J Countries

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Sponsor:
ECE IPI CTR
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I am delighted to present the 2010 J Project Booklet and the output of our physician education and clinical research collaboration program in the last year. Six J Project Booklets have been published in the past years to present the development of the primary immunodeficiency (PID) awareness program in Eastern and Central Europe (ECE). The previous editions listed the venues, the organizers and programs, and summarized the main focus and special features of the particular meetings. The 2010 booklet has additional and unique messages. We report in this issue on the J Project Steering Committee (SC) Meeting organized in Budapest. At this Meeting the history, the current state and the future challenges of the J Project was discussed. We considered the possibility of organizing regions in the territory of the so-called “J Countries”. Based on traditions of medical care and similarities of culture and languages, 5 or 6 regions might be organized. This kind of organization may allow rotation of leadership of the J Project. There was also agreement about organizing every year a J Project Juniors’ Meeting to promote further education of young immunologists in ECE. Selection of the venue of the Juniors’ Meeting was proposed to be made by application. Biotest GmbH representatives proposed that the Company may take care of financing of these meetings from a separate budget. They were also committed to offer mobility grants for ECE scientists and young doctors to spend short periods in the Debrecen PID Center in order to learn specific diagnostic and research assays. The year 2010 was also featured by the very first award given for to Professor Liudmyla Chernyshova, an outstanding J Project activist and fan (page 14).

Debrecen, March 15, 2011

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 2010

CITY, Country       DATE
1. Tirana (Albania)  April 8-9
2. Porto Marina (Egypt)  April 16-17
3. Kharkov (Ukraine)  May 19-20
4. Zlatibor (Serbia)  June 1-2
5. Zagreb (Croatia)   July 2-3
6. Chelyabinsk (Russia)  August 30-31
7. Budapest (Hungary)  September 9-10
8. Tehran (Iran)       October 11-12

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.
Tirana, Albania
8-9 April, 2010

Host:
Georgina Kuli-Lito (gkuli_lito@hotmail.com)

Venue:
Hotel “Tirana International”

Main Topic:
Primary Immunodeficiencies in Albania
PROGRAM

8 April

Welcome reception Restorant “Piazza”

9 April

Registration
Opening of the meeting

First Session

Presentation of project of primary immunodeficiencies (J Project)
Maródi L. (Debrecen)

Laboratory diagnosis of Primary Immune Deficiencies in Albania
Ylli Z., Prifti M., Selimaj V., Sulcebe G. (Tirana)

The frequency and characteristics of PID in Konya, Turkey
Reisli I. (Konya)

Coffee break

Second session

Case report of three boys with Agammaglobulinemi
Muci M., Veveçka E., Sulçebe G. (Tirana)

Diagnosis and follow up of first case with Bruton in Albania-France
d’Angely S. (Paris)

Case report of a child with cellular immunodeficiencies
Bizhga M., Serbo L., Sulcebe G. (Tirana)

First case with lymphoploiforative syndrome in Sarajevo
Mulaosmanović V. (Sarajevo)

Questions and discussions
Lunch

Third session

Antibody Deficiency Disorder associated with abnormal B cell function
Xhuke M., Kota L., Sulcebe G., Mati L., Sala E., Agastra M., Borici S. (Tirana)

Secondary immunodeficiency caused by medication, complicated by varicella acute myocarditis
Bejiqi R., Bejiqi H., Retkoceri R., Kelmendi M., Zeka N., Dedinca S. (Kosovo)

Chronic granulomatoz diseases. Case report.

Data on ICF syndrome.
Face M., Nastas E., Godo A., Bali D., Xhafa M.

Questions and discussions
Conclusions

10 April

Sight seeing tour of Tirana and Durresi
Lunch
The Albanian Pediatric Society, in collaboration with the J Project and with the support of USAID, held on 9th of April 2010, a national conference on primary immunodeficiency diseases (PIDs).

PIDs are inborn errors of immunity manifested at an early age and, if diagnosed late, may cause severe or fatal infections in children. The number of reported cases of PIDs in Albania is low and this conference was aimed to raise awareness of pediatricians and other professionals with regard to improve diagnoses and treatment, and to establish a national PID database. The lack of appropriate laboratory facilities is one of the obstacles in establishing the precise diagnosis especially genetic causes of these diseases. Until now there has been a national registry of PIDs in Albania.

Although the conference had a very much focused topic, it was attended by about 340 pediatricians and family doctors from primary, secondary and tertiary level of health institutions from all over the country of Albania as well as from the new Republic of Kosovo. The J Project launched in 2004 in Eastern Europe with the support of the Jeffrey Modell Foundation and Biotest Hungary seeks to diagnose patients with PIDs and raise awareness and promote continuing medical education in this field. To date close to 40 meetings have been organized in Central and Eastern Europe and recently, in Asia and Africa. This first J Project Meeting in Albania may be considered as a milestone in improving a better care of PID patients. It is our hope that we shall establish a strong collaboration between the Albanian specialist and activists of the J Project as well as the European Society for Immunodeficiencies (ESID).

We are grateful to USAID, the main supporter of several recent conferences organized by the Albanian Pediatrics Association, that provided funding to draft of the pediatrics protocol’s publication, as well as, funding of several studies conducted in the field of public health.

The conference concluded that:

1) PIDs should be considered as an important group of diseases to be diagnosed and treated in children with recurrent and severe infections as early as possible

2) Establishment of national registry for PID in Albania is needed.

3) Collaboration between the University Hospital Center of Tirana and the JMF Reference Center in Debrecen should be started to assure genetic diagnosis of PIDs as early as possible.

We are grateful to USAID, the main supporter of several recent conferences organized by the Albanian Pediatrics Association, that provided funding to draft of the pediatrics protocol’s publication, as well as, funding of several studies conducted in the field of public health.

Georgina KULI-LITO
L→R: László Maródi (Debrecen), Georgina Kuli-Lito (Tirana), Evda Vevecka (Tirana), Zhaneta Shatri (USAID), Petrit Vasili (Minister of Health in Albania), Ismail Reisli (Konya)

Participants of the Meeting
Host:  
Aisha El-Marsafy (Aisha_mars@hotmail.com)  
Mohammed El Guindi  

Venue:  
Alexandria-Porto Marina-North Coast  

Main Topic:  
Gastrointestinal Manifestations of Primary Immunodeficiencies
PROGRAM – Major Presentations

Symposium on primary immunodeficiency disease links to gastrointestinal disease in pediatrics

Diarrhoea, is it primary immunodeficiency disease?
El-Marsafy A. (Cairo)

SUMMARY AND CONCLUSIONS

The 46th J Project Meeting was organized as separate session of the Fifteenth National and International Conference of the Egyptian Society of Pediatric Gastroenterology, Hepatology and Nutrition. Accordingly gastrointestinal manifestations of various primary immunodeficiency disorders were discussed.

Major presentations were given by experts from the Cairo University and the Ain Shams University in Cairo.

Most immunodeficiency disorders predispose patients to GI tract infections in addition to respiratory tract diseases as presented by Dr. Reem Elfiki. In particular, Prof. El-Marsafy covered this area focusing to a number of diseases like IPEX, SCID and CD40L deficiency.

Liver involvement in patients with CGD and CD40 ligand deficiency was specifically focused in the presentations by Dr. Jeannette Azmy. The complexity of mucosal immunity and the importance of mucosal barrier to maintain normal immuno homeostasis was discussed by Dr. Nermeen Galal.

The PID session was attended by gastroenterologists, hepatologists and nutrition experts working in the pediatric field as well as by general pediatricians from Egypt, Jordan, Syria, Lebanon, Tunisia, Saudi Arabia, United Arab Emirates, Sudan, and United States of America. The session was well acknowledged and raised a lot of interest among participants.

Aisha EL-MARSAFY

L→R: Aisha El-Marsafy, Jeannette Azmy, Nermeen Galal (Cairo University)
Kharkov, Ukraine
19-20 May, 2010

Host:
Liudmyla Chernyshova (chernyshova@ukr.net)

Venue:
Kharkov State University named after Karazin

Main Topic:
Role of J Project Meetings in improvement diagnostics of PID in children in Ukraine

The Statue of Taras Shevchenko
PROGRAM

Opening ceremony

Primary immunodeficiencies: Worldwide health problem.
*Marodi L.* (Debrecen)

Medical management of children with primary immunodeficiency. Integration of pediatric immunology in European medical field.
*Chernyshova L.* (Kiev)

Perspective of using of stem cells in correction of immunodeficiencies
*Goltsev A.M.* (Kharkov)

Should we always trust in molecular genetic diagnosis? Case of identification of two different nosologic forms of SCID in one family.
*Samarin D.* (Kiev)

PID with chromosome instability
*Kostyuchenko L.* (L’viv)

IgG subclasses deficiency
*Volokha A.* (Kiev)

Compliance for treatment in PID
*Bondarenko A.* (Kiev)

Modern possibilities of diagnosis and treatment of SCID (family case)
*Savchak I.* (L’viv)

Coffee break.

Main directions of activities of Kharkov Clinical Immunology Center
*Savvo O., Tymokhina N.I.* (Kharkov)

Lui-Barr syndrome: diagnostics and management
*Lapiy F.* (Kiev)

Session of PID patients’ parents section

Master-class “Diagnosis and treatment of PID in children”

Symposium of regions chief pediatric immunologists

Closing

L→R: Anastasia Bondarenko, László Maródi, Liudmyla Chernyshova, Olesya Rabosh, Yuriy Stepanovskyi, Svetlana Yakimovich, Dimitryi Samaran, Alla Volokha
SUMMARY AND CONCLUSIONS

According to the East-European Initiative “J Project” the 6th Ukrainian Meeting has taken place in Kharkov (Ukraine) at May 20, 2010. The meeting was organized by chief pediatric immunologists of MOH of Ukraine Chernyshova L. and chief immunologist of Kharkov region Savvo O. Kharkov is the 2nd largest city in Ukraine after Kiev with 1.5 million population. In the 1920-ies Kharkov was the capital of Ukraine. The immunologic center of the city was organized many years ago, which now serves as a center for 3 regions of Ukraine (Kharkov, Sumy, Lugansk). Although the number of diagnosed PID cases is increasing, making the definite diagnosis is still a problem, as well as the use of up-to-date treatment. The main problem is that doctors of first line contact have limited knowledge about PIDs and the laboratory background is insufficient. For these reasons educational programs are necessary to improve the recognition of PIDs. Achievements in diagnostics of PIDs in Ukraine and elsewhere, demonstration of clinical cases of PIDs (antibody deficiencies, Nijmegen breakage syndrome, Wiskott-Aldrich syndrome, ataxia telangiectasia), the possibilities of genetic diagnostics and treatment of PIDs were the basic directions of the presented reports. Prof. L. Maródi noted the great contribution of Prof. L. Chernyshova in the development of clinical immunology in Ukraine and international cooperation, and also handed to her the award “Pro Collaboratione” conferred in 2010 by Debrecen Regional Committee of the Hungarian Academy of Arts and Sciences.

Besides, in his presentation L. Maródi acquainted the participants with the latest advances in genetic diagnostics of PID and the discovery of new diseases and mutations. The goal of the seminar was to increase awareness of PIDs and discuss of treating experiences in the Kharkov region. The leading immunologists from different regions of Ukraine and about 120 pediatricians from Kharkov region have taken part in the meeting. On patient’s session patients and their parents talked about their history of disease, path to diagnosis and treatment problems. Their speeches were memorable and impressive. At the end of the seminar was held a workshop on the diagnosis of the most common PIDs. In her report L. Chernyshova stressed that J Project Meetings gave much help in improvement of recognition of PIDs in Ukraine which is now supported by statistical data from the Jeffrey Modell Foundation presented also by L. Maródi. The numbers of PID patients in Ukraine are now approaching 2000 which includes transitory hypogammaglobulinemia and selective IgA deficiency as well.
Zlatibor, Serbia
1-2 June, 2010

Host:
Srdjan Pasic (Pasics@ikomline.net)

Venue:
Zlatibor Congress Center

Main Topic:
Primmary immunodeficiency diseases

Landscape of Zlatibor
PROGRAM

Progress in PID
Maródi L. (Hungary)

Chronic mucocutaneous candidiasis: an update
Lilić D. (GB)

A single center experience in HSCT for PID
Seger R. (Switzerland)

Break

HSCT in autoimmune diseases
Abinun M. (GB)

Nijmegen Breakage syndrome
Erdős M. (Hungary)

Nijmegen breakage syndrome: case report
Pasi S. (Serbia)

Hereditary angioedema in Serbia
Andrejevic S. (Serbia)

Recurrent fever with pericarditis: case report
Ristic G. (Serbia)

PID care in Ukraine
Volokha A. (Ukraine)
SUMMARY AND CONCLUSIONS

This one-day J Project Meeting was held at Zlatibor, Serbia on June 1st, 2010. This meeting was attended by 70 pediatricians who were participants of traditional Pediatric Serbian Summer School.

Professor Laszlo Marodi gave an introductory lecture about J project achievements and goals. Dr Desa Lilic, University of Newcastle, United Kingdom introduced the participants about new knowledge about pathogenesis of chronic mucocutaneous candidiasis and APECED. Dr Mario Abinun presented the data regarding outcome of autologous hematopoetic stem cell transplantation in juvenile arthritis.

Dr Srdjan Pasic, Mother and Child Health Institute, Belgrade gave a lecture about autoimmune features in Nijmegen breakage syndrome. Dr Sladjana Andrejevic, University Clinical Center, Belgrade presented new treatment options for hereditary angioedema. Finally, Ms Dragana Koruga, the chair of Serbian patient’s organization presented current problems affecting the families of children with PID and possible solutions.

Srdjan PASIC

“Quo vadis immunodeficiency”
Zagreb, Croatia
18-19 June, 2009

Host:
Darko Richter (darkorichter@hotmail.com)

Venue:
Croatian Medical Association, Main Hall

Main Topic:
Primary immunodeficiency in Croatia
PROGRAM

1 July
Get together dinner

2 July
Welcome & opening remarks
Richter D., Maródi L.

Overview of primary immunodeficiencies
Overview of primary immunodeficiencies in Croatia
Richter D. (Croatia)

PIDs: Current concepts and future challenges
Maródi L. (Hungary)

Immunodeficiency in the adult
Artuković M. (Croatia)

Coffee break

Familial hemophagocytic lymphohistiocytosis
Bilić E. (Croatia)

Molecular genetics and clinical presentation of dyskeratosis congenita
Erdős M. (Hungary)

Immunodeficiency syndromes with autoimmunity
Jelušić-D. M. (Croatia)

Congenital neutropenias
Kelečić J. (Croatia)

Lunch break

Vaccination of immunodeficient children
Pneumococcal conjugate vaccine in immunodeficient children
Tešović G. (Croatia)

Vaccination of children with primary immunodeficiency
Harjaček M. (Croatia)

Coffee break

Case presentations
(CGD, AT, Hermansky-Pudlak, HIDS, Blau, X-SCID, neutropenia & hypogammaglobulinemia)

Concluding remarks and farewell
Richter D., Maródi L.

Mihaela Bataneant presenting her case
SUMMARY AND CONCLUSIONS

The 43rd J Project meeting was simultaneously the 1st Croatian meeting on primary immunodeficiency. It was held in Zagreb on Thursday 1 July and Friday 2 July under the title: „Primary immunodeficiency in Croatia“. After the get together dinner on Thursday attended by all the lecturers and invited guests, including Prof. Ivan Malčić, the Head of the Dept. of Pediatrics, University Hospital Center Zagreb, the main body of the meeting was held on Friday. There were 56 registered participants from Croatia, mainly pediatricians and pediatric interns, and to lesser extent adult medicine specialists. Three distinguished guests from the J Project countries have taken part: Prof. Laszlo Maródi, Dr. Melinda Erdős and Dr. Mihaela Bataneant. Pediatric nurses and families of PID patients have also taken part in the Meeting.

The choice of lecturers and topics has aimed at including the majority of professionals who, in their daily practice, are expected to come across primary immunodeficiency patients: pediatric hematologists, pediatric rheumatologists, pediatric respiratory specialists, internal medicine specialists, and, last, but not least, pediatric allergists and clinical immunologists. They should have their awareness raised as to recognize primary immunodeficiency patients and refer them to the center capable of offering comprehensive diagnostic and therapeutic care. This is located in the Dept. of Pediatrics of the University Hospital Center Zagreb, and is coordinated by Dr. Darko Richter.

The morning was devoted to somewhat lengthy overview lectures, but the afternoon was a lively show of various case reports that has spurred up a lot of discussion. A parallel session of the IPOPI took place during the morning hours in the side hall gathering several Croatian PID families. During the lunch break 19 pediatric allergists and clinical immunologists from Croatia founded the Section on Allergy and Clinical Immunology of the Croatian Pediatric Society. This is now a formal body that will continue to promote the knowledge and implementation of comprehensive diagnostic and therapeutic care for primary immunodeficiency in Croatia. The Croatian Medical Chamber has awarded active participants with 10 CME credits, and the passive participants with 8 CME credits.

The Meeting was organized under the auspices of the Croatian Pediatric Society – Regional Organization of the City of Zagreb. From now on, the organizer will be the Section on Allergy and Clinical Immunology of the Croatian Pediatric Society. It is felt that the local J Project meetings will return at a frequency of once in 12-18 months.

Darko RICHTER
Doc. Dr. Drago Buneta (middle), the pioneer of PID field in Croatia with several of his former students: Jadranka Kelečić (Zagreb), left; Marija Radonić (Dubrovnik), right; in the back (L→R) Tamara Voskresensky-Baričić and Darko Richter, both from Zagreb. Dr. Buneta described a Wiskott-Aldrich syndrome patient, the first PID case in Croatia in 1972.
Host:  
Irina Tuzankina (I.Tuzankina@iip.uran.ru)

Venue:  
Chelyabinsk State Medical Academy

Main Topic:  
Diagnostics of primary immunodeficiencies in the Urals region
PROGRAM

Inflammation and immunity
Chereshnev V. (Russia)

Importance of PID in clinical medicine
Maródi L. (Hungary)

Regional register of PID in Sverdlovsk Region
Tuzankina I. (Russia)

Immune response development of fetus and newborns
Prodeus A. (Russia)

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

Laboratory diagnostics of PID
Pashnina I. (Russia)

Experience of PID patients in Chelyabinsk region
Pischalnikov A. (Russia)

Apoptosis. Method of flow cytometry
Pashnina I. (Russia)

Case reports
Moiseeva T. (Russia)

Valery Chereshnev
Director of Institute of Immunology and Physiology,
Urals Branch, Russian

Chelyabinsk State Medical Academy
SUMMARY AND CONCLUSIONS

In August 2010 a meeting of the J Project for PID was held in Chelyabinsk. The meeting was organized as the part of “International School of flow cytometry in clinical and laboratory practice”. It was held on 9th day of School (31 August).

Audience was more than 200 people. They were pediatricians and immunologists from different cities of Russia, Uzbekistan and Kazakhstan (172 pediatricians from Chelyabinsk region and 98 doctors from other cities and countries).

Participation in conference was equal to supplementary post-graduate education of “Clinical laboratory diagnostics” and “Allergology and Immunology”.

The meeting was very helpful for practical diagnostics and management of patients with PID in Chelyabinsk region. It was the first pediatric forum where a lot of specialists of our city exchanged experience in management of patients with PID.

The experiences of this meeting will undoubtedly improve the situation of diagnostics and management of patients with primary immunodeficiency in our region. The scientific and social program of the meeting was rated as highly successful and we would like to thank all the participants for their contributions.

Irina TUZANKINA

"Sphere of Love"
Host:
László Maródi (lmarodi@dote.hu)
Miklós Szolnoky (Biotest@biotest.hu)

Venue:
Hotel Ramada, Budapest

Main Topic:
J Project Steering Committee Meeting
PROGRAM

Friday 9th September; 19:00

Welcome reception and dinner at Restaurant Kéhli

Friday 10th September; 9:00 – 15:30

Opening and address
Maródi L. (Hungary)

Maródi L. (Hungary)

Reports on national J Project activities
All Delegates

Lunch

Developing strategic plans
All Delegates

Regional J Project sections
All Delegates

Steering Committee members from regions
All Delegates

New activities (Juniors’ Meeting, mobility grants, registry, society, journal)
All Delegates

Any other business

19:00 Dinner at Restaurant Gundel
L→R: Igor Shavchak (Ukraine), Gasper Markelj (Slovenia), Darko Richter (Croatia)

L→R: Michaela Bataneant (Romania), Ioan Gherghina (Romania), Melinda Erdős (Hungary)

L→R: Jiri Litzman (Czech Republic), Aisha El-Marsafy Egypt), Irina Vitvitskaya (Russia), Ismail Reisli (Turkey), Margit Serban (Romania), László Maródi (Hungary), Irina Tuzankina (Russia)
L→R: Georgina Kuli-Lito (Albania), Sirje Velbri (Estonia), Anna Scherbina (Russia)

L→R: Jan Sun (Germany), Michael Becker (Germany)

Anastasia Bondarenko (Ukraine)  
Malgorzata Pac (Poland)
Marina Guseva (Russia)

Tatjana Prokofjeva (Latvia)

Mikhail Belevtsev (Belarus)

Irina Kondratenko (Russia)

L→R: Kristina Mironska (Rep. of Macedonia), Jiri Litzman (Czech Republic), Aisha El-Marsafy (Egypt), Ismail Reisli (Turkey)
SUMMARY AND CONCLUSIONS

The first J Project Steering Committee (SC) Meeting which was held on September 10-11, 2010 in Budapest. The SC is the group of colleagues who have organized J Project meetings over the past 6 years. The Meeting was organized together with the Jeffrey Modell Primary Immunodeficiency Center in Debrecen and Biotest Hungary, and was hosted by Biotest GmbH, Dreieich, Germany. The meeting was attended by leaders of PID centers from:

- Albania (G. Kuli-Lito),
- Belarus (M. Belevtsev),
- Croatia (D. Richter),
- Czech Republic (J. Litzman),
- Egypt (A. El-Marsafy),
- Estonia (S. Velbri),
- Germany (K. Schaefer, M. Becker, J. Sun),
- Hungary (M. Erdős, L. Maródi, M Szolnoky, Z. Becze),
- Latvia (T. Prokofjeva),
- Poland (M. Pac, A. Szafalarska),
- Republic of Macedonia (K. Mironksa),
- Romania (M. Serban, I. Gherghina, M. Bataneant),
- Russia (I. Kondratenko, I. Tuzankina, M. Guseva, A. Shcherbina, I. Vitvitskaya),
- Slovenia (G. Markelj),
- Turkey (I. Reisli), and
- Ukraine (A. Bondarenko, I. Savchak).

The major purpose of the meeting was to summarize professional and scientific achievements over the past six years and to outline future directions, concepts and plans. Center leaders provided an overview of the past six years’ achievements and results in their countries. It was pointed out that “The J Project is for everyone and the emphasis is on the activity and communication of the centers.”

The participants all believed that the J Project should go on in future. In particular, the Project should do more for young immunologist in ECE, particularly by empowering them to become active members in PID patient care and research. It is for this reason that the SC has committed to educating and integrating young doctors into the future work of the J Project in our mainstream work and promoting their involvement in the J Project program in general and in the activities of J Project Countries and Centers in particular. The SC decided to launch the J Project Juniors’ Meeting series with one meeting each year. The purposes of this meeting are: 1) to promote stronger participations of young doctors within the J Project Program; 2) to strengthen the capacity of PID centers to work with and get involve young doctors in patient care; 3) the brainstorming with the youth representatives on future strategies of the J Project. Juniors’ Meeting participants should have the opportunity to actively participate in the program, to outline the agenda, and to build partnerships with each other’s institutions. They should understand how important PID working groups, PID registries and patient groups are to establish cooperation between PID centers in ECE.

We also discussed organizing working groups like Program Committee (PC), Diagnostics Working Group (DWG), Therapeutics WG (TWG), Registry WG (RWG), Juniors WG (JWG) and PR Committee (PRC). The discussion went as far as to suggest the possible establishment...
of J Project Society (JPS) and starting with a new PID-focused journal. The general consensus was that the discussion on all the above listed issues should be continued and final decisions should be made on the basis of general support by the participating countries. It was also suggested that the size of the SC should be decreased and one way to achieve it may be to organize J Project regions representing themselves in the SC by their leaders elected for a defined period of time.

The SC represented a new milestone in the history of the J Project having been a platform program the role of which continues to be promotion of a better diagnoses and treatment of PID patients in ECE. We are grateful to Biotest GmbH and Biotest Hungary for having made this event possible through a special grant. I believe the meeting has been very successful enhancing communication between J Project Centers.

László MARÓDI
Tehran, Iran
11-12 October, 2010

Host:
Ali Rabbani (rabbania@tums.ac.ir)
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

Moderators: Aghamohammadi A. (Tehran), Friedrich W. (Germany)

Diagnosis and treatment of primary immunodeficiency diseases
Aghamohammadi A. (Tehran)

Hematopoietic cell transplantation in the treatment of primary immunodeficiency disorders
Friedrich W. (Germany)

Moderators: Mansouri D., Rezaei N. (Tehran)

Primary immunodeficiencies presenting in adults
Mansouri D. (Tehran)

Clinical phenotypes of primary immunodeficiencies
Rezaei N. (Tehran)

Hyper-IgM syndrome (HIGM)
Kashef S. (Tehran)

Intravenous immunoglobulin replacement therapy
Mohammadzadeh I. (Tehran)

Updates on Autoimmune Lymphoproliferative syndrome (ALPS) and a case report
Cheraghi T. (Tehran)

The role of T regulatory cells in primary immunodeficiencies and autoimmunity
Soheili H. (Tehran)

Ataxia-telangiectasia in three patients presenting with Hyper-Immunoglobulin M syndrome
Nasiri R. (Tehran)

Oral and dental health in Iranian patients with primary antibody deficiency disorder
Javanbakht H. (Tehran)

L–R: Sina Nikaeen, Mohammad Gharagozlou, Masoud Movahedi, Ali Rabbani (Head of the meeting), Asghar Aghamohammadi (Scientific Director of the Meeting), Arndt Borkhardt, Mostafa Moin, Wilhelm Friedrich, Nima Rezaei (Executive Director of the meeting), Asghar Ramyar, Iraj Mohammadzadeh
CORRIGENDUM:  We apologize that the pictures on this page were accidentally shown on page 39 of the 2009 J Project Booklet. The Editors.
SUMMARY AND CONCLUSIONS

Each year an International Congress on Pediatrics takes place in Tehran and several PID experts attended these scientific congresses in Iran during last decade. 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 also 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 is also considered as The 46th J Project Meeting, is organized by the Department of Pediatrics, Children's Medical Center (Pediatrics Center of Excellence in Iran), Tehran University of Medical Sciences with collaboration of research centers, located at the Children’s Medical Center. The meeting focuses on the linkages of fundamental sciences and patient-oriented research under the main theme of clinical immunology, allergy and immunodeficiency diseases, whilst improving the awareness of physicians, pediatricians and other specialists was the main objective of the meeting.

The major part of audiences includes general practitioners, pediatricians, and pediatric sub specialists; however, medical students, pediatric residents and fellows in the field of clinical immunology and infectious diseases are expected to actively participate in the meeting.

Ali RABBANI,
Asghar AGHAMOHAMMADI &
Nima REZAEI

Tehran
J Project Meetings in Central and Eastern-Europe
2004-2010

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: Elisaveta 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, April 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 Mironksa
21. Iasi, Romania, October 12-13, 2007; Organizer: Aurica Ruginu
22. Ljubljana, Slovenia, November 16-17, 2007; Organizer: Tadej Avcev
23. Odessa, Ukraine, April 10, 2008; Organizer: Lyudmila Chernyshova
24. Sunny Beach, Bulgaria, May 22-23, 2008; Organizer: Elisaveta Naumova
25. Sarajevo, Bosnia-Herzegovina, October 10-11, 2008; Organizer: Velma Mulaosmanovic
27. Riga, Latvia, November 27-28, 2008; Organizer: Tatjana Prokofjeva
28. Timishoara, 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 Reisi
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
39. Tirana, Albania, April 8-9, 2010; Organizer: Gerorgina 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
## J Project Meetings in 2011 N° 47-58

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<td>47</td>
<td>Pushkin’s Hills (Russia)</td>
<td>Marina Guseva; <a href="mailto:gusevamarina@mail.ru">gusevamarina@mail.ru</a>; Areg Totolian <a href="mailto:totolian@spbraaci.ru">totolian@spbraaci.ru</a></td>
<td>Feb 3-4</td>
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<td>Mar 10-11</td>
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<td>Moscow (Russia)</td>
<td>Irina Kondratenko <a href="mailto:ikondratenko@rambler.ru">ikondratenko@rambler.ru</a></td>
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Debrecen Jeffrey Modell PID Reference Center

Genetic testing of PID genes in 2004-2010

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Summary: 414 81 43 20 23 18 2 2 3 3 2 1 5 1 618
Genes available for genetic testing in 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 (FASG)
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-Infecion-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 (CLEC6A)
33. Chronic mucocutaneous candidiasis (CLEC7A)
34. Chronic mucocutaneous candidiasis (MRC1)
35. Mucocutaneous candidiasis (CARD9)

LYSOSOMAL STORAGE DISORDERS (Genes)

36. Fabry disease (GLA)
37. Gaucher disease (GBA)
38. Niemann-Pick disease (SMPD1)

OTHERS (Genes)

39. Von Hippel-Lindau syndrome (VHL)
40. X-linked congenital adrenal hypoplasia (DAX1)
41. Galectin 3 deficiency (LGALS3)
42. Alström syndrome (ALMS1)
43. Atopic eczema (FLG)