# BRITISH MEDICAL JOURNAL

# British Medical Journal

## Volume 3, No.1, January 2023

**Internet address:** http://ejournals.id/index.php/bmj
**E-mail:** info@ejournals.id
Published by British Medical Journal
Issued Bimonthly
3 knoll drive. London. N14 5LU United Kingdom
+44 7542 987055



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# CLINICAL-FUNCTIONAL CHARACTERISTICS OF CYSTIC FIBROSIS DISEASES IN ARAL SEA REGION - ACTUAL PROBLEM.

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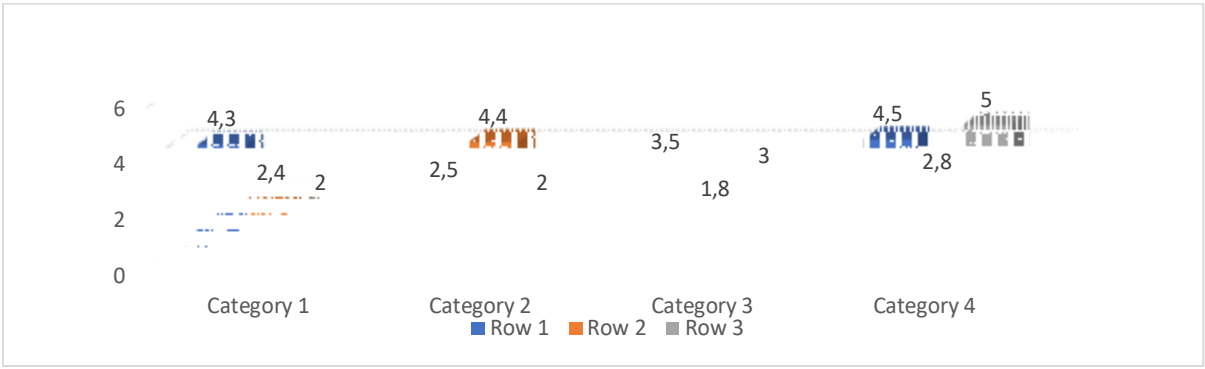
*Abstract: Studies have been carried out to determine the mechanisms of the effect of vitamin D deficiency on the clinical manifestation, course and prognosis of intestinal diseases in children in the Aral Sea region, as well as to develop substantiated methods of corrective therapy. Based on the data obtained, it was concluded that the analysis of the results in the dynamics of observation will make it possible to predict the course and outcome of the disease, evaluate the effectiveness of the therapy, and, if*

*Keywords: Aral sea region,bowel disease, children, cystic fibrosis .*

Among all forms of cystic disease, the intestinal form is the most common. The basis of this disease is a hereditary pathology, which is manifested by damage to the glands of the digestive system, in particular, the pancreas and liver. Among the diseases of cystic fibrosis, the intestinal form (KFM) is accompanied by serious disorders of the gastrointestinal tract and respiratory system (in a mixed form). This disease occurs with the same frequency in boys and girls. In Asian countries, the frequency of intestinal form of this disease is about 1:2700. 1:1100 IN THE IN ARAL SEA REGION

KFM is caused by a mutation. A mutation common to all forms of cystic disease involves the gene responsible for the excretory function of the glands. As a result, the sweat, salivary, lacrimal, bronchial glands, as well as the pancreas begin to produce a thick, viscous secretion that is difficult to separate. In addition, with cystic disease, the transport of electrolytes through cell membranes (mainly sodium and potassium) is disturbed, as a result of which the severity of metabolic diseases in the body increases.

A gene mutation can occur completely by itself, it is influenced by harmful environmental factors such as smoking, maternal alcohol consumption, and poor environmental conditions.




Even if both parents had cystic disease in the family, such a family may have healthy children. The probability of having a sick child in such a family is 25%. The inheritance of cystic fibrosis is distributed as follows: the probability of having a sick child is 25%, the probability of having a healthy child with a mutant gene is 50%, and the probability of having a child with a completely normal set of genes is 25%.

**Clinical manifestations**

Cystic fibrosis has the following forms:

1.form (in which the gastrointestinal tract and the bronchopulmonary system are simultaneously affected).

2.Mainly lung shape.

3.Mainly intestinal form.

4.Atypical or deleted form.

5.Space obstruction.

In most cases, the intestinal form of cystic disease appears not from the first day of life, but from 8-9 months, when the child begins to introduce a sufficient amount of complementary foods or when the child is transferred to artificial feeding. With this form of the disease, most importantly, the functioning of the intestines and pancreas is disturbed. The child develops a deficiency of pancreatic enzymes, which leads to breakdown and absorption of fats and proteins.

**Diagnostics**

Neonatal diagnosis. The child is carefully examined and examined in the maternity ward, immediately after birth and during the entire period of stay in this medical institution. Neonatologists rule out birth defects, including intestinal defects. Special attention is paid to the timely release of meconium. Usually, the first part of meconium should pass in 8-10 hours of the child's life, meconium can be released for 2-3 days. In the intestinal form of cystic fibrosis, the meconium is very thick and sticky, and it is difficult to pass it.

In severe cases, a specific form of intestinal obstruction develops - meconial ileus. Biochemical screening. This study can be conducted for children in the first month of life, for most regions (including Ukraine, Russia, Belarus) screening for cystic fibrosis is mandatory in maternity hospitals, and it is carried out for all children without exception. The level of specific pancreatic enzyme - immunoreactive trypsinogen in the blood is checked. In children with cystic fibrosis, the amount of this enzyme is several times higher than normal.

Sweat sample. If any form of cystic fibrosis is suspected in a child, it is already done. In this disease, the child's sweat glands also produce a more concentrated secretion, and the child's skin contains more salt than healthy children. It should be remembered that at least three sweat tests must be performed to make a final diagnosis.

Stool analysis. It allows to indirectly determine the functional activity of enzymes of the liver and pancreas (the amount of unseparated proteins or fats is determined).

**Treatment**

Fermentotherapy plays a key role in the treatment of the intestinal form of cystic fibrosis. It is appointed by courses and is held throughout his life. Children who regularly receive enzyme therapy do not differ from their peers. Growth and mental development in such children are completely normal. Recently, microspherical drugs such as Creon or Pancitrate have been chosen for enzyme therapy. A special feature of these drugs is that the active substances in them are placed in microspheres surrounded by a gelatin capsule. Under the influence of enzymes, the capsule dissolves in the upper parts of the gastrointestinal tract, and microspheres containing enzymes mix with food, ensuring its uniform and complete digestion.

Antibiotic therapy is not used as a rule, as in the intestinal form of cystic fibrosis, but it can be used to treat the mixed form of this disease (when both the lungs and the intestine are affected). In such a situation, along with antibiotics, sputum thinners are also taken, which make it easier to remove thick sputum from the lungs.

Hepatoprotectors (Urosan, Ursofalk, etc.) can be included in the treatment regimen.

In addition to having a positive effect on the morphofunctional state of hepatocytes, these drugs help dilute bile, which is very important in the intestinal form of cystic fibrosis, which is often accompanied by cholelithiasis.

Vitamin therapy is also a very important part of the treatment regimen for the intestinal form of cystic fibrosis, because the digestion and absorption of fat-soluble vitamins in the intestines of these sick children is impaired. Therefore, these vitamins should enter the child's body not only with food, but also as part of medicines. In particular, it is necessary to ensure that the child receives a sufficient amount of vitamins A, D, K and E.

Diet therapy. Currently, two different approaches are used to formulate a diet for children with cystic fibrosis.

The first (more modern) approach is used in children who regularly take enzyme preparations. Such children are offered improved nutrition (up to 150% of the age norm), in which the percentage of fats should be at least 30%.

It is believed that a child who is on permanent enzyme therapy can eat any food allowed for his age. It is allowed to use fatty sour cream, cottage cheese, butter and sunflower oil. The child should receive a large amount of fluid, while maintaining the normal urinary function of the kidneys, it is allowed to drink more than twice the age norm. In addition, taking into account the large amount of sodium chloride excreted with the sweat of cystic fibrosis children, such children should salt their food (1 to 5 grams of table salt per day is allowed as prescribed by the doctor).

Since children with cystic fibrosis are, in most cases, on permanent enzyme therapy, the second approach, which has been used everywhere until now, is now considered obsolete. If, for some reason or another, there is no enzyme therapy, it is necessary to limit fat and predominate protein foods in the child's diet.

The best option for feeding babies is breast milk, because it contains not only the right amount of proteins, fats and carbohydrates (with proper nutrition of a nursing mother), but also enzymes for their digestion. Bottle-fed children usually receive medicinal mixtures with protein hydrolyzate, which are more easily absorbed and help to quickly restore weight.

**Features of care and lifestyle**

Given the severity of the disease and the lack of a complete cure for children from this pathology, children with cystic fibrosis require constant care and attention.

Medicines and medical care for such children are free in almost all countries, but upon confirmation of this diagnosis, parents must immediately apply to the VKK (medical advisory commission) to give the child the status of a disabled child, only in this case the child can use the benefits provided by the state.

Such children are vaccinated according to the usual schedule. Before vaccination, it is necessary to conduct a general blood and urine analysis to rule out a hidden inflammatory process that may occur in the child's body. In addition, the child should be examined by a pediatrician immediately before vaccination. If there are any, even the smallest manifestations of Arvi or the main disease worsens, vaccination is postponed to the next day. Care of children with cystic fibrosis also has its own characteristics. After one year, children should eat at least 6 times a day, with the constant use of medication during the day, so all family members should strictly follow all the recommendations of the attending physician regarding dietary therapy and drug treatment.

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