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Meeting report

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Winter Specialized Immunology School, “Inborn Errors of Immunity”

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On December 15–17, 2022, the Winter Specialized Immunology School, “Inborn Errors of Immunity,” was held in Uzhgorod of Zakarpattia Region with the support of Danylo Halytsky Lviv National Medical University, the International Interdisciplinary Association of Medical Professionals, the All-Ukrainian Association of Pediatric Immunology NGO, and the International European University. This school aims to analyze new approaches to the early diagnosis of primary/congenital disorders of the immune response in the context of the multidisciplinary approach, as well as to discuss the strategy of therapeutic tactics of such patients. The scientific event was held in the case marathon and direct communication formats. At the beginning of the event, a round table was held with the participation of regional experts in pediatric and adult immunology from different parts of Ukraine. More active cooperation with European colleagues and the creation of a national registry of immunodeficient patients with impaired antibody synthesis were noted. More than 20 speeches by clinical immunologists/pediatric immunologists were presented during two days of intensive work. During this school, results of Biopharma activities, a domestic producer of immunoglobulins, and plans for the future were presented in the field of domestic immunology development and diagnosis of patients with inborn errors of immunity. Discussions during the Winter School revealed the need to actively protect persons with congenital disorders of the immune system against infections by means of timely systematic seasonal vaccination, replacement immunoglobulin therapy, prophylactic antimicrobial therapy, and coordinated activities of specialists from various fields to provide immunological aid to patients in war conditions. At the end of the “Inborn Errors of Immunity” Winter School, organizers expressed confidence that most efforts and ideas highlighted during the professional meeting will be successfully implemented.

Keywords: Vocational school, primary immunodeficiency.

On December 15–17, 2022, the Winter Specialized Immunology School, “Inborn Immunity Faults,” was held in Uzhgorod in Zakarpattia Region, in the format of the case marathon and face-to-face communication.

Event organizers were the International Interdisciplinary Association of Medical Professionals, the All-Ukrainian Association of Pediatric Immunology NGO, Danylo Halytsky Lviv National Medical University, and the International European University represented by opinion leaders in the diagnosis and medical support of primary immunodeficiencies (inborn errors of immunity) – Professors Anastasiia Bondarenko and Valentyna Chopyak, Associate Professors Yuriy Stepanovskyi and Khrystyna Lishchuk-Yakymovych.

Inborn errors of immunity are characterized by quantitative and/or qualitative disorders in various parts of the human immune system resulting in the body’s inability to resist microbial aggression and/or the development of autoimmune, autoinflammatory, allergic, lymphoproliferative, and other pathological processes. Patients with various immune system defects require highly specialized care. The course of these diseases in patients with primary immunodeficiency (PID) can be severe and accompanied by high mortality. Such patients require inpatient treatment using various types of antimicrobial, immunosupportive, immunomodulatory, immunosuppressive therapy, and immunoprophylaxis to regulate the impaired functions of the immune system. The need for long, intensive, and systematic hospital treatment is frequent. Patients with certain PIDs are prescribed antimicrobial prophylaxis and vaccination as one of the important means of protection against infectious diseases. With the rapid development of immunology and molecular genetic research methods, the spectrum of nosological PID forms in the world is growing continuously, and we must be ready for these challenges.

At the beginning of the event, a round table was held with the participation of regional experts in pediatric and adult immunology from various parts of Ukraine, chaired by Professor Valentyna Chopyak, an immunology expert of the Ministry of Health of Ukraine, regarding the current problems faced in the field of clinical immunology in the conditions of war in Ukraine. The main prospects for using directive documents of the immunological service in the conditions of war and the provision of replacement immunotherapy, namely immunoglobulins, for patients with PID were discussed. More active cooperation with European colleagues and the creation of a national registry of immunodeficient patients with impaired antibody synthesis were noted. The areas of further activities of the immunological service during martial law were outlined. The focus was placed on protecting persons with congenital immune system disorders against infections by means of timely systematic seasonal vaccination, replacement immunoglobulin therapy, prophylactic antimicrobial therapy, and coordinated activities of specialists from various fields to provide immunological aid to patients in war conditions.

Professor Anastasiia Bondarenko and Associate Professor Khrystyna Lishchuk-Yakymovych presented to the world “novelties” of diagnosis, treatment, and peculiarities of immunoprophylaxis in patients with primary immunodeficiencies: the new 2022 IUIS (International Union of Immunologic Societies) classification, the place of inborn errors of immunity in the International Classification of Diseases, 11th Revision (ICD-11), materials of IPIC and ESID immunological congresses held in 2022. To date, more than 450 nosological forms of PID have been described differing by the nature of immune system dysfunction and the degree of immunosuppression, which predetermines the susceptibility to infections, response to immunization, and risks of possible complications.

During the school, results of Biopharma activities, a domestic producer of immunoglobulins, and plans for the future were presented in the field of domestic immunology development and diagnosis of patients with inborn errors of immunity. Initiated diagnostic projects with PID, including support for molecular genetic examinations, bear great significance in early diagnosis and ensuring high quality of life for patients, screening programs, etc. The Biopharma plant is the only plasma fractionator in Eastern Europe. What is more, it is located in Ukraine. Immunoglobulin

replacement therapy is one of the leading therapeutic agents for inborn errors of immunity. Therefore, the presence and support of a domestic manufacturer are significant in continuous therapy.

Laboratory diagnostics and the correct interpretation of immunological research results play a crucial role in making an accurate diagnosis and the future fate of patients with inborn errors of immunity. During the master class “Interpretation of Immunological Studies,” complex issues of differential diagnosis between primary and secondary immunodeficiencies against the background of severe kidney, intestinal and oncological diseases and/or immunosuppressive therapy were discussed. Analysis of the indicators of the immunological examination compared to the clinical picture, data from the anamnesis, and treatment received will help avoid erroneous interpretations. During the master class, immunologists had the opportunity to talk with a specialist who is directly involved in conducting immunological research and understands the intricacies of evaluating immunogram indicators on a flow cytometer. The tandem of a clinician and a qualified laboratory diagnostics specialist creates a successful path to a complex diagnosis of primary immunodeficiency, allowing for determining the correct treatment and ensuring a decent quality of life for patients.

Participants were selected competitively based on the most interesting clinical cases of PID. The most interesting and problematic cases were included in the event program. The chosen format of the case marathon involved presentations by participants and their active discussion with the audience, shaping conclusions, examination, and treatment algorithms in groups, which allowed all participants to be actively engaged in the process. Moderators (Professor Valentyna Chopyak, Professor Anastasiia Bondarenko, Associate Professor Khrystyna Lishchuk-Yakymovych, and Associate Professor Yuriy Stepanovskyi) participated in discussing clinical cases with simultaneous training and practice for immunologists in the field of modern algorithms for the diagnosis and treatment of primary immunodeficiencies – inborn errors of immunity using Standards for the Diagnosis and Treatment of Primary Immunodeficiencies, which were previously approved by the Ministry of Health of Ukraine (Order No. 2952 dated December 31, 2021).



Figure 1. Participants of the Winter Specialized Immunology School, “Inborn Errors of Immunity.”

During the presentation of clinical cases, problems were discussed. The following were particularly interesting: various clinical manifestations of general variable immunodeficiency (infectious, autoimmune, oncological, hematological), the differential diagnosis between primary and secondary immunodeficiency of antibody formation, a long path to the diagnosis in hypergammaglobulinemia syndrome E. Collaboration with related specialists, particularly hematologists, rheumatologists, and pulmonologists, is essential for practice due to various manifestations of primary immunodeficiencies, including lymphomas, juvenile idiopathic arthritis, pulmonary fibrosis, etc. Autoinflammatory syndromes constitute a relatively new group of inborn errors of immunity, often presenting significant diagnostic challenges (hypergammaglobulinemia D syndrome, Schnitzler syndrome, familial Mediterranean fever, macrophage activation syndrome). Current problems include interpretations of molecular genetic examination results for variants of uncertain significance and/or incomplete penetrance, management of the infectious syndrome in chronic granulomatous disease, the course and consequences of COVID-19 in patients with inborn errors of immunity, timely prenatal diagnosis and management of severe combined immunodeficiency, and the selection of the tactics for managing patients with diagnoses potentially requiring bone marrow transplantation.

During school, immunologists practiced the following in detail:

- the wording of the diagnosis of primary immunodeficiency according to modern classifications (ICD-10, IUIS 2022, ICD-11)
- warning signs – criteria of a primary immunodeficiency for doctors of various specialties because the clinical picture of PID can be diverse
- interpretation of genetic examination results
- principles for prescribing replacement therapy with immunoglobulins (regime, duration, monitoring of efficiency)
- selecting target therapy
- recommendations for bone marrow transplantation in primary immunodeficiencies
- the patient's route to bone marrow transplantation.



Figure 2. The first day of the Specialized School



Figure 3. The second day of the Specialized Immunology School

The Winter Specialized Immunology School, “Inborn Errors of Immunity,” has become a sip of live communication in a relatively safe part of Ukraine – Zaparattia Region. Brilliant clinical cases that this event composed of, relaxed communication, and being on the same wavelength as part of a large joint conference enabled full immersion in the professional atmosphere. It brought together adult and pediatric immunology specialists in their exchange of experience, expansion of scientific and practical knowledge, and improvement of practical work in providing immunological assistance to patients with PID, especially in times of war.