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Peculiarities of the course of respiratory infection among children with α -1-antitrypsin deficiency.

Background. α -1-antitrypsin deficiency (A1AD) is the reason of orphan disease among children which is elucidated only in single publications. In the childhood the pulmonary form of the diseases manifests in the form of reinfection of respiratory tract, also it can occur in the form of a recurring syndrome of wheezing which is followed by the transformation into bronchiectasis and emphysema in adults. At early stage of the disease the diagnostics is hard. Therefore, A1AD is a rare disease and it is important to create the international registries of such patients.

Case. A female 6-year-old child is under our supervision in the pulmonology center from the age of 3 years. The anamnesis shows that she was born from the 4th pregnancy, weighing 3,100 g. Until 3-years-old, she grew and developed according to her age, was not sick and she was vaccinated timely. Recurrent respiratory infections (bronchitis, pneumonia) and wheezing syndrome (3-4 times a year) have been observing since 3-years-old.

Positive dynamics was observed only after the appointment of antibacterial therapy.

Increasing of body temperature to 38, decreasing in body weight (BMI 10.4), severe respiratory failure, moderate manifestations of deformation of nails by type "watch glasses" and deformation of fingers in the form of "drum" wands" was noted during the last admission to the pulmonary center.

The clinical blood test showed accelerated ESR and leukocytosis. Leukocytes up to 50 were observed as a result of sputum test, and the bacterial diagnostic showed the presence of *St. Pneumonia* and *St. Haemolyticus*.

Chest CT scan showed cylindrical bronchiectasis, mainly in the reed segments on the left, in the middle lobe on the right and in the lower parts of the both lungs. The function of external respiration was the respiratory obstruction of the 1st degree. Oxygen saturation was 88%

AAT and ALAT were 2 times higher of the age norm. Fibroelastography of the liver did not reveal fibrosis.

Blood α -1-antitrypsin was sharply reduced to 0.38 g/l (with a norm of 0.78-2.0 g/l).

The diagnosis of cystic fibrosis was ruled out.

α -1-antitrypsin deficiency and cylindrical bronchiectasis were diagnosed.

Discussion. The pulmonary form of A1AD cannot be diagnosed for a long time and it can leak under the guise of respiratory tract infections, chronic obstructive pulmonary disease (COPD). Diagnostic search for α -1-antitrypsin deficiency should pass in children with repeated bronchitis, pneumonia, recurrent wheezing syndrome, respiratory failure and persistent microbial state of sputum.