# Lupusnephritis Registry (LN Registry)

General information on Lupusnephritis Registry
LN Registry is a joint project between European Society for Paediatric Nephrology (ESPN) and The European Rare Kidney Disease Reference Network (ERKNet). In this network several expert pediatric and adult nephrology centers are working together to provide the best care to patients with rare kidney diseases.

One of the objectives of ERKNet is to improve knowledge on rare kidney diseases. Several registries (ERKReg) are started to gather information on the natural course and long-term outcome of the patients. For some rare kidney disease more specified databases are constructed. All data are anonymized following the GDPR rules and legacy.

The main purposes of the ERKReg are: (1) to collect data how many patients with rare kidney diseases are treated across Europe (2) to find and inform physicians on the availability of novel therapies (3) to help optimizing and monitoring the quality of patient care, (4) to provide a platform for sub-registries of rare kidney diseases for which more detailed knowledge is needed. ERKNet partners, but also centers from outside the Network can join ERKReg.

https://www.erknet.org/index.php
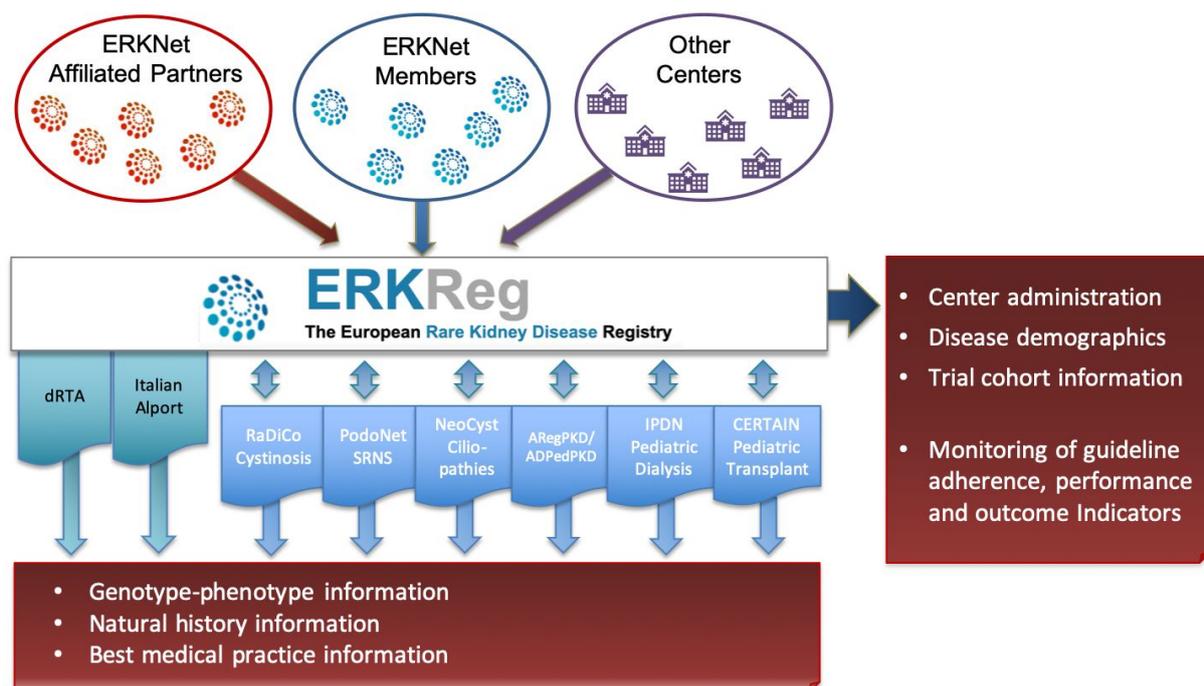
LN registry is a secured web-based application. The access of authorized users is controlled by assignment of a secure, individualized password. The data entered will only be visible to the investigators of the center. The data are stored on a commercial server in Germany, inaccessible to non-authorized personnel. Regular back-ups are made and are kept in a secured location. Data will be kept in the database for at least 15 years, these data will not be destroyed without the permission of the user.
The lead investigator of the center is authorized to download at any time all data of his/her patients and can use these data without restrictions for analytical purposes.

The first requirement of the user is to request a login. This login can be requested by e-mail to the help desk member Tanja Wlodkowski (tanja.wlodkowski@med.uni-heidelberg.de).

Patients must give a consent for data use in ERN registries and databases including the subregistries (Please find attached the informed consent). Non- ERN members will also have to give consent for data use for the LN registry.
Every patient enrolled will have a unique ID code (number of center- ID number as 001-001). A local document with name, surname, date of birth, date of informed consent and ID, will be stored on the hospital server in a secured location.

Basic data collection
The general basic information includes the date of informed consent (1) the consent to data being shared for clinical care, (2) the consent to coded data being included in one or more ERN databases and (3) the consent to being contacted about research projects.

Inclusion criteria will be a diagnosis of LN under the age of 18 years and biopsy proven LN. The basic data are the sex, the date of birth (month and birth year), the ethnicity (popup menu), the date of first symptoms and the date of the first presentation to center.
Renal diagnosis questions include details on type of renal disease (histopathology) and renal function and immunological parameters. Initial clinical symptoms will be asked based on the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI, https://rheumatologie.at/pdf/sledai.pdf) .
When basic data are saved, the unique ID number will be provided and a visit (annual) can be filled in. Annual basic questions encompasses data on anthropometrics and renal function, therapy, extracorporal therapy and complications (hypertension, steroid associated, disease associated comorbidity).

LN Registry
Systemic lupus erythematosus (SLE) is a chronic inflammatory autoimmune disease of unclear etiology, which can be directly mediated by autoantibodies or indirectly affect all organ systems through immune complex deposition. Kidney involvement occurs in 60- 80% of paediatric SLE patients. Children presenting with lupus nephritis (LN) are treated with various immunosuppressive protocols including long-term steroid treatment. Pediatric patients are mostly treated off-label since Data on renal outcome and treatment toxicity are scare. Development specific requirements of paediatric patients are not taken into account. Not only growth and development, but also psychological factors such as puberty and family structures differentiate this group of patients from adults.

The objective of ESPN - ERKNet LN registry is to collect data on long term renal outcome and its determining factors in childhood onset LN (including transition) and detect disease and treatment associated comorbidities in order to allow benchmarking, develop research hypothesis, improve treatment and outcome. The register will be prospectively, retrospective data since 2015 will only be included in the basic sheet, where the symptoms and data at time of diagnosis will be asked. Visit sheets cannot be filled in retrospectively.

LN Registry is an affiliated Registry of ERKReg. This registry can be filled in when patient is included in the basic data sheet. Additionally, patients from Non-ERKReg members will be included in collaboration with ESPN centres.
Specific data are: Biopsy results (histopathology), induction and maintainance therapy, pubertal status, height of the parents.

At every visit (annually) the following data are requested:
- Anthropometric data
- Current therapy : conservative (immunosuppressant, antihypertensive and supportive therapy), dialysis, transplantation
- Activity Index (SLEDAI) including disease related symptoms
- Disease and therapy related comorbidities: Blood pressure, ophthalmologic examination, echocardiogram, fever, diarrhea, gingiva hyperplasy, menstrual disorder, anemia, thrombocytopenia, leukocytopenia and others, steroid associated complications (cushingoid appearance, striae distensae, cataract, osteonecrosis)
- Biochemical data: blood (creatinine, cystatin c, albumine, urea, blood count, immunological parameters: ANA, ANCA, dsDNA-antibody, C3, C4), urine (albumin, protein, hematuria, erythrocyte casts)
- Sonographic data
- Relapses or flare