Editors: Melinda Erdős and László Maródi

Co-Editors:
Mihaela Bataneant, Mikhail Belevtsev, Ewa Bernatowska, Mikhail Bolkov, Peter Ciznar, Marina Guseva, Katalin Haraszti, Sevan Iritsyan, Natalja Kurjane, Noémi Miltner, Kristina Mironska, Marianna Murdjeva, Gulnara Nasrullayeva, Elissaveta Naumova, Bubusaira Orozbekova, Malgorzata Pac, Karaman Pagava, Lidija Pocek, Ismail Reisli, Nima Rezaei, Svetlana Sciuca, Margit Serban, Svetlana Sharapova, Irina Tuzankina, and Alla Volokha

June 2021
Primary immunodeficiency was a well-established clinical and research discipline when the European Federation of Immunological Societies (EFIS) brought to life the Day of Immunology (DoI) on April 29, 2005. Soon after this, the World Primary Immunodeficiency Week (WPIW) was established and dedicated to increasing global awareness on primary immunodeficiency disorders (PIDs) each year, on April 22-29, 2011. It is therefore reasonable to say that April is a “month of immunologists” worldwide. The ambition of the PID professional community to dedicate a week instead of a day could perhaps be attributed to the underdevelopment of PID diagnostics and patient care compared to other major fields of clinical immunology, like allergy and autoimmunity. These annual events are necessarily supplemented with even more challenging educational programs like the J Project. Not surprisingly, April has always seen a higher number of JP Meetings attributable to the DoI and the WPIW. Still, the record number of 16 meetings this year is outstanding and unique (Table 1). Although it was spontaneous, it would be hard not to attribute it to the 10th anniversary of the WPIW. This has been a silent celebration by the J Project community showing sensitively how traditions may change our professional life and contribute to progress of our wonderful discipline. Table 1 shows the list of 16 JP meetings and our “April heroes” organizing these events and celebrating the 10th WPIW in a fully professional way, on behalf of the whole JP community. We have also reached the 300th JP meeting in this grandiose month and welcome Montenegro, the youngest member country. Figure 1 shows the number of JP meetings per year since 2004 and a peak number in 2020. The consistent increase of the meeting number since 2016 is not proportional with the number of JP member countries. Rather, it represents the strong devotion of most leaders of the J Project in the 32 member countries to provide a better care of PID patients in Eurasia. We are grateful to ESID, JMF, FCI, and a few pharmaceutical companies for their generous support of the J Project.

László Maródi
### Table 1. Sixteen J Project meetings in April, 2021

<table>
<thead>
<tr>
<th>City, Country</th>
<th>Organizer(s)</th>
<th>Date</th>
<th>Subject</th>
</tr>
</thead>
<tbody>
<tr>
<td>Konya, Turkey</td>
<td>Ismail Reisli</td>
<td>Apr 6</td>
<td>PID and infections</td>
</tr>
<tr>
<td>Warsaw, Poland</td>
<td>M Pac, E Bernatowska</td>
<td>Apr 10</td>
<td>Immunization in PID</td>
</tr>
<tr>
<td>Debrecen, Hungary</td>
<td>N Miltner, L Maródi</td>
<td>Apr 17</td>
<td>PID for students</td>
</tr>
<tr>
<td>Kiyv, Ukraine</td>
<td>Alla Volokha</td>
<td>Apr 22</td>
<td>PID and rheumatology</td>
</tr>
<tr>
<td>Timisoara, Romania</td>
<td>Mihaela Bataneant</td>
<td>Apr 22</td>
<td>PID – general features</td>
</tr>
<tr>
<td>Plovdiv-Bulgaria</td>
<td>M Murdjeva, E Naumova</td>
<td>Apr 22</td>
<td>PID and COVID-19</td>
</tr>
<tr>
<td>Budapest, Hungary</td>
<td>K Haraszti, L Maródi</td>
<td>Apr 23</td>
<td>PID for teachers</td>
</tr>
<tr>
<td>Riga, Latvia</td>
<td>Natalja Kurjane</td>
<td>April 24</td>
<td>PID care in Latvia</td>
</tr>
<tr>
<td>Tehran, Iran</td>
<td>Nima Rezaei</td>
<td>April 26</td>
<td>PID and COVID-19</td>
</tr>
<tr>
<td>Baku, Azerbaijan</td>
<td>Gulnara Nasrullayeva</td>
<td>April 28</td>
<td>PID and COVID-19</td>
</tr>
<tr>
<td>Bratislava, Slovakia</td>
<td>Peter Ciznar</td>
<td>April 28</td>
<td>PID diagnostics</td>
</tr>
<tr>
<td>St. Petersburg, Russia</td>
<td>Marina Guseva</td>
<td>April 29</td>
<td>PID and COVID-19</td>
</tr>
<tr>
<td>Yerevan, Armenia</td>
<td>Sevan Iritsyan</td>
<td>April 29</td>
<td>PID and COVID-19</td>
</tr>
<tr>
<td>Podgorica, Montenegro</td>
<td>Lidija Pocek (1st JP)</td>
<td>April 29</td>
<td>PID – joining the JP</td>
</tr>
<tr>
<td>Minsk, Belarus</td>
<td>Mikhail Belevtsev</td>
<td>April 29</td>
<td>6th PID - JP Meeting</td>
</tr>
<tr>
<td>Osh city, Kyrgyzstan</td>
<td>B Orozbekova, I Tuzankina</td>
<td>April 29</td>
<td>PID – general features</td>
</tr>
</tbody>
</table>

**Figure 1**

- 300 J Project meetings
<table>
<thead>
<tr>
<th>CITY</th>
<th>COUNTRY</th>
<th>ORGANIZER(S)</th>
<th>DATE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ekaterinburg</td>
<td>Russia</td>
<td>Irina Tuzankina</td>
<td>February 11-12</td>
</tr>
<tr>
<td>Riga</td>
<td>Latvia</td>
<td>Natalja Kurjane</td>
<td>February 19</td>
</tr>
<tr>
<td>Konya</td>
<td>Turkey</td>
<td>Ismail Reisli</td>
<td>February 24</td>
</tr>
<tr>
<td>Konya</td>
<td>Turkey</td>
<td>Ismail Reisli</td>
<td>April 6</td>
</tr>
<tr>
<td>Warsaw</td>
<td>Poland</td>
<td>Ewa Bernatowska</td>
<td>April 10</td>
</tr>
<tr>
<td>Debrecen</td>
<td>Hungary</td>
<td>Noémi Miltner &amp; László Maródi</td>
<td>April 17</td>
</tr>
<tr>
<td>Kiev</td>
<td>Ukraine</td>
<td>Alla Volokha</td>
<td>April 22-23</td>
</tr>
<tr>
<td>Timisoara</td>
<td>Romania</td>
<td>Mihaela Bataneant</td>
<td>April 22-23</td>
</tr>
<tr>
<td>Budapest</td>
<td>Hungary</td>
<td>Katalin Haraszti</td>
<td>April 23</td>
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<tr>
<td></td>
<td></td>
<td>Honváriné &amp; László Maródi</td>
<td></td>
</tr>
<tr>
<td>Plovdiv-Sofia</td>
<td>Bulgaria</td>
<td>Marianna Murdjeva &amp; ElissavetaNaumova</td>
<td>April 23</td>
</tr>
<tr>
<td>Riga</td>
<td>Latvia</td>
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<tr>
<td>Podgorica</td>
<td>Montenegro</td>
<td>Lidiya Poczek</td>
<td>April 29</td>
</tr>
<tr>
<td>Minsk</td>
<td>Belarus</td>
<td>Mikhail Belevtsev</td>
<td>April 29-30</td>
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<tr>
<td>Osh</td>
<td>Kyrgyzstan</td>
<td>Bubusayra Orozbekova</td>
<td>April 29-30</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Irina Tuzankina</td>
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</tr>
<tr>
<td>Tbilisi</td>
<td>Georgia</td>
<td>Karaman Pagava</td>
<td>June 5</td>
</tr>
<tr>
<td>Jalal-Abad</td>
<td>Kyrgyzstan</td>
<td>Bubusayra Orozbekova</td>
<td>June 17-19</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Irina Tuzankina</td>
<td></td>
</tr>
</tbody>
</table>
EKATERINBURG, RUSSIA          FEBRUARY 11-12

Host: Irina Tuzankina
Venue: Webinar
Topic: International forum «Immunology 2021», dedicated to the 35th anniversary of Service of Clinical Immunology in the Ural

PROGRAM

11 February
I Plenary Session - Conference opening

Chereshnev Valery Aleksandrovich, Ekaterinburg
Chadova Elena Anatolyevna, Ekaterinburg
Olga Petrovna Kovtun, Ekaterinburg
Solovieva Olga Eduardovna, Ekaterinburg
Kiselev Arkady Yuryevich, Ekaterinburg
Video presentation with participation of Timur Bekmambetov, founder of the Sunflower Charitable Foundation for Children and Adults with Immunity Disorders, Moscow
László Maródi, Budapest, Hungary
Jean-Laurent Casanova, New York, USA; Paris, France

Martine Pergent, Paris, France
Aripova Tamara Uktamovna, Tashkent, Uzbekistan.
Nifadiev Vladimir Ivanovich, Bishkek, Kyrgyzstan
Orozbekova Bubusaira Tolobaevna, Bishkek, Kyrgyzstan.
Uzakbaev Kamchybek
Askarbekovich, Bishkek, Kyrgyzstan
Snemshchikova Irina Anatolyevna, Orel
Vakhlova Irina Veniaminovna, Ekaterinburg
Averyanov Oleg Yurievich, Ekaterinburg
Presentations

Problems of medicine of the XXI Century
Chereshnev Valery Aleksandrovich

Neonatal window as a basis for the development of the immune response
Rumyantsev Alexander Grigorievich

Inborn immunity errors in children of the Sverdlovsk Region: Service history
Tuzankina Irina Aleksandrovna

Omix technologies as the basis for the diagnosis of primary immunodeficiencies
Belevtsev Mikhail Vladimirovich

Hematological manifestations of PID in children
Sharapova Svetlana

PID in Konya, Turkey
Ismail Reisli

The role of inflammation markers in the development and progression of metabolic syndrome
Aripova Tamara Uktamovna

J Project in Uzbekistan
Nabieva Umida Pulatjanzona

Clinical and epidemiological features of primary immunodeficiencies in the Republic of Kazakhstan
Kovzel Elena Fedorovna

COVID-19 Immunopathogenesis
Simbirtsev Andrey Semenovich

Results of seroepidemiological studies of coronavirus infection
COVID-19 in the Republic of Kyrgyzstan
Nurmatov Zurdin Sharipovich, T.E. Kuchuk, Orozbekova Bubusaira Tolobaevna
The level of some cytokines at different periods of gestation in women with COVID-19
Musakhodjaeva Diloram Abdullaevna, Jabbarova Yulduz Kasymovna, Yeshimbetova Gulsara Zakirovna, Dzhumaev Kamil Chorievich, Ismoilova Shoira Tolkunovna, Yuldasheva Gulchekhra Rakhimovna

Principles of immunomodulatory therapy for systemic connective tissue diseases and with COVID-19 in children
Uzakbaev Kamchybek Askarbekovich

Immunorehabilitation of patients during a pandemic
Fedoskova Tatyana Germanovna

Current problems of immunopharmacology
Kozlov Ivan Genrikhovich

Childhood immunology in a pandemic
Prodeus Andrey Petrovich

How to define immunodeficiency
Korsunsky Ilya Anatolyevich

Genotype-phenotype correlations in primary immunodeficiencies
Kondratenko Irina Vadimovna

Clinical Immunology in Rheumatology
Popov Artem Anatolyevich

Immunotropic drugs in real pediatric practice
Tsarkova Sofya Anatolyevich

Immunoregulation of reparative processes in the oral mucosa
Sarkisyan Narine Grishaevna

Immunological studies in determining the activity of pulmonary tuberculum
Berdyugina Olga Viktorovna

Possibilities of medical preparations
Tuzankina Irina Aleksandrovna
II Plenary Session
Moderators: Valery Cherepnev, Elena A. Chadova, Oleg Yuryevich Averyanov, Irina A. Tuzankina

Provision of specialized immunological medical care in the Sverdlovsk region - the value of the register of PIDs for the diagnostic and treatment process
Vlasova Elena Viktorovna

Organization of specialized medical care for patients with hereditary angioedema in the Sverdlovsk region
Beltyukov Evgeniy Kronidovich

Laboratory methods for the diagnosis of immune-dependent pathology in the Sverdlovsk region
Pashnina Irina Aleksandrovna

Laboratory methods for the diagnosis of autoimmune pathology
Krivolapova Irina Mikhailovna

Immunodeficiencies in Slavic Childre
Sharapova Svetlana

Present and future of molecular genetic diagnostics of hereditary immunopathology in the Sverdlovsk region
Deryabina Svetlana Stepanovna

Congenital errors of immunity without signs of immunodeficiency
Bolkov Mikhail Artemovich

Immunological and genetic aspects of changes in the mineral composition of bones
Sannikova Oksana Yurievna

Welcome speech
Shcherbina Anna Yurievna

PID Epidemiology in the Russian Federation
Mukhina Anna Aleksandrovna

PID Diagnostic Algorithm
Natalya Borisovna Kuzmenko

SCID - Diagnostic News
Rodina Yulia Aleksandrovna

Variety of autoimmune diseases
Kozlova Anna Leonidovna

Discussion
Assessment of indicators of morphogenetic proteins of the WNT signaling pathway in patients with coronary heart disease
Plotnikova Maria Olegovna,
Snemshchikova Irina Anatolyevna,
Afonina Irina Aleksandrovna

When Nutrition Becomes Treatment in the Practice of a Pediatrician
Ipatova Maria Georgievna

The effectiveness of sublingual immunotherapy with birch pollen allergen for cross-reactive food allergy in children
Lepeshkova Tatyana Sergeevna

Molecular basis of allergy pathology in children
Bass Evgeniya Aronovna

Molecular genetic and phenotypic manifestations of PIDs with congenital heart defects
Cheremokhin Dmitry Andreevich

Dental pathology in patients with primary immunodeficiency
Dolgikh Maria Andreevna

Bioinformatics and biotechnology - two sides of one analysis
Shinwari Khyber
INTERREGIONAL SCIENTIFIC AND PRACTICAL CONFERENCE
"PATIENT-ORIENTED APPROACH IN MEDICINE"

Moderators: Chadova Elena Anatolyevna, Voronina Lyudmila Ivanovna, Tuzankina Irina Aleksandrovn

Video presentation of the Belarusian Society of Patients with PID "Let's Save Immunity"

Quality of life of patients diagnosed with PID
Martine Pergent (IPOPI)

Longitudinal study of the quality of life of patients with PID
Alexander Runov

Questions of administrative interaction of patients with PID, with the management bodies of medical organizations
Voronina Lyudmila Ivanovna

Legal issues of patients with congenital immunity errors and patient communities
Andrey Vadimovich Nechkin

Delegates from Bishkek, Kyrgyzstan
InternationaL event of patient organizations

Moderators: Chadova Elena Anatolyevna, Tuzankina Irina Aleksandrovna, Berseneva Natalia Nikolaevna

Experience of organizing patients with PID in Russia and issues of interaction of patient organizations with administrative bodies and society
Berseneva Natalya Nikolaevna

The Role of Patient Organizations
Zhulev Yuri Aleksandrovich

The scope of the practice of applying the legislation on drug provision and the organization of treatment for patients with PID

Emelin Anton Evgenievich

Correct preparation of documents for the ITU
Ivancha Elena Viktorovna

Experience of the patient community in Belarus
Dirko Iya Leontievna

Providing patients with primary immunodeficiency with medical nutrition prescribed for health reasons
Berseneva Natalya Nikolaevna

Speeches by representatives of various patient communities.

Closing

Summary

On February 11 and 12, 2021, the 1st International Congress "Doctor-Patient: Immunology 2021" was held under the initiative of the Institute of Immunology and Physiology of the Ural Branch of the Russian Academy of Sciences (Ekaterinburg) and the Ministry of Health of the Sverdlovsk Region. The purposes of this congress were to call the attention of professional societies and organizations to the problems of sick people, to form an information space in the field of medicine, to promote the development of the level of medical knowledge among both medical specialists and patients, to develop and improve the interaction between medical organizations, government entities, and public organizations, to improve the quality of life of patients, to discuss the specifics of the work of medical organizations during the COVID 19 pandemic, to create a platform for discussion and search for solutions on the interaction of the medical community, patient and other public organizations with each other and with authorities.

The organizers of the Congress were as follows:

Information support of the Congress was: websites of the Russian Scientific Society of Immunologists https://rnoi.ru, the Institute of Immunology and Physiology of the Ural Branch of the Russian Academy of Sciences, the Ministry of Health of the Sverdlovsk Region, the Regional Children's Clinical Hospital in Yekaterinburg. The technical provider of the event was Kamkor-Integration Group of Companies.

1328 people from 12 countries registered in the International Congress. Among the participating countries were: Belarus, Hungary, Kazakhstan, Uzbekistan, Kyrgyzstan, Tajikistan, Latvia, Ukraine, France, Netherlands, Sweden, Turkey, and the USA. Congress participants represented 128 cities and regions of the Russian Federation, including Moscow, St. Petersburg, Novosibirsk, Ekaterinburg and the Sverdlovsk region, Tomsk, Omsk, Krasnoyarsk, Kemerovo, Orenburg, Vladivostok, Kazan, Ivanovo, Tyumen, Khabarovsk, Nizhny Novgorod, Ufa, Perm, Chelyabinsk and Chelyabinsk region, Rostov-on-Don, Kaliningrad, Irkutsk, Saratov, Angarsk, Berdsk, Novokuznetsk, Izhevsk, Grozny, Makhachkala.

Medical workers - representatives of practical health care who took part in the Congress both as speakers and and listeners (more than 1220 people), received certificates of professional development. Representatives of public organizations became active participants in the Congress, sharing their experience of their work. Among them are representatives of patient organizations: the International Organization of Patients with Primary
Immunodeficiency (IPOPI), the Patient Organization of Patients with Primary Immunodeficiency in the Sverdlovsk Region ("Immune Academy Valery Chereshnev Umida Nabieva, Tashkent, Uzbekistan) the All-Russian Union of Public Associations of Patients, the All-Russian Organization of Patients with Primary Immunodeficiency (OPPID), the All-Russian Society hemophilia, the Belarusian Society of Patients with Primary Immunodeficiencies, "Let's Save Immunity", charitable foundations (the Sunflower Charitable Foundation for Children and Adults with Immunity disorders), a video greeting was presented with the participation of Timur Bekmambetov, co-founder of this Charitable Foundation for helping children and adults with Immunity disorders "Sunflower".

The Congress was attended by Russian leading experts in the field of medicine from Yekaterinburg, Moscow, St. Petersburg, Orel and other cities of the Russian Federation, as well as foreign experts from Belarus (Minsk, Vitebsk), Uzbekistan (Tashkent, Samarkand, Bukhara), Kyrgyzstan
(Bishkek, Osh), Kazakhstan (Nur-Sultan, Astana) France (Paris), Hungary-USA (Budapest-New York), Turkey (Konya), Kenya, Ukraine (Odessa, Kiev, Ternopil, Lugansk), Tajikistan, the Netherlands, Sweden, Iran.

Foreign and leading Russian scientists, doctors greeted the participants of the congress online, among them:

- **Jean-Laurent Casanova**, Head of the St. Giles Laboratory of Genetics of Human Infectious Diseases, Rockefeller University, NY

- **László Maródi**, Head of the J Project, Semmelweis Univ, Budapest

- **Martine Pergent**, President of IPOPI, Paris, France
Within the framework of the International Congress, 43 reports of leading Russian and foreign scientists were presented. Within the framework of the plenary sessions, reports were made on the problems of medicine in the XXI century, the current level of knowledge on the problems of immunology - the regulator of life, which forms the basis for the formation of various human pathologies, the history of the development of clinical immunology in the world, in Russia and in the Sverdlovsk region, the prospects for the development of science and practice, the role of immune mechanisms in ensuring health and in the formation of pathology, technologies of diagnosis, treatment and rehabilitation, as well as the possibility of immunotropic interventions in real pediatric practice and other fields of medicine are discussed. Reports were made on laboratory methods for the diagnosis of immune-dependent and autoimmune pathology, the organization of specialized medical care for patients with congenital errors of immunity and other immune-dependent pathologies in the Sverdlovsk region, on the present and future of molecular genetic diagnostics of hereditary immunopathology, congenital errors of immunity, algorithms for diagnosing diseases associated with congenital errors of immunity, etc. The issues of pathogenesis, diagnosis, treatment and rehabilitation of the newly emerged infection COVID-19, complex diagnostic issues and vaccination opportunities were actively discussed, experience of work in various regions of the world was presented. It was decided to make this congress regular, declare Ekaterinburg as the venue for the next forums, develop a logo for the event, and consolidate the name “Doctor-patient-society: Immunity of the XXI century”.

Irina Tuzankina
Mikhail Bolkov
his J Project meeting was focused on various diagnostic aspects and approved therapies of **Cytopenia in clinical practice**, and it was organized in Riga, Latvia. The meeting was online as a webinar due to limitations by the coronavirus pandemic. More than 400 doctors of different clinical specialties, mostly pediatricians, internists, family doctors, and neurologists were registered. The first lecturer was Svetlana Sharapova, an active member of J Project from Belarus and an outstanding expert in the field, with a comprehensive lecture about **Cytopenia as a clinical sign of primary immunodeficiencies**. The second lecturer was Natalja Kurjane, from
Paula Stradins Clinical University Hospital in Riga, Latvia. Her talk was about *Neutropenia in children and adults*. She focused mostly primary genetic causes of neutropenia and discussed acquired neutropenia from the differential diagnostic point of view. Finally, Zanna Kovaleva, pediatric hematologist from the Children’s Clinical University Hospital in Riga Latvia, presented an instructive clinical case of a 3 years old patient, who had several autoimmune disorders, and was transplanted with the diagnosis of “undefined inborn error of immunity”. We appreciate the active participation of the speakers and the ambitious discussion by many clinicians. We believe that this meeting further contributed to the increased awareness on primary immunodeficiencies in Latvia and indicated the ongoing participation of our country in the J Project.

The next J Project meeting in Riga is scheduled for 24th of April, 2021. The subject of the meeting is proposed to be: **Primary immunodeficiencies in clinical practice**. We plan to invite local speakers, mostly clinicians to present instructive clinical cases of various primary immunodeficiencies. The organizers are devoted to invite speakers from the other Baltic countries, as well as lecturers from other J Project countries. We expect speakers also from the leadership of the European Society for Immunodeficiencies (ESID).

*Zanna Kovaleva, Natalja Kurjane and Svetlana Sharapova*
KONYA, TURKEY

Host: Ismail Reisli
Venue: Webinar
Topic: The approach to patients with recurrent infections and suspected PIDs

PROGRAM

The RUMI JP Meeting

Sık Enfeksiyonlu Hastaya Yaklaşım

The approach to a patient with recurrent infections

Düzenleme Kurulu (Organization)
Prof. Ismail Reisli

Communication
Prof. Ismail Reisli, MD
Necmettin Erbakan University
Pediatric Immunology and Allergy
42080, KONYA-TURKEY
ireisli@hotmail.com
Tel: +90 352 225 66 50

Öğretim ve Araştırma Merkezi,
SIEM, KONYA (Konya University)

Chair: Ilhan Tezcan, Ismail Reisli

19:00-19:15 Açılış / Opening
19:15-19:30 Sık Enfeksiyonlu Hastaya Yaklaşım
Approach to a patient with recurrent infections
Dr. Ismail Reisli

19:30-19:45 Primer İmmün Yetmezlik
Primary Immunodeficiency
Dr. Sevgi Keleş

19:45-20:00 Tedavi Yaklaşımı
Therapy
Dr. Şükrü Nail Güner

20:00-20:40 Olgularla Primer İmmün Yetmezlik
Case Presentations with PID
Çocuk hastalar / in pediatric patients
Dr. Hasibe Artaç

Yetişkin hastalar / in adult patients
Dr. Şevket Arslan

20:40-21:00 Tartışma / Discussion

Katılım Ücretsizdir
This online Conference is free of charge
Prof Keles (below) was talking about the pathophysiology of the Immune System
This online J Project Meeting was organized together with the Clinical Immunology Society and RUMI Pediatric Society in Konya to increase awareness on primary immunodeficiency (PID) diseases among pediatricians, internalists and family physicians. The main topic was ‘The approach to a patient with recurrent infections’. This Meeting was held on February 24, 2021 and it was the 285th JP Meeting since the beginning in 2004. The meeting was co-organized with the Necmettin Erbakan and Selçuk Universities and primarily focused on diagnosis and treatment of challenging PID cases diagnosed by pediatricians and internalists.

The moderators of RUMI J Project Meeting were Prof Ilhan Tezcan from Hacettepe University and Prof Ismail Reisli from Necmettin Erbakan University. Prof Tezcan is also the President of Clinical Immunology Society and he focused his talk on the importance of early diagnosis and treatment for the patients with PID and thanked to Prof Reisli for his valuable effort to organize a serious of J Project Meetings as well as three JP Congresses in Turkey. After Prof Reisli gave his lecture about the approach to a patient with recurrent infections, Prof Keles and Assoc. Prof Guner talked about current IUIS classification of primary immunodeficiencies and their treatment modalities. Prof Artac and Assoc Prof Arslan presented their pediatric and adult cases. These cases were mainly about Bruton’s disease, severe combined immunodeficiency and common variable immunodeficiency.

There were 175 participants attended this online meeting. The participants discussed the importance of early diagnosis and early treatment of the patients with PID. I would like to thank all participants for their participation to this J Project Meeting, and also thank CSL Behring Company for their sponsorship to make this organization possible.

Ismail Reisli
Leader of the J Daughter Anatolia Project
Konya-Turkey
Host: Ismail Reisli
Venue: Webinar
Topic: Approach to a patient with recurrent infections

PROGRAM

GÜNEYDOĞU ANADOLU J PROJECT TOPLANTISI
Southeast Anatolia J Project Meeting
6 Nisan 2021 Salı / 6th April 2021 Tuesday

Sık enfeksiyon geçiren hastaya yaklaşım
Approach to a patient with recurrent infection

OTURUM BAŞKANI / CHAIR
Dr. Kazım Doğan Eroğulları
Gastgebärende Ältere Heilkundlich Demiçile Reisli (GADH)
TTB Merkez Korusu Üyesi

19:30 - 19:50
Çocuklarda Primer İmmün Yetmezlikler
Primary immunodeficiency in Children
Dr. Sevgi Koçak

19:00 - 19:15
Sık Enfeksiyon Geçiren Hastaya Yaklaşım
Approach to a patient with recurrent infections
Dr. Ismail Reisli

19:15 - 19:30
İmmün Sistemi ve Primer İmmün Yetmezlikler
Immun system and Primary Immunodeficiency
Dr. Ilhan Tuzcan

19:50 - 20:05
Yetkin Çanıkta Primer İmmün Yetmezlikler
Primary Immunodeficiency in Adults
Dr. Zafer Çağlaçaner

20:05 - 20:15
Primär İmmün Yetmezliklerinin Tedavisi
Treatment of Primary Immunodeficiency
Dr. Süleyman Nall Günsen

20:15 - 20:30 TARTIŞMA / Discussion

*Bağışıklık hizmeti hastaların sağlık durumlarını iyileştirmek için anormallikleri ifade eden proteinlerin analizini, hastaların hastalık riskini belirlemek, hastaların yaşam kalitesini artırmak, tedavi maliyetlerini de düşüktür. Bu amaçla, Avrupa İmmün Yetmezlikler Derneği (ESID), Ortadoğu ve Doğu Avrupa ülkelerinde Primer İmmün Yetmezlik Hastalarına Farkındalık Projesi (J-Project) adı altında bir proje başlatmıştır. Bu proje ile 2004 yılından itibaren, önceleri Avrupa ülkelerinde eğitim toplantıları düzenlemeye ve 2010 yılı itibariyle tüm Avrupa ülkelerinde eğitim toplantıları düzenleme, 2018 yılında primer hekimler arasında immün yetmezlik bilinmesi ve farkındalık düzeyini artırmak, özellikle bu hastaların erken tanısı ve tedavisi gerçekleştirilmektr.
The speakers of Southeast Anatolia J Project Meeting
This online J Project Meeting (No 286) was organized together with the Clinical Immunology Society and Gaziantep Society of Family Physician to increase awareness on primary immunodeficiency (PID) diseases among pediatricians and family physicians. The main topic was ‘Approach to a patient with recurrent infections’. This Meeting was held on April 6, 2021. The meeting was co-organized with the Necmettin Erbakan and primarily focused on diagnosis and treatment of PID diagnosed by pediatricians and family physicians.

The moderator of Southeast Anatolia J Project Meeting was Kazım Doğan Eroğullari, MD, from Gaziantep Society of Family Physicians. The first speaker was Prof Reisli and gave his lecture about the approach to a patient with recurrent infections. The second speaker was Prof İlhan Tezcan who is
the President of Clinical Immunology Society. Prof Tezcan focused on immune system components and IUIS classification of PID. Prof Sevgi Keles and Prof Zafer Caliskaner talked about the PID in children and adulthood and the importance of early diagnosis and treatment for the patients with PID, respectively. The last speaker, Assoc. Prof Sukru Nail Guner talked about the current treatment modalities of PID.

There were 304 participants attended this online meeting. The participants discussed the importance of early diagnosis and early treatment of the patients with PID. I would like to thank all participants for their participation to this J Project Meeting, and also thank Pharmada Company for their sponsorship to make this organization possible.

Ismail Reisli
Leader of the J Daughter Anatolia Project
Konya-Turkey
Waraw, Poland

April 10

Host: Ewa Bernatowska
Venue: The Department of Immunology, Children’s Memorial Health Institute, Warsaw
Topic: Vaccination in primary immunodeficiency and healthy children in the era of COVID-19

XII National Immunization Day
http://www.ogolnopolskidzienszczepien.pl/
Vaccines have brought us closer, and will bring us closer again…
World Immunization Week – 2021

The conference was organized under the patronage of Polish Paediatric Society, Polish Society for Experimental and Clinical Immunology, Polish Academii of Science, J Project and JMF Foundation

PROGRAM

Emilia Skirmuntt, Oxford

Preventing the spread of SARS-CoV-2 infection - own experience of the Children’s Memorial Health Institute,
Marek Migdał, Warsaw

Clinical pictures of COVID-19 in primary immunodeficiency patients.
Małgorzata Pac, Warsaw

Recommendations on management of vaccines for risk groups.
Ewa Bernatowska, Warsaw

Universal vaccinations against rotavirus infection national consultant in the field of paediatrics.
Teresa Jackowska, Warsaw

Risk of pertussis infection - the need for prophylaxis at every stage of life
Ernest Kuchar, Warsaw
Annual conference was organized as one of the events, the part of the National Immunization Day [http://www.ogolnopolskidzienszczepien.pl/]. The main topic this year was the vaccination in primary immunodeficiency and healthy children in the era of COVID-19. During the second year of the pandemic of SARS-CoV-2, herd immunity as a form of indirect protection seems to be still unsatisfied. Emilia Skirmuntt from Department of Zoology, Oxford University gave a first lecture on SARS-CoV-2 pandemic and vaccination against COVID-19: prognosis and limitations. Doctor Emilia Skirmuntt is an expert on molecular epidemiology and evolutionary genetics of infectious diseases. She presented the current epidemiology of SARS-CoV-2, the new variants of coronavirus, progress in the development of the new generation of protein vaccine, which gave hope together with currently using vaccines for achieving herd immunity. The second lecture presented by Marek Migdał general director of the Children’s Memorial Health Institute. He summarised our own experience of the Children’s Memorial Health Institute, in preventing the spread of SARS-CoV-2 infection. The medical staff, 1 800 physicians, nurses, laboratory workers and administration were vaccinated with 2 doses of Comirnaty. One percent of them got infected with SARS-CoV-2. In the course of COVID 19, no one of 212 hospitalised patients died. Clinical pictures of COVID-19 in primary immunodeficiency patients, treated in Department of Immunology, the Children’s Memorial Health Institute was presented by Małgorzata Pac. The clinical course of disease in 19 children with SARS-CoV-2 infection, including 8 patients with antibody deficiency, 5 with CID, 5 with autoinflammatory diseases and 1 patient with neutropenia. All of them presented the mild course diseases, in more than have of them, no clinical symptoms was noted. Ewa Bernatowska presented the guideline created to provide primary care and specialty clinicians with evidence-based guidelines for active immunization of patients with altered immunocompetence and their household contacts in order to safely prevent vaccine-preventable infections. Follow by guidance on routine immunization services during the COVID-19 pandemic in the WHO European Region of 20 March 2020 underlines, universal vaccinations against rotavirus infection was presented by Teresa Jackowska - national consultant in the field of paediatrics. President of Polish Vaccinology Society Ernest Kuchar presented the risk of pertussis infection, not only in children’s population, but also in adults. He postulated the need for prophylaxis at every stage of life.

Ewa Bernatowska
Webinar chaired by Ewa Bernatowska and Małgorzata Pac, Children’s Memorial Health Institute, Warsaw

Emilia Skirmuntt, Department of Zoology, University of Oxford

Marek Migdał, director of the Children’s Memorial Health Institute, Warsaw
Debrecen, Hungary

Host: Noémi Miltner & László Maródi
Venue: Webinar
Topic: Basic Immunology and PID/HIV-1 - HIV-2 dual infection lecture for the high school students across Hungary

Program

Noémi Miltner (Debrecen)
Coordinator of J Project
Introduction of J Project

István Máté Sütő (Debrecen)
Basic immunology
Primary immunodeficiencies
Anti-viral immune responses
10 warning signals of PID

Zsófia Szojka (Lund, Sweden)
Secondary immunodeficiencies
HIV-1 and HIV-2 infections
AIDS

Beáta Soltész (Debrecen)
Genetical background of PIDs
Results of Hungarian PID research
Hungarian PID panels

Péter Hajdú (Debrecen)
Nutritionist Relationship between
PID and healthy intestinal flora

Noémi Miltner (Debrecen)
Summary and future plans
On 17 April 2021, the J Project Hungary event was held online to celebrate World PID Week and World Immunology Day. This year, the target audience was secondary school students and their teachers, with the aim of teaching basic immunology and primary immunodeficiencies (PID).

This year, due to the COVID pandemic, the event was again held online, opened by Noémi Miltner, Assistant Research Fellow at the Institute of Biochemistry and Molecular Biology, Faculty of Medicine, University of Debrecen, who introduced the J Project to the audience.

István Máté Sütő, PhD candidate at the Institute of Immunology and the Doctoral School of Molecular Cell and Immunobiology, University of Debrecen, continued with his presentation entitled: Immunology primer. We were given an insight into the wonderful world of immunology, then we learned more about PID, the anti-viral immune response and we also introduced the students and their teachers to the 10 warning signs of PID.

Then, Zsófia Ilona Szojka, postdoctoral fellow at Institute of Medical Microbiology, Lund University (Sweden) continued with an interactive presentation on secondary immunodeficiencies, HIV-1 and HIV-2 infections, and AIDS.

Beáta Soltész, postdoctoral fellow at Department of Human Genetics, University of Debrecen, followed with a presentation on the genetic background of PID. We also gained insights into basic molecular biology methods.

Finally, the conference was closed by Péter Hajdú nutrition specialist and PhD student at the School of Nutrition and Food Science, University of Debrecen, who talked about the relationship between PID and healthy gut flora.

The conference was attended by many teachers (e.g. biology-chemistry), high school students from both county and rural schools, PID patients and other people interested in the topic.

Noémi Miltner
KIEV, UKRAINE  APRIL 22-23

Host: Alla Volokha
Venue: Park Hotel Favor
Topic: Rheumatic diseases in Primary Immunodeficiencies

PROGRAM

April 22

Moderator: Alla Volokha
Opening of the event. Greetings.

Autoimmunity and immunodeficiency
Alla Volokha

PID Registry: PID with immune dysregulation in Ukrainian PID registry
Anastasia Bondarenko

The implementation of on-line PID Registry
Valentyna Chopiak

The update from the Slovenia PID registry
Gasper Markelj

Extended indications to bone marrow transplantation in PID
Liudmila Chernyshova

Modern principles of treatment of rheumatic diseases
Yaryna Boyko

Moderator: Anastasia Bondarenko
Joint syndrome in primary immunodeficiencies
Oksana Boyarchuk

Immune cytopenia in PID
Svetlana Sharapova

Organ-specific lesions in APECED
Larisa Kostyuchenko

Annual experience of transition from intravenous introduction of immunoglobulin to subcutaneous: features and benefits (Octapharma)
Galina Koval

April 23

Rheumatic diseases as the main manifestation of PID (clinical cases)
Moderator: Anna Hilfanova

Idiopathic juvenile arthritis as the first manifestation of Bruton's disease
Anna Hilfanova
Joint syndrome as the main manifestation of primary combined immunodeficiency
Tetiana Marushko

Nonspecific intestinal disease as a manifestation of X-linked lymphoproliferative syndrome
Daria Yanushevska, Raisa Mostovenko

Systemic lupus erythematosus in patients with complement deficiency
Olena Mukvich

Autoinflammatory diseases
Moderator: Yuri Stepanovsky

Periodic disease. Questions and answers
Olena Oshlyanska

PFAPA or FMF? How to correctly interpret the results of genetic testing?
Yuri Stepanovsky

A clinical case of Schnitzler's syndrome
Victoria Vasylets

Experience of canakinumab use in Ukraine in patients with auto-inflammatory diseases
Yuri Stepanovsky

Clinical case of autoinflammatory undifferentiated syndrome in a child with glycogenosis type 1A
Solomiya Timchyshyn

Vaccination in PID and rheumatic diseases or in patients with PID receiving immunosuppressive therapy
Fedir Lapiy

L-R: Anastasia Bondarenko, Fedir Lapiy, Yuri Stepanovsky, Alla Volokha
SUMMARY

On April 22-23, 2021, the annual Ukrainian XVIII J Project symposium "Rheumatic diseases in Primary Immunodeficiencies" was held, organized by the Department of Pediatric Infectious Diseases and Pediatric Immunology of Shupyk NMAPE and Ukrainian Association of Pediatric Immunology within the framework of the Central- Eastern-European J Project.

The main objectives of the event were education, training and dissemination of knowledge on primary immunodeficiency (PID), as well as discussion of the diagnosis and treatment of PID. This year's symposium drew attention to rheumatic diseases as the frequent manifestation of primary immunodeficiency. It was aimed to strengthen the interdisciplinary interaction of pediatric immunologists and rheumatologists in order to improve the diagnosis and management of patients with primary immunodeficiency. The important additional topic of the symposium was the introduction of the National on-line register of primary immunodeficiencies.

The scientific symposium "Rheumatic diseases in Primary Immunodeficiencies" aroused great interest in the medical community due to the quality of information content and consideration of current practical issues. The event was attended by 630 doctors different specialities: pediatricians, family doctors, rheumatologists, as well as immunologists from all over Ukraine. The speakers of the conference were leading experts, immunologists and rheumatologists from Ukraine, as well as specialists from Slovenia and Belarus.

In addition to the main program of the conference, master classes were held on the use of subcutaneous immunoglobulin drugs in patients with severe hypogammaglobulinemia.

Alla Volokha
TIMISOARA, ROMANIA

Host: Mihaela Bataneant
Venue: “Louis Turcanu” Children's Emergency Clinical Hospital + Webinar
Topic: Primary immunodeficiencies in and out of the COVID pandemic

UMF “Victor Babeș” Timișoara
Centrul “Jeffrey Modell” Timișoara
Spitalul Clinic de Urgență pentru Copii Louis Turcanu

PROGRAM

Diana Deleanu (Cluj, adult patients)
Ciprian Jurcut (Bucharest, adult patients)
Alexis Cochino (Bucharest, pediatric patients)
Aurica Rugina (Iasi, pediatric patients)
Mihaela Bataneant (Timisoara, pediatric patients)

April 22 – J Project Junior
“Louis Turcanu” Children's Emergency Clinical Hospital

J Project junior meeting:
Educational lectures and instructive clinical presentations by residents

The role of Project J in the development of primary immunodeficiencies in Romania
Mihaela Bătăneanț, Timisoara

Hypogamaglobulinemia in children - differential diagnosis illustrated by case presentations
Sabrina Danciu, Timișoara

Combined immunodeficiencies - the importance of early diagnosis
Cristina Popa, Timișoara

3 cases with phagocyte defects (severe congenital neutropenia, chronic granulomatous disease and WHIM syndrome)
Andreea Beloia, Timișoara

Key elements in the diagnosis of primary immunodeficiencies
Mihaela Bătăneanț, Timisoara
Hemophagocytic lymphohistiocytosis - a potentially fatal disease
Silvana Târziu, Timișoara

April 23

J Project meeting with seniors: Educational lectures on pediatric and adult patients

A complex case of IgA deficiency
Aurica Rugina, Iasi

Allergy in primary immunodeficiencies
Bianca Matiș, Timișoara

ENT manifestations suggestive of immunodeficiency
Bianca Petcu, Timișoara

Possibilities for diagnosis, monitoring and treatment of the patient with primary immunodeficiency in a city hospital
Maria Savu, Deva

Gastroenterological manifestations in patients with primary immunodeficiencies
Victor Baldea, Timisoara

Vaccination in primary immunodeficiencies
Mihaela Bataneanț, Timisoara

Q&A; Discussions

Conference of patients with primary immunodeficiencies – online

Opening of the conference

Virtual exhibition made by the High School of Fine Arts Timisoara,
Anca Delia Gyemant, profesor coordonator

Primary immunodeficiencies and pandemic. Cortisone in COVID - his friend or enemy?
Alexis Cochino, București

Infection with the new Corona virus
Irena Pintea, Cluj-Napoca

News and perspectives for patients in Romania
Ciprian Jurcut, București

Vaccination against COVID-19 in primary immunodeficiencies
Mihaela Bătăneanț, Timișoara

Ensuring the continuity of treatment of patients who depend on plasma-derived therapies
Marilena Stănescu, PVA Head Takeda România

The situation of human immunoglobulin supply in Europe and Romania
Florin Hozoc, CEO BPHD Life Solutions

Discussions and closing of the conference
SUMMARY

On April 22-23, 2021, the 10th J Project Meeting from Romania was held in Timisoara, Romania in a hybrid model. The meeting was organized by the University of Medicine and Pharmacy “Victor Babes” Timisoara and “Louis Turcanu” Children's Emergency Clinical Hospital.
The participants included residents in pediatrics and neonatology, pediatricians, general practitioners, laboratory doctors, allergist, otorhinolaryngologist, gastroenterologist, infectious disease and internal medicines doctors, from Romania: Timisoara, Deva, Iasi, Bucharest, Cluj. The J Project Meeting was entitled: Primary immunodeficiencies in and out of the COVID pandemic.

We started with juniors meeting in “Louis Turcanu” Children's Emergency Clinical Hospital with almost 60 participants. After a brief presentation of the suggestive signs for PID, there were presented and discussed clinical cases from main topics of PID. On April 23rd we had webinar with clinician and with patients and ARPID (Romanian Association of patients with PID). Because of the pandemics, the access of patients with primary immunodeficiencies to referral centers was difficult. In this period the local doctors had a crucial role in the management of these patients. From this reason we decided to involve doctors from several specialties, the main aims being to increase and broaden knowledge of PID and to establish professional collaboration and to facilitate early diagnosis and treatment. The invited speakers were Conf. Dr. Rugina Aurica from Iasi, Dr. Maria Savu, pediatrician from Deva, Hunedoara county, Matis Bianca allergist, Bianca Petcu ENT-ist and Victor Baldea, resident in gastroenterology in Timisoara, whose mother has CVID. There were discussed the main clinical manifestations in PID. The last part of the meeting was held also online. Starting with a virtual exhibition made by the High School of Fine Arts Timisoara, we continued with 2 topics: PID and COVID and the status of access to immunoglobulin in our country. Invited speakers were from Bucharest: Dr Alexis Cochino (pediatrician) and Dr. Jurcut Ciprian (internal medicine), from Cluj: dr. Irena Pintea (allergist) and Dr. Bataneant Mihaela.

The patient’s organization – ARPID was an important part of this meeting, and its president gave us information about their achievements and activities. Also, they invited two representatives of immunoglobulin companies and distributors and we discussed about immunoglobulin supply status in Romania. The scientific program and discussions of this meeting were highly successful, and we would like to thank to all participants.

Mihaela Bataneant
BUDAPEST, HUNGARY  
APRIL 23

Host: Katalin Haraszti Honváríné & László Maródi  
Venue: Webinar  
Topic: PID for students, parents and teachers

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**ONLINE IMMUNOLÓGIAI ELŐADÁSOK**

**ELŐADÓK ÉS TÉMÁK:**

- Dr. Lányi Árpád  - A csodálatos immunrendszer
- Dr. Kovács Anikó  - Kamaszkori bőrproblémák, tetoválások veszélyei
- Dr. Szolnoky Miklós  - Immunbetegségek jelei és tünetei
- Dr. Tóth Beáta  - Gének és immunitás, génmutáció

Taksony Vezér Német Nemzettségi Általános Iskola, 7-8. osztályosok részére

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**PROGRAM**

The wonderful immune system
*Arpad Lanyi*

Adolescent skin problems, the dangers of tattoos
*Aniko Kovacs*

Signs and symptoms of immune disorders
*Miklos Szolnoky*

Genes and immunity, mutations
*Beata Toth*
SUMMARY

A miniseries of four lectures was designed to introduce the concept of immune defense, to 6-8 graders who had no previous education in Immunology. Beyond the basics, lectures also discussed regulation of the immune response, inborn errors of immune response genes causing immunodeficiencies or autoimmune disease.

The first 20 minutes talk by Árpád Lányi PhD., associate professor at the Department of Immunology of the University of Debrecen gave an overview of innate and acquired immunity, immune cell functions, and basic vaccination strategies. In the final part of his talk Dr. Lanyi discussed allergy, and contemporary approaches for the induction of allergen-specific immune tolerance.

Aniko Kovács, dermatologist at the Department of Dermatology, Venereology & Dermatooncology, Semmelweis University, Budapest spoke about adolescent skin problems.

Miklós Szolnoky, immunologist, Szent János Hospital, Budapest summated the signs and symptoms of immune disorders. Beáta Tóth, molecular immunologist, University of Debrecen talked about the disease causing genes of the immune system. She gave an overview of laboratory methods using to test DNA mutations.
In front: Katalin Haraszti Honváriné
PROGRAM

Session 1
COVID-19, etiology, immunity and treatment
Marianna Murdjeva, Medical University-Plovdiv

PID and COVID-19
Spaska Lesichkova, Alexandrovska University Hospital, Sofia

COVID-19 and Post-COVID-19 complications in immunocompromised children
Maria Spasova, University Hospital “St. George”, Plovdiv

COVID-19 epidemiology measures
Jordanka Stoilova, Medical University– Plovdiv
Cutaquig – New generation subcutaneous immunoglobulin for treatment of patients with PID – Octapharma AG
Angel Stoimenov

Session 2
Consultations and discussions - patient questions and answers from medical doctors.
Moderators : M. Murdjeva, M. Spasova, E. Naumova, I. Stoilova, S. Mihailova, S. Lesichkova

SUMMARY

The Fifth school on Primary Immune Deficiencies in Bulgaria was organized online due to the COVID-19 pandemic. It was organized and supported by: 1) Bulgarian Association of Clinical Immunology (BACI), 2) Medical University-Plovdiv, 3) Functional Expert Center “Jeffrey Model”, 4) Expert center for rare diseases-PID, University Hospital Alexandrovska, Sofia, 5) J Project and 6) Bulgarian Association of patients with PID. It was addressed to patients with PID and parents of children with PID.

Prof. Elissaveta Naumova opened the meeting by introducing the audience to the progress made in diagnosis and treatment of PID in Bulgaria and the significance of raising awareness of defects of the immune system besides the existing difficult pandemic situation. She announced the new
opportunity for online consultations during the pandemic with a digital questionnaire developed for this purpose for symptoms suspected of PID.

Prof. Marianna Murdjeva, presented a lecture about the etiology, immunity and treatment of SARS-Cov-2 related illness.

COVID-19 infection in patients with PID was presented by Dr. Spaska Lesichkova. She made an overview of indications and contraindications of treatment protocols. Dr. Lesichkova also commented on COVID-19 vaccinations for patients with PID.

Assoc. Prof. Maria Spasova presented case reports on COVID-19 infection in children who develop MISC Syndrome, differential diagnosis and treatment options.

Prof. Jordanka Stoilova (epidemiologist) presented a lecture about the prophylactic measures - 3Ds – disinfection, distance and discipline in minimizing COVID-19 infection.

Dr Angel Stoimenov, presented Cutaquig, a new generation human immunoglobulin solution for subcutaneous administration. In his review he emphasized the advantages and better quality of the new medicine and the excellent place which Bulgaria holds in Europe and worldwide.

After the lectures a dialogue and discussion with patients and medical doctors took place, where several important questions were presented: epidemiology measures for Bulgarian Christian rituals in the churches; educational process and exams for children with PID in the pandemic situation; plasma treatment for COVID-19 in patients with PID; treatment with the new SCIG- Cutaquig. That was an opportunity to discuss different aspects of COVID-19 disease and vaccination process.

Marianna Murdjeva & Elissaveta Naumova
Host: Natalja Kurjane
Venue: Webinar
Topic: Primary immunodeficiency in clinical practice in adults and children: analysis of clinical cases
The online JP Meeting "Primary immunodeficiency in clinical practice in adults and children: analysis of clinical cases" was held on the 24th of April in Riga, Latvia. This JP Meeting was focused on the discussion of challenging clinical cases of primary deficiencies. We aimed to increase the awareness and knowledge about different clinical aspects of primary immunodeficiencies among the doctors of different specialties. This meeting was co-organized with the Latvian Medical Association.

The moderator of this webinar was Natalja Kurjane, from Paula Stradins Clinical University Hospital Riga, Latvia. The first speaker was Tatjana Prokofjeva, a pediatrician and immunologist from Childrens’ Clinical University Hospital in Riga, Latvia, who spoke about the epidemiology of primary immunodeficiencies in Latvia. The second lecture was given by Inese Mihailova, a nephrologist and immunologist from Paula Stradins Clinical University Hospital in Riga, Latvia, who focused her talk on various aspects of autoimmunity in patients with primary immunodeficiencies. Then the floor was given to Zanna Kovaleva, a pediatric hematologist from the Childrens’ Clinical University Hospital, who presented an instructive clinical case of a patient with X-linked lymphoproliferative disease. The next lecturer was Ieva Nokalna, a pediatrician and immunologist from Childrens’ Clinical University Hospital, who presented several educational cases on chronic granulomatous disease and gave a summary about an abscess as a clinical manifestation of primary immunodeficiency. After this lecture, Lota Ozola, a pediatric pulmonologist and immunologist from Childrens’ Clinical University Hospital, gave a speech that concentrated mainly on differential diagnostics of recurrent sinopulmonary tract infections in children. The next speaker was Edita Gasiuniene, an immunologist and allergologist, the Head of Primary Immunodeficiency Center in Hospital of Lithuanian University of Health Science Kauno Klinikos, Lithuania. She presented a unique clinical case about a patient with common variable immunodeficiency and lung transplantation. After that Zane Lucane, a general practitioner and resident in clinical immunology from Paula Stradins Clinical University Hospital, presented a clinical case about recurrent pericarditis in a patient with the autoinflammatory disease and gave a short summary about this group of diseases. The next speaker was Adine Kanepa, an internist, allergologist and resident in clinical immunology from Paula Stradins Clinical University Hospital, who focused her talk on the differential diagnosis of angioedema and presented an instructive clinical case about hereditary angioedema. The next lecturer was Pavels Gordins, an allergologist and immunologist, the Head of the East Yorkshire Regional Immunology and Allergy Unit, Castle
Hill Hospital, East Yorkshire, United Kingdom. His lecture was focused on a practical approach to diagnosing primary immunodeficiency for non-immunologists. After that, the floor was given to Inga Nartisa, a laboratory doctor and geneticist from Childrens’ Clinical University Hospital. In her lecture, she concentrated on the possibilities of genetic testing of primary immunodeficiencies in Latvia. Finally, Natalja Kurjane, an internist and immunologist from Paula Stradins Clinical University Hospital, concluded the meeting with a lecture about the follow-up and treatment of primary immunodeficiency patients in immunologist's everyday practice.

More than 150 doctors of different specialties, mostly pediatricians, internists, family doctors, and neurologists attended this meeting. We appreciate the active participation of the speakers and the ambitious discussion by many clinicians.

Natalja Kurjane
TEHRAN, IRAN

APRIL 26-27

Host: Nima Rezaei
Venue: Virtual Congress
Topic: 12th International Congress on Immunodeficiency Diseases (ICID)” J Project 2021: In Memory of Prof. Asghar Aghamohammadi

PROGRAM
April 26th

Challenges of Inborn Errors of Immunity

Opening session - in memory of Prof. Asghar Aghamohammadi
*Nima Rezaei*, Iran

Probing Common Variable Immunodeficiency - in memory of Prof. Asghar Aghamohammadi
*Klaus Warnatz*, Germany

Playing with fire - Type-I interferons in health and disease
*Tim Niehues*, Germany

Diagnosis of PID: from easy tests to molecular studies
*Teresa Español*, Spain

The HSCT decision in patients with Profound Combined Immunodeficiency (P-CID): a prospective cohort study
*Stephan Ehl*, Germany

Genetic and immunological causes of life-threatening COVID-19
*Jean-Laurent Casanova*, USA

COVID-19 in PID
*Kathleen Sullivan*, USA

COVID-19 in Patients with PID and SID
*Fabian Peissker*, Germany

Earliest Possible Diagnosis – How You Can Make it Happen; How You Can Make it Matter; How You Can Do it Now!
*Vicki Modell and Fred Modell*

April 28th

Inborn Errors of Immunity and Associated Conditions

CD19 deficiency: From beginning to date
*Ismail Reisli*, Turkey

Hyperactive STAT1 signaling operates dysregulated immune profile in STAT1 GOF disease partially reversing with ruxolitinib
*Safa Baris*, Turkey

Pathology of the organs of vision in patients with primary immunodeficiency
*Gulnara Nasrullayeva*, Azerbaijan
Inborn errors of immunity predominantly affecting the gut
Ahmet Ozen, Turkey

Monogenic Intestinal Epithelium Defects and the Development of Inflammatory Bowel Disease
Mahnaz Sadegh-Shabestari, Tabriz, Iran

Probiotic application in Immunodeficiency disorders: Beneficial or harmful?
Hamid Ahanchian, Mashhad, Iran

Early IBD and primary immunodeficiencies
Tooba Momen, Isfahan, Iran

Enteropathy and primary immunodeficiency disorders
Marziyeh Tavakol, Karaj, Iran

An overview of novel autoinflammatory cases in a cohort study of Iranian patients with Inborn Error of Immunity between 1976 and 2020
Roya Sherkat, Isfahan, Iran

EBV-associated primary immunodeficiency diseases
Samin Sharafian, Tehran, Iran

Childhood Chronic Mucocutaneous Candidiasis
Sepideh Darougar, Iran

Updates of Primary immunodeficiency and chronic mucocutaneous candidiasis
Abbas Khalili, Iran

Questions and Answers

April 29th

Regional Challenges of Inborn Errors of Immunity

COVID-19 Presentations in Pediatric Patients with Inborn Errors of Immunity
Nasrin Moazzen, Mashhad, Iran

COVID-19 and the Immune System
Zahra Daneshmandi, Iran

Mendelian Susceptibility to Mycobacterial Disease (MSMD)
Alireza Mahdaviani, Tehran, Iran

New updates about Hyper IgE
Javad Ghafari, Mazandaran, Iran

Virological status of chronic diarrhea in patients with Primary Immunodeficiency
Kian Darabi, Iran

IVIG Infusion side Effects: A long Term study in Shiraz
Hossein Esmaeilzadeh, Shiraz, Iran

Complement deficiency
Mojgan Kiani Amin, Tehran, Iran

Evaluation of T and B Cell Subsets in Patients with Ataxia Telangiectasia
Reza Yazdani, Tehran, Iran

Report of two cases with PGM 3 deficiency and review of PID associated with a TH2 diathesis
Zahra Chavoshzadeh, Tehran, Iran

PID in Yazd: a historical cohort study of 97 Primary immune deficiency patients during 10 years
Nasrin Behniafard, Yazd, Iran

Malignancy in Primary Antibody Immunodeficiency
Narges Eslami, Iran

The influence of COVID-19 pandemic on patients with PID in Fars province, Southeast Iran
Soheila Alyasin, Shiraz, Iran

Questions and Answers

Concluding Remarks
SUMMARY

On 26-29 April 2021, a virtually scientific webinar, entitled “12th international Congress for Immunodeficiency Diseases (ICID)” (http://rcidcong.tums.ac.ir/) was organized by Universal Scientific Education and Research Network (USERN) (https://usern.tums.ac.ir/) and Research Center for Immunodeficiencies, the referral center of primary immunodeficiency diseases (PID) in Iran for both children and adult patients (http://rcidcong.tums.ac.ir/). More than 400 people participated in this meeting, which aimed to increase awareness of general physicians and pediatricians about the principal manifestations of PID and the correct approach to those signs and symptoms.

To briefly mention the history of the event, the first International Congress on Immunodeficiency Disorders was organized in 2005. That congress was a great event to further develop bilateral scientific exchange of Iranian scientists with other researchers of the world. In October 2009, alongside with the 21st International Congress of Pediatrics, a joint meeting on Immunodeficiency Diseases was established. Since 2011, as of establishment of the Research Center for Immunodeficiencies, the Meetings is organized annually in April during the PID week.

On 26th April, the congress began with a lecture in memory of Prof. Asghar Aghamohammadi by the head of RCID. Afterward, the congress keeps on with the presentation of keynote lectures by distinguished professors in immunology: Kalus Warnatz, Tim Niehues, Teresa Espanol, Estephan Ehl, Jean- Laurent Casanova, Kathleen Sullivan, Fabian Peissker, and Viki and Fred Model.

On 27th, the congress continued with PID presentations and the relation between PID and COVID-19. Some well-known professors in immunology
and pediatrics, Hirokazu Kanegane, Bodo Grimbacher, Waleed Al-Herz, Mirjam van der Burg, Sergio Rosenzweig, Jordan S. Orange, presented their latest research on this issue. The program was continued by a number of lectures from regional and national speakers on April 28th and 29th.

PI week organizers participated in several TV and media interviews and informed the public about the prevalence of PIDs in Iran, warning signs of PIDs, and treatment of PID patients.

Nima Rezaei
Host: Gulnara Nasrullayeva
Venue: Webinar
Topic: Detection and treatment of primary immunodeficiencies

PROGRAM

Moderator: Nasrullayeva Gulnara. Professor, Head of Scientific-Research Immunology Laboratory of AMU

Opening remarks

Detection and treatment of Primary Immune Deficiencies in Azerbaijan
G Nasrullayeva, AMU, Department of Children Diseases

Diagnosis of X-dependent agammaglobulinemia
V Mammadova, AMU, Scientific-Research Immunology Laboratory

Auto-ABs against type I INFs in patients with critical COVID-19 pneumonia (Patients with critical COVID-19 pneumonia)
Paul Bastard, Paris University, Laboratory of human genetics of infectious diseases, The Rockefeller University (New York).

The effect of intrauterine infection on the immune system
N Mollayeva, AMU, Department of Children Diseases

Common variable immune deficiency in pediatric practice
B Babayeva, Scientific Research Institute of Pediatrics named by K. Faradjeva
Batndaxili İnfeksiyanın İmmun sistemə təsirı

Azerbaiyanda Birinci İmmun Çatışmazlıqların aşkarlanması və müalicəsi

Azerbaiyanda Birinci İmmun Çatışmazlıqların aşkarlanması və müalicəsi
Within the framework of World Primary Immunodeficiency Week, on April 29, 2021, in the capital of Azerbaijan, Baku, we held an online webinar dedicated to the detection and treatment of primary immunodeficiency (PID). This webinar was hosted by the PID Center with the support of the Medical Media online medical platform and was broadcast live on Facebook and YouTube. The webinar was attended by about 100 participants, mainly doctors of various specialties from hospitals and clinics in different cities and regions of the country, interns and residents, as well as the parents of patients. The announcement of the webinar was previously posted on our Facebook page "Help for Patients with Primary Immunodeficiency - IIXXY".

Speaking at the webinar, doctors from Azerbaijan and France spoke about the relevance of PID, the importance of timely and accurate diagnosis of diseases. They described their performances in the case description.

Opening the webinar, Professor Gulnara Nasrullayeva spoke about the work carried out in Azerbaijan over the past 11 years to identify and register PID patients, and introduce modern examination methods. She noted that over the years, educational seminars and meetings with specialist doctors were regularly organized, our employees participated in international conferences and television broadcasts, textbooks and scientific recommendations, articles in newspapers and magazines were published in order to reach a wider audience of the population.

The second speaker was the Ph.D. Vafa Mammadova. She provided information on the pathology associated with antibody immunodeficiency, which is most common both in the world and in Azerbaijan, and presented a report on a patient with Bruton's syndrome. The speaker dwelled on the clinic, diagnosis, immunological examination and treatment of such patients in detail, noted the importance of genetic testing in the diagnosis of the disease.

At a time when the whole world is fighting the COVID-19 pandemic, the problem of COVID-19 in PID patients is of particular importance. For this reason, Paul Bastard, Ph.D. from the University of Paris and Rockefeller University, made a very interesting presentation on the role of auto-antibodies against type I INF in patients with critical pneumonia in COVID-19. The speaker explained the important role of pro-inflammatory cytokines in the infectious process, noted that COVID has consequences that are more
serious in older people with autoimmune diseases. His speech was presented by simultaneous translation of the Azerbaijani audience.

The next speaker was Nushaba Mollaeva, Ph.D of the Department of Childhood Diseases of the 1st AMU, who presented the results of a study on the influence of intrauterine infection on the development of PID and emphasized the importance of early prenatal diagnosis.

Ass. prof. Bayaz Babaeva told the participants of the webinar about the clinical picture and features of the course of general variable immunodeficiency in children, explained the differences between this immunodeficiency and other types of antibody deficiency.

The webinar generated a lot of interest. At the end of the webinar, answers were given to questions from the audience online. It was emphasized that timely and correct diagnosis of the disease plays an important role in the lives of patients and their lifestyle.

Gulnara Nasrullayeva
BRATISLAVA, SLOVAKIA

Host: Peter Ciznar
Venue: Webinar
Topic: Early diagnosis and access to care of primary immunodeficiencies in Slovakia and surrounding countries – achievements and challenges

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Sympózium o primárnych imunodeficienciách 2021
(Symposium on primary immunodeficiencies 2021)

Včasá diagnostika a starostlivosť o pacientov s primárnou imunodeficienciou na Slovensku a v okolitých krajínach – úspechy a výzvy
(Early diagnosis and access to care of primary immunodeficiencies in Slovakia and surrounding countries – achievements and challenges)

Wednesday 28 April, 2021
on-line

15:00 Úvod (Introduction)

15:15 – 16:45 Časť 1 (Part 1)
Bernatowska E.: BCG-Associated Complications in Primary Immunodeficiency Patients - Thirty-Nine Years of Experience in the Department of Immunology, Children’s Memorial Health Institute, Warsaw
Forstner-Waldl E., Pummer L.: Immunodeficiency phenotype in syndromic diseases, Vienna AKH
Volokha A.: IPEX syndrome in Ukraine
Kriván G.: Genetic diagnosis in PID, Szent László Hospital, Budapest

16:45 Key note: Sedivá A. COVID u pacientov s PID (COVID complications in PID patients)

17:15 – 18:45 Časť 2 (Part 2)
Hrubíško M. Sledovanie stavu bunkovej imunity pacienta po prokonaní ochorenia COVID-19
Jesenáč M., Bobčáková A., Kapustová O., Petrovičová O., Bánovčin P.: Skúsenosti so zriedkavými primárnymi komplementopatiami
Ostro R. EBV asociované PID - XLP syndromy
Čižnár P., Huľínková I., Melicharová J., Kuková Z.: Primárne imunodeficiencity s atopickým fenotypom

18:45 - 20:00 Časť 3 (Part 3)
Čižnár P.: Substitučná liečba imunoglobulínmi a darcovstvo plazmy
Imunodeficit.sk – Pacientkska organizácia pre primárne imunodeficiencie

Nadácia JMF – story, film

20:15: Diskusia a ukončenie (Final conclusions)
SUMMARY

After a one-year forced break, the PID-J Project Symposium with international participation was held formally in Bratislava again, though via an on-line format thanks to the Solen agency for Medical Education.

It turned out, however, that the online format was more accessible to a larger number of participants. While in the past it was attended by about 100 participants, this year 289 were registered from all around Slovakia. Thanks to the online format, participants were able to listen to lectures and participate in discussions with leading experts from countries around Slovakia. The invitation was accepted by prof. Bernatowska from Warsaw, prof. Šedivá from Prague, prof. Forster-Waldl from Vienna, prof. Volokha from Kiev and Dr. Kriván from Budapest.

The symposium was divided into 3 parts. Included was the topic of COVID-19 infection and inborn errors of immunity, which was discussed in the keynote lecture of prof. Šediva and in the lecture of prof. Hrubisko, the formal Head of the Slovak Clinical Immunology Society. In the third part a short presentation of the Slovak PID Patients’ Organization was held by Mr. Bodor a representative of the organization. There was a lively discussion on the topic of plasma donation in Slovakia. The country lags significantly behind in plasma collection, and clinical immunologists and hematologists are working to change the legislation. This is an important step in improving the availability of the immunoglobulin products.

A spectrum of PIDs is associated with mycobacterial infection complications and increased susceptibility to BCG live vaccine. Poland is one of few European countries continuing with early whole population introduction of BCG vaccine. Prof. E.Bernatowska presented 39 year of experiences. According to Polish CMHI study SCID patients presented in 42% local adverse events in place of inoculation, and 15% of disseminated BCG complication. This finding seems to be less than reported by Marciano et. al., with 34% of disseminated forms in 119 of 349 SCID patients. In CGD group of patients the Duch or Pasteur substrain of BCG vaccine has been responsible for abscess or skin complication in 8% patients. Prof. Bernatowska stressed the difference of vaccine strain and PID type. Replacement of highly reactogenic vaccines (Russia, Sweden, Danish and Pasteur) with less virulent (Japan, Moreau, Glaxo, Prague) lead to great
reduction in complications. However, early diagnosis with introduction of newborn screening, should prevent BCG vaccination in any SCID patient. A spectrum of PIDs is associated with mycobacterial infection complications and increased susceptibility to BCG live vaccine. Poland is one of few European countries continuing with early whole population introduction of BCG vaccine. Prof. E.Bernatowska presented 39 year of experiences. According to Polish CMHI study SCID patients presented in 42% local adverse events in place of inoculation, and 15% of disseminated BCG complication. This finding seems to be less than reported by Marciano et. al., with 34% of disseminated forms in 119 of 349 SCID patients. In CGD group of patients the Dutch or Pasteur substrain of BCG vaccine has been responsible for abscess or skin complication in 8% patients. Prof. Bernatowska stressed the difference of vaccine strain and PID type. Replacement of highly reactogenic vaccines (Russia, Sweden, Danish and Pasteur) with less virulent (Japan, Moreau, Glaxo, Prague) lead to great reduction in complications. However, early diagnosis with introduction of newborn screening, should prevent BCG vaccination in any SCID patient.

Professor Volokha presented a review and a case of IPEX syndrome. Immune dysregulation associated with severe enteropathy, exfoliative erythroderma, polyendocrinopathy with hypothyroidism lead to genetic confirmation of mutation in FOXP3 gene in an infant referred to their center. Introduction of corticosteroids reduced the enteropathy and bronchitis and allowed to start the HSCT as the only curative therapy recently available. Prof. Volokha discussed the variability of clinical presentation, including atypical IPEX which must be considered even if onset of symptoms is in late childhood. Diagnostic steps and management processes have been presented according to Barzaghi and Passerini, 2018. Cited large multicenter study suggested that HSCT at time of mild organ impairment results in resolution of autoimmunity.

Genetic results may substantially affect the primary clinical diagnosis and the therapy, as documented by Dr. Krivan. A case of CTLA-4 defect, SYK gain of function defect and G6PC3 deficiency have been excellently documented. Genetic testing was performed in CEMM laboratories of K. Boztug in Vienna. In all three cases a new biologic treatment has been presented. CTLA-4 functions as an immune checkpoint and downregulates immune responses. Variants in this gene have been associated with autoimmune diseases and germline haploinsufficiency leads to CHAI disease (lymphoproliferation, autoimmunity and hypogammaglobulinemia). Dr. Krivan presented experience with abatacept.
This biologic drug modulates immune response by preventing costimulatory binding of naïve T cells. In a second case tyrosine kinase SYK mutation have been diagnosed in a patient with frequent respiratory infections, enteritis and lymphadenopathy. Genetic diagnosis has led to switch from immunosuppressive methylprednisolone and CSA to fostamatinib, a SYK inhibitor formally used for treatment of refractory chronic immune thrombocytopenia. Finally, a case of agranulocytosis was presented. G6PC3 deficiency lead to accumulation of toxic product 1,5AG6P which intoxicated neutrophils. Inhibition of renal sodium glucose cotransporter with empagliflozin leads to glycosuria and urinary excretion of 1,5AG. The result was an increase in the pool of G6P, improvement in respiratory burst and protein glycosylation. Introduction of empagliflozin allow to stop previous G-CSF treatment, lead to granulocyte function normalization and resolution of thrombocytopenia.

Combined immunodeficiencies associated with syndromic features were presented by Prof. Forster-Waldl. The typical one represents: congenital thrombocytopenia (i.e. WAS), DNA repair defects (ataxia telangiectasia, Ligase IV-deficiency), thymus defects with additional congenital anomalies (MDS 22q11), Immuno-ossseous dysplasia, Hyper-IgE syndromes, Defects of vitamin B12 and folate metabolism and anhidrotic ectodermodyplasia with immunodeficiency. The presentation by prof. Forster-Waldl stressed the importance of performing immunological investigation in patients with chromosomal aberrations and recurrent ENT or airway infections to identify potential immunodeficiency, that can be specifically treated. A significant proportion of these patients have antibody deficiency requiring immunoglobulin replacement and antibiotic prophylaxis.

The COVID epidemic has also affected lives of hundreds of PID patients with a different outcome as showed by professor Šedivá from Prague PID center. The role of interferon type 1 in antiviral immunity have been extensively documented. Immunodeficiencies affecting interferon response showed to be associated with more severe course of COVID-19. The 3 major studies have been cited on impact of coronavirus disease in patients with inborn errors of immunity. I. Meyts et al. (ESID) study of 94 PID cases, I.Quinti et. (Italy) study and a Cunningham-Rundles 2021 (New York) study of 16 cases. In all of them it seems that nonspecific factors like age, chronic lung disease and pre-infection lymphopenia were considered more deleterious than PID itself. However, IFN defects, defects of cytotoxic immunity and dysregulation syndromes with autoantibodies against IFNs...
are associated with severe complications. Prof. Šedivá also cited an exceptional project named COVID human genetic effort led by J-L. Casanova, targeted to identify variation in genes leading to increased susceptibility to severe COVID disease. Finally experiences of the Prague PID centers have been also presented.

M. Hrubisko reported the first experience with tests of cellular immunity response of patients after COVID-19 infection. He stressed that intensity of the immune response is modulated by the severity of infection; 11% of patients with asymptomatic or mild disease do not form neutralizing antibodies. Unfortunately, the commercially available test in Slovakia use N antigen response which do not reflect a postvaccination antibody formation. However, no response does not necessarily mean any response. SARS-CoV-2 specific T-cells could be detected even in seronegative patients. He noted that T-cell response could be a more sensitive indicator of virus exposure. Prof Hrubisko further reported the first experience with T-SPOT test against SARS-CoV-2; 96,6% of COVID-19 PCR positive patients show positive T-SPOT results 60 day after exposure; and 83,3% 60 to 240 days after exposure. He concluded that cellular response seems to be a more sensitive and persistent test for SARS-CoV-2 exposure in hypogammaglobulinemia patients.

M. Jesenak presented a review of complement functions and primary immunodeficiency disorders. Complementopaties represent a rare form of PID, approximately 1-2% of all PIDs. Infectious and noninfectious complications make a heterogeneous clinical picture in affected patients. Also, therapies vary according to the specific defects from antibiotic prophylaxis, immunosuppression and HSCT to targeted biological treatments. Vaccination strategy should be an important tool for severe bacterial complication in terminal complement pathway malfunction. Authors presented clinical experiences with five cases of complement deficient patients, excluding their large experience with hereditary angioedema cohort. Presentation include a case of 12-year and 29-year old with recurrent *Neisseria meningitidis*, a 3-year-old with C2 deficiency and transitory hypogammaglobulinemia, a 17-year-old with atypical hemolytic uremic syndrome and factor I deficiency case. Experience with eculizumab treatment have been discussed.
R. Ostro reviewed Epstein Barr virus associated immunodeficiencies, namely XLP syndromes. EBV is a ubiquitous virus and nearly all humans are exposed throughout their life. Usually it runs asymptomatic or in the case of adolescents and young adults it can result in the clinical picture of infectious mononucleosis. EBV-infected B lymphocytes are controlled primarily by NK cells, CD4+T and CD8+T lymphocytes. Several genetic disorders affecting T and NK cell function result in failure to control EBV infection. Mutations in SH2D1A (XLP1), BIRC4 (XLP2), ITK, CD27, MAGT1, CORO1A, and LRBA are virus specific and do not result in increased susceptibility to other viruses. Mutations in genes associated with hemophagocytic lymphohistocytosis, PRF1, STXBP2, and UNC13D, can also predispose to severe chronic active EBV disease. Immunodeficiencies with severe EBV infection but also with susceptibility to other viral, bacterial and fungal infections include mutations in PIK3CD, CTPS1, STK4, GATA2, MCM4, FCGR3A, CARD11, ATM, and WAS. In addition, patients with severe combined immunodeficiency, which can be due to mutations in a number of different genes, are at high risk for EBV B lymphocyte lymphomas. Studying these defects allowed to better understand the role of different proteins important for control EBV and other viral diseases.

P. Ciznar focused on monogenic defects associated with a clinical presentation of an atopic phenotype. Since the description of Hyper-IgE syndrome by R. Buckley in 1975, a huge number of defects have been recognized in various signaling pathways resulting in very high plasma IgE concentrations or in severe atopic skin conditions. Knowledge has grown to understand the key pathways like T and B cell signaling, cytokine signaling, skin barrier functions or mast cell functions. Although most atopic conditions are environmentally driven conditions in some cases allergy alone is a presenting symptom of primary immunodeficiency, suggesting a monogenic etiology. Authors present clinical experiences - the diagnostic process and therapeutic outcome with eight patients. Their common signs include eczema, IgE over 2000 IU/ml and eosinophilia. STAT3 LOF have been confirmed in 6 cases, dominant negative IL6ST mutation in one case and dominant negative STAT5b mutation in another case. Authors conclude to focus on atypical atopy symptoms since early childhood, infectious complications, family history and non-immune conditions like growth delay and skeletal abnormalities. These patients should be analyzed in more
detailed way by genetic methods (NGS). HSCT and new targeted treatments could be offered to diagnosed patients improving the clinical outcome and quality of life.

Finally, the meeting ended with a JMF film “Do Something” projection. A strong story that encouraged medical participants to continue their daily efforts and encouraged patients to fight their disease. The symposium was for the PID community in Slovakia once again an excellent opportunity to share experiences, share rare cases and recent information about diagnosis and treatment of primary immunodeficiencies.

_Using Ciznar_

Speakers of the PID-J Project symposium:
(L→R) P Ciznar-Slovakia, E Bernatowska-Poland, A Sediva-Czech Republic, E Förster-Walldl-Austria, A Volokha-Ukraine
Past, present and future

Sympózium o primárnych imunodeficienciách 2021

Presentation format

Sympózium o primárnych imunodeficienciách 2021

BCG-Associated Complications in Primary Immunodeficiency Patients - Thirty-Nine Years of Experience in the Department of Immunology, Children's Memorial Health Institute, Warsaw

Ewa Bernatowska
ST. PETERSBURG, RUSSIA

APRIL 29

Host: Marina Guseva
Venue: Webinar
Topic: Diagnosis, routing and treatment aspects of patients with primary immunodeficiency

PROGRAM

Introduction
MN Guseva

Primary immunodeficiency in pediatrician practice
MN Guseva

Genetics of primary immunodeficiency in 2021

EN Suspitsyn

Rheumatological masks of primary immunodeficiency
MM Kostik

Discussion

SUMMARY

On 21th April, another educational project on primary immunodeficiency was held in St. Petersburg on the basis of the St. Petersburg Pediatric Medical University.

As in the previous year, due to the coronavirus pandemic, it was held online. The program covered the diagnosis, routing and treatment aspects of patients with primary immunodeficiency for pediatricians of all specialties. The consultative and diagnostic center of the Pediatric Medical University has accumulated extensive experience in the management of patients with primary immunodeficiency and works closely with clinics. Much attention was paid to new international achievements in the study of coronavirus infection, and the latest publications on this topic were presented.

In the report of MN Guseva, the latest international news on the diagnosis of PID was heard on the example of hereditary angioedema, new interesting clinical cases were presented. In the report of the geneticist EN Suspitsyn, the news in the genetic diagnosis of PID in the world practice was reported. Detailed information was given about the possibilities of TREC and KREC research, genetic research by the method NGS and the possibilities of performing a clinical exome at the Department of Medical Genetics of the St. Petersburg Pediatric Medical University. The report of MN Kostik covered in detail the rheumatological masks of primary immunodeficiency and orphan clinical cases of primary immunodeficiency. The online mode will allow doctors from many regions of Russia to connect.

Marina Guseva
Масштаб проблемы

• Частота – не менее 1:10000
• В мире – примерно 6 000 000 пациентов с ПИД, 70-90% не диагностированы
• Генетически детерминированные дефекты иммунитета, приводящие к повышенной частоте инфекционных, онкологических и аутоиммунных осложнений
• Постоянно расширяющийся спектр проявлений (волчанка, тяжелая пищевая аллергия, крапивница, отеки, ВЭК и т.д.)

Online-конференция
«Первичные иммунодефициты (ПИД) в практике педиатра. В рамках международной недели информирования о ПИД.»

14:50
As every year, we organized a meeting for J project. This year the main audience of the event were hematologists. The 297th event was a start for a new collaboration with the National Center of Hematology in field of primary immunodeficiency.

The main topic of the meeting was related with covid-19 and laboratory abilities for covid-19, PID and genetic testing. In this view the talk of Arsen Arakelyan (Director of the Institute of Molecular Biology) was an excellent and profound immersion in sequencing of genome of SARS CoV-2 from samples of Armenian patients. As a subject for further collaboration the possibility of using nanopore sequencing machine as diagnostic tool for PID was discussed. The results of the sequencing was compare with data of epidemiologic service of the National center of hematology. The immunization status of healthcare workers was introduced by Ruben Hovhannisyan (Head of the epidemiologic service of National Center of Hematology). The role of covid-19 antibody tests are staying unclear, but most of the staff had antibodies against SARS CoV-2.

This year we had a pleasure to attend a talk from Svetlana Sharapova (Minsk, Belarus). She prepared a beautiful presentation about
lymphoproliferative disorders in time of PID. This talk was the central as it linked the field of hematology. This type of talks are bridges between fields and could influence to further collaboration.

The last part of the event was about novel dysimmune features of MDS/CMML and Vexas syndrome. The subject was presented by Arsen Mekinian (Professeur des Universités-Praticien Hospitalier, Paris, France). The topic was a mix of rheumatology, clinical immunology and hematology. The real interdisciplinary view to the novel angiopathy. Arsen present his teamwork and encourage the young specialist to join his team and be published in ranked medical journals.

*Sevan Iritsyans*
Ruben Hovhannisyan, The immunization status of healthcare workers

Svetlana Sharapova, Hematologic features of PID
On April 29th the first J Project meeting was organized in Montenegro. The aim of the meeting was to form a national working group in Montenegro, that will be the new member of the J project network. Lidija Pocek, pediatric allergologyst informed her colleagues about the J Project network and share important informations.

The members of the working group in Montenegro will be:
- Veselinka Djurisic, pediatric gastroenterologyst
- Lidija Pocek, pediatric allergologyst
- Tamara Zivkovic, pediatric reumatologyst
- Milica Gazivoda, pediatrician, resident of allergology

The members of the working group agreed to start to make a PID register according to model from Slovenia. The working group have a collaboraton with „Dr Vukan Cupic Institute for Health Protection of Mother and Child Serbia“ from Belgrade regarding diagnostic and treatment procedures about PID patients. We also discussed the possibility to include geneticists in the working group. We agreed to have some lectures regarding PID in September 2021. I hope so that we will have possibillity to meet in some annual meeting and to share new informations.

Lidija Pocek
MINSK, BELARUS

Host: Mikhail Belevtsev
Venue: Business center of hotel Belarus
Topic: Topical issues of primary immunodeficiency
School-seminar on clinical immunology

Ministry of Health of the Republic of Belarus
Republican Scientific and Practical Center for Pediatric Oncology, Hematology and Immunology

VI conference with international participation
“Topical issues of primary immunodeficiency.
School-seminar on clinical immunology ”

PROGRAM

April 29
Opening of the conference
Lazar Dmitry Nikolaevich – Deputy Head of the Main Directorate of the organization Medical Assistance - Head of the Department of Medical Assistance to Mothers and Children Ministry of Health
Solntseva Anzhelika Viktorovna – MD, DSc, Professor, Director of the Republican Scientific and Practical Center for Pediatric Oncology, Hematology and Immunology, pediatric endocrinologist of the Ministry of Health

Section "PRIMARY IMMUNODEFICIENCIES”
Moderators: MV Belevtsev, AP Salivonchik

Primary immunodeficiencies in Republic of Belarus. PID &COVID19
Michael Belevtsev

Registers of primary Immunodeficiencies (PID).
Experience of ESID register. S Sharapova

Search for the severe combined immunodeficiency among children, deceased under 1 year E Polyakova

Diagnosis severe combined immunodeficiency in adults E Latysheva
Section "PRIMARY IMMUNODEFICIENCIES: DIFFICULTIES IN DIAGNOSTICS"
Moderators: SO Sharapova, TA Uglova

PID management: still to be improved
N Brodszki

Primary immunodeficiencies with impaired antibody production
TA Uglova

Difficulties in diagnosing CVID in a patient with autoimmune manifestation (a clinical case of a patient with an NFKB1 mutation)
YS Zharankova

Difficulties in diagnosis and therapeutic tactics in patients with autoinflammatory syndrome
S Aleshkevich

Section "PRIMARY IMMUNODEFICIENCIES" (continued)
Moderators: SO Sharapova, TV Shman

Autoinflammatory diseases in children, manifestations, diagnosis, approaches to treatment
Y Stepanovsky

Differential diagnosis of IPEX - syndrome and SCID
A Volokha

Family mediterranean fever in Armenia
V Mukuchyan

DADA2 deficiency in children and adults, manifestations, approaches to diagnosis
S Sharapova

DADA2 family case, different age of manifestation, different genetics, the result of bone marrow transplantation
A Bondarenko

Defects in leukocyte migration in patients with primary immunodeficiency
I Sakovich

Clinical and laboratory features of the diagnosis of selective IgA deficiency
M Shitikova

Round table "PRIMARY IMMUNODEFICIENCIES IN THE REPUBLIC OF BELARUS: PROBLEMS AND THEIR SOLUTION"
Moderators: MV Belevtsev, AP Salivonchik, TA Uglova

Features of early diagnostics of primary immunodeficiencies in the regions of the Republic of Belarus and the city of Minsk
M Belevtsev

Primary immunodeficiencies in adults. Problems and solutions
A Salivonchik

Treatment problems for patients with PID
TA Uglova

Vaccination of PID patients from Covid19
YS Zharankova

The work of the society “Let's save immunity”
IL Dirko

Discussion. Project acceptance resolution
TA Uglova

April 30
**SCHOOL-SEMINAR IN CLINICAL IMMUNOLOGY**
Moderators: MM Zafranskaya, MP Potapnev

Immunological bases of formation tolerance
*M Zafranskaya*

Heterogeneity of T-lymphocytes and their role in immunopathology
*D Nizhegorodova*

Immunity and COVID19
*T Shman*

Cytokine "storm". Causes and consequences
*M Potapnev*

Endocrinopathies and Immunity
*A Solntseva*

Immunity and microbiota
*M Belevtsev*

**SCHOOL-SEMINAR IN CLINICAL IMMUNOLOGY**

**IMMUNOLOGY**
Moderators AE Goncharov, AN Meleshko

Gene therapy: from idea to practical implementation
*V Grinev*

Immunological and clinical efficacy the use of pooled mesenchymal stem cells in the treatment of diseases associated with an excessive immune response
*A Goncharov*

Current trends in antineoplastic vaccinations
*M Stegantseva*

Tumor-mediated immunosuppression as obstacle to immunotherapy
*A Meleshko*

Immunological basis of CAR-T therapy oncohematological diseases
*A Migas*

Discussion

**SUMMARY**

About 100 specialists arrived from different regions of Belarus and from Minsk. Representatives of the Ministry of Health were present. The first day of the conference was devoted to primary immunodeficiencies, the second day was devoted to an educational program in clinical immunology. There were many lecturers from other countries who presented online. Lecturers from Ukraine (A Bondarenko, A Volokha, Y Stepanovskiy) and from Russia (E Latysheva, A Mukhina). The main topics were SCID and discussion of the PID registry. I devoted part of my report to congratulations and information about the J PROJECT.

*Mikhail Belevtsev*
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Four important J PROJECT milestones 2004-2009
OSH, KYRGYZSTAN

Host: Bubusayra Orozbekova
Venue: Webinar
Topic: 3rd international scientific and practical school-conference in the Kyrgyz Republic “Clinical Immunology, Allergology and Infectology”

PROGRAM

The conference in the Kyrgyz Republic "CLINICAL IMMUNOLOGY, ALLERGOLOGY AND INFECTOLOGY"
Date of the first day: April 29, 2021 It starts at 10:00. Time of Bishkek, Kyrgyzstan. (Bishkek's time zone is UTC+6 (GMT+6).
First day: 29.04.2021. It starts at 10:00.
Registration of participants 09.00. to 10.00.
GRAND OPENING OF THE SCHOOL-CONFERENCE
Moderators: Orozbekova B. T.
Welcome speech:
1. Kozhobekov Kudaiberdi Gaparalievich, Rector of Osh State University, Doctor of Mathematical Sciences, Professor
2. Valery Chereshnev, President of the RNOI, Scientific Director of the Institute of Immunology and Physiology of the Ural Branch of the Russian Academy of Sciences, Academician of the Russian Academy of Sciences, Doctor of Medical Sciences, Professor
3. Tuzankina Irina Aleksandrovna, Gen.secretary of the RNOI, PhD, Institute of Immunology and Physiology of the Ural Branch of the Russian Academy of Sciences, ZDN RF, Doctor of Medical Sciences, Professor
4. Zhusupbekova Nurida Erkinbekovna, Head of the Department of
Organization of Medical Care and Public Health of the Ministry of Health of the Kyrgyz Republic, Candidate of Medical Sciences, Associate Professor
5. Zarifyan Anes Gurgenovich, Dean of the Medical Faculty of KRSU, Professor
6. Uzakbayev Kamchybek Askarbekovich, Director of NCOMiD, MD, Professor
7. Usenov Keneshbek Zhumabekovich, Rector of the Zhlala-Abad State University, Doctor of Technical Sciences, Professor. Alybaev Kurmanbek Sarmanovi Vice-rector for Scientific Work
8. Osumbekov Bayyshbek Ziyaidinovich, Vice-rector for Scientific Work of Osh State University, Doctor of Medical Sciences, Professor.

11.00-11.30.
1. Valery Chereshnev, President of the RNOI, Scientific Director of the Institute of Immunology and Physiology of the Ural Branch of the Russian Academy of Sciences, Academician of the Russian Academy of Sciences, Doctor of Medical Sciences, Professor. "Diseases and immunopathology: prospects in the 21st century"

11.30-12.00.
2. Tuzankina Irina Aleksandrovna, General Secretary of the RNOI, Chief Researcher of the Institute of Immunology and Physiology of the Ural Branch of the Russian Academy of Sciences, Honored Scientist of the Russian Federation, Doctor of Medical Sciences, Professor. "Primary immunodeficiency – innate errors of immunity".

12.00-12.30.
3. Mikhail Artemovich Bolkov, Member of the RNOI secretariat, Senior Researcher of the Institute of Immunology and Physiology of the Ural Branch of the Russian Academy of Sciences, Candidate of Medical Sciences. "Classification of innate errors of immunity".

12.30 - 13.00.
4. Olga Fedoruk, manager of the company "Lamira", Minsk, Republic of Belarus. "Modern approaches to the choice of doses and forms of correction of Vit D deficiency".

13.00. - 14.00. Break

14.00-14.30.
5. Irina Pashnina, Head of the clinical and diagnostic laboratory of the State Autonomous Healthcare Institution of the Sverdlovsk Region "Regional Children's Clinical Hospital", Doctor of Biological Sciences. "Organization of laboratory immunological examination: where to start".
6. Deryabina Svetlana Stepanovna, senior scientific. sotr. Institute of Immunology and Physiology of the Ural Branch of the Russian Academy of Sciences, Candidate of Biological Sciences. "Molecular genetic diagnostics of immunopathology"

7. Irina Pashnina, Head of the clinical and diagnostic Laboratory of the Regional Children's Clinical Hospital, Doctor of Biological Sciences. "Laboratory diagnostics of primary immunodeficiency – from simple to complex".

8. Vlasova Elena Viktorovna, Head of the Immunological Department of the Regional Children's Clinical Hospital, Candidate of Medical Sciences. "Rare forms of primary immunodeficiency. Clinical analysis".

9. Baltabaev Mir-Ali Kurbanalievich. (co-author. Baltabaev A.M.) Head of the Department of Dermatovenerology and Phthisiology of KRSU, MD, Professor, (Department of Dermatovenerology and Phthisiology of KRSU, Associate Professor, PhD) Clinical and pathogenetic aspects and improvement of therapy of true acantholytic pemphigus.

10. Maynazarova Elmira Sydykovna, Associate Professor of the Department of Epidemiology and Immunology of the KRSU, Candidate of Medical Sciences. Development of the immunological system in the Kyrgyz Republic.


### Second day: 30.04.2021. It starts at 09:00.

**Moderators:** Orozbekova B. T., Mainazarova E.

<table>
<thead>
<tr>
<th>Time</th>
<th>Session</th>
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<tbody>
<tr>
<td>09.00-09.30</td>
<td>1. Tyumonbaeva Nasira Bektursunovna. researcher at the Institute of Mountain Physiology and Medicine of the National Academy of Sciences of the Kyrgyz Republic, Candidate of Biological Sciences, Regional features of cellular and humoral immunity in residents of various mountain heights of the Kyrgyz Republic.</td>
</tr>
<tr>
<td>09.30 - 09.50</td>
<td>2. Bokonbaeva Syrgajoomartovna, Head of the Department of Pediatrics of KRSU, MD, Professor. Features of the immune response of young children from large families with community-acquired pneumonia</td>
</tr>
<tr>
<td>09.50-10.20</td>
<td>3. Umida Pulatjanovna Nabieva, Head of the laboratory of autoimmune conditions of the Institute of the Institute of Human Immunology and Genomics of the Academy of Sciences of the Republic of Uzbekistan, Associate Professor of the Department of Clinical Allergology, Immunology and Microbiology of the Tashkent Pediatric Medical Institute, MD &quot;Primary immunodeficiency in Uzbekistan&quot;.</td>
</tr>
<tr>
<td>10.20 - 10.50</td>
<td>4. Nurmatov Zuriddin Sharipovich, Head of the Republican Scientific and Practical Center for the Control of Viral infections of the NGO &quot;Preventive Medicine&quot;, MD, PhD, Epidemiological situation of COVID-19 in the Kyrgyz Republic.</td>
</tr>
<tr>
<td>10.50 - 11.20</td>
<td>5. Sulaimanov Shairbek Alibaevich, Department of Pediatrics of the KRSU, Doctor of Medical Sciences, Professor, Clinical, diagnostic and epidemiological characteristics of COVID-19 in the KR.</td>
</tr>
<tr>
<td>11.20 - 11.40</td>
<td>6. Sadyrova Nurgul Adylgazievna, Head of the Department of Pediatrics and Obstetrics of ZHAGU, Candidate of Medical Sciences, Associate Professor. Classification of immunomodulators.</td>
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<tr>
<td>11.40 - 12.00</td>
<td>7. Bubusaira Tolobaevna Orozbekova, Head of the Department of Epidemiology and Immunology of the KRSU, Doctor of Medical Sciences, Professor. The use of Lycopide in athletes.</td>
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<td>12.00. - 13.00 Break</td>
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<td>13.00. - 13.30</td>
<td>8. Gulim Rakhmatovna Saatova, Head of the Department of Cardiorheumatology, NCOMiD, Doctor of Medical Sciences,</td>
</tr>
</tbody>
</table>
Professor. Immunological reactivity in children of Kyrgyz residents

13.30-14.00.
9. Abdyldaev Taalaibek Taabaldievich, head. course of allergology and immunology, KSMIPiPK named after S. B. Daniyarov, Doctor of Medical Sciences, Professor. Prognosis of the prevalence of bronchial asthma in children in the Kyrgyz Republic.

10. Fedoskova Tatyana Germanovna, Head of the Laboratory of Molecular Mechanisms of Allergy Federal State Budgetary Institution of the State Scientific Center "Institute of Immunology" Federal Medical and Biological Agency of Russia, Corresponding Member of the Russian Academy of Sciences, MD, Professor. Repeated respiratory diseases in children. Management tactics. Moscow.

14.30. - 14.50
11. Kazybekova Astra Azhymudunovna. Researcher at the Institute of Mountain Physiology and Medicine of the National Academy of Sciences of the Kyrgyz Republic. Age-related assessment of immune reactivity in residents of mountainous technogenic areas

14.50. - 15.10
12. Kadyralieva Zuurakan Kubanychbekovna, senior lecturer of the Department of Epidemiology and Immunology of KRSU. The immune mechanism of the development of arthritis is complicated by COVID-19.

15.10-15.30
13. Sorombayeva Nadira Orozbekovna, S. B. Daniyarov KSMIPiPK Department of "Public Health with a course of infection control", Candidate of Medical Sciences, Associate Professor.
Uulke Syrbybekovna Asyryanova, Republican Scientific and Practical Center of the NGO "Preventive Medicine", Organization of monitoring of the infection control system in hospitals of the Kyrgyz Republic

15.30. - 15.50
14. Raimkulov Kursanbek Mamasalievich acting associate Professor of the Department of Medical Biology, Genetics and Parasitology of the I. K. Akhunbayev KSMU. candidate of Biological Sciences Vera Sadvakasovna Toigombayeva, Head of the Department of General and Clinical Epidemiology of the I. K. Akhunbayev KSMU, MD, Professor, Invasion of the population of endemic regions of the Kyrgyz Republic by echinococoses

15.50. - 16.10
15. Imankulova Kyyal Turusbekovna postgraduate student of the Department of General and Clinical Epidemiology of the I. K.
SUMMARY

The conference was held with the participation of the Kyrgyz Republic, the Russian Federation, the Republic of Uzbekistan, Hungary, and the United States.

Organizers:
1. Ministry of Health of the Kyrgyz Republic
2. Kyrgyz-Russian Slavic University, Faculty of Medicine, Department of Epidemiology and Immunology
3. I.K. Akhunbaev Kyrgyz State Medical Academy
4. Russian Scientific Society of Immunologists
5. National Center for Maternal and Child Health Protection, Ministry of Health of the Kyrgyz Republic
6. Kyrgyz State Medical Institute for Retraining and Professional Development named after S. B. Daniyarov
7. Osh State University, Faculty of Medicine
8. J Project - International Primary Immunodeficiency Education Project
9. Jalal-Abad State University, Faculty of Medicine
10. ESID - European Society for Primary Immunodeficiencies
The conference was attended by 604 specialists (198 active full-time participants, and 406 as listeners in remote access), including practitioners of healthcare of the Kyrgyz Republic, specialists in the field of cardiology, rheumatology, allergology, immunology, pediatrics, epidemiology, neurology, from various countries and from the different regions of Kyrgyz Republic - Bishkek, Osh, Jalalabad, as well as Talas, Naryn, Chui, Batken and Issyk-Kul regions.

The main directions of the conference: organizational and methodological foundations for the development and implementation of innovative technologies on clinical immunology and primary immunodeficiencies in pediatrics, topical issues of immunopathology, genetics of immunodeficiencies, immune mechanisms of neuroendocrine disorders, improvement of organizational technologies for training personnel in the field of clinical immunology.

Within the J Project symposium, we discussed topical issues of pediatrics and clinical immunology, prevention of the development of immune-dependent pathology of actual infections, primary immunodeficiencies, and auto-inflammatory syndromes.

We discussed the situation with COVID-19, the features of its development and the consequences of the transferred infection, the informativeness of diagnostic methods, the effectiveness of various methods of its treatment, the state and potential of the laboratory services of the region.

Based on the results of the discussion of the reports heard and the holding of meetings of the working groups on clinical immunology and primary immunodeficiencies, the conference participants came to the following decisions:

1. Recognize the work of the conference as successful, well-organized, and productive.
2. To resume the work of the "Society of Immunologists in the Kyrgyz Republic", to convene a general meeting of immunologists.
3. To contribute to the progress of the development of medical science and practice in the Kyrgyz Republic, the continuation of joint work to promote knowledge with the efforts of specialists from different countries.
4. Expand and deepen joint work in the field of science and practice with neighboring countries of the Central Asian region.
5. Conduct the 4th online/full-time school conference on clinical immunology in the Kyrgyz Republic in June 2021 in Jalal-Abad on the
basis of the medical faculty of Jalal-Abad State University to implement the methodological foundations for innovative technologies and practical approach in diagnostics and treatment of primary immunodeficiency in the Kyrgyz Republic.

Orozbekova Bubusaira Tolobaevna, MD, prof., Head of the Department of Epidemiology and Immunology, Kyrgyz-Russian Slavic University

Mainazarova Elmira Sydykavna, Candidate of Medical Sciences, Associate Professor of the Department of Epidemiology and Immunology, Kyrgyz-Russian Slavic University
L→R:

1. Alymkulova Akylbubu Jamalovna, Researcher at the Institute of Cardiology of the Kyrgyz Republic
2. Orozbekova Bubusaira Tolobaevna, MD, prof., Head of the Department of Epidemiology and Immunology, Kyrgyz-Russian Slavic University
3. Mainazarova Elmira Sydykavna, Candidate of Medical Sciences, Associate Professor of the Department of Epidemiology and Immunology, Kyrgyz-Russian Slavic University
4. Kadyralieva Zuurakan Kubanchbekovna, Senior Lecturer, Department of Epidemiology and Immunology, Kyrgyz-Russian Slavic University

Bubusaira Orozbekova
Tbilisi, Georgia

June 5

Host: Karaman Pagava
Venue: Webinar
Topic: IV Scientific Conference on Primary Immunodeficiencies

Scientific-Practical Conference “Child and Adolescent Medicine -XIV”
IV Scientific Conference on Primary Immunodeficiencies
(with international participation)
The conferences are dedicated to 100th Anniversary of the Pediatric Education in Georgia

PROGRAM
Opening of the Conference, Greetings

100th Anniversary of the Pediatric Education in Georgia (Apropos of the first Academic Department of Child Diseases)
Karaman Pagava, Tbilisi

Main problems of COVID-19 infection in children
Manana Khochava, Tbilisi

Multi-system inflammatory syndrome in children in relation to COVID-19 infection
Karaman Pagava, Tbilisi

Autoimmune enteropathies in primary immunodeficiencies
Alla Volokha, Kiev

Modern possibilities of diagnosing primary immunodeficiencies. Experience of the Republic of Belarus
Mikhail Belevtsev, Minsk

Defects of leukocyte adhesion. Diagnostic approaches
Svetlana Sharapova, Minsk

E-Poster session
Usage of Aprinol in children respiratory diseases
Karaman Pagava, Tbilisi

Proportion of frequently ill children and types of diseases among hospitalized pediatric patients
Irma G. Korinteli, Karaman Pagava, Tbilisi

Discussion
SUMMARY

The 4th J Project meeting in Georgia was dedicated not only to the Day of Immunology and the World Primary Immunodeficiency Week but also to the 100th anniversary of pediatric education in the country. The opening presentation was given by K Pagava about the history of child medicine in ancient Georgia (Colkhis & Iberia) and of the establishment of academic pediatric departments in modern Georgia. There were underlined the main scientific achievements, particularly of last decades (bacteriophage therapy, rare diseases, incl. primary immunodeficiencies, fuzzy logic in medicine, antibiotic stewardship, autoimmune and autoinflammatory diseases, adolescent medicine, low intensive laser therapy, interferon therapy) and also significant contribution to the health education and promotion of healthy life style. The changes in clinical and laboratory manifestations of COVID-19 infection in children from beginning of the epidemic until now were discussed by M Khochava, I Shalamberidze, and T Jokhtaberidze. The communication was based on the literature data, but a special attention was paid to the results of clinical analysis of the cases in Georgia. Meta-analysis of publications regarding the multisystem inflammatory syndrome in children (MIS-C) in relation to COVID-19 infection using own cases was presented by K Pagava and IG Korinteli. There was given detailed description of immune enteropathies in primary immunodeficiencies – epidemiology, diagnostic approaches and management by A Volokha. A special attention was given to the treatment – the following approaches were underlined – prevention and treatment of
infections, IVIG, immunomodulation, avoiding of live vaccines, parenteral nutrition, transplantation of stem cells of bone marrow. Current state of immunological and genetic diagnosis of primary immunodeficiencies in Belarus was presented by M Belevtsev. The following topics were underlined - late diagnostics of PID, date of diagnostics and beginning of specific treatment, oncological diseases, autoimmune diseases, COVID-19 and PID, postvaccination reactions, implementation of target therapy, organization of regional centers and educational resources. Differential diagnosis of defects of leukocyte adhesion was presented by S Sharapova. A list of different manifestations suspicious on LAD was given. For diagnostics of LAD the following tests were recommended: investigation of CD18+ and CD15s, definition of leukocyte migration and genetic tests. There were presentation by IG Korinteli and K Pagava on results of open clinical trial on Aprinol in outpatients aged from 1 till 15 years with respiratory diseases. High effectivity of this medicine was confirmed. There was presentation by IG Korinteli and K Pagava on proportion of frequently ill children (0-17 years) in different hospitals of Tbilisi during one year. 103,436 medical histories were analyzed. Re-hospitalization occurred in 25.1% cases, among them 58.7% were hospitalized 3 and more times, 20.4% were treated in the intensive care units. It has been concluded that particularly these patients should be referred for further investigations for PID.

Karaman Pagava

Levan Dvali
JALAL-ABAD, KYRGYZSTAN  
JUNE 17-19

Host: Bubusayra Orozbekova  
Venue: Webinar  
Topic: 3rd international scientific and practical school-conference in the Kyrgyz Republic “Clinical Immunology, Allergology and Infectology”

PROGRAM

17 June
Solemn opening of the school - conference
Moderators: Orozbekova Bubusayra Tolobaevna, Majanazarova Elmira Richikovna

Greetings
Usenov Keneshbek Zhumabekovich, Rector of Jalal-Abad State University, Doctor of Technical Sciences, Professor
Chereshnev Valeriy Alexandrovich, RSI President, Scientific Director of the Institute of Immunology and Physiology of the Ural Branch of the Russian Academy of Sciences, Academician RAS, Doctor of Medical Sciences, Professor
Nifad’ev Vladimir Ivanovich, Rector of the Kyrgyz-Russian Slavic University, Academician of the National Academy of Sciences, Doctor of Technical Sciences, Professor
Kozhobekov Kudyoberdi Gararalievich, Rector of the Osh State University, Doctor of Mathematical Sciences, Professor
Zarifiyan Anez Gurgenovich, Dean of the Medical Faculty of Kyrgyz-Russian Slavic University, Professor

Immunity as the coherent whole
Chereshnev Valery Alexandrovich

Cells of the Immune System, clusters of differentiation, antibodies, cytokines
Bolkov Mikhail Artemovich

Immunogram as a tool for diagnosing various immunopathology
Pashnina Irina Aleksandrovna

COVID-19: problems and solutions
Chereshnev Valery Alexandrovich
Autoimmune manifestations at COVID-19  
*Nabiyeva Umida Puladjanovna*

Basics of genetics and immunopathology  
*Bolkov Mikhail Artemovich*

Laboratory diagnostics of rheumatic diseases: is it always the key to success?  
*Mikhailova Anastasia Sergeevna*

Modern methods for determining autoantibodies for the diagnosis of autoimmune diseases  
*Krivolapova Irina Mikhailovna*

Errors of the pre-analytic stage of laboratory research and ways to eliminate them  
*Pashnina Irina Aleksandrovna*


**18 June**

Moderators:  
Orozbekova Bubusayra Tolobaevna, Majanazarova Elmira Richikovna

Some aspects of pathogenesis, clinics and improving the treatment of peeled pressing forms  
*Baltabaev Mir-Ali Kurban-Alievich*  
(in collaboration with AM Baltabaev)

Immunodeficiency. Congenital immunopathology  
*Tuzankina Irina Aleksandrovna*

Immunological methods for diagnosing primary immunodeficiency  
*Pashnina Irina Aleksandrovna*

Methods of diagnosis and treatment of primary immunodeficiency.  
*Tuzankina Irina Aleksandrovna*

Relationships of clinical doctors and a genetic laboratory: tasks and opportunities  
*Deryabina Svetlana Stepanovna*

Hereditary angioedema in children  
*Tuzankina Irina Aleksandrovna*

Humoral immunity defects on the clinical examples of agammaglobulinemia  
*Vlasova Elena Viktorovna*

Genetic diagnosis of X-linked agammaglobulinemia on clinical examples  
*Cheremokhin Dmitriy Andreevich* (in collaboration with SS Deryabina)

Rare clearly defined primary immunodeficiency syndromes (ATA, Nijmegen, DiGeorge syndromes)  
*Vlasova Elena Viktorovna*

Primary immunodeficiency with defects of immunoregulation (APECED, IPEX and IPEX-like syndromes)  
*Bolkov Mikhail Artemovich*

Autoinflammatory syndromes  
*Bolkov Mikhail Artemovich*

Using of Likopid at the athletes  
*Orozbekova Bubusayra Tolobaevna*

Problems of patients with primary immunodeficiency  
*Berseneva Natalia Nikolaevna*

Discussions

**19 June**

Moderators:  
Orozbekova Bubusayra Tolobaevna, Majanazarova Elmira Richikovna
Modern approaches to choosing doses and forms of correction of the deficit of Vitamin D
Zuridinova Aida Ashhelievna

Atopic dermatitis and allergies in children
Bass Evgenia Aronovna

Modern methods of diagnosis and therapy of allergic diseases
Sulayimanov Shayirbek Alibaevich

The forecast of the prevalence of bronchial asthma in Kyrgyzstan and bronchial asthma clinical protocols
Abdyldaev Talaybek Tabaldievich

Epidemiological Features The prevalence of COVID-19 in the Kyrgyz Republic
Nurmatov Zurdin Sharipovich

Structure, the dynamics of allergic diseases in the Kyrgyz Republic
Asheraliev Mukhtar Esenzhanovich

Repeated respiratory diseases in children. Tactics of reference
Fedoskova Tatiana Germanovna

Immunomodulating therapy in gynecology: probable and evidence
Stakeev Cholpon Askarovna

Study of the incidence of enterobiosis in samples of children with allergic diseases in the Kyrgyz Republic
Arinbayev Beckbolot Saparbekovich

Molecularly gene characteristic of a viral hepatitis Delta in the Kyrgyz Republic
Nogoybaeva CalysByubyu Asanbekovna

Cytokine status in chronic heart failure ischemic genesis with a reduced ejection fraction in elderly and senile people
Tsai Larisa Grigorievna

Discussions

SUMMARY

From June 17 to 19, 2021, the IV Visiting International Scientific and Practical School-Conference in the Kyrgyz Republic "Clinical Immunology, Allergology and Infectology" was held in Jalal-Abad (Kyrgyzstan). The organizers of this event were:
1. Ministry of Health of the Kyrgyz Republic
2. Kyrgyz-Russian Slavic University, Faculty of Medicine, Department of Epidemiology and Immunology
3. Kyrgyz State Medical Academy named after I. K. Akhunbayev
4. Russian Scientific Society of Immunologists
5. National Center for Maternal and Child Health, Ministry of Health of the Kyrgyz Republic
6. Kyrgyz State Medical Institute of Retraining and Advanced Training named after S. B. Daniyarov
7. Jalal-Abad State University, Faculty of Medicine
8. Osh State University, Faculty of Medicine
9. J Project-International educational project on primary immunodeficiency
10. ESID-European Society for Primary Immunodeficiency
11. Institute of Immunology and Physiology of the Ural Branch of the Russian Academy of Sciences

Various issues of fundamental and clinical immunology were covered, including problems of infectious, allergic, autoimmune and auto-inflammatory pathology. A separate series of reports supported by ESID and JProject was devoted to the problem of diagnosis and treatment of primary immunodeficiency or innate errors of immunity. Reports on the epidemiological situation in the Kyrgyz Republic were also presented.

The school conference was held in a face-to-face and online format with a broadcast via the Internet and the opportunity to ask a question and get an answer. The conference was attended by doctors and scientists from Kyrgyzstan, the Russian Federation, Uzbekistan and Kazakhstan. The conference was attended by 245 specialists (98 active participants and 147 remote access listeners).

Bubusaira Orozbekova
1. Sadyrova Nurgul Adylgazievna, Ph.D., Head of the Department of Pediatrics and Obstetrics-Gynecology, Jalal-Abad State University named after B.Osmonov

2. Orozbekova Bubusaira Tolobaevna, Doctor of medical science, Ph.D., prof., Head of department of epidemiology and immunology, Kyrgyz-Russian Slavic University named after B. N. Yeltsin


4. Alybaev Kurmanbek Sarmanovich, doctor of physical and mathematical sciences, prof., Vice-Rector for Science of Jalal-Abad State University named after Osmonov
1. **Mainazarova Elmira Sydykovna**, PhD, Associate Professor of the Department of epidemiology and immunology, Kyrgyz-Russian Slavic University named after B. N. Yeltsin

2. **Orozbekova Bubusaira Tolobaevna**, Doctor of medical science, Ph.D., prof., Head of department of epidemiology and immunology, Kyrgyz-Russian Slavic University named after B. N. Yeltsin
Sadyrova Nurgul Adylgazievna, Ph.D., Head of the Department of Pediatrics and Obstetrics-Gynecology, Jalal-Abad State University named after B.Osmonov

1. Tumonbaeva Nasira, Ph.D., Associate Professor, Senior Researcher at Department of Immunology and Physiology, National Academy of Sciences of the Kyrgyz Republic

2. Mainazarova Elmira Sydykovna, PhD, Associate Professor of the Department of epidemiology and immunology, Kyrgyz-Russian Slavic University named after B. N. Yeltsin

3. Orozbekova Bubusaira Tolobaevna, Doctor of medical science, Ph.D., prof., Head of department of epidemiology and immunology, Kyrgyz-Russian Slavic University named after B. N. Yeltsin

4. Bolkov Mikhail Artemovich, PhD, senior researcher at Institute of immunology and physyology UB RAS (Ekaterinburg, Russia).

5. Vershinina Irina Leopoldovna, physical therapy and sports medicine doctor (Ekaterinburg)

6. Alymkulova Akylbubu Zhamalovna, a researcher at the National Center for Cardiology and Therapy named after academician Mirsaid Mirrakhimov under the Ministry of Health of the Kyrgyz Republic

7. Coordinator at the Jalal-Abad State University named after B.Osmonov
# J PROJECT MEETINGS 2004-June 2021

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<th>Year</th>
<th>City (Country)</th>
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<td>2004</td>
<td>1. Targu Mures, Romania</td>
<td>Csilla Todea</td>
<td>March 11-12</td>
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<td></td>
<td>2. Prague, Czech Republic</td>
<td>Anna Sediva</td>
<td>May 10-11</td>
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<td>3. Belgrade, Serbia/Montenegro</td>
<td>Srdjan Pasic</td>
<td>June 11-12</td>
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<td>4. Skopje, Macedonia</td>
<td>Katarina Stavrik</td>
<td>September 17-18</td>
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<td>5. Kyiv, Ukraine</td>
<td>Alla Volokha</td>
<td>November 18-19</td>
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<td>6. Zakopane, Poland</td>
<td>Ewa Bernatowska</td>
<td>December 16-17</td>
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<tr>
<td>2005</td>
<td>7. Sofia, Bulgaria</td>
<td>Elissaveta Naumova</td>
<td>April 15-16</td>
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<td>8. Prague, Czech Republic</td>
<td>Anna Sediva</td>
<td>May 9-10</td>
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<td>9. Gdansk, Poland</td>
<td>Ewa Bernatowska</td>
<td>September 23-24</td>
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<td>10. Debrecen, Hungary</td>
<td>László Maródi</td>
<td>November 4-5</td>
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<td>11. Lviv, Ukraine</td>
<td>Larysa Kostyuchenko</td>
<td>November 25</td>
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<td>12. Oradea, Romania</td>
<td>Zoltan Ellenes</td>
<td>December 20</td>
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<td>14. Prague, Czech Republic</td>
<td>Anna Sediva</td>
<td>May 8-9</td>
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<td>15. Bucharest, Romania</td>
<td>Nicolae Iagaru</td>
<td>June 9-10</td>
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<td>16. Minsk, Belarus</td>
<td>Michael Belevtsev</td>
<td>November 9-10</td>
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<td></td>
<td>17. Debrecen, Hungary</td>
<td>László Maródi</td>
<td>December 8-9</td>
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<tr>
<td>2007</td>
<td>18. Zaporozhzhye, Ukraine</td>
<td>Lyudmila Chernyshova</td>
<td>April 19-20</td>
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<td>19. St. Petersburg, Russia</td>
<td>Marina Guseva</td>
<td>May 29-30</td>
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<td>20. Ohrid, Macedonia</td>
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<td>Aurica Rugina</td>
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<td>Lyudmila Chernyshova</td>
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<td>24. Sunny Beach, Bulgaria</td>
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<td>25. Sarajevo, Bosnia-Herzegovina</td>
<td>Velma Mulaosmanovic</td>
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<td>27. Riga, Latvia</td>
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<td>29. Sevastopol, Ukraine</td>
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<td>Marina Guseva</td>
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<td>Ismail Reisli</td>
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<td>33. Krakow, Poland</td>
<td>Danuta Kowalczyk</td>
<td>September 24-25</td>
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<td>34. Tallinn, Estonia</td>
<td>Sirje Velbri</td>
<td>October 7-8</td>
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<td>35. Ljubljana, Slovenia</td>
<td>Nataša Toplak</td>
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<td>Nermeen Galal</td>
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<td>Michael Belevtsev</td>
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<td>László Maródi.</td>
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<td>Nima Rezaei</td>
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<td>May 12</td>
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<td>Prague, Czech Republic</td>
<td>Anna Sediva</td>
<td>May 14-15</td>
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<td>Panteley Popandopulo</td>
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<td>Irina Tuzankina</td>
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<td>Mihaela Bataneant</td>
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<td>Jiri Litzman</td>
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<td>Zsuzsa Horváth</td>
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145. Kaliningrad, Russia Irina Tuzankina June 27-30
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151. Konya, Turkey Ismail Reisli April 18
152. Kropivnitskyi, Ukraine Liudmyla Chernyshova April 20-21
153. Tehran, Iran Nima Rezaei April 22-29
154. Kaunas, Lithuania Brigita Sitkauksiene April 26
155. Bratislava, Slovakia Peter Ciznar May 3
156. Eskisehir, Turkey Ismail Reisli May 5
157. Ljubljana, Slovenia Tadej Avcin May 13
158. Baku, Azerbaijan Gulnara Nasrullahova June 1
159. Warsaw, Poland Ewa Bernatowska June 7
160. Tsigov Chark, Bulgaria Marianna Murdjeva June 16-18
161. Chelyabinsk, Russia Irina Tuzankina August 27
162. Sofia, Bulgaria Elissaveta Naumova September 9
163. **Edinburgh, UK 9th SC Meeting**
László Maródi September 12
& Katalin Vámos
164. Budapest, Hungary László Maródi September 22
165. Astana City, Kazakhstan Elena Kovzel October 6-7
166. Olomouc, Czech Republic Anna Sediva October 10
167. Van, Turkey Ismail Reisli October 13-14
168. Moscow, Russia Irina Kondratenko October 25
169. Chisinau, Rep. of Moldova Svetlana Sciucu November 3
170. Pristina, Kosovo Vlora Ismaili-Jaha December 16

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171. **Budapest, Hungary 10th SC Meeting**
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173. Minsk, Belarus Michael Belevtsev April 19-21
174. Tehran, Iran Nima Rezaei April 21-23
175. Bratislava, Slovakia Peter Ciznar April 25
176. St. Petersburg, Russia Marina Guseva April 26
177. Almaty, Kazakhstan Zhanat Ispayeva April 26-27
178. Kiev, Ukraine Liudmyla Chernishova April 26-27
& Alla Volokha
179. Kaunas, Lithuania Brigita Sitkauksiene May 2
180. Batman, Turkey Ismail Reisli May 3
181. Ljubljana, Slovenia Tadej Avcin May 5
182. Warsaw, Poland Ewa Bernatowska May 10-11
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248. Debrecen Hungary Noémi Miltner June 10
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255. Chisinau R. of Moldova Svetlana Scuica September 8
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Dushanbe Tajikistan Irina Tuzankina September 15
257. Skopje N. Macedonia Kristina Mironska September 24
258. Vladivostok Russia Galina Tcyvkina September 24
259. Nakhoda Russia Galina Tcyvkina September 28
260. Konya Turkey Ismail Reisli September 29
261. Kiev Ukraine Alla Volokha October 15-16
262. Skopje N. Macedonia Kristina Mironska October 17
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275. Yerevan Armenia Sevan Iritsyan November 27
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276. Warsaw Poland Malgorzata Pac November 27
& Ewa Bernatowska
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<td>Slovakia</td>
<td>Peter Ciznar</td>
<td>April 28</td>
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<td>April 29</td>
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<td>Bubusayra Orozbekova</td>
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<td>Georgia</td>
<td>Karaman Pagava</td>
<td>June 5</td>
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