



## **ESID Online Registry**

### **User Manual**

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## 1 Introduction

Welcome to the ESID Online Registry for Primary Immunodeficiencies. This web-based database system allows you to enter and access patient data in a secure decentralized way through the internet via your own browser. This manual includes a detailed guide to the documentation process.

All pages in the system include detailed descriptions (tool tips and other explanations). Therefore, most of the features should be self-explanatory. With this manual, we mainly want to describe the general documentation process and those features which are probably not completely “intuitive”.

The ESID Registry project is managed at the Center for Chronic Immunodeficiency at University Medical Center Freiburg.

**Current project leader:** Prof. Mikko Seppänen, Chairperson ESID Registry Working Party


**Project coordination:** Dr. Gerhard Kindle

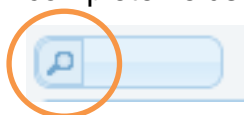
## 2 Working with the Database System

Before we start this tour, we would like to remind you that you are working with real patient data. Therefore, please observe the following user principles:


- every user of the system (= every user name) corresponds to one person only.
- never log into the system with someone else's user name.
- close the session by logging out after finishing your work.
- keep your password in a secure place
- use a secure password
- change your password regularly

## 2.1 General principles

- Please make a selection **for every field** – this is necessary in all forms of the Registry (if not stated otherwise).
- To save the data, you **must** click the “Create” or “Save” button below every form.
- Some centres use a **personalized version** of the system, i.e. they can work with patient names. The additional features of this version are described in 2.9
- Buttons with a question mark:  contain definitions for all fields where necessary.
- The system contains “classic” drop down menus as well as searchable auto-complete fields. The latter are marked by a magnifier like this:

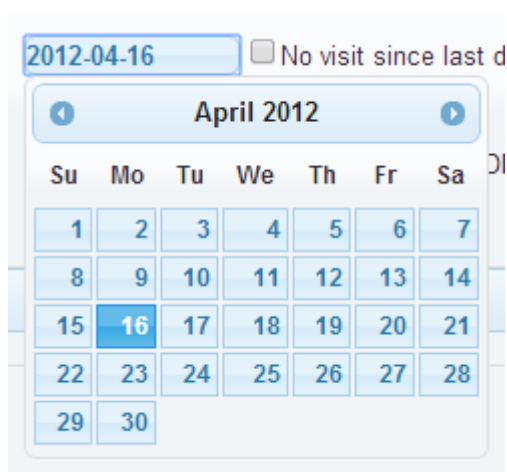


To use this type of field

1. Enter two or more characters
  2. Wait until a list of matching entries is displayed and select one of these
- Some drop down menus – especially in unPAD or APDS forms – have an extra -  button next to them. If you like to add a new entry to the drop down list please click on this button.

If you click on the “send mail” link a new window of your standard email application should open. If not please copy the provided information and send an email manually.

- Date fields:
  - Fields that ask for a complete date contain a date picker that appears upon selection of the field:



- o Date fields that are split into three fields:

Year:  Month:  Day:

Here, it is possible to leave month and day open. Please only enter the date as precisely as known. Do not use dates like 2000-1-1 or 2000-15-6 to signal that month and day are unknown. This only leads to flaws in data quality.

- Some fields only appear after you have selected a specific field, e.g.:

Familial Case  Yes  No  Unknown

Familial Case  Yes  No  Unknown

Index patient

Index patient is the patient's

- If you have not completed a field or entered inconsistent data, messages are displayed on top of the page:

**⚠ The field Sex cannot be empty. Please enter or select a value.**

**⚠ Please enter a value for the field Country of Birth**

**Create Patient**

Patient Consent  Full consent  Research only  Not applicable (deceased)

Date of birth  Year:  Month:

Country of Birth

Sex  Male  Female  Unknown

### 2.1.1 Two types of “unknown”

Since version 1.6, the ESID Registry offers two fields to indicate that information for an item is not available:

Currently unk.  Truly unk.

These have been introduced to enable the documenting centres to differentiate between items that they currently don't have at hand but may be able to enter it later after further investigation, and items where data is really not available no matter how hard they look. Before this was introduced, there was only one option “unknown” which did not offer this possibility. So, please note the following

#### Definitions:

**Currently unk.** This means that you do not have the data at hand at the moment, but if you ask the treating physician or go through the patient files in detail, you may find and enter it later.

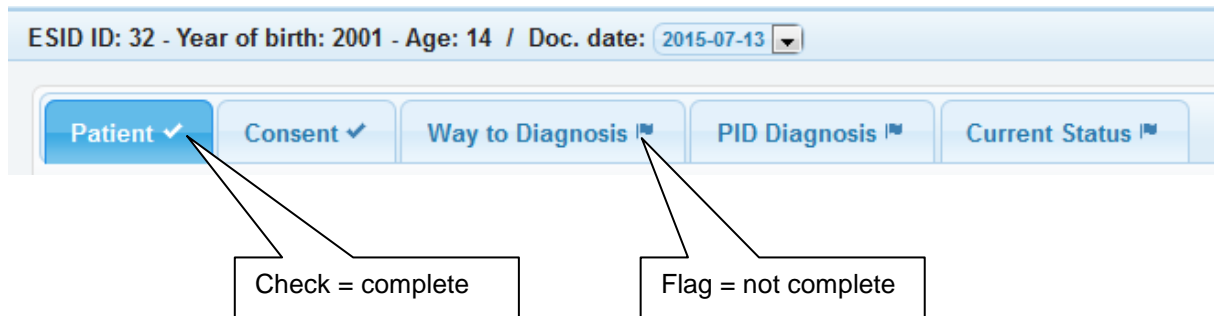
**Truly unk.** This means that data is really not available, and in effect you should waste no more time looking for it.

All previously entered “unknown” items have been converted to “currently unknown” when switching to version 1.6 (March 13, 2015).

## 2.1.2 Complete or not complete?!

To complete a form you have to answer all questions in the form.

The tabs themselves carry checks and flags that signal whether the form(s) has already been completed.



Currently (June 2019) there are three types of datasets part of the ESID-Registry:

- Level 1 (standard forms for every patient)
- Level 2 (unPAD)
- Level 3 (APDS)

The main difference in general is that you must not answer questions with “currently unknown” in unPAD or APDS forms to mark them as complete. In Level 1 forms this takes no effect.

## 2.2 Accessing the system

Enter the following URL in your browser:

<https://cci-esid-reg.uniklinik-freiburg.de/EERS/>

The screenshot shows the login interface of the ESID Registry System. At the top, there is a header with the ESID logo and the text 'The new ESID Registry System'. On the right side of the header, there is a 'HELP' link. The main content area contains a 'Login' form with the following elements: a 'Login' button, a 'Username' input field, a 'Password' input field, a 'Login' button, and a link for 'Forgot your login or password?'.

Enter the login credentials you received from the ESID Registry team.

When you log in for the first time, you will be asked to change your password:

The screenshot shows the 'Create new Password' form. At the top, there is a red error message: 'Sorry, your password has expired. Please create a new one.' Below the error message, there is a 'Create new Password' section. The form contains the following fields and buttons: 'Username' (test.cci), 'Current Password' (with a help icon), 'New Password' (with a help icon and a lock icon), 'Repeat new Password' (with a help icon), 'Generate password' button, 'Save' button, and 'Reset' button. Two callouts are present: one pointing to the lock icon with the text 'Click on this lock to display the password as clear text.' and another pointing to the 'Generate password' button with the text 'Optional: Click this button to let the system generate a password.'

The new password has to have at least

- 1 upper case letter,
- 1 lower case letter,
- 1 digit,
- 1 special character of this list: +\_.,:;=?!

and has to be between 14 and 24 characters long.

In case you have trouble logging in, you find our contact details in the HELP menu (top right).



## 2.3 The main page (patient list)

After logging in, the system shows a list of your patients. This is the view that the system also returns to if you click “Select patient” in the top left corner.

The following columns are displayed:

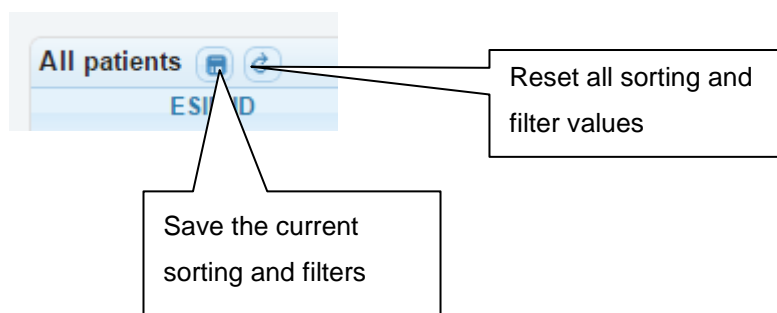
- ESID ID: The unique patient ID
- Year of Birth
- Living Status (alive, deceased...)
- Sex
- PID Diagnosis
- Gene
- Last date of docu: The date of the last documentation timepoint
- Date of last news: The date that the most recent documentation is based on (as entered in form “Current status”)
- Level 1 Complete: “Yes” if level 1 was completed at the last date of docu

Each column can be used to **sort or filter** the list, e.g.:

ESID ID	Year of Birth	Living Status	Sex	PID Diagnosis	Gene	Last Date Of Docu	Date of Last News	Level 1 Complete
30000	2000	All		CVID		2014-06-04		Yes
30926	2003	Deceased		SCID	IL7Ralpha	2014-05-13	2014-05-01	Yes
30927	2004	Lost to follow-up		Shwachman-Diamond-syndr	SBDS	2014-04-30	2012-04-16	No
30928	1998	Discharged after complete recovery	Female	Agammaglobulinemia		2014-04-30	2014-04-22	No
30930	2000	Alive	Female	Combined ID	PNP	2014-04-29	2014-04-01	Yes
30931	2006	Alive	Male	Congenital neutropenia		2014-05-02	2012-06-11	No
30933	2000	Alive	Female	CVID		2014-04-29	2014-04-08	No
30934	1996	Alive	Male	Unclassified IDs		2014-05-05		No
30936	2002	Alive	Male	Combined ID	LIG4	2014-05-05	2014-05-01	Yes
30938	2002	Alive	Female	CVID		2014-05-05	2014-02-05	No
30940	2003	Alive	Female	CVID		2014-06-04	2014-04-01	No
30944	2001	Alive	Male	ALPS		2014-04-29	2014-04-01	No
30952	1994	Alive	Male	CSR / HGM (Hyper-IgM)	UNG	2014-05-02	2014-05-01	No
30955	2001	Alive	Female	Shwachman-Diamond-syndr	SBDS	2014-05-12	2014-05-01	No
30956	1999	Alive	Female	Shwachman-Diamond-syndr	SBDS	2014-05-12		No

### Saving your sorting and filters:

If you would like to save your current filters, so that the list is loaded the same way the next time you call it, you can do so using the buttons on the left top side of the list:



The list can be exported in different formats using the buttons displayed below the list

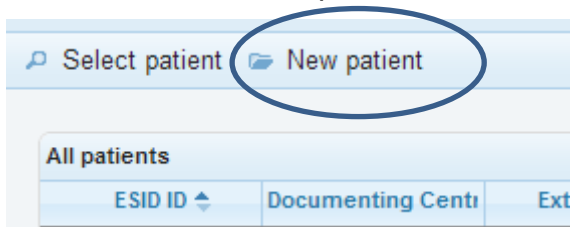


By default, the list shows 15 entries per page. You can set a personal default value of up to 100 entries in your account settings (2.10).

For users who enter data for more than one center, an additional column “Documenting Centre” is visible in the patient list.

## 2.4 Adding a new patient

In order to add a new patient, select “New patient” on the top left of the page:



The following page shows up. Please enter the details for the patient.

Explanations for the fields are available via the “?” buttons.

**Create Patient**

Patient Consent  Full consent  Research only  Not applicable (deceased)

Date of birth  Year:  Month:

Country of Birth   Unknown

Country of current residence   Unknown

Sex  Female  Male  Unknown

Death before initial registration  Please only report deceased patients if this is required by your centre.

Familial Case  No  Yes  Unknown

Twin  No  Yes: identical  Yes: non-identical  Yes, but heredity unknown  Unknown

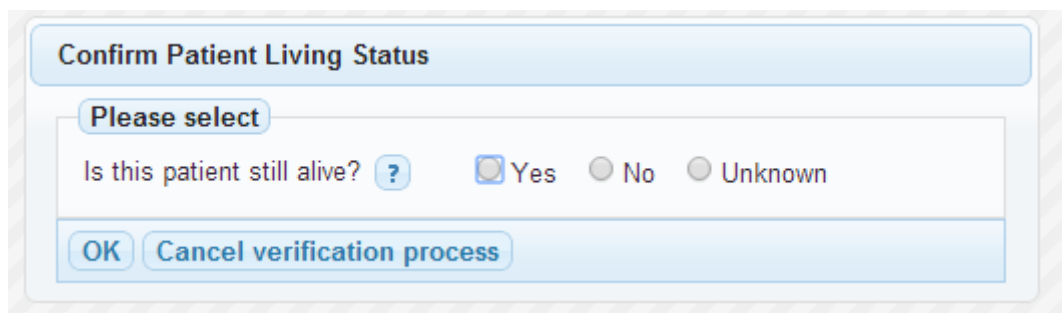
Consanguinity of parents  No  Yes  Probable  Unknown

Documenting Centre  Test Centre

After you have successfully created the patient entry, the standard patient tabs are shown (see 2.6)

## 2.5 Verifying an imported patient

In order to verify an existing entry, click on the entry in the list. You are then asked to confirm that the patient is still alive (This does not apply for deceased patient entries transferred from another registry).



**Confirm Patient Living Status**

Please select

Is this patient still alive? ?  Yes  No  Unknown

OK Cancel verification process

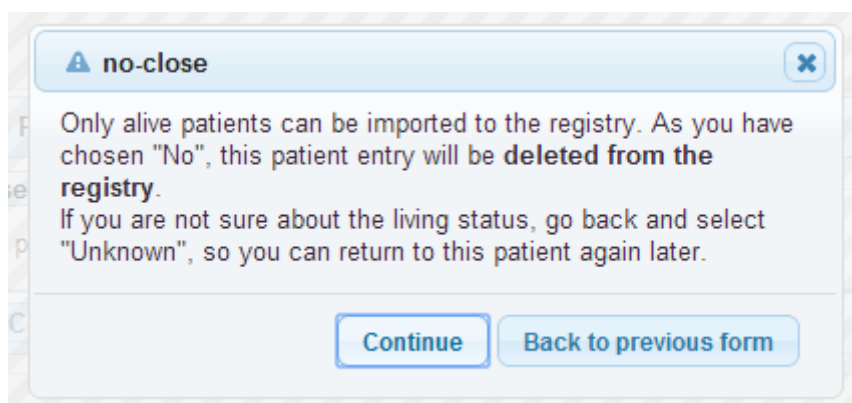
If the patient was still alive at the last time you heard of him, select "Yes". You can enter the time point of last news later on in the "Current status" form (2.6.4). This also means that: If the last time you saw him was a long time ago (e.g. two or three years), but your center still follows this patient, select "Yes".

If the last time you saw him was a long time ago (e.g. more than three years) and your center does NOT follow him anymore, select "No". This is because you should only register patients currently followed by your center.

If you know that the patient is now followed by another center, and this is an ESID Registry Documenting Center, please send us an email with the patient ID. We can arrange a transfer of this entry to the other center then.

If you select "No", the entry will be deleted from the system. This is because the system is not intended for retrospective documentation of deceased patients.

The system asks you for confirmation before the entry is deleted:



**no-close**

Only alive patients can be imported to the registry. As you have chosen "No", this patient entry will be **deleted from the registry**.

If you are not sure about the living status, go back and select "Unknown", so you can return to this patient again later.

Continue Back to previous form

Next, depending on the type of diagnosis, the system asks you to confirm or change the diagnosis, e.g.:

**Confirm clinical criteria**

**Please select**

The clinical criteria for **Autoimmune lymphoproliferative syndrome (ALPS)** are:

At least one of the following:  
 \*splenomegaly  
 \*lymphadenopathy (>3 nodes, >3 months, non-infectious, non-malignant)  
 \*autoimmune cytopenia (>= 2 lineages)  
 \*history of lymphoma  
 \*affected family member

AND at least one of the following:  
 \*TCRab+CD3+CD4-CD8- of CD3+ T cells>6%  
 \*elevated biomarkers (at least 2 of the following):  
 \*\*\*sFASL > 200pg/ml  
 \*\*\*Vitamin B12 > 1500ng/L  
 \*\*\*IL-10 > 20pg/ml  
 \*\*\*impaired FAS mediated apoptosis

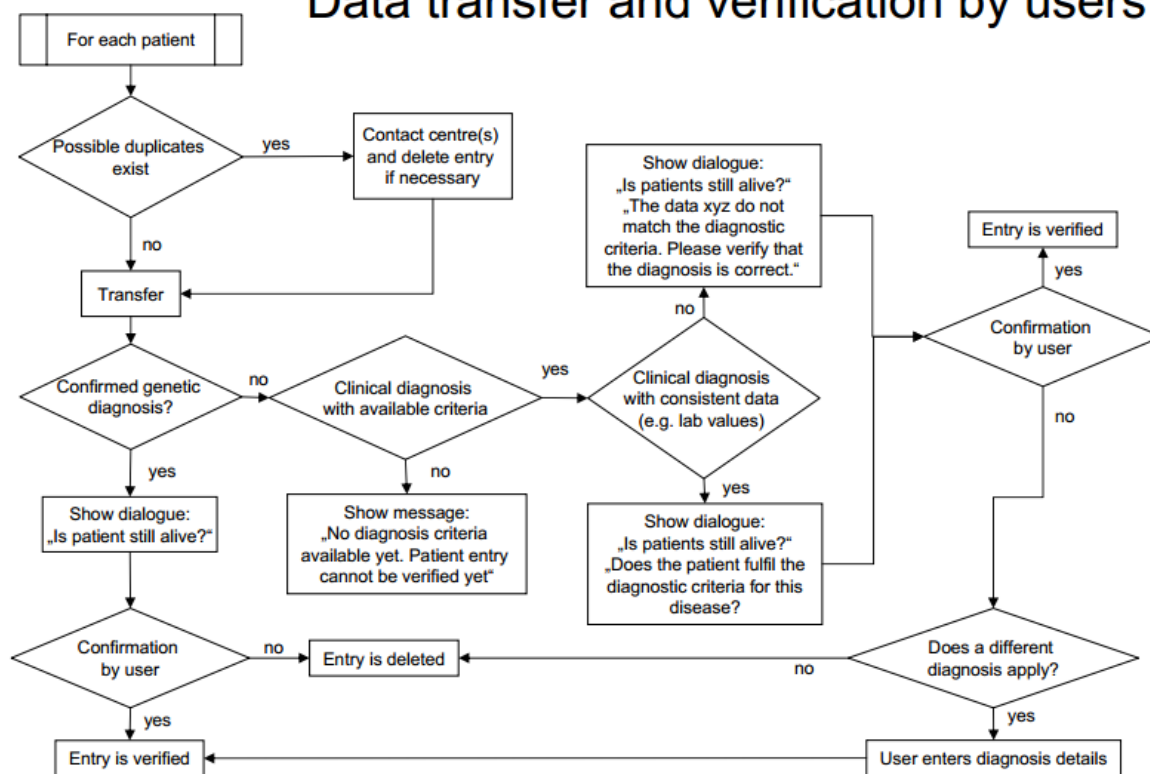
Does the patient fulfil the diagnostic criteria for this disease?

Yes  No  Unknown

For patients with lymphoproliferation and/or autoimmunity who do not fulfil these criteria, please consider the foll...

The underlying process is depicted here:

## Data transfer and verification by users



If you have successfully verified the entry, the patient tabs are shown for further editing (see 2.6).

ESID ID: 31004 - Diagnosis: CGD - Year of birth: 1975 - Age: 40 / Doc. date: 2015-02-12

**i** Patient entry verified. Date of first documentation set to: 2015-02-12

**Patient** ✓ | Way to Diagnosis | PID Diagnosis | Current Status

**Edit Patient**

Patient Consent  ?  Full consent  Research only  Not applicable (deceased)

Date of birth  ? Year: 1975 Month: 6

Country of Birth  ?   Unknown

Country of current residence  ?   Unknown

Sex  Female  Male  Unknown

Familial Case  ?  No  Yes  Unknown

Twin  No  Yes: identical  Yes: non-identical  Yes, but heredity unknown  Unknown

Consanguinity of parents  ?  No  Yes  Probable  Unknown

Documenting Centre Test Centre

## 2.6 The Level 1 tabs

The Level 1 Tabs are the standard view for a patient.

There is a header row that shows the patient's unique ID, the year of birth and current age, as well as the currently selected Documentation date:

ESID ID: 32 - Year of birth: 2001 - Age: 14 / Doc. date: 2015-07-13

**Patient** ✓ | **Consent** ✓ | Way to Diagnosis | PID Diagnosis | Current Status

If you are editing a patient that has been imported, some forms may already contain data, but the tabs are not marked as complete (cf. 2.1.2), because selections are missing for some items.

In order to complete a patient, please select every tab marked by a flag and enter the necessary data.

The Documentation date which is shown on the top right cannot be edited manually. It is the time point when the patient was created (or verified, see 2.5) and inserted automatically by the system.

Some of the tabs (forms) are described in detail in the following chapters.

## 2.6.1 Patient

If you select “Yes” for familial case, you can select the ID of the index patient for this family (if he/she has been registered). The **index patient** is the first patient diagnosed with this immunodeficiency in this family. If you link several patients in this way, you are able to switch between patients belonging to this family by clicking on “show family list”:

The screenshot shows the 'Familial Case' form with the following fields:

- Familial Case:  No  Yes  Unknown
- Index patient:  [show family list](#)
- Index patient is the patient's:
- This is the index patient:

The 'List of relatives' table below shows the following data:

ESID ID	Year of Birth	Status	Sex	Is index	Index rel.	PID	ClinDx year	GeneticDx year
2	1973	Alive	Female	Yes			1995	
3	2010	Alive	Female	No	grandson/-daughter	Agammaglobulinem	2012	2014

Export options: CSV, EXCEL, ODS, PDF, RTF, XML

## 2.6.2 Consent (ICF)

Since version 1.8, the system requests detailed information on the ICF (informed consent form) that the patient has signed. Note that you can add several consent forms for a patient, like here:

The 'Consent List' table shows the following data:

Date of signature	Consent version	Version unknown	Research Option	Pharma Option
2015-07-02	Dutch version: 1.1, 2015-07-0	No	No	Yes
		Yes	Yes	Yes

Export options: CSV, EXCEL, ODS, PDF, RTF, XML

For all registered patients until the end of version 1.7 (2015-07-13), the ICF version has been set to "unknown", so that datasets are still evaluated to complete.

Please note that you can select from all consent forms that have ever been in use at your centre. The consent forms are each distinct and follow this pattern:

Language - Version number - Version date - ethics committee (EC) that approved this form

**If a version is missing**, please send an email to us, we will add it for you.

### 2.6.3 PID Diagnosis

In the PID Diagnosis form, it is possible to enter a diagnosis with known or unknown genetic defect:

- Known genetic defect:

You can either select a clinical diagnosis (“Current PID diagnosis”), and the possible genes for this diagnosis are displayed:

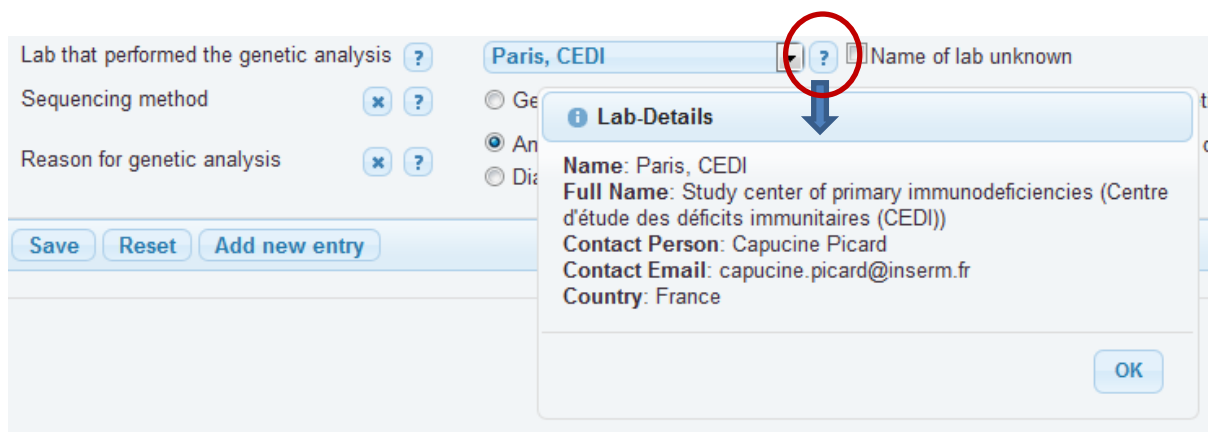
OR you can first enter the gene and then select a suitable PID diagnosis:

- Unknown genetic defect:

Use the “Current PID Diagnosis” field to select a PID diagnosis. Next, you will be asked to confirm the clinical diagnosis criteria (as described in the verification process (2.5)).

### Genetic laboratories:

There is a field that asks for the name of the “Lab that performed the genetic analysis”. You can select one of the names from the list and see the details for the lab by clicking on the question mark if you want to make sure that this entry is correct:



The screenshot shows a web form with the following elements:

- Lab that performed the genetic analysis**: A dropdown menu with "Paris, CEDI" selected. A question mark icon next to the dropdown is circled in red. A blue arrow points from this icon to a pop-up window.
- Sequencing method**: A field with a red 'x' and a question mark icon.
- Reason for genetic analysis**: A field with a red 'x' and a question mark icon.
- Buttons**: "Save", "Reset", and "Add new entry".
- Lab-Details Pop-up**:
  - Name**: Paris, CEDI
  - Full Name**: Study center of primary immunodeficiencies (Centre d'étude des déficits immunitaires (CEDI))
  - Contact Person**: Capucine Picard
  - Contact Email**: capucine.picard@inserm.fr
  - Country**: France
  - OK** button



## 2.6.4 Current status

Patient, Way to Diagnosis and PID Diagnosis only have to be completed once (at the first documentation = baseline documentation). The Current Status form has to be completed at every documentation timepoint.

This is what the Current Status form looks like at baseline:

Please note the following:

- You must enter a “date of last visit” of the patient at your centre or the “date of last news” from the patient. This is important to know which timepoint the reported data refers to.
- If you select “Ig replacement”, an additional tab (form) appears.
- The same is valid for HSCT and Gene therapy.

Additional forms

## 2.6.5 Ig replacement

The form for immunoglobulin replacement includes many automatic checks as well as calculations in order to ensure that data is entered complete and correct.

Some of these features are described here:

- Only appropriate routes are shown for each brand:

Current brand name   Unknown  
 Current route of administration  Intravenous  Subcutaneous  Intramuscular

Current brand name  **Hizentra / CSL Behring**  
 Current route of administration  Subcutaneous  Intramuscular

Current brand name  **Endobulin / Grifols**  
 Current route of administration  Intravenous

- Automatic calculation of dose:

Enter mg/kg OR absolute dose OR BOTH

This read-only value is calculated from dose & interval & weight

The interval can be entered in two different formats – please select one of these for each entry:

Patient's current weight  kg  Unknown  
 Current dose  mg/kg body weight Dose per month:   
 Current dose (total amount)   grams  milliliters  
 Dose unknown   
 Interval for this dose  Select interval-type first:  every x weeks or days  x time(s) per week, month or year  
 Interval unknown

every   weeks  days  
 time(s) per  week  month  year

Example of automatic calculation:

The weight is needed to calculate the mg/kg dose

Patient's current weight	<input type="text" value=""/>	kg	<input type="checkbox"/> Unknown
Current dose	<input type="text" value=""/>	mg/kg body weight	Dose per month: <input type="text" value=""/>
Current dose (total amount)	<input type="text" value="10.3"/>	<input checked="" type="radio"/> grams	<input type="radio"/> milliliters
Dose unknown	<input type="checkbox"/>		
Interval for this dose	Select interval-type first: <input checked="" type="radio"/> every x weeks or days		
	<input type="text" value="5"/>	<input checked="" type="radio"/> weeks	<input type="radio"/> days

Patient's current weight	<input type="text" value="35"/>	kg	<input type="checkbox"/> Unknown
Current dose	<input type="text" value="294"/>	mg/kg body weight	Dose per month: <input type="text" value="235"/>
Current dose (total amount)	<input type="text" value="10.3"/>	<input checked="" type="radio"/> grams	<input type="radio"/> milliliters

These two values are calculated and stored by the system

If you enter BOTH mg/kg dose and absolute dose, the system calculates whether the two match. If not, a dialog box is shown as follows:

<input type="text" value="300"/>	mg/kg body
<input type="text" value="100"/>	grams
every <input type="text" value="5"/>	weeks
<input type="checkbox"/>	
<input type="text" value="35"/>	kg <input type="checkbox"/> Unknown

no-close
✕

The absolute dose does not match the dose in mg/kg. The calculated value is 143 mg/kg

Take calculated and save
Take yours and save
Return to form

If you enter the dose in milliliters, the brand name is needed for the calculation, because each brand has a specific IgG concentration:

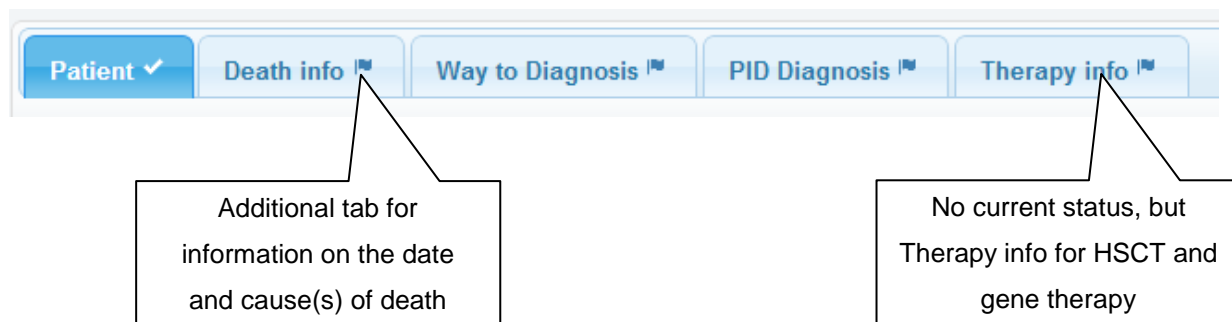
Current brand name	<input type="text" value="Subcuvia / Baxter"/>	<input type="checkbox"/> Unknown
Current route of administration	<input type="radio"/> Subcutaneous <input type="radio"/> Intramuscular <input type="radio"/> Unknown	
Current place of administration	<input type="radio"/> Home <input type="radio"/> Hospital <input type="radio"/> Hospital: inpatient <input type="radio"/> Hospital: outpatient	
Patient's current weight	<input type="text" value="35"/>	kg <input type="checkbox"/> Unknown
Current dose	<input type="text" value="294"/>	mg/kg body weight    Dose per month: <input type="text" value="365"/>
Current dose (total amount)	<input type="text" value="100"/>	<input type="radio"/> grams <input checked="" type="radio"/> milliliters

## 2.7 Deceased patients

If you are

- a) working on an imported entry of a deceased patient or
- b) have selected “Death before initial registration” for a new patient,

the Patient tabs look slightly different:

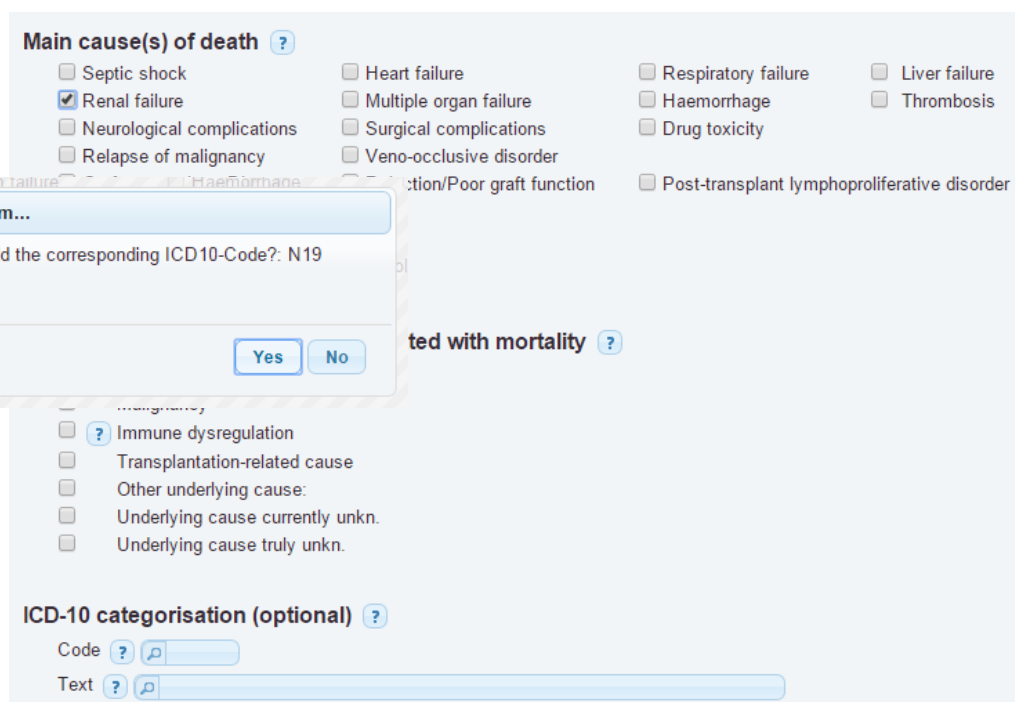


Please note that no follow-ups can be entered for deceased patients. Previous entries for Ig replacement remain stored. Data on HSCT and gene therapy can still be entered.

### 2.7.1 ICD10 coding death causes

The “Death info” tab contains an additional section for ICD10 codes. This section is – in contrast to all other items in the Registry – optional. It was added because some centres need this categorization for internal purposes.

In order to simplify the adding of ICD10 codes, corresponding codes for the “Main cause of death” are suggested by the system when one of these options is selected.



If confirmed, the respective entry is added to the ICD10 section:

Underlying cause currently unkn.  
 Underlying cause truly unkn.

**ICD-10 categorisation (optional)**

ICD10 Code List - Click on a row to edit data or add a new entry: [Add New](#)

ICD10 Code	ICD10 Text
N19	Unspecified kidney failure

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If an option is de-selected, the system will ask if the corresponding ICD10 entry shall be removed:

**Please Confirm...**

Do you want to remove the responding ICD10-Code?: N19

Yes No

Of course, you can also add and edit ICD10 codes manually. You can also delete them by selecting an entry and then click the “Delete” button:

**Edit ICD10**

**Edit ICD10 Code of Death Cause**

Code ?

Text ?

Save Reset **Delete**

## 2.8 Follow-up documentation

If you have completed a documentation timepoint for a patient, it is possible to add a follow-up documentation. The system will start sending email reminders if a patient has not been updated for more than one year (365 days).

In any case, you can enter follow-ups as often as you like (but only one per day).

Please note though that only one documentation timepoint is possible per day. This also implies that you cannot enter baseline data and follow-up data on the same day, because the documentation date is automatically generated and stored by the system.

This is an example for a complete documentation timepoint:

- All tabs have a check sign
- “+ Follow up” is displayed next to the current documentation date

ESID ID: 30936 - Year of birth: 2002 - Age: 12 / Documentation date: 2014-04-29 + Follow up

Patient ✓ Way to Diagnosis ✓ PID Diagnosis ✓ Current Status ✓ Ig-Replacement ✓ HSCT ✓ Gene therapy ✓

The following dialog box is shown:

**Add Follow Up** [X]

Add follow up documentation for this patient?

OK Cancel

Next, the patient tabs are shown, and the Current Status form is selected:

Patient ✓ Way to Diagnosis ✓ PID Diagnosis ✓ **Current Status** HSCT ✓ Gene therapy ✓

**Create Current Status**

Date of last news from patient ? [ ]  No news from patient since last documentation

This is the date of the ?  last clinical visit  last news (phone, letter etc.)  unknown

Current status ?  Alive  Deceased  Lost to follow-up  Discharged after complete recover

Changes to diagnosis ?  No Change  PID diagnosis has changed  No PID after all

Current Ig replacement ?  Yes  No  Unknown

HSCT ?  Yes  No  Unknown SCETIDE ID ? dfw3 EBMT

Gene therapy ?  Yes  No  Unknown

Save Reset

As you can see, all other tabs do NOT have to be documented again, except for Ig replacement. Only the following items require documentation:

Date of last news from patient

(or alternatively the date of last news from the patient)

Current status (the previous entry is selected):

Alive  Deceased  Lost to follow-up  Discharged after complete recovery

Changes to diagnosis (No change is pre-selected)

No Change  PID diagnosis has changed  No PID after all

Therapy items:

Current Ig replacement	<input type="radio"/> Yes	<input type="radio"/> No	<input type="radio"/> Unknown
Haematopoietic stem cell transplantation (HSCT)	<input type="radio"/> Yes	<input checked="" type="radio"/> No	<input type="radio"/> Unknown
Gene therapy	<input checked="" type="radio"/> Yes	<input type="radio"/> No	<input type="radio"/> Unknown

You need to change HSCT and gene therapy only if it was “No” or “Unknown” previously, because the previous selection is already displayed (so “Yes” remains “Yes” and does not have to be selected again at every documentation).

Please also note that all HSCT and gene therapies are only entered once and can be edited at every documentation timepoint.

### 2.8.1 Changing the PID diagnosis

If you have found a genetic mutation in your patient, you can simply enter this into the PID diagnosis. However, if the **clinical diagnosis (“PID Diagnosis”)** has changed, this has to be documented as a new diagnosis entry, in order to preserve the previous data:

The screenshot shows the 'Current Pid Diagnosis' section of the ESID Online Registry. It includes fields for 'Current PID diagnosis' (set to 'Unclassified synd'), 'Affected gene', 'Additional genes', 'Genetic tests performed?' (set to 'Yes, but no mutatio'), and 'Lab that performed the genetic analysis' (set to 'Newcastle'). At the bottom, there are three buttons: 'Save', 'Reset', and 'Add new entry'. The 'Add new entry' button is circled in orange, indicating that a new diagnosis entry should be added when the clinical diagnosis changes.

**New Pid Diagnosis**

**Create PID Diagnosis**

Current PID diagnosis

Affected gene

Additional genes

Genetic tests performed?   Yes, but no mutation found  Not genetically tested  Result pending  History of genetic tests unknown

Previous diagnosis entries are displayed as a list below the current diagnosis:

**Current Pid Diagnosis**

Current PID diagnosis

Affected gene

Additional genes

Date of genetic diagnosis Year:  Month:  Day:   Date unknown

Reason for genetic analysis   Analysis following clinical diagnosis  Family screening  Prenatal diagnosis  
 Diagnosis by neonatal screening  Unknown

Sequencing method   Gene sequencing  Whole exome/genome sequencing  Unknown

Lab that performed the genetic analysis   Name of lab unknown

**Pid Diagnosis-List of former diagnoses**

Current PID diagnosis	Affected gene	Additional genes	Diagnosis valid from	Diagnosis valid until
Unclassified syndromic immunodeficiencies			2014-04-28	2014-06-04

Page 1 of 1 | 15 | View 1 - 1 of 1

Export:

Documentation timepoint when the former diagnosis was entered

Documentation timepoint when a new diagnosis was entered instead



## 2.8.2 Loading previous data for Ig replacement

In order to minimize the documentation burden, you can load the Ig replacement data from the previous documentation timepoint into the form:

The screenshot shows the 'Create Ig-Replacement' form with the following fields and options:

- Current brand name: [?]
- Current route of administration:  Subcut
- Current place of administration: [?],  Home
- Patient's current weight: [?] kg
- Current dose: [?]
- Current dose (total amount): [x] [?]
- Dose unknown: [?],
- Interval for this dose: [?], Select inte
- Interval unknown: [?],
- Current side effects: [?],  Yes

At the bottom, there are three buttons: 'Save', 'Reset', and 'Load previous data'. The 'Load previous data' button is circled in orange.



The screenshot shows the 'Create Ig-Replacement' form with data loaded from 2014-08-13. The fields are populated as follows:

- Current brand name: [?] Flebogamma 10% / Grifols  Unknown
- Current route of administration:  Intravenous  Unknown
- Current place of administration: [?],  Home  Hospital  Hospital: inpatient  Hospital: out
- Patient's current weight: [?] 55 kg  Unknown
- Current dose: [?] 91 mg/kg body weight Dose per month: 1274 [?]
- Current dose (total amount): [x] [?] 5  grams  milliliters
- Dose unknown: [?],
- Interval for this dose: [?], Select interval-type first:  every x weeks or days  x time(s)
- every [x] 2  weeks  days

Please also note that the date of the (very) first Ig replacement is only requested at the baseline documentation:

The screenshot shows the 'Date of first Ig replacement' field with the following options:

- Date of first Ig replacement: [?], Year: [ ], Month: [ ], Day: [ ],  Date unknown

## 2.9 Personalized version

If your centre uses the personalized version, i.e. patient names are included, there are additional features.

### 2.9.1 Patient list with names

In the patient list, you can display patients' names. You can enable this feature by clicking on "Show patient names":



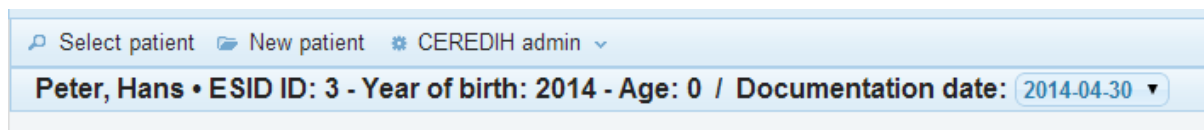
The screenshot shows a web interface for a patient list. At the top, there is a button labeled "Show patient names" which is highlighted with a blue arrow. Below this is a table with the following columns: ESID ID, Documenting Centre, Year of Birth, Living Status, and a fifth column for gender. The table contains several rows of patient data, including ESID IDs like 30000, 30926, 30927, 30928, 30930, 30931, and 30932, all from "Test Centre".

ESID ID	Documenting Centre	Year of Birth	Living Status	
<input type="text"/> x	Test <input type="text"/> x	<input type="text"/> x	All <input type="text"/> x	All
30000	Test Centre	2000	Alive	Female
30926	Test Centre	2003	Alive	Female
30927	Test Centre	2004	Alive	Male
30928	Test Centre	1998	Alive	Female
30930	Test Centre	2000	Alive	Female
30931	Test Centre	2006	Alive	Male
30932	Test Centre	2000	Alive	Female

Please note that if your centre has a large number of patients, it may take up to 15 seconds to load the patients' names because the personal patient data is stored on another server and loaded into the page.

Therefore, you can switch between "Show patient names" and "Hide patient names" as you like. You can also set a default value for this feature in the "Account settings" (2.7).

The patient's name is always displayed on top of the page, independent of the patient list setting:



The screenshot shows a header bar with navigation links: "Select patient", "New patient", and "CEREDIH admin". Below this, the patient's name "Peter, Hans" is displayed along with "ESID ID: 3 - Year of birth: 2014 - Age: 0 / Documentation date: 2014-04-30".

Select patient	New patient	CEREDIH admin
Peter, Hans • ESID ID: 3 - Year of birth: 2014 - Age: 0 / Documentation date: 2014-04-30		

#### 2.9.1.1 Improving the loading time

In your account settings (see 2.10) you can define that your complete patient list will be saved locally on your computer. Note that first name, last name and complete birth date will NOT be saved, but this caching method still reduces the loading time for the patient list with names considerably, especially if you have more than 1000 patient entries. Please also note that after you have activated this feature, the first time that

you load the patient list, it will not be faster, because then the cache is being generated. On all subsequent loads, the loading time should be faster.

### 2.9.1.2 Downloading the patient list with names

You can also **download** the patient list with names, using the button below the list:



Currently, only CSV format is available. Depending on your browser, you will need to save and rename the downloaded file to a .csv file. In order to open it with Excel, please use the Excel text import function.

Please make sure to set the CHARSET in the Excel import settings to UTF-8, otherwise special characters like Umlauts will not be displayed correctly.

### 2.9.1.3 Sorting and filters

Please note that the sorting and filters that you set and save on the patient list WITH names is NOT visible in the patient list WITHOUT names, and vice versa. This means that in fact, you can save two sets of filters.

## 2.9.2 Create a new patient

If you create a new patient, the system redirects you to the other server in order to enter the patient name etc. (screenshots next page).

This other server has two purposes:

1. It stores the patient name, complete date of birth and place of residence so this data can be loaded into the ESID Registry
2. It performs a so called “record linkage” and thereby prevents reporting of duplicate entries.

For the second task, it is important that you really enter as many details as possible, and not only the patient names.

## Enter personal data for a new patient

**Information**

This web application manages and stores identifying patient data (IDAT) for the ESID Registry, consisting of the fields below. It uses the data entered in these fields to **find possible matches** in the current list of entries. After you have submitted the data below, your web browser will **return to the main server pages**. In case of a direct match, you will be redirected to the matching entry, in case this patient belongs to your centre. Please make sure to read the messages displayed after returning to the main server.

- If first or last name consist of multiple components, enter all components, separated by spaces or hyphens, in the appropriate field.
- In case of compound names, check if they are written as one word (like "Annalena") or separated ("Anna-Lena").
- Enter the birth name only if it differs from the last name.
- The fields marked with \* are mandatory.
- Currently, the following (non-ASCII) special characters are supported: ` ç í à á é è ê ó ô õ ñ ã Ä Ö Ö Ü ß

**Personal data**

First name(s):  \*

Last name :  \*

Birth name :  \* (if different)

Date of birth :  \*

City of residence :  (e.g. The Eyrie)

(Postal code / City)

Please also enter zip code and place of residence as well as maiden name (if available) in order to make your entry duplicate-proof.

This service is operated by: ESID Online Registry coordination, CCI, University Medical Center Freiburg, Tel. +49 7141 271-1000  
Mainzeiliste Copyright © 2013-2015 Martin Lablans, Andreas Borg and Frank Ückert. Licensed under the GNU Affero General Public License (AGPLv3+). More

After you have added the data, select “Add patient”, and the system will take you back to the main server.

If the patient has not been registered before, there will be a message in the top row saying:

**i** Please create this patient now by filling in and saving the form below.

**Create Patient**

Patient Consent  ?  Full consent  Research only

Date of birth  ? Year:  Month:

Country of Birth

If the patient exists and has been registered at your centre, the message will say:

**i** This patient already exists. You have been redirected to his or her entry.

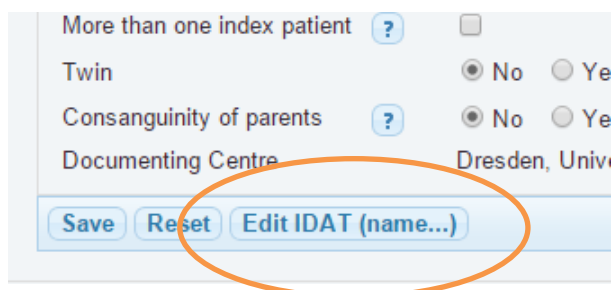
If the entry belongs to **another** centre, you are redirected to your patient list, and there is a **message** that tells you the name of the center that this patient belongs to.

### 2.9.3 Edit personal information

Sometimes, you realize that part of the personal information is incorrect, e.g.

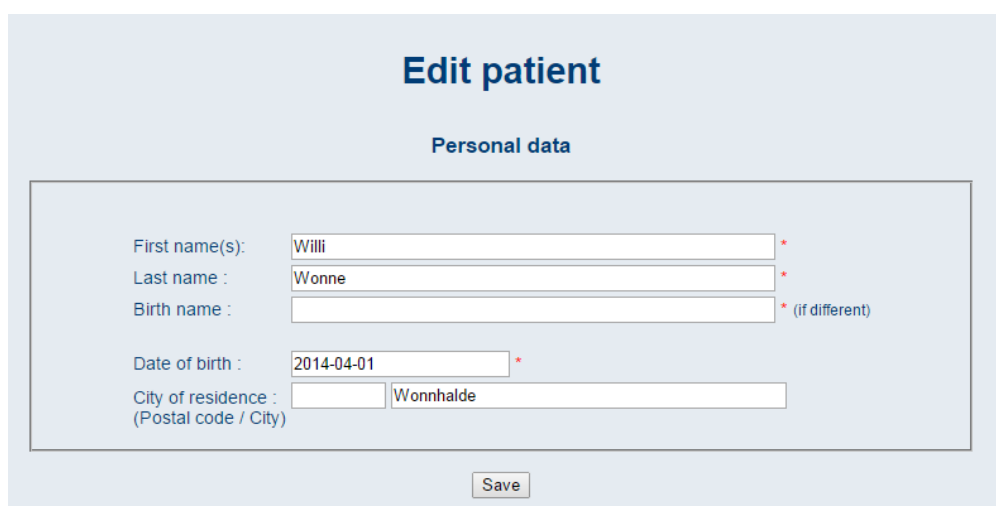
- The patient has married and changed his/her family name
- There is a typo in the entry

You can then edit the personal information by selecting the patient and clicking on the “Edit IDAT<sup>1</sup> (name...)” button in the Patient tab:



More than one index patient  ?  
Twin  No  Yes  
Consanguinity of parents  No  Yes ?  
Documenting Centre Dresden, Univ

This results in a redirect to the other server, where you can edit all of the data:



**Edit patient**

Personal data

First name(s):  \*

Last name:  \*

Birth name:  \* (if different)

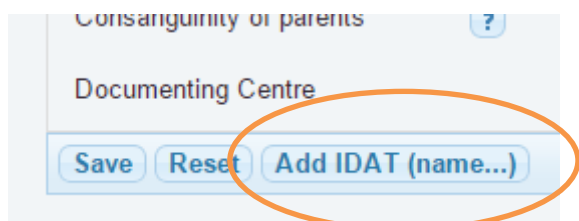
Date of birth:  \*

City of residence:  (Postal code / City)

When you are finished, “Save” takes you back to the standard view (Patient tabs).

### 2.9.4 Add personal information to existing patient

It may happen that some of your patients have been registered without names, e.g. because you recently switched to the personalized version. In these cases, you can add the IDAT by clicking on the following button in the “Patient” tab:

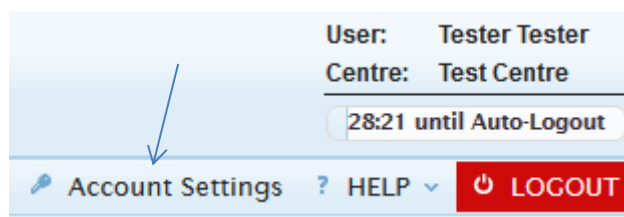


Consanguinity of parents  ?  
Documenting Centre

<sup>1</sup> „IDAT“ is short for the technical term „identifying data“

## 2.10 Account settings

You can edit your account and change your password by clicking the link on the top right of the screen:



**Edit user data**

Username	user
Change Password	<a href="#">Change Password</a>
Centre	Test Centre
Title	<input type="text"/>
First name	<input type="text" value="Tester"/>
Last name	<input type="text" value="Tester"/>
Gender (sex)	<input type="radio"/> Female <input checked="" type="radio"/> Male <input type="radio"/> Currently unk. <input type="radio"/> Truly unk.
Profession	<input type="text"/>
Email	<input type="text" value="esid-registry@uniklinik-frankfurt.de"/>
Alternative email address	<input type="text"/>
Telephone	<input type="text"/>
Fax number	<input type="text"/>
Address	<input type="text"/>
Postal code	<input type="text"/>
Town	<input type="text"/>
Country of living	<input type="text" value="Netherlands"/>
Affiliation (for publications)	<input type="text"/>
Display names in list	<input type="checkbox"/> <a href="#">?</a>
Cache patient list locally	<input type="checkbox"/> <a href="#">?</a>
Patient List size	<input type="text" value="15"/> <a href="#">?</a>
Language	<input type="text" value="English"/> <a href="#">?</a>
App Theme	<input type="text" value="Theme: cupertino"/>
Display animation in info dialogs	<input type="checkbox"/> <a href="#">?</a>
Hover-Effect in forms	<input type="checkbox"/> <a href="#">?</a>

These contact details are used by the ESID Registry team to contact you e.g. send you a new password, invite you to join a study etc.

If you use the personalized version, you can set the default patient list behavior here, as well as the caching method (see 2.9.1.1)

Define a standard number of patient entries per page in the patient list.

The so called "Hover-Effect" may increase the user experience in filling out forms. Give it a try. Test it with different "App Themes" until you've found your favorite.

## 2.11 Level 2 Documentation

Some PIDs are prepared for entering a so called level 2 dataset. These are additional forms that have to be documented for every documentation date.

Documentation start for level 2 is always the currently last documentation date.

You will be asked by a pop-up if the patient exists already:

**Confirm level 2 documentation**

Please select

Do you want to add level 2 documentation for this patient? ?

Level 2 documentation is necessary for the unPAD study which currently covers the PIDs:  
 "Deficiency of specific IgG (Specific antibody deficiency - SPAD)",  
 "IgA with IgG subclass deficiency",  
 "Isolated IgG subclass deficiency",  
 "Selective IgM deficiency",  
 "Selective IgA deficiency",  
 "Unclassified antibody deficiency"  
 AND "Common variable immunodeficiency (CVID)".  
 Please refer to the registry study section on [www.esid.org](http://www.esid.org) for further information.

Yes  No  Ask again later

OK

Zurich, IZZ

Or if you create the first PID Diagnosis for a new patient:

**Create PID Diagnosis**

Current PID diagnosis ?

Affected gene ?

Additional genes ?

Genetic tests performed? x ?  Yes, but no mutation found  Not genetically tested  Result pending  
 Currently unk.  History of genetic tests truly unk.

**Do you want to add level 2 documentation? ?**  Yes  No  Ask again later

This decision can be made on a patient level, i.e. you can decide for every patient, if you will provide L2 data. Please be aware that you need to add Level 2 data for this patient for all subsequent documentation dates as well, if you choose yes.

If you are uncertain at the moment about the participation you might want to choose the option 'Ask again later'. You will be asked again when you access the dataset next time.

## 2.12 Entering lab values

You can set all date-field values like the first one by clicking on this button.

If configured you can switch all values from percent to absolute and vice versa.

You can convert values by clicking on this button.

Lab-values are grouped in lab-panels where every panel has to be saved separately.

If one lab-value depends on another the relation will be automatically calculated.

The screenshot shows a form titled "Create Latest Blood Count" with a table of lab values. The table has columns for Name, Date of Sample, Value, calculated, and Value not available. The rows include Leukocytes, Lymphocytes, CD3+, CD4+, CD8+, CD19+, and CD 56+/CD16+. Below the table are radio buttons for "Data obtained in the absence of immunosuppressive therapy?" and a "Save" button.

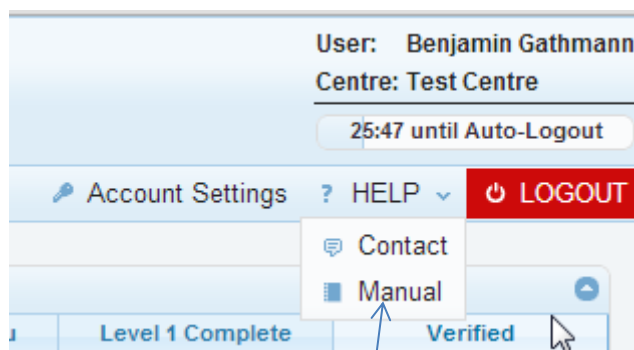
Name	Date of Sample	Value	calculated	Value not available
Leukocytes	2016-07-10	= 33 10 <sup>9</sup> /L		
Lymphocytes	2016-07-10	= 16 10 <sup>9</sup> /L	48.48 %	
CD3+	2016-07-10	= 10 <sup>9</sup> /L		
CD4+	2016-07-10	= 10 <sup>9</sup> /L		
CD8+	2016-07-10	= 10 <sup>9</sup> /L		
CD19+	2016-07-10	= 10 <sup>9</sup> /L		
CD 56+/CD16+	2016-07-10	= 10 <sup>9</sup> /L		

If a unit for value conversion does not exist, please ask your administrator to add it.  
 The calculated values are not saved in the database. They are shown for informative use only.



### 3 Contacting the Registry administrator

For contacting us, you can either send an email or call us (details see below) – or you can use the contact form inside the Registry which is available in the “HELP” menu:



As you can see, this manual itself is also available in the “HELP” section.

Use the contact-form below or contact us using the contact details below.

**Get in contact**

Topic

Subject

Your message

Upload attachment (max. 2MB)

---

You can reach us by email: [esid-registry@uniklinik-freiburg.de](mailto:esid-registry@uniklinik-freiburg.de)

---

or by telephone: +49 (0)761 270-36961

---

or by FAX: +49 (0)761 270-36960

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Contact address: Gerhard Kindle  
 Center for Chronic Immunodeficiency - CCI  
 Breisacher Strasse 115  
 D-79106 Freiburg  
 Germany

#### Contact details:

University Medical Center Freiburg  
 Center for Chronic Immunodeficiency (CCI)  
 Breisacher Strasse 115  
 79106 Freiburg; Germany  
 Tel.: +49-761-270-36961  
 Email: [esid-registry@uniklinik-freiburg.de](mailto:esid-registry@uniklinik-freiburg.de)