

THE J PROJECT



EDITORS:

László Maródi & Katalin Vámos

with the contribution of

*Asghar Aghamohammadi, Mónika Apáti Nagy, Tadej Avcin, Ewa Bernatowska,
Peter Ciznar, Liudmila Cerempei, Liudmyla Chernyshova, Maria Guseva,
Demet Hafizoglu, Sorin Iurian, Tatiana Nasyrina, Elissaveta Naumova, Ismail Reisli,
Nima Rezaei, Anna Sediva, Irina Tuzankina, Dilara Kocacik Uygun*

December, 2014

Contents

Foreword	3
ECE IPI CTR	4
J Project Meetings in 2014	5
Antalya, Turkey	6
Antalya, Turkey	11
Zagazig, Egypt	13
Tehran, Iran	14
Poltava, Ukraine	16
Bratislava, Slovakia	18
Debrecen, Hungary	19
St. Petersburg, Russia	20
Varna, Bulgaria	21
Catez, Slovenia	22
Tehran, Iran	24
Yekaterinburg, Russia	26
Warsaw, Poland	29
Pardubice, Czech Republic	31
Nizhny Novgorod, Russia	32
Sibiu, Romania	33
Erzurum, Turkey	35
Chisinau, Moldova	36
J Project Meetings 2004–2014	39
J Project Steering Committee members	42
Genetic testing in the Debrecen PID Center	44
Links	46



*The PID team in Debrecen. L → R: Melinda Erdős,
Gabi Csorba, Beáta Soltész, Vera Gulácsy, Adrien Sarkadi,
László Maródi, Beáta Tóth, Szilvia Taskó, Zsuzsa Pistár*

The 100th J Project Meeting and the Antalya Declaration

All those who have played a role in the remarkable success and fascinating professional aspects of the J Project over the last 10 years must consider themselves fortunate to have been caught up in the spirit of the project and a love of the primary immunodeficiency (PID) field, and to have been involved in the care of patients suffering from inborn errors of immunity. The organization of 100 J Project meetings, to educate physicians and scientists and to facilitate exchanges between us to improve our understanding of PIDs was little more than a dream 10 years ago. The genetic analysis of 1000 DNA samples at the Debrecen PID Center was also inconceivable. Nevertheless, all these pipe dreams had become a reality by the time of the Antalya meeting in Turkey, in March 2014, when we celebrated the 10th birthday of the Project.

We were honored to cohost this jubilee meeting with Ismail Reisli, providing a forum for discussions about our achievements over the last 10 years in ECE and parts of Asia and Egypt. We also discussed the current situation and the paths we should take to improve PID patient care further and to advance research in the near future in this large area of Europe and the extended area covered by the J Daughters Project.

The Antalya Meeting was attended by more than 300 participants, 250 of whom were from J Project countries. We also had 30 invited speakers from Western countries including Jean-Laurent Casanova the keynote speaker of the Meeting. This provided an excellent forum for learning more about basic and clinical aspects of PIDs. The program included 65 oral presentations and 153 posters, 218 presentations in all, with almost all the professional participants presenting at least a case report poster. I think this provides a clear indication of the remarkable level of clinical research activity in the J Project area and highlights the strong development of PID patient care in the region.

The Antalya Declaration was accepted as a minimum requirement for PID patient care in J Project countries and beyond, as a means of finding new strategies and approaches to achieving the original goals of the J Project. This declaration focuses on five critical areas for further development to improve PID care, with a harmonization of standards and practices in ECE countries and elsewhere (see below).

We thank Biotest, not only for providing platinum sponsorship, but also for continual support of the J Project program over the last 10 years. We also thank our gold sponsors, Baxter and Octapharma Nordic for their generous support, and other sponsors, including Kedrion, for their contribution to the success of this meeting.

Last, but not least, we thank all those who have contributed to the success of the J Project by organizing one or more of the 100 J Project Meetings, taking part in our clinical research activity, or serving on the JP Steering Committee over the last 10 years.

László Maródi



University of Debrecen, Clinical Center

The **East-Central-European Infectious and Pediatric Immunology Center for Training and Research (ECE IPI CTR)** was established on December 22, 1999. From educational and scientific points of view, the Center is closely associated with the **Department of Infectious and Pediatric Immunology** at the **University of Debrecen**. The Department has a strong profile of immunology including research into antimicrobial host defense mechanisms. The main fields of research activity of the Department are as follows:

- Molecular pathology of primary immune deficiency diseases
- Host defense mechanisms against bacteria and fungi
- Mechanisms of action of intravenous immunoglobulin

The Center provides modern educational utilities, demonstrational materials, and rooms that are suitable for practical seminars as well as lecture room. ECE IPI CTR functions as an institutional and regional infectious and pediatric immunology center. Its essential role is the initiation of regular postgraduate courses for specialists in infectious diseases and immunology in countries of East Europe.

From the conception aspects, ECE IPI CTR is tightly connected with the **Interregional Association of the Carpathian Euro-region**, one of the main aims and functions of which is the formulation of educational and scientific co-operation within the region of Eastern and Central Europe.

J Project Meetings in 2014

City (<i>Country</i>)	Date
1. Antalya (<i>Turkey</i>)	March 12–14
2. Antalya (<i>Turkey</i>)	March 15
3. Zagazig (<i>Egypt</i>)	April 17
4. Tehran (<i>Iran</i>)	April 22–29
5. Poltava (<i>Ukraine</i>)	April 24–25
6. Bratislava (<i>Slovakia</i>)	April 25
7. Debrecen (<i>Hungary</i>)	April 26
8. St. Petersburg (<i>Russia</i>)	May 28
9. Varna (<i>Bulgaria</i>)	October 6
10. Catez (<i>Slovenia</i>)	October 11–12
11. Tehran (<i>Iran</i>)	October 18–19
12. Yekaterinburg (<i>Russia</i>)	October 24–25
13. Warsaw (<i>Poland</i>)	October 24–25
14. Pardubice (<i>Czech Republic</i>)	November 11
15. Nizhny Novgorod (<i>Russia</i>)	November 26
16. Sibiu (<i>Romania</i>)	November 27–28
17. Erzurum (<i>Turkey</i>)	December 6
18. Chisinau (<i>Rep. of Moldova</i>)	December 12



Szarvas

Hosts: László Maródi (*lmarodi@med.unideb.hu*),

Ismail Reisli (*ireisli@hotmail.com*)

Venue: Rixos Lares Hotel, Antalya

PROGRAM

March 12

Laboratory diagnostic approaches to primary B cell deficiencies

Mary Ellen Conley, (Memphis, USA)

Specific antibody deficiencies with normal immunoglobulins

Ricardo Sorensen, (New Orleans, USA)

Recent advances in understanding immunoglobulin class switch deficiencies

Anne Durandy, (Paris, France)

Update on CVID and IgA deficiency

Lennart Hammarström, (Stockholm, Sweden)

Current understanding of immunopathology of XLP

Stuart Tangye, (Sydney, Australia)

Pathophysiology of and molecular genetics of periodic fever syndromes

Tadej Avčin, (Ljubljana, Slovenia)

Conventional and novel genetic approaches to PIDs

Hans Ochs, (Seattle, USA)

Laboratory evaluation of TLR-mediated immunity

Xavier Bossuyt, (Leuven, Belgium)

PID patients with neutropenia: differential diagnostic algorithms

Nima Rezaei, (Tehran, Iran)

Novel adhesion protein deficiencies

Amos Etzioni, (Haifa, Israel)

Genetic forms of Omenn syndrome

Anna Villa, (Milan, Italy)

BMT/HSCT: a success story of treating PID patients

Andrew Cant, (Newcastle upon Tyne, UK)



Life-threatening infectious diseases of childhood: single-gene inborn errors of immunity

Jean-Laurent Casanova, (Paris-New York, France-USA)

March 13

Patients' needs and wants

Jose Drabwell, (London, UK)

Established treatment options – the good and bad of IVIG and SCIG

Jiří Litzman, Brno, (Czech Republic)

Potential new options in IG delivery: facilitated subcutaneous IG therapy

Stephen Jolles, (Cardiff, UK)

Novel agammaglobulinemia genotype

Mary Ellen Conley, (Memphis, USA)

Activated phosphoinositide 3-kinase δ syndrome

Sergey Nejentsev, (Cambridge, UK)

- Gain-of-function mutations in PIK3CD and senescent T cells
Gulbu Üzel, (Bethesda, USA)
- Immunodeficiency due to CD19 mutations
İsmail Reisli, (Konya, Turkey)
- Clinical and molecular characteristics of 200 Iranian CVID patients
Asghar Aghamohammadi, (Tehran, Iran)
- Class switch recombination defects and unique cases of CD40 and CD40L deficiencies
Necil Kütükçüler, (Izmir, Turkey)
- Progressive degenerative damage of the central nervous system in X-linked agammaglobulinemia patients
Irina Tuzankina, (Ekaterinburg, Russia)
- Identification of novel B cell primary immunodeficiencies associated with autoimmunity
Kaan Boztug, (Vienna, Austria)
- IVIG/SCIG replacement: the golden standard of treatment
Vincent R Bonagura, (New York, NY)
- Chances and limitations of Ig replacement in the care of patients with CVID
Klaus Warnatz, (Freiburg, Germany)
- IVIG treatment of patients with CVID and agammaglobulinemia
Tatiana Latysheva, (Moscow, Russia)
- GWAS analyses in CVID patients
Reinhold Schmidt, (Hanover, Germany)
- Clinical and immunological efficacy of SCIG treatment of children and adult
Anna Szaflarska, (Cracow, Poland)
- Clinical study of a novel 10% intravenous immunoglobulin preparation
Gergely Kriván, (Budapest, Hungary)
- Molecular defects underlying impaired humoral immune responses in primary immunodeficiencies
Stuart Tangye, (Sydney, Australia)
- Nijmegen Breakage syndrome: long-term outcome and treatment
Beata Wolska-Kuśnierz, (Warsaw, Poland)
- CVID – from T cell dysfunction to B cell disorder
Elissaveta Naumova, (Sofia, Bulgaria)
- Moreau strain BCG disease in 60 SCID and other Polish patients – 30 years of experience
Ewa Bernatowska, Małgorzata Pac, (Warsaw, Poland)
- SCID before and after neonatal BCG vaccination in Slovakia
Peter Čížnár, (Bratislava, Slovakia)
- T and B cell development in DiGeorge syndrome
Adam Klocperk, (Prague, Czech Republic)
- How should we treat patients with TAP deficiency
Figen Doğu, (Ankara, Turkey)
- CD25 deficiency: Clinical and laboratory features
Aydan İkinçioğulları, (Ankara, Turkey)
- PID network research development in Latin-America
Ricardo Sorensen, (New Orleans, LA, USA)
- The “A” Project research collaboration program in Africa
Ahmed Aziz Bousfiha, (Casablanca, Morocco)
- Opportunities for finding PID in the HIV and Tb epidemic
Monika Esser, (Cape Town, South Africa)
- PID care and research in Saudi-Arabia
Hamoud Al-Mousa, (Riyadh, Saudi Arabia)
- PID as an evolving area of medical research in India
Surjit Singh, (Chandigarh, India)
- Exome sequencing in the Asian PID Network
Yu-Lung Lau, (Hong Kong, China)
- Developing PID care in Central-Asia: The J Central Asia Project
Elena Kovzel, (Astana City, Kazakhstan)

Primary immunodeficiency in Malaysia:
Present status and future direction
Lokman M Noh, (Kuala Lumpur, Malaysia)

March 14

DOCK8 regulation of skin immunity
Helen Su, (Bethesda, MD, USA)

Aicardi-Goutières syndrome
Yanick J Crow, (Manchester, UK)

Primary OX40 deficiency and genetic etiologies of Kaposi sarcoma
Minji Byun, (New York, NY, USA)

Novel STAT1 GOF mutations in ECE patients with CMCD
Beáta Soltész, (Debrecen, Hungary)

Different severity of phenotypes in patients with STAT1 gain-of-function mutations
Alla Volokha, (Kiev, Ukraine)

The effect of GOF-STAT1 mutation on STAT3 function
Desa Lilic, (Newcastle upon Tyne, UK)

Anti-IL-17A antibodies, may be correlated with the severity of mucocutaneous candidiasis in APECED patients
Adrien Sarkadi, (Debrecen, Hungary)

Regulatory T cells in a family with IPEX-like manifestations
Dalia Abd Elaziz, (Cairo, Egypt)

Hyperimmunoglobulinemia D syndrome in two children with periodic fever
Georgina Kuli-Lito, (Tirana, Albania)

STAT2 deficiency

Sophie Hambleton, (Newcastle upon Tyne, UK)

Challenges of HSCT to treat patients with phagocytic cell defects

Reinhard Seger, (Zurich, Switzerland)

Chronic mild neutropenia revealing MonoMac syndrome

Mihaela Bataneant, (Timisoara, Romania)

Lung and brain aspergillosis in a patient with AR CGD

Anastasia Bondarenko (Kiev, Ukraine)

HIES genotypes and phenotypes

Bodo Grimbacher, (Freiburg, Germany)

HSCT for severe autoimmune and auto-inflammatory syndromes in children

Mario Abinun, (Newcastle upon Tyne, UK)

Novel and recurrent WASP mutations in Eastern and Central European patients with WAS

Vera Gulácsy, (Debrecen, Hungary)

Thrombopoietin therapy in WAS

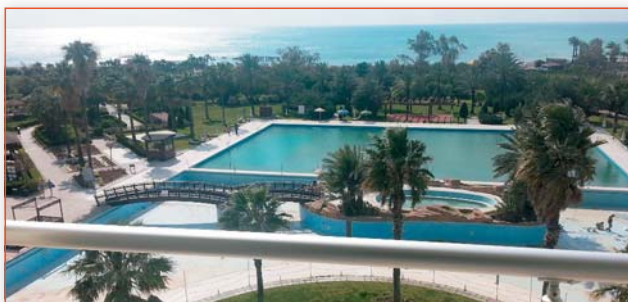
Anna Shcherbina, (Moscow, Russia)

Characterizations of CGD patients in Turkey

Yavuz Köker, (Kayseri, Turkey)

Mitochondrial pathology in autoinflammatory disorders

Anna Šediva, (Prague, Czech Republic)



Park of Rixos Lares



Najwa Elbashir, László Maródi and Nahla Erwa



*Marina Guseva, László Maródi
and Beáta Tóth*



*Ismail Reisli, László Maródi,
and Sükrü Güner*



*László Maródi, Jose Drabwell,
Martine Pergent,
Peter Spath, Dragana Koruga*



*Svetlana Sharapova, Mikhail Belevtsev,
Andrey Salivonchi*



*Sükrü Güner, Surjit Singh, Ismail Reisli,
Asghar Aghamohammadi, Nima Rezai,
Hassan Abolhassani*

THE ANTALYA DECLARATION

Primary immunodeficiency (PID) disorders constitute a major and growing health problem in individuals of all ages, worldwide. They may lead to severe, life-threatening infections and death in early childhood or prolonged, recurrent infections beginning at any age. These conditions affect at least 1 in 1,000 individuals, in all countries. Over the past 10 years, much has been accomplished in Eastern and Central Europe (ECE), under the auspices of the J PROJECT program, which aims to increase awareness of these conditions among physicians and to improve medical training in this domain. However,



*The Presidency of the Antalya Declaration:
Ismail Reisli, Jose Drabwell,
Amos Etzioni, Jean-Laurent Casanova,
László Maródi*

much remains to be done if we are to achieve the goals outlined in 2004. The last decade has seen an unprecedented increase in our knowledge of the clinical, immunological and genetic features of PIDs. The number of known PIDs now exceeds 250, and more than 200 PID disease-causing genes have been described. The PID expert representatives of the J PROJECT met in Lara, Antalya, Turkey on 12–15 March, 2014. They unanimously agreed on the following recommendations and urged their presentation and approval for implementation in ECE countries and elsewhere, throughout the world. National governments and health departments should take responsibility for supporting healthcare providers, thereby facilitating the diagnosis and treatment of PID disorders early in life, to decrease the heavy burden of disease and mortality rates. Specific plans and programs for the prevention, diagnosis and treatment of PID disorders and their infectious and noninfectious complications, such as cancer, inflammatory and autoimmune diseases and allergy, should be formulated at national and regional levels. Such programs should be implemented through partnerships between professional organizations, patient groups and healthcare providers. The Antalya Declaration calls for prompt and effective action, to ensure the application of current knowledge to the diagnosis and treatment of PID patients, so as to improve the quality of life of these patients considerably. To this end, the following five objectives have been targeted for attainment by the end of 2015:

1. **EDUCATION** of medical students and healthcare professionals should be improved, with the inclusion in the curriculum of PID diseases as an emerging and increasingly important field of clinical medicine. Continuing medical education is also required for physicians practicing in primary care settings and hospitals.
2. **PUBLIC AWARENESS** should be raised to publicize the fact that most PID patients remain undiagnosed and untreated.
3. **PID GENETIC DIAGNOSTICS** should be managed free of charge in each of the J Project countries.
4. **A PID REGISTER** should be established in every J Project country and following diagnosis, data for all PID patients should be entered into national and international databases.

5. **PERSONALIZED IMMUNOGLOBULIN REPLACEMENT and HEMATOPOIETIC STEM CELL TRANSPLANTATION** should be made available to PID patients, and should not be restricted by funding or infrastructure constraints.

To help to achieve these goals, PID research collaboration should receive preferential support from national funding agencies or bodies. These recommendations should be endorsed by national health departments, PID-related professional societies and patient groups. All the points outlined above should result in measurable reduction of co-morbidity and mortality of PID patients. They should also result in the lag between the first manifestation and definitive diagnosis of PID being less than one year for all patients. **The Antalya Declaration calls for urgent and decisive action and should be signed by medical or regulatory bodies to support this declaration and goals written herein.**

ANTALYA, TURKEY

March 15

Host: Ismail Reisli (*ireisli@hotmail.com*)

Venue: Hotel Rixos Lares



Ismail Reisli (left) opening the 101th J Project meeting and paving the future right next day after closing the Jubilee 100th J Project Meeting. The participants were mostly from Turkey.

L→R, László Maródi, Zafer Caliskaner, Fazil Orhan. Speaker, Safa Baris

PROGRAM

■ 8.30–8.45

AÇILIŞ

Prof.Dr. İsmail REİSLİ

BİRİNCİ OTURUM

OTURUM BAŞKANLARI

Prof. Dr. Güzide AKSU

Prof. Dr. Mustafa YILMAZ

■ 8.45–9.05

AĞIR KOMBİNE İMMÜN YETMEZLİK: Tanıda geç kalınca...

Doç. Dr. Safa BARIŞ

Marmara Üniversitesi Eğ. ve Araş. Hast. Pediatrik Allerji ve İmmünoloji Bölümü

■ 9.05–9.25

AĞIR KOMBİNE İMMÜN YETMEZLİK: Erken tanının sağladıkları...

Doç. Dr. Deniz ÇAĞDAŞ AYVAZ

Hacettepe Üniversitesi Tıp Fakültesi Pediatrik İmmünoloji BD

■ 9.25–9.45

KRONİK GRANÜLOMATÖZ HASTALIK

Doç. Dr. Hasibe ARTAÇ

Selçuk Üniversitesi Tıp Fakültesi Pediatrik İmmünoloji BD

■ 9.45–10.05

HİPERİMMÜNGLOBULİN E SENDROMU

Uzm. Dr. Esra HAZAR SAYAR

Yrd. Doç. Dr. Şükrü Nail GÜLER

Necmettin Erbakan Üniversitesi Meram Tıp Fakültesi Pediatrik İmmünoloji ve Allerji BD

■ 10.05–10.25

KAHVE ARASI

İKİNCİ OTURUM

OTURUM BAŞKANLARI

Prof. Dr. Uğur ÖZBEK

Prof. Dr. Uğur MUŞABAK

■ 10.25–10.45

ERİŞKİN HASTADA YAYGIN DEĞİŞKEN İMMÜN YETMEZLİK

Uzm. Dr. Fevzi DEMİREL

Doç. Dr. Mustafa GÜLEÇ

GATA Gülhane Tıp Fakültesi İmmünoloji ve Allerji Hastalıkları BD

■ 10.45–11.05

ERİŞKİN HASTADA SELEKTİF IgA VE C3 EKSİKLİĞİ

Uzm. Dr. Şevket ARSLAN

Prof. Dr. A. Zafer ÇALIŞKANER

Necmettin Erbakan Üniversitesi Meram Tıp Fakültesi İmmünoloji ve Allerji Hastalıkları BD

■ 11.05–11.25

NORMAL İMMÜNGLOBULİN DÜZEYLİ SPESİFİK ANTİKOR EKSİKLİĞİ

Doç. Dr. Nesrin GÜLEZ

Doktor Behçet Uz Eğ. ve Araş. Hastanesi

■ 11.25–11.45

MOLECULAR DIAGNOSTIC FOR PRIMARY IMMUNODEFICIENCIES IN TURKEY

Yrd. Doç. Dr. Yuk Yin NG

Dep. of Genetics Institute for Exp. Medicine (DETAE) Istanbul University

Dep. of Genetics and Bioengineering, Istanbul Bilgi University

■ 11.45–12.00

KAPANIŞ VE ÖĞLE YEMEĞİ

Host: Mohammed Almalky (*mohammed_malky@yahoo.com*)

Venue: Zagazig University, Zagazig

PROGRAM

What is PID?

Aisha Almarsafy, (Cairo University)

Lab in PID Present and future

Safaa Meshael, (Cairo University)

Case (1) Agammaglobulinamia

Nermeen Galal, (Cairo University)

Case (2) Ataxia Telengactesia

Sanaa Mahmod, (Zagazig University)

Case (3) CGD

Mayada Sabry, (Mansora University)

Case (4) HIES

Aisha Almarsafy, (Cairo University)

Case (5) CGD

Usama Elsafy, (Zagazig University)

Case (6) LADI

Dalia Salah, (Cairo University)

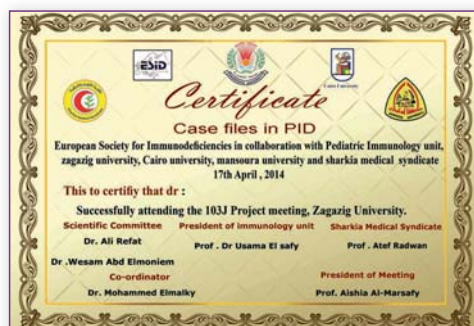
Case (7) Undiagnosed cases in PID

Mohamed Almalky, (Zagazig University)

SUMMARY

This J-Project Meeting was organized in Zagazig University – Egypt on Thursday 17th of April as an early event of PID week in collaboration with Cairo University, Mansoura University and our local Medical Syndicate. The Meeting was organized by the Department of Pediatric Hematology and Immunology, Zagazig University. The meeting was attended by about 180 physicians including house officers, general practitioners, pediatricians, laboratory doctors, dermatologists. and there were speakers from Cairo, Mansoura and Zagazig Universities. The program was directed to increase the awareness on PID among general practitioners and pediatricians by learning the basic clinical signs and symptoms of most common disorders and to present some of these cases during the meeting. The speakers were from three centers of PID from three different Universities Zagazig, Cairo and Mansoura. Discussion was organized among immunologists through interactive communication.

Mohamed Almalky



Hosts: Nima Rezaei (*rezaei_nima@yahoo.com*)

Asghar Aghamohammadi (*aghamohammadi@tums.ac.ir*)

Venue: Children's Medical Center Hospital, Tehran, Iran

SUMMARY

Iran joined the efforts of numerous organizations around the world to improve the awareness of PID. The Iranian NMO and Research Center for Immunodeficiencies (RCID) carried out different activities to commemorate the World PI Week (WPIW) for the fourth consecutive year on 22–29 April 2014 at the Children's Medical Center Hospital, Tehran, Iran. Patients and their family meeting, scientific lectures, expert PID meeting, junior scientists meeting, panel discussion and Interviewing with media are parts of the activities that take place during the Wold PI week in Iran.

On the first day of WPIW, a social activity of walking from the Hospital to the Laleh Park was organized to distribute 10 warning signs poster in the public. Members of the hospital attracted people's attention and handed out postcards of the World PI Week and leaflets with the 10 warning signs of primary immunodeficiency.

The morning report of the Children's Medical Center Hospital was dedicated to PID during which lecture on primary immunodeficiency and approach to a case with CVID were given for pediatricians, pediatric residents and medical students. This was continued by two lectures by Dr. Aghamohammadi and Dr. Rezaei on approach to PID patients and PID activities in Iran, respectively.

On 23 April, another scientific meeting was organized in Rasoul Akram Hospital,



one of the main general hospitals in Tehran. Indeed PID patients with their families and a large number of people participated in the ceremony which was held by the Iranian NMO in the Children's Medical Center. Children's TV artists are guests of this ceremony who perform joyful programs for patients.

On 24 April, A ceremony was held at Children's Medical Center Hospital in order to appreciate people supporting primary immunodeficiency patients.

On 26 April, a theater was held at the conference hall of Children's Medical Center Hospital. All proceeds from the theater went directly to patients care and were used to buy medications and purchase supplies for patients.

On 27 April, another scientific meeting was organized in the Mofid Children's Hospital.

During the WPIW, different programs were conducted in several children's hospital in Tehran. The aim of these programs was to increase awareness of medical staff about diagnosis and treatment of PIDs and also difficulties faced by patients to get their drugs especially intravenous immunoglobulin. Dr Aghamohammadi, Head of Research Center for Immunodeficiencies and Dr Rezaei, the Deputy President of Re-





search Center for Immunodeficiencies participated in several television and radio interviews and spoke about prevalence of PID in Iran, warning signs of primary immunodeficiency and treatment of PID patients.

POLTAVA, UKRAINE

April 24–25

Host: Liudmyla Chernyshova (*vchernysova@ukr.net*)

Venue: Poltava

PROGRAM

April 24

Clinical features allowing to suppose primary immunodeficiency of antibody formation – on examples of patients
Volokha A. (Ukraine)

Clinical features of combined immunodeficiency – on examples of patients
Kostyuchenko L., Romanyshin Y. (Ukraine)

New generations of Ukrainian immunoglobulins for intravenous injection pharmacopeia requirements for their production
Karbovskiy V., Rybakov A. (Ukraine)

New problems in diagnostics and treatment of PID in Ukraine
Chernyshova L. (Ukraine)

Congenital Immunity Defects – NEMO syndrome
Belevtsev M. (Belarus)

Questions of immunodeficiency in educational program at pre-diploma period of medical training
Kryuchko T. (Ukraine)

April 25

Complement system's deficiency manifestation
Polich T., Bondarenko A. (Ukraine)

Hemophagocytic syndrome
Savchak I. (Ukraine)

Immunization in medical care of children with PID
Lapyi F. (Ukraine)

Local immunity in children with PID of antibody formation
Yakimovich (Ukraine)

SUMMARY

The 10th Ukrainian J-Project Meeting «Modern possibilities for diagnosis and treatment of primary immunodeficiency in children» was organized in Poltava (Ukraine) on the 24–25 of April during the World PI Week 2014. The Meeting was organized by the Department of Pediatric Infectious Diseases and Pediatric Immunology of Shupik NMAPE (Head of Department – Professor L.I. Chernyshova) with the assistance of the Department of Health Poltava Regional State Administration and sponsorship of domestic manufacturer of intravenous immunoglobulin “Biopharma”. The meeting was attended by 200 pediatricians from the Poltava region, pediatric immunologists from most regions of Ukraine, as well as guest speakers from Belarus. The Belarusians Mikhail Belevtsev and Svetlana Sharapova were the only foreign guests who supported Ukrainian J Project meeting this year. The program consisted of two parts. The first part was designed to increase awareness on PID among general practitioners and pediatricians by learning the basic clinical signs and symptoms for timely referral of patients to a pediatric immunologist. Lectures by leading immunologists of Ukraine (Liudmyla Chernyshova, Alla Volokha, Yaryna Romanyshin) were presented followed by an interactive poll to check the learned knowledge. In the second part more specific issues of diagnostics and treatment of PID were discussed. Possibilities of laboratory diagnostics of PID in Ukraine were presented by young enthusiastic laboratory immunologist Dariia Osypchuk working under the supervision of Head of Laboratory of Immunology in Pediatric, Obstetrics and Immunology Institute in Kiev professor Victor Chernyshov. Report concerning possibilities of diagnostics of new PID was done by Mikhail Belevtsev (Belarus). Many reports were dedicated to clinical presentation of cases. Anastasiia Bondarenko, Yuriy Stepanovsky, Anna Gilfanova prepared interactive reports. Svetlana Sharapova reported the results of treatment some of Ukrainian patients in Belarus. Discussion was organized among immunologists through interactive communication. After the end of the meeting tour around Poltava was organized for participants. Poltava is famous for the Battle of Poltava in 1709 between Peter I of Russia and the Swedish forces. Under Field Marshal Carl Gustav Rehnskiöld during the Great Northern War. So the obligatory social event was attending the historical and cultural reserve “Poltava battle field”, which is part of an international association of military history museums in the world (IAMAM) operating under the auspices of UNESCO. Relaxed atmosphere and hospitality of Poltava helped to create the best impression of the meeting.

Liudmyla Chernishova



*At the monument of the Poltava battle.
L→R in the first row: Liudmyla Chernishova,
Liubov Nikonets, Anastasia Bondarenko,
Ihor Savchak*

Host: Peter Ciznar (*ciznar@dfnsp.sk*)

Venue: Children's Clinic, Slovak Postgraduate Academy of Medicine, Bratislava

Main topic: Management of primary immunodeficiencies

PROGRAM

Treatment of hypogammaglobulinemia in EU countries

Sediva A., (Czech Republic)

Severe form of atopic dermatitis associated with hypogammaglobulinemia

Kayserová J., Kubešová H., Sediva A. (Czech Republic)

Combined immunodeficiency in patients with HLHS

Hlaváčková E., Humlová D. (Czech Republic)

Risk of autoimmune complications in CVID

Straková J. (Slovakia)

Management of autoimmune cythopenias in CVID

Raffáč Š., Gabzdilová J., Mitniková M. (Slovakia)

Severe combined immunodeficiency in Serbia and Montenegro

Srdjan Pasič (Serbia)

Severe combined immunodeficiency in Slovakia

Ciznar P., (Slovakia)

Chronic granulomatous disease in ECE countries

Markej G., (Slovenia)

Chronic granulomatous disease, conservative treatment

Kayserová H., (Slovakia)

Chronic granulomatous disease, transplantation therapy

Boďová I., (Slovakia)

Inflammatory bowel – like diseases associated with PI

Jeseňák M., Bánovčin P., Havlíčková Z., Szépeová R. (Slovakia)

Diagnostic tests for complement functions

Zuzulová M. (Slovakia)

Diagnostics of antibody deficiencies

Ciznar P. (Slovakia)

Subcutaneous replacement in systemic immunoglobulin adverse reactions

Eštóková T. (Slovakia)

Registry of PI in Children University Hospital and SSAK

Veghová L. L. (Slovakia)



Hosts: Mónika Apáti Nagy (*apati.monika@nitak.hu*)László Maródi (*lmarodi@med.unideb.hu*)**Venue:** University of Debrecen, Debrecen

PROGRAM

- | | |
|---|--|
| 10.00 Registration | |
| 10.30 Greetings by the presidents of MIBE and HUSID | placement from the perspectives of patients |
| <i>Mónika Apáti Nagy and László Maródi</i> | <i>Adrien Apáti Nagy, Bella Csépké, Mónika Gillich</i> |
| 11.00 Primary immunodeficiency disorders in childhood | 12.00 Discussion |
| <i>Adrien Sarkadi</i> | 12.30 Lunch |
| 11.30 Subcutaneous immunoglobulin re- | 13.00 Talent spotter competition |
| | 15.00 Announcement of awards |
| | 15.15 Balloon launch |

SUMMARY

This Meeting was organized together with Hungarian Society for Immunodeficiencies (HUSID) and the Hungarian Organization for Patients with Immunodeficiencies (HOPI) during the World Primary Immunodeficiency Week and was devoted to the management of patients with PID. We discussed the recent development and the field of immunoglobulin substitution. In particular, presentations by patients and doctors were focused on advantages and disadvantages of subcutaneous immunoglobulin (SCIG) therapy. Patients who had been on SCIG reported their own experiences with this treatment. The interactive meeting was followed by the balloon launch in front of the main building of University of Debrecen.

László Maródi*Mónika Apáti Nagy**Mónika Gillich**Bella Csépké*



Preparation for balloon launch



Adrien Sarkadi

ST. PETERSBURG, RUSSIA

May 28

Host: Tatiana Nasyrina (tatiana.nasyrina@biotest.com)
Venue: St.Petersburg, Mira str., 14, “Research Institute of Epidemiology and Microbiology, Pasteur Institute”
Main topic: Primary immunodeficiencies, diagnosis and treatment

1-st State Medical University named Pavlov (St. Petersburg)
Research Institute of epidemiology and microbiology, named Pasteur (St. Petersburg)
State Pediatric Medical Academy (St. Petersburg)
All-Russian Centre of emergency and radiation medicine of the Ministry of emergency of Russia (St. Petersburg)

PROGRAM

PID Center in St. Petersburg
Areg Totolian, (St. Petersburg)
 PID in the practice of the therapeutics and principles of therapy
Tatiana Latisheva, (Moscow)
 Primary immunodeficiencies in children. Major clinical guidelines
Mikhail Yartsev, (Moscow)
 Primary immunodeficiencies associated with the deficit of adaptive immunity. B-cell immunodeficiencies
Natalia Kalinina, (St. Petersburg)

Primary immunodeficiencies associated with deficit of the innate immunity. Chronic granulomatous disease
Natalia Davidova, (St. Petersburg)
 Autoinflammatory syndroms
A. Shcherbina, (Moscow)
 Multi-step diagnostic Protocol for screening patients for PID designed for doctors-nonimmunological specialization
M Guseva, (St. Petersburg)
 DNA-diagnosis of primary immunodeficiency
A. Semenov, (St. Petersburg)

SUMMARY

May 28, 2014 year in St. Petersburg under the patronage of the 1-st St. Petersburg State Medical University named Pavlov, St-Petersburg Research Institute of epidemiology and microbiology, named Pasteur, St-Petersburg State Paediatric Medical Academy, All-Russian Centre of emergency and radiation medicine of the Ministry of emergency of Russia took place “J Project meeting “Diagnosis and treatment of primary immunodeficiency”.

Presentations made leading experts from Moscow and St. Petersburg. Conference attended by 70 doctors, including Pediatricians, Infectious diseases specialists, Neurologists. For the first time managed to attract General practitioners from different cities and villages of the North-Western administrative district. All lectures listened with interest and asked questions. In addition to the reports we shown Biotest film: “PID in adults”. After the Conference doctors thanked the organizers and expressed hope for continued this program in other cities in the North-West region.

Tatiana Nasyrina

VARNA, BULGARIA

October 6

Host: Elissaveta Naumova (*naumovaej@gmail.com*)

PROGRAM

Primary immunodeficiency in Bulgaria
– present and future

Naumova E. (Bulgaria)

Diagnosis and management of primary
antibody deficiencies

Avcin T., (Slovenia)

Outcomes of treatment in children with
severe combined immunodeficiency
(SCID) in Slovakia.

Čížnár P., (Slovakia)

Common variable immune deficiency at
the university hospital “ALEXAND-

ROVSKA”-one and a half year experi-
ence

*Mihailova S., Gesheva N., Lesichkova
S., Baleva M., Temelkov A., Naumova
E. (Bulgaria)*

Application of dynabeads® human t-ac-
ticator cd3/cd28 kit for assessment of
t-cell function in patients with com-
mon variable immunodeficiency

*Gesheva N., Lesichkova S., Mihailova
S., Mihaylova A., Naumova E., (Bul-
garia)*

SUMMARY

This J Project has been held together with the 4th National Congress of Immunology organized by the Bulgarian Association of Clinical Immunology and Immunology Society as well. The immunological audience presented at the congress was informed about both the current status of diagnosis and treatment of PID patients in Bulgaria and PID registry activities. Peter Čížnár from Slovakia presented an interesting talk about

Outcomes of treatment in children with SCID in Slovakia while Tadej Avcin from Slovenia has focused in his presentation on current issues related to management of patients with primary antibody deficiency requiring immune globulin intravenous or subcutaneous replacement therapy, early evaluation of infections, prophylactic antibiotic in refractory infections and monitoring of the patient's pulmonary function. During the discussion a progress in management of PID in Bulgaria has been considered.

Elissaveta Naumova



Elissaveta Naumova and her audience

CATEZ, SLOVENIA		October 11–12
Host:	Slovenian Pediatric Association and Slovenian Society for children with immune deficiencies	
Scientific chair:	Prof. Tadej Avčin, MD, PhD (tadej.avcin@kclj.si)	

PROGRAM

October 10
Scientific meeting and patient discussion with visiting professor Andrew Cant, ESID President elect

October 11
Advances in management of children with primary immunodeficiency diseases
A. Cant, (Newcastle upon Tyne, GB)
What should a pediatrician know about bone marrow transplant in children
J. Jazbec, (Slovenia)

Fever as a symptom of autoinflammatory diseases
(N. Toplak, (Slovenia))
Infections in immunocompromised children
M. Pokorn, (Slovenia)
Slovenian registry of patients with PID
Š. Blazina, (Slovenia)
Clinical and genetic characteristics of patients with CGD in central and eastern European countries
G. Markelj, (Slovenia)



Natasa Toplak, Andrew Cant, Gasper Markelj, Tadej Avcin, and Stefan Blazina

SUMMARY

Slovenian autumn J-Project Meeting was organized as a part of the Slovenian paediatric congress to reach broad scientific public. The meeting was organized by the Slovenian Pediatric Association and Slovenian Society for children with immune deficiencies. The president of scientific committee was Prof. Tadej Avcin, Head of the Department of Allergology Rheumatology and Clinical Immunology at the University Children's Hospital Ljubljana which is the only Jeffrey Modell Centre for Primary Immunodeficiencies in Slovenia. The meeting was attended by 400 pediatricians from all parts of Slovenia. Pediatric immunologists, infectious disease specialists and physicians from pediatric BMT centre were invited as speakers. We were very pleased that prof. Cant from Newcastle upon Tyne responded to invitation to participate in scientific discussion of patients with yet undefined PID and patients who are difficult to treat because of complicated disease course. After each case presentation we had vivid discussion about diagnostic and treatment plans in our most challenging patients. Prof. Cant gave his keynote lecture on overview of progress in the field of BMT in patients with PID in the last 25 years. Probability of 5-year survival has increased with better methods of donor selection and reduced intensity conditioning protocols. Advantages of new methods as TCR alpha/beta depleted transplants were discussed among others. One of important messages from prof. Cant's lecture was that BMT improves patient's quality of life and should be taken into account when considering a transplant as a treatment option. Prof. Jazbec is a head of the Slovenian pediatric transplant centre and has many years of experiences in BMT in children. Five patients with SCID had successful BMT in this centre in the last years. Prof. Jazbec explained basic concepts of treatment with BMT, most common complications and results from our centre. Nataša

Toplak who has special interest in auto inflammatory diseases presented classification of these diseases and major clinical and laboratory features. Although these are rare diseases it is important to consider them as differential diagnosis in patients with signs of inflammation. Marko Pokorn shared with the audience his experiences with infections in patients with PID as infectious disease specialist. He also gave an overview of most common infections that are typical for different defects of immune system. Štefan Blazina presented data from the Slovenian national PID registry that show expanding spectrum of PID diagnoses and increasing number of newly diagnosed patients with PID in Slovenia. Nearly half of the patients in the registry have genetically confirmed diagnosis. Gašper Markelj presented clinical and genetic data from one of the largest cohorts of patients with CGD from central and eastern European countries. Prevalence of diagnosed CGD in Slovenia is higher than in other countries.

Meeting was considered as very successful as regards raising awareness about PID especially among primary physicians who are the first who come in contact with patients with PID and should consider PID as a differential diagnosis. One of the important messages from the congress was also that PID is not a synonym of recurrent infections but often presents with autoimmune manifestations or even allergy.

Tadej Avcin

TEHRAN, IRAN		October 18–19
Host:	Nima Rezaei (<i>rezaei_nima@yahoo.com</i>)	
Venue:	Research Center for Immunodeficiencies, Children’s Medical Center Tehran University of Medical Sciences, Tehran	
Main topic:	7 th Meeting on Clinical Immunology & Immunodeficiencies in Iran	

SUMMARY

The 7th Meeting on Clinical Immunology and Immunodeficiencies, which also considered as the 108th J Project Meeting, was organized in October 18–19, 2014 (Tehran, Iran), along with 26th International Congress of Pediatrics. In 2005, first International Congress on Immunodeficiency Disorders was organized in Tehran. A number of PID experts from different countries (USA, UK, Germany, France, Italy, Sweden, Spain, Japan, and Turkey) attended the congress to present an update in this field, whereas many scientists and researchers took part in this congress to increase their knowledge. This 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. That two days meeting was supported by the Jeffrey Modell Foundation (JMF). The great success of these conferences brought up the idea of organizing the Clinical Immunology, Allergy and Immunodeficiency Diseases Meeting this year, just before the 22nd International Congress

of Pediatrics. This meeting, which was considered as the 46th J Project Meeting, is organized by the Research Group for Immunodeficiencies in collaboration with the Department of Pediatrics, Children's Medical Center Hospital, Tehran University of Medical Sciences.

Since 2011, as of establishment of the Research Center for Immunodeficiencies, the Meetings on Clinical Immunology and Immunodeficiencies is organized annually along with International Congress of Pediatrics in October. This year, the 7th Meeting on Clinical Immunology and Immunodeficiencies, also considered as the 108th J Project Meeting, was organized along with the 26th International Congress of Pediatrics to focus on the linkages of fundamental sciences and patient-oriented research under the main theme of clinical immunology and immunodeficiency diseases. Improving the awareness of physicians, pediatricians and other specialists would be the main objective of the meeting. We are very pleased to host a total of more than 1,500 delegates in this meeting this year. The major part of audiences included general practitioners, pediatricians, and pediatric subspecialists; however, medical students, pediatric residents and fellows in the field of clinical immunology and infectious diseases have been actively participated in the meeting.

The meeting was organized in three sessions in three different halls. The main session of the meeting was started by a key-note lecture by Prof. Arndt Borkhardt, Germany who presented a talk on "Novel development in pediatric stem cell transplant", followed by a main lecture by Prof. Siamak Bahram, France "The new genetics of the major histocompatibility complex with applications to transplantation and HLA-disease associations". Dr. Asghar Aghamohammadi and Dr. Nima Rezaei presented two other main lectures, followed by 15 other short talks, where nine selected posters were also presented during the meeting. A fund-raising social event by Hossein Alizadeh, concert director, with singing of Mohammad Motamedi was also an unforgettable even during the meeting. All the proceeds from this event will be spent for patients with primary immunodeficiency diseases.

Nima Rezaei, MD, PhD
Asghar Aghamohammadi, MD, PhD



*L→R: Arndt Borkhardt,
Nima Parvaneh, Nima Rezaei,
Asghar Aghamohammadi,
Siamak Bahram*



*The HamAvayan band heading
by Hossein Alizadeh
and Mohammad Motamedi*



YEKATERINBURG, RUSSIA

October 24–25

Host: Irina Tuzankina (*ituzan@yandex.ru*)

PROGRAM

October 24

History of the study of patients with primary immunodeficiency in Russia

Yartsev M.N. (Moscow, Russia)

Primary immunodeficiencies in the Urals region

Tuzankina I.A. (Yekaterinburg, Russia)

Recent developments in understanding genetics of chronic mucocutaneous candidiasis

László Maródi (Debrecen, Hungary)

Clinical manifestations and molecular genetics of CGD

Gašper Markelj (Ljubljana, Slovenia)

Rare forms of primary immunodeficiency

Sherbina A.Y. (Moscow, Russia)

Clinical features of the Di Giorgi syndrome. Family case

Vlasova E.V., Tuzankina I.A., Deryabina S.S. (Yekaterinburg, Russia)

Analysis of genealogical markers in adult patients with PID

Karakina M.L., Tuzankina I.A., Shershnev V.I. (Yekaterinburg, Russia)

Clinical and immunological phenotypes in Common Variable Immunodeficiency

Latysheva T.V. (Moscow, Russia)

Tolerability of IVIg for primary immunodeficiency treatment

Fabrizio Fabrizzi (Florence, Italy)

Opportunities of replacement immunoglobulin therapy in restoring immune

dysfunctions of patients with X-linked agammaglobulinemia (XLA)

Sizyakina L.P. (Rostov-on-Don, Russia)

Primary immunodeficiency. EBV. Malignant lymphoproliferation

Belevtsev M.V. (Minsk, Belarus)

Omen Syndrome in East Slavic countries

Sharapova S.O. (Minsk, Belarus)

Results and prospects of the Primary Immunodeficiency center in the Republic of Kazakhstan

Kovzel E.F., Otarbayev N.K. (Astana, Kazakhstan)

Pharmacoeconomic study of newborn screening testing (TREC)

Bolkov M.A., Tuzankina I.A. (Yekaterinburg, Russia)

Pharmacoeconomics in PID

Smirnova I.N. (Moscow, Russia)

October 25

B cell immunodeficiencies

Anne Durandy (Paris, France)

Small forms of immunodeficiency and their diagnostics

Korsunskiy I.A. (Moscow, Russia)

Transplantation in PID. Results

Balashov D.N. (Moscow, Russia)

Autoinflammatory diseases

Kozlova A.L. (Moscow, Russia)

PID's Genetics

Kuz'menko N.B. (Moscow, Russia)

Determination of TREC and KREC in 1-year-old children

Deryabina S.S., Vlasova E.V., Tuzankina I.A., Prodeus A.P., Gordukova M.A. (Yekaterinburg – Moscow, Russia)

SUMMARY

Eurasian J Project meeting, held on 24 and 25 October 2014 in Yekaterinburg in the framework of Russian sciences forum in the Urals with international participation «Topical issues of fundamental medicine».

In total, the Forum was attended by 496 people, including Rolf Martin Zinkernagel, Nobel Prize winner. There were 27 academicians and corresponding members of the Russian Academy of Sciences, including the main organizer and Chairman of the Science and High technology Committee, Director of the Institute of Immunology and physiology, Russian Academy of Sciences V.A.Chereshnev. And 151 MMD and PHD students, health practitioners, students, graduate students – 318 people.

In Eurasian J Project meeting participated experts of Primary Immunodeficiencies – leading immunologists, clinicians,

students and graduate students. Among these participants were people from Belarus, Uzbekistan, Kazakhstan, Switzerland, France, Germany, Hungary, Slovenia, Japan, Italy, more than 20 Russian cities (Moscow, St. Petersburg, Ekaterinburg, Novosibirsk, Chelyabinsk, Tyumen, Omsk, Kazan, Krasnodar, Perm, Rostov-on-Don, Orenburg, Orel, Samara, Astrakhan, Berdsk, Izhevsk, Forest, Barrow et al.).



Valery Aleksandrovich Chereshnev chairing the Keynote session



Rolf M. Zinkernagel presenting his Keynote lecture

Internationally recognized scientists and young researchers presented their lectures and reports in 3 sessions. Debates and discussions about presented material were active and interesting, was attended by both well-known Russian scientists and Nobel laureate Rolf Zinkernagel Martin.

Pleasant and warm end of this international meeting of colleagues was a visit to the historic cultural and educational complex on the border between Europe and Asia, which did not interrupted even by sudden-onset of cold and heavy snowfall.

Irina Tuzankina



L→R: Andrey Pihlak, Yokota Shumpei, Valeriy Chereshevnev, Margarita Cheresheva, Mikhail Belevtsev, Svetlana Sharapova, Boris Pinegin, Andrey Prodeus, Elena Glebova, László Maródi, Areg Totolyan

Host: Ewa Bernatowska (ewa.bernatowska@gmail.com)

Venue: Villa Park Wesola

PROGRAM

October 24

Polish Society of Experimental and Clinical Immunology (PTDiK), and the Central European Journal of Immunology – the creation of new sections within the PTIDiK, cooperation with Polish Working Group for the Primary Immunodeficiency Disorders

Jacek Roliński, Ewa Bernatowska

The development of clinical immunology in the country, efforts aimed at improving the care of patients with primary immunodeficiencies in Poland; Number of specialists in the field of clinical immunology, drug programs, reimbursement of medicines

Maciej Siedlar

Biological drugs in the treatment of primary immunodeficiency disorders

Karina Janz-Różyk

Polish Working Group for primary immunodeficiency disorders – actions

for growth of detection of primary immune deficiency; – how registry of primary immunodeficiency disorders should look like; – vaccinations in children and adults – are they safe and effective

Ewa Bernatowska

Replacement therapy with intravenous and subcutaneous gamma globulins preparations

Małgorzata Pac

Adverse events after therapy with gamma globulins preparations

Barbara Pietrucha, Edyta Heropolińska-Pliszka, Małgorzata Pac

European registry of primary immunodeficiency disorders – ESID registry on line – now available; HSCT registry of Polish Working Group for the Primary Immunodeficiency Disorders

Beata Wolska-Kuśnierz

Common variable immunodeficiency; eti-



ology, disturbances of B-cell maturation

Barbara Piątosza

Diagnostic criteria of common variable immunodeficiency

Jacek Roliński

October 25

Common variable immunodeficiency; immunological research, clinical picture, replacement therapy – a single center experience

Jacek Roliński, Rafał Becht, Ewa Więsik-Szewczyk, Magdalena Strach, Hanna Suchanek, Anna Pituch-Noworolska, Aleksandra Uszyńska, Zbigniew Santorski

Round Table Discussion – Present and future of diagnosis and treatment of humoral primary immunodeficiencies – from birth to elderly age. Conference of the parents and children with primary immunodeficiency disorders organized by the Polish Working Group with the participation of patients and their parents

Adverse events after therapy with gamma globulins preparations

Barbara Pietrucha, Edyta Heropolitańska-Pliszka, Małgorzata Pac

Skin changes in primary immunodeficiency disorders

Edyta Heropolitańska-Pliszka, Barbara Pietrucha



Ewa Bernatowska (middle) and her associates

SUMMARY

Conference has been organized in the framework of J Project in cooperation with patients organization – IMMUNOPROTECT (see program). The venue for the conference has been located in Wesola, in the forest area, near Warsaw.

Topic of the first session has been focused on present activity of Central European Journal of Immunology, an official journal of the Polish Society of Experimental and Clinical Immunology, which has been associated with JProject. The new editor pro-

fessor Urszula Demkow was replaced professor Włodzimierz Marcinkiewicz on this position.

The optimistic news is, that Central European Journal of Immunology will be a member of PubMed database, but is still on waiting list. During the afternoon session, the updated on the Polish registry of primary immunodeficiencies (PID) has been presented. It has been shown that number of the diagnosed patients with PIDs increased in 2014 to 4,360. Since first of September program of replacement therapy for adult has been established as a National Therapeutic Program for Adults, which is fully guaranteed by National Health Fund.

The national project with the aims of improving patient care, resulting in early clinical recognition and improved diagnostic and treatment strategies, and raising awareness of PID across the country initiated by Polish immunologists has been consolidated more and more. The Polish Working Group for PID was established in March, 2005. The Group was set up from 6 main paediatric centres for the diagnosis and therapy of PID, and growing up to 15 centres. During the last years, the Group has been assembled from 12 adults centers also, covering the whole of Poland.

Topic of the next day session has been focused on diagnosis and therapy of PID adults patients. Lectures have been done by leading immunologists from adults centers. This year sixtyfive clinical immunologists, both pediatricians and internists attend the conference. The group of 5 colleagues from Ukraine was participated actively in the meeting.

Ewa Bernatowska

PARDUBICE, CZECH REPUBLIC

November 11

Host: Anna Sediva (anna.sediva@fnmotol.cz)

Venue: Pardubice, Hotel Lave

PROGRAM

Humorální imunodeficiency

Jiří Litzman

Pneumopatologie u nemocných léčených pro běžnou variabilní imunodeficiency (CVID)

Pavla Králíčková

Přístup k pacientům s bronchiectáziemi

Zuzana Antušová

Běžný variabilní imunodeficiency a autoimunita

Tomáš Milota

10 varovných znaků primárních imunodeficiency

Andrea Poloučková

Terapie imunoglobuliny, praktické aspekty

Radana Zachová

Screening SCID

Anna Šedivá

Transplantace kmenových buněk krvetvorby u dětí s primárním imunodeficiency

Renata Formánková

Cytometrické vyšetřování lymfocytů při imunodeficiency – EuroFlow PID

Tomáš Kalina

Sekvenování nové generace u imuno-
deficitů

Froňková Eva

Molekulárně genetické vyšetření primár-
ních imunodeficiencí

Tomáš Freiburger

Marek a Marek aneb jak je důležité mít
bratrance

Jiří Bronský

Nový autoinflamatorní syndrom s recidi-
vujícími horečkami, těžkou anémií a
B-buněčným imunodeficitem

Jana Kayserová

SUMMARY

Our meeting in Pardubice was another one in a serie of meetings rotating around the Czech Republic.

The main aim of these meetings is raising awarness about PIDs and to approach lo-
cal physicians of the first contact with patients. This year we organized our meeting in
East Bohemian region.

The meeting took part in Pardubice, on 10. and 11. November 2014.

The first day of the meeting was on invitation which was distributed to members of
PID group. established within Czech Society for Allergy and Clinical Immunology.
Total 15 members actively participated and agreed on a program for the next year.

The second day was open to all attendants. The program is attached, covered humor-
al and cellular immunodeficiencieis, novel topics such as next generation sequencing
and advanced flow cytometry were also presented. The program was complemented
by interesting case reports.

Total 35 people participated in this activity, awarded also by educational credit by a
chambre of physicians.

The meeting for 2015 is already in preparation, would take part in Moravia in Sep-
tember 2015.

Anna Sediva

NIZHNY NOVGOROD, RUSSIA

November 26

Host: Tatiana Nasyrina (*tatiana.nasyrina@biotest.com*)

Venue: Conference Hall State institution Nizhny Novgorod Regional
Children's clinical hospital

Main topic: Educational seminar-school for physicians, "Primary immunode-
ficiencies, introduction to practice"

Organizers: The Ministry of Health of the Nizhny Novgorod region; Nizhny
Novgorod State Medical Academy; Public organization "Asthma.
Allergy. Immunity"

PROGRAM

Film demonstration: “The human immune system: protection against infections»
“Primary immunodeficiencies, classification, principles of diagnosis»

I. Kondratenko

Film demonstration: “The Right to Life”
Principles of therapy of patients with primary immunodeficiencies

T. Latisheva, Moscow

Ways and the difficulties of identifying patients with primary immunodeficiencies

M. Abelevich, Nizhniy Novgorod



I. Kondratenko



T. Latisheva



M. Abelevich

SIBIU, ROMANIA

November 27–28

Host: Sorin Iurian (*iurian_sorsab@hotmail.com*)

Venue: Hotel Continental, Sibiu

PROGRAM

1st Session

Clinical and immunological presentation of combined immunodeficiencies

Alessandro Plebani, (Brescia, Italy)

Severe Combined Immunodeficiency.
Case Presentation

Mihaela Bataneant (Timisoara, Romania)

A Case of Combined Immunodeficiency Complicated by Stroke

Alexis Cochino, Bucharest, Romania

Safety through Quality – The Production Process of Intratect Intravenous Immunoglobulin

Fabian Peissker, (Biotest)

2nd Session

Discovery of Novel Genetic Etiologies of Chronic Mucocutaneous Candidiasis Disease

Laszlo Marodi, (Debrecen, Hungary)

Diagnosing primary immunodeficiency:
(almost) all you need is pattern recognition

Esther de Vries, (Hertogenbosch, The Netherlands)

Primary Immunodeficiencies in National Programme

Diana Deleanu, (Cluj Napoca, Romania)

Hyper IgM Syndrome. Case Report

Iurian Sorin (Sibiu, Romania), Laszlo Marodi, (Debrecen, Hungary)

3rd Session

ARPID, a Journey to Future

Artemisa Baldea, (Romania)

Home Care for Patients with Primary Immunodeficiencies

Mircea Cinteza, (Bucharest, Romania),

Mr. Florin Hozoc, (Besmax Pharma Distribution)

4th Session

Hereditary angioedema: a diagnostic, therapeutic and management challenge

Dumitru Moldovan, (Tîrgu Mures, Romania)

Autoimmune and atopic diseases in paediatric patients with selective IgA deficiency

Zoltán Ellenés-Jakabffy, (Oradea, Romania)

A case of Familial Hemophagocytic Lymphohistiocytosis

M. Bataneant, (Timisoara, Romania)

Round table. PID Diagnosis in Romania. Conclusions.

SUMMARY



Front row (L→R), Michail Becker, Esther de Vries,
Alessandro Plebani, László Maródi.

Second row (L→R), Fabian Preissker, Sorin Iurian,
Diana Deleanu, Margit Serban

The 116th J Project Meeting has been held in Sibiu, Romania on November 28, 2014 and represented the 3rd meeting held in Sibiu. During the XV–XVIII centuries, Sibiu became the most powerful fortress of Transylvania and one of the most flourishing and prosperous citadels. Pope Eugen the IVth called it “The Red Citadel” from the colour of the bricks used for its fortifications. Sibiu is one of the most important cultural centres of Romania and along with the city of Luxembourg, it was designated a European Capital of Culture for the year 2007. Formerly the centre of the Transylvanian Saxons, the old city of Sibiu was ranked as “Europe’s 8th most

idyllic place to live” by Forbes. In order to improve the knowledge about PIDs, The J Project Meetin was preceded by a course about basics in immunology and PIDs and was dedicated to general practitioners, residents in pediatrics, medicine students. More than 60 physicians attended J Project Meeting, including professionals in PID field (Prof. Laszlo Marodi – Hungary, Prof. Esther de Vries – Netherlands, Prof. Alessandro Plebani – Italy), specialists and residents in pediatrics and general practitioners from many Romanian counties (Sibiu, Timisoara, Cluj-Napoca and Bucharest). We remarked an important participation of ARPID members (Romanian Association of Patients with Primary Immunodeficiencies) that shared their experience regarding PIDs. There were also presented many clinical cases, representing an opportunity to interact and receive useful information from colleagues with experience in PIDs. On behalf of Romanian Group of Primary Immunodeficiencies, we are so grateful to professor Laszlo Marodi for his support regarding the diagnosis of PID patients from Romania. The social program included participation to a concert sustained by Sibiu Philharmonic Orchestra and a visit to Bruckenthal National Museum, making this conference an experience to remember.



*Together with parents and children
from Brassow*

Sorin Iurian

ERZURUM, TURKEY

December 6

Hosts: Dilara Kocacık Uygun, Demet Hafizoğlu

SUMMARY

After we had planned the Eastern Anatolia J Project to increase the the awareness of primary immunodeficiency diseases among the physicians, we had organized J Projects in Diyarbakır, Urfa and Malatya. J-Project Meeting in Erzurum was the fourth meeting of Eastern Anatolia J Project, and also the seventh J-Project Meeting in Turkey. Erzurum J Project Meeting was held on December 6, 2014 in Turkey. The meeting was organized with Dr. Dilara Kocacık Uygun and Dr. Demet Hafizoğlu from Erzurum. The meeting was primarily focused on the awareness, clinical signs and treatment of PID among the medical students and physicians (pediatricians, family physicians), also to discuss the importance of early diagnose in PID. More than 60 participants attended the meeting. The meeting was started with an opening lec-



ture by Prof. Dr. Ismail Reisli focused on the awareness of primary immunodeficiency. Two case reports were presented in the meeting about patients with PID which were based on diagnostic problems, clinical signs, screening and treatment of PID by Dilara Kocacık Uygun and Demet Hafızoğlu. The third part of the meeting were dedicated to the warning signs of PID in adults by Prof. Dr. Zafer Çalışkaner.

I would like to thank all speakers and participants for their contributions, especially the our sponsor (Gen Ilac) for their financial help which made it

Ismail Reisli

CHISINAU, MOLDOVA

December 12

Host: Liudmila Cerempei (*lcerempei@rambler.ru*)

Venue: Chisinau Congress Center

Main topic: Primary immunodeficiency in children: peculiarities of diagnosis and treatment

PROGRAM

Possibilities and limitations in diagnosis and treatment of PID in Moldova. PID syndromes. Louis-Bar Syndrome (case presentation)

Liudmila Cerempei, (Moldova)

From The J Project to the Antalya Declaration

László Maródi, (Debrecen, Hungary)

Peculiarities of the immune system I children. Methods for primary immunodeficiency diagnosis in children

Sergiu Ghinda, (Moldova)

Discovery of Novel Genetic Etiologies of Chronic Mucocutaneous Candidiasis Disease

László Maródi, (Debrecen, Hungary)

Recurrent febrile syndrome associated with pancytopenia

Mihaela Bataneant, (Timișoara, Romania)

Evolution of the ENT diseases in children with PID

Svetlana Diacova, (Moldova)

Bruton's disease (case presentation)

Rodica Selevestru, (Moldova)

DiGeorge syndrome in children (case presentation)

Ludmila Grițco, (Moldova)

Allergy “masks” in PIDs –

Tatiana Gorelco, Healthcare (Moldova)

Kostmann syndrome in children (case presentation)

Ala Jivolcovschi, (Moldova)

Secondary immunodeficiency in children

Liudmila Cerempei, (Moldova)

SUMMARY

The J Project symposium, which took place in Chisinau on 12th December 2014 hosted 111 pediatricians from whole Moldova.

At the moment, in the Republic of Moldova there are 26 diagnosticated pediatric patients with primary immunodeficiencies (PID):

- 21 with IgA selective deficit;
- 2 patients with Louis-Barr syndrome;
- 1 patient with Di George;
- 1 patient with Bruton disease;
- 1 patient with Kostmann syndrome.

The Symposium was opened by the Health Minister Mr. Cekan, who underlined the importance of such activities for Moldovan pediatrics, and mentioned could increase the rate of early detection, treatment and supervision of patients with primary immunodeficiencies. The symposium's program included 10 reports (the program is attached).

Prof. László Maródi, Coordinator of the “J Project”, Department of Infectology and Pediatric Immunology, University of Debrecen (Hungary) greeted the participats with



L→R, Svetlana Cemirtan, Olga Cîrstea, Mihaela Bataneant, László Maródi, Olga Motpan, Liudmila Cerempei, Cristina Grosu, Ana Clima

an introducing report “From The J Project to the Antalya Declaration”, mentioning J Project’s successes in Esten and Westen Europe.

Prof. Liudmila Cerempei, Coordinator of the “J Project” in Moldova (Pediatric Department, SMPHU “N. Testemițanu”) gave a speech on the topic “Possibilities and limitations in diagnosis and treatment of PID in Moldova. PID syndromes. Louis-Bar Syndrome (case presentation)”. The actuality of genetic tests in patients with persistent muco-cutaneous candidiasis was disclosed by Prof. Laszlo Marody in his lecture “Discovery of Novel Genetic Etiologies of Chronic Mucocutaneous Candidiasis Disease”. His discourse was highly appreciated by attending doctors. Dr. Mihaela Bataneant, University “Victor Babes”, Timișoara, România also participated at the symposium with two reports „Recurrent fever associated with pancytopenia – relevant for primary immunodeficiency” and “Diagnostic alorythm in primry immunodeficiencys through a clinical case”.

Finally, we approved the following objectives:

1. To organize a PID Center, that will assure early diagnosis, treatment, supervision and counseling of PID patients.
2. To ensure the patients with PID with the necessary treatment (i/v immunoglobulin in high titers).
3. To found the Republican Society of PID for doctors and patients.
4. To apply PID patient’s registry.
5. To register ourselves as ESID members.
6. To register the PID patients in ESID.

Liudmila Cerempei



Liudmila Cerempei and her audience

J Project Meetings 2004-2014

1. **Targu Mures**, Romania, March 11–12, 2004; Organizer: *Csilla Todea*
2. **Prague**, Czech Republic, May 10–11, 2004; Organizer: *Anna Sediva*
3. **Belgrade**, Serbia/Montenegro, June 11–12, 2004; Organizer: *Srdjan Pasic*
4. **Skopje**, Macedonia, September 17–18, 2004; Organizer: *Katarina Stavrik*
5. **Kiev**, Ukraine, November 18–19, 2004; Organizer: *Alla Volokha*
6. **Zakopane**, Poland, December 16–17, 2004; Organizer: *Ewa Bernatowska*
7. **Sofia**, Bulgaria, April 15–16, 2005; Organizer: *Elissaveta Naumova*
8. **Prague**, Czech Republic, May 9–10, 2005; Organizer: *Anna Sediva*
9. **Gdansk**, Poland, September 23–24, 2005; Organizer: *Ewa Bernatowska*
10. **Debrecen**, Hungary, November 4–5, 2005; Organizer: *László Maródi*
11. **Lviv**, Ukraine, November 25, 2005; Organizer: *Larysa Kostyuchenko*
12. **Oradea**, Romania, December 20, 2005; Organizer: *Zoltán Ellenés*
13. **Debrecen**, Hungary, January 16, 2006; Organizer: *László Maródi*
14. **Prague**, Czech Republic, May 8–9, 2006; Organizer: *Anna Sediva*
15. **Bucharest**, Romania, June 9–10, 2006; Organizer: *Nicolae Iagaru*
16. **Minsk**, Belarus, November 9–10, 2006; Organizer: *Mikhail Belevtsev*
17. **Debrecen**, Hungary, December 8–9, 2006; Organizer: *László Maródi*
18. **Zaporozhzhye**, Ukraine, Apr 19–20, 2007; Organizer: *Liudmyla Chernyshova*
19. **St. Petersburg**, Russia, May 29–30, 2007; Organizer: *Marina Guseva*
20. **Ohrid**, Macedonia, September 20–21, 2007; Organizer: *Kristina Mironska*
21. **Iasi**, Romania, October 12–13, 2007; Organizer: *Aurica Rugina*
22. **Ljubljana**, Slovenia, November 16–17, 2007; Organizer: *Tadej Avcin*
23. **Odessa**, Ukraine, April 10, 2008; Organizer: *Liudmyla Chernyshova*
24. **Sunny Beach**, Bulgaria, May 22–23, 2008; Organizer: *Elissaveta Naumova*
25. **Sarajevo**, Bosnia-Herzegovina, October 10–11, 2008; Organizer: *Velma Mulaosmanovic*
26. **Chisinau**, Rep. of Moldova, Oct. 31–Nov. 1, 2008; Organizer: *Lyudmila Cerempei*
27. **Riga**, Latvia, November 27–28, 2008; Organizer: *Tatjana Prokofjeva*
28. **Timisoara**, Romania, March 19–20, 2009; Organizer: *Margit Serban*
29. **Sevastopol**, Ukraine, April 9–10, 2009; Organizer: *Liudmyla Chernyshova*
30. **Ekaterinburg**, Russia, May 13–14, 2009; Organizer: *Irina Tuzankina*
31. **St. Petersburg**, Russia, June 9–10, 2009; Organizer: *Marina Guseva*
32. **Konya**, Turkey, June 25–26, 2009; Organizer: *Ismail Reisli*
33. **Krakow**, Poland, September 24–25, 2009; Organizer: *Danuta Kowalczyk*
34. **Tallinn**, Estonia, October 7–8, 2009; Organizer: *Sirje Velbri*
35. **Ljubljana**, Slovenia, October 9–10, 2009; Organizer: *Natasa Toplak*
36. **Tehran**, Iran, October 11–12, 2009; Organizer: *Asghar Aghamohammadi, Nima Rezaei*
37. **Cairo**, Egypt, October 18–19, 2009; Organizer: *Nermeen Galal*
38. **Minsk**, Belarus, October 22–23, 2009; Organizer: *Mikhail Belevtsev*
39. **Tirana**, Albania, April 8–9, 2010; Organizer: *Georgina Kuli-Lito*
40. **Porto Marina**, Egypt, April 16–17, 2010; Organizer: *Aisha El-Marsafy*
41. **Kharkov**, Ukraine, May 19–20, 2010; Organizer: *Liudmyla Chernyshova*

42. **Zlatibor**, Serbia, June 1–2, 2010; Organizer: *Srdjan Pasic*
43. **Zagreb**, Croatia, July 2–3, 2010; Organizer: *Darko Richter*
44. **Chelyabinsk**, Russia, August 30–31, 2010; Organizer: *Irina Tuzankina*
45. **Budapest**, Hungary, September 9–10, 2010; Organizers: *László Maródi, Miklós Szolnoky*
46. **Tehran**, Iran, October 11–12, 2010; Organizers: *Asghar Aghamohammadi, Nima Rezaei*
47. **Pushkin Hills**, Russia, February 3–4, 2011; Organizers: *Marina Guseva, Areg Totolian*
48. **Zakopane**, Poland, February 24–26, 2011; Organizer: *Ewa Bernatowska*
49. **Oradea**, Romania, March 1, 2011; Organizer: *Ladislau Ritli*
50. **Debrecen**, Hungary, March 10–11, 2011; Organizer: *László Maródi*
51. **Moscow**, Russia, March 31–April 1, 2011; Organizer: *Irina Kondratenko*
52. **Baku**, Azerbaijan, April 22–23, 2011; Organizer: *Gulnara Nasrullayeva*
53. **Bratislava**, Slovakia, April 28–29, 2011; Organizer: *Peter Ciznar*
54. **Lutsk**, Ukraine, May 11–12, 2011; Organizer: *Liudmyla Chernyshova*
55. **Sibiu**, Romania, June 10–11, 2011; Organizer: *Sorin Iurian*
56. **Chelyabinsk**, Russia, June 26–27, 2011; Organizer: *Irina Tuzankina*
57. **Ohrid**, Rep. Macedonia, October 8, 2011; Organizer: *Kristina Mironska*
58. **Tehran**, Iran, October 15, 2011; Organizer: *Nima Rezaei*
59. **Trabzon**, Turkey, November 19–20, 2011; Organizers: *Fazil Orhan, Ismail Reisli*
60. **Riga**, Latvia, November 26, 2011; Organizer: *Tatjana Prokofjeva*
61. **Lillafüred**, Hungary, January 20–21, 2012; Organizer: *Miklós Szolnoky*
62. **Budapest**, Hungary, January 27–28, 2012; Organizer: *Miklós Szolnoky, Judit Szabó*
63. **Rostov-on-Don**, Russia, March 16, 2012; Organizer: *Ludmila Sizyakina*
64. **Moscow**, Russia, March 29, 2012; Organizer: *Irina Kondratenko*
65. **Chernigov**, Ukraine, April 5–6, 2012; Organizer: *Liudmyla Chernyshova*
66. **Tallinn**, Estonia, April 19, 2012; Organizer: *Krista Ress*
67. **Tehran**, Iran, April 22, 2012; Organizers: *Asghar Aghamohammadi, Nima Rezaei*
68. **Bratislava**, Slovakia, April 27, 2012; Organizers: *Peter Ciznar*
69. **Diyarbakir**, Turkey; May 12, 2012; Organizer: *Ismail Reisli*
70. **Prague**, Czech Republic; May 14–15, 2012; Organizer: *Anna Sediva*
71. **Konya**, Turkey; May 26–27, 2012; Organizer: *Ismail Reisli*
72. **Warsaw**, Poland; 2012; Organizer: *Ewa Bernatowska*
73. **Sibiu**, Romania; June 1–2, 2012; Organizer: *Sorin Iurian*
74. **Tzigov Chark**; June 8–9, 2012; Organizer: *Elissaveta Naumova*
75. **Astana City**, Kazakhstan, June 12–15, 2012; Organizer: *Panteley Popandopulo*
76. **Tyumen**, Russia–Siberia, July 1–2, 2012; Organizer: *Irina Tuzankina*
77. **Ljubljana**, Slovenia, September 14–15, 2012; Organizer: *Tadej Avcin*
78. **Tehran**, Iran, October 13–14, 2012; Organizer: *Nima Rezaei*
79. **Timisoara**, Romania, October 25–26, 2012; Organizer: *Mihaela Bataneant*
80. **Minsk**, Belarus, November 17, 2012; Organizer: *Mikhail Belevtsev*
81. **Ufa**, Russia, November 23, 2012; Organizer: *Irina Tuzankina*
82. **Sanliurfa**, Turkey, November 25, 2012; Organizer: *Ismail Reisli*
83. **Krakow**, Poland, December 15, 2012; Organizer: *Ewa Bernatowska*

84. **Mukachevo**, Ukraine, April 18, 2013; Organizer: *Liudmyla Chernyshova*
 85. **Tehran**, Iran, April 23, 2013; Organizer: *Nima Rezaei*
 86. **Bratislava**, Slovakia, April 24, 2013; Organizer: *Peter Ciznar*
 87. **Debrecen**, Hungary, April 26, 2013; Organizer: *László Maródi*
 88. **Ljubljana**, Slovenia, May 9, 2013; Organizer: *Tadej Avčin*
 89. **Prague**, Czech Republic, May 13–14, 2013; Organizer: *Anna Sediva*
 90. **Moravia**, Czech Republic, May 31, 2013; Organizers: *Jiri Litzman, Anna Sediva*
 91. **Budapest**, Hungary, May 31–June 1, 2013; Organizer: *László Maródi*
 92. **Cracow**, Poland, September 20–21, 2013; Organizer: *Ewa Bernatowska*
 93. **Szolnok**, Hungary, October 5, 2013; Organizer: *Zsuzsa Horváth*
 94. **Tehran**, Iran, October 12–13, 2013; Organizer: *Nima Rezaei*
 95. **Ptuj**, Slovenia, October 18–19, 2013; Organizer: *Tadej Avčin*
 96. **Krasnoyarsk**, Siberia, Russia, October 22, 2013; Organizer: *Irina Tuzankina*
 97. **Malatya**, Turkey, November 16, 2013; Organizer: *Ismail Reisli*
 98. **Budapest**, Hungary, November 22, 2013; Organizer: *László Maródi*
 99. **Pushkin Hills**, Russia, February 5, 2014; Organizer: *Areg Totolian, Marina Guseva*
 100. **Antalya**, Turkey, March 12–14, 2014; Organizer: *László Maródi, Ismail Reisli*
 101. **Antalya**, Turkey, March 15, 2014; Organizer: *Ismail Reisli*
 102. **Zagazig**, Egypt, April 17, 2014; Organizer: *Mohamed Almaky*
 103. **Tehran**, Iran, April 22–29, 2014; Organizer: *Nima Rezaei, Asghar Aghamohammadi*
 104. **Poltava**, Ukraine, April 24–25, 2014; Organizer: *Liudmyla Chernyshova*
 105. **Bratislava**, Slovakia, April 25, 2014; Organizer: *Peter Ciznar*
 106. **Debrecen**, Hungary, April 26, 2014; Organizer: *László Maródi*
 107. **St. Petersburg**, Russia, May 28, 2014; Organizer: *Tatiana Nasyrina*
 108. **Varna**, Bulgaria, October 6, 2014; Organizer: *Elissaveta Naumova*
 109. **Catez**, Slovenia, October 11–12, 2014; Organizer: *Tadej Avčin*
 110. **Tehran**, Iran, October 18–19, 2014; Organizer: *Nima Rezaei*
 111. **Yekaterinburg**, Russia, October 24–25, 2014; Organizer: *Irina Tuzankina*
 112. **Warsaw**, Poland, October 24–25, 2014; Organizer: *Ewa Bernatowska*
 113. **Pardubice**, Czech Republic, November 11, 2014; Organizer: *Anna Sediva*
 114. **Nizhny Novgorod**, Russia, November 26, 2014; Organizer: *Tatiana Nasyrina*
 115. **Sibiu**, Romania, November 27–28, 2014; Organizer: *Sorin Iurian*
 116. **Erzurum**, December 6, 2014; Organizer: *Dilara Kocacık Uygün, Demet Hafizoğlu*
 117. **Chisinau**, Moldova, December 12, 2014; Organizer: *Liudmila Cerempei*
-



Cambridge, UK

THE J PROJECT

Steering Committee Members

1. Albania	Tirana	Georgina Kuli-Lito gkuli_lito@hotmail.com
2. Azerbaijan	Baku	Gulnara Nasrullayeva g.nasrullayeva@hotmail.com
3. Belarus	Minsk	Mikhail Belevtsev belevtsev_m@mail.ru
4. Bosnia-Herzegovina	Sarajevo	Velma Selmanović velmamulaosmanovic@hotmail.com
5. Bulgaria	Sofia	Elissaveta Naumova naumovaej@gmail.com
6. Croatia	Zagreb	Jadranka Kelečić jkelecic17@gmail.com
7. Czech Republic	Prague	Anna Šediva anna.sediva@fnmotol.cz
8. Czech Republic	Brno	Jiří Litzman jiri.litzman@fnusa.cz
9. Egypt	Cairo	Aisha El-Marsafy aisha_mars@hotmail.com
10. Estonia	Tallinn	Sirje Velbri velbrisir@hotmail.com
11. Estonia	Tallinn	Krista Ress krista.ress@itk.ee
12. Hungary	Debrecen	László Maródi lmarodi@med.unideb.hu
13. Hungary	Debrecen	Melinda Erdős melinda.erdos@yahoo.de
14. Hungary	Debrecen	Beáta Tóth toth.beata.anna@freemail.hu
15. Iran	Tehran	Asghar Aghamohammadi aghamohammadi@Tums.ac.ir
16. Iran	Tehran	Nima Rezaei rezaei_nima@yahoo.com
17. Kazakhstan	Astana City	Nurzhan Otarbayev otarbaev@rambler.ru
18. Latvia	Riga	Tatjana Prokofjeva monja@balticom.lv
19. Lithuania	Kaunas	Brigita Sitkauskienė brigita.sitkauskienė@kaunoklinikos.lt
20. Macedonia	Skopje	Kristina Mironska mironska2002@yahoo.com
21. Moldova	Chisinau	Liudmila Cerempei lcerempei@rambler.ru
22. Poland	Warsaw	Ewa Bernatowska ewa.bernatowska@gmail.com

23. Poland	Warsaw	Malgorzata Pac malgorzata.pac@wp.pl
24. Poland	Cracow	Anna Szaflarska miszafla6@wp.pl
25. Poland	Warsaw	Malgorzata Skomska malgosia.skomska@gmail.com
26. Romania	Timisoara	Mihaela Bataneant mihabataneant@yahoo.com
27. Romania	Timisoara	Margit Serban mserban@spitalcopiitm.ro
28. Russia	St. Petersburg	Marina Guseva gusevamarina@mail.ru
29. Russia	Moscow	Irina Kondratenko ikondratenko@rambler.ru
30. Russia	Chelyabinsk	Alexander Pishchalnikov pau6277@yandex.ru
31. Russia	Ekaterinburg	Irina Tuzankina ituzan@yandex.ru
32. Serbia	Belgrade	Srdjan Pašić pasics@ikomline.net
33. Slovakia	Bratislava	Peter Čiznár ciznar@mail.t-com.sk
34. Slovenia	Ljubljana	Tadej Avčin tadej.avcin@kclj.si
35. Turkey	Konya	İsmail Reisli ireisli@hotmail.com
36. Turkey	Konya	Zafer Çalışkaner caliskaner@yahoo.com
37. Ukraine	Kiev	Anastasiia Bondarenko nastyabond@bk.ru
38. Ukraine	Kiev	Liudmyla Chernyshova chernyshova@ukr.net
39. Ukraine	Lviv	Larysa Kostyuchenko lkostyuchenko@ukr.net
40. Ukraine	Kiev	Alla Volokha volokha@gmail.com



*Delegates from Ukraine and Hungary in Antalya.
L → R Alla Vokha, Adrien Sarkadi, Beáta Soltész, Anastasiia Bondarenko, Liudmyla Chernyshova, Beáta Tóth, Gabi Csorba, Zsuzsa Horváth*

Genetic testing in the Debrecen PID Reference Center

PRIMARY IMMUNODEFICIENCIES (*GENES*)

1. X-linked agammaglobulinemia (*BTK*)
2. AR agammaglobulinemia (*IGHM*)
3. Common variable immunodeficiency subtype 1 (*ICOS*)
4. Common variable immunodeficiency subtype 2 (*TACI*)
5. X-linked hyper-IgM syndrome (*TNSF5=CD40L*)
6. Hyper-IgM syndrome (*UNG*)
7. Activation induced cytidine deaminase deficiency (*AICDA*)
8. Recombination-activating gene 1 deficiency (*RAG1*)
9. Recombination-activating gene 2 deficiency (*RAG2*)
10. X-linked severe combined immunodeficiency (*SCID1*) (*IL-2RG*)
11. T-, B+, NK+ severe combined immunodeficiency (*IL7RA*)
12. X-linked chronic granulomatous disease (*CYBB*)
13. Autosomal recessive chronic granulomatous disease (*NCF1*)
14. Wiskott-Aldrich syndrome (*WAS*)
15. AR-Hyper-IgE syndrome (*TYK2*)
16. AD-Hyper-IgE syndrome (*STAT3*)
17. Severe congenital neutropenia type 1 (*ELANE*)
18. Severe congenital neutropenia type 2 (*GFI1*)
19. Severe congenital neutropenia type 3 (*HAX1*)
20. Severe congenital neutropenia type 4 (*G6PC3*)
21. Hereditary neutrophilia (*CSF3R*)
22. Shwachman-Diamond syndrome (*SBDS*)
23. Familial hemophagocytic lymphohistiocytosis syndromes (*PRF1*)
24. Warts, hypogammaglobulinemia, infections and myelokathexis (*WHIM*) (*CXCR4*)
25. Complement component 2 deficiency (*C2*)
26. X-linked lymphoproliferative syndrome (*SH2D1A*)
27. X-linked lymphoproliferative syndrome type 2 (*XIAP*)
28. DOCK8 deficiency (*DOCK8*)
29. Nijmegen breakage syndrome (*NBN*)
30. Chronic mucocutaneous candidiasis disease – DC-SIGN (*CLEC4L*)
31. Chronic mucocutaneous candidiasis disease – dectin-1 (*CLEC7A*)
32. Chronic mucocutaneous candidiasis disease – dectin-2 (*CLEC6A*)
33. Chronic mucocutaneous candidiasis disease (*MRC1*)
34. Chronic mucocutaneous candidiasis disease (*CARD9*)
35. Chronic mucocutaneous candidiasis disease (*STAT1*)
36. Chronic mucocutaneous candidiasis disease (*IL17RA*)
37. Chronic mucocutaneous candidiasis disease (*IL17F*)
38. Chronic mucocutaneous candidiasis disease (*TRAF3IP2*)
39. Cartilage hair hypoplasia (*RMRP*)
40. X-linked Dyskeratosis congenita (*DKC1*)
41. Autoimmune lymphoproliferative syndrome type 1A (*CD95=FASG*)

42. Autoimmune lymphoproliferative syndrome type 1B (*CD178=FASLG*)
43. Autoimmune polyendocrine syndrome (*AIRE*)
44. Hyper-IgD syndrome (*MVK*)
45. IL18R1 deficiency (*IL18R1*)
46. X-linked ectodermal dysplasia, incontinentia pigmenti (*IKBKG*)
47. Familial Mediterranean fever (*MEFV*)
48. Familial Mediterranean fever (*CIAS1*)
49. Familial Mediterranean fever (*TNFRSF1A*)
50. Mendelian susceptibility to mycobacterial disease (*IL12RB1*)
51. Mendelian susceptibility to mycobacterial disease (*IL12B*)
52. Mendelian susceptibility to mycobacterial disease (*IFNGR1*)
53. Mendelian susceptibility to mycobacterial disease (*IFNGR2*)
54. MYD88 deficiency (*MYD88*)
55. WIP deficiency (*WIPF1*)
56. Ikaros deficiency (*IKAROS*)
57. X-linked autoimmunity-allergic dysregulation syndrome (*IPEX*) (*FOXP3*)
58. IRAK4 deficiency (*IRAK4*)
59. PIK3CD deficiency (*PIK3CD*)

LYSOSOMAL STORAGE DISEASES (*GENES*)

60. Fabry disease (*GLA*)
61. Gaucher disease (*GBA*)
62. Niemann-Pick Disease (*SMPD1*)
63. Morquio A syndrome (*GALNS*)

OTHER DISEASES (*GENES*)

64. Von Hippel-Lindau Syndrome (*VHL*)
65. X-linked adrenal hypoplasia congenital (*DAX1*)
66. Galectin 3 deficiency (*LGALS3*)
67. Alström syndrome (*ALMS1*)
68. Atopic eczema (*FLG*)
69. Myhre-syndrome (*SMAD4*)



Richárd Juha: At the gate of dream

Links

The J Project (JP)

<http://jproject.dote.hu>

Jeffrey Modell Foundation (JMF)

<http://www.info4pi.org>

European Society for Immunodeficiencies (ESID)

<http://www.esid.org>

International Patient Organisation for Primary Immunodeficiencies (IPOPI)

www.ipopi.org

Immune Deficiency Foundation

<http://primaryimmune.org>

International Nursing Group for Immunodeficiencies (INGID)

<http://www.ingid.org>

African Society for Immunodeficiencies (ASID)

<http://www.asid.ma>

Latin American Society for Immunodeficiencies (LASID)

<http://www.lasid.org>

International Union of Immunological Societies (IUIS)

<http://www.iuisonline.org>

Federation of Clinical Immunology Societies (FOCIS)

<http://www.focisnet.org>

European Federation of Immunological Societies (EFIS)

<http://www.efis.org>

Clinical Immunology Society (CIS)

<http://www.clinimmsoc.org>

American Academy of Allergy & Asthma and Immunology (AAAAI)

<http://www.aaaaai.org>

