ERKReg The European Rare Kidney Disease Registry

The European Rare Kidney Disease Registry

Bartter – Gitelman Syndromes Sub-registry user manual

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INTRODUCTION

The Bartter Gitelman (BG) sub-registry is one of several sub-registries collection disease 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.

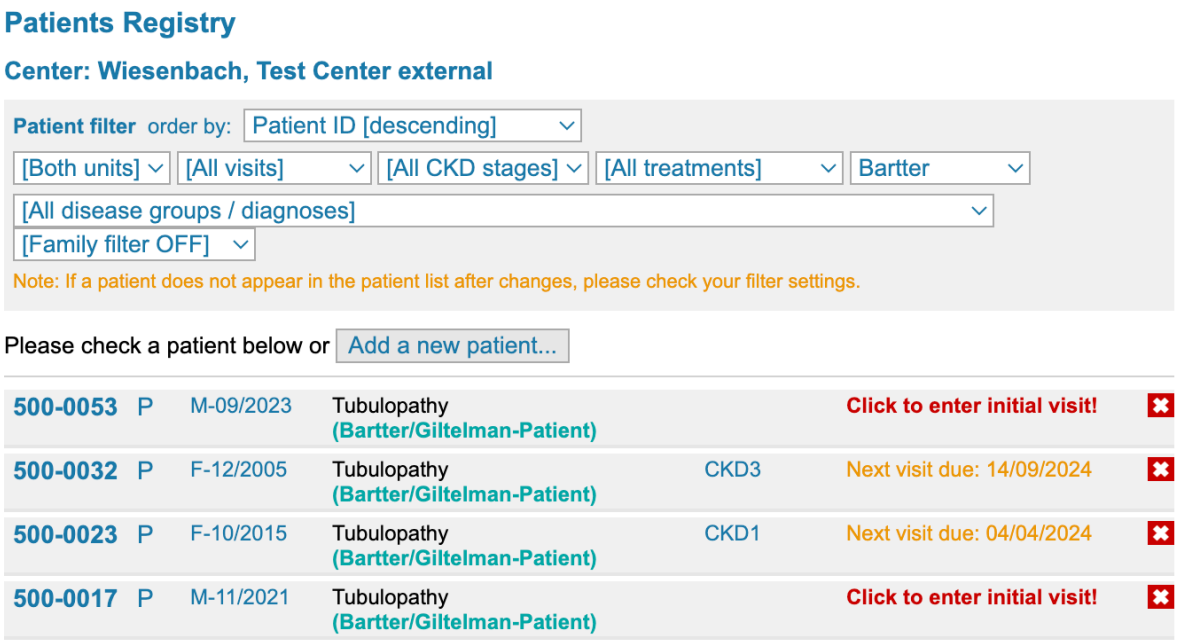

FIGURE 1: ERKReg data entry page

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.

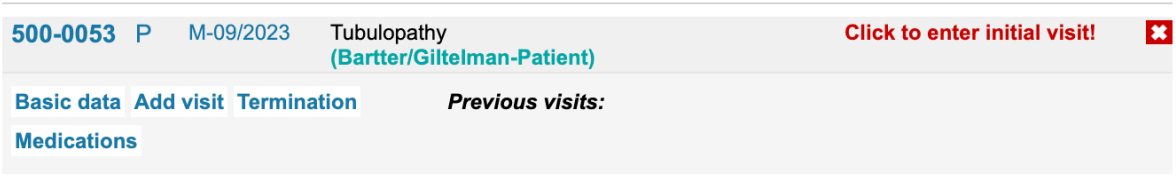

FIGURE 2: Options available upon selection of a BG sub-registry patient.

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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 BG sub-registry (Figure 3).

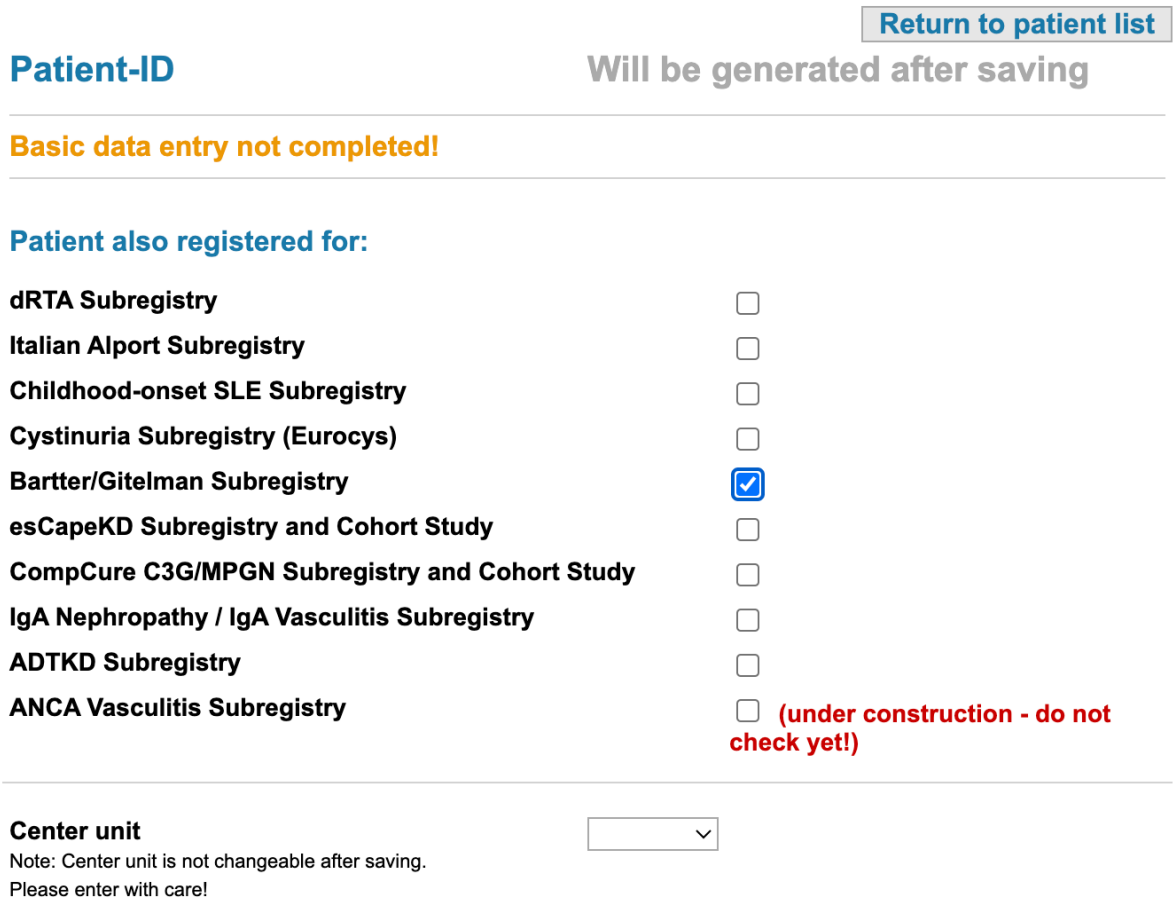

FIGURE 3: Adding a new patient to the BG sub-registry.

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 BG 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 BG sub-registry are depicted in Figure 5.

Prenatal history

Number of weeks of gestation: ☐ weeks

Prenatal manifestation: ☐

Birth weight ☐ grams

Acute renal failure in neonatal period ☐

Family history

Family members affected? ☐

Height father ☐ cm

Height mother ☐ cm

Clinical presentation at time of diagnosis

Polydipsia (Thirst) / Polyuria ☐

Failure to thrive ☐

Growth retardation ☐

Salt craving ☐

Muscle weakness ☐

Muscle cramps / Tetany / Carpopedal spasms ☐

Paresthesia ☐

Joint pain/Chondrocalcinosis ☐

Cardiac arrhythmia ☐

Ataxia ☐

Hearing impairment/Sensorineural deafness ☐

Incidental discovery of electrolyte abnormalities ☐

Seizure(s) ☐

Other ☐

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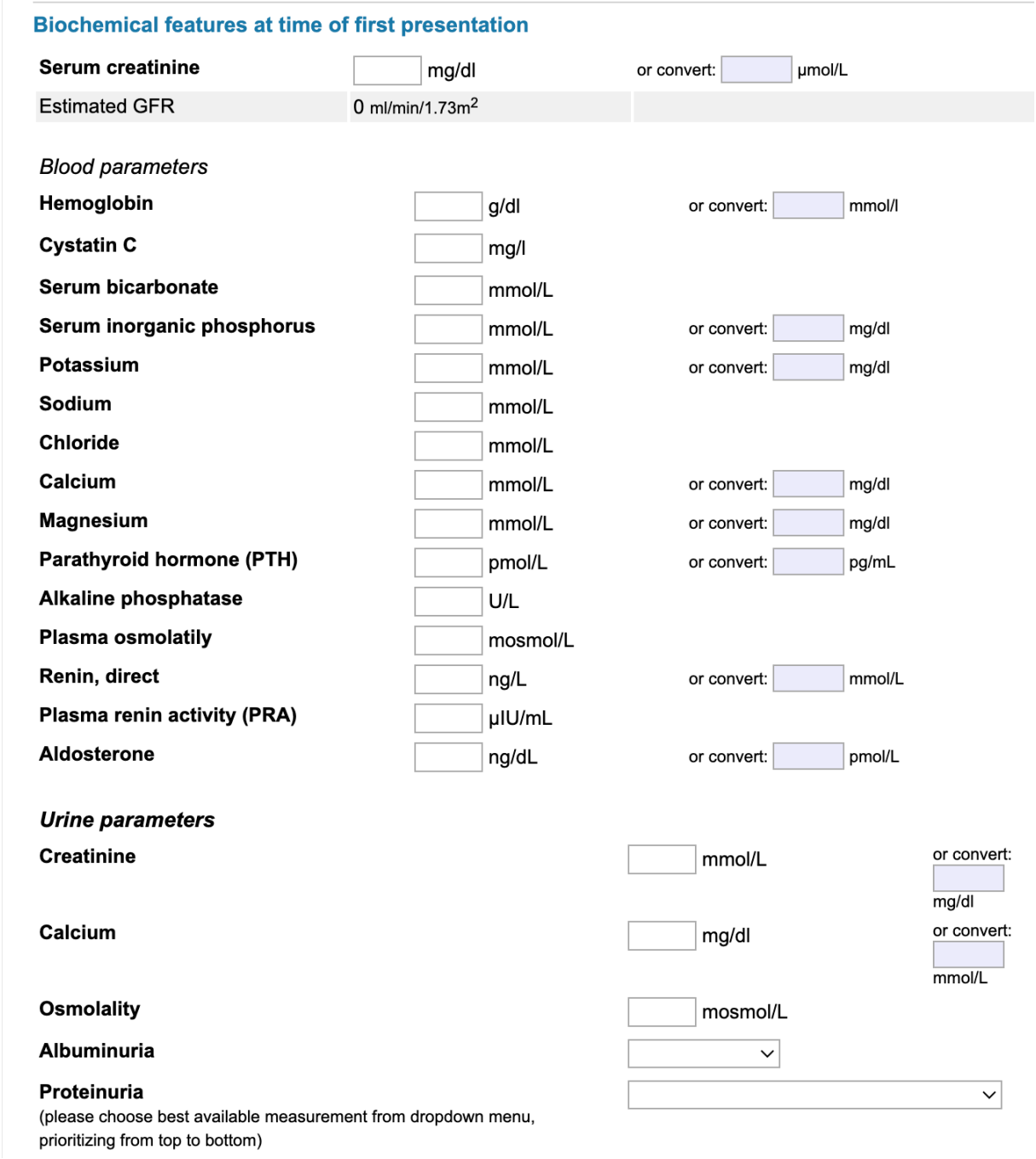

FIGURE 5: BG sub-registry specific data fields collected in the sub-registry.

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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 creatinine 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.

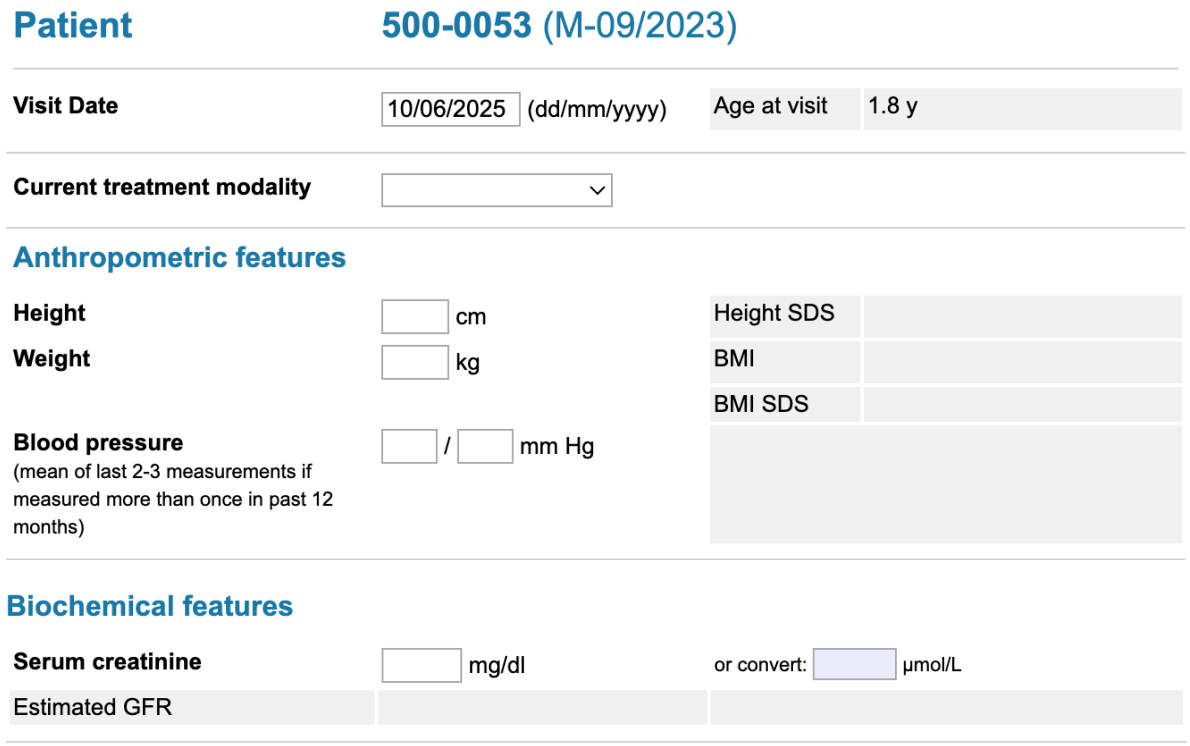

FIGURE 6: BG sub-registry entering visit data.

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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Clinical features

Diarrhea	
Fatique, weakness (severe)	
Abdominal pain	
Sensorineural hearing loss	
Polydipsia (Thirst) / Polyuria	
Failure to thrive	
Salt craving	
Muscle weakness	
Muscle cramps / Tetany / Carpopedal spasms	
Paresthesia	
Joint pain/Chondrocalcinosis	
Cardiac arrhythmia	
Ataxia	
Seizure(s) [info]	
Other	

Blood parameters

Hemoglobin		g/dl	or convert:	mmol/l
Cystatin C		mg/l		
Serum bicarbonate		mmol/L		
Serum inorganic phosphorus		mmol/L	or convert:	mg/dl
Potassium		mmol/L	or convert:	mg/dl
Sodium		mmol/L		
Chloride		mmol/L		
Calcium		mmol/L	or convert:	mg/dl
Magnesium		mmol/L	or convert:	mg/dl
Parathyroid hormone (PTH)		pmol/L	or convert:	pg/mL
Alkaline phosphatase		U/L		
Plasma osmolatily		mosmol/L		
Renin, direct		ng/L	or convert:	mmol/L
Plasma renin activity (PRA)		µIU/mL		
Aldosterone		ng/dL	or convert:	pmol/L

Urine parameters

Creatinine		mmol/L	or convert: mg/dl
Calcium		mg/dl	or convert: mmol/L
Osmolality		mosmol/L	
Albuminuria			
Proteinuria (please choose best available measurement from dropdown menu, prioritizing from top to bottom)			

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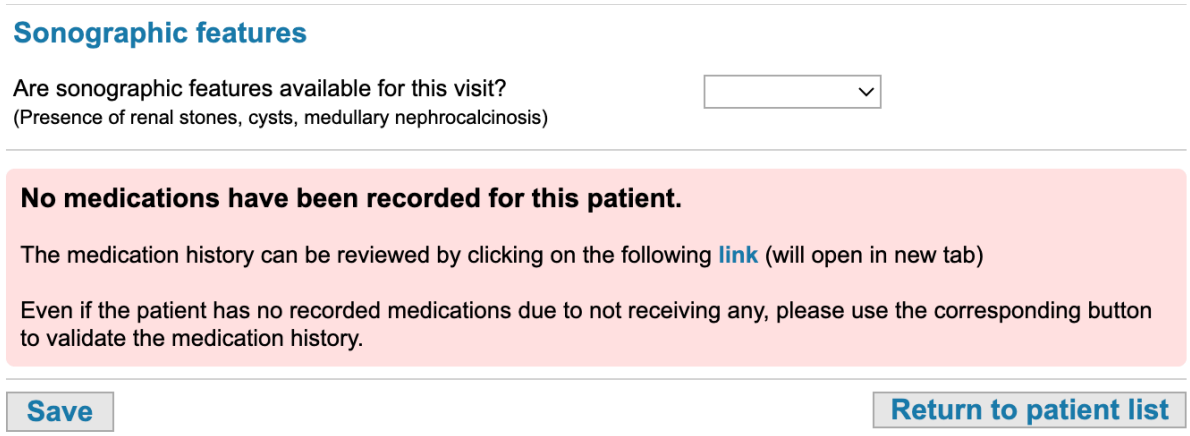

FIGURE 7: BG sub-registry entering visit data.

TERMINATION

Patients can be terminated if the follow-up does not take place anymore or if the patient passes away (Figure 8).
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.

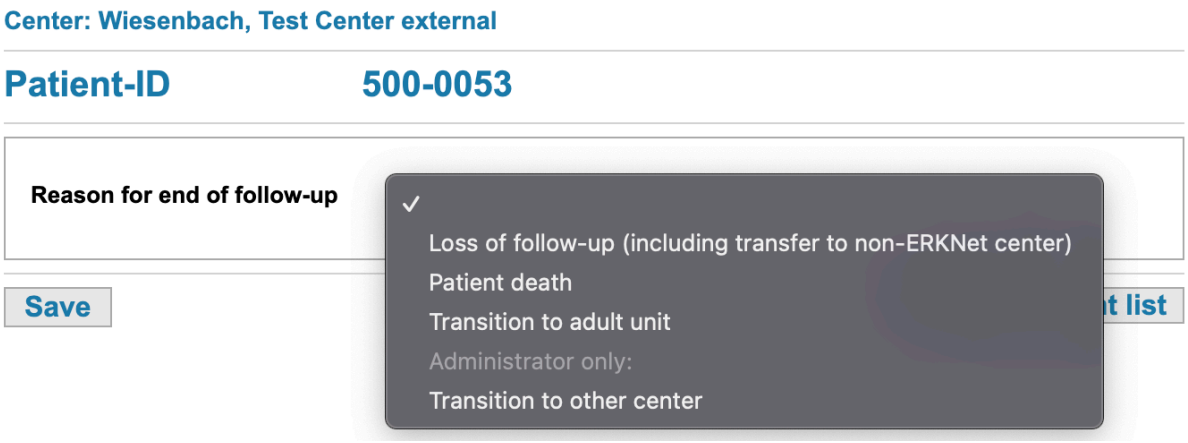

FIGURE 8: BG sub-registry entering visit data.

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MEDICATIONS

Under the medications module, specific medications usage information is collected (Figure 9).
New medications can be added and existing data can be modified (by clicking on medication name) to reflect the current treatment of the patient.
For each drug, specify the prescribed dosage, route of admission, frequency of usage and the patient weight at the time of prescription (the weight is used to calculate the dosage/kg/day field displayed in the table in Figure 9).
For ongoing treatments, the stop date should be left empty.
If a change in dosage occurred, please enter a stop date for the current dosage and readd the drug using the new dosage.

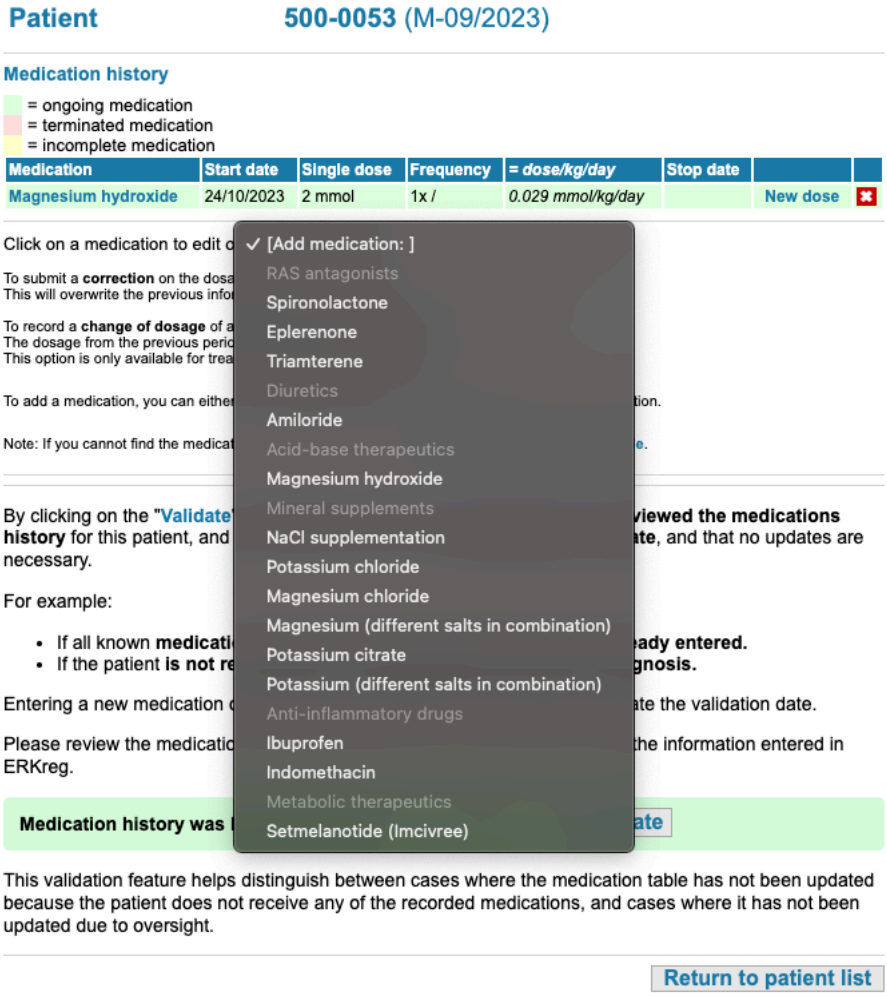

FIGURE 9: BG sub-registry entering medication.

In case of a missing medication – please contact us by email using the contact information found at the end of the manual.

CONTACT

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

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