



# IPOPI 4<sup>TH</sup> REGIONAL ASIAN PID MEETING

19-20 NOVEMBER 2022  
KUALA LUMPUR, MALAYSIA

an IPOPI event

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GLOBAL ALLIANCE EDUCATIONAL SEMINAR



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# APSID/SEAPID regional registry update: Where are we and where would we like to be?

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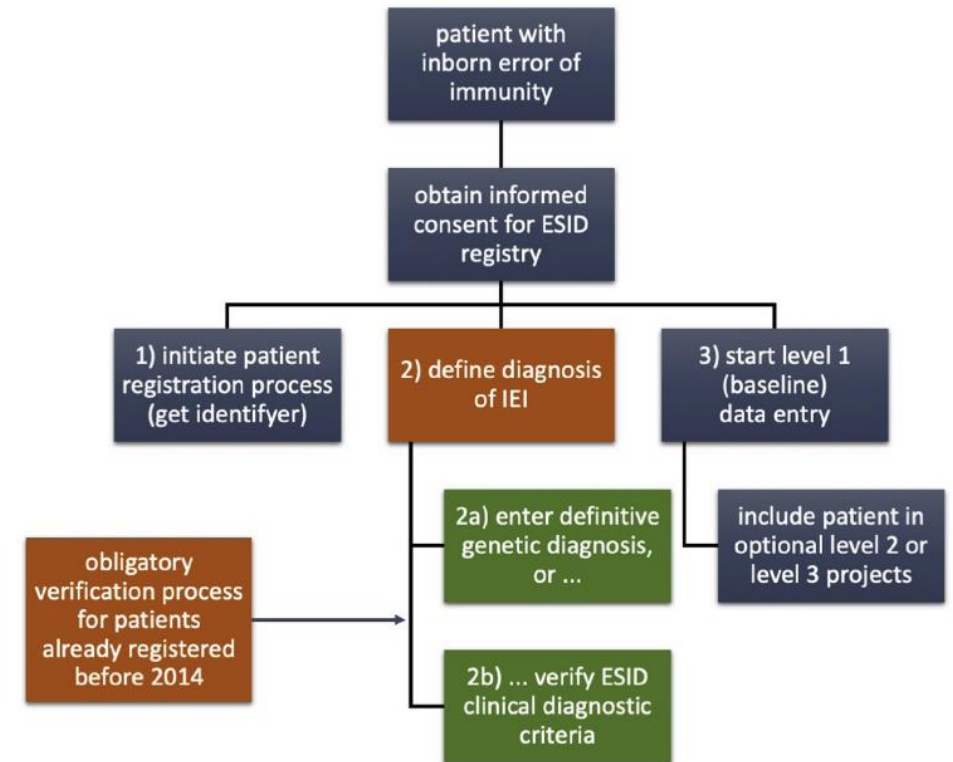
*APLAR Centre for Excellence in Rheumatology (2020-2023)*

*President, Asia Pacific Society for Immunodeficiencies (2020-2024)*

*ICMR Centre for Advanced Research in Primary Immunodeficiency Diseases (2015-2020)*

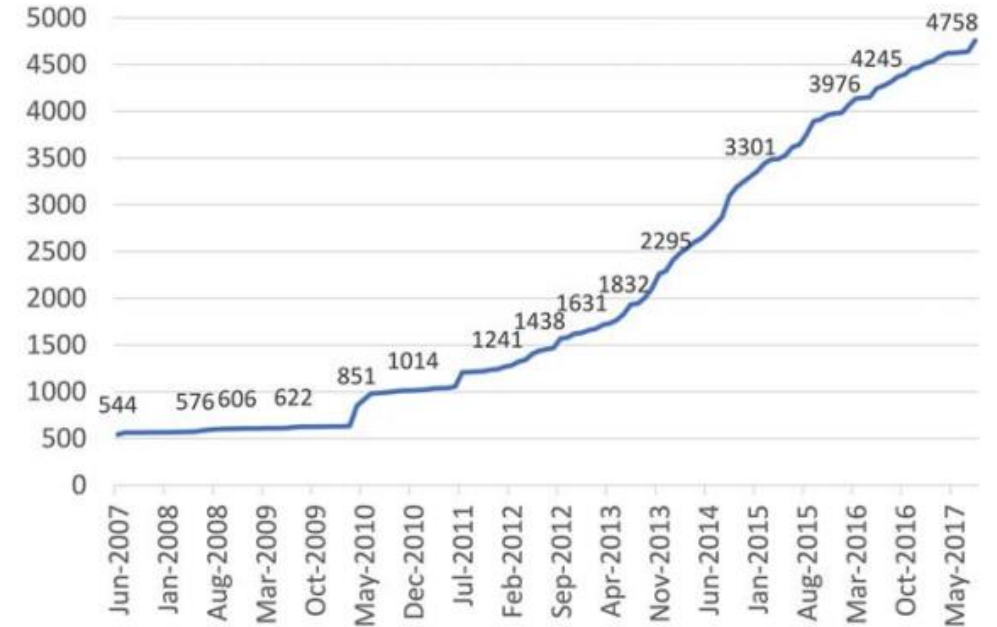
# European Society for Immunodeficiencies (ESID)

- ESID set up an internet-based patient registry in 2004
- 25,000 patients with inborn errors of immunity (IEI) registered as of 2019



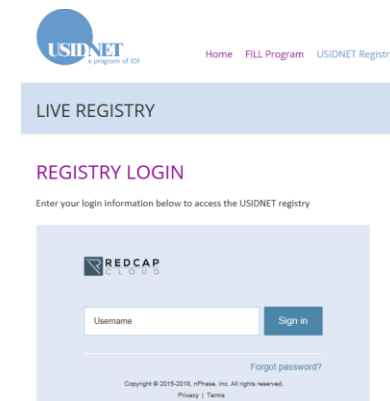
# United Kingdom PID (UKPID)

- Based on ESID registry
- Accessible to users through the website of the **United Kingdom Primary Immunodeficiency Network (UKPIN)**
- 95% (36/38) of centers in the UK have engaged with this project
- As of August 2017, 4758 patients have been enrolled
- Data are presented on the:
  - range of diagnosis
  - estimated disease prevalence
  - geographical distribution
  - age at presentation
  - treatment modalities
  - evidence of their monitoring and effectiveness



## Clinical Immunology Society (CIS)

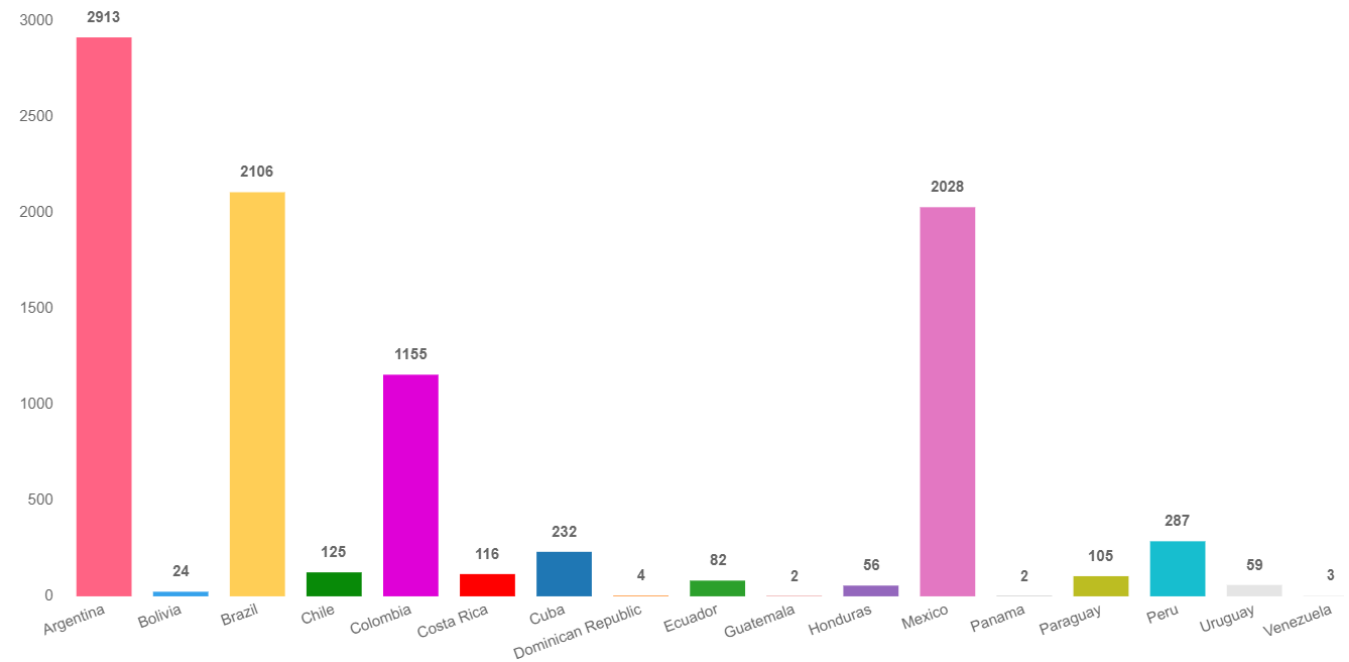
- PI Diseases Registry at US Immunodeficiency Network (USIDNET)
- The purpose of this project is to assemble and maintain a patient-consented registry of validated clinical, molecular, and laboratory data from PIDD patients in the United States and Canada (North America)
- The registry **began in 1992** as the '*Registry of U.S. Residents with Chronic Granulomatous Disease (CGD)*'



<https://usidnet.org/usidnet-registry/>

# Latin American Society for Immunodeficiencies (LASID)

- The LASID registry was **established in April 2009** with the support of the ESID, Jeffrey Modell Foundation, Baxter Bioscience and Conselho Nacional de Pesquisa e Desenvolvimento Científico (CNPq, Brazil)
- 9299 registered cases in LASID as of October 2022



# Asia Pacific Society for Immunodeficiencies (APSID) Registry (I)

- **2015 APSID (Osaka)**
  - *'PID Registry and Regional Network'* was formed at APSID Preparatory meeting
- **2016 APSID (Hong Kong)**
  - Current status of PID registry in Japan, Iran, Thailand and South Asia PID (SEAPID) were presented



**PID Registry and Regional Network (2016 APSID)**

# APSID Registry (II)

## Registry and Research Working Party

**2016-2020**

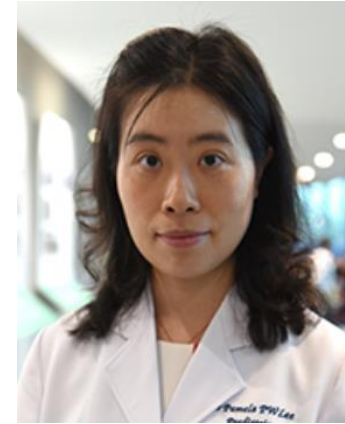


**Dr. Woei Kang Liew**

**2020-2024**



**Prof. Yae-Jean Kim**



**Dr. Pamela Lee**

# Milestones of APSID Registry (III)

- **2017: *Country Representatives / Participating Centers*** identified
- **2020:** Funding for APSID registry secured
- Pre-existing **registry in Hong Kong University** was converted to **APSID registry**  
<https://apsid.hku.hk/login.php>
- Updated the web-page format by the collaboration between HKU IT team and Korean IT team who developed KINET registry program: similar to that of ESID
  - Baseline and follow-up data
  - Level 1 and Level 2 data
  - Updated based on ESID diagnostic criteria and 2019 IUIS classification
- Delays because of COVID-19 pandemic
- **2021-2022:** APSID researchers participated in **collaborative research** on **Hyper-IgE syndrome** with researchers in **UK** through coordination of APSID registry network, and study was presented at 2022 ESID meeting

# Future of APSID Registry (IV)

- Will **update** APSID registry program according to 2022 IUIS classification
- Will **invite** more APSID centers, currently mostly HK patients (~190 patients)
- **Share** common English IRB form and consent form and translate to the local language in each participating country
- Update **structure and operating process** of APSID registry and working party
  - Ethics and publication / Protocol and data monitoring / Registry program education
- Need **collaboration** with existing registries in APSID countries
- **Funds** need to be secured
- **Goals for APSID registry:**
  - **Immediate:** summarizing epidemiologic data on major PIDs in APSID area (e.g., SCID, XLA, WAS, CGD, etc)
  - **Long-term:** regular and continuous updates on registry data and collaborative study on specific question per request through APSID registry network inside APSID and with other continent registries

# Current APSID Registry website (V)



TOLERANCE TO BALANCE...  
...CREATIVITY FROM DIVERSITY

## Study

Subject list

Enroll

Blank Form

### Icon Description:

- No data entered
- Currently entering
- Open query
- Query Answered
- (#) Open query count

Study
 Setting
 Export

Blank Form

Blank Form

Baseline	<a href="#">(Level 1) Baseline form</a>
Baseline	<a href="#">(Level 1) Way to diagnosis</a>
Baseline	<a href="#">(Level 1) PID Diagnosis</a>
Baseline	<a href="#">(Level 1) Stem cell / gene therapy</a>
Baseline	<a href="#">(Level 1) Immunoglobulin (Ig) replacement</a>
Baseline	<a href="#">(Level 2) Clinical manifestations</a>
Baseline	<a href="#">(Level 2) Diagnosis Form</a>
Baseline	<a href="#">(Level 2) Laboratory Values</a>
Baseline	<a href="#">(Level 2) Instrumental Data</a>
Baseline	<a href="#">(Level 2) Treatment</a>

Follow Up	<a href="#">(Level 1) Follow-up short form</a>
Follow Up	<a href="#">(Level 1) PID Diagnosis</a>
Follow Up	<a href="#">(Level 1) Stem cell / gene therapy</a>
Follow Up	<a href="#">(Level 1) Immunoglobulin (Ig) replacement</a>
Follow Up	<a href="#">(Level 2) Diagnosis Form</a>
Follow Up	<a href="#">(Level 2) Laboratory Values</a>
Follow Up	<a href="#">(Level 2) Instrumental Data</a>
Follow Up	<a href="#">(Level 2) Treatment</a>
Death Report	<a href="#">(Level 1) Death report form</a>

# Primary Immunodeficiency Database in Japan (PIDJ)

- Established in **2007**
- A **joint research project** with research groups associated with the **Ministry of Health, Labor and Welfare**; the **RIKEN Research Center for Allergy and Immunology (RCAI)**; and the **Kazusa DNA Research Institute (KDRI)**
- **PIDJ** serves as a database and as a patient consultation service connecting general physicians with PID specialists and specialized hospitals
- Since 2017, PIDJ ver.2 has been used
- As of 2021, 4,481 patients have been enrolled, of whom 64% have been subjected to genetic analysis



Mitsui-Sekinaka K et al., Front Immunol. 2022 Jan 10;12:805766<http://pidj.rcai.riken.jp/index.html>

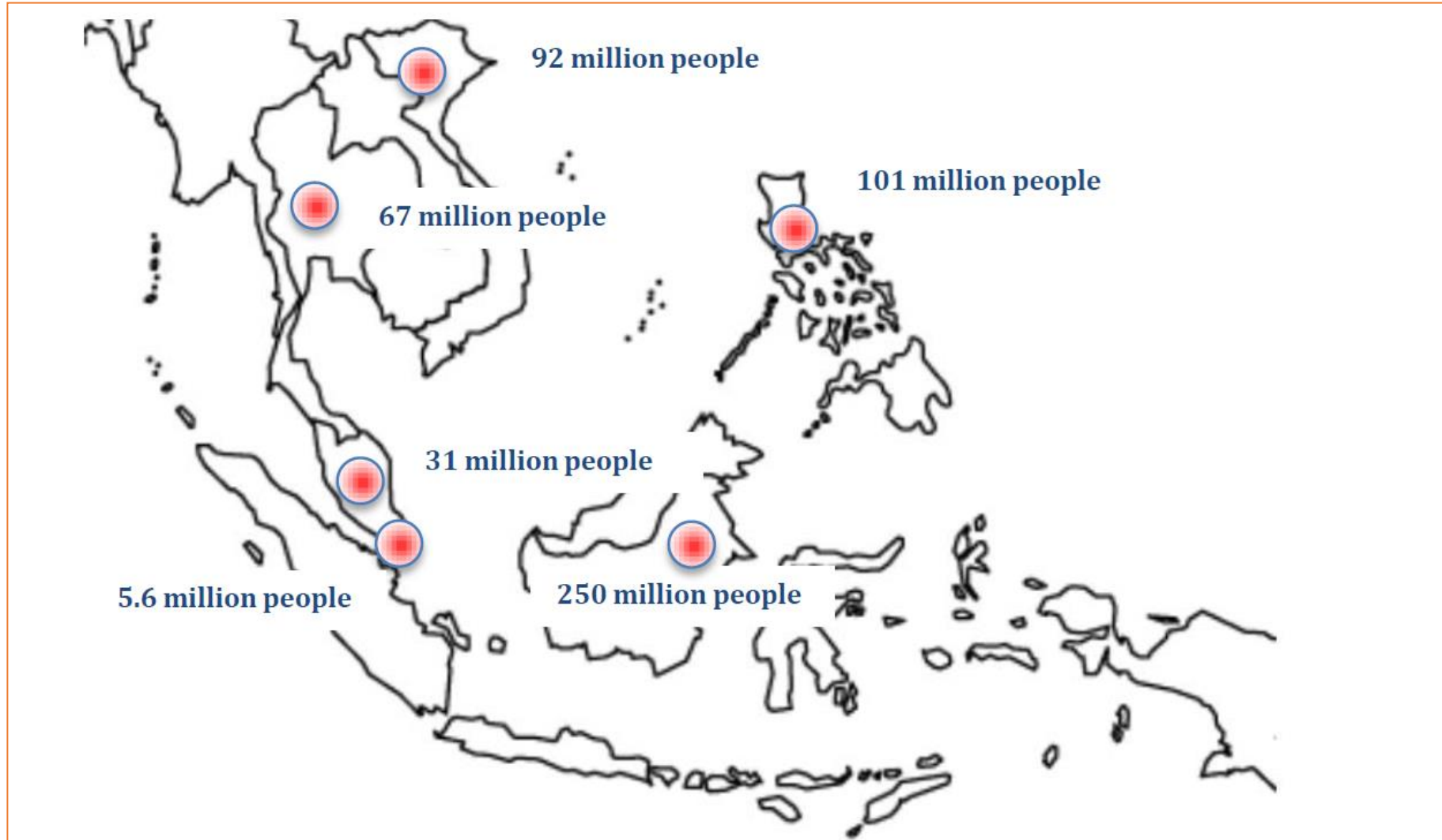
# Korean Immunodeficiency Network (KINET)

## Samsung Medical Center, Seoul, Korea (PI, Yae-Jean Kim)

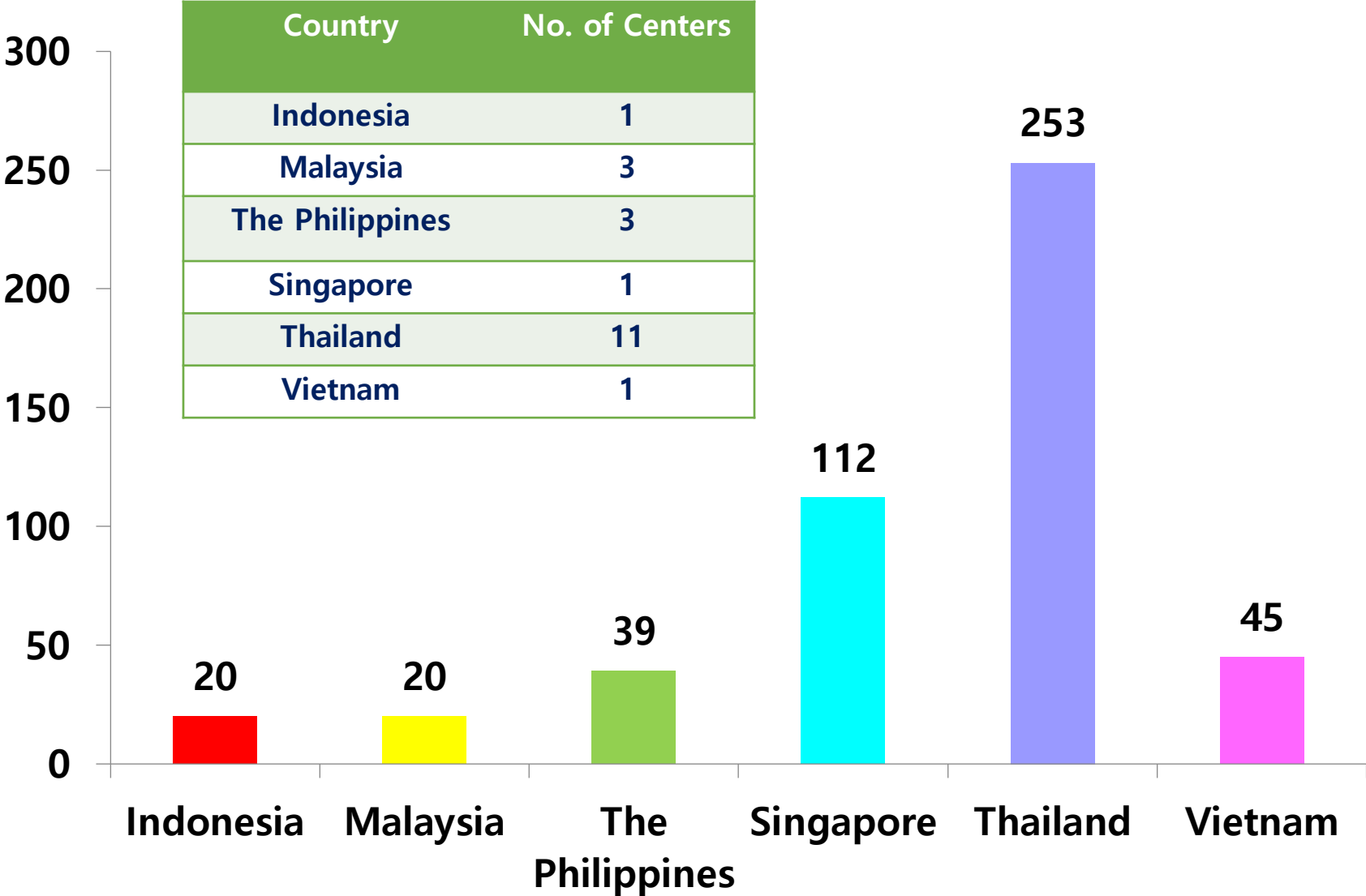
- **June 2017:** IT company selected, and first meeting to develop web-based PID registry
  - Four additional meetings in 2017 and 2018
- **May 2018:** First web-based Korean PID registry program established: <http://kpid.re.kr>
  - Web-based, user friendly
  - Included both ESID diagnostic criteria and IUIS classification
  - Started to enroll patients at Samsung Medical Center
- **2022:** Grant secured for national registry
  - Korean immunodeficiency Network (KINET) is established <http://kinet.or.kr>
  - Expanded to 5 centers in Seoul metropolitan area in 2022
  - Will expand to nationwide registry in 2023



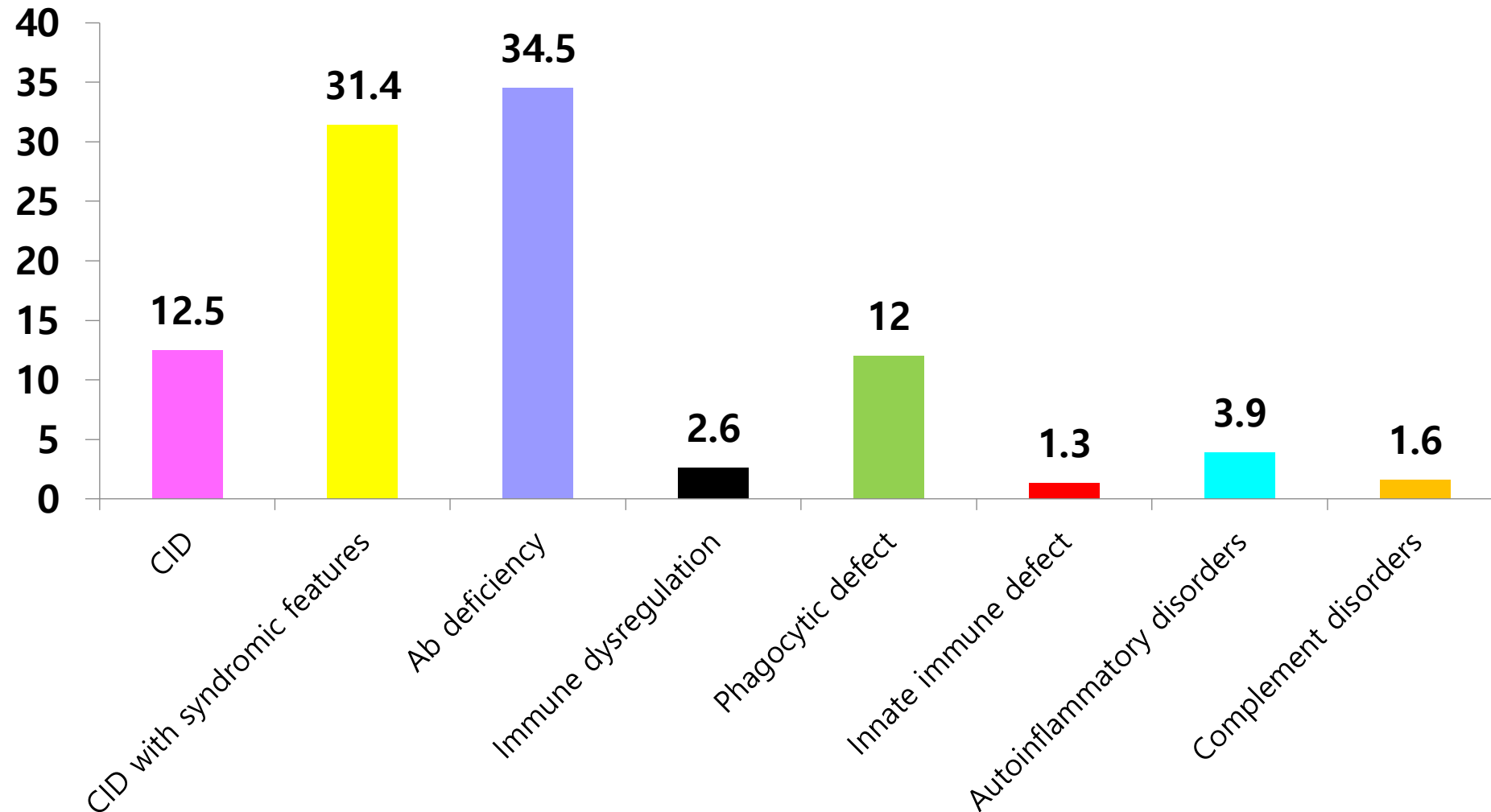
# SEAPID (Southeast Asia PID) Network



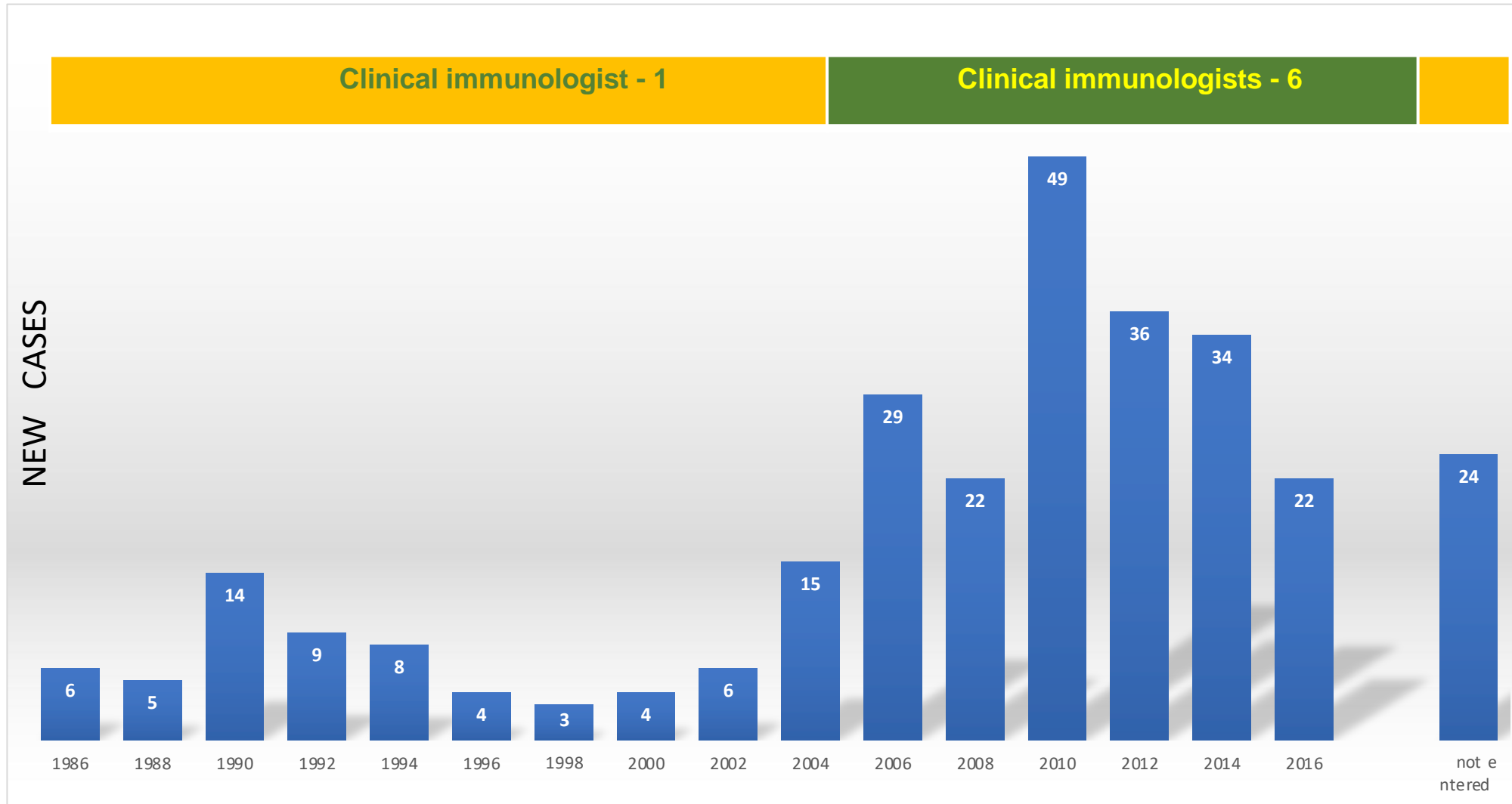
# Numbers of PID from SEAPID survey (2016)



# PID categorized by IUIS classification: SEAPID survey (2016)



# New PID cases in MyPIN Clinical Immunology Clinics Malaysia (1986-2017)



MyPIN

Malaysian Primary Immunodeficiency Network

Dr Lokman M Noh  
[ex prof]. UKM, USM, UPM ]

Dr Amir H Abd Latiff,  
Pantai Hosp K Lumpur, Mal

Assoc Prof Intan Hakimah  
Ismail`  
Univ Putra Malaysia, Serdang

Dr Kent Wu  
Gleneagle Hosp .Klumpur, Mal

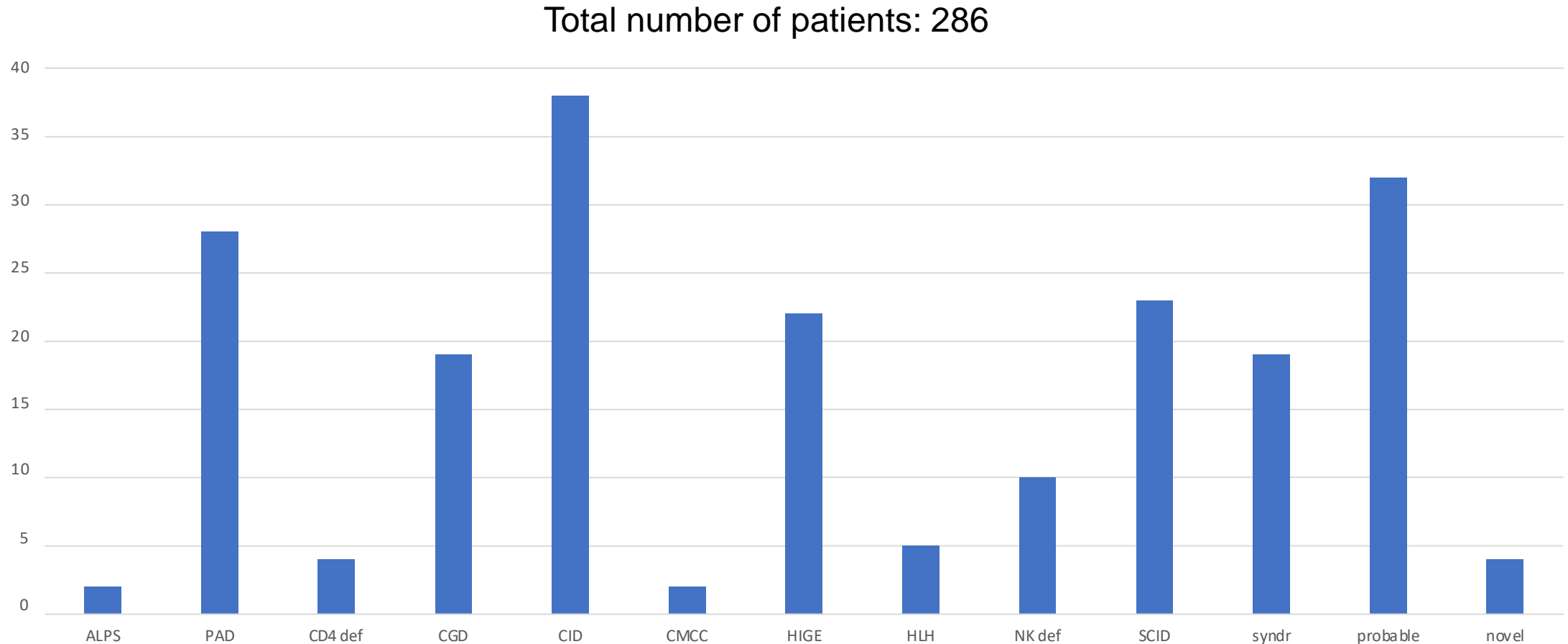
Dr Intan Juliana Hamid  
Univ Sains Malaysia, Penang  
[AMDI]

Assoc Prof Adli Ali  
Univ Kebangsaan Malaysia, K  
Lumpur

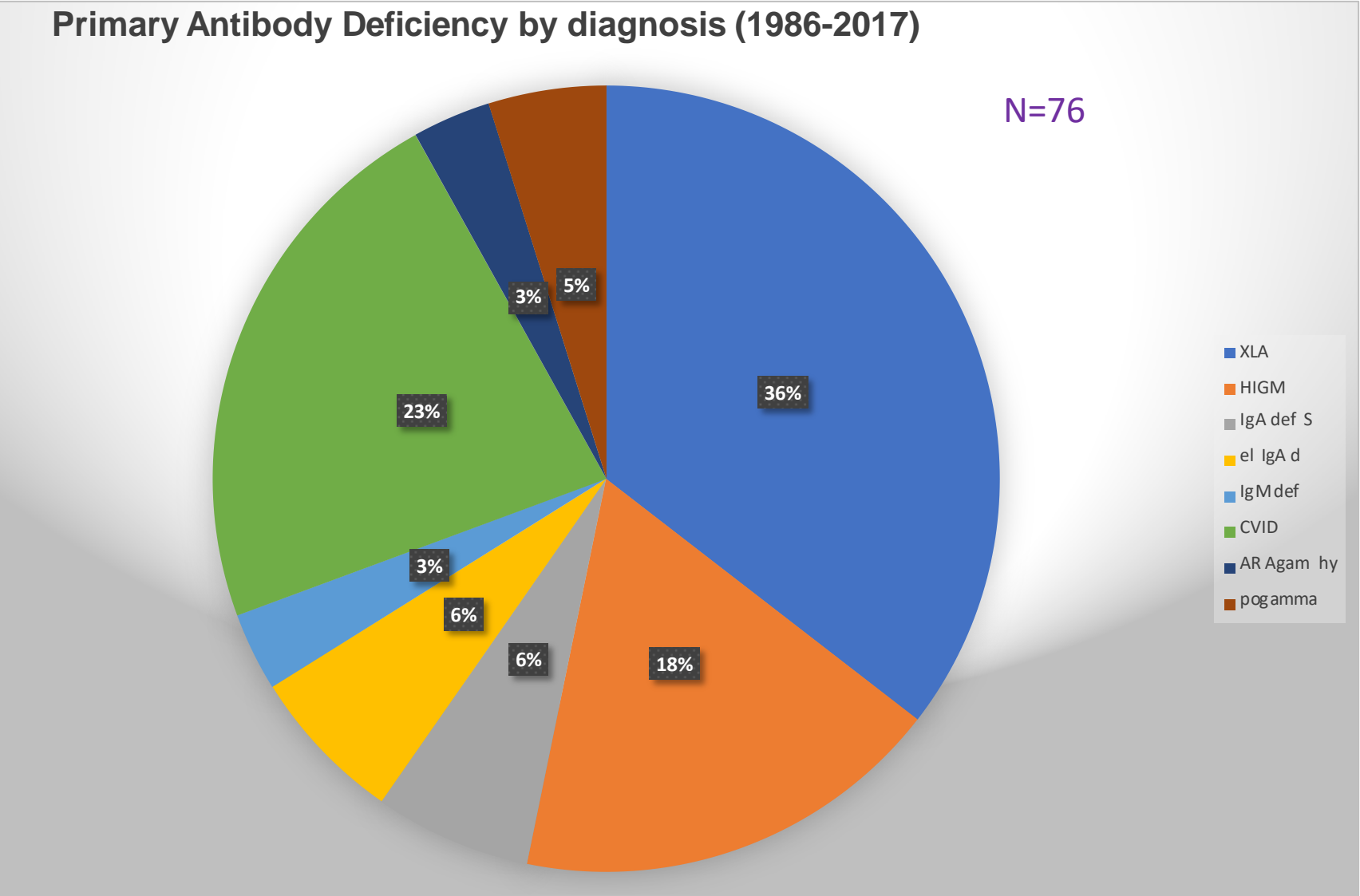
Year

Lokman M Noh. HTA K Lumpur Malaysia 2022

# Types of Primary Immunodeficiencies/ Inborn Errors of Immunity Malaysia MyPIN Registry (1986-2017)



# Primary Antibody Deficiency (PAD) in Malaysia (1986-2017)



# **Indian National Registry for Primary Immunodeficiency Diseases**

**Indian Council of Medical Research  
National Rare Disease Registry**

# Background

- Major objectives of the registry: collection of epidemiological, clinical, laboratory, and treatment data on important rare diseases
- Overall aim of facilitation of policy making, estimation of unmet medical needs, outcome assessment, facilitation of collaboration between stakeholders, multi-centric research

# Rare diseases included in the registry

- **Primary Immunodeficiency Diseases**

- *Haemoglobinopathies*
- *Bleeding disorders*
- *NMD*
- *LSD*
- *Skeletal dysplasia*
- *Small molecule disease*

# Structure of the registry

- **Central coordinating Center:**
  - Division of Reproductive, Biology, Maternal and Child Health, Indian Council of Medical Research, New Delhi
- **Coordinating center for QA/ QC:**
  - Division of Genetics, Department of Pediatrics, AIIMS New Delhi
- **Nodal centers:**
  - PGI Chandigarh
  - ICMR-NIIH, Mumbai
- **Participating centers**
  - 21 centers across India

# Proforma for Registry

- Socio-demographic details
- Clinical details
- Family history
- Investigations including molecular analysis
- Treatment options
- Ten diagnostic categories / Unclassified

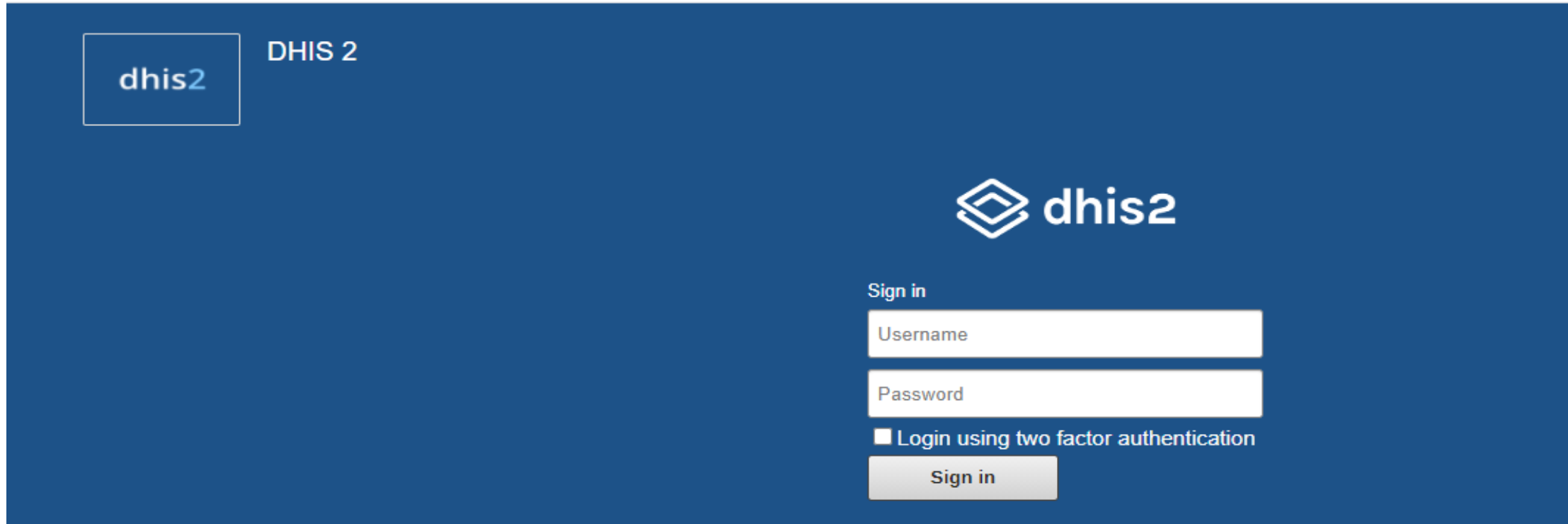
# Criteria for Inclusion in Registry

- Molecular defect proven

OR


- Patients satisfying ESID Working definitions for clinical diagnosis of PID

# The online portal



The image shows the login page of the DHIS 2 online portal. The background is a solid dark blue. In the top left corner, there is a small white box containing the text 'dhis2' in blue, followed by the text 'DHIS 2' in white. In the center right, there is a white logo consisting of a stylized 'd' made of three stacked diamonds, followed by the text 'dhis2' in white. Below the logo, the text 'Sign in' is displayed in white. Underneath, there are two white input fields: the first is labeled 'Username' and the second is labeled 'Password'. Below these fields is a checkbox with the text 'Login using two factor authentication'. At the bottom of the login section is a grey button with the text 'Sign in' in white.

dhis2 DHIS 2

 dhis2

Sign in

Username

Password

☐ Login using two factor authentication

Sign in

# The online portal

Enrolling organisation unit

National Institute of Immunohaematology

2021-02-16

Complete

Deactivate



Delete

## Tabular Data Entry

1. Socio  
Demographic  
Details

2. PID

Report date \*

2021-03-05

Date of onset of symptoms: yyyy-MM-dd

Age of onset of symptoms:

Years

Months

Disease Information:

Infection	<input type="radio"/> Yes <input type="radio"/> No	
Site of Infection	Number of infections	Number of infections in last year
1. Meningitis		

## Profile

Patient Name

test entry

Father's Name

Mother's Name

Patient's Aadhaar

Father's Aadhaar

Mother's Aadhaar

Gender

Date of Birth

Date of

Years

Months

Days



Nationality

State

District

City / Town of Birth



# Malaysia PID mortality - comparison

	WCH-HTA KL	MYPIN	WORLD
<b>All PID</b>	6/25 <b>(24 %)</b> [2017-2020]	na	China [12.3 %]. <sup>1</sup> 2004-2009
<b>SCID</b>	-	21/23 <b>(91 %)</b> (1992-2015)	S Africa. 11/14 <b>(79 %)</b> <sup>2</sup> (1983-2009) US. 5 %. <sup>3</sup>
<b>CGD</b>	-	5/20 (25 %) <sup>5</sup> 1991- 2016	ITALY 10 %. <sup>4</sup>
<b>XLA</b>		3/6 (50%) 1987-2007	

## Mortality:

- **Dengue:** 0.08%. (2021). Malaysia [KJ .NST Mar 2022
- **COVID 19:** 1.58 % 2020 [DGH . STAR Apr 9,2020]

1. Wang LL 2011. J Clin Immunol
2. Naidoo R. 2011, J Clin Immunol
3. R Buckley. SCID is a Pediatric Emergencies. NEJM 2000; 343(18): 1313
4. Martire B et al.. Clin Immuno 2008; 126:155-164
5. Noh LM et al.Allergy ,Asthma Clin Immunology . 2021;17:50

# Responsibilities of participating centres

- Ethics approval
- Identify a nodal person responsible for taking consent, filling of hard copy proformas and coordinating with the nodal centre for submission and feedback
- Ensure timely data sharing with nodal centre
- For avoiding duplication of patient data, **Aadhar number** to be recorded for all enrolled patients
- Data for more than one rare disease can be entered by each centre