

Asia Pacific Society for Immunodeficiencies

Inaugural APSID Scientific Congress cum Hong Kong Spring School
27 April – 1 May 2016 • Hong Kong

REPORT ON CONGRESS HIGHLIGHTS



TOLERANCE TO BALANCE...
CREATIVITY FROM DIVERSITY



From Conception to Delivery: The Birth of APSID

The Asia Pacific Society for Immunodeficiencies (APSID) Inaugural Scientific Congress was held in Hong Kong, 28th April – 1st May 2016. The idea of establishing APSID was conceived in Osaka, Japan in April 2015, when over 60 Asian paediatricians and scientists met and discussed the visions and missions of APSID. With the dedications, passion and determinations of leading PID clinicians and scientists in Asia, APSID was inaugurated on 29th April 2016 in the historical Béthanie Campus, Pokfulam.

The 4 Key Missions of APSID are:

1. To care and cure patients with PID
2. To share our PID experience so as to promote collaboration & education
1. To improve PID management through understanding its genetics & pathogenesis
2. To advocate and advance the care of PID patients through engaging governments, patient organizations & industry

The Inauguration Ceremony was graced by the presence of Prof Andrew Cant, President of the European Society for Immunodeficiencies (ESID); Prof Luigi Notarangelo, immediate Past President of the Clinical Immunology Society, USA; Prof Hans Ochs, adviser of APSID since the time of conception in 2015; and Mrs Jose Drabwell, President of the International Patient Organization for Primary Immunodeficiencies (IPOPI), together with the Overseas Organizing Committee and over 150 participants of the APSID Congress.



A moment of joy: Yu-Lung Lau, President of APSID, officiating the Inauguration Ceremony with the Overseas Organizing Committee – Luigi Notarangelo (USA), Hans Ochs (USA), Andrew Cant (UK), Amos Etzioni (Israel), Xiqiang Yang (mainland China), Xiaodong Zhao (Mainland China), Kohsuke Imai (Japan), Surgit Singh (India), Amir Hamzah Abdul Latiff (Malaysia), Asghar Aghamohammadi (Iran), Nima Rezaei (Iran), Le Thi Minh Huong (Vietnam), Wen-I Lee (Taiwan), Woei-Kang Liew (Singapore), Hoque Mahbulul (Bangladesh) and Kamchaisarian Wasu (Thailand)



APSID Inaugural Congress Opening



The opening of the APSID Congress began with **Prof Yu-Lung Lau's** Opening Speech 'The Dream' – the collective dream of APSID members to propel PID care, education and research forward for the benefit of our patients with PID, through the collaborative infrastructure and the various Working Parties of APSID to implement the dreams. This was followed by the sharing of the History and Development of ESID and CIS, by **Prof Andrew Cant** and **Prof Luigi Notarangelo**, respectively. Prof Cant emphasized the important roles of the ESID Working Parties which are key to the growing impact of ESID on PID care, education and research. Summer Schools (or Spring and Winter Schools) are highly successful educational activities organized by both ESID and CIS in promoting the development of the next generation of clinicians and scientists in PID. Partnership of APSID with ESID, CIS and other professional societies including LASID, ASID and the J Project will form a global network of PID clinicians and researchers.

Mrs Jose Drabwell, President of IPOPI, reported the exciting developments in PID patient organizations in Southeast Asia, and the wonderful work that has been done to improve PID awareness in Asia. In Hong Kong, the PID patient advocacy group PID League was established in 2015. The journey was narrated by **Mr Geoffrey Yu**, Vice-President of PID League. Since Jose's first visit to Hong Kong in December 2013, PID League has been maintaining close contacts with IPOPI and participated in the IPOPI meeting during the 2014 ESID Biennial Conference in Prague. Locally, PID League actively promotes PID awareness and advocacy to medical professionals and the general public.



Patient Care & Research Frontiers in PID

The APSID Plenary Lecture was delivered by **Prof Luigi Notarangelo**, on the use of induced pluripotent stem cells (iPS) in PID care and research. Using patient-derived iPS, Prof Notarangelo's lab investigates the pathogenetic mechanisms of PID such as herpes simplex encephalitis in TLR3 signaling defects, bone marrow failure associated with genetic defects of the non-homologous end joining pathway, and faulty myeloid differentiation in reticular dysgenesis. In addition, patient-derived iPS serves as excellent tools to investigate efficacy and safety of novel therapeutic approach such as gene editing, with great promise in translational applications .



In another lecture, Prof Notarangelo updated us on the current status of newborn screening for SCID based on enumeration of T-cell receptor excision circle (TREC) in the USA. Babies with SCID identified through newborn screening have superior survival as appropriate protective measures can be provided in the pre-symptomatic stage while expediting haematopoietic stem cell transplantation (HSCT) which is the definitive curative procedure.

Graft-versus-host disease (GVHD) is a major cause of morbidity and mortality associated with HSCT. **Prof Wenwei Tu** (Hong Kong) gave a talk on regulatory T-cell therapy in HSCT. Using a humanized mouse model, his research group demonstrated the efficacy of ex vivo generated human antigen-specific Treg in controlling acute GVHD without compromising general immunity and graft-versus-tumor activity, showing promise for clinical applications.

Dr Wanling Yang (Hong Kong) reported the applications of whole exome sequencing (WES) technology in identifying rare PID genetic defects in patients referred to the Asian PID (APID) Network based in The University of Hong Kong. WES has facilitated the identification of PID which were rarely described in Asians, diagnosing PID presenting with atypical clinical and immunophenotype, as well as discovering novel PID genetic defects within the APID Network.

Pathogenetic Mechanisms of PID: Autoimmunity, Inflammation & Immunodysregulation

This symposium focused on the spectrum of PIDs associated with autoimmune and inflammatory manifestations. **Prof Hans Ochs** explained the molecular basis of autoimmunity in Wiskott-Aldrich syndrome, CD40L deficiency, ALPS, IPEX and other IPEX-like disorders such as STAT1 and STAT3 gain-of-function mutations, CTLA4 deficiency and LRBA deficiency. The recognition of the underlying genetic defects has important implications for specific therapies, including immunosuppression, kinase inhibitors, specific agonists (e.g. CTLA4-Ig) or HSCT.

An overview of autoinflammatory syndromes – From CAPS to HIDS, FMF, Blau and so on, was given by **Prof Yu-Lung Lau**. Hereditary autoinflammatory syndromes are characterized by systemic and organ-specific inflammation due to single gene defects in the innate immune pathways. Therapeutic options including corticosteroids, methotrexate, biologics such as anti-IL1 therapy and HSCT were discussed.

Inflammatory manifestations in two specific forms of PID, namely leukocyte adhesion deficiency (LAD) and sarcoidosis in granulomatous common variable immunodeficiency (CVID), were presented by **Prof Amos Etzioni** (Israel) and **Prof Martin van Hagen** (Netherlands). Taken together, this Symposium highlighted immunodysregulation as a consequence of genetic defect in the immune system, accounting for clinical manifestations which could be presented to clinicians working in various clinical disciplines, including haematologists, rheumatologists, gastroenterologists and more.

Infections

The symposium on infections comprised Fungal Infections (**Dr Pamela Lee**, Hong Kong) and Mycobacterial Infections (**Dr Wen-I Lee**, Taiwan) as Indicators for PID. These two talks highlighted the susceptibility to *Pencillium marneffe* infections in patients with gain-of-function STAT1 mutations, and the range of PIDs predisposing to BCG disease which is an important issue as BCG is given universally to neonates in Asia where TB is endemic. These talks are followed by the lecture on the Use of Anti-fungal and Anti-mycobacterial Agents in PID by **Prof Pak-Leung Ho** (Hong Kong), a renowned microbiologist who provided the audience on practical aspects in managing these difficult-to-treat infections in immunocompromised patients. Finally, A Practical Guide on Vaccination in PID was given by **Prof Yu-Lung Lau**, pointing out when live vaccines are contraindicated, while emphasizing situations when vaccines could benefit patients with PID when used correctly and should be promoted.

Clinical Care and Therapy for PID

Genetic heterogeneity in CVID and hyper-IgM syndromes, and their implications on diagnostics and management, were discussed by **Prof Asghar Aghamohammadi** (Iran) and **Prof Kohsuke Imai** (Japan), respectively. The patient population studied by Prof Aghamohammadi, with high rate of consanguinity, revealed a significant proportion of CVID patients diagnosed with LRBA deficiency. On the other hand, Prof Imai identified over 20 patients with activated PI3 kinase delta syndrome (APDS) with CVID / hyper-IgM phenotype.

Prof Xiaodong Zhao (China) and **Prof Surjit Singh** (India) reported the recent achievements in PID development in mainland China and India, the two most populated countries in the world. There remains lots of challenges in improving the outlook of PID patients, including access to specialist care, diagnostics, therapies including immunoglobulin replacement and HSCT. Public health resources and health insurance policies are major issues to be tackled.

Prof Hirokazu Kanegane (Japan) discussed the current recommendations on immunoglobulin (Ig) replacement for PID patients, and discussed the benefits and drawbacks of intravenous (IV) and subcutaneous (SC) Ig. SCIG can be administered by the patients at home, is efficacious and offers a better quality of life. Patient's choice and lifestyle should be taken into consideration.

Prof Andrew Cant provided an update on the current practice on HSCT for PID. The availability of better HLA matching of unrelated donors and use of cord blood HSC, technology in manipulating mismatched HSC, reduced intensity conditioning and more effective treatment for GVHD have significantly improved post-HSCT survival, and opening up therapeutic options for a wider range of PID. Prof Cant emphasized that the success of HSCT for PID depends on teamwork, attention to detail, and the special skills and expertise that develop from these.

APSID General Assembly

The First APSID General Assembly was held on 30th April 2016. 142 delegates registered as Founding Members of APSID and participated in the General Assembly. The Constitution, which had been proposed and discussed at the 2015 Preparatory Meeting in Osaka, was approved. Prof Yu-Lung Lau, the Convenor of the APSID Preparatory Meeting in Osaka and the APSID Inaugural Congress, received unanimous support to be the President of APSID for a 4-year term. Together with the President, the following members constituted the **Executive Board**:



Secretary: Prof Surjit Singh
Treasurer: Prof Godfrey Chan
Chair, Cellular Therapy and Transplant Working Party: Prof Kohsuke Imai
Chair, Clinical Care Working Party: Prof Xiaodong Zhao
Chair, Education and Training Working Party: Prof Shigeaki Nonoyama
Chair, Genetics and Genomics Working Party: Dr Pamela Lee
Chair, Registry and Research Working Party: Dr Woei-Kang Liew



In addition, the APSID Juniors Working Party was formed to empower young PID trainees and fellows to be future leaders. The APSID Juniors are led by two enthusiastic PID trainees, **Dr Alric Mondragon** (Philippines) and **Dr Youjia Zhong** (Singapore) who participated in both the Hanoi Winter School and the Hong Kong Spring School. The APSID Juniors will be involved in organizing APSID Schools, and will be represented in the Executive Board.

Representing the APSID Juniors:
Dr Alric Mondragon and Dr Youjia Zhong delivering the vote of thanks at the Inauguration Dinner

The APSID Registry and Researching Working Party in Action

A symposium on 'PID Registry and Regional Network' was held on Day 2 of the APSID Congress. **Dr Nizar Mahlaoui**, Chair of the ESID Registry Working Party, introduced the history, structure and recent developments of the ESID Patient Registry. This was followed by talks on the current status of PID Registry in Japan, Iran,

Thailand and the Southeast Asia PID (SEAPID) by **Prof Kohsuke Imai**, **Prof Nima Rezaei**, **Dr Wasu Kamchaisatian** and **Dr Narissara Suratannon**. An interactive discussion with the audience was led by Dr Woei-Kang Liew, Chair of the APSID Registry Working Party.



Education and Training: The APSID Spring School



Following the success of the APSID Hanoi Winter School in Nov 2015, the APSID Spring School was held on 28-29th April before the Main Congress. The Spring School was attended by 51 enthusiastic trainees from 16 countries and regions, as well as over 50 APSID delegates who joined the Spring School as observers. 27 oral and 26 poster presentations were delivered by the trainees, covering a wide spectrum of PIDs with challenges in diagnosis and management.

The Spring School consisted of educational lectures delivered by the Faculty, including Approach to a Child with Neutropenia (**Prof Amos Etzioni**), SCID – Classification and diagnosis (**Prof Luigi Notarangelo**), Use of Flow Cytometry in PID Diagnosis (**Prof Hirokazu Kanegane**), Antibody Deficiency Syndromes (**Prof Hans Ochs**), Illustrated Guide to Infection in PID (**Prof Andrew Cant**), and ABC guide to HSCT – The Asian Perspectives (**Dr Pamela Lee**). Each lecture was followed by oral presentations given by the trainees, who were required to identify 3 key learning issues for discussion. Each trainee was assigned to an adviser (**Prof Reinhard Seger** [Switzerland], **Prof Bee-Wah Lee** [Singapore], **Dr Woei-Kang Liew** [Singapore], **Dr Jonie Santos-Ocampo** [Philippines], **Dr Hsin-Hui Yu** [Taiwan] and **Dr Narissara Suratannon** [Thailand]) who assisted them with preparing their presentations, so the learning process actually started well before the School. At the Spring School, the Faculty offered suggestions on further investigations for cases without a definitive diagnosis yet, and comments on practical management issues. The setting of the Spring School was conducive to collaborative learning, and the audience actively participated in the discussion.

The Spring School ended with a sharing session ‘From “Nothing” to “Something” – My Journey in PID Development’ by Prof Surjit Singh, Prof Bee-Wah Lee and Prof Yu-Lung Lau. Trainees were deeply impressed by their enthusiasm and passion, and their remarkable stories of how they built the foundation for PID care, research and education in their countries inspired and empowered the younger generation of doctors to strive for a better future for PID patients.



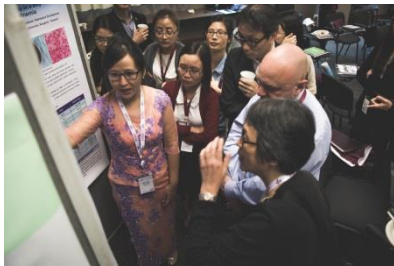
Comments from Spring School participants

Please state one or more aspect(s) which you find the APSID Spring School helpful to your clinical practice:



1. learn more knowledge about PID
2. Meet many friends in immunological field
3. Discuss my case with professors and know what to do next
4. I want to join APSID and do something useful

Trainees from Bangladesh, India, Cambodia and Indonesia receiving their certificates from Prof Hans Ochs and Prof Amos Etzioni at the APSID Inauguration Dinner Reception



"Discussion by the experts that happened during case discussion help a lot"

"Good to seeing how other immunologists approach patients with immunodeficiency"

Poster Walk

"All the cases were interesting and learned a lot from the discussion and comments by the faculty"



The Faculty enjoyed that too!

1. I was able to learn the rare PIDs which I only get to read in books
2. I am glad I was able to get the chance to interact w/ the experts on PID.
3. I am truly inspired by all of the faculty. I admire their hard work and compassion in taking care of PID pts. I hope, someday, we will be to increase the awareness of PID in our country and establish a collaboration with the other subspecialties

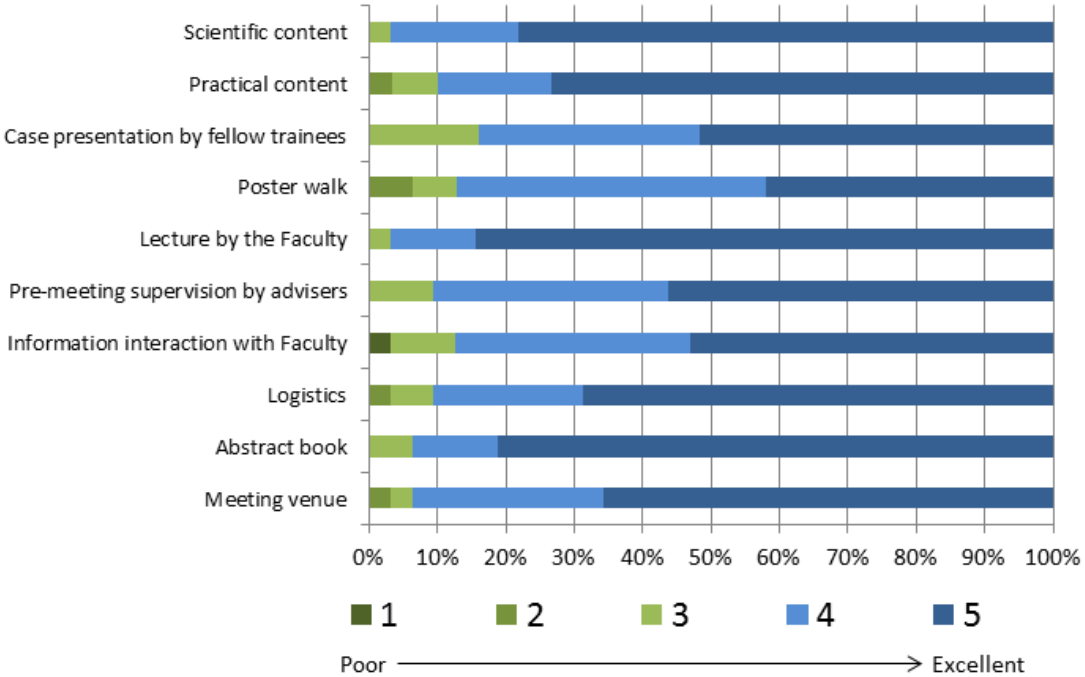
Connecting people.....



Spring School Evaluation

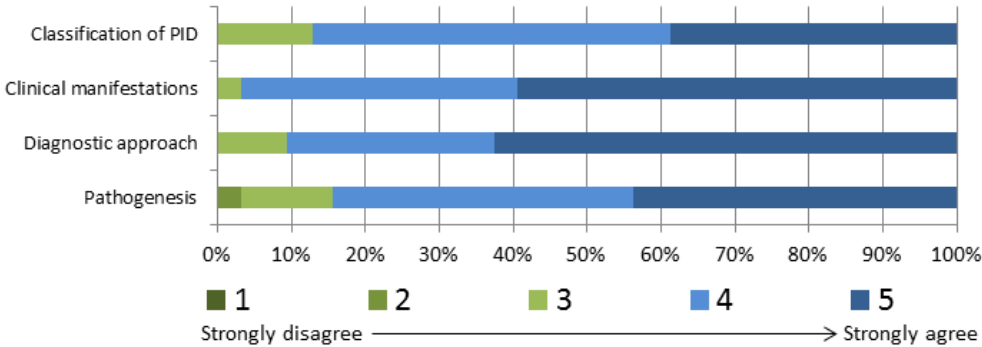
Overall rating

Please rate the following on a scale of 1-5



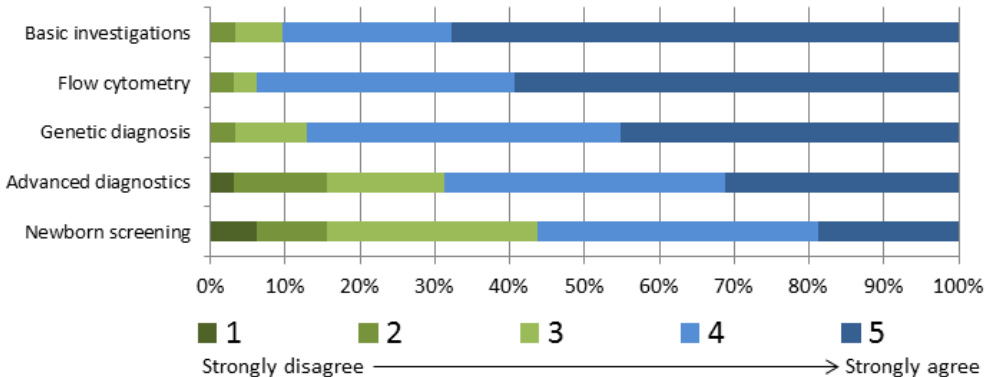
Basic concepts in PID

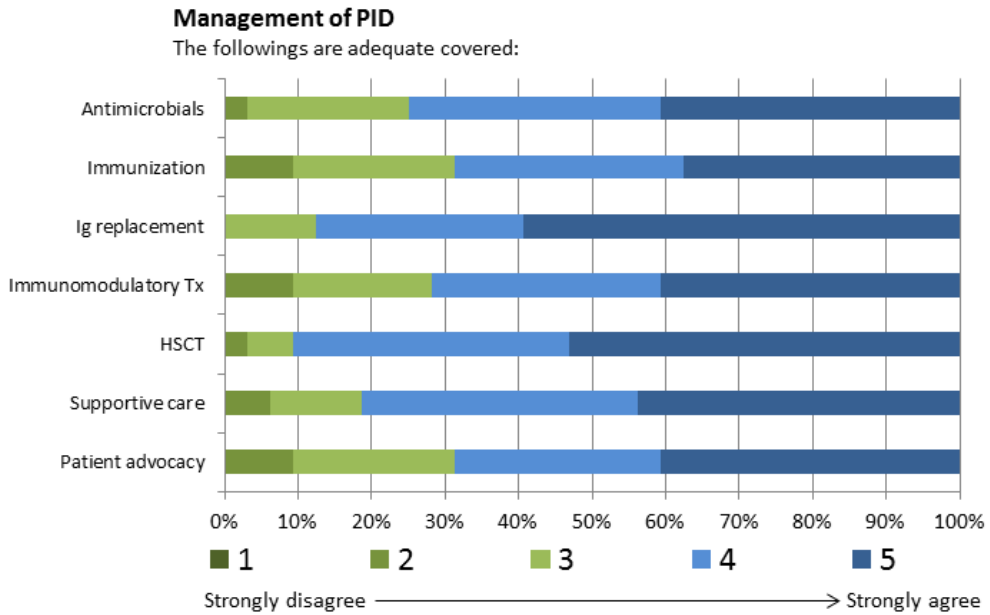
The followings are adequate covered:



Investigations related to PID

The followings are adequate covered:



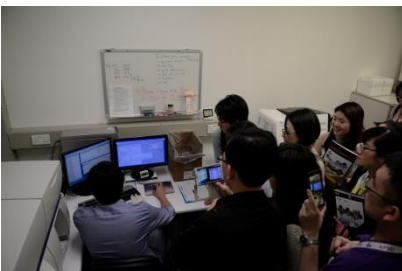


Educational Workshops

Two lunchtime workshops were held during lunch time on day 2 of the APSID Main Congress. In response to the quest for knowledge sharing in next generation sequencing and bioinformatics from the Genetics and Genomics Working Party, **Dr Wanling Yang** led a practical workshop on Genetics, using real case scenarios to demonstrate the steps in performing NGS data analysis. Participants were able to make use of on-line tools, databases and software to have better understanding about data processing and applications in clinical settings.



The Flow Cytometry Workshop was led by **Dr Jian Zheng** and **Dr Tai Lam** (Hong Kong). Using real human blood samples, participants had the opportunity to observe how specimens were prepared, and to gain understanding about the principle of panel design, fluorochrome selection and instrument set-up.



Patient Advocacy and Public Engagement



Primary Immunodeficiency League, the PID patient organization in Hong Kong, celebrates its first year birthday in 2016 and members were invited to join the APSID Congress Dinner in Loke Yew Hall, HKU. The joyous moment was shared by clinicians and nurses looking after our PID patients throughout the years, their parents and their loved ones. Some of the patient representatives also took part in the IPOPI Asian Regional Patient Meeting held prior to the APSID Main Congress at the Cyberport Hotel .

Five patients were previously invited to participate in the First International Festival of Pediatric Patients Paintings (IFPPP), and their paintings were selected for publication in the IFPPP book. They were awarded prizes for their wonderful work by Prof Nima Rezaei, representing the Health And ART (HEART) Group from Tehran, Iran.



The Congress Dinner was a special occasion for us to thank the Hong Kong Society for the Relief of Disabled Children (HKSRDC), who generously supported the genetic diagnosis for over 1,500 patients referred to The University of Hong Kong from more than 60 hospitals in Asia. An appreciation plaque was presented to the Mr TT Cheung, Vice-President of HKSRDC.

Working towards a better future in PID care requires support and participation from all stakeholders – patients, families, healthcare professionals, industry and government.



Mr TT Cheung and Prof YL Lau



Patients and families



APSID Traveling Award recipients



Industry sponsors

Planning for the Future: The APSID Executive Board

APSID had the first Executive Board Meeting on 1st May 2016. APSID is committed to organizing biennial scientific congress, and the next one will be hosted by mainland China in 2018. Two PID Schools will be held every year, with the upcoming Autumn School to be held in Kuala Lumpur, Malaysia in conjunction with the Asia Pacific Association of Allergy, Asthma and Clinical Immunology (APAAACI) and Asia Pacific Association of Pediatric Allergy, Respiriology & Immunology (APAPARI).

The Chairperson of each Working Party presented and affirmed the action plans and targets as discussed with their members in the 2015 Osaka Meeting. The ultimate goal is not just to connect PID centers in the Asia Pacific Region to develop clinical care protocols, to promote education and training, and to set up patient registries, but also to establish inter-professional collaborations with geneticists and transplant physicians to improve diagnostics and HSCT for PID.



Acknowledgement

The success of the APSID Inaugural Congress and Spring School was the hard work of the Local Organizing Committee, with full support from the Faculty, the Overseas Organizing Committee, our generous sponsors and all participating delegates.



Local Organizing Committee (from left): Dr Brian Chung, Dr Patrick Chong, Dr Mike Kwan, Dr Ka-Yau Lai, Dr Agnes Leung, Dr Joshua Wong, Dr Gilbert Chua, Dr Pamela Lee, Prof Yu-Lung Lau, Prof Godfrey Chan, Dr Marco Ho, Dr Wanling Yang, Prof Wenwei Tu, Ms Ada Tang and Mr Wilfred Wong



The local team from Queen Mary Hospital, with nurses Miu Ling (right) and Sau Man (left)



Support team: Raymond, Eva, Ivy, Chi, Chung, Ada, Vivian, Dora and helpers from sponsors

