The diagnostic significance of pulmonary scintigraphy in children with cystic fibrosis

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Abstract

Semnificația diagnostică a scintigrafiei pulmonare la copii cu fibroză chistică

Scopul acestui studiu constă în caracteristica perfuziei pulmonare prin metoda scintigrafică cu Tc-99 la 17 copii cu fibroză chistică. Examenul scintigrafic a arătat prezența tulburărilor de perfuzie la majoritatea copiilor cu diferite grade de severitate a bolii. Reducerea perfuziei (în unele cazuri, absența perfuziei) a fost observată în toate zonele pulmonare afectate, mai frecvent localizate în plămânul drept (lobii superior și mediu), o incidență mai rară fiind constatată la nivelul plămânului stâng (zonele superioare și inferioare). Tulburarea severă de perfuzie la copiii cu fibroză chistică este un indice indirect, care arată gradul de intensitate și extensie a proceselor fibroase.
Cuvinte cheie: fibroză chistică, scintigrafie pulmonară, copii

Abstract

The aim of this study consists in the characteristic of the pulmonary perfusion by scintigraphic method with Tc-99 in 17 children with cystic fibrosis. Scintigraphic exam showed the presence of perfusion disturbances in the most of the children with the different severity grade. The perfusion reducing (in some cases absence of the perfusion) was observed in the all pulmonary areas with more often location in the right lung (upper and middle lobes) and more rare incidence in the left lung (upper and lower zones). The severe perfusion disorder in children with cystic fibrosis is an indirect index, which shows intensity grade and extensity of fibrous processes.
Keywords: cystic fibrosis, pulmonary scintigraphy, children

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Introduction

Cystic fibrosis (CF) is an acquired abnormality with the autosom-recessive type of transmission, which is determined by the CFTR (conductor of the Cystic Fibrosis transmembrane regulator) gene change (mutation) that causes cystic fibrosis. This is a progressive, chronic disease, in which the mucus becomes dry, adherent and viscous. Mucus builds up and blocks the passages from various organs, especially in the lung and pancreas. At the lung, the mucus can cause severe respiratory problems and lung disease emergence. At the pancreas, mucus causing digestive impairment and malnutrition, which may result in impaired growth and normal development [3, 6, 7].

Congenital mutations of the CFTR gene cause changes of the protein, which is responsible for the transport of ions through the cell membrane. It causes the secretion of exocrine gland disturbances (these glands secret the level areas covered by epithelium, and not in the bloodstream) especially at the respiratory system and gastrointestinal tract [5, 7].

Only a small percentage of patients with cystic fibrosis live more than 40 years. The incidence in Moldova constitute 1:2000-1:3000 of newborns, and this percentage is growing. The severity determines the progress of the disease and requires confirmation of medical technique that will appreciate the structural changes of the lungs [1, 5, 6].

Pulmonary scintigraphy, imaging method, is being used for the evaluation of pulmonary perfusion [2, 4]. This diagnostic imaging technique is based on the principle of detecting gamma radiation emitted as a result of the injection of a radioactive isotope with a particular tropism for organ, or lesion and converting photons emitted into electrical signals, which are viewed at the oscilloscope, in the form of scintigraphic image. This method has an important role in the early identification of pulmonary vascular defects, especially in cystic fibrosis and other fibrosing chronic lung disease [3, 4]. Repeat scintigraphy can provide data about pulmonary revascularization in affected areas.

Aim. Assessment of pulmonary perfusion in children with cystic fibrosis to evaluate the intensity of the fibrosis process and spread of bronchiectasis.

Materials and methods

The team of researchers from the Pneumology clinic, Mother and Child Institute, were investigated 18 children with cystic fibrosis, in ages between 2 and 18 years old, among whom 4 girls and 14 boys. Most of them has a mixed form of the disease (pulmonary and intestinal). The children were examined in detail to confirm the diagnosis of cystic fibrosis. All children had the increased values of sweat test (>60 mEq/L, Macroduct, USA). ΔF508 mutation has been confirmed in 9 children, 1 child with mutation R334W and 1 child – L551D.

Lung perfusion in children with cystic fibrosis has been studied through pulmonary scintigraphy using Gamma-camera (Siemens, Germany) with the Tc-99 MAA. The imaging examination included ultrasound results, pulmonary radiography, thoracic HRCT.

Results. Most children with cystic fibrosis have had severe disease evolution (12 children), 5 children had moderate degree. The colonization of a chronic bronchial tree with *Staphylococcus aureus* was present at 10 children with cystic fibrosis, *Haemophilus influenzae* – 9 children, *Peudomonas aeruginosa* – in 5 cases. In some cases it is found microbial agents association. Clinical manifestations were dominated by severe pulmonary disease with pulmonary fibrosis to 8 children, bronchiectasis (5 cases), fibrous atelectasis (2 cases), chronic bronchitis (11 children). The severity of respiratory manifestations in children with cystic fibrosis was confirmed by respiratory failure with restrictive and obstructive pulmonary disorder II-III degree (10 children). In 2/3 cases were present growth disorders.

Scintigraphic examination identified pulmonary perfusion disturbances in 17 children with different characteristics and the degrees of severity. Moderate reduction in pulmonary perfusion of the right lung in the upper sectors (fig. 1) was detected in 76.5% of cases, the medium sectors have pulmonary perfusion disorders at 47.1% of children, involving lower lung zones with the focal character were revealed in 35.3%. In accordance with data from the literature on the right upper lobe impairment is an important criterion for imaging diagnosis of pulmonary manifestations in cystic fibrosis. In the left lung the severity of perfusion disorders were less expressed, but in 35.3% of the cases have been observed serious changes in the upper segments, in 41.2% - lower lung area, less pronounced in the medium sectors – 29.4%.

The perfusion was absent in pulmonary sectors of the lung (fig. 2) in 3 children. Severe problems with the total absence of perfusion was reflected in the right lung: upper lobe – 4 children, medium lobe – 3 children, lower lobe – 1 case. The left lung was affected in unique cases with upper lobe involvement (1 child) and medium sectors (1 child). Only in a child with cystic fibrosis lung perfusion has not suffered pathological changes. Contrasting the results of pulmonary radiography obtained by radiographic methods (chest radiography, pulmonary computed tomography) with scintigraphic exploratory data revealed a correlation of these diagnostic procedures for the identification of the pulmonary fibrosis, bronchiectasis.

Infusion disorders caused in children with cystic fibrosis is an indirect index that can show the intensity and spread of pulmonary fibrosis.

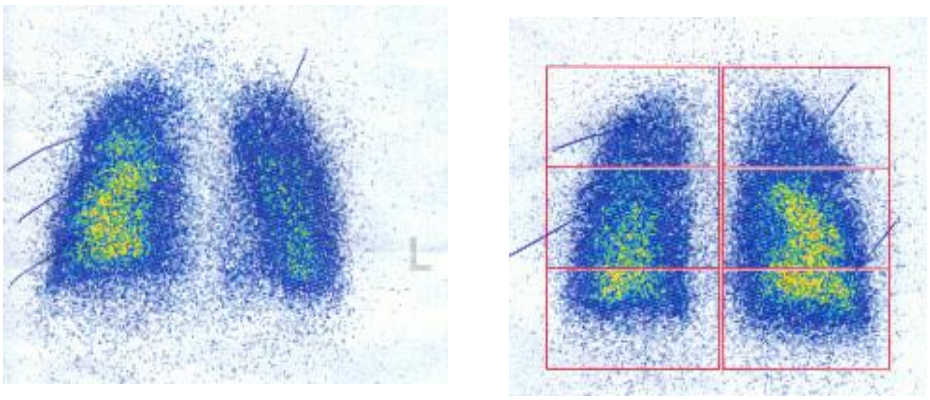

Fig. 1. Pulmonary scintigraphy in children with cystic fibrosis. Moderate reduction in pulmonary perfusion of the right lung

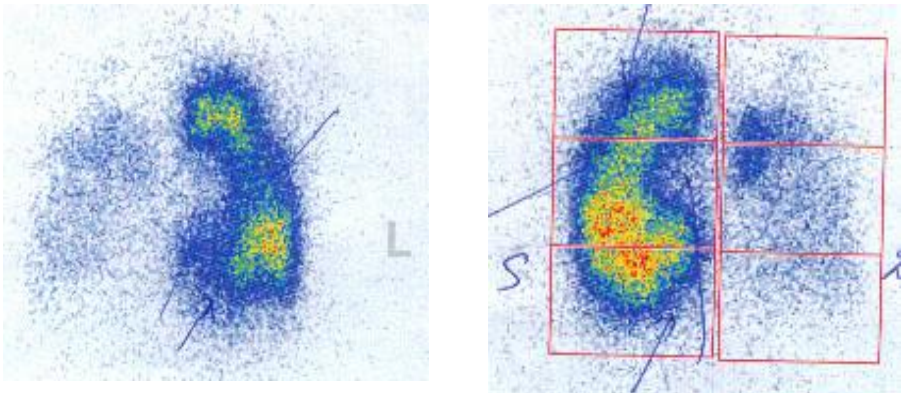

Fig. 2. Pulmonary scintigraphy in children with cystic fibrosis. Absent perfusion in pulmonary sectors of the lung

Conclusion. Pulmonary perfusion in children with cystic fibrosis is reduced in all areas with diffuse lung disorders, the most severe in the right upper lobe. Pulmonary scintigraphy can be recommended as a screening test for the evaluation of intensity of lung fibrosis in children with cystic.

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