



PKD FOUNDATION
Polycystic Kidney Disease

Understanding your risk for ADPKD progression

ADPKD=autosomal dominant polycystic kidney disease.

Understanding ADPKD

Autosomal dominant polycystic kidney disease, or ADPKD, is a progressive, inherited disease (passed from an affected parent to their child) that causes cyst growth in the kidneys that gets worse over time.

Each child of a parent with ADPKD has a **50%** chance of inheriting the disease.

Approximately **10%** of people diagnosed with ADPKD have no family history.

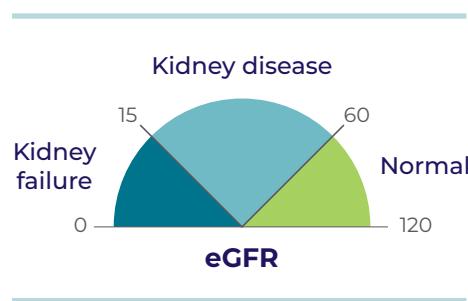
Cysts grow in or on the kidney and as they get larger they reduce the kidneys' ability to filter your blood over time.

Labs may not show that your kidneys are getting bigger

Your doctor may use something called **eGFR, or estimated glomerular filtration rate**, to measure how well your kidneys are doing their job of filtering your blood.

This test shows how well your kidneys are working in that moment, but depending on the stage of your disease, it may not show if your ADPKD is getting worse over time.

By only measuring eGFR, your kidneys may *appear* to function normally, even though the cysts may be growing and your kidneys may be getting bigger. Because patients can experience ADPKD differently—even those in the same family—some patients may not see a decline in eGFR for years. It is important to get imaging tests done to understand the status of your disease and to help predict the rate of your disease progression.



► Even if you have a family member who has ADPKD, your disease may be a little different. That is why it is important to know how your own disease is changing over time and the rate at which it is progressing.

► Learn more about the stages of ADPKD at pkdcure.org/stages-of-adpkd.

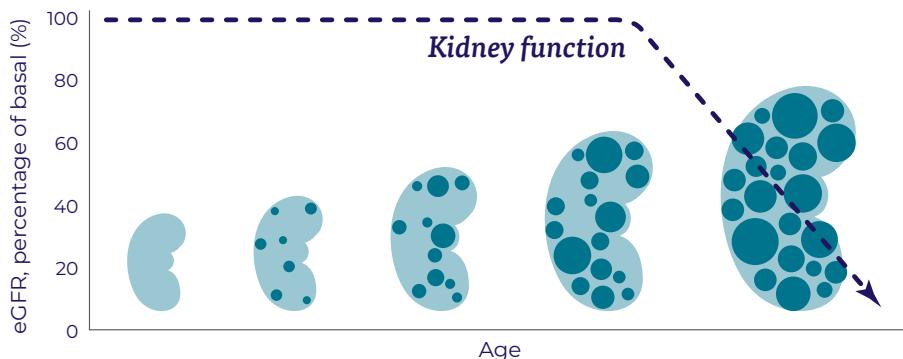
Kidney size is a predictor of disease progression for ADPKD

A normal kidney weighs about a third of a pound. Because of the cysts, kidneys affected by ADPKD can grow to weigh many pounds. This increase in **total kidney volume, or TKV**, can be up to 4 times that of normal kidneys. As cysts grow, the kidneys continue to get bigger, and the damage continues to get worse.

Patients with bigger kidneys are more likely to experience more rapid loss of kidney function than patients with smaller kidneys.

► Some patients experience pain. Talk to your nephrologist and visit pkdcure.org for information on how you can manage this pain.

Your eGFR may remain stable while your kidneys are growing



Adapted from Grantham JJ, et al. *Nat Rev Nephrol.* 2011;7(10):556-566

► Growing cysts can cause damage to your kidneys that is permanent. It is important to talk to your nephrologist about how regular testing can help monitor for growing cysts.

On average, your TKV as measured by magnetic resonance imaging (MRI) will increase by about 5% per year, even though your kidney function may stay in the normal range for decades.

At diagnosis

Initially, patients may present with symptoms such as pain, high blood pressure, and blood in the urine. These symptoms alone are not enough to diagnose ADPKD.

► If you are not seeing a nephrologist, you can ask your doctor for a referral.

Your nephrologist, who specializes in kidney disease, will determine if you have ADPKD based on:



Your **family history** of the disease



Imaging tests

- If your kidneys are bigger than they should be
 - The number of cysts you have in your kidneys
-

A positive family history is known in about 75% of patients with ADPKD. This is helpful to identify other family members who may be at risk. Genetic testing is available to help confirm a diagnosis of ADPKD.

It may be helpful to bring someone to your appointments. You can also refer to pkdcure.org for resources and support.

There are 3 imaging tests your doctor can use to assess ADPKD.

If you have ADPKD, these tests will show how many cysts you have, how big they are, and the progression of your disease.

1. Ultrasound

- Most common imaging test done to screen for PKD
- Is a safe test that uses sound waves and does not require the use of radiation or contrast dye to be injected
- It is the least costly screening method
- Is an accurate test for diagnosing but may lack precision for detecting changes in kidney volume
- Can be used to measure kidney size (length)

2. Computed tomography (CT)

- CT scans are a sophisticated form of an X-ray
- CT scans involve radiation or may also require iodinated contrast dye, which can be harmful to the kidneys
- A CT scan is more sensitive than an ultrasound for detecting small cysts
- May be used to look at complications of ADPKD

3. Magnetic resonance imaging (MRI)

- MRIs take pictures of the body using a magnet
- An MRI is another test that is more sensitive than ultrasounds
- May cost more than ultrasound, depending on your insurance coverage
- Not widely available
- MRIs provide an accurate reading of kidney volume
- May also be used to look at complications of ADPKD

► *In the beginning, these tests will help your doctor diagnose you with ADPKD, but they can continue to be important in understanding the progression of your disease as well. Speak with your doctor about your current test results.*

Know the risk of your disease progression

Progression of ADPKD looks different for everyone, even people in the same family, so it's important to understand if you are at risk of your ADPKD progressing at a faster rate.

If you have a faster rate of progression, your kidney function will get worse and cysts will grow more quickly, leading to earlier kidney failure. Larger kidneys can lead to earlier high blood pressure, more pain, earlier kidney damage, and early kidney failure.

TKV can help predict disease progression in ADPKD

Doctors can measure your kidney volume based on imaging tests or scans and will adjust based on your height. Then, they will see if the size of your kidneys puts you at risk for rapid disease progression.

For instance, 2 people might have the same size kidneys. But in a shorter person they would be a greater risk. Or, if 2 people of the same height have the same size kidneys, the younger person may be at risk for rapid disease progression.



Each of these patients has a combined kidney volume, also called total kidney volume (TKV), of 800 mL. Each of their kidneys is about 400 mL, which is slightly more than 13 fluid oz.

► Your kidneys may continue to function normally, even as cysts grow and damage is getting worse. Imaging your kidneys allows your nephrologist to understand the status of your ADPKD.

► Knowing your kidney size is important to understanding your disease. Imaging and scans can tell your doctor how big your kidneys are and if you are at risk of rapid progression.

In patients with rapid disease progression, this can mean problems with cysts at a young age. This includes pain, high blood pressure, blood in your urine, and rapid loss of kidney function.

You may be at higher risk for rapid disease progression if you:



Have high blood pressure before age 35



Experience a fast drop in eGFR



Have blood in the urine before age 35



Are overweight ($BMI \geq 25$)



Have a family history of kidney failure, such as the need for dialysis or kidney transplant, before age 58



Have certain types of genetic mutations

► *Growing cysts can cause scarring and damage leading to reduced kidney function. It is important to talk to your doctor about how regular imaging tests can help monitor for growing cysts.*

Partner with your nephrologist

Your nephrologist can better understand the progression of your ADPKD by:

- ✓ Knowing your risk factors for rapid disease progression.
- ✓ Performing a TKV or total kidney volume measurement test.

Measuring TKV can give your nephrologist a lot of information about your disease and how quickly it might get worse. Your nephrologist will request an ultrasound, MRI, or CT scan to get a more detailed picture of your kidneys. TKV may be calculated from measurements taken from imaging tests. Depending on the type of test your nephrologist orders, your nephrologist will work with their radiologist to take specific measurements of your kidneys.

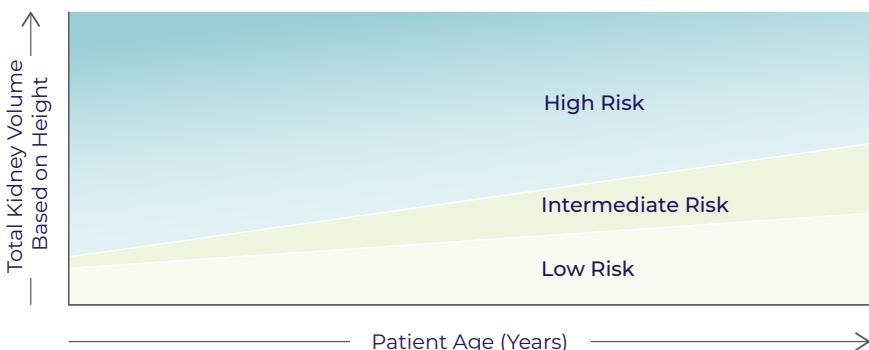
Normal TKV is 250-350 mL. Recent studies have shown that TKV predicts the risk of kidney function decline, making it a verified metric for nephrologists. Because of this, TKV is used to evaluate disease outcomes in clinical trials of novel treatments for ADPKD.

► *Talk to your nephrologist about his or her experience treating ADPKD, as some have more experience than others. You may have a choice to select a nephrologist who understands your disease and is aligned with your goals. Find one who is proactive in your disease management goals.*

Your TKV can help your nephrologist determine your classification to understand your risk for rapid disease progression. Once you have these measurements, your nephrologist may be able to determine your risk for decline by plotting it on a graph, similar to the one below.

Your nephrologist will also assess your eGFR to see how well your kidneys are working.

ADPKD imaging classification can help predict your rate of disease progression



Mayo Clinic classification diagram. Republished with permission of the American Society of Nephrology, from Imaging classification of autosomal polycystic kidney disease: a simple model for selecting patients for clinical trials. *J Am Soc Nephrol.* 2015;26(1):160-172

It is important for you and your nephrologist to know if you are at risk for rapid disease progression in order to help choose the right disease management strategy for you.

► Your nephrologist may work closely with the radiologist to ensure the right data is collected to calculate your TKV.

► This is not a diagnostic tool. Your nephrologist can use this assessment tool to help predict future decline.

What if I feel fine?

You may feel okay and your eGFR may be normal, but there is a “growing problem” with your kidneys and your disease may be progressing. Working with your nephrologist to learn more about your kidneys is the best way to determine the rate at which your disease is progressing.

The following are some questions you may want to ask your nephrologist so you can understand your disease better and create an action plan together:

Where am I on the Mayo Clinic Classification?

How big are my kidneys? And what is my TKV?

What is the rate of my disease progression?

What did my latest MRI/CT show in terms of progression?

► It may help to take a family member or friend along, if possible, to help you advocate for yourself or to remember the information you receive.

How will we track the progression of my ADPKD?

How do we manage my disease over time?

What are the best ways I can manage my disease?

What should I expect next?

My action plan

How to take care of yourself and your disease

There are a number of things you and your doctor can do to help manage your disease as best as possible. Be sure to discuss the following with your nephrologist:



How to manage hypertension (high blood pressure) or pain.



Maintain a kidney-friendly diet. Discuss what this means with your nephrologist.



It is important to stay hydrated, but discuss the appropriate level of water intake.



Exercise is an important part of maintaining overall good health.



Make sure you are scheduling regular visits with your doctors (ideally your nephrologist).



Build a strong support system of family and friends.



How you may help slow the progression of your disease.

For support and resources, visit:

Foundations and Informative Sites

PKD Foundation

Dedicated to PKD research and education

pkdcure.org

American Association of Kidney Patients (AAKP)

Education and advocacy for kidney patients in the United States

aakp.org

National Kidney Foundation

A resource for all people affected by kidney disease

kidney.org

Other Support and Resources

AAKP Renal Support Groups

A list of renal support groups, broken down by state

American Kidney Fund

Providing programs of prevention, early detection, financial support, disease management, clinical research, innovation, and advocacy

PKD Connect

A community of care and resource center

PKDRecipes.com

Recipes for polycystic organ health



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