

A First Step: The Importance of Patient Registries

Report from the ESID Registry of Primary Immunodeficiencies

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The prevalence of immunodeficiency disorders varies extensively depending on the type of disease. Many of these disorders are quite rare and only a few patients are found in each country. Furthermore, multiple etiologies and distinct clinical subtypes exist for some diseases where each subtype is represented by a limited number of patients in each major referral center.

Genetic and/or therapeutic studies usually involve a large number of patients. Identification and localization of patients is therefore necessary for the successful completion of these studies. Access to a sufficiently large number of patients thus constitutes the basis for clinical trials and it is only by pooling the resources from different countries that we will solve the molecular basis of these disorders and this will be able to institute

efficient therapeutic measures.

The formation of a main registry of patients with various forms of immunodeficiency in Europe is a first step to achieve a foundation on which future experiments and trials can be based. A working party within ESID has started to build such a registry. The center is currently located in Sweden (Lennart Hammarström and Mohammad R. Abedi) with a close collaboration with additional cen-

ters in England (Gareth Morgan and Hockchye Gooi), Italy (Roberto Paganelli), and Spain (Nuria Matamoros). Data are submitted to the main registry on a simple form which contains all pertinent types of diagnosis, easily computerized and with a simple mode of data entry and retrieval.

The registry was started in January 1994 and was initially supported by a European Community grant (PL 1321; see previous

ESID newsletters or ESID registry home page on Internet for more details). The computer hardware used for registry work has been donated by Pharmacia & Upjohn, Sweden.

Until now, 9707 patients in various European countries have been entered into our files. After the start, there has been a steady flow of the patient reports to the registry. Altogether, we have received data from 26 countries, and the registry is nearing completion in terms of country participation. In countries where a national registry already exists, data may be submitted either to the national registry only or to both the national and the ESID registries at the same time. Data from the existing national registries will in turn be submitted to the central ESID registry on a collaborative basis. The continued growth of the registry is now mainly due to the submission of individual patient reports from the different ESID countries. So far, patients with primary antibody deficiencies comprise a majority of the reported patients, although a considerable variation is seen between reports from different ESID membership countries.

The ESID main registry has reached the size where it is useful for various epidemiologic, therapeutic, and genetic studies. Multiple requests for information have been processed and confidential patient codes, derived from the registry, are included in publications based on these patients. It also forms the basis of various subregistries (Btk, WAS, CD40L, CGD) which have recently been started. These subregistries, which are independently maintained and funded, contain detailed genetic and clinical information and thus greatly enhance the effectiveness of the ESID registries. Additional subregistries on, among other diseases, ataxia telangiectasia and common variable immunodeficiency, are currently being planned. Contacts have also been made with representatives of the American, South African, Australian, and far east (Chinese) registries in order to be able to establish a future collaboration which may benefit our goals. ●