



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

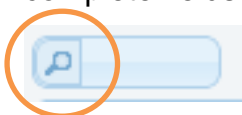
2 Working with the Database System

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:

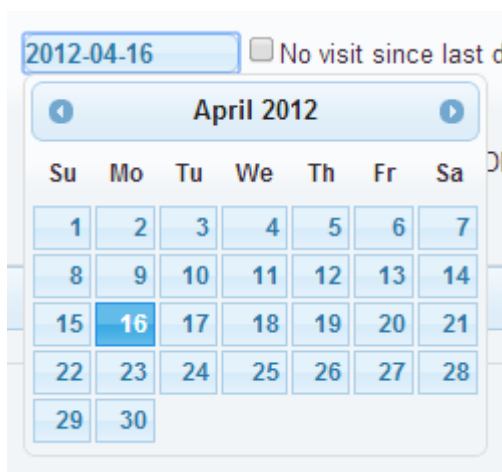


To use this type of field

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:



- You can also enter the date manually in those date picker fields.
You then must use the format: YEAR(4-digits)-MONTH(2-digits)-DAY(2-digits) – delimited by “-” (hyphen) – just like in the screenshot above.
This format is also described as “YYYY-MM-DD” (ISO 8601).
- 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.

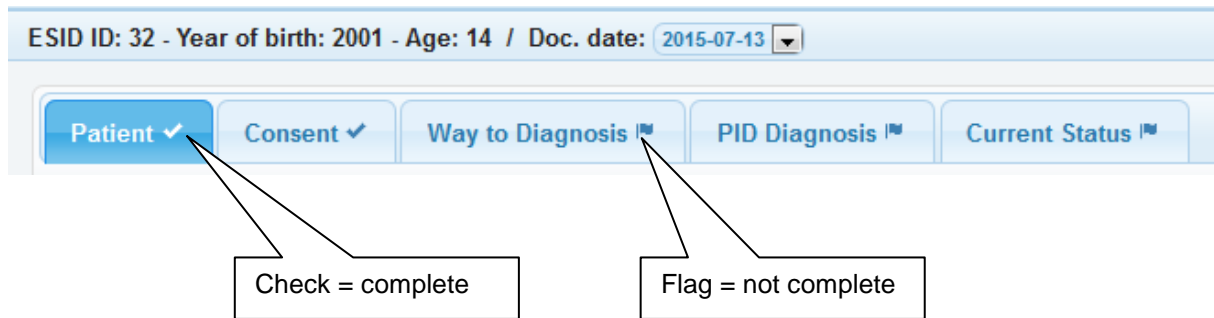
☐ **Truly unk.** This means that data is really not available, and in effect you should waste no more time looking for it.

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/>

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 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).

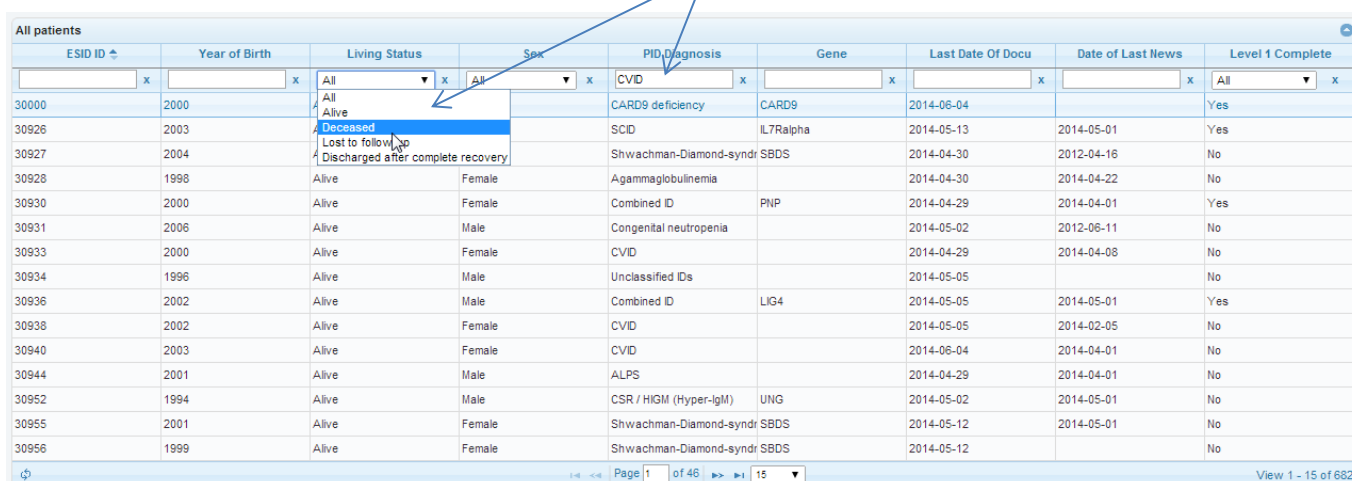
2.3 The main page (patient list)

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

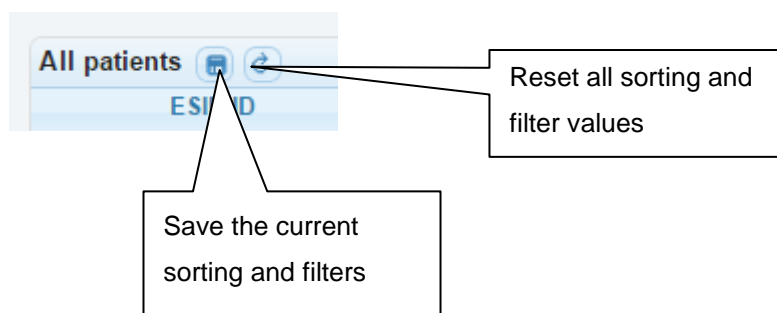
Each column can be used to **sort or filter** the list, e.g.:



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	All	CVID		2014-06-04		Yes
30926	2003	Alive		SCID	IL7Ra	2014-05-13	2014-05-01	Yes
30927	2004	Deceased		Shwachman-Diamond-syndr	SBDS	2014-04-30	2012-04-16	No
30928	1998	Lost to follow-up		Agammaglobulinemia		2014-04-30	2014-04-22	No
30930	2000	Discharged after complete recovery	Female	Combined ID	PNP	2014-04-29	2014-04-01	Yes
30931	2006	Alive	Male	Congential 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 / HIGM (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

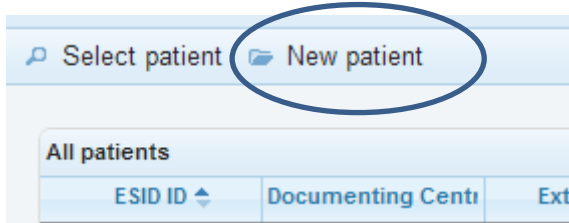


By default, the list shows 15 entries per page. You can set a personal default value of up to 100 entries in your account settings (2.10).

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.

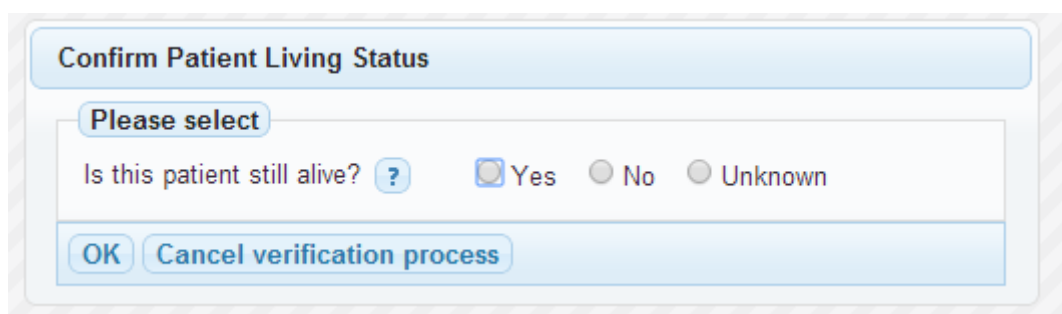
Explanations for the fields are available via the “?” buttons.

 A screenshot of the 'Create Patient' form. It contains various fields for patient information, each with a question mark icon for help. The fields include: Patient Consent (radio buttons for Full consent, Research only, Not applicable (deceased)), Date of birth (Year and Month dropdowns), Country of Birth (dropdown with Unknown checkbox), Country of current residence (dropdown with Unknown checkbox), Sex (radio buttons for Female, Male, Unknown), Death before initial registration (checkbox), Familial Case (radio buttons for No, Yes, Unknown), Twin (radio buttons for No, Yes: identical, Yes: non-identical, Yes, but heredity unknown, Unknown), Consanguinity of parents (radio buttons for No, Yes, Probable, Unknown), and Documenting Centre (Text input). A callout box points to the 'Not applicable (deceased)' option with the text: 'Please only report deceased patients if this is required by your 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).



A dialog box titled "Confirm Patient Living Status". It contains a "Please select" label, a question "Is this patient still alive?" with a question mark icon, and three radio buttons labeled "Yes", "No", and "Unknown". The "Yes" radio button is selected. At the bottom, there are two buttons: "OK" and "Cancel verification process".

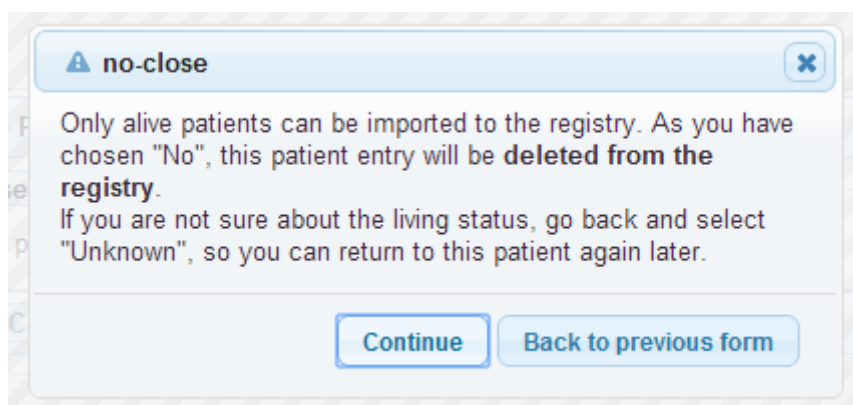
If the patient was still alive at the last time you heard of him, select "Yes". You can enter the time point of last news later on in the "Current status" form (2.6.4). This also means that: If the last time you saw him was a long time ago (e.g. two or three years), but your center still follows this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



A warning dialog box with a title bar that says "no-close" and a close button. The text inside reads: "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." At the bottom, there are two buttons: "Continue" and "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

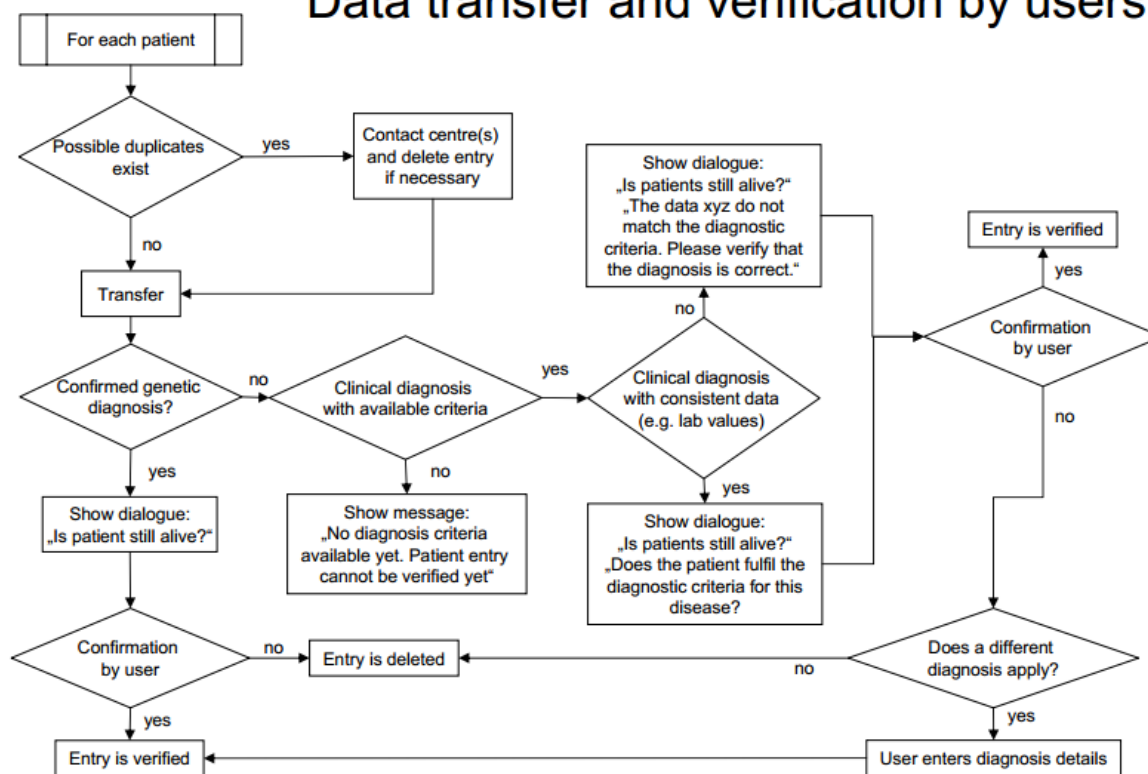
Does the patient fulfil the diagnostic criteria for this disease?

☒ Yes ☐ No ☐ Unknown

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

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

Save **Reset**

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:** Radio buttons for No, Yes (selected), and Unknown.
- Index patient:** A dropdown menu showing the value '2'.
- Index patient is the patient's:** A dropdown menu showing the value 'grandson/-daughter'.
- This is the index patient:** A checkbox.

A red circle highlights the 'show family list' button. An arrow points from this button to the 'List of relatives' table below.

List of relatives

Click on a row to switch to this patient

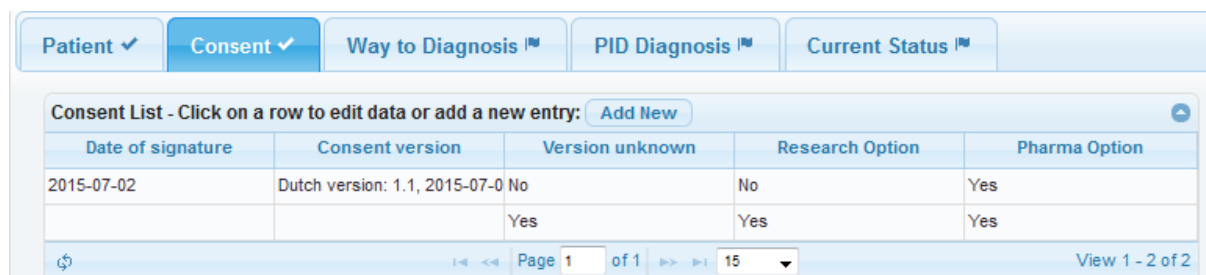
ESID ID	Year of Birth	Status	Sex	Is index	Index rel.	PID	ClinDx year	GeneticDx year
2	1973	Alive	Female	Yes			1995	
3	2010	Alive	Female	No	grandson/-daughter	Agammaglobulinem	2012	2014

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Export: 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 screenshot shows the 'Consent List' table with the following columns:

- Date of signature**
- Consent version**
- Version unknown**
- Research Option**
- Pharma Option**

The table shows two rows of consent 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

Page 1 of 1 | 15 | View 1 - 2 of 2

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:

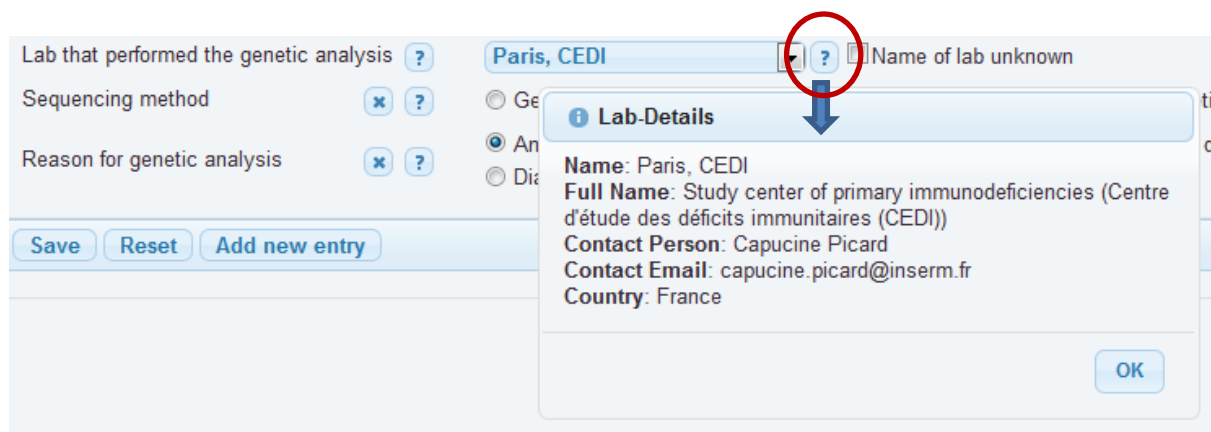
OR you can first enter the gene and then select a suitable PID diagnosis:

- 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 for entering genetic analysis data. The field 'Lab that performed the genetic analysis' has a dropdown menu with 'Paris, CEDI' selected. A red circle highlights a question mark icon next to the dropdown, with a blue arrow pointing down to a 'Lab-Details' pop-up window. The pop-up window displays the following information:

- 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

The background form includes fields for 'Sequencing method' and 'Reason for genetic analysis', each with a search icon and a question mark. At the bottom, there are buttons for 'Save', 'Reset', and 'Add new entry'. An 'OK' button is located at the bottom right of the 'Lab-Details' pop-up.

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:

The first screenshot shows the 'Current brand name' dropdown with a question mark icon and an 'Unknown' checkbox. The 'Current route of administration' has three radio buttons: Intravenous, Subcutaneous, and Intramuscular.

The second screenshot shows 'Hizentra / CSL Behring' selected in the brand name dropdown. The 'Current route of administration' now only has two radio buttons: Subcutaneous and Intramuscular.

The third screenshot shows 'Endobulin / Grifols' selected in the brand name dropdown. The 'Current route of administration' now only has one radio button: Intravenous.

- Automatic calculation of dose:

The screenshot shows the following fields and options:

- Patient's current weight:** A text input field followed by 'kg' and an 'Unknown' checkbox.
- Current dose:** A text input field followed by 'mg/kg body weight'.
- Dose per month:** A text input field with a question mark icon.
- Current dose (total amount):** A text input field with a multiplication icon and a question mark icon, followed by radio buttons for 'grams' and 'milliliters'.
- Dose unknown:** A checkbox with a question mark icon.
- Interval for this dose:** A text input field with a question mark icon, followed by the text 'Select interval-type first:' and two radio buttons: 'every x weeks or days' and 'x time(s) per week, month or year'.
- Interval unknown:** A checkbox with a question mark icon.

Three callout boxes provide additional information:

- Top Left:** Enter mg/kg OR absolute dose OR BOTH (pointing to the 'Current dose' field).
- Top Right:** This read-only value is calculated from dose & interval & weight (pointing to the 'Dose per month' field).
- Bottom:** The interval can be entered in two different formats – please select one of these for each entry:
 - every [input] weeks or days
 - [input] time(s) per week or month or 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: ?
Current dose (total amount)	x ?	<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="radio"/>		
	x	every <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)	x ?	<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:

300 mg/kg body weight
100 grams
every 5 weeks
☐
35 kg ☐ Unknown
☐ Yes ☐ No ☐ 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:

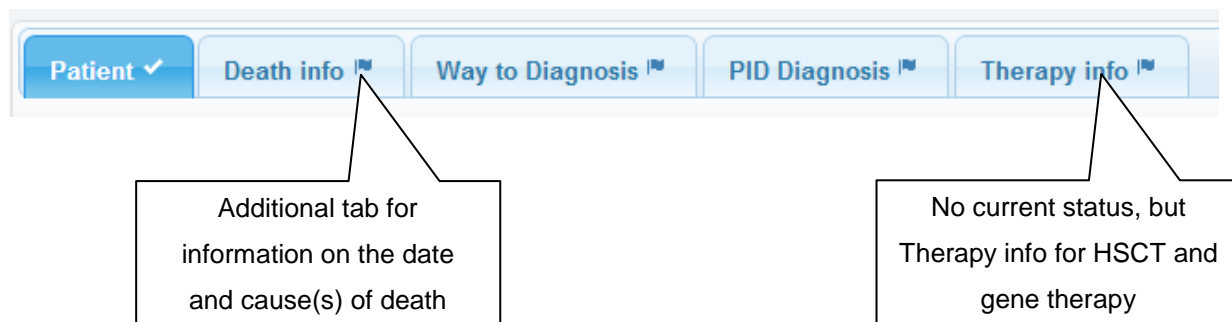
Current brand name	?	<input type="text" value="Subcuvia / Baxter"/>	<input type="checkbox"/> Unknown
Current route of administration		<input type="radio"/> Subcutaneous <input type="radio"/> Intramuscular <input type="radio"/> Unknown	
Current place of administration	?	<input type="radio"/> Home <input 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)	x ?	<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.

The screenshot shows the 'Main cause(s) of death' form with a grid of checkboxes for various causes. A confirmation dialog box is overlaid on the form.

Main cause(s) of death

<input type="checkbox"/> Septic shock	<input type="checkbox"/> Heart failure	<input type="checkbox"/> Respiratory failure	<input type="checkbox"/> Liver failure
<input checked="" type="checkbox"/> Renal failure	<input type="checkbox"/> Multiple organ failure	<input type="checkbox"/> Haemorrhage	<input type="checkbox"/> Thrombosis
<input type="checkbox"/> Neurological complications	<input type="checkbox"/> Surgical complications	<input type="checkbox"/> Drug toxicity	
<input type="checkbox"/> Relapse of malignancy	<input type="checkbox"/> Veno-occlusive disorder	<input type="checkbox"/> Post-transplant lymphoproliferative disorder	

Please Confirm...
Do you want to add the corresponding ICD10-Code?: N19
Yes No

ICD-10 categorisation (optional)

Code:
Text:

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

Page 1 of 1 15

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

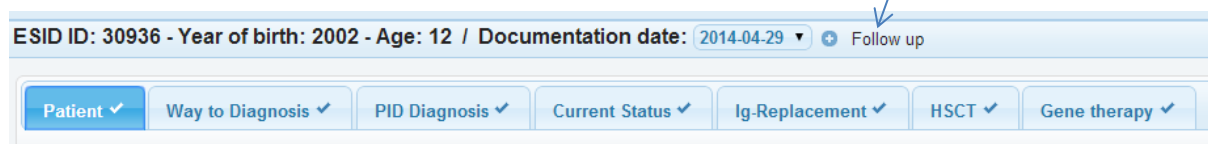
If you have completed a documentation timepoint for a patient, it is possible to add a follow-up documentation. The system will start sending email reminders if a patient has not been updated for more than one year (365 days).

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:

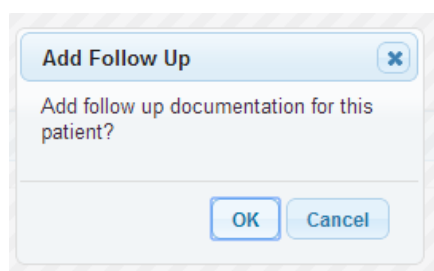
- All tabs have a check sign
- “+ Follow up” is displayed next to the current documentation date



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 'PID Diagnosis' tab selected. Under 'Current Pid Diagnosis', the following fields are visible:

- Current PID diagnosis: ?
- Affected gene: ?
- Additional genes: ?
- Genetic tests performed?: x ? ☒ Yes, but no mutation
- Lab that performed the genetic analysis: ?

At the bottom, the 'Add new entry' button is circled in orange.

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 View 1 - 1 of 1

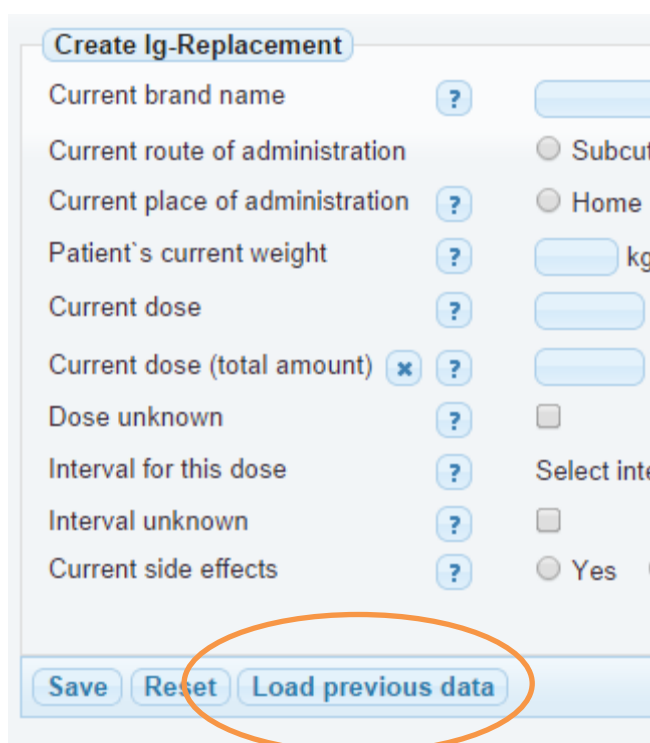
Export:

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

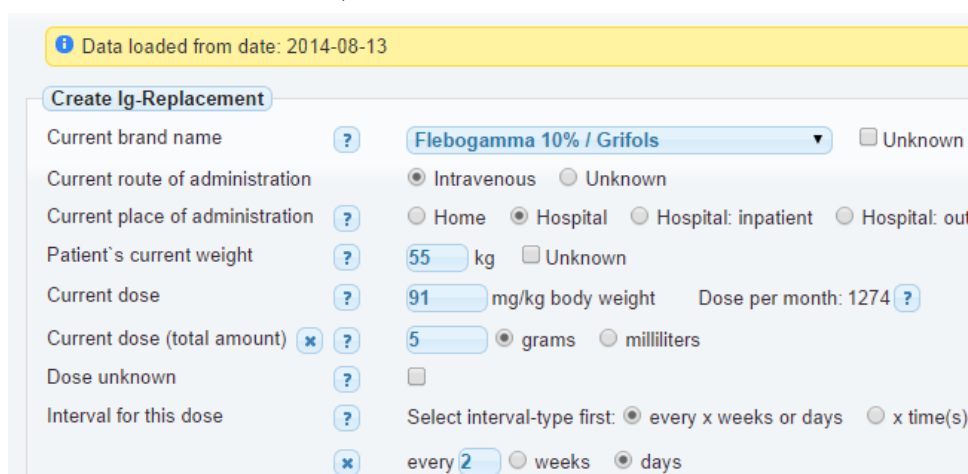
In order to minimize the documentation burden, you can load the Ig replacement data from the previous documentation timepoint into the form:



The screenshot shows the 'Create Ig-Replacement' form. The 'Load previous data' button at the bottom is circled in orange. The form fields include:

- 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 interval
- Interval unknown: ? ☐
- Current side effects: ? ☐ Yes

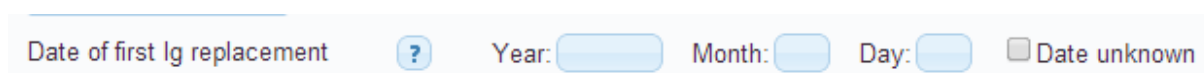
Buttons at the bottom: Save, Reset, Load previous data.

The screenshot shows the 'Create Ig-Replacement' form after loading previous data. A yellow banner at the top indicates 'Data loaded from date: 2014-08-13'. The form 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 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. It includes a question mark icon, a Year dropdown, a Month dropdown, a Day dropdown, and a checkbox for '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":



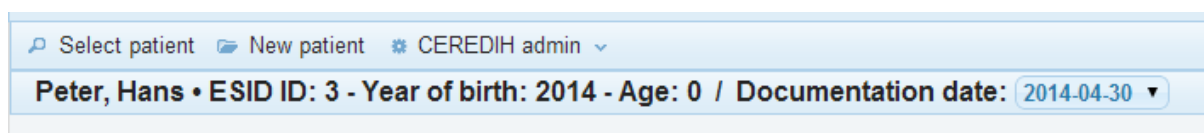
The screenshot shows a web interface for a patient list. At the top, there is a header bar with 'All patients' and a button labeled 'Show patient names'. Below this is a table with columns: ESID ID, Documenting Centre, Year of Birth, Living Status, and a fifth column for patient names. The table contains several rows of patient data. A blue arrow points from the text 'Show patient names' in the paragraph above to the 'Show patient names' button in the interface.

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 these links, the patient's name 'Peter, Hans' is displayed, followed by '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 â á é ê ë ð ó Ñ ñ ä Å ö Ö ü Û 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)

This service is operated by: ESID Online Registry coordination, CCI, University Medical Center Freiburg, Tel. +49 7 1 240-1234
Mainzeilliste Copyright © 2013-2015 Martin Lablans, Andreas Borg and Frank Ückert. Licensed under the GNU Affero General Public License (AGPLv3+). More

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

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

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.

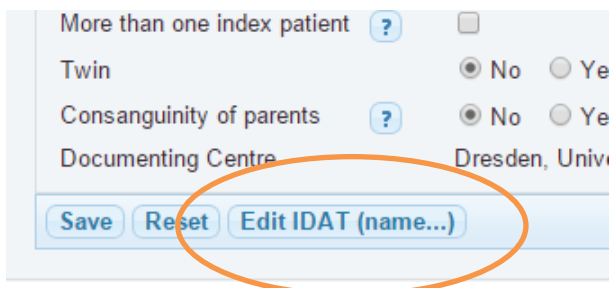
If the entry belongs to **another** centre, you are redirected to your patient list, and there is a **message** that tells you the name of the center that this patient belongs to.

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 IDAT¹ (name...)” button in the Patient tab:



More than one index patient ? ☐

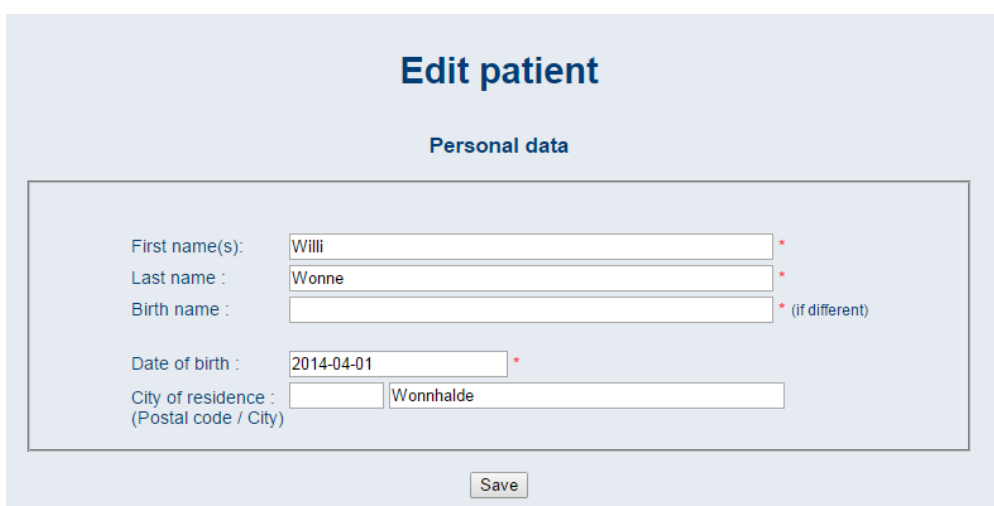
Twin ☒ No ☐ Yes

Consanguinity of parents ? ☒ No ☐ Yes

Documenting Centre Dresden, Univ

Save Reset **Edit IDAT (name...)**

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



Edit patient

Personal data

First name(s): Willi *

Last name : Wonne *

Birth name : * (if different)

Date of birth : 2014-04-01 *

City of residence : Wonnhalde

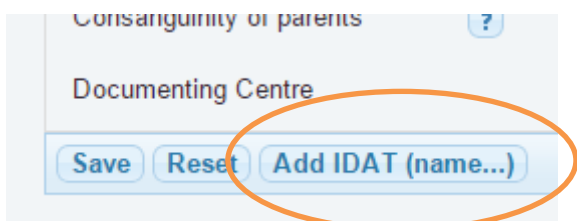
(Postal code / City)

Save

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 ?

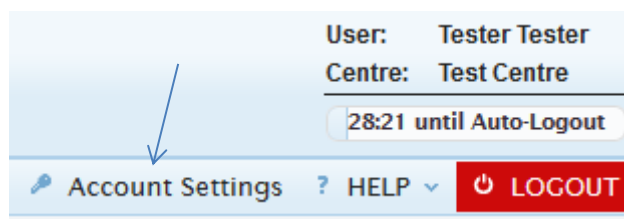
Documenting Centre

Save Reset **Add IDAT (name...)**

¹ „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	Change Password
Centre	Test Centre
Title	<input type="text"/>
First name	Tester
Last name	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	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	Netherlands
Affiliation (for publications)	<input type="text"/>
Display names in list	<input type="checkbox"/>
Cache patient list locally	<input type="checkbox"/>
Patient List size	15
Language	English
App Theme	Theme: cupertino
Display animation in info dialogs	<input type="checkbox"/>
Hover-Effect in forms	<input type="checkbox"/>

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 for further information.

☐ Yes ☐ No ☐ Ask again later

OK

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

Create PID Diagnosis

Current PID diagnosis ? **Common variable immunodeficiency disorders (CVID)**

Affected gene ? **Search PID by 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

Save **Reset**

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.

Create Latest Blood Count

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

Data obtained in the absence of immunosuppressive therapy? ☐ Yes ☐ No ☐ Currently unk. ☐ Truly unk.

If no, please specify

Save **Reset**

Convert Value

Please choose your unit:

Convert 12 /mm³ to 10⁹/L

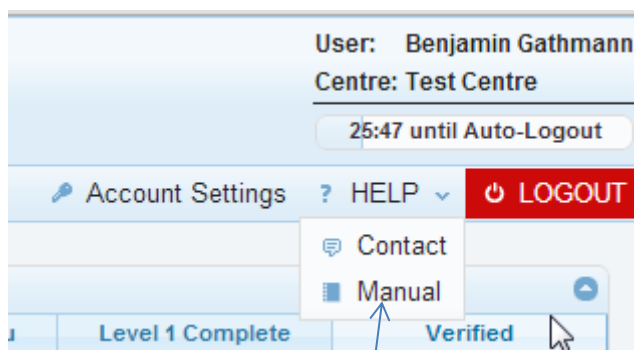
OK **Cancel**

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) Keine Datei ausgewählt.

You can reach us by email: esid-registry@uniklinik-freiburg.de

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

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

**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