ERKNet
The European Rare Kidney Disease Reference Network

Frequently Asked Questions

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1. What is ERKReg?

The ERKNet Registry, also known as ERKReg, is a secure web-based application which has four major aims:

(i) to generate epidemiological information,
(ii) to identify current patient cohort for clinical research,
(iii) to explore diagnostic and therapeutic management practices and
(iv) to monitor treatment performance and patient's outcomes.

Moreover, the registry allows to integrate detailed disease-specific registries as extensions to the core database.

2. Who can see the data I enter in the registry?

Are my data visible to the investigators of other Centres?

Access of authorized users to the registry is controlled by assignment of a secure, individualized password. A hierarchical access authorization system is implemented with super-administrator, administrator, sub-administrator andcentre user levels.

Project Management Team ('super-administrator')	This group will have access to the global database.
Sub-Registry Leads ('sub-administrator')	This group will have access to all data in the associated sub-registry.
Center users	This group will have access to their own center´s data.
Investigators	This group may propose analyses of registry data by submitting an Analysis Request Form for review and approval by the Data access committee.

The data entered will only be visible to the investigators of your own centre and will not be shared with other users of the registry, except in aggregated format for benchmarking purposes. Sub-administrators are authorized download data excerpts from patients with the disease(s) of their interest.

3. Where are the data stored?

The data are stored on a commercial server in Germany, unaccessible to non-authorised personnelor entities. Regular back-ups are made. These back-ups are kept in a secured location. They guarantee the protection of (identifiable) data and the security of all information.

4. For how long will data be kept in the ERKReg database?

The data will be kept in the database for at least 15 years. Data will not be destroyed without permission of the user.

5. Can I download the data of my centre?

The lead investigator of the centre is authorized to download at any time all data of the patients tagged as participating in the Registry/Affiliated Registry and use these data without restrictions foranalytical purposes.

6. Which browser is best to use ERKReg?

To maximise your experience with ERKReg, users are highly advised to use modern web-browsers. Recent versions of Google Chrome, Mozilla Firefox and Safari provide the best user experience. Microsoft Edge and Internet Explorer support ERKReg.

7. How to get started with ERKReg?

- ✓ Submit the patient informed constent form to your data protection office/ ethics board for local approval
- ✓ Multilingual research-oriented ICF provided by the coordination office
- ✓ Supportive infomation available from the coordination office
- ✓ Obtain the patient consent in your center
- ✓ Train your doctors in the ambulance and ward in proper informing the patients about ERKReg → hand out the patient information flyer
- ✓ Elaborate an internal system how to track the patients and their next visit
- ✓ Identify a staff member in charge for the data entries into the online registry
- ✓ Keep a local list of the patient`s identity (e.g. excel list with ERKReg pseudonym, name, date of birth)
- ✓ Contact person for centre queries
- ✓ Review your centre performance in patient management
  Continuously online in centre dashboard
  Upon receiving the bi-annual KPI reports

8. How can I request access to ERKReg?

To be able to start, the first requirement for the user is to request a login. For this purpose, pleasesend an e-mail request to the ERKReg Support Team: erkreg@erknet.org

9. How can I log in?

The application can be accessed through web-browsers. For best experience, make sure you are using a supported browser. ERKReg is supported to its full potentials on Google Chrome, MozillaFirefox and Safari, preferably latest version. You can access the registry by the ERKNet page webpage by clicking on www.erkreg.org

When you have an ERKReg login account (which can be used to access multiple ERKNet services),enter username and the password provided by the help desk member and click on the button “Login” .

10. Is patient consent mandatory for enrolling patients in the ERKReg?

Yes, all patients must give at least a consent for data use in ERN registries and databases that willbe recorded by ticking a checkbox indicating the minimum has been collected. The consent formscan be obtained by the ERKNet Central Office.

11. How can I enroll a patient into ERKReg?

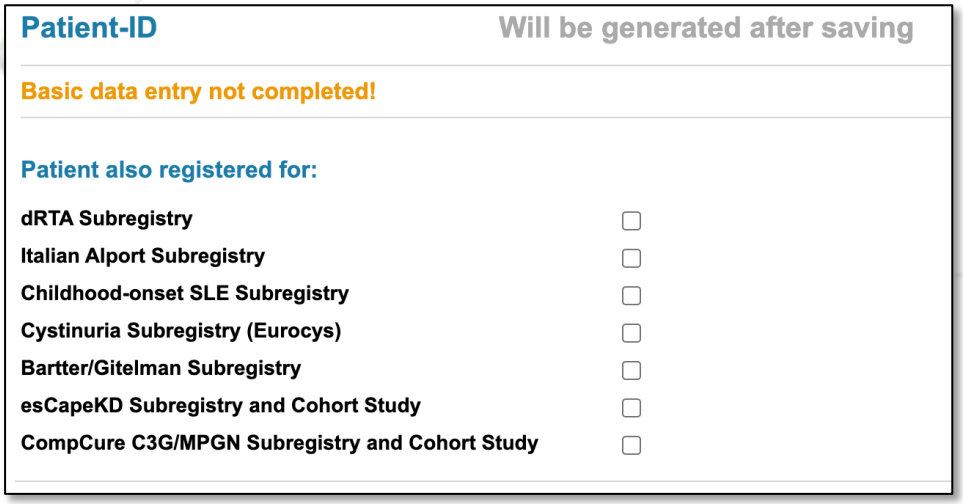
To enroll a new patient from your centre, please click on “Data entry “.

12. How do I know the ID code of my patient?

The patient ID is automatically generated by the system after saving the data. You can see the patient-ID on the left of the menu. We recommend generating a local Excel document with the Name, Surname, date of birth, date of informed consent, together with the ERKReg patient ID. Thiskey document should only be stored on a save hospital server and only be shared among the local users of the registry.

13. Do all ERKReg patients have to be registered in the sub-registries?

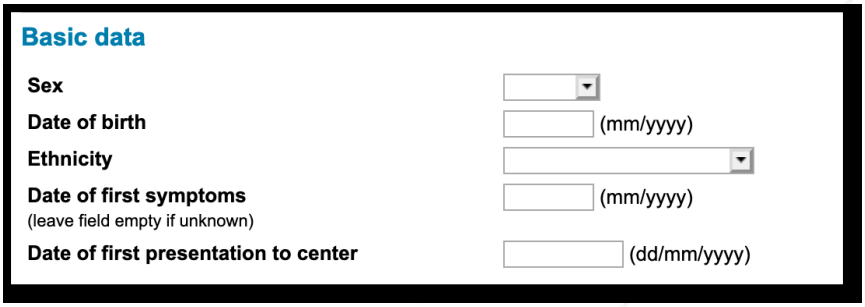
No, only patients with a rare kidney disease for which a sub-registry has been established can be entered into these affiliated registries by selecting the corresponding sub-registry in the basic data form.



14. What date should I indicate in the box 'date of first symptoms’?

The date of first symptoms onset is usually related to the underlying disease. Sometimes the early symptoms of a rare kidney disease coincide with the first presentation at the hospital. Sometimes the first symptoms may appear long before the patient sees the doctor. For most of the CAKUT, the diagnosis can already be made during the pregnancy, as signs can be detected by ultrasound (e.g. oligohydramnios in the case of PUV). In this case, it is possible to indicate a date before birth as "date of the first symptom". If the date of first symptoms is not specified in the medical records, we ask the user

to carefully read patients' medical history and assess when the symptoms started. If it is not possible to trace the date, please leave the box blank.



15. Can I change the DOB of a patient if it would incorrectly be entered and saved?

Yes, no problem! You can change this at any moment. Watch out: If the date of birth is subsequently changed because it may have been entered incorrectly, all visits must be saved again so that the SDS value for height and BMI for weight can be recalculated.

16. How can I choose the "primary renal diagnosis"?

Before registering a patient, we recommend checking the "Renal Orphacode list" (available in Downloads menu). The list contains all the kidney diseases in the registry to clarify the user which diseases can be entered and which not.

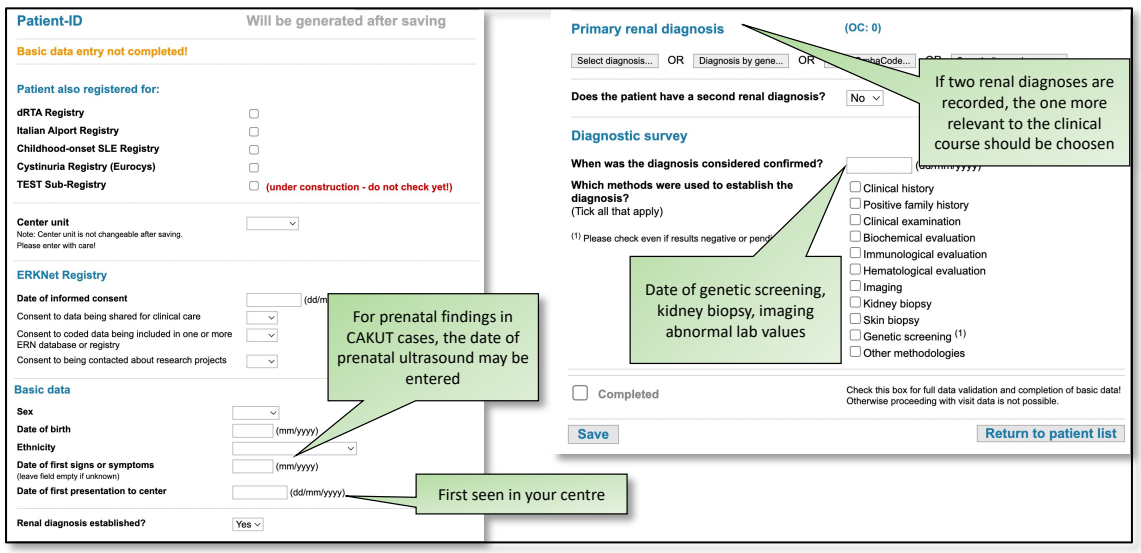
In the data entry menu, the "Primary renal diagnosis" can be chosen through 4 different searchbars:

-"Select diagnosis": the search is made along the Orphanet diagnosis tree, choosing in hierarchicalorder the disease groups down to the entity of choice.

-"Diagnosis by gene": Type in the search bar the name of the mutated gene that caused the disease. The disease(s) associated with the gene will pop up.

- "Enter ORPHA code": Type the Orphanet disease code in the search bar.

- "Search diagnosis name": Enter at least the first 4 letters of the disease name into the search bar.

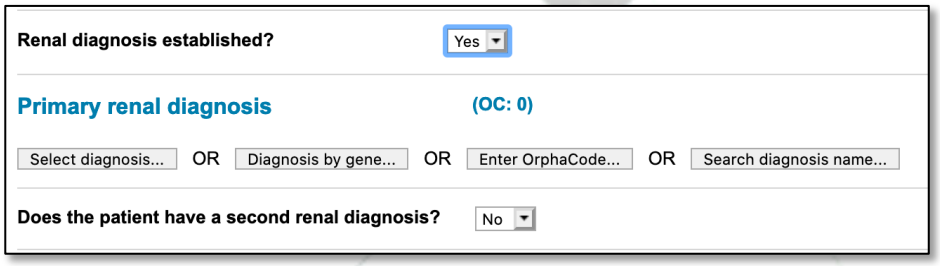


17. What should I do when the renal diagnosis of a patient has not been established yet?

If the kidney diagnosis has not been established, mark "no" in the respective field of the basic entry menu. You can come back and change this entry and at additional information at any time once therenal diagnosis has been established.

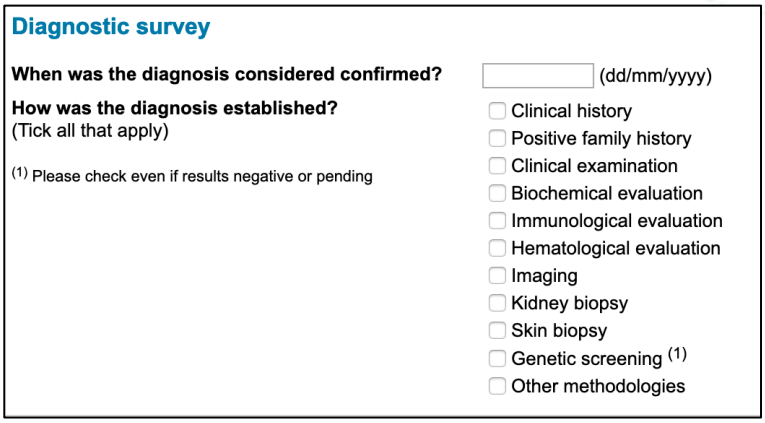
18. How do I know which is the primary and which is the secondary renal diagnosis?

Usually, the primary diagnosis is the one confirmed by genetics. If genetics has not been performed, the primary diagnosis is generally stated first in the patient's letter. In case it is still unclear, which is the main diagnosis, please contact the referring doctor of your ERKNet Centre for clarification.

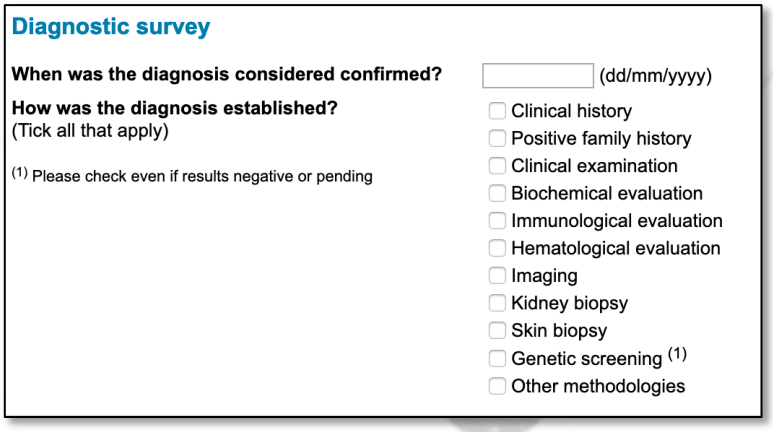


19. How do I know when the diagnosis was confirmed?

This depends on the type of disease: sometimes it requires a genetic test or a kidney biopsy result,in other cases the physician in charge can make a diagnosis based on ultrasound or on clinicalgrounds. To help in a general way, we can state that a glomerulopathy´s diagnosis is usually confirmed by genetics or less frequently by biopsy results. For tubulopathies, ciliopathies and metabolic nephropathies, the diagnosis is confirmed by genetics. For the thrombotic microangiopathies, the diagnosis is confirmed by immunohistological results and/or genetics results. For CAKUT by cystouretrography or radiological examination. Sometimes, it takes years before the genetic cause is found; in these cases, it makes sense to choose an earlier date with different criteria.
It is always advisable to ask the referring physician of the ERKNet Centre for support in case of doubt.



20. How do I know how the diagnosis was established?



Diagnostic survey

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

How was the diagnosis established?
(Tick all that apply)

(1) Please check even if results negative or pending

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

Medical diagnosis is the process of determining which disease or condition explains a person's symptoms and signs. The information required for diagnosis is typically collected from a history and physical examination of the person seeking medical care. Often, one or more diagnostic procedures, such as medical tests, are also done during the process. Here below you will find listed a number of variables that could help you to establish a diagnose.

Clinical history: a narrative or record of past events and circumstances that are or may be relevant to a patient's current state of health. For instance, a patient with Alport disease could tell that he may could experience the presence of blood in urine.

Positive family history: A family medical history can identify people with a higher-than-usual chance of having common diseases. These disorders are influenced by a combination of genetic factors, environmental conditions, and lifestyle choices. For instance, the Autosomal dominant polycystic kidney disease (ADPKD) is an inherited condition. A patient with ADPKD could have one or more relatives who are affected by the same disease.

Clinical examination (or physical examination): helps the medical doctor to determine the general status of patient's health. The exam also gives the chance to talk to patients about any ongoing pain or symptoms that they are experiencing. For instance, signs and symptoms of nephrotic syndrome include: Severe swelling (edema), particularly around the eyes and in ankles and feet. Weight gain due to fluid retention.

An evaluation of biochemical, hematological and immunological parameters helps the doctor to establish a diagnosis.
For Biochemical we intend an electrolyte panel, also known as a serum electrolyte test, is a blood test that measures levels of the body's main electrolytes. This could be important in a Tubulopathy where we expect variation of the electrolytes in blood and urine (Na, K, Ca, Mg, etc.). We intend also all the other lab test (e.g. LDH, renal function, pancreatic and liver enzymes) that are not hematological or immunological parameters.

Hematological parameters are red blood cell count (RBC), white blood cell count (WBC), platelet count, hemoglobin, hematocrit, mean volume (MCV), etc. For instance, a Hemolytic-uremic syndrome (HUS) is an acute, fulminant disorder characterized by thrombocytopenia (low platelet count) and microangiopathic hemolytic anemia (low RBC).

For Immunologic blood test, are intended any of a group of diagnostic analyses of blood that employ antigens (foreign proteins) and antibodies (immunoglobulins) to detect abnormalities of the immune system. These tests are used for detecting autoimmune disorders. Please take a look here below at the immunological diagnostics it may be requested.

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Glomerulopathies:

Diagnosis	C3 glomerulonephritis	Dense deposit disease	Primary IgA nephropathy (Berger disease)	Immune complex associated membranoproliferative glomerulonephritis, with complement abnormalities	Immune complex associated membranoproliferative glomerulonephritis, not otherwise specified	Membranous nephropathy	Lupus like glomerulopathy ('full house' nephropathy)	Adult-onset Still disease	Buerger disease	Dermatomyositis
Orphacode	329918	93571	9999982	329903	9999975	97560	9999981	829	36258	221
Serum C3	not tested/normal/low	not tested/normal/low	not tested/normal/low	not tested/normal/low	not tested/normal/low	not tested/normal/low	not tested/normal/low			
Serum C4	not tested/normal/low/elevated	not tested/normal/low/elevated		not tested/normal/low/elevated	not tested/normal/low/elevated	not tested/normal/low/elevated	not tested/normal/low/elevated			
Serum IgA			not tested/normal/elevated							
dsDNA							not tested/negative/positive			
ANA				not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	t tested/negative/positi	not tested/negative/positive	not tested/negative/positive
Anti-beta2 Glycoprotein IgC									not tested/negative/positive	
Anti-beta2 Glycoprotein IgA									not tested/negative/positive	
Lupus anticoagulant									not tested/negative/positive	
Anti-Ro (SSA)										
Anti-RNP										
Anti-phospholipase A2										
Anti-phospholipid/Anti-car	iolipin								not tested/negative/positive	
Anti-CCP										
RF								not tested/negative/positi	not tested/negative/positive	
ACA									not tested/negative/positive	
cANCA				not tested/negative	not tested/negative					
pANCA				not tested/negative	not tested/negative					
Myeloperoxidase-ANCA										
Antiproteinase 3-ANCA										
PLA2 antibodies						not tested/negative/positive				
C3 nephritis factor	not tested/negative/positive	not tested/negative/positive								
CFH autoantibodies	not tested/negative/positive	not tested/negative/positive				not tested/negative				
Anti-scl70									not tested/negative/positive	
Anti PM-Scl										
Anti-RNA polymerase III										
Anti-SM										
Anti-Mi-2										not tested/negative/positive
Anti-GBM										
Cryoglobulin				not tested/negative/positive	not tested/negative/positive					
Anti-Jo-1										not tested/negative/positive

Diagnosis	Dermatomyositis	Juvenile dermatomyositis	Polymyositis	Juvenile polymyositis	Systemic sclerosis	Systemic Lupus Erytematosus	Pediatric systemic lupus erythematosus	Reynolds syndrome	C3 glomerulopathy secondary to MGRS	Proliferative glomerulonephritis with monoclonal immune deposits (PGNMID)	Anti-glomerular basement membrane disease	Immunoglobulin A vasculitis
Orphacode	221	93672	732	93568	90291	93552	93552	779	999970	999971	375	761
Serum C3						not tested/normal/low	not tested/normal/low		not tested/normal/low	not tested/normal/low		
Serum C4						not tested/normal/low/elevate	not tested/normal/low/elevated			not tested/normal/low/elevated		
Serum IgA												not tested/normal/elevated
dsDNA						not tested/negative/positive	not tested/negative/positive					
ANA	not tested/negative/positive				not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive				
Anti-beta2 Glycoprotein IgC						not tested/negative/positive	not tested/negative/positive					
Anti-beta2 Glycoprotein IgA						not tested/negative/positive	not tested/negative/positive					
Lupus anticoagulant						not tested/negative/positive	not tested/negative/positive					
Anti-Ro (SSA)						not tested/negative/positive	not tested/negative/positive					
Anti-RNP						not tested/negative/positive	not tested/negative/positive					
Anti-phospholipase A2												
Anti-phospholipid/Anti-car						not tested/negative/positive	not tested/negative/positive					
Anti-CCP												
RF								not tested/negative/positive				
ACA					not tested/negative/positive			not tested/negative/positive				
cANCA											not tested/negative/positive	
pANCA											not tested/negative/positive	
Myeloperoxidase-ANCA											not tested/negative/positive	
Antiproteinase 3-ANCA											not tested/negative/positive	
PLA2 antibodies												
C3 nephritis factor												
CFH autoantibodies						not tested/negative/positive	not tested/negative/positive					
Anti-scl70					not tested/negative/positive			not tested/negative/positive				
Anti PM-Scl					not tested/negative/positive							
Anti-RNA polymerase III					not tested/negative/positive							
Anti-SM								not tested/negative/positive				
Anti-Mi-2	not tested/negative/positive	tested/negative/positive										
Anti-GBM											not tested/negative/positive	
Cryoglobulin								not tested/negative/positive				
Anti-Jo-1	not tested/negative/positive	tested/negative/positi	tested/negative/positi	tested/negative/positive								

Diagnosis	Granulomatosis with polyangiitis (Wegener)	Eosinophilic granulomatosis with polyangiitis	Microscopic polyangiitis	Pauci-immune glomerulonephritis	Polyarteritis nodosa	Bechet disease
Orphacode	900	183	727	93126	767	117
Serum C3					not tested/normal/low	
Serum C4					not tested/normal/low/elevated	
Serum IgA						
dsDNA						
ANA					not tested/negative/positive	
Anti-beta2 Glycoprotein IgC					not tested/negative/low	ot tested/negative/p
Anti-beta2 Glycoprotein IgA					not tested/negative/low	ot tested/negative/p
Lupus anticoagulant					not tested/negative/low	ot tested/negative/p
Anti-Ro (SSA)					not tested/negative/positive	
Anti-RNP						
Anti-phospholipase A2						
Anti-phospholipid/Anti-car						
Anti-CCP					not tested/negative/positive	
RF					not tested/negative/positive	
ACA						
cANCA	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive		
pANCA	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	
Myeloperoxidase-ANCA	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive		
Antiproteinase 3-ANCA	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive		
PLA2 antibodies						
C3 nephritis factor						
CFH autoantibodies						
Anti-scl70						
Anti PM-Scl						
Anti-RNA polymerase III						
Anti-SM						
Anti-Mi-2						
Anti-GBM	not tested/negative/positive		not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	
Cryoglobulin						
Anti-Jo-1						

Tubulopathies:

Diagnosis	Auto-immune distal renal tubular acidosis	Sjögren disease	TINU Syndrome
Orphacode	9999994	289390	91500
Serum C3	not tested/normal/low	not tested/normal/low	
Serum C4	not tested/normal/low/elevated	not tested/normal/low/elevated	
Serum IgA			
dsDNA	not tested/negative/positive		
ANA	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive
anti-Ro (SSA)	not tested/negative/positive	not tested/negative/positive	
anti-RNP	not tested/negative/positive		
Anti-phospholipase A2			
antiphospholipid antibodies	not tested/negative/positive		
anti-CCP			
RF		not tested/negative/positive	not tested/negative/positive
ACA			
cANCA			
pANCA			not tested/negative/positive
myeloperoxidase-ANCA			
antiproteinase 3-ANCA			
PLA2 antibodies			
C3 nephritis factor			
CFH autoantibodies			
Anti-scl70			
anti PM-Scl			
Anti-RNA polimerasis III			
Anti-Mi-2			
Ati-GBM			
anti-PR3			
Cryoglobulin			
Anti-Jo-1			

TMA:

Diagnosis	Infection-related hemolytic uremic syndrome	Atypical HUS	Methylmalonic acidemia with homocystinuria, type cbl	Hemolytic uremic syndrome with DGKE deficiency	Thrombotic thrombocytopenic purpura
Orphacode	54448	2134	79282	357008	54057
Serum C3	not tested/normal/low	not tested/normal/low	not tested/normal/low	not tested/normal/low	not tested/normal/low
Serum C4	not tested/normal/low/elevated	not tested/normal/low/elevated	not tested/normal/low/elevated	not tested/normal/low/elevated	not tested/normal/low/elevated
anti-dsDNA antibodies					
anti nuclear antibodies (ANA)					
anti-Ro (SSA)					
anti-RNP					
Anti-phospholipase A2					
antiphospholipid antibodies					
C3 nephritis factor					
CFH autoantibodies	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive	not tested/negative/positive
ADAMTS13 activitiy	not tested/10-100%	not tested/10-100%	not tested/10-100%	not tested/10-100%	not tested/<10%
Shigatoxin PCR	not tested/negative/positive	not tested/negative	not tested/negative	not tested/negative	not tested/negative
STEC stool culture	not tested/negative/positive	not tested/negative	not tested/negative	not tested/negative	not tested/negative
STEC serum antibodies	not tested/negative/positive	not tested/negative	not tested/negative	not tested/negative	not tested/negative
Homocysteine			not tested/eleveted		

Imaging: Diagnostic imaging describes various techniques of viewing the inside of the body to help figure out the causes of an illness and confirm a diagnosis. For example, Fetal Posterior Urethral Valves (PUV) are usually detected in a fetus during pregnancy thanks ultrasounds diagnostic or MRI.

A kidney biopsy involves taking one or more tiny pieces (samples) of kidney to look at with special microscopes. The microscopes make it possible to see the samples in greater detail. This investigation is important in the differential diagnosis of glomerulopathies.

A skin biopsy is used to diagnose or rule out skin conditions and diseases. It may also be used to remove skin lesions. This investigation is important to perform in diseases involving kidney and skin like Alport Syndrome or Granulomatosis with polyangiitis.

Genetic screening is a type of medical test that identifies changes in chromosomes, genes, or proteins. The results of a genetic test can confirm or rule out a suspected genetic condition or help determine a person's chance of developing or passing on a genetic disorder. If you do not have yet the results of this test, please click on "pending". You will have the opportunity to enter the results any time!!!

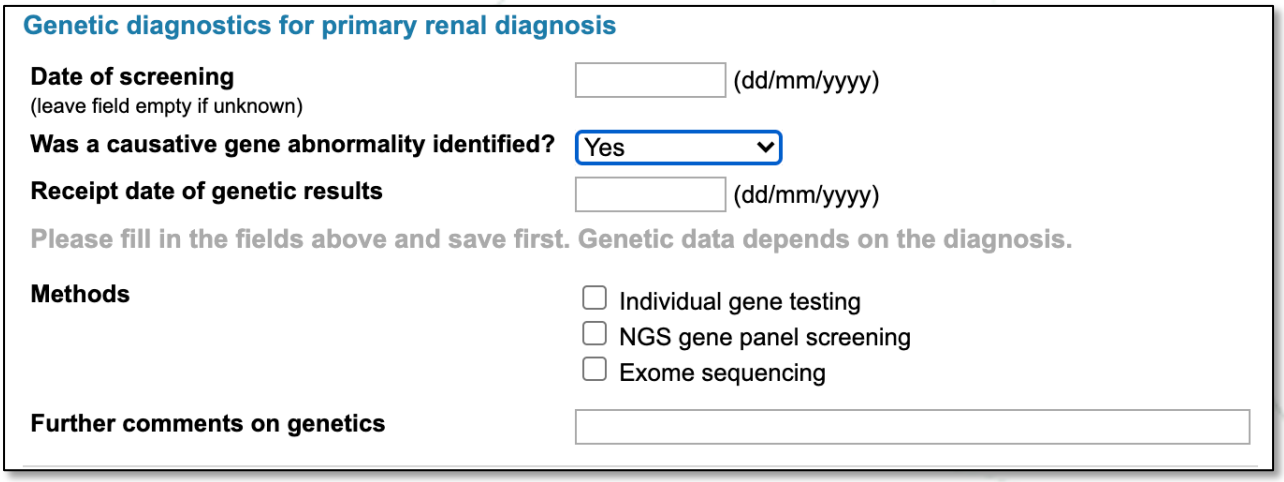
By Other methodologies we mean all analyses that do not include those mentioned above.

21. How can I choose between the different Methods of genetic diagnostics?

There are different methods to perform a genetic diagnostic.
Individual gene testing: Single gene testing is performed to identify changes or variants in a single gene. Basically, when we have a suspect of a disease, for most of the diseases we already know on which gene we could find the mutation. We expect that the mutation is located in that single gene and the investigator test just that single one.

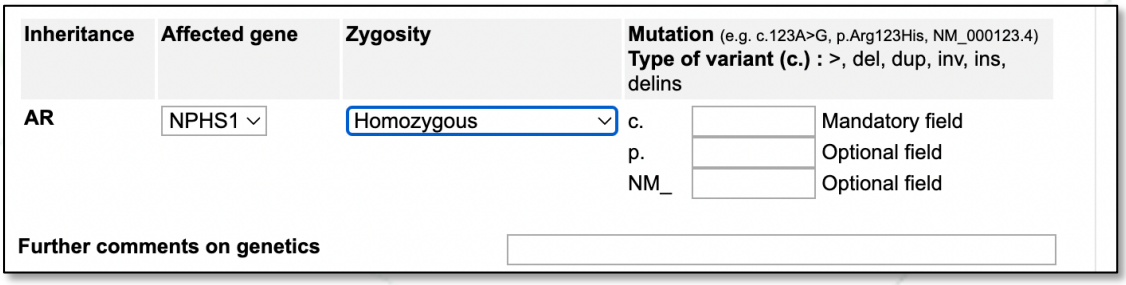
NGS panel screening: The next generation sequencing techniques is the targeted enrichment step where gene panels focus on a limited number of genes. We expect that the mutation is possibly located on 2 or 3 different genes which are usually involved in that disease and we test just these ones.

Exome sequencing: Exome sequencing, also known as whole exome sequencing (WES), is a genomic technique for sequencing all the protein-coding regions of genes in a genome (known as the exome). The goal of this approach is to identify genetic variants that alter protein sequences, and to do this at a much lower cost than whole-genome sequencing. This is a untargeted method and is usually used when the investigator has not idea which gene could be affected.
Nowadays the most used methods are the NGS gene panel screening together the individual gene testing.



22. How do I enter the genetic results?

When you have identified a causative gene mutation, you will be asked to enter the details. Depending on the selected diagnosis, only associated genes will appear in the dropdown list. Please choose the inheritance, the affected gene and the zygosity.
For the mutation you will be asked to enter the coding DNA sequence change (→ c.). This is a mandatory field. Information on the amino acid sequence change (→p.) and the Gen Bank cDNA reference sequence (→NM_) are optional.
If you are unsure, please consult the treating clinician or the geneticist.



23. What to do if my gene is not in the dropdown list

It might happen that new gene mutations have been identified, which are not yet in our list. Please contact us at erkreg@erknet.org to check whether we can add this to the database.

24. What should I indicate when the genetic diagnostic has not been performed?

You should not check "Genetic Screening" under "How was the diagnosis established?".This information may possibly be added in the future if the test should be performed at a later stage.

25. How to enter CAKUT patient?

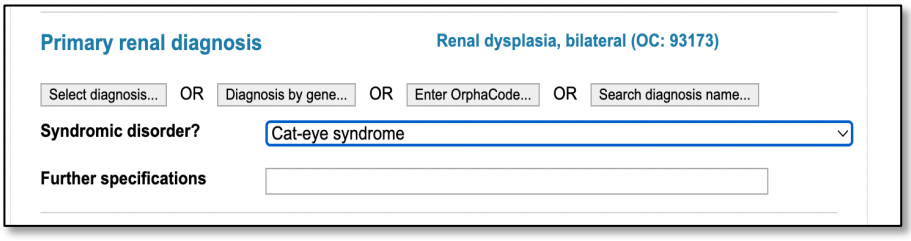
- ✓ All syndromic CAKUT
- ✓ All familial CAKUT
- ✓ All CAKUT cases with impaired renal mass, as evidenced by CKD stage 2 and higher and/or bilateral renal parenchyma abnormalities on ultrasound (hypo/dysplasia, hyperecogenicity)
- ✓ Single kidney (unilateral renal agenesis) with associated anomaly (hypodysplasia and/or urinary tract abnormality) of existing kidney
- ✓ Cystic-multidysplastic kidney with associated anomaly (hypodysplasia and/or urinary tract abnormality) of contralateral kidney (hypertrophic kidney as single symptom)
- ✓ Bilateral VUR 4 or 5 → no unilateral
- ✓ Posterior urethral valves and all other Lower Urinary Tract Obstruction (LUTO)
- ✓ For CAKUT patient ≤2 years information on ultrasound diagnostics is mandatory to fill in

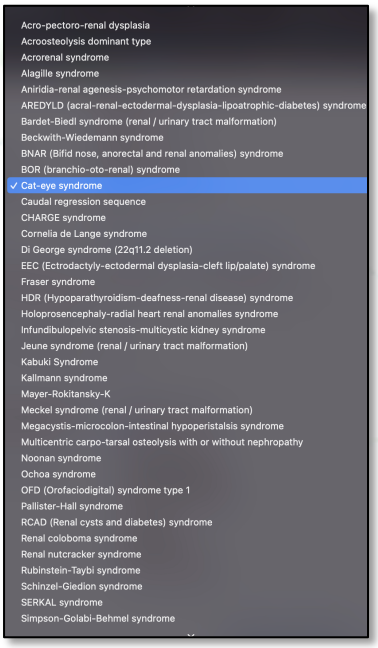
Renal agenesis, unilateral & unilateral multicystic dysplastic kidney

→These patients will have “two renal diagnoses”: please, indicate as first “renal agenesis unilateral” and a second diagnosis to indicate the associated anomaly of the existing kidney (e.g. “hypodysplasia” or “VUR”)

26. How to enter syndromic CAKUT patients

- ✓ Enter the renal phenotype as primary renal diagnosis first
- ✓ Upon saving the entry, you will be asked whether the diagnosis is part of a syndromic disorder
- ✓ → It is technically not possible to select a syndromic CAKUT disorder as primary renal diagnosis





27. How do I choose the date of the “initial visit”?

The date of the initial visit of a registry patient may coincide with the date of the signature of the informed consent but cannot be more than six weeks prior to the date of the informed consent.

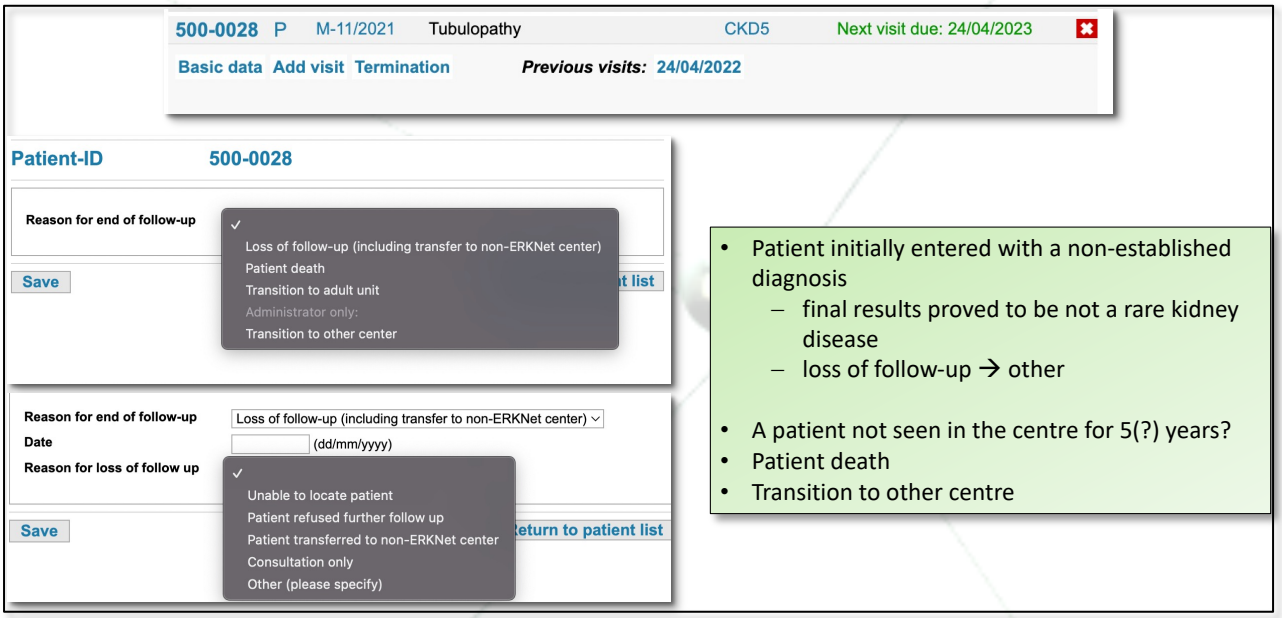
28. How do I know when the next visit is due?

The date of the next visit to be documented is indicated in the patient dashboard.

29. Why is "next visit due" highlighted in different colors?

The statement "next visit due" can be highlighted in different colors depending on the status of the registration. Green means that the patient has been correctly saved in the system. Yellow means that the patient has been saved correctly but needs to be updated or has missing data. Red means that the initial visit of the patient has not been entered yet.

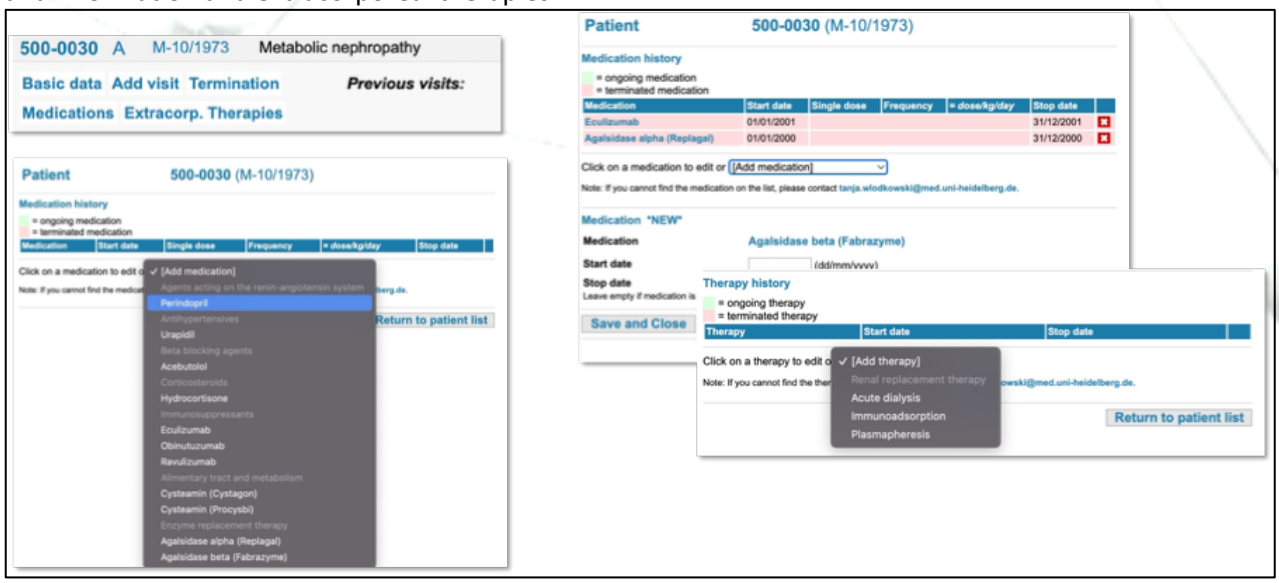
30. When do I have to terminate a patient and how can I do it?



- Patient initially entered with a non-established diagnosis
  - final results proved to be not a rare kidney disease
  - loss of follow-up → other
- A patient not seen in the centre for 5(?) years?
- Patient death
- Transition to other centre

31. What is the medication and therapy database?

For some diagnoses (cystinosis, aHUS, cystinuria, pediatric SLE) we are collecting the medication history and information and extracorporeal therapies.

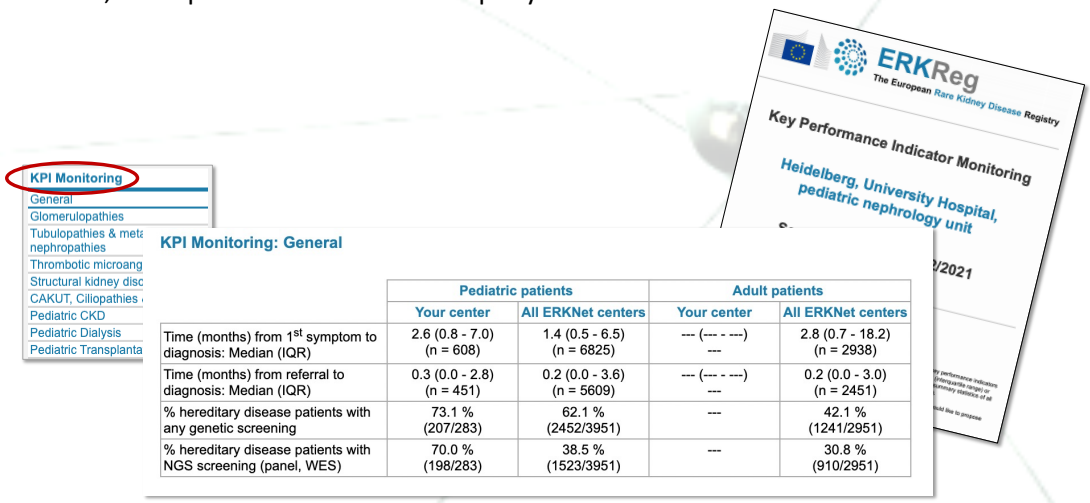


32. What "ID number" should I indicate in the informed consent?

This space must be completed with the automatically generated system patient ID once the patient has been successfully registered.

33. What is the Key Performance Indicator (KPI) Monitoring System

To harmonize patient care across Europes, ERKNet expert centres agreed to adhere to endorsed clinical practice guidelines with respect to diagnostics and treatment of rare kidney diseases. The KPI system allows to monitor the guideline adherence and patient management in those centres.
In addition to basic information, some disease-specific data elements will be asked in ERKReg to measure the KPIs. Each centre has continuous access to its own performance statistics via the ERKReg website. In addition, KPI reports are sent out twice per year to all centres.



	Pediatric patients		Adult patients	
	Your center	All ERKNet centers	Your center	All ERKNet centers
Time (months) from 1st symptom to diagnosis: Median (IQR)	2.6 (0.8 - 7.0) (n = 608)	1.4 (0.5 - 6.5) (n = 6825)	--- (--- - ---) ---	2.8 (0.7 - 18.2) (n = 2938)
Time (months) from referral to diagnosis: Median (IQR)	0.3 (0.0 - 2.8) (n = 451)	0.2 (0.0 - 3.6) (n = 5609)	--- (--- - ---) ---	0.2 (0.0 - 3.0) (n = 2451)
% hereditary disease patients with any genetic screening	73.1 % (207/283)	62.1 % (2452/3951)	---	42.1 % (1241/2951)
% hereditary disease patients with NGS screening (panel, WES)	70.0 % (198/283)	38.5 % (1523/3951)	---	30.8 % (910/2951)

34. How can I contact the IT support?

In case you have any technical difficulty logging into the ERKReg or using any of its functions, please contact the support team via email erkreg@erknet.org. Please sendyour feedback, suggestions, or complaints to the support team.