FEATURES OF THE COURSE AND COMPLICATIONS OF CYSTIC FIBROSIS DEPENDING ON AGE

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Abstract: *Cystic fibrosis (CF) is a hereditary disease caused by systemic dysfunction of the exocrine glands. Cystic fibrosis is most common among Caucasians, but cases have been reported in representatives of all races. Among the Caucasian population, one case of the disease is registered per 2,000 to 3,000 newborns, and among the black African and Japanese populations, its frequency is one case per 100,000.*

Key words: *cystic fibrosis , exocrine part, secretion, glands.*

The disease was first described relatively recently, in 1938 by a pathologist Dorothy Anderson described cystic degeneration of the pancreas in combination with pulmonary pathology in young children [1]. Then the English name of the disease arose - Cystic fibrosis (cystic fibrosis). In 1946, Farber proposed the term " Mucoviscidosis " (from the Latin mucus - mucus, viscus - viscous), indicating the role of increased viscosity of the excreta secreted by the exocrine glands. However, even earlier, in old German tales, it was indicated that if a child's kiss is accompanied by a salty taste, the child is doomed [2]. Cystic fibrosis affects the exocrine part of the pancreas. In this regard, we can consider cystic fibrosis as one of the causes of diabetes in children. The high prevalence of the disease, a pronounced tendency to further growth, severe progressive course, early disability due to various complications put diabetes mellitus on a par with the most important problems of medicine and require further study of the pathogenetic mechanisms of its development and progression [3].

Every twentieth European is a carrier of the cystic fibrosis gene , and if both parents have the gene, the chance of having a sick child is 25%. In Russia, cystic fibrosis is diagnosed in one out of 5,000 newborns. In Europe and North America, cystic fibrosis occurs in 1 case per 2,500 newborns, while its incidence in Africa and Asia is low. The frequency of births of children with this disease is 0.5%. It is responsible for up to 6% of deaths in early childhood.

Moreover, patients often live significantly less than their peers. In the past , it was believed that this disease affects the lungs and digestive system , but now it is known that it affects most organs.

A genetically determined disorder of the excretory function of the exocrine glands, mainly the respiratory system and gastrointestinal tract. It is caused by mutations in the gene encoding the membrane protein CFTR, which is a channel for chlorine ions in the membranes of epithelial cells, a regulator other ion channels, and is also responsible for the transport of bicarbonates. The absence of synthesis or the synthesis of pathological protein causes a blockade or deterioration in the transport of chlorine from the cell and an increase in the absorption of sodium into the cell, which leads to a decrease in the water content in the secretions of the exocrine glands. The low volume of paraciliary fluid secreted by the epithelium makes adequate mucociliary cleansing impossible, and the high concentration of NaCl and a change in the pH of the secretion reduce the activity of antibacterial peptides.

According to a number of authors, an increase in the level of immunoreactive trypsinogen in MB occurs as a result of blockage of the ducts of the pancreatic glands with viscous secretion, which prevents trypsinogen from penetrating into the lumen of the small intestine, where it is normally converted to trypsin. This leads to the release of trypsinogen into the blood [4].

In addition, the cause! of an increase in the level of IRT in. In addition to CF, the blood of newborns may contain a number of congenital and hereditary pathologies, such as: intrauterine hypoxia of the fetus, intrauterine infections, perinatal stress, fetal immaturity, conjugation jaundice of newborns, chromosomal rearrangements, etc., as well as heterozygous carriage of mutations

in the CFTR gene, as a consequence of functional insufficiency of the pancreas [5].

The aim of the study: to study the characteristics of the course and frequency of complications of Cystic Fibrosis in children.

Materials and methods. Of all the sick children admitted to the Samarkand branch of the Russian Scientific Center for Emergency Medicine, 38 patients with Cystic Fibrosis were identified in the children's department from 2018 to 2020 , who had clinical symptoms (wheezing, shallow breathing, persistent cough with viscous, sometimes purulent sputum, pale skin, dry mouth, frequent "oily" foul-smelling stool, intestinal colic, hypotrophy). The diagnosis of " Cystic fibrosis " was made based on the ultrasound of the pancreas, as a result of the increased content of chlorine in sweat fluid, Na and Cl in the blood and nail plates, according to results of a genetic study to identify partial mutations.

Results: During the observation period, the number of children hospitalized for Cystic Fibrosis remained stable : 16 in 2018, 13 in 2019, and 9 in 2020. At the time of diagnosis, 15.7% (6) of children were aged 0–6 months, 5% (2) in the age groups 6–12 months and 1–3 years. It should be noted that neonatal screening was performed only in 21% of cases (8 children), while it was not performed in 30% of patients, and there was no data on its implementation for two patients, therefore they were not included in the statistical processing. The diagnosis of Cystic Fibrosis , mixed form was made in 73% of cases. The data by severity were distributed as follows: in 2018, 11 patients (68.7%) were observed with severe Cystic Fibrosis , 31.2% (5) — moderate severity; in 2019 — 61.5% (8) — severe, 30.7% (4) — moderate (in 1 case there were no data on severity); in 2020 — 66% (6) — severe, 33% (3) — moderate severity. In the structure of complications, the leader was chronic pancreatic insufficiency: in 2018 and 2020 it complicated the underlying disease in 100% of cases, in 2019 — 50%. The next most common complications were bronchiectasis : in 2018 — 62.5% (5 out of 8), in 2019 — 50% (3 out of 6), in 2020 — 80% (4 out of 5). Next was pneumofibrosis: 2018 — 12.5% (2), 2019 — 10% (1), 2020 — 33% (3).

Pulmonary hypertension: in 2018 — 31.2% (5), 2019 — 30% (4), 2020 — 11% (1).

Conclusion. Thus, the diagnosis of Cystic Fibrosis is most often detected in children aged 0–6 months (15.7%). Neonatal screening was performed in 21% of cases. The predominant form of cystic fibrosis - mixed (73%), severe. The most common complications are chronic pancreatic insufficiency (50-100%), bronchiectasis (50–80%), pneumofibrosis (10–33%), pulmonary hypertension (11–31.2%).

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