SODIUM DEPLETION IN CYSTIC FIBROSIS PATIENTS OLDER THAN 24 MONTHS

Ivan Bambir*, Bambir Ivan, Omerza Lana, Sapina Matej, Vukic Dugac Andrea, Tjesic – Drinkovic Duska. University Hospital Centre Zagreb

Abstracts

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). 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 <17 mmol/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 144 mmol/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


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 secretolitics 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