



## The ESID Online Database Network for PIDs

### An international platform for PID epidemiology and research

Since 2004, ESID (European Society for Immunodeficiencies, [www.esid.org](http://www.esid.org)) has been running an online database for primary immunodeficiencies. The ESID database is a secure, internet-based patient registry which combines both clinical and laboratory data of PID patients. It provides a common data set for, currently 214 different PID entities and is updated regularly, according to progress in research. This data set includes diagnosis, therapy, medical history, laboratory data and health outcome indicators.

With this database, ESID is striving to answer important questions on the epidemiology of PID: What is the incidence and prevalence of PID in different countries and in Europe as a whole? What is the mortality rate and how is it changing with progress in diagnosis and therapy? What are the prevalent treatment strategies? What is the diagnostic delay in different countries and for different PID? Other registries in Europe have been addressing these questions before, mostly on the national level, such as in Spain (REDIP, <http://web.hsd.es/redip/>) and Italy (IPINET, [ww.aieop.org](http://ww.aieop.org)). The ESID Online Database offers a platform to unite these efforts and collect reliable epidemiologic data for all European countries.

However, the ESID database is not merely a single time point survey restricted to epidemiology. It also serves as a research platform with long-term documentation. Using the database, researchers have the possibility to identify patient cohorts for genetic screening and multi-centre trials. Datasets can be flexibly extended for studies using the database as a platform for their reporting forms.

#### Current status

Since its start in August 2004, a total of 7,499 patients from 54 individual centres and national registries have been documented in the ESID Database. The current documentation progress is shown in Fig. 1.

Although this figure is still far from presenting a complete picture of PID in Europe, the current results already give valuable insights into the epidemiology of PID, in particular in those countries with a high documentation rate.

For example, after reading the article “Joint Nordic meeting in Sweden” (e-update September 2008, p.10), we recently analysed the gender distribution of PID patients in children vs. adults. While the article stated that “70% of adult PID-patients are women”, our results from the ESID Database do not support this hypothesis:



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We analysed the gender of all patients who had not deceased and were not lost to follow-up. The total number of these was 6,771. Fig. 2 shows the gender distribution in total as well as in the groups of patients younger than 16 years (“children”) and 16 years or older (“adults”). The female share was indeed higher in the adult group, but men still represented a majority with 56.4%.

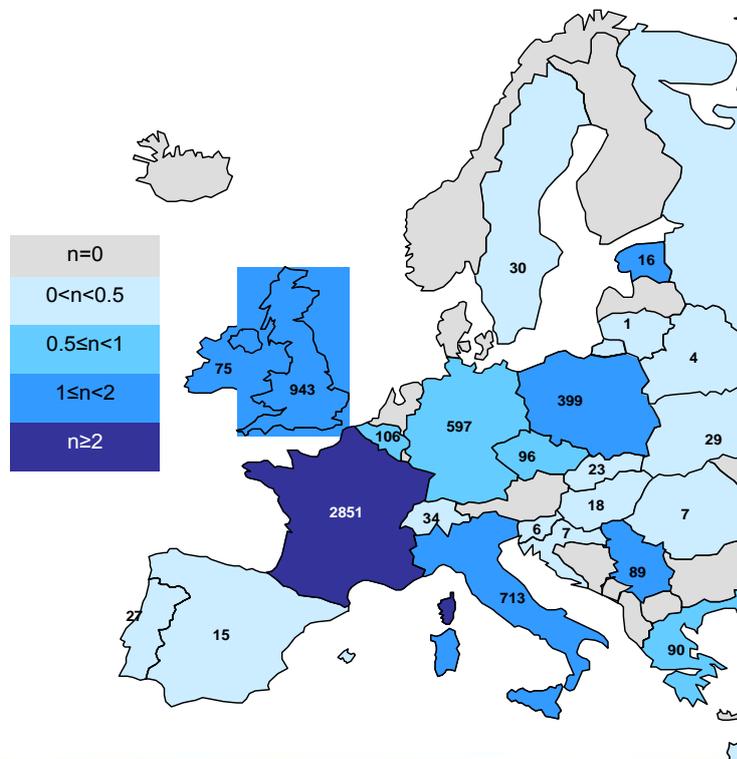
In terms of research, the ESID Database has already been used in surveys on X-linked thrombocytopenia (XLT) patients and neuroendocrine carcinoma in patients with CD40L deficiency. Multi-centre clinical trials on common variable immuno-deficiency (CVID) and X-linked agammaglobulinaemia (XLA) are currently under way.

## Public awareness

We believe that running the ESID database is essential to raising public awareness of PID. While information on the prevalence of PID is often based on estimates the ESID database will hopefully soon contain enough documented patients to provide reliable epidemiological data to the public as well as to health authorities thus showing that PID are perhaps not as rare as often believed.

The ESID Database team is already collaborating with several patient organisations on the national level, such as DSAI in Germany and PIA in the United Kingdom. We are looking forward to establishing more of these collaborations and invite any organisation interested to contact us at [registry@esid.org](mailto:registry@esid.org).

**Benjamin Gathmann, ESID Online Database Team**

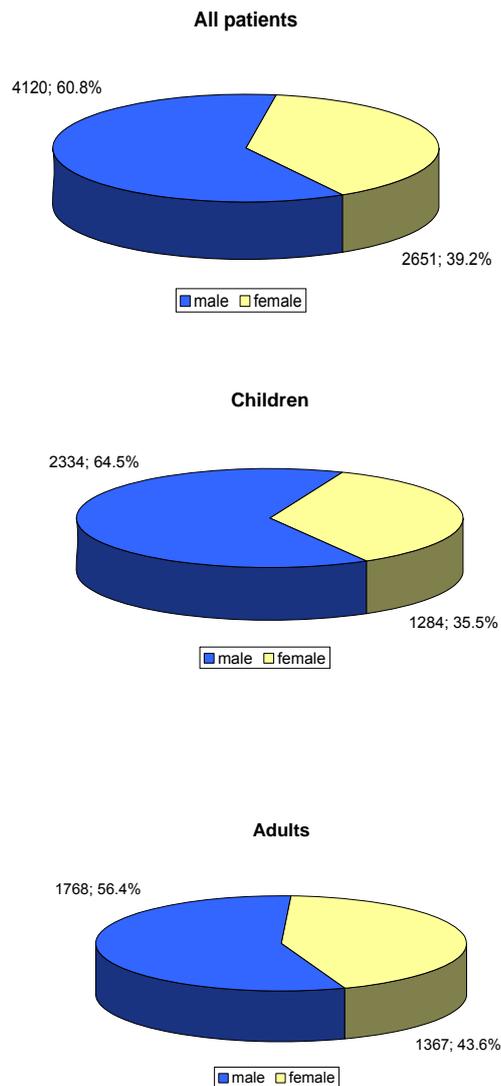


**Fig. 1 Registration progress by countries**

The total number of patients documented in each country is displayed on the map. The colours represent the number of registered patients in relation to the country population, according to the colour legend. (n patients per 100,000 inhabitants)



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**Fig. 2 Gender distribution**

Gender distribution of patients in the ESID Online Database. “Children” group: patients aged 15 years or younger. “Adults” group: patients aged 16 years or older. The charts show the absolute number of patients per gender, followed by the relative share in %.



## The UK PID registry - A national approach to data collection with a view to international research

In the UK like in several other countries the necessity for a National PID Registry has become more and more apparent over the years. Patients and healthcare professionals share an interest in such a registry which will be an essential instrument for high-quality research and ultimately will contribute to the improvement of patient care. Another key objective of data collection is to raise public awareness and to strengthen PID's position towards key stakeholders like the media and the government.

The British patient organisation PIA (Primary Immunodeficiency Association, [www.pia.org.uk](http://www.pia.org.uk)) and UK PIN (The UK Primary Immunodeficiency Network, [www.ukpin.org.uk](http://www.ukpin.org.uk)) have combined forces to set up a National Registry. Obviously, a state-of-the-art technology was required and the UK centres decided to benefit from the experience and reliability of an established and proven system, namely the ESID Online Database for Primary Immunodeficiencies. This system has been running successfully since 2004 and provides all required features. It collects data on clinical as well as laboratory and quality of life data and enables long-term observations through follow-up documentation. A more detailed description of the ESID Registry can be found in this editions of e-Update on pages 8 - 10.

However, the UK centres opted for a solution that hosts this registry on a dedicated server in the UK. ESID fully supports National Registries in this decision. Thus, the UKPIN registry steering group has initiated all necessary processes to set up a server with a copy of the ESID database in the near future. The necessary ethics approval has been obtained and we are in the process of finalising data protection policies together with University College London, where the server will be hosted.

### Current status

Currently, eight UK centres already contribute their patients' data. Data of 943 PID patients has already been entered on the ESID platform (as of December 12<sup>th</sup> 2008).

As soon as the server is established, many more UKPIN member centres are expected to join. PIA supports the project not only by providing the necessary server hardware, but mainly by funding one person to visit the documenting centres and help them with data entry. This has proven to be crucial in other countries already because obviously it does mean a certain amount of additional work for the centres and often a lack of time and manpower has been an obstacle. Some centres are lucky to have dedicated persons who make sure the data is updated regularly, whereas others rely on additional help.

Another advantage of using the ESID system is that although the UK National Registry is an independent system and can be adapted and amended according to UK needs, it still can communicate with the international version and thus enables networking and research on a much wider scale. The patient can choose whether he wants to make his data available for research within the UK only or whether he wants to make it available for potential international research projects by exporting the data once a year to the international ESID platform. For further information you are welcome to contact Viviane Knerr, database coordinator at the Royal Free Hospital, University College London: [v.knerr@medsch.ucl.ac.uk](mailto:v.knerr@medsch.ucl.ac.uk).

A total number of 36 UKPIN member Trusts shall be added  
Currently 8 centres have entered 940 patients

