Background. Myoclonus-dystonia is a movement disorder that typically affects the neck, torso, and arms. Individuals with this condition experience quick, involuntary muscle jerks or twitches (myoclonus). About half of individuals with myoclonus-dystonia develop dystonia, which is involuntary tensing of various muscles that causes unusual positioning. In myoclonus-dystonia, dystonia often affects one or both hands, causing writer's cramp, or the neck, causing the head to turn (torticollis).
Case report. A 26-year-old man consulted his doctor because of involuntary movement of the eyeball and involuntary muscular contractions on the left side of the body. It progressed to involuntary movement of the neck (torticollis) and left arm. Postural tremor is also present in the left arm. The laboratory results showed slighty diminished ceruloplasmin enzyme and blood copper, but the urine copper was three times higher than normal. The patient has been suspected of Wilson's disease but the genetic test came out negative and treatment with cuprinil has been ineffective. The patient also tried taking Isicom (levodopa), Romparkin (central anticholinergic), Haloperidol (neuroleptic) with no benefit. In 2014 he suffered a surgery for herniated disc C5-C6 with no symptom improvement. The cerebral MRI and electromyography has shown no significant changes. The Wilson disease diagnosis can be excluded, but to confirm Myoclonus dystonia, it's needed to make another genetic molecular testing for SGCE. This gene translates a transmembrane protein in the dystrophin associated glycoprotein complex found in skeletal muscle and neurons. If the test is positive, the most effective treatment would be a surgical intervention of Deep Brain Stimulation (DBS) in internal globus pallidus and the central intermediate nucleus of the thalamus, which can cure both the myoclonus and dystonia.
Conclusions. This case illustrates the difficulty of assigning a clear diagnosis regarding dystonia and the complexity of etiologies. Nonetheless, a correct diagnosis and treatment can improve the patient's life quality substantially.
Key words: myoclonic-dystonia, torticollis, Wilson's disease, DBS

35. SPONTANEOUS PNEUMOTHORAX: CONGENITAL CYSTIC ADENOMATOID MALFORMATION OF THE LUNG

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Background. Congenital cystic adenomatoid malformation (CCAM) is a rare developmental abnormality of the lung with an incidence of 1 in 25,000 to 1 in 35,000 pregnancies and represents 25% of congenital lung malformations and 95% of congenital lung lesions, males and females being equally affected. It is characterized by multicystic mass arising in the lung parenchyma from abnormal proliferation of the immature terminal respiratory bronchioles. The clinical spectrum is wide and ranges from silent incidental lesions to severe congenital respiratory distress or stillbirths. The diagnosis of CCAM can be made in utero by prenatal ultrasongraphy and postnatally by imaging radiography and CT. We report a case of a CCAM, which provoked spontaneous pneumothorax.
Case report. Eight month old patient was delivered to the Emergency Department at Institution of Mother and Child with pneumonia and left spontaneous pneumothorax. From the anamnesis, after birth was discovered CCAM after which the patient took a conservative treatment. On examination the patient was agitated, presented acrocyanosis, the general condition of the patient was severe. Chest X-ray showed a large left-sided pneumothorax with mediastinal shift to right and air-containing cystic area of left lower lobe. Pneumothorax was drained immediately, drain tube was seen through left 5th intercostal space. CT scan control of the chest showed expanded left lung with multiple cystic lesion in the lower lobe. After pre-operative preparation lower left

lobectomy was perfomed. According classification by Stocker (1977), our patient had CCAM, tip I.
Conclusions. Long term outcome is good in surgically managed asymptomatic patients with some studies showing only slight decrease in lung volume. Congenital cystic adenomatoid malformation of lung can cause severe respiratory distress but early diagnosis and surgical intervention can improve the condition and prevent death due to respiratory failure. The potential postnatal complications of CCAM include spontaneous pneumothorax, haemopneumothorax and pyopneumothorax, increased likelihood of malignancies such as bronchoalveolar carcinoma.
Key words: congenital cystic adenomatoid malformation, diagnosis, pneumothorax

DEPARTMENT OF ANESTHESIOLOGY AND INTENSIVE CARE no.2

36. ANESTHETIC AND INTENSIVE CARE MANAGEMENT IN A TRANSORAL SURGICAL APPROACH OF A PATIENT WITH ATLANTOAXIAL DISLOCATION

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Background. Transoral approach is an advanced neurosurgical technique in the treatment of atlantoaxial dislocation with spinal cord compression. Good cooperation between the surgical and the anesthetic teams during the treatment of this pathology plays a key role in achieving a positive result.
Case report. Child M, 7 years old, presented with dyspnea at rest, pain in the cervical region, lower limbs, headaches, upper limbs plegia, lower limbs paresis. Diagnosis: Osteoblastic/lithic process of C1 - C2 vertebrae with C2 fracture, with anterior and left paravertebral solid conglomerate, stenosis of the magnum foramen, compression of the medulla oblongata, signs of poststenotic myelopathy. The patient had a BMI of 16.5, severe spastic tetraplegia with an upper limb accent, cervical myelopathy and moderate hypercapnic respiratory failure, a positive bilateral Babinski sign and bilateral plantar clonus. The decision of performing a medullary decompression with spinal stabilization was taken. The monitored values were those of patient's: BP (invasive), CVP, HR, Ps, SpO2, diuresis, ABB, ionogram, coagulogram, and the results of neuromonitoring. Ist stage of the surgery was the occipito-cervical internal segmental fixation of C0-C1-C3-C4, with open C1-C2 distension, laminectomy and TIVA (14 hours). A volume of 2400 ml of blood was lost. The IInd stage of the surgery was performed after 12 days, and consisted of C1 and C2 corpectomy, with transoral cranio-cervical prosthesis and TIVA (12 hours). The patient was discharged after 21 days from the 2nd surgery.
Conclusions. An acute respiratory failure developed as a result of the postoperative polyneuropathy (the right hemithorax did not participate in the respiratory act, the left one showed only low amplitude trips). The ventilator weaning was difficult and was successful only after tracheostomy was installed and only when a combination of cholinolytic and anticholinesterasic therapies was used.
Key words: atlantoaxial dislocation, case report

INTERNAL MEDICINE I

ENDOCRINOLOGY