

<b>ADPedKD: detailed description of data which will be collected in this registry</b>
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**I. Basic data**

1. Patient ID: will be given automatically
2. Personal information
  - Date of informed consent: DD/MM/YYYY
  - Date of birth: MM/YYYY
  - Sex
  - Date of diagnosis (incl. prenatal)?, week of gestational age (if prenatal diagnosis)
  - Date of initial visit at doctor (incl. Obstetrics and Gynecology)
3. Pre- and perinatal period
  - a. Pregnancy
    - Spontaneous or assisted pregnancy? If assisted, specify why: due to fertility problems or due to the parent's wish for genetic testing
    - Were there any prenatal abnormalities? If yes: at which gestational age?
    - Increased renal echogenicity in ultrasound? If yes: at which gestational age?
    - Enlarged kidneys? If yes: at which gestational age?
    - Renal cysts? If yes: at which gestational age?  
If yes: number of cysts left and right? (0; 1 – 5; 6 – 10; > 10; unknown), and size of largest cyst (cm)
    - Other renal abnormalities? If yes, please specify
    - Amniotic fluid: Normal; Oligohydramnios; Polyhydramnios; unknown
    - Other prenatal abnormalities or other relevant information (e.g. fetal MRI)? If yes: please specify
  - b. Perinatal period
    - Gestational age at birth
    - Birth weight
    - Birth length
4. Initial diagnosis
  - a. Initial visit
    - Reason for initial visit : postnatal accidental finding; family screening (due to index patient); presenting features incl. prenatal or unknown
    - ADPedKD-ID of index patient (if available)
    - Method of diagnosis: Ultrasound; CT; MRI; Genetic testing; if other: please specify
    - Imaging method: Ultrasound; CT; MRI or unknown
    - Date of diagnosis (DD/MM/YYYY)
    - Specify if the following is/are present at initial visit (multiple selections possible):  
Abnormal prenatal US /Hematuria / Urinary tract infection / Enuresis /Flank or back pain /Cyst complications (bleeding, infection ...)/Proteinuria or albuminuria /Hypertension / Urolithiasis /Heart valvular abnormalities /Others - please specify

- b. Imaging history: Please add available results of the first imaging (kidney size, cysts...) to the imaging history

5. Genetics

a. Patient's genetic testing

- Has a molecular genetic testing been performed?
- add information on the following genes: is this gene sequenced, if yes: is a sequence alteration/mutation detected? If yes, please specify  
*PKD1, PKD2, PKHD1, HNF1B*
- Other sequenced genes: which? Gene Sequence alteration / mutation? If yes, please specify
- Other genetic data (e.g. gene panel)?

b. Genetic testing of family members

Genetic testing performed in family members?

Please add each family member where genetic testing was performed

6. Family history

a. Parents / Siblings

- Country of origin of father
- Country of origin of mother
- Father's year of birth (YYYY)
- Mother's year of birth (YYYY)
- Consanguinity of parents
- Number of siblings
- Assisted conception for any sibling? If yes, no. of siblings with assisted conception
- If yes: specify fertility treatment and reason for assisted conception: due to fertility problems or the parent's wish for genetic testing unknown
- Miscarriages of patient's mother: if yes, at which gestational age
- Induced terminations of pregnancy of mother: If yes, at which gestational age and for what reason

b. Further family history

- Family history of Tuberous Sclerosis? If yes, please specify affected family member(s)
- Family history of Diabetes mellitus? If yes, please specify affected family member(s)
- Family history of intracranial aneurysm? If yes, please specify affected family member(s)
- Family history of gout? If yes, please specify affected family member(s)
- Family history of colon diverticulosis? If yes, please specify affected family member(s)
- Family history of heart valvular defects? If yes, please specify affected family member(s)
- Family history of liver cyst? If yes, please specify affected family member(s)
- Family history of pancreatic cyst? If yes, please specify affected family member(s)
- Family history of kidney cyst other than ADPKD? If yes, please specify affected family member(s): ARPKD, CAKUT, medullary sponge disease, nephronophthisis, HNF1B-nephropathy, other or unknown disease.

c. ADPKD family history

Please add each family member with ADPKD

## II. Initial visit: visit date (DD/MM/YYYY)

### 1. Patient's status

- a. Patient's status: alive or deceased
- b. Body measurements
  - Height
  - Weight
  - Based on these; BMI, height SDS, BMI SDS will be calculated / updated automatically after saving
  - RR (systolic/diastolic) (RR SDS will be calculated / updated automatically after saving)
  - Heart rate
  - Tanner stages
- c. Participation in study  
Has the patient been included in any other study or registry? If yes, please add name of study and date of inclusion (DD/MM/YYYY)

### 2. Renal Manifestations

- a. Current or previous symptoms
  - Macroscopic Hematuria?
  - Microscopic Hematuria?
  - Proteinuria/albuminuria?
  - Urinary tract infections? no yes unknown
  - Abdominal or flank pain? no yes unknown
  - Urolithiasis? no yes unknown
  - Enuresis? no yes unknown
  - Hypertension? no yes under treatment unknown
  - Cyst complication (infection/bleeding)? no yes under treatment unknown
  - Other renal symptoms? no yes unknown
- b. Imaging history (Ultrasound, CT, MRI)  
Please add each available imaging result to the imaging history
  - imaging date
  - imaging method
  - kidney sizes
  - presence of extrarenal cysts
  - presence of cyst complications

### 3. Extrarenal Manifestations

- a. Cardiovascular involvement
  - Instrumental examination done or organ-related symptoms
  - Ambulant 24hr RR
  - ECG
  - Echocardiography
  - Other cardiovascular symptoms?
- b. CNS

- Instrumental examination done or organ-related symptoms
  - Neurological symptoms? (headache, epilepsy...)
  - MRI CNS done? If yes: Intracranial aneurysm present?
- c. Eyes
- Instrumental examination done or organ-related symptom
  - Hypertensive Retinopathy?
  - Other ophthalmological symptoms? no yes unknown
- d. Other Organs (e.g. lungs, infections)
- Instrumental examination done or organ-related symptoms?
  - Finding / symptom: Date (DD/MM/YYYY) and please specify

#### 4. Laboratory values

- a. Laboratory values: Examination done? If yes:
- Date of examination DD/MM/YYYY
  - Lab taken: after fasting or not
- b. Blood: please mark and fill available parameters (unit or conversion calculator is provided on the website)
- Hematology:
    - WBC (total)
    - Neutrophils
    - Lymphocytes
    - Monocytes
    - Eosinophils
    - Basophils
    - Hb
    - HCT
    - MCV
    - MCH
    - MCHC
    - Platelets
    - Erythrocytes
  - Electrolytes
    - Potassium
    - Sodium
    - Calcium
    - Phosphate
  - Kidney
    - Serum Creatinine
    - Cystatin C
    - Urea or BUN
- GFR: Bedside Schwartz Equation or CKiD Schwartz Equation will be automatically calculated
- Liver
    - GPT
    - GOT
    - Gamma-GT
    - Bilirubine
    - Alkaline Phosphatase
    - Cholinesterase

Quick  
INR  
Albumine  
- Other  
Glucose  
Magnesium  
Ferritin  
Transferrin saturation  
Uric acid  
CRP  
PTH  
HCO<sub>3</sub>  
pH  
Total cholesterol  
HDL  
LDL  
VLDL  
Triglycerides  
Bile acids  
25-OH-Vitamin D  
Osmolality

- c. Urine: please mark and fill available parameters  
- Urine sample (quotients will be automatically calculated)

Sodium  
Creatinine  
Protein  
Albumine  
Protein-creatinine-quotient  
Albumine-creatinine-quotient  
β<sub>2</sub>-Microglobuline  
Osmolality

- Urine 24 h collection (quotients will be automatically calculated)

Volume of 24 h  
Sodium  
Creatinine  
Protein  
Albumine  
Protein-creatinine-quotient  
Albumine-creatinine-quotient  
β<sub>2</sub>-Microglobuline  
Calcium  
Phosphate  
Osmolality

5. Medications

Please add each known medication to the medications history. If medication modalities changes, enter end date for this medication and add a modified new one.

Add: generic name (roll-out system is provided on the website); start date; single dose; frequency; end date

6. Therapies (incl. RRT)

Please add each known therapy to the therapies history, with date of therapy

Kidney: Cyst fenestration, dialysis, nephrectomy, renal transplantation, loss of renal transplant

Other: other urological/surgical procedure: please specify

7. Further developments

Open field box is provided on the website to add any further symptoms, further performed diagnostic procedures (e.g. renal biopsy, ophthalmological examination, lung function test), further developments

III. **Follow-up visits: visit date (DD/MM/YYYY); to be filled in for each follow-up visit over time**

1. Patient's status

a. Patient's status: alive or deceased

b. Body measurements

- Height
- Weight
- Based on these; BMI, height SDS, BMI SDS will be calculated / updated automatically after saving
- RR (systolic/diastolic) (RR SDS will be calculated / updated automatically after saving)
- Heart rate
- Tanner stages

c. Participation in study

Has the patient been included in any other study or registry SINCE THE LAST VISIT? If yes, please add name of study and date of inclusion (DD/MM/YYYY)

2. Renal Manifestations

a. Symptoms SINCE THE LAST VISIT

- Macroscopic Hematuria?
- Microscopic Hematuria?
- Proteinuria/albuminuria?
- Urinary tract infections? no yes unknown
- Abdominal or flank pain? no yes unknown
- Urolithiasis? no yes unknown
- Enuresis? no yes unknown
- Hypertension? no yes under treatment unknown
- Cyst complication (infection/bleeding)? no yes under treatment unknown
- Other renal symptoms? no yes unknown

b. Imaging history (Ultrasound, CT, MRI) SINCE THE LAST VISIT

Please add each available imaging result to the imaging history

- imaging date

- imaging method
- kidney sizes
- presence of extrarenal cysts
- presence of cyst complications

3. Extrarenal Manifestations SINCE THE LAST VISIT

- a. Cardiovascular involvement
  - Instrumental examination done or organ-related symptoms
  - Ambulant 24hr RR
  - ECG
  - Echocardiography
  - Other cardiovascular symptoms?
- b. CNS
  - Instrumental examination done or organ-related symptoms
  - Neurological symptoms? (headache, epilepsy...)
  - MRI CNS done? If yes: Intracranial aneurysm present?
- c. Eyes
  - Instrumental examination done or organ-related symptom
  - Hypertensive Retinopathy?
  - Other ophthalmological symptoms? no yes unknown
- c. Other Organs (e.g. lungs, infections)
  - Instrumental examination done or organ-related symptoms?
  - Finding / symptom: Date (DD/MM/YYYY) and please specify

4. Laboratory values SINCE THE LAST VISIT

- a. Laboratory values: Examination done? If yes:
  - Date of examination DD/MM/YYYY
  - Lab taken: after fasting or not
- b. Blood: please mark and fill available parameters (unit or conversion calculator is provided on the website)
  - Hematology:
    - WBC (total)
    - Neutrophils
    - Lymphocytes
    - Monocytes
    - Eosinophils
    - Basophils
    - Hb
    - HCT
    - MCV
    - MCH
    - MCHC
    - Platelets
    - Erythrocytes
      - Electrolytes
        - Potassium
        - Sodium
        - Calcium

Phosphate

- Kidney

Serum Creatinine

Cystatin C

Urea or BUN

GFR: Bedside Schwartz Equation or CKiD Schwartz Equation will be automatically calculated

- Liver

GPT

GOT

Gamma-GT

Bilirubine

Alkaline Phosphatase

Cholinesterase

Quick

INR

Albumine

- Other

Glucose

Magnesium

Ferritin

Transferrin saturation

Uric acid

CRP

PTH

HCO<sub>3</sub>

pH

Total cholesterol

HDL

LDL

VLDL

Triglycerides

Bile acids

25-OH-Vitamin D

Osmolality

c. Urine: please mark and fill available parameters

- Urine sample (quotients will be automatically calculated)

Sodium

Creatinine

Protein

Albumine

Protein-creatinine-quotient

Albumine-creatinine-quotient

β<sub>2</sub>-Microglobuline

Osmolality

- Urine 24 h collection (quotients will be automatically calculated)

Volume of 24 h

Sodium

Creatinine



Protein  
Albumine  
Protein-creatinine-quotient  
Albumine-creatinine-quotient  
β2-Microglobuline  
Calcium  
Phosphate  
Osmolality

d. Medications SINCE THE LAST VISIT

Please add each known medication to the medications history. If medication modalities changes, enter end date for this medication and add a modified new one.

Add: generic name (roll-out system is provided on the website); start date; single dose; frequency; end date

e. Therapies (incl. RRT) SINCE THE LAST VISIT

Please add each known therapy to the therapies history, with date of therapy

Kidney: Cyst fenestration, dialysis, nephrectomy, renal transplantation, loss of renal transplant

Other: other urological/surgical procedure: please specify

f. Further developments SINCE THE LAST VISIT

Open field box is provided on the website to add any further symptoms, further performed diagnostic procedures (e.g. renal biopsy, ophthalmological examination, lung function test), further developments

#### IV. Termination entry or loss of follow-up

Please fill in date of termination entry/loss of follow-up.

If reason for termination = death, please enter date of death (DD/MM/YYYY)

If not: please add reason:

- patient moved to another pediatric center, not participating in ADPedKD
- patient moved to adult department
- patient's wish (withdrawal of informed consent)
- loss of follow-up
- other, please specify
- unknown

#### V. Transfer of patient to another ADPedKD center

Before using this functionality you should inform the destination center about the transfer. After filling in and saving the form, the patient will be shown in your patient registry as "waiting for transfer". While waiting for transfer, no changes of this patient's data will be possible.

In the destination center's registry the transfer can be accepted or declined:

Accept: The patient's data will completely transferred to the new center (patient will disappear from your registry).

Decline: The patient remains in your registry, the "waiting for transfer" indicator disappears.