The J Project
2009

J Countries
The J Project
2009

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March 2010
I am delighted to introduce the 2009 J Project Booklet reporting on primary immunodeficiency conferences organized in Eastern and Central Europe over the last year. Since the start of the J Project five years ago in Targu Mures, Romania, there have been five to six J Meetings per year. In 2009, considerable progress was made in terms of the number of J Project conferences held and the Project crossed the frontiers of Europe. The first meeting was organized by Margit Serban and Mihaela Bataneant, in Timisoara and Buzias, of the site of the Christian Serban memorial. Margit Serban and Ioan Gherghina very sensitively described advances and difficulties in PID care in various regions of Romania. The next meeting was organized by Liudmyla Chernishova, the 2010 winner of the “Pro Cooperatione” award of the Debrecen Committee of the Hungarian Academy of Sciences, in the city of heroes, Sevastopol, in the Ukraine. Ismail Reisli, the head of the PID centers in the Konya region, took notes from Liudmyla Chernishova on how best to organize a J Project meeting. He then invited the J Project community to Konya in June. Ismail Reisli provided us with a great scientific program, and his exceptional hospitality ensured that we were touched by the wisdom of Mevlana, the culture of Sema ceremony, and the impressive history of the Anatolian Selcuk Empire. However, this was not the first excursion of the J Project into Asia. The Ekaterinburg PID center headed by Irina Tuzankina decided to join the J Project and she organized a wonderful meeting in Regional Children’s Hospital No. 1 in the city, from which you can see Europe to the left and Asia to the right. An agreement was reached with the leading physicians in Ekaterinburg and PID experts from Moscow that Ekaterinburg, the gateway to Asia, should be established as a PID center for Siberia. The Cracow J Meeting was memorable because it was a really a subspeciality-oriented conference of immunologists and hematologists interested
in PID. It seems to me that this meeting was the first to show how subspeciality-specific warning signs of PID may facilitate the earlier recognition of primary immunodeficiencies. The meeting in Tallin, Estonia, was organized by Sirje Velbri, one of the most enthusiastic and devoted members of the J Project community. The highlights of the program she put together included presentations on APECED (APS-1), a PID studied in Nordic countries by a number of clinical research groups. The meeting in Ljubljana, organized jointly by Tadej Avcin and Natasa Toplak, was entirely devoted to autoinflammatory diseases, a recently classified new group of PID. The invitation of internationally recognized scientists in the field ensured that this meeting attained high standards. After the J Project meeting in Turkey, it was a real pleasure to see Nima Rezaei and Asghar Aghamohammadi organized the first J Meeting in Iran, with invited speakers from Europe. Remarkable progress in the field of PID care in Iran was presented and discussed. Aisha El Marsafy of Cairo University also gave me a pleasant surprise when she informed me that she wished to organize the first J Meeting in Egypt. The meeting was elegant, of a high standard and very stimulating, thanks to the support of the PID community for this event. We ended the long list of J meetings this year in Minsk, Belarus, where Michael Belevtsev, a molecular biologist has long been one of the advocates of children with PID. It is hard to believe that we could have a better and more successful year than 2009 this year, but I hope to be proved wrong on this point.

March 15, 2010

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

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

The Centre provides modern educational utilities, demonstrational materials, and rooms that are suitable for practical seminars as well as lecture rooms. ECE IPI CTR functions as an institutional and regional Infectious and Pediatric Immunology Centre. 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 Euroregion, one of the main aims and functions of which is the formulation of educational and scientific co-operation within the region of East-Central-Europe.
The J Project

PID Awareness Meetings in 2009

CITY, Country                      DATE
1. Timisoara & Buzias (Romania)     March 19-20
2. Sevastopol (Ukraine)             April 9-10
3. Yekaterinburg (Russia)           May 13-14
4. St. Petersburg (Russia)          June 9-10
5. Konya (Turkey)                   June 18-19
6. Krakow (Poland)                  September 24-25
7. Tallinn (Estonia)                October 7-8
8. Ljubljana (Slovenia)             October 9-10
9. Tehran (Iran)                    October 11-12
10. Cairo (Egypt)                   October 18-19
11. Minsk (Belarus)                 October 22-23

Aims

1. To organize professional meetings on PID and related diseases in regions of East-European countries with low number of registered PID patients.
2. To discuss diagnostic and therapeutic practices and problems, and to define specific areas to be improved and to be supported by other European groups, institutions, companies, and foundations.
3. Updating national PID registries.
4. Establishing PID professional working groups.
5. Establishing PID patients, groups.

Structure of the Meetings

1. Informal discussion on the day before the meeting.
2. Introductory lectures by invited and local speakers.
3. Case reports mostly by local speakers.
4. PID WG and patients’ group activity in the country.
Timisoara & Buzias, Romania
19-20 March, 2009

Host:
Margit Serban (mserban@rdstm.ro)

Venue:
3rd Pediatric Clinic, Timisoara & Central Medical Clinic “Cristian Serban”

Main Topic:
Primary immunodeficiencies from theory to practice, Bone marrow transplantation in PIDs, PID registry in Romania
**PROGRAM**

19 March

Opening ceremony

**Session I**

The molecular genetic approach to primary immunodeficiency diseases  
*Maródi L. (Debrecen)*

Bone marrow transplant in PIDs  
*Serban M. (Timisoara)*

XLP: an insidious primary immunodeficiency disorder  
*Erdős M. (Debrecen)*

PID registry in Romania  
*Batanean M. (Timisoara)*

**Session II - Case presentations**

Type I and IV hypersensibility in a case with IgA and α1-antitripsin deficiency  
*Iurian S. (Sibiu)*

Diagnosis of the Wiskott-Aldrich syndrome without genetic analysis – “to be or not to be”  
*Iagaru N. (Bucharest)*

A case of CVID in a family with a history of type 1 diabetes mellitus  
*Miron N. (Cluj-Napoca)*

Hereditary angioedema: a subestimated PID  
*Moldovan D. (Targu Mures)*

A CVID case presentation  
*Miculschi G. (Oradea)*

The evolution in a case of agammaglobulinemia  
*Marc M. (Cluj-Napoca)*

General assembly meeting of Romanian PID working group

**Session III - Case presentations**

Successful hematopoietic stem cell transplantation for chronic granulomatous disease complicated with pulmonary and vertebral aspergillosis – a challenge for treatment. A case report  
*Jinca C. (Timisoara)*

Severe combined immune deficiency by purin nucleotid phosphorylase deficiency – first case diagnosed in Romania  
*Cochino A. (Bucharest)*

Hyper-IgD syndrome – a case presentation  
*Ellenes J.Z. (Oradea)*

Limits and diagnosis difficulties in a case of muco-cutaneous hypopigmentation  
*Boeriu E. (Timisoara)*

A case with IgA deficiency, celiac disease, Ehlers-Danlos syndrome, recurrent wheezing and hypogonadism  
*Bulucea D. (Craiova)*

Evolution in a case with IgM deficiency and hyper IgE  
*Marc M. (Cluj-Napoca)*

Diagnosis difficulties in a case with chronic pneumonia and hypogammaglobulinemia  
*Batanean M. (Timisoara)*

Iron deficiency anemia that mimic minor β-talasemia in a child with IgA deficiency, recurrent parotiditis and dysmorphism  
*Bulucea D. (Craiova)*

Conclusions
SUMMARY AND CONCLUSIONS

The 28th Meeting of the J Project on Primary Immunodeficiencies was held on 19-20 March 2009 in Timisoara and Buzias (Romania), two cities located in the western part of the country, along the borders of Hungary and Serbia.

More than 70 participants, dedicated immunologists but also young doctors, representing all the historical regions of Romania (Banat, Transylvania, Muntenia, Oltenia, Moldova, Bihor and Maramures) attended the meeting. The meeting was organized within the framework of the J Project. Prof. László Maródi, and Melinda Erdős from Debrecen, Hungary also participated, and a colleague from the Republic of Moldova also joined us. The agenda of the conference which comprised two parts was generous. The first part was held in Timisoara, with the following topics: Preconference educational session, and Patient’s Group activity. The second part was organized in Buzias, with the following main topics: Bone marrow transplantation in PIDs, PID registry in Romania, and PID case presentations.

A pre-conference educational session was also organized dedicated to post-graduate education of young physicians (PhD students, residents, general practitioners, pediatricians, and laboratory specialists) with the aim of providing a useful tool for a comprehensive approach of immunological processes and immunodeficiencies. It was followed by patients’ group discussions regarding difficult and controversial clinical situations (Chediak-Higashi syndrome, CVID syndrome with severe atypical mycobacteria lung infection, cluster of autoimmunity in the family of a child with CVID syndrome) focusing on diagnosis and differential diagnosis, concluding on the importance of bio-molecular investigation for a definite diagnosis.

The second day meeting took place in Buzias. Prof. László Maródi presented a review on the history of J Project, highlighting its achievements, development and perspectives. Its hallmark is the steadily increasing number of participant countries; started in Hungary, implying at first the neighboring countries; and now it succeeded to expand the project further in Asia (Yekaterinburg) and Africa (Cairo). The J project spread out knowledge and skills in the field of diagnosis, intending an improved approach to therapy of these rare diseases. All these involved countries maintained an active connection with Debrecen, the center of J Project, and could share progress in this field, having access to a molecular diagnosis.

Prof. Margit Serban, president of the local organizing committee, presented general data regarding to BMT in PID – an important and the only curative treatment for many severe life threatening PIDs. Dr. Michaela Bataneant, the secretary of the local organization committee, informed the audience about the progress and changes of the National PID registry in Romania. Interesting case reports about the first PNP Romanian patient, Wiskott Aldrich syndrome, XLP syndrome, hyper-IgD syndrome were also presented.
As deliverables, all participants received a booklet with the main abstracts of the presentations made at the meeting and an electronic course material based on ESID recommendations for diagnostic approach of PID. The meeting was officially recognized by the local Medical Collegium and accredited with 10 CME credits.

The meeting was well appreciated by the participants and received feedbacks have ascertained the importance of such an event for the intellectual and scientific development of the medical community in this region of Europe. All such meetings will be welcomed in the future in Romania and periodic workshops on PID will definitely benefit all those involved in these programs.

Margit SERBAN & Mihaela BATANEANT

To the memory of Christian Serban

Timisoara
Sevastopol, Ukraine
9-10 April, 2009

Host:
Liudmyla Chernyshova (chernyshova@ukr.net)

Venue:
National Medical Academy of Post-graduate Education named after P.L. Shupik Dep. of Pediatric Infectious Diseases and Clinical Immunology

Main Topic:
Diagnostics and management of children with primary immunodeficiencies
PROGRAM

9 April

Opening Ceremony

Hyper IgE syndrome (Job’s syndrome)
Bondarenko A. (Kiev)

Clinic and treatment of primary immunodeficiency of IL-12 receptor
Boyko Y. (Lviv)

Neutropenia syndrome. Differential diagnostics and treatment
Samarin D. (Kiev)

Immunologic methods in PID diagnostics
Chernyshov V. (Kiev)

Chronic mucocutaneous candidosis
Nikonets L. (Donetsk)

Principles of diagnostics of PIDs
Kondratenko I. (Moscow)

Hyper IgM syndrome: case report
Lapti F. (Kiev)

Congenital neutropenia: case reports
Halyabar O. (Lviv)

10 April

General Assembly of Ukrainian Association for “Pediatric Immunology”

Closing Ceremony

PID in Ukraine: Early diagnostic and treatment possibilities
Chernyshova L. (Kiev)

Molecular genetic approach to PIDs
Maródi L. (Debrecen)

Ukrainian immunoglobulin preparations
Peculiarities of manufacturing
Ryadskaya L. (Kiev)

Diagnostics and management of children with primary IgA immunodeficiency
Volokha A. (Kiev)

Dinner

XLP: an insidious PID
Erdős M. (Debrecen)

Primary combined immunodeficiencies
Kostyuchenko L. (Lviv)

The frequency and characteristics of PID in Turkey
Reisli I. (Konya)
SUMMARY AND CONCLUSIONS

According to East-European Initiative “J Project” the 5th Ukrainian Symposium «Primary Immunodeficiencies» has taken place in Sevastopol (Crimea, Ukraine) at April 8-9 2009. The members of the organizing committee were Prof. L. Chernyshova, Prof. L. Maródi, and Dr. E. Kushnir.

The detectibility of primary immunodeficiencies (PID) has improved during the last few years in Ukraine, but the lower occurrence of PID in compare to other countries is still a big problem. Ukraine is a large country and the development of immunology in the different regions is very different. Crimea or the Autonomous Republic of Crimea is a peninsula situated on the south of Ukraine (northern coast of the Black Sea) which area is 26,200 square kilometres and its population is 1,973,185. Despite this we have very few patients from this area. The goal of seminar was to improve the knowledge about PIDs, including the diagnostic approach and treatment possibilities. More than 300 pediatricians from 25 regions of Crimea and chief immunologists from all regions of Ukraine took part in the meeting. The reports were done by Prof. L. Maródi, Prof. L. Chernyshova, leading immunologists of Ukraine and other invited speakers from Hungary and Turkey. In total in school-seminar work about 350 participants took part. The main topics of the presentations were the achievements in diagnostics of PIDs in Ukraine, the current possibilities of genetic diagnostics in the country, and the treatment of PIDs. Interesting clinical cases were also presented about antibody deficiencies, Duncan syndrome, combined immunodeficiency, hyper-IgE syndrome, and neutropenia.

Sevastopol has wide and interesting history. The Navy bases of Russia and Ukraine are also situated here. During the Crimea war between Russia and Turkey (1854-1855) famous author of “War and Peace” Lev Tolstoy defended one of the Sevastopol’s bastions. Guests also had a nice excursion to the Panorama of Crimean War museum.

Liudmyla CHERNYSHOVA
Local televisions were also interested in the meeting

VISITING SEVASTOPOL and BAKHCHISARAY

Panorama of Sevastopol

Visiting Panorama of Crimean War museum

BAKHCHISARAY

A memory place of Lev Tolstoy the author of “War and Peace”
Yekaterinburg, Russia
13-14 May, 2009

Host:
Irina Tuzankina (I.Tuzankina@iip.uran.ru)

Venue:
Regional Children Clinical Hospital №1, Yekaterinburg

Main Topic:
Primary immunodeficiencies: modern vision and problems

Irina Tuzankina and László Maródi (middle) with colleagues in front of the Regional Children Clinical Hospital №1 Yekaterinburg
PROGRAM

13 May

Acquaintance with the regional hospital and center of clinical immunology
Case studies of patients with PID
Excursions

Laboratory diagnostics of PID
Pashnina I.A. (Ekaterinburg)

Postmortal diagnostics of PID
Belikov E.S. (Ekaterinburg)

Coffee break

Marrow transplantation with PID: experience and opportunities
Balashov D.N. (Moscow)

Supervision of patient with severe combined immunodeficiency in post-transplantation period
Vyatkin I.N. (Ekaterinburg)

Differential diagnostics of patients with feverishness (syndromes and hypogranulocytosis)
Prodeus A.P. (Moscow)

Histiocytosis in regional register of Sverdlovsk regional
Perina F.G. (Ekaterinburg)

General Discussion

Closing the Conference

14 May

Welcome reception

Introduction - Salutatory address

The importance of primary immunodeficiencies in clinical medicine
Maródi L. (Debrecen)

Regional register of PID in Sverdlovsk region
Tuzankina I.A. (Ekaterinburg)

Fetus and newborns immune response development
Prodeus A.P. (Moscow)

Clinical difficulties in diagnostics of several PID forms
Vlasova E.V. (Ekaterinburg)

Laboratory diagnostics of PID
Pashnina I.A. (Ekaterinburg)

Postmortal diagnostics of PID
Belikov E.S. (Ekaterinburg)

Coffee break

Marrow transplantation with PID: experience and opportunities
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Supervision of patient with severe combined immunodeficiency in post-transplantation period
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General Discussion

Closing the Conference
SUMMARY AND CONCLUSIONS

On May 13-14, 2009, a J Project primary immunodeficiency (PID) meeting entitled “Primary immunodeficiencies: an emerging field of modern clinical medicine“ was organized at the Regional Center for Clinical Immunology of the Regional Clinical Childrens Hospital №1 in Yekaterinburg. Organizers of this PID event were Prof. I.A. Tuzankina, head of the Center, and Prof. V.A. Chereshnev, director of the Institute of Immunology and Physiology, and member of the Ural Branch of the Russian Academy of Sciences.

The conference was attended by Prof. L. Maródi from Debrecen, Hungary, and professors A.P. Prodeus, E.D Pashanov, and D.N. Balashov, all from Moscow. The more than 200 participants of the conference included guests from the Tyumen, Chelyabinsk, Perm, and Yekaterinburg regions, heads of departments of Ural State Academy of Medicine, doctors and heads of departments of Yekaterinburg hospitals, post-graduate students of various institutes, and residents and students of Ural State Medical Academy.

On the first day we organized a whole day clinical consultation and discussion of problematic PID cases cared at the Clinical Immunology, Onco-hematology and BMT Divisions of the Regional Clinical Childrens Hospital №1. Eg., a rare progressive neurodegenerative manifestation of X-linked agammaglobulinemia in a 5 year-old boy, a variant case of hyper-IgM syndrome in a 4 year-old girl, a young boy with ataxia-telangiectasia and a 16 year-old male patient with possible autoimmune lymphoproliferative syndrome were discussed, and diagnostic and treatment plans were made.

The second day the conference started with an introductory lecture by Prof. L. Maródi about “The importance of primary immunodeficiency patient care in clinical medicine”. The development of the normal immune responses in utero and in newborns, and the problem of the proper use of immunotropic and antibacterial drugs in the pediatric practice was presented by A.P. Prodeus, who also discussed the differential diagnostic approach of autoinflammatory diseases in children. The regional PID registry of the Yekaterinburg region that includes 154 cases of different syndromes was presented by Prof. I.A. Tuzankina. The current data of the registry about histiocytosis were summarized by F.G. Perina, and O.V. Streneva. The difficulties of the diagnosis of several PIDs were presented by E.V. Vlasova. The current possibilities in the laboratory and pathology diagnosis of PIDs in Yekaterinburg were described by I.A. Pashnina, E.S. Belikov, and J.M. Kobeleva. Lectures by Drs. Pashanov and Balashov were given on bone marrow and stem cell transplantation. Experiences and results of the local hospitals in bone marrow transplantation were also summarized. An interesting case with severe combined immunodeficiency was also presented by I.N. Vyatkin, and N.G. Maysheva).

At the end of the conference Prof. L. Maródi made the proposal to establish a center for molecular and genetic diagnostics of PID in Yekaterinburg, along with the one already existing in Moscow, to complete the diagnostic circle in the Ural region. Currently, there are diagnostic facilities and qualified specialists, but a large number of patients as well, who are waiting for medical assistance and verification of diagnosis in western Siberia. Prof. L. Maródi and representatives of the Ministry of Health of Yekaterinburg region got information about the structure of the regional center of clinical immunology. Prof. Maródi offered collaboration between the Debrecen and Yekaterinburg centers in further training of the medical doctors and in the molecular genetic tests.

Irina A. TUZANKINA
J PROJECT: in good direction without borders

Andrei Prodeus and László Maródi

Visiting the memorial of the Czar
St. Petersburg, Russia
9-10 June, 2009

Host:
Marina Guseva (gusevamarina@mail.ru)

Venue:
Anichkov Palace, St. Petersburg

Main Topic:
Early diagnosis of primary immunodeficiencies and improvement of patients’ quality of life
PROGRAM

9 June

Roundtable discussion
Patients with primary immunodeficiency disorders and organizations of patients
Drabwel J. (United Kingdom)
Kalinina N.M. (St. Petersburg)

IPOPI achievements in the last few years. Why it is important to be part of IPOPI?
Drabwel J. (United Kingdom)

Work experience of organization of patients with PID in Serbia
Koruga D. (Serbia)

National Association “Genetics”: Goals, achievements, projects
Karimova S.I. (St. Petersburg)

Organization of patients with disorders of PID in Russia
Moskalev M.I. (Moscow)

Rare diseases and orphaned medical technologies – a new paradigm of modern medicine
Sokolov A.A. (St. Petersburg)

Information portal on rare diseases, medicines to orphans and seldom applied medical technologies (information for patients)
Dembrovskiy V.N. (St. Petersburg)

Discussion

Diagnostics and conducting patients with primary immunodeficiency disorders in Russia
Prodeus A.P. (Moscow)

The Organization of rendering of medical aid to children with primary immunodeficiencies of Belorussia.
Information-analytical system of early revealing of patients with PID
Belevtsev M.V. (Minsk)

The Register of patients with PID in Sverdlovsk region: questions of diagnostics of PID, conducting patients and organization of the service
Tuzankina I. (Yekaterinburg)

The Register of patients with PID in Bashkorostan Republic
Hairullina R.M. (Ufa)

Information portal on rare diseases, medicines to orphans and seldom applied medical technologies (information for doctors)
Dembrovskiy V.N. (St. Petersburg)

DNA diagnostics of hereditary immunodeficiencies
Polyakov A.V. (Moscow)

Laboratory diagnostics of PID. Role of flow cytometry in diagnostics of PID
Sharapova S.O. (Minsk)

Break

Session 2

XLP: An insidious primary immunodeficiency disorder
Erdős M. (Debrecen)

Functional disorders of polymorphonuclear neutrophils. Clinical case of chronic granulomatosis disease
Kalinina N.M. (St. Petersburg)

Defects of fagocitosis as possible reasons of atopic diseases
Nisheva E.S. (St. Petersburg)
Hyper-IgE syndrome: the case report (from the syndrome of Iova to defect of STAT 3 gene)
Yartcev M.N. (Moscow)

Syndrome of Marshal
Baranova O.V. (Moscow)

The syndrome of chronic diarrhoea in children with PID
Cerempei L.G. (Chisinau)

Combination of XLA with Mohr-Tranebjaerg syndrome
Velbri S. (Tallinn)

Clinical case of Omenn syndrome
Volkova N.L. (St. Petersburg)

Complexities of identification of rare forms of PIDs: Skin-eye form of albinism with PID
Guseva M.N. (St. Petersburg)

Break

Session 3

A case of Wiskott-Aldrich syndrome in a 7 month old child
Prokoffjeva T. (Riga)

The molecular-genetic and clinical characteristic of group of patients with Wiskott-Aldrich syndrome
Sabirova S.S. (Moscow)

Features of surgical tactics in congenital immunodeficiencies with generalized microbacterial infection
Mushkin A.Y. (St. Petersburg)

Immunodeficiencies with chronic candidiasis of skin and mucous membranes
Shabashova N.V. (St. Petersburg)

Researches of molecular-genetic nature of FHL and XLP syndrome in groups of Russian patients
Poltavec N.A. (Moscow)

Vaccination of children with PID
Dovletbaeva G.A. (Ufa)

Imunogloblin. What is this? Questions of diagnostics, use and administration
Prodeus A.P. (Moscow)

Rational therapy with antibodies in primary immunodeficiencies
Bologov A.A. (Moscow)

Laboratory monitoring of immunoresolved therapy on the basis of immunoglobulins
Reznikov Y.P. (Moscow)
SUMMARY AND CONCLUSIONS

This conference was the second J project meeting in St. Petersburg. The first meeting was in May 2007, which was the beginning point of all J project meetings in Russia. The main purpose of the conference was the early diagnosis of primary immunodeficiencies and improvement of patients’ quality of life.

The roundtable discussion “Patients with primary immunodeficiency disorders and patient organizations” took place on June 9, 2009 in library of Anichkov Palace. Jose Drabwell, chairperson of IPOPI, talked about IPOPI achievements in the last few years and the importance to be take part in IPOPI. Dragana Koruga shared her experiences in the organization of the Serbian patient’s group. Two Russian patient organizations, the “National Association of Genetics” (St. Petersburg) and “Voluntary Society” (Moscow) talked about their goals, achievements and future projects. General aspects of diagnostics of primary immunodeficiencies and the treatment were presented in the report of Prof. A.A. Sokolov and M.N. Guseva. An information portal for rare diseases [www.rarediseases.ru](http://www.rarediseases.ru) was showed in two presentations.

Three sessions of the Conference were held on June 10, in Gold Saloon of Anichkov Palace. In the beginning of the first session Prof. L. Maródi talked about the J project and the new concepts in the field of immunodeficiencies. The report of Prof. A.P. Prodeus about diagnostics and PID patient care in Russia was also very interesting. Prof. I. Tuzankina presented the PID registry of the Sverdlovsk region of Russia. The report of A.V. Polyakov about the DNA diagnostics of hereditary immunodeficiencies was very informative. The reports of doctors from main hospitals from St. Petersburg and Moscow were also presented on this Conference. We thank our guests from Latvia, Estonia and Belorussia for giving talks about the information-analytical system of early recognition of PIDs (M.V. Belevtsev), role of flow cytometry in the diagnostics of PID (S.O. Sharapova), case reports of Wiskott-Aldrich syndrome (T. Prokofjeva), and combination of XLA with Mohr-Tranebjaerg syndrome (S. Velbri). The participants of the Conference received materials on diagnostics and treatment of PIDs.

The guests enjoyed our beautiful city during the excursion by boat during the night under the bridges. We thank companies Biotest and Oktafarm for help in organization the meeting. The Conference will undoubtedly improve the situation of diagnostics and treatment of patients with PIDs in St. Petersburg. We hope that cooperation between hospitals, centers in Russia and other countries will be continued.

Marina GUSEVA
Konya, Turkey
18-19 June, 2009

Host:
Ismail Reisli (ireisli@hotmail.com)

Venue:
Selcuk University Meram Medical Faculty, Conference Hall

Main Topic:
The difficulties in diagnosis, treatment and management of primary immunodeficiencies
PROGRAM

18 June
Opening lecture and Guitar concert

Living with CVID
Gozubatik G.

Immunologic organization in Turkey
Deniz G. (Istanbul)

PID diagnostics and treatment in Hungary
Maródi L. (Debrecen)

Warning signs in primary immunodeficiencies
Artac H. (Konya)

PIDs in Konya
Keles S. (Konya)

Coffee Break

Combined immunodeficiencies
Turul T. (Ankara)

B cell immunodeficiencies
Kütükçüler N. (Izmir)

Defects of Phagocyte function
Aksu G. (Izmir)

Defects of the complement proteins

Lunch

Defects of Innate immune system
Yeğin O. (Antalya)

Bone marrow transplantation in PID
İkincioğulları A. (Ankara)

Hyper-IgE syndrome
Barlan İ. (Istanbul)

Periodic fever syndromes
Sebnem Kılıc S. (Bursa)

Coffee Break

Experience of our center in PID
Aytekin C. (Ankara)

Asthma and chronic pulmonary disease in PID
Pekcan S. (Konya)

Hematologic complications in PID
Çalışkan U. (Konya)

Oncologic complications in PID
Köksal Y. (Konya)

Social program and Dinner

19 June
Sightseeing tour and Lunch
SUMMARY AND CONCLUSIONS

The J Project meeting entitled “The difficulties in diagnosis, treatment and management of primary immunodeficiencies” was held in Konya, on June 18-19, in 2009, which was the first J Project meeting in Turkey. The meeting was organized by Prof. L. Maródi and Assoc. Prof. I. Reisli within the framework of the J Project.

The main targets of the meeting were to improve the awareness of primary immunodeficiencies (PIDs) among the physicians, pediatricians and other specialists, to discuss PID diagnostic problems, clinical symptoms, treatments, and to clarify importance of early diagnosis of PID. We have also aimed to improve the awareness of PIDs among the medical students.

The first part of meeting was started with a lecture about immunologic organisation in Turkey by G. Deniz (President of Turkish Society of Immunology) from Istanbul University. Prof. L. Maródi reported on the diagnostics and treatment of PIDs in Hungary. In the continued part of first panel, H. Artaç (Selcuk University, Konya) was talked about the warning signs of PID and S. Keles (Selcuk University, Konya) was talked about “the PID in Konya”. The second session were dedicated to combined immunodeficiency, B cell deficiency, phagocyte system defects and complement defects presented by T. Turul (Hacettepe University, Ankara), N. Kutukculer (Ege University, Izmir), G. Aksu (Ege University, Izmir) and I. Reisli (Selcuk University, Konya), respectively. In the third part, I. Barlan (Marmara Univesity, Istanbul), S. Kilic (Uludag University, Bursa) and A. Ikinciogulları (Ankara University, Ankara) gave their lectures about hyper-IgE syndrome, periodic fever syndromes and bone marrow transplantation in PID, respectively. The last part of the meeting was about the complications of PID including the lungs (S. Peckan, Selcuk University), haematology (U. Caliskan, Selcuk University) and oncology (Y. Koksal, Selcuk University, Konya). During the panel, C. Aytekin (Sami Ulus State Hospital, Ankara) and O. Ardeniz (Ege University, Izmir) have also shared their experiences about the the follow-up and the complications of PID.
In the evening, the shuttle transferred the participants to a nice restaurant situated near to “Museum Mevlana” and they had a traditional Konya dinner after “Sema Ceremony”. During the sightseeing of Konya the participants visited the Mevlana Museum, the Alaaddin Mosque and Palace (from Anatolian Selcuk Empire), and Karatay and Ince Minare Medrese (Universities in 13th century in Konya).

We would like to thank all the speakers and participants for their contributions, especially Prof. L. Maródi and our main sponsor (GlaxoSmithKline) for their financial help which made it possible to organize this meeting.

Ismail REISLI
Krakow, Poland
24-25 September, 2009

Host:
Danuta Kowalczyk (dkowalcz@cm-uj.krakow.pl)

Venue:

Main Topic:
Hematological problems in primary immunodeficiencies
PROGRAM

25 September
Official check-in
Dinner

26 September
Opening Ceremony

Session I

Hematological aspects of primary immune deficiency
*Lange A.* (Wroclaw)

Primary immunodeficiencies: An evolving clinical discipline
*Maródi L.* (Debrecen)

Hematology disorders at antibodies deficiency
*Guseva M.* (St. Petersburg)

Hematological malignancies in children with primary immunodeficiencies - a single Romanian center report
*Bataneant M.* (Timisoara)

Autoimmune hematological versus non-hematological complications in PID
*Hanini S.* (Timisoara)

Coffee break

Anemias at primary immunodeficiency disorders
*Kalinina N.* (St. Petersburg)

B-cell subsets in humoral immunodeficiencies
*Bukowska-Strakova K.* (Krakow)

Double negative T lymphocytes - diagnosis and clinical significance
*Pituch-Noworolska A.* (Krakow)

The first registered case of Wiskott-Aldrich syndrome in Latvia
*Prokoffjeva T.* (Riga)

Specificity of infectious complications of pre- and posttransplant course in primary immunodeficiencies treated with allogeneic bone marrow transplantation
*Krasowska-Kwiecień A.* (Krakow)

Lunch

Session II

Clinical management of patients with primary CXCR4 deficiency (WHIM syndrome)
*Erdős M.* (Debrecen)

AK2 deficiency- reticular dysgenesis
*Schwartz K.* (Ülm)

Nijmegen breakage syndrome in Romania
*John J.* (Timisoara)

Autosomal dominant hyper-IgE syndrome – a multidisciplinary disease
*Heropolitańska-Pliszka E.* (Warszawa)

Inflammatory bowel disease in children with agammaglobulinemia
*Pac M.* (Warszawa)

IL-12/IFNγ axis disturbance complicated by recurrent M. kansasii infection, interstitial lung disease and B-cell lymphoma in an adult patient
*Bazan-Socha S.* (Krakow)

Preparations of intravenous immunoglobulins diminish the number and proinflammatory response of CD14+CD16+ monocytes in common variable immunodeficiency patients
*Siedlar M.* (Krakow)

Intravascular large B cell lymphoma mimicking CNS vasculitis in rheumatoid arthritis female patient
*Korkosz M.* (Krakow)

Clinical and therapeutic problems of patients suffering from CVID
*Lewandowicz-Uszyńska A.* (Wrocław)
Host: Sirje Velbri (velbrisir@hotmail.com)

Venue: Estonian National Library

Main Topic: Primary immunodeficiency: from physician education to genetic databases
PROGRAM

Registration and coffee

The Project of Primary Immunodeficiency
Maródi L. (Debrecen)

Primary immunodeficiency in Estonia
Sirje Velbri (Tallinn)

Clinical presentation and molecular genetic forms of hyper-IgE syndrome
Maródi L. (Debrecen)

Common variable immunodeficiency in children
Ress K. (Tartu)
Putnik U. (Tallinn)

Lunch

XLA associated with Mohr-Traneberg syndrome
Ilisson J., Rägo T. (Tartu)
Einberg Ü. (Tallinn)

Common variable immunodeficiency in adults
Varik M. (Tallinn)

DiGeorge syndrome
Ordania R. (Tallinn)

Chronic mucocutaneous candidiasis
Kisand K., Näriska M. (Tartu)

Coffee break

Hereditary angioedema - C1 inhibitor deficiency
Vasar M., Savisaar M. (Tartu)

Netterton syndrome
Kukk T. (Tartu)

H. simplex encephalitis
Ilmoja M. (Tallinn)
Kisand K., Ress K. (Tartu)
SUMMARY AND CONCLUSIONS

The J Project Meeting in Tallinn, Estonia was held on 7-8 October 2009. The meeting was organized by different Estonian Societies (Pediatric Association, Society of Allergology and Immunology and Infectious Diseases), and by the J Project – Prof. L. Maródi. The aim of this meeting was to improve the diagnostics and treatment of patients with primary immunodeficiencies (PIDs) in Estonia.

At first Prof L. Maródi introduced J Project, its aim, achievements and importance for early recognition and treatment of PIDs. S. Velbri gave a summary about PIDs in Estonia: she talked about the possibilities of diagnostics and therapy. Other reports focused on different forms of PIDs. Prof L. Maródi gave a detailed review about hyper-IgE-syndrome. The doctors from Tallinn and Tartu presented CVID in children (K. Ress, U. Putnik) and adults (M. Varik), DiGeorge syndrome (R. Zordania), chronic mucocutaneous candidiasis (K. Kisand, M. Närkska) and hereditary angioedema (M. Vasar, M. Savisaar). Rare cases of PIDs were also discussed: XLA associated with Mohr-Traneberg syndrome (Ü. Einberg), Netherton syndrome (T. Kukk) and H simplex encephalitis (M. Ilmoja).

It was concluded that it is necessary to improve awareness about PIDs and to have more cooperation with the centers of other countries for detailed diagnostics (molecular genetic diagnostics etc.) and therapy of complicated PID cases.

Sirje VELBRI

Sirje Velbri (Tallinn) and Tatjana Prokfjeva (Riga) at the St. Petersburg J Project meeting in 2009
Host:  
Nataša Toplak (natasa.toplak@ardi.si)

Venue:  
University Children’s Hospital Ljubljana, Main Hall

Main Topic:  
Periodic fever syndromes and other autoinflammatory diseases
PROGRAM

9 October

Introduction
Kržišnik C. (Ljubljana)

General overview, basic mechanisms and differential diagnosis of Autoinflammatory syndromes

Periodic fevers assembly in middle/eastern/south Europe
Toplak N. (Slovenia)

PRES working parties
Martini A. (Italy)

Autoinflammatory Diseases’ Working Party PRES, Eurofever project and registry
Gattorno M. (Italy)

Periodic and recurrent fever as differential diagnostic challenge
Toplak N. (Slovenia)

Basic mechanisms of autoinflammatory syndromes
Gattorno M. (Italy)

Systemic autoimmune diseases as a differential diagnosis of periodic fever syndromes
Avčin T. (Slovenia)

Periodic fever- approach of a paediatric infectious specialist
Pokorn M. (Slovenia)

Periodic fever- do we have to worry about malignant diseases?
Kitanovski L. (Slovenia)

Coffee break

Periodic fever syndromes

Familial Mediterranean Fever
Demirkaya E. (Turkey)

Hyper IgD syndrome
Zulian F. (Italy)

TRAPS
Gattorno M. (Italy)

PFAPA syndrome
Dolezalova P. (Czech Republic)

Cryopyrin- associated periodic fevers
Lepore L. (Italy)

Coffee break

Studies and case presentations

Mutations in MEFV gene in patients with Henoch-Schönlein purpura—study presentation
Kolnik M., Debeljak M., Toplak N., Avčin T. (Slovenia)

A Turkish boy with a long and awful history
Aktay N. (Turkey)

Periodic fever- familiar Mediterranean fever, PFAPA or other syndrome?
Bilić I., Rožmanić V., Babić-Božović I., Banac S., Čače N. (Croatia)

Recurrent fever with nephritis and anemia: a challenging diagnosis
Taddio A., Lepore L., Tommasini A. (Italy)

Periodic fever syndrome in a 58 years old man- first TRAPS case in Slovenia
Sinožić D., Toplak N. (Slovenia)
Keynote lecture
Historical overview and molecular genetic diagnostics of Periodic fever syndromes
Aksentijevich I. (USA)

Welcome reception

10 October

The expanding spectrum of Auto-inflammatory diseases I
Early sarcoidosis- Blau syndrome with case presentation
Harjacek M. (Croatia)
Systemic JIA and recurrent pericarditis
Martini A. (Italy)
CRMO
Girschick H. (Germany)
Behçet syndrome
Kone-Paut I. (France)

Short coffee break

The expanding spectrum of Auto-inflammatory diseases II
Schnitzler syndrome
Sediva A. (Czech Republic)
Inflammatory bowel disease
Orel R. (Slovenia)

Case presentations
PFAPA syndrome- the first case report from Lithuania
Panaviene V., Rusoniene S. (Lithuania)
Behcet disease- case report
Grujovska S., Kuzmanovska D., Todorovski G. (Macedonia)
Chronic recurrent multifocal osteomyelitis-case report
Makay B. (Turkey)

Coffee break

Future perspective and conclusions
How to optimize expensive genetic testing of autoinflammatory diseases?
Gattorno M. (Italy)
Problems with establishing the diagnosis of periodic fever in Slovakia and similar middle and eastern European countries
Vesely R. (Slovakia)
Novel treatments in autoinflammatory diseases
Aksentijevich I. (USA)
Clinical practice guidelines / suggestions for the approach to the patients with suspected periodic fever syndrome (round table discussion)
Moderator: Avčin T., Discussants: Toplak N., Doležalova P., Aksentijevich I., Gattorno M., Martini A.
SUMMARY AND CONCLUSIONS

Periodic fever syndromes belong to a larger family of autoinflammatory diseases. These syndromes are rare and this group of diseases is relatively new. The development, especially in genetic diagnostic, is rapid. New diseases from this spectrum are being discovered, such as deficiency of the interleukin-1-(IL-1)receptor antagonist (DIRA). In the last 10 years the genetic background of periodic fever syndromes has been discovered and the number of diagnosed patients is increasing but probably there are still many patients unrecognized especially in countries there these diseases are rare or only thought to be rare. To improve the recognition, spreading the knowledge is essential. With the purpose to improve the knowledge about Periodic fever syndromes and other autoinflammatory diseases in the region of Eastern and Central European (ECE) countries educational meeting in Ljubljana, Slovenia, was organized in October 2009. The idea for the meeting arose as a part of Eurofever project in the Autoinflammatory Diseases’ Working Party of Pediatric Rheumatology European Society (PRES). Experts in the field of autoinflammatory diseases were invited as speakers. 121 participants from 22 countries were registrated for the meeting.

The lectures were divided in 5 parts: General overview, basic mechanisms and differential diagnosis of autoinflammatory syndromes, periodic fever syndromes, studies and case presentations, the expanding spectrum of autoinflammatory diseases and future perspective on periodic fever syndromes and other autoinflammatory diseases in the region of ECE countries. The keynote lecture on historical overview and molecular genetic diagnostics of Periodic fever syndromes was given by Aksentijevich I. from National Institute of health (NIH), USA.

The opportunity for presenting interesting cases was given to participants from the region of ECE countries. Seven interesting cases of periodic fevers and autoinflammatory diseases were reported from Croatia, Italy, Lithuania, Macedonia, Slovenia and Turkey. A study of mutations in MEFV gene in patients with Henoch-Schonlein purpura in Slovenia was presented by our group.

On the second day of the meeting extended spectrum of autoinflammatory diseases was presented. In the last part of the meeting future perspective on this interesting group of diseases was discussed. Novel therapies were presented by Aksentijevich I. How to optimize expensive genetic testing for periodic fever syndromes and other autoinflammatory diseases was presented by Gattorno M. Clinical practice guidelines and suggestions for the approach to the patients with suspected periodic fever syndrome was discussed in round table discussion moderated by Gattorno M. and Avčin T. Simplified algorithm for approach to a patient with suspected periodic fever was presented by Toplak N., Ljubljana. During the meeting we discussed the possibility of genetic testing for hereditary periodic fever syndromes. Free genetic testing in specialized centers was offered to countries in which genetic testing can't be done.

The first educational meeting on periodic fever syndromes and other autoinflammatory diseases in the region of ECE countries was organized in Slovenia without a fee so that doctors from the countries of the region would be able to join the meeting. Although we were surprised by the number of registrated participants, mainly coming from Slovenia and neighbor countries, we hopped that more colleagues from eastern countries would join us. Perhaps this will come true in the future. We hope that this meeting was only the first one in line of the meetings that will follow in other countries of the region although we are aware of the fact that meetings like this, especially
without the fee, are very difficult to organize.

We would like to thank all sponsors for their significant contribution. We would also like to thank to all the invited speakers for excellent lectures and to the persons in organizing committee who showed great enthusiasm in organizing this meeting.

Natasa TOPLAK

Maruša Debeljak (Ljubljana), Ivona Aksentijevich (USA), Anna Sediva (Prague), Marco Gattorno (Genova), Pavla Dolezalova (Prague), Alberto Martini (Genova), Nataša Toplak (Ljubljana), Isabelle Kone-Paut (Paris), and Erkan Demirkaya (Ankara)

Participants
Tehran, Iran
11-12 October, 2009

Host:
Asghar Aghamohammadi (aghamohammadi@sina.tums.ac.ir)
Nima Rezaei (rezaei_nima@yahoo.com)

Venue:
Children’s Medical Center, Tehran University of Medical Sciences

Main Topic:
New advances in primary immunodeficiencies
**PROGRAM**

**11 October**

**Session 1: Primary Immunodeficiency Diseases; General Aspects**

General Aspects of primary immunodeficiencies
*Geha R. (USA)*

Primary immunodeficiency diseases in adults
*Mansouri D. (Iran)*

Break & Poster presentation

**Session 2: Allergy**

Pediatric Asthma
*Moin M. (Iran)*

Allergic disorders in infant and adults
*Movahedi M., (Iran)*

Management of Allergic disorders
*Gharagozlou M. (Iran)*

Lunch & Poster presentation

**Session 3: Predominantly Antibody Deficiencies**

Primary antibody deficiencies
*Lennart Hammarström, Sweden*

Principles and practice of vaccination in primary antibody deficiency
*Read R. (UK)*

Common variable immunodeficiency
*Aghamohammadi A. (Iran)*

**Key presentations**

Approach to the children with recurrent infections
*Aghamohammadi A. (Iran)*

Approach to the children with chronic cough
*Moin M. (Iran)*

**12 October**

**Session 1: Combined Immunodeficiencies**

Inherited defects in class switch recombination
*Durandy A. (France)*

Cellular and molecular mechanisms of the Wiskott Aldrich syndrome
*Geha R. (USA)*

Severe combined immunodeficiency
*Parvaneh N. (Iran)*

Break & Poster presentation
Session 2: Defects of Innate Immunity

Congenital neutropenia
_Klein C. (Germany)_

Mendelain susceptibility to mycobacterial infection
_Picard C. (France)_

Neutropenia and primary immunodeficiency diseases
_Rezaei N. (Iran)_

Lunch & Poster presentation

**Key presentations**

Novel monogenic disorders of the immune system - from genes to novel therapies
_Klein C. (Germany)_

Antibiotic therapy in respiratory and urinary infections
_Karimi A. (Iran)_

Clinical manifestations and complications of primary immunodeficiencies
_Al-Herz W. (Iran)_

Primary immunodeficiency diseases associated with neutropenia
_Rezaei N. (Iran)_

Immunological aspects of patients with disseminated BCG in North West Iran
_Sadeghi Shabestari M. (Iran)_

Odd presentation of chronic granulomatous disease
_Sherkat R. (Iran)_

Some moments of the meeting
SUMMARY AND CONCLUSIONS

The 36th J Project meeting on primary immunodeficiency diseases (PIDs) was held on 11-12 October 2009 in Tehran, Iran.

Each year an International Congress on Pediatrics takes place in Tehran and several PID experts have attended these scientific congresses in Iran during the last decade. In 2005, the 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 Congress of Pediatrics, a joint meeting on Immunodeficiency Diseases was established. In order to have mutual scientific exchange with other researcher of the world, the scientific committee arranged a 2-days meeting, which is organized by Department of Pediatrics, Children's Medical Center (Pediatrics Center of Excellence in Iran), Tehran University of Medical Sciences. The Jeffrey Modell Foundation (JMF) also supported the meeting.

The meeting focused on the linkages of fundamental sciences and patient-oriented research under the main theme of immunodeficiencies, whilst improving the awareness of physicians, pediatricians and other specialists on PID was the main objective of the meeting, which was attended by more than 300 researchers and clinicians. The major part of the audience includes general practitioners, pediatricians, and pediatric subspecialists; however, medical students, pediatric residents and fellows in the field of clinical immunology and infectious diseases were also actively participated in the meeting.

Some important topics that presented during this meeting were as follow:
- Approach to the children with recurrent infections
- Approach to the children with chronic cough
- Approach to the children with fever
- Approach to a patient with neutropenia
- Antibiotic therapy in respiratory and urinary infections
- Clinical manifestations and complications of primary immunodeficiencies
- Primary immunodeficiency diseases associated with neutropenia
- Immunological aspects of patients with disseminated BCG in North West Iran
- Odd presentation of chronic granulomatos disease

A poster session was also organized in which the posters were presented in 5 minutes. It should be mentioned that at first night of the meeting, a ceremony was organized by the Iranian Pediatric Hematology and Oncology Group, where Prof Christoph Klein presented an interesting lecture "Novel monogenic disorders of the immune system - from genes to novel therapies".

Finally, we believe that the J Project is essential and useful, and we would like to thank Prof. László Maródi for his initiative in this regard. We consider the event in Tehran as a successful J Project event, and we plan to repeat it in mid October, 2010 after the ESID meeting in Istanbul. Therefore we invite all the ESID members to take part in our next J Meeting, as it would be easy to travel to Tehran from Istanbul; to have a memorable time in both Turkey and Iran.

We are looking forward to continuing the successful cooperation and seeing you in Tehran.

Asghar AGHAMOHAMMADI &
Nima REZAIEI
Cairo, Egypt
18-19 October, 2009

Host:
Nermeen Galal (nermeengalal@gmail.com)
Aisha El Marshafy (aisha_mars@hotmail.com)

Venue:
Cairo University Children Hospital (Abu El-Reech, Japanese Hospital)

Main Topic:
Primary Immunodeficiency disorders in children
Cairo University Children Hospital

PROGRAM

One Day Symposium on Primary Immunodeficiency

19 October

Introductory remarks
Elmarsafy A. (Cairo)

How does PID relate to your practice?
Azmy J. (Cairo)

T cell defects
Reda S. (Cairo)

Predominantly Antibody deficiency
Galal N. (Cairo)

ESID, PID CDWP, J project
Maródi L. (Debrecen)

PID Registry
Gathmann B. (Germany)

Case presentations
Ghoneimy D. (Cairo)

Coffee Break

Laboratory component
Nabil H., Afifi H. (Cairo)

Infections in PID
El-Kholy A. (Cairo)

Genetic diagnosis of PID
El-Sawy M. (Cairo)

BMT
Haddad A. (Cairo)

IVIG, Shortcoming Future
Moufah F. (Blood Bank)

Discussion

Aisha El Marsafy (second from right), guest speakers, and participants
SUMMARY AND CONCLUSIONS

Minutes of the 37th J project meeting – European Society of Immunodeficiency Disorders (ESID)

Attendees: 101
- Teams of Immunodeficiency units at Cairo, Ain Shams and Mansoura Universities
- Cairo and Ain Shams University Professors with the following specialities:
  - Allergy, Pulmonology, Hematology, Oncology, Gastroenterology, Hepatology, Endocrinology, Intensive care, Microbiology and Clinical Pathology Departments
- Graduate study candidates (M.Sc. Degree), Nursing diploma, visitor residents on training programs from Libya and Palestine
- Representatives of several Ministry of Health operated hospitals: Ahmed Maher, Helwan, Mounira, Om ElMasreyeen
- Military Medical hospitals, Ministry of interior affairs Hospital, National Research center, Abu Reesh insurance hospital for school children
- National Blood transfusion Center
- VACSERA (the holding company for vaccines and biological products)
- Pharmaceutical representatives

The first session was chaired by:
- Professor Yehia Elgamal, head of ESPAI (Egyptian Society of Pediatric Allergy and Immunology), member of World Allergy Organization
- Professor László Maródi, head of PID in development WP, ESID
- Professor Aisha Marsafy, Head of Immunodeficiency practice at Cairo University Children’s Hospital.

Prof. Marsafy gave some introductory remarks stating why the global attention is shifting to PID and how the disorders are still hugely under reported in Egypt as well as in Africa. A case of hyper-IgM syndrome was presented as an example of cases whose care gets blocked because of lack of investigations. The second talk was given by Azmy J. consultant pediatrician, Cairo University who focused on the warning signs of PIDs, and how PIDs relate to clinical practice of different specialists. Reda S., professor of pediatrics, Ain Shams University gave a talk about combined immunodeficiency disorders and how SCID is a forgotten pediatric emergency and advocated national screening for SCID plus a stem cell bank as well as increased awareness among GP. Galal N, associate professor of pediatrics, Cairo University later gave a talk about need to know information for pediatricians about antibody deficiency stressing on the interpretation of results and the common mistakes.

Prof. Maródi L. talked about the evolving practice of PID and about new concepts of disorders like HUS, Crohn’s disease and PAP. He went through the accomplishments of the J Project and the goal of having PID patients benefiting from the most advanced molecular therapy all over the world. Gathmann B. presented the entries submitted by the two documenting centers in the registry totaling to 150 patients with massive under reporting. He also highlighted the potential routes for collaboration between centers. Elghoneimy D., lecturer of pediatrics, Ain Shams University presented a case about chronic granulomatous disease. Hassan R., assistant lecturer of pediatrics, Ain Shams University demonstrated a case of hyper-IgE syndrome.

The second session was started by Mouftah F., general director of blood transfusion services, Ministry of Health, Egypt who elaborated on the available services, methods of fractionation, near future perspectives for regional preparation of IVIG (end of 2010 -2011). Nabil H., associate professor of Clinical Pathology and Immunology, Cairo University gave a presentation about flow cytometry and how to bridge the gap between clinicians and laboratory partners. Afifi H., professor of
Clinical Pathology at Ain Shams University demonstrated the available services and markers for PID cases. Elkholy A., professor of Clinical Pathology at Cairo University elaborated on the microbiological aspect pertaining to PID disorders. Haddad A., professor of Pediatric Oncology, National Cancer Institute, Cairo University talked about his team’s experience with BMT for PID patients, the current obstacles, referral routes and outcome.

The meeting was concluded with the following recommendations:

• Follow up meeting to further cooperate between the existing teams in various centers.
• Trials of having each center administer or add specific diagnostic tests which will be offered to patients from other centers.
• Set up of satellite centers with people who have interests in PID to increase awareness.
• Better documentation and set up of a national registry.

CAIRO
Minsk, Belarus
22-23 October, 2009

Host:
Michail Belevtsev (belevtsev_m@mail.ru)

Venue:
Lesnoy Belarussian Center for Pediatric Oncology and Hematology

Main Topic:
Actual questions of primary immunodeficiency
PROGRAM

22 October

Opening and introduction
TBA Ministry of Health Republic of Belarus

Plenary session I

Medical care organization and perspectives for children with primary immunodeficiency in Belarus
Aleinikova O.V. (Minsk)

Diagnostics of primary immunodeficiency, common principles.
Kondratenko I.V., Bologov A.A. (Moscow)

Evolution of human immune system. Modern view.
Leliavskii A.A. (Minsk)

Human immune system. Principle of immunological diagnostics of primary immunodeficiency. Sharapova S.O., Belevtsev M.V. (Minsk)

Genetic methods of diagnostics and treatments of primary immunodeficiency. Prenatal diagnostics.
Migas A.A. (Minsk)

Operation analytical system for comprehensive diagnostics of Primary Immunodeficiency
Krasko O.V. (Minsk)

Lunch

Plenary session II

Features of agammaglobulinemia in children and adults
Yanchenko V.V., Novikov D.K. (Vitebsk)

Diagnostics and medical care of patients with primary immunodeficiency in Bashkortostan
Hamatdinova Z.R., Proligina D.D (Ufa)

Chromosomes instability syndromes
Rodina U.A., Bologov A.A. (Moscow)

Hyper-IgM syndromes. Case reports.
Prokopheva T. (Riga)

Novel immunodeficiencies relevant to internal medicine
Maródi L. (Debrecen)

Clinical diversity of phagocyte defects
Aleshkevich S.N. (Minsk)

Discussion

23 October

Plenary session I

Clinical and immunological features of diagnostics and treatments patients with ataxia-telangiectasia
Gurmchuk I.E. (Minsk)

Diagnostics of DiGeorge Syndrome in Belarus
Hurs O.M., Politiko A.D. (Minsk)

Study of genetics Familial Hemophagocytic lymphohistiocytosis and X-linked Lymphoproliferative syndrome
Poltavec N.V. (Moscow)

Coffee break

Principle of treatment of Primary Immunodeficiency.
Deripapa E.V. (Moscow)

Application of intravenous immunoglobulin in pediatrician practice
Uglova T.A. (Minsk)
Hematopoietic stem cell transplantation for treatment of primary immunodeficiency.  
Kachan G.L. (Misk)  

Diagnostic SCID (deficiency IL-7α-R). Reconstitution of patient’s immune system after HSCT.  
Sharapova S.O. (Minsk)  

Lunch  

SUMMARY AND CONCLUSIONS  

On 22-23rd October, 2009 in Minsk Conference on Primary Immunodeficiencies has been organized within the limits of J Project. More than 150 pediatricians and immunologists from all regions of Belarus participated in the conference. Results of work of the Center of Primary Immunodeficiencies on the basis of Belarussian Center for Pediatric Oncology and Hematology have been reported at the conference. The leading experts of the center: Sharapova S.O, Aleshkevich S.O., Migas A.A., Belevtsev M.V have given reports. As a result of the center’s work the primary immunodeficiency was diagnosed in 90 patients, methods of advanced immunological diagnostics are introduced and genetic investigation on 13 genes was carried out.  

Irina Kondratenko from Russia, László Maródi from Hungary, and Tatiana Prokofjeva from Latvia are also participated in the conference with their reports.  

Michail BELEVITSEV  

See You soon at the 2010 J Project meetings!
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. Kyiv, 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: Zoltan Ellenes
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: Michael Belevtsev
17. Debrecen, Hungary, December 8-9, 2006; Organizer: László Maródi
18. Zaporozhzhye, Ukraine, Apr 19-20, 2007; Organizer: Lyudmila 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: Lyudmila Chernyshova
24. Sunny Beach, Bulgaria, May 22-23, 2008; Organizer: Elissaveta Naumova
25. Sarajevo, Bosnia-Herzegovina, October 10-11, 2008; Organizer: Velma Mulaosmanovic
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. Yekaterinburg, 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: Nataša Toplak
36. Tehran, Iran, October 11-12, 2009; Organizer: Asghar Aghamohammadi and Nina Rezaei
37. Cairo, Egypt, October 18-19, 2009; Organizer: Nermeen Galal
38. Minsk, Belarus, October 22-23, 2009; Organizer: Michael Belevtsev
### J Project Meetings in 2010 № 39-46

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<td>Georgina Kuli-Lito</td>
<td>April 8-9</td>
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<td>Aisha El-Marsafy</td>
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<td>Liudmyla Chernyshova</td>
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<td>Srdjan Pasic</td>
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<td>43. Zagreb (Croatia)</td>
<td>Darko Richter</td>
<td>July 2-3</td>
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<td>44. Chelyabinsk (Russia)</td>
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<td>45. Budapest (Hungary)</td>
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<td><a href="mailto:lmarodi@dote.hu">lmarodi@dote.hu</a>; László Maródi</td>
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<td>Asghar Aghamohammadi</td>
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# THE J PROJECT

## Steering Committee Members

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Genetic testing of PID genes in 2004-2009

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Summary : 351 81 33 20 23 18 2 2 3 3 2 1 5 1 556
Genes sequenced at the Debrecen Jeffrey Modell PID Reference Center

PRIMER IMMUNODEFICIENCIES (Genes)
1. X-linked agammaglobulinemia (BTK)
2. X-linked hyper-IgM syndrome (TNSF5=CD40L)
3. Activation induced citidine deaminase deficiency (AICDA)
4. Common variable immunodeficiency (TACI)
5. Common variable immunodeficiency (ICOS)
6. Hyper-IgD syndrome (MVK)
7. Hyper-IgE syndrome (STAT3)
8. X-linked severe combined immunodeficiency (IL-2RG)
9. Recombinase activator gene 1 deficiency (RAG1)
10. Recombinase activator gene 2 deficiency (RAG2)
11. Familial hemophagocytic lymphohistiocytic syndrome (PRF1)
12. X-linked lymphoproliferative syndrome (SH2D1A)
13. Autoimmune lymphoproliferative syndrome (FAS)
14. Autoimmune lymphoproliferative syndrome (FASLG)
15. Autoimmune polyendocrinopathy I syndrome (AIRE)
16. Dyskeratosis congenita (DKC1)
17. Cartilage-hair hypoplasia (RMRP)
18. Nijmegen breakage syndrome (NBS1)
19. Shwachman-Diamond syndrome (SBDS)
20. Warts-Hypogammaglobulinemia-Infection-Myelokathexis syndrome /WHIM syndrome (CXCR4)
21. Wiskott-Aldrich syndrome (WAS)
22. Kostmann syndrome (HAX1)
23. Ciclic neutropenia (ELA2)
24. Severe congenital neutropenia (GFI1)
25. Severe congenital neutropenia (CSF3R)
26. Severe congenital neutropenia (G6PC3)
27. X-linked chronic granulomatous disease (CYBB)
28. Autosomal recessive chronic granulomatous disease (NCF1=p47phox)
29. C2 deficiency (C2)
30. IL18R deficiency (IL18RG)
31. Chronic mucocutaneous candidiasis (CLEC4L)
32. Chronic mucocutaneous candidiasis (CLEC7A)
33. Chronic mucocutaneous candidiasis (MRC1)

LYSOSOMAL STORAGE DISORDERS (Genes)
34. Fabry disease (GLA)
35. Gaucher disease (GBA)
36. Niemann-Pick disease (SMPD1)

OTHERS (Genes)
37. Von Hippel-Lindau syndrome (VHL)
38. X-linked congenital adrenal hypoplasia (DAX1)
39. Galectin 3 deficiency (LGALS3)
40. Alström syndrome (ALMS1)
AIM: To increase awareness on PID