SINDROAME DE AFECTARE RESPIRATORIE LA COPILUL CU IMUNODEFICIENȚE PRIMARE

RESPIRATORY SYNDROMES IN CHILDREN WITH PRIMARY IMMUNODEFICIENCY

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Summary

Aim: evaluation of respiratory syndromes found in children with primary immunodeficiencies.

Methods: in the study, were evaluated 14 children with primary immunodeficiencies, diagnosed in the Pneumology Clinic of IMSP Mother and Child Institute. The patients in the study were divided according to the type of PID. The patients were evaluated by methods: determination of serum Ig, determination of cellular immunity, imaging examination of the chest by Rx or CT.

Results. Of 14 patients evaluated in study, 46.8% were diagnosed with primary immunodeficiencies due to antibody deficiency, and 53.2% presented with combined primary immunodeficiencies (PID). Pulmonary infections, especially pneumonia, were the most common clinical manifestations in all patients with PID. Until the establishment of the diagnosis all patients (100%) present at least one episode of pneumonia, the number of episodes being directly proportional to the age of the diagnosed.

Conclusion: Complications of respiratory system diseases are frequent in primary immunodeficiencies in children, PID due to antibody deficiency being mainly associated with irreversible bronchopulmonary processes, with severe evolution.

Introduction.

Primary immunodeficiencies (PIDs) represent a group of genetic diseases characterized by quantitatively and/or qualitatively defects of different components of the innate and adaptive immune system [11,13]. The number of PIDs has grown exponentially with the advancement of genetic diagnostic methods. Nowadays more than 350 entities divided into 8 groups have been described, antibody deficiencies being the most common, comprising approximately 70-75% of the number of IDPs [1,2,8].

PIDs manifest as a broad spectrum of clinical manifestations, including increased susceptibility to infections, autoimmune processes, and malignancies.

Recurrent infections of the upper and/or lower respiratory tract are often the first and most frequent manifestation in the clinical picture of primary immunodeficiencies, especially in IDP with humoral involvement (CVID, XLA, HIgM, selective IgA deficiency). The recurrence and severity of these infections lead to complications in the respiratory system such as: bronchiectasis, mucus plug formation, fibrotic changes, emphysema, abscess, empyema, and pulmonary destructive processes [3-7]. In this context, any child with recurrent respiratory infections with severe evolution that are difficult to respond to treatment should be analyzed as a possible patient with IDP [12]. Because respiratory impairment is a significant cause of morbidity and the leading cause of mortality (30–65%) in both children and adults with IDP, early diagnosis and appropriate therapy can ameliorate or at least slow the progression of these complications.

Aim: evaluation of respiratory syndromes found in children with primary immunodeficiencies.

Methods. In the study, were evaluated 14 children with primary immunodeficiencies, diagnosed in the Pneumology Clinic of IMSP Mother and Child Institute. The patients in the study were divided according to the type of PID. The patients were evaluated by a complex of paraclinical methods: determination of serum

immunoglobulin values by the ELISA immunoenzymatic method, determination of cellular immunity, imaging examination of the chest and upper respiratory tract by radiography or CT.

Results and discussions. Of 14 patients evaluated in study, 46.8% were diagnosed with primary immunodeficiencies due to antibody deficiency, and 53.2% presented with combined primary immunodeficiencies. Following the evaluation of the immunological profile, in the group of children with PID due to antibody deficiency, the average concentration of serum immunoglobulins was considerably reduced, and in the case of patients with combined IDP, the serum level of immunoglobulins was at values comparable to age norms.

Pulmonary infections, especially pneumonia, were the most common clinical manifestations in all patients with PID. Until the establishment of the diagnosis all patients (100%) present at least one episode of pneumonia, the number of episodes being directly proportional to the age of the diagnosed.

Table 1. Frequency of pneumonia in studied patients according to PID entities

PID	Absolute number	Frequency
Combined PID	7	50%
CVID	1	7.1%
WAS	2	14.2%
HyperIgM	1	7.1%
XLA	3	21.4%

Pulmonary pathology was most frequently associated with some complications at the time of diagnosis [9]. Respiratory infections with various locations were not an

exception for the patients in our study, they went to the doctor because of severe infections (mainly pneumonia), complicated with acute processes (pulmonary destruction, empyema, confluent foci of pulmonary infiltration) or chronic pathologies upper and lower respiratory tract.

According to the results obtained, fibroatelectases and bronchiectasis are the most common complication recorded, they predominate in the group of patients with PID due to antibody deficiency. This phenomenon is explained by the importance of immunoglobulins in the protection of mucous membranes, especially the respiratory one, which is the most exposed to contact with pathogenic microorganisms. Similar data were also obtained in international studies, where the frequency of bronchiectasis varies between 37.6-75%, predominating in the group of humoral immunodeficiencies [3,6,9-10,12-15]. After analyzing a group of 65 patients, Membrila-Mondragon et al. reported that the most common complications were bronchiectasis (15 patients, p=0.12) and pulmonary fibrosis (3 cases, p=0.52) [4].

B lymphocytes and immunoglobulins A, M, G play an active role in the protection of the mucosal barrier and in protection against ENT infections, their deficiency resulting in recurrent ENT infections (19-98%) [7,13], associated with subsequent chronicity. In the patients evaluated in the study, chronic ENT changes were recorded with predominance in the IDP group due to antibody deficiency - chronic otitis (33.3%) and chronic sinusitis (66.7%).

Conclusion.

Complications of respiratory system diseases are frequent in primary immunodeficiencies in children, IDP due to antibody deficiency being mainly associated with irreversible bronchopulmonary processes, with severe evolution.

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