ERKReg The European Rare Kidney Disease Registry

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Distal renal tubular acidosis (dRTA) Sub-registry user manual

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INTRODUCTION

The Distal renal tubular acidosis (dRTA) sub-registry is one of several sub-registries collecting specific information in ERKReg and this document provides a step-by-step user manual to enter a new patient and add the patient data.

Once logged-in to ERKReg, please select the Data Entry option from the left-side menu to enter the Patients Registry page (Figure 1).

The Patients Registry page displays the existing patients from your center in ERKReg with the current CKD classification and the next scheduled visit. You may also use the top section of the page to filter and identify patients. Of special note is the option to filter patients by association to a sub-registry.

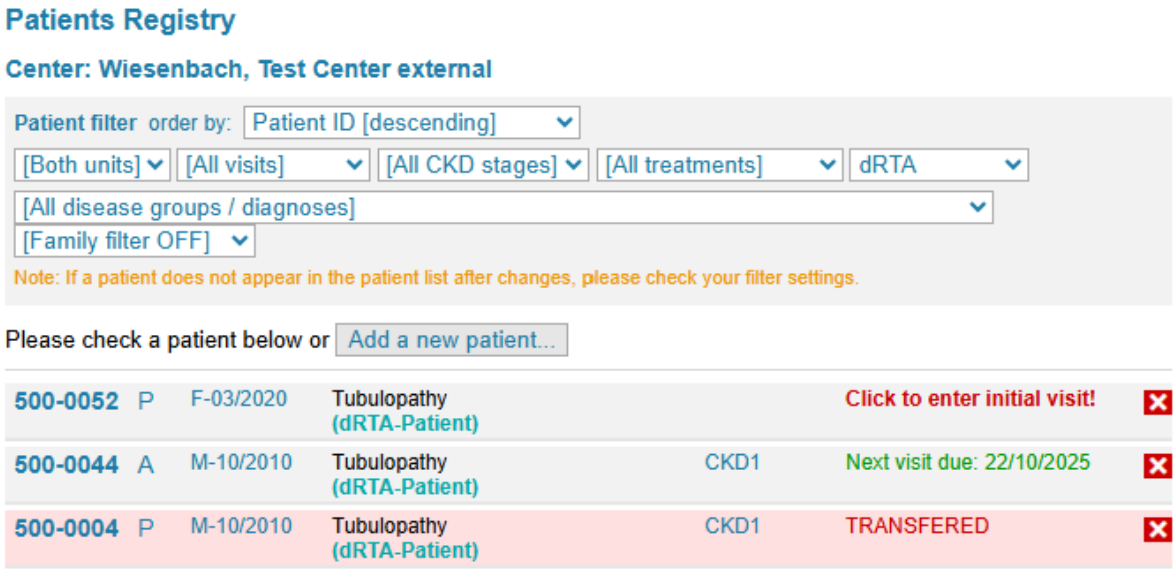

FIGURE 1: ERKReg data entry page

When a patient line is selected, the patient menu appears (Figure 2), which allows to modify the basic patient data, add or modify visit data, and medications.

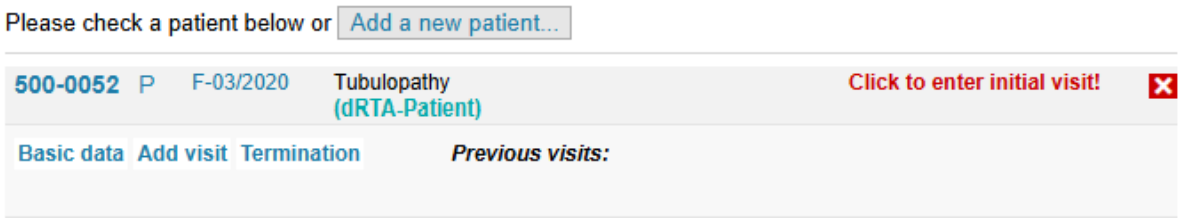

FIGURE 2: Options available upon selection of a dRTA sub-registry patient.

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BASIC DATA

When a new patient is added to the registry, the basic data module is presented, where you are required to enter the center unit, and can select to include the patient in the DRTA sub-registry (Figure 3).

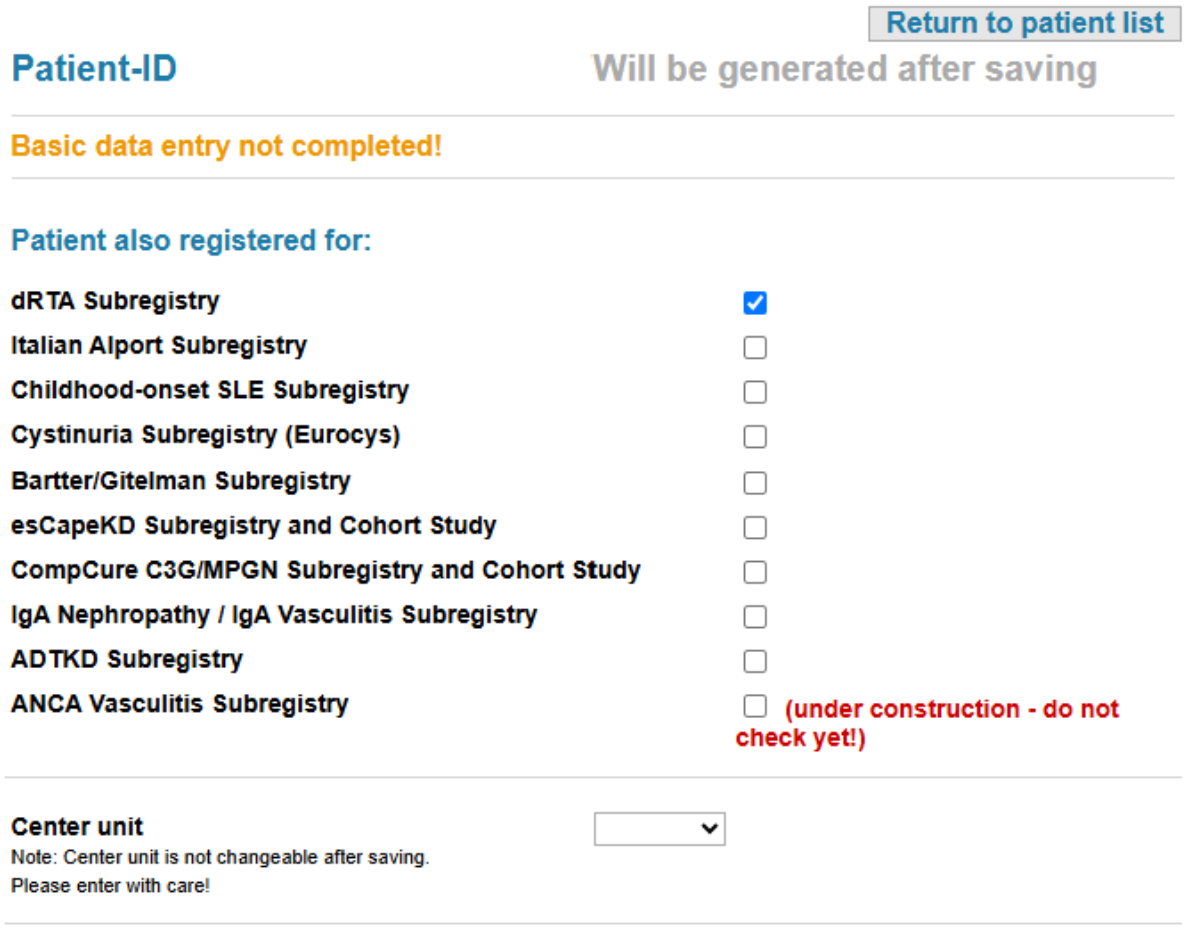

FIGURE 3: Adding a new patient to the dRTA sub-registry.

Please note, for existing patients in the registry, you may modify the basic data by selecting the "Basic data" button (see Figure 2), you may then add them to the DRTA sub-registry. After which, disease specific fields will be presented (Figure 4).
For each patient in the registry, basic information is collected - including the date of consent, the patient background and information regarding the disease diagnosis (Figure 4).

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ERKNet Registry

Date of informed consent (dd/mm/yyyy)

Consent to coded data being included in one or more ERN database or registry

Consent to being contacted about research projects or clinical trials

Consent to pseudonymized data being shared to support commercial projects aimed at improving healthcare. [info]

Consent to pseudonymized data being shared with researchers outside the European Union. [info]

Note:
• Please use the updated consent form, which contains the previous two items.

Basic data

Sex

Date of birth (mm/yyyy)

Ethnicity

Date of first signs or symptoms (mm/yyyy)
(leave field empty if unknown)

Date of first presentation to center (dd/mm/yyyy)

Renal diagnosis established? Yes

Primary renal diagnosis (OC: 0)

Select diagnosis... OR Diagnosis by gene... OR Enter OrphaCode... OR Search diagnosis name...

Note: To record a syndromic form of CAKUT, please assign its non-syndromic form first. Only then it will be possible to specify the syndromic form.

Does the patient have a second renal diagnosis? No

Diagnostic survey

When was the diagnosis considered confirmed? (dd/mm/yyyy)

Which methods were used to establish the diagnosis?
(Tick all that apply)

(1) Please check even if results negative or pending

- [ ] Clinical history
- [ ] Positive family history
- [x] Clinical examination
- [x] Biochemical evaluation
- [ ] Immunological evaluation
- [ ] Hematological evaluation
- [ ] Imaging
- [ ] Kidney biopsy
- [ ] Skin biopsy
- [ ] Genetic screening (1)
- [ ] Other methodologies

FIGURE 4: Basic information collected for all registry patients.

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For sub-registry patients, additional disease specific fields are collected, the additional fields for dRTA sub-registry are depicted in Figure 5.

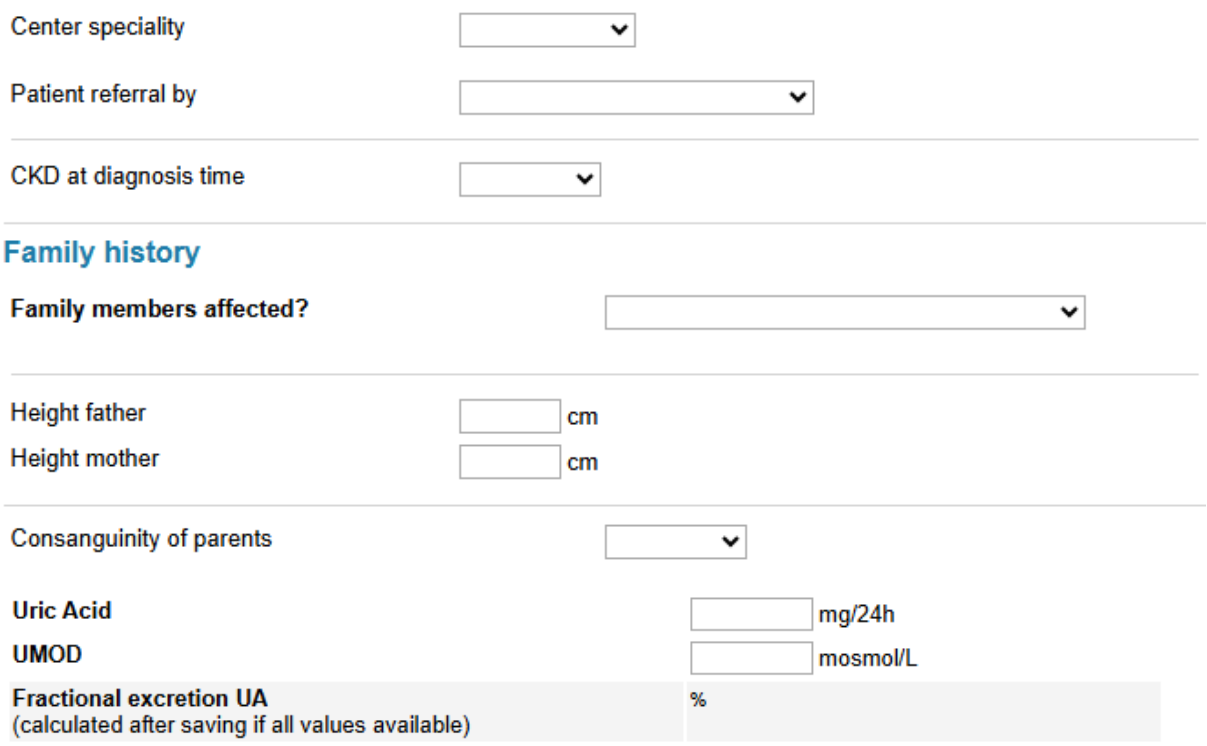

FIGURE 5: dRTA sub-registry specific data fields collected in the subregistry.

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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 6). 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 and **the data is only saved using the SI units.** Please ensure that you have selected the correct unit used in your centre (**Standard International or Imperial, e.g. mmol/l or mg/dl).**

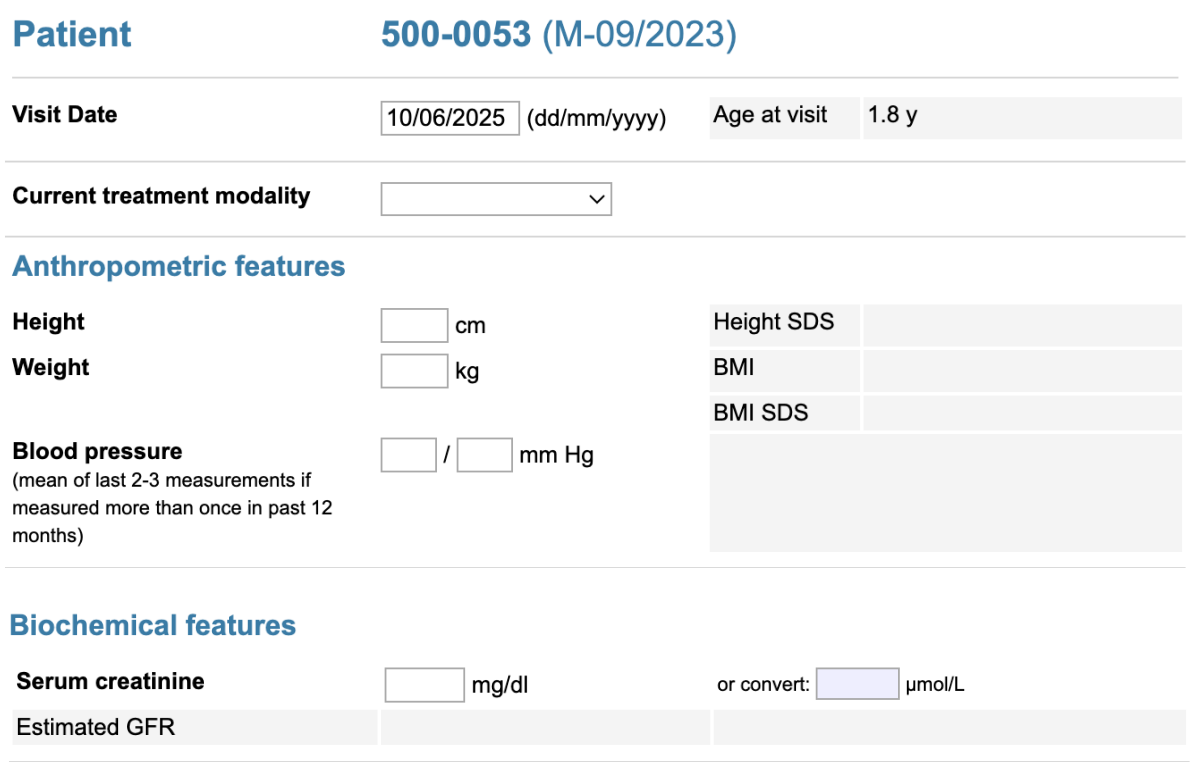

FIGURE 6: dRTA sub-registry entering visit data.

Additional biochemical parameters can then be added, if the measurements were performed (Figure 5). If the measurements were not conducted, please leave the fields empty (Figure 7). When relevant, additional conversion fields are available to convert from molar units to SI units.

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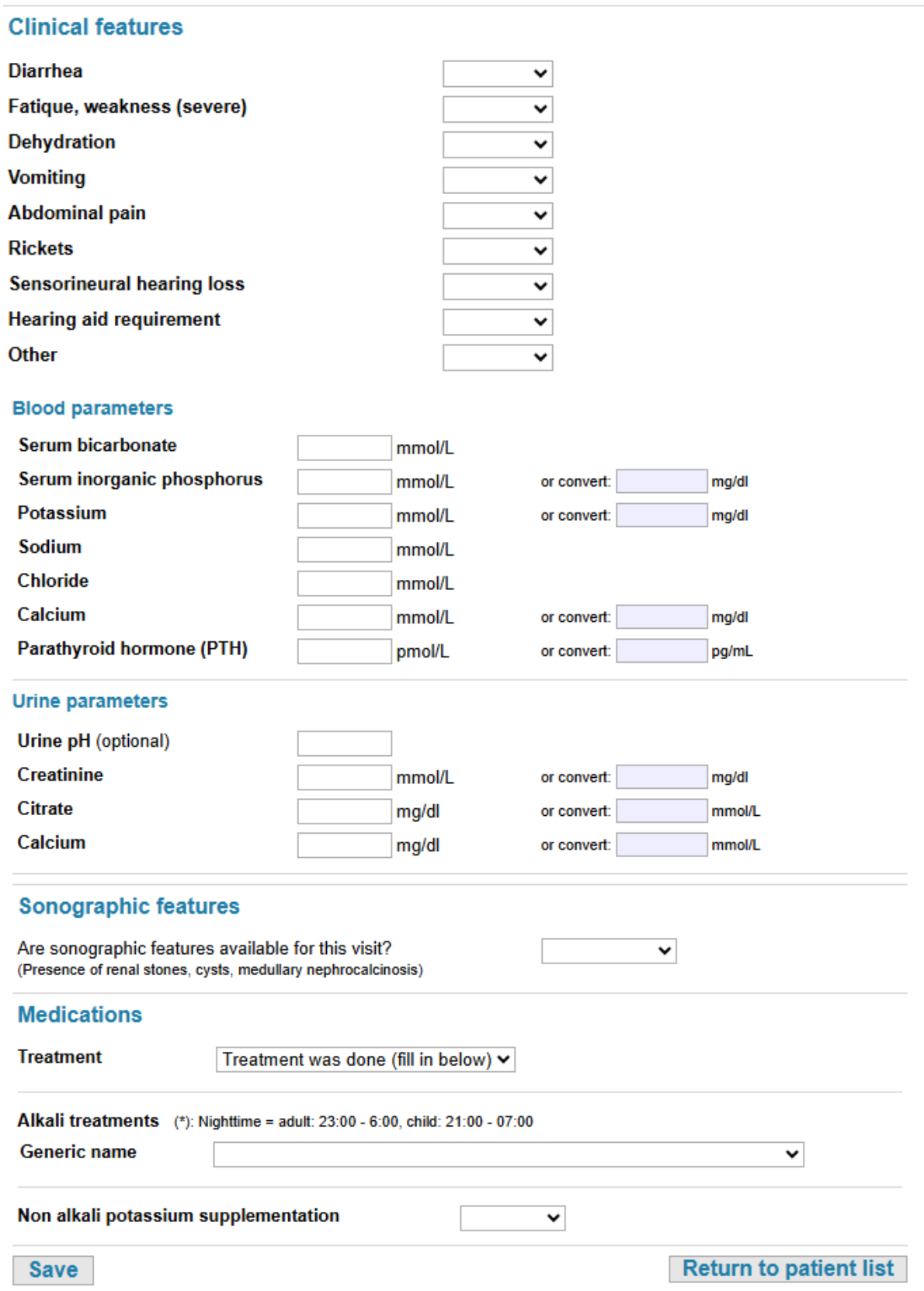

FIGURE 7: dRTA sub-registry entering clinical features on visit data information.

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IMPORTANT CONSIDERATIONS

Please ensure, you have selected the correct drug. In our experience, the commercial name is the easiest way to calculate the alkali dose. A list of products was compiled from public sources; however, note that in many centers galenic (custom-compounded) preparations are used (Figure 8). The most common citrate solutions are:

- Tripotassium citrate (324.4 mg=1 mmol= 3 mEq alkali)
- Polycitra (Na-K-citrate), which typically comes at a mixture containing 550 mg K-citrate and 500 mg Na-citrate per 5 ml for a total of 3.3 mmol citrate (10 mEq alkali) in 5 ml.
- Sodium bicarbonate, 500 mg = 6 mmol =6 mEq alkali
- Potassium bicarbonate 1000mg is =10 mmol =10 mEq alkali)
- Shohl's solution is typically 1 mEq alkali per ml, whereas the modified Shohl's solution is typically 2 mEq/ml.

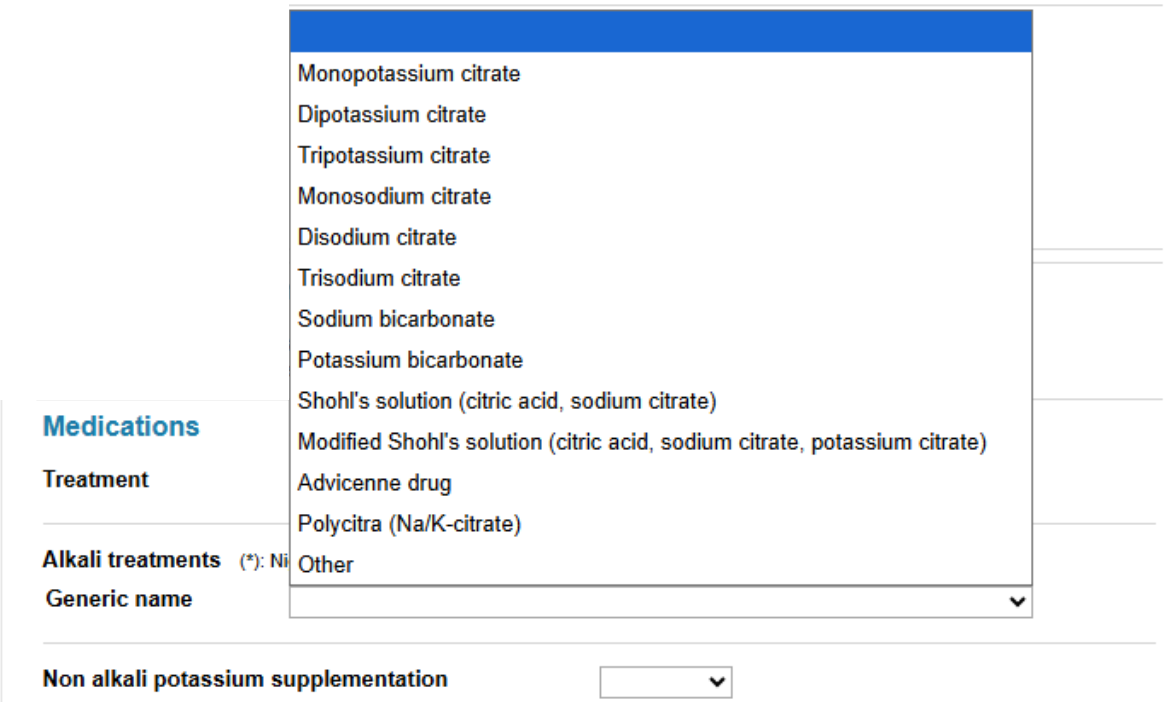

FIGURE 8: dRTA sub-registry alkali treatments.

IMPORTANT

Note that you need to enter the total daily dose and then separately the number of intakes for day and night. Thus, if the patient takes 10 mEq alkali 3 times during the day, you should enter "30 mEq" and indicate that this occurs in three-day doses and zero-night doses.

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TERMINATION

Patients can be terminated if the follow-up does not take place anymore or if the patient passes away (Figure 9).
If the patient care is transferred to another center – please contact us using the contact information found at the end of the manual, we will transition the patient to the new center and assign them with a new patient id.

Please note - termination does not delete the patient data from the database.

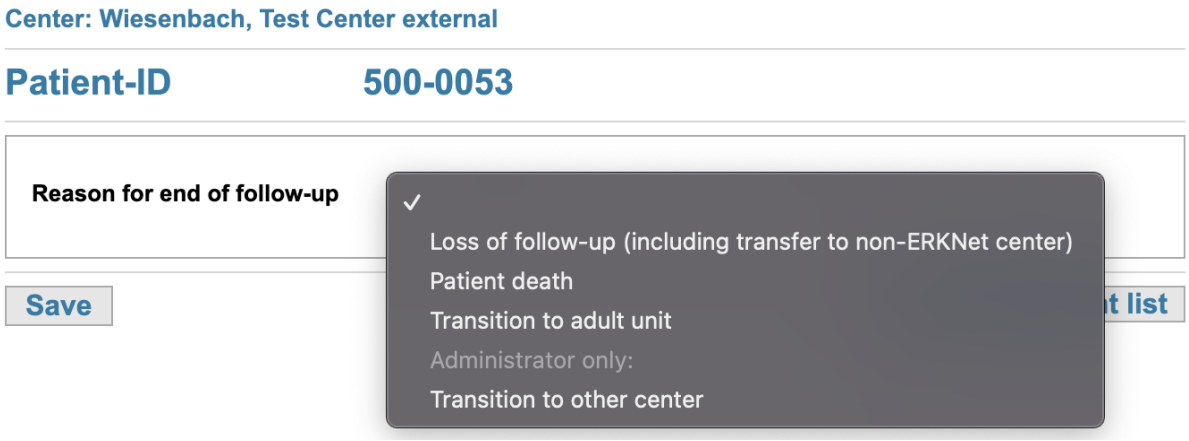

FIGURE 9: dRTA sub-registry patient termination entry.

CONTACT

If you have any issue, please contact the ERKReg project manager: contact@erknet.org

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