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President's Corner

Dear Friends,

Well, time is running and here we are at the summer just before the vacation. Indeed I believe that many of us deserve this vacation due to the hard work that was done in the field of PID.

As you may recall last month was the "World PID week" which I hope increase the awareness for immunodeficiency worldwide.

The J project, run by Prof. Lazlo Marodi just had their 50th meeting! We all congratulate Lazlo for his efforts and success. PID meetings for young physician/researchers were also done this winter in Prague and in Finland and the feedback that we got from these events were excellent.

After the successful meeting in Istanbul, Eleonora and the scientific committee have already started in organizing the 2012 ESID meeting in Florence which I'm sure will be an exciting one. So many new genes are discovered, which causes PID (for example look at NEJM, 2011 or Nature 2011), that I 'm looking forward in great expectation to our meeting in Italy.

The ESID board met in Geneva last month in order to reformulate the mission and vision of our society. This was a stimulating event and we are now finalizing the report and hopefully in the next newsletter, it will be published. Wishing you all a happy summer vacation

Amos Etzioni ESID President

Secretary Report

Dear ESID members,

The Executive Board met in Geneva on 20 May 2011 for two days. The main purpose of the Executive meeting was to further consolidate the future of the Society and reach a consensus on the organisation and programme of the next Biennial ESID Meeting, which will be held in Florence in 2012. Many topics were discussed:

- ESID support for scientific meetings: because of the increasing numbers of requests by meeting organisers asking to advertise their events in the "Scientific meeting" section of the ESID website, the Board has decided that the Educational Working Party will have the right of the final decision. In particular, it will be asked the organizers to submit the scientific programme. ESID logo will be granted only upon the approval of the ESID board.
- The ESID Board has decided to finance the travel expenses of Amos Etzioni and Christoph Klein to the next LASID meeting in Mexico City next October 2011.
- The ESID website: it is now urgent to improve our website in order to make it more attractive and user firendly. We are trying to optimise the search engine, although it is not so easy to find the "perfect" website. Our plan is to investigate the websites created by other scientific societies. Please email me or Susanne Schimidt any suggestions and comments you may have. We think that your contribution to the new website design will be crucial.
- Our Treasurer, Eleonora Gambineri, is working hard to finalise the scientific programme of the next biennial ESID meeting. The Board discussed the programme at the IUS congress in New York at the beginning of June and proposed emerging scientific fields.



Finally I would like to mention several events that have already taken place or will be taking place in the near future.

ESID Summer School

The next ESID Summer School, which represents an excellent opportunity to update PID knowledge. It will be held in Tuscany, Italy (Resort II Ciocco) next 28 September. The applications closed in the middle of June.

Joint Scientific events

Other scientific events have been organised in different European countries in conjunction with the ESID. A few examples: the ESID Junior Symposium organised in Finland (16-17 June 2011) by Crina Samarghite and and Mauno Vihinen.

The 10th ESID Prague Meeting organized by Anna Sediva on 9-10 May at the University Hospital Motol, Prague, Czech Republic.

A number of travel grants have been given to attend these workshops.

Education

In June, short-medium and long term fellowships were offered to physicians or scientists interested in learning diagnostic/therapeutic procedures or lab techniques in the field of PID.

ESID top registry publication award 2010

ESID 2010 awards went to the French PID study group CEREDIH for its study of a large cohort of patients (more than 3000 PID cases) and to a German group directed by Dr. M. Albert for an analysis of 173 patients affected by X-linked trombocytopenia (XLT).

I would like to ask all of you to support our Society by convincing new members to join ESID. I would like to remind you the benefits of ESID membership:

- Reduced registration fee to ESID biennial congress
- Reduced registration fee to FOCIS meetings New!
- Access to a privileged network of PID professionals
- Eligibility to receive awards: travel grants, fellowships, publications
- Preference to take part in ESID PID Schools
- ESID Newsletter (quarterly)
- Access to ESID website members only area

With my best for a wonderful summer All the best

Anna Villa ESID Secretary

News: ESID Working Parties

BMT & Gene Therapy

Dear All,

It's the middle of the year and it seems to have gone by very quickly indeed. We had a successful working party session in Paris at EBMT and we are already moving onto the Autumn meeting which will be held in Belgrade on 6th - 8th October.



Mario Abinun is hosting this year's event and it is our first venture into the Balkans. We are hoping to try and encourage attendance from some of the Balkan and eastern European nations so that we promote the optimal management of patients with severe immunodeficiencies in these countries.

The J project under Lazlo Marodi's has done a tremendous job in promoting education and awareness of PIDs but we need to take this onto the next level with the aim of providing the best management possible. Details of the meeting and the programme will be available on the website soon.

I am happy to say that the EBMT IEWP guidelines for transplantation of SCID and other severe immunodeficiencies are now available on the ESID and EBMT websites. Thank you to all the different contributors and I hope these guidelines will help in providing the best treatment protocols for our patients. In Belgrade we will need to assess how well they are being followed and used. Personally I have received far fewer e-mails asking for how patients should be transplanted. That may mean that the guidelines document is doing its job or just that people really don't want my advice anymore!

I also recently went to a meeting at the EU organised by IPOPI to try and convince EU members that all EU nations should adopt newborn screening for SCID. Newborn screening is now one of the major issues surrounding management of SCID. it is absolutely clear that identifying SCID at birth and preventing infants from contracting opportunistic infections leads to a better survival outcome (see Brown et al., Blood 2011 Mar 17;117(11):3243-6). It will also tell us truly what the incidence of SCID in Europe is. The USA have already mandated that all states should adopt newborn screening and 4 states currently screen all newborns. The data coming out is already extremely interesting and highlights that SCID may be far more frequent than we originally imagined. The technology to screen is already available and I think we should all be putting pressure on national screening committees to adopt SCID screening. The sooner screening is adopted, the sooner we will be able to improve their chances of survival. The EU meeting will, I hope, be the start of the process to try and raise this issue and exert some influence on member states to adopt this screening programme.

I hope you all have a good summer.

Best wishes Bobby Chairperson ESID WP Genetics

Clinical

Meeting on how to update diagnostic criteria in PID

New York, June 4, 2011

Participants:

Helen Chapel, Mary Ellen Conley, Luigi Notarangelo, Klaus Warnatz Absent: Amos Etzioni

Aim of the meeting:

To discuss the first steps for the update and development of diagnostic criteria of PID.

Discussion:

There was general agreement that several diagnostic criteria need to be updated and some to be newly developed. Currently there is no accepted standard procedure how to achieve this. In order to facilitate the operational procedures an executive board needs to be founded. For the general acceptance of the procedure a broad geopraphical representation and involvement of different societies and especially IUIS



is important.

Suggestions:

An executive Board of five people with experience in PID from different parts of the world and belonging to various organisations with an interest in PID should be nominated.

Suggested participants:

Antonio Condino-Neto (Latin America) Mary Ellen Conley (North America, IUIS) Yu-Lung Lau (Asia) Reinhold Schmidt (Europe, IUIS Committee Chairman Clinical Immunology) Mimi Tang (Australia, IUIS) Klaus Warnatz (Europe, ESID) All participants were addressed and agreed to participate.

Tasks of the board:

- 1) Agree on the purpose and intended use of the diagnostic criteria.
- 2) Produce a SOP for the development of updated or new diagnostic criteria in PID. This SOP needs to be communicated to IUIS and the continental societies of immunodeficiency.
- Publish the SOP through the societies and invite experts to found committees to suggest areas of need and develop diagnostic criteria.
- 4) Develop a master copy for diagnostic criteria which will be used by the experts committees.
- 5) Supervise the ongoing activities by ensuring that experts in the field are including for each disorder, the correct procedure according to SOP, time lines (if not sufficiently controlled by expert committees), communication of the proposed draft of diagnostic criteria to societies, a correct review process (needs to be defined by the board in agreement with societies), by confirming a correct procedure and by communicating the final draft to societies.

Mode of action:

Decision processes should be based on consensus and no veto. Communication is by Email, tele- or videoconference.

Genetics

ESID Genetic WP second announcement

The ESID Genetic Working Party wants to help to connect and support the collaborative clinical, genetic laboratories and research units associated with the ESID society.

The exchange of genetic diagnostic tools will improve the diagnosis and the genetic counselling of patients with PIDs which are important for the patients' support and care. Moreover, in the context of the technological advances in genomics, it is an exciting challenge to develop these new applications for the diagnosis and research of new PIDs. The ESID Genetics WP wants to be a link between clinicians, immunologists and scientists to help in the diagnosis of new PIDs and also to develop trials (i.e. on clinical and genetic correlation of specific PIDs). The Genetics working party closely collaborates with the other ESID working parties.

Please respond to the following questions:



How to participate?

Dear Colleagues,

We would like to be able to help direct our fellow ESID members when they have questions regarding laboratories that can be contacted for genetic testing of potential primary immunodeficiencies. In this regard, we would be grateful if you could answer the two questions below by return email (capucine.picard@inserm.fr):

- Do you perform genetic testing for primary immunodeficiency diseases?
- If yes, for which genes are you performing tests?

Thank you in advance for your rapid response.

Our best, Capucine PICARD for the Genetics WP of ESID

Education

ESID Summer School 2011

ESID SUMMER SCHOOL for Primary Immunodeficiency Diseases 28 September - 02 October, 2011, II Ciocco, Tuscany, Italy



Latest Update on ESID Summer School 2011

ANNOUNCEMENT FINAL SELECTION ESID Summer School

The ESID SUMMER SCHOOL for Primary Immunodeficiency Diseases will take place from September 28 to October 2, 2011, in the II Ciocco Hotel in Tuscany, Italy.

We had more than a hundred applications, and the selection process was a difficult one! It is good to see there are so many excellent young applicants interested in PID, but unfortunately, we can only accommodate 30.



The following participants were selected:

North-America: Christina Yee

Australia: Arihia M. Brewerton

Middle East: Dalia El-Ghoneimy

South-America:

Juan Aldave Becerra Saul O.L. Reyes Africa: Imen Ben Mustapha

Far East: Amit Rawat Deepti Suri

Eastern Europe: Jana Semberova Pavlina Kralickova Vera Goda Natalja Kurjane Alexis-Virgil Cochino Western Europe:

Laia Alsin Mike Recher Elisangela Santos-Valente Maria Raptaki Alessandra Magnani Stephanie Humblet-Baron Claudia Wehr Margje Haverkamp Lisette van de Corput Clementien Vermont Joao Neves Joana Soares Consuelo Anzilotti Helene Martini Marina Vignoli Francesca Ferrua Christian Nielsen

ANNOUNCEMENT FINAL SELECTION ESID Grants/Awards

We also had quite a few applications for the ESID fellowship grants, and - again - a difficult selection process.

This year the ESID Awards and Grants will go to

SHORT term:

Ana Merkler (Croatia) Rofida Nofal (Egypt)

MEDIUM term:

Maria Castiello (Italy) for her project on Characterization of the role of B cells in the pathogenesis of the Wiskott Aldrich Syndrome

LONG term:

Hannelore Bax (The Netherlands) for her project on Phenotypic correction of immunodeficiencies through modification or degradation of transcripts

You will read more about the ESID Summer School in future issues of the ESID Newsletter, and, of course, the fellowship grant awardees will write about the results of their project next year!

Best regards, Esther de Vries Chairperson Educational Working Party

2011 Summer School

The ESID Educational Working Party is pleased to announce the 2011 Summer School for Primary Immunodeficiency Diseases!

This event, which has received very positive feedback from many participants in the past who found the course invaluable, will run from September, 28, to October, 2, 2011, in Il Ciocco, Tuscany, Italy!

Faculty



For five days a well known international Faculty will be leading case-based discussions and give lecture presentations on primary immunodeficiencies. Educational Working Party Chairperson Esther de Vries and Marta Rizzi as the ESID Junior representative, together with Andrew Cant, Helen Chapel, Jacques van Dongen, Steve Holland, Georg Hollander, Eleonora Gambineri, and Klaus Warnatz will be some of the names appearing in this year's Summer School faculty.

Accelerate Your Career

If you are working on a career in primary immunodeficiencies, whether in the clinic, the lab, or a combination, this will be an excellent opportunity to update your knowledge from teachers at the forefront of PIDs, hear about the latest diagnostic tools, and learn from leaders in the field who see and manage patients with the whole spectrum of primary immunodeficiency, while also networking and socialising with your colleagues.

A little scoop...

The detailed Summer School programme will be finalized at a later stage but as ESID member you will receive web mails with updates and more detailed information on a regular basis.

Location—II Ciocco

"There's a place in Tuscany where you can lose yourself in the open spaces, walk in the woods and run over green meadows while our mountain watches over you!" - Il Ciocco is set in a natural, 2000 hectare park in the Serchio Valley near Barga/Lucca and offers exclusive meeting and leisure facilities in a naturally and culturally rich environment.

Application Procedure

Each year the Society selects between 30 and 35 ESID junior members based on their background and interest in PID. For successful applicants ESID will cover accommodation, meals and onsite transfers. Incoming travel and personal expenses are not covered. The online application procedure is fully automated; applications in any other form are not accepted!

What we need from you is:

- Be a current ESID junior member (Attention: age will not be an absolute limit)
- If you are not an ESID member, you can become one! Kindly click here)
- Curriculum Vitae and a list of publications and/or meeting presentations
- Letter of support from your Head of Department or similar
- Statement of career goals (typewritten, max. 500 words) and why attendance at this meeting will benefit your career now and in the future
- Case of PID (or suspected PID) that is educational and not just "tricky" (max. 3 PowerPoint slides)!

APPLICATIONS CLOSED

RESULTS WILL BE ANNOUNCED by JUNE 30th

For questions, write us at: esid.admin@kenes.com

Venue Official Website

Factsheet - ESID Summer School 2011 (118k)

Feature about Barga, Italy







Officially Sponsored by



Juniors

LAUNCH - Map of European Centers- NEW!!!!

Dear ESID Juniors,

the summer is approaching and it's a good time to relax, take sometime and plan for the activities of the near future.

To help the planning of our career development we launch in the beginning of July, in the Juniors' section of our ESID website, available only for members, the already announced <u>MAP of EUROPEAN CENTERS</u> available for short-middle and long- term exchange.



Take some time to navigate through it, you can click on the map and look the information of a center in a precise area of Europe, or you can use the filter function and select for example for your preferred internship in length. Or you can simply scroll over the whole list of centers and see if you can get inspiration for a short (up to 1 month), middle (up to 6 months) or long (up to 1 year) visit in one of the centers that are scattered all over Europe.



For financing the visit, you might use the fellowship available from ESID (the second call for the fellowship will be November 15th 2011), you can check if the chosen center has available funding, or you can look up for your own funding. The map of centers is a work in progress, you might want to list your center too in the website, in that case you can **download the questionnaire below, fill it up and send it to esid.admin@kenes.com**,and we will add the info into the map.

I really hope this tool will help ESID Juniors and ESID members to improve their experience and expand their knowledge, and the research and clinical network. I have to acknowledge the great contribution of each center, and of the juniors supporting this initiative, with a special mention for Ales Janda, his contribution has been fundamental for the starting of the project, and I have to acknowledge also the professional help of our web-masters and of Susanne Schmitt.

Map of Centres - Questionnaire (64k)

During the past May and June, we had both very successful and appreciated meeting for Juniors, the Prague spring meeting, at its 10th anniversary, and the Tampere meeting, you can read in this newsletter the report over this two events. We want to congratulate Anna Sediva and Crina Samarghitean for the great success of their meeting and thank both of them for the engagement for the ESID Juniors.

At the end of September is waiting for 35 selected Juniors the great experience of the Summer school, this year in Italy. I am looking forward to meet and discuss with all of you!

I wish you all a great summer!

Marta Chair of ESID Junior WP

Young Researcher's Corner

Kindly contributed by Brigida Immacolata

ELISPOT, a new tool for research and diagnostic

Dear all, this is my turn to give a contribution to the Young Research Corner and the topic is: ELISPOT, a new tool for research and diagnostic.

The Enzyme-linked immunosorbent spot (ELISPOT) assay was developed by Cecil Czerkinsky in 1983, as a method to monitor immune responses in humans and animals. Similarly to an ELISA immunoassay, it was originally used to count antibody-specific secreting B cells and then to also identify cytokine-producing cells at the single cell level.

Moreover, it allows the detection of low frequencies of cytokine secreting cells. This assay provides evidence of the secretory product of individual activated or responding cells. During the assay, each developing spot represents a single reactive cell that reacts to the substrate and the final number of spots per well provides evidence of how many antigen-specific cells are present in the tested cell population. Thus, the ELISPOT assay provides both qualitative (type of immune protein) and quantitative (number of responding cells) information. The good resolution of the number of secreting cells allows the capacity to test also specific cell subsets, often present at lower percentages among the general population.

The ELISPOT assay is more sensitive than tetramers/pentamers stainings or intracytoplasmic stainings for detectable cytokines by cytofluorimetry in which one can have only the idea of how much the specific cytokine is produced by or expressed in a specific cell population, even this population is rare (i.e. antigen-



specific response). To over-come the problems of release of the cell product, its dilution in the supernatant and its degradation, ELISPOT assays took advantage on the use of nitrocellulose or polyvinylidene fluoride (PVDF) membranes to capture and identify spots formed by cytokine secreting cells, which directly sit on the membrane. Thus, the cytokine is captured around the secreting cell as it is released where the local concentration of the cytokine is high, before dilution of the cytokine in the supernatant. Moreover, the detection of spots required a short incubation time with the substrate. This makes ELISPOT assays much more sensitive than conventional ELISA measurements.

To detect antibody/cytokine-specific cells, the plates are coated with monoclonal (more specificity) or polyclonal capture antibodies, chosen for their specificity for the analyte to detect. After blocking of aspecific bounds with a serum protein that is non-reactive with any of the antibodies in the assay, the cells of interest are plated at different numbers, along with antigen or mitogen and then placed into incubator for a specified time period. Then, the coated antibody on the membrane captures cytokine/Ig or other cell products secreted by activated cells locally. After washing to remove cells, debris, and media components, a biotinylated polyclonal antibody specific for the compound that will be detected and reactive to a distinct epitope of the target cytokine/Ig is added to the wells and thus is used to detect the captured cytokine/Ig. After washing to remove the excess of un-bound antibody, the detected analyte is visualized with a streptavidin-HRP and a precipitating substrate, revealing a coloured spot (usually a blackish blue) that represents an individual cytokine/Ig-producing cell.

The spots can be counted with an automated reader to capture the microwell images and to analyze spot number and size.

Applications in T or B cells studies offer the possibility to measure the magnitude and the quality of cell immunity at single cell level by detecting individual events of Ag-specific T cells that engage the secretion of cytokines and effector molecules such as GZ-B and or perforin or specific Ig-secreting plasmablasts. Compared to intracytoplasmic stainings, the ELISPOT assay provides information of the frequency and the effector function of T or B cells. Moreover it is possible to use both freshly isolated cells like PBMC or thawed cells, with the final goal to detect how many cells produce a determined compound. Furthermore, it is possible to use the same live cells for other purpose (i.e. FACS staining, cell culture, freezing).

This offers the possibility to test the same cells for different assays, representing a good option for diseases in which the cell number is a critical limiting factor or for patients with a seriously compromised immune system (i.e. HIV or CVID patients).

By Immacolata Brigida

References

- "A solid-phase enzyme-linked immunospot (ELISPOT) assay for enumeration of specific antibodysecreting cells." Czerkinsky CC, Nilsson LA, Nygren H, Ouchterlony O, Tarkowski A. J Immunol Methods. 1983 Dec 16;65(1-2):109-21.
- "Comparison of a limiting dilution assay and ELISpot for detection of memory B-cells before and after immunisation with a protein-polysaccharide conjugate vaccine in children." Blanchard-Rohner G, Galli G, Clutterbuck EA, Pollard AJ. J Immunol Methods. 2010 Jun 30;358(1-2):46-55.
- "Upregulation of IL-21 Receptor on B Cells and IL-21 Secretion Distinguishes Novel 2009 H1N1 Vaccine Responders from Nonresponders among HIV-Infected Persons on Combination Antiretroviral Therapy." Pallikkuth S, Pilakka Kanthikeel S, Silva SY, Fischl M, Pahwa R, Pahwa S. J Immunol. 2011 Jun 1;186(11):6173-81.



"Antibody forming cells and plasmablasts in peripheral blood in CVID patients after vaccination." Chovancova Z, Vlkova M, Litzman J, Lokaj J, Thon V. Vaccine. 2011 May 31;29(24):4142-50.

"Anergic responses characterize a large fraction of human autoreactive naive B cells expressing low levels of surface IgM." Quách TD, Manjarrez-Orduño N, Adlowitz DG, Silver L, Yang H, Wei C, Milner EC, Sanz I. J Immunol. 2011 Apr 15;186(8):4640-8.

PID Care in Development

Spring period of each year is particularly important for increasing awareness about primary immunodeficiencies. April 29th is traditionally proclaimed Day of Immunology.

The purpose of Day of Immunology, in 2011organized for 7th time already, is to raise awareness amongst the public, the press, politicians and decision makers about the critical importance of the immune system in everybody's everyday life. To further specify all these important aspects of awareness campaign in the field of primary immunodeficiencies, the new concept of PID awareness week was raised for the first time in 2011 and ESID was among the founders of the event! The first ever PID week included a number of important activities, for further information <u>click here</u>.

The same site provides also a link to very useful materials that can be used in any country for further improvement of PID awareness, diagnosis, care and management. PID week is open to everybody and active participation of ESID members would further foster its significance.

One of the registered events of 2011 PID week was also ESID Prague Spring Meeting (report see attachment), held in Prague May 9 and 10, 2011. ESID Prague Spring Meeting traditionally contributes to an increase of knowledge and awareness about PIDs particularly among ESID juniors. Its scope perfectly fits the goals of PID awareness week. ESID PIDCD WP has PID awareness week among its priorities and would notify ESID members ahead the event next year, to give everybody the opportunity to participate and contribute to better complex management of PIDs in Europe.

ESID PIDCD calls for active participation in similar activities and would appreciate news about such events for their central ESID registration.

Continuous effort to increase of awareness of PID in Central and Eastern Europe by J project series of meetings went on this year with meetings in Russia and Ukraine, for more information<u>click here</u>.

As always, we welcome any comments, suggestions and short reports from locally organized activities to be published in ESID Newsletter, to share the knowledge and further help PIDs patients.

For ESID PIDCD Anna Sediva Prague, May 20, 2011



10th ESID Prague Spring Meeting Report

Department of Immunology, 2nd Medical School, Charles University, University Hospital Motol, Prague, Czech Republic

The tenth ESID Prague Spring Meeting was organized at the University Hospital Motol, Prague, Czech Republic, on May 9 and 10, 2011. The meeting was attended by 39 participants from 12 countries, majority of them from Czech Republic, further from Austria, Bulgaria, Egypt, France, Germany, Hungary, the Netherlands, Romania, Slovakia, Spain, and United Kingdom. This anniversary meeting has further strengthened very active participation of ESID juniors on this event.

Since its launch in 2002 the Prague ESID meeting has been devoted to the exchange of information on primary immunodeficiencies (PIDs) between Western and Central and Eastern Europe. Continuous and successful transfer of knowledge from West to East was clearly demonstrated during the meeting, with steadily rising quality of scientific presentations particularly by Eastern European participants.

The meeting was very nicely supported by excellent contributions of invited speakers who encouraged very vivid discussions during all scientific sessions.

This year's main topic, CVID, was covered by Helen Chapel from UK. Helen Chapel was the initiator and founder of this series of meetings and was invited specifically for 10th anniversary meeting as a distinguished guest. Andrew Cant, also from UK, contributed greatly to the friendly atmosphere of entire event. Kaan Boztug from Austria brought top level science to the meeting and in the same time offered working and PhD positions. Such opportunities to find PhD students or to get PhD positions become gradually one of the most valuable benefits of ESID Prague Spring Meetings.

Social program was very carefully planned on the occasion of 10th anniversary meeting. Short stroll through Prague, followed by Czech dinner and later by a visit to night Zoo underlined junior character of the meeting and was appreciated by all participants. Students of Academy of Fine Arts in Prague also contributed to the festive character of anniversary event by designing graphics specifically for Prague Spring Meeting participants. Graphic was given to everybody as a reminder of the decade of Prague events.

The meeting was traditionally organized as a part of the activities related to the Day of Immunology, declared by EFIS on 29th April, 2011 and to PI week, for the first time organized in 2011.

ESID Prague Spring Meeting was also held on the occasion of opening of Jeffrey Modell Diagnostic and Therapeutic Center for Primary Immunodeficiencies, awarded to Prague center in 2011. The meeting was supported by the Charles University, 2nd Medical School, Prague and by University Hospital Motol, Prague. Substantial contributions came from the traditional general sponsors – Olympus, from Roche and Pražská Plynárenská a.s., and from companies Baxter, Exbio, Grifols, Novartis, Scintila, Synlab and Shire, in alphabetical order, and from Czech Immunology Society.

We thank all the participants for their contributions and we are looking forward to the next meetings in Prague.

Anna Šedivá, Andrea Polouèková Prague, May 13, 2011



Report - ESID Juniors Symposium/Immunology Day 2011

16-17 June, 2011, Tampere, Finland by Dr. Crina Samarghitean



Conference Venue: FinMedi 5, Auditorium, Institute of Biomedical Technology, University of Tampere

On behalf of European Society of Immunodeficiencies (ESID) Juniors Working Party and Bioinformatics group, Institute of Biomedical Technology, University of Tampere we organized for the first time in Finland, ESID Juniors Symposium/Immunology Day 2011.

The program followed a special twist this year. During two days in FinMedi 5, Auditorium, Institute of Biomedical Technology, we could assist to state of the art lectures, hot topics in primary immunodeficiencies (PID), demo sessions, pharma exhibition, a PIDart exhibition and possibility to meet with other IT, biotech, pharma and healthcare opinion leaders. We had lively discussions and interactions between clinicians, lab scientists and computer scientists. More than 60 participants from 16 countries, mainly from Finland, further from United Kingdom, Hungary, Italy, Slovenia, Slovakia, Norway, Sweden, Netherlands, Belarus, Belgium, Switzerland, Poland, Macedonia, Kosovo were registered to our symposium. We received more than 20 abstracts and 21 PIDart entries for PIDart exhibition.

Many new findings have been presented, and new possibilities for treatment in the near future as well as developments with impact that lay further ahead were lively discussed. The core of this symposium was the question how to bridge the gap between clinical, laboratory and computer aided diagnosis in PIDs as well as how to develop national PID patient registries and make the most of it. The new chair of Finnish patient organization, Riina Riihiniitty, updated us with the latest changes in their organization and made an open call for further collaborations with all the parties interested.

We were very pleased that so many top scientists and clinicians at the front of PIDs agreed to speak during this symposium and share their knowledge and views with us and each other. In the first day we had the chance to listen an overview on the latest developments in flow cytometry by Dr. Mirjam van der Burg, Netherlands, followed by lively discussions around diagnostic protocols in primary immunodeficiencies, covered by Dr. Esther de Vries, MD, PhD, Netherlands. Prof. Mauno Vihinen presented the latest developments in bioinformatics tools in PIDs, followed by a lecture on decision support systems and PIDexpert covered by Assist. Prof. Kati Iltanen and Dr. Crina



Samarghitean, Finland.

The second day started with an outstanding lecture on Latest biotherapies on PIDs and neonatal screening covered by Prof. Lennart Hammarström, Karolinska Institute, Stockholm,Sweden. The session on biobanks and PID registries were covered by Dr. Tiina Laatikainen, from National Institute for Health and Welfare, Department of Chronic Disease Prevention, Helsinki, Finland and Dr. Annarosa Soresina, Brescia Children's Hospital, Italy. We hope that the success story of IPINet will serve as a model for the Finnish PID registry, which might be integrated within other national registries and biobanks of Finland and abroad.

The outstanding keynote lecture: Primary immunodeficiencies 1981-2011: An evolving discipline, covered by Prof. Laszlo Marodi, Medical and Health Science Center, Department of Infectious and Pediatric Immunology, University of Debrecen, Hungary brought not only a lot of knowledge but also enthusiasm and passion for PID for the new generation.

We were also very pleased that we were able to offer a platform for the younger generation of scientists/clinicians in this field with a number of selected orals and the poster sessions. A number of travel awards kindly provided by European Society of Immunodeficiencies (ESID), European Federation of Immunodeficiencies (EFIS)/European Journal of Immunology (EJI) allowed to reinforce the juniors network, mobility and collaborations between different countries of Europe.

This year the winners of those three travel ESID awards went to: Ning Wang, Sweden, Ceri Roberts, United Kingdom and Linda Veghova, Slovakia. The six travel EFIS/EJI travel awards went to: Maaike Kusters, Netherlands, Larisa Fedarushchanka, Belarus, Gaspar Markejl, Slovenia, Nicola Read, United Kingdom, Arriana Troilo, Italy and Peder Ydalus, Norway. All these winners have been officially congratulated in the award ceremony and received also some books, kindly provided by CSL Behring. These books will help them further in the management of PID patients.

We also wanted to stimulate excellence in research and clinical practice and this year the 'Best poster award' went to Ning Wang from Karolinska Institutet, Stockholm, Sweden for 'IgA deficiency in autoimmune diseases' poster. The 'Best presentation award' went to Nicola Read, Brighton and Sussex Medical School, Brighton, UK, for 'Paediatric Hereditary Angiooedema: Variations in management within the UK and impact of the disease'. We congratulate again the winners and thank to our sponsor Sanquin for this session. We also wanted to stimulate the use of decision support tools, and a 'special prize for the most clinical cases submitted in electronic format', kindly provided by Baxter, Finland branch, was awarded to Ceri A Roberts, Oxford Radcliffe Hospitals NHS Trust, United Kingdom.

The PIDart exhibition/contest 'Visionary PIDs' received a lot of interest and entries this year too not only from clinicians and lab people but also from computer scientists and even PID patients. The best three PIDartworks 2011 were: 'Hepatosplenomegaly' by Larisa Fedarushchanka, Belarusian Medical Academy of Post-Graduate Education, Minsk, Belarus; Scientific Impressionism II, by Nicholas Hubbart image kindly provided by Prof. Hans Ochs, Seattle Children's Reseearch Institute, USA; and Cell locomotion (a demo cell simulation), by Peder Ydalus, , University of Oslo, Norway. Between the favorites were also, Immunoglobulins on flower impressions and Spiral of my life before and after Immunology, by Anonymous adapted by Crina Samarghitean, Finland and Bright immune interaction, by Dr. Maaike Kusters, Jeroen Bosch Hospital, the Netherlands.

The social program was also very interactive and dynamic and well appreciated by the participants. We continued our 'bright interactions' in a Viking environment, enchanted by the trills of the Finnish folk music, birds songs, performed by Paula Siren and Adina Dumitrescu and a well known troubadour of Argentinean tango, Martin Alvarado.

We demonstrated successfully that it is possible to have also brilliant lectures, enthusiastic clinicians/lab scientists/computer scientists under the same roof with artists and be able to keep the ESID/EFIS Juniors spirit alive and celebrate in a new way Immunology Day 2011.

We thank once again to all the participants, organizers and our golden sponsors: CSL Behring, Baxter, Sanquin



and DAF and our supporters: EFIS/EJI, ESID, OrphanEurope, FlowJo and BD Biosciences who made all these possible. Thank you for your great contribution!

On behalf of the organisers Crina Samarghitean MD, MSc, PhDc Chair of ESID Juniors Symposium/Immunology Day 2011 Chair ESID Juniors WP (2008-2010)

PedPAD study

We would like to invite you to participate in the PedPAD Study concerning **Clinical and laboratory characteristics** of children with hypogammaglobulinemia as documented in the ESID online Registry.

Most of the currently published clinical data on hypogammaglobulinemia concern adult patients; because of their maturing immune system, these data cannot simply be extrapolated to children. An overview of pediatric data will be helpful for doctors caring for these patients, and may form the basis for further clinical studies. We want to use the data collected in the ESID Registry to give an overview of all clinical, laboratory and geographical features of children with hypogammaglobulinemia, and describe possible diagnostic delay. Please let us know whether you want to participate with the data from your centre. Please download the invitation letter and the study protocol for more information. Dr. Esther de Vries June 2011

PedPAD Invitation letter (51k) PedPAD research protocol (347k)

DOCK8 Deficiency Therapy Survey

On behalf of our working group we kindly invite you to join our survey.

Online submission via the ESID Database is now open!

The data forms are available both in the subregistry DOCK8 (Combined ID) and Hyper IgE syndrome (HIES).

In addition, submission on paper is also available.

Please find more information in the invitation letter.

We are looking forward to your help!

- The DOCK8 Working Group

Mail contact: Ellen.Renner@med.uni-muenchen.de

Version 2011-03-30 POCK 8 Case Report Form (40k) Version 2011-12-21