

USIDNET Registry Guide for Enrolling Patients

First and foremost, the United States Immunodeficiency Network (USIDNET) would like to thank you for contributing patient data to the registry.

How to begin:

- (1) Submit your protocol to your local IRB for approval.
For your reference, approved protocol documents from participating institutions can be found at www.USIDNET.org under Disease Registry. If you do not have a local IRB, you may enroll your patients under the Western IRB. All patients to be enrolled in the Registry must sign an approved informed consent document unless the enrolling physician has obtained a waiver of consent by a controlling IRB.
- (2) Primary Investigators must submit a signed copy of the USIDNET Online Registry User Agreement which addresses rules and regulations for using the Registry. This form can be found at www.USIDNET.org.
- (3) Submit a signed copy of the USIDNET Online Registry Username and Password Application. The Username and Password Application must be signed by each person you wish to grant access to the Registry. The PI must co-sign each application.
- (4) Enrollment
A USIDNET Registry Enrollment Training Guide with screen shots has been developed to walk you step-by-step through the online enrollment process.

Enrollment can be done either directly into the online Registry via www.USIDNET.org under Disease Registry or by filling out the electronic forms (also under Disease Registry) and emailing them to Contact@USIDNET.org.

- Each patient requires a Core form and for patients with a known diagnosis, a Disease-Specific form, if available, should be filled out.
- For patients with no clear diagnosis (or those with diagnoses for which Disease-Specific forms are not available), only the Core form is needed.

Disease-specific forms exist for:

Agammaglobulinemias, X-linked and AR
Chronic Granulomatous Disease
Complement deficiencies
CVID and other Hypogammaglobulinemias
DiGeorge Syndromes

Hyper IgM Syndromes
Leukocyte Adhesion Defect
NEMO and NF- κ B related disorders
Severe Combined Immune Deficiency / CID
Wiskott-Aldrich Syndrome

Additional comments and/or relevant laboratory data can be entered into the MEMO section found on the last page of the Core form. Please fill out the forms as completely as possible. The quality of the data entered determines the quality of the data retrieved.

Filling out the forms:

Core form – used for all patients irrespective of their diagnosis

The Core form has several sections.

- A) The first section collects demographic information on the submitting physician and on the patient.
- B) The next section deals with infections. Here you will be asked whether the infections predated the diagnosis, were observed at some time, or were prominent.
- C) The longest section allows you to indicate systemic or organ specific conditions seen in the patient. Although the section is long, it is usually a simple matter to check the appropriate boxes.
- D) Allergies and vaccine-related adverse events follow and are listed separately.
- E) The next section requests information on treatment modalities including standard therapy, transplant procedures and complementary medicine.
- F) Next is a brief grid asking for your best assessment of the level of function of the patient. There are separate grids for adults and children. This need not be specifically related to the immune deficiency. For example, if the patient is disabled from a malignancy or congenital defect, you would indicate their best level of function taking into account all conditions.
- G) The last section collects basic laboratory information on the patient. Different labs utilize different measures and different normative values. To help make the most of these data, please include normal ranges when requested. The post-pneumococcal vaccine titers request the number of serotypes to which the patient responded that were in the vaccine given and were tested. The protective range is usually given as 1.0 to 1.3 ug/ ml for each serotype.

Disease-specific forms

Because there are approximately 200 specific disorders listed by the IUIS, diseases with relatively similar findings are grouped into a shared data collection form. The development of additional data entry forms is welcomed and should be discussed with the USIDNET Project Manager at Contact@USIDNET.org.

Each disease-specific form requests information on how the diagnosis was established. If you are using the form for a related disease, you may ignore this Diagnostic Criteria section. The subsequent sections are quite brief and request some additional information and laboratory data on specific conditions. There are also brief grids related to disease-specific management.