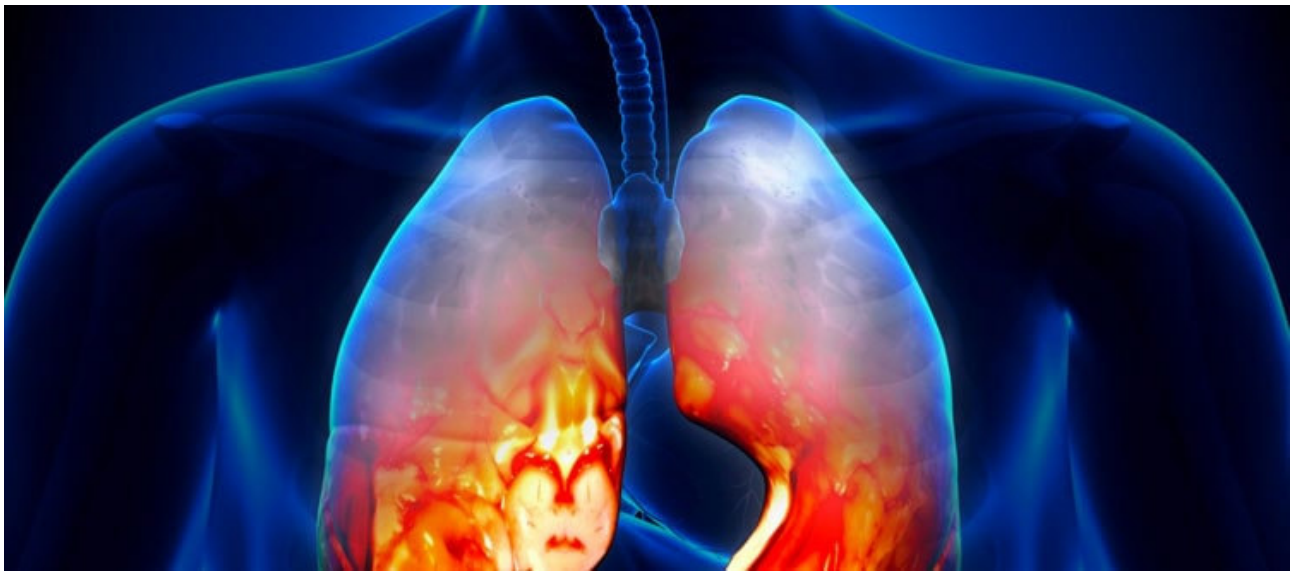
Cystic fibrosis

Mucoviscidosis is the serious congenital disease which appears as damaged tissues and suppressed secretory function of exocrine glands. The disease has a negative impact on function of many body systems, in particular, gastrointestinal and respiratory systems. Therefore, mucoviscidosis can be categorized into lung, gastrointestinal and combined forms.
The lung disease (cystic fibrosis) is the respiratory type of mucoviscidosis characterized by destructive changes in bronchopulmonary system, breathing difficulties, presence of sputum and bacterial infection.
A mucoviscidosis patient has the following symptoms of lung disease:

- Fits of cough, which may be accompanied by vomiting.
- Solid sputum and coughing up difficulties.
- Sputum might be mucusy or purulent.
- Breathing difficulties with possible dyspnea and pale skin.
- Fatigue.



Diagnosis and disease course particulars
In children, mucoviscidosis diagnosis can be established even in the earliest days of life. Presence of the gene mutation (cause of the disease) appears as intestinal obstruction, provoked by excessively dense meconium. The disease can be identified by blood laboratory testing which detects elevated immunoreactive trypsin levels. Also, Gibson-Cooke sweat testing can be conducted to establish the precise diagnosis. Radiography, bronchoscopy, spirometry and other examinations are helpful in defining severity and possible complications of the disease. The lung disease has progressive course that can be described by the four stages:

- The first stage lasts from birthday to 8-10 years of age. The symptoms include dry cough, fatigue, and dyspnea following slight physical activity. In children, the lung form of mucoviscidosis may be accompanied by sleep disorders, lack of appetite, apathy.
- The second stage follows the first one and may last between 3 to 15 years. During this period, sputum appears solid, so coughing up is getting more difficult. Dyspnea is getting worse.
- The third stage is characterized by complications. The disease affects functioning of various body systems. Secondary destructive changes in bronchi are seen. Stage duration is 3 to 5 years.
- The fourth stage. During this period, frequency of cardiovascular insufficiency increases, which often leads to fatal outcomes.

Treatment of mucoviscidosis

Health-promotion procedures, supportive and symptomatic therapy are recommended for the patients suffering from cystic fibrosis. Mucoviscidosis treatment involves some efficient physiotherapeutic measures, such as massage, breathing exercises, drainage. A patient should eat healthy food, keep proper hours, and take all the necessary vitamins.

The symptomatic treatment of the lung disease includes administration of the following medicines:

- Glucocorticosteroids to treat oedema, spasm, and to relieve breathing.
- Mucolytics to thin solid sputum; they contribute to expectoration (recombinant human deoxyribonuclease, 7% sodium chloride solution).
- Antiseptics (inhaled).
- Antibacterial medicines, when bacterial infection is revealed.

The treatment option recommended for cystic fibrosis patients is inhalation therapy. Efficient inhalation should be carried out with a mouthpiece and include the following medicines:

- The Okistar Hyal 7; inhalation sessions are twice daily or as necessary.
- If the inhaled medicines listed above cause discomfort, less concentrated sodium chloride solutions should be used (Lorde® Hyal).