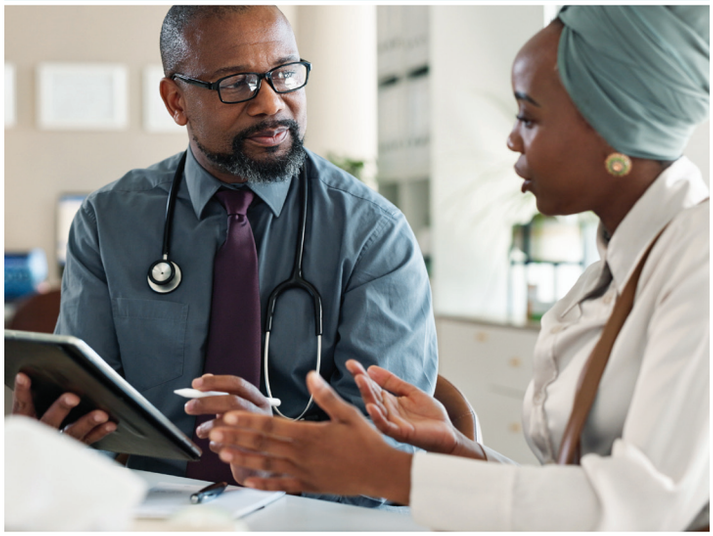
Understanding the KDIGO 2025 ADPKD Clinical Guideline

A Plain Language Guide



PKD FOUNDATION
Polycystic kidney disease

PKD FOUNDATION
Polycystic kidney disease

pkdcure.org

1.800.PKD.CURE

This guide was created by the PKD Foundation as a plain language summary of the **KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of Autosomal Dominant Polycystic Kidney Disease (ADPKD).** It is intended to assist anyone impacted by ADPKD, including patients, care partners, and families with understanding the basic recommendations provided in the guideline and support shared decision-making conversations with healthcare providers.

Published December 2025

Acknowledgements

PKD Foundation Staff Writers and Reviewers:

- **Paul Scribner, MSW,** Vice President of Community Programs and Education
- **Chris Chen, Ph.D.,** Vice President of Research
- **Nicole Harr,** Senior Director of Community Programs and Education
- **Caitlin Lasky,** Director of Marketing and Communications
- **Sydney Johnston,** Marketing Strategist
- **Nicole Torres, R.N.,** Community Education Strategist

External Reviewers:

- **Dwight Odland,** Los Angeles Community PKD Connect Ambassador, PKD Foundation Board member, Contributor to KDIGO 2025 Guideline for ADPKD
- **Ronald D. Perrone, M.D.,** Co-Director, Polycystic Kidney Disease Outcomes Consortium; Nephrologist, Division of Nephrology, Tufts Medical Center; Contributor to KDIGO 2025 Guideline for ADPKD
- **Amy Earley,** Guideline Development Director, KDIGO (Kidney Disease: Improving Global Outcomes)
- **Danielle Green,** Chief Executive Officer, KDIGO (Kidney Disease: Improving Global Outcomes)
- **Michael Cheung,** Chief Scientific Officer, KDIGO (Kidney Disease: Improving Global Outcomes)

How to Use This Guide

This guide is here to help you understand **autosomal dominant polycystic kidney disease (ADPKD)** and feel more prepared to manage your health. It explains the most important points from the **KDIGO 2025 Clinical Practice Guideline for ADPKD** in easy-to-understand language.

You don't need to read it all at once. Start with the parts that matter most to you right now, like learning about your diagnosis, managing symptoms, or planning for future treatment.

Use the **key recommendations** and **questions to consider asking your doctor**, provided in each section, to get ready for your doctor visits. You can even bring this guide with you to appointments and ask your care team to go over it with you.

This guide is intended for:

- People living with ADPKD
- Care partners and family members
- Friends of people impacted by ADPKD

This guide can help you:

- Understand what to expect with ADPKD
- Ask informed questions at medical visits
- Make decisions together with your doctor
- Feel more confident about your care

Everyone's journey with ADPKD is different. Use this guide to learn, stay organized, and speak up for the care that fits your needs.

If you want to do a deeper dive into the KDIGO Guideline you can always view it at https://kdigo.org/guidelines/autosomal-dominant-polycystic-kidney-disease-adpkd/

Table of Contents

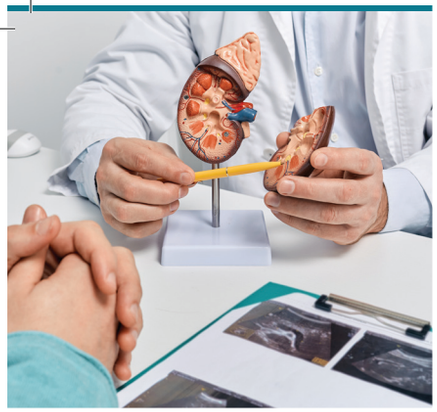
What is the KDIGO Guideline for ADPKD and Why Does It Matter?

Officially titled, *KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of Autosomal Dominant Polycystic Kidney Disease (ADPKD)*, the guideline is a trusted resource developed to help provide clear, evidence-based practice guidance for the care and treatment of those impacted by ADPKD.

These guidelines were created by KDIGO (Kidney Disease: Improving Global Outcomes), an international organization of doctors, researchers, and patient advocates. To develop the guideline, they reviewed the best available science, medical practices, and lived experiences of those impacted by ADPKD to provide the most up-to-date recommendations for managing the condition at every stage.

Whether you are newly diagnosed, have been living with ADPKD for many years, have received a kidney transplant, or are a care partner of someone affected by ADPKD, this guide is here to help you:

1. Understand the essential recommendations provided in each guideline chapter.
2. Have more informed conversations with your healthcare team about each recommendation so you can best manage ADPKD for yourself or a loved one.



Chapter 1:

Understanding ADPKD – What It Is, How It's Diagnosed, and What to Expect

What This Chapter Covers

This chapter explains what ADPKD is, how it's diagnosed, how common it is, and what we know about how it usually progresses. It also describes the naming and classification system doctors use to predict how quickly a person's ADPKD may progress.

Autosomal dominant polycystic kidney disease (ADPKD) is a lifelong condition where fluid-filled cysts grow and multiply in the kidneys, causing them to enlarge and lose function over time, which can result in kidney failure. The disease can also affect other parts of the body, including the liver, pancreas, heart, and brain. It can also lead to high blood pressure and other complications.

ADPKD is a genetic disorder that is estimated to affect millions of people worldwide, regardless of age, race, or ethnicity. It's usually passed from parent to child. If one parent has the condition, each child has a 50% chance of inheriting it. ADPKD is usually caused by a mutation in one of two genes; however, mutations in other genes have also been shown to cause PKD.

The two primary genes mutated in ADPKD are:

- ***PKD1***, which often causes more rapid disease progression
- ***PKD2***, which usually progresses more slowly

In a small number of people, PKD may be found accidentally when kidney imaging is done for a separate health concern. If this person has no obvious family history of PKD, it's recommended that testing be done on parents to confirm there is no family history. If no family history is confirmed, this type of PKD is termed "de novo," meaning it is a newly formed gene variant of PKD. Confirmation of diagnosis can be done through genetic testing, and further imaging is commonly performed to monitor cyst growth (Figure 1).

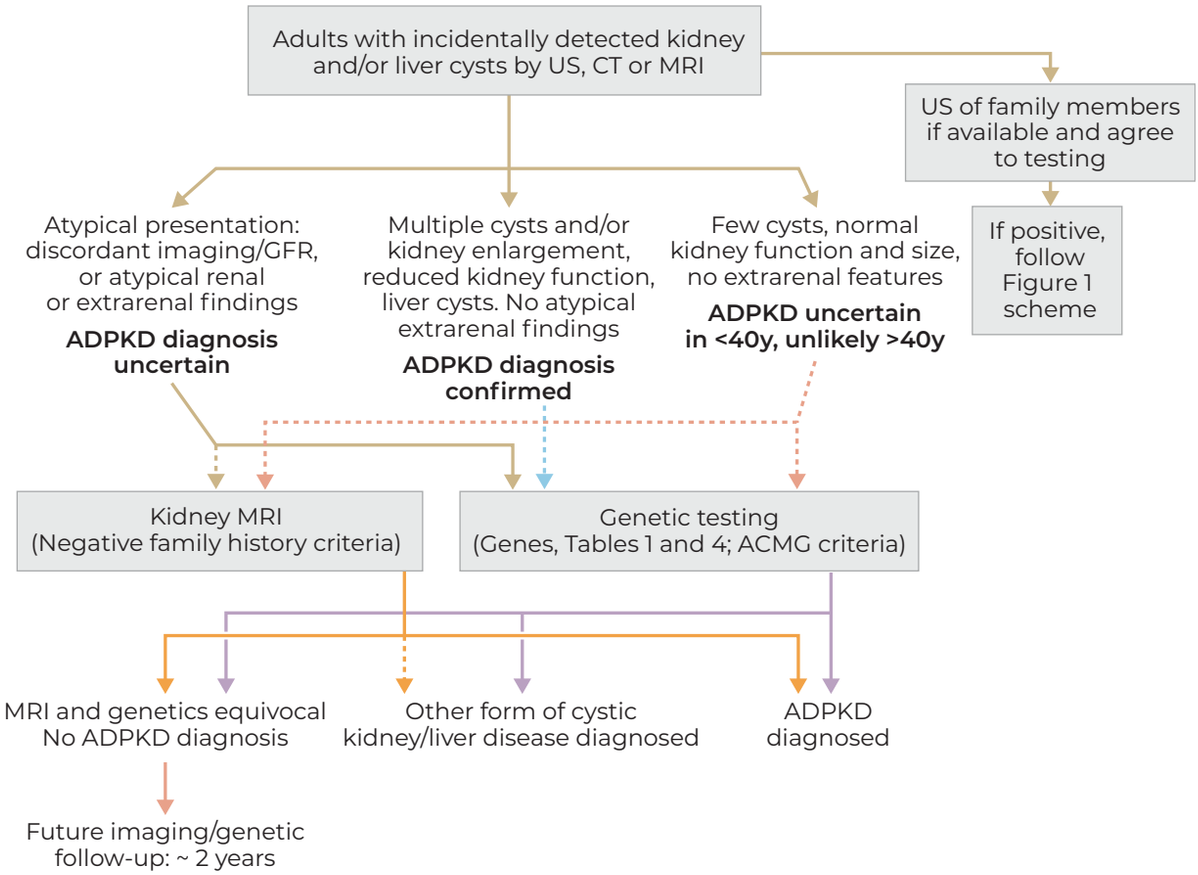


Figure 1 Steps used to diagnose adults who are found to have kidney or liver cysts by chance and don't have a known family history of autosomal dominant polycystic kidney disease (ADPKD). Refer to the *KDIGO 2025 ADPKD Guideline* for "Figure 1 scheme" and "Tables 1 and 4" noted in this diagram. This figure is included in the *KDIGO 2025 ADPKD Guideline* and is being used in this document with permission.

US= Ultrasound, CT= Computed tomography, MRI= Magnetic resonance imaging

Doctors use imaging (like ultrasounds, CT scans, or MRIs) to check for cysts and measure kidney size and growth. For individuals ages 16-40, and a positive family history of ADPKD with gene variants *PKD1* or *PKD2*, these imaging tests can be used to confirm diagnosis and estimate how fast the disease may progress.

Genetic testing is recommended if:

- An individual presents with kidney cysts and has no known family history of ADPKD
- Imaging results are unclear
- Someone is considering having children or donating a kidney

Symptoms often start between **ages 30 and 50**, but the disease can present much earlier, especially if there is a history of early onset end stage renal disease (ESRD).

ADPKD affects about 1 in every 1,000 people. But not everyone's journey is the same. These tools help personalize your care.

Key Recommendations for People with ADPKD and Caregivers

Diagnosis and Imaging Basics

- **Ultrasound is often the first step in diagnosing ADPKD.** It's a simple and safe way to look for cysts in your kidneys.
- **CT scans or MRIs may be used to get a more detailed picture of kidney size.** This is especially helpful for determining risk of disease progression.
- **Genetic testing can confirm a diagnosis in many cases.** It's especially useful when imaging is unclear or for family planning.

Understanding the Names and Labels

- **ADPKD is the most accurate name for the condition.** It describes a dominant genetic form of PKD that usually starts in adulthood.
- **Doctors may also describe your disease by its genetic form (*PKD1* or *PKD2*).** This can help predict how quickly your condition may change. For example, ADPKD-*PKD1*.

Understanding Progression

- **Some people progress quickly, while others progress slowly.** Your doctor may use tools like the Mayo Imaging Classification (MIC), which looks at your age and the size of your kidneys, to help predict your path.
- **Many factors are associated with the severity of ADPKD.** The type of gene variant, sex (male or female), and environmental factors (obesity, salt intake) all play a role.
- **Knowing your risk level can help guide treatment.** It helps to determine how often you need monitoring and whether you might benefit from certain medications.

How Common Is It?

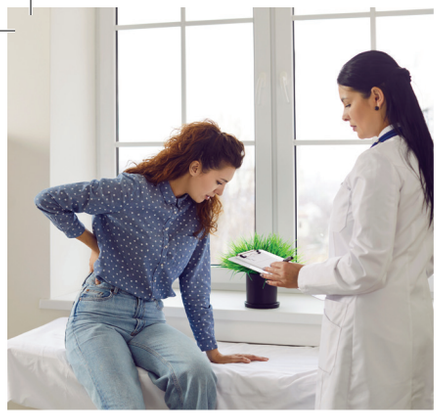
- **ADPKD is the most common inherited kidney condition.** It affects people of all backgrounds, with symptoms often starting between ages 30 and 50. As many as 10 in 100 people with kidney failure worldwide have ADPKD.

It is recommended that all patients with ADPKD see a nephrologist (kidney doctor) with expertise in treating PKD.

Talking with Your Healthcare Team

Here are some important questions to consider asking your doctor:

- Can you explain what my imaging results show about my kidney size and cyst growth?
- Would genetic testing give us useful information for confirming diagnosis or for family planning?
- Do I have *PKD1* or *PKD2* (or another genetic variant), and what might this mean for me?
- What's my risk for disease progression, and how will we keep track of it over time?



Chapter 2:

Managing Kidney Symptoms and Complications

What This Chapter Covers

This chapter explains the most common **kidney-related symptoms** people with ADPKD experience, like:

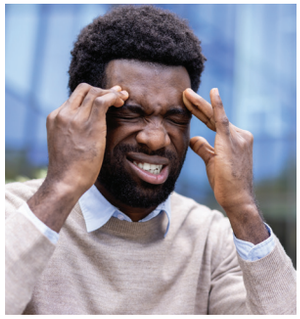
- High blood pressure
- Kidney pain
- Blood in the urine
- Kidney cyst infections and urinary tract infections (UTIs)
- Kidney stones
- Gout

It also covers how to monitor these symptoms and what can be done to manage them effectively. While many of these symptoms are common, **the good news is that they're treatable**—especially when recognized early.

Knowing what's normal for your body and when to contact your care team can help you stay ahead of problems and feel more confident managing your ADPKD.

Key Recommendations for People with ADPKD and Caregivers

Know Blood Pressure (hypertension) Basics



High blood pressure is one of the earliest and most common symptoms of ADPKD. It can make kidney disease progress faster if left untreated. High blood pressure often has no symptoms until it becomes more severe. Having no symptoms, however, doesn't mean it should be ignored.

Some common symptoms of more severe high blood pressure may include:

- Headache
- Dizziness
- Blurry vision
- Chest pain and/or irregular heartbeat
- Nosebleed

Maintaining a target blood pressure is important and reduces the risk of kidney damage and heart complications. Routine blood pressure monitoring (at home and in your doctor's office), along with dietary and lifestyle changes, pain management, and use of medication, can help manage blood pressure (Figure 2).

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 (protein in the urine), 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 Blood pressure management recommendations for autosomal dominant polycystic kidney disease (ADPKD). Below is a list of abbreviations used in this table and what they mean. This figure is included in the KDIGO 2025 ADPKD Guideline and is being used in this document with permission.

BP= Blood pressure, HBPM= Home blood pressure monitoring, ABPM= Ambulatory blood pressure monitoring, LVH=Left ventricular hypertrophy, RAS=Renin-angiotensin system, ACEi=Angiotensin-converting enzyme inhibitor, ARB=Angiotensin receptor blocker

Target blood pressure recommendations are based on your current blood pressure, age, and level of chronic kidney disease (CKD).

- For those ages 18-49 with CKD levels 1-2 and blood pressure over 130/85 mm Hg, it's recommended to keep your blood pressure ≤ 110/75 mm Hg.
- For those over age 50 with any stage of CKD, it's suggested to keep your top blood pressure number (systolic blood pressure) ≤ 120 mm Hg.

IMPORTANT NOTE: It's always best to talk with your doctor to determine your ideal target blood pressure and ways to manage it based on your individual needs.

Manage Your Kidney Pain

- **Kidney pain is a common symptom in ADPKD but it's treatable.** It can be felt in the back, flank (sides), or abdomen (stomach) and may be caused by large cysts, a burst cyst, infection, or kidney stones. Abdominal pain may also be due to liver cysts.
- **Track your pain.** Pay attention to where it is, how strong it feels, and what triggers it. Keeping a pain diary can help your doctor find the cause and determine appropriate treatment.
- **Be involved in your pain management plan.** There are many options available to manage pain (Figure 3). The general recommendation is to start with noninvasive therapies like heat and ice application, meditation, acupuncture, and acetaminophen (Tylenol) before using more invasive or risky treatments like opioids and surgery.
    - We understand that the figure below may seem overwhelming with so many options to choose from. We recommend discussing these treatment options with your doctor to determine what's right for you, as approaches may be different for each person.
- **Avoid overusing medications like ibuprofen (Advil, Motrin).** These non-steroidal anti-inflammatory drugs (NSAIDs) may harm your kidneys over time. Acetaminophen (Tylenol) is generally safer but talk with your doctor first.
- **If pain doesn't improve, ask for help.** You may need a new pain management strategy, a referral to a pain specialist, or more advanced imaging to understand what is going on.

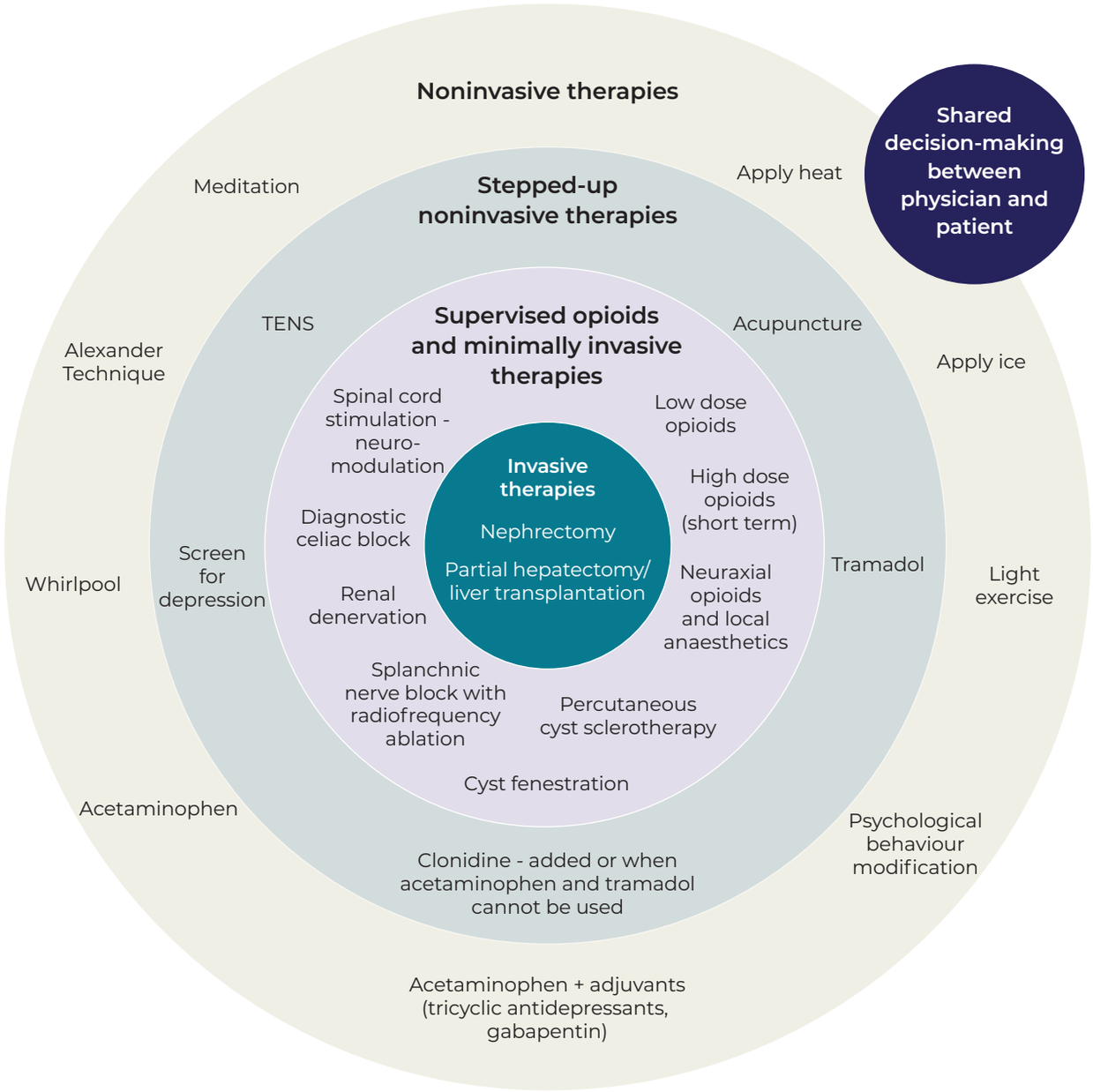


Figure 3 It's recommended to start with the most widely used noninvasive therapies for managing chronic kidney pain. If needed, progress to stepped-up noninvasive therapies, then to supervised opioids and minimally invasive therapies. If these don't help, invasive therapies may be considered. This step-by-step approach supports shared decision-making to find the safest, most effective care. This figure is included in the *KDIGO 2025 ADPKD Guideline* and is being used in this document with permission.

TENS= Transcutaneous electrical nerve stimulation

What if You See Blood in Your Urine (Hematuria)?

- **Blood that's visible in the urine is a common and expected symptom of ADPKD.** Your urine might look pink, red, or brownish. It may or may not be associated with pain. If no other symptoms are present, it can often resolve within one to two days.
- **Blood in the urine may also be caused by physical trauma, cyst rupture, cyst bleeding, or kidney stones.** People with larger kidneys, high blood pressure, and advanced kidney disease are more likely to experience this.
- **If you're unsure, contact your doctor to determine a course of treatment.** Bedrest, hydration (drinking lots of water), and pain management may be recommended depending on the cause.

Always call your doctor if:

- The blood in your urine lasts more than a few days
- You're in pain or passing blood clots
- You have a fever (this could mean infection)

Manage Your Kidney Cyst Infections and Urinary Tract Infections (UTIs)

- **People with ADPKD should receive an urgent medical evaluation for kidney cyst infection if they have the following signs and symptoms and no other possible cause of infection:**
  - Fever
  - Abdominal, back, or side pain
  - Chills, nausea, and feeling very unwell
  - Blood tests showing higher than normal white blood cells or signs of inflammation
- **Treating a kidney cyst infection usually involves taking antibiotics for four to six weeks.** Specific antibiotics are given for kidney cyst infections that can get into the cyst and fully clear the infection. For kidney cysts, this can take longer than it does for treating other infections in the body because it can be harder for the medicine to reach the inside of a cyst.
- **Don't ignore UTIs.** Treating urinary tract infections quickly may help prevent them from spreading to the kidneys. They can typically be treated with a shorter course of antibiotics compared to treating a kidney cyst infection.

Understand Kidney Stone Basics

- **ADPKD increases your risk of developing kidney stones.** These can cause sudden sharp pain, nausea, or blood in the urine.
- **Drinking enough fluids can help prevent stones.** Talk with your doctor about how much you should drink based on your kidney function.
- **You may need imaging (like an ultrasound or CT scan)** if you have symptoms of a stone.

Understand and Manage Gout

- **ADPKD does not increase your risk of developing gout.** Gout is more common in people with chronic kidney disease, so those with ADPKD should be aware of the symptoms and treatment recommendations available.
- **Factors that increase the risk for developing gout include being male, older in age, overweight, and having decreased kidney function.** Talk with your doctor if you experience joint pain, swelling, or redness.
- **Treatment options are similar to those for individuals without CKD, but the stage of your kidney disease may influence the course of treatment.** Your doctor may suggest changes to your diet and weight, along with medications based on your stage of CKD and individual health needs.

Talking with Your Healthcare Team

Here are some important questions to consider asking your doctor:

- What is my current blood pressure and what is my target blood pressure?
- What symptoms should I watch for and report right away?
- What can I do to prevent kidney infections, UTIs, and kidney stones?
- I'm having pain—can we talk about where it's coming from and what might help?
- What should I do if I see blood in my urine again?

"I always urge my patients to tell me about how symptoms affect their daily life—physically and emotionally. This includes things like ongoing pain, infections, fear of flare-ups, and management of treatments. I especially want to know if they're struggling, feeling overwhelmed or having symptoms of anxiety or depression. There are great support groups, therapists, and patient mentors who can help. I can't help if I don't know what's going on."

Chapter 3:

Managing Chronic Kidney Disease (CKD) in ADPKD – Progression, Kidney Failure, and Kidney Replacement Therapy

What This Chapter Covers

As ADPKD progresses, it may lead to **chronic kidney disease (CKD)**, a gradual loss of kidney function over time (Figure 4). Some people may eventually reach **kidney failure**, also known as **end-stage renal disease (ESRD).** This chapter helps you understand:

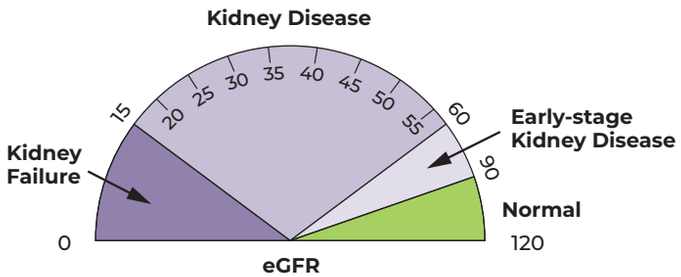
- How kidney function is measured
- What to expect as your kidney function changes
- When to plan for **kidney replacement therapy** (like **transplant** or **dialysis**)
- How to prepare emotionally and practically for what may lie ahead

Not everyone with ADPKD will need dialysis or a transplant—but if you do, starting the conversation early gives you more choices and support.

Key Recommendations for People with ADPKD and Caregivers

Track Your Kidney Function

- **Your kidney function is measured with a test called eGFR (estimated glomerular filtration rate).** This number shows how well your kidneys are filtering waste from your blood.



- **As your eGFR gets lower, your care plan may need to change.** Ask your doctor to explain what your current eGFR means and how it's trending over time.
- **Imaging tests can also help predict how your ADPKD will progress.** MRI and ultrasound tests show how large your kidneys are and help your healthcare team know how fast your cysts are growing.

Other recommendations provided in the KDIGO 2025 ADPKD Guideline for managing chronic kidney disease (CKD) in ADPKD are similar to those for other forms of chronic kidney disease and include:

- Controlling blood pressure
- Following a healthy diet, with attention to protein and salt intake
- Staying active
- Managing diabetes effectively
- Treating anemia (a low red blood cell count) appropriately
- Lowering cholesterol to prevent heart disease

Prepare Early for Kidney Failure (If It Happens)

- **Not everyone with ADPKD reaches kidney failure.** But if your eGFR falls below a certain level, your kidney function can be partially replaced through dialysis or a kidney transplant.
- **Start planning early—before your kidney function gets too low.** Typically, this should be considered when the eGFR falls to 25-30%. This helps avoid emergencies and gives you time to explore options.
- **Options include:**
  - **Kidney transplant –** This is often the preferred treatment for kidney failure and is ideally done before starting dialysis.
  - **Dialysis –** A process that removes fluid and waste from the body with the use of a machine when the kidney function is severely reduced. There are two options, so speak with your nephrologist about what is right for you. Options include:
    - **Hemodialysis (HD) –** Usually done at a clinic, several times a week
    - **Peritoneal dialysis (PD) –** Usually done at home, this treatment uses the lining of your belly, called the peritoneum, as a natural filter to clean your blood

When to See a Kidney Specialist about Transplant or Dialysis

- **Ask for a referral to a specialist when your eGFR drops below 30 ml/min/1.73m².** Earlier referrals mean more time to prepare and fewer complications.
- **You may also want to meet with a transplant team and social worker.** They can help with financial planning, emotional support, and choosing a dialysis option, if needed.

Other Things to Know About Kidney Transplant and Living Donation

- **The guideline suggests that the preferred treatment for kidney failure in people with ADPKD is getting a kidney transplant from a living donor—ideally before starting dialysis.** It can offer a better quality of life, longer-lasting transplanted kidney compared to one from a deceased donor, fewer dietary restrictions, and fewer complications than dialysis.
- **A healthy family member or friend may be able to donate a kidney.** Living donation can shorten your wait time and improve outcomes. If possible, begin discussions about donation at least 12 months before you think you'll need dialysis. This will help you find a potential living donor and prepare for transplant.
- **Some complications following a kidney transplant are more common for people with ADPKD.** Before you have a transplant, make sure you talk with your doctor about potential complications. This will help you know what signs and symptoms to look for after transplant and can help you even prevent certain complications.
- **You don't need to decide everything right away.** Learning about your options early helps you feel more in control if the time comes.

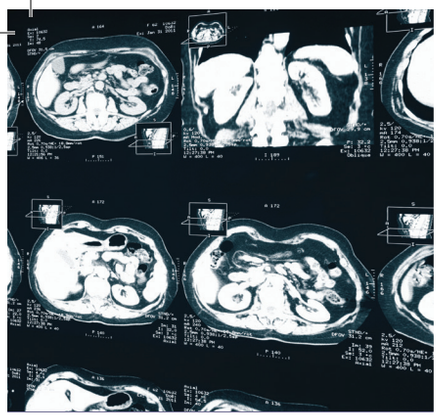
Will Your Existing (Native) Kidneys be Removed?

- Native kidneys in people with ADPKD are sometimes removed. **Native nephrectomy is the surgical removal of one or both existing cystic kidney(s). Ideally, it's done after beginning dialysis or after transplantation.** Talk with your nephrologist and transplant team about your specific situation and what's best for you.
- **Specific reasons for native nephrectomy can include:**
  - Recurrent and/or severe kidney infection
  - Recurrent and/or severe kidney cyst bleeding
  - Kidney stones that cause symptoms
  - Kidney pain
  - Suspicion of kidney cancer
  - Not enough space for a kidney transplant
  - Other health problems caused by extremely large kidneys

Talking with Your Healthcare Team

Here are some important questions to consider asking your doctor:

- What is my current eGFR and how has it changed over time?
- How will my kidney function change over time based on the size of my kidney(s) and the number of cysts I currently have?
- Am I at risk of needing a transplant or dialysis in the future?
- When should I start planning for kidney replacement therapy?
- Can you refer me to a nephrologist or transplant center now?
- What are my options for receiving a living kidney donation and how does the process work?



Chapter 4:
Slowing Down the Progression of ADPKD

What This Chapter Covers

This chapter is about how to **slow the growth of kidney cysts** and delay the loss of kidney function in ADPKD. It focuses on three key strategies (Figure 4):

- Managing high blood pressure
- Considering medications like **tolvaptan**, which can slow cyst growth in some people
- Maintaining healthy lifestyle practices

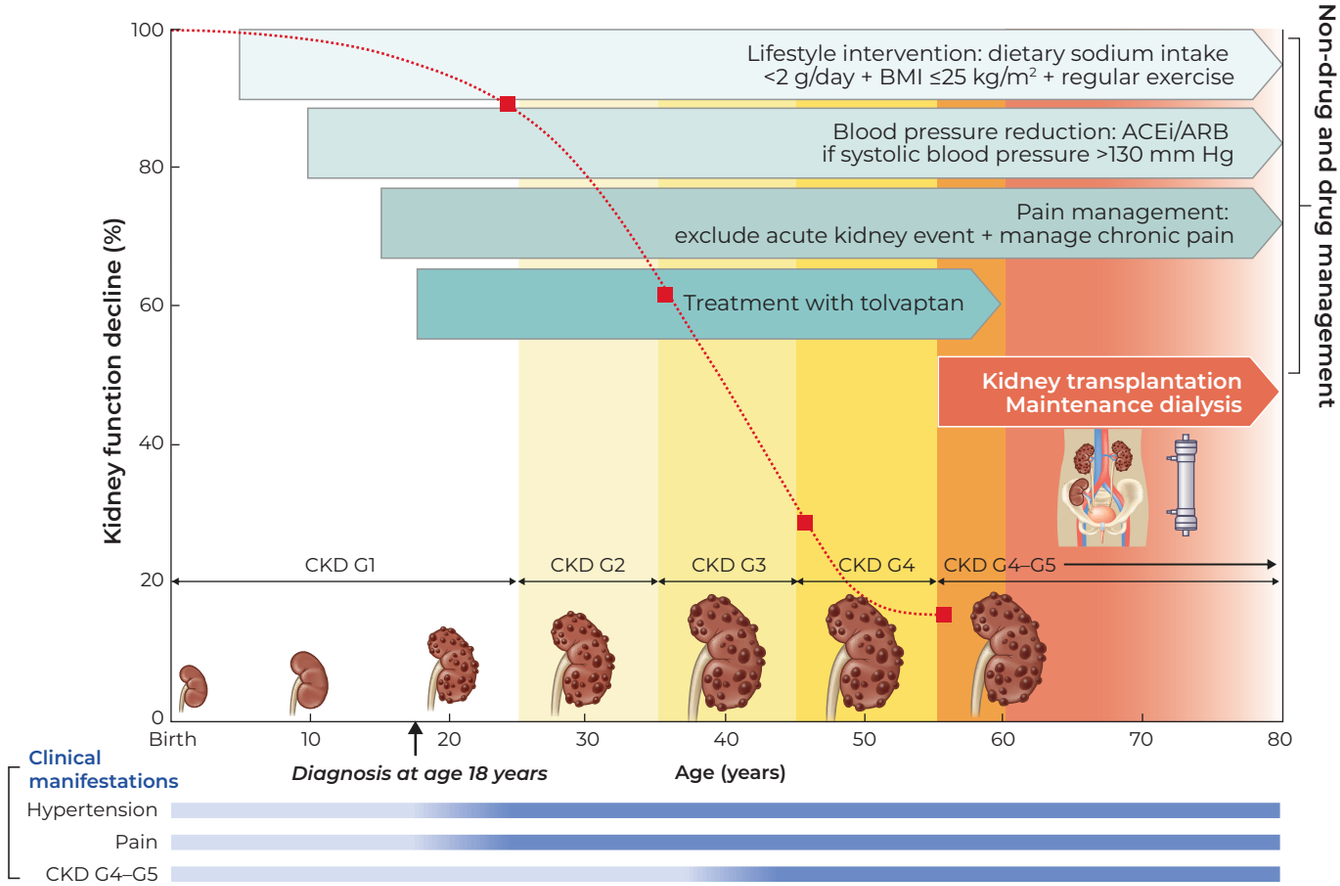


Figure 4 This is a timeline of typical ADPKD presentation with age on the horizontal line, going from left to right. White/yellow/red color shades indicate progressing CKD stage. The red curve shows the decline in kidney function as a patient ages. Blue bars at the bottom show progression of clinical symptoms over time. Green and red arrow bars show timing of recommended clinical interventions. This figure is included in the *KDIGO 2025 ADPKD Guideline* and is being used in this document with permission.

Key Recommendations for People with ADPKD and Caregivers

Manage Blood Pressure

- **High blood pressure is common in ADPKD and can speed up kidney damage.** Keeping it under control is one of the most important steps in slowing disease progression.
- **The best medications to start with are ACE inhibitors or ARBs.** These drugs help lower blood pressure and protect the kidneys.
- **Talk to your doctor to determine your ideal target blood pressure and ways to manage it based on your individual needs (Figure 2).**

Consider if Tolvaptan is Right for You

- **Tolvaptan is a medication that can slow cyst and kidney growth and kidney function decline.** It's most helpful for people with fast-progressing ADPKD and with an eGFR greater than or equal to 25 ml/min/1.73 m^2.
- **Your doctor may use imaging, age, kidney function, and family history to see if tolvaptan is a good fit for you.** Tolvaptan may not be an option for some individuals with ADPKD.
- **You'll need regular blood tests to check your liver while taking tolvaptan.** It also makes you urinate more often, so it's important to stay well hydrated.

Other Health Practices to Maintain Kidney Function

Evidence from multiple research studies suggests that following these health and dietary practices can help delay kidney disease progression and extend the lives of people with ADPKD.

- **Maintain a healthy weight.** This helps improve overall kidney and cardiovascular health. Obesity may increase cyst growth and cause kidneys to get worse more quickly.
- **Follow a low salt diet.** This can help control blood pressure, reduce fluid retention, and have a positive impact on both kidney health and cyst progression in ADPKD.
- **Stay well hydrated by drinking enough water every day.** Start with at least eight cups. The exact amount you should drink can vary and depend on many factors, including your current kidney function. For example, someone with severely reduced eGFR and on dialysis should not drink lots of water. **Always consult with your healthcare provider for specific recommendations as there are situations where drinking a lot of water may not be recommended for people with ADPKD.**

Talking with Your Healthcare Team

Here are some important questions to consider asking your doctor:

- Is my blood pressure in the right range for ADPKD?
- Would tolvaptan help slow my disease and what are the side effects?
- How do we decide if I'm a good candidate for tolvaptan?
- Are there other dietary or health practices that I should be considering?

Chapter 5:

Understanding Liver Cysts and Polycystic Liver Disease (PLD)

What This Chapter Covers

Polycystic liver disease (PLD) is the most common **extrarenal manifestation** (non-kidney medical issue) in people with ADPKD and occurs at earlier ages with more aggressive cyst growth in women. Most people won't have major liver problems, but some may develop symptoms when the liver becomes very enlarged and uncomfortable (Figure 5).

Most people with ADPKD have liver cysts that increase with age. PLD is characterized by the presence of 10 or more cysts seen in the liver.

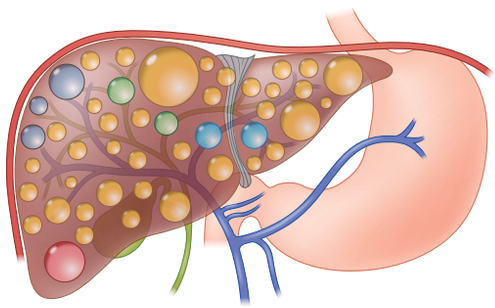
This chapter explains what to watch for, how to manage symptoms, and when to consider treatments.

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



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

Cyst complications

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

Figure 5 Symptoms and complications of polycystic liver disease (PLD). This figure is included in the *KDIGO 2025 ADPKD Guideline* and is being used in this document with permission.

Key Recommendations for People with PLD and Caregivers

- **Most liver cysts don't cause symptoms.** When they do, they may lead to bloating, fullness, or discomfort.
- **Women, especially those who've had multiple pregnancies or used estrogen, may be more affected.**
- **Liver imaging (MRI or CT) can show the size and number of cysts.**
- **Treatment is only needed if the symptoms are severe and affect your quality of life.** Options include medications, draining large cysts, surgery, or liver transplant.
- **Liver cyst infections are relatively uncommon, but they occur more frequently in individuals on dialysis or those who've received a kidney transplant.** Liver cyst infections are a serious complication that can result in severe health problems. Common symptoms include fever, abdominal pain, and abnormal blood tests. Your doctor can prescribe antibiotics to treat the infection.
- **There's evidence that using somatostatin analogues (a synthetic form of the naturally occurring hormone) slows down liver growth, as measured by total liver volume (TLV).** Your doctor may suggest starting this medication if you're experiencing severe symptoms of liver enlargement.

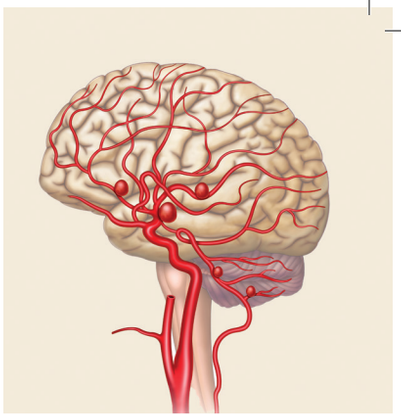
Talking with Your Healthcare Team

Here are some important questions to consider asking your doctor:

- Should my liver cysts be monitored, and if so, how often?
- Are there recommendations available for slowing the growth of liver cysts?
- What can I do if I feel bloated or uncomfortable from liver enlargement?
- Are there treatments that can shrink or remove liver cysts if needed?

Chapter 6:

Understanding Brain Aneurysms and Other Non-Kidney Complications



What This Chapter Covers

ADPKD can affect parts of the body **outside of the kidneys and liver**, including the brain and blood vessels. The most serious concern is **intracranial (brain) aneurysms (or ICA for short)**, which are weak spots in blood vessels that can balloon out and rupture.

An ICA rupture can lead to a **subarachnoid hemorrhage (SAH)**. This is when there's bleeding in the space between the brain and skull. It is considered a life-threatening event.

People with ADPKD have an increased risk of ICAs and SAH. If a family member has had one of these issues, your risk is even higher. Talk with your nephrologist about your risk factors, if you should be screened, and what symptoms to look for.

This chapter helps you identify complications of ADPKD (including aneurysms), understand your risk, review how to monitor signs and symptoms, and provides guidance on what to do if they occur.

Key Recommendations for People with ADPKD and Caregivers

ADPKD brings a risk of brain aneurysm. You may be at increased risk if you have a family history of brain aneurysm, or if you smoke, have uncontrolled high blood pressure, or drink large quantities of alcohol. Symptoms of a brain aneurysm represent a medical emergency and can include (Figure 6):

- Thunderclap headache (a severe headache that comes on suddenly)
- Nausea or vomiting
- Seizures
- Confusion and/or dizziness
- Loss of consciousness

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 6 Key features of a thunderclap headache and possible symptoms of a brain aneurysm, and what to do if they occur. This figure is included in the *KDIGO 2025 ADPKD Guideline* and is being used in this document with permission.

- **Talk to your doctor about whether regular screening (like an MRI of your brain) is right for you.** Brain imaging isn't needed for everyone—but it may be helpful if you have symptoms or a strong family history.
- **Talk with your doctor about the possible complications of ADPKD, as it can affect many organs and tissues throughout the body.** It's important to be aware of potential issues such as:
  - Heart valve issues
  - Blood vessel issues in other parts of the body
  - Abdominal hernias
  - Abdominal or chest discomfort from enlarged organs
  - Cysts on the pancreas and/or spleen
  - Lung issues

Talking with Your Healthcare Team

Here are some important questions to consider asking your doctor:

- Do I need a brain scan based on my family history?
- What symptoms should I look for that might suggest an aneurysm or another complication?
- Are there other body systems I should be monitoring as part of my ADPKD care?

Chapter 7:

Living Well with ADPKD – Lifestyle and Emotional Health

What This Chapter Covers

This chapter is all about you—**your quality of life, mental health, relationships, and day-to-day well-being.** ADPKD affects more than your kidneys. Managing emotional health, reducing stress, staying active, and eating well can make a real difference.

Talk with your healthcare team about a personalized care plan based on your medical, physical, and emotional needs (Figure 7).

Key Recommendations for People with ADPKD and Caregivers

- **Mental health matters.** Depression, anxiety, and fear of the future are common. Talk to your doctor about your emotional well-being at every visit and request a referral to see a counselor if you need more ongoing support.
- **Stay physically active if you can.** Regular movement helps with blood pressure, weight management, mood, and energy.
- **Establish a well-rounded medical team to provide a comprehensive care approach.** This may include the following health professionals:
  - Nephrologist
  - Transplant team
  - Social worker
  - Dietitian
  - Physical therapist
  - Therapist or counselor
- **A healthy, well-balanced diet can support your kidneys.** Ask your doctor or dietitian if you should limit salt, protein, potassium, or phosphorus based on your kidney function. It's recommended that people with ADPKD eat the following foods regularly:
  - Fruits and vegetables
  - Whole grains
  - Fiber
  - Plant-based proteins
  - Unsaturated fats

- **Find the support that fits your needs.** Work with your healthcare team and social worker to help find support and resources related to:
  - Finances
  - Clinical research opportunities (clinical trials)
  - Job training, or help discussing ADPKD with your employer
  - Genetic counseling
  - Family planning guidance
  - Health insurance navigation
- **Connect with others.** Support groups—online or in person—can offer encouragement and practical tips.

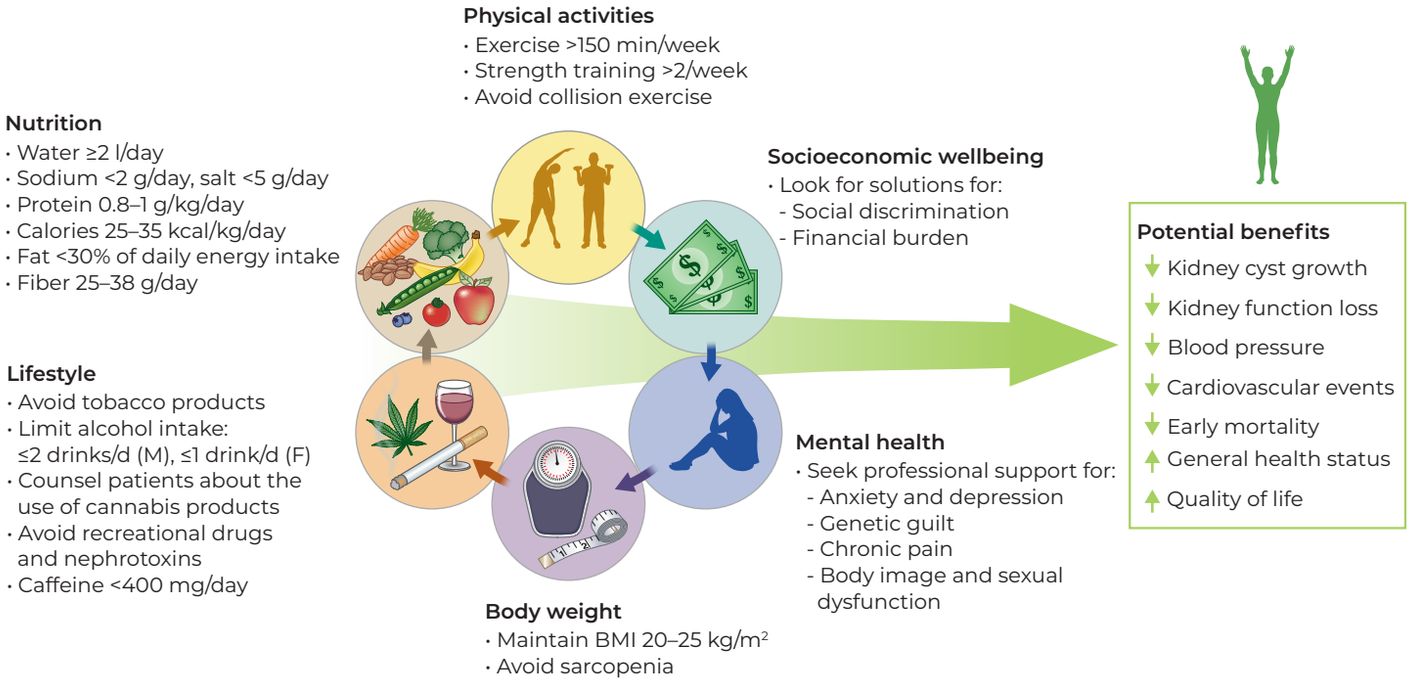


Figure 7 Recommendations for lifestyle, nutrition, physical and emotional wellness, and socioeconomic management aimed at promoting health benefits, including slowing cyst growth and improving quality of life. This figure is included in the *KDIGO 2025 ADPKD Guideline* and is being used in this document with permission.

Talking with Your Healthcare Team

Here are some important questions to consider asking your doctor:

- How can I manage stress and anxiety related to my ADPKD?
- Are there patient support groups or counselors I can connect with?
- Can we talk about a healthy lifestyle plan that supports my kidney health?

Chapter 8:

Understanding ADPKD and Pregnancy

What This Chapter Covers

This chapter explains **what to consider if you're pregnant or thinking about having children** while living with ADPKD. Pregnancy is possible for most people with ADPKD, but it does come with extra risks—especially if you have high blood pressure or reduced kidney function.

Hormone therapy for women with ADPKD and PLD is also discussed, as estrogen-containing contraceptives and hormone replacement therapy may affect growth of liver cysts. Talk to your doctor for more information and special considerations.

Key Recommendations for People with ADPKD and Caregivers

- **Talk with your healthcare team before getting pregnant (Figure 8).** A nephrologist, high-risk OB-GYN (maternal-fetal medicine specialist), and genetic counselor can help you plan for:
  - Medication changes
  - Health risks during pregnancy
  - Reproductive options (sperm or egg donation, preimplantation genetic testing (PGT), or fetal testing for ADPKD)
  - Possible health risks for the child who has a 50% chance of inheriting the condition
  - The risk of inheriting ADPKD for each pregnancy

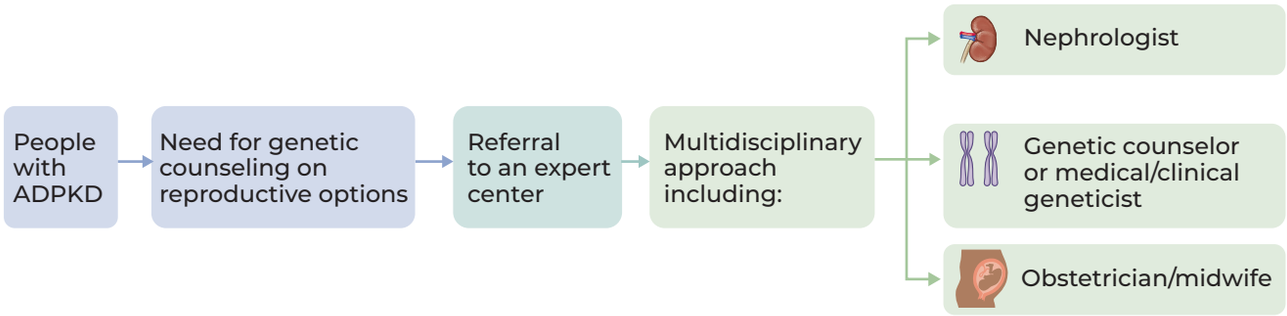


Figure 8 A care approach to family planning involves collaboration with a nephrologist, genetic counselor, and obstetrician for individuals with autosomal dominant polycystic kidney disease (ADPKD). This figure is included in the *KDIGO 2025 ADPKD Guideline* and is being used in this document with permission.

- **High blood pressure and declining kidney function can make pregnancy riskier.** The following is generally recommended during pregnancy:
    - Monthly kidney function and blood pressure testing
    - Maintaining blood pressure below 135/85 mmHg
    - Starting low dose aspirin from weeks 12-36 of pregnancy
    - Monthly screening for a urinary tract infection (UTI)
    - Staying well-hydrated
- **Some blood pressure medications (ACEi and ARB) and other medications, like tolvaptan, must be stopped before pregnancy.** Make a medication plan with your doctor that includes what you'll do before, during, and after pregnancy.
- **Genetic counseling can help if you're concerned about passing ADPKD to your child or would like to explore options to prevent this.**
- **You should see your nephrologist within six months after giving birth.** Talk to your doctor about whether you need to change any of your current medications, if you can start taking any medications you stopped during pregnancy, and how often you should check your blood pressure (Figure 9).
- **Partner with your healthcare team to find the support you need during and after pregnancy.** Seeing a specialist can support both your emotional and physical health during and after pregnancy, including help with nutrition, lactation, and pelvic floor therapy.

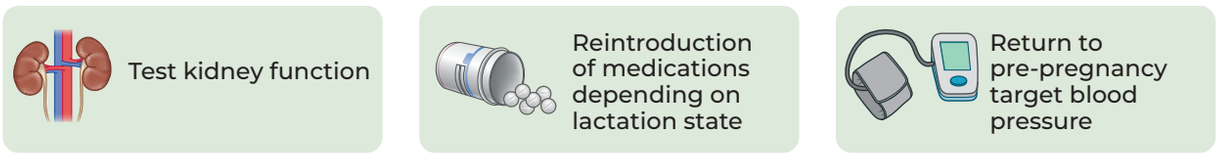


Figure 9 Health recommendations your nephrologist might suggest to support your health after childbirth. This figure is included in the *KDIGO 2025 ADPKD Guideline* and is being used in this document with permission.

Talking with Your Healthcare Team

Here are some important questions to consider asking your doctor:

- Is it safe for me to get pregnant based on my kidney health?
- Do I need to change my medications before trying to conceive?
- How will my blood pressure and kidney function be monitored during and after my pregnancy?
- Are there any other complications from ADPKD to monitor during pregnancy?
- Does having ADPKD put my child at risk for any health issues during and after pregnancy?
- Can I talk to a genetic counselor about genetic testing options and the risk of passing on ADPKD while pregnant?
- What options are available to reduce the risk of passing on ADPKD to future children?

Chapter 9:

Supporting Children with ADPKD

What This Chapter Covers

Although ADPKD is usually considered an adult-onset condition, it can sometimes be diagnosed before birth, or during infancy, childhood, or adolescence. There are currently no approved therapies available for children with ADPKD.

Very early onset ADPKD (VEO-ADPKD) is characterized by an individual presenting with symptoms resulting from ADPKD before 18 months of age.

Early onset ADPKD (EO-ADPKD) is characterized by an individual presenting with symptoms resulting from ADPKD between 18 months and 15 years of age.

It's important that parents use a shared decision-making approach with their child and healthcare team to determine if diagnostic testing is needed and/or appropriate for an at-risk child with a family history of ADPKD.

This chapter also provides guidance for families on how to monitor and care for children with ADPKD.

Key Recommendations for Parents and Caregivers

- **Blood pressure monitoring is important, even in young children.** Here's what you need to know:
  - High blood pressure can start early and may be the first sign of disease.
  - Blood pressure should be checked yearly by your pediatric nephrologist or pediatrician.
  - The recommended target blood pressure for children with ADPKD is less than, or equal to, the 50th percentile for age, sex, and height, or less than 110/70 mmHg in adolescents.
  - Blood pressure medication may be recommended for high blood pressure in children with ADPKD with the use of an ACE inhibitor or ARB as the first choice of therapy.

- **Follow-up care for children with ADPKD is based on the child's clinical presentation and specific needs.** A child's care plan is influenced by factors such as blood pressure, kidney function, imaging results, and the presence of symptoms.
- **Talk with a pediatric nephrologist.** They can guide you through testing, treatment, and support, and provide lifestyle and dietary recommendations for your child.
- **Be mindful of your child's emotional well-being.** Having a genetic condition can bring up fear, guilt, or worry—for both the child and family.
- **Your child will eventually transition to seeing an adult nephrologist when they grow up.** Begin discussing and preparing for this transition in early adolescence with your child, pediatrician, and pediatric nephrologist (Figure 10).

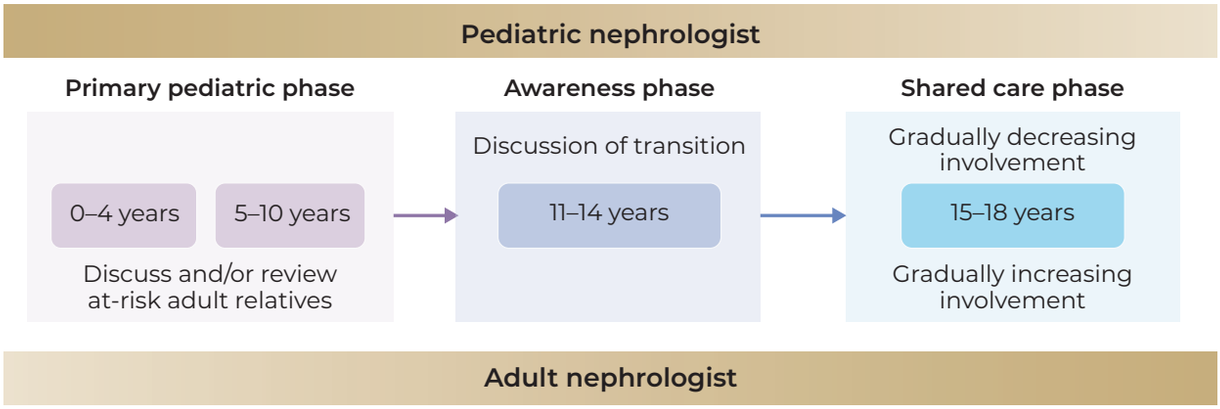


Figure 10 A three-phase approach to transitioning from child to adult kidney care, starting with a primary pediatric phase, followed by an awareness phase, and ending with a shared care phase—using ongoing discussions and gradually increasing patient involvement from early childhood through age 18. This figure is included in the *KDIGO 2025 ADPKD Guideline* and is being used in this document with permission.

Talking with Your Child's Healthcare Team

Here are some important questions to consider asking your doctor:

- Should my child be screened for ADPKD?
- What signs should I watch for as my child grows?
- How do I talk to my child about ADPKD in an age-appropriate way?
- What symptoms should I watch for that may indicate high blood pressure or other complications related to ADPKD?
- When should we see a pediatric nephrologist?

Chapter 10:

Putting It All Together – A Personalized Care Plan for ADPKD

What This Chapter Covers

This final chapter brings everything together. It focuses on creating a **personalized and comprehensive long-term care plan** based on your age, symptoms, disease progression, beliefs, values, and life goals (Figure 11). It also encourages shared decision-making, where patients, families, and providers work as a team to determine the best care possible.

It's important to have a healthcare team you trust—one that supports your active role in managing your health. Being fully aligned with the care plan you and your team create helps you achieve the best possible health outcomes.

Key Recommendations for People with ADPKD and Caregivers

- **No two ADPKD journeys are alike.** Your care plan should reflect your health, values, and personal preferences.
- **Check in regularly with your care team to adjust your plan.** What works now may change over time.
- **Don't be afraid to speak up and advocate for your physical needs and emotional wellbeing.** A multidisciplinary healthcare team (a group of healthcare professionals with different specialties) can help provide holistic and specialized care to meet the needs of those affected by ADPKD. Talk with your doctor for recommendations and referrals.
- **Don't wait to talk about long-term planning.** Transplant, dialysis, or supportive care can all be part of your journey. You deserve time to prepare and choose what's right for you.
- **Ask your doctor about additional resources that can enhance your healthcare.** You may be able to participate in national registries and clinical trials to help improve care for yourself and others with ADPKD.

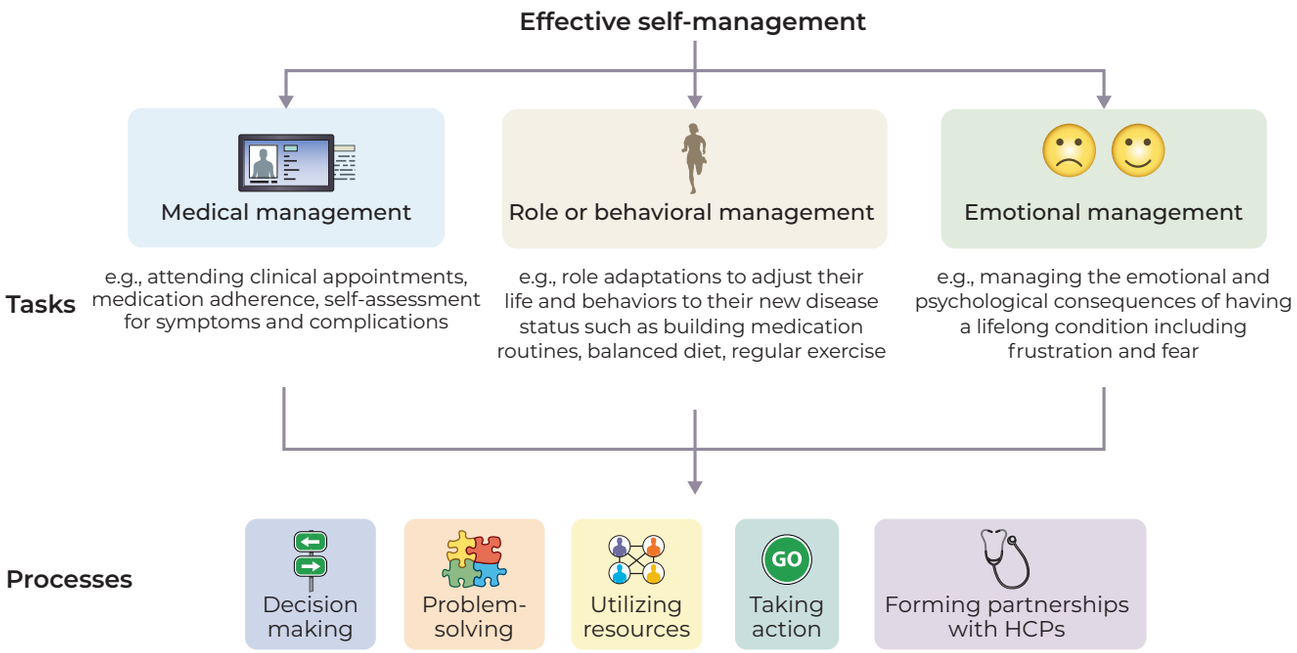


Figure 11 Steps to successfully manage your care by completing medical, emotional, and behavioral tasks—using processes like decision-making and forming partnerships with your healthcare team. This figure is included in the *KDIGO 2025 ADPKD Guideline* and is being used in this document with permission.

HCPs= Healthcare professionals

Talking with Your Healthcare Team

Here are some important questions to consider asking your doctor:

- Can we create a plan based on how my ADPKD is progressing?
- What do we need to think about now and what can wait until later?
- Can we schedule regular check-ins to make sure my care plan is still right for me?
- Which specialists should I consult for both the emotional and physical aspects of ADPKD and will I need a referral to see them?
- What support groups and organizations are available to help me?

For more information on ADPKD including educational resources, community and peer support opportunities, and financial assistance resources visit: https://pkdcure.org/for-patients/

Resources for More Help

Clinical Trials Information

Clinical Trials
www.clinicaltrials.gov

- This is a site managed by the U.S. National Institutes of Health (NIH) where you can learn about clinical studies from around the world.

PKD Clinical Trials
https://pkdcure.org/research/clinical-trials/

- Search for PKD-specific clinical trials and sign up for the Accelerating Clinical Trials (ACT) Alerts.

PKD Foundation Support

ADPKD Registry
go.pkdcure.org/KDIGO_ADPKDRegistry

- If you have ADPKD, your experiences could be the key to helping future generations. It's the nation's first dedicated ADPKD Registry and one of the first patient registries of any kind to integrate patient-provided health records. By sharing your disease experience and connecting your electronic health records, your data can help power the path to a cure.

PKD HOPE Line
https://pkdcure.org/get-connected/hope-line/

- Connect with our support team by dialing **844-PKD-HOPE (844-753-4673)**. Our team is available Monday through Friday from 8 a.m. – 5 p.m. (CT) to answer your questions and direct you to the resources you need.

PKD Parents Community
https://pkdcure.org/get-connected/community/parents/

- The PKD Parents Community offers support, compassion, and guidance and can help connect families with experts and other local PKD families.

PKD Thrive Community
https://pkdcure.org/for-parents/pkd-thrive/

- The PKD Thrive Community was launched in 2022 to bring together young adults with PKD. PKD Thrive offers support, compassion, and a safe place to connect and interact with others who understand where you are in your PKD journey. Email **volunteers@pkdcure.org** for more information and to get involved.

PKD Connect Peer Mentors
https://pkdcure.org/get-connected/peermentors/

- PKD Connect Peer Mentors provide resources, guidance, motivation, and emotional support to people impacted by PKD.

PKD Communities
https://pkdcure.org/get-connected/community/

- Communities are groups of PKD patients, family members, and loved ones - who want to learn, connect, and take action with others.

Area for notes when visiting your healthcare team

PKD FOUNDATION
Polycystic kidney disease

pkdcure.org

1.800.PKD.CURE

Scan for More Information for Patients

Scan for More Information for Clinicians

PKD FOUNDATION

Polycystic kidney disease

www.pkdcure.org