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 fist 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 45years 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 trombocytosis 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 GENETICPREDICTORS 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.