS

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Valvular bronchoblocation is a minimally invasive non–drug method of treating pulmonary tuberculosis and its complications. The method is based on the creation of therapeutic hypoventilation in the affected area of the lung while maintaining the drainage function of the bronchus by installing an endobronchial valve in its lumen. The article describes a case of effective application of the valvular bronchoblocation method in an acute progressive form of tuberculosis - caseous pneumonia, in a patient with multidrug-resistant tuberculosis pathogen (MDR MBT).

Keywords: tuberculosis, method of treatment, valvular bronchoblocation, effectiveness of treatment, multidrug resistance, causative agent of tuberculosis.

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Introduction: One of the main obstacles to achieving success in eliminating tuberculosis is multidrug-resistant tuberculosis (MDR-TB) [4]. Valvular bronchoblocation (CBB) is a minimally invasive non–drug method used in the complex treatment of pulmonary tuberculosis and its complications. The method is based on the creation of therapeutic hypoventilation in the affected area of the lung while maintaining the drainage function of the bronchus by installing an endobronchial valve (EC) into its lumen. The EC is designed in such a way that with

intense exhalation and coughing, air and bronchial contents exit through it from the blocked area of the lung, and when inhaled, atmospheric air does not enter there, this leads to local collapse of the lung, sometimes up to atelectasis. The valve is installed during fibrobronchoscopy under general or local anesthesia [5].

Also in recent years, the installation of endobronchial valves has been used to accelerate the repair processes in patients with destructive tuberculosis with multiple and broad drug resistance (MDR and XDR) [2]. The possibilities of conservative treatment in such patients are often exhausted, and surgical treatment is possible only in 15% of them due to concomitant diseases and/or the prevalence of the process [3]. Currently, there are already many reports on the effectiveness of using the valvular bronchoblocation method, among the first were publications on the treatment of decay cavities in the infiltrative form of tuberculosis, including drug–resistant mycobacterium tuberculosis (MBT) [1].

Here is an example of successful treatment of a patient with an acute progressive form of pulmonary tuberculosis - caseous pneumonia in the phase multidrug-resistant MBT inseminations with the use of CBB during the intensive phase of chemotherapy in inpatient settings.

A clinical example. Patient A. Age: 30 years old, mechanic-car mechanic.

Anamnesis of the disease: He became acutely ill, had an increase in body temperature to 38.5 ° C, was treated independently. Before the disease, FLH did not pass for 2 years. I went to the polyclinic and as a FLG (+) was sent for a computed tomography (CT) of the chest organs (OGK). Conclusion dated 02/21/2022: Infiltrative-destructive-focal changes in the upper lobe of the right lung, infiltrative-focal changes in S4,5,6,9,10 of the right lung and in S1-2, 4,5,6 of the left lung.

Anamnesis of life: Born in ulus. Secondary education, worked as a mechanic. He is married and has 2 children. Bad habits: smokes half a pack a day; drinks alcohol, has been registered with a narcologist since 2020. Drug use: denies. Previous diseases: acute respiratory viral infections. The presence of hepatitis, venereal, oncological diseases in oneself and relatives: denies. Chronic diseases: gastritis. Allergic history: not available for food, not available for medicines. Blood transfusion: denies. Frostbite at the age of 12 of the right hand.

Epidemiological history: he was not ill with a new coronavirus infection (NCVI). I have not received vaccinations against covid-19, influenza. Over the past 14 days, he has denied contact with infectious patients. I have not traveled outside the RS. According to him, he has not been in contact with tuberculosis patients.

He was admitted for inpatient treatment on 03/05/2022. with a diagnosis of Caseous pneumonia of the upper lobe of the right lung with insemination. MBT (+), MDR MBT was taken for the following drugs: isoniazid, rifampicin, streptomycin, ethambutol, pyrazinamide, levofloxacin, cyclocerin, ethionamide, ofloxacin (H,R, S,E,Z, Lfx,Cs,Eto,Ofx).

Concomitant diseases: Chronic bronchitis. Amputation stumps of the fingers of the right upper and lower extremities n/a frostbite n/a Peptic ulcer of the stomach. the ulcer of the prepiloric department is associated with HP+ in the scarring stage. Duodenogastric reflux. Duodenite.

Upon admission, he complained of coughing with foamy sputum, an increase in body temperature to 39.5 °, weakness, chest pain, weight loss of 10 kg per month, poor appetite.

According to the examination, anemia was noted in the general blood test (hemoglobin – 99 g/ l), lymphocytopenia – 9%, ESR – 36 mm per hour. In a biochemical blood test, an increase in CRP to 216 mg / l.

Microscopic examination of sputum on MBT is positive. MDR to H,R,S,E,Z,Lfx-,Cs,Eto,Ofx from 03/9/12 by seeding on dense nutrient media.

He was treated in the department of multidrug-resistant tuberculosis. In the hospital, he received chemotherapy for intensive phase IV of the standard regimen – 240 doses (5 drugs); bedaquiline (Bq) – 400 mg per day, linezolid (Lzd), - 600 mg 1 time per day intravenously, moxifloxacin (Mfx) - 400 mg, 1 time per day orally, amikacin (Am).– 930 mg 1 time per day, orally, delamanide (Dlm) – 100 mg 2 times a day.

Due to the prevalence of the process and resistance to 9 anti-tuberculosis drugs, the patient was selected for valvular bronchoblocation (CBB) at a medical commission with the participation of the attending physician, the head of the department and an endoscopist. CBB was

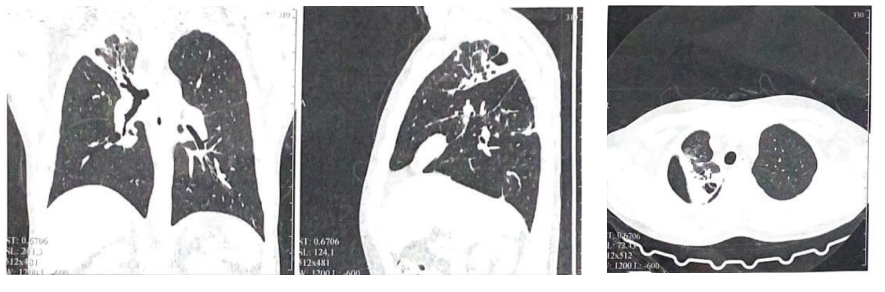

Fig. 1. Computed tomography of the chest organs from 11.09.22. to the installation of CBB

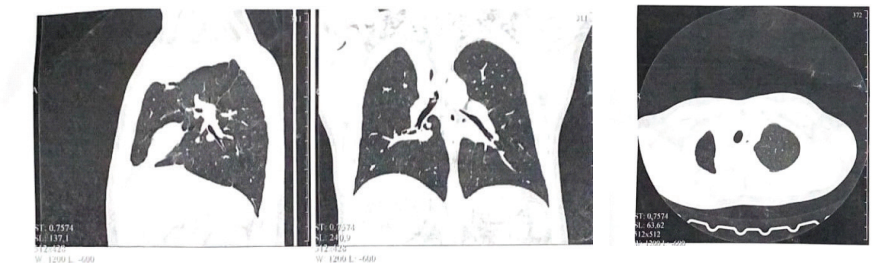

Fig. 2. Computed tomography of the chest organs from 10/31/12 after the installation of CBB

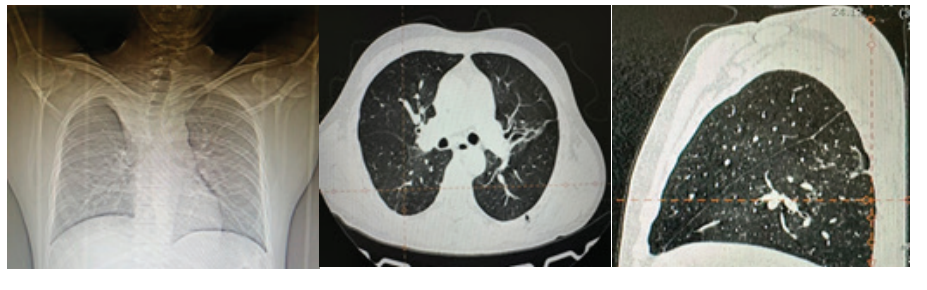

Fig. 3. Computed tomography of the chest organs from 04/15.24

performed on 11.10.2022 under combined endotracheal anesthesia, FB15 V(3) device; tracheoscope No.14.

From the protocol: intubation with tracheoscope No. 14. The mucous membrane of the trachea is hyperemic. Karina is sharp and mobile. The bronchial mucosa is hyperemic. There is a viscous mucosa of sputum in the lower parts. Sanitation, aspiration. Into the lumen of the right upper lobe bronchus (PVDB) KBB No. 11 is installed, the fixation is good, the KBB is functioning. The mouths of the remaining bronchi can be traced to 4-5 orders of magnitude in the lumen of mucous sputum, aspiration.

Conclusion: KBB PVDB. Recommended: Antitussive drugs, R-control.

Objective examination: Heart rate 70 in 1 min. BDD 16 in 1 min. Blood pressure 120/70 mmHg. t° 37.5°.Objectively: general condition: moderate severity, Consciousness: clear Position of the patient: active, Peripheral lymph nodes: enlarged, Skin: clean, moist, normal color. Auscultation breathing: carried out in all fields to the right of the wheezing from the KBB. Heart tones: muted, rhythmic. Tongue: moist, clean. Abdomen: not enlarged, soft, painless, Liver: not enlarged, elastic, consistency Spleen: not enlarged. Peripheral edema: no, Urination: free, painless, Stools: daily, every other day, decorated.

Prior to the esTableishment of the CBB, in the protocol of the OGK CT examination dated 11.09.22. (photo 1) it was revealed that the upper lobe of the right lung is reduced in volume. In S1, S2, and partially S3 of the right lung, a compaction of the lung tissue with cavities (up to 2.1x2.6 cm in size) and bronchial lumen in the structure is revealed. Foci of low and medium intensity can be traced in the adjacent lung tissue. In S3, S4, S5 of the right lung, there is a compaction of lung tissue with bronchial lumen in the structure. In S5, S6, S10 of the right lung, S6, S9 of the left lung, foci of medium intensity up to 0.5 cm in size are determined. Bronchi are visualized to a sub-segmental level, the lumen of the segmental bronchi is not changed.

In the upper mediastinum, the lymph node of the retrocaval space is up to 1.0 cm in size, the tracheobronchial space is up to 0.9 cm in size. There is no free fluid in the pleural cavity.

In the protocol of the OGK CT examination dated 31.10.22. (photo 2) the positive dynamics of the process has been revealed. In dynamics, there is a complete resorption of areas of inflammatory pulmonary tissue compaction by the type of frosted glass, without clear contours in S4,8,9 of the right lung. The upper lobe of the right lung is reduced in volume, atelectatized, and KBB is traced at the level of PVDB. In S3, S4, S5 of the left lung, there is a decrease in the volume and compaction of lung tissue with lumen of expanded bronchi in the structure, due to fibrous changes. In S5, S6, S10 of the right lung, S6 of the left lung, scattered foci of various sizes of medium intensity are detected against the background. In dynamics, in S6 of the right lung, foci decreased in number, in S10 of the right lung, a small thin-walled cavity decreased in size to 0.5x0.3 cm. Bronchi are visualized to a subsegmental level, the walls of segmental bronchi are compacted.

The roots are structural. The mediastinum is slightly shifted to the right, the trachea is also slightly curved to the right. In the mediastinum there is a lymph node of retrocaval space up to 0.6 cm in size, tracheobronchial space up to 0.8 cm in size. There is no free fluid in the pleural cavity.

Since June 2022, he has had 13 negative sputum culture results on MBT. Since 11/29/12, he has been abacillated, the patient has been removed from the bacillarity register. The patient was discharged from the hospital on 11/30/12, transferred to the continuation phase up to 310 doses on an outpatient basis. Terizidone (Trd) was prescribed – 500 mg per day, sparfloxacin (Sfx) – 200 mg per day, aminosalicylic acid (Pas) – 800 mg per day, up to 310 doses. Extract from the minutes of the meeting of the medical commission No. 234 MDR dated 05/16/2023 Diagnosis: A16.0 Cirrhotic tuberculosis of the right lung. KBB PVDB dated 10.11.22 A18.3 Intestinal tuberculosis in the subsiding stage I MBT(-) MDR HRSEZLfxCsEtoOfx from 03/9/12

In the protocol of the OGK CT examination dated 04/15.24. (photo 3) in dynamics, the upper lobe of the right lung is reduced in volume, atelectatized, and the valvular bronchoblocker remains in the PVDB. S3, S4, S5 of the left lung are reduced in volume due to fibrous changes with expanded bronchial lumen in the structure. In conclusion, atelectasis of the upper lobe of the right lung, the condition after CBB of the right upper lobe bronchus. Formation of fibroatelectasis in S3, S4,S5 of the left lung. Foci in S5, S6, S10 of the right lung. Positive dynamics.

At this time, the patient is being monitored in the III group of dispensary supervision, the patient has no adherence to treatment, did not visit a phthisiologist regularly, deblocking was scheduled for the end of April 2024.

Conclusion. It should be noted the high efficiency of this method in the complex treatment of the patient. Based on the presented clinical observation, it can be concluded that in cases where the patient has multidrug resistance of Mycobacterium tuberculosis and a widespread tuberculous process in the lungs, in our example caseous pneumonia, it is advisable to use valvular bronchoblocation. The method of temporary occlusion of the bronchi of CBB allows to achieve closure of the decay cavities, sanitation of the pleural cavity and in many cases to avoid surgical intervention.

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