



## **REGISTRY WP ESID 2015 Newsletter DECEMBER Edition**

**Paris, Nov. 25<sup>th</sup> 2015**

Dear all,

The ESID registry is the largest database on PID worldwide and one of the major component of our Society. Since its creation in 2004, it has gathered information on a growing number of patients collected through 100+ centers across Europe.

Together with **the Registry WP-Steering Committee Elect** (Bodo Grimbacher, Germany - Isabella Quinti, Italy - Matthew Buckland, United Kingdom - Markus Seidel, Austria - Joris van Montfrans, The Netherlands) and Mrs. Jose Drabwell, IPOPI President, along with Gerhard Kindle from Freiburg University, we continue to work on the new Registry platform.

### **Some recent achievements**

**1. Data transfer to the new registry (Level 1):**

Since the launch of the new system, the number of validated patients has continued to grow along with the number of newly registered, among whom more than 1,200 are newly registered patients.

Several centers have expressed interest in joining the ESID Registry such as the Swedish PID Group, along with some Russian centers.

**2. ESID Registry Clinical Diagnosis Criteria:**

The ESID Registry WP has developed new working clinical diagnosis criteria for PID patients lacking a genetic documentation (<http://esid.org/Working-Parties/Registry/Diagnosis-criteria>) in order to enhance the quality of registration of patient.

**3. ESID Database data access and publication rules:**

These have been recently updated.

Please visit our website for further information.

**4. ESID Registry website section is full of resources:**

- a. New ESID Registry
- b. Diagnosis criteria
- c. Registry publications
- d. Studies
- e. Documents
- f. Steering Committee
- g. Documenting centers
- h. Old Registry
- i. ESID Database Statistics
- j. Informed patient consents
- k. Contact info



## REGISTRY WP ESID 2015 Newsletter DECEMBER Edition

### 5. Level 3 studies:

- a. **The APDS project** (PIs: Prof Stephan Ehl - Freiburg, Prof. Alison Condliffe - Sheffield, Dr. Sven Kracker - Paris). This collaborative study is financially supported by 3 pharmaceutical companies: Novartis, HSK and UCB. More information here: <http://esid.org/Working-Parties/Registry/Studies/APDS-Registry>
- b. **The unPAD** (unclassified Predominantly Antibody Disorder, PI: Prof. Esther de Vries). The initiation of this study is financially supported by PPTA.

### 6. Nine papers using the ESID Registry or the ESID clinical criteria have been published this year so far:

1. Marschall K et al. *Clin Exp Immunol*. **The Swiss National Registry for Primary Immunodeficiencies: Report on the first 6 years' activity 2008-2014.** (PMID: 26031847)
2. Brignier AC et al. *J Allergy Clin Immunol*. **Early-onset hypogammaglobulinemia: A survey of 44 patients** (PMID: 25959671)
3. Grimbacher B; ESID Registry Working Party. *Clin Exp Immunol*. **The European Society for Immunodeficiencies (ESID) registry 2014** (PMID: 25546747)
4. Jonkman-Berk BM et al. *Clin Immunol*. **Primary immunodeficiencies in the Netherlands: National patient data demonstrate the increased risk of malignancy.** (PMID: 25451158)
5. Turley AJ et al. *J Clin Immunol*. **Spectrum and Management of Complement Immunodeficiencies (Excluding Hereditary Angioedema) Across Europe.** (PMID: 25663093)
6. Aydin SE et al. *J Clin Immunol*. **DOCK8 Deficiency: Clinical and Immunological Phenotype and Treatment Options - a Review of 136 Patients.** (PMID: 25627830)
7. Salvator H et al. *Eur Respir J*. **Pulmonary manifestations in adult patients with CGD.** (PMID: 25614174)
8. Suarez F et al. *J Clin Oncol*. **Incidence, presentation, and prognosis of malignancies in ataxia-telangiectasia: a report from the French national registry of primary immune deficiencies.** (PMID: 25488969)
9. Ludviksson BR et al. *J Clin Immunol*. **Epidemiology of Primary Immunodeficiency in Iceland.** (PMID: 25315263)

### **In the near future, we plan to:**

1. Complete the working diagnostic criteria
2. Implement the first level 2 registration forms (for Combined Immunodeficiencies)



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3. Implement the link to a high quality genetics module, along with the ESID Genetics WP.
4. Foster clinical and translational research on PIDs
5. Implement the APDS Registry Level 3 study eCRFs
6. Perform the first analysis of the new registry (epidemiological data, therapy data...)
7. Secure the Registry funding through Level 3 projects along other potential sources, including the ESID financial support
8. Continue to offer the most updated website section for all users and visitors
9. Renew a call for grants for helping centers to document their patients in the Registry

You can have **more information here:** <http://esid.org/Working-Parties/Registry>

**And last but not least: Thank you to all contributors to the ESID Registry!**

Best wishes,

**Nizar Mahlaoui**

REGISTRY WP Chairman, on behalf of the ESID Registry WP Steering Committee.

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