intensive care unit, drainage of the pleural effusion was performed with the application of alteplase for three days, continued treatment with antibiotics and symptomatics after confirmation of isolation of *S. Pneumoniae* from blood culture. On the tenth day of treatment, the boy became febrile with clinical deterioration. Radiologically, cavitation of the lung parenchyma was found, 4x5 cm with an aeroliquid level, which corresponded to a lung abscess. Treatment continued with modification of antibiotic therapy, vancomycin was introduced. Parenteral antimicrobial treatment lasted for about 5 weeks and then continued orally (amoxicillin + clavulanic acid) for a total of 8 weeks. During the treatment, regression of pulmonary changes was monitored using ultrasound, and a control lung scan was taken after two months confirming the abscess regression.

Pulmonary abscess as a complication of pneumonia is not common but should be considered in any severe clinical presentation of inflammation accompanied by the development of complications, especially in children who have not been vaccinated against pneumococcus as was the case with our patient.

414 SODIUM DEPLETION IN CYSTIC FIBROSIS PATIENTS OLDER THAN 24 MONTHS

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Sodium depletion is a well-recognized complication of cystic fibrosis in infants and toddlers, due to which they are routinely prescribed sodium supplementation. It exists in normonatremic subjects as well. Data on sodium homeostasis in older age groups is scarce. Spot urine sodium/creatinine ratio (UNa/Cr) strongly correlates with the fractional excretion of sodium

(FENa) and is considered to be a marker of normonatremic sodium depletion (NNaD).¹ This is a pilot observational study investigating the (UNa/Cr) in subjects older than 24 months, as a possible sign of sodium depletion.

Participants were children and adolescents seen between January 1st and October 31st 2019. at the UHC Zagreb – CF centre, Dept. of paediatrics.

Basic demographic, anthropometric clinical and laboratory data were collected. UNa/Cr ratio was considered pathological if <17mmol/mmol, corresponding to minimal target FENa of 0.5%. Descriptive statistics and Mann Whitney U test were used for data analysis; the significance set at $p < 0.05$.

30 patients (13 male) aged from 2.25 to 21.5 years entered the study. Only one patient had UNa <10 mmol/l. The calculated UNa/Cr ratio ranged from 1.85 - 52.8 mmol/mmol. UNa/Cr below the lower limit of 17 mmol/mmol was found in 20/30 patients (median 6.41 mmol/mmol; IQR 4.24 – 7.39). One third of subjects (10/30) had UNa/Cr above the cut-off value with the median value 26.04 mmol/mmol (IQR 23.55 – 50.54). We observed an almost significant difference in the age of patients with and without decreased UNa/Cr: median age in decreased UNa/Cr ratio group was 14.65 years (IQR 8.24 – 19.76) vs. 8.675 years (IQR 6.1 – 12.5) in the normal reference group ($p = 0,0556$).

Plasma sodium concentration was measured in 20/30 patients and all samples were within the normal reference range (135 mmol/l to 144mmol/l), regardless whether the

UNa/Cr was normal ($n=7$), or below 17 mmol/mmol ($n=13$).

Measurement of UNa alone is a rough parameter of sodium balance; in our set of patients, this criterion identified only 1/30 subjects as sodium depleted. However, we found a significant proportion of tested subjects (20/30) with an abnormally low UNa/Cr, suggesting an abnormally low fractional excretion of sodium – a parameter that reflects sodium status more appropriately. The plasma sodium concentrations remained normal. We suggest that this is due to an increase in sodium reabsorption to control hyponatremia. NNaD may be an under-recognized condition among CF patients beyond toddlers' age.

REFERENCE

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415 PRIMARY CILIARY DYSKINESIA – A THERAPEUTIC CHALLENGE

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Primary ciliary dyskinesia (PCD) is a hereditary heterogeneous pulmonary disease characterized by abnormalities of ciliary biogenesis, ultrastructure and function, leading to the development of bronchiectasis and chronic sinusitis, with situs inversus in about half of the patients. Diagnosis is based on a combination of phenotypic characteristics and diagnostic tests.

Treatment is directed to preserve pulmonary function and delay chronic changes leading eventually to lung transplantation. Therapeutic recommendations are based on the experience regarding treating diseases with similar mucociliary clearance disorders.

Here we describe the treatment course of a girl with PCD with situs viscerum inversus, whose diagnosis was confirmed at the age of 6 months by biopsy of the airway mucosa and electron microscopy showing lack of inner dynein arms.

The patient had respiratory difficulties since birth and chronic cough. The chest X-ray repeatedly showed pulmonary paracardial shading of the left lung. Ad-hoc physical therapy, secretolitics, inhaled steroids, salbutamol and, if necessary, antibiotics were prescribed. At 7 years of age, MSCT of the thorax revealed advanced left-sided pathology: parenchymal consolidation of the middle lobe with air bronchogram and ectatic bronchi and in the lower lobe ectatic bronchi and small-nodular interstitial infiltrates.

According to these findings, therapeutic measures were intensified with daily physiotherapy including high-frequency thoracic drainage (Vest machine), Flutter, manual drainage and encouraging physical activity. The use of secretolytics with antibiotics (orally, i.v., inhalations) according to sputum microbiology was regular and macrolide as immunomodulatory therapy was introduced. Checkups with spirometry and sputum analysis were performed every 3 months and ENT controls 1-2 times yearly. She was vaccinated against pneumococcus and yearly against influenza.

After 3.5 years of this regular treatment and monitoring, a new CT-scan showed significant resolution of the consolidation in middle pulmonary lobe with now visible bronchiectasis and wall thickening, as well as complete resolution of parenchymal lesions in the posterobasal segment of the left inferior lobe. In

the meantime, genetic testing proved homozygosity for the pathogenic variant of the SPAG1 gene.

The treatment of PCD is challenging because of low evidence-based recommendations. However, this case report shows that with regular monitoring and therapy adjustments depending on the circumstances, it is possible to maintain the airways state and pulmonary function at a satisfactory level even when the profound chronic lung changes exist.

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ABNORMAL LOCALISATION OF THE RADIAL SPOKE HEAD PROTEIN 4 AS A CAUSE OF PRIMARY CILIARY DYSKINESIA- CASE REPORT

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Primary ciliary dyskinesia (PCD) is a rare genetic heterogeneous disease with autosomal recessive inheritance. The estimated prevalence is 1:10 000 to 1:40 000 live-born. To date, more than 40 different genes were identified to be involved. The most common form of PCD is defects in the outer dynein arms and radial spokes. PCD is characterized by chronic upper and lower respiratory tract disease, infertility, neurological manifestations and abnormality in left-right asymmetry. The severity of disease varies between patients. When growing up, symptoms become more pronounced with the presence of destructive changes in the lungs, bronchiectasis, and the development of respiratory failure.

In this report we present a brother and sister, from young, healthy parents, who have recurrent respiratory symptoms and respiratory infections from infancy: chronic cough, recurrent bronchopneumonia and bronchitis. The first medical examinations, hospitalisations and laboratory testing for cause of recurrent infections were done in infancy. The laboratory findings were inconclusive. In May 2018., the boy, aged 11, is underfed (BMI 14.39kg/m², 5. ct) with clinical problems of chronic cough, and auscultator findings of crackles. We done the laboratory examination and tried to found out the underlining cause. The laboratory findings reveal no increase in inflammatory tests, negative microbiological samples, with the exclusion of humoral and cellular immunodeficiency's, sarcoidosis, aspergillosis, tuberculosis, heart disease and gastroesophageal reflux. We observed allergic sensitization, obstructive changes in pulmonary function test and bronchial hyperreactivity. High resolution computed tomography (HRCT) of the chest showed bronchiectasis in the lower lobe of the left and middle lobe of the right lung. Pulmonary function tests demonstrated obstructive ventilation functional impairment. Diagnostic bronchoscopy with mucosal sampling was performed. The immunofluorescence test showed RSPH9 abnormal localization in the ciliary axonemes. This finding confirms the diagnosis of PCD. At the same time, we follow up the boy's younger sister, at the age of 8, with the similar clinical symptoms. Pulmonary function tests showed obstructive-restriction functional impairment, while the HRCT scan of the chest revealed atelectasis and bronchiectasis in the segments of the lower pulmonary lobe to the right and V segments to the left. According to the PID diagnosis in older brother, we presumed that girl has the same disease, respectively.

Primary ciliary dyskinesia is a rare disorder with no etiological treatment.

Early diagnosis is important for introduction of early pulmonary rehabilitation and management of exacerbations in order to preserve the pulmonary parenchyma for as long as possible

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THE EFFECT OF NIV ON THE CLINICAL OUTCOME IN SEVERAL DIFFERENT PROFILES OF PEDIATRIC PATIENTS WITH CHRONIC RESPIRATORY FAILURE

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Noninvasive ventilation (NIV), refers to the administration of ventilatory support without the use of intubation and is a widely used and efficient method for treating patients with chronic respiratory insufficiency. Recent studies show that the spectrum of patients who could be candidates for NIV may require further evaluation, because, if started as early as possible, patients could benefit from less complications and a better quality of life.

This retrospective study included 7 patients with chronic respiratory insufficiency due to neurological and respiratory disorders as part of a complex genetic disorder which were followed and treated at Srebrnjak Children's Hospital and ultimately received NIV as the main therapy of choice for respiratory failure. Laboratory findings and clinical status, as well as incidence of respiratory symptoms and frequency of hospital admissions were analyzed.

All of our patients fulfilled diagnostic criteria for chronic respiratory failure (requiring daily oxygen support, having baseline SaO₂ < 88% on room air or pCO₂ > 50 with a normal pH value).

Of our 7 patients 2 (28,6%) were female and 5 (71,4%) were male. Only one patient had no confirmed genetic diagnosis while 6 (85,7%) had proven chromosomal aberrations or genetic mutations. The most frequently diagnosed respiratory difficulties were recurrent respiratory infections of the upper airways (N=7, 100%), lower airways with bronchial obstruction (N=5, 71,4%) and recurrent pneumonias (N=2, 73,4%).

Results show that all of the included patients had a better respiratory status (measured by pulse oxymetry and acid-base status) and a significant decrease in the frequency of respiratory infections was also observed.

We conclude that noninvasive ventilation is the therapy of choice for children with chronic respiratory insufficiency. We suggest that NIV should be started as early as possible after the initial diagnostic procedures were performed and the clinical and diagnostic criteria have been met. The indications for NIV should be further extended as to prevent severe complications in patients with chronic diseases that affect respiratory muscle strength and would also be beneficial for their quality of life.

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CYSTIC ADENOMATOID LUNG MALFORMATION – CAUSE OF RECURRENT PNEUMONIA

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