Bronchiectasis

Methodic materials for international students (IV-VI year)

Author: N.A.Filippova, assistant professor

Published: 2004

Definition

Bronchiectasis is an irreversible local dilatation of the bronchi due to the destruction and neural-muscular tone disturbances of their walls/

Types of bronchiectases:

	Primary	Secondary
Aethiology	In case of absence of underlying lungs diseases (pathomorphological basis of bronchoectatic disease)	Bronchiectases as a complication of other diseases: chronic pulmonary abscess, tuberculosis caverna, foreign body of bronchus, COPD
Localization	More common - lower lobe of the left lung	More common in middle lobe of the right lung

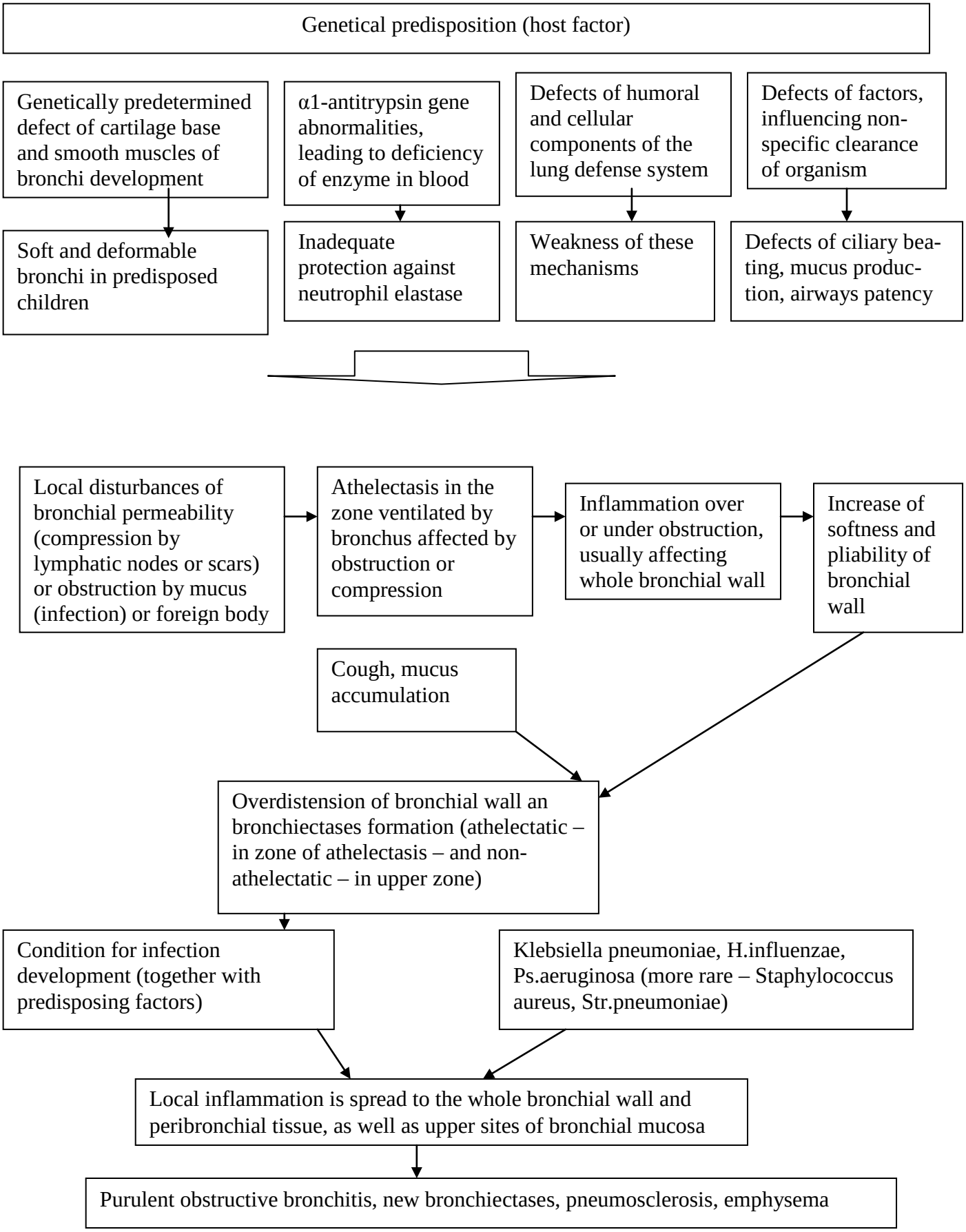
Bronchiectatic disease:

Acquired disease, appearing in childhood or adolescence, which is characterized by localized chronic purulent inflammation in regionally dilated bronchi, first of all in lower segments of lungs.

Prevalence

Bronchiectatic disease is 3 times more common in males.

Pathogenesis



Morphology

1. Usually localized in small bronchi
2. Microscopically there is a varying degree of damage to the bronchial epithelium:

- areas of ulceration
- patch-like replacement of columnar ciliated epithelium by squamous epithelium.
- goblet cell and mucus gland hyperplasia
- peribronchial connective tissue is often damaged or lost (leading to dilated but collapsible airways)
- there may be microabscesses in the bronchial wall and evidence of pulmonary vascular dilatation, occlusion, or hypertension.
- usually extensive inflammatory cell infiltration which consists of neutrophils (predominantly in the lumen) and mononuclear cells (in the bronchial wall).
- variable degree of tissue oedema and fibrosis.

Morphological types of bronchiectasis

Destructive bronchiectasis	Atelectatic bronchiectasis	Retentional bronchiectasis
- less common - associated with the most severe damage and large saccular dilatation with loss of bronchial subdivisions (saccular broncіectasis). - destruction of whole depth of bronchial wall may be present (muscles, glands, cartilages, elastic tissue) with affection by inflammation or fibrosis of respiratory tissue Usually saccular	- usually lobar or segmental - occurs in absence of other forms - usually related to proximal bronchial distortion or occlusion - inflammatory changes are less marked histologically Both cylindric and saccular	- due to reduction due to decrease of muscular tonus of bronchi (often due to chronic bronchitis progression, more rare – in case of bronchi hypoplasia in childhood) Often cylindric

Some textbooks also define follicular bronchiectasis: characterized by the formation of multiple lymphoid follicles which may distend and project into the bronchial tree. They are associated with extensive loss of elastic tissue in the bronchial wall and occur in most of cases of

bronchiectasis. The follicles themselves contain activated lymphocytes which are predominantly CD8 positive cytotoxic T cells. In addition, there is an increase in B lymphocytes in the bronchial wall. At present it is known that many of these produce the IgA1 and IgA2 subclasses of immunoglobulins. It is unknown whether B lymphocytes producing the IgG subclasses are also increased in number, but there is evidence of the local production of the IgG subclasses 1 to 4.

Classification of bronchiectatic disease

1. Form: cylindric, saccular, mixed
2. Localization: one-sided, both-sided, singular, numerous
3. Severity: mild, moderate, severe

Criteria	Mild	Moderate	Severe
Frequency of relapses	1-2 times a year	3-4 times a year	>4 times a year
Intoxication	Minimal	Moderate	Significant
Delay of growth and sexual development (in children	No	Possible	Marked
Respiratory function disturbances	No	Moderate	Severe
Localization	One-sided	2-sided	Diffuse

4. Complications (present, absent):

- pneumonia
- chronical purulent obstructive bronchitis
- bleeding
- pulmonary emphysema
- pneumosclerosis
- respiratory failure
- cor pulmonale
- secondary amyloidosis
- seronegative arthropathy, particularly during exacerbations of the disease. Recent studies have suggested that the arthropathy may be the result of immune complexes formed between high levels of circulating immunoglobulin and bacterial antigens.

- Vasculitis: cutaneous vasculitis has been described in severe bronchiectasis with and without cystic fibrosis. Again, the cause is thought to be circulating immune complexes which may be deposited in peripheral vessels.

Clinical manifestations

1. **Cough:** excessive (more than 100 ml, more in saccular, less in cylindrical bronchiectases) purulent sputum which is divided in 2 layers while kept in ordinary conditions. Cough and expectoration are increased in position, when the bronchus which is enabling bronchiectasis drainage has vertical downwards position.
2. **Haemophthysis:** present in 95%. Revealed even if there are "dry" bronchectases (without purulent inflammation) – Bezanson's syndrome
3. **Dyspnea**: expiratory, due to bronchial obstruction and emphysema. Appears at the rather progressive stage of the disease ("appears late but doesn't go away after appearance").
4. **Emphysema**: objective signs
5. **Intoxication**: perspiration, weakness, fever during exacerbations. Intoxication, at first, is present in exacerbation only, then it becomes constant. Delay of growth and sexual development may be present in children. Clubbing fingers and nails may be revealed.
6. **Objective signs of bronchial inflammation:** medium to coarse crackles may be heard over the affected zone
7. **Objective signs of bronchial obstruction:** breath with prolonged expiration (breath itself may be weakened due to emphysema)
8. **Signs of pleura affection may be present:** pleura friction rub

Laboratory and instrumental diagnosis

1. **Signs of inflammation:** WBC increase with shift of the neutrophils formula to the left; ESR increase, increase of α2- and γ-globulins, fibrinogen level, CRP.
2. **Secondary erythrocytosis** may be present
3. **Spirogram:** both obstructive and restrictive changes (decrease of both VC and FEV1)
4. **Chest X-ray**: reticular deformation of lungs picture, pneumofibrosis signs, which also may lead to reduced lungs volume

5. **CT and bronchograms are the main method of the diagnosis**
6. **Bronchoscopy** may give possibility to suspect the diagnosis (pus coming from one of the bronchi)

Diagnostic formula example:
Bronchiectatic disease, moderate course, exacerbation. Bronchiectases of basal segments of right and left lung. Chronic obstructive bronchitis. Complication: kidneys amyloidosis, proteinuric stage

Differential diagnosis
Tuberculosis, bronchial cancer and pulmonary abscess

Course and outcomes
Usually progressive course with complications development

Treatment

I. Approach to surgical treatment

Is indicated in moderate course (in about 40% of patients) in one-sided localized bronchiectases (pulmonary resection).
Palliative operation is used in 2-sided localized bronchiectases (resection at the most affected side)

Lung and heart-lung transplantation is being undertaken successfully in cystic fibrosis patients with severe endstage lung disease with or without cor pulmonale. A similar approach may be indicated in bronchiectasis without cystic fibrosis. However, the time course of the disease is longer in the absence of cystic fibrosis and respiratory failure is more likely to occur in middle to late middle age, if at all. Nevertheless, this option should be considered for any severely affected younger patient.

II. Medications

1. **Antibiotics:**

- depends on the results of bacteriological investigation (more often – wide spectrum penicillin, augmentin, III generation cephalosporins, aminoglycosides).
- intravenous and intramuscular use of antibiotics.
- Endobronchial bronchoscopic clearance with removing of pus and infusion of antibacterial drugs (0.1% furacilline or 1% dioxidine)

2. Drainage

- postural drainage – 6-8 times a day – being in position with the bronchus enabling drainage is in vertical downwards position
- mucolytics
- broncholytics (β2-mimetics, cholynolytics)
- endobronchial bronchoscopic clearance

3. Infusions: metabolic and desintoxication treatment

- glucose solution 5% - 1-3 liters daily with potassium and magnesium preparations
- protein preparations: protein hydrolysates (hydrolysin), aminoacids solution (polyamine, panamin), human albumin (100 ml twice a week); human plasma of the same blood group
- desintoxication solutions: dextranes (haemodes, reopolyglucin); extracorporal detoxication (plasma exchange therapy, sorbtion technologies, UV irradiation of blood)

4. Immune system correction

- human plasma of the same blood group (300-500 ml every 2^{nd} day – 3-5 times)
- immune globulins (passive immunization, used in exacerbation and marked intoxication): normal human immune globulin (25-50 ml daily or every 2^{nd} day – 5-7 injections; anti-flu Ig – 3 doses intramuscular, 5-7 days; staphylococcus γ-globulin – 3-7 ml intramuscular daily or every 2^{nd} day -5-7 injections)
- T-cells stimulation (at the end of exacerbation): T-activin 100 mkg once daily - 5-7 times; thymalin – 10 mg 5-7 times