



ERKReg

The European Rare Kidney Disease Registry

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IgAN / IgAV Subregistry User Manual





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INTRODUCTION

The IgAN / IgAV subregistry is a subregistry of ERKReg. 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 subregistry.

Patients Registry

Center: Wiesenbach, Test Center external

Patient filter order by:

Note: If a patient does not appear in the patient list after changes, please check your filter settings.

Please check a patient below or

500-0032	A	F-03/2000	Glomerulopathy (IgA Vasc/Nephr-Patient)		Click to enter initial visit!	
500-0031	P	M-09/2023	Glomerulopathy (IgA Vasc/Nephr-Patient)	CKD2	Next visit due: 01/03/2026	

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, medications and extracorporeal therapies.

500-0031	P	M-09/2023	Glomerulopathy (IgA Vasc/Nephr-Patient)	CKD2	Next visit due: 01/03/2026	
<input type="button" value="Basic data"/> <input type="button" value="Add visit"/> <input type="button" value="Termination"/>			Previous visits: 29/02/2024			
<input type="button" value="Medications"/> <input type="button" value="Extracorp. Therapies"/>						

Figure 2: Options available upon selection of an IgAN / IgAV subregistry patient.





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 IgAN/IgAV subregistry (Figure 3).

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 IgAN/IgAV subregistry. After which, disease specific fields will be presented (Figure 4).

Patient-ID

Will be generated after saving

Basic data entry not completed!

Patient also registered for:

- dRTA Subregistry
- Italian Alport Subregistry
- Childhood-onset SLE Subregistry
- Cystinuria Subregistry (Eurocys)
- Bartter/Gitelman Subregistry
- esCapeKD Subregistry and Cohort Study
- CompCure C3G/MPGN Subregistry and Cohort Study
- ANCA Vasculitis Subregistry
- IgA Nephropathy / IgA Vasculitis Subregistry

Center unit

Note: Center unit is not changeable after saving.
Please enter with care!

Figure 3: Adding a new patient to the IgAN / IgAV subregistry.

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).

For subregistry patients, additional disease specific fields are collected, the additional fields for IgAN/IgAV are depicted in Figure 5.





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...

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)
- Clinical history
 - Positive family history
 - Clinical examination
 - Biochemical evaluation
 - Immunological evaluation
 - Hematological evaluation
 - Imaging
 - Kidney biopsy
 - Skin biopsy
 - Genetic screening ⁽¹⁾
 - Other methodologies
- ⁽¹⁾ Please check even if results negative or pending

Clinical presentation at time of diagnosis

Height cm

Height SDS	
------------	--

Weight kg

BMI	0 kg/m ²
-----	---------------------

Blood pressure / mm Hg

BMI SDS	
---------	--

Clinical presentation [\[info\]](#)

Figure 4: Basic information collected for all registry patients.





Biochemical features at time of diagnosis

Serum creatinine mg/dl or convert: $\mu\text{mol/L}$
 Estimated GFR 0 ml/min/1.73m²

Comorbidities

HIV \downarrow
 Celiac Disease \downarrow
 Inflammatory Bowel Disease (IBD) \downarrow
 Liver Cirrhosis \downarrow
 Familial Mediterranean Fever (FMF) \downarrow
 Rheumatic Disease \downarrow
 Other

Habits

Smoking Status \downarrow

Surgeries

Did the patient undergo tonsillectomy? \downarrow

Histopathological diagnostics

Date of kidney biopsy (dd/mm/yyyy)

Oxford Classification

Is the Oxford Classification available? \downarrow

Immunofluorescence results

Complement 3 (C3) \downarrow
 Complement 4d (C4d) \downarrow
 Immunoglobulins \downarrow

Proteinuria at time of biopsy g/24h or convert: g/m²/d
 Urine Protein:Creatinine ratio at time of biopsy g/g or convert: g/mol
 Urine Albumin:Creatinine ratio at time of biopsy g/g or convert: mg/mmol

Thrombotic microangiopathy (TMA) \downarrow
 Immunosuppression prior to biopsy \downarrow
 RASi at time of biopsy \downarrow

IgAN-PT (Prognosis score) 0

Figure 5: IgAN / IgAV specific data fields collected in the subregistry.

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 $\mu\text{mol/L}$ 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 $\mu\text{mol/L}$ and the data is only saved using the SI units.

Patient

500-0032 (F-03/2000)

Visit Date

(dd/mm/yyyy)

Age at visit

y

Current treatment modality

Anthropometric features

Height

cm

Weight

kg

BMI

Blood pressure

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

/ mm Hg

Biochemical features

Serum creatinine

mg/dl

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

Estimated GFR

Figure 6: Entering visit data information.

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.



Blood parameters

Serum LDL cholesterol

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

mg/dl

or convert: mmol/l

Serum bicarbonate

mmol/L

Hemoglobin

g/dl

or convert: mmol/l

Immunomarker parameters

Serum IgA

mg/dL

Quality and Performance Indicators

Proteinuria

(please choose best available measurement from dropdown menu, prioritizing from top to bottom)

Date of proteinuria measurement

(dd/mm/yyyy)

Protein/Creatinine ratio

g/g or convert: g/mol

Hematuria

Is patient currently on statin therapy?

Is the patient receiving RAS antagonist therapy?

(ACE inhibitor / AT1 receptor blocker / mineralocorticoid receptor blocker)

Does the patient receive SGLT-2 inhibitor therapy?

(e.g. empaglifozin, dapaglifozin, canaglifozin)

Is the patient receiving any blood pressure lowering treatment?

Next follow-up scheduled in:

Figure 7: Biochemical parameters for patients enrolled in the IgAN / IgAV subregistry.

At the end of the visit, please enter the next scheduled follow-up visit (as depicted in Figure 7). Please use the following instructions for the next scheduled visit interval:

- 3 Months – Patients with a rapidly-progressive disease course (defined as $\geq 50\%$ decline in eGFR over three months or less) or first visit after transplantation.





- 6 Months – Patients within the first year of diagnosis or first visit after the initiation of a new medication, or patients with a $\geq 25\%$ decline in eGFR over a year or less or an increase of 0.5 in urine protein:creatinine over a year or less.
- 1 Year – The default interval.
- 2 Years – IgAV patients with eGFR > 90 ml/min/1.72m² and at least two years without significant proteinuria (defined as urine protein:creatinine < 0.2 for children or < 0.5 for adults), hematuria or clinical features suggesting of IgAV (palpable purpuric rash, arthralgia/arthritis or episodic abdominal pain not otherwise explained).

If a biopsy was performed, additional fields about the biopsy information are presented – including the Oxford MEST-C classification (for both IgAN and IgAV) and additional parameters required for the IIgAN-PT prognosis score (Figure 8).

Please note - the score is only calculated once all fields are filled in.

Biopsy parameters

Biopsy performed since last visit?	<input type="text" value="Yes"/>
Date of kidney biopsy	<input type="text"/> (dd/mm/yyyy)
Serum creatinine at time of biopsy	<input type="text"/> mg/dl or convert: <input type="text"/> $\mu\text{mol/L}$
24h proteinuria	<input type="text"/> g/24h or convert: <input type="text"/> g/m ² /d
Is the Oxford Classification available?	<input type="text" value="Yes"/>

Oxford Classification:

M - Mesangial Cellularity	<input type="text" value="M0"/>
E - Endocapillary Hypercellularity	<input type="text" value="E0"/>
S - Segmental Sclerosis	<input type="text" value="S0"/>
T - Interstitial Fibrosis/Tubular Atrophy	<input type="text" value="T0"/>
C - (fibro)Cellular Crescents	<input type="text" value="C0"/>

IIgAN-PT (Prognosis score)	0
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Necessary parameters for the calculation of the score:

- Patient blood pressure.
- Proteinuria values.
- Patient height and weight (if providing proteinuria values in g/m²/d).
- Information on RASi and Immunosuppression treatment.
- Oxford classification for biopsy.

Figure 8: Biopsy information for patients in the IgAN / IgAV subregistry.



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.

Patient termination entry

Center: Wiesenbach, Test Center external

Patient-ID 500-0032

Reason for end of follow-up

- Loss of follow-up (including transfer to non-ERKNet center)
- Patient death
- Administrator only:
- Transition to other center

Save

Figure 9: Patient termination entry

EXTRACORPORAL THERAPIES

In extracorporal therapies, you may enter information about specific treatment modalities: plasmapheresis and renal replacement therapies (Figure 10).

By clicking on an existing therapy, you may modify the saved information.



Patient

500-0032 (F-03/2000)

Therapy history

- = ongoing therapy
- = terminated therapy

Therapy	Start date	Kidney affected	Number of sessions	Stop date
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Click on a therapy to edit or

✓ [Add therapy]

Note: If you cannot find the therapy

Renal replacement therapy

owski@med.uni-heidelberg.de.

Plasmapheresis

Figure 10: Extracorporeal therapies for patients enrolled in the IgAN / IgAV subregistry.

MEDICATIONS

Under the medications module, specific medications usage information is collected (Figure 11).

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 11).

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.

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



Patient

500-0031 (M-09/2023)

Medication history

= ongoing medication
 = terminated medication

Medication	Start date	Single dose	Frequency	= dose/kg/day	Stop date	
Enalapril	09/01/2024		x /	/kg/day		✘

Click on a medication to edit or v

To add a medication, you can either search directly in the list or type the first letters of the medication.

Note: If you cannot find the medication on the list, please contact giulia.bassanese@med.uni-heidelberg.de.

Medication *NEW*

Medication Lisinopril

Start date (dd/mm/yyyy)

Single dose mg v

Frequency v per day v

Route p.o. v

Patient's weight kg

Stop date (dd/mm/yyyy)
Leave empty if medication is ongoing

Save and Close

Cancel

Figure 11: Medication module for patients enrolled in the subregistry.

CONTACT:

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

