Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Nomenclature, diagnosis, prognosis, and prevalence

1 Definition of ADPKD

ADPKD is defined as a group of dominantly inherited disorders associated with kidney cysts and extrarenal manifestations, caused by a pathogenic variant in a gene associated with this disease. The disease is progressive, usually manifesting clinically in adulthood, often resulting in kidney failure.

2 Prevalence

ADPKD is the most prevalent monogenic kidney disease associated with kidney failure, accounting for 5%–10% of individuals with kidney failure worldwide. ADPKD affects all populations, with generally no common pathogenic variant enriching the disease in a geographic area or racial and/or ethnic group.

3 Diagnosing ADPKD

Abdominal ultrasound should initially be employed to screen adults at risk of ADPKD (usually ones with affected parent). Age-specific cyst number criteria have been employed to diagnose or exclude ADPKD. Kidney MRI or CT can also be useful, if available, and genetic testing can be of value (Figure 1).

4 Genetic testing

Although often not required to diagnose ADPKD in a person with a typical presentation, genetic testing can be particularly informative for people with an uncertain diagnosis based on kidney imaging and can aid in obtaining a definitive diagnosis in those with a negative or unknown family history. Due to the heterogeneity in the genetic causes of ADPKD, genetic testing should screen a panel of known PKD genes, not just *PKD1* and *PKD2*.

5 Prognostic markers

Methods to assess risk of rapid progression vary in different regions of the world. Where imaging is available to assess the height-adjusted total kidney volume (htTKV) for age in people with typical disease, the Mayo Image Classification (MIC) is currently the most straightforward way to identify patients with rapidly progressive disease. The slope of eGFR decline and the Predicting Renal Outcome in Polycystic Kidney Disease (PROPKD) score, employing genetics, sex, and clinical factors, are useful if quantitative imaging is not available.

6 Nomenclature

Use a common nomenclature including the disease name followed by the gene name (e.g., ADPKD-*PKD1*) when a genetic diagnosis is available and pathogenicity is well supported. This nomenclature uses the familiar disease name but, by adding the gene name, provides more specific details of the disease in the affected individual. A similar nomenclature is recommended for autosomal dominant polycystic liver disease or ADPLD (e.g., ADPLD-*SEC63*).

7 Incidentally detected kidney and/or liver cysts: Obtaining a diagnosis

In a person with incidentally detected kidney and/or liver cysts (without a family history), detailed clinical and imaging assessment and genetic testing can help obtain a firm diagnosis (Figure 2).

8 Factors associated with disease progression

A range of clinical, genetic, lifestyle, environmental, and chance factors are thought to influence the rate of disease progression in ADPKD (Figure 3).

9 Considerations when testing for ADPKD

Testing for ADPKD in an at-risk adult either by imaging or genetics should employ a patient-centered approach. This should prioritize patients' values and preferences with an understanding of possible negative, as well as positive, outcomes from the testing.

Figure 1

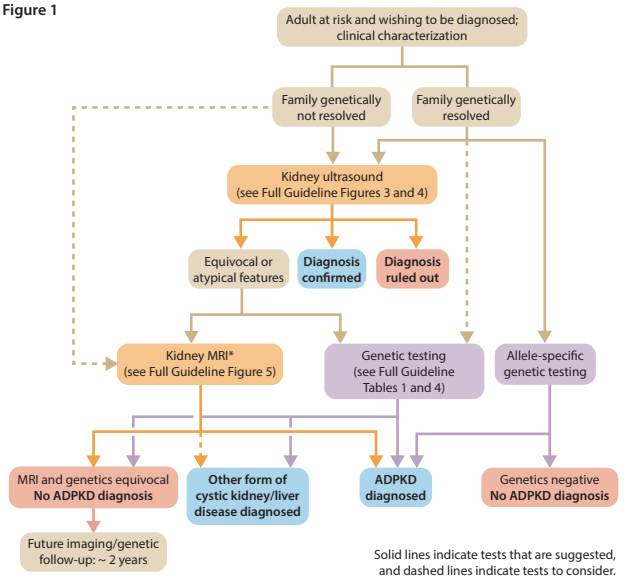
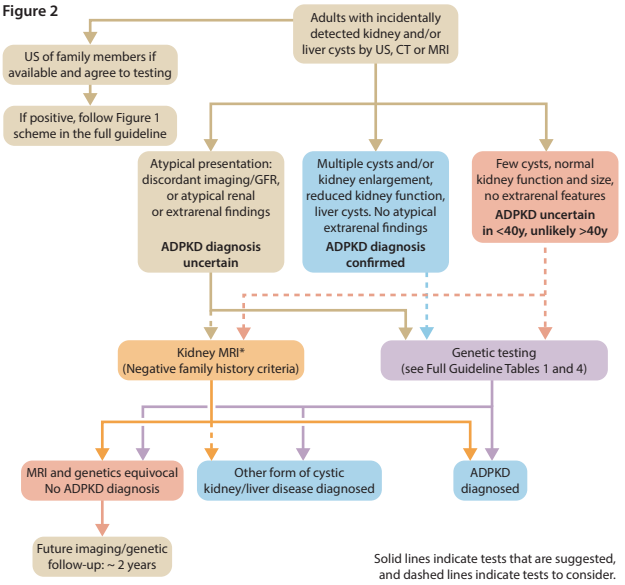


Figure 2



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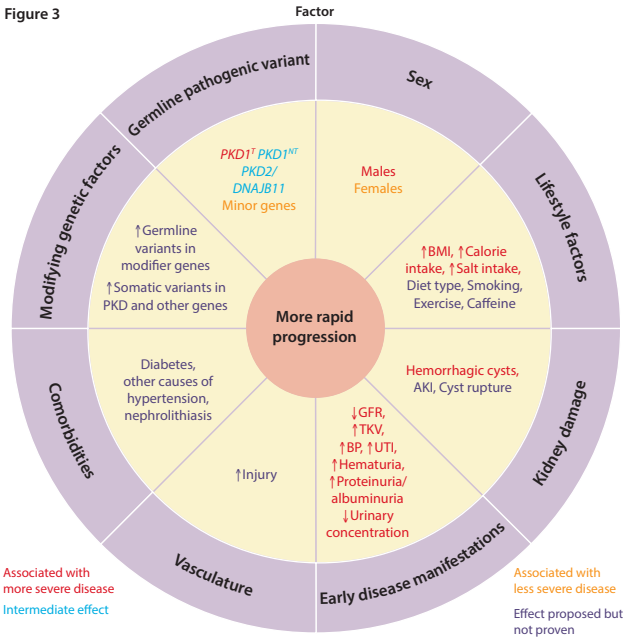


ADPKD, autosomal dominant polycystic kidney disease; CT, computed tomography; MRI, magnetic resonance imaging

Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Kidney manifestations

1 Blood pressure management

Management of high blood pressure (BP) in people with ADPKD should include regular BP monitoring, preferably with home BP measurements (HBPM), dietary and lifestyle modifications, and pharmacotherapy, if indicated (Figure 1).

2 Target blood pressure

- For people with ADPKD aged 18–49 years with CKD G1–G2 and high BP (>130/85 mm Hg), we recommend a target BP of ≤110/75 mm Hg, as measured by HBPM, if tolerated.
- For people with ADPKD aged ≥50 years, with any stage of CKD, we suggest a target systolic BP < 120 mm Hg, as assessed using standardized office BP measurement, if tolerated

3 Pain management

Shared decision-making between the healthcare provider and the person with ADPKD or their caregiver should guide pain-management strategies in ADPKD (Figure 2). This process is expected to reduce the patient's anxiety, increase the patient's cooperation, and respect the patient's personal choices and views.

4 Gross hematuria

Healthcare providers should be aware of the causes and natural history of gross hematuria in people with ADPKD to provide proper guidance and, if appropriate, reassurance.

5 Kidney cyst infection

People with ADPKD who present with fever, abdominal or flank pain, and increased white blood cells and/or CRP should be worked up for kidney cyst infection (Figure 3). In people with ADPKD and kidney cyst infection, we suggest treatment with 4–6 weeks of antibiotic therapy rather than a shorter course.

6 Kidney stones

The management of kidney stones and gout in people with ADPKD should be similar to the general population. Obstructing kidney stones are more challenging to treat in people with ADPKD and should be managed by centers of expertise.

Figure 1

Hypertension in ADPKD		
Monitoring	**Non-pharmacologic interventions**	**Medical management**
• Standardized office BP measurement in preference to routine office BP measurement • HBPM is preferred to office only measurements • Consider ABPM in children and adults with difficult BP control, LVH, proteinuria, or declining kidney function but normal office BP readings • Consider work up for secondary high BP when >3 BP medications are needed in the setting of medication and dietary compliance	• Reduce dietary sodium including minimizing processed foods • Optimize body weight with a healthy diet and regular exercise • Optimize pain management	• Inhibition of RAS provides the cornerstone of BP management and includes the use of an ACEi or ARB • Optimize BP with a 2nd-line agent, if needed • Individualized therapy is indicated

Figure 2

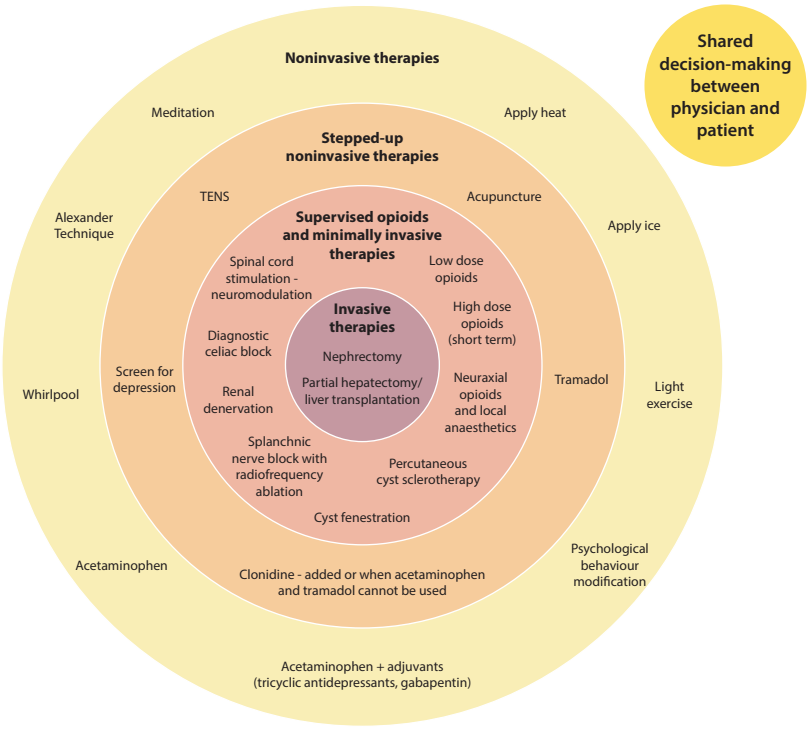
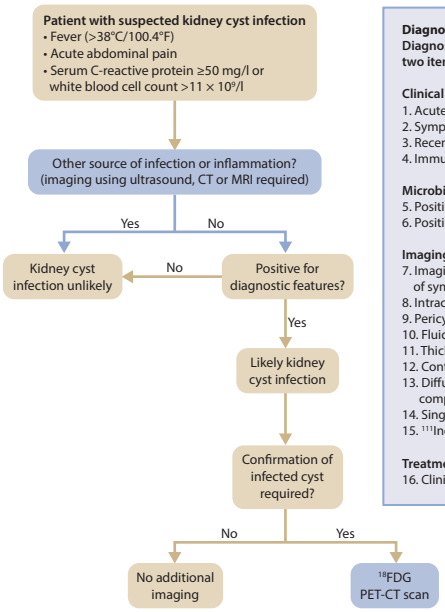


Figure 3



Diagnostic features
Diagnostic features considered positive in the presence of at least two items from at least 2 categories:

Clinical factors
1. Acute pain or tenderness in kidney area
2. Symptoms of urinary tract infection
3. Recent instrumentation of urinary tract
4. Immune compromised patient (including patients on dialysis)

Microbiology
5. Positive urine and/or blood culture
6. Positive cyst fluid culture

Imaging
7. Imaging (ultrasound, CT, or MRI) before and after onset of symptoms demonstrating a new complex cyst
8. Intracystic gas (ultrasound, CT, or MRI)
9. Pericystic inflammation (CT or MRI)
10. Fluid-fluid levels in a cyst (MRI)
11. Thickened cyst wall (CT or MRI)
12. Contrast enhancement in the lining of cyst walls (CT or MRI)
13. Diffusion weighted imaging showing increased cyst density compared to normal cysts
14. Single-photon emission CT with Ga-67 abnormal uptake by a cyst
15. ^{111}Indium-white blood cell scan showing accumulation in a cyst

Treatment
16. Clinical response to antibiotic treatment

ADPKD, autosomal dominant polycystic kidney disease; CKD, chronic kidney disease; CRP, C-reactive protein

Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: CKD management and progression, kidney failure, and kidney replacement therapy (KRT)

1 CKD management

Management of the complications of CKD and lipid lowering therapy for the primary prevention of cardiovascular disease in people with ADPKD should be similar to those in other kidney diseases. HIF-PHIs should not be used to manage anemia in people with ADPKD who are not receiving dialysis as they may theoretically exacerbate cystogenesis. Diabetes management should be the same than in general population. SGLT2i and GLP-1 RAs are not recommended until further data in people with ADPKD are available.

2 Native nephrectomy

Native nephrectomy should only be performed for specific indications, usually in people with ADPKD receiving a kidney transplant (Figure 1). Native nephrectomy should be considered by a multidisciplinary team in a center with sufficient surgical experience; the benefit must outweigh the risk. Ideally, native nephrectomy should not precede KRT and should be unilateral when appropriate.

3 Transplantation

Living-related donor transplantation done preemptively is the optimal treatment for kidney failure in people with ADPKD. Several complications after kidney transplantation are more common in ADPKD (Figure 2). Due to reduced availability of living, related donors in ADPKD families, there is value in evaluation of the extended family, and the wider circle of friends, coworkers, and acquaintances. Potential at-risk, blood-related donors should undergo thorough evaluation to exclude ADPKD.

4 Accurate assessment of weight and BMI

The transplant team should take into consideration the weight (volume) of enlarged cystic organs. During the health-screening phase before kidney transplantation, the body weight should be adjusted for the estimated polycystic kidney and liver weights to arrive at a more accurate indication of the BMI (Figure 3)

5 Dialysis

Peritoneal dialysis and hemodialysis are acceptable dialysis modalities for people with ADPKD with comparable long-term mortality. People at high risk of abdominal hernias should be counseled to avoid peritoneal dialysis. There are no objective criteria for total kidney volume that would predict success or failure of peritoneal dialysis.

6 Clinical trials

Voluntary participation in clinical trials of interventions to slow progression of ADPKD should be offered to all eligible people with ADPKD.

Figure 1

Recurrent and/or severe kidney infection
Symptomatic nephrolithiasis
Recurrent and/or severe kidney cyst bleeding
Intractable pain
Suspicion of kidney cancer
Insufficient space for insertion of a kidney graft
Ventral hernia in the setting of massively enlarged kidneys
Severe symptoms related to massively enlarged kidneys

Figure 2

Post-transplant complication	
New-onset diabetes	Pooled RR 1.92; 95% CI: 1.36–2.70
Erythrocytosis	Recipients with post-transplant erythrocytosis were more likely to have PKD than other kidney diseases (17% vs. 6%; P <0.001)
Valvular heart disease	Greater risk for worsening of tricuspid, mitral and aortic valve regurgitation
Aortic root dilatation	Greater risk for dilation of sinus of Valsalva and ascending thoracic aorta
Subarachnoid hemorrhage	3.8/1000 hospital admission in kidney transplant recipients with ADPKD compared to 0.9/1000 in kidney transplant recipients without ADPKD
Thromboembolic events (DVT, PE)	8.6% of 534 patients with ADPKD vs. 5.8% of 4779 patients without ADPKD after kidney transplantation ($P = 0.009$)
Skin cancers: SCC, BCC, melanoma	Adjusted ORs 1.22, 1.30, 1.21, respectively
Urinary tract infections	Weak evidence only
Cyst infection	Cumulative IR 3%, 6 % and 12% (63% kidney, 37% liver) at 1, 5 and 10 years after transplantation (1.6 episodes per 100 person-years). Increased risk with history of cyst infection before transplantation, HR: 3.47; 95% CI: 1.29–9.31
Colon diverticulitis	Prevalence (2006–2013) in kidney transplant recipients with compared to without ADPKD (2.6% vs 0.8%)

Figure 3

$$\text{Adjusted BMI (ADPKD)} = \frac{\text{Adjusted body weight (kg)*}}{\text{Height (m)}^2}$$

*Adjusted body weight = Measured body weight (kg) – TKV (in kg) – TLV (in kg) + weight of normal kidneys (kg) and liver (kg)

ADPKD, autosomal dominant polycystic kidney disease; BMI, body mass index; GLP-1 RA, glucagon-like peptide-1 receptor agonists; HIF-PHIs. hypoxia-inducible factor-prolyl hydroxylase inhibitors; SGLT2i, sodium-glucose cotransporter-2 inhibitors

Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Therapies to delay the progression of kidney disease

1 Basic actions to delay disease progression

Patients with ADPKD should be encouraged to follow 4 basic actions to delay disease progression: maintain optimal blood pressure and body weight, follow a low-salt diet, and maintain a high-water intake. Evidence from observational studies and clinical trials strongly suggests that achieving these goals delays disease progression and extends the survival of the kidneys and people with ADPKD.

2 Vasopressin receptor antagonist (tolvaptan)

Tolvaptan is recommended in adults with ADPKD and eGFR ≥25 ml/min/1.73 m² at risk of rapidly progressive disease and who have no absolute contraindications (i.e., planning pregnancy/pregnant/breast-feeding; impaired ability to manage aquaretic side effects; urinary tract obstruction; requirement for strong CYP3A inhibitors; significant liver disease other than polycystic liver disease; Figure 1). Potential benefits, harms, and uncertainties regarding long-term treatment with tolvaptan should be discussed.

3 Methods to evaluate rapid disease progression for tolvaptan

The Mayo Imaging Classification (MIC) is recommended to assess rapid progression when imaging is available to measure kidney volumes with subclass 1C to 1E as a criterion for starting tolvaptan (Figure 1). Yearly decline of eGFR ≥3 ml/min/1.73 m² over the previous 3–5 years where there are no other reasons for the decline or a PROPKD score >6 are alternative methods.

4 Management of aquaretic side effects of tolvaptan

Patients starting tolvaptan should be informed that it causes an immediate increase in urine output that requires a matched water intake to avoid dehydration. To improve tolerability, tolvaptan is started at a low dose and titrated upwards (Figure 2); however, the dose can be down-titrated if necessary. Patients should be assured that with time, polyuria decreases to some extent, and the high urine output becomes more accustomed and tolerated. Patients should be counselled to hold tolvaptan when access to water or ability to drink is limited because of travelling or illness, or in situations when large extrarenal losses of water are present, such as episodes of gastroenteritis or in very hot weather.

5 Management of elevations in liver enzymes due to tolvaptan

Liver enzyme elevations occur in approximately 5% of patients within the first 18 months of treatment. To avoid the risk of severe liver injury, all patients should have liver function tests at baseline, monthly for first 18 months, every 3 months thereafter (Figure 3). Tolvaptan should be held immediately when elevations in enzymes occur and discontinued permanently if the elevations reach 3 or more times the upper limit of normal in the absence of other explanations.

6 Other potential adverse events of tolvaptan

Patients should also be informed that tolvaptan causes a rapid but slight increase in serum creatinine which is reversible when the drug is discontinued. Other rare and relatively minor risks of tolvaptan treatment include flares of gout and myalgias with elevations of creatine kinase. All people with ADPKD should have a "sick day plan" that includes skipping doses where there is a transient risk of volume depletion.

7 Increased water intake in the absence of tolvaptan

Increased water intake suppresses vasopressin release and is therefore recommended in all patients with an eGFR ≥25 ml/min/1.73 m². Patients should be provided individualized counseling to drink 2–3 liters of water per day unless there is a contraindication such as a medical condition or a medication interfering with the capacity to dilute urine. Increased water intake is not an alternative to tolvaptan in patients with rapid disease progression.

8 Other therapies

Other therapies (mTOR inhibitors, metformin, statins, somatostatin analogs, SGLT2i, ketogenic interventions, complementary) have not been proven to slow the decline in kidney function in ADPKD, and should not be used for this purpose unless further evidence becomes available.

Figure 1

Initiation of tolvaptan should be offered to adults with ADPKD and:
eGFR ≥25 ml/min per 1.73 m²

AND

Risk of rapid disease progression as indicated by either:
Mayo class 1C to 1E
OR
Historical rate of eGFR decline (≥3 ml/min per 1.73 m² per year)

Figure 2

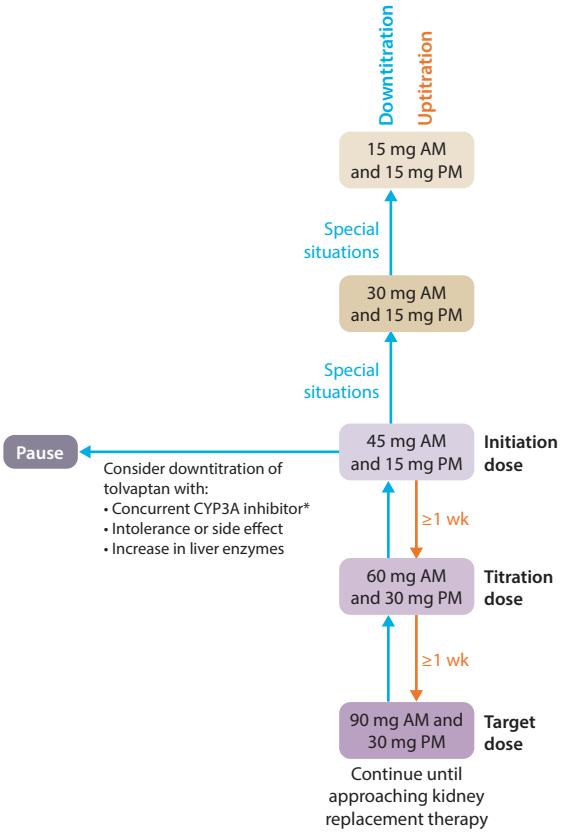
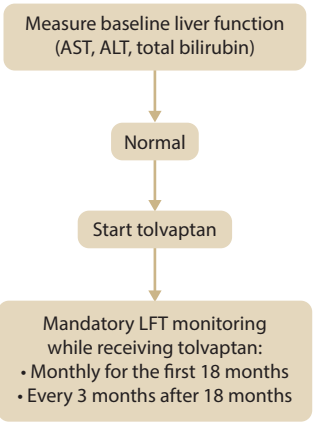


Figure 3



ADPKD, autosomal dominant polycystic kidney disease; CYP3A, cytochrome P450, 3A inhibitors; eGFR, estimated glomerular filtration rate; LFT, liver function tests; mTOR, mammalian target of rapamycin; PROPKD, Predicting Renal Outcomes in Polycystic Kidney Disease; SGLT2i, sodium-glucose cotransporter-2 inhibitors

Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Polycystic liver disease

1 Polycystic liver disease (PLD)

PLD is the most common extrarenal manifestation of ADPKD. It is a hereditary disease characterized by the presence of >10 fluid-filled cysts scattered throughout the liver (Figure 1). PLD most often causes no symptoms and does not impact the synthetic or secretory capacity of the liver. In some cases, symptoms may develop due to mass effects causing abdominal fullness, distension and mechanical back pain, or compression of other organs such as lungs and stomach, or of veins such as the hepatic or portal veins or inferior vena cava, or of bile ducts. Symptoms may also occur with cyst hemorrhages and infections.

2 Abdominal imaging

People with ADPKD should have abdominal imaging, using ultrasound, CT, or MRI, to evaluate both liver and kidney phenotype. When liver cysts are found, patients should be advised of the likely outcomes and possible symptoms.

3 Women with PLD

PLD develops earlier and is more severe in women than in men. Women with PLD, should be counselled to minimize or avoid sex hormone therapy, as appropriate depending on the extent of the liver disease. Observational studies have shown that exposure to estrogen-containing oral contraceptives is associated with a 15.5% greater liver volume for each decade of use. The growth of polycystic livers decreases after menopause, but increases again if estrogen replacement therapy is initiated.

4 Treatment of PLD

Most patients with PLD have no symptoms and require no treatment; however, people with PLD who experience cyst-related symptoms negatively impacting their quality of life or who have severe disease likely to develop symptoms should receive treatment. The choice of treatment in people with symptomatic PLD should be based on symptoms, liver cyst characteristics, total liver volume, and treatment availability. Treatment may involve interventional radiology, surgery, or liver transplantation and should be done at centers of expertise if possible.

5 Somatostatin analogues

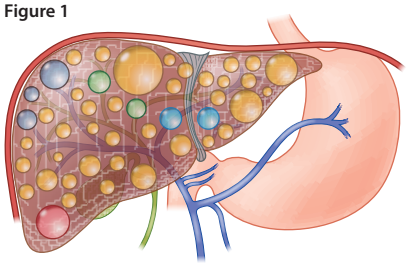
Long-acting somatostatin analogues should be given to people with ADPKD and markedly enlarged polycystic liver with volume-related symptoms to complement other therapies or when other therapies are not available. Long-acting somatostatin analogs are usually well tolerated but some adverse effects are possible (e.g., gallstones, bradycardia). Premenopausal women, who experience a faster liver growth than postmenopausal women, have a better response. Liver volume and disease-specific symptom questionnaires, such as PLD-Q and POLCA, may serve as measures to assess treatment outcomes.

6 Liver cyst infection

Liver cyst infections should be suspected in the presence of a triad of fever, localized abdominal pain and marked elevation of C-reactive protein or leukocytosis, supported by imaging consistent with infection, sometimes requiring 18FDG-PET-CT scanning, and confirmed by diagnostic features in at least 2 categories, such as clinical factors and microbiology. Treatment should be initiated promptly with a 3rd-generation intravenous cephalosporin with or without a fluoroquinolone; cyst drainage is required in severe (i.e., sepsis, immunosuppression) or refractory cases. Antibiotics (intravenous or oral) should be continued for ≥4 weeks (Figures 2 & 3).

ADPKD, autosomal dominant polycystic kidney disease; CT, computed tomography; 18FDG-PET-CT, 18F-fluorodeoxyglucose integrated with positron emission tomography/computed tomography; MRI, magnetic resonance imaging; PLD-Q, Polycystic Liver Disease Questionnaire; POLCA, Polycystic Liver Disease Complaint-specific Assessment

Figure 1



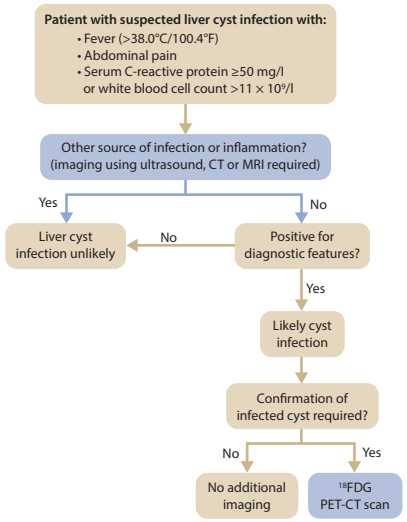
Pressure against diaphragm and lungs
1. Shortness of breath
2. Fatigue

Pressure against the stomach
1. Lack of appetite or early satiety
2. Acid reflux
3. Nausea and vomiting
4. Involuntary weight loss

Cyst complications
1. Intracystic
   a. Recurrent cyst infection
   b. Recurrent cyst hemorrhage
2. Extracystic
   a. Jaundice
   b. Hepatic venous outflow obstruction and portal hypertension

Overall liver size
1. Abdominal fullness
2. Limited mobility
3. Fatigue
4. Anxiety
5. Concern or dissatisfaction with abdomen size
6. Problems with intercourse
7. Mechanical back pain

Figure 2



Diagnostic features
Diagnostic features considered positive in the presence of at least two items from at least 2 categories:

Clinical factors
1. Pain presenting as acute pain or tenderness in liver area
2. History of cyst infection
3. Recent instrumentation of biliary tract
4. Immune compromised patient (including patients on dialysis)

Microbiology
5. Positive blood culture
6. Positive cyst fluid culture

Imaging
7. Imaging showing changes before and after onset of symptoms (ultrasound, CT or MRI)
8. Intracystic gas (ultrasound, CT or MRI)
9. Pericystic inflammation (CT or MRI)
10. Fluid-fluid levels in a cyst (MRI)
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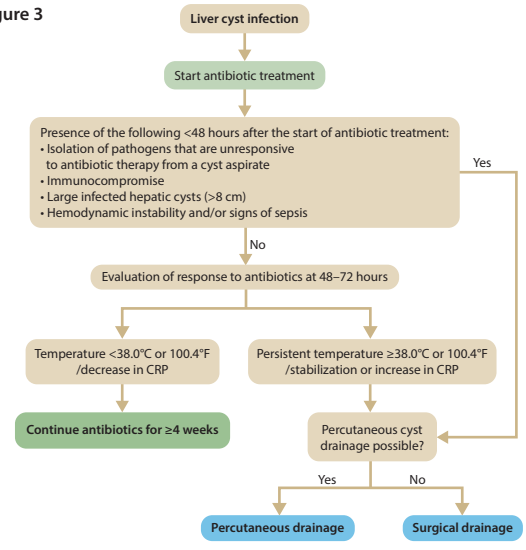
15. ^{111}indium-white blood cell scan showing accumulation in a cyst

Treatment
16. Clinical response to antibiotic treatment

Figure 3



Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Intracranial aneurysms and other extrarenal complications

1 Increased risk awareness

Intracranial aneurysms (ICAs) constitute the most life-threatening association of ADPKD. The prevalence of ICAs and the incidence of ICA ruptures causing subarachnoid hemorrhage (SAH) are approximately 4 and 7 times higher in ADPKD than in the general population (Figure 1). In both populations, the prevalence and incidence are higher when there is a family history of ICA and especially SAH. Adults with ADPKD should be informed of their elevated risk for ICAs and SAH, with education on recognizing symptoms like thunderclap headaches that require immediate medical attention (Figure 2).

2 Comprehensive risk assessment

A detailed personal history of SAH and a family history of ICA, SAH, or unexplained sudden death are crucial in identifying people with ADPKD who are at higher risk for ICA, guiding screening decisions.

3 Screening recommendations

Screening for ICAs is recommended in people with ADPKD and a personal history of SAH, or a positive family history of ICA or SAH, or unexplained sudden death, especially in those eligible for treatment and with a reasonable life expectancy. Consider ICA screening in specific clinical contexts, such as during evaluation for kidney and/or liver transplantation or prior to major elective surgery in people with ADPKD.

4 Advantages and limitations of presymptomatic screening

Presymptomatic screening may allow for intervention if an ICA at risk of rupture is identified, which may prevent death or significant comorbidity. It may also allow adequate imaging follow-up if an ICA with low risk of rupture is identified, and it may reduce anxiety and provide reassurance when no ICA detected. However, screening may also lead to the identification of ICA with very low risk of rupture that do not require intervention but will require long term follow-up. Screening does not exclude the risk of *de novo* ICA development and rupture after screening and may also lead to procedures with possible treatment failure or complications, including death or significant morbidity, and may cause anxiety when an ICA is identified. It may also limit access to life insurance, loans, driver's licenses, or potential work opportunities.

5 Shared decision-making

Shared decision-making is essential in ICA screening. People with ADPKD not considered at increased risk who wish to be screened should be provided access after being adequately informed about the benefits and risks.

6 Imaging for ICA detection

When screening is pursued, time-of-flight (TOF) magnetic resonance angiography (MRA) without gadolinium enhancement is recommended. High-resolution computed tomography angiography (CTA), which requires contrast administration, may be used as an alternative. In high-risk people with ADPKD and negative initial ICA screening results, the timing of rescreening should be individualized, with intervals of 5–10 years, depending on age, risk factors, and life expectancy.

7 Lowering the risk of ICA development and rupture

Modifiable factors that increase the risk of ICA development and rupture include smoking, uncontrolled hypertension, and alcohol in large quantities. In the last decade, the risk of SAH in the general population has decreased worldwide, likely due to less smoking and better blood pressure control. Therefore, smoking cessation and blood pressure control are critical.

8 Other vascular and extrarenal complications

ADPKD is a systemic disease and involves almost every organ and tissue. Physicians taking care of people with ADPKD should be aware of these associations and address them appropriately when they present. Presymptomatic screening for these extrarenal associations is not indicated except in families with non-ischemic idiopathic cardiomyopathies, thoracic aortic aneurysms, or coronary artery dissections.

Figure 1

	General population	General population with family history of ICA or SAH	ADPKD population	ADPKD population with family history of ICA or SAH
Prevalence of ICA (95% CI)	2.9% (1.9–4.5)	3.4 (1.9–5.9) higher risk	12.9% (10.4–15.4)	17.1% 13.4–21.1)
Incidence rates of SAH (per 1000 person-years, 95% CI)	0.079 (0.069–0.09)	3–7 higher risk	0.57 (0.19–1.14)	Likely higher (based on data from general population)

Figure 2

Thunderclap headache

Definition:
- Strikes suddenly
- Intense pain: "worst headache in my life"
- Reaches maximal intensity within 60 seconds

May be associated with or followed by:
- Nausea or vomiting
- Seizures
- Altered mental state/loss of consciousness

What to do:
- Seek immediate medical attention
- Have evaluation in an emergency department equipped with CT scan
- Inform caregivers about the increased risk for subarachnoid hemorrhage associated with ADPKD

Figure 3

Organ	Association	Screening
Heart	Valvular heart disease, non-ischemic cardiomyopathies, congenital heart disease, atrial fibrillation, pericardial effusion	Non-ischemic cardiomyopathies (when there is a family history)
Arteries	Aortic root dilatation, thoracic aortic aneurysms (TAA), coronary artery dissections (CAD) and aneurysms, other aneurysms	TAA or CAD (when there is a family history)
CNS	Arachnoid cyst, dural diverticula	No
Bowel	Colon diverticulosis and diverticulitis, small bowel and duodenal diverticula	No
Liver	Dilatation of common bile duct, congenital hepatic fibrosis, cholangiocarcinoma	No
Pancreas	Cysts	No
Spleen	Cysts	No
Lung	Bronchiectasis, pleural effusion	No
Abdominal wall	Hernias	No
Genital	Seminal vesical cyst, sperm abnormalities	No

ADPKD, autosomal dominant polycystic kidney disease; ICA, intracranial aneurysms; SAH, subarachnoid hemorrhage

Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Lifestyle and psychosocial aspects

1 Individualized approach

ADPKD is a complex genetic disorder, affecting many organs, with variable rates of progression between and within families; therefore an individualized and comprehensive approach to evaluation and management is needed, including attention to nutrition and lifestyle, physical activity and body weight, and socioeconomic wellbeing and mental health (Figure 1).

2 Diet & lifestyle

No specific diet has been proven to delay progression of ADPKD. A generally healthy diet with emphasis on fresh fruits and vegetables, fish, whole grains and nuts, and limitation of salt, sugar, fat and animal proteins is recommended to maximize general and cardiovascular health. Salt should be restricted to 5 g daily. Recreational toxins (tobacco, drugs, or excessive alcohol) should also be avoided.

3 Weight management

Avoidance of overweight and obesity is strongly recommended for general health benefits; there is also preclinical and clinical evidence that overweight and obesity might accelerate kidney growth and ADPKD progression.

4 Water & caffeine

High water intake (2–3 liters/day) is recommended for people with ADPKD and CKD G1-G3 to avoid dehydration, reduce the risk of kidney stones, and possibly slow the progression of the cystic disease. Caffeine intake should not exceed the recommended amount for the general population, which is <400 mg/day (e.g., 4 cups of brewed coffee/day).

5 Exercise

Appropriate exercise is important for maintenance of general health and strength, and it also contributes to psychosocial well-being. The recommended intensity should be individualized according to the patient circumstances, bearing in mind that people with very large kidneys or liver could be vulnerable to direct organ injury from contact sports.

6 Stressors for people with ADPKD

Having a slowly progressive genetic disease presents unique psychosocial stressors for people with ADPKD, which should be elicited and addressed during healthcare encounters (Figure 2). Some patients may require referral to specialized services for management of depression/anxiety, job training, or financial counseling. Patient information and education about self-care should be integrated into all patient care encounters. (Tools for evaluation are available in Appendix 1 of the full guideline at www.kdigo.org)

7 Multidisciplinary team

Implementation of an individualized and comprehensive approach to evaluation and management requires a multidisciplinary team that includes physicians, nurse educators, accredited nutrition providers or registered dietitians, physical therapists, social workers and other caregivers as needed. Potential benefits of this multidisciplinary approach include a reduction in the rate of progression of the disease, better blood pressure control, reduction in cardiovascular events and early mortality, and better general health and quality of life.

8 Comprehensive information about all aspects of the disease

People, caregivers and families affected with ADPKD should be provided comprehensive information about all aspects of the disease and the resources available. These resources should be provided to people, caregivers and families affected by ADPKD (Figure 3) and should include disease information, education about basic management and self-care, prognostic assessment, information on kidney-protective pharmacotherapy including clinical trial opportunities, adequate planning to manage the psychosocial and financial impact of the disease including family planning, and when appropriate discussion of kidney replacement options, research and resources for social support, among others.

ADPKD, autosomal dominant polycystic kidney disease;
CKD, chronic kidney disease

Figure 1

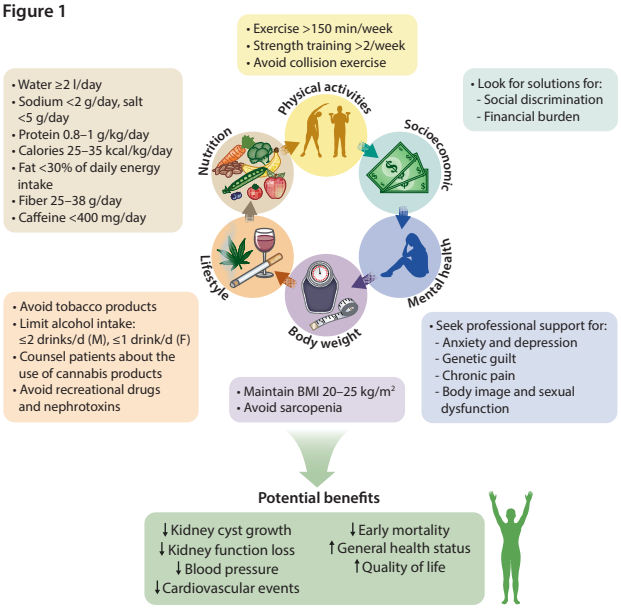


Figure 2

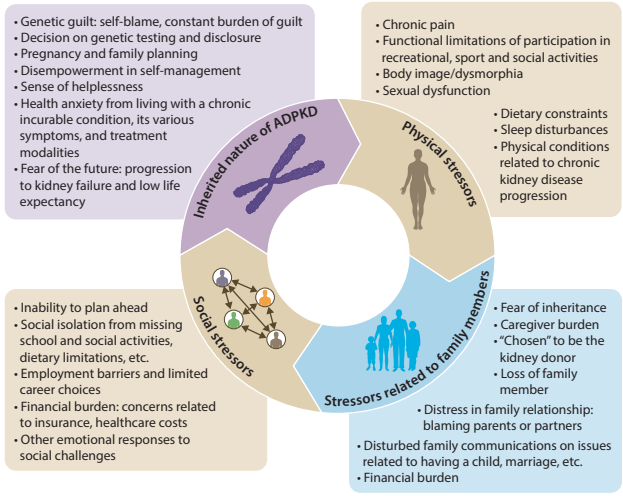


Figure 3

Disease information	• Explanation of the disease and its potential course and manifestations
Basic management and self-care	• Self-management: water intake, low-salt diet, low-protein diet (where appropriate), weight control, lifestyle (e.g., exercise), smoking cessation, caffeine intake, etc. • Cardiovascular risk management: importance, antihypertensive therapy, cholesterol-lowering therapy • Situations for contacting clinic (e.g., pain, complications)
Prognostic assessment	• Rationale, interpretation and implications of prognostic risk score
Specific kidney-protective pharmacotherapy	• Indication, rationale/benefit, adverse effects, monitoring requirements • Clinical trial opportunities
Managing disease impact	• Potential impact of the disease on activity (e.g., work and lifestyle) • Psychological impact and support available • Discussing ADPKD with employers • Issues regarding health insurance and mortgage applications • Family planning, including genetic counselling and preimplantation genetic diagnosis, contraception, and pregnancy issues
Kidney replacement therapy	• Dialysis and transplantation options (according to clinical situation and availability)
Research	• Registry entry, clinical trials, patient-reported outcome data collection
Resources for social support	• Details of financial burden of ADPKD and how to get socio-financial support • Details of ADPKD patient organizations
Hereditary nature of ADPKD	• The most common hereditary kidney disease and its genetic transmission • Importance of kidney imaging in the diagnosis of ADPKD • Possible benefits and harms of genetic screening

Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Pregnancy and reproductive issues

1 Hormone therapy

When considering hormone therapy in women with ADPKD, liver imaging should be performed when feasible to document liver cysts and possible PLD, to inform discussion about options for contraception, hormonal replacement, and other indications (Figure 1).

2 Preconception counseling

Preconception counseling for people with ADPKD addresses a range of topics, including medication adjustments for women, information on reproductive options and potential pregnancy outcomes, and the distinct risks anticipated for both the mother and a child at risk of inheriting the condition. It should be offered to both men and women with ADPKD who are of reproductive age (Figure 1).

3 Reproductive options

Reproductive options without testing of fetus/embryo include accepting 50% chances of offspring with ADPKD, or using egg (in the case of an affected female) or sperm (in the case of an affected male) donation from a donor not affected by ADPKD. Reproductive options with testing of fetus/embryo include preimplantation genetic testing and prenatal testing (Figure 2).

4 Management during pregnancy

During pregnancy, BP, kidney function, soluble fms-like tyrosine kinase-1-to-placental growth factor ratio (sFlt-1/PlGF), and proteinuria should be monitored in women with ADPKD, similar to women with CKD. Low-dose aspirin (75–150 mg daily) should be prescribed from week 12 to week 36 in pregnant women with ADPKD (Figure 1).

5 Blood pressure monitoring

More frequent BP monitoring, preferably weekly home BP monitoring, is advised in all women with ADPKD who become pregnant, most importantly, in those with preexisting hypertension or hypertension diagnosed during their pregnancy.

6 Postpartum review

Women with ADPKD should be seen by a nephrologist within 6 months after delivery for a postpartum kidney review. Women with ADPKD may have bladder instability or urinary incontinence after delivery and should be offered pelvic-floor physical therapy (Figure 3).

Figure 1

Women with ADPKD of childbearing age

Hormone therapy

- Counsel about risk/benefit of estrogen/progesterone therapy in ADPKD women with regard to PLD
- IUDs (including levonorgestrel-releasing IUD) and gestagen OCPs may be preferred for women with PLD

Preconception counseling

- Discontinue potential teratogenic drugs before becoming pregnant (e.g., tolvaptan, RASi)
- Review the risks of preeclampsia, pregnancy induced hypertension, and premature delivery in ADPKD women
- Genetic counseling. Information on risk of inheritance of ADPKD for each pregnancy, nature of fetal/childhood outcomes in affected offspring, and the potential risk/benefit of PGT/PT/egg-sperm donation

Management during pregnancy

- Regular monthly assessment of BP, kidney function, and proteinuria by a health care provider
- Home BP monitoring is encouraged
- Suggested target BP <135/85 mm Hg
- Low dose of aspirin from week 12 to week 36 is recommended for all pregnant ADPKD women
- Monthly screening for UTI is advised. Those with positive urine cultures should be treated adequately
- Encourage increased fluid intake

Management after pregnancy

- Tolvaptan is contraindicated during breastfeeding and should not be prescribed during this time
- Some ACEi such as enalapril or captopril have very low penetration into human milk and can be used with careful monitoring of the infant for signs of hypotension, if other agents are not adequately controlling blood pressure.
- Women with bladder instability or urinary incontinence after pregnancy should be offered pelvic floor physical therapy, especially when tolvaptan will be prescribed

Figure 2

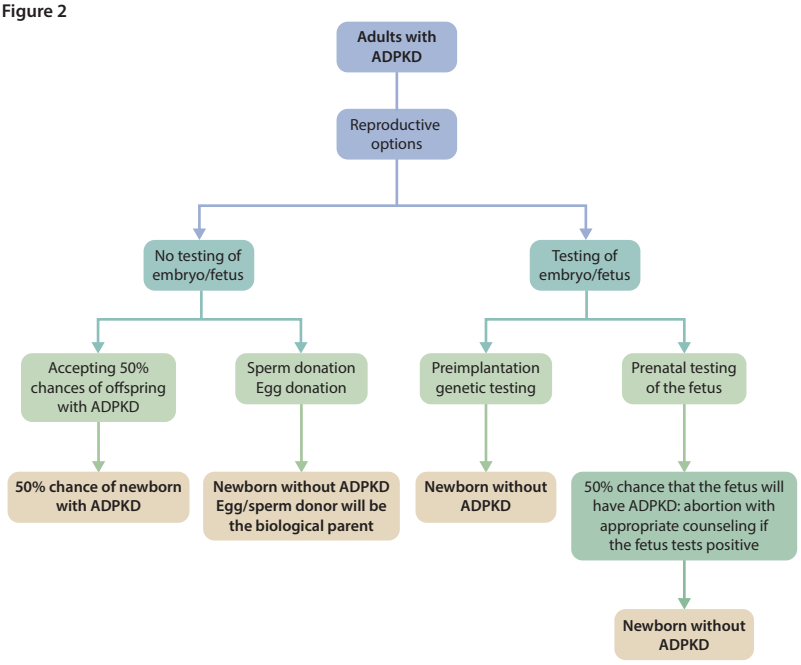
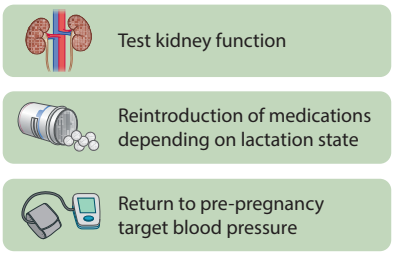


Figure 3



ADPKD, autosomal dominant polycystic kidney disease; BP, blood pressure; PLD, polycystic liver disease

Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Pediatric issues

1 Shared decision-making

Shared decision-making should be undertaken when discussing the benefits and harms related to screening/diagnosis of at-risk children in families with ADPKD, including the parents/legal guardians and the mature child (Figure 1).

2 Blood pressure control

Standardized office BP should be assessed annually in children (≥5 years) and adolescents with or at risk for ADPKD. Annual 24-hour ABPM should be performed in children and adolescents (≥5 years and height ≥120 cm) with VEO-ADPKD or EO-ADPKD and in children and adolescents with or at risk for ADPKD with BP ≥75th percentile.

3 Blood pressure target

We recommend targeting BP to ≤50th percentile for age, sex, and height or ≤110/70 mm Hg in adolescents with ADPKD. RASi (i.e., ACEi or ARBs) is the first-line therapy for high BP in children and adolescents with ADPKD.

4 Diet and exercise

Children with ADPKD should follow general recommendations for a healthy diet, consistent with WHO guidelines, and should maintain a healthy body weight and physical activity.

5 Treatment of ADPKD in children

There is currently insufficient evidence to support use of targeted or disease-modifying therapies for ADPKD in children beyond antihypertensive treatment.

6 Follow-up

The follow-up of children with diagnosed ADPKD should be performed by a pediatrician or pediatric nephrologist, tailored based on clinical indications, such as BP, kidney function, urine studies, and ultrasound (Figure 2).

7 Transition from pediatric to adult patient

As children enter young adulthood, a formal transition process should be developed for all children diagnosed with or at risk for ADPKD. Assessment for extrarenal manifestations should be recommended, as indicated for adults with ADPKD.

Figure 1

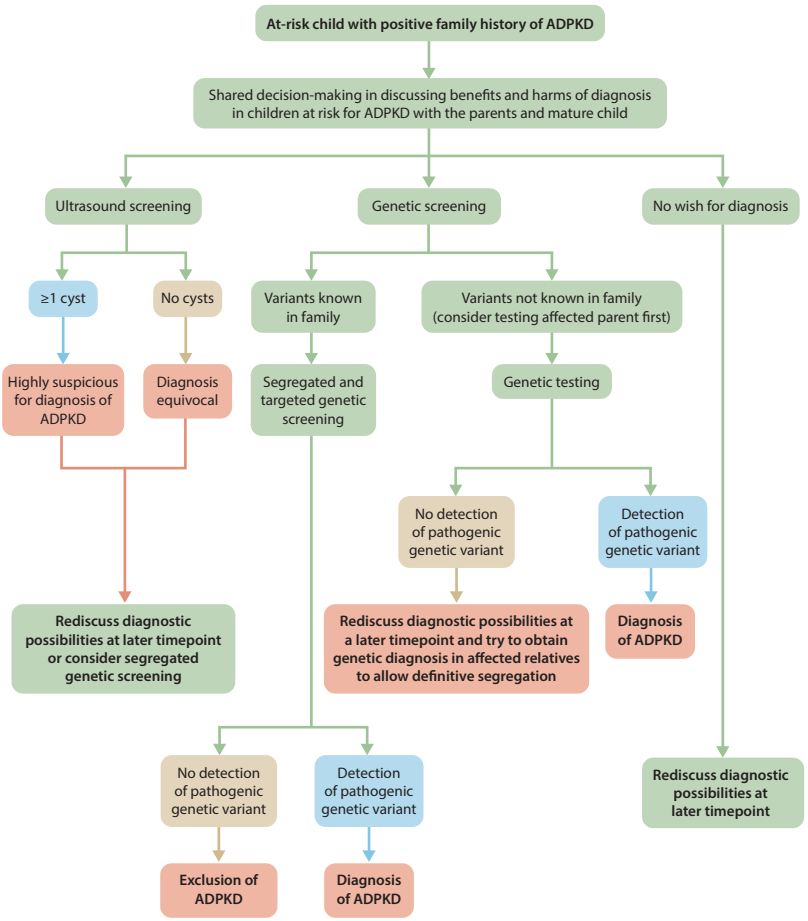
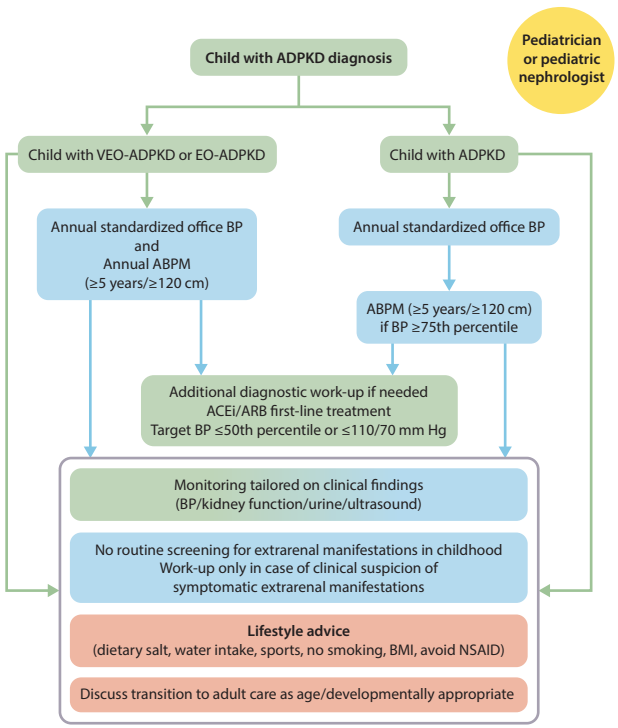


Figure 2



ACEi, angiotensin-converting enzyme inhibitor; ADPKD, autosomal dominant polycystic kidney disease; ARB, angiotensin II receptor blocker; BP, blood pressure; EO, early onset; RASi, renin-angiotensin system inhibitors; VEO, very early onset; WHO, World Health Organization

Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Approaches to the management of people with ADPKD

1 Shared decision-making

Shared decision-making should be the cornerstone of patient-centered management in people with ADPKD (Figure 1). By employing shared decision-making when working with people with ADPKD you allow them to play an active role in their own care.

2 Care pathway

The required lifelong management of people with ADPKD should follow a comprehensive, multidisciplinary, and holistic health care pathway. Healthcare systems should provide care coordination or patient navigation for people with ADPKD to ensure holistic health care during their disease journey.

3 Self-management program

Healthcare systems should implement a structured self-management program for people with ADPKD that includes a system of processes to address medical, behavioral, and emotional management (Figure 2).

4 Patient support

ADPKD-focused patient organizations, national kidney federations, and patient support groups can help enhance the care of people with ADPKD and their families. Healthcare systems should promote the participation of people with ADPKD in registries that gather outcome data using standardized data definitions to improve the future care of ADPKD individuals.

ADPKD, autosomal dominant polycystic kidney disease

Figure 1

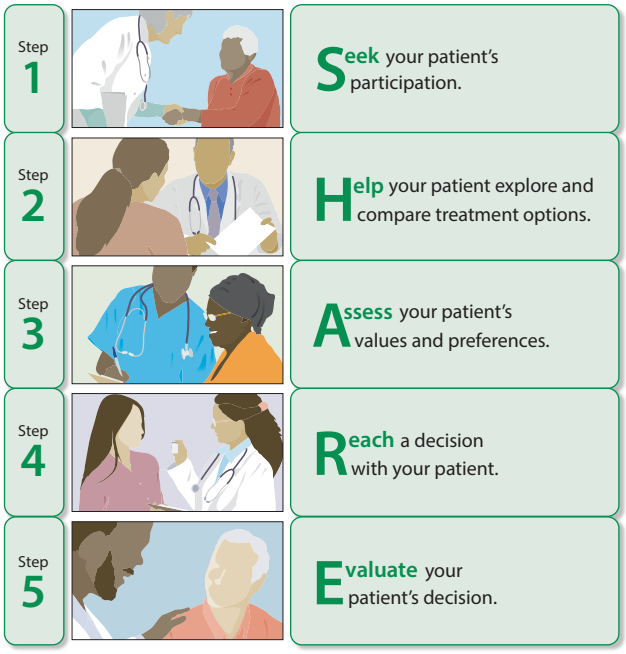


Figure 2