A ten-year-old boy was referred to the Clinic for further treatment after frequent bronchopneumonia in the last 4 years. Each time on the X-ray, basal right, the shadow on the lungs was verified. After antibiotic therapy, the control X-ray always showed incomplete regression of the shadow. The boy was born with esophageal atresia with tracheoesophageal fistula and was successfully operated on the eighth day of life.

Post-operative dilatation of esophagus has been successfully performed twice. The boy was in a good general health condition, except he was often tired after the activity. With the auscultation of the lungs, basal right, crepitation could be heard. CT of the chest, in the area of the posterobasal segment of the right lower lung lobe, showed a cluster of cystic air forms with condensed pulmonary parenchyma. The described change corresponded to developmental lung anomaly – cystic adenomatoid lung malformation. Right thoracotomy and lobectomy of the right lower lung lobe were performed. By surgery, the boy had no recurrent bronchopneumonia. Approximately one-third of CPAMs are diagnosed after the neonatal period. These lesions typically are CPAM types 1, 2 or 4, and tend to be smaller than CPAMs that present with respiratory symptoms at birth. A common presentation in older children is recurrent pneumonia. Other presenting complaints include cough, dyspnea and/or cyanosis. Findings on physical examination include decreased breath sounds over the lesion, hyperresonance and chest wall asymmetry with a bulge on the affected side.

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SUCCESSFUL TREATMENT OF PRIMARY SPONTANEOUS PNEUMOTHORAX WITH VIDEO-ASSISTED THORACOSCOPIC PLEURECTOMY

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Pneumothorax is defined as a collection of air that is located between the visceral and parietal pleura. Spontaneous pneumothorax occurs in the absence of any identified trauma. It is subdivided into primary and secondary types. The annual incidence of primary spontaneous pneumothorax (PSP) in the general population is estimated to be 5 to 10 per 100000. The peak incidence occurs between 16 and 24 years of age. We present the case of a sixteen-year-old boy who manifested on several occasions with right-sided spontaneous pneumothorax. The boy was actively involved in sports. Until spontaneous pneumothorax occurred, he had never been ill. He did not take medication. He had no allergies. A total of three times the right chest was drained. At each arrival, pneumothorax manifested with abrupt pain. The X-ray confirmed the diagnosis each time. After a third spontaneous pneumothorax, a CT of the chest was done. CT showed small apical blebs. The trachea was neatly positioned, neatly branching. The right main bronchus seemed wider than the left. The vascular structures were of a neat appearance. The heart was neatly positioned, with a morphologically neat appearance. Surgical treatment was initiated. After collapse of the right lung, an incision was made in the third intercostal space to the right in the anterior axillary line. An angle camera was installed. Upon placement of the camera, the trocar is placed in the seventh intercostal space in the median axillary line. After identifying the apical blebs changes with a linear endostapler,

a resection is performed. A partial parietal pleurectomy is then performed. Negative pressure drainage is set. In prophylaxis, the boy received cefazolin. A one-year follow-up did not result in relapse.

Surgery for pneumothorax consists of stapling ruptured blebs and resection of abnormal lung tissue. The approaches used include VATS (video-assisted thoracoscopic surgery), mini-thoracotomy and conventional thoracotomy. We usually use VATS, which provides adequate exposure for resection or stapling and an opportunity for pleurectomy, abrasion or chemical pleurodesis. The morbidity of VATS is less than with conventional or mini-thoracotomy, and recurrence rates are approximately 5%, although open thoracotomy and pleurectomy have the lowest recurrence rate. Given that in our previous cases, after repeated tube drainage, we did not have recurrences, we did not opt for VATS immediately after the first recurrence. Reading the literature, in the future we will be guided by the algorithm to initiate VATS after the first recurrence of pneumothorax.

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INTERSTITIAL LUNG DISEASE DUE TO SFTPC MUTATION – A RARE CASE REPORT

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Interstitial Lung Disease due to SFTPC mutation -a rare case report Mutations of the surfactant protein (SP)-C gene SFTPC have been associated with neonatal respiratory distress syndrome and interstitial lung disease.

If accurate diagnosis and proper management are delayed, irreversible respiratory failure may occur. A girl born following dizygotic twin pregnancy born at 37+3 weeks, born in good condition, no active resuscitation. However noted to have a fast-respiratory rate. Subsequently has had several admissions to hospital with faltering growth, persistent tachypnoea and oxygen requirement. High resolution CT angiogram showed atelectatic changes in the dependant portions, genetics work up identified a variation in a gene called SFTPC.

Introduction Pulmonary surfactant is a complex mixture of phospholipids and proteins that reduces surface tension of the alveolus by forming a surface-active film at the air-liquid interface. Of the four surfactant associated proteins the hydrophobic SP-B and ASP-C are especially essential in the dynamics of phospholipids by promoting the transition between the storage form and the functional surface film.

SP-C is synthesized from a 197 –amino acid precursor (proSP-C) which is encoded by SFTPC on human chromosome 8p21.3. In this report we present a case of a child who has childhood interstitial lung disease due to SFTPC mutation (c.289G>T).

Child Details A 5-week-old initially presented with possible aspiration; she was noted to be persistently tachypneic (up to 90 breaths/min) needing minimal oxygen to maintain saturations. She was reviewed weekly, where she remained tachypneic associated with feeding issues and faltering growth, was extensively investigated (GI/cardiac/metabolic). Had a prolonged stay at hospital and was subsequently discharged home on 0.5L O₂, high calorie NJ feeds and anti-reflux medications (post barium study).

Over the course she has had several admissions to the hospital, she has been treated with methylprednisolone 6 doses,

commenced on hydroxychloroquine and requires 3-4LO2 during day and Opti flow 6L/min at night. She gets blue with slightest agitation, precipitous desaturations and accompanying hypoxic attacks. She has marked finger clubbing and tachypnoea but no obvious increased work of breathing/chest deformity.

Of note her first cousin (maternal side) has suspected interstitial lung disease along with Bardet-Biedl, she has CT-changes but no surfactant gene abnormalities. Her maternal grandfather died at 45 years with pulmonary fibrosis, also her maternal great grandmother died young with pulmonary fibrosis. Her two half siblings are healthy.

Conclusion We report a child with SFTPC mutation with interstitial lung disease.

421 ASSOCIATION BETWEEN THROMBOCYTOSIS AND DISEASE SEVERITY IN CHILDREN WITH BRONCHIOLITIS

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The objective of this study was to investigate the frequency of secondary thrombocytosis in children during RSV (respiratory syncytial virus) bronchiolitis and bronchiolitis caused by other cause. Predictive value of secondary thrombocytosis in relation with severity of clinical features and the outcome of the treatment in bronchiolitis affected children has also been studied.

In this retrospective study, medical records of 136 infants treated at Pediatrics department of County General Hospital Požega were used.

Demographic and other patients' features with bronchiolitis were collected from their medical records. The data was statistically examined.

The mean age of participants was 117,5 day. There were 72 (52,9%) boys and 64 (47,1%) girls. 94 (64,1%) of children had positive rapid antigen testing (immunoassay) for RSV from nasopharyngeal aspirate. Statistically significant association between duration of hospitalization and oxygen application in therapy, with thrombocytosis, was determined with logistic regression. The risk for thrombocytosis was increased with every day of hospitalization by 18,5%, and with the oxygen application in therapy by 33,1%. Need for hospitalization and development of pneumonia statistically do not have an influence on the development of thrombocytosis. There wasn't statistically significant difference in thrombocytosis according to RSV infection.

Children with bronchiolitis and thrombocytosis had a more severe course of the disease, which was presented with longer duration of hospitalization and the need for oxygen application in therapy.

422 MODERN VIEW OF A PATIENT WITH BRONCHOPULMONARY DYSPLASIA

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To analyze the new model of a patient with bronchopulmonary dysplasia (BPD) based on the analysis of 70 case histories

of infants hospitalized at the National Medical Research Center for Children's Health Federal state autonomous institution of the Russian Federation Ministry of Health from 2020 to 2021.

Collection and analysis of anamnestic data, as well as clinical and laboratory examination of 70 infants with BPD. The analysis of the results was carried out using STATISTICA 10.0 program.

In total, all studied infants (n = 70) were diagnosed with a new form of bronchopulmonary dysplasia. Among them there were 35 boys (50%) and 35 girls (50%). The mean gestational age was 26,4 weeks [SD, 1.9 weeks]. All premature infants were born with extremely low and very low birth weight (85,7% and 14,3%, respectively). The median birth weight was 745 g (Interquartile range (IQR): 650 – 920). The median Apgar score at the 1st minute was 5 (IQR: 4 – 5), at the 5th minute – 6 (IQR: 5 – 6). 67 infants were intubated after birth, the median of duration of mechanical ventilation was 21 days (IQR: 6 – 36); CPAP treatment was initiated in 33 children, the median was 8 days (IQR: 5 – 13), BIPAP – 39 patients, nasal cannulas – 47 infants, nasal mask – 14, incubator – 19. Median duration of oxygen dependence was 54 days (IQR: 45 – 70). The period of oxygen dependence had an inverse relationship with anthropometric data and gestational age. All patients received surfactant therapy, postnatal corticosteroids – 32 infants (46%). In addition, pneumonia was observed in 35 children (50%), intraventricular hemorrhage in 60 patients (86%), necrotizing enterocolitis in 34 children (49%). Pulmonary hypertension, as one of the complications of BPD, occurred only in 7 patients (10%).

The current model of a patient with a new form of BPD is a premature infant with extremely low or very low body weight, born at a gestational age of no more than 28 weeks. The morphological and functional immaturity of these children, combined with the impact of new resuscitation technologies on their respiratory system, led to the pathomorphosis of BPD and the predominance of a new form in the population. Actually the study of the clinical features and long-term outcomes of BPD is an urgent problem of pediatric pulmonology and requires close attention in the future.

423 GENETIC PREDICTORS OF A NEW FORM OF BRONCHOPULMONARY DYSPLASIA

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Bronchopulmonary dysplasia (BPD) is a multifactorial disease with a significant genetic component. Novel genes and associated pathways may play an important role in susceptibility for the development of bronchopulmonary dysplasia in preterm infants. Our aim was to identify rare genetic variants contributing to the new form of BPD phenotype by full exome sequencing.

Full exome sequencing was performed on 39 DNA samples from patients with moderate and severe new BPD and 30 DNA samples from control group without clinical signs of BPD. After mapping and annotation, each sample showed an average of 40,000 genetic variants with a reading depth of at least 70x.