



**International Union of Immunological Societies  
Primary Immunodeficiency Expert Committee**

**Information Pack**  
**Expert Committee on Primary**  
**Immunodeficiency**  
**2014**

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## **IUIS Introduction**

The International Union of Immunological Societies (IUIS) is an umbrella organization for many of the regional and national societies of immunology throughout the world. As articulated in its Constitution, the objectives of IUIS are:

- Organize international co-operation in immunology and to promote communication between the various branches of immunology and allied subjects
- Encourage within each scientifically independent territory co-operation between the Societies that represent the interests of immunology
- Contribute to the advancement of immunology in all its aspects.

There are currently 69 Member Societies of IUIS, many of which belong to one of four Regional Federations encompassing Europe, Latin America, Africa and Asia-Oceania. Links with other international scientific bodies are promoted through the International Affiliate Members of IUIS, membership of the International Council for Science (ICSU) and formal liaison with the World Health Organization.

Activities of IUIS are overseen by the Officers and Council, who are elected at the triennial General Assembly and are broadly representative of the interests and geographical distribution of member societies. Several Committees have also been established to conduct activities of ongoing interest to IUIS, currently in the areas of clinical immunology, education, nomenclature, quality assessment and standardization, and veterinary immunology.

International Congresses of Immunology are held every three years under the auspices of IUIS. IUIS also contributes to the staging of regular congresses and conferences by each of the four Regional Federations and to various educational activities in immunology.

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Primary Immunodeficiency Expert Committee  
2014

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## IUIS Primary Immunodeficiency Expert Committee

### **Introduction**

The Primary Immunodeficiency Expert Committee was originally set up under the auspices of the World Health Organization in 1973, to describe and classify the types of primary immune defects affecting humans. The classification of those diseases recognized at the time was published in a report (Cooper MD et al NEJM 288 966-967) and revised every two years subsequently. However in the 1990s the WHO made a policy decision to concentrate on more common diseases, so the IUIS agreed to take on the remit of this committee; this was formalized at the 54th IUIS Council meeting in Taipei in 2008.

### **Mission Statement**

- Provide an up-to-date classification of all primary immunodeficiency diseases (PIDs)
- Assist with the identification, diagnosis and management of patients with these uncommon conditions
- Support diagnostic and therapeutic guidelines developed by national societies and others, to assist healthcare providers
- Promote awareness, diagnosis and treatment of PIDs in all regions of the world
- Produce ad hoc reports on any aspect of PIDs and to assist in the welfare of patients with these conditions

### **Classification Tables**

An up-to-date report on an ever-increasing list of primary immunodeficiencies (PID) is produced biennially. The latest version of the “Classification of Primary Immunodeficiency Diseases by the International Union of Immunological Societies (IUIS) Expert Committee on Primary Immunodeficiency” was published in *Frontiers of Immunology* in April of 2014 (*Front. Immunol*, 2014 vol. 5 pp. 162) with a recent corrigendum, and is freely available in pdf or HTML format for all at <http://journal.frontiersin.org/Journal/10.3389/fimmu.2014.00162/abstract>.

The process of considering reports of new primary immune deficiencies, in order to classify them appropriately, continues at biennial meetings held in different continents. The current classification was produced as a result of the work that started in the meeting that was held in New York in April of 2013.

This current report highlights over 230 primary immune deficiencies and provides important definitions for clinical management and standardized research, as well as for patients. Readers of the report include clinicians who use this document in their daily practice in many countries and scientists investigating the immunologically mechanisms that prevent infectious diseases, autoimmunity, inflammation, cancer and allergy in humans. Many national and international healthcare providers use this document for the basis of funding therapies, and professional medical bodies have based their diagnostic guidelines on this classification.

About 20 or more new PID are described every year. The use of modern tools of genetics such as genome and exome sequencing, SNP arrays, CGH along with powerful algorithms of bioinformatics to name a few, is providing is resulting in more inborn errors of immunity being described per year (*J. Exp. Med.*, 2014 vol. 211 (11) pp. 2137-49). In addition, these technologies are facilitating the recognition of the same genetic defects in unrelated patients. However, the number of single patients left without candidate genes shared by other patients is also growing. Therefore, it is expected that improvements in these technologies coupled to more sophisticated tools of molecular biology (transfection and knockdown of genes, induced-pluripotent stem cells -iPSC-, gene editing, etc) will allow a significant number of new PID will be characterized in detail in the next few years.

The Committee is holding its biannual business meeting along the European Congress of Immunodeficiencies in Prague in October of 2014. We will define place and date for the next biannual meeting sometime in the first semester of 2015.



## IUIS PID-EC 2014

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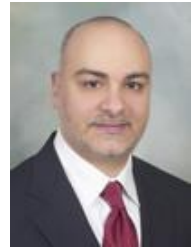
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Lennart Hammärstrom, Alain Fischer, Helen Chapel, Mary Ellen Conley.

### **IUIS PID-EC Report 2014**

Al-Herz W, Bousfiha A, Casanova JL, Chatila T, Conley ME, Cunningham-Rundles C, Etzioni A, Fischer A, Franco JL, Gaspar HB, Holland SM, Klein C, Nonoyama S, Ochs HD, Oksenhendler E, Picard C, Puck JM, Sullivan K, Tang MLK. Primary immunodeficiency diseases: an update on the classification from the international union of immunological societies expert committee for primary immunodeficiency. *Front. Immun.* (2014) vol. 5 pp. 162

DOI: 10.3389/fimmu.2014.00162

**The full report can be found on the IUIS website:**

<http://journal.frontiersin.org/Journal/10.3389/fimmu.2014.00162/abstract>

### **IUIS PID-EC Report 2011**

Al-Herz W, Bousfiha A, Casanova JL, Chapel H, Conley ME, Cunningham-Rundles C, Etzioni A, Fischer A, Franco JL, Geha RS, Hammarström L, Nonoyama S, Notarangelo LD, Ochs HD, Puck JM, Roifman C, Seger R, Tang MLK. Primary Immunodeficiency Diseases: an update on the Classification from the International Union of Immunological Societies Expert Committee for Primary Immunodeficiency. *Frontiers In Immunol*, 2011, 2: 1-26.

DOI: 10.3389/fimmu.2011.00054

**The full report can be found on the IUIS website:**

<http://journal.frontiersin.org/Journal/10.3389/fimmu.2011.00054/abstract>

**Other Publications related to the PID-EC for the 2011 Classification tables:**

#### **Editorial comments:**

1. Chapel H. **Classification of primary immunodeficiency diseases by the International Union of Immunological Societies (IUIS) Expert Committee on Primary Immunodeficiency 2011.** *Clin Exp Immunol*, 2012. 168(1): 58.

Comment: Editorial note to inform the community about the new classification.

[Al-Herz W](#), [Notarangelo LD](#). **Classification of primary immunodeficiency disorders: one-fits-all does not help anymore.** *Clin Immunol*. 2012 Jul; 144(1): 24-5.

**Comment:** Letter to the editor highlighting the improvements the Classification and limitations.

Bousfiha AA, Jeddane L, Ailal F, Al-Herz W, Conley ME, Cunningham-Rundles C, Etzioni A, Fischer A, Franco JL, Geha RS, Hammarström L, Nonoyama S, Ochs HD, Roifman CM, Seger R, Tang MLK, Puck JM, Chapel H, Notarangelo LD, Casanova JL. **A Phenotypic Approach for IUIS PID Classification and Diagnosis: Guidelines for Clinicians at the Bedside.** J Clin Immunol, 2013 vol. 33 (6) pp. 1078-87.

**Comment:** Simplified version as a practical reference tools for Physicians, Scientists and Patient Organizations

## IUIS PID EC Activities

### Previous activities

- Report on classification of PIDs published biennially since 1973. For the latest version of the Classification see page 12 of this report and follow the link. **For previous versions of the classification and other publications by the Committee see appendix 1 (page 24).**
- Availability of immunoglobulin – Successful application to WHO Essential Medicines List 2007 and WHO Essential Medicines List for Children 2008
- Safety of immunoglobulin therapies – Position paper published in Clinical and Experimental Immunology in 1999

### Current activities

- Revision of national and international guidelines for the diagnosis and or management of immune deficiencies by expert groups on request. *Liaison – Bobby Gaspar*
- Assistance with the revision of diagnostic criteria for primary immunodeficiencies with the IUIS Clinical Immunology committee and the European Society for Immune Deficiencies.
- Production of simplified versions of the PID Classification Tables for the general public. *Liaison – Aziz Bousfiha and Jean Laurent Casanova*
- Assistance to Orphanet and the World Health Organization in the revision of the International Classification of Diseases (ICD) codes for the primary immune deficiencies, version 11, due to be published in 2015. *Liaison – Mimi Tang*
- Assurance of continued inclusion of therapeutic immunoglobulin in the World Health Organization Essential Medicines Lists. *Liaison – Jose Franco*
- To encourage, with the International Patient Organisation for Primary Immunodeficiencies (IPOPI), the implementation of the recommendations from the IUIS PID position paper on the Safety and Availability of Immunoglobulin (1999) including: the avoidance of blood donation by individuals who have received blood or blood products in the past; recording of all batch numbers for tracing in the event of any pathogen transmission; to attempt (with the patient organizations) to obtain ante-mortem permission for post-mortem examination in all patients with primary immune deficiencies in countries in which post-mortem examinations are permitted. Advice on the routine immunisations needed for patients with primary immune deficiencies. *Liaison – Jose Franco*

### **Relationship with IUIS**

The PID committee liaises with the following IUIS committees:

- *IUIS Committee on Clinical Immunology* - particularly in relation to diagnostic criteria and availability of new therapies.
- *IUIS Committee on Education* - advertisement of PID schools (winter, spring and summer schools) for young investigators run by International PID Organizations: European Society for Immune Deficiencies, United States Clinical Immunology Society, Latin American Society for Immunodeficiencies (LASID) and the South Asia Pacific group. The objectives are to facilitate the exchange of students from different regions, encourage new groups (such as the African Society for Immunodeficiencies) to establish similar educational opportunities for PID.
- *IUIS Committee on Quality Assessment and Standardization* – particularly in relation to QA for diagnostic procedures in PIDs, such as test immunization responses.

### **Sponsors**

The PID-EC is grateful to the following for their continued support: Octapharma, Baxter and Kedrion, We also thank previous sponsors: Grifols, Talecris and CSL-Behring. Without them, the committee would not had be able to meet these responsibilities.

## **IUIS PID-EC Job Description – Chair**

### **Organization**

IUIS / Primary Immunodeficiency Expert Committee

### **Position Title**

Chair of the Primary Immunodeficiency Expert Committee (PID-EC)

### **Appointment Process**

Nominations received from the PID-EC members and a vote taken.

### **Term**

The Chair of the committee will start as Co-Chair for a term of 2 years, followed by 2 years of acting as Chair, followed by 2 years of acting as Past-Chair.

### **Position Summary**

The Chair of the Committee leads and oversees the operations and affairs of the PID-EC.

### **Key Responsibilities and Duties**

- Responsible for coordinating the completion and revision of the classification of Primary Immunodeficiency Diseases, typically every two years.
- Initiates and leads the Committee's annual report, budget and financial statement to the IUIS Council.
- Responsible for coordinating revision of the ICD codes that pertain to Primary Immunodeficiency Diseases. In this task, the Chair will be assisted by the Secretary and by all Committee members.
- The Chair shall represent the Committee with other scientific organizations and patient advocacy groups.
- The Chair shall be responsible for calling and chairing all meetings of the Committee, assigns work to the Committee members, sets the agenda and runs the meetings, and ensures distribution of meeting minutes.
- The Chair shall serve as the official spokesperson of the Committee at the IUIS Council meeting. The Chair may delegate to be represented by the Secretary or by another Committee member, upon approval by the majority of the Committee.
- Provides leadership and direction for Committee activities.
- Sets tone for the Committee work.
- Oversees the logistics of Committee's operations.
- Builds an effective team and inspires the members to actively participate.
- Ensures that members have the information needed to do their jobs.



- Works to ensure succession planning of the members and the chair.
- Initiates and leads the Committee's annual evaluation.
- Performs other duties as determined by the IUIS Council.

**Reports to**  
IUIS Council

## **IUIS PID-EC Job Description – Co-Chair**

### **Organization**

IUIS / Primary Immunodeficiency Expert Committee

### **Position Title**

Co-Chair of the Primary Immunodeficiency Expert Committee (PID-EC)

### **Appointment Process**

Nominations received from the PID-EC members and a vote taken.

### **Term**

The Co-Chair will start as a regular member for the term of 2 years, followed by 2 years of acting as Chair, followed by 2 years of acting as Past-Chair.

### **Position Summary**

The Co-Chair of the Committee assists with leads and oversees the operations and affairs of the PID-EC.

### **Key Responsibilities and Duties**

- Supports the activities of the chair to initiate the Committee's annual report, budget and financial statement to the IUIS Council.
- Assist the chair with coordinating revision of the ICD codes that pertain to Primary Immunodeficiency Diseases.
- The Co-Chair shall represent the Committee with other scientific organizations and patient advocacy groups in case the Chair is absent.
- The Co-Chair shall be responsible for assisting the Chair in all meetings of the Committee, as well as to assign work to the Committee members, sets the agenda and runs the meetings, and ensures distribution of meeting minutes.
- Ensure that all the members are attending the meetings.
- The Co-Chair shall represent the Chair and serve as an official replacement spokesperson of the Committee at the IUIS Council meeting.
- Provide support and direction for Committee activities as the Chair is absent.
- Oversees the communication of Committee's operations.
- Participate in the Chair's task to build an effective team and inspire the members to actively participate in any activities of the committee.
- Ensures the supervision of members' tasks and provide them with the information needed to do their jobs.
- Works to ensure succession planning of the members and the chair.
- Performs other duties as determined by the PID-EC Chair or the IUIS Council.

**Reports to**  
Chair of the PID-EC

## **IUIS PID-EC Job Description – Secretary**

### **Organization**

IUIS / Primary Immunodeficiency Expert Committee

### **Position Title**

Secretary of the Primary Immunodeficiency Expert Committee (PID-EC)

### **Appointment Process**

Nominations received from the PID-EC members and a vote taken.

### **Term**

6 years

### **Position Summary**

The Secretary of the Committee leads the administrative affairs of the PID-EC.

### **Key Responsibilities and Duties**

- Works with the Treasurer to generate income for the PID-EC.
- Maintains accurate records of the work of the PID-EC.
- Reports to the IUIS Council yearly as required, ahead of the Council meeting.
- Liaises sub-committees of the IUIS PID-EC with other IUIS Committees.
- Liaises with IPOPI and other organizations.
- Ensures that activities of members are carried out, implemented and concluded in a timely manner.
- Oversees the arrangements of committee meetings and manages the minute taking and record keeping of the meetings and ensure their distribution in a timely manner.
- Generate the annual report of activities for the IUIS annual Council meeting and with the Treasurer provide feedback to the financial report in a timely manner.
- Ensures that the Chair has the information needed to make decisions about the PID-EC's future strategy.
- Keeps accurate records to ensure succession planning of the Secretary role.
- Provides information for the Committee's annual evaluation.
- Performs other duties as determined by the PID-EC Chair or IUIS Council.

### **Reports to**

Chair of the PID-EC

## **IUIS PID-EC Job Description – Treasurer**

### **Organization**

IUIS / Primary Immunodeficiency Expert Committee

### **Position Title**

Treasurer of the Primary Immunodeficiency Expert Committee (PID-EC)

### **Appointment Process**

Nominations received from the PID-EC members and a vote taken.

### **Term**

6 years

### **Position Summary**

The Treasurer of the Committee leads and oversees the financial operations and affairs of the PID-EC.

### **Key Responsibilities and Duties**

- Manages the PID-EC bank account income and expenditure.
- Works with the Secretary to generate income for the PID-EC.
- Oversees the financial impact of the Committee's activities.
- Sends requests for funds for the following year with Secretary's annual report to IUIS.
- Claims financial allocation from IUIS in a timely manner.
- Ensures that the Chair has the information needed to make financial decisions about the PID-EC's future strategy.
- Keeps accurate records to ensure succession planning of the Treasurer role.
- Provides financial information for the Committee's annual evaluation.
- Performs other duties as determined by the PID-EC Chair or IUIS Council.

### **Reports to**

Chair of the PID-EC

## **IUIS PID-EC Job Description – Member**

### **Organization**

IUIS Primary Immunodeficiency Expert Committee

### **Position Title**

Member of the IUIS Primary Immunodeficiency Expert Committee (PID-EC)

### **Appointment Process**

Election by current members of the committee after an election organized by the Secretary

### **Term**

One will serve a term of 8 years, with an option to renew for a following 4 years.

### **Position Summary**

Committee members are required to participate in ongoing and new projects on matters pertaining to the IUIS PID-EC Committee mission.

### **Key Responsibilities and Duties**

- Makes a serious commitment to participate actively in Committee work. He/she must commit the time, energy and creativity needed to assist the Committee in realizing its goals and objectives.
- Regularly participates in Committee conference calls and attends meetings. If s/he fails to attend two meetings consecutively, s/he will be deemed to have resigned.
- Stays informed about Committee matters; is well prepared for meetings and reviews and comments on minutes and reports.
- Actively participates in planning and implementing short and long-range goals and projects of the Committee.
- Leads a particular project at the request of the committee.
- Is responsible for responding to communications from the Executive Committee in a timely manner and accepting and performing tasks assigned to him/her by the Chair.
- Promotes the IUIS and the PID-EC to colleagues and other organizations. Serves as an advocate of the IUIS.
- Takes a genuine interest in other Committee members and builds a collegial working relationship that contributes to consensus.
- Brings concerns to the Committee's table if they could negatively impact any work of the IUIS and/or the IUIS PID-EC.

**Reports to**

Chair of the PID-EC

**Other Factors**

- Seeks guidance from Committee Chair when needed.
- Requests the information needed to do his/her jobs from Committee Chair and/or Secretary.
- Maintains active communications with Committee Chair and/or Secretary.

**APPENDIX 1**

1. Rosen FS, Eibl M, Roifman C, Fischer A, Volanakis J, Aiuti, F, Notarangelo L, Kishimoto T, Resnick IG, Hammarstrom L, Seger R, Chapel H, Cooper MD, Geha RS, Good RA, Waldmann TA, Wedgwood RJP. **Primary immunodeficiency diseases. Report of an IUIS Scientific Committee. International Union of Immunological Societies.** *Clin Exp Immunol* (1999) vol. 118 Suppl 1 pp. 1-28.
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## **APPENDIX 2**

### **Guidelines for updating the Classification Tables**

1. The application for creating and updating the tables is MS Word
2. Always use “track changes” so others are able to see the changes
3. Only the Group Chair will accept the track changes
4. The font to be used is Arial 8
5. When there are multiple gene defects, these should be split into separate cells of the table
6. In the final tables, any reference to “mice” should be left out
7. A legend explaining abbreviations should be put at the bottom of the document, below the table
8. The use of the asterisk is to indicate rare cases (ten or fewer unrelated cases reported in the literature). These should be put at the bottom of the document below the legend
9. Footnotes should be put at the bottom of the document below the legend, and below the asterisk

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