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DIAGNOSTIC POSSIBILITIES OF PID IN THE REPUBLIC OF MOLDOVA

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Awareness activities: PID are rather rare disorders but they are more common than it was estimated. **The rate** of recognition and diagnosis of PID is directly dependent on awareness of medical staff (JMF manuscript 2011).

According to V.Modell (2011), founder of the JMF, the awareness of medical staff and patients concerning PID remains low all over the world and the majority of patients with recurrent infections are not diagnosed or underdiagnosed. In fact, PID prevalence levels exceed the official data. The handbook entitled "Primary Immunodeficiencies in children" was elaborated for family practitioners, pediatricians and medical residents, and published in 2012. It covers the following topics: general information about **children** immune system, 10 warning signs for children with PID, clinical presentation, immunological and genetic features of the most common PID syndromes, diagnostic algorithms**, and** tables with reference values of immunological tests.

New diagnostic possibilities of PID in Moldova (2012). Determination of lymphocyte subpopulations using cytoflowmetric analysis, assessment of the IgG subclasses, IgD, evaluation of the C1-estherase activity, phagocytic burst-test using dihydrorhodamine , phagocyitosis killing activity (E.coli) using cytoflowmetric analysis.. Algorithm diagnostic of PID in Moldova**:** Clinical screening – 10 warning signs of PID JMF(clinical features characteristic for *well-defined syndromes of PID* are also taken **into** consideration). At risk of PID patients primary are evaluated:: family history, clinical course of disease (especially infectious syndrome characteristic for different forms of PID), documented presence of other features of PID (autoimmunity, malignancy),documented presence of other conditions which can be cause of infectious susceptibility (structural abnormalities, cystic fibrosis, etc).Children with **major risk** of IDP are selected and laboratory screening: WBC manual count is made, total IgA, IgG, IgM, Ig E. Patients with major risk of PID are examined using different diagnostic protocols depending on clinical presentation.

- Different immunological tests are carried out according to the Practice Parameter for the diagnosis and management of PID (Bonilla F. et al., 2005).
- ESID criteria for PID are used to establish the possible, probable and definitive diagnosis of PID if appropriate.

Current situation:

6 children are diagnosed with PID in Moldova currently:

- 5 patients with IgA selective deficiency (6-8 years old)
- 1 patient with Di George syndrome (4 years old)

Currently more than 150 genetic defects determining severe disorders of the immune system have been described. According to the European Society for Primary Immunodeficiencies database most of the reported immune deficiencies occur with a frequency of not less than 1:100000.

The improvement of the diagnosis PID will help to achieve in 2013-2014 the Project "**Complex diagnostic approach for patients with rare forms of primary immunodeficiency"** which will be achieved with Belarus Republic.

Objective of the Project: **To elaborate effective diagnostic approach** based on the analysis of retrospective and prospective clinical, immunological and genetic data of patients with rare primary immunodeficiency syndromes in the territory of the Republic of Moldova and the Republic of Belarus.

It is aimed to perform an in-depth analysis of the disease history, clinical data and immunological disorders in patients with rare (1:100000 to 1:1000000) primary Immunodeficiencies in Moldova and the Republic of Belarus. At least 50 patients with 20 primary immunodeficiency syndromes of rare incidence will be included.

On the basis of the received data in Moldova and the Republic of Belarus a similar algorithm and check-list for clinical and immunological assessment will be developed for children with rare primary immunodeficiencies. Therefore, a complex diagnostic approach must be elaborated for rare primary immunodeficiency syndromes, which will include clinical, immunological and genetic features, and will contribute to the same effective as for common immunodeficiency syndromes early diagnosis and appropriate treatment.

Activities which will help to improve PID diagnosis in Moldova:

- Implementation of lectures on childhood PID topics in the curricula of postgraduate training for family doctors and pediatricians.
- PhD research on PID problems.
- Organization of the J Project meeting in Moldova in the nearest future.