

FOREWORD



This Booklet is designed to entice and challenge you with the concept of **The J Project**. To introduce you to this concept, let me give you some ideas and details.

In countries with lower socio-economic development, patients with rare diseases are often neglected and inadequately treated. Because of the limitations of diagnostic tools and treatment facilities, patients with rare primary immunodeficiency diseases (PID) either die or suffer throughout their life. The European Society for Immunodeficiencies (ESID) guidelines for the diagnosis of many PIDs are now available, and it is highly desirable to disseminate them so that the number of diagnosed and treated PID patients should increase.

The East-Central European Infectology and Pediatric Immunology Centre for Training and Research (ECE IPI CTR) was established in Debrecen in 1999 to develop a collaborative network of infectologists and immunologists in the East-Central European region. One of the recent initiatives of the Centre is to set up a PI information network and disseminate diagnostic and treatment guidelines among medical professionals as well as in the lay community in ECE countries. **The J Project** has been supported, in addition to ECE IPI CTR, by the ESID, the European Commission, the Jeffrey Modell Foundation and several pharmaceutical companies. The Project also embraces established physician education programs like the ones in the Czech Republic or Poland.

At an early stage of its development, I envisaged this Project being used primarily as a physician education program. However, a reasonable progression and an equally important application of this project may be a closer scientific collaboration between research groups of the so-called J-Countries. The task we jointly completed in 2004 with the concept of **The J Project** has lead us to outline an extended project with more emphasis on molecular genetic diagnosis of PIDs in ECE countries and joint research on PIDs in this region. I hope that our Project gives you the same excitement of exploration and sheer delight in the creative generation of ideas that we ourselves experienced in 2004.

Debrecen, March 15, 2005

László Maródi

THE J PROJECT

PID Awareness Meeting Series East-Central Europe 2004



| | CITY, Country | Date |
|----|---------------------------------|-----------------------|
| 1. | TARGU MURES, Romania | 11-12 March, 2004 |
| 2. | PRAGUE, Czech Republic | 10-11 May, 2004 |
| 3. | BELGRADE, Serbia and Montenegro | 11-12 June, 2004 |
| 4. | SKOPJE, Macedonia | 17-18 September, 2004 |
| 5. | KIEV, Ukraine | 18-19 November , 2004 |
| 6. | ZAKOPANE, Poland | 16-17 December, 2004 |

AIMS

To organize professional meetings on PID and related diseases in East-European countries with low number of registered PID patients.

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.

Updating national PID registries.

Establishing PID professional working groups.

Establishing PID patients' groups.

STRUCTURE OF MEETINGS

Informal discussion the night before the meeting.

Introductory lectures by invited and local speakers.

Case reports mostly by local speakers.

PID WG and patients' group activity in the country.

National registry update.



University of Debrecen
Medical and Health Science Centre

The **East-Central-European Infectology and Pediatric Immunology Centre for Training and Research (ECE IPI CTR)** was officially established on 22 December, 1999. From educational and scientific points of view, the Centre is closely associated with the **Department of Infectology 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 the Centre are as follows:

host defense mechanisms against bacteria
characteristics of immune defense mechanisms against fungi
molecular pathology of primary immune deficiency diseases
mechanisms of action of intravenous immunoglobulins
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 a regional Infectology 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-Central 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 function of which is the formulation of educational and scientific co-operation within the region of East-Central-Europe.

TARGU MURES, ROMANIA

11-12 MARCH, 2004

HOST: CSILLA TODEA

todea.cs@rdslink.ro



ORGANIZING COMMITTEE:

Despina Baghiu (Targu Mures)

Maria Cucuruz (Timisoara)

Zoltán Ellenés-Jakabffy (Oradea)

Eva Kiss (Targu Mures)

László Maródi (Debrecen)

Csilla Todea (Targu Mures)

VENUE: 'Bod Peter' Deaconesses' House, Targu Mures

MAIN TOPIC:

Perspectives of PID Patient Care in Romania



PROGRAMME: 12 March

Presentations

Primary Immune Deficiencies in the Paediatric Clinical Practice

Ioan Gherghina, Nicolae Iagaru, Adrian Cochino (Bucharest)

Epidemiological aspects of PID in Romania

Margit Serban, Maria Cucuruz, Mihaela Bataneant, Mihai Gafencu (Timisoara)

Diagnostic difficulties in some forms of PID

Maria Cucuruz, Margit Serban, Estera Boeriu, Mihaela Bataneant, Mihai Gafencu (Timisoara)

Rules and regulation in administration of intravenous immunoglobulin (IVIG) in primary immunodeficiency diseases

Despina Baghiu (Targu Mures)

Organization of PID patient care in Hungary

Laszlo Marodi (Debrecen)

Diagnosis and treatment of PID patients in Macedonia

Sonja Peova (Skopje)

Registered PID cases of the 2nd Paediatric Unit of Cluj

Mariana Marc, Nicolae Miu, Cristina Marii (Cluj)

The Paediatric PID Registry of Bihar County

Zoltan Ellenes-Jakabffy¹, Laszlo Marodi², Ligia Burta¹, Timea Evanics¹, Corina Nagy¹, Laszlo Ritli¹, Gabriela Miculschi¹, Rodica Mihalceanu¹, Radu Spineanu¹, Maria Praja¹ (¹Oradea, ²Debrecen)

Primary immunodeficiency cases hospitalized in the 2nd Paediatric Clinic of Iasi

Aurica Rugina, Stela Gotia (Iasi)

Infections in PID patients with IVIG therapy

Zoltan Ellenes-Jakabffy¹, Laszlo Marodi² (¹Oradea, ²Debrecen)

Necrotizing fasciitis in patient with hyper-IgE syndrome

Melinda Erdos (Debrecen)

C1-esterase inhibitor deficiency-case presentation

Gabriela Miculschi¹, Zoltan Ellenes-Jakabffy², Laszlo Ritli¹, Maria Cucuruz² (¹Timisoara, ²Oradea)

C4 deficiency-clinical aspects

Laszlo Ritli, Claudia Cladovan, Zoltan Ellenes-Jakabffy, Gabriela Miculschi, Ana Popa (Oradea)

PID case diagnosed at the Gheorgheni City Hospital

Maria Orosz (Gheorgheni)

PID cases diagnosed at the 1st Paediatric Unit of the Targu Mures University Medical School

Csilla Todea, Timea Imre (Targu Mures)

PID case report

Mihaela Bataneant (Timisoara)

SUMMARY AND CONCLUSIONS:

The first PID awareness event of the J Project, initiated by the ECE IPI CTR, was organized in Targu Mures, Romania, together with the Romanian Immunodeficiency Working Group. Dr. Csilla Todea of Targu Mures took care of the local organization. She did a wonderful job in putting together the program, inviting doctors from all over Romania and from abroad, and in selecting a great venue for the meeting in the Bod Peter Deaconesses' house. The meeting was attended by more than 60 pediatricians interested and/or involved in PID patient care. Doctors from other East-European countries including Ukraine (Dr. Alla Volokha, Kiev), Macedonia (Drs. Sonja Peova and Katarina Stavrik, both from Skopje), Yugoslavia-Monte Negro (Dr. Srdjan Pasic, Belgrade) were also attending. ECE IPI CTR was represented by Prof. Laszlo Marodi and Dr. Melinda Erdos both from Debrecen.

Reports on recent developments and current problems in PID patient care in Romania were presented. There were several, very nicely presented case reports (Drs. Zoltan Ellenes-Jakabffy, Melinda Erdos, Gabriela Miculschi) as well. The Romanian Primary Immunodeficiency Working Group has been established on April 2, 2003, in Oradea. That time the number of PID patients registered in the country was very limited. In Targu Mures, less than a year later, Dr. Maria Cucuruz and her colleagues presented 173 patients in the national PID registry! This number showed what an active search for immunodeficient patients has been going during the past year in Romania. Importantly, most of the patients registered are from Transylvania indicating that professional activity in the field is not balanced yet in the country. From this perspective, it must be emphasized how important it was to have senior pediatricians from the Bucharest area (Prof. Ioan Ghergina) and from Iasi (Prof. Aurica Rugina).

The first steps were taken to organize the Romanian parents' group as well. A couple from Targu Mures, parents of a patient with XLA, joined the meeting and appeared to be ready to organize the group. Drs. Todea and Ellenes promised to help their work. It was proposed to apply for a WIN grant offered specifically for parents' groups by the Jeffrey Modell Foundation.

Diagnosis and treatment of PID patients in Romania is inappropriate. There are areas in which diagnostic reagents are available only for 5-6 months of the year, even for screening assays. Only a few patients are on regular IVIG, and bone marrow transplantation as a treatment facility has just become available for pediatric oncology patients only. Therefore, it was offered that:

Serum samples for immunochemical analysis (measurement of concentrations of IgG, IgA, IgM, IgE, C3, C4, anti-tetanus antibody, anti-pneumococcus antibody and activity of CH50) should be sent to Debrecen to ECE IPI CTR which will cover the expenses until the limitation of its budget.

Six young doctors from different areas of the country should become member of ESID. The membership fees for two years will be covered by ECE IPI CTR. ECE IPI CTR will organize laboratory training for a small group of Romanian colleagues in Debrecen.

It was proposed to target health authorities in Romania in order to improve the availability of treatment.

It was suggested to send an abstract describing Romanian PID registry to the coming ESID meeting in Versailles and to apply for travel grants from the organizers.

Finally, everyone was encouraged to attend to the next J Project events in Prague (May) and in Belgrade (June).

László MARÓDI

MEDIA RESPONSE:

Maria Cucuruz, Csilla Todea: Positive evolutions in the diagnosis of primary Immunodeficiencies. Viata Medicala. p.1-4, 7, 2004.

Maria Cucuruz, Csilla Todea: Primary immunodeficiencies - guide for diagnosis and treatment. Viata Medicala. p.2, 11, 2004.

Annamaria Nagy: Immunologiai kerekasztal-megbeszeles. Marosvasarhelyi Nepujzag. VI: 15626, 56, 2004.

Gyongyi Bodolai: Ismerni es kezelni kell a betegeket. Marosvasarhelyi Nepujzag. VI: 15630, 60, 2004.

SPONSORS: Baxter, Biotest Pharma GmbH, ECE IPI CTR, EURO PID NAS, Glaxo, Humanserum, Sankio

PRAGUE, CZECH REPUBLIC

10-11 MAY, 2004

HOST: ANNA SEDIVA

annasediva@hotmail.com



ORGANIZING COMMITTEE: Ales Janda (Prague)

Anna Sediva (Prague)

VENUE: Institute of Immunology, 2nd Medical School, Charles University, Prague

MAIN TOPIC:

ESID Prague Spring Meeting 2004



PROGRAMME:

10 May

Presentations

ESID Register of PID

Bodo Grimbacher (Freiburg)

The Paediatric PID Register of Bihar County

Zoltan Ellenes-Jakabffy (Oradea)

Problems and Reality of Russian Registry of Primary Immunodeficiencies

Olga Paschenko (Moscow)

View on the German Support Group DSAI Deutsche Selbsthilfe Angeborene Immundefekte e. V. - Subcutaneous Immunoglobulin Treatment - Experience of a Patient

Eva Soergel (Nuremberg)

Peripheral and Central Autoimmunity due to Immune Dysregulation

Hans D. Ochs (Seattle)

APECED - Autoimmune Endocrinopathy-Candidiasis-Ectodermal Dystrophy

Anna Sediva (Prague)

Hematological manifestations of primary immunodeficiencies

Anna Yurasova (Moscow)

X - Linked Lymphoproliferative disease (Case report)

Marina Praprotnik (Ljubljana)

11 May

Presentations

Clinical and Laboratory Presentation of IgM Deficiency

Jiri Litzman (Brno)

Recognition Receptors of Innate Immune System

Radek Spisek (Prague)

Unusual presentation of IRAK4 deficiency

Helen Chapel (Oxford)

Severe Combined Immunodeficiency due to Rag and Artemis defects

Anna Villa (Segrate)

Frequency of MBL Defective Haplotypes in Czech Population

Tomas Freiburger (Brno)

Fatal Outcome in Two Siblings with Thymic Hypoplasia and severe combined immunodeficiencies (Case report)

Rasa Duobiene, Radvile Malickaite (Vilnius)

Successful Allogenic SCT in Infant with Familial Haemophagocytic Lymphohistiocytosis, Dilemmas in Using Young "Unaffected" Sibling

Petr Sedlacek (Prague)

Suspected INF Gamma Receptor Deficiency

Laszlo Vaszil (Budapest)

Peculiarity of Mycobacterial Infection in Primary Immunodeficiency Patients

Elizabeth Galkina (Moscow)

Hyper IgE Syndrome (Case report)

Hana Kayserova, Alena Hlavata, Petr Ciznar (Bratislava)

Preterm Baby Suspected of Omenn Syndrome (Case report)

Pavel Rozsival, Eva Parizkova (Hradec Kralove)

**Bone Marrow Transplantatiton in X-linked T-B+NK- SCID patient with
Pneumocystis carinii pneumonia (Case report)**

Lucie Sramkova, Ales Janda (Prague)

Social programme

Bike trip "Prague-Brno"



SUMMARY AND CONCLUSIONS:

On May 10 and 11, 2004, the third ESID Prague Spring meeting was held in the University Hospital Motol, Prague. This year, 35 participants from 10 countries, namely Czech Republic, Germany, Hungary, Italy, Lithuania, Slovakia, Slovenia, Romania, Russia and USA, attended and actively participated in the event. The mean age of the participants was 36 years (with a range from 26 to 68), thus demonstrating the continuous exchange of knowledge between the generations among immunologists in Eastern European countries.

The third ESID Prague Spring Meeting was part of the J Project, a coordinated activity of Eastern European countries, represented by a chain of successive meetings in these countries, with the aim to increase the information about PID in Eastern Europe.

The main task of the Prague ESID Meeting has traditionally been the attempt to

facilitate the exchange of information on PID between Western and Eastern Europe. This task was accomplished this year by an excellent attendance from these formerly separated, now mostly unified regions. Invited participating speakers for 2004 were Helen Chapel, Hans Ochs, Anna Villa and Bodo Gribbacher.

Topics for the first day were targeted mainly at the registries of PID in the countries of the participants. The topic was opened by the presentation of the European PID registry by Bodo Gribbacher. The program continued with the introduction of a new cluster of diseases on the border between primary immunodeficiencies and autoimmune disorders, namely the group IPEX-APECED-ALPS, presented by Hans Ochs. News on SCID and the disorders of innate immunity were on the program on the second day of the meeting. A large part of the program was devoted to case reports. Unusual and interesting cases were valued most among all participants. This year's meeting introduced a new section concerning the involvement of patients in the PID area.

The meeting offered complementary activities, composed of information about the hosting facility, the 2nd Medical School and Charles University and University Hospital Motol in Prague, and an excellent cultural and social program. The meeting was followed by a bike trip that further enriched the basic motto of Prague Spring Meetings - to facilitate the spread of information of PID among immunologists. Eight participants took part in the bike trip that started in Prague and finished in Brno, in the laboratory specializing in the molecular diagnosis of PID. The meeting was supported by EURO-PID-NAS project. Ten travel grants were provided for East-European participants. This year's meeting confirmed the still growing scientific quality of the conference and the usefulness of these educational activities. The plans for the next year have already been initiated.

The programme with PowerPoint presentations (with the consent of the authors) and photo documentation is available at <http://imunologie.lf2.cuni.cz/esid2004/program.htm>.

Anna SEDIVA

SPONSORS: Baxter, EURO PID NAS, Immunotech, University Hospital Motol

BELGRADE, SERBIA AND MONTENEGRO

11-12 JUNE, 2004

HOST: SRDJAN PASIC

bg.pasic@eunet.yu



ORGANIZING COMMITTEE: Gordana Bunjevacki (Belgrade)
Koruga Dragana, YUGOPID (Belgrade)
Laszlo Marodi (Debrecen)
Aleksandra Minic (Belgrade)
Srdjan Pasic (Belgrade)
Jelena Tomic (Novi Sad)

VENUE: Mother and Child Health Institute, Belgrade

MAIN TOPIC:

4th Primary Immunodeficiency Awareness Meeting



PROGRAMME: 12 June

Presentations

The J Project: An East-Central European perspective

Laszlo Marodi (Debrecen)

Primary antibody deficiency syndromes: our experience

Srdjan Pasic (Belgrade)

Respiratory care in PID patients

Predrag Minic (Belgrade)

Polish registry of PID DNA repair defects

Malgorzata Pac (Warsaw)

University Childrens' Clinic experience in PID

Dragana Janic (Belgrade)

Current status of PID in Ukraine

Alla Volokha (Kiev)

Round table: **National PID registries**

Hyper IgE syndrome (Case report)

Jelena Tomic (Novi Sad)

Hyper IgE syndrome: Review and case presentation (Case report)

Melinda Erdos (Debrecen)

Hyper IgA (Case report)

Sonja Peova (Skopje)

Hemophagocytic lymphohistiocytosis in PID

Milos Kuzmanovic (Belgrade)

SUMMARY AND CONCLUSIONS:

The 4th meeting of the Central East European initiative of awareness in PID, the J Project took place at the Mother and Child Health Institute, Belgrade, Serbia and Montenegro on the 12th of June, 2004. An introductory lecture about Central East European perspectives in PID was given by Prof. Laszlo Marodi, Debrecen, Hungary.

Antibody deficiency syndromes in Serbia and Montenegro were presented by Dr. Srdjan Pasic and Dr. Predrag Minic, Mother and Child Health Institute, Belgrade, while Dr. Dragana Janic, University Children Clinic, Belgrade presented a group of patients with presumed or proven PID. Our invited guests were Dr. Malgorzata Pac, Warsaw, Poland who gave a lecture about the Polish registry of DNA breakage syndromes and Prof. Alla Volokha from Kiev, Ukraine who presented an increasing number of patients with recognized PID in Ukraine. The other participants from regional centers in Hungary, Serbia and Macedonia were Dr. Melinda Erdos, Dr. Jelena Tomic, Dr. Milos Kuzmanovic and Prof. Sonja Peova, who presented case reports of patients affected with PID.

This one-day meeting was important for making contact among physicians who are engaged in diagnostics and care of PID patients. The participants also agreed that existing hospital registries of PID patients will be updated and the data should be joined together into a national registry.

Srdjan PASIC

SPONSOR: Jugohemija AA

SKOPJE, MACEDONIA

17-18 SEPTEMBER, 2004

HOST: KATARINA STAVRIK

kstavric@hotmail.com



ORGANIZING COMMITTEE:

Lidija Kareva (Skopje)

Laszlo Marodi (Debrecen)

Kristina Mironska (Skopje)

Sonja Peova (Skopje)

Katarina Stavric (Skopje)

VENUE: University Children's Hospital, Clinical Centre, Skopje

MAIN TOPIC:

Teaching Pediatric Immunology in East-Central Europe



PROGRAMME:

18 September

Presentations

The J project

Laszlo Marodi (Debrecen)

Teaching pediatric immunology in Macedonia

Sonja Peova (Skopje)

Possibilities for Immunodeficiency Diagnosis at the Institute of Immunobiology and Human Genetics, Faculty of Medicine, Skopje

Mirko Spirovski (Skopje)

Molecular basis of HIGM syndrome phenotype

Laszlo Marodi (Debrecen)

De George Syndrome

Anna Sediva (Prague)

Selective antibody deficiency syndrome

Melinda Erdos (Debrecen)

HCV in children with PID

Kristina Mironska (Skopje)

MHC class II Deficiency (Bare Lymphocyte syndrome) (Case report)

Snejina Mihailova (Sofia)

Hiper IgE Syndrome (Case report)

Lidija Kareva (Skopje)

CVID (Case report)

Katarina Stavric (Skopje)

Round table: **World Immunodeficiency Network**

Social programme

Sightseeing of Skopje



SUMMARY AND CONCLUSIONS:

The 4th PID event was organized in Skopje, Macedonia, by Dr. Stavric, Dr. Mironska, Dr. Kareva and Dr. Peova, members of the Macedonian Immunodeficiency Working Group. The meeting venue was the University Children's Hospital in Skopje. In addition to the organizers and local doctors, 20 pediatricians and immunologists involved in PID patient care and PID laboratory diagnosis attended from Macedonia. Doctors Shumkov and Mihailova from Sofia, Bulgaria, Prof. Laszlo Marodi and Dr. Melinda Erdos from Debrecen, Hungary, and Prof. Dr. Anna Sediva from Prague, Czech Republic attended from abroad.

The University Children's Hospital has a primary role in the education of paediatricians in Macedonia. The Department of Immunology of the University Children's Hospital, was established in 1976 to assure clinical and laboratory investigation of children with Immunodeficiencies. It provides both ambulatory and inpatient consultation services for children with all forms of immune

disorders. Over the years, our Department became a centre for PID patients in Macedonia. Children with PID are registered by our Department. We have registered 32 patients until now (selective IgA deficiencies are not taken into account). The number of PID patients is limited because we miss the adult patients with PID. It was therefore important at this meeting that we invited internists and specialists for infectious diseases to increase the awareness of PID in adults. It was decided that Dr Mironska will organise the Macedonian PID Registry and will report the patients to the European Registry. She will attend the conference in Versailles, thanks to the ECE IPI CTR in Debrecen covering her registration fee. Also five doctors from Macedonia should become members of ESID. ECE IPI CTR will cover the membership fee in concert with the J Project initiative.

We visited the Institute of Immunobiology and Human Genetics, Faculty of Medicine before the Meeting was started. The Institute was established in 1990 to improve genetic diagnosis of inherited diseases. Report on possibilities for immunodeficiency diagnosis at the Institute of Immunobiology and Human Genetics was discussed in detail. Current program for teaching pediatric immunology, PID patient care, review on Hyper IgM syndrome and case reports were presented and discussed. Importantly, a mother of a patient with PID attended the meeting and she is prepared to organize the Macedonian PID Parent's Group with help of Dr. Stavic. It was decided to apply for WIN grant offered by Jeffrey Modell Foundation. Finally it was encouraged to attend the next meeting in Kiev in November where the two months activities from the meeting in Skopje will be reported. ECE IPI CTR will cover the travel grant for one or two Macedonian doctors.

Katarina STAVRIC

SPONSORS: ECE IPI CTR, EURO PID NAS, GlaxoSmithKline, Krka-lek, Libra 1

KIEV, UKRAINE

18-19 NOVEMBER, 2004

HOST: ALLA VOLOKHA

volokha@carrier.kiev.ua



ORGANIZING COMMITTEE: Ludmila Chernyshova (Kiev)

Fedir Lapij (Kiev)

Laszlo Marodi (Debrecen)

Dmitriy Samarin (Kiev)

Alla Volokha (Kiev)

VENUE: Kiev Medical Academy of Postgraduate Education

MAIN TOPIC:

Management of Infections in Children with Cellular Immunodeficiencies



PROGRAMME:

19 November

Presentations

Development of PID care in Ukraine

Ludmila Chernyshova (Kiev)

PID care in Charkiv Immunological Center

Ludmila Pushkarenko (Charkiv)

PID care in Lwiw Immunological Center

Larisa Kostyuchenko (Lwiw)

Progress in activities of PID Working group in Macedonia

Katarina Stavric (Skopje)

X-linked lymphoproliferative disease

Laszlo Marodi (Debrecen)

HIGM Syndrome

Irina Kondratenko (Moscow)

Twenty five years of experiences with intravenous (IVIG) and subcutaneous (SCIG) immunoglobulin substitution therapy in antibody deficient children in Poland

Malgorzata Pac (Warsaw)

Bone marrow transplantation in children with PID

Evgeniy Pashanov (Moscow)

Treatment and prophylaxis of infections in children with PID

Alla Volokha (Kiev)

Interleukin-1 receptor associated kinase-4 deficiency

Melinda Erdos (Debrecen)

Complement deficiency

Andrey Prodeus (Moscow)

The clinical picture, diagnosis and therapy of hereditary angioedema (HAE)

Henriette Farkas (Budapest)

Establishment of the Central-Eastern European Center of HAE

Lilian Varga (Budapest)

Difficulties of diagnostics in some forms of PID

Fedir Lapij, Dmitriy Samarin (Kiev)

Diagnostic facilities of PID in Ukraine

Viktor P. Chernyshov (Kiev)

Case report - CVID

Larisa Vasilchenko (Lwiw)

Social programme

Ukrainian-style dinner and Sightseeing of Kiev



SUMMARY AND CONCLUSIONS:

The 5th PID awareness meeting in East-Central Europe referred to as the J Project was organized in Kiev, Ukraine, together with the Department of Pediatric Infectious Diseases and Immunology, Kiev Medical Academy of Postgraduate Education. The Meeting was attended by more than 100 pediatricians, most of them pediatric immunology specialists who are interested and involved in PID patient care in this large country of 48 million inhabitants.

The participants were from all 27 regions of Ukraine. Doctors from East-Central Europe attending the Meeting were: Malgorzata Pac, Warsaw (Poland), Katarina Stavric, Skopje (Macedonia), Irina Kondratenko, Andrey Prodeus, and Evgeniy Pashanov from Moscow (Russia), Henriette Farkas and Lilian Varga from Budapest (Hungary). ECE IPI CTR and the Debrecen Jeffrey Modell Center were represented by Laszlo Marodi and Melinda Erdos from Debrecen (Hungary). According to the traditions at J Project meetings, most presentations were in the local language, but the speakers presented figures and tables with English text. English presentations by invited speakers were translated into Ukrainian to assure optimal targeting of the messages. This worked out very well here, as well as at previous meetings.

Currently, there are three acknowledged pediatric immunology centers in Ukraine. Ludmila Chernyshova presented data on recent developments and current problems in PID patient care in Ukraine, in general, and in Kiev in particular. There were also reports from the other two centers, Kharkiv and Lwiw,

by Ludmila Pushkarenko and Larisa Kostyucsenko, respectively, on recent achievements in diagnosis and treatment of PID patients in Eastern and Western regions of the country. Katarina Stavric reported on impressive achievements and increase of the overall activity of the PID Working group after the recent J Project Meeting in Macedonia, including the establishment of the Parents' group. The Macedonian Parents' group is applying for a WIN grant, and provides help to initiate parents' activity in two neighbouring countries, Bulgaria and Serbia-Montenegro.

Irina Kondratenko updated us on the immunologic and genetic spectrum of hyper-IgM syndrome, and presented data of patients in Russia. Laszlo Marodi gave a review on XLP and presented a Hungarian family with two male patients who died of FIM. A case report on IRAK-4 deficiency was presented by Melinda Erdos. These presentations emphasized the importance of molecular genetics in carrier detection and prenatal diagnosis in X-linked PIDs. It was announced that genetic analysis of 10 PIDs is now available in Debrecen for the J Project countries.

Malgorzata Pac from Warsaw gave an overview on twenty-five years of experience with IVIG substitution, and she discussed the perspective of SCIG substitution. Evgeniy Pashanov shared with us his experience on bone marrow transplantation in children with primary immunodeficiency in Russia. I gave a lecture on treatment and prophylaxis of infections in immunocompromised patients.

There were several presentations on complement deficiencies, including complement in health and disease by Andrey Prodeus. Henriette Farkas reviewed the diagnosis and treatment of hereditary angioedema (HAE) and Lilian Varga presented data on the importance of developing centers for diagnosis and management of this disease. They are able to help East-European doctors to make the diagnosis of HAE.

It is remarkable how active the audience was during the discussions following the presentations and continuing during breaks and dinner. Participants of this meeting received useful information about new approaches to diagnosis and treatment of primary immuno-deficiencies. I strongly believe that the J Project Meeting in Kiev was a very successful event not only because it provided a useful educational program but also because we could summarize current problems and outline future responsibilities in PID patient care in Ukraine.

Alla VOLOKHA

SPONSORS: Biopharma, ECE IPI CTR, EURO PID NAS

ZAKOPANE, POLAND

16-18 DECEMBER, 2004

HOST: EWA BERNATOWSKA

bernatowskae@yahoo.com



ORGANIZING COMMITTEE: Anna Berglof (Stockholm)
Ewa Bernatowska (Warsaw)
Teresa Espanol (Barcelona)
Andrzej Mackiewicz (Poznan)
Malgorzata Pac (Warsaw)

VENUE: Hotel 'Nosalowy Dwor', Zakopane

MAIN TOPIC:

Progress in Education in Clinical Immunology



PROGRAMME:

18 December

Presentations

New specialization in medical laboratory immunology

Andrzej Mackiewicz (Poznan)

Invitation to Congress of Immunology in Lublin, 18-22.05.05

Jacek Rolinski (Lublin)

The J project

Ewa Bernatowska (Warsaw)

EURO- POLICY- PID

Anna Berglof (Stockholm)

New ESID online registry

Barbara Frisch (Freiburg)

Safety and effectiveness of IVIG preparations in PID patients

Zoltan Ellenés-Jakabffy (Oradea)

PID registry in Ukraine

Alla Volokha (Kiev)

CVID - classification with B memory cells

Teresa Espanol (Barcelona)

V (D) J recombination defects in T/B SCID patients

Mirjam van der Burg (Rotterdam)

HIGM syndrome in Polish Registry

Edyta Heropolitanska-Pliszka (Warsaw)

19 December

Presentations

Diagnostic strategies for PID patients

Mirjam van der Burg (Rotterdam)

SCID patients; genotype, phenotype and therapy

Beata Wolska-Kusnierz (Warsaw)

Di George syndrome - Polish subregistry

Malgorzata Pac (Warsaw)

Social programme

Walk in Zakopane

SUMMARY AND CONCLUSIONS:

The second Autumn - Winter School of Clinical Immunology was organized in Zakopane, a beautiful small town lying at the foot of the highest Polish mountains - Tatra. There were fifty-two participants at this meeting. A group of young immunologists from both Central-Eastern and from Western European countries also attended the meeting, among which young Polish immunologists were also represented, together with physicians specializing in clinical immunology. This group has grown and now is composed of 26 clinicians.

The topics of the meeting ranged from the current status of primary immunodeficiency diseases in individual countries, including national registers, to the newly created ESID Online Registry. Advances in molecular diagnosis of primary immune deficiencies and therapy including long-term follow-up of patients who had received bone marrow transplantation, as well as recent advances in diagnosis and classification of CVID and wide spectrum of CVID cases, were presented. The diagnostic and therapeutic guidelines of some primary immune deficiencies were also discussed. This year's invited speakers were Teresa Espanol, Mirjam van der Burg, Barbara Frisch and Anna Berglof.

The second Autumn - Winter School of Clinical Immunology has brought the J Project to an end. A chain of successive meetings organized in Central-Eastern European countries offered tight co-operation between countries, and increased their attitude to future collaboration.

The conference was located opposite to a well-built ski area. During the lunch break some speakers and young colleagues were skiing, others visited the colourful crowd of the village.

The next meeting will be held in Gdansk at the Baltic coast, September 23-24, 2005. For more information, please, contact us at immuno@czd.waw.pl.

Ewa BERNATOWSKA

MEDIA RESPONSE: RFMFN - radio report, December 18, 2004 on "Infectious diseases and immune response"

SPONSORS: EURO PID NAS, Octapharma, PERFECT

WE INVITE YOU TO THE FORTHCOMING MEETING OF THE J PROJECT!

SOFIA, BULGARIA

9-10 MAY, 2005

HOST: ELISSAVETA NAUMOVA

immun@medun.acad.bg



VENUE: Medical University, Sofia

MAIN TOPIC:

Recent Advantages in the Diagnosis and Therapy of PID

SCHEDULE OF THE PROGRAMME:

15 April

18:00-19:00 Opening ceremony & Introductions

19.30 Welcome dinner

16 April

9:00-10:30 European Projects:
The J project
EURO- POLICY- PID

10:30-11:30 **National Registries** (Country Representatives - 15 min each)

11:30-11:45 **Molecular defects associated with PID**

11:45-12:00 **Mutant genes in PID and clinical use of molecular genetic testing**

12:00-12:15 **Role of flow cytometry in the diagnostic evaluation of PID**

12:15-12:30 **Diagnostic strategies for PID patients**

12:30 Visit at the Central Laboratory of Clinical Immunology,
University Hospital "Alexandrovska"

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| 13:30-14:30 | Lunch |
| 14:30-16:30 | Case reports (15 min each) |
| 16:30-17:00 | Coffee break |
| 17:00-17:20 | Bone marrow transplantation in PID treatment strategies |
| 17:20-17:40 | IVIG therapy in PID diseases |
| 17:40-18:00 | Gene therapy |
| 18:00-18:20 | Network for molecular genetics diagnosis of PID |
| 18:20-19:00 | Concluding remarks |
| 20:00 | Dinner and social event |