ME 5.2.1: The HCP team adopts and implements clinical practice guidelines and decision-making tools developed or adapted by the ERN

Tentative list of performance and outcome indicators

How to get to the KPI-List for your center:
Open ERKReg → Login → KPI-Monitoring

1.	**General**
1.1	Time (months) from 1st symptom to diagnosis: Median (IQR)
1.2	Time (months) from referral to diagnosis: Median (IQR)
1.3	% hereditary disease patients with any genetic screening
1.4	% hereditary disease patients with NGS screening (panel, WES)
2	**Glomerulopathies**
2.1	% patients with hereditary glomerulopathies with genetic screening
2.2	% patients with multidrug resistant NS with comprehensive genetic screening
2.3	% normotensive patients (CKD1-3)
2.4	% patients with office blood pressure in target range
2.5	% patients with immunological glomerulopathies in clinical remission
2.6	% patients with persistent proteinuria who receive RAS antagonist therapy
2.7	% patients with hereditary and multidrug resistant NS on conservative treatment who are prescribed RAS antagonist therapy
2.8	% adult patients with statin therapy
2.9	% adult patients with LDL cholesterol <100 mg/dl
2.10	% children with steroid sensitive idiopathic nephrotic syndrome who are obese
2.11	% children with steroid sensitive idiopathic nephrotic syndrome with height < 3rd percentile
3	**Tubolopathies & metabolic nephropathies**
3.1	% patients with hereditary nephropathy with genetic confirmation
3.2	% children (<16y) with normal length/height SDS
3.3	% patients with renal tubular acidosis maintaining normal serum bicarbonate
3.4	% cystinosis patients in whom at least one cystine blood level has been obtained during the past 12 months
3.5	% cystinosis patients with leukocyte cystine level < 1 nmol/mg protei
3.6	% cystinosis patients with leukocyte cystine level < 2 nmol/mg protein
3.7	% cystinuria patients free of new stones in past 12 months
3.8	% Fabry disease patients with at least one proteinuria measurement in past 12 months
3.9	% adult Male Fabry disease patients receiving enzyme replacement therapy (Replagal, Fabrazyme)
3.10	% Bartter/Gitelman patients with serum K > 2.5 mmol/l
4	**Thrombotic microangiopathies**
4.1	% new-onset HUS patients with complete initial diagnostics pediatric: ADAMTS13, STEC stool culture, Shigatoxin PCR adult: ADAMTS13 only
4.2	% aHUS patients with genetic/autoantibody screening (NGS, CFH autoantibodies
5	**Structural kidney disorders**
5.1	% ADPKD patients with genetic screening
5.2	% ADPKD patients with at least one total kidney volume measurement by MRI
5.3	% normotensive ADPKD patients (CKD1-4) by office BP
5.4	% ADPKD patients screened for intracranial aneurysm when family history is positive

5.5	% TSC patients treated with mTOR inhibitors for indication AML>3 cm
5.6	% TSC patients treated with mTOR inhibitors for neurological indication (SEGA/Epilepsy)
5.7	% VHL patients with children or planning a pregnancy offered prenatal genetic counseling
5.8	% VHL patients with regular audiology assessment (every 2 years, starting at age 5)
6	**CAKUT, Ciliopathies & OUP**
6.1	% familial cystic disease patients with genetic screening
6.2	% patients with PUV detected by prenatal ultrasound
6.3	Mean (SD) of febrile UTIs in past 12 months in OUP / VUR patients
7	**Pediatric CKD**
7.1	% children (<16y) with height > 3rd percentile
7.2	% children (1-16y) with height < 3rd percentile on growth hormone therapy
7.3	% children with BMI < 5th percentile
7.4	% children with BMI > 85th percentile
7.5	Children < 3 years with BMI < 5th percentile receiving enteral feeds
7.6	% patients with office systolic BP < 75th percentile
7.7	% patients with office systolic BP < 50th percentile
7.8	% patients with hemoglobin > 11 g/dl
7.9	Serum phosphorus in normal range for age
7.10	% patients with serum bicarbonate > 20 mmol/L
8	**Pediatric Dialysis**
8.1	% children (<16y) with height > 3rd percentile
8.2	% children (1-16y) with height < 3rd percentile on growth hormone therapy
8.3	% children with BMI < 5th percentile
8.4	% children with BMI > 85th percentile
8.5	Children < 3 years with BMI < 5th percentile receiving enteral feeds
8.6	Serum phosphorus in normal range for age
8.7	% patients with hemoglobin >= 11 g/dl
8.8	% patients with serum bicarbonate >= 20 mmol/L
8.9	PD peritonitis rate
9	**Pediatric Transplantation**
9.1	% children with pre-emptive transplantation
9.2	% children (<16y) with height > 3rd percentile
9.3	% children (1-16y) with height < 3rd percentile on growth hormone therapy
9.4	% normotensive children (by office BP)
9.5	% patients with hemoglobin > 10 g/dl
9.6	% patients without severe metabolic acidosis
9.7	% patients with biopsy proven rejection