ERKReg
The European Rare Kidney Disease Registry

Cystinuria Subregistry User Manual

This report is part of the project '777304 / ERK-REG', which has received funding from the EU Health Programme (2014-2020).

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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.

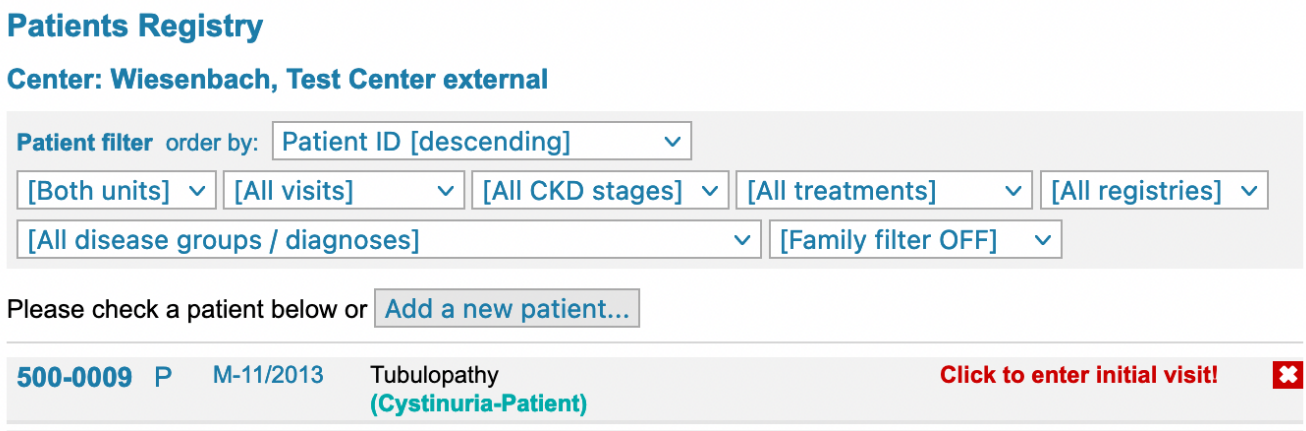

Figure 1: ERKReg data entry page

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

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).

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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 creatine was measured in µmol/L rather than in mg/dl, it is possible to directly make the conversion using the conversion field. Please note that data entered into mg/dl will not be converted into µmol/L. Please also note that the data in the database is only saved in SI units.

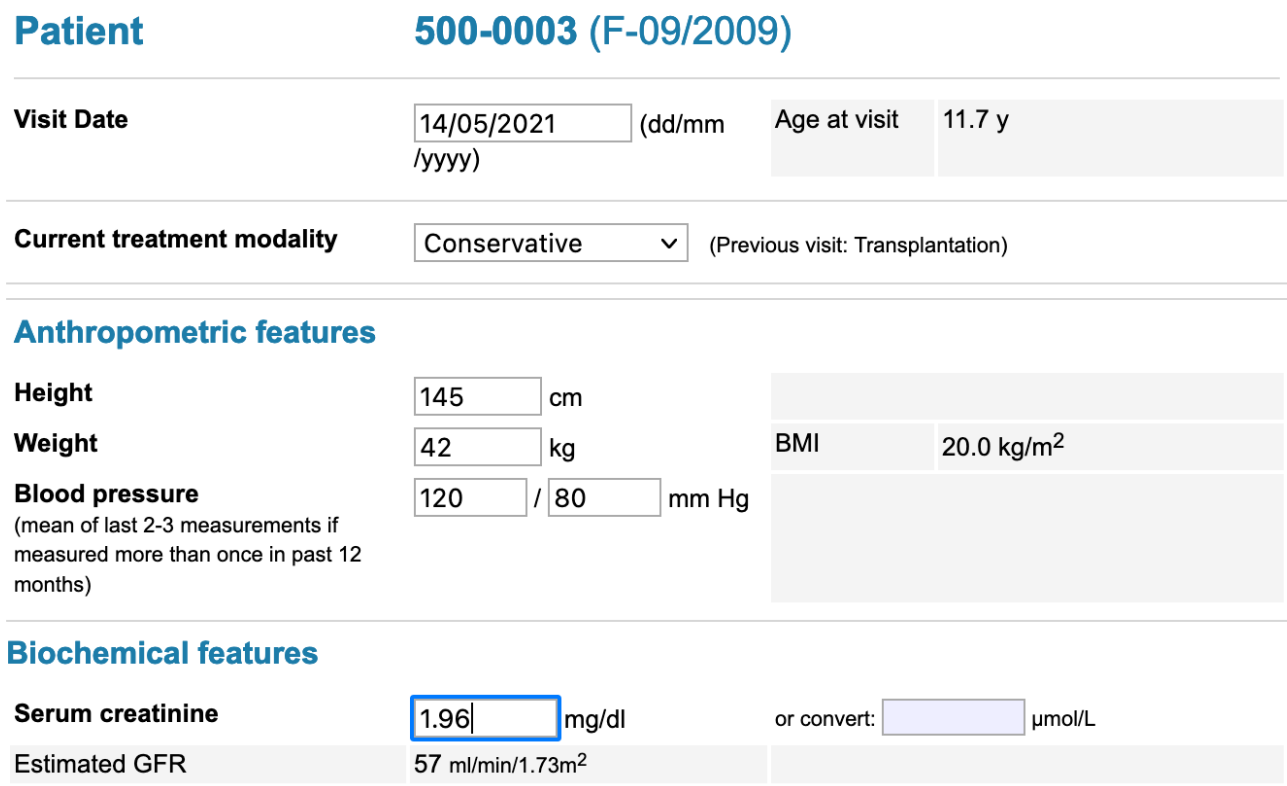

Figure 3: Mandatory data in patient visit.

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.

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24h urinary samples: (Fill what applies)

Urinary volume	1358 mL/24h		
Creatininuria	g/24h	or convert:	mmol/24h
Cystinuria	mg/24h	or convert:	µmol/24h
Natriuresis	mmol/24h		
Kaliuresis	mmol/24h		
Urea	30 g/24h	or convert:	mmol/24h

Urinary spot samples: (Fill what applies)

Proteinuria	g/L	
Cystine crystals	No	
Phosphate amorphus calcium carbonate	Yes	
Urine pH	8.7	Method used: Dipstick
Urine specific gravity		
Creatininuria	900 g/L	or convert: mmol/L
Cystinuria	µmol/mmol of creatinine	
Cystine capacity	mmol/L	

Calculi: (Fill what applies)

Spontaneous stone passage	Yes	Date (dd/mm/yyyy): 12/12/2018 Number of calculis: 5
Radiology (inclusion/follow-up)	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.

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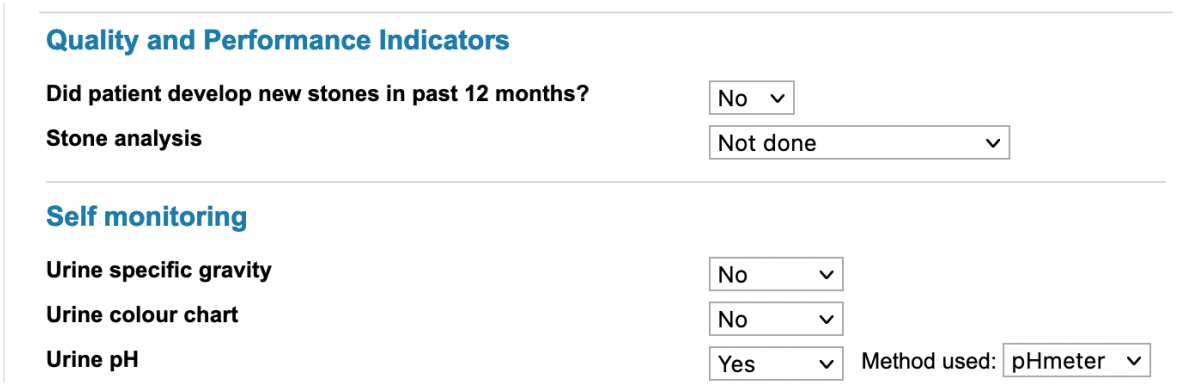

Figure 5: Self-monitoring and Quality indicators for the patients enrolled in the Cystinuria subregister.

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.

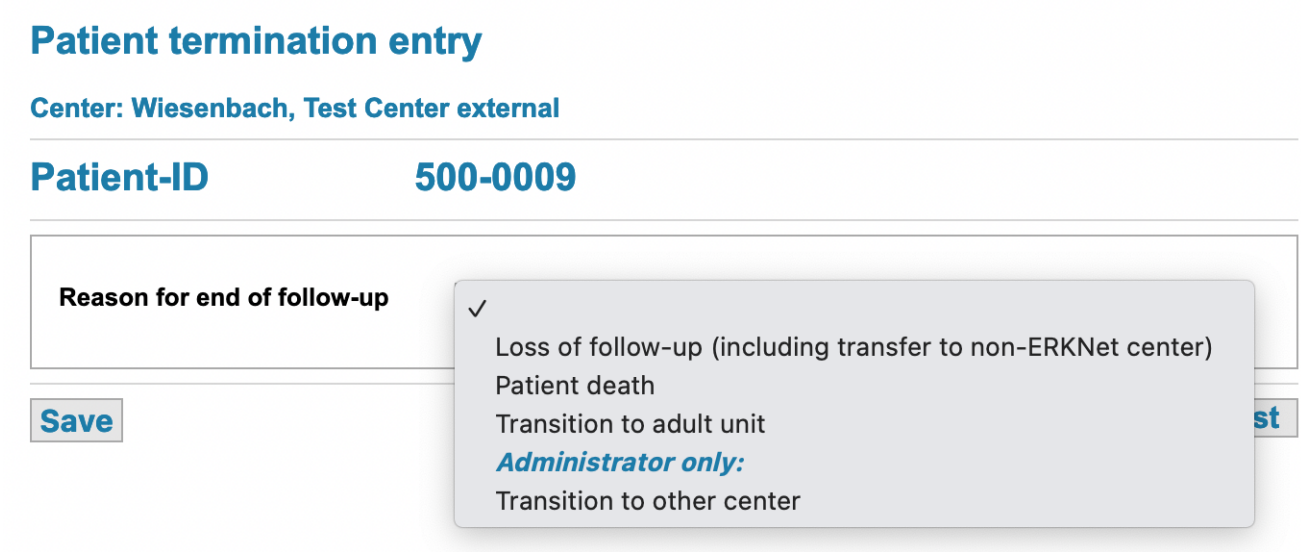

Figure 6: Patient termination entry

EXTRACORPORAL THERAPIES

In extracorporal 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).

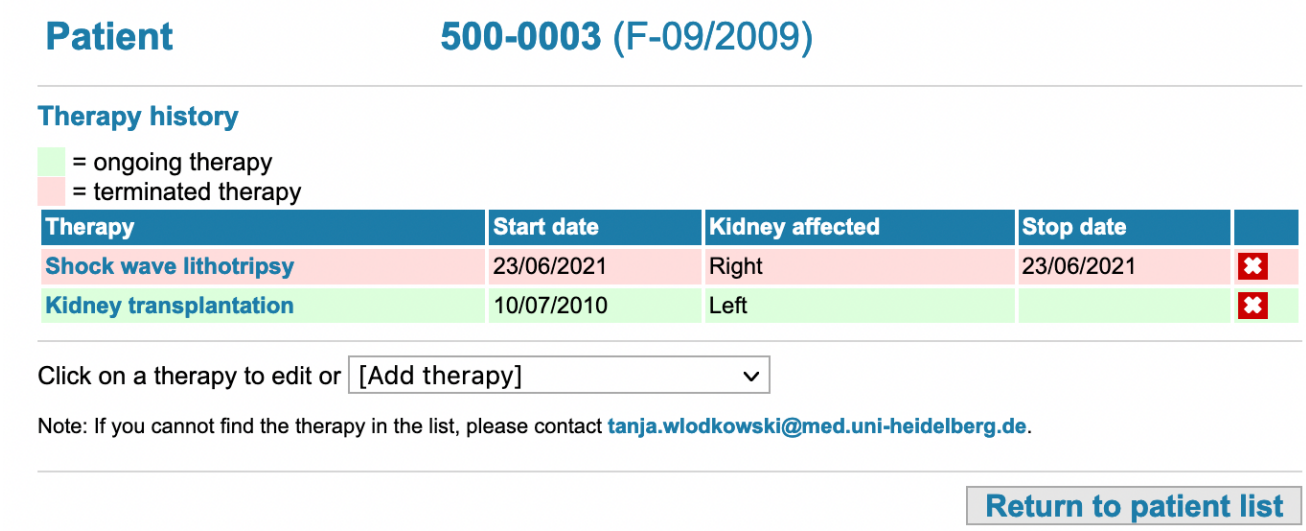

Figure 7: Extracorporal therapies for patients enrolled in the Cystinuria subregister.

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).

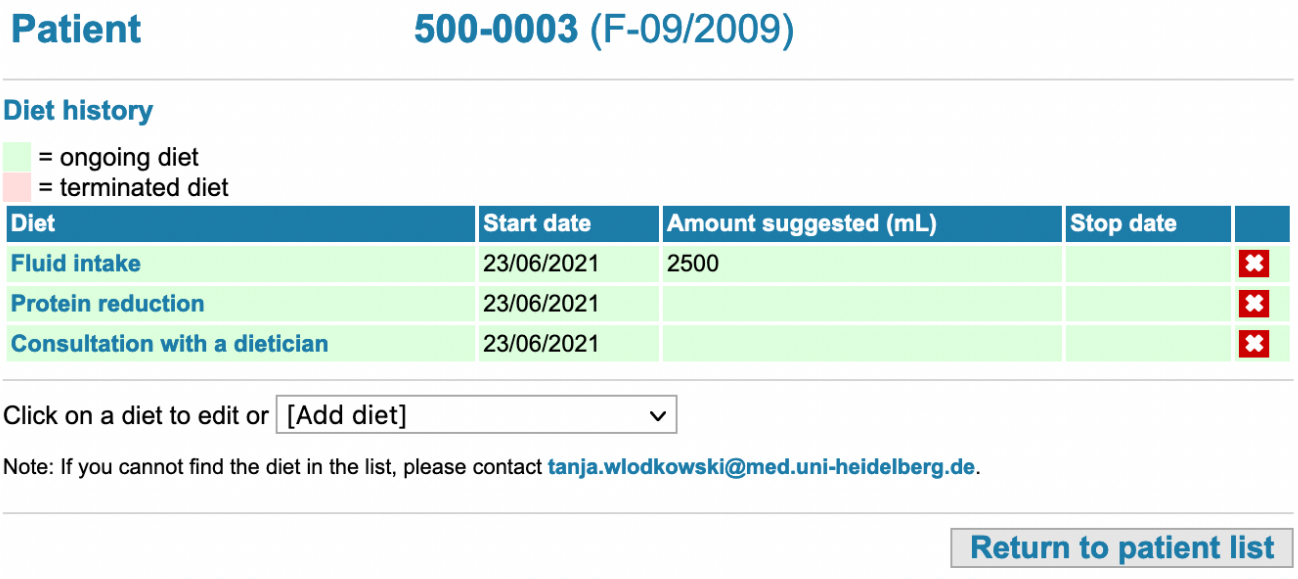

Figure 8: Diets for patients enrolled in the Cystinuria subregister.

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.

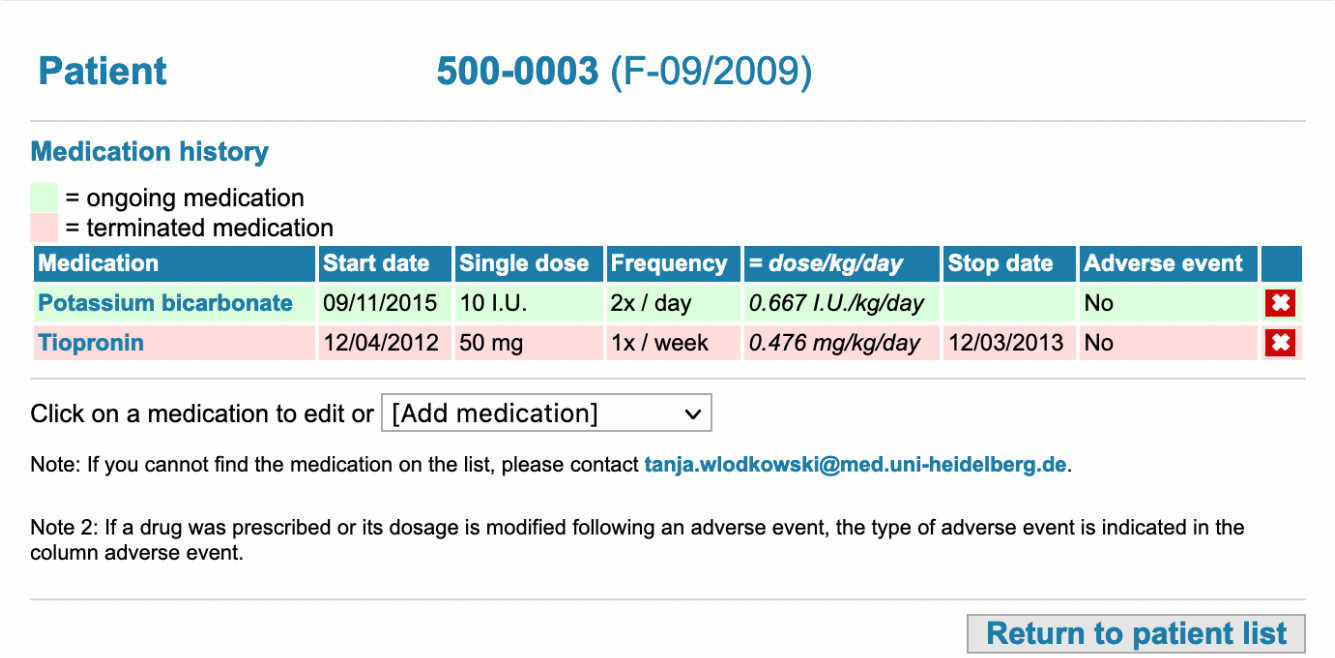

Figure 9: Medication module for patients enrolled in the Cystinuria subregister.

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).

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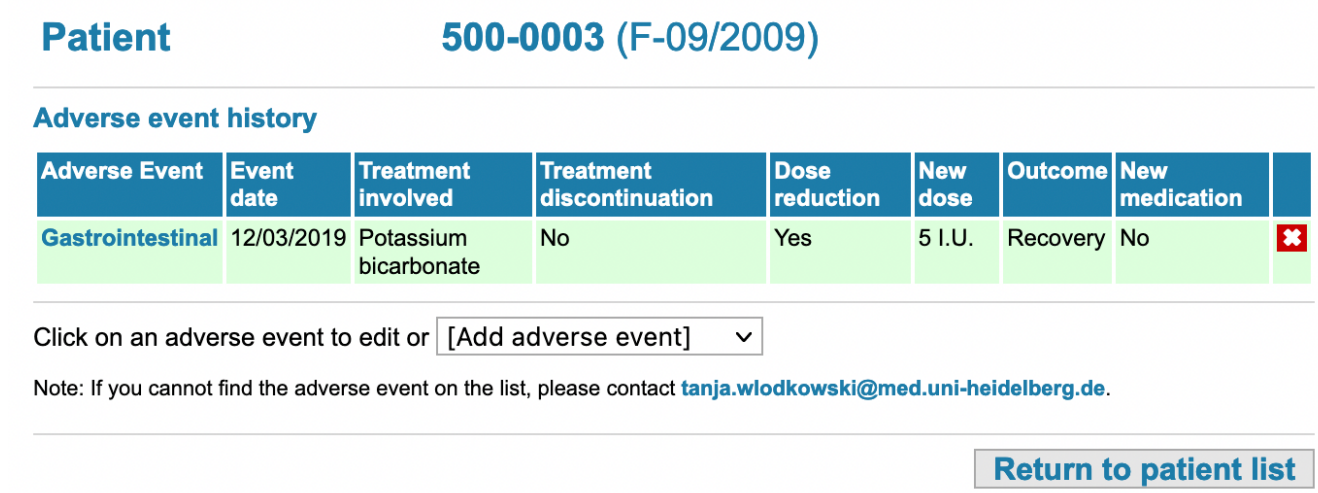


Patient **500-0003** (F-09/2009)

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	✖

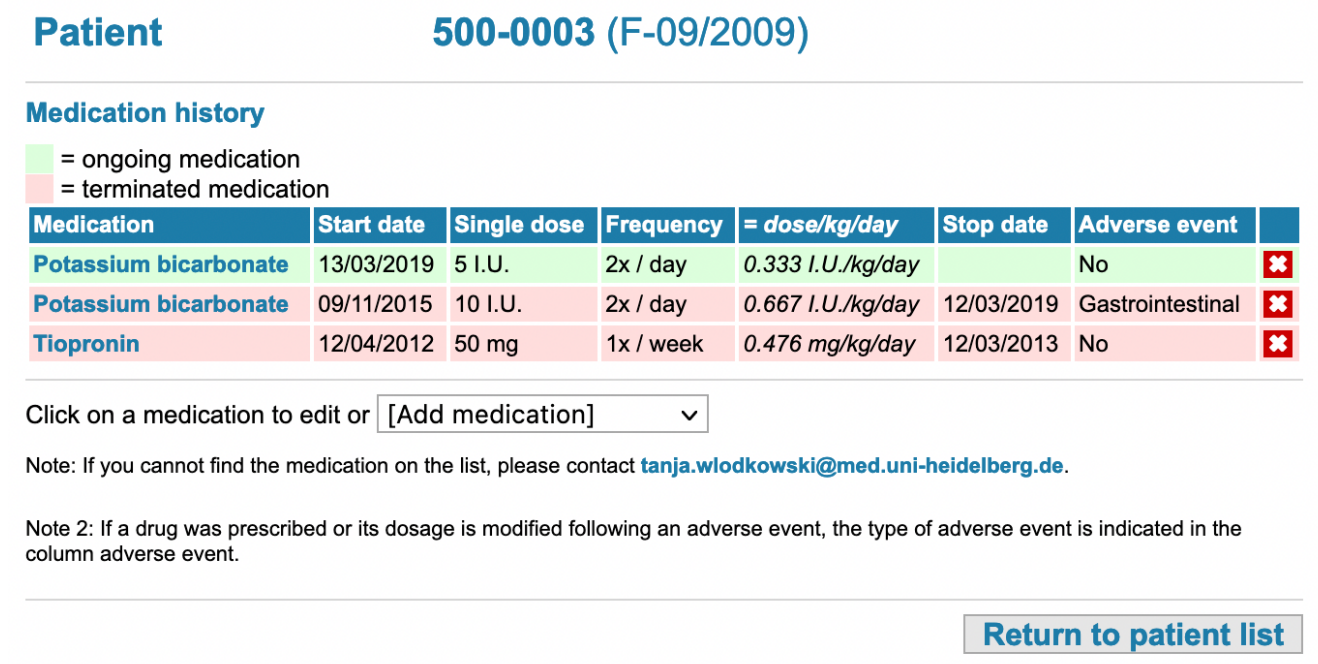
Click on an adverse event to edit or [Add adverse event]

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

Return to patient list

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 [Add medication]

Note: If you cannot find the medication on the list, please contact 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.

Return to patient list

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

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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.

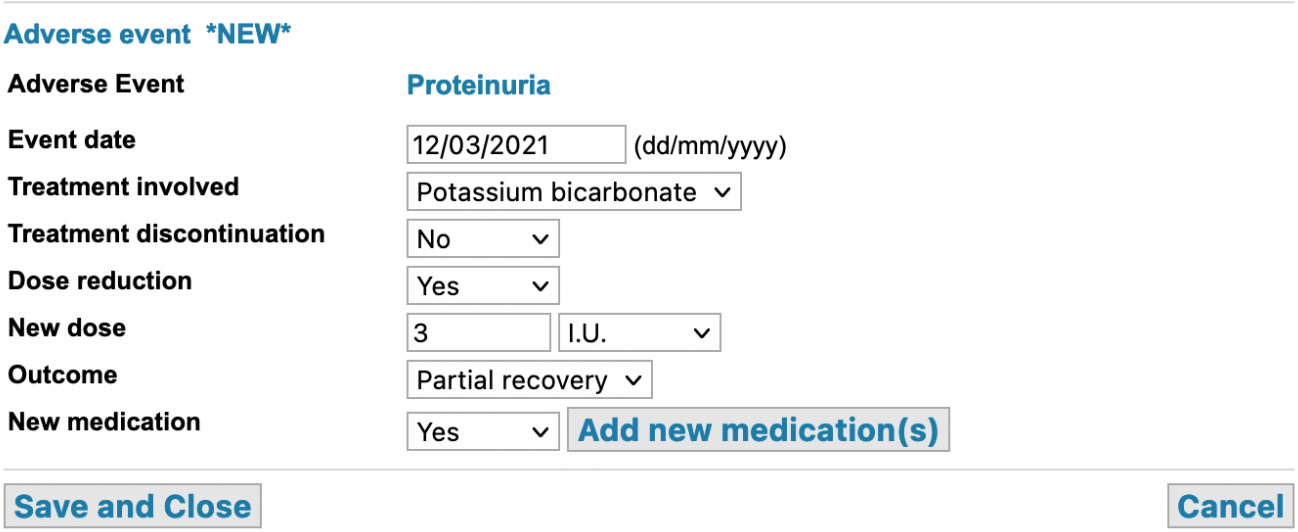

Figure 12: How to add a new medication following an adverse event.

CONTACT:

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