



ESID online clinical and research database

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The European Society for Immunodeficiencies (ESID) is a non-profit organisation which seeks to facilitate the exchange of information between people concerned with Primary Immunodeficiencies (PID) and supports the research on causes, mechanisms and treatment of PID. Since 2004, ESID is running an online-based research patient registry on a platform capable of performing clinical and post marketing surveillance. This is made possible through the financial support of five PPTA member companies and the support of the European Union within the 6th framework program.

The ESID database is a secure, internet-based patient registry, which brings together clinical and laboratory data of PID patients. It improves diagnosis, classification, prognosis and therapy by enabling the exchange of information among centres, assuring detailed long-term documentation and facilitating large genetic and therapeutic projects. The database allows the documentation of patients with a primary immunodeficiency into an online system which provides a common dataset for almost 180 different PIDs. This so-called core dataset includes diagnosis, therapy, laboratory data and secondary outcome measures. All documenting centres receive an annual incentive of 10 Euro for every complete core dataset. This payment is meant to compensate for the administrative work of entering the data.

After accessing the website www.esid-registry.org, the user selects the sub-registry in which he/she wants to document a new patient. The large number of PIDs is grouped into seven categories which are: Predominantly antibody disorders, predominantly T-cell deficiencies, phagocytic disorders, complement deficiencies, Other well defined PIDs, autoimmune and immunedysregulation syndromes and unclassified immunodeficiencies.

In addition to the common core dataset, experts from all over Europe work on the creation of large disease-specific data models. So far, the following 8 sub-registries feature an extended dataset: Common variable immunodeficiency,

DiGeorge syndrome, Nijmegen breakage syndrome, Centromere instability and facial abnormality syndrome, Hyper IgE-syndromes, ICOS deficiency, IPEX syndrome and Secondary hypogammaglobulinemia. Seven more are presently being programmed (X-linked agammaglobulinemia, Severe combined immunodeficiency, Hyper IgM Immunodeficiency, IgA deficiency, Osteopetrosis, Ataxia teleangiectatica, TACI deficiency). A novel tool allows the deposition of genetic mutation data in the ESID database as well as in the "IDbases" in the Institute of Medical Technology in Tampere, Finland. This tool currently already works for x-chromosomal and autosomal dominant mutations and will be extended to approximately 90 sub-registries.

In 2005, the focus of the database team lay mostly on the acquisition of documenting centres and on the increase of patient numbers. Today, 50 documenting centres in 24 different countries have already signed up. The Italian National PID Register (AIEOP) has been included electronically. The United Kingdom decided to set up a National Registry within the ESID database in 2006. French patients will be entered by a National Centre in Paris, whereas Spain has set up its own online registry based on a one-time registration. The Spanish data is intended to be imported electronically into the ESID system. Additional National Registries are planned in Poland, Belgium and the Netherlands. The ESID CVID-register compiled by Dr. Lennart Hammarstrom in Sweden with 519 patients will be integrated successively in the ESID-online CVID subregistry once patients have consented.

With 1,123 patients in November 2005, the database already offers a considerable cohort of patients, so that in 2006 we will concentrate more on output-features. The database also contains valuable anonymized data for PPTA member companies, e.g. on therapy (drug groups, route of administration, compliance, side effects) and on the quality of life (days missed at school/work, days in hospital). Therefore survey requests of our sponsors are very welcome.

Visit the ESID website at: www.esid.org
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