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PRIMARY IMMUNODEFICIENCY CONDITIONS

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Abstract
One of the most pressing problems in medicine today is the growth of immune-dependent pathologies, i.e. diseases based on immunopathological mechanisms [1, 7]. Primary immunodeficiencies are a large group of severe, genetically determined diseases, the cause of which is a violation of the body's immune response cascade [2, 4]. Diseases with immunopathological conditions are characterized by early chronicity, reluctance to traditional methods of therapy and the need for immunorehabilitation [1]. This article is devoted to studying the problem of diagnosis and treatment of primary immunodeficiencies, with the aim of reducing the mortality of patients and improving their quality of life.

Keywords: primary immunodeficiency, main groups, autoimmune diseases, immunoglobulins, treatment tactics.

Introduction

Primary immunodeficiency is a group of diseases that affect the immune system. People with primary immunodeficiency have a weakened immune system, making them more susceptible to infections, autoimmune diseases, and other health conditions. Primary immunodeficiency is a relatively rare disease, but it can have significant consequences for those affected. Timely and accurate diagnosis is essential to ensure that people with primary immunodeficiency receive proper care and treatment.

Despite the fact that there are numerous studies on immunology, and in particular on primary immunodeficiencies, the alertness of doctors regarding primary immunodeficiency is low. This is due to the fact that primary immunodeficiencies are considered quite rare and complex diseases. However, data from the International Primary Immunodeficiency Foundation (Jeffry Modell Fondation) indicate that at least 10 million people on Earth suffer from one form or another of primary immunodeficiency [3, 13].

From the group of primary immunodeficiencies, based on the available data on the mechanisms of development of these diseases, 4 main groups can be distinguished:

-humoral or B-cell immunodeficiencies;

- combined immunodeficiencies, in which the cellular and humoral components of immunity are affected;

- phagocytosis defects;

- complement defects.

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The onset of clinical signs of primary immunodeficiency occurs in childhood, however, for example, with a hypomorphic mutation, the typical clinical picture of the disease may appear at a later date [3, 9]. Moreover, the time period between the initial visit to a pediatrician or general practitioner and the diagnosis of primary immunodeficiency ranges from 9 months to 4-7 years [2, 17].

Symptoms of primary immunodeficiency can vary widely depending on the specific disease and the severity of the immune system disorder. Common symptoms of primary immunodeficiency include recurrent infections such as ear infections, sinusitis, pneumonia and skin infections. In addition, people with primary immunodeficiency may experience chronic diarrhea, persistent thrush or fungal infections, and an inability to develop normally. They may also be more susceptible to autoimmune diseases such as lupus or rheumatoid arthritis [3, 6, 8].

Diagnosing primary immunodeficiency can be challenging because symptoms are often nonspecific and may be similar to those of other conditions. Doctors typically begin by conducting a thorough medical history and physical examination. They may also order a series of blood tests to evaluate immune system function, including a complete blood count, immunoglobulin levels, and antibody function tests. Imaging tests, such as a chest x-ray or CT scan, may also be used to evaluate signs of infection.

In some cases, genetic testing may be used to identify specific genetic mutations associated with certain forms of primary immunodeficiency. This testing may be especially useful in cases where the diagnosis is unclear or when symptoms are atypical. Genetic testing can also help identify individuals who may be at risk of developing primary immunodeficiency and can help guide family planning decisions [2, 14, 16].

Once primary immunodeficiency is diagnosed, treatment will depend on the specific disease and the severity of the immune system disorder. In some cases, simple measures such as prophylactic antibiotics, vaccinations and good hygiene may be enough to prevent infections. In more severe cases, immunoglobulin replacement therapy, bone marrow transplantation, or gene therapy may be required to restore immune system function [1, 12, 15].

Even earlier, knowledge about primary immunodeficiency was insignificant, and treatment options were minimal. Currently, the situation has changed dramatically. The emergence of immunoglobulins, which have been actively used as replacement therapy for primary immunodeficiency with antibody deficiency, has significantly changed the prognosis of patients. Timely initiation of regular replacement therapy in an adequately selected dose allows patients to significantly reduce the risk of developing severe infections, lead an active social life and even have healthy offspring. The emergence of the opportunity to perform bone marrow transplantation in patients with primary immunodeficiency gave them a chance not only to survive, but also not to have symptoms of the disease during treatment [2, 5, 10, 11].

Conclusions

In conclusion, diagnosing primary immunodeficiency is a complex process that requires a comprehensive assessment of the patient's medical history, physical examination, and laboratory results. Clinicians should be vigilant for signs of immunodeficiency, especially in patients with recurrent infections or autoimmune disorders. An accurate diagnosis is essential to ensure that affected individuals receive appropriate care and treatment and to minimize the long-term health consequences of primary immunodeficiency. With proper diagnosis and treatment, people with primary immunodeficiency can lead healthy and fulfilling lives.

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