Registry Working Party
The redesign of the ESID Registry

A summary explaining the development and design of the new ESID Registry

May 8, 2013

Dear users of the ESID Online Registry,

We would like to update you with progress on the redesign of the ESID Online Database.

We are delighted with the support you have shown for the ESID Registry which now has more than 18,000 patient entries from 29 different countries. This success is due to the enthusiasm and hard work of all the participating centres, and we greatly appreciate your continued commitment to this important initiative.
Over the last two years we have carefully reviewed the structure and operation of the Registry. We recognise that there is a need to make data entry simpler, whilst ensuring the highest quality of data input.
By making it simpler, we hope to encourage more data entry and by reviewing all aspects of quality we aim to ensure that we can generate much more reliable data on e.g. prevalence, incidence, diagnostic delay and Ig replacement.
We have also identified specific problems including some poorly defined data fields, which are therefore difficult to complete. A need for well defined case definitions to assist accurate diagnostic data entry and the need to ensure long term follow up data is maintained.
Technology has also moved on since the original system was established in 2004 and we needed to consider that in our re-design.

A steering committee comprised of representatives from national PID registries in Europe (see: Registry Steering Committee) was established in 2011, and has been considering the establishment of this new system.

To date, this redesign consists of the following components:

- A redesign of the data model (level 1-3, see below)
- A redesign of the technical platform (the data entry pages on the internet)
- Improved data quality control
- Automatic requests for a (short) yearly follow-up documentation of each patient

This means that

- There will be a new data entry system replacing the current one.
- All data fields that exist in the new system will be transferred from the old to the new system.
- There will be additional fields (e.g. on first clinical symptoms) which must be entered for each patient to make his or her entry complete in the new system.

It is important to mention that

- The current system will remain available as a read-only for accessing previous data.
- All current data will therefore remain available to you (also possible as e.g. csv or xls files)

The data transfer to the new system will include a quality check and verification process

- A quality check of existing data will be executed by the database administration before transfer to the new system (e.g: In IgA deficient patients, is IgA <0.07 g/l etc).
- There will be a requirement to verify the diagnosis for patient entries with no genetic diagnosis in the new system. For this purpose, we are developing ESID Registry working criteria for the clinical diagnosis of immunodeficiencies in collaboration with the ESID Clinical Working Party (for example: ALPS, Hyper IgE syndrome, CVID).
- Furthermore, you will need to verify that the patient is still alive.
- Entries of deceased patients will not be transferred to the new system, unless you specifically require this for some purpose. These data will remain available on the old system

The overall purpose of this data checking process is to improve the quality of data in the new system, so that we will have a patient Registry which is as reliable as can reasonably be expected, by ourselves, patients, reviewing authorities and sponsors alike.

Level 1 data model

Please go to the New ESID Registry section to view the first data models of the new ESID Registry.

These are termed "Level 1 dataset". Level 1 is mandatory for each patient. All parts of it must be completed. At initial registration ("baseline"), there are sections on diagnosis, HSCT and Ig replacement that need to be completed. At follow-up, the system asks for a status update and for changes in therapy. If the patient has died, or the diagnosis has changed, details on these changes can be recorded

For all of your patient entries that are transferred to the new system, you will need to complete some new fields on level 1 that do not exist in the current system.

The new system will ask you to update your patients once a year. If you miss to update some patients, the system will send a reminder at the end of the year with a list of these patients.

Level 2 and level 3 data models

As you can see, at level 1 we do not ask for lab values, clinical features and other treatments than Ig replacement and HSCT. These will be covered on the second level. The level 2 data models will be category (or disease)-specific (based on the IUIS classification). They are under development.

Level 2 is optional. If you do not have the resources, you will not have to work on level 2 at all. Depending on your research interests or the type of patients you attend, you can also choose to document level 2 for selected categories only. However, if you decide to use level 2 for one or more categories, patients will only be considered fully registered if you complete all fields for these patients (just as at level 1).

In addition, there shall be dedicated studies on a third level. These shall have a fixed time frame (e.g. two years) and address specific questions defined by a study protocol, including a statistical evaluation plan and a „feasibility assessment". The Registry Steering Committee will review such proposals and decide whether they should be added in the Registry.

There is no fixed date for the new system going live yet, but we expect it to be available in the second half of 2013.
As for ethical approvals and informed patient consents, it should not be necessary to collect these anew. However, regulations differ from centre to centre and country to country, so to be on the safe side, you should check with your local authorities once the new system is up and running.

If you have any comments, please send them to esid.registry@kenes.com or contact your representative in the steering committee (a list of he members is given on the website cited above).

We believe that the redesign of the registry will make it a very valuable tool and hope for your further commitment and support of the project.

Yours sincerely,

Prof. Stephan Ehl
Chairman ESID Registry Working Party

Benjamin Gathmann
ESID Registry coordinator

Additional note (July 2013):

In order to simplify the change to the upcoming new ESID Database, we have added the new fields contained in level 1 to the current system on June 14, 2013.

These fields are restricted to the "Diagnosis" form of the core dataset. The new fields include
-reason for genetic diagnosis
-sequencing method
-approximate age of onset in years (alternative to the date of onset)
-first symptom
-lab abnormalities
-"unknown" options for date of clinical and genetic diagnosis
-new options "no mutation found" and "not genetically tested"
The new fields are displayed in bold letters.

You have now got the opportunity to complete these fields for all of your patients (or as many as possible) before the data is transferred to the new system. This way, you will have less work once the new system goes productive. It is projected to be available this fall.