Autosomal Dominant Polycystic Kidney Disease (ADPKD)

Diagnosis and risk stratification, BP targets, tolvaptan eligibility/dosing, kidney and liver manifestations, intracranial aneurysm screening, and lifestyle.

2025 KDIGO Clinical Practice Guideline
Evaluation, Management, and Treatment of ADPKD
KDIGO ADPKD Work Group · Kidney Int
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FORMAT *Condensed by clinical theme from recommendations + practice points across 10 guideline chapters.*

1 Diagnosis & Risk Stratification

–	GUIDANCE
Screening	Abdominal ultrasound first-line in at-risk adults, with family history/kidney function/comorbidities (1B); MRI/CT/genetic testing to clarify equivocal cases
US criteria (+FHx)	Diagnose: age 15–39 ≥3 total cysts, 40–59 ≥2/kidney, ≥60 ≥4/kidney. Exclude: 15–39 <1 total cyst, 40–59 <2 total
Genetic testing	Not required for typical presentation; useful for equivocal imaging, negative family history, very-early-onset disease, living-donor evaluation, family planning
Prognosis	Use Mayo Imaging Classification (MIC) via height-adjusted TKV on MRI/CT to predict eGFR decline and kidney failure timing (1B); PROPKD score >6 supports rapid-progression risk when MIC/eGFR trend indeterminate

2 Blood Pressure Management

–	GUIDANCE
Measurement	Standardized office BP preferred (1B); home/ambulatory BP to complement (2B)
Age 18–49, G1–G2	If BP >130/85: target ≤110/75 mm Hg by home monitoring, if tolerated (1D)
Age ≥50, any stage	Target mean SBP <120 mm Hg by office measurement, if tolerated (2C)
First-line agent	RASi (ACEi or ARB) (1C); avoid combining ACEi+ARB+direct renin inhibitor (1B)
Resistant HTN	Investigate secondary causes if ≥3 agents required despite adherence

3 Tolvaptan — Eligibility, Dosing & Monitoring

–	GUIDANCE
Indication	eGFR ≥25 + risk of rapid progression (Mayo class 1C–1E OR eGFR decline ≥3 mL/min/1.73m²/y) (1B)
Dosing	Start 45 mg AM/15 mg PM → uptitrate q≥1wk → target 90 mg AM/30 mg PM
Hold/reduce if	Concurrent strong/moderate CYP3A inhibitor, intolerance, or rising liver enzymes
LFT monitoring	Baseline, then monthly × 18 mo, then every 3 mo (mandatory)
Sick-day plan	Skip doses with volume-depletion risk (diarrhea, vomiting, fever, limited water access, heat); temporary "drug holiday" for travel
Stop before	Pregnancy, lactation, and initiation of KRT; may continue despite age >55 or eGFR falling below 25 if already established

4 Water Intake & Other Agents — NOT Recommended

–	GUIDANCE
Water (no tolvaptan)	2–3 L/d spread through the day if eGFR ≥30 and no contraindication to solute excretion (2D); G4–G5: drink to thirst
mTOR inhibitors	Do NOT use to slow progression (1C)
Statins	Do NOT use specifically to slow progression (2D) — general CV indications still apply
Metformin	Do NOT use specifically to slow progression in nondiabetics (1B)
Somatostatin analogues	NOT for the sole purpose of slowing eGFR decline (2B); may consider for severe symptoms from massively enlarged kidneys
SGLT2i / keto diets	Not established to slow eGFR decline in ADPKD specifically; ketogenic interventions not recommended outside trials

5 Kidney Manifestations

–	GUIDANCE
Chronic pain	Rule out other causes (stones, infection, malignancy); stepwise nonpharm → pharm → minimally invasive (dominant-cyst decortication/sclerotherapy, nerve block) → nephrectomy (last resort)
Nephrolithiasis	Individualize screening; target urine output ≥2.5 L/d; manage per general-population stone guidelines
Gout	Don't treat asymptomatic hyperuricemia. First flare + CKD ≥G3/urate >9 mg/dL/urolithiasis: consider urate-lowering therapy; allopurinol first-line, start low-dose and titrate
Hematuria	Counsel at diagnosis that gross hematuria can occur — avoids unnecessary alarm
UTI / cyst infection	Don't treat asymptomatic bacteriuria. Suspect cyst infection with fever + flank/abdominal pain + CRP ≥50 mg/L or WBC >11 ×10⁹/L. Treat with 4–6 wk of a lipid-soluble antibiotic (fluoroquinolone or TMP-SMX) (2D)

6 Polycystic Liver Disease

–	GUIDANCE
Severe symptoms	Long-acting somatostatin analogues for markedly enlarged livers with severe volume-related symptoms (1B); reassess at 6–12 mo, stop if no benefit
Not effective	Ursodeoxycholic acid, mTOR inhibitors, and V2-receptor antagonists should not be used to slow liver growth
Transplant	Liver transplant for massive PLD without alternatives; combined kidney–liver transplant if eGFR <30 plus liver transplant indication
Liver cyst infection	Empiric 3rd-generation IV cephalosporin ± fluoroquinolone (target Enterobacteriaceae); switch to oral fluoroquinolone after stabilization; duration ≥4 wk; consider percutaneous drainage if unresponsive, immunocompromised, cyst >8 cm, or hemodynamically unstable

7 Intracranial Aneurysm & Other Extrarenal

–	GUIDANCE
Education	All patients educated on thunderclap headache (sudden, "worst of my life," peaks <60 s) → emergency evaluation with CT
Screening indication	Personal SAH history, or family history of ICA/SAH/unexplained sudden death, in those eligible for treatment with reasonable life expectancy (1D); imaging = MRA without gadolinium or CTA
Rescreening	Every 5–10 y if initial screen negative and risk remains high
Aortic/cardiac	Screen for aortic aneurysm only with a family history; manage with smoking cessation, statin, and BP control. Baseline echo if severe/uncontrolled HTN, murmur, cardiac symptoms, or family history of TAA/cardiomyopathy

8 Lifestyle & Diet

–	GUIDANCE
Diet	Water ≥2 L/d; sodium <2 g/d; protein 0.8–1 g/kg/d (avoid >1.3 g/kg/d); calories 25–35 kcal/kg/d; plant-based proteins preferred
Activity	Moderate-intensity activity ≥150 min/wk + strength training ≥1 h twice weekly; caution re: direct organ injury with large kidneys/liver during contact activity
Tobacco / alcohol	Counsel cessation of all tobacco (modifiable ICA risk factor); alcohol ≤1 drink/d (women), ≤2 drinks/d (men)
Adjusted BMI	Subtract estimated total kidney/liver weight from body weight for accurate BMI when organs are massively enlarged

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