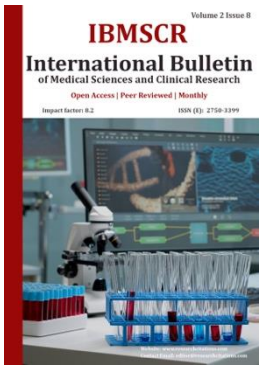

BRONCHIECTASIS IN CHILDREN: REVIEW OF CURRENT CLINICAL GUIDELINES

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Annotation: the article is devoted to the problem of bronchiectasis in pediatric patients and reflects the most current approaches to the diagnosis and management of patients with this pathology. Information about the true prevalence of bronchiectasis both abroad and in Uzbekistan varies. At the same time, bronchiectasis can be either an independent disease or a manifestation of another pathology, which necessitates the need for an integrated multidisciplinary approach not only for diagnosis, but also in the management of patients with this pathology.

Key words: bronchiectasis, children, congenital lung malformations, allergic bronchopulmonary aspergillosis, immunodeficiency, computed tomography, cystic fibrosis, chronic bronchitis.

BRIEF INFORMATION

Definition Bronchiectasis is a localized irreversible dilatation of the bronchi, accompanied by inflammatory changes in the bronchial wall and surrounding parenchyma with the development of fibrosis.

Etiology and pathogenesis

The development of bronchiectasis can be facilitated by numerous pathological factors - congenital structural defects of the walls of the bronchial tree, compression of the bronchus due to various reasons (for example, enlarged lymph nodes or a foreign body), as well as inflammation, which damages the elastic tissues and cartilage of the bronchus.

Inflammation of the bronchial wall may be a consequence of a respiratory tract infection, exposure to toxic damaging substances, or one of the manifestations of autoimmune diseases.

The lungs normally have a system of primary and secondary defenses, which allows them to maintain sterility, therefore bronchiectasis is usually caused by various congenital and acquired conditions, although the term “idiopathic bronchiectasis” is still used.

Bronchiectasis, as a result of destruction of the bronchial wall due to inflammation, is caused by damage to the bronchial epithelium by bacterial toxins, and then by inflammatory mediators that are released from neutrophils, which leads to disruption of physiological protective mechanisms, mainly the ascending flow of mucus.

As a result, favorable conditions are created in the bronchi for the growth of bacteria, and a vicious circle arises: inflammation–damage to the epithelium–disturbance in the ascending flow of mucus–infection–inflammation [1–5].

Bronchiectasis can be observed in patients with the following pathologies [1–7]:

1) with congenital structural anomalies of the bronchopulmonary system, such as:

• Williams–Campbell syndrome (ballooning bronchiectasis);

• Mounier–Kuhn syndrome (tracheobronchomegaly);

• tracheomalacia;
• bronchomalacia;
• stenosis of the trachea and/or bronchi;
• bronchogenic cysts;
• tracheal bronchus;
• pulmonary sequestration;
• cystic adenomatous malformation;
2) toxic damage to the respiratory tract:
• when inhaling toxic substances;
• aspiration syndrome due to gastroesophageal reflux;
• aspiration due to muscular dystrophy;
• aspiration due to the presence of a tracheoesophageal fistula;
3) bronchial obstruction:
• caused by external causes (lymphadenopathy, abnormal vessel, tumor);
• intrabronchial obstruction by a foreign body due to aspiration;
• intrabronchial obstruction by a space-occupying formation (tumor, granuloma, etc.);
4) obstructive pulmonary diseases:
• with 1-antitrypsin deficiency;
5) disorders of mucociliary clearance:
• with primary ciliary dyskinesia;
• cystic fibrosis (including atypical forms);
6) infection, such as:
• whooping cough;
• measles;
• adenoviral infection;
• pneumonia;
• tuberculosis;
• non-tuberculous mycobacteriosis, including in HIV infection;
7) primary immunodeficiency conditions, such as:
• agammaglobulinemia;
• common variable immunodeficiency;
• selective deficiency of immunoglobulin (Ig) A;
• selective deficiency of immunoglobulin G subclasses;
• severe combined immunodeficiency, ataxiatelangiectasia (Louis-Bar syndrome);
• Jobe's syndrome (hyper-IgE syndrome);
• chronic granulomatous disease;
• deficiency of transporters associated with the presentation of antigens;
• complement defects;
8) secondary immunosuppression caused by:
• oncohematological diseases;
• allogeneic transplantation, including bone marrow;
• use of immunosuppressive drugs;
9) allergic bronchopulmonary aspergillosis;
10) systemic diseases, such as:
• rheumatoid arthritis;

• systemic scleroderma;
• relapsing polychondritis (Meyenburg-Altherr-Ulinger syndrome);
• ankylosing spondylitis;
• sarcoidosis;
• Ehlers–Danlos, Marfan syndromes;
• Young's syndrome;
• “yellow nails” syndrome;
• metaphyseal chondrodysplasia, McKusick type;
11) inflammatory bowel diseases, such as:
• ulcerative colitis;
• Crohn's disease;
12) idiopathic bronchiectasis.

Sputum culture in children with bronchiectasis may reveal microorganisms such as Haemophilus influenzae, Streptococcus pneumoniae, Moraxella catarrhalis, Staphylococcus aureus, Pseudomonas aeruginosa.

Colonization of P. aeruginosa in bronchiectasis in children is less common than in adults, is detected mainly in patients with cystic fibrosis, and is usually associated with a more severe course of the disease [4–7].

A number of patients with bronchiectasis may develop bronchial obstruction, the genesis of which is complex and multicomponent: not only irreversible structural changes in the bronchial tree play a role in the formation of bronchial obstruction, but also the influence of inflammatory mediators [5–7].

Bronchial hyperreactivity is detected in 40% of patients with bronchiectasis, a positive test with a bronchodilator when examining external respiratory function is detected in 20–46% of patients [5, 8].

Classification

According to the accepted Classification of clinical forms of bronchopulmonary diseases in children, bronchiectasis and bronchiectasis, which are a manifestation of another pathology, are distinguished.

Bronchiectasis (J47) is an acquired chronic inflammatory disease of the bronchopulmonary system, characterized by a purulent-inflammatory process in dilated deformed bronchi with infiltrative sclerotic changes in the peribronchial space. Due to the fact that one patient may experience different types of bronchiectasis, the prevalence and localization of changes within specific bronchopulmonary segments is of greater importance.

Cylindrical bronchiectasis occurs mainly with sclerosis of the bronchial walls. In this case, the lumen of the bronchus expands evenly over a fairly large area. Most often this occurs against the background of other lung diseases (secondary bronchiectasis). The cylindrical shape does not contribute to the accumulation of a large volume of pus, therefore the general condition of patients, as a rule, is not too severe; in some cases, such bronchiectasis can regress when the cause that caused them is eliminated (foreign body aspiration, atelectasis, infections).

Saccular bronchiectasis is a single spherical or oval expansion on one side of the bronchus. Often this form occurs with congenital defects in the development of lung tissue. The bags are blind protrusions of the wall that can reach large sizes. A significant amount of sputum and pus accumulates here. The course of the disease in such patients is usually severe.

A common variant of the development of bronchiectasis is partial obstruction of a large bronchus by a tumor, foreign body, scar, or compression from the outside by enlarged lymph nodes. Such bronchiectasis occurs in the area of partial or complete atelectasis and is designated as atelectatic.

In the mechanism of development of bronchiectasis, a certain role is played by the traction of the bronchial wall by fibrous cords from the surrounding fibrous tissue, and therefore the concept of traction bronchiectasis has been established in scientific terminology [5–7, 12, 13].

During bronchiectasis, two phases are distinguished.

The exacerbation phase is an active inflammatory process with the accumulation of pus. During this period, the symptoms of the disease are most pronounced. In some cases, in the absence of adequate treatment, a rapid deterioration in the patient's condition may occur: the inflammatory process goes beyond the dilated bronchus and pneumonia develops. The frequency of exacerbations can vary - from several episodes per year to several within one month.

The remission phase is characterized by the absence of acute symptoms. Bronchiectasis persists. In the presence of multiple dilations of the bronchi and concomitant pneumosclerosis, a dry or wet cough and signs of respiratory failure may be observed in the remission phase.

DIAGNOSTICS

Due to the fact that bronchiectasis can occur both as an independent disease and as a manifestation of another pathology, the diagnostic approach should be multidisciplinary.

The presence of bronchiectasis in a child can be assumed if the following clinical symptoms are present:

• chronic cough (productive or without sputum) for more than 8 weeks;
• persistent wheezing in the lungs that cannot be explained by other reasons;
• incomplete resolution of pneumonia after adequate therapy, or repeated pneumonia of the same location;
• “asthma”, torpid to adequately prescribed and administered therapy;
• the presence of respiratory symptoms in children with structural and or functional disorders of the gastrointestinal tract and upper respiratory tract;
• hemoptysis.

Surgery

Surgical treatment of bronchiectasis (resection of part of the lung) is recommended:

• for localized bronchiectasis (prevalence of no more than one lobe - limited process), which is a source of frequent exacerbations of lower respiratory tract infections, significantly worsening the patient’s quality of life;
• dangerous (more than 200 ml/day) bleeding or hemoptysis (uncontrolled by conservative therapy) from the local affected area. An alternative to resection in the latter case is bronchial artery embolization [1–5, 7, 15]

A comment. Surgical treatment is performed extremely rarely in patients with local bronchiectasis, persistent recurrent pneumonia of the same localization, frequent bleeding, infection or prolonged segmental collapse of the lung, usually only in cases where the possibilities of conservative therapy have been exhausted. The decision to operate depends on a number of factors - the frequency and severity of hemoptysis, the location of bleeding

(from the area of bronchiectasis), the presence of local or diffuse bronchiectasis. Their relapses occur in 20% of patients after surgery.

CONCLUSION

In the pediatric population, bronchiectasis is not as common, but the exact prevalence figures are unknown. The detection of bronchiectasis in the absence of specific symptoms for this nosology should be ensured by diagnostic measures with good quality visualization and high professional awareness of specialists about this problem. The modern differential diagnostic approach and principles of management of such patients should be multidisciplinary, which will ensure timely diagnosis and improve the prognosis for children with this pathology.

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