



**ERKReg**

The European Rare Kidney Disease Registry

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**Cystinuria Subregistry User Manual**





# ERKReg

## The European Rare Kidney Disease Registry

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### INTRODUCTION

The Cystinuria subregistry is a subregistry from ERKReg. This document provides a step-by-step user manual to enter a new patient and add the data associated to the patient.

Once logged in, if you click on Data Entry (left menu), you will be redirected to the list of patients that you have already entered into the subregistry (see Figure 1). You can easily filter the data using the filtering options available (see Figure 1). You can also see when the next visit to the center is due, and what is the current CKD of the patient.

### Patients Registry

Center: Wiesenbach, Test Center external

Patient filter order by:  [v]

[Both units] [v] [All visits] [v] [All CKD stages] [v] [All treatments] [v] [All registries] [v]

[All disease groups / diagnoses] [v] [Family filter OFF] [v]

Please check a patient below or

<b>500-0009</b>	P	M-11/2013	Tubulopathy (Cystinuria-Patient)	<b>Click to enter initial visit!</b>	
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Figure 1: ERKReg data entry page

If you click on a patient, you will be able to modify the basic patient data, add or modify some visit data, add medications, extracorporeal therapies, diets and adverse events.

<b>500-0003</b>	P	F-09/2009	Tubulopathy (Cystinuria-Patient)	CKD4 / T1	Next visit due: 14/05/2022	
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**Basic data** **Add visit** **Termination**      **Previous visits:** 14/05/2021 14/05/2020 14/05/2019

**Medications NEW**

**Extracorp. Therapies**

**Diet**

**Adverse Event**

Figure 2: Options available upon the entry of a Cystinuria patient.

### BASIC DATA

When a new patient is added, you need first to fill the basic data, where you are to specify the consent given by the patient, the patient basic data (date of birth, sex, ethnicity, date of first symptoms, date of diagnosis), the patient diagnosis and specific questions linked to the diagnosis (for example, specific questions about the family history are asked for patients enrolled in the Cystinuria subregister).





### ADD VISIT

Once the basic data is entered, you will be able to add a visit for the patient. It is mandatory to fill the date of the visit, the current treatment modality, the height, the weight, the blood pressure and the serum creatinine (see Figure 3). If the serum creatinine was measured in  $\mu\text{mol/L}$  rather than in  $\text{mg/dl}$ , it is possible to directly make the conversion using the conversion field. Please note that data entered into  $\text{mg/dl}$  will not be converted into  $\mu\text{mol/L}$ . Please also note that the data in the database is only saved in SI units.

#### Patient

500-0003 (F-09/2009)

Visit Date

14/05/2021 (dd/mm/yyyy)

Age at visit

11.7 y

Current treatment modality

Conservative

(Previous visit: Transplantation)

#### Anthropometric features

Height

145 cm

Weight

42 kg

BMI

20.0  $\text{kg/m}^2$

Blood pressure

(mean of last 2-3 measurements if measured more than once in past 12 months)

120 / 80 mm Hg

#### Biochemical features

Serum creatinine

1.96 mg/dl

or convert:   $\mu\text{mol/L}$

Estimated GFR

57  $\text{ml/min/1.73m}^2$

Figure 3: Mandatory data in patient visit.

Additional biochemical parameters can then be added, if the measurements were performed (see Figure 4). If the measurements were not conducted, please leave the fields empty (see Figure 4). Conversion fields are also available to convert from molar units to SI units.



### 24h urinary samples: (Fill what applies)

<b>Urinary volume</b>	<input type="text" value="1358"/>	mL/24h	
<b>Creatininuria</b>	<input type="text"/>	g/24h	or convert: <input type="text"/>
<b>Cystinuria</b>	<input type="text"/>	mg/24h	or convert: <input type="text"/>
<b>Natriuresis</b>	<input type="text"/>	mmol/24h	
<b>Kaliuresis</b>	<input type="text"/>	mmol/24h	
<b>Urea</b>	<input type="text" value="30"/>	g/24h	or convert: <input type="text"/>

### Urinary spot samples: (Fill what applies)

<b>Proteinuria</b>	<input type="text"/>	g/L	
<b>Cystine crystals</b>	<input type="text" value="No"/>		
<b>Phosphate amorphous calcium carbonate</b>	<input type="text" value="Yes"/>		
<b>Urine pH</b>	<input type="text" value="8.7"/>		<b>Method used:</b> <input type="text" value="Dipstick"/>
<b>Urine specific gravity</b>	<input type="text"/>		
<b>Creatininuria</b>	<input type="text" value="900"/>	g/L	or convert: <input type="text"/>
<b>Cystinuria</b>	<input type="text"/>	µmol/mmol of creatinine	
<b>Cystine capacity</b>	<input type="text"/>	mmol/L	

### Calculi: (Fill what applies)

<b>Spontaneous stone passage</b>	<input type="text" value="Yes"/>		<b>Date (dd/mm/yyyy):</b> <input type="text" value="12/12/2018"/>
			<b>Number of calculi:</b> <input type="text" value="5"/>
<b>Radiology (inclusion/follow-up)</b>	<input type="text" value="No"/>		

Figure 4: Optional biochemical parameters for patients enrolled in the Cystinuria subregister.

Finally, you will be required to provide further information regarding the type of self-monitoring used by the patient and the development of new stones over the 12 months prior to the visit (see Figure 5). These fields are mandatory to fill.



### Quality and Performance Indicators

Did patient develop new stones in past 12 months?

No ▾

Stone analysis

Not done ▾

### Self monitoring

Urine specific gravity

No ▾

Urine colour chart

No ▾

Urine pH

Yes ▾

Method used: pHmeter ▾

Figure 5: Self-monitoring and Quality indicators for the patients enrolled in the Cystinuria subregister.

## TERMINATION

Patients can be terminated if the follow-up does not take place anymore, if the patient is transferred to another center or if the patient passes away. Please note that this does not delete the patient data from the database.

### Patient termination entry

Center: Wiesenbach, Test Center external

Patient-ID 500-0009

Reason for end of follow-up



Loss of follow-up (including transfer to non-ERKNet center)

Patient death

Transition to adult unit

**Administrator only:**

Transition to other center

Save

st

Figure 6: Patient termination entry

## EXTRACORPORAL THERAPIES

In extracorporeal therapies, you have the possibility to enter therapies (surgeries or renal replacement therapies). You can click on a therapy to edit it or to add a new therapy (see Figure 7).





### Patient 500-0003 (F-09/2009)

#### Therapy history

  = ongoing therapy  
  = terminated therapy

Therapy	Start date	Kidney affected	Stop date	
Shock wave lithotripsy	23/06/2021	Right	23/06/2021	✘
Kidney transplantation	10/07/2010	Left		✘

Click on a therapy to edit or  ▼

Note: If you cannot find the therapy in the list, please contact [tanja.wlodkowski@med.uni-heidelberg.de](mailto:tanja.wlodkowski@med.uni-heidelberg.de).

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Figure 7: Extracorporeal therapies for patients enrolled in the Cystinuria subregister.

### DIET

In diet, you have the possibility to enter diets (alimentary restrictions, consultations with a dietician and surgeries). You can click on a diet to edit it or to add a new diet (see Figure 8).

### Patient 500-0003 (F-09/2009)

#### Diet history

  = ongoing diet  
  = terminated diet

Diet	Start date	Amount suggested (mL)	Stop date	
Fluid intake	23/06/2021	2500		✘
Protein reduction	23/06/2021			✘
Consultation with a dietician	23/06/2021			✘

Click on a diet to edit or  ▼

Note: If you cannot find the diet in the list, please contact [tanja.wlodkowski@med.uni-heidelberg.de](mailto:tanja.wlodkowski@med.uni-heidelberg.de).

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Figure 8: Diets for patients enrolled in the Cystinuria subregister.





### MEDICATIONS

In medications, you have the possibility to enter medications (mineral supplements, cystine-binding drugs and alkaline beverage). You can click on a medication to edit it or to add a new medication (see Figure 9). For each drug, you are required to indicate the dosage prescribed to the patient, the frequency at which the patient must take his treatment and the patient weight (that will be used to automatically compute the dosage/kg/day).

Please note that if the medication was stopped or the dosage modified following an adverse event, the type of adverse event is indicated in the column Adverse Event.

**Patient** **500-0003 (F-09/2009)**

#### Medication history

  = ongoing medication  
  = terminated medication

Medication	Start date	Single dose	Frequency	= dose/kg/day	Stop date	Adverse event	
Potassium bicarbonate	09/11/2015	10 I.U.	2x / day	0.667 I.U./kg/day		No	✘
Tiopronin	12/04/2012	50 mg	1x / week	0.476 mg/kg/day	12/03/2013	No	✘

Click on a medication to edit or

Note: If you cannot find the medication on the list, please contact [tanja.wlodkowski@med.uni-heidelberg.de](mailto:tanja.wlodkowski@med.uni-heidelberg.de).

Note 2: If a drug was prescribed or its dosage is modified following an adverse event, the type of adverse event is indicated in the column adverse event.

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Figure 9: Medication module for patients enrolled in the Cystinuria subregister.

### ADVERSE EVENT

In adverse event, you have the possibility to enter adverse events related to prescribed medications. You need therefore to complete first the medication data. The adverse events can be of several types: Gastrointestinal, Cutaneous, Hematological, Hepatic, Proteinuria, Pulmonary or Other. You can click on an adverse event to edit it or to add a new adverse event (see Figure 10).







### Patient

500-0003 (F-09/2009)

#### Adverse event history

Adverse Event	Event date	Treatment involved	Treatment discontinuation	Dose reduction	New dose	Outcome	New medication	
Gastrointestinal	12/03/2019	Potassium bicarbonate	No	Yes	5 I.U.	Recovery	No	✘

Click on an adverse event to edit or  ▼

Note: If you cannot find the adverse event on the list, please contact [tanja.wlodkowski@med.uni-heidelberg.de](mailto:tanja.wlodkowski@med.uni-heidelberg.de).

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Figure 10: Adverse event module for the patients enrolled in the Cystinuria subregister.

If you add a new adverse event of type “Other”, you will be able to enter a comment in the Remark field.

### Patient

500-0003 (F-09/2009)

#### Medication history

= ongoing medication  
 = terminated medication

Medication	Start date	Single dose	Frequency	= dose/kg/day	Stop date	Adverse event	
Potassium bicarbonate	13/03/2019	5 I.U.	2x / day	0.333 I.U./kg/day		No	✘
Potassium bicarbonate	09/11/2015	10 I.U.	2x / day	0.667 I.U./kg/day	12/03/2019	Gastrointestinal	✘
Tiopronin	12/04/2012	50 mg	1x / week	0.476 mg/kg/day	12/03/2013	No	✘

Click on a medication to edit or  ▼

Note: If you cannot find the medication on the list, please contact [tanja.wlodkowski@med.uni-heidelberg.de](mailto:tanja.wlodkowski@med.uni-heidelberg.de).

Note 2: If a drug was prescribed or its dosage is modified following an adverse event, the type of adverse event is indicated in the column adverse event.

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Figure 11: If an adverse event (e.g. Gastrointestinal) leads to a dosage reduction, the medication table is automatically adapted.

If the adverse event leads to a dosage reduction or a treatment discontinuation, the medication table is automatically adapted (see Figure 11). In the case of a dosage reduction, please note that the new dosage must be strictly lower than the original dosage. It is important to notice that the frequency of the medication, the medication route and the patient weight are automatically kept to the same values than





for the original medication. In case these also need to be modified, you need to do so directly in the medication table.

If a new medication is prescribed following an adverse event, you directly click on Add new medication(s) once you filled the previous fields (see Figure 12). It will redirect you towards the medication table, where you will be able to enter a new medication. In the medication table, please select the right adverse event associated to the prescription of the new medication.

### Adverse event **\*NEW\***

<b>Adverse Event</b>	<b>Proteinuria</b>
<b>Event date</b>	<input type="text" value="12/03/2021"/> (dd/mm/yyyy)
<b>Treatment involved</b>	<input type="text" value="Potassium bicarbonate"/> ▾
<b>Treatment discontinuation</b>	<input type="text" value="No"/> ▾
<b>Dose reduction</b>	<input type="text" value="Yes"/> ▾
<b>New dose</b>	<input type="text" value="3"/> <input type="text" value="I.U."/> ▾
<b>Outcome</b>	<input type="text" value="Partial recovery"/> ▾
<b>New medication</b>	<input type="text" value="Yes"/> ▾ <b>Add new medication(s)</b>

**Save and Close**

**Cancel**

Figure 12: How to add a new medication following an adverse event.

### **CONTACT:**

If you have any issue, please contact the ERKReg project manager: [tanja.wlodkowski@med.uni-heidelberg.de](mailto:tanja.wlodkowski@med.uni-heidelberg.de).

