

INTERSTITIAL LUNG DISEASES IN CHILDREN

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Article history:	Abstract:
Received:February 10th 2022Accepted:March 11th 2022Published:April 30th 2022	There is an undoubted link between chronic lung diseases in children and adults. This is a single clinical problem for pediatricians and therapists, it has not only medical, but also social significance. Chronic respiratory diseases that began in childhood often lead to disability of patients in adulthood, and sometimes to dramatic outcomes.
Keywords: Lung Angiomatosis, Fibrosis, Bronchiectasis, Pulmonary	

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Hereditary diseases among children with chronic inflammatory lung diseases are diagnosed in 5-6% of patients. The IBD group includes various etiological forms of the disease: alveolitis (exogenous allergic, granulomatosis toxic, fibrosing); (sarcoidosis, disseminated tuberculosis, etc.), dissemination of malignant nature (lung carcinomatosis, etc.); rare forms of dissemination in the lungs (pulmonary hemosiderosis, Goodpasture syndrome, alveolar proteinosis, lung angiomatosis, etc.) interstitial fibrosis of the lungs in systemic diseases (collagenosis, cardiogenic pneumosclerosis in chronic hepatitis, etc.).

The initial pathogenic impulse triaaers pathological immune reactions involving various types of cells that produce proteases and oxidants that damage interstitial and parenchymal structures of lung tissue. Stereotypical changes in pulmonary infection develop in the form of inflammatory infiltration of varying degrees of severity, productive alveolitis, and fibrosis is subsequently formed (the picture of a "cellular lung").

As a result of a decrease in the diffusion capacity of the lungs, the development of ventilation-perfusion imbalance in patients, arterial hypoxemia is determined in the early stages of the disease only with physical exertion. As the process progresses, hypoxemia is registered at rest, accompanied by hypocapnia. Hypercapnia appears in far-reaching cases of the disease. When studying the function of external respiration, a predominantly restrictive type of ventilation disorder, a decrease in the main pulmonary volumes, is detected.

The most important diagnostic methods for IBD are chest X-ray examination methods. High-resolution computed tomography is becoming increasingly important. In the early stages of the disease, mainly the strengthening and deformation of the pulmonary pattern, a decrease in the transparency of the pulmonary fields by the type of "frosted glass", finefocal shadows are determined. As the process progresses, the deformation of the pulmonary pattern becomes more pronounced, signs of interstitial

fibrosis, cavities are revealed, and a picture of a "cellular lung" is formed.

Upon termination of contact with the antigen, complete recovery is possible in a few days or weeks. With repeated contacts, relapses of the disease develop, which may be subacute in nature, remain unrecognized, which leads unexpectedly for the patient and the doctor to the transition of the disease into a chronic form. The main sign of the subacute form is shortness of breath, which persists for several weeks or months.

In the chronic form, constant shortness of breath, cough with the separation of mucosal sputum are typical in the clinic. With physical exertion, shortness of breath increases, cyanosis develops. During auscultation, constant crepitating wheezes are heard. Gradually the state of health worsens, weakness, fatigue, decreased appetite, weight loss, decreased motor activity appear. During the examination, the deformation of the chest is determined in the form of its flattening, changes in the type of "drumsticks" and "watch glasses" develop.

High-resolution computed tomography reveals early changes in the pulmonary parenchyma with great accuracy. The chest X—ray shows changes in the form of small (miliary) focal shadows located mainly in the area of the middle parts of the lungs, sometimes a decrease in the transparency of the lung tissue is a symptom of "frosted glass". There may be multiple infiltrative oblacoid or denser shadows with reverse development over weeks or months.

Chest radiography. 5-10% of patients at the first visit to the doctor have no changes on the X-ray (stage 0), 35-45% have bilateral enlargement of the basal lymph nodes (stage I), 25% have retinal nodular lesion, linear shadows, bilateral enlargement of the basal lymph nodes (stage II), 25% have limited darkening in the lungs (stage III). The final stage of the disease (stage IV) is manifested by irreversible changes in the lungs - pneumosclerosis, displacement of the roots of the lungs, bronchiectasis, emphysema. Occasionally, single or multiple cavities, focal shadows,



pneumothorax, unilateral pleural effusion, calcification of lymph nodes may be observed.

Crystals of orthophosphate and calcium carbonate with an admixture of iron salts with a size of 50-200 microns, having a concentric structure, are formed in the alveoli. About 400 cases have been described, more often in people of Turkish origin. The progression of the process leads to fibrous changes in the interalveolar septa and respiratory failure in middle age. In most cases, the disease is asymptomatic, in children it is more often diagnosed by accident: symmetrical small calcifications on the background of a diffusely enhanced pattern. A chest X-ray reveals a pathognomonic symptom of a "sandstorm". The course of the disease is long.

The auscultative picture in the lungs in interstitial diseases is quite specific. In patients, gentle crepitating wheezes are heard on inspiration. With obliterating bronchiolitis, a weakening of breathing, moist shallow and medium-bubbly wheezing is often detected, crepitation is possible. Characteristic, according to the observations of leading pediatric discrepancy pulmonologists, is the between pronounced shortness of breath and relatively small physical changes in the lungs. In addition, in most cases, shallow breathing can be detected. Percussion usually reveals a dulling of the percussion sound over the lower parts of the lungs.

Usually, arterial hypoxemia is detected in patients, which is detected in the early stages in patients only with physical exertion. As the process progresses, hypoxemia is registered at rest.

An extremely promising area of diagnostics is the study of the gas composition of exhaled air and the biochemical spectrum of its condensate, which is especially important in children and patients who cannot be used invasive technologies for vital indications.

Traditional radiography remains the main method that gives a visual representation of the pathological process in the lungs in IBD. In the early stages of the disease, the radiological signs of IBD are mainly the strengthening and deformation of the pulmonary pattern, a decrease in the transparency of the pulmonary fields by the type of "frosted glass", the detection of fine-focal shadows. As the processes, the deformation of the pulmonary pattern become more pronounced - signs of interstitial fibrosis appear, different-sized cavities, a picture of a "cellular lung" is formed.

There is no unified classification of interstitial lung diseases. They are conditionally divided into diseases

with a known etiology (these include exogenous allergic alveolitis, toxic alveolitis) and diseases with an unidentified etiological factor (fibrotic alveoli, sarcoidosis, various vasculitis). In addition, there are so-called secondary interstitial lung diseases that develop with systemic lesions of connective tissue.

Despite the polymorphism of clinical and morphological variants and manifestations of interstitial lung diseases, most of them begin with fairly stereotypical changes in the pulmonary interstitium in the form of inflammatory infiltration of varying degrees of severity, productive alveolitis, subsequently fibrosis is formed, the rate of progression of which may be different, and the picture of a "cellular lung" develops.

Among the clinical manifestations of the disease, respiratory failure plays a decisive role. Shortness of breath is the main symptom of almost all IBD, occurs in most patients, especially in young children, and is the earliest sign of the disease. Respiratory failure initially occurs or increases with physical exertion, has a steadily progressive character. In some patients, shortness of breath is accompanied by wheezing. These manifestations of the disease can be mistaken for bronchial asthma.

Arterial hypoxemia is determined with great constancy in patients, detected in the early stages of the disease only with physical exertion, and as the process progresses, hypoxemia is registered at rest, accompanied by hypocapnia, reflecting the features of the respiratory pattern of patients - frequent shallow breathing (rapid shallow breathing), hypercapnia appears in the advanced stages of the disease. The main mechanism of hypoxemia is a ventilationperfusion imbalance, a decrease in the diffusion capacity of the lungs.

It should be emphasized that diseases belonging to the group of idiopathic interstitial pneumonia are extremely rare in children. This also applies to our observations. The symptoms of severe respiratory failure are dominant in the clinical picture. The disease has a steadily progressive course, while corticosteroid therapy is ineffective, which can serve as an additional criterion for a differential diagnosis.

In conclusion, it should be said that interstitial lung diseases are characterized by a variety of nosological forms in children and are currently the most important problem, especially urgent is the diagnosis of various interstitial lung diseases in the early, still reversible stages of the disease, the search for new modern approaches to the treatment of these life-threatening diseases of the child.



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