CONDITION

Autosomal dominant polycystic kidney disease

Learn how autosomal dominant polycystic kidney disease affects the kidneys, how it is inherited, and how monitoring and treatment can reduce complications.

WHAT IT IS
Inherited disease causing multiple kidney cysts

BODY SYSTEM
Kidneys and urinary system

MAIN SYMPTOMS
Side or back pain, blood in urine, urinary infections, kidney stones

WHAT CAUSES IT
Inherited gene change

HOW IT IS DIAGNOSED
Kidney ultrasound, MRI or CT

MAIN TREATMENT
Blood pressure control and complication management

CAN IT BE CURED?
No, treatments only slow progression

IS IT AN EMERGENCY?
Sometimes, with a sudden worst-ever headache

LONG-TERM OUTLOOK
Variable, from lifelong kidney function to kidney failure

Overview

Autosomal dominant polycystic kidney disease, or ADPKD, is an inherited condition in which many fluid-filled sacs called cysts develop in the kidneys. The cysts can grow over time, making the kidneys larger and interfering with their ability to filter waste and extra fluid from the blood.

ADPKD is different from isolated simple kidney cysts, which become more common with age and usually do not cause the same pattern of kidney enlargement or inherited disease. It is also different from autosomal recessive polycystic kidney disease, a rarer condition that usually begins in infancy or childhood.

The course of ADPKD varies widely. Some people maintain useful kidney function throughout life, while others develop chronic kidney disease and eventually kidney failure. High blood

pressure is common and may appear before kidney function falls. ADPKD can also affect other parts of the body, especially the liver and blood vessels in the brain.

Early recognition matters because blood pressure control, healthy daily habits, appropriate monitoring, and treatment for people at risk of rapid progression can help protect kidney function and reduce complications.

Symptoms

ADPKD often causes no symptoms during its early stages. A person may learn they have it after an imaging test for another reason, during family screening, or after developing high blood pressure.

Possible symptoms include:

Pain may come from an enlarging kidney, bleeding into a cyst, infection, or a kidney stone. Blood in the urine can have several causes, so it should not automatically be assumed to come from ADPKD.

Liver cysts are common, particularly with increasing age, but they usually do not prevent the liver from working. A person with extensive liver cysts may develop abdominal fullness, discomfort, or early fullness when eating.

A sudden, extremely severe headache is not a routine ADPKD symptom. It can be a sign of bleeding from a brain aneurysm and requires emergency care, especially if it occurs with vomiting, fainting, neck stiffness, confusion, weakness, or trouble speaking.

- Pain or pressure in the side, abdomen, or lower back
- Blood in the urine, which may make urine look pink, red, or brown
- Frequent urinary tract infections
- Kidney infections, sometimes with fever and side pain
- Kidney stones
- A feeling of abdominal fullness as the kidneys or liver become larger
- Headaches related to high blood pressure
- Tiredness, swelling, nausea, itching, or changes in urination when kidney disease becomes advanced

Causes and risk factors

ADPKD is caused by a disease-related change in a gene involved in the structure and function of kidney cells. Most cases involve the PKD1 or PKD2 gene. Changes in several less common genes can cause similar or milder cystic kidney disorders.

"Autosomal dominant" describes the inheritance pattern. A person who has an ADPKD-causing gene change has a 50% chance of passing that change to each child. The chance is the same for every pregnancy and does not depend on the child's sex.

Not everyone with ADPKD has a known family history. The condition may result from a new genetic change, or relatives may have had mild or unrecognized disease. Family members with the same genetic change can also have different ages of onset and different rates of kidney decline.

Factors associated with a greater chance of faster progression include certain PKD1 variants, large kidneys for a person's age and height, high blood pressure or urinary complications at a younger age, and a family history of kidney failure at a relatively young age. These factors help clinicians estimate risk, but they cannot predict an individual's course with certainty.

Lifestyle does not cause ADPKD. However, smoking, uncontrolled blood pressure, high dietary sodium, and other conditions that strain the kidneys may add to the risk of kidney or cardiovascular problems.

Diagnosis

Diagnosis usually begins with a medical history, family history, physical examination, blood pressure measurement, blood tests, and kidney imaging.

Ultrasound is often the first imaging test. It uses sound waves to look for kidney cysts and does not involve radiation. The number of cysts needed to support a diagnosis depends on the person's age and whether a parent has confirmed ADPKD. Age matters because isolated simple cysts become more common later in life.

Magnetic resonance imaging, or MRI, provides more detailed images and can detect smaller cysts. Computed tomography, or CT, may also show cysts but involves radiation and may require contrast material. MRI or CT may be used when ultrasound findings are unclear or when a clinician needs to measure kidney size.

For some adults, MRI measurements of total kidney volume can help estimate how quickly the disease may progress. This information is interpreted together with age, kidney function, genetic findings, and medical history. It is not needed for every person.

Blood tests measure creatinine, which is used to estimate the glomerular filtration rate, or eGFR. The eGFR is an estimate of how well the kidneys filter blood. Urine testing can look for

blood, infection, or albumin, a protein that may enter the urine when kidneys are damaged.

Genetic testing may be considered when imaging is uncertain, there is no clear family history, a related person is being evaluated as a potential kidney donor, or the results could affect family planning or treatment decisions. A negative genetic test does not always exclude ADPKD because testing may not identify every disease-causing change.

Testing for brain aneurysms is not automatically needed for everyone with ADPKD. Screening with magnetic resonance angiography may be discussed for people with a personal history of aneurysm or brain bleeding, a family history of aneurysm or unexplained sudden death, or other circumstances in which knowing the result would affect major decisions.

Treatment and management

There is no single treatment that removes all kidney cysts. Management focuses on slowing kidney damage, controlling symptoms, and preventing or treating complications.

Blood pressure control is a central part of care. Angiotensin-converting enzyme inhibitors like lisinopril or ramipril, called ACE inhibitors, or angiotensin receptor blockers, called ARBs, are commonly used when medication is needed. Blood pressure goals and medicine choices depend on age, kidney function, pregnancy plans, and other health conditions. These medicines require clinician supervision and periodic blood tests.

Tolvaptan is a medicine that may be offered to selected adults who are at risk of rapidly progressing ADPKD. It blocks the effect of vasopressin, a hormone involved in water balance and cyst growth. Clinical trials found that it can slow kidney enlargement and loss of kidney function, but it does not cure ADPKD.

Tolvaptan causes increased urination and thirst and can cause serious liver injury. People taking it need scheduled liver blood tests and guidance about fluid intake, illness, and other medicines. Whether its potential benefit outweighs its burdens depends on an individualized assessment of progression risk.

Daily habits that may support kidney and cardiovascular health include:

Fluid advice must be individualized. Drinking large amounts of water may be unsafe for some people, including those with advanced kidney disease, heart problems, or medicines that affect fluid balance.

Nonsteroidal anti-inflammatory drugs like ibuprofen or naproxen , or NSAIDs, include ibuprofen and naproxen. Frequent or prolonged use can reduce blood flow to the kidneys. A clinician can help identify safer pain strategies. Persistent pain may require evaluation for infection, stones, cyst bleeding, or another cause. In selected cases, pain specialists or surgical

procedures may be considered.

Urinary tract and kidney infections are treated with antibiotics like amoxicillin or doxycycline chosen for the infection and the person's kidney function. An infected cyst may require a longer course of treatment or drainage. Kidney stones may be managed with pain control, hydration guidance, testing of the stone or urine, and procedures when a stone blocks urine flow or does not pass.

If kidney failure develops, treatment options include dialysis and kidney transplantation. ADPKD does not usually return in a transplanted kidney. Very large native kidneys are removed only in selected situations, such as repeated infection, severe symptoms, bleeding, or lack of space for a transplant.

People who are pregnant or planning pregnancy should discuss medicines and risks with their care team in advance. Some medicines used for blood pressure or ADPKD are not appropriate during pregnancy.

- Keeping dietary sodium within the limit recommended by the care team
- Staying physically active within personal abilities
- Maintaining a healthy weight
- Avoiding tobacco
- Drinking enough fluid to prevent dehydration when medically appropriate
- Reviewing nonprescription medicines and supplements with a clinician

Complications

Possible complications include chronic kidney disease, kidney failure, high blood pressure, kidney stones, cyst bleeding, and kidney or cyst infections.

ADPKD can also cause problems outside the kidneys. Liver cysts are the most common. Less frequent complications include changes in heart valves, abdominal wall hernias, and diverticular disease of the colon.

An intracranial aneurysm is a weakened, bulging area in an artery in the brain. Most aneurysms do not rupture, but rupture can cause life-threatening bleeding around the brain. Risk assessment and screening decisions should consider personal and family history rather than the ADPKD diagnosis alone.

Long-standing high blood pressure and reduced kidney function also increase the risk of heart and blood vessel disease. Managing blood pressure, cholesterol, diabetes, tobacco exposure,

and physical activity remains important throughout care.

Follow up

Routine follow-up may involve a primary care clinician, nephrologist, or both. A nephrologist is a doctor who specializes in kidney disease. The timing of visits depends on blood pressure, symptoms, kidney function, progression risk, and treatment.

Monitoring may include:

Contact a clinician promptly for fever with side or back pain, painful urination, persistent visible blood in the urine, blood clots in the urine, sudden severe flank pain, repeated vomiting, or a major decrease in urine output. These symptoms may reflect infection, bleeding, a stone, or urinary blockage.

Seek emergency care for a sudden "worst-ever" headache, fainting, seizure, new weakness or numbness, confusion, trouble speaking, severe chest pain, or severe difficulty breathing. These are not routine follow-up concerns.

- Blood pressure checks
- Creatinine, eGFR, electrolytes, and urine testing
- Review of pain, infections, stones, and blood in the urine
- Review of medicines, supplements, and possible kidney side effects
- Imaging when it is likely to change risk assessment or treatment
- Counseling about pregnancy, family testing, or genetic testing when relevant

Sources

- Kidney Disease: Improving Global Outcomes. KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of Autosomal Dominant Polycystic Kidney Disease . Kidney International. 2025;107(2S):S1-S239.
- National Institute of Diabetes and Digestive and Kidney Diseases. Autosomal Dominant Polycystic Kidney Disease . National Institutes of Health.
- Harris PC, Torres VE. Polycystic Kidney Disease, Autosomal Dominant . GeneReviews, University of Washington, Seattle. Updated September 29, 2022.



- Cornec-Le Gall E, Alam A, Perrone RD. Autosomal dominant polycystic kidney disease. The Lancet . 2019;393(10174):919-935.
- Otsuka America Pharmaceutical, Inc. Jynarque (tolvaptan) tablets: U.S. prescribing information . DailyMed, U.S. National Library of Medicine.