IPOPI
First Indonesian PID Patients-Doctors National Meeting
Held at the Grand Sahid Jaya Hotel,
Jakarta, Indonesia
9.00-15.30hrs
2nd May 2015

Report

1. Introduction

The first IPOPI Indonesian PID Patients-Doctors National Meeting took place at the Grand Sahid Jaya Hotel on Saturday 2nd May 2015. The meeting was organised in collaboration with Cipto Mangunkusumo Hospital (CMH) and the Faculty of Medicine Universitas Indonesia (FMUI). The meeting was held as part of the celebrations for the 2015 World Primary Immunodeficiencies Week (WPIW). A visit had been held prior to the meeting earlier in the week at Cipto Mangunkusumo Hospital where Prof. Van Hagen, member of IPOPI's Medical Advisory Panel (MAP), delivered a guest lecture on primary immunodeficiencies diagnosis and treatment to the doctors and PhD students.

The meeting brought together 55 participants from various regions in Indonesia including patients, family members, Indonesian doctors and guest lecturers from The Netherlands and Malaysia.

The meeting was opened by Dr Dina Muktiarti (CMH) and Mrs Jose Drabwell (IPOPI President). Special thanks were given to all representatives from the various regions of Indonesia, patients and their families, and IPOPI's MAP members: Prof Van Hagen (The Netherlands) and Dr Adli Ali (Malaysia). Mrs Drabwell extended IPOPI's thanks to Dr Dina Muktiarti and Dr Nina Dwi Putri without whose help the meeting would not have been organised so efficiently. She also highlighted the need to have both children and adult immunologists in the future and more paediatricians in Indonesia. Lastly she thanked Octapharma for their support to the meeting. A formal welcome address followed by the Director of CMH and the Dean of the Faculty of Medicine Universitas Indonesia (FMUI).

2. Scientific session: presentations

The meeting started with three scientific presentations. Prof Van Hagen (IPOPI MAP Member, Netherlands and Thailand) commenced the session with a presentation on "The immune system and PIDs" designed to provide an overview to the patients and families in attendance. The presentation described the basic function and various components of the innate immune system and the adaptive immune system. The function of cytokines, the varying degree of the immune 'repertoire' in different people, and interestingly the links between the immune system, hormones and the nervous system were explored. Prof Van Hagen also provided examples on specific issues encountered in the South East Asian region such as patients producing antibodies against interferon gamma cytokynes leading to microbacterial infections. Importantly he highlighted the essential interaction that should take place between paediatric specialists, adult immunologists and specialists and laboratories to ensure appropriate diagnosis and management of PID patients.

Dr Adli Ali (IPOPI MAP Member, Malaysia) then followed with a most interesting presentation reviewing the history of PIDs in South East Asia (SEA) from the first diagnosis made in the 1970s in Malaysia, to the first use of immunoglobulin therapies (early 1980's) and bone marrow transplants (1996, Singapore) to treat PIDs. The presentation then concentrated on the recently published *IPOPI PID Principles of Care* identifying gaps in the diagnosis and care framework for PIDs in SEA. Based on SEAPID national registries it is estimated there are 703 identified patients in the SEA region when based on a prevalence of 1:1,200 (Boyle et al) it should be 533,333 patients. He stressed the importance of having efficient registries and reliable databases put in place in SEA countries. International collaborations, work with IPOPI's national patient organisations in SEA and the role of SEAPID the newly formed SEA PID doctors group were among other highlighted key aspects to improve PID care in the region.

Dr Nia Kurniati (Universitas Indonesia/Cipto Mangunkusumo Hospital) followed with a presentation outlining PID care in Indonesia. She explored several case study examples and went on to highlight different diagnostic pathways, from detection of PIDs based on clinical exams, to basic laboratory tests and the need for advance laboratory tests and initially collaboration with Europe and the USA to get them set up. Typical PID warning signs were reviewed and it was noted that a specific Indonesian definition of such signs is needed. The clinical manifestations of PIDs linked to specific cell function were also presented. The results of a survey on laboratory facilities carried out in different hospitals in Indonesia were then summarized and an overview of the PID patients from Cipto Mangunkusumo Hospital and allergy-immunology specialists in Indonesia was also provided.

3. Patients session: Presentations

The patients session first featured a presentation by Mr Johan Prévot (IPOPI Executive Director) who provided an overview of IPOPI's activities and support programmes available to help NMOs with their national campaigns and actions. He stressed the great milestones recently achieved in South East Asia through the IPOPI Bob Le Bien Asian development programme with the launch of new NMOs in Malaysia, Thailand, Hong Kong, and more recently Singapore where a new patient group was recently formed. The creation of SEAPID at the meeting which IPOPI co-organized with the Care-for-Rare Foundation in Bangkok in early 2015 was highlighted as a key development for the region. The presentation was followed by a PID patient panel session in which Ghifary Muhammed, a young patient living with agammaglobulinaemia, his mother and the mother of Abel Mehola who lives with Hyper IgE syndrome provided their testimonies on living with a PID. Ghifary was born in good health but rapidly started to develop recurring infections. Later in life these episodes affected his school life resulting in many weeks and months of absence until he was referred to Cipto Mangunkusumo Hospital where finally he was diagnosed with PID. At first his condition improved after getting on immunoglobulin replacement therapy however his situation worsened as soon as treatment was stopped due to the fact his family could not afford covering the costs. Fortunately the costs of his treatment are now covered and Ghifary is doing much better. The mother of Abel then explained to the audience the struggle her family has gone through going from doctor to doctor and from misdiagnosis to misdiagnosis. At one point, the family was told that their son had HIV and later on leukaemia, until finally a PID diagnosis was made: Hyper IgE syndrome. The patient stories demonstrated the importance of having a patient organisation and increasing awareness of PIDs in Indonesia to improve diagnosis rates and provide appropriate treatments to patients in need. Mr Agustaman F Wirawan and his colleague Ms. Novi Riandini followed with a presentation outlining their experience and work with the Indonesian Haemophilia Society. They described having a rare disorder such as PID and Haemophilia without a National Patient Organisation as having "a child without parents". In their presentation they went over the history of the Haemophilia Society from its start as an informal group of parents and patients to a formal national group now member of the World

Federation of Haemophilia. The benefits which the Haemophilia patient group has brought were highlighted, from increasing diagnosis rates, to better access to treatment and greater public awareness. This experience further demonstrated the need for a PID patient organisation in Indonesia.

4. Discussion, Facts and Key Priorities

The presentations and panel session were followed by a discussion. Several facts and issues were identified during the meeting:

- There are 250 million inhabitants in Indonesia
- There are 3000 paediatricians for the whole country
- There are 27 allergy-immunology consultants for the whole country
- There are 55 known PID Patients in Indonesia (Note: *at the start of the meeting this number was thought to be 15. However through information sharing during the meeting between doctors from different regions, 55 were identified*)
- 45 of those PID patients are managed through paediatrics units
- 10 remaining PID patients are managed through internal medicine units
- Prevalence based on international figures (1:1,200) is likely to be 208,333 patients in Indonesia
- The most prevalent types of PIDs in Indonesia based on the report (n=15) provided at the meeting are:
    - Predominently antibody deficiencies (agammaglobulinaemia, IgA deficiency and transient hypoagammaglobulinaemia): 60%
    - Well defined syndromes (Wiskott Aldrich Syndrome, Hyper IgE): 20%
    - Combined B and T cell deficiencies (SCID): 6,66%
    - Autoinflammatory disorders (Blau syndrome): 6,66%
    - Awaiting definite diagnosis: 6,66%
- IVIG is not automatically reimbursed by the national healthcare system, only through some hospitals (ie . Cipto Mangunkusumo)
- SCIG is not available yet
- No BMTs for PIDs have been performed to date
- Lack of diagnosis facilities and expertise is a key issue to tackle
- Availability of laboratory facilities / tests in Indonesian hospitals overview based on local survey:

Complete blood count	*Available in all hospitals*
Immunoglobulin (IgG, IgA, IgM, IgE)	*Available in most hospitals*
Lymphocyte subset	*Available in most hospitals*
Complement	*Available in most hospitals*
Lymphocyte proliferation test	*Not Available*
Granulocyte function-NBT/DHR	*Not Available*
Pneumococcal antibody	*Only in 1 research lab*
Genetic testing	*Not Available*

- There is no national PID registry yet
- The Need for adult PID specialists was highlighted

☐ The importance of increasing PID awareness among doctors was highlighted

☐ The need for patients support was also identified

5. Conclusions

The meeting was the first of its kind in Indonesia, bringing together doctors and patients from different regions of the country. It provided a unique opportunity to meet together and discuss the importance of starting a patient organisation in Indonesia. The main outcome of the discussion was a unanimous agreement between the patients and the doctors that a national patient organisation should be launched as soon as possible. It was agreed that the first step should be to identify additional patients willing to participate in a future group through communication between the different centres and doctors reaching out to their patients and families who will then be invited to meet together with a view to launch an IPOPI Indonesian NMO. It was also agreed to form a network between the different centres to share diagnosis facilities and exchange specimens. IPOPI indicated they would be happy to facilitate a follow up meeting with additional patients and families.

The process to apply for IPOPI membership in the near future was reviewed. It was agreed that IPOPI would stay in close contact to guide this process and ensure that an Indonesian NMO is launched in the near future. IPOPI looks forward to further supporting this objective, which will benefit the whole PID community in the country.

The meeting was organised with the invaluable support of:

octapharma
For the safe and optimal use of human proteins

And

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