**TEAM 3**

**NEPHROLOGY (KIDNEYS AND HYPERTENSION)**

Electrolyte Disorder

Renal artery stenosis

hypertensive nephrosclerosis

renovascular hypertension

polycystic kidney disease

glomerulonephritis

chronic kidney disease

nephrotic syndrome

kidney stone

kidney cancer

acute kidney infection

end stage renal disease

endocrine related hypertension

urinary tract infection

LUPUS NEPhritis

IGA nephropathy

Minimal change disease

Amyloidosis

Hemolytic uremic syndrome

Henoch schonlein purpura

Primary hyperoxaluria

Pyelonephritis

Medullary cystic kidney disease

Nephrogenic systemic fibrosis

Malarial nephropathy

Abderhalden kaufmann lignac syndrome

Alport syndrome

Interstitial nephritis

NODULAR GLOMERULOSCLEROSIS

# **Electrolyte Disorder**

Other names: electrolyte imbalance

## **Definition and Description**

Electrolytes are chemicals naturally occurring in your body fluids. They include chloride, phosphate, potassium, sodium, and calcium. These are important for normal body functions and should be present in certain concentrations.

The kidneys maintain a balance of electrolytes by shifting sodium levels as the body requires.

When the level of electrolytes in your body is too low or too high, the resulting condition is called an electrolyte imbalance.

Maintaining an electrolyte balance is vital for your body to function smoothly.

### **What are the types of electrolyte disorders?**

Electrolyte disorders have different names based on which mineral is out of balance.

They also use a prefix based on whether the electrolyte level is too high or too low:

* **Hyper-** means too high.
* **Hypo-** means too low.

The most common types of electrolyte disorders are:

* **Hypercalcemia** — Calcium levels are too high.
* **Hypocalcemia** — Calcium levels are too low.
* **Hyperchloremia** — Chloride levels are too high.
* **Hypochloremia** — Chloride levels are too low.
* **Hyperkalemia** — Potassium levels are too high.
* **Hypokalemia** — Potassium levels are too low.
* **Hypermagnesemia** — Magnesium levels are too high.
* **Hypomagnesemia** — Magnesium levels are too low.
* **Hypernatremia** — Sodium levels are too high.
* **Hyponatremia** — Sodium levels are too low.
* **Hyperphosphatemia** — Phosphate levels are too high.
* **Hypophosphatemia** — Phosphate levels are too low.

### **Electrolytes are essential. They help:**

* Regulate fluid levels in the body and blood plasma
* Keep the blood pH in the normal range
* Allow muscle contractions, including heartbeat
* Transmit messages from the muscles, nerve cells, heart, and other cells
* Help in blood clotting
* Form new tissues

## **Causes and Risk Factors**

### **Causes**

Water makes up more than half of your body’s weight. Blood and fluid in and around cells (called fluid compartments) hold most of this water. Your kidneys and liver, as well as other organs and tissue, continually move electrolytes in and out of cells to adjust fluid levels within the compartments.

Certain health conditions can affect your body’s ability to move and balance electrolytes. When fluid compartments have too many or too few electrolytes, you have an electrolyte imbalance.

Some of the common causes of electrolyte disorders seen in clinical practices are:

* Hyponatremia: low dietary sodium intake, primary polydipsia, syndrome of inappropriate antidiuretic hormone secretion (SIADH), heart failure, cirrhosis, adrenal insufficiency, prolonged hyperglycemia, and severe dyslipidemia.
* Hypernatremia: unreplaced fluid loss via the skin or gastrointestinal tract, osmotic diuresis, or hypertonic saline administration.
* Hypokalaemia: hyperaldosteronism or the use of loop diuretics.
* Hyperkalaemia: metabolic acidosis, insulin deficiency, hypoaldosteronism, prolonged beta-blocker use, or acute or chronic kidney disease.
* Hypercalcemia: malignancy, hyperparathyroidism, or chronic granulomatous diseases such as tuberculosis or sarcoidosis.
* Hypocalcaemia: acute pancreatitis, iatrogenic parathyroid dysfunction, resistance to parathyroid hormone, hypomagnesemia, or sepsis.
* Hypermagnesemia: increased oral magnesium intake.
* Hypomagnesemia: increased renal losses with diuretics, alcohol use disorder, or gastrointestinal losses.
* Bicarbonate level: increases in primary metabolic alkalosis or compensation to primary respiratory acidosis and decreases in primary metabolic acidosis or compensation to primary respiratory alkalosis.
* Hyperchloremia: excessive normal saline infusion.
* Hypochloremia: increased gastrointestinal or renal losses.
* Hypophosphatemia: refeeding syndrome, vitamin D deficiency, or hyperparathyroidism.
* Hyperphosphatemia: hyperparathyroidism or chronic kidney disease.

### **Risk Factor**

Infants, young children and older adults are more prone to changes in electrolyte levels, but an imbalance can happen to anyone.

Certain conditions can also throw off your body’s electrolyte levels. You may be more likely to develop an electrolyte imbalance if you have:

* Burns.
* Cancer.
* Cardiovascular disease, heart failure or high blood pressure.
* Dehydration due to not drinking enough liquids or from excessive vomiting, diarrhea, sweating (hyperhidrosis) or fever.
* Over hydration or water intoxication (drinking too much water).
* Eating disorders.
* Kidney disease.
* Liver disease like cirrhosis.
* Substance use disorder.

Certain medications can also affect electrolyte levels. These include:

* Antibiotics.
* Chemotherapy drugs.
* Corticosteroids.
* Diuretics and laxatives.

## **Signs and symptoms**

Since electrolytes are needed for essential body functions, an increase or decrease in their number is quickly noticeable. Some symptoms of electrolyte imbalance are:

* Cramps
* Dizziness
* Irregular heartbeat
* Mental confusion
* agitation
* dry mouth and thirst
* restlessness
* confusion or difficulty with cognition
* muscle weakness or spasms
* numbness or tingling
* fatigue
* heart palpitations
* constipation
* nausea or vomiting
* slow or irregular heart rate
* difficulty breathing
* low or high blood pressure
* itching

## **Diagnosis Methods**

Some electrolyte disorders do not cause noticeable symptoms and are diagnosed during routine physical exams or by testing for other conditions. If you have symptoms, your doctor will perform a physical exam, talk to you about your health history, and order blood and urine tests to diagnose electrolyte imbalances.

* **Anion gap blood test** — Checks your body’s acid-base balance and looks for electrolyte imbalances.
* **Electrolyte panel** — Checks levels of electrolytes in your blood.
* **Urine electrolyte test** — Checks levels of electrolytes in your urine.

Your doctor may also order an estimated glomerular filtration rate (eGFR) test, which shows how efficiently your kidneys are functioning based on your age, creatinine level, and sex.

A simple blood test can measure the levels of electrolytes in your body. A blood test that looks at your kidney function is important as well.

Your doctor may want to perform a physical exam or order extra tests to confirm a suspected electrolyte imbalance. These additional tests will vary depending on the condition in question.

For example, hypernatremia (too much sodium) can cause skin elasticity loss due to significant dehydration. Your doctor can perform a pinch test to determine whether dehydration affects you.

An electrocardiogram (ECG or EKG), an electrical tracing of your heart, may also be useful to check for any irregular heartbeats, rhythms, or ECG or EKG changes brought on by electrolyte problems.

## **Treatment options**

Treatment varies depending on the type of electrolyte imbalance and the underlying condition causing it. Certain treatments are generally used to restore the proper balance of minerals in the body. These include:

### **Intravenous (IV) fluids**

Intravenous (IV) fluids, typically containing sodium chloride, can help rehydrate the body. This treatment is commonly used in cases of dehydration resulting from vomiting or diarrhea. Electrolyte supplements can be added to IV fluids to correct deficiencies.

### **Certain IV medications**

IV medications can help your body restore electrolyte balance quickly. They can also protect you from negative effects while being treated by another method.

The medication you receive will depend on the electrolyte imbalance you have. Medications that may be administered include calcium gluconate, magnesium sulfate, and potassium chloride.

### **Oral medications and supplements**

Oral medications and supplements are often used to correct chronic mineral abnormalities in your body. This is more common if you’ve been diagnosed with ongoing kidney disease.

Depending on your electrolyte imbalance, you may receive medications or supplements such as:

* calcium (gluconate, carbonate, citrate, or lactate)
* magnesium oxide
* potassium chloride
* phosphate binders, which include sevelamer hydrochloride (Renagel), lanthanum (Fosrenol), and calcium-based treatments such as calcium carbonate

These can help replace depleted electrolytes on a short- or long-term basis, depending on the underlying cause of your disorder. To correct the imbalance, your doctor will usually treat the underlying cause.

### **Hemodialysis**

Hemodialysis is a type of dialysis that uses a machine to remove waste from your blood.

One way to get the blood to flow to this artificial kidney is for your doctor to surgically create a vascular access, or an entrance point, into your blood vessels.

This entrance point will allow Trusted Source a larger amount of blood to flow through your body during hemodialysis treatment. This means more blood can be filtered and purified.

Hemodialysis can be used to treat an electrolyte imbalance. Your doctor may also decide on hemodialysis treatment if the electrolyte problem has become life threatening.

## **Prevention Tips**

Maintaining a balance of electrolytes in your body is relatively easy:

**Drink water.** Make sure you are drinking water two hours before going to the gym or doing any other kind of physical activity.

Also, try to drink at least four to six ounces of water after every 20 minutes of physical activity, and drink water after exercising.

Drinking water is the simplest and the best way to prevent electrolyte imbalance complications.

Other fluids that help balance your electrolytes include:

**Coconut water.** Coconut water has a low sugar level and will not cause a sugar spike in your blood. Still, it has more calories than tap water. If possible, opt for unsweetened coconut water to cut down calories.

**Electrolyte or sports drinks.** Sports drinks also help make up for the lost electrolytes since they contain electrolytes. Most of them have potassium chloride and sodium chloride. If you work out for less than 75 minutes, standard water should work fine.

If you are exercising for a longer interval, an electrolyte drink will replenish your electrolyte count quickly. Typically, eight ounces of an electrolyte drink has 100 milligrams of sodium and 30 milligrams of potassium. Make sure to read the label to check if the drink has electrolytes, though, since some of them don't.

**Eat electrolyte-rich foods.** You can also eat certain foods to increase electrolyte levels in the body, such as potatoes, avocados, oranges, bananas, strawberries, turkey, and spinach.

‌ Don't rehydrate with carbonated or energy drinks. They may cause a sudden spike in your blood sugar levels.

To prevent electrolyte imbalance, drink plenty of water during physical activity. Eat a balanced diet containing electrolyte-rich foods. Don't engage in strenuous activity outdoors during hot weather. If you're working out inside, don't do it without an air conditioner, especially if you sweat heavily.

## **Prognosis**

Prognosis depends upon the severity of the deficiency, the arrival of the patient at the right time, efficiency of the diagnosis, and management of the disorder. Usually, the electrolytes imbalance is easily diagnosed by the blood test and corrected on time. However, the chances of deaths are high in significantly older adults, very young children, and critically ill patients.

## **Possible complications**

Without treatment, electrolyte disorders can become life-threatening and cause:

* Cardiac arrest.
* Comas.
* Seizures.

## **When to See a Doctor/ Red Flag**

You should call your healthcare provider if you experience:

* Changes in heart rate.
* Extreme fatigue.
* A prolonged bout of diarrhea or vomiting.
* Signs of dehydration.
* Unexplained confusion, muscle cramps, numbness or tingling.

## **Differential diagnosis**

The signs and symptoms of electrolyte imbalance can also be due to the following disorders, which should be ruled out to progress towards an efficient treatment;

* Any severe infection
* Neurological infections or encephalopathy
* Dehydration
* Adrenal Crisis
* Alcoholism
* Cardiogenic Pulmonary Edema
* Cirrhosis
* Hypothyroidism
* Hypoalbuminemia
* Metabolic Alkalosis

## **Any Severe Infection**

## A serious systemic or localized infection that can cause significant physiological disturbance, including sepsis, septic shock, or organ dysfunction. Severe infections often require urgent medical intervention and can lead to multi-organ failure if untreated.

## **Neurological Infections or Encephalopathy**

## Infections of the central nervous system such as meningitis or encephalitis, or metabolic/toxic disturbances causing brain dysfunction (encephalopathy), presenting with altered mental status, seizures, or focal neurological deficits.

## **Dehydration**

## A state of excessive loss of body water, with or without electrolyte imbalance, leading to hypovolemia and impaired organ perfusion. Clinical signs include dry mucous membranes, hypotension, tachycardia, and altered consciousness.

## **Adrenal Crisis**

## An acute, life-threatening condition due to insufficient cortisol production, often precipitated by stress or infection in patients with adrenal insufficiency. Symptoms include hypotension, shock, abdominal pain, vomiting, and electrolyte abnormalities.

## **Alcoholism**

## Chronic excessive alcohol consumption leading to multisystem effects including liver disease, cardiomyopathy, neuropathy, and increased infection risk.

## **Cardiogenic Pulmonary Edema (CPE**)

## Pulmonary edema caused by elevated hydrostatic pressure in pulmonary capillaries secondary to cardiac dysfunction, particularly left ventricular failure. It results in accumulation of low-protein fluid

## **Cirrhosis**

A chronic liver disease characterized by fibrosis and architectural distortion of the liver parenchyma, leading to portal hypertension, liver dysfunction, and complications such as ascites, variceal bleeding, and hepatic encephalopathy.

**Hypothyroidism**

A condition of deficient thyroid hormone production causing generalized slowing of metabolism. Clinical features include fatigue, weight gain, cold intolerance, bradycardia, constipation, dry skin, and cognitive slowing.

**Hypoalbuminemia**

A low serum albumin level indicating poor nutritional status, liver dysfunction, or protein loss (e.g., nephrotic syndrome). It reduces plasma oncotic pressure, potentially contributing to edema and fluid shifts.

**Metabolic Alkalosis**

A disturbance characterized by elevated blood pH due to increased bicarbonate or loss of hydrogen ions. Causes include vomiting, diuretic use, and mineralocorticoid excess. It can impair oxygen delivery and cause neuromuscular irritability.

## **Epidemiology**

Electrolyte imbalance is usually found in people working in stressful conditions like soldier’s training, wars, and patients admitted in the ICU. The US armed forces reported 7.2 cases per 100,000 person-years from 2003 through 2018 of hyponatremia. According to a review by Tsipotis et alo, 21% of the patients had community-acquired hypernatremia, while 25.9% of patients had hospital-acquired hypernatremia. Calcium imbalances are found mostly in patients with breast cancer, lung cancer, and multiple myeloma due to bone destruction. Hypokalaemia frequency is 1% in the US, while hyperkalaemia is found rarely in the general population and around 1-10% in hospitalized patients. It is more common in people with muscle destruction like military recruits, sickle cell patients, drug abusers.

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**Renal Artery Stenosis**

Other names: **RAS**

## **Definition and Description**

**Renal artery stenosis** is a narrowing of **arteries** that carry **blood** to one or both kidneys. Most often seen in older people with **atherosclerosis** (hardening of the arteries), renal artery stenosis can worsen over time and often leads to **hypertension** (**high blood pressure**) and kidney damage. The body senses less **blood** reaching the **kidneys** and misinterprets that as the body having **low blood pressure**. This signals the release of hormones from the kidney that lead to an increase in **blood pressure**. Over time, renal artery stenosis can lead to **kidney failure**.

### **What’s the link between renal artery stenosis and peripheral artery disease?**

Renal artery stenosis affects the **renal arteries** that carry blood to your **kidneys**. **Peripheral artery disease (PAD)** affects the arteries that carry blood from your heart to your arms and legs. Atherosclerosis is usually the cause of both. Studies show that between 14% to 35% of people with Peripheral artery disease also have Renal artery stenosis.

Research also suggests that chronic kidney disease, which can result from RAS, may put people at a higher risk for PAD. People with both chronic kidney disease and PAD are at an increased risk of Myocardial infarction, **stroke**, limb loss and other serious health complications.

## **Causes and Risk Factor**

### **Causes**

The two main causes of renal artery stenosis include:

* **Buildup on kidney (renal) arteries.** Fats, cholesterol and other substances (plaque) can build up in and on your kidney, artery walls (atherosclerosis). As these deposits get larger, they can harden, reduce blood flow, cause kidney scarring and eventually narrow the artery. Atherosclerosis occurs in many areas of the body and is the most common cause of renal artery stenosis.
* **Fibromuscular dysplasia.** In fibromuscular dysplasia, the muscle in the artery wall doesn't grow as it should. This often begins in childhood. The renal artery can have narrow sections alternating with wider sections, giving a bead-like appearance in images of the artery.

The renal artery can narrow so much that the kidney doesn't get enough blood. This can lead to high blood pressure at a young age. This can happen in one or both kidneys. Experts don't know what causes fibromuscular dysplasia, but the condition is more common in women and may be something that's present at birth (congenital).

Narrowed kidney arteries and fibromuscular dysplasia can affect other arteries in your body as well as your kidney arteries and cause complications.

Rarely, renal artery stenosis results from other conditions such as inflammation of the blood vessels or a growth that develops in your abdomen and presses on your kidneys' arteries.

Between 60% and 90% of RAS cases result from atherosclerosis. Fibromuscular dysplasia usually causes the remaining cases. Fibromuscular dysplasia occurs when there’s abnormal cell growth on artery walls, causing the arteries to narrow. It’s much more common in females — and it may result from genetics or hormones.

### **Risk Factors**

Most cases of renal artery stenosis result from narrowed kidney arteries. Risk factors that make narrowed arteries more likely in your kidneys and other parts of your body include:

* Aging
* Being Female
* High blood pressure
* High cholesterol
* Diabetes
* Obesity
* Smoking and other tobacco use
* A family history of early heart disease
* Lack of exercise

## **Signs and Symptoms**

Renal artery stenosis often doesn't cause any signs or symptoms until it's advanced. The condition may be discovered incidentally during testing for something else. Your health care provider may also suspect a problem if you have:

* High blood pressure that begins suddenly or worsens without explanation
* High blood pressure that begins before age 30 or after age 50

As renal artery stenosis progresses, other signs and symptoms may include:

* High blood pressure that's hard to control
* A whooshing sound as blood flows through a narrowed vessel (bruit), which your doctor hears through a stethoscope placed over your kidneys
* Elevated protein levels in the urine or other signs of a problem with kidney function
* Worsening kidney function during treatment for high blood pressure
* Fluid overload and swelling in your body's tissues
* Treatment-resistant heart failure

Symptoms of poor kidney function may include:

* Concentration problems or confusion.
* Difficulty sleeping.
* Edema (swelling due to fluid buildup).
* Fatigue.
* Headaches.
* Loss of appetite.
* Muscle cramps.
* Nausea and vomiting.
* Shortness of breath (dyspnea).
* Skin changes, such as dry, itchy or darkened skin.
* Unexplained weight loss.
* Urinating often.

## **Diagnosis Methods**

Sometimes, healthcare providers detect and diagnose renal artery stenosis incidentally. This means it happens during the process of diagnosing or treating another disease. If your healthcare provider suspects RAS, they may perform a variety of tests:

* **Physical exam:** Your provider checks your blood pressure, looks for swelling in your limbs and listens to your breathing. They may put a stethoscope near your kidneys to listen as blood flows through your arteries. Blood flowing through narrowed arteries often makes a whooshing sound. Unexplained high blood pressure is one of the most common indicators of RAS.
* **Kidney function tests:** Blood and urine tests, called kidney function tests, can tell how well your kidneys are working. Increased levels of protein, creatinine, nitrogen and other waste products in your body fluids indicate that your kidneys aren’t filtering waste from your blood efficiently.
* **Imaging scans:** There are a variety of imaging scans that can evaluate the size of your kidneys and how well blood is getting to your kidneys. Your provider may perform a renal scan, duplex ultrasound, computerized tomographic angiography (CTA) or magnetic resonance angiogram (MRA).

Imaging tests commonly done to diagnose renal artery stenosis include:

* Doppler ultrasound: High-frequency sound waves help your doctor see the arteries and kidneys and check their function. This procedure also helps your doctor find blockages in the blood vessels and measure their severity.
* CT scan. During a CT scan, an X-ray machine linked to a computer creates a detailed image that shows cross-sectional images of the renal arteries. You may receive a dye injection to show blood flow.
* Magnetic resonance angiography (MRA). MRA uses radio waves and strong magnetic fields to produce detailed 3D images of the renal arteries and kidneys. A dye injection into the arteries outlines blood vessels during imaging.
* Renal arteriography. This special type of X-ray exam helps your doctor find the blockage in the renal arteries and sometimes open the narrowed part with a balloon or stent. Before an X-ray is taken, your doctor injects a dye into the renal arteries through a long, thin tube (catheter) to outline the arteries and show blood flow more clearly. This test is mainly done if it's also likely that you need a small tube (stent) placed in your blood vessel to widen it.

## **Treatment options**

The most common treatments for RAS are lifestyle changes and medication. Surgery may be an option if someone develops severe stenosis in their renal arteries, is at risk of arterial occlusion (blockage), has unmanaged high blood pressure resistant to medications or has progressive loss of kidney function.

Your healthcare provider will likely recommend a variety of lifestyle changes to help manage RAS and lower high blood pressure, including:

* Eating a healthy diet low in fat, cholesterol, sodium and sugar.
* Getting regular exercise.
* Losing weight.
* Quitting smoking.

**How can medication help me manage renal artery stenosis?**

Medication, combined with lifestyle changes, can help regulate high blood pressure and slow or prevent the progression of kidney disease. Your healthcare provider may recommend a combination of drugs:

* **Angiotensin-converting enzyme (ACE) inhibitors and angiotensin receptor blockers (ARBs)** block certain hormones that may cause your blood vessels to narrow.
* **Aspirin** thins your blood, so it flows more easily through arteries.
* **Beta-blockers and calcium channel blockers** reduce high blood pressure.
* **Diuretics,** or “water pills,” help your kidneys remove extra water from your blood.
* **Statins** reduce high cholesterol.

**What surgeries are available for renal artery stenosis?**

Severe RAS not managed by medication may require surgery. The two main types of vascular surgeries for RAS include:

* **Angioplasty and stenting:** A surgeon inserts a catheter (thin, flexible tube) into your narrowed renal artery. They inflate a tiny balloon inside of your artery to widen it. Next, they place a stent (small mesh tube) in your artery to keep it open and help blood continue flowing to your kidney. Angioplasty and renal artery stenting is a minimally invasive procedure and is the most common type of surgery for RAS.
* **Renal artery bypass:** A surgeon reroutes blood flow to your kidney by bypassing the narrowed or blocked renal artery. They use a blood vessel graft (usually from your leg) or an artificial tube. The graft or tube is sewn to an existing healthy artery and then attached to the blocked artery (at a place between the blockage and the kidney).
* **Renal endarterectomy:** A surgeon opens your narrowed renal artery and removes plaque and other substances that may be blocking blood flow.

The condition may require three or more different drugs to control high blood pressure. Patients may also be asked to take other medications, such as cholesterol-lowering drugs and aspirin.

For a small number of people, an intervention such as angioplasty, often with stenting or surgery, may be recommended. With angioplasty, a catheter is inserted into the body through a blood vessel and guided to the narrowed or blocked renal artery. A balloon on the catheter is then inflated to open up the inside of the artery. A stent can then be placed to keep the area open.

Surgery to bypass the narrowed or blocked portion of the artery and/or remove a non-functioning kidney may be needed for some patients. But this procedure is not often done.

If you're diagnosed with renal artery stenosis, it's important to discuss the risks of the different treatments with your doctor. The side effects of blood pressure medications may include dizziness, sexual problems, headache, and cough. Complications of angioplasty include bruising, bleeding, additional kidney damage, and the possibility that the arteries can close again.

## **Prevention Tips**

You can reduce your risk of RAS by:

* Eating a balanced diet.
* Exercising regularly.
* Managing your blood pressure.
* Maintaining a healthy body weight.
* Not smoking.

## **Prognosis**

Renal artery disease (RAS) is a progressive condition, worsening over time. The outlook for someone with RAS depends largely on the severity of the disease. Research shows that the four-year survival rate for people with renal artery blockage of 95% or more is only 48%. So, only about half of people with near-total artery occlusion survive for four years after diagnosis.

The consequences of renal artery stenosis are hypertension, which may be particularly difficult to control or may require multiple antihypertensive agents (with increased adverse effects), and progressive loss of kidney function (ischemic nephropathy).

In addition, the discovery of atherosclerotic renovascular disease (RVD) frequently occurs in the setting of generalized vascular disease (i.e., cerebral, cardiac, peripheral), with the clinical consequences of disease in those vascular beds. Thus, any therapeutic intervention for renal artery stenosis should logically consider the underlying prognosis associated with these comorbidities.

Researchers have studied the natural history of atherosclerotic renal artery stenosis by obtaining images from sequential abdominal aortography or duplex ultrasound scans in patients with documented renal artery lesions who have been treated medically. Most studies show that progressive arterial obstruction occurs in 42-53% of patients with atherosclerotic renal artery stenosis, often within the first 2 years of radiographic follow-up. The incidence rate of progression to complete renal artery occlusion in these studies ranges from 9-16%; this often occurs in patients with a high-degree stenosis. In a study of 85 patients at the Cleveland Clinic who were followed for 3-172 months, patients with mild-to-moderate stenosis remained unchanged on follow-up, and 39% of patients with greater than 75% lesions progressed to total occlusion.

## **Possible Complication**

Potential complications of RAS include:

* Chronic kidney disease.
* Coronary artery disease.
* Kidney atrophy (reduced kidney size).
* Peripheral artery disease (PAD).
* Renal hypertension (high blood pressure in your renal arteries).
* High blood pressure
* Kidney failure, requiring treatment with dialysis or a kidney transplant
* Fluid retention in your legs, causing swollen ankles or feet
* Shortness of breath due to a sudden buildup of fluid in the lungs

## **When to see a Doctor/ Red Flag**

Make an appointment with your doctor if you have any persistent signs or symptoms that worry you.

**DIFFERENTIAL DIAGNOSIS**

* Acute kidney injury
* Azotemia
* Chronic Glomerulonephritis
* Hypersensitivity Nephropathy
* Hypertension
* Malignant Hypertension
* Nephrosclerosis
* Renovascular hypertension
* Uremia

## Acute Kidney Injury (AKI)

A sudden decline in kidney function occurring over hours to days, resulting in impaired waste elimination, fluid and electrolyte imbalance, and accumulation of nitrogenous waste products (azotemia). AKI may present with reduced urine output (oliguria), fluid overload, electrolyte disturbances (e.g., hyperkalemia), metabolic acidosis, and symptoms such as fatigue, nausea, and confusion. Causes are classified as:

* Prerenal: Due to decreased renal perfusion (e.g., dehydration, heart failure, sepsis).
* Intrinsic renal: Due to direct kidney damage (e.g., acute tubular necrosis, glomerulonephritis, nephrotoxic drugs).
* Postrenal: Due to urinary tract obstruction (e.g., stones, enlarged prostate).  
  Diagnosis involves serum creatinine measurement, urine output monitoring, urine studies, imaging, and sometimes biopsy. Treatment targets the underlying cause, fluid and electrolyte management, and may require dialysis in severe cases

**Azotemia**

An elevation of blood urea nitrogen (BUN) and serum creatinine due to impaired renal excretion of nitrogenous waste. It is a laboratory finding indicative of renal dysfunction and can be prerenal, renal, or postrenal in origin.

**Chronic Glomerulonephritis**

A progressive inflammatory disease of the glomeruli leading to chronic kidney damage and scarring. It manifests with proteinuria, hematuria, hypertension, and gradual loss of renal function, potentially progressing to end-stage renal disease.

**Hypersensitivity Nephropathy**

An immune-mediated interstitial nephritis often triggered by drugs or infections, characterized by inflammation of the renal interstitium and tubules. Clinical features include fever, rash, eosinophilia, and renal impairment.

**Hypertension**

A common condition of persistently elevated blood pressure that can cause renal damage (hypertensive nephropathy) and contribute to progression of chronic kidney disease.

**Malignant Hypertension**

A severe form of hypertension with rapid onset and marked elevation of blood pressure associated with vascular damage, including fibrinoid necrosis and hyperplastic arteriolosclerosis, leading to acute kidney injury and end-organ damage.

**Nephrosclerosis**

Hardening and sclerosis of small renal arteries and arterioles due to chronic hypertension or aging, causing ischemic glomerular and tubular injury and gradual decline in renal function.

**Renovascular Hypertension**

Hypertension caused by narrowing of renal arteries (e.g., atherosclerosis, fibromuscular dysplasia), leading to renal ischemia, activation of the renin-angiotensin system, and secondary hypertension.

**Uremia**

A clinical syndrome resulting from accumulation of urea and other nitrogenous waste products due to severe renal failure. It causes systemic symptoms including fatigue, anorexia, nausea, vomiting, neurological disturbances, pericarditis, and bleeding diathesis.

## **Epidemiology**

In patients with mild hypertension, the prevalence of renal artery stenosis is probably less than 1%, while in those with acute as high as 10 % to 40% in patients with acute, severe, or refractory hypertension, the prevalence may be as high as 10-40%. Studies suggest that ischemic nephropathy may be responsible for 5-22% of advanced kidney disease in all patients older than 50 years.

A review of a random sample of Medicare claims data (patients 67 years of age and older) found that the incidence of atherosclerotic renovascular disease was 3.7 per 1000 patient-years. The prevalence decreased with advancing age; the adjusted odds ratio (OR) was 0.86 for patients aged 75 to 84 years and 0.44 for those aged ≥ 85 years, compared with those aged 67 to 74 years. The prevalence was highest in whites (adjusted OR for Blacks, 0.66).

RVD is less common in Blacks than in Whites. The incidence rate in two studies of patients with severe hypertension was 27-45% in Whites versus 8-19% in Blacks.

Although the incidence of atherosclerotic RVD is independent of sex, Crowley et al showed that female sex (as well as older age, elevated serum creatinine level, coronary artery disease, peripheral vascular disease, hypertension, and cerebrovascular disease) is an independent predictor of RVD progression.

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**Hypertensive Nephrosclerosis**

Other names: hypertensive kidney disease, hypertensive nephropathy (HN), nephroangiosclerosis

**Definition and Description**

The term hypertensive nephrosclerosis has traditionally been used to describe a clinical syndrome characterized by long-term essential hypertension, hypertensive retinopathy, left ventricular hypertrophy, minimal proteinuria, and progressive kidney failure. Most cases are diagnosed based solely on clinical findings. In fact, most of the literature dedicated to hypertensive nephrosclerosis assumes that progressive kidney failure in a patient with long-standing hypertension, moderate proteinuria, and no evidence suggesting an alternative diagnosis characterizes hypertensive nephrosclerosis.

The lack of firm criteria on which to base a histologic diagnosis and the lack of a clear demonstration that hypertension initiates the development of kidney failure likely indicate that the true prevalence of hypertensive nephrosclerosis has been overestimated. The paradoxical results of increasing incidence of kidney failure despite wider antihypertensive drug therapy and reduction in hypertensive target events, such as stroke and cardiovascular disease, raises questions about the causal role of hypertension in this disorder. Indeed, Carriazo et al suggest that hypertensive nephrosclerosis as a cause of end-stage renal disease (ESRD) may not exist at all.

As reported by Zuccalà and Zucchelli (1996), part of the confusion in the classification of hypertensive nephrosclerosis stems from the use of the word nephrosclerosis.Coined almost a century ago by Theodor Fahr, nephrosclerosis literally means "hardening of the kidney." In the United States and Europe, the terms hypertensive nephrosclerosis, benign nephrosclerosis, and nephroangiosclerosis are commonly used to describe the same clinical condition.

## **Cause and Risk Factors**

### **Causes**

Hypertensive arteriolar nephrosclerosis is progressive kidney damage caused by long-standing, poorly controlled high blood pressure (hypertension).

Chronic high blood pressure puts great strain on blood vessels. There are greater blood flows in the kidney and heart than other organs. Over time, renal vessels would suffer damage---vessels begin to thicken and harden. The narrowing vessels couldn’t provide enough oxygen to tissues (ischemia) and tissues start to die from ischemia. Not only do the kidneys have trouble filtering blood quickly enough because of the reduced flow, but their function also declines because of the tissue death. Owing to kidney damage, some metabolic waste can’t excrete in daytime, so patients will have nocturia to discharge waste. With aggravation of condition, proteinuria follows on.

Hypertensive" refers to high blood pressure and "nephropathy" means damage to the kidney; hence this condition is where chronic high blood pressure causes damage to kidney tissue; this includes the small blood vessels, glomeruli, kidney tubules and interstitial tissues. The tissue hardens and thickens which is known as nephrosclerosis. The narrowing of the blood vessels means less blood is going to the tissue and so less oxygen is reaching the tissue resulting in tissue death (ischemia).

Risk factors for HN include poorly controlled, moderate-to-severe hypertension, older age, other kidney disorders, and Afro-Caribbean background, whose exact cause is unclear, as it may be due to either genetic susceptibility or poor health management among people of Afro-Caribbean descent.

### **Risk Factor**

* Older age
* Poorly controlled high blood pressure
* Presence of other kidney disorders (for example, diabetic nephropathy)

Black people are at increased risk, but it is unclear if the risk is increased because poorly treated high blood pressure is more common among them or because they are more genetically susceptible to kidney damage caused by high blood pressure.

**Signs and symptoms**

loss of appetite, nausea, vomiting, itching, sleepiness or confusion, weight loss, and an unpleasant taste in the mouth, may develop.

## **Diagnosis Methods**

### **Urine test**

Microalbuminuria (moderate increase in the levels of urinary albumin) is a non-specific finding in patients with vascular disease that is associated with increased risk of cardiovascular events. The majority of patients with benign nephrosclerosis have proteinuria in the range from 0.5 to 1 g/ 24hr. In the case of glomerular damage occurring in HN, hematuria can occur as well.

### **Definitive diagnosis**

The definitive diagnosis of HN requires morphological examination. Common histological features can be identified in the renal and glomerular vasculature. Glomerulosclerosis is often present, either locally or globally, which is characterized by hardening of the vessel walls. Also, luminal narrowing of the arteries and arterioles of the kidney system. However, this type of procedure is likely to be preceded by a provisional diagnosis based on laboratory investigations.

### **Future diagnostic approaches**

Increasing access to, and use of, genome profiling may provide opportunity for diagnosis based on presentation and genetic risk factors, by identifying ApoL1 gene variants on chromosome 22.

Upon physical examination, evidence of hypertension-related target organ damage includes hypertensive changes in the retinal vessels and signs of left ventricular hypertrophy.

On ophthalmologic examination, hemorrhages or exudates are characteristic of accelerated hypertension, and papilledema is a feature of malignant hypertension.

## **Treatment Options**

In addition to kidney disease, Hypertensive Nephropathy patients also suffer other health problems, because high blood pressure is also hard on the heart and lung. Such patients may benefit from measures to reduce blood pressure, including diet and exercise as well as medications to force the pressure down.

### **ACEI and ARBs**

ACEI and ARBs are recommended to Hypertensive Nephropathy patients, as these medicines can lower the blood pressure, and meanwhile they can reduce protein urine to protect kidney function. What’s more, these medicines are also beneficial to vessels in the heart, reducing the risk of related disease.

Several antihypertensive medications, including thiazide diuretics, beta-blockers, angiotensin-converting enzyme (ACE) inhibitors, angiotensin receptor blockers (ARBs), and calcium channel blockers, in principle, can be used as initial monotherapy in patients with hypertension. However, controversy regarding which drug to use as a first-line therapy continues partly due to the belief that antihypertensive medications have benefits beyond lowering of blood pressure (BP).

### **Dialysis or kidney transplantation**

Hypertensive Nephropathy may progress to the point where the kidneys are no longer able to function well enough to support the lives of the patients. In these situations, dialysis or kidney transplantation may become necessary. Nephrologists could help patients choose the best treatment options considering the patient's own condition and side effects of treatment.

## **Preventive Tips**

For hypertension complicating primary kidney disease, considerations for prevention include the following:

* Systemic hypertension clearly induces or accelerates the progression of kidney disease in experimental models. In these models, blood pressure (BP) control reduces proteinuria and prevents deterioration of kidney function.
* Similarly, in a variety of primary human kidney diseases, hypertension strongly predicts a faster decline in the glomerular filtration rate.
* As demonstrated by the MDRD study, even small differences in mean arterial pressure between the usual BP control group and the low-BP group had significant effects in reducing kidney disease progression.

## **Prognosis**

Regarding the target BP, the Working Group Report on Hypertension and Diabetes recommended a BP goal of less than 130/80 mm Hg to preserve kidney function and to reduce cardiovascular events in patients with hypertension and diabetes. Lower BPs are recommended for patients with proteinuria greater than 1 g/d and kidney insufficiency, regardless of etiology. The optimal BP goal to slow the progression of kidney failure in patients with hypertensive nephrosclerosis currently is unknown.

Hypertensive nephrosclerosis remains a poorly defined entity. Researchers continue to search for a clear definition, a pathophysiologic mechanism, and optimal treatment for patients with this condition. As suggested by Meyrier (1996), hypertensive nephrosclerosis may conceivably be a primary microvascular nephropathy.

Uncontrolled hypertension can accelerate the decline of kidney function in patients with primary renal disease; however, whether mild-to-moderate essential hypertension can cause ESRD in white people is uncertain. The available data do not support the hypothesis that high BP is the only factor determining ESRD in these patients.

Medical treatment is indicated in patients younger than 80 years with BP higher than 140/90 mm Hg. In these patients, antihypertensive treatment has proven to reduce the risk of stroke and cardiovascular mortality. Data from HYVET showed decreased strokes, heart failure, and all-cause mortality from the treatment of patients older than 80 years with BP less than 160 mm Hg.

## **Possible Complication**

Traditionally, nephrosclerosis was considered the consequence of long-term hypertension. This premise is based on observations of rapidly progressive kidney failure developing in some patients with malignant hypertension. Such individuals demonstrate arterial and necrotizing lesions in the kidneys, which may be reversed with effective BP control. However, less severe hypertension, per se, is suggested to cause kidney failure only rarely, and progressive kidney impairment is usually secondary to undiagnosed primary kidney disease.

Madhavan et al (1995) followed the cases of 2125 men with mild-to-moderate hypertension for 5 years and found no change in serum creatinine values.Similarly, Tomson et al (1991) followed the cases of 176 patients with essential hypertension for more than 14 years and found no change in serum creatinine values, with none of the patients developing kidney failure.

In the Baltimore Longitudinal Study on Aging, the cases of 446 patients who are predominantly white and of middle or upper socioeconomic status were followed over a 13-year period. In this study, patients with hypertension had a decline in their glomerular filtration rates (GFRs) at a faster rate than normotensive subjects (0.92 mL/min/y vs 0.75 mL/min/y). Although this study showed that patients with hypertension lost kidney function at a faster rate with aging than normotensive subjects, the rate of decline in kidney function was small and unlikely to result in end-stage renal disease (ESRD). More importantly, this study failed to determine whether the decline in kidney function was secondary to essential hypertension or was the result of undiagnosed primary kidney disease.

## **Differential Diagnosis**

Other problems to consider in the differential diagnosis of nephrosclerosis include the following:

* Renal atherosclerotic disease
* Cholesterol microembolization
* Malignant hypertension
* Mildly active primary kidney disease
* Lead nephropathy

**Renal Atherosclerotic Disease**

A condition characterized by atherosclerotic plaque buildup in the renal arteries, leading to narrowing (renal artery stenosis) and reduced renal blood flow. It is the most common cause of renovascular hypertension and can result in chronic kidney disease due to ischemic injury. Symptoms often include resistant hypertension, decline in kidney function, and occasionally flash pulmonary edema. Diagnosis is by imaging such as Doppler ultrasound, CT angiography, or MR angiography. Treatment includes lifestyle modification, blood pressure control, and in some cases revascularization via angioplasty or surgery.

**Cholesterol Microembolization**

A complication of atherosclerosis where cholesterol crystals dislodge from atherosclerotic plaques and embolize to small arteries, including renal arterioles. This causes ischemic injury and inflammation, leading to acute or subacute kidney injury, livedo reticularis, and other systemic signs. It may occur spontaneously or after vascular procedures.

**Malignant Hypertension**

A severe form of hypertension characterized by rapid and extreme elevation of blood pressure leading to vascular injury including fibrinoid necrosis and hyperplastic arteriolosclerosis in renal arterioles. It causes acute kidney injury, encephalopathy, and retinal hemorrhages. Urgent blood pressure control is essential.

**Mildly Active Primary Kidney Disease**

Refers to kidney diseases with ongoing but low-grade inflammation or injury, often presenting with mild proteinuria or hematuria and relatively preserved renal function. Examples include early or stable glomerulonephritis. Close monitoring and appropriate treatment are required to prevent progression.

**Lead Nephropathy**

Chronic kidney disease caused by prolonged lead exposure leading to tubular injury, interstitial fibrosis, and nephrosclerosis. It manifests as reduced renal function, hypertension, and sometimes gout. Diagnosis is supported by elevated blood lead levels and history of exposure. Management includes removal from exposure and chelation therapy.

## **Epidemiology**

The incidence of hypertensive nephropathy varies around the world. For instance, it accounts for as many as 25% and 17% of patients starting dialysis for end-stage kidney disease in Italy and France respectively. Contrastingly, Japan and China report only 6 and 7% respectively. Since the year 2000, nephropathy caused by hypertension has increased in incidence by 8.7%, these figures may be even higher, as hypertension is not always reported as the specific cause of kidney disease.

It has been recognized that the incidence of hypertensive nephropathy varies with ethnicity. Compared to Caucasians, African Americans in the USA are much more likely to develop hypertensive nephropathy. Of those who do, the proportion who then go on to develop end-stage kidney failure is 3.5 times higher than in the Caucasian population. In addition to this, African Americans tend to develop hypertensive nephropathy at a younger age than Caucasians (45 to 65, compared to >65).

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**Polycystic Kidney Disease**

**Definition and description**

Polycystic kidney disease (PKD) is a condition in which clusters of cysts grow in the body, mainly in the kidneys. Over time, the cysts may cause the kidneys to get bigger and stop working. PKD is most often passed through families. This is called an inherited condition.

Cysts are round sacs with fluid in them. They are not cancerous. In PKD, the cysts vary in size. They can grow very large. Having many cysts or large cysts can damage the kidneys.

Polycystic kidney disease also can cause cysts to grow in the liver, the pancreas and other places in the body. The disease can cause serious complications, including high blood pressure and kidney failure.

PKD varies greatly in how bad it is. It's possible to prevent some complications. Lifestyle changes and treatments might help reduce damage to the kidneys.

**Causes**

**Genes**

Gene changes cause polycystic kidney disease. Most often, the condition runs in families. Sometimes, a gene change happens on its own in a child. This is known as a spontaneous gene change. Then neither parent has a copy of the changed gene.

There are two main types of polycystic kidney disease. They're caused by different gene changes. The two types of PKD are:

* **Autosomal dominant polycystic kidney disease (ADPKD).** This is the most common type of ongoing kidney disease that's passed through families, also called inherited. Symptoms of ADPKD often start between the ages of 30 and 40.

Only one parent needs to have the condition to pass it to the children. If one parent has ADPKD, each child has a 50% chance of getting the condition. This is the more common type of polycystic kidney disease.

* **Autosomal recessive polycystic kidney disease (ARPKD).** This type is far less common than is ADPKD. The symptoms often appear soon after birth. Sometimes, symptoms don't appear until later in childhood or during the teen years.

Both parents must have gene changes to pass on this form of the condition. If both parents carry a changed gene, each child has a 25% chance of getting the condition.

**Risk factors**

The biggest risk factor for getting polycystic kidney disease is getting the gene changes that cause the disease from one or both parents.

**Signs & Symptoms**

* High blood pressure.
* Belly, side or back pain.
* Blood in the urine.
* A feeling of fullness in the belly.
* Increased size of the belly from enlarged kidneys.
* Headaches.
* Kidney stones.
* Kidney failure.
* Urinary tract or kidney infections.

**Diagnosis methods**

For polycystic kidney disease, certain tests can detect the size and number of kidney cysts you have. Tests also can show how much healthy kidney tissue you have. Tests include:

* **MRI scan.** As you lie inside a large cylinder, magnetic fields and radio waves show views of your kidneys. This method most often is used to know how badly PKD affects the kidneys, liver or pancreas. MRI can help measure total kidney volume, which helps healthcare professionals know more about your condition.
* **Ultrasound.** This involves putting a wand-like device called a transducer on your body. It gives off sound waves that go back to the transducer. A computer turns the sound waves into images of your kidneys.
* **CT scan.** You lie on a table that goes into a big, doughnut-shaped device. The device uses X-ray beams to show images of your kidneys.

**Prevention tips**

If you have polycystic kidney disease and you're thinking about having children, a genetic counsellor can help you know your risk of passing the disease to your children.

Keeping your kidneys as healthy as possible may help prevent some of the complications of this disease. It's most important to manage your blood pressure.

manage your blood pressure using the following guidelines

* Take the blood pressure medicines your healthcare professional prescribes as directed.
* Eat a low-salt diet that has plenty of fruits, vegetables and whole grains.
* Get to and stay at a healthy weight.
* Exercise regularly. Aim for at least 30 minutes of moderate physical activity most days of the week.
* Limit alcohol use.
* Don't smoke.

**Treatment options**

How bad polycystic kidney disease varies from person to person. That's true even among people in the same family. Often, people with PKD reach end-stage kidney disease between ages 55 and 65. But some people with PKD have mild diseases. They might never get end-stage kidney disease.

Treating polycystic kidney disease involves dealing with the following symptoms and complications in their early stages:

* **Kidney cyst growth.** The medicine tolvaptan (Jynarque, Samsca) may be used for adults at risk of ADPKD that's getting worse fast. Tolvaptan is a pill that you swallow that slows how fast kidney cysts grow. It also slows the decline in how well your kidneys work.

Tolvaptan carries a risk of serious liver injury. And it can interact with other medicines you take. It's best to see a specialist in kidney health, called a nephrologist. A nephrologist can watch for side effects and possible complications of the medicine.

* **High blood pressure.** Keeping high blood pressure under control can slow the disease and kidney damage. Eating a low-sodium, low-fat diet that's moderate in protein and calories and drinking more fluids may help control blood pressure.

Other helpful lifestyle changes include not smoking, moving more and easing stress. Smoking can greatly harm the kidneys. It also can speed up the start of kidney failure.

Medicines most often are needed to control high blood pressure. Medicines called angiotensin-converting enzyme (ACE) inhibitors or angiotensin II receptor blockers (ARBs) are often used to control high blood pressure.

* **Loss of kidney function.** To help your kidneys stay as healthy as possible for as long as possible, experts suggest being at a healthy weight and body mass index. Drinking water and fluids throughout the day may help slow the growth of kidney cysts. This could slow the loss of kidney function. Eating a low-salt diet with less protein might let kidney cysts respond better to more fluids.
* **Pain.** You might be able to control the pain of polycystic kidney disease with medicines available without a prescription, such as acetaminophen (Tylenol, others). Don't take nonsteroidal anti-inflammatory medicines such as ibuprofen (Advil, Motrin IB, others) or naproxen sodium (Aleve). Long-term use of nonsteroidal anti-inflammatory medicines can affect how your kidneys work.

For worse pain, a healthcare professional might use a needle to draw out cyst fluid and put in a medicine to shrink kidney cysts. The medicine is called a sclerosing agent.

Or you may need surgery to remove cysts if they're large enough to cause pressure and pain. The surgery is called cyst fenestration.

* **Bladder or kidney infections.** Treating infections quickly with antibiotics can help prevent kidney damage. You might have a simple bladder infection or a more complicated cyst or kidney infection. For more-complicated infections, you may need to take antibiotics longer.
* **Blood in the urine.** Drink lots of fluids as soon as you notice blood in your urine. It's best to drink water to dilute the urine. This might help prevent clots from forming in your urinary tract.

Most often, the bleeding stops on its own. If it doesn't, call your healthcare professional.

* **Kidney failure.** Your kidneys can stop removing waste and extra fluids from your blood. Then you need either dialysis or a kidney transplant. That's why you need to see your healthcare team regularly.

You may be able to have a kidney transplant before your kidneys fail. Then you wouldn't need to have dialysis. This is called preemptive kidney transplantation.

* **Aneurysms.** If you have polycystic kidney disease and a family history of brain aneurysms that burst, your healthcare team may want to do regular screenings for brain aneurysms.

If you have an aneurysm, surgical clipping might reduce the risk of bleeding. This depends on the size of the aneurysm. Nonsurgical treatment of small aneurysms may involve controlling high blood pressure and high blood cholesterol, as well as quitting smoking.

Early treatment offers the best chance of slowing the progress of polycystic kidney disease.

**Prognosis**

The outlook for Autosomal dominant polycystic kidney disease (ADPKD)is highly variable. Some people experience kidney failure soon after the condition is diagnosed, whereas others may live the rest of their life with their kidneys working relatively well.

As well as kidney failure, ADPKD can also cause several other potentially serious problems, such as heart attacks and strokes caused by high blood pressure, or bleeding on the brain (subarachnoid haemorrhage) caused by a bulge in the wall of a blood vessel in the brain (brain aneurysm).

**Possible complications**

* **High blood pressure.** This is common in polycystic kidney disease. Not treated, high blood pressure can cause more damage to the kidneys and increase the risk of heart disease and strokes.
* **Loss of kidney function.** The kidneys' losing their ability to do their work is one of the most serious complications of polycystic kidney disease. Nearly half of people with the condition have kidney failure by age 60. But for some people, it starts in the early 30s.
* **Pain.** It's common to have pain with polycystic kidney disease. Pain often is in the side or back. The pain can come and go or be ongoing. The pain may be linked to bleeding into a cyst, a urinary tract infection, a kidney stone or, less often, cancer.
* **Cysts in the liver.** The older people with polycystic kidney disease get, the more likely it is they'll get cysts in the liver. With cysts, the liver most often keeps working.

Women tend to get larger cysts than do men. Hormones and pregnancies might be part of the reason.

* **Brain aneurysm.** A balloon-like bulge in a blood vessel, called an aneurysm, in the brain can cause bleeding if it bursts. People with polycystic kidney disease have a higher risk of aneurysms. People with a family history of aneurysms seem to be at highest risk.

Ask your healthcare professional if you need screening. If screening doesn't show an aneurysm, your healthcare professional may suggest screening again in a few years. The timing of repeat screening depends on your risk.

* **Pregnancy complications.** Most people with polycystic kidney disease can have success with pregnancy. But sometimes, they can get a life-threatening condition called preeclampsia during pregnancy. Those most at risk have high blood pressure or a loss of kidney function before they become pregnant.
* **Heart valve conditions.** As many as 1 in 4 adults with polycystic kidney disease gets mitral valve prolapse. When this happens, the heart valve no longer closes well. This lets blood leak backward.
* **Colon conditions.** People with polycystic kidney disease may get weaknesses and pouches or sacs called diverticula in the wall of the colon. This condition is called diverticulosis. Diverticula most often don't cause symptoms, but they may bleed or get infected.

**Differential diagnosis**

Renal cysts are common in adults. Finding renal cysts in an ultrasound in an asymptomatic subject raises several options for the diagnosis like simple cysts, ADPKD or acquired renal cysts. Careful history and physical examination, as well as other clinical findings such as hypertension, liver cysts or renal failure facilitate the differential diagnosis between these common entities. However, in certain circumstances, particularly with atypical clinical presentations the following entities should be excluded:

* **Multiple benign simple cysts:** Prevalence of simple renal cysts increases with age, and they are commonly detected with sensitive imaging methods such as computed tomography (CT) or magnetic resonance imaging (MRI). MRI-based series detect at least one renal cyst in 93% of subjects 45-59 years of age. Spiral CT detected simple renal cysts in 41% of 617 patients. Renal cysts are present in approximately 50% of men (mean age 66 years) and 35% of women (mean age 63 years)
* **Localised renal cystic disease:** Localised renal cystic disease is a rare and benign condition. Patients may present hypertension, flank pain, haematuria or flank mass. Clinical features distinguish this disease from ADPKD because of unilateral location, negative family history, no progression to chronic renal failure and no extrarenal involvement
* **Medullary sponge kidney:** Medullary sponge kidney is a cystic renal disease with unknown inheritance characterised by malformation of the distal collecting tubules with nephrolithiasis, impairment of renal function, tubular acidosis and recurrent urinary tract infections.
* **Bilateral parapelvic cysts***:* Parapelvic cysts are a subset of simple cysts that arise within the renal parenchyma adjacent to the renal sinus and account for 5% of all renal cysts in adults. Clinically they may manifest through obstruction of the ureter or renal pelvis
* **Von Hippel-Lindau syndrome:** Autosomal dominant-inherited von Hippel-Lindau (VHL) syndrome is characterised by a combination of hemangioblastomas (retina and cerebellum), renal cell cancers and, less frequently pancreatic, endocrine tumours and pheochromocytoma .In the early stage, precancerous renal cysts may occur, which result in enlargement of the kidneys and may be misdiagnosed as PKD. However, kidney failure is not a major feature in VHL syndrome.
* **Autosomal dominant medullary cystic disease:** Autosomal dominant medullary cystic disease may be present in adulthood (30–60 years) with renal dysfunction and, occasionally, renal cysts. The gene involved is uromodulin (encoding Tamm-Horsfall protein) on chromosome 16.
* **Orofaciodigital syndrome type I:** This is an X-linked disease due to mutation of the OFD1 gene, and craniofacial and digital defects are associated with polycystic kidney disease
* **Bardet-Biedl syndrome:** Bardet-Biedl syndrome is a very rare disease with incidence Polycystic kidney disease coexists with several extrarenal defects, such as vision loss due to retinal degeneration, childhood obesity, mental retardation, malformation of the urogenital tract and polydactyly (
* **Renal cysts and diabetes syndrome:** Type 5 MODY (Maturity Onset Diabetes of the Young) is a rare monogenic disease resulting from mutations in hepatocyte nuclear factor 1β (HNF-1β) that is associated with renal cysts. About 50% of this autosomal-dominant disease results from de novo mutations and presents with renal cysts or malformation in 90% of patients. It also presents with diabetes mellitus in 45% of patients, genital tract abnormalities and hyperuricemia in 20%, hypomagnesaemia in 40% and elevated liver enzymes in 15%

**PREVENTIVE TIPS**

## **Manage high blood pressure**

Managing blood pressure is one of the most critical things to focus on if you have PKD. Most people with PKD have high blood pressure, also called hypertension, that can cause damage to the kidneys and cardiovascular system. Known as the silent killer, high blood pressure makes arteries less elastic, puts strain on the heart, and increases the risk of heart attack and stroke.

Being aware of your blood pressure is vital to managing it well. You can check your blood pressure at home to help you hit the targets experts recommend. For those between 18 and 50 years of age who still have relatively healthy kidney function, the target is 110/75 millimetres of mercury (mm Hg). For those who are over 50, the target blood pressure is 120/80 mm Hg.

Diet and lifestyle are the two primary ways of controlling high blood pressure. Two commonly prescribed types of hypertension medication — known as angiotensin-converting enzyme (ACE) inhibitors and angiotensin 2 receptor blockers (ARBs) — also are frequently recommended for controlling high blood pressure in people with PKD.

## **Cut back on salt**

Salt intake can lead to increased growth of kidney cysts and high blood pressure. Limiting dietary salt to 2,300 mg a day can slow the progression of ADPKD and help prevent high blood pressure that can damage your heart and kidneys.

High Dietary salt, also called sodium, leads to high blood pressure. Salt also stimulates the body to produce a hormone called vasopressin. If you have PKD, vasopressin has the effect of stimulating the growth of kidney cysts. Researchers have found that increased salt consumption is associated with faster decline in kidney function in PKD.

When cutting back on salt, it’s natural to focus on using the saltshaker less. While this is an important step, most of the sodium in most diets is hidden inside processed and fast foods. Reading food labels carefully can help you understand which food products are high in sodium. Avoiding these products will likely reduce the amount of salt you consume.

## **Drink lots of fluid**

Consuming substantial amounts of water keeps your urine dilute so your body does not release vasopressin, a hormone that maintains the fluid balance in your body and stimulates the growth of kidney cysts in PKD. When urine is diluted, the body does not release as much vasopressin. Typically, people with PKD are advised to drink 3 to 4 quarts of water over the course of a day.

Your healthcare professional may run tests to measure how dilute your urine is. If you see that your urine is dark yellow or orange, that often means it is not dilute enough to suppress vasopressin. This likely means your fluid intake needs to increase unless your healthcare team has provided instructions about restricting your fluid intake.

[Request an Appointment](https://www.mayoclinic.org/appointments?mc_id=us&utm_source=mcppkd&utm_medium=l&utm_content=mcorgmcprst&utm_campaign=mayoclinic&geo=national&placementsite=enterprise&invsrc=other&cauid=191437)

**Maintain a healthy body weight**

PKD can be slowed by maintaining a healthy body weight through diet and exercise. The goal for a person with PKD is a body mass index (BMI) of 25 or lower. If you don’t know your BMI, Mayo Clinic has an online calculator that uses height and weight to produce an estimate.

A healthy body weight helps control high blood pressure, which can prevent or reduce damage to your kidneys and cardiovascular system.

In addition to helping to control body weight, reducing calorie intake if you are overweight also can slow the progression of PKD. Reducing calories cuts energy to cells that produce cysts. Researchers have found that people with ADPKD who are overweight tend to experience more-rapid decline of kidney function and greater kidney growth.

## **Eat a PKD-friendly diet**

Appropriate calorie intake — as well as low salt and moderate protein intake — are the cornerstones of a diet that can help manage PKD. If your kidney function is below relatively healthy levels, your healthcare team may advise you to reduce your protein consumption. Like salt, protein may stimulate the release of vasopressin, a hormone that can stimulate growth of cysts. For people with kidney function that’s below relatively healthy levels, experts recommend limiting protein consumption below 1.3 grams for every kilogram of weight.

## **If you smoke, stop**

Smoking makes PKD worse. It raises blood pressure and can increase damage to kidneys. If you smoke, ask your care team for recommendations on how you can successfully quit.

## **Watch for other effects**

PKD can cause several effects in the kidneys or other parts of the body that are good to be aware of, including:

### **Brain aneurysm.**

Learn about the risk of aneurysm, a potentially dangerous bulging of blood vessels in the brain that is more common among people with PKD. Call for immediate medical attention if an extremely severe headache comes on very quickly, such as within a minute. This can be a so-called thunderclap headache, a medical emergency that can signal the rupture of a brain aneurysm.

### **Low red blood cell count, also called anaemia.**

People with lower kidney function often have anemia. This might make you feel very tired. Iron supplements or special hormones called erythropoietin-stimulating agents that are taken under the supervision of your healthcare team can help remedy anemia.

### **Low phosphorus levels.**

Levels of phosphorus, an essential mineral, can decline when kidney function declines. This can be detected with blood tests and treated with vitamin supplements or medication.

### **Symptoms of low kidney function.**

Be aware of what symptoms could arise if your kidney function becomes very low. These are called uremic symptoms and could include fatigue, loss of appetite, mental fogginess, metallic taste in the mouth, buildup of fluids in the legs, called Edema, and fluid buildup in the lungs, which can lead to shortness of breath.

**Recent guideline**

* Nutrition

At present, no specific diet is known to prevent cysts from developing in patients with PKD. Reducing salt intake helps control blood pressure in patients with PKD who have high blood pressure. A diet low in fat and moderate in calories is recommended to maintain a healthy weight. Speak to your doctor or a dietitian about other changes to your diet, such as avoiding caffeine.

* Exercise

Physical exercise is recommended for people with PKD, however exercises that are potentially harmful to the kidney, such as contact sports, should be avoided. It is important not to become too dehydrated during any physical activity.

* Preparing for your appointment

In addition to your primary care doctor, you should also find a nephrologist who has experience treating patients with PKD. You and your healthcare professional can work together to choose treatment options that are best for you.

**Statistics**

* Ages 15 to 29 years: 2 or more cysts, unilateral or bilateral.
* Ages 30 to 59 years: 2 or more cysts in each kidney.
* Ages 60 years or older: 4 or more cysts in each kidney.
* Symptoms typically begin starting at age 30.
* Incidence of 1:400 - 1:1,000. (Relatively common.)
* End Stage Kidney Disease (ESKD) develops in 50% of patients who reach 20 years of age.

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**Glomerulonephritis**

**Definition and description**

Glomerulonephritis is inflammation of the tiny filters in the kidneys (glomeruli). The excess fluid and waste that glomeruli remove from the bloodstream exit the body as urine. Glomerulonephritis can come on suddenly (acute) or gradually (chronic).

Severe or prolonged inflammation associated with glomerulonephritis can damage the kidneys. Treatment depends on the type of glomerulonephritis you have.

**Causes and risk factors**

**Autoimmune diseases**

Autoimmune diseases are illnesses caused by the immune system attacking healthy tissues. Autoimmune diseases that may cause glomerulonephritis include:

* **Lupus.** A chronic inflammatory disease, systemic lupus erythematosus can affect many parts of your body, including your skin, joints, kidneys, blood cells, heart and lungs.
* **Goodpasture's syndrome.** In this rare disorder, also known as anti-GBM disease, the immune system creates antibodies to tissues in the lungs and kidneys. It can cause progressive and permanent damage to the kidneys.
* **IgA nephropathy.** Immunoglobulin A (IgA) is an antibody that's a first line of defence against infectious agents. IgA nephropathy occurs when deposits of the antibody accumulate in the glomeruli. The inflammation and subsequent damage may go undetected for a long time. The most common symptom is blood in the urine.

**Infections**

Infectious diseases can directly or indirectly lead to glomerulonephritis. These infections include:

* **Post-streptococcal glomerulonephritis.** Glomerulonephritis may develop a week or two after recovery from a strep throat infection or, rarely, a skin infection caused by streptococcal bacteria (impetigo). Inflammation occurs when antibodies to the bacteria build up in the glomeruli. Children are more likely to develop post-streptococcal glomerulonephritis than are adults, and they're also more likely to recover quickly.
* **Bacterial endocarditis.** Bacterial endocarditis is an infection of the inner lining of your heart's chambers and valves. It isn't clear whether the inflammation in the kidneys is the result of immune system activity alone or other factors.
* **Viral kidney infections.** Viral infections of the kidney, such as hepatitis B and hepatitis C, cause inflammation of the glomeruli and other kidney tissues.
* **HIV.** Infection with HIV, the virus that causes AIDS, can lead to glomerulonephritis and progressive kidney damage, even before the onset of AIDS.

### **Vasculitis**

Vasculitis is inflammation of blood vessels. Types of vasculitis that can cause glomerulonephritis include:

* **Polyarteritis.** This form of vasculitis affects medium and small blood vessels in many parts of your body, including the kidneys, skin, muscles, joints and digestive tract.
* **Granulomatosis with polyangiitis.** This form of vasculitis, formerly known as Wegener's granulomatosis, affects small and medium blood vessels in your lungs, upper airways and kidneys.

### **Sclerotic conditions**

Some diseases or conditions cause scarring of the glomeruli that results in poor and declining kidney function. These include:

* **High blood pressure.** Long-term, poorly managed high blood pressure can cause scarring and inflammation of the glomeruli. Glomerulonephritis inhibits the kidney's role in regulating blood pressure.
* **Diabetic kidney disease (diabetic nephropathy).** High blood sugar levels contribute to scarring of the glomeruli and increase the rate of blood flow through the nephrons.
* **Focal segmental glomerulosclerosis.** In this condition, scarring is scattered among some of the glomeruli. This may be the result of another disease, or it may occur for no known reason.

### **Other causes**

Infrequently, chronic glomerulonephritis runs in families. One inherited form, Alport syndrome, also might impair hearing or vision.

Glomerulonephritis is associated with certain cancers, such as gastric cancer, lung cancer and chronic lymphocytic leukemia.

**Signs & Symptoms**

Your first indication that something is wrong might come from the results of a routine urine test (urinalysis).

Glomerulonephritis signs and symptoms may include:

* Pink or cola-coloured urine from red blood cells in your urine (hematuria).
* Foamy or bubbly urine due to excess protein in the urine (proteinuria).
* High blood pressure (hypertension).
* Fluid retention (Edema) with swelling evident in your face, hands, feet and abdomen.
* Urinating less than usual.
* Nausea and vomiting.
* Muscle cramps.
* Fatigue.

**Possible complications**

Glomerulonephritis affects the ability of nephrons to filter the bloodstream efficiently. The breakdown in filtering results in:

* Accumulation of wastes or toxins in the bloodstream.
* Poor regulation of essential minerals and nutrients.
* Loss of red blood cells.
* Loss of blood proteins.

Possible complications of glomerulonephritis include:

* **Acute kidney failure.** Acute kidney failure is the sudden, rapid decline in kidney function, often associated with an infectious cause of glomerulonephritis. The accumulation of waste and fluids can be life-threatening if not treated promptly with an artificial filtering machine (dialysis). The kidneys often resume typical function after recovery.
* **Chronic kidney disease.** Persistent inflammation results in long-term damage and declining function of the kidneys. Chronic kidney disease is generally defined as kidney damage or decreased function for three or more months. Chronic kidney disease may advance to end-stage kidney disease, which requires either dialysis or a kidney transplant.
* **High blood pressure.** Damage to the glomeruli from inflammation or scarring can lead to increased blood pressure.
* **Nephrotic syndrome.** Nephrotic syndrome is a condition in which there is too much blood protein in urine and too little in the bloodstream. These proteins play a role in regulating fluids and cholesterol levels. A drop in blood proteins results in high cholesterol, high blood pressure and swelling (Edema) of the face, hands, feet and abdomen. In rare instances, nephrotic syndrome may cause a blood clot in a kidney blood vessel.

**Diagnosis methods**

Glomerulonephritis may be identified with tests if you have an acute illness or during routine testing during a wellness visit or an appointment managing a chronic disease, such as diabetes. Tests to assess your kidney function and make a diagnosis of glomerulonephritis include:

* **Urine test.** A urinalysis can reveal signs of poor kidney function, such as red blood cells and proteins that should not be in urine or white blood cells that are a sign of inflammation. There also may be a lack of the expected levels of waste products.
* **Blood tests.** Analysis of blood samples can reveal higher than expected levels of waste products in the bloodstream, the presence of antibodies that may indicate an autoimmune disorder, bacterial or viral infection, or blood sugar levels indicating diabetes.
* **Imaging tests.** If your doctor detects evidence of kidney disease, he or she may recommend imaging tests that may show an irregularity in the shape or size of the kidney. These tests may be an X-ray, an ultrasound exam or a CT scan.
* **Kidney biopsy.** This procedure involves using a special needle to extract small pieces of kidney tissue to look at under a microscope. A biopsy is used to confirm a diagnosis and to assess the degree and nature of tissue damage.

**Treatment options**

Treatment of glomerulonephritis and your outcome depend on:

* Whether you have an acute or chronic form of the disease.
* The underlying cause.
* The type and severity of your signs and symptoms.

Some cases of acute glomerulonephritis, especially those that follow an infection with streptococcal bacteria, might improve on their own and require no treatment. If there's an underlying cause — such as high blood pressure, an infection or an autoimmune disease — treatment will be directed to the underlying cause.

In general, the goal of treatment is to protect your kidneys from further damage and to preserve kidney function.

**Lifestyle and home remedies**

* Lower your salt intake to prevent or minimize fluid retention, swelling and hypertension.
* Consume less protein and potassium to slow the buildup of wastes in your blood.
* Maintain a healthy weight.
* Take your medications as directed by your healthcare provider.
* Control your blood sugar level if you have diabetes.
* Quit smoking.

### **Therapies for associated kidney failure**

Kidney failure is the loss of 85% or more of kidney function. Acute kidney failure due to infection-related glomerulonephritis is treated with dialysis. Dialysis uses a device that works like an artificial, external kidney that filters your blood.

End-stage kidney disease is chronic kidney disease that can only be managed by regular kidney dialysis or a kidney transplant.

**Management**

1. Keeping track of the renal function tests (RFTs), serum albumin, and urine protein excretion rate.

By controlling the BP and inhibiting the renin-angiotensin axis through Loop diuretics, which serve two purposes; the removal of excess fluid and the correction of hypertension. Angiotensin-converting enzyme inhibitors (ACEIs) are frequently the first choice for managing hypertension and chronic kidney disease (CKD). Angiotensin 2 receptor blockers (ARBs) have been found to halt CKD progression in diabetic or nondiabetic renal disease cases, much like ACEIs.

1. For individuals with severe/refractory hypertension with/without encephalopathy, vasodilators (e.g., nitroprusside and nifedipine) can be used.
2. Clinicians can manage the complications associated with progressive chronic disease, including anaemia, bone mineral disorders, acidosis, cardiovascular disease, and restless legs/cramps.
3. Appropriate counselling regarding diet.
4. Preparation for renal replacement therapy (RRT), if needed.

**Prevention tips**

* Seek prompt treatment of a strep infection with a sore throat or impetigo.
* To prevent infections that can lead to some forms of glomerulonephritis, such as HIV and hepatitis, follow safe-sex guidelines and avoid intravenous drug use.
* Control high blood pressure, which lessens the likelihood of damage to your kidneys from hypertension.
* Control your blood sugar to help prevent diabetic nephropathy.

**Differential diagnosis**

* Acute kidney injury
* Crescentic glomerulonephritis
* Diffuse proliferative glomerulonephritis
* Focal segmental glomerulonephritis
* Glomerulonephritis associated with Non streptococcal infection
* Goodpasture syndrome
* Lupus nephritis
* Membranoproliferative glomerulonephritis
* Poststreptococcal glomerulonephritis
* Response to corticosteroid therapy.
* Rapidly progressive glomerulonephritis

The following renal syndromes frequently mimic the early stages of acute GN:

* Idiopathic haematuria
* Chronic GN with an acute exacerbation
* Anaphylactoid purpura with nephritis
* Familial nephritis

## Acute Kidney Injury (AKI)

A rapid decline in kidney function over hours to days, resulting in accumulation of nitrogenous wastes (azotemia), fluid and electrolyte imbalances, and variable urine output. Causes are classified as:

* Prerenal: Due to decreased renal perfusion (e.g., dehydration, heart failure).
* Intrinsic renal: Due to direct kidney damage (e.g., acute tubular necrosis, glomerulonephritis).
* Postrenal: Due to urinary tract obstruction.  
  Symptoms may include reduced urine output, edema, nausea, confusion, and in severe cases seizures or coma. Diagnosis relies on serum creatinine, urine studies, imaging, and sometimes biopsy. Treatment targets the underlying cause, supportive care, and dialysis if needed.

Crescentic Glomerulonephritis

A severe form of rapidly progressive glomerulonephritis characterized histologically by crescent formation in Bowman's space due to proliferation of parietal epithelial cells and infiltration of macrophages. Clinically present with rapid loss of renal function and nephritic syndrome. Causes include anti-GBM disease, ANCA-associated vasculitis, and immune complex diseases.

Diffuse Proliferative Glomerulonephritis

A form of glomerulonephritis with widespread proliferation of glomerular cells and immune complex deposition, often seen in lupus nephritis (class IV). It leads to nephritic syndrome with hematuria, proteinuria, and impaired renal function.

Focal Segmental Glomerulonephritis (FSGS)

A pattern of glomerular injury affecting some glomeruli (focal) and portions of those glomeruli (segmental), leading to proteinuria and progressive renal failure. It can be primary or secondary to infections, drugs, or other diseases.

Glomerulonephritis Associated with Non-Streptococcal Infection

Glomerulonephritis triggered by infections other than streptococcus, such as bacterial endocarditis or viral hepatitis, causing immune complex deposition and inflammation.

Goodpasture Syndrome

An autoimmune disease characterized by antibodies against glomerular and alveolar basement membranes, causing rapidly progressive glomerulonephritis and pulmonary hemorrhage.

**Lupus Nephritis**

Renal involvement in systemic lupus erythematosus marked by immune complex deposition in glomeruli, causing a spectrum of glomerular injury from mild mesangial involvement to diffuse proliferative nephritis.

**Membranoproliferative Glomerulonephritis (MPGN)**

A pattern of glomerular injury with mesangial cell proliferation and thickening of capillary walls due to immune complex or complement deposition, leading to nephritic or nephrotic syndrome.

**Poststreptococcal Glomerulonephritis**

An immune complex-mediated glomerulonephritis occurring after streptococcal infection, presenting with hematuria, edema, hypertension, and reduced renal function.

**Response to Corticosteroid Therapy**

Many glomerulonephritides, especially those with immune-mediated mechanisms like lupus nephritis or FSGS, may respond to corticosteroids and immunosuppressive agents, which reduce inflammation and immune activity.

**Rapidly Progressive Glomerulonephritis (RPGN)**

A clinical syndrome of rapid loss of renal function over days to weeks, often associated with crescentic glomerulonephritis on biopsy. Causes include anti-GBM disease, ANCA-associated vasculitis, and severe immune complex diseases.

Renal Syndromes Mimicking Early Acute GN

* **Idiopathic Hematuria**: Persistent microscopic hematuria without overt glomerular inflammation or renal dysfunction.
* **Chronic GN with Acute Exacerbation**: A chronic glomerular disease presenting with sudden worsening of symptoms and renal function.
* **Anaphylactoid Purpura with Nephritis**: Also known as Henoch-Schönlein purpura nephritis, a small vessel vasculitis causing purpura and renal involvement.
* **Familial Nephritis**: Genetic forms of glomerulonephritis such as Alport syndrome presenting with hematuria and progressive renal failure.

**Prognosis**

Post-streptococcal glomerulonephritis (PSGN) has an excellent prognosis, especially in children with complete recovery, usually occurring within 6 to 8 weeks. In adults, around 50% of the patients continue to have reduced renal function, hypertension, or persistent proteinuria.

Frequently IgA nephropathy has a benign course. Others gradually progress to End Stage Renal Disease (ESRD), with ESRD frequency increasing with age.

Additionally, on presentation, nephrotic range proteinuria, hypertension, high serum creatinine level, and widespread intestinal fibrosis of the kidneys indicate a poor prognosis.

Membranoproliferative glomerulonephritis progresses to ESRD inevitably, despite therapy. Also, the frequency of recurrence is high even after a kidney transplant.

**WHEN TO SEE A DOCTOR**

### **When should I see my healthcare provider?**

Contact your healthcare provider if you have symptoms like:

* Blood in your pee (hematuria) or other changes in the appearance of your pee.
* Changes in how often you pee.
* Joint pain.
* Swelling in your legs or face.
* Shortness of breath.

**Statistics**

* Glomerulonephritis constitutes 25% to 30% of all end-stage renal disease cases—about a quarter of patients present with nephritic syndrome.
* IgA nephropathy has been found to be the most common cause of glomerulonephritis worldwide. However, post-streptococcal glomerulonephritis remains much more prevalent in regions such as the Caribbean, Africa, India, Pakistan, Papua New Guinea, South America, and Malaysia.
* Acute glomerulonephritis affects males more than females, with a male-to-female ratio of 2 to 1.

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**Renovascular Hypertension**

**Definition and description**

## **Alternative Names**

Renal hypertension; Hypertension - renovascular; Renal artery occlusion; Stenosis - renal artery; Renal artery stenosis; High blood pressure – renovascular

Renovascular hypertension happens when the blood flow to the kidneys is compromised, leading to high blood pressure through a hormonal response by the affected kidney. This is a serious condition and can lead to complications such as heart attack, stroke, and even death. Renovascular hypertension involves decreased perfusion to the kidney and activation of the renin-angiotensin-aldosterone (RAAS) pathway. Activation of the RAAS pathway results into the following:

· Vasoconstriction, mostly in the heart, kidney, and vascular smooth muscle

· Sympathetic nervous stimulation causing a presynaptic release of norepinephrine

· Stimulates secretion of aldosterone by the adrenal cortex, which in turn causes sodium and water retention, thereby raising blood pressure.

· It also causes the increased synthesis of collagen type I and III in fibroblasts, leading to thickening of the vascular wall and myocardium, and fibrosis

· It has been shown to have a growth effect on renal cells, which has been implicated in the development of glomerulosclerosis and tubulointerstitial fibrosis

**Causes and risk factors**

Renal artery stenosis:

Renal artery stenosis secondary to atherosclerosis is the most common cause and is mostly seen in older adults (>65 years). It has a higher prevalence in patients with known atherosclerotic disease (such as those with coronary artery disease, peripheral artery disease, or carotid artery stenosis) and autopsy studies have revealed that "greater than 25% of all patients who die of cardiovascular disease have some degree of RAS.”

Fibromuscular dysplasia (FMD): Fibromuscular dysplasia (FMD) is usually seen in young women and accounts for around 10% of renovascular hypertension and 5.8% of secondary hypertension. FMD can affect any arterial bed but most commonly affects the distal two-thirds of the renal artery.

Buildup on kidney (renal) arteries. Fats, cholesterol and other substances (plaque) can build up in and on your kidney, artery walls (atherosclerosis). As these deposits get larger, they can harden, reduce blood flow, cause kidney scarring and eventually narrow the artery. Atherosclerosis occurs in many areas of the body and is the most common cause of renal artery stenosis

**Risk factors**

· Aging

· High blood pressure

· High cholesterol

· Diabetes

· Obesity

· Smoking and other tobacco use

· A family history of early heart disease

· Lack of exercise

**Signs & Symptoms**

· High blood pressure that begins suddenly or worsens without explanation

· High blood pressure that begins before age 30 or after age 50

As renal artery stenosis progresses, other signs and symptoms may include:

· High blood pressure that's hard to control

· A whooshing sound as blood flows through a narrowed vessel (bruit), which your doctor hears through a stethoscope placed over your kidneys

· Elevated protein levels in the urine or other signs of a problem with kidney function

· Worsening kidney function during treatment for high blood pressure

· Fluid overload and swelling in your body's tissues

· Treatment-resistant heart failure

**Possible complications**

Complications of Renovascular hypertension are mostly due to uncontrolled blood pressure and include:

* Renal failure
* Myocardial infarction
* Stroke
* Pulmonary Edema
* Retinopathy
* Left ventricular hypertrophy
* Congestive heart failure
* Aneurysm
* Vascular dementia

**Diagnosis methods**

Laboratory Tests

1. Urine analysis: To check for proteinuria, hematuria, and casts. The presence of proteinuria indicates the presence of renal parenchymal disorder, whereas the presence of hematuria or red blood cell (RBC) casts indicates the presence of glomerulonephritis.

2. Blood urea nitrogen and serum creatinine: To assess baseline kidney function.

3. Basal metabolic profile: To assess for electrolyte disturbances and acid-base balance.

4. Complement levels and autoimmune profile: In suspected cases of autoimmune diseases affecting the renal vasculature.

5. Plasma free metanephrines or 24-hour urinary fractionated metanephrines and normetanephrine to rule out pheochromocytoma

6. Plasma renin-aldosterone ratio to rule out hyperaldosteronism

7. 24-hour urinary free cortisol or low dose dexamethasone suppression test to rule out Cushing's syndrome.

**Imaging**

8. Doppler ultrasound. High-frequency sound waves help your doctor see the arteries and kidneys and check their function. This procedure also helps your doctor find blockages in the blood vessels and measure their severity.

9. CT scan. During a CT scan, an X-ray machine linked to a computer creates a detailed image that shows cross-sectional images of the renal arteries. You may receive a dye injection to show blood flow.

10. Magnetic resonance angiography (MRA). MRA uses radio waves and strong magnetic fields to produce detailed 3D images of the renal arteries and kidneys. A dye injection into the arteries outlines blood vessels during imaging.

11. Renal arteriography. This special type of X-ray exam helps your doctor find the blockage in the renal arteries and sometimes open the narrowed part with a balloon or stent. Before an X-ray is taken, your doctor injects a dye into the renal arteries through a long, thin tube (catheter) to outline the arteries and show blood flow more clearly. This test is mainly done if it's also likely that you need a small tube (stent) placed in your blood vessel to widen it.

**Treatment options**

The management of renovascular hypertension aims to treat the underlying cause. Several options are available, which include pharmacological and invasive therapy.

Pharmacological therapy: Entails the use of antihypertensive medications to control blood pressure. Since RAAS is the most prominent pathway contributing to hypertension in these disorders, ACE-Is and angiotensin receptor blockers (ARBs) form the cornerstone of managing renovascular hypertension (Class 1a indication). Often more than one medication will be needed to control the blood pressure. Calcium channel blockers, thiazides, beta-blockers, and hydralazine have been shown to be effective to control blood pressure in patients with RAS.

Direct renin inhibitors:

Direct renin inhibitors such as aliskiren have been studied as monotherapy or in combination with ACEIs/ARBs to treat hypertension. Though it has been shown to be effective for the treatment of hypertension there is not enough data to prove its efficacy in treating renovascular hypertension.

ACEIs and ARBs inhibit the action of angiotensin II, thereby causing vasodilation and promoting sodium and water excretion. However, these medications are contraindicated in patients with a single functioning kidney or bilateral lesions as they can cause efferent arteriolar vasodilation leading to interruption in autoregulation and thereby decreasing glomerular filtration. While these medications are effective in controlling blood pressure, they can also lead to worsening renal function.

Percutaneous angioplasty: Percutaneous angioplasty is the treatment of choice for renovascular hypertension due to FMD and for patients with atherosclerotic renal artery stenosis that is not controlled with medications.

1. Patients with hemodynamically significant RAS and recurrent, unexplained congestive heart failure or sudden, unexplained pulmonary Edema (class Ia)

2. Hemodynamically significant RAS and accelerated hypertension, resistant hypertension, malignant hypertension or hypertension with an unexplained unilateral small kidney, and hypertension with intolerance to medication (Class IIa)

3. Patients with bilateral RAS and progressive chronic kidney disease or a RAS to a solitary functioning kidney (Class IIa)

4. Patients with hemodynamically significant RAS and unstable angina (class IIa)

5. Asymptomatic bilateral or solitary viable kidney with hemodynamically significant RAS (Class IIb)

6. Patients with RAS and chronic renal insufficiency with unilateral RAS (class IIb)

7. In addition to angioplasty, renal stent placement is indicated for patients with ostial atherosclerotic lesions (Class I).

Patients with FMD and renovascular hypertension are also treated with percutaneous intervention with or without a stent. Multiple studies have shown a decrease in baseline blood pressure after intervention for FMD.

Renal artery bypass surgery. During a bypass procedure, doctors graft a substitute blood vessel to the renal artery to make a new route for blood to reach your kidneys. Sometimes this means connecting the renal artery to a vessel from somewhere else, such as the liver or spleen. These operations are most often done if angioplasty isn't successful, or when there's a need for additional surgical procedures.

Management:

Primary management of renovascular hypertension should aim to correct the underlying cause. Renovascular hypertension due to atherosclerotic renal artery stenosis should be primarily managed medically as multiple studies have failed to show renal or cardiovascular benefits with invasive management.

**Differential diagnosis**

The differential diagnosis for renovascular hypertension includes potential causes of secondary hypertension, such as:

· Pheochromocytoma: Usually presents as a constellation of symptoms such as flushing, headache, tachycardia, and episodic uncontrolled hypertension.

· Primary hyperaldosteronism: Presents with persistent hypokalemia and metabolic alkalosis.

· Obstructive sleep apnea: Usually seen in obese males with increased neck circumference and a history of snoring. Diagnosed with polysomnography/sleep study.

· Coarctation of the aorta: Patients usually have a systolic murmur, radio-femoral delay, and upper extremity hypertension. The diagnosis is with MRA or CTA.

· Cushing syndrome: Associated with moon facies, buffalo hump, proximal myopathy, glucose intolerance, abdominal striae, and central obesity.

**Prognosis**

Atherosclerotic renal artery stenosis is a progressive disorder that can lead to worsening stenosis and, ultimately, renal failure.

High blood pressure (systolic >160 mm hg),

diabetes mellitus, and high-grade stenosis (>60% obstruction) have been shown to be associated with an increased rate of progression.

Untreated renovascular hypertension can also lead to end-stage renal failure with a median survival time of 25 months and a 4-

**EPIDEMIOLOGY**

## United States statistics

RVHT is a common type of potentially correctable secondary hypertension. Although it accounts for less than 1% of mild hypertension, the prevalence may be as high as 38% in patients with severe hypertension and general atherosclerotic vascular or peripheral vascular disease.

The incidence of hypertension in children is reported to be 1-5%, and in adolescents may be as high as 10%. In children, unlike adults, 70-80% of hypertension may be secondary hypertension, which is often correctable. RVHT is second only to coarctation of the aorta as a surgically correctable cause of hypertension in children.

### International statistics

According to the World Health Organization (WHO), among the regional populations, the African region has the highest prevalence of hypertension put at 27%, which is a major contributor to the burden of hypertension in LMICs; with the high prevalence attributable to an increase in the risk factors for hypertension in Africans in the last five decades . The reported prevalence of hypertension varies in various parts of the world and is probably influenced by many factors including genetic, racial and environmental factors. In the year 2000, high blood pressure was estimated to affect about 972 million persons globally, thus giving a worldwide prevalence of 26.4% in the adult population (26.6% of men and 26.1% of women), with 333 million affected persons in developed countries and 639 million persons in the developing world. It is projected that in the year 2025, 1.56 billion persons will be affected, an increase of 60% from the figure in 2000. Most of the expected increase in the prevalence of hypertension will occur in the developing countries where nearly three-quarters of all hypertensives will be found by the year 2025.

The prevalence of RVHT internationally is not clear, but it likely accounts for the sole etiology in a relatively small percentage of unselected patients with hypertension. Significant geographic differences in the overall prevalence of RVHT have not been reported, though the etiology does appear to vary geographically.

In the western hemisphere, FMD is the most common cause of pediatric RVHT. Reports from Asia and South Africa identify Takayasu arteritis affecting the renal artery as the most common cause of RVHT in children. One pediatric study in south Asia found that 87% of the patients presenting with RVHT had arteritis.

**NIGERIA**

The overall age-standardized prevalence of hypertension was 38.1% and this varied across the geo-political zones as follows: North-Central, 20.9%; North-East, 27.5%; North-West, 26.8%; South-East, 52.8%; South-South, 44.6%; and South-West, 42.1%. Prevalence rate did not differ significantly (p > 0.05) according to place of residence; 39.2% versus 37.5 %; urban vs rural. Prevalence of hypertension increased from 6.8% among subjects less than 30 years to 63.0% among those aged 70 years and above. Awareness was better (62.2% vs. 56.6%; P = 0.0272); treatment rate significantly higher (40.9 % vs. 30.8%; P < 0.0001) and control similar (14 vs. 10.8%) among urban compared to rural residents. Women were more aware of (63.3% vs. 52.8%; P < 0.0001); had similar (P > 0.05) treatment (36.7 vs. 34.3%) and control (33.9% vs. 35.5%) rates of hypertension compared to men.

results suggest a large burden of hypertension in Nigeria and a closing up of the rural-urban gap previously reported. This calls for a change in public health policies anchored on a primary health care system to address the emerging disease burden occasioned by hypertension.

There has been a recently reported very high increase in prevalence of hypertension in Nigeria between 1995 and 2000 where it increased by over 540% from four million to twenty-eight million individuals. The age adjusted HTN prevalence in 2020 was 32.5%, quite higher than the 28% prevalence reported in 2010 in an earlier study. Murthy *et al.* reported a very high prevalence of hypertension among the Ibos and Nupe communities in the southeast and northcentral at 40.4% and 50.5% in 2013. Previously, the prevalence of hypertension in the rural communities was low with the burden of the disease associated predominantly with urban dwellers. Recent studies suggest otherwise with the prevalence of hypertension recording an exponential rise in the rural populations, 27.6% was reported in south-east Nigeria in 2023. Possibly widespread changes in sociocultural and lifestyle habits that have deeply permeated even the rural communities are responsible for the rising prevalence of hypertension among all adult populations in the country with studies reporting association between socio-demographic and lifestyle factors with hypertension.

Owerri, like many other parts of Nigeria, is experiencing rapid developmental and socio-economical changes with associated adoption of western lifestyles, including dietary patterns, which no doubt can impact on the prevalence of NCDs such as hypertension in the populace. A high prevalence of 42.7% was reported in adults in a rural community, a suburb of Owerri in 2021.

Globally, about half (46%) of adults with hypertension are not aware they have the disease.

The apparent “silent” nature of hypertension in most sufferers, contributes to the high level of unawareness among the subjects.

Several risk factors for essential hypertension have been identified and they include older age, a positive family history of hypertension, diabetes, heavy alcohol intake, obesity, excessive dietary salt intake, sedentary lifestyle, cigarette smoking as well as hyperlipidaemia

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**ACUTE KIDNEY INJURY(AKI) OR ACUTE RENAL FAILURE**

**Description**

Acute kidney injury happens when the kidneys suddenly can't filter waste products from the blood. When the kidneys can't filter wastes, harmful levels of wastes may build up. The blood's chemical makeup may get out of balance. Acute kidney injury (AKI) can happen within a few hours or a few days. For most people, AKI develops within 48 hours, but sometimes it can take as long as 7 days.

This replaces an older term “acute renal failure” (or ARF) which you may still see or hear occasionally.

Acute kidney injury is most common in people who are in the hospital, mostly in people who need intensive care.

Acute kidney injury ranges from mild to severe. If severe, ongoing and not treated, it can be fatal. But it also can be reversed. People in otherwise good health may get back typical or nearly typical use of their kidneys.

the change of serum creatinine and urine output, as follows

* Rise in serum creatinine ≥0.3 mg/dL within 48 hours
* Rise in serum creatinine ≥1.5 times baseline, which is known or presumed to have occurred within the prior seven days
* Urine output < 0.5 mL/kg/hour for six hours.

## **Pathophysiology**

The driving force for glomerular filtration is the pressure gradient from the glomerulus to the Bowman space. Glomerular pressure depends primarily on renal blood flow (RBF) and is controlled by the combined resistances of renal afferent and efferent arterioles. Regardless of the cause of AKI, reductions in RBF represent a common pathologic pathway for a decrease in the glomerular filtration rate (GFR). The etiology of AKI consists of 3 main mechanisms: prerenal, intrinsic, and obstructive (postrenal).

In prerenal failure, GFR is depressed by compromised renal perfusion. Tubular and glomerular functions remain normal.

Intrinsic failure includes diseases of the kidney itself, predominantly affecting the glomerulus, interstitium, or tubule, that are associated with the release of renal afferent vasoconstrictors. Ischemia is the most common cause of intrinsic kidney failure. Patients with chronic kidney disease (CKD) may also present with superimposed AKI from prerenal failure and obstruction, as well as intrinsic kidney disease.

Obstruction of the urinary tract initially causes an increase in tubular pressure, which decreases the filtration driving force. This pressure gradient soon equalizes, and maintenance of a depressed GFR then depends on renal efferent vasoconstriction.

**Causes**

Acute kidney injury can happen when:

* You have a condition that slows blood flow to your kidneys.
* You have damage to your kidneys.
* Your kidneys' urine drainage tubes, called ureters, get blocked.

### **Slowed blood flow to the kidneys**

Conditions that may slow blood flow to the kidneys and lead to kidney injury include:

* Loss of too much body fluid, called dehydration.
* Infection with or without sepsis or septic shock.
* Medicines such as aspirin, ibuprofen (Advil, Motrin IB, others) or naproxen sodium (Aleve).
* Blood or fluid loss.
* Severe low blood pressure from blood pressure medicines.
* Heart attack.
* Heart failure or heart disease.
* Liver cirrhosis or failure.
* Bad allergic reaction, called anaphylaxis.
* Bad burns.

### **Damage to the kidneys**

The following may damage the kidneys and lead to acute kidney injury:

* Swelling and irritation, called inflammation, of the tiny filters in the kidneys. This is called glomerulonephritis (glove-mer-u-loe-nuh-FRY-tis).
* Medicines, such as certain chemotherapy drugs, antibiotics and dyes used during imaging tests.
* Infection, such as with the virus that causes coronavirus disease 2019 (COVID-19).
* Toxins, such as alcohol, heavy metals and cocaine.
* An immune system condition called lupus that causes glomerulonephritis.
* Blood clots in the veins and arteries in and around the kidneys.
* Cholesterol deposits that block blood flow in the kidneys.
* A condition that results from red blood cells being destroyed too early, called hemolytic uremic syndrome.
* A group of rare diseases affecting the skin and connective tissues called scleroderma.
* A rare blood disorder called thrombotic thrombocytopenic purpura.
* Muscle tissue breakdown, called rhabdomyolysis. The toxins from the muscle being destroyed leads to kidney damage.
* Breakdown of tumor cells called tumor lysis syndrome. This leads to the release of toxins that can injure the kidneys.

### **Urine blockage in the kidneys**

Conditions that keep urine from leaving the body are called urinary obstruction. These can lead to acute kidney injury. They include:

* Kidney stones.
* Enlarged prostate.
* Blood clots in the urinary tract.
* Bladder cancer.
* Prostate cancer.
* Cervical cancer.
* Colon cancer.
* Growth pushing on the ureters.
* Nerve damage of the nerves that control the bladder.

Another common cause for AKI is when your body is reacting to an urgent or emergent health concern (such as heart surgery or COVID-19 infection).

Usually, AKI happens because of a combination of factors. This is especially true for older adults who are at higher risk given their age.

**Risk factors**

Acute kidney injury almost always is linked to another medical condition or event. Conditions that can increase your risk of acute kidney injury include:

* Ongoing kidney disease, also called chronic kidney disease.
* Older age, but it does happen to children.
* Being in the hospital, most often for a serious condition that needs intensive care.
* Blockages in the blood vessels in your arms or legs, called peripheral artery disease.
* Diabetes, especially if it's not controlled.
* High blood pressure.
* Heart failure.
* Liver diseases.
* Certain cancers and their treatments.

**Symptoms**

The signs and symptoms of AKI can differ depending on many factors like the cause, severity, and your other health conditions

Symptoms of acute kidney injury may include:

* Less urine output. (pee) than usual or no urine.
* High blood pressure
* Fluid buildup, which can cause shortness of breath and swelling in the legs, ankles or feet.
* Tiredness.
* Confusion or fogginess.
* Nausea.
* Pain in the belly or in the side below the rib cage.
* Weakness.
* Irregular heartbeat.
* Itching.
* Loss of appetite.
* Chest pain or pressure.
* Seizures or coma in severe cases.

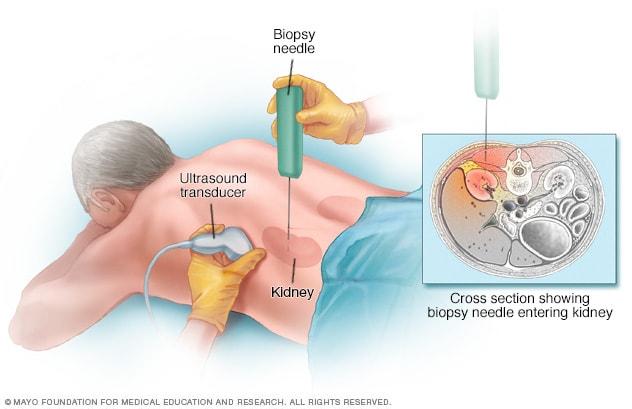
Sometimes acute kidney injury causes no symptoms. Then it may be found through lab tests done for something else.

**Complications**

Complications of acute kidney injury may include:

* **Fluid buildup.** A buildup of fluid in your lungs can cause shortness of breath.
* **Chest pain.** The lining that covers your heart, called the pericardium, can get inflamed. This can cause chest pain.
* **Muscle weakness.** This can result from the body's fluids and minerals in the blood called electrolytes being out of balance.
* **Permanent kidney damage.** Sometimes, acute kidney injury causes lifelong loss of the use of the kidneys, called end-stage renal disease. People with end-stage renal disease need either lifelong treatments to remove waste from the body, called dialysis, or a kidney transplant to survive.
* **Death.** Acute kidney injury can cause the kidneys to stop working.

## **Diagnosis**

**Kidney biopsy Enlarge** image

You might have the following tests to diagnose acute kidney injury:

* **Blood tests.** A sample of your blood may show fast-rising levels of urea and creatinine. This helps show how your kidneys are working.
* **Urine output measures.** Measuring how much urine you pass in 24 hours may help find the cause of your kidney failure.
* **Urine tests.** A sample of your urine may show something that suggests a condition that might explain kidney failure. This is called urinalysis.
* **Imaging tests.** Imaging tests such as ultrasound and CT scans can show your kidneys.
* **Removing a sample of kidney tissue for testing.** Your healthcare professional may suggest removing a small sample of your kidney tissue for lab testing. This is called a biopsy. A needle put through your skin and into your kidney removes the sample.

**TREATMENT**

Treatment for acute kidney injury most often means a hospital stay. Most people with acute kidney injury are already in the hospital. How long you'll stay in the hospital depends on the reason for your acute kidney injury and how quickly your kidneys recover.

### **Treating the cause of your kidney injury**

Treatment for acute kidney injury involves finding the illness or injury that damaged your kidneys. Your treatment depends on the cause. It might involve stopping a medicine that's damaging your kidneys.

### **Treating complications until your kidneys recover**

Your healthcare team also works to prevent complications and give your kidneys time to heal. Treatments that help prevent complications include:

* **Treatments to balance fluids in your blood.** If a lack of fluids in your blood is the cause of your acute kidney injury, you may need fluids through a vein, called intravenous (IV) fluids.

If acute kidney injury causes you to have too much fluid, this may lead to swelling in your arms and legs. Then you may need medicines called diuretics, which cause your body to get rid of extra fluids.

* **Medicines to control blood potassium.** Your kidneys might not filter potassium from your blood well enough. Potassium regulates blood pressure and other body functions.

You might need medicines called potassium binders to keep potassium from building up. These include sodium zirconium cyclosilicate (Lokelma) or patiromer (Veltassa). Too much potassium in the blood can cause irregular heartbeats, called arrhythmias, and muscle weakness.

* **Medicines to restore blood calcium levels.** If the levels of calcium in your blood drop too low, you might need to get calcium through a vein, called an infusion.
* **Treatment to remove poisons from your blood.** If wastes build up in your blood, you may need hemodialysis for a time. Also called dialysis, it helps remove poisons and excess fluids from your body while your kidneys heal.

Dialysis also may help remove excess potassium from your body. During dialysis, a machine pumps blood out of your body through an artificial kidney, called a dialyzer, that filters out waste. The blood is then returned to your body.

Depending on your level of kidney injury you may be:

* Careful monitoring of fluid intake and output, diet, drugs, prevention of infections.
* **Given additional fluids, often via a drip, if you are short of fluid**
* **Given medication to treat or prevent infections and taken off certain drugs (such as ibuprofen) until your kidney function improves, or given alternatives**
* **Given treatments to relieve any blockages**
* Continuous blood purification treatments – such as continuous dialysis or hemofiltration, may be given, especially in intensive care units. These don’t necessarily work better but can be easier in patients who are critically ill.

##### **Treatments for the cause**

* **Obstruction** – relief of the blockage usually makes the situation much better.
* **Pre-renal** – once ATN is established, there is no treatment that is proven to speed recovery, but early treatment with fluids etc may help this developing.
* **Renal** – some causes benefit from urgent treatment. Examples include:

Acute interstitial nephritis  
Crescentic nephritis  
Glomerulonephritis – may need special treatments too.

**Prevention**

You might cut your risk of acute kidney injury by taking care of your kidneys. Try to:

* **Get treated quickly for bad infections.**
* **Work with your healthcare team to manage kidney and other ongoing conditions.** Kidney disease, diabetes or high blood pressure increases your risk of acute kidney injury. If you have one of these, do what your healthcare team tells you to manage your condition.

If you have risk factors for kidney disease, check with your healthcare team to be sure that prescription medicines you take are safe for your kidneys.

* **Read labels when taking pain medicines available without a prescription.** Do what the label says when taking medicines such as aspirin, acetaminophen (Tylenol, others), ibuprofen (Advil, Motrin IB, others) and naproxen sodium (Aleve). Taking too much of these medicines may increase your risk of kidney injury. This is especially true if you already have kidney disease, diabetes or high blood pressure.
* **Live a healthy lifestyle.** Be active and eat a healthy, balanced diet. If you drink alcohol, drink only in moderation.

**Lifestyle and home remedies**

During your recovery from acute kidney injury, a special diet can help support your kidneys and limit the work they must do. Your healthcare team may send you to a dietitian. A dietitian can look at what you eat and suggest ways to make your diet easier on your kidneys.

Your dietitian may suggest that you:

* **Choose foods lower in potassium.** These include apples, peaches, carrots, green beans and white bread and white rice. Eat them instead of foods higher in potassium. These include potatoes, bananas, tomatoes, oranges, beans and nuts.
* **Don't eat foods with added salt.** This includes many packaged foods, such as frozen dinners, canned soups and fast foods. Other foods with added salt include salty snack foods, canned vegetables, and processed meats and cheeses.
* **Limit phosphorus.** Phosphorus is a mineral found in foods, such as dark-colored sodas, milk, oatmeal and bran cereals. Too much phosphorus in your blood can weaken your bones and cause your skin to itch.

As your kidneys get better, you may no longer need a special diet. But healthy eating still is important.

**DIFFERENTIAL DIAGNOSIS**

Differentials to consider in AKI include the following:

* Abdominal aneurysm
* Alcohol toxicity
* Alcoholic ketoacidosis
* Chronic kidney disease
* Dehydration
* Diabetic ketoacidosis
* Gastrointestinal (GI) bleeding
* Heart failure
* Metabolic acidosis
* Obstructive uropathy
* Protein overloading
* Renal calculi
* Sickle cell anemia
* Steroid use
* Urinary obstruction
* Urinary tract infection

## **Abdominal Aortic Aneurysm (AAA)**

An abdominal aortic aneurysm is a localized dilation or bulging of the abdominal aorta, typically defined as a diameter greater than 3 cm or more than 50% larger than normal. Most AAAs are asymptomatic and discovered incidentally during imaging for other reasons or through screening programs, especially in older men and smokers.

**Alcohol Toxicity**

Adverse effects of excessive alcohol consumption causing multi-organ damage including liver disease, cardiomyopathy, neuropathy, and metabolic disturbances.

**Alcoholic Ketoacidosis**

A metabolic acidosis occurring in chronic alcoholics with recent binge drinking and poor nutrition, characterized by elevated ketones, dehydration, and electrolyte imbalances.

**Chronic Kidney Disease (CKD)**

Progressive loss of kidney function over months to years, leading to accumulation of waste products, electrolyte disturbances, anemia, and complications such as hypertension and cardiovascular disease.

**Dehydration**

Excessive loss of body water leading to hypovolemia, electrolyte imbalances, and impaired organ perfusion.

**Diabetic Ketoacidosis (DKA)**

A life-threatening complication of diabetes characterized by hyperglycemia, ketosis, metabolic acidosis, dehydration, and electrolyte abnormalities.

**Gastrointestinal (GI) Bleeding**

Bleeding originating anywhere in the gastrointestinal tract, presenting with hematemesis, melena, or hematochezia, potentially causing anemia and hemodynamic instability.

**Heart Failure**

A clinical syndrome caused by structural or functional cardiac disorders impairing ventricular filling or ejection, leading to symptoms of fluid overload and poor perfusion.

**Metabolic Acidosis**

A disturbance characterized by decreased blood pH due to accumulation of acids or loss of bicarbonate, seen in conditions like renal failure, ketoacidosis, or lactic acidosis.

**Obstructive Uropathy**

Blockage of urinary flow causing hydronephrosis and potential renal impairment.

**Protein Overloading**

Excessive protein filtration or reabsorption in the kidney, potentially leading to tubular injury.

**Renal Calculi**

Kidney stones cause obstruction, pain, hematuria, and risk of infection.

**Sickle Cell Anemia**

A hereditary hemoglobinopathy causing vaso-occlusion, hemolysis, and multisystem complications including renal papillary necrosis.

**Steroid Use**

Chronic corticosteroid therapy can cause metabolic disturbances, immunosuppression, and increased risk of infections.

**Urinary Obstruction**

Blockage in the urinary tract causing impaired urine flow, hydronephrosis, and kidney damage.

**Urinary Tract Infection (UTI)**

Infection of the urinary tract, commonly causing dysuria, frequency, urgency, and sometimes systemic symptoms.

## **Epidemiology**

In the United States, approximately 1% of patients admitted to hospitals have AKI at the time of admission. The estimated incidence rate of AKI during hospitalization is 2-5%. AKI develops within 30 days postoperatively in approximately 1% of general surgery cases and arises in more than 50% of intensive care unit (ICU) patients.In recipients of solitary kidney transplants, 21% developed AKI within the first 6 months after transplantation.

hospitalization rates for dialysis-requiring AKI in adults increased considerably while mortality decreased. In adults with diabetes, rates increased from 26.4 to 41.1 per 100,000 population, with relative increases greater in younger versus older adults. In adults without diabetes, rates increased from 4.8 to 8.7 per 100,000 population between 2000 and 2009 and then plateaued. Mortality declined significantly in patients both with and without diabetes.12 - 35 new AKI cases are seen per year. A single-center study puts cAKI and hAKI incidences at 9.8 per million children population (pmcp)/year (0.46%) and 3.7 pmcp/ year (0.17%), respectively; cAKI and hAKI prevalence rates were 49.2 pmcp (2.23%) and 18.3 pmcp (0.84%), respectively IN NIGERIA.

Leading causes of AKI, accounting for 80.0% of all etiologies, were nephrotoxins (29.0%), infection (20.0%), intravascular volume depletion (17.9%), and glomerular disease (13.1%).

Financial constraints, late presentation, presence of ≥ 2 comorbidities, need for dialysis, non-dialysis when indicated, severe hypertension, white cell count > 15 000/mm3, and platelet count < 100 000/mm3 are significant mortality risk factors in childhood AKI in our environment.

Children aged one month to 15 years managed for AKI (identified by pediatric RIFLE criteria) from January 2017 to December 2017 were followed up for a short period of four weeks following the AKI. An annual prevalence of 26 AKI cases per 1000 children was recorded with 43 AKI cases from 1634 children seen during the 12-month period. The median age was 48 months. Twenty-two were males (51.2%). Sepsis (20, 46.6%), acute glomerulonephritis (5, 11.6%), diarrheal dehydration (5, 11.6%), severe falciparum malaria (4, 9.3%), and hemolytic uremic syndrome (4, 9.3%) were the major causes of the AKI. Fourteen children were managed conservatively, while 29 children that required dialysis had access to it. Thirteen children died (percentage mortality of 30.2%). The hazard of dying was eight times more in male gender [95% confidence interval (CI); 1.03-72.9, P = 0.017] and was lower in children without pulmonary edema by 0.14 (95% CI; 0.03-0.63, P = 0.01). In Nigeria, mortality from AKI is still high, and male children and those with pulmonary edema should be closely managed for AKI to reduce this high mortality.

**Prognosis**

The prognosis for patients with AKI is directly related to the cause of the injury and mostly to the presence or absence of preexisting kidney disease (estimated GFR [eGFR] < 60 mL/min), as well as to the duration of kidney dysfunction prior to therapeutic intervention. In the past, AKI was thought to be completely reversible, but long-term follow-up of patients with this condition has shown otherwise.

However, the greatest impact on mortality was seen in individuals with an eGFR of greater than 60 mL/min who developed AKI. Those with stage 3 AKI had a mortality rate of 50%, while mortality in individuals with an eGFR of greater than 60 mL/min but who did not develop AKI was only 3%. Among individuals with an eGFR of less than 30, the mortality rate was 12.1% in those who did not develop AKI, versus 40.7% among patients with stage 3 AKI.

**WHEN TO SEE A DOCTOR**

It’s essential that AKI is detected early, for a better chance of the kidneys fully recovering. Causes of AKI include dehydration, infections, certain medications and blockage of one or both tubes leading from the kidneys to the bladder. AKI is different to chronic kidney disease (CKD) when there is a reduction in the function of the kidneys that does not get better. You should contact your GP urgently if you have severe (or ongoing) vomiting and/or diarrhea, especially if you are on blood pressure medications, or are unable to keep fluids down; or if you notice a reduction in the amount of urine you are passing. In some situations, your GP may tell you to stop certain medications, for example blood pressure tablets, water tablets or certain anti-inflammatory drugs or painkillers while you are unwell. Your GP may also ask for a blood test to make sure your kidneys are working properly. Occasionally it may be necessary to be admitted to hospital for treatment of AKI if it is not getting better.

**Questions to ask**

What are my biggest risk factors for AKI?

What can I do to help lower my risk for AKI?

Are there any medications I should avoid (either now or in the future) due to my kidneys?

[If having potential symptoms of AKI] Should I go to the emergency room for my symptoms?

Was the cause of my AKI preventable? If so, what can I do to prevent it from happening again?

When should I follow up after my AKI treatment is done to check my kidney health?

**REFERENCES**

<https://www.kidney.org/kidney-topics/acute-kidney-injury-aki>

<https://www.mayoclinic.org/diseases-conditions/kidney-failure/symptoms-causes/syc-20369048>

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# **chronic kidney disease (CKD) chronic renal disease**

# Your kidneys do many important jobs. Some of the ways they keep your whole body in balance include:

Removing natural waste products and extra water from your body

Help make red blood cells

Balancing important minerals in your body

Help maintain your blood pressure

Keep your bones healthy

Kidney disease is called “chronic” because kidney function slowly decreases over time. CKD can lead to kidney failure, which is also called end-stage kidney disease. Not everyone with CKD will develop kidney failure, but the disease will often worsen without treatment. There’s no cure for chronic kidney disease. But there are steps you can take to slow kidney damage. Treatments like dialysis and transplantation are options for kidney failure (end-stage kidney disease).

So, CKD is [divided into 5 stages](https://www.kidney.org/atoz/content/stages-chronic-kidney-disease-ckd) to help guide treatment decisions.The stages are based on how well your kidneys can filter out waste from your blood. Blood and urine tests determine which stage of CKD you’re in.

The stages range from very mild (stage 1) to kidney failure (stage 5). Healthcare providers determine the stage of your kidney function according to the glomerular filtration rate (GFR). Your GFR is a number based on the amount of creatinine, a waste product, found in your blood.

| **Stage** | **GFR (mL/min)** | **What It Means** |
| --- | --- | --- |
| Stage 1 | 90 and higher | Your kidneys are working well but you have signs of mild kidney damage. |
| Stage 2 | 60 to 89 | Your kidneys are working well but you have more signs of mild kidney damage. |
| Stage 3a | 45 to 59 | Your kidneys aren’t working as well as they should and show mild to moderate damage. This is the most common stage. You may notice symptoms at this stage. |
| Stage 3b | 30 to 44 | Your kidneys show moderate damage and don’t work as well as they should. With the right treatment, many people can stay in this stage and never advance to stage 4. |
| Stage 4 | 15 to 29 | You have very poor kidney function; your kidneys are severely damaged and close to not working. |
| Stage 5 | Less than 15 | Your kidneys are very close to failing or have stopped working. You may need kidney dialysis or a kidney transplant at this stage. |

#### **Signs and symptoms**

Many people living with CKD do not have any symptoms until the more advanced stages and/or complications develop. If symptoms do happen, they may include:

Foamy urine

Urinating (peeing) more often or less often than usual

Itchy and/or dry skin

Feeling tired

Nausea

Loss of appetite

Weight loss without trying to lose weight

People who have more advanced stages of CKD may also notice:

Trouble concentrating

Numbness or swelling in your arms, legs, ankles, or feet

Achy muscles or cramping

Shortness of breath

Vomiting

Trouble sleeping

Breath smells like ammonia (also described as urine-like or “fishy”)

## **Causes and Risk Factors**

## Anyone can develop CKD - at any age. However, some people are at a higher risk than others. The most common CKD risk factors are:

Diabetes

High blood pressure (hypertension)

Heart disease and/or heart failure

Obesity

Over the age of 60

Family history of CKD or [kidney failure](https://kidney.org/atoz/content/KidneyFailure)

Personal history of [acute kidney injury (AKI)](https://www.kidney.org/atoz/content/AcuteKidneyInjury)

Smoking and/or use of tobacco products

For many people, CKD is not caused by just one reason. Instead, it is a result of many physical, environmental, and [social factors](https://www.kidney.org/atoz/content/social-determinants-health-and-chronic-kidney-disease). Early detection is important – CKD often begins without causing any noticeable symptoms. Knowing the risk factors can help you know your level of risk and if you should get checked for CKD.

### **Other causes**

CKD can also be caused by many other conditions or circumstances. Some examples include:

**Glomerular diseases:** [glomerulonephritis](https://www.kidney.org/atoz/content/glomerul), [IgA nephropathy (IgAN)](https://www.kidney.org/atoz/content/iganeph), and [HInephropathy](https://www.kidney.org/atoz/content/hiv-and-chronic-kidney-disease-what-you-need-know)

**Inherited conditions:** [polycystic kidney disease](https://www.kidney.org/atoz/content/polycystic)

**Autoimmune conditions:** [lupus (lupus nephritis)](https://www.kidney.org/atoz/content/lupus)

**Severe infections:** [sepsis](https://www.kidney.org/atoz/content/sepsis) and [hemolytic uremic syndrome (HUS)](https://www.kidney.org/atoz/content/hemolytic)

**Other causes:** [kidney cancer](https://www.kidney.org/atoz/content/kidney-cancer), [kidney stones](https://www.kidney.org/atoz/content/kidneystones), frequent untreated and/or long-lasting [urinary tract infections (UTIs)](https://www.kidney.org/atoz/content/uti), [hydronephrosis](https://www.kidney.org/atoz/content/hydronephrosis), and kidney and urinary tract abnormalities before birth.

## **Complications**

## As CKD worsens, the risk of getting complications goes up. Some examples include:

Cardiovascular disease (heart disease and/or stroke)

High blood pressure

Anemia (low levels of red blood cells)

Metabolic acidosis (buildup of acid in the blood)

Mineral and bone disorder (when blood levels of calcium and phosphorus are out of balance leading to bone and/or heart disease)

Hyperkalemia (high levels of potassium in the blood)

Kidney failure

Some conditions, like cardiovascular disease and high blood pressure, can also cause or worsen CKD.

## **Diagnosis**

### **Tests**

### Checking for CKD is easy with two simple tests:

a **blood test** known as the [estimated glomerular filtration rate (eGFR)](https://www.kidney.org/atoz/content/gfr)

a **urine test** known as the [urine albumin-creatinine ratio (uACR).](https://www.kidney.org/atoz/content/uacr)

Both tests are needed to have a clear picture of your kidney health. Having an eGFR under 60 and/or a uACR over 30 for three months or more is a sign you may have kidney disease.

The eGFR is an estimate of how well your kidneys are removing waste products from the blood. It is calculated using your [serum creatinine](https://www.kidney.org/atoz/content/what-creatinine) level, age, and sex. It can also be calculated using your [cystatin C](https://www.kidney.org/atoz/content/cystatinC) level. A “normal” eGFR varies according to age – it decreases as you get older. For this test, a **higher number** is better. Your eGFR number is used to determine your [stage of CKD](https://www.kidney.org/atoz/content/stages-chronic-kidney-disease-ckd).

The uACR measures the amount of two different substances in your urine – albumin (protein) and creatinine. Healthy kidneys keep the albumin in your blood while filtering the creatinine out into the urine. So, there should be very little or no albumin in your urine. The uACR is calculated by dividing the amount of urine albumin by the amount of urine creatinine to find the ratio. For this test, a **lower number** is better. Your uACR number is used to test for albuminuria - a significant risk factor for complications.

In some cases, your healthcare professional may order [additional tests](https://www.kidney.org/atoz/content/tests-to-check-your-kidney-health) to get more information about your kidney health. Some examples include a [kidney biopsy](https://www.kidney.org/atoz/content/kidney-biopsy) or medical imaging (CT scan, ultrasound, or MRI)

**Treatment**

Managing CKD is focused on four very important goals:

Managing the disease(s) or condition(s) that are most likely causing the CKD (for example, your diabetes, high blood pressure, or IgA nephropathy)

Taking steps to slow down the CKD disease process directly (also known as “slowing CKD progression”)

Lowering your risk of cardiovascular disease (having a heart attack or stroke)

Treating any complications that you may have because of your CKD

Specific treatment recommendations depend on your [stage of CKD](https://www.kidney.org/atoz/content/stages-chronic-kidney-disease-ckd) and what other health conditions you have (including any CKD complications). Below are recommendations that apply to most people with CKD. No two people are the same, so talk with your healthcare professional about recommendations tailored to you.

### **Medications**

Your healthcare professional may prescribe one or more medicines to help slow down or stop your CKD from getting worse. These medicines can include an ACE inhibitor/ARB, an SGLT2 inhibitor and/or an nsMRA.

Your healthcare professional may also prescribe a statin (cholesterol medicine). Guidelines recommend a statin for people with CKD who also have diabetes, a history of heart disease, or are age 50 or older. Even if you do not have high cholesterol, a statin can help lower your risk of having a heart attack or stroke.

You may also need to take additional medications or supplements to manage any CKD complications you might have (if applicable).

**Nutrition**

It is important to limit your [sodium (salt)](https://www.kidney.org/atoz/content/sodiumckd) intake to less than 2300 mg per day (about 1 teaspoon of salt from **all** the food and drinks you consume each day). This recommendation is very important if you also have high blood pressure. Your healthcare professional may advise an even lower target depending on your other health conditions. This means a lot more than not using a salt shaker but also limiting foods with high levels of sodium listed on their nutrition facts label. Some foods that don’t taste salty can have a surprising amount of sodium when you check their nutrition facts label. Based on the results of your blood tests, your healthcare professional or kidney dietitian may also advise you to change how much [potassium](https://www.kidney.org/atoz/content/about-potassium), [phosphorus](https://www.kidney.org/atoz/content/phosphorus), and/or calcium you might be getting through your diet.

Meeting with a dietitian can be especially helpful if you also have other health conditions like high blood pressure, diabetes, or heart failure where it is even more important to integrate a healthy diet into your lifestyle to help prevent complications. It can feel overwhelming to keep track of so many changes, and a dietitian can help you identify what works best for you.

**Lifestyle recommendations**

Now is a great time to make healthier lifestyle choices:

If you smoke and/or use tobacco products, stop. Smoking can speed up the kidney disease process and increase your risk of getting kidney failure. It also increases your risk for other serious health problems, including high blood pressure, heart disease, cancers, and stroke.

Exercise regularly. Remember, it’s okay to start slowly – taking short walks is a great way to begin.

Sleeping well is important, too. Try to get enough sleep so you are well-rested.

If you are [overweight](https://www.kidney.org/atoz/content/obesity), losing weight through a balanced diet and physical activity can help improve your health in many ways.

Find ways to [reduce and manage stress](https://www.kidney.org/atoz/content/Stress_and_your_Kidneys) in your life.

**Other ways to lower your risk**

Taking steps to manage other health conditions you may also have can also help your CKD. This includes [high blood pressure](https://www.kidney.org/atoz/what-high-blood-pressure), [diabetes](https://www.kidney.org/atoz/content/diabetes), and high cholesterol.

People with CKD should also avoid certain pain medicines known as **non-steroid anti-inflammatory drugs (NSAIDs)**. These can be harmful to your kidneys, especially at higher doses and/or with long-term use. Some examples include:

ibuprofen (Motrin, Advil)

indomethacin (Indocin)

naproxen (Aleve, Naprosyn)

diclofenac tablets or capsules (Cata flam, Zipsor)

celecoxib (Celebrex)

meloxicam (Mobic)

aspirin (only if more than 325 mg per day)

Many of these NSAID medicines are available over the counter (OTC) and may be sold under a different name or be mixed with other ingredients (like cough & cold medicines). Sometimes it may not be possible to avoid using these products depending on your other health conditions. Always ask your healthcare professional before using any products with these drug names or if the word “NSAID” is printed on the product’s label. In general, acetaminophen, also called Tylenol, is safe for your kidneys at recommended doses - but check with your healthcare professional first to determine the cause of your pain and the best way to treat it.

If your healthcare professional says you have [metabolic acidosis](https://www.kidney.org/atoz/content/metabolic-acidosis), increasing the number of fruits and vegetables you eat everyday can reduce the effect.

**Questions to ask**

What are my eGFR and uACR numbers? What is my CKD stage?

How high is my level of risk for developing heart disease or a stroke? What can I do to lower my risk?

When should I have my eGFR and uACR tested again?

Am I at a healthy weight?

Is my blood pressure within the recommended goal range?

Do I have diabetes or prediabetes? If so, is my A1C within the recommended goal range?

Do I have albuminuria?

Are there any changes I should make to my diet?

Should I take any medication(s) to help lower my risk for CKD getting worse?

**EPIDEMIOLOGY**

Even though robust knowledge of CKD epidemiological studies remains inadequate, the reported prevalence of CKD in Asia, which accounts for more than half of the global population (60%), is among the highest in the world. The Thai Screening and Early Evaluation of Kidney Disease (SEEK) study found an 18% prevalence of KD, which was higher than previous estimates of 14%. In Malaysia, the prevalence has been reported to be approximately 10%, although, in China, estimates vary from 10% to 19% in the Tibetan regions. In addition, one of the largest population-based prevalence studies to date included 47,204 individuals from various regions of China, and it reported a prevalence of 11%, equating to nearly 120 million people living with CKD in China. In Southeast Asia, numerous studies from Bangladesh, India, and Pakistan have reported CKD prevalence estimates near or exceeding 20% in some communities, and the prevalence appears to be between 10% and 20% in Nepal and Sri Lanka. The global age-standardized CKD prevalence is 10.4% in men and 10% in women, hence, developing nations having a higher prevalence than developed countries. Recent systematic reviews reported a prevalence of 13.9%and 10.1% in sub-Saharan Africa (SSA). West Africa had the highest pooled prevalence of CKD on the continent, at 16%.

Importantly, CKD is characterized in Africa by the young age of patients, with significant morbidity and premature deaths; and approximately 90% of CKD patients die within 90 days of procuring dialysis. CKD prevalence in sub-Saharan Africa may be comparable to or exceed that of many high-income countries in several countries. Although data are limited and imperative, several community-based surveys from the Democratic Republic of Congo, Tanzania, Ghana, and Senegal have since reported prevalence estimates ranging from 5% to 17%, as well as very low awareness.

Despite Nigeria's large population (over 200 million people), however, little is known about the epidemiological data of CKD in the general population. Yet, the peak prevalence of CKD has been observed to be between the third and fifth decades of life, contributing to a labor shortage and economic waste, and the incidence of CKD in Nigeria has been found to range between 1.6% and 12.4%, respectively. CKD in Nigeria determined that ESRD cases accounted for 8% of all medical admissions and 42% of renal admissions in a teaching hospital in the southeast of Nigeria. These studies, however, are hospital-based and may have excluded many patients who do not have access to hospital care; and there is no national data on CKD prevalence; and only a few community-based studies have been undertaken in some regions of the country.

A recent systematic review identified seven population-based studies, five from the Southern part of the country and two from the Northern part, and the prevalence of CKD ranged from 2.5% to 26%. A similar study conducted in South-East Nigeria found a prevalence of 11.4% in rural dwellers and 11.7% in semi-urban dwellers. Besides, another study from North-West Nigeria in a recent review documented CKD prevalence of 26%, implying an underlying high prevalence of CKD and clearly stating the need for more studies.

Several studies in recent decades have reported that KD risk factors are associated with a vast multitude of biological and social factors such as genetic, clinical factors, sociodemographic, and environmental risk factors of KD. In most societies around the world, the prevalence of KD is consistently related to socially defined conditions. This phenomenon is well documented in developed nations, where the disease affects racial/ethnic minorities and persons with poor socioeconomic position. Racial and ethnic minorities (e.g., African Americans in the United States) are disproportionately affected by advanced and progressive KD, as demonstrated by several extensive data. The relationship between socioeconomic status and the likelihood of developing CKD and eventually kidney failure has also been amply studied, with individuals with a lower socioeconomic position bearing the brunt of the burden.CKD outcomes have also been attributed to clinical health problems such as diabetes and hypertension, as well as lifestyle habits such as dietary behaviors.

According to studies, a high prevalence of KD is associated with a lack of kidney health services and health insurance coverage for KD patients, particularly in low-resource nations like Nigeria. Although a lot of clinical- and community-based studies have documented public KD knowledge level and risk-inducing lifestyles in Nigeria and in Lagos State.

### **When to see a doctor**

Make an appointment with your doctor if you have signs or symptoms of kidney disease. Early detection might help prevent kidney disease from progressing to kidney failure.

If you have a medical condition that increases your risk of kidney disease, your doctor may monitor your blood pressure and kidney function with urine and blood tests during office visits. Ask your doctor whether these tests are necessary for you.

### **What does it feel like when something is wrong with your kidneys?**

You typically don’t have any signs of kidney disease, especially in its early stages. Once you begin having symptoms, the first sign something is wrong may involve swelling in your hands and feet, itchy skin or needing to pee more often. Since symptoms vary, it’s best to call your healthcare provider if you believe there’s something wrong.

**Is kidney disease hereditary?**

Yes, kidney disease can run in biological families. Risk factors for CKD, like diabetes, also tend to run in families.

#### **What is kidney dialysis?**

Because there’s no cure for CKD, if you’re in end-stage kidney disease, you and your healthcare team must consider additional options. Complete kidney failure will result in death if it’s left untreated. Options for end-stage kidney disease include dialysis and kidney transplantation.

Dialysis is a procedure that uses machines to remove waste products from your body when your kidneys are no longer able to perform this function. There are two major types of dialysis:

* **Hemodialysis:** With hemodialysis, your blood is circulated through a machine that removes waste products, excess water and excess salt. The blood is then returned to your body. Hemodialysis requires four-hour treatments, three times a week.
* **Peritoneal dialysis:** In peritoneal dialysis, a dialysis solution is placed directly into your abdomen through a catheter. The solution absorbs waste and then is removed via the same catheter. Fresh solution is added to continue the process of cleaning. You can perform this type of dialysis yourself. There are two types of peritoneal dialysis: continuous ambulatory peritoneal dialysis (CAPD), which involves a change in dialysis solution four times a day; and continuous cycling peritoneal dialysis (CCPD). CCPD uses a machine to automatically fill, remove and refill the fluid during the nighttime.

#### **What is a kidney transplant?**

Kidney transplantation involves replacing an unhealthy kidney with a healthy kidney. Kidneys for transplantation come from two sources: living donors and deceased donors. Living donors are usually family members, partners or friends. A living kidney donor is possible because a person can live well with one healthy kidney.

Deceased donor kidneys usually come from people who are organ donors. All donors are carefully screened to make sure there’s a suitable match and to prevent any transmissible diseases or other complications.

On average, people wait about three to five years for a kidney from a deceased donor. It’s usually quicker to receive a kidney from a living donor.

**Can kidney disease be prevented?**

Seeing your healthcare provider on a regular basis throughout your life is a good start for preventing kidney disease. About 1 in every 3 people in the United States is at risk for kidney disease. People at high risk may have regular tests to check for CKD so it’s detected as early as possible. Some other things you can do to prevent CKD are:

* Manage your high blood pressure.
* Manage your blood sugar if you have diabetes.
* Eat a well-balanced diet.
* Don’t smoke or use tobacco.
* Be active for 30 minutes at least five days a week.
* Maintain a healthy weight.
* Take nonprescription pain relievers only as directed. Taking more than directed can damage your kidneys.
* Limit alcohol-containing beverages.

**Outlook / Prognosis**

If you have kidney disease, you can still live a productive home and work life and enjoy time with your family and friends. To have the best outcome possible, it’s important for you to become an active member of your treatment team.

Early detection and appropriate treatment are important in slowing the disease progression, with the goal of preventing or delaying kidney failure. You’ll need to keep your medical appointments, take your medications as prescribed, stick to a nutritious diet and monitor your blood pressure and blood sugar.

### **How long can someone live with chronic kidney disease?**

While CKD can lead to death, many people with the condition live long and happy lives after diagnosis. Most people who seek treatment for kidney disease and manage their condition never progress to kidney failure or death. That’s why it’s important to attend all your checkups and work with your healthcare provider on a treatment plan.

The leading cause of death in people with CKD is actually heart disease, a complication of CKD. Managing other health conditions that negatively impact your kidneys is also key to maintaining your kidney function.

### **Additional Common Questions**

### **What foods are bad for kidneys?**

In people with healthy kidneys, there aren’t necessarily bad foods or foods that hurt your kidneys. But, if you have CKD, your healthcare provider may recommend a kidney-friendly diet. Elements of a kidney-friendly diet may include:

* **Avoiding foods that are high in salt**. This also helps control blood pressure.
* **Eating the right amount of protein**. Protein creates more waste than other food groups. So, since your kidneys remove waste, lowering protein can help preserve their function.
* **Eating heart-healthy foods**.
* **Eating foods low in phosphorus**. This includes fresh fruits and vegetables and whole grains. Foods like dairy and beans are high in phosphorus.
* **Avoiding foods high in potassium** like bananas, oranges and potatoes.

Since following a kidney-friendly diet is hard to understand and to do, it’s always a good idea to consult a dietitian as part of your treatment plan. They can help make sure you’re eating the right types of food if you have chronic kidney disease.

### **What color is urine when your kidneys are failing?**

Your pee shouldn’t change color, but may be foamy or frothy, which means there’s excess protein in your pee. Excess protein means your kidneys aren’t filtering toxins from your body.

## **Differential Diagnosis**

## When evaluating a patient for CKD, it is crucial to consider other potential diagnoses that may present with similar symptoms and clinical findings. The differential diagnoses include:

* Acute kidney injury
* Alport syndrome
* Anti Glomerular basement membrane disease
* Diabetic nephropathy
* Multiple myeloma
* Nephrolithiasis
* Rapidly progressive glomerulonephritis
* Renal artery stenosis

## **Acute Kidney Injury (AKI)**

A sudden decline in kidney function occurring over hours to days, characterized by impaired waste elimination, fluid and electrolyte imbalance, and accumulation of nitrogenous wastes (azotemia). AKI may present with reduced urine output (oliguria), fluid overload, electrolyte disturbances, metabolic acidosis, and symptoms such as fatigue, nausea, and confusion. Causes are classified as prerenal (e.g., hypovolemia), intrinsic renal (e.g., acute tubular necrosis, glomerulonephritis), and postrenal (e.g., obstruction). Diagnosis involves serum creatinine, urine studies, imaging, and sometimes biopsy. Treatment focuses on addressing the underlying cause, supportive care, and dialysis if needed.

**Alport Syndrome**

A rare inherited disorder caused by mutations in genes encoding type IV collagen (*COL4A3*, *COL4A4*, *COL4A5*), essential for the structural integrity of glomerular basement membranes (GBM). This leads to progressive damage of the glomeruli, resulting in hematuria (often microscopic), proteinuria, hypertension, and eventual chronic kidney disease progressing to end-stage renal disease. Associated features include sensorineural hearing loss and ocular abnormalities (e.g., anterior lenticonus). Diagnosis is based on clinical features, family history, urine tests, kidney biopsy, and genetic testing. Management includes blood pressure control (ACE inhibitors) and supportive care; kidney transplantation is required in advanced disease.

**Anti-Glomerular Basement Membrane (Anti-GBM) Disease**

An autoimmune disorder characterized by circulating antibodies targeting the glomerular and alveolar basement membranes, causing rapidly progressive glomerulonephritis and pulmonary hemorrhage (Goodpasture syndrome). It presents with hematuria, proteinuria, rapidly declining renal function, and hemoptysis. Diagnosis is confirmed by detection of anti-GBM antibodies and kidney biopsy showing crescentic glomerulonephritis. Treatment includes plasmapheresis, immunosuppression with corticosteroids and cyclophosphamide.

**Diabetic Nephropathy**

A microvascular complication of diabetes mellitus characterized by progressive glomerular damage due to hyperglycemia-induced metabolic and hemodynamic changes. Clinical features include persistent albuminuria, declining glomerular filtration rate, hypertension, and eventual end-stage renal disease. Histologically, it shows mesangial expansion, glomerular basement membrane thickening, and nodular glomerulosclerosis (Kimmelstiel-Wilson lesions). Management includes glycemic and blood pressure control, use of ACE inhibitors or ARBs, and lifestyle modifications.

**Multiple Myeloma**

A plasma cell malignancy that can cause kidney injury through several mechanisms including light chain cast nephropathy, amyloidosis, and hypercalcemia. Renal impairment manifests as elevated creatinine, proteinuria (Bence Jones proteins), and electrolyte disturbances. Diagnosis involves serum and urine protein electrophoresis, bone marrow biopsy, and imaging. Treatment targets the underlying malignancy.

**Nephrolithiasis**

The formation of stones (calculi) within the kidneys, often causing flank pain, hematuria, and urinary obstruction. Stones may be composed of calcium oxalate, uric acid, struvite, or cystine. Diagnosis is by imaging (ultrasound, CT scan). Management includes pain control, hydration, and sometimes surgical removal or lithotripsy.

**Rapidly Progressive Glomerulonephritis (RPGN)**

A clinical syndrome of rapid loss of renal function over days to weeks, often associated with crescentic glomerulonephritis on biopsy. Causes include anti-GBM disease, ANCA-associated vasculitis, and severe immune complex glomerulonephritis. Presents with hematuria, proteinuria, hypertension, and oliguria. Requires urgent immunosuppressive therapy.

**Renal Artery Stenosis**

Narrowing of one or both renal arteries, most commonly due to atherosclerosis or fibromuscular dysplasia, leading to renal ischemia and secondary hypertension (renovascular hypertension). It can cause progressive renal impairment. Diagnosis is by Doppler ultrasound, CT or MR angiography. Treatment includes medical management of hypertension and revascularization procedures in selected cases.

**RECENT GUIDELINES**

Top 10 Takeaways for People Living with CKD from the KDIGO 2024 Clinical Practice Guideline for the Evaluation and Management of Chronic Kidney Disease Combining a healthy diet, exercise, stopping smoking, controlling weight, and lowering blood pressure and lipids with appropriate medications can slow down how quickly CKD progresses and reduce the risks of kidney failure, heart attack, stroke, and heart failure .

Comprehensive care Your doctor may do additional tests to understand the cause of your kidney disease and, based on the level of GFR and amount of protein in your urine, will determine your stage of CKD (severity and risk). CKD diagnosis and staging

Eating a balanced, healthy diet that suits reduced kidney function may bene t complications related to CKD

• Consume diets higher in vegetables, fruits, whole grains, ber, legumes, plant-based proteins, unsaturated fats, and nuts; and lower in processed meats, and ultra processed foods which are high in salt, sugars and carbohydrates.

• Salt intake should be <5 grams per day (equivalent to 2 grams of sodium) and protein ≤0.8 grams of protein per kg bodyweight per day.

• Initially, potassium rich foods like fruits are encouraged but as kidney function deteriorates, diet will need to be reviewed to prevent heart rhythm issues.

• Diet should be reviewed with a dietician or nutrition provider and monitored on a regular basis. Healthy and diverse diet

The best care for you is a holistic approach built by knowledge and trust in your healthcare team. Have the confidence to discuss your physical, social and emotional health and lifestyle issues with them. You will have different priorities depending on where you are in your lifespan. Holistic approach to care BP control is at the heart of CKD care and can be controlled by BP-lowering therapy customized for you together with treatment targets that are adjusted if you are frail, at high risk of falls, have very limited life expectancy, or suffer from low blood pressure or dizziness when you stand up. As kidney function falls, BP can be elevated by water overload from eating and drinking as the kidney produces reduced levels of urine, usually identified by swelling at the ankles.

Use of diuretic drugs (water pills) help increase water excretion. BP control There is significant evidence to support drugs shown to slow down or limit progression of kidney and cardiovascular disease.

• RASi (ACEi or ARBs) have kidney protective effects and should be given if you have high BP or albuminuria

• SGLT2i reduce the risk of kidney failure and cardiovascular disease and have proven benefit in CKD with and without diabetes

• Moderate- or high-intensity statins are also part of first-line therapy RASi, SGLT2i, Statins

Understanding your condition and creating a partnership through shared decision-making with your healthcare team will be of great benefit. Become a proactive and empowered patient. This will help you protect kidney function and reduce the risk of side effects.

• Ask for access to your test results and understand your parameters.

• Understand each medication you are required to take and why you are taking it.

• Ask for a medication review if you feel you are overwhelmed by the number of pills you must take.

• Ask for clear medical advice on drug therapy during acute episodes of illness especially dehydration.

• For healthcare team appointments, write down questions to ask about your care.

• Understand your risk factors and what are good lifestyle choices, now and in the long term.

• Research and ask for early education about dialysis and transplantation options. Positive approach by patients to managing CKD As CKD progresses, patients report more symptoms as the kidney struggles to clear toxins and water, and to send the correct signals to your endocrine system. Having a positive partnership with your healthcare team can help identify your symptom burden, and nd possible treatment strategies that are best for you

• Tiredness can be very challenging, so reduce stress by setting small, daily goals to help manage what needs to be done. Symptoms of tiredness from anemia may require a regimen of iron and possibly epoetin.

• Losing appetite is very common from acidosis and can limit healthy eating, so research meals you can enjoy taken in smaller portions and with less uid intake.

• Gout can be very debilitating and painful and needs urgent and long-term drug control. Symptom control kidney disease is challenging and can be isolating, but coping strategies may allow you to achieve life priorities while living with kidney disease. You may struggle to appreciate medical advice if you are struggling with mental health or social, welfare, and financial issues.

• Identify specialist welfare and financial advice services

• Ask or join kidney charities and patient groups who can offer a range of help, grants, counselling and lived experience.

• CKD treatment is universally agreed, so using trusted websites from all health care systems can help educate and answer many common patient questions.

Coping strategies for patients Untreated CKD can lead to problems with heart function, blood vessel health, diabetes, and bone issues. It is important to work with your team to help them keep you in the best health possible. Understand long-term risks Promote participation in high-quality research in CKD across the lifespan Sick Identities dehydrating illness Recalls or retrieves list of pills to stop Identities and stops pills Recovers and resumes pills Plant-based foods Absorption rate 50%–60%

Plant-based foods may have low absorption rate, net alkalizing and carbohydrate content encourages K+ shifts into intracellular space, minimizing impacts on serum K+

Animal-based foods Absorption rate 70%–90% Animal-based protein has higher absorption and net acid results in higher amounts of K+ remaining in serum

Processed foods Absorption rate 90% Potassium salts (often found in processed foods) absorption rate has been reported to be 90% Animal proteins Meat, poultry, seafood, eggs: 28 g (1 oz) = 6–8 g protein 1 egg = 6–8 g protein Dairy, milk, yogurt, cheese: 250 ml (8 oz) = 8–10 g protein 28 g (1 oz) cheese = 6–8 g protein

Plant proteins Legumes, dried beans, nuts, seeds: 100 g (0.5 cup) cooked = 7–10 g protein Whole grains, cereals: 100 g (0.5 cup) cooked = 3–6 g protein Starchy vegetables, breads: 2–4 g protein Decreased appetite Prevalence 42% Severity score: 19.8 Leg swelling Prevalence 45% Severity score: no data Fatigue Prevalence 70% Severity score: 22.8 Shortness of breath Prevalence 42% Severity score: 15 Muscle cramps Prevalence 46% Severity score: no data Heartburn Prevalence 46% Severity score: no data Drowsiness Prevalence 53% Severity score: 22.5 Pain Prevalence 53% Severity score: 22.5 Poor mobility Prevalence 56% Severity score: 19 Bone/joint pain Prevalence 55% Severity score: no data Poor sleep Prevalence 49% Severity score: 23.8 Sexual dysfunction Prevalence 48% Severity score: 56.4 Itching Prevalence 46% Severity score: 25 Healthy diet Weight management Stop use of tobacco products Physical activity SGLT2i continue until dialysis or transplant Aim for SBP <120 mm Hg RAS inhibitor\* at maximum tolerated dose (if HTN) Statin-based therapy moderate- or high-intensity statin Manage hyperglycemia as per the KDIGO Diabetes Guideline, including use of GLP-1 RA where indicated Use ns-MRA in people with diabetes and an indication for use Dihydropyridine CCB and/or diuretic if needed to achieve individualized BP target Antiplatelet agent for clinical ASCVD risk, lipids BP Lifestyle First-line drug therapy for most patients + Targeted therapies for complications Steroidal MRA if needed for resistant hypertension if eGFR ≥45 Ezetimibe, PCSK9i indicated based on ASCVD risk and lipids Manage anemia, CKD-MBD, acidosis, and potassium abnormalities, where indicated Use the same principles to diagnose and manage ASCVD and atrial fibrillation as in people without CKD Regular risk factor reassessment (every 3–6 months) ACEi, angiotensin-converting enzyme inhibitor; ARB, angiotensin II receptor blocker; ASCVD, atherosclerotic cardiovascular disease; BP, blood pressure; CCB, calcium channel blocker; CKD, chronic kidney disease; CKD-MBD, chronic kidney disease-mineral and bone disorder; eGFR, estimated glomerular alteration rate; GLP-1 RA, glucagon-like peptide-1 receptor agonist; HTN, hypertension; KDIGO, Kidney Disease: Improving Global Outcomes; MRA, mineralocorticoid receptor antagonist; ns-MRA, nonsteroidal mineralocorticoid receptor antagonist; PCSK9i, proprotein convertase subtilisin/Kexin type 9 inhibitor; RAS(i), renin-angiotensin system

**REFERENCE**

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<https://www.mayoclinic.org/diseases-conditions/chronic-kidney-disease/symptoms-causes/syc-20354521>

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# **End Stage Renal Disease (ESRD)**

## **End-stage renal failure, also known as end-stage renal disease (ESRD)**, is the final, permanent stage of chronic kidney disease, where kidney function has declined to the point that the kidneys can no longer function on their own. A patient with end-stage renal failure must receive dialysis or kidney transplantation to survive for more than a few weeks. Patients may experience a wide variety of symptoms as kidney failure progresses. These include fatigue, drowsiness, decrease in urination or inability to urinate, dry skin, itchy skin, headache, weight loss, nausea, bone pain, skin and nail changes and easy bruising. Doctors can diagnose the disease with blood tests, urine tests, kidney ultrasound, kidney biopsy, and CT scan. According to the National Center for Chronic Disease Prevention and Health Promotion, about 30 million people, or 15% of adults, in the U.S. are estimated to have chronic kidney disease. Chronic kidney disease can often be treated before it progresses to end-stage renal failure or leads to other health problems. Some of the risk factors for developing chronic kidney disease—that could ultimately lead to end-stage renal failure—include diabetes, high blood pressure, heart disease, drug abuse, blockages in the urinary tract, family history, inflammation, and some genetic disorders. Additionally, having chronic kidney disease and not properly managing it can cause the disease to progress to the point that it becomes end-stage.

## **Causes**

Kidney disease occurs when a disease or condition impairs kidney function, causing kidney damage to worsen over several months or years. For some people, kidney damage can continue to progress even after the underlying condition is resolved.

Diseases and conditions that can lead to kidney disease include:

* Type 1 or type 2 diabetes
* High blood pressure
* Glomerulonephritis (gloe-mer-u-low-nuh-FRY-tis) — an inflammation of the kidney's filtering units (glomeruli)
* Interstitial nephritis (in-tur-STISH-ul nuh-FRY-tis), an inflammation of the kidney's tubules and surrounding structures
* Polycystic kidney disease or other inherited kidney diseases
* Prolonged obstruction of the urinary tract, from conditions such as enlarged prostate, kidney stones and some cancers
* Vesicoureteral (ves-ih-koe-yoo-REE-tur-ul) reflux, a condition that causes urine to back up into your kidneys
* Recurrent kidney infection, also called pyelonephritis (pie-uh-low-nuh-FRY-tis)

**Risk factors**

Certain factors increase the risk that chronic kidney disease will progress more quickly to end-stage renal disease, including:

* Diabetes with poor blood sugar control
* Kidney disease that affects the glomeruli, the structures in the kidneys that filter wastes from the blood
* Polycystic kidney disease
* High blood pressure
* Tobacco use
* Black, Hispanic, Asian, Pacific Islander or American Indian heritage
* Family history of kidney failure
* Older age
* Frequent use of medications that could be damaging to the kidney

**Symptoms**

Early in chronic kidney disease, you might have no signs or symptoms. As chronic kidney disease progresses to end-stage renal disease, signs and symptoms might include:

* Nausea
* Vomiting
* Loss of appetite
* Fatigue and weakness
* Changes in how much you urinate
* Chest pain, if fluid builds up around the lining of the heart
* Shortness of breath, if fluid builds up in the lungs
* Swelling of feet and ankles
* High blood pressure (hypertension) that's difficult to control
* Headaches
* Difficulty sleeping
* Decreased mental sharpness
* Muscle twitches and cramps
* Persistent itching
* Metallic taste

Signs and symptoms of kidney disease are often nonspecific, meaning they can also be caused by other illnesses. Because your kidneys can make up for lost function, signs and symptoms might not appear until irreversible damage has occurred.

**Acute** (Symptoms of acute renal failure depend largely on the underlying cause.):

Hemorrhage

Fever

Weakness

Fatigue

Rash

Diarrhea or bloody diarrhea

Poor appetite

Severe vomiting

Abdominal pain

Back pain

Muscle cramps

No urine output or high urine output

History of recent infection (a risk factor for acute renal failure)

Pale skin

Nosebleeds

History of taking certain medications (a risk factor for acute renal failure)

History of trauma (a risk factor for acute renal failure)

Swelling of the tissues

Inflammation of the eye

Detectable abdominal mass

Exposure to heavy metals or toxic solvents (a risk factor for acute renal failure)

**Chronic:**

Poor appetite

Vomiting

Bone pain

Headache

Insomnia

Itching

Dry skin

Malaise

Fatigue with light activity

Muscle cramps

High urine output or no urine output

Recurrent urinary tract infections

Urinary incontinence

Pale skin

Bad breath

Hearing deficit

Detectable abdominal mass

Tissue swelling

Irritability

Poor muscle tone

Change in mental alertness

Metallic taste in mouth

The symptoms of acute and chronic renal failure may resemble other conditions or medical problems. Always consult your doctor for a diagnosis.

**DIAGNOSIS**

To diagnose end-stage renal disease, your health care provider may ask you about your family's and your medical history. You may also have physical and neurological exams, along with other tests such as:

* **Blood tests,** to measure the amount of waste products, such as creatinine and urea, in your blood
* **Urine tests,** to check the level of the protein albumin in your urine
* **Imaging tests,** such as ultrasound, MRI or CT scan, to assess your kidneys and look for unusual areas
* **Removing a sample of kidney tissue (biopsy),** to examine under a microscope to learn what type of kidney disease you have and how much damage there is

Certain tests might be repeated over time to help your provider follow the progress of your kidney disease.

### **Stages of kidney disease**

There are five stages of kidney disease. To determine what stage you have, your health care provider performs a blood test to check your glomerular filtration rate (GFR). The GFR measures how much blood the kidneys filter each minute, recorded as milliliters per minute (mL/min). As the GFR declines, so does your kidney function.

When your kidneys no longer work at a level that's necessary to keep you alive, you have end-stage renal disease. End-stage renal disease usually occurs when kidney function is less than 15% of typical kidney function.

As a part of kidney disease staging, your provider also might test whether you have protein in your urine.

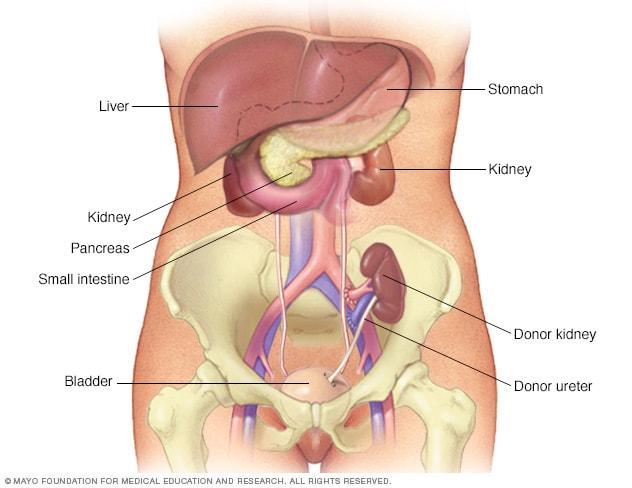
| **Kidney disease stage** | **GFR, mL/min** | **Kidney function** |
| --- | --- | --- |
|  | | |
| **Stage 1** | 90 or above | Healthy kidney function |
| **Stage 2** | 60 to 89 | Mild loss of kidney function |
| **Stage 3a** | 45 to 59 | Mild to moderate loss of kidney function |
| **Stage 3b** | 30 to 44 | Moderate to severe loss of kidney function |
| **Stage 4** | 15 to 29 | Severe loss of kidney function |
| **Stage 5** | Less than 15 | Kidney failure |

## **Treatment**

### End-stage renal disease treatments include:

* Kidney transplant
* Dialysis
* Supportive care

### **Kidney transplant**

**Kidney transplant Enlarge** image

A kidney transplant is a surgical procedure to place a healthy kidney from a live or deceased donor into a person whose kidneys no longer function properly. A kidney transplant is often the treatment of choice for end-stage renal disease, compared with a lifetime on dialysis.

The kidney transplant process takes time. It involves finding a donor, living or deceased, whose kidney best matches your own. You then have surgery to place the new kidney in your lower abdomen and attach the blood vessels and ureter — the tube that links the kidney to the bladder — that will allow the new kidney to function.

You may need to spend several days to a week in the hospital. After leaving the hospital, you can expect frequent checkups to monitor your progress as your recovery continues. You may take several medications to help keep your immune system from rejecting your new kidney and to reduce the risk of post-surgery complications, such as infection.

After a successful kidney transplant, your new kidney filters your blood, and you no longer need dialysis.

Specific treatment for renal failure will be determined by your doctor based on:

Your age, overall health, and medical history

Extent of the disease

Type of disease (acute or chronic)

Underlying cause of the disease

Your tolerance for specific medications, procedures, or therapies

Expectations for the course of the disease

Your opinion or preference

Treatment may include:

Hospitalization

Administration of intravenous (IV) fluids in large volumes (to replace depleted blood volume)

Close monitoring of important electrolytes such as potassium, sodium, and calcium

In some cases, patients may develop severe electrolyte disturbances and toxic levels of certain waste products normally eliminated by the kidneys. Patients may also develop fluid overload. Dialysis may be indicated in these cases.

Medications (to help with growth, prevent bone density loss, and/or to treat anemia)

Diuretic therapy or medications (to increase urine output)

Specific diet restrictions or modifications

**Dialysis: What is dialysis?**

Dialysis is a procedure that is performed routinely on persons who suffer from acute or chronic renal failure, or who have ESRD. The process involves removing waste substances and fluid from the blood that are normally eliminated by the kidneys. Dialysis may also be used for individuals who have been exposed to or ingested toxic substances to prevent renal failure from occurring. There are two types of dialysis that may be performed, including the following:

**Peritoneal dialysis.** Peritoneal dialysis is performed by surgically placing a special, soft, hollow tube into the lower abdomen near the navel. After the tube is placed, a special solution called dialysate is instilled into the peritoneal cavity. The peritoneal cavity is the space in the abdomen that houses the organs and is lined by two special membrane layers called the peritoneum. The dialysate is left in the abdomen for a designated period which will be determined by your doctor. The dialysate fluid absorbs the waste products and toxins through the peritoneum. The fluid is then drained from the abdomen, measured, and discarded. There are three different types of peritoneal dialysis: continuous ambulatory peritoneal dialysis (CAPD), continuous cyclic peritoneal dialysis (CCPD), and intermittent peritoneal dialysis (IPD).  
  
CAPD does not require a machine. Exchanges, often referred to as *passes*, can be done three to five times a day during waking hours. CCPD requires the use of a special dialysis machine that can be used in the home. This type of dialysis is done automatically, even while you are asleep. IPD uses the same type of machine as CCPD, but treatments take longer. IPD can be done at home but usually is done in the hospital.  
  
Possible complications of peritoneal dialysis include an infection of the peritoneum, or peritonitis, where the catheter enters the body. Peritonitis causes fever and stomach pain. Your diet for peritoneal dialysis will be planned with a dietitian, who can help you choose meals according to your doctor's orders. Generally:

You may have special protein, salt, and fluid needs.

You may have special potassium restrictions.

You may need to reduce your calorie intake, since the sugar in the dialysate may cause weight gain.

**Hemodialysis.** Hemodialysis can be performed at home or in a dialysis center or hospital by trained health care professionals. A special type of access, called an arteriovenous (AV) fistula, is placed surgically, usually in your arm. This involves joining an artery and a vein together. An external, central, intravenous (IV) catheter may also be inserted, but is less common for long-term dialysis. After access has been established, you will be connected to a large hemodialysis machine that drains the blood, bathes it in a special dialysate solution which removes waste substances and fluid, then returns it to your bloodstream.  
  
Hemodialysis is usually performed several times a week and lasts for four to five hours. Because of the length of time hemodialysis takes, it may be helpful to bring reading material, to pass the time during this procedure. During treatment you can read, write, sleep, talk, or watch TV.  
  
At home, hemodialysis is done with the help of a partner, often a family member or friend. If you choose to do home hemodialysis, you and your partner will receive special training.  
  
Possible complications of hemodialysis include muscle cramps and hypotension (sudden drop in blood pressure). Hypotension may cause you to feel dizzy or weak, or sick to your stomach. Side effects are avoided by following the proper diet and taking medications, as prescribed by your doctor. A dietitian will work with you to plan your meals, according to your doctor's orders. Generally:

You may eat foods high in protein such as meat and chicken (animal proteins).

You may have potassium restrictions.

You may need to limit the amount you drink.

You may need to avoid salt.

You may need to limit foods containing mineral phosphorus (such as milk, cheese, nuts, dried beans, and soft drinks).

### **Palliative care**

If you choose not to have a kidney transplant or dialysis, you can choose palliative or supportive care to help you manage your symptoms and feel better. You also can combine palliative care with kidney transplant or dialysis.

Without either dialysis or a transplant, kidney failure progresses, eventually leading to death. Death can occur quickly or take months or years. Supportive care might include management of symptoms, measures to keep you comfortable and end-of-life planning.

**Lifestyle and home remedies**

As part of your treatment for kidney disease, your health care provider might recommend that you follow a special diet to help support your kidneys and limit the work they must do. Ask for a referral to a registered dietitian with expertise in kidney disease to learn ways to make your diet easier on your kidneys.

Depending on your situation, kidney function and overall health, your dietitian might recommend that you:

* **Avoid products with added salt.** Lower the amount of sodium you eat each day by avoiding products with added salt, including many convenience foods, such as frozen dinners, canned soups and fast foods. Other foods with added salt include salty snack foods, canned vegetables, and processed meats and cheeses.
* **Choose lower potassium foods.** Your dietitian might recommend that you choose lower potassium foods at each meal. High-potassium foods include bananas, oranges, potatoes, spinach and tomatoes.

Examples of low-potassium foods include apples, cabbage, carrots, green beans, grapes and strawberries. Be aware that many salt substitutes contain potassium, so you generally should avoid them if you have kidney failure.

* **Limit your protein.** Your dietitian will estimate the grams of protein you need each day and make recommendations based on that amount. High-protein foods include lean meats, eggs, milk, cheese and beans. Low-protein foods include vegetables, fruits, breads and cereals.

**Coping and support**

Learning you're in kidney failure can come as a shock, even if you've known about your kidney disease for a while. It might be difficult to manage the treatment schedule if you're on dialysis.

To help you cope, consider trying to:

* **Connect with other people who have kidney disease.** It might help you to talk to other people with end-stage renal disease. Ask your doctor about support groups in your area. Or contact organizations such as the American Association of Kidney Patients, the National Kidney Foundation or the American Kidney Fund for groups in your area.
* **Maintain your routine, when possible.** Try to continue to work and do the activities you enjoy, if your condition allows.
* **Be active most days of the week.** With your provider's approval, aim for at least 30 minutes of physical activity most days of the week. This can help you with fatigue and stress.
* **Talk with someone you trust.** It might help to talk about your feelings with a friend or family member, a faith leader, or someone else you trust. Your provider might be able to recommend a social worker or counselor.

**Preparing for your appointment**

For end-stage renal disease, you'll likely continue to see the same health care provider and care team you've been seeing for treatment of chronic kidney disease. If you're not already being cared for by a doctor who specializes in kidney problems (nephrologist), you might be referred to one as your disease progresses.

### **What you can do**

To get ready for your appointment, ask if there's anything you need to do ahead of time, such as make changes to your diet. Then take note of:

* **Your symptoms,** including any that seem unrelated to your kidneys or urinary function, and when thy began
* **All your medications and doses,** vitamins or other supplements you take
* **Your key medical history,** including other medical conditions and family history of kidney disease
* **Questions to ask your provider**

Take a family member or friend along, if possible, to help you remember the information you're given.

For end-stage renal disease, some basic questions to ask your provider include:

* What's the level of damage to my kidneys?
* Is my kidney function worsening?
* Do I need more tests?
* What's causing my condition?
* Can the damage to my kidneys be reversed?
* What are my treatment options?
* What are the potential side effects of each treatment?
* I have these other health conditions. How can I best manage them together?
* Do I need to eat a special diet?
* Can you refer me to a dietitian who can help me plan my meals?
* Are there brochures or other printed material I can have? What websites do you recommend?
* How often do I need to have my kidney function tested?

Don't hesitate to ask any other questions you have.

### **What to expect from your healthcare provider**

Your provider may ask you questions, such as:

* Have you noticed changes in your urinary habits or unusual fatigue?
* Have you been diagnosed or treated for high blood pressure

## **Complications**

Kidney damage, once it occurs, can't be reversed. Potential complications can affect almost any part of your body and can include:

* Fluid retention, which could lead to swelling in your arms and legs, high blood pressure, or fluid in your lungs (pulmonary edema)
* A sudden rise in potassium levels in your blood (hyperkalemia), which could impair your heart's ability to function and may be life-threatening
* Heart disease
* Weak bones and an increased risk of bone fractures
* Anemia
* Decreased sex drive, erectile dysfunction or reduced fertility
* Damage to your central nervous system, which can cause difficulty concentrating, personality changes or seizures
* Decreased immune response, which makes you more vulnerable to infection
* Pericarditis, an inflammation of the saclike membrane that envelops your heart (pericardium)
* Pregnancy complications that carry risks for the mother and the developing fetus
* Malnutrition
* Irreversible damage to your kidneys (end-stage kidney disease), eventually requiring either dialysis or a kidney transplant for survival

**Prevention**

If you have kidney disease, you may be able to slow its progress by making healthy lifestyle choices:

* Achieve and maintain a healthy weight
* Be active most days
* Limit protein and eat a balanced diet of nutritious, low-sodium foods
* Control your blood pressure
* Take your medications as prescribed
* Have your cholesterol levels checked every year
* Control your blood sugar level
* Don't smoke or use tobacco products
* Get regular checkups

**PROGNOSIS**

People with ESRD are living longer than ever. Dialysis treatments (both hemodialysis and peritoneal dialysis) are not cures for ESRD but will help you feel better and live longer. Over the years, ESRD can cause other problems such as bone disease, high blood pressure, nerve damage, and anemia (having too few red blood cells). You should discuss prevention methods and treatment options for these potential problems with your doctor.

### **When to seek care**

Make an appointment with your health care provider if you have signs or symptoms of kidney disease.

If you have a medical condition that increases your risk of kidney disease, your care provider is likely to monitor your kidney function with urine and blood tests and your blood pressure during regular office visits. Ask your provider whether these tests are necessary for you.

**EPIDEMIOLOGY**

53 patients (56.6% male), median age 11 (inter quartile range 8.5-12) years were studied. Mean annual incidence of ESRD in Ibadan for children aged 14 years and below was 4 per million age related population (PMARP) while for those aged 5-14 years it was 6.0 PMARP. Glomerulonephritis was the cause in 41 (77.4%) patients amongst whom, 29 had chronic glomerulonephritis and 12 had nephrotic syndrome. Congenital anomalies of the kidneys and urinary tract (CAKUT) accounted for 11 (21.2%) cases, posterior urethral valves being the most common. Acute hemodialysis, acute peritoneal dialysis or a combination of these were performed in 33 (62.3%), 6 (11.3%) and 4 (7.5%) patients respectively. Median survival was 47 days and in-hospital mortality were 59%. Incidence of pediatric ESRD in Ibadan is higher than previous reports from sub-Saharan Africa. Glomerulonephritis, and then CAKUT are the most common causes. Mortality is high, primarily due to lack of resources. Preventive nephrology and chronic RRT programs are urgently needed.

The incidence of CKD (chronic kidney disease) in Nigeria has been shown by various studies to range between 1.6 and 12.4%. We have shown that the burden of renal disease in Nigeria is probably significantly higher than any previous study on end-stage renal disease (ESRD) has documented, as most studies are hospital-based and fail to include the many patients who do not have access to hospital care. The increased prevalence of ESRD among blacks in the United States and South Africa compared with other races also suggests that ESRD may be more prevalent in Africa than in the United States and other developed nations. Common causes of CKD in Nigerian adults are glomerulonephritis and hypertension, while common causes in children are glomerulonephritis and posterior urethral valves. In the United States, diabetes and hypertension are the commonest causes of CKD and glomerulonephritis plays a less important role. Access to renal replacement therapy (RRT) in Nigeria is limited, and mortality rates are very high, ranging between 40 and 50%. Important steps towards improving the situation are the development of prevention programmes and increased funding to ensure increased availability of RRT. To achieve this, health policies concerning CKD must be formulated, and the lack of a renal registry makes it difficult for this to be done. There is a need for the development of a functional organizational structure for the reporting of CKD in Nigeria, the Nigerian Renal Registry.

In Africa, most cases of ESRD likely remain undiagnosed and untreated, leading to almost certain mortality. Limited aggregate data exist to accurately characterize ESKD rates, which are likely quite high, and steps to establish a continent-wide registry are ongoing. The prevalence of treated ESKD in sub-Saharan Africa (SSA) is lower than that of other developing countries (less than 100 pmp), despite comparable incidence rates, and is likely due to limited access to KRT (only ~10% of adults with incident ESKD remained on dialysis ≥ 3 months). KRT access generally requires self-funding, even in wealthier countries like South Africa, which only provides government funding for KRT if a patient is eligible for transplant.

### Global variation in kidney replacement therapy modality and practice patterns

Although kidney transplantation is the preferred treatment for eligible ESKD patients, dialysis is the predominant therapy in most countries. Considerable variation exists in access to and use of kidney transplantation. In 2013, transplantation for ESKD patients ranged from 57–72% in Nordic countries, Estonia, and the Netherlands, to less than 10% in some Asian and eastern European countries. Countries with the highest transplantation rates—mostly Nordic and several other European countries—also have some of the lowest ESKD incidence rates. In such countries, transplantation may be offered to a higher proportion of ESKD patients because of the relatively low number of incident cases. In some countries, by focusing on transplantation or home dialysis, <1/3 of ESKD patients used in-center HD. These include Hong Kong, Estonia, the Netherlands, New Zealand, and some Nordic countries. This differs from many East and Southeast Asian countries where ≥85% of patients receive in-center HD. Japan is notable because it has a large and mature ESKD program with excellent clinical outcomes, but very low transplantation and home dialysis use. In-center HD is favored over home dialysis partly for historical reasons (dialysis facilities are available and easily accessible, with many places intentionally near public transportation stops), and kidney donation rates are low, in part due to spiritual beliefs.

CKD is a global health challenge, especially in LMICs. Most people in developing countries have limited incomes and cannot afford health insurance, which risks personal financial crises from out-of-pocket medical costs for both CKD care and KRT. There is a greater prevalence of KRT among groups of people with a higher income level which is consistent with the notion that KRT access is highly dependent on health-care expenditures and discontinuation and death once resources are exhausted. In a single-center study of 320 ESKD patients initiated on maintenance HD in Nigeria, >80% of the patients funded dialysis treatments from out-of-pocket payment. Within 12 weeks of initiation, 98% had dropped out of the program through deaths and abandonment, and only 2% were able to fund treatments beyond 12 weeks.

Disparity in access to KRT is not limited to LMICs. Some of the most explicit examples of inequity are evident in undocumented immigrant ESKD care. In the US, undocumented immigrants with ESKD (currently estimated between 5,500 and 8,857) are ineligible for Medicare and coverage decisions are made at state or local levels. The two main treatment options, emergency-only hemodialysis (EOHD) and chronic outpatient dialysis, highlight the dilemma between principles of justice and societal standards. Some patients on EOHD are dialyzed once to twice weekly while others just once a month. Not surprisingly, EOHD is associated with psychosocial distress, life-threatening physical symptoms, and poor outcomes with a mean dialysis vintage of 16 months at time of death

At a community/systems level, reductions in environmental toxins (air pollutants, heavy metals, agrichemicals, contaminated water and soil), improved access to healthy foods, education and healthy living conditions by ensuring equitable access to housing and employment, health-care provider capacity building, health system organization, and government policy grounded in environmental, social and economic justice are necessary. Allocation and policy development.

**Differential Diagnosis**

The clinical features of end-stage renal disease mimic many other disorders, and many diseases lead to end-stage renal disease. Therefore, the following differentials should be considered whenever assessing a patient with end-stage renal disease.

* Chronic glomerulonephritis
* Chronic pyelonephritis
* Rapidly progressive glomerulonephritis
* Nephropathy of pregnancy/pregnancy toxemia
* Unclassifiable nephritis
* Polycystic kidney disease
* Nephrosclerosis
* Malignant hypertension
* Diabetic nephropathy
* Systemic lupus erythematosus nephritis
* Amyloidal kidney
* Gouty kidney
* Renal failure due to a congenital abnormality of metabolism
* Renal/urinary tract tuberculosis
* Renal/urinary tract calculus
* Renal/urinary tract tumor
* Obstructive urinary tract disease
* Myeloma
* Renal hypoplasia

**Chronic Glomerulonephritis**

A progressive inflammatory disease of the glomeruli characterized by irreversible scarring and fibrosis, leading to gradual loss of kidney function. It often follows acute glomerulonephritis or arises from autoimmune or infectious causes. Symptoms include fatigue, hypertension, edema, proteinuria, hematuria, and impaired renal function. Diagnosis involves urinalysis, blood tests, and kidney biopsy. Without treatment, it can progress to end-stage renal disease.

**Chronic Pyelonephriti**s

A long-standing bacterial infection of the renal pelvis and parenchyma, often due to recurrent urinary tract infections or urinary obstruction. It leads to renal scarring, tubular atrophy, and impaired kidney function. Symptoms may be subtle, including flank pain, hypertension, and urinary abnormalities. Diagnosis is by imaging and urine cultures.

**Rapidly Progressive Glomerulonephritis (RPGN)**

A clinical syndrome marked by rapid decline in renal function over days to weeks, often associated with crescent formation in glomeruli on biopsy. Causes include anti-GBM disease, ANCA-associated vasculitis, and severe immune complex glomerulonephritis. Presents with hematuria, proteinuria, hypertension, and oliguria. Requires urgent immunosuppressive treatment.

**Nephropathy of Pregnancy / Pregnancy Toxemia**

Renal impairment during pregnancy, often related to preeclampsia or eclampsia, characterized by hypertension, proteinuria, and edema. It can cause acute kidney injury and requires close monitoring and management of maternal and fetal health.

**Unclassifiable Nephritis**

Cases of nephritis that do not fit established categories due to atypical clinical or histological features. Diagnosis relies on kidney biopsy and immunological studies.

**Polycystic Kidney Disease (PKD)**

A genetic disorder characterized by the development of numerous cysts in both kidneys, leading to progressive renal enlargement, hypertension, hematuria, and eventual renal failure. Autosomal dominant PKD is the most common form.

**Nephrosclerosis**

Hardening and thickening of renal arterioles and small arteries due to chronic hypertension or aging, causing ischemic glomerular and tubular injury and gradual loss of kidney function.

**Malignant Hypertension**

A severe, rapidly progressive form of hypertension causing vascular injury including fibrinoid necrosis and hyperplastic arteriolosclerosis, leading to acute kidney injury and end-organ damage.

**Diabetic Nephropathy**

A microvascular complication of diabetes characterized by progressive glomerular damage, proteinuria, hypertension, and declining renal function. Histologically marked by mesangial expansion and nodular glomerulosclerosis.

**Systemic Lupus Erythematosus (SLE) Nephritis**

Renal involvement in SLE with immune complex deposition causing a spectrum of glomerular injuries from mild mesangial to diffuse proliferative glomerulonephritis, leading to hematuria, proteinuria, and renal impairment.

Amyloidal Kidney

Renal deposition of amyloid fibrils causing proteinuria, nephrotic syndrome, and progressive renal failure. It can be primary (AL amyloidosis) or secondary to chronic inflammation.

**Gouty Kidney**

Renal damage due to deposition of urate crystals causing interstitial nephritis, tubular obstruction, and chronic kidney disease.

**Renal Failure Due to Congenital Abnormality of Metabolism**

Kidney impairment resulting from inherited metabolic disorders affecting renal function, such as cystinosis or Fabry disease.

**Renal/Urinary Tract Tuberculosis**

A chronic granulomatous infection of the kidneys and urinary tract caused by *Mycobacterium tuberculosis*, leading to scarring, strictures, and renal dysfunction.

**Renal/Urinary Tract Calculus**

Formation of stones in the urinary tract causing obstruction, pain, hematuria, and risk of infection.

**Renal/Urinary Tract Tumor**

Benign or malignant neoplasms of the kidney or urinary tract presenting with hematuria, flank pain, or mass.

**Obstructive Urinary Tract Disease**

Blockage of urine flow due to stones, strictures, tumors, or prostatic hypertrophy, causing hydronephrosis and renal impairment.

**Myeloma**

A plasma cell malignancy causing renal impairment through light chain deposition, cast nephropathy, and hypercalcemia.

**Renal Hypoplasia**

Congenital underdevelopment of one or both kidneys resulting in reduced nephron number and potential renal insufficiency.

**REFERENCES**

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### **Nephrotic Syndrome (NS)**

### Nephrotic Syndrome (NS) is not a disease itself, but rather an umbrella term for the collection of signs and symptoms that result from damage in the kidney’s filters, called glomeruli. This damage causes protein to leak into the urine, a condition called proteinuria. The glomeruli filter blood as it passes through the kidneys, separating things the body needs from those it doesn’t. One of these things your body needs is protein. Healthy glomeruli keep blood protein (mainly albumin) from passing through and spilling into the urine. Damaged glomeruli allow too much blood protein to pass through the kidneys and leave the body in the urine, leading to Nephrotic Syndrome.

In healthy kidneys, the glomeruli filter out the waste products. They allow your blood to retain the cells and proteins your body needs to function regularly.

Damaged glomeruli allow blood proteins to leak into your pee, including albumin. If you have nephrotic syndrome, your damaged glomeruli allow three or more grams (g) of protein to leak into your pee over 24 hours. Three grams is a little heavier than a U.S. penny. It’s 20 times the amount of protein that healthy glomeruli allow.

#### **Is nephrotic syndrome life-threatening?**

Nephrotic syndrome is a serious condition. Without treatment, nephrotic syndrome can affect your life expectancy due to secondary complications.

#### **Who does nephrotic syndrome affect?**

Nephrotic syndrome can affect anyone. However, it’s most common in people with diabetes-related kidney disease, people who have allergies and people who have a biological family history of kidney disease with nephrotic syndrome.

#### **How common is nephrotic syndrome?**

Nephrotic syndrome is relatively rare.

Nephrotic syndrome occurs in about 1 in every 50,000 children each year. Most children receive a nephrotic syndrome diagnosis between the ages of 2 and 5. Boys are about twice as likely to have nephrotic syndrome as girls.

It occurs in 3 in every 100,000 adults each year.

#### **How does nephrotic syndrome affect my body?**

If you have nephrotic syndrome, losing different proteins may cause various problems.

Some proteins help prevent blood clots. When you lose those proteins in your pee, blood clots can form.

Immunoglobulins are proteins that help your immune system fight diseases and infections. When you lose immunoglobulins, you’re at a greater risk of general infections that affect different body systems, including:

* Pneumonia.
* Cellulitis.
* Peritonitis.

## **Risk factors**

Factors that can increase your risk of nephrotic syndrome include:

* **Medical conditions that can damage your kidneys.** Certain diseases and conditions increase your risk of developing nephrotic syndrome, such as diabetes, lupus, amyloidosis, reflux nephropathy and other kidney diseases.
* **Certain medications.** Medications that might cause nephrotic syndrome include nonsteroidal anti-inflammatory drugs and drugs used to fight infections.
* **Certain infections.** Infections that increase the risk of nephrotic syndrome include HIV, hepatitis B, hepatitis C and malaria.

## **Complications**

Possible complications of nephrotic syndrome include:

* **Blood clots.** The inability of the glomeruli to filter blood properly can lead to loss of blood proteins that help prevent clotting. This increases your risk of developing a blood clot in your veins.
* **High blood cholesterol and elevated blood triglycerides.** When the level of the protein albumin in your blood falls, your liver makes more albumin. At the same time, your liver releases more cholesterol and triglycerides.
* **Poor nutrition.** Loss of too much blood protein can result in malnutrition. This can lead to weight loss, which can be masked by edema. You may also have too few red blood cells (anemia), low blood protein levels and low levels of vitamin D.
* **High blood pressure.** Damage to your glomeruli and the resulting buildup of excess body fluid can raise your blood pressure.
* **Acute kidney injury.** If your kidneys lose their ability to filter blood due to damage to the glomeruli, waste products can build up quickly in your blood. If this happens, you might need emergency dialysis — an artificial means of removing extra fluids and waste from your blood — typically with an artificial kidney machine (dialyzer).
* **Chronic kidney disease.** Nephrotic syndrome can cause your kidneys to lose their function over time. If kidney function falls low enough, you might need dialysis or a kidney transplant.
* **Infections.** People with nephrotic syndrome have an increased risk of infections.

**Symptoms and Causes**

Common nephrotic syndrome symptoms include:

* Large amounts (greater than 3.5 grams) of the protein albumin in your pee (albuminuria).
* High fat and cholesterol levels in your blood (hyperlipidemia).
* Swelling (edema), usually in your legs, feet or ankles. Swelling may also occur in your hands or face.
* Low levels of albumin in your blood (hypoalbuminemia).
* Loss of appetite.
* Feeling unwell or sick.
* Abdominal pain (pain anywhere from your ribs to your pelvis).
* Foamy pee.

Another symptom of nephrotic syndrome is a loss of minerals and vitamins that are essential to your health and development, including calcium and vitamin D. In children with nephrotic syndrome, this may affect their growth. You may develop osteoporosis, which weakens your hair and nails.

**CAUSES**

Kidney diseases often damage your glomeruli. The diseases target your glomeruli, though healthcare providers and medical researchers aren’t sure why. Damaged glomeruli are the primary cause of nephrotic syndrome. These diseases include:

* **Amyloidosis**. This is a disease in which amyloid proteins build up in your vital organs. Amyloidosis most commonly occurs in your kidneys, affecting their ability to filter.
* **Diabetes-related nephropathy**. “Nephropathy” means that your kidney isn’t working properly. In diabetes-related nephropathy, diabetes causes damage or dysfunction to one or more of the nerves in your kidneys. It typically causes numbness, tingling, muscle weakness and pain in your affected area.
* **Focal segmental glomerulosclerosis (FSGS)**. In FSGS, scarring affects small areas (segments) of some of your glomeruli. It may cause swelling, kidney failure and loss of proteins in your pee.
* **Lupus**. Lupus is an autoimmune disease that causes inflammation, swelling and pain throughout your body, including your kidneys.
* **Membranous nephropathy**. In membranous nephropathy, your body’s immune system attacks the filtering membranes in your kidneys.
* **Minimal change disease (MCD)**. This is a type of nephropathy in which your kidneys aren’t working properly. However, a kidney biopsy shows little or no damage to your glomeruli or kidney tissue. MCD may occur at any age, but it’s most common in children.

## **Diagnosis and Tests**

### The following tests and procedures help diagnose nephrotic syndrome:

#### **Urinalysis tests**

A urinalysis (urine test) examines the visual, chemical and microscopic aspects of your pee. Your healthcare provider may recommend a few different types of urinalysis tests.

During a dipstick test, you’ll pee into a special container at your healthcare provider’s office or a hospital. Then, a healthcare provider will place a strip of paper coated with special chemicals (dipstick) into the container. The dipstick will change color if there’s albumin in your pee.

If your healthcare provider needs a more precise measurement, they may recommend urine protein tests. Urine protein tests may include a single urine sample or a 24-hour collection of urine.

In a single urine sample, your container is sent to a lab. Lab technicians compare how much albumin and creatinine are in your pee (albumin-to-creatinine ratio). If your urine sample has more than 30 milligrams (mg) of albumin for each gram of creatinine, it may signal a problem.

In a 24-hour urine collection, your healthcare provider will give you a container to collect your pee from home. On the day of the test, you’ll:

* Pee in the toilet as usual when you first wake up.
* Pee in the container the rest of the day until you go to sleep.
* Pee in the container one last time when you first wake up the following day.

You’ll then drop your sample off at your healthcare provider’s office or a lab. Lab technicians will only measure the amount of albumin in your sample.

#### **Blood tests**

During an albumin blood test, your healthcare provider will use a thin needle to withdraw a small amount of blood from a vein in your arm. The blood sample goes to a lab for testing. A low level of albumin or other proteins may indicate nephrotic syndrome.

Lab technicians may also test your blood cholesterol and blood triglyceride levels. Those levels may increase if your blood albumin level is low.

#### **Kidney biopsy**

During a kidney biopsy, your healthcare provider will remove a small piece of your kidney tissue to examine at a lab under a microscope.

Your healthcare provider will first numb the area with a local anesthetic, so you won’t feel any pain. They’ll also give you a light sedative to help you relax. Then, they’ll insert a needle through your skin and into your kidney to collect the tissue sample.

If you have diabetes and your healthcare provider suspects you have nephrotic syndrome, you likely won’t need a kidney biopsy. Your medical history, urine tests and blood tests are often enough to help them diagnose nephrotic syndrome because of your diabetes.

**Management and Treatment**

### Nephrotic syndrome isn’t curable. However, nephrotic syndrome often goes away in children once they reach their late teenage years or early 20s.

Treatment helps relieve your nephrotic syndrome symptoms and prevents further damage to your kidneys.

Nephrotic syndrome treatment includes addressing the underlying cause and taking steps to reduce high blood pressure, high cholesterol, swelling and infection risks. Treatment usually includes medications and changes to your diet.

Some blood pressure medications can slow down a kidney disease that causes nephrotic syndrome, including:

* Angiotensin-converting enzyme (ACE) inhibitors.
* Angiotensin receptor blockers (ARBs).

These medications reduce the pressure inside your glomeruli, which reduces albuminuria. Many people require two or more medications to regulate their blood pressure.

In addition to an ACE inhibitor or an ARB, your healthcare provider may recommend a diuretic. Diuretics help your kidneys remove fluid from your blood. Diuretics also help reduce blood pressure and swelling. Other medications that help lower your blood pressure include beta-blockers and calcium channel blockers.

To lower your cholesterol, your healthcare provider may recommend statin medications.

If you have nephrotic syndrome, it’s also a good idea to get a yearly influenza (flu) vaccine and a pneumococcal (*new-ma-cah-cole*) vaccine. A pneumococcal vaccine helps protect your body from a pneumococcus (*new-ma-cah-cuss*) bacterial infection. These bacteria may cause ear infections, pneumonia and meningitis.

In some cases, your healthcare provider may recommend medications that help prevent blood clots ([anticoagulants](https://my.clevelandclinic.org/health/treatments/22288-anticoagulants) or blood thinners). If you have nephrotic syndrome, you’ll only take these medications if you develop a blood clot. They don’t prevent nephrotic syndrome or any symptoms of nephrotic syndrome.

### **Should I avoid any foods or drinks if I have nephrotic syndrome?**

Diet and nutrition don’t cause or prevent nephrotic syndrome in adults. However, if you have nephrotic syndrome, changes to your diet may help relieve some of your symptoms.

You can help reduce swelling by limiting the amount of sodium in your diet. Most sodium in your diet comes from salt. You can also help reduce swelling by drinking more fluids.

Reducing saturated fat and cholesterol in your diet can help manage hyperlipidemia. It’s a good idea to limit foods like full-fat dairy, red meat, processed meat, cheese, fried foods, baked goods and sweets.

### **How do I take care of myself?**

The best way to manage your symptoms is to take your medications as prescribed by your healthcare provider. It’s also a good idea to maintain a diet with appropriate amounts of potassium and protein but low amounts of sodium, saturated fat and cholesterol.

#### **How soon after treatment will I feel better?**

It depends on the cause of your nephrotic syndrome. Some people may start to feel better a few days after treatment, while it may take others a few weeks or even months.

## **Prevention**

### You can’t prevent nephrotic syndrome. However, you can improve the symptoms of nephrotic syndrome by treating its cause and making changes to your diet.

**Self-care**

Changes to your diet might help with nephrotic syndrome. Your doctor might refer you to a dietitian, who might recommend that you do the following:

* Choose lean sources of protein. Plant-based protein is helpful in kidney disease.
* Reduce the amount of fat and cholesterol in your diet to help control your blood cholesterol levels.
* Eat a low-salt diet to help control swelling.
* Reduce the amount of liquid in your diet.

## **WHEN TO SEE A DOCTOR**

Start by seeing your primary care doctor. If your doctor suspects you or your child has a kidney problem, such as nephrotic syndrome, you might be referred to a doctor who specializes in the kidneys (nephrologist).

Here's some information to help you get ready for your appointment.

### **What you can do**

When you make the appointment, ask if there's anything you need to do in advance, such as restrict your diet. Take a family member or friend along, if possible, to help you remember the information you'll be given.

Make a list of:

* **Your or your child's symptoms** and when they began
* **Key personal information,** including major stresses or recent life changes
* **All medications, vitamins or other supplements** you or your child takes, including doses
* **Questions to ask** your doctor

For nephrotic syndrome, some questions to ask include:

* What's the most likely cause of my or my child's nephrotic syndrome?
* What tests do I or my child need?
* Is this condition likely temporary?
* What are the treatment options? And which do you recommend?
* Are there changes I can make to my or my child's diet? Could consulting a dietitian help?
* How can I best manage this condition with my or my child's other medical conditions?
* Are there brochures or other printed material that I can have? What websites do you recommend?

### **What to expect from your doctor**

Your doctor is likely to ask you questions, such as:

* Do symptoms come and go, or do you have them all the time?
* How severe are the symptoms?
* Does anything seem to improve the symptoms?
* What, if anything, appears to worsen the symptoms?

**Outlook / Prognosis**

With proper diagnosis and treatment, the outcome for people with nephrotic syndrome is good. Most people respond well to treatment, and nephrotic syndrome often goes into remission.

If you have nephrotic syndrome as a child, it often goes away by your early adult years. Contact your healthcare provider as soon as you notice any kidney symptoms that last longer than a few days, especially if your pee is foamy.

**What questions should I ask my healthcare provider?**

* How do you know that I have nephrotic syndrome?
* Do I need to take any tests to confirm your diagnosis?
* If I don’t have nephrotic syndrome, what other condition might I have?
* What condition caused me to develop nephrotic syndrome?
* Do you think my nephrotic syndrome will go into remission with treatment?
* Do you think my child’s nephrotic syndrome will go away when they get older?
* What medications or treatments do you recommend?
* Should I make any changes to my diet?

**EPIDEMIOLOGY**

Childhood nephrotic syndrome, if left untreated, leads to progressive kidney disease or death. We quantified the prevalence of steroid-sensitive nephrotic syndrome, steroid-resistant nephrotic syndrome, and histological types as the epidemiology of nephrotic syndrome in Africa remains unknown yet impacts outcomes. Methods: Primary outcomes included steroid response, biopsy defined minimal change disease, and focal segmental glomerulosclerosis (FSGS) by both pooled and individual proportions across regions and overall. Findings: There were 81 papers from 17 countries included. Majority of 8131 children were steroid-sensitive (64% [95% CI: 63–66%]) and the remaining were steroid-resistant (34% [95% CI: 33–35%]). Of children biopsied, pathological findings were 38% [95% CI: 36–40%] minimal change, 24% [95% CI: 22–25%] FSGS, and 38% [95% CI: 36–40%] secondary causes of nephrotic syndrome. Interpretation: Few African countries reported on the prevalence of childhood nephrotic syndrome. Steroid-sensitive disease is more common than steroid-resistant disease although prevalence of steroid-resistant nephrotic syndrome is higher than reported globally.

Nephrotic syndrome is an important chronic disease in children. The estimated annual incidence of nephrotic syndrome in healthy children is two to seven new cases per 100,000 children less than 18 years of age. It is more common in boys than girls at younger ages, but once adolescence is reached, there is no significant difference between genders. Increased incidence and more severe diseases are seen in African American and Hispanic populations.

We will look at the statistics from different regions of the world.

**United States Statistics**

Diabetic nephropathy associated with nephrotic syndrome is most common, with an estimated rate of around 50 cases per million population. In the pediatric population, nephrotic syndrome could occur at a rate of 20 cases per million.

**International Statistics**

In India and Turkey, biopsy results in children with nephrotic syndrome have revealed similar histology types compared to what would be expected in Western countries. In Pakistani adult patients with nephrotic syndrome, the histological patterns of kidney biopsies are like those seen in western countries.

In parts of the Middle East and Africa, glomerular diseases have also been linked with urogenital schistosomal infection. However, tropical nephrotic syndrome due to parasitic diseases such as malaria or schistosomiasis may be non-existent.

Doe et al. reported causes of nephrotic syndrome in the African pediatric population where kidney biopsy most often revealed typical histologic findings, such as minimal change disease and focal and segmental glomerulosclerosis. Nephrotic syndrome due to quartan malaria is not a very well-established phenomenon. In Congo, Pakasa and Sumaili call attention to the fall of parasite-associated nephrotic syndrome.

**Race-, sex-, and Age-related Demographics**

Because diabetes mellitus is one of the major causes of nephrotic syndrome, American Indians, African Americans, and Hispanics have an increased incidence of nephrotic syndrome than White persons. HIV-associated nephropathy is a consequence of HIV infection that is uncommon in Whites; however, it is frequently seen in African Americans because of their greater prevalence of the ApoL1 alleles. Focal glomerulosclerosis seems to be overrepresented as one of the causes of nephrotic syndrome in African Americans as opposed to White children. There is a male predominance in nephrotic syndrome, as seen in chronic kidney disease in general. This pattern is also observed in paraneoplastic membranous nephropathy. However, lupus nephritis affects mostly women.

## **Differential Diagnosis**

The differential diagnosis for nephrotic syndrome include the following:

* Hepatic: insufficiency, hepatocellular cirrhosis, Budd-Chiari syndrome
* Digestive: exudative enteropathy, lymphangiectasia, malnutrition
* Cardiac: hereditary angioneurotic edema
* Immune: anaphylaxis
* Renal: chronic glomerulonephritis, diabetic nephropathy, focal segmental glomerulosclerosis, HIV-associated nephropathy, IgA nephropathy, membranous glomerulonephritis, minimal change disease.

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**KIDNEY STONE**

**You may hear healthcare professionals refer to kidney stones as renal calculi, nephrolithiasis or urolithiasis.**

Kidney stones are solid masses or crystals that form from substances (like minerals, acids and salts) in your kidneys. They can be as small as a grain of sand or — rarely — larger than a golf ball. Kidney stones are also called renal calculi or nephrolithiasis.

Depending on the size of your kidney stone (or stones), you may not even realize that you have one. Smaller stones can pass through your urinary tract in your pee with no symptoms. Large kidney stones can get trapped in your ureter (the tube that drains urine from your kidney down to your bladder). This can cause pee to back up and limit your kidney’s ability to filter waste from your body. It can also cause bleeding.

It can take as long as three weeks for kidney stones to pass on their own. Even some small stones can cause extreme pain as they go through your urinary tract and out of your body. You may need a provider to break up and remove a stone that can’t pass on its own. About 1 in 10 people will get a kidney stone during their lifetime. They’re most common in men in their 30s and 40s. They’re also more common among non-Hispanic white people.

**Symptoms**

A kidney stone usually doesn't cause symptoms until it moves around within the kidney or passes into one of the ureters. The ureters are the tubes that connect the kidneys and bladder.

If a kidney stone gets stuck in one of the ureters, it may block the flow of urine and cause the kidney to swell and the ureter to spasm. That can be very painful. At that point, you may have these symptoms:

* Serious, sharp pain in the side and back, below the ribs.
* Pain that spreads to the lower stomach area and groin.
* Pain that comes in waves and varies in how intense it feels.
* Pain or a burning feeling while urinating.

Other symptoms may include:

* Pink, red or brown urine.
* Cloudy or foul-smelling urine.
* A constant need to urinate, urinating more often than usual or urinating in small amounts.
* Upset stomach and vomiting.
* Fever and chills if an infection is present.
* Bloody pee
* Pain when you pee

Pain caused by a kidney stone may change as the stone moves through your urinary tract. For instance, the pain may shift to a different part of the body or become more intense.

**Causes**

Kidney stones often have no definite, single cause. But many factors may raise your risk.

Kidney stones develop when the urine contains more crystal-forming substances than the fluid in the urine can dilute. These substances include calcium oxalate, calcium phosphate and uric acid. At the same time, the urine may lack substances that prevent crystals from sticking together. That creates an ideal setting for kidney stones to form.

### **Types of kidney stones**

Knowing the type of kidney stone you have helps your healthcare professional figure out its cause and the right treatment for you. This information also can give clues on how to prevent more kidney stones. If you can, try to save your kidney stone if you pass one. Then bring it to your healthcare professional, who can check on what type of kidney stone it is.

Types of kidney stones include:

* **Calcium stones.** Most kidney stones are calcium stones. They're usually made of the chemical compound calcium oxalate. Oxalate is a substance made daily by the liver or absorbed from diet. Some fruits and vegetables, as well as nuts and chocolate, have high amounts of oxalate.

Dietary factors, high doses of vitamin D, intestinal bypass surgery and many conditions that affect metabolism can make calcium or oxalate more concentrated in urine.

Calcium stones also can be made of calcium phosphate. This type of stone is more common in metabolic conditions such as renal tubular acidosis. It also may be linked with some medicines for migraines or seizures such as topiramate (Topamax, Trokendi XR, others).

* **Uric acid stones.** Uric acid stones can form in people who lose too much fluid because of ongoing diarrhea or people who have trouble absorbing nutrients from food; those who eat a high-protein diet or lots of organ meats or shellfish; and those with diabetes mellitus or metabolic syndrome. Some genetic factors also may raise the risk of uric acid stones.
* **Struvite stones.** Struvite stones form in response to a urinary tract infection. These stones can grow quickly and become quite large, sometimes with few symptoms or little warning.
* **Cystine stones.** These stones form in people with a rare genetic condition called cystinuria that causes the kidneys to leak too much of a protein building block called cystine.

**Risk factors**

Factors that raise your risk of kidney stones include:

* **Family or personal history.** If someone in your family has had kidney stones, you're more likely to develop stones too. If you've already had one or more kidney stones, you're at higher risk of getting another.
* **Dehydration.** Not drinking enough water each day can raise your risk of kidney stones. People who live in warm, dry climates and those who sweat a lot may be at higher risk than others.
* **Some diets.** Eating a diet that's high in oxalate, protein, sodium and sugar may raise your risk of some types of kidney stones. This is especially true with a high-sodium diet. Too much sodium raises the amount of calcium the kidneys must filter. And that greatly raises the risk of kidney stones.
* **Obesity.** This complex disease involves having too much body fat, and it's been linked with a higher risk of kidney stones.
* **Digestive diseases and surgery.** Gastric bypass surgery, inflammatory bowel disease or ongoing diarrhea can cause changes in the digestive process. These changes affect how the body absorbs calcium and water. That in turn increases the amounts of stone-forming substances in the urine.
* **Other health conditions** such as renal tubular acidosis, cystinuria, hyperparathyroidism and repeated urinary tract infections also can raise the risk of kidney stones. A rare genetic condition called primary hyperoxaluria raises the risk of calcium oxalate stones.
* **Some supplements and medicines.** These include vitamin C, dietary supplements, overuse of laxatives, calcium-based antacids, and some medicines for migraines or depression.

#### **Medical conditions that increase kidney stone risk**

#### Certain health conditions can put you at a higher risk for kidney stones. These include:

Cystic fibrosis.

Cystinuria, a genetic disorder that causes a buildup of cystine.

Diabetes.

Gout.

High blood pressure.

High calcium levels in your urine (hypercalciuria).

Inflammatory bowel disease (IBD).

Kidney cysts.

Obesity.

Osteoporosis.

Parathyroid disease.

Primary hyperoxaluria.

Hemiplegia or paraplegia (types of paralysis).

**COMPLICATIONS**

Kidney stones can put you at risk for: A blockage that backs pee up into your kidney, causing it to swell (hydronephrosis).

Kidney infection (pyelonephritis).

Acute kidney injury (a type of kidney failure that can be reversible).

Frequent urinary tract infections (UTIs).

Chronic kidney disease (CKD).

**Diagnosis and Tests**

Diagnosis involves the steps that your healthcare professional takes to find out if you have kidney stones. Diagnosis also can include testing to find the cause and chemical makeup of kidney stones. Your healthcare professional starts by giving you a physical exam. You also may need tests such as:

* **Blood tests.** Blood tests may reveal too much calcium or uric acid in your blood. Blood test results help track the health of your kidneys. These results also may lead your healthcare professional to check for other health conditions.
* **Urine testing.** Your healthcare professional may ask you to collect samples of your urine over 24 hours. The 24-hour urine collection test may show that your body is releasing too many stone-forming minerals or too few substances that prevent stones. Follow your healthcare professional's instructions closely. Collecting the urine appropriately is key to making changes in your treatment plan to prevent new stones from forming.
* **Imaging.** Imaging tests such as CT scans may show kidney stones in your urinary tract. An advanced scan known as a high-speed or dual energy CT scan may help find tiny uric acid stones. Simple X-rays of the stomach area, also called the abdomen, are used less often. That's because this kind of imaging test can miss small kidney stones.

Ultrasound is another imaging option to diagnose kidney stones.

* **Analysis of passed stones.** You may be asked to urinate through a strainer to catch any stones that you pass. Then a lab checks the chemical makeup of your kidney stones. Your healthcare professional uses this information to find out what's causing your kidney stones and to form a plan to prevent more kidney stones.
* **Genetic testing.** Some rare conditions that pass from parent to child make kidney stones more likely. For instance, having cystinuria raises the risk of cystine stones. Primary hyperoxaluria raises the risk of calcium oxalate stones. If your healthcare professional thinks you might have such a condition, your healthcare professional may recommend genetic testing to find out for sure.

## **Management and Treatment**

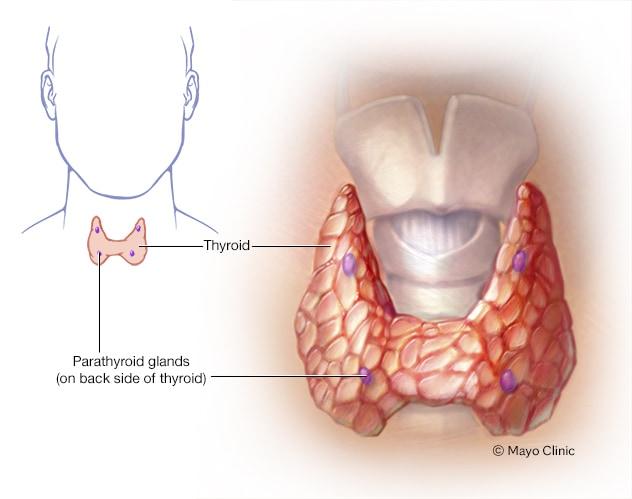
Treatment for kidney stones varies. It depends on the type of stone and the cause.

### **Small stones with few symptoms**

Most small kidney stones don't require invasive treatment such as surgery. You may be able to pass a small stone by:

* **Drinking water.** Drinking as much as 2 to 3 quarts (1.8 to 3.6 liters) a day likely will keep your urine dilute and may prevent stones from forming. Unless your healthcare professional tells you otherwise, drink enough fluid. It's ideal to mainly drink water to produce clear or nearly clear urine.
* **Pain relievers.** Passing a small stone can cause mild to serious discomfort. To relieve mild pain, your healthcare professional may recommend pain relievers such as ibuprofen (Advil, Motrin IB, others) or naproxen sodium (Aleve). For serious pain, other treatments in the emergency room may be needed.
* **Other medicines.** Your healthcare professional may give you medicine to help pass your kidney stone. This type of medicine is known as an alpha blocker. It relaxes the muscles in your ureter. This helps you pass the kidney stone more quickly and with less pain. Examples of alpha blockers include tamsulosin (Flomax) and the drug combination dutasteride and tamsulosin (Jalyn).

### **Large stones and those that cause symptoms**

**Parathyroid glands** Enlarge image

Kidney stones that are too large to pass on their own may need more-extensive treatment. So might stones that cause bleeding, kidney damage or ongoing urinary tract infections. Treatments may include:

* **Using sound waves to break up stones.** For some kidney stones, your healthcare professional may recommend a treatment called extracorporeal shock wave lithotripsy. This also is known as ESWL. But it depends on the size and location of your stones.

ESWL uses sound waves to create strong vibrations called shock waves that break the stones into tiny pieces that can be passed in urine. The treatment lasts about 45 to 60 minutes. It can cause moderate pain, so you may be given medicines to prevent pain or help relax you.

ESWL can cause blood in the urine and bruising on the back or stomach area. It also can cause bleeding around the kidney and around other nearby organs. It can cause discomfort as the stone fragments pass through the urinary tract too.

* **Surgery to remove very large stones in the kidney.** A surgery called percutaneous nephrolithotomy (nef-row-lih-THOT-uh-me) involves removing a kidney stone using small telescopes and tools inserted through a small cut in the back or side.

You receive medicine called a general anesthetic that prevents pain and puts you in a sleep-like state during the surgery. You'll likely recover in the hospital for 1 to 3 days afterward. Your healthcare professional may recommend this surgery if ESWL doesn't help you enough.

* **Using a scope to remove stones.** To remove a smaller stone in your ureter or kidney, your surgeon may use a thin lighted tube called a ureteroscope. This instrument is equipped with a camera. The surgeon places the ureteroscope through the urethra and bladder to the ureter.

Once the stone is found, special tools can snare the stone or break it into pieces that will pass in the urine. Then the surgeon may place a small tube called a stent in the ureter to relieve swelling and support healing. You may need general or local anesthesia during this procedure.

* **Parathyroid gland surgery.** Some calcium phosphate stones are caused by overactive parathyroid glands. These glands are located on the four corners of the thyroid gland, just below Adam's apple. When these glands make too much parathyroid hormone, that's a condition known as hyperparathyroidism. The condition can cause calcium levels to become too high, and kidney stones may form as a result.

Hyperparathyroidism sometimes happens when a small tumor that isn't cancer forms in one of the parathyroid glands. Or hyperparathyroidism can happen if you develop another condition that leads these glands to make more parathyroid hormone. Removing the tumor from the gland stops kidney stones from forming. Or your healthcare professional may recommend treatment of the condition that's causing your parathyroid gland to make too much of the hormone.

**Prevention**

Prevention of kidney stones may include a mix of lifestyle changes and medicines.

### **Lifestyle changes**

You may lower your risk of kidney stones if you:

* **Drink water throughout the day.** This is the most important lifestyle change you can make. If you've had kidney stones before, your healthcare professional may tell you to drink enough fluids to pass about 2.1 quarts (2 liters) of urine a day or more. You may be asked to measure how much urine you pass to make sure that you're drinking enough water.

If you live in a hot, dry climate or you exercise often, you may need to drink even more water to produce enough urine. If your urine is light and clear, you're likely drinking enough water.

* **Eat fewer oxalate-rich foods.** If you tend to form calcium oxalate stones, your healthcare professional may recommend limiting foods rich in oxalates. These include rhubarb, beets, okra, spinach, Swiss chard, sweet potatoes, nuts, tea, chocolate, black pepper, sesame or tahini products, and soy products. Reviewing your diet with a dietitian with expertise in kidney stones is usually helpful.
* **Choose a diet low in sodium and animal protein.** Lower the amount of sodium you eat. And choose protein sources that don't come from meat or fish, such as legumes. Think about using a salt substitute to flavor foods.
* **Keep eating calcium-rich foods but use caution with calcium supplements.** Calcium in food doesn't have an effect on your risk of kidney stones. Keep eating calcium-rich foods unless your healthcare professional recommends otherwise.

Ask your healthcare professional before taking calcium supplements. These have been linked with a higher risk of kidney stones. You may lower the risk by taking supplements with meals. Diets low in calcium can make kidney stones more likely to form in some people.

**Outlook / Prognosis**

Around 90% of small kidney stones (smaller than 6 mm) and 60% of large stones (larger than 6 mm) pass on their own. If you have a large kidney stone or one that’s blocking the flow of pee, you’ll need to have a procedure to break up and/or remove it. Sometimes, smaller stones that were expected to pass on their own can grow or move to create a blockage.

If you’ve had kidney stones, you’re likely to get more in the future. You’ll likely need to work on preventing them with changes to the foods you eat and, sometimes, medication.

If your provider thinks your kidney stone can pass on its own, you should drink plenty of water to help flush it out. Take any medications as prescribed and follow your provider’s recommendations on what to eat and drink (and what to avoid).

**Living With**

### Kidney stones shouldn’t stop you from going about your daily activities or drastically reduce your quality of life. Thanks to passing them while you urinate, and thanks to treatment options, kidney stones aren’t permanent.

### If you’ve had kidney stones, you’re at a higher risk for more kidney stones and chronic kidney disease. Kidney stones don’t cause death.

### **When to see a doctor**

Make an appointment with your healthcare professional if you have any symptoms that worry you.

Get a healthcare checkup right away if you have:

* Pain so bad that you can't sit still or find a comfortable position.
* Pain along with upset stomach and vomiting.
* Pain along with fever and chills.
* Blood in your urine.
* Trouble passing urine.

### **What questions should I ask my healthcare provider?**

You may want to ask your provider:

Do I have a kidney stone or is there another reason for my symptoms?

What type of kidney stone do I have?

What size is my kidney stone?

Where’s my kidney stone located?

How many kidney stones do I have?

Do I need treatment, or will I be able to pass the kidney stone?

Should I be tested for kidney disease?

What changes should I make to my diet?

What type of procedure should I have to get rid of the stones?

Kidney stones can be frustrating at best and agonizingly painful at worst. To stop your situation from getting worse, you should be evaluated by a healthcare provider as soon as possible. The pain can get severe, and surgery might be necessary. Remember: Don’t skip your prescriptions, drink lots of water and follow any dietary guidelines. Also, remember that kidney stones are a temporary condition. They won’t bother you forever.

**Medications**

Medicines can control the amount of minerals and salts in the urine. They may be helpful in people who form certain kinds of stones. The type of medicine that your healthcare professional prescribes depends on the kind of kidney stones you have. Here are some examples:

* **Calcium stones.** To help prevent calcium stones from forming, your healthcare professional may prescribe a thiazide diuretic or potassium citrate. If you have calcium oxalate stones due to the rare genetic condition primary hyperoxaluria, you may need other treatments to lower the amount of oxalate in your blood. Your healthcare professional may recommend that you take vitamin B6, also called pyridoxine. Or you may need prescription medicines such as lumasiran (Oxlumo) or nedosiran (Rivfloza).
* **Uric acid stones.** Your healthcare professional may prescribe allopurinol (Zyloprim, Aloprim, others) to lower uric acid levels in your blood and urine. You also may be prescribed potassium citrate. Sometimes, these medicines may dissolve existing uric acid stones.
* **Struvite stones.** To prevent struvite stones, your healthcare professional may recommend ways to keep your urine free of bacteria that cause infection. For instance, you may be told to urinate more often and to drink fluids to keep your urine flow good. Rarely, long-term use of antibiotics in small or occasional doses may help achieve this goal. For instance, your healthcare professional may suggest that you take an antibiotic before and for a while after surgery to treat your kidney stones. Medicines called acetohydroxamic acid also may help prevent struvite stones.
* **Cystine stones.** A diet that's lower in sodium and protein may help prevent cystine stones. Your healthcare professional also may recommend that you drink more fluids so that you urinate more. If those changes alone don't help, medicines called thiol drugs or other newer medicines also may be prescribed. They might make crystals less likely to form.

**Epidemiology**

• Estimated prevalence of 3% in all individuals

• Affects up to 12% of the population during their lifetime

• Stone recurrence rates approach 50% at 10 years

• Caucasian males have the highest incidence in the US

• Incidence highest in the “Stone Belt,” i.e. southeastern and central southern US.

Calcium-containing stones continue to comprise the most common KS composition globally . In the population analysis of Olmsted County, Minnesota, 94% of the incidence stones were calcium stones, of which 76% were mainly calcium oxalate, 18% were mainly calcium phosphate, 5% were uric acid, 1% were struvite–magnesium ammonium phosphate, and 0.1% were cystine. In Germany, the analysis of 45,783 urinary stones  
in the period from 2007 to 2020 produced similar results. Calcium oxalate (CaOx) was the most frequent type of stone, with 71.4%, calcium phosphate was 10.2%, and uric acid  
was 8.3% .Uric acid nephrolithiasis accounts for 8–10% and is increasing globally. It mostly presents in obese cases or individuals with metabolic syndrome, and its increasing incidence corresponds to the increasing prevalence of metabolic syndrome, obesity, and diabetes  
worldwide. Uric acid stone prevalence is higher in older ages. Hyperuricosuria is not the essential causality: acidic urine is .  
In a retrospective analysis of 1516 patients in a large stone center in the USA, the percentage of uric acid stones relative to the total number of kidney stones increased significantly from 7% to 14% over the period 1980–2015. In this study, uric acid stone formers were older and had a higher BMI and a lower urinary pH than calcium stone formers . In another US study with 4339 kidney stones covering patients from seven states, uric acid stones comprised 12%. This study also showed that, with the exception of Florida, the stone composition did not differ across US regions. An increase in uric-acid-containing stones from 2.0% 40 years ago to 9.1% in the 2014–2017 period was also reported in a Norwegian surgical cohort.  
Struvite stones containing magnesium ammonium phosphate, also known as infection-related stones, comprise 7–8% of stones worldwide. They are the result of the ammoniage-nesis caused by urea-splitting bacteria secondary to infection .  
Rare inherited metabolic disorders are often linked to the presentation of kidney stones in early childhood, causing a high burden of recurrence. These include cystinuria, primary hyperoxaluria, distal renal tubular acidosis (RTA), xanthinuria, Lesch–Nyhan syndrome, Dent disease, and adenine phosphoribosyl transferase (APRT) deficiency (a cause of dihydroxyadenine stones).

**Differential Diagnosis**

• Obstructing renal or ureteral stone

• Hydronephrosis (ureteropelvic junction obstruction, stricture, ureteral/ renal malignancy)

• Bacterial cystitis or pyelonephritis

• Acute abdomen (bowel, biliary, pancreas, or aortic abdominal aneurysm)

• Radicular pain (L1 herpes zoster, sciatica)

• Depending on the patient gender, primary gonadal pathology – Women: ectopic pregnancy, ovarian torsion – Men: testicular torsion, orchitis

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## **Endocrine Related Hypertension**

## **Endocrine hypertension** is a specific type of high blood pressure caused by disorders in the endocrine system, the network of glands that produce hormones. These hormones regulate various functions in the body, and when they become imbalanced, they can lead to elevated blood pressure.

## There are two types of hypertension—primary (essential) and secondary. Most hypertension is the primary type. Its cause is unknown, but genetics and factors such as excessive salt intake, obesity, lack of exercise, and the use of tobacco and alcohol play a role. Primary hypertension is more common among African Americans and among older adults.

## Secondary hypertension is due to medical conditions such as kidney disease and obstructive sleep apnea, as well as endocrine disorders such as primary aldosteronism, [**Cushing’s syndrome**](https://www.endocrine.org/patient-engagement/endocrine-library/cushings-syndrome-and-cushing-disease), and pheochromocytoma. In addition, several prescription and over-the-counter medications, such as oral contraceptives, nonsteroidal inflammatory agents, steroids, and decongestants, can cause secondary hypertension.

## **Endocrine Connection**

## The endocrine system is a series of glands that secrete [**hormones**](https://www.endocrine.org/patient-engagement/endocrine-library/hormones-and-endocrine-function) that the body uses for a wide range of functions, including regulating blood pressure.

**Adrenal glands**: If the adrenal glands make too much aldosterone, cortisol, or hormones like adrenaline, it can cause high blood pressure.

**Thyroid gland**: High blood pressure can be caused by an underactive (hypothyroidism) or overactive (hyperthyroidism) thyroid gland.

**Pituitary gland**: Sometimes problems with the adrenal glands and thyroid gland are due to problems with the pituitary gland. If the pituitary gland sends too much signal to the adrenal glands or thyroid gland, it can result in high blood pressure.

**Parathyroid glands**: If the parathyroid glands make too much parathyroid hormone, it can cause high blood pressure.

**Pancreas**: High blood pressure in adults with obesity may be partially due to elevated insulin levels and insulin resistance. Insulin is made in the pancreas.

As mentioned above, secondary hypertension is due to a medical condition. Below are some conditions that can affect your blood pressure:   
  
**Primary aldosteronism**: Primary aldosteronism is due to the adrenal glands making too much of the hormone aldosterone. In addition to high blood pressure, it can cause low potassium levels in the blood (hypokalemia).   
  
**Pheochromocytoma**: Pheochromocytoma is a rare cause of secondary hypertension caused when the body makes too many hormones similar to adrenaline. In addition to high blood pressure, pheochromocytomas can present with episodes of headache, sweating, and a racing heartbeat.   
  
**Cushing’s syndrome**: Cushing’s syndrome is another rare cause of secondary hypertension caused when the adrenal glands make too much of the hormone cortisol. In addition to high blood pressure, it can present with weight gain in the trunk, muscle weakness, and purple stretch marks.   
  
**Thyroid problems**: Both low thyroid hormone levels (hypothyroidism) and high thyroid hormone levels ([**hyperthyroidism**](https://www.endocrine.org/patient-engagement/endocrine-library/hyperthyroidism)) can cause high blood pressure. Hypothyroidism typically elevates the diastolic pressure more than the systolic pressure, whereas hyperthyroidism typically elevates the systolic pressure more than the diastolic pressure. Other symptoms of hypothyroidism include fatigue, feeling cold all the time, weight gain, constipation, hair loss, and dry skin. Other symptoms of hyperthyroidism include feeling hot all the time, tremors, racing heartbeat, and weight loss.   
  
**Primary hyperparathyroidism**: Primary hyperparathyroidism is caused when the parathyroid glands make too much parathyroid hormone, which results in elevated blood calcium levels. In addition to high blood pressure, it can also cause kidney stones, nausea, constipation, and excessive thirst and urination.   
  
**Obesity**: High blood pressure is very common in adults who are overweight or obese.

Endocrine Hypertension Mechanism

Endocrine hypertension involves the mechanism where certain hormones, when produced excessively or insufficiently by the endocrine glands, disrupt normal blood pressure regulation. Understanding this mechanism is crucial for diagnosing and treating this type of high blood pressure. Hormones from glands such as the adrenal and thyroid impact how salts and fluids are handled by the kidneys, how blood vessels constrict or relax, and how the heart functions. These processes contribute collectively to blood pressure control.

**Hormone:** A chemical messenger produced by glands in the endocrine system, which regulates physiological processes from metabolic rate to mood.

Two key players in endocrine hypertension include:

Aldosterone: By promoting sodium retention, it indirectly increases blood volume and pressure.

Cortisol: In excess, it leads to sodium and water retention, plus vessel constriction, raising blood pressure.

It's important for healthcare professionals to consider these hormonal influences when treating hypertension.

In conditions like primary aldosteronism, patients often exhibit resistant hypertension, which does not easily respond to conventional blood pressure medications. This resistance occurs because the excess aldosterone continuously stimulates sodium retention and increases blood volume.

### **Causes**

The causes of endocrine hypertension are varied, often stemming from either excess or insufficient hormone production. Some common causes include:

* **Primary Aldosteronism:** Overproduction of aldosterone leads to excessive sodium and water retention, resulting in increased blood volume and hypertension.
* **Pheochromocytoma:** This is a tumor of the adrenal gland that causes an excess release of adrenaline, thereby increasing heart rate and blood pressure.
* **Cushing Syndrome:** High levels of cortisol, another hormone produced by the adrenals, can result from tumors or prolonged use of corticosteroid medications, leading to hypertension.
* **Thyroid Disorders:** Both **hyperthyroidism** (excess **thyroid hormone**) and **hypothyroidism** (insufficient **thyroid** hormone) can affect **blood pressure regulation**.

For instance, if you have pheochromocytoma, a rare tumor, it can trigger sudden episodes of high blood pressure, headache, sweating, and rapid heart rate. These symptoms occur due to the adrenal gland releasing excess adrenaline into your bloodstream.

### **Symptoms**

Identifying the symptoms of endocrine hypertension can help in its early detection and treatment. Common symptoms include:

* Severe headaches and dizziness
* Changes in vision
* Sudden episodes of sweating
* Unexplained weight gain or loss
* Palpitations or irregular heartbeats

**DIAGNOSIS**

For **diagnosis**, healthcare providers may use blood and urine tests to measure hormone levels, imaging tests like CT or **MRI** scans to detect tumors, and specialized tests to check adrenal or thyroid gland function.

Blood pressure can be measured both at the doctor’s office and at home. In addition, sometimes a doctor might order a 24-hour blood pressure monitor that can be worn at home. After a hypertension diagnosis, you should discuss with your doctor whether they think you have primary or secondary hypertension, as this will affect the diagnostic tests that are done.

After a diagnosis of hypertension, you should have regular blood pressure checks to see how well your treatment is working. The goal is typically to lower your systolic blood pressure to less than 130 mm Hg and your diastolic blood pressure to less than 80 mm Hg. However, depending on your health and medical conditions, your doctor may have a different blood pressure goal for you. Your doctor can help guide your treatment goals based on your risk factors.   
  
It is best to monitor your blood pressure at home and bring a written record to the doctor at each visit. This information can help in adjusting the medication dose, if needed, and making treatment effective.

Endocrine Hypertension Treatment

Treating **endocrine hypertension** involves addressing the underlying **hormonal imbalance** responsible for high blood pressure. The approach typically varies depending on the specific endocrine disorder in play. Here are some standard treatment methods used to manage this condition:

* **Medication:** Drugs that block overproduction of hormones or that counteract their effects can help balance hormone levels.
* **Surgery:** Removal of hormone-secreting tumors (such as those in pheochromocytoma) can often provide dramatic relief from symptoms.
* **Lifestyle Changes:** Diet, exercise, and stress-management techniques can support medication therapy.
* **Radiofrequency Ablation:** A newer technique used for certain **adrenal tumors**.
* **Surgery:** A procedure that involves cutting and removing a part of the body, often used to remove tumors causing overproduction of hormones.

For a patient with adrenalectomy, the removal of one or both adrenal glands can significantly lower hormone production related to hypertension, often resolving the condition effectively.

You can prevent hypertension by making the following lifestyle changes:

Keep a healthy weight [(body mass index, or BMI, of 18.5 to 24.9 kg/m2)]

Reduce the amount of saturated and total fat in your diet. Eat lots of fruits and vegetables and choose low-fat dairy products.

Reduce salt (sodium) in your diet

Exercise (such as brisk walking) at least 30 minutes a day, most days of the week

Limit alcohol intake (men: no more than two drinks a day; women and lightweight men: no more than one drink a day)

Quit smoking

**Risk factors**

* Family history
* Overweight/obesity
* Lack of exercise
* High salt diet
* Alcohol use
* Smoking
* History of pituitary, thyroid, parathyroid, or adrenal problems

Although there is no cure for primary hypertension, it usually can be controlled. Doctors often prescribe a combination of medication and lifestyle changes. It is very important to take the medication(s) exactly as prescribed daily. Missed doses can increase blood pressure and the risk of heart attack or stroke.

**DIFFERENTIAL DIAGNOSIS**

Essential (95%) Secondary causes (5%) Endocrine Hypertension Adults Cushing’s Syndrome Primary aldosteronism Pheochromocytoma Hyperthyroidism Hypothyroidism Hyperparathyroidism Acromegaly Insulin Resistance Children CAH: 11beta-hydroxylase deficiency CAH: 17 alpha-hydroxylase deficiency Apparent mineralocorticoid excess Liddle syndrome Pseudohypoaldosteronism type 2 Glucocorticoid Resistance Insulin Resistance Constitutive activation of the MR (Geller syndrome)

Non-Endocrine Hypertension Polycystic kidney disease Glomerular disease Renovascular

• Atherosclerosis (older individuals)

• Fibromuscular dysplasia (women)

• Other: Scleroderma, vasculitis (PAN) Medications (Contraceptive drugs, NSAIDs, nasal decongestants with adrenergic effects, MAOIs, steroids, methamphetamine, cocaine) Obstructive sleep apnea Coarctation of aorta Pre-eclampsia, eclampsia Polycythemia vera

**PREVENTION**

You can prevent hypertension by making the following lifestyle changes:

Keep a healthy weight [(body mass index, or BMI, of 18.5 to 24.9 kg/m2)]

Reduce the amount of saturated and total fat in your diet. Eat lots of fruits and vegetables and choose low-fat dairy products.

Reduce salt (sodium) in your diet

Exercise (such as brisk walking) at least 30 minutes a day, most days of the week

Limit alcohol intake (men: no more than two drinks a day; women and lightweight men: no more than one drink a day)

Quit smoking

**PROGNOSIS**

Endocrine-related hypertension can have varied prognosis depending on the underlying cause. For example, primary aldosteronism, a common endocrine cause of hypertension, often presents with resistant or difficult-to-control hypertension and can be associated with normo- or hypokalemia. If diagnosed and treated appropriately, the prognosis can be favorable, with potential for complete cure in cases where the condition is caused by a surgically removable adrenal tumor.

Pheochromocytoma, another endocrine cause, can present with severe symptoms such as headaches, palpitations, excessive sweating, and anxiety-like attacks. **Early diagnosis and treatment can significantly improve the prognosis, but if left untreated, it can lead to severe complications including cardiovascular events**.

In cases of Cushing's syndrome, timely diagnosis and management can help control blood pressure and reduce the risk of complications such as cardiovascular disease and osteoporosis. However, the prognosis can be influenced by the underlying cause of the syndrome, whether it is due to a pituitary tumor, adrenal tumor, or ectopic ACTH production.

Hyperparathyroidism, when associated with hypertension, can lead to complications such as hypercalcemia and renal damage. Proper management, which may include surgery or medical therapy, can improve the prognosis.

Overall, the prognosis of endocrine-related hypertension is influenced by the specific endocrine disorder, the presence of complications, and the effectiveness of the treatment regimen. Early and accurate diagnosis is crucial for optimal management and improved outcomes.

**EPIDEMIOLOGY**

The search yielded a total of 1748 hits from which 45 relevant studies met the inclusion criteria for the review. The overall crude prevalence of hypertension ranged from 0.1% (95%CI:-0.1 to 0.3) to 17.5% (95% CI: 13.6 to 21.4) in children and 2.1% (95%CI: 1.4 to 2.8) to 47.2% (95%CI: 43.6 to 50.8) in adults depending on the benchmark used for diagnosis of hypertension, the setting in which the study was conducted, sex and ethnic group. The crude prevalence of hypertension ranged from 6.2% (95%CI: 4.0 to 8.4) to 48.9% (95%CI: 42.3 to 55.5) for men and 10% (95%CI: 8.1 to 12) to 47.3% (95%CI: 43 to 51.6%) for women. In most studies, prevalence of hypertension was higher in males than females. In addition, prevalence across urban and rural ranged from 9.5% (95%CI: 13.6 to 21.4) to 51.6% (95%CI: 49.8 to 53.4) and 4.8% (95%CI: 2.9 to 6.7) to 43% (95%CI: 42.1 to 43.9) respectively.

The prevalence of hypertension is high among the Nigerian population. Appropriate interventions need to be developed and implemented to reduce the preventable burden of hypertension especially at Primary Health Care Centers which is the first point of call for over 55% of the Nigerian population.

**REFERENCE**

**https://www.endocrine.org/patient-engagement/endocrine-library/endocrine-related-hypertension**

**https://pmc.ncbi.nlm.nih.gov/articles/PMC4603956/**

## **Hypertensive Crisis OR Malignant hypertension**

A hypertensive crisis means you suddenly have very high blood pressure. Normally, your blood pressure should be:

* Top number (systolic) below 120 mm Hg
* Bottom number (diastolic) below 80 mm Hg

During a hypertensive crisis, it jumps much higher:

* Top number: 180 mm Hg or higher, AND/OR
* Bottom number: 120 mm Hg or higher

**A hypertensive crisis is a medical emergency. Go to the emergency room (ER) for treatment.**

Most people who have a hypertensive crisis have high blood pressure (hypertension). This means their blood pressure is higher than it should be from day to day. [Experts](https://pubmed.ncbi.nlm.nih.gov/38985116/) estimate that about 1% to 2% of people with high blood pressure experience a hypertensive crisis. But such crises can also affect people who’ve never had blood pressure problems before.

### **Types of this condition**

There are two main types of hypertensive crises:

* **Hypertensive urgency**. You have very high blood pressure, but no signs of organ damage. Some healthcare providers call this “uncontrolled hypertension” instead of hypertensive urgency. No matter the term used, the goal is to get your numbers to a healthy range.
* **Hypertensive emergency**. You have very high blood pressure that’s causing new or worsening organ damage. Examples of affected organs include your heart, [aorta](https://my.clevelandclinic.org/health/body/17058-aorta-anatomy), eyes, brain and kidneys. Severe preeclampsia and eclampsia are also considered hypertensive emergencies in pregnant patients.

You might also hear your provider use the term “malignant hypertension.” This is a historical term from the early twentieth century. It describes sudden damage to your kidneys and eyes from extremely high blood pressure. Out of habit, some providers still use this term when talking about hypertensive emergencies. But providers generally prefer the term “hypertensive emergency” because it has a more specific definition today.



A hypertensive emergency is when you have blood pressure that’s at least 180/120, along with symptoms like chest pain, dizziness or severe headache.

### **Symptoms of a hypertensive crisis**

Hypertensive crisis symptoms depend on whether there’s organ damage.

Hypertensive urgency (very high blood pressure without organ damage) usually doesn’t have symptoms. The only way you know your blood pressure is very high is by checking it. But it’s also possible to have symptoms like:

Anxiety

Mild headache

Nosebleed

Shortness of breath

Symptoms of a hypertensive emergency (high blood pressure with organ damage) include:

Altered mental status

Chest pain

Dizziness

Edema (swelling)

Heart palpitations

Peeing less than usual

Seizures

Severe headache

Symptoms and signs of stroke, such as sudden facial droop, slurred speech or sudden weakness in your arms and/or legs

Vision changes, including eye pain, loss of vision or sudden blurry vision

During an exam, your provider may notice these signs of a hypertensive emergency:

Bulging neck veins (jugular venous distention)

Crackling sounds in your lungs

Heart murmurs that you didn’t have before

Blood vessel damage in the back of your eye

Unusual asymmetric weakness that raises suspicion for a stroke

**CAUSES**

Hypertensive crisis causes include:

* Not taking your blood pressure medicines as prescribed, for any reason (most common cause)
* Suddenly stopping the use of your blood pressure medicines
* Medicines interacting with each other

Kidney disease

* Endocrine issues
* Preeclampsia or eclampsia during pregnancy

Use of certain addictive substances

Head trauma

Brain tumor

* Certain medicines you take for other conditions

Medicines that may cause a hypertensive crisis include:

Steroids

Medicines for depression

Cyclosporine

* Pseudoephedrine (an ingredient in some cold and flu products)

#### **Risk factors**

You may have a higher risk of having a hypertensive crisis if you:

* Have obesity
* Are male
* Are Black
* Don’t take your blood pressure medicines consistently
* Use stimulant drugs that aren’t prescribed for you

### **Complications of a hypertensive crisis**

Dangerously high blood pressure can lead to:

* A sudden, rapid decline in heart function (acute heart failure)
* Sudden fluid buildup in your lungs (acute pulmonary edema)
* Sudden loss of kidney function (acute kidney failure)
* A tear in your largest artery (aortic dissection)
* Bleeding around your brain (intracranial hemorrhage)
* Lack of blood flow to your heart (heart attack)
* Lack of blood flow to your brain (ischemic stroke)
* Temporary brain dysfunction (hypertensive encephalopathy)

## **Diagnosis and Tests**

### **How doctors diagnose this condition**

A healthcare provider will take your blood pressure in both arms to diagnose a hypertensive crisis. They’ll also review your medical history, talk to you about any symptoms you have and do a physical exam. You may need some tests to help your provider find the cause.

Possible tests include:

* Blood tests
* Urine tests
* Electrocardiogram (EKG/ECG)
* Fundoscopic exam
* Neurological exam
* Transthoracic echocardiogram
* Computed tomography (CT) scan of your chest and head

Your provider will diagnose you with a hypertensive emergency if you have signs of new or worsening organ damage. They’ll diagnose you with hypertensive urgency if these signs aren’t there.

## **Management and Treatment**

Treatment for a hypertensive crisis happens in the emergency room. Healthcare providers give you medicine to bring your blood pressure down to a safe level.

You may go home the same day (with medicines to take on your own) if you don’t have signs of organ damage. But if you’re having a hypertensive emergency, you’ll need to stay in the intensive care unit (ICU) for a couple of days. Providers will give you medicine directly into your veins (through an IV). They’ll also continuously monitor your blood pressure.

Your care team will decide how quickly to bring down your blood pressure based on what other medical conditions you have. In some cases, lowering your blood pressure too quickly can prevent your organs and tissues from getting enough blood. So, your providers may lower it gradually over 24 to 48 hours.

But they’ll bring down your blood pressure more quickly during a hypertensive crisis if you have certain conditions like aortic dissection, severe preeclampsia or eclampsia. In these cases, the benefits of rapid lowering outweigh any risks.

#### **Medications**

Medicines for hypertensive crisis treatment include:

Captopril

Clevidipine

Clonidine

Esmolol

Hydralazine

Labetalol

Nicardipine

Nifedipine

Nitroglycerin

Nitroprusside

Providers choose the right medicine for your needs. They consider your medical conditions and other medicines you’re taking.

### **When should I see my healthcare provider?**

Call the local emergency services number if your blood pressure is 180/120 mm Hg or higher. You need treatment right away.

After receiving treatment for a hypertensive crisis, follow up with your usual healthcare provider. They may want to adjust your blood pressure medicines. They’ll also talk to you about any changes you should make in your daily life.

For example, they may recommend changing what you eat (like following the DASH diet) or adding more physical activity to your routine.

## **Outlook / Prognosis**

A hypertensive crisis is a warning sign that your blood pressure isn’t where it needs to be. Your provider will help you lower your blood pressure and keep it at healthy levels. Make sure to keep all your follow-up appointments and take your medicines exactly as your provider prescribes them.

[**Surgery**](https://www.vaia.com/en-us/explanations/medicine/surgery/)**:** A procedure that involves cutting and removing a part of the body, often used to remove tumors causing overproduction of hormones.

For a patient with adrenalectomy, the removal of one or both adrenal glands can significantly lower hormone production related to hypertension, often resolving the condition effectively.

Managing endocrine hypertension also involves a comprehensive assessment and monitoring plan:

| **Intervention** | **Purpose** |
| --- | --- |
| Medications | Regulate hormone levels and lower blood pressure. |
| Surgery | Remove dysfunctional gland/tumor. |
| Lifestyle Modifications | Improve overall heart health and reduce stress. |

Each patient's treatment plan should be tailored based on their medical history, specific hormonal imbalances, and the presence of

**pheochromocytoma**

A pheochromocytoma (pronounced FEE-oh-KROH-moh-sy-TOH-muh) is a rare tumor that forms in the center of one or both of your adrenal glands (adrenal medulla). The tumor is made of a certain type of cell called a chromaffin cell, which produces and releases the hormones that cause the “fight or flight” response.

Usually, pheochromocytoma affects only one adrenal gland, but it can affect both glands. Sometimes there’s more than one tumor in one adrenal gland.

Most pheochromocytomas are benign (not cancerous). Approximately 10% to 15% of pheochromocytomas may be malignant (cancerous). There's no standard staging system for pheochromocytoma if it’s cancerous. Instead, it’s described as the following:

* **Localized pheochromocytoma:** The tumor is in one or both adrenal glands only.
* **Regional pheochromocytoma:** The cancer has spread to lymph nodes or other tissues near your adrenal glands.
* **Metastatic pheochromocytoma:** The cancer has spread to other parts of your body, like your liver, lungs, bone or distant lymph nodes.
* **Recurrent pheochromocytoma**: The cancer has recurred (come back) after it has been treated. It may come back in the same place or in another part of your body.

#### **What are adrenal glands?**

You have two adrenal glands, one on top of each kidney in the back of your upper abdomen. They are part of your endocrine system. Each adrenal gland has two parts. The outer layer of your adrenal gland is called your adrenal cortex. The center of your adrenal gland is called your adrenal medulla.

Your adrenal medullae make hormones called catecholamines that help regulate the following important bodily functions and aspects:

* Heart rate.
* Blood pressure.
* Blood sugar (blood glucose).
* The way your body responds to stress (the “fight or flight” response).

The primary catecholamines include:

* Dopamine.
* Epinephrine (adrenaline).
* Norepinephrine (noradrenaline).

Sometimes a pheochromocytoma can release extra adrenaline and noradrenaline into your blood, causing certain symptoms.

Anyone at any age can get a pheochromocytoma, but they occur most often in people between 30 and 50 years of age. Approximately 10% of cases occur in children.

**Pheochromocytomas**

Pheochromocytomas are rare tumors. The true number of pheochromocytoma cases is unknown since many people with pheochromocytomas don’t have symptoms and go undiagnosed. Less than 1% of people who have high blood pressure have a pheochromocytoma.

**Symptoms**

**What are the symptoms of pheochromocytoma?**

Signs and symptoms of pheochromocytoma happen when the tumor releases too much adrenaline (epinephrine) or noradrenaline (norepinephrine) into your blood. However, some pheochromocytoma tumors don’t make extra adrenaline or noradrenaline and don’t cause symptoms (are asymptomatic).

Common symptoms of pheochromocytoma include:

* High blood pressure (hypertension).
* Headache.
* Excessive sweating for no known reason.
* A pounding, fast or irregular heartbeat.
* Feeling shaky.

Less common symptoms of pheochromocytoma include:

* Pain in your chest and/or abdomen.
* Being much paler than usual.
* Nausea and/or vomiting.
* Diarrhea.
* Constipation.
* An extreme drop in blood pressure upon standing suddenly (orthostatic hypotension).
* Unexplained weight loss.

You may experience signs and symptoms of pheochromocytoma after certain events, including:

* Intense physical activity.
* A physical injury or intense emotional stress.
* Childbirth.
* Going under anesthesia.
* Surgery.
* Eating foods high in tyramine, like red wine, chocolate and cheese.

### **Can symptoms of pheochromocytoma come and go?**

A person with a pheochromocytoma could have sustained high blood pressure (the most common symptom of pheochromocytoma) or it may come and go.

People with pheochromocytomas may also experience paroxysmal “attacks,” which are chronic episodes of high blood pressure that often lead to headaches, irregular heartbeats (palpitations) and excessive sweating (diaphoresis). These episodes can happen anywhere from several times a day to a couple of times a month.

### **causes of pheochromocytoma**

In most cases of pheochromocytoma, the exact cause is unknown, and it occurs randomly.

Approximately 25% to 35% of people who have pheochromocytoma have a hereditary condition (passed through the family) that’s linked to pheochromocytoma, including:

* Multiple endocrine neoplasia 2 syndrome, types A and B (MEN2A and MEN2B).
* Von Hippel-Lindau (VHL) disease.
* Neurofibromatosis type 1 (NF1).
* Hereditary paraganglioma syndrome.
* Carney-Stratakis dyad [paraganglioma and gastrointestinal stromal tumor (GIST)].
* Carney triad (paraganglioma, GIST and pulmonary chondroma).

Pheochromocytomas may also be caused by mutations (changes) of one of at least 10 different genes.

## **Diagnosis and Tests**

Since pheochromocytoma is a rare tumor and is sometimes asymptomatic, it can be difficult to diagnose. Healthcare providers sometimes find pheochromocytomas when a test or procedure is done for another reason.

A healthcare provider may suspect a diagnosis of pheochromocytoma after reviewing the following factors:

* A detailed medical history, including previous pheochromocytoma cases in your family.
* A thorough physical and medical evaluation.
* Certain symptom characteristics, such as paroxysmal attacks and high blood pressure that’s unresponsive to standard treatment.

### **What tests are used to diagnose pheochromocytoma?**

Your healthcare provider may use the following tests and procedures to diagnose pheochromocytoma:

* **24-hour urine test:** This type of urine (pee) test involves collecting your urine for 24 hours to measure the level of catecholamines (adrenal hormones) in your urine. Substances that result from the breakdown of these hormones are also measured. Higher-than-normal amounts of certain catecholamines in your urine may be a sign of pheochromocytoma.
* **Blood catecholamine tests:** These tests measure the level of catecholamines in your blood. Substances that result from the breakdown of these hormones are also measured. Higher-than-normal levels of certain catecholamines in your blood may be a sign of pheochromocytoma.
* **CT scan (computer tomography scan):** A CT scan is an imaging procedure that takes a series of X-ray images from different angles to provide detailed pictures of areas inside your body. Your provider may recommend a CT scan so that they can look at your adrenal glands.
* [**MRI (magnetic resonance imaging)**](https://my.clevelandclinic.org/health/diagnostics/4876-magnetic-resonance-imaging-mri)**:** An MRI is an imaging procedure that uses a magnet, radio waves and a computer to make a series of detailed pictures of areas inside your body. Your provider may recommend an MRI so that they can look at your adrenal glands.

After your provider has diagnosed pheochromocytoma, they’ll likely perform additional tests to see if the tumor is benign or malignant and if it has spread to other parts of your body.

### **Is there genetic testing for pheochromocytoma?**

If you’re diagnosed with pheochromocytoma, your provider may recommend genetic counseling to find out your risk for having an inherited syndrome and other related cancers.

Your healthcare provider may recommend genetic testing if any of the following situations apply to you:

* You have a personal or family history of traits linked with inherited pheochromocytoma or paraganglioma syndrome.
* You have tumors in both of your adrenal glands.
* You have more than one tumor in one adrenal gland.
* You have signs or symptoms of higher-than-normal catecholamine levels in your blood.
* You’ve been diagnosed with pheochromocytoma before age 40.

If your genetic counselor finds certain gene changes in your testing results, they'll likely recommend that your family members who are at risk but do not have signs or symptoms be tested as well.

## **Management and Treatment**

The best treatment option is surgery, when feasible.

Treatment options for pheochromocytoma depend on several factors, including:

* The size of the tumor.
* If the tumor is benign (not cancer) or malignant (cancer).
* If you have symptoms of catecholamines that are higher than normal.
* If the tumor is in one area only or has spread to other places in your body (metastasized).
* If the tumor has been diagnosed for the first time or has come back (recurred).

If you have pheochromocytoma that causes symptoms due to excess adrenal hormones, your healthcare provider will likely recommend medication to manage the symptoms. Medications may include:

* Medication that keeps your blood pressure normal, such as alpha-blockers.
* Medication that keeps your heart rate normal, such as beta-blockers.
* Medication that blocks the effect of the excess hormones released by your adrenal gland(s).

Treatment options for pheochromocytoma include:

* Surgery.
* Radiation therapy.
* Chemotherapy.
* Ablation therapy.
* Embolization therapy.
* Targeted therapy.

Together, you and your healthcare team will determine a treatment plan that works best for you and your situation.

#### **Surgery**

Surgery is the main form of treatment for pheochromocytoma. Approximately 90% of pheochromocytomas are successfully removed by surgery.

If you have a pheochromocytoma, your provider may recommend a type of surgery called adrenalectomy to remove one or both of your adrenal glands. During the surgery, your surgeon will check the surrounding tissue and lymph nodes to see if the tumor has spread. If it has, your surgeon will remove the affected tissue(s) as well, if possible.

After surgery, your provider will check the catecholamine levels in your blood or urine. Normal catecholamine levels are a sign that all the pheochromocytoma cells were removed.

If your surgeon removes both of your adrenal glands, you’ll need life-long hormone therapy to replace hormones made by your adrenal glands.

#### Radiation therapy

Radiation therapy is a cancer treatment that focuses strong beams of energy to destroy cancer cells or keep them from growing while sparing as much surrounding healthy tissue as possible.

There are two types of radiation therapy:

* External radiation therapy: This therapy uses a machine outside your body to send radiation toward the cancer.
* Internal radiation therapy: This therapy uses a radioactive substance sealed in needles, seeds, wires or catheters that a healthcare provider places directly into or near the cancer.

The type of radiation therapy your provider may recommend depends on whether your cancer is localized, regional, metastatic or recurrent. Providers most often use external radiation therapy and/or 131I-MIBG therapy to treat malignant pheochromocytoma. 131I-MIBG is a radioactive substance infusion that collects in certain kinds of tumor cells, killing them with the radiation that it gives off.

#### **Chemotherapy**

Chemotherapy is a cancer treatment that uses drugs to stop the growth of cancer cells by killing the cells or by preventing them from dividing and multiplying. Chemotherapy is usually given intravenously through a vein (intravenously). It’s usually an effective treatment, but it can cause side effects.

#### **Ablation therapy**

Ablation therapy is a minimally invasive treatment option that uses very high or very low temperatures to destroy tumors. Ablation therapies that can help kill cancer cells and abnormal cells include:

* Radiofrequency ablation: This therapy uses radio waves to heat and destroy cancer cells and abnormal cells. The radio waves travel through electrodes (small devices that carry electricity).
* Cryoablation: This therapy uses liquid nitrogen or liquid carbon dioxide to freeze and destroy cancer cells and abnormal cells.

#### **Embolization therapy**

Embolization therapy is a pheochromocytoma treatment that blocks the artery leading to your adrenal gland. Blocking the blood flow to your adrenal glands helps kill the cancer cells that are growing there.

#### **Targeted therapy**

Targeted therapy is a treatment option that uses medications or other substances to attack specific cancer cells without harming healthy cells. Healthcare providers use targeted therapies to treat metastatic and recurrent pheochromocytoma.

Researchers are currently studying sunitinib, a type of tyrosine kinase inhibitor, for the treatment of metastatic pheochromocytoma. Tyrosine kinase inhibitor therapy is a type of targeted therapy that prevents tumors from growing.

## **Prevention**

Unfortunately, you can’t prevent developing a pheochromocytoma. However, if you’re at risk for developing a pheochromocytoma due to certain inherited syndromes and genes, genetic counseling can help screen for pheochromocytoma and potentially help you catch it in its early phases.

Talk to your healthcare provider if you have any first-degree relatives (siblings and parents) that have been diagnosed with pheochromocytoma and/or any of the following genetic conditions:

* Multiple endocrine neoplasia 2 syndrome.
* Von Hippel-Lindau (VHL) disease.
* Neurofibromatosis type 1 (NF1).
* Hereditary paraganglioma syndrome.
* Carney-Stratakis dyad.
* Carney triad.

## **Outlook / Prognosis**

### **T**he prognosis (outlook) for pheochromocytoma is usually good if it’s treated. Approximately 90% of pheochromocytomas are successfully removed by surgery.

If pheochromocytomas are left untreated, they can potentially cause serious, life-threatening complications, including:

* Heart muscle disease (cardiomyopathy).
* Inflammation of your heart muscle (myocarditis).
* Uncontrolled bleeding in your brain (cerebral hemorrhaging).
* Accumulation of fluid in your lungs (pulmonary edema).

Some people with a pheochromocytoma may also be at risk of developing a stroke or heart attack (myocardial infarction).

## **When should I see my healthcare provider?**

If you’ve been diagnosed with a pheochromocytoma and are experiencing concerning symptoms, contact your healthcare provider.

If you’re experiencing symptoms of pheochromocytoma, such as high blood pressure and headaches, talk to your provider. Even though pheochromocytoma is rare and the likelihood of having it is low, it’s important to treat high blood pressure.

If you’ve recently found out that one of your first-degree relatives (siblings and parents) has a genetic syndrome, such as multiple endocrine neoplasia 2 syndrome or von Hippel-Lindau (VHL) disease, that puts you at a higher risk of developing pheochromocytoma. You should contact your provider about genetic testing.

### **What questions should I ask my doctor?**

If you’ve been diagnosed with a pheochromocytoma, it may be helpful to ask your healthcare provider the following questions:

* What caused my pheochromocytoma?
* Could my children and/or relatives develop a pheochromocytoma?
* What are my treatment options?
* What are the side effects of different treatment therapies?
* How can I manage my symptoms?

**Complications**

A pheochromocytoma can lead to other health problems. The high blood pressure linked with a pheochromocytoma can damage organs, especially tissues of the heart and blood vessel system, brain and kidneys. This damage can cause dangerous conditions, including

Heart disease.

* Stroke.
* Kidney failure.
* Vision loss.

### **Cancerous tumors**

Rarely, a pheochromocytoma spreads to other parts of the body. This tumor is then considered cancerous, also called malignant, and is referred to as metastatic pheochromocytoma. Cancer cells from a pheochromocytoma or paraganglioma most often travel to the lymph system, bones, liver or lungs.

**REFERENCE**

<https://www.mayoclinic.org/diseases-conditions/pheochromocytoma/symptoms-causes/syc-20355367>

<https://my.clevelandclinic.org/health/diseases/23373-pheochromocytoma>

### **Hyperaldosteronism**

Hyperaldosteronism is a condition in which one or both of your adrenal glands produce too much aldosterone. Aldosterone is a hormone that helps regulate your blood pressure by controlling the levels of potassium and sodium in your blood.

Your adrenal glands are part of your endocrine system. They make hormones your body needs to carry out daily functions. You have two adrenal glands — one atop each kidney.

There are two main types of hyperaldosteronism:

**Primary hyperaldosteronism (Conn’s syndrome)**: An issue within your adrenal glands causes them to release too much aldosterone.

* **Secondary hyperaldosteronism**: An issue somewhere else in your body causes your adrenal glands to make too much aldosterone.

Hyperaldosteronism causes high blood pressure (hypertension) and low potassium levels in your blood.

### **Who does hyperaldosteronism affect?**

Hyperaldosteronism mostly affects people 30 to 50 years old. It more often affects women than men.

**How common is hyperaldosteronism?**

It’s difficult for researchers to estimate how common hyperaldosteronism is. Some studies suggest that 5% to 10% of people with high blood pressure have primary hyperaldosteronism. Experts estimate that as many as 25% of people who have medication-resistant high blood pressure may have hyperaldosteronism.

**Symptoms**

The symptoms of hyperaldosteronism can vary based on the severity of the condition. Some people with mild cases of hyperaldosteronism have no symptoms (are asymptomatic).

The most common symptom of hyperaldosteronism is high blood pressure (hypertension), especially medication-resistant hypertension.

If you experience other symptoms, they’ll probably be caused by having moderate to severe high blood pressure and/or low potassium levels (hypokalemia).

Symptoms of high blood pressure include:

Headaches.

* Dizziness.
* Vision changes.
* Difficulty breathing.

Symptoms of low potassium include:

Muscle weakness (which can lead to temporary paralysis in severe cases).

Muscle spasms.

Tingling and numbness.

Fatigue.

* Extreme thirst (polydipsia).
* Frequent urination (peeing).

### **causes**

### Hyperaldosteronism has different causes depending on the type: primary or secondary.

### **Causes of primary hyperaldosteronism**

Primary hyperaldosteronism happens when there’s an issue within your adrenal glands that causes them to produce too much aldosterone.

Adrenal adenomas (noncancerous tumors) are the most common cause of primary hyperaldosteronism.

Rarer causes of primary hyperaldosteronism include:

Unilateral adrenal hyperplasia (one enlarged adrenal gland).

Aldosterone-producing adrenocortical carcinomas (cancerous tumors).

Familial hyperaldosteronism type 1 (a condition you inherit from your biological parents).

**Causes of secondary hyperaldosteronism**

Reduced blood flow to your kidneys causes secondary aldosteronism.

To understand why this happens, it’s important to know that aldosterone is part of a complex chain of hormone reactions that regulates your blood pressure.

This is known as the renin-angiotensin-aldosterone system, and it involves the following steps:

Your kidneys release [renin](https://my.clevelandclinic.org/health/body/22506-renin) (an enzyme) when your body detects low blood pressure or low sodium in your blood. Renin converts angiotensinogen (a precursor of angiotensin that’s produced by your liver) to angiotensin I (angiotensin is a hormone that narrows your blood vessels).

Angiotensin I is converted to angiotensin II.

Angiotensin II narrows your blood vessels and stimulates the release of aldosterone.

Reduced blood flow to your kidneys “mistakenly” triggers the renin-angiotensin-aldosterone system, which results in excess aldosterone in your body.

Causes of reduced kidney blood flow and secondary hyperaldosteronism include:

Obstructive renal artery disease.

Renal hypertension.

Conditions that cause fluid retention (edema), such as heart failure, cirrhosis of the liver and nephrotic syndrome.

**Diagnosis and Tests**

A healthcare provider will diagnose hyperaldosteronism with blood tests. However, many people never have hyperaldosteronism diagnosed because several conditions and risk factors can cause high blood pressure.

General signs of hyperaldosteronism include medication-resistant high blood pressure and the following results of an electrolyte blood panel:

Mildly high sodium level (hypernatremia).

Mildly low magnesium level (hypomagnesemia).

If your healthcare provider thinks you might have hyperaldosteronism based on these signs and your symptoms, they’ll likely order one of two blood tests: plasma renin concentration (PRC) or plasma renin activity (PRA).

If you have primary hyperaldosteronism, your PRC and PRA levels will be lower than normal. In secondary hyperaldosteronism, the levels will be higher than normal.

You may also need an aldosterone suppression test. This test involves consuming a certain amount of sodium (salt) orally or through an IV over a certain amount of time. You’ll then provide urine (pee) samples over a 24-hour period so that a laboratory can measure the amount of aldosterone in your pee.

If these tests confirm you have hyperaldosteronism, your provider will order additional tests to determine the cause. For example, they may recommend an imaging test such as a CT (computed tomography) scan to check for a tumor that could be causing hyperaldosteronism.

## **Management and Treatment**

The treatment of hyperaldosteronism depends on what’s causing it. But the main goal is to manage your blood pressure.

Healthcare providers usually recommend treating primary hyperaldosteronism caused by an adrenal gland tumor by surgically removing the tumor. In some cases, these tumors can be treated with only medication. Even after surgery, you might still have high blood pressure and need to take medicine to manage it.

Providers treat secondary hyperaldosteronism by managing your blood pressure with medications and treating the underlying cause (such as heart failure).

Medications that can help treat hyperaldosteronism include:

Spironolactone (Aldactone®).

Eplerenone (Inspra®).

Amiloride (Midamor®).

Men may experience erectile dysfunction and gynecomastia (enlarged male breast tissue) with long-term use of medicines that block the effects of aldosterone, such as spironolactone.

**Prevention**

### **Can hyperaldosteronism be prevented?**

In most cases, there’s nothing you can do to prevent hyperaldosteronism.

## **Outlook / Prognosis**

The prognosis of hyperaldosteronism varies depending on what caused it.

The outlook for primary hyperaldosteronism is generally good if it’s diagnosed and treated early. The outlook for secondary hyperaldosteronism depends on the cause of the condition.

The most common complications of hyperaldosteronism are cardiovascular issues caused by high blood pressure, including:

Atrial fibrillation.

Left ventricular hypertrophy.

Heart attack.

Stroke.

**Living With**

### **When should I see my healthcare provider about hyperaldosteronism?**

If you’ve been diagnosed with hyperaldosteronism, you’ll need to see your healthcare provider regularly to make sure your medication dosage is working.

Visit a healthcare provider if you notice any new symptoms or if your symptoms are changing.

nts

REFERENCE

https://my.clevelandclinic.org/health/diseases/24470-hypertensive-crisis

<https://www.mayoclinic.org/diseases-conditions/kidney-stones/diagnosis-treatment/drc-20355759>

**URINARY TRACT INFECTION**

**Definition and description**

A urinary tract infection (UTI) is an infection of your urinary system. This type of infection can involve your:

* Urethra (urethritis).
* Kidneys (pyelonephritis).
* Bladder (cystitis).

Urine (pee) is a byproduct of your blood-filtering system, which your kidneys perform. Your kidneys create pee when they remove waste products and excess water from your blood. Pee usually moves through your urinary system without any contamination. However, bacteria can get into your urinary system, which can cause UTIs.

#### **What is the urinary tract?**

The urinary tract makes and stores pee. It includes your:

* Kidneys. Kidneys are small, bean-shaped organs on the back of your body, above your hips. Most people have two kidneys. They filter water and waste products from your blood, which becomes pee. Common wastes include urea and creatinine.
* Ureters. Your ureters are thin tubes that carry pee from your kidneys to your bladder.
* Bladder. Your bladder is a balloon-like organ that stores pee before it leaves your body.
* Urethra. The urethra is a tube that carries pee from your bladder to the outside of your body.

#### **How common are urinary tract infections?**

UTIs are very common, especially in females. About half of females will have a UTI at some point during their lives. Males can also get UTIs, as well as children, though they only affect 1% to 2% of children. Healthcare providers treat 8 million to 10 million people each year for UTIs.

**Causes**

UTIs typically occur when bacteria enter the urinary tract through the urethra and begin to spread in the bladder. The urinary system is designed to keep out bacteria. But the defenses sometimes fail. When that happens, bacteria may take hold and grow into a full-blown infection in the urinary tract.

The most common UTIs occur mainly in women and affect the bladder and urethra.

* **Infection of the bladder.** This type of UTI is usually caused by Escherichia coli (E. coli). E. coli is a type of bacteria commonly found in the gastrointestinal (GI) tract. But sometimes other bacteria are the cause.  
  Having sex also may lead to a bladder infection, but you don't have to be sexually active to develop one. All women are at risk of bladder infections because of their anatomy. In women, the urethra is close to the anus. And the urethral opening is close to the bladder. This makes it easier for bacteria around the anus to enter the urethra and to travel to the bladder.
* **Infection of the urethra.** This type of UTI can happen when GI bacteria spread from the anus to the urethra. An infection of the urethra can also be caused by sexually transmitted infections. They include herpes, gonorrhea, chlamydia and mycoplasma. This can happen because women's urethras are close to the vagina.

**Risk factors**

UTIs are common in women. Many women experience more than one UTI during their lifetimes.

Risk factors for UTIs that are specific to women include:

* **Female anatomy.** Women have a shorter urethra than men do. As a result, there's less distance for bacteria to travel to reach the bladder.
* **Sexual activity.** Being sexually active tends to lead to more UTIs. Having a new sexual partner also increases risk.
* **Certain types of birth control.** Using diaphragms for birth control may increase the risk of UTIs. Using spermicidal agents also can increase risk.
* **Menopause.** After menopause, a decline in circulating estrogen causes changes in the urinary tract. The changes can increase the risk of UTIs.

Other risk factors for UTIs include:

* **Urinary tract problems.** Babies born with problems with their urinary tracts may have trouble urinating. Urine can back up in the urethra, which can cause UTIs.
* **Blockages in the urinary tract.** Kidney stones or an enlarged prostate can trap urine in the bladder. As a result, the risk of UTIs is higher.
* **A suppressed immune system.** Diabetes and other diseases can impair the immune system — the body's defense against germs. This can increase the risk of UTIs.
* **Catheter use.** People who can't urinate on their own often must use a tube, called a catheter, to urinate. Using a catheter increases the risk of UTIs. Catheters may be used by people who are in the hospital. They may also be used by people who have neurological problems that make it difficult to control urination or who are paralyzed.
* **A recent urinary procedure.** Urinary surgery or an exam of your urinary tract that involves medical instruments can both increase the risk of developing a UTI.

**Signs and symptoms**

A UTI causes inflammation in the lining of your urinary tract. The inflammation may cause the following problems:

* Pain in your flank, abdomen, pelvic area or lower back.
* Pressure in the lower part of your pelvis.
* Cloudy, foul-smelling pee.
* Urinary incontinence.
* Frequent urination.
* Urge incontinence.
* Pain when you pee (dysuria).
* Blood in your pee (hematuria).

Other UTI-associated symptoms may include:

* Pain in your penis.
* Feeling extremely tired (fatigue).
* Fever.
* Chills.
* Nausea and vomiting.
* Mental changes or confusion.

**Diagnosis methods (tests, lab work, imaging, etc.)**

Tests and procedures used to diagnose urinary tract infections include:

* **Analyzing a urine sample.** Your health care provider may ask for a urine sample. The urine will be looked at in a lab to check for white blood cells, red blood cells or bacteria. You may be told to first wipe your genital area with an antiseptic pad and to collect the urine midstream. The process helps prevent the sample from being contaminated.
* **Growing urinary tract bacteria in a lab.** Lab analysis of the urine is sometimes followed by a urine culture. This test tells your provider what bacteria are causing the infection. It can let your provider know which medications will be most effective.
* **Creating images of the urinary tract.** Recurrent UTIs may be caused by a structural problem in the urinary tract. Your health care provider may order an ultrasound, a CT scan or MRI to look for this issue. A contrast dye may be used to highlight structures in your urinary tract.
* **Using a scope to see inside the bladder.** If you have recurrent UTIs, your health care provider may perform a cystoscopy. The test involves using a long, thin tube with a lens, called a cystoscope, to see inside the urethra and bladder. The cystoscope is inserted in the urethra and passed through to the bladder.

**Treatment**

Antibiotics usually are the first treatment for urinary tract infections. Your health and the type of bacteria found in your urine determine which medicine is used and how long you need to take it.

### **Simple infection**

Medicines commonly used for simple UTIs include:

* Trimethoprim and sulfamethoxazole (Bactrim, Bactrim DS)
* Fosfomycin (Monurol)
* Nitrofurantoin (Macrodantin, Macrobid, Furadantin)
* Cephalexin
* Ceftriaxone

The group of antibiotics known as fluoroquinolones isn't commonly recommended for simple UTIs. These drugs include ciprofloxacin (Cipro), levofloxacin and others. The risks of these drugs generally outweigh the benefits for treating uncomplicated UTIs.

In cases of a complicated UTI or kidney infection, your health care provider might prescribe a fluoroquinolone medicine if there are no other treatment options.

Often, UTI symptoms clear up within a few days of starting treatment. But you may need to continue antibiotics for a week or more. Take all of the medicine as prescribed.

For an uncomplicated UTI that occurs when you're otherwise healthy, your health care provider may recommend a shorter course of treatment. That may mean taking an antibiotic for 1 to 3 days. Whether a short course of treatment is enough to treat your infection depends on your symptoms and medical history.

Your health care provider also may give you a pain reliever to take that can ease burning while urinating. But pain usually goes away soon after starting an antibiotic.

### **Frequent infections**

If you have frequent UTIs, your health care provider may recommend:

* Low-dose antibiotics. You might take them for six months or longer.
* Diagnosing and treating yourself when symptoms occur. You'll also be asked to stay in touch with your provider.
* Taking a single dose of antibiotic after sex if UTIs are related to sexual activity.
* Vaginal estrogen therapy if you've reached menopause.

### **Severe infection**

For a severe UTI, you may need IV antibiotics in a hospital

If your infection doesn’t respond to treatment, a provider may order the following tests to examine your urinary tract for a disease or injury:

* Ultrasound. An ultrasound is an imaging test that helps your provider look at your internal organs. An ultrasound is painless and doesn’t require any preparation.
* Computed tomography (CT) scan. A CT scan is another imaging test. It’s a type of X-ray that takes cross-section images of your body — like slices — that create 3D images of the inside of your body. A CT scan is more precise than a standard X-ray.
* Cystoscopy. A cystoscopy uses a cystoscope to look inside your bladder through your urethra. A cystoscope is a thin instrument with a lens and a light at the end.

**Treatment options**

What is the best thing to do for a urinary tract infection?

The best thing to do for a urinary tract infection is to see a healthcare provider. You need antibiotics to treat a UTI. Your provider will select an antibiotic that works best against the bacteria responsible for your infection.

Once you get a prescription for antibiotics, it’s very important that you follow the directions for taking them. Be sure to take the full course of antibiotics, even if your symptoms go away and you start feeling better. If you don’t finish all your medicine, the infection can return and be more challenging to treat.

If you get UTIs a lot, a provider may recommend that you take antibiotics:

Every day.

Every other day.

After sex.

At the first sign of symptoms.

Talk to a provider about your best treatment option if you have a history of frequent UTIs.

What specific antibiotics are used to treat a urinary tract infection?

Healthcare providers commonly prescribe the following antibiotics to treat UTIs:

Nitrofurantoin.

Sulfonamides (sulfa drugs), such as sulfamethoxazole/trimethoprim.

Amoxicillin.

Cephalosporins, such as cephalexin.

Doxycycline.

Fosfomycin.

Quinolones, such as ciprofloxacin or levofloxacin.

If you get UTIs often, a healthcare provider may give you low-dose antibiotics for a short time to prevent the infection from coming back. The provider may recommend this cautious approach to treat frequent UTIs because your body can develop resistance to the antibiotic, and you can get other types of infections, including C. diff colitis. This practice isn’t very common.

Can I become immune to the antibiotics used to treat a UTI?

Sort of. Every time you use antibiotics to treat a UTI, the infection adapts and can become harder to fight (antibiotic resistance). But the infection becomes immune to the antibiotics, not you. Antibiotics may not always be the best solution. As a result, a healthcare provider may suggest alternative treatments if you get frequent urinary tract infections. These may include:

Waiting. Your provider may suggest a “watch and wait” approach to your symptoms. During this time, it’s a good idea to drink plenty of fluids (especially water) to help flush out your system.

Intravenous (IV) treatment. In some complicated cases, a UTI may be resistant to antibiotics, or the infection may move to your kidneys. You may need treatment at a hospital, where providers will give you medicine through a needle they insert into a vein, usually in your arm (intravenously). Once you return home, you may need to take oral antibiotics for a period to rid yourself of the infection completely.

\* **Prevention tips**

#### **Does cranberry juice prevent a urinary tract infection?**

Cranberry juice that you can buy at the grocery store doesn’t prevent a UTI. However, cranberry extract supplements (vitamin pills) may decrease your chances of getting a UTI.

If you get UTIs often, methenamine hippurate is another nonantibiotic alternative that helps prevent infections.

#### **Can a UTI go away on its own?**

Minor urinary tract infections can sometimes get better on their own. However, most UTIs need antibiotics to go away. You absolutely need antibiotics if you have a UTI as well as:

* A fever.
* Chills.
* Nausea and vomiting.

### **How soon after treatment will I feel better?**

Most people feel better within a few days after starting antibiotics to treat a UTI.

The following lifestyle changes can help prevent urinary tract infections:

#### **Practice good hygiene**

Practicing good hygiene is one of the best ways to help prevent UTIs. This is especially important if you have a vagina because your urethra is much shorter, and it’s easier for *E. coli* to move from your rectum back into your body. Always wipe from front to back after a bowel movement (pooping) to avoid this.

During your menstrual cycle, it’s also a good idea to regularly change your period products, including pads and tampons. You should also avoid using any deodorants on your vagina.

#### **Drink plenty of fluids**

Drinking extra fluids — especially water — each day can help flush out bacteria from your urinary tract. Healthcare providers recommend drinking six to eight glasses of water daily.

#### **Change your peeing habits**

Peeing can play a big role in getting rid of bacteria from your body. Your pee is a waste product, and each time you empty your bladder, you help remove that waste from your body.

Peeing frequently can reduce your risk of developing an infection, especially if you get UTIs a lot.

You should also try to pee right before and right after having sex. Sex can introduce bacteria to your urethra, and peeing before and after sex helps flush it out. If you can’t pee, wash the area with warm water.

#### **Change your birth control**

You may have an increased risk of developing a UTI if you use a diaphragm for birth control. Talk to a healthcare provider about other birth control options.

#### **Use a water-based lubricant during sex**

If you use lubricant during sex, make sure it’s water-based. You should also avoid spermicide if you have frequent UTIs.

#### **Change your clothing**

Tight-fitting clothing can create a moist environment, which promotes bacterial growth. You can try loose-fitting clothing and cotton underwear to prevent moisture from accumulating around your urethra.

#### **Medications**

If you’re postmenopausal, a healthcare provider may suggest a vaginal cream that contains estrogen. These creams may help reduce your risk of developing a UTI by changing the pH of your vagina. Talk to a healthcare provider if you’re postmenopausal and get a lot of UTIs.

Over-the-counter (OTC) supplements — including cranberry extract and probiotics — may also help prevent UTIs. Talk to a healthcare provider before you start taking any supplements.

## **Outlook / Prognosis**

The outlook for urinary tract infections is good. Most UTIs usually respond very well to treatment. A UTI can be annoying or uncomfortable before you start treatment. However, once a healthcare provider identifies the bacteria and prescribes the appropriate antibiotic, your symptoms should improve quickly.

It’s important to finish all of the antibiotics that your healthcare provider prescribes. If you have frequent UTIs or your symptoms aren’t improving, your provider may test to see if your infection is resistant to antibiotics. Antibiotic-resistant infections may require IV antibiotics or other treatments.

### **When should I go to the doctor for a UTI?**

Call a healthcare provider if you have symptoms of a UTI. Call them again if they diagnosed you with a UTI and your symptoms worsen. You may need a different treatment.

#### **When should I go to the ER?**

Go to the emergency room if you have a UTI and develop the following symptoms:

* Fever.
* Back pain.
* Vomiting.

### **What questions should I ask my healthcare provider?**

* How do you know that I have a urinary tract infection?
* If I don’t have a urinary tract infection, what other condition do I have?
* What bacteria are responsible for my urinary tract infection?
* What antibiotic will you prescribe to treat my urinary tract infection?
* Are there any special directions I need to follow while taking antibiotics?
* How long will it take to feel better?
* Do I need to schedule a follow-up appointment?
* What can I do to help relieve my symptoms at home?

## **Additional Common Questions**

### **What is the difference between a urinary tract infection and cystitis?**

A UTI is an infection in your urinary system, which may include your kidneys, ureters, bladder or urethra. Cystitis is a type of UTI. It’s an infection in your bladder, and it’s the most common type of UTI.

### **How can you tell the difference between a urinary tract infection and a bladder infection?**

A urinary tract infection is a more general type of infection. There are many parts of your urinary tract. A UTI is a term for an infection that takes place throughout your urinary tract.

A bladder infection is a specific infection that makes its way to your bladder and causes inflammation. Another name for a bladder infection is cystitis.

Not all UTIs become bladder infections. It’s important to treat a UTI quickly to prevent the infection from spreading to other areas of your urinary tract. The infection can spread not only to your bladder but also to your kidneys. Kidney infections are a more complicated type of infection. Another name for a kidney infection is pyelonephritis.

### **Types of urinary tract infections**

Each type of UTI may result in more-specific symptoms. The symptoms depend on which part of the urinary tract is affected.

| **Part of urinary tract affected** | **Signs and symptoms** |
| --- | --- |
| Kidneys | * Back or side pain * High fever * Shaking and chills * Nausea * Vomiting |
| Bladder | * Pelvic pressure * Lower belly discomfort * Frequent, painful urination * Blood in urine |
| Urethra | * Burning with urination * Discharge |

## **Complications**

When treated promptly and properly, lower urinary tract infections rarely lead to complications. But left untreated, UTIs can cause serious health problems.

Complications of a UTI may include:

* Repeated infections, which means you have two or more UTIs within six months or three or more within a year. Women are especially prone to having repeated infections.
* Permanent kidney damage from a kidney infection due to an untreated UTI.
* Delivering a low birth weight or premature infant when a UTI occurs during pregnancy.
* A narrowed urethra in men from having repeated infections of the urethra.
* Sepsis, a potentially life-threatening complication of an infection. This is a risk especially if the infection travels up the urinary tract to the kidneys.

## **Lifestyle and home remedies**

Urinary tract infections can be painful, but you can take steps to ease discomfort until antibiotics treat the infection. Follow these tips:

* **Drink plenty of water.** Water helps to dilute your urine and flush out bacteria.
* **Avoid drinks that may irritate your bladder.** Avoid coffee, alcohol, and soft drinks containing citrus juices or caffeine until the infection has cleared. They can irritate your bladder and tend to increase the need to urinate.
* **Use a heating pad.** Apply a warm, but not hot, heating pad to your belly to help with bladder pressure or discomfort.

**Alternative medicine**

Many people drink cranberry juice to prevent UTIs. There's some indication that cranberry products, in either juice or tablet form, may have properties that fight an infection. Researchers continue to study the ability of cranberry juice to prevent UTIs, but results aren't final.

There's little harm in drinking cranberry juice if you feel it helps you prevent UTIs, but watch the calories. For most people, drinking cranberry juice is safe. However, some people report an upset stomach or diarrhea.

But don't drink cranberry juice if you're taking blood-thinning medication, such as warfarin (Jantovin).

**Preparing for your appointment**

Your primary care provider, nurse practitioner or other health care provider can treat most UTIs. If you have frequent UTIs or a chronic kidney infection, you may be referred to a health care provider who specializes in urinary disorders. This type of doctor is called a urologist. Or you may see a health care provider who specializes in kidney disorders. This type of doctor is called a nephrologist.

### **What to expect from your doctor**

Your health care provider will likely ask you several questions, including:

* When did you first notice your symptoms?
* Have you ever been treated for a bladder or kidney infection?
* How severe is your discomfort?
* How often do you urinate?
* Are your symptoms relieved by urinating?
* Do you have low back pain?
* Have you had a fever?
* Have you noticed vaginal discharge or blood in your urine?
* Are you sexually active?
* Do you use contraception? What kind?
* Could you be pregnant?
* Are you being treated for any other medical conditions?
* Have you ever used a catheter?

Unusual urinary cloudiness (turbidity) and odor can be influenced by several factors, including:

* Amorphous phosphates
* Foods (see below)
* Hormonal changes (eg, pregnancy)
* Hydration status
* Liver failure
* Medications (eg, sulfonylurea)
* Renal failure
* Sexually transmitted infections
* Trimethylaminuria
* Vaginal infections
* Vitamins
* Voiding dysfunction unrelated to infection

Foods that can cause urinary odor include:

* Asparagus
* Brussels sprouts
* Fish (eg, salmon)
* Garlic
* Onions
* Spices
* Sulfur-containing foods

**Differential diagnosis**

The differential diagnoses of an uncomplicated UTI include:

* Bladder stones
* Complicated UTI
* Food or dietary issues
* Herpes simplex infection
* Medication adverse effects
* Overactive bladder
* Pelvic inflammatory disease
* Prostatitis
* Pyelonephritis
* Recurrent UTI
* Relapsing UTI
* Renal infarction
* Renal stones
* Sexually transmitted infections
* Urethritis
* Vaginitis

## Bladder Stones

Bladder stones are hard mineral concretions that form in the urinary bladder, typically when urine remains in the bladder for prolonged periods, allowing minerals like salt and protein waste to crystallize and aggregate. They are more common in men, especially older men with conditions causing incomplete bladder emptying such as benign prostatic hyperplasia (BPH) or neurogenic bladder.

Symptoms:

* Lower abdominal pain or discomfort
* Pain or burning sensation during urination
* Frequent urination, especially at night
* Difficulty starting urination or interrupted urine flow
* Blood in the urine (hematuria)
* Cloudy or dark urine
* Urinary incontinence or retention in some cases

Causes and Risk Factors:

* Urinary stasis due to bladder outlet obstruction (e.g., enlarged prostate)
* Neurogenic bladder from nerve damage (stroke, spinal injury)
* Recurrent urinary tract infections (UTIs) causing bladder inflammation
* Foreign bodies or medical devices in the bladder
* Kidney stones that migrate into the bladder and grow
* Dehydration leading to concentrated urine

Complications:

* Recurrent UTIs
* Bladder inflammation or damage
* Urinary retention

Diagnosis:

* History and physical exam (including rectal exam in men)
* Urinalysis and urine culture
* Imaging: ultrasound, X-ray, CT scan
* Cystoscopy for direct visualization

Treatment:

* Small stones may pass spontaneously with increased fluid intake
* Larger stones often require removal via cystolitholapaxy (endoscopic stone fragmentation and extraction) or, rarely, open surgery
* Addressing underlying causes (e.g., prostate enlargement) is essential to prevent recurrence
* Antibiotics may be needed if infection is present

**Complicated Urinary Tract Infection (UTI)**

A UTI associated with structural or functional abnormalities of the urinary tract, or occurring in patients with comorbidities, often requiring more aggressive treatment.

**Food or Dietary Issues**

Certain dietary factors can influence urinary tract health, stone formation, or infection risk.

**Herpes Simplex Infection**

A viral infection causing painful genital or urethral lesions, sometimes associated with urethritis.

**Medication Adverse Effects**

Some drugs can cause urinary symptoms or predispose to infections or stone formation.

**Overactive Bladder**

A syndrome characterized by urinary urgency, frequency, and sometimes urge incontinence without infection or other obvious pathology.

**Pelvic Inflammatory Disease (PID)**

Infection of the female upper genital tract causing pelvic pain, fever, and vaginal discharge; may affect urinary symptoms.

**Prostatitis**

Inflammation or infection of the prostate gland causing pelvic pain, urinary symptoms, and sometimes systemic signs.

**Pyelonephritis**

A bacterial infection of the kidney and renal pelvis presenting with flank pain, fever, and urinary symptoms.

**Recurrent UTI**

Repeated episodes of urinary tract infection requiring evaluation for underlying causes.

**Relapsing UTI**

Reinfection with the same organism shortly after treatment, often indicating persistent source.

**Renal Infarction**

Ischemic injury to kidney tissue due to arterial occlusion, causing flank pain and impaired renal function.

**Renal Stones**

Calculi formed in the kidneys that may cause pain, hematuria, and obstruction.

**Sexually Transmitted Infections (STIs)**

Infections transmitted sexually that can cause urethritis, vaginitis, and other urinary symptoms.

**Urethritis**

Inflammation of the urethra, often due to infection, causing dysuria and discharge.

**Vaginitis**

Inflammation of the vagina causing discharge, itching, and discomfort.

## **Epidemiology**

## UTIs occur at least 4 times more frequently in females than males. The following are key statistics regarding UTIs:

* Approximately 40% of women in the United States will develop a UTI during their lifetime.
* Approximately 10% of women experience a UTI annually.
* Recurrences are common, with nearly 50% of patients experiencing a second infection within a year.
* UTIs are most common in women between the ages of 16 and 35.

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**KIDNEY CANCER**

**Definition and description**

Kidney cancer is a growth of cells that starts in the kidneys. The kidneys are two bean-shaped organs, each about the size of a fist. They're located behind the abdominal organs, with one kidney on each side of the spine.

In adults, renal cell carcinoma is the most common type of kidney cancer. Other, less common types of kidney cancer can happen. Young children are more likely to develop a kind of kidney cancer called Wilms tumor.

The number of kidney cancers diagnosed each year seems to be increasing. One reason for this may be the fact that imaging techniques such as CT scans are being used more often. These tests may lead to the incidental discovery of more kidney cancers. Kidney cancer is often found when the cancer is small and confined to the kidney.

**Causes**

It's not clear what causes most kidney cancers.

Kidney cancer happens when cells in the kidney develop changes in their DNA. A cell's DNA holds the instructions that tell the cell what to do. In healthy cells, the DNA gives instructions to grow and multiply at a set rate. The instructions tell the cells to die at a set time. In cancer cells, the DNA changes give different instructions. The changes tell the cancer cells to make many more cells quickly. Cancer cells can keep living when healthy cells die. This causes too many cells.

The cancer cells form a mass called a tumor. The tumor can grow to invade and destroy healthy body tissue. In time, cancer cells can break away and spread to other parts of the body. When cancer spreads, it's called metastatic cancer.

**Risk factors**

Factors that may increase the risk of kidney cancer include:

* **Older age.** The risk of kidney cancer increases with age.
* **Smoking tobacco.** People who smoke have a greater risk of kidney cancer than those who don't. The risk decreases after quitting.
* **Obesity.** People who are obese have a higher risk of kidney cancer than people who are considered to have a healthy weight.
* **High blood pressure.** High blood pressure, also called hypertension, increases the risk of kidney cancer.
* **Certain inherited conditions.** People who are born with certain inherited conditions may have an increased risk of kidney cancer. These conditions may include von Hippel-Lindau disease, Birt-Hogg-Dube syndrome, tuberous sclerosis complex, hereditary papillary renal cell carcinoma and familial renal cancer.
* **Family history of kidney cancer.** The risk of kidney cancer is higher if a blood relative, such as a parent or sibling, has had the disease.

**Symptoms**

Kidney cancer doesn't usually cause symptoms at first. In time, signs and symptoms may develop, including:

* Blood in the urine, which may appear pink, red or cola colored.
* Loss of appetite.
* Pain in the side or back that doesn't go away.
* Tiredness.
* Unexplained weight loss.

**Diagnosis methods (tests, lab work, imaging, etc.)**

Kidney cancer diagnosis often begins with a physical exam and a discussion of your health history. Blood and urine tests as well as imaging tests may be used. A sample of tissue may be taken for lab testing.

Tests and procedures used to diagnose kidney cancer include:

### **Blood and urine tests**

Tests of your blood and urine may give your healthcare team clues about what's causing your symptoms. Blood tests may check for the number of red blood cells in the body. Urine tests may look for substances in the urine, such as blood, bacteria and cancer cells.

### **Imaging tests**

Imaging tests make pictures of the body. They can show the location and size of kidney cancer. Tests might include ultrasound, CT or MRI.

### **Biopsy**

A biopsy is a procedure to remove a sample of tissue for testing in a lab. For kidney cancer, a thin needle is inserted into the kidney or other body part such as the lymph nodes. A healthcare professional uses the needle to remove a sample of tissue. A biopsy may not be needed if imaging tests show enough information to make a diagnosis.

### **Kidney cancer staging**

If you're diagnosed with kidney cancer, the next step is to determine the cancer's extent, called the stage. Your healthcare team uses the cancer staging test results to help create your treatment plan. Staging tests for kidney cancer may include additional CT and MRI scans.

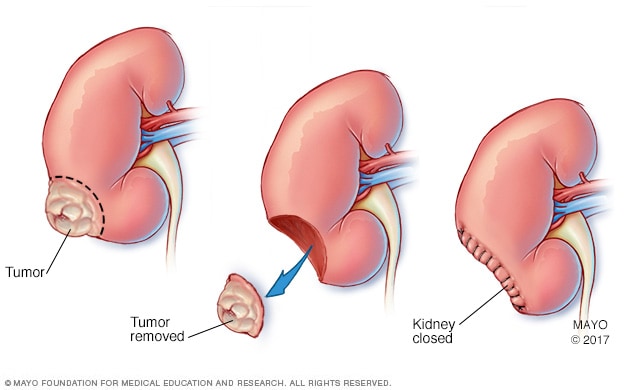
The stages of kidney cancer range from 1 to 4. A stage 1 kidney cancer is small and confined to the kidney. As the cancer gets larger, the stages get higher. A stage 4 kidney cancer has grown beyond the kidney or spread to other parts of the body.

**Treatment options**

Kidney cancer treatment sometimes begins with surgery to remove the cancer. For cancers confined to the kidney, this may be the only treatment needed. Sometimes medicine is given after surgery to lower the risk that the cancer will come back. If the cancer has spread beyond the kidney, surgery might not be possible. Other treatments may be recommended.

Your healthcare team considers many factors when creating a treatment plan. These factors may include your overall health, the type and stage of your cancer, and your preferences.

### **Surgery**

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**Partial nephrectomy**

For most cancers confined to the kidney, surgery is the first treatment. The goal of surgery is to remove the cancer while preserving kidney function, when possible. Operations used to treat kidney cancer include:

* **Removing the affected kidney.** A complete nephrectomy, also known as a radical nephrectomy, involves removing the entire kidney and a border of healthy tissue around it. Nearby tissues such as the lymph nodes, adrenal gland or other structures also may be removed.  
  The surgeon may perform a nephrectomy through a single incision in the abdomen or side, called an open nephrectomy. The surgeon also may use a series of small incisions in the abdomen, known as laparoscopic or robot-assisted laparoscopic nephrectomy.
* **Removing the cancer from the kidney.** A partial nephrectomy involves removing the cancer and a small margin of healthy tissue that surrounds it rather than the entire kidney. This procedure also is called kidney-sparing or nephron-sparing surgery. It can be done as an open procedure, laparoscopically or with robotic assistance.  
  Kidney-sparing surgery is a common treatment for small kidney cancers and it may be an option if you have only one kidney. When possible, kidney-sparing surgery is generally preferred over a complete nephrectomy to preserve kidney function. It also may reduce the risk of later complications, such as kidney disease and the need for dialysis.

The type of surgery you have is based on your cancer and its stage, as well as your overall health.

### **Cryoablation**

Cryoablation is a treatment to freeze cancer cells. During cryoablation, a special hollow needle is inserted through the skin and into the kidney cancer using ultrasound or other image guidance. Cold gas in the needle is used to freeze the cancer cells.

Cryoablation can treat small kidney cancers in certain situations. It might be used in people who have other health concerns that make surgery risky.

### **Radiofrequency ablation**

Radiofrequency ablation is a treatment to heat cancer cells. During radiofrequency ablation, a special probe is inserted through the skin and into the kidney cancer using ultrasound or other imaging to guide placement of the probe. An electrical current is run through the needle and into the cancer cells. This causes the cells to heat up or burn.

Radiofrequency ablation can treat small kidney cancers in certain situations. It might be used in people who have other health concerns that make surgery risky.

### **Radiation therapy**

Radiation therapy treats cancer with powerful energy beams. The energy can come from X-rays, protons or other sources. During radiation therapy, you lie on a table while a machine moves around you. The machine directs radiation to precise points on your body.

Radiation therapy can be used on the kidney to kill the cancer cells. It also can help control or reduce symptoms of kidney cancer that has spread to other areas of the body, such as the bones and brain.

### **Targeted therapy**

Targeted therapy for cancer is a treatment that uses medicines that attack specific chemicals in the cancer cells. By blocking these chemicals, targeted treatments can cause cancer cells to die.

### **Immunotherapy**

Immunotherapy for cancer is a treatment with medicine that helps the body's immune system kill cancer cells. The immune system fights off cancer and other cells that shouldn't be in the body. Cancer cells survive by hiding from the immune system. Immunotherapy helps the immune system cells find and kill the cancer cells.

For kidney cancer, immunotherapy may be used after surgery to kill any cancer cells that might remain. It also may be used when the cancer grows very large or spreads to other parts of the body.

### **Chemotherapy**

Chemotherapy treats cancer with strong medicines. Many chemotherapy medicines exist. Most are given through a vein. Usually, kidney cancers are resistant to chemotherapy. However, it may be used for certain rare types of kidney cancer.

### **Palliative care**

Palliative care is a special type of healthcare that helps you feel better when you have a serious illness. If you have cancer, palliative care can help relieve pain and other symptoms. A healthcare team that may include doctors, nurses and other specially trained health professionals provides palliative care. The care team's goal is to improve the quality of life for you and your family.

Palliative care specialists work with you, your family and your care team. They provide an extra layer of support while you have cancer treatment. You can have palliative care at the same time you're getting strong cancer treatments, such as surgery, chemotherapy, immunotherapy, targeted therapy or radiation therapy.

The use of palliative care with other proper treatments can help people with cancer feel better and live longer.

**Alternative medicine**

Alternative medicine therapies can't cure kidney cancer. But some integrative treatments can be combined with your healthcare team's care to help you cope with side effects of cancer and its treatment, such as distress.

People with cancer often experience distress. If you're distressed, you may have difficulty sleeping and find yourself constantly thinking about your cancer.

Discuss your feelings with your healthcare team. Specialists can help you find ways of coping. In some cases, medicines may help.

Integrative medicine treatments also may help you feel better, including:

* Acupuncture.
* Art therapy.
* Exercise.
* Massage therapy.
* Meditation.
* Music therapy.
* Relaxation exercises.
* Spirituality.

Talk with your healthcare team if you're interested in these treatment options.

**Prevention**

There's no sure way to prevent kidney cancer, but you may reduce your risk if you:

### **Drink alcohol in moderation, if at all**

If you choose to drink alcohol, do so in moderation. For healthy adults, that means up to one drink a day for women and up to two drinks a day for men.

### **Eat more fruits and vegetables**

Choose a healthy diet with a variety of fruits and vegetables. Food sources of vitamins and nutrients are best. Avoid taking large doses of vitamins in pill form, as they may be harmful.

### **Exercise most days of the week**

Aim for at least 30 minutes of exercise on most days of the week. If you haven't been active lately, ask your healthcare professional whether it's OK and start slowly.

### **Maintain a healthy weight**

If your weight is healthy, work to maintain that weight. If you need to lose weight, ask a healthcare professional about healthy ways to lower your weight. Eat fewer calories and slowly increase the amount of exercise.

### **Stop smoking**

Talk with your healthcare team about strategies and aids that can help you quit. Options include nicotine replacement products, medicines and support groups. If you've never smoked, don't start.

### **Control high blood pressure**

Ask your healthcare professional to check your blood pressure at your next appointment. If your blood pressure is high, you can discuss options for lowering your numbers. Lifestyle measures such as exercise, weight loss and diet changes can help. Some people may need to add medicines to lower their blood pressure. Discuss your options with your healthcare team.

**Prognosis**

If you have kidney cancer, you may have questions about your prognosis. A prognosis is the doctor’s best estimate of how cancer will affect someone and how it will respond to treatment. Prognosis and survival depend on many factors. Only a doctor familiar with your medical history, the type and stage and other features of the cancer, the treatments chosen and the response to treatment can put all of this information together with survival statistics to arrive at a prognosis.

A prognostic factor is an aspect of the cancer that the doctor will consider when making a prognosis. A predictive factor influences how a cancer will respond to a certain treatment. Prognostic and predictive factors are often discussed together. They both play a part in deciding on a treatment plan and a prognosis.

The following are prognostic and predictive factors for kidney cancer.

## **Stage**

The stage of kidney cancer is the most important prognostic factor. People who have tumours that are only in the kidney have a better prognosis than people with cancer that has spread outside the kidney.

## **Grade**

Low-grade tumours have a better prognosis than high-grade tumours. Low-grade tumours are less likely to spread because they grow slowly. High-grade tumours are more aggressive and tend to spread quickly.

## **Type of kidney cancer**

Papillary and chromophobe types of renal cell carcinoma have a better prognosis because they are often low grade.

Collecting duct carcinoma and renal medullary carcinoma have a poor prognosis because they are often very aggressive.

## **Level of risk**

The most common system used to predict prognosis for people with metastatic renal cell carcinoma is the International Metastatic Renal Cell Carcinoma Database Consortium (IMDC). It uses 6 predictors:

* The Karnofsky performance status scale
* is less than 80.
* There was less than 1 year between diagnosis and treatment of metastatic cancer.
* The blood calcium level is abnormally high.
* The red blood cell count is lower than normal (called anemia).
* The platelet count is greater than normal (called thrombocytosis).
* There is an abnormally high level of neutrophils
* ( called neutrophilia).

These predictors are combined to develop a level of risk:

* Favourable risk means the person has none of the predictors.
* Intermediate risk means the person has 1 or 2 predictors.
* Poor risk means the person has 3 or more predictors.

## **Types**

### **Renal cell carcinoma**

Renal cell carcinoma (RCC) is the most common type of kidney cancer in adults. RCC usually starts in the lining of tiny tubes in the kidney called renal tubules. RCC often stays in the kidney, but it can spread to other parts of the body, most often the bones, lungs, or brain.

### **Clear cell renal cell carcinoma**

Clear cell renal cell carcinoma, also known as ccRCC or conventional renal cell carcinoma, is the most common form of kidney cancer. Clear cell renal cell carcinoma is named after how the tumor looks under the microscope. The cells in the tumor look clear, like bubbles.

In adults, ccRCC makes up about 80% of all renal cell carcinoma cases. ccRCC is more common in adults than children. Renal cell carcinoma makes up 2% to 6% of childhood and young adult kidney cancer cases."

### **Rare types of kidney cancer**

Rare kidney cancers occur most frequently in children, teenagers, and young adults.

**Papillary renal cell carcinoma (PRCC)**

* 15% of all renal cell carcinomas
* Tumor(s) located in the kidney tubes
* Type 1 PRCC is more common and grows slowly
* Type 2 PRCC is more aggressive and grows more quickly

**Translocation renal cell carcinoma (TRCC)**

* Accounts for 1% to 5% of all renal cell carcinomas and 20% of childhood causes
* Tumor(s) located in the kidney
* In children, TRCC usually grows slowly often without any symptoms
* In adults, TRCC tends to be aggressive and fast-growing

### **Benign (non-cancerous) kidney tumors**

Benign, or noncancerous kidney tumors grow in size but do not spread to other parts of the body and are not usually life-threatening. Surgical removal is the most common treatment and most tumors do come back.

**Papillary renal adenoma**

* The most common benign kidney tumor
* Tumors are small, slow growing, often without any symptoms
* Usually an incidental finding on an imaging test done for a different reason

**Oncocytoma**

* Tumors start in the cells of the kidney collecting ducts and tumors can grow in one of both kidneys
* The tumors can grow to a large size about the These tumours can grow quite large, starting at just over an inch (walnut) and growing up to 4 inches (grapefruit)

**Angiomyolipoma**

* Benign fatty tumors can also be be due to overgrowth of blood vessel and smooth muscle tissue cells
* Tumors are non-cancerous, but they can become very large and destroy surrounding tissue
* Tumors that are over an inch and a half can cause internal bleeding

**Complications**

Complications from kidney cancer may include:

* Kidney failure
* Local spread of the tumor with increasing pain
* Spread of the cancer to lung, liver, and bone

For kidney cancer, some basic questions to ask include:

* Do I have kidney cancer?
* What is the stage of my kidney cancer?
* Has my kidney cancer spread to other parts of my body?
* Will I need more tests?
* What are the treatment options?
* How much does each treatment increase my chances of a cure or prolong my life?
* What are the potential side effects of each treatment?
* How will each treatment affect my daily life?
* Is there one treatment option you believe is the best?
* What would you recommend to a friend or family member in my situation?
* Should I see a specialist?
* Are there any brochures or other printed material that I can take with me? What websites do you recommend?
* What will determine whether I should plan for a follow-up visit?

Don't hesitate to ask other questions.

### **What to expect from your doctor**

Be prepared to answer questions, such as:

* When did your symptoms begin?
* Have your symptoms been continuous or occasional?
* How severe are your symptoms?
* What, if anything, seems to improve your symptoms?
* What, if anything, appears to worsen your symptoms?

**Differential diagnosis**

The differentials include

Wilms tumor,

rhabdoid kidney disease,

polycystic kidney disease,

pheochromocytoma,

dysplastic kidney,

hydronephrosis,

nephroma,

angiomyolipoma and rhabdomyosarcoma.

## **Wilms Tumor (Nephroblastoma)**

Wilms tumor is the most common kidney cancer in children, typically diagnosed around 3.5 years of age. It presents most often as a painless abdominal mass, sometimes accompanied by abdominal pain, hematuria, hypertension, fever, or symptoms related to metastases (commonly lungs). Diagnosis involves imaging (ultrasound, CT, MRI) and histologic confirmation, usually after nephrectomy. Treatment typically includes surgical removal of the affected kidney (nephrectomy), chemotherapy (vincristine, dactinomycin, and others), and sometimes radiotherapy. Prognosis is generally excellent with modern multimodal therapy, with about 90% survival. Bilateral or very large tumors may require preoperative chemotherapy and nephron-sparing surgery.

**Rhabdoid Kidney Tumor**

A rare, aggressive renal tumor in young children characterized by rhabdoid cells. It has a poor prognosis compared to Wilms tumor and requires intensive multimodal treatment including surgery, chemotherapy, and radiotherapy.

**Polycystic Kidney Disease (PKD)**

A genetic disorder characterized by the development of numerous fluid-filled cysts in the kidneys, leading to progressive renal enlargement and dysfunction. Autosomal dominant PKD is the most common form, presenting in adulthood, while autosomal recessive PKD presents in infancy or childhood.

**Pheochromocytoma**

A rare catecholamine-secreting tumor arising from adrenal medulla chromaffin cells, causing episodic hypertension, headaches, sweating, and palpitations. Diagnosis involves biochemical testing for catecholamines and imaging. Surgical removal is the treatment of choice.

**Dysplastic Kidney**

A congenital malformation where the kidney develops abnormally with cysts and disorganized tissue, often leading to nonfunctional renal tissue. It may be unilateral or bilateral and can cause renal insufficiency.

**Hydronephrosis**

Dilation of the renal pelvis and calyces due to obstruction of urine flow, leading to increased pressure and potential kidney damage. Causes include congenital abnormalities, stones, tumors, or strictures.

**Nephroma**

A benign renal tumor, such as metanephric adenoma or congenital mesoblastic nephroma, typically presenting as a renal mass in children or adults.

**Angiomyolipoma**

A benign renal tumor composed of blood vessels, smooth muscle, and fat. It is often associated with tuberous sclerosis and may cause bleeding if large.

**Rhabdomyosarcoma**

A malignant tumor of skeletal muscle origin, commonly occurring in the genitourinary tract in children, including the bladder and prostate. It presents with mass effect symptoms and requires multimodal treatment including surgery, chemotherapy, and radiotherapy.

**Epidemiology data**

Rate of New Cases and Deaths per 100,000: The rate of new cases of kidney and renal pelvis cancer was 17.5 per 100,000 men and women per year. The death rate was 3.4 per 100,000 men and women per year. These rates are age-adjusted and based on 2018–2022 cases and 2019–2023 deaths.

Lifetime Risk of Developing Cancer: Approximately 1.8 percent of men and women will be diagnosed with kidney and renal pelvis cancer at some point during their lifetime, based on 2018–2021 data, excluding 2020 due to COVID.

Prevalence of This Cancer: In 2022, there were an estimated 676,631 people living with kidney and renal pelvis cancer in the United States.

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## **pyelonephritis**

**DEFINITION AND DESCRIPTION**

Pyelonephritis is the medical term for a kidney infection. Most often it is caused by bacteria that have traveled to the kidney from an infection in the bladder.

Bladder infections are much more common in women compared to me. That's in large part because the distance to the bladder from skin, where bacteria normally live, is quite short and direct. While most bacterial bladder infections stay in the bladder, the high incidence of female bladder infections means women will also be more prone to pyelonephritis.

A woman is more likely to develop pyelonephritis when she is pregnant. Pyelonephritis and other forms of urinary tract infection increase the risk of premature delivery. A man is more likely to develop the problem if his prostate is enlarged, a common condition after age 50.

Both men and women are more likely to develop pyelonephritis if they have any of the following conditions:

* an untreated urinary tract infection
* diabetes
* nerve problems that affect the bladder
* kidney stones
* a bladder tumor
* abnormal backflow of urine from the bladder to the kidneys, called vesicoureteral reflux
* an obstruction related to an abnormal development of the urinary tract.

Tests or procedures that involve the insertion of an instrument into the bladder also increase the risk of urinary tract infections and pyelonephritis.

Children sometimes develop pyelonephritis because of an abnormality in the bladder that allows urine there to flow backward (reflux) into the ureter, the connection between the kidney and bladder. This can lead to scarring of the kidney.

## **Symptoms of pyelonephritis**

The two primary symptoms of pyelonephritis are pain in one flank, the area just beneath the lower ribs in the back, and fever. The pain can travel around the side toward the lower abdomen. There also can be shaking chills and nausea and vomiting. The urine may be cloudy, tinged with blood or unusually strong or foul-smelling. You may need to urinate more often than normal and urinating may be painful or uncomfortable.

### **causes**

Bacteria that enter the urinary tract through the urethra can multiply and travel to your kidneys. This is the most common cause of kidney infections.

Bacteria from an infection in another part of the body also can spread through the bloodstream to the kidneys. In rare cases, an artificial joint or heart valve that becomes infected can cause a kidney infection.

Rarely, a kidney infection happens after kidney surgery.

### **How do you get a kidney infection?**

Your kidneys make pee (urine) to get rid of waste. The pee moves through tubes (ureters) to your bladder (a pouch that holds your pee until you go to the bathroom). From there, it moves through another tube (urethra) to leave your body. This usually cleans out any bacteria or other germs with it.

Sometimes, bacteria can move upwards into your body and infect parts of your urinary tract, including your urethra, bladder (cystitis) or ureters. From there, they can move into one or both of your kidneys, causing a kidney infection. Bacteria that get into your blood from another part of your body can also infect your kidneys.

#### **Risk factors for kidney infections include:**

* Blockage. Anything that keeps you from emptying the pee out of your urinary tract can allow bacteria to grow and back up into your kidneys. This includes kidney stones, enlarged prostate and uterine prolapse. Pressure on your bladder during pregnancy can also increase your risk.
* Vesicoureteral reflux. This is a condition where pee goes the wrong way and backs up from your bladder.
* Conditions that put you at increased risk for infections. These include diabetes, HIV and being on immunosuppressive medications.
* Your anatomy. Women have a shorter urethra, which makes it easier for bacteria to move up to their bladder and kidneys.

## **Diagnosing pyelonephritis**

If your doctor is concerned that you have a kidney infection, he or she will ask you about other medical problems, any past infections and your recent symptoms. He or she will check your vital signs (temperature, heart rate, blood pressure), and will press on your abdomen and flanks to see if there is tenderness near the kidney. In women, the symptoms of pyelonephritis may be similar to those of certain sexually transmitted diseases, so your doctor may recommend that you have a pelvic examination.

To diagnose pyelonephritis, your doctor will order urine tests to look for white cells in the urine and for culture to determine the type of bacteria causing the infection. Usually your doctor will also order blood tests to determine your white blood cell count and to make sure your kidney function is normal. Your doctor may also order a blood culture because some people with pyelonephritis have bacteria in their blood as well as their urine. Antibiotics are started prior to the culture results and will be adjusted once the bacterial species is identified in 24 to 48 hours.

## **Expected duration of pyelonephritis**

Most patients with uncomplicated cases of pyelonephritis find that their symptoms begin to improve after one to two days of treatment with antibiotics. However, even after symptoms improve, antibiotics are usually prescribed to complete a 7 to 10 day course.

## **Preventing pyelonephritis**

To help prevent pyelonephritis if you have had a previous episode or are at risk:

* Drink several glasses of water each day. Water discourages the growth of infection-causing bacteria by flushing out your urinary tract. This flushing also helps to prevent kidney stones, which can increase the risk of pyelonephritis.
* If you are a woman, wipe from front to back. To prevent the spread of intestinal and skin bacteria from the rectum to the urinary tract, women should always wipe toilet tissue from front to the back after having a bowel movement or urinating.
* Decrease the spread of bacteria during sex. Women should urinate after sexual intercourse to flush bacteria from the bladder. Some women who have frequent urinary tract infections after sexual activity can take antibiotics around the time of intercourse to prevent an infection.

If there is a structural problem with the urinary system, such as blockage from a stone, or a developmental abnormality, surgery can be done to restore normal urinary function and prevent future episodes of pyelonephritis.

## **Treating pyelonephritis**

Doctors treat pyelonephritis with antibiotics. In most uncomplicated cases of pyelonephritis, the antibiotic can be given orally (by mouth), and treatment usually lasts for 7 to 10 days. Commonly used oral antibiotics include trimethoprim with sulfamethoxazole (Bactrim and others), ciprofloxacin (Cipro) or levofloxacin (Levaquin), but the choice of antibiotic will depend on your history of allergies and laboratory testing of the bacteria causing the infection.

If you have high fever, shaking chills or severe nausea and vomiting, you are more likely to become dehydrated and may be unable to take oral antibiotics. In that case, you may require hospital treatment so that antibiotics can be given intravenously (into a vein). High fever and shaking chills also may be signs that your kidney infection has spread to your bloodstream and can travel to other parts of your body. If your doctor is concerned that you may have an obstruction (such as a kidney stone that is stuck in the ureter) or a structural abnormality in your urinary system, other tests may be ordered, such as a computed tomography (CT) scan or ultrasound.

## **When to call a professional**

Call your doctor immediately if you have symptoms of pyelonephritis (particularly fever and flank pain, with or without urinary symptoms), especially if you are pregnant.

## **Prognosis**

A single episode of uncomplicated pyelonephritis rarely causes permanent kidney damage in an otherwise healthy adult. However, repeated episodes of pyelonephritis can cause chronic (long-lasting) kidney disease in children, people with diabetes, and adults who have structural abnormalities of the urinary tract, or nerve diseases that disrupt bladder function. Pyelonephritis can become chronic if an infection cannot be cleared easily, as in a person with a kidney stone or a developmental abnormality of the urinary system.

### **complications of a kidney infection**

Sometimes, kidney infections can lead to life-threatening complications, especially in people with a weakened immune system or other underlying health issues. These include:

* Emphysematous pyelonephritis. This is a condition where bacteria start destroying parts of your kidneys and create gas. It’s most common in people with diabetes.
* Renal papillary necrosis. This is a condition that damages your kidneys.

**EPIDEMIOLOGY**

Acute pyelonephritis in the United States is found at a rate of 15 to 17 cases per 10,000 females and 3 to 4 cases per 10,000 males annually, with an annual total of 250,000 cases annually reported in the US. One large study of over 750,000 patients in Sweden found that uncomplicated UTI/cystitis developed into pyelonephritis 0.47% of the time with antibiotic treatment. This risk rose to 1.43% if an antibiotic prescription was not filled within 5 days of cystitis diagnosis.

Young, sexually active women are most often affected by acute pyelonephritis due to their higher incidence of UTIs, but men have a higher mortality rate. Men are more likely to have diabetes, nephrolithiasis, or kidney disease than women. Groups with extremes of age, such as older adults and infants, are also at higher risk. Acute pyelonephritis has no racial predisposition.

Pregnant women are also considered a high-risk group due to physiologic changes predisposing them to an increased risk of UTI. Acute pyelonephritis leads to maternal complications and, in some studies, also preterm delivery and low birth weight. Asymptomatic bacteriuria occurs in 2% to 7% of pregnant women. While clinical guidelines in North America and Europe have recommended screening for and treating asymptomatic bacteriuria in pregnant patients to avoid pyelonephritis, these guidelines are based on studies now considered low-quality from the 1960s and 1980s. More recent data found no significant difference in cases of pyelonephritis with treatment of asymptomatic bacteriuria, and overall events of pyelonephritis were low (0 vs 1 event in the treated and untreated groups, respectively). Therefore, more recent data supports not treating asymptomatic bacteriuria in pregnant women, especially given that antibiotics can have potential adverse effects. More high-quality randomized controlled trials are needed in this area.

**DIFFERENTIAL DIAGNOSIS**

When diagnosing acute pyelonephritis, it is initially wise to keep the differential broad. Physicians should consider other disorders as well when patients present with fever, flank pain, and costovertebral angle tenderness. Because symptoms can be variable (unilateral, bilateral, radiating, sharp, dull) and because pyelonephritis can progress to sepsis and shock, the differential diagnoses associated with pyelonephritis can be extensive.

Common mimics of acute pyelonephritis can include but are not limited to the following:

* Appendicitis
* Cholecystitis
* Costochondritis
* Diverticulitis
* Ectopic pregnancy
* Endometritis
* Focal nephronia
* Herpes zoster
* Lobar pneumonia
* Nephrolithiasis
* Ovarian cyst pathology
* Pancreatitis
* Pelvic inflammatory disease
* Perinephric abscess
* Pyonephrosis (obstructive pyelonephritis)
* Renal abscess
* Rib fracture
* Ureterolithiasis
* Ureteropelvic junction obstruction
* Urolithiasis
* Xanthogranulomatous pyelonephritis

**Appendicitis**: Inflammation of the appendix, usually due to blockage, causing abdominal pain typically starting near the belly button and moving to the lower right abdomen; requires urgent surgery to prevent rupture.

**Cholecystitis**: Inflammation of the gallbladder, often caused by gallstones blocking the cystic duct, leading to right upper abdominal pain, fever, and nausea.

**Costochondritis**: Inflammation of the cartilage connecting ribs to the breastbone, causing localized chest pain worsened by movement or pressure.

**Diverticulitis**: Inflammation or infection of diverticula (small pouches) in the colon, causing lower abdominal pain, fever, and digestive changes.

**Ectopic Pregnancy**: Implantation of a fertilized egg outside the uterus, most commonly in a fallopian tube, causing abdominal pain and vaginal bleeding; a medical emergency.

**Endometritis**: Inflammation of the uterine lining, often due to infection after childbirth or miscarriage, presenting with fever, pelvic pain, and abnormal discharge.

**Focal Nephronia:** Localized bacterial infection of the kidney tissue causing a mass-like lesion, fever, and flank pain; considered a severe form of pyelonephritis.

**Herpes Zoster**: Reactivation of varicella-zoster virus causing painful, blistering rash in a dermatomal distribution.

**Lobar Pneumonia**: Infection of a large, continuous area of a lung lobe, causing fever, cough, chest pain, and consolidation on imaging.

**Nephrolithiasis**: Formation of kidney stones causing severe flank pain, hematuria, and possible urinary obstruction.

**Ovarian Cyst Pathology**: Fluid-filled sacs on the ovary that may cause pelvic pain, bloating, or be asymptomatic; complications include rupture or torsion.

**Pancreatitis**: Inflammation of the pancreas, often due to gallstones or alcohol, presenting with severe upper abdominal pain radiating to the back, nausea, and vomiting.

**Pelvic Inflammatory Disease (PID)**: Infection of female reproductive organs, usually from sexually transmitted infections, causing pelvic pain, fever, and abnormal vaginal discharge.

**Perinephric Abscess**: Collection of pus around the kidney due to infection, causing flank pain, fever, and systemic illness.

**Pyonephrosis (Obstructive Pyelonephritis**): Pus accumulation in an obstructed kidney collecting system, leading to severe infection and flank pain.

**Renal Abscess**: Localized pus collection within the kidney due to bacterial infection, causing fever, flank pain, and systemic symptoms.

**Rib Fracture**: Break in one or more ribs usually from trauma, causing localized chest pain worsened by breathing or movement.

**Ureterolithiasis**: Presence of stones in the ureter causing severe colicky flank pain, hematuria, and possible urinary obstruction.

**Ureteropelvic Junction Obstructio**n: Congenital or acquired blockage at the junction of the ureter and renal pelvis, causing hydronephrosis and flank pain.

**Urolithiasi**s: Formation of stones anywhere in the urinary tract, causing pain, hematuria, and urinary symptoms.

**Xanthogranulomatous Pyelonephritis**: Chronic kidney infection with destruction of renal tissue and granuloma formation, often associated with obstruction and stones, causing flank pain and systemic illness.

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**Hyperoxaluria**

**DEFINITION AND DESCRIPTION**

Hyperoxaluria (hi-pur-ok-suh-LU-ree-uh) happens when you have too much oxalate in your urine. Oxalate is a natural chemical the body makes. It's also found in some foods. But too much oxalate in the urine can cause serious problems.

Hyperoxaluria can be caused by a change in a gene, an intestine disease or eating too many foods that are high in oxalate. The long-term health of your kidneys depends on finding hyperoxaluria early and getting it treated quickly.

Oxalosis (ok-suh-LOW-sis) happens after the kidneys stop working well in people who have primary and intestine-related causes of hyperoxaluria. Too much oxalate collects in the blood. This can lead to oxalate buildups in blood vessels, bones and organs.

### **What age does hyperoxaluria start?**

Symptoms of hyperoxaluria can develop anytime from infancy to adulthood. The average age that symptoms appear is 5 years old.

People with primary hyperoxaluria tend to get recurring kidney stones as a child or teenager (usually before age 20). If infants develop primary hyperoxaluria, the condition tends to be severe. About 50% of these children will experience kidney failure by age 15. About 80% will experience kidney failure by age 30.

People with enteric and dietary hyperoxaluria tend to develop kidney stones and other symptoms in adulthood.

#### **How common is hyperoxaluria?**

Hyperoxaluria is rare, but healthcare providers can’t be entirely sure of how rare. Less than 1,000 people have primary hyperoxaluria in the United States. Enteric and dietary hyperoxaluria are less common.

**CAUSES**

Hyperoxaluria happens when too much of a chemical called oxalate builds up in the urine. There are different types of hyperoxaluria:

* **Primary hyperoxaluria.** This type is a rare inherited disease, which means that it's passed down in families. It's caused by changes in a gene. With primary hyperoxaluria, the liver doesn't make enough of a certain protein that prevents too much oxalate from being made. Or the protein doesn't work as it should. The body gets rid of excess oxalate through the kidneys, in urine. The extra oxalate can combine with calcium to form kidney stones and crystals. These can damage the kidneys and cause them to stop working.  
  With primary hyperoxaluria, kidney stones form early. They most often cause symptoms from childhood through age 20. The kidneys of many people with primary hyperoxaluria stop working well by early to middle adulthood. But kidney failure can happen even in babies with this disease. Others with primary hyperoxaluria may never have kidney failure.
* **Enteric hyperoxaluria.** Some intestine problems cause the body to absorb more oxalate from foods. This can then increase the amount of oxalate in the urine. Crohn's disease is one intestine problem that can lead to enteric hyperoxaluria. Another is short bowel syndrome, which can happen when parts of the small intestine are removed during surgery.  
  Other health problems make it hard for the small intestine to absorb fats from food. If this happens, it might leave oxalate more available for the gut to absorb. Usually, oxalate combines with calcium in the gut and exits the body through stools. But when there is increased fat in the gut, calcium binds to the fat instead. This allows oxalate to be free in the gut and absorbed in the bloodstream. It's then filtered by the kidneys. Roux-en-Y gastric bypass surgery also can lead to trouble absorbing fat in the gut, which raises the risk of hyperoxaluria.
* **Hyperoxaluria tied to eating foods with lots of oxalate.** Eating large amounts of foods high in oxalate can raise your risk of hyperoxaluria or kidney stones. These foods include nuts, chocolate, brewed tea, spinach, potatoes, beets and rhubarb. It's important to stay away from high-oxalate foods if you have diet-related or enteric hyperoxaluria. Your doctor also may tell you to limit these foods if you have primary hyperoxaluria.

#### **Risk factors for hyperoxaluria**

Having a biological parent with hyperoxaluria is the biggest risk factor for developing the condition yourself. Even people who don’t have symptoms can pass primary hyperoxaluria on to their children. Your healthcare provider may offer you genetic testing if your biological siblings have primary hyperoxaluria.

You may also be at greater risk if you:

* Have recurring kidney stones.
* Had kidney stones as a child (even one time).
* Have a family history of kidney stones.
* Have calcium deposits on your kidneys

**SYMPTOMS**

Often, the first sign of hyperoxaluria is a kidney stone. Kidney stone symptoms can include:

* Sharp pain in the back, side, lower stomach area or groin.
* Urine that looks pink, red or brown due to blood.
* Frequent urge to pee, also called urination.
* Pain when peeing.
* Not being able to urinate or peeing only a small amount.
* Chills, fever, upset stomach or vomiting.

DIAGNOSIS

You'll likely have a thorough physical exam. You might be asked questions about your health history and eating habits.

Tests to diagnose hyperoxaluria may include:

* **Urine tests,** to measure oxalate and other substances in the urine. You're given a special container to collect your urine over 24 hours. It's then sent to a lab.
* **Blood tests,** to check how well your kidneys work and measure oxalate levels in the blood.
* **Stone analysis,** to find out what kidney stones are made of after you've passed them through urine or gotten them removed with surgery.
* **Kidney X-ray, ultrasound or computerized tomography (CT) scan,** to check for any kidney stones or calcium oxalate buildup in the body.

You may need more tests to find out for sure if you have hyperoxaluria and see how the disease has affected other parts of your body. These tests may include:

* **DNA testing** to look for the gene changes that cause primary hyperoxaluria.
* **Kidney biopsy** to check for buildup of oxalate.
* **Echocardiogram,** an imaging test that can check for oxalate buildup in the heart.
* **Eye exam** to check for oxalate deposits in the eyes.
* **Bone marrow biopsy** to check for buildup of oxalate in the bones.
* **Liver biopsy** to look for low levels of proteins, also called enzyme deficiencies. This test is needed only in rare cases when genetic testing doesn't show the cause of hyperoxaluria.

If you learn you have primary hyperoxaluria, your siblings also are at risk of the disease. They should have tests as well. If your child has primary hyperoxaluria, you may want to get genetic testing if you and your partner plan to have more children. Medical genetics counselors who have experience with hyperoxaluria can help guide your decisions and testing.

**TREATMENT**

Treatment depends on the type of hyperoxaluria you have, the symptoms and how serious the disease is. How well you respond to treatment also helps your health care team decide how else to manage your condition.

### **Reducing oxalate**

To lower the amount of calcium oxalate crystals that form in your kidneys, your doctor may suggest one or more of these treatments:

* **Medicine.** Lumasiran (Oxlumo) is a medicine that lowers the level of oxalate in children and adults with primary hyperoxaluria. Prescription doses of vitamin B-6, also called pyridoxine, can help reduce oxalate in the urine in some people with primary hyperoxaluria. Phosphates and citrate prepared by a pharmacy and taken by mouth help keep calcium oxalate crystals from forming.  
  Your doctor also may give you other medicines, such as thiazide diuretics. It depends on which other unusual signs are found in your urine. If you have enteric hyperoxaluria, your doctor also may recommend a calcium supplement to take with meals. This could make it easier for oxalate to combine with calcium in the gut and leave the body through stool.
* **Drinking lots of fluids.** If your kidneys still work well, your doctor will likely tell you to drink more water or other fluids. This flushes the kidneys, prevents oxalate crystal buildup and helps keep kidney stones from forming.
* **Diet changes.** In general, it's more important to pay attention to your food choices if you have enteric or diet-related hyperoxaluria. Diet changes may help lower the levels of oxalate in your urine. Your health care team may suggest that you restrict foods high in oxalates, limit salt and eat less animal protein and sugar. But diet changes may not help all people with primary hyperoxaluria. Follow your care team's suggestions.

**COMPLICATION**

Without treatment, primary hyperoxaluria can damage the kidneys. Over time the kidneys may stop working. This is called kidney failure. For some people, this is the first sign of the disease.

Symptoms of kidney failure include:

* Peeing less than usual or not peeing at all.
* Feeling ill and tired.
* Not feeling hungry.
* Upset stomach and vomiting.
* Pale, ashen skin or other changes to skin color tied to having a low number of red blood cells, also called anemia.
* Swelling of hands and feet.

Oxalosis happens if you have primary or enteric hyperoxaluria and your kidneys stop working well enough. The body can no longer get rid of the extra oxalate, so the oxalate starts building up. First it builds up in the blood, then in the eyes, bones, skin, muscles, blood vessels, heart and other organs.

Oxalosis can cause many health problems outside the kidneys in its late stages. These include:

* Bone disease.
* Anemia.
* Skin ulcers.
* Heart and eye problems.
* In children, serious problems developing and growing.

**WHEN TO SEE A DOCTOR**

It's not common for children to get kidney stones. Kidney stones that form in children and teenagers are likely to be caused by a health problem, such as hyperoxaluria.

All young people with kidney stones should have a checkup. The checkup should include a test that measures oxalate in the urine. Adults who keep getting kidney stones also should be tested for oxalate in the urine.

**EPIDEMIOLOGY**

The lifetime risk of nephrolithiasis in the developed world is about 10% to 15%. Men have an increased incidence compared to women. In the United States, the prevalence is about 10% in men and 7% in women, with a 10-year recurrence rate of 50%. Whites have higher rates of nephrolithiasis and hyperoxaluria than Blacks.

Calcium stones comprise about 80% of all kidney stone diseases, with calcium oxalate predominating at approximately 75%. The risk of recurrence with calcium stones is about 60% in 10 years without appropriate preventive measures.

The estimated overall incidence of secondary hyperoxaluria appears to be increasing over time. Typical rates of hyperoxaluria range from 25% to 45% in all recurrent calcium stone-formers. They are also higher in non-American populations; Asian countries typically have higher rates of hyperoxaluria than Western countries. The reasons for this are unclear but likely due to cultural, genetic, and dietary issues. This significantly increased incidence of hyperoxaluria is also considered a significant contributing factor to the observed rise in global nephrolithiasis rates. Further studies are needed to confirm this finding.

There does appear to be a protective effect from female sex hormones on oxalate excretion, while testosterone has a detrimental effect. The exact reason for this is unclear and will require more studies to elucidate.

Greater body weight appears to increase urinary oxalate, but there are conflicting data on whether this is relatively proportionate between the sexes. However, obesity is associated with higher urinary oxalate levels in both men and women. Among stone formers, patients with obesity are found to have oxalate levels about 33% higher than stone-forming patients who are not obese.

Urinary oxalate excretion is higher in Whites after a controlled, high-oxalate meal than in Blacks. The reason for this is unclear but is thought to be due to genetic factors, with Whites having a higher rate of intestinal oxalate absorption than Blacks.There does not appear to be an age-related factor concerning oxalate absorption.

PH is quite rare. While usually diagnosed in the pediatric age group, it is often diagnosed very late into the course of the disease, usually only after the development of nephrocalcinosis or ESRD. PH type I is the most common and severe form of this rare disorder, accounting for 80% of those diagnosed with the condition.

The prevalence of the disease ranges from 1 to 3 per 1 million population in the US, with an approximate incidence rate of approximately 1 in 100,000 live births per year in Europe and 1 in 58,000 population worldwide. Higher rates are reported from inbred populations and in developing countries. PH accounts for less than 1% of the pediatric ESRD population in USA, UK, and Japan registries.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis should include conditions that lead to nephrolithiasis, specifically calcium oxalate stones and excess deposition in tissues leading to nephrocalcinosis. Some of these conditions are hypocitraturia, medullary sponge kidney, nephrocalcinosis of prematurity, and renal tubular acidosis.

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**Amyloidosis**

Amyloidosis (am-uh-loi-DO-sis) is a rare disease that occurs when a protein called amyloid builds up in organs. This amyloid buildup can make the organs not work properly.

Organs that may be affected include the heart, kidneys, liver, spleen, nervous system and digestive tract.

Some types of amyloidosis occur with other diseases. These types may improve with treatment of the other diseases. Some types of amyloidosis may lead to life-threatening organ failure.

Treatments may include chemotherapy with strong drugs used to treat cancer. Other types of medications can reduce amyloid production and control symptoms. Some people may benefit from organ or stem cell transplants.

#### **How common is amyloidosis?**

Amyloidosis is a rare disease. Healthcare providers estimate that in the U.S., there are only 1,275 to 3,200 new cases of AL amyloidosis diagnosed each year.

**CAUSES**

There are many different types of amyloidosis. Some types are hereditary. Others are caused by outside factors, such as inflammatory diseases or long-term dialysis. Many types affect multiple organs. Others affect only one part of the body.

Types of amyloidosis include:

* **AL amyloidosis (immunoglobulin light chain amyloidosis).** This is the most common type of amyloidosis in developed countries. AL amyloidosis is also called primary amyloidosis. It usually affects the heart, kidneys, liver and nerves.
* **AA amyloidosis.** This type is also known as secondary amyloidosis. It's usually triggered by an inflammatory disease, such as rheumatoid arthritis. It most commonly affects the kidneys, liver and spleen.
* **Hereditary amyloidosis (familial amyloidosis).** This inherited disorder often affects the nerves, heart and kidneys. It most commonly happens when a protein made by your liver is abnormal. This protein is called transthyretin (TTR).
* **Wild-type amyloidosis.** This variety has also been called senile systemic amyloidosis. It occurs when the TTR protein made by the liver is normal but produces amyloid for unknown reasons. Wild-type amyloidosis tends to affect men over age 70 and often targets the heart. It can also cause carpal tunnel syndrome.
* **Localized amyloidosis.** This type of amyloidosis often has a better prognosis than the varieties that affect multiple organ systems. Typical sites for localized amyloidosis include the bladder, skin, throat or lungs. Correct diagnosis is important so that treatments that affect the entire body can be avoided.

**RISK FACTOR**

Factors that increase the risk of amyloidosis include:

* **Age.** Most people diagnosed with amyloidosis are between ages 60 and 70.
* **Sex.** Amyloidosis occurs more commonly in men.
* **Other diseases.** Having a chronic infectious or inflammatory disease increases the risk of AA amyloidosis.
* **Family history.** Some types of amyloidosis are hereditary.
* **Kidney dialysis.** Dialysis can't always remove large proteins from the blood. If you're on dialysis, abnormal proteins can build up in your blood and eventually be deposited in tissue. This condition is less common with more modern dialysis techniques.
* **Race.** People of African descent appear to be at higher risk of carrying a genetic mutation associated with a type of amyloidosis that can harm the heart.

**SYMPTOMS**

You may not experience symptoms of amyloidosis until later in the course of the disease. Symptoms may vary, depending on which organs are affected.

Signs and symptoms of amyloidosis may include:

* Severe fatigue and weakness
* Shortness of breath
* Numbness, tingling, or pain in the hands or feet
* Swelling of the ankles and legs
* Diarrhea, possibly with blood, or constipation
* An enlarged tongue, which sometimes looks rippled around its edge
* Skin changes, such as thickening or easy bruising, and purplish patches around the eyes

**DIAGNOSIS**

Amyloidosis is often overlooked because the signs and symptoms can mimic those of more-common diseases.

Early diagnosis can help prevent further organ damage. Precise diagnosis is important because treatment varies greatly, depending on your specific condition.

### **Laboratory tests**

Blood and urine may be analyzed for abnormal protein that can indicate amyloidosis. People with certain symptoms may also need thyroid and kidney function tests.

### **Biopsy**

A tissue sample can be checked for signs of amyloidosis. The biopsy may be taken from the fat under the skin on the abdomen or from bone marrow. Some people may need a biopsy of an affected organ, such as the liver or kidney. The tissue can be tested to see what type of amyloid is involved.

### **Imaging tests**

Images of the organs affected by amyloidosis may include:

* **Echocardiogram.** This technology uses sound waves to create moving images that can show how well the heart is working. It can also show heart damage that can be specific to particular types of amyloidosis.
* **Magnetic resonance imaging (MRI).** MRI uses radio waves and a strong magnetic field to create detailed images of organs and tissues. These can be used to check the structure and function of the heart.
* **Nuclear imaging.** In this test, tiny amounts of radioactive material (tracers) are injected into a vein. This can reveal early heart damage caused by certain types of amyloidosis. It can also help distinguish between different types of amyloidosis, which can guide treatment decisions.

**TREATMENT**

There's no cure for amyloidosis. But treatment can help manage signs and symptoms and limit further production of amyloid protein. If the amyloidosis has been triggered by another condition, such as rheumatoid arthritis or tuberculosis, treating the underlying condition can be helpful.

### **Medications**

* **Chemotherapy.** Some cancer drugs are used in AL amyloidosis to stop the growth of abnormal cells that produce the protein that forms amyloid.
* **Heart medications.** If your heart is affected, you may need to take blood thinners to reduce the risk of clots. You may also need medications to control your heart rate. Drugs that increase urination can reduce the strain on your heart and kidneys.
* **Targeted therapies.** For certain types of amyloidosis, drugs such as patisiran (Onpattro) and inotersen (Tegsedi) can interfere with the commands sent by faulty genes that create amyloid. Other drugs, such as tafamidis (Vyndamax, Vyndaqel) and diflunisal, can stabilize bits of protein in the bloodstream and prevent them from getting transformed into amyloid deposits.

### **Surgical and other procedures**

* **Autologous blood stem cell transplant.** This procedure involves collecting your own stem cells from your blood through a vein and storing them for a short time while you have high-dose chemotherapy. The stem cells are then returned to your body via a vein. This treatment is most appropriate for people whose disease isn't advanced and whose heart isn't greatly affected.
* **Dialysis.** If your kidneys have been damaged by amyloidosis, you may need to start dialysis. This procedure uses a machine to filter wastes, salts and fluid from your blood on a regular schedule.
* **Organ transplant.** If amyloid deposits have severely damaged your heart or kidneys, you might need surgery to replace those organs. Some types of amyloid are formed in the liver, so a liver transplant could stop that production.

**COMPLICATION**

Amyloidosis can seriously damage the:

* **Heart.** Amyloid reduces the heart's ability to fill with blood between heartbeats. Less blood is pumped with each beat. This can cause shortness of breath. If amyloidosis affects the heart's electrical system, it can cause heart rhythm problems. Amyloid-related heart problems can become life-threatening.
* **Kidneys.** Amyloid can harm the kidneys' filtering system. This affects their ability to remove waste products from the body. It can eventually cause kidney failure.
* **Nervous system.** Nerve damage can cause pain, numbness, or tingling of the fingers and feet. If amyloid affects the nerves that control bowel function, it can cause periods of alternating constipation and diarrhea. Damage to the nerves that control blood pressure can make people feel faint if they stand up too quickly.

## **Outlook / Prognosis**

### **What can I expect if I have amyloidosis?**

Your healthcare provider can treat symptoms, slow the disease’s progress and, in some cases, they can help reverse amyloidosis. Still, some amyloidosis types may cause life-threatening organ damage without treatment. This is why early diagnosis and prompt treatment are so important.

## **Living With**

### **How do I take care of myself?**

There are several types of amyloidosis, which means there isn’t a one-size-fits-all approach to managing this condition. Ask your healthcare provider about steps that make sense for you.

This may include taking care of your physical health by eating nutritious foods and exercising regularly. Caring for yourself also involves prioritizing your mental health. Ask your provider to recommend support groups for people who have amyloidosis. Connecting with others can help you cope with the emotional challenges of living with a rare disease.

### **When should I see my healthcare provider?**

It can be tough to know when to see a provider, as the symptoms of amyloidosis are so varied. As a general rule, schedule an appointment if you’re experiencing unexplained symptoms that don’t improve. Your symptoms may or may not be related to amyloidosis. But if they are, the sooner you’re diagnosed, the better your outlook.

**EPIDEMIOLOGY**

AL amyloidosis has an incidence of 1 case per 100,000 person-years in Western countries. In the United States, there are approximately 1275 to 3200 new cases per year. The annual proportion of new cases with AL is 78%.

Familial transthyretin-associated amyloidosis (ATTR) is a less common systemic type of amyloidosis with unknown incidence, but approximately 10% to 20% of diagnosed cases in tertiary centers are secondary to ATTR amyloidosis. Of these cases, seven percent are hereditary and result from mutated transthyretin (ATTRm) and approximately six percent represent the acquired age-related, wild-type ATTRwt amyloidosis. ATTRwt is seen more commonly in males and was formerly known as senile systemic amyloidosis. Secondary or AA amyloidosis represents 6% of all the amyloidosis cases diagnosed each year. AA amyloidosis is an acquired process and reactive due to chronic inflammation.

**DIFFERENTIAL DIAGNOSIS**

Chronic inflammatory conditions and systemic conditions are associated with amyloidosis. Systemic or secondary amyloidosis often coexists with immunoglobulin M associated disorders.

The following are the differential diagnosis;

* Familial Renal Amyloidosis
* Immunoglobulin-Related Amyloidosis
* Membranous Glomerulonephritis
* Renal Vein Thrombosis due to amyloid
* Cutis Verticis Gyrata
* Mastocytosis
* Nodular Localized Cutaneous Amyloidosis
* Pseudoxanthoma Elasticum

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**lupus nephritis**

**DEFINITION**

Lupus nephritis is a problem that occurs often in people who have systemic lupus erythematosus, also called lupus.

Lupus is a disease in which the body's immune system attacks its own cells and organs, called autoimmune disease. Lupus causes the immune system to make proteins called autoantibodies. These proteins attack tissues and organs in the body, including the kidneys.

Lupus nephritis occurs when lupus autoantibodies affect parts of the kidneys that filter out waste. This causes swelling and irritation of the kidneys, called inflammation. It might lead to blood in the urine, protein in the urine, high blood pressure, kidneys that don't work well or even kidney failure.

### **Who gets lupus nephritis?**

Only adults and children with lupus can develop lupus nephritis. You’re more likely to get lupus if you:

* Are a woman (9 out of 10 people with lupus are women) between the ages of 15 and 44, though men are more likely to develop lupus nephritis.
* Are of Black, Native American, Hispanic/Latino, Pacific Islander or of Asian descent.
* Come in contact with certain infections, viruses, toxic chemicals or pollutants in the environment.
* Have a family history of the disease.
* Have another autoimmune disease.

### **How common is lupus nephritis?**

About 50% of adults with lupus will develop lupus nephritis. About 80% of children with lupus will develop this kidney condition

**CAUSES**

As many as half of adults with systemic lupus get lupus nephritis. Systemic lupus causes the body's immune system to damage the kidneys. Then the kidneys can't filter out waste as they should.

**RISK FACTOR**

The only known risk factors for lupus nephritis are:

* **Being male.** Women are more likely to get lupus, but men get lupus nephritis more than women do.
* **Race or ethnicity.** Black people, Hispanic people and Asian Americans are more likely to have lupus nephritis than are whites.

SYMPTOMS

Symptoms of lupus nephritis tend to develop about five years after lupus symptoms first appear. But lupus nephritis can be the first — and sometimes the only — manifestation of systemic lupus erythematosus (SLE). Lupus nephritis can cause:

* Edema (swelling due to fluid buildup) in your lower body or around your eyes.
* Fever with no known cause.
* Hematuria (blood in the urine).
* High blood pressure.
* Increased urination, especially at night.
* Joint pain or swelling.
* Muscle pain.
* Proteinuria (protein in the urine), which often causes your urine to look foamy.
* Red skin rash on the face.
* Weight gain due to excess fluid in your body.

DIAGNOSIS

Tests to diagnose lupus nephritis include:

* **Blood and urine tests.** Besides the usual blood and urine tests, urine collected over 24 hours might be tested. These tests measure how well the kidneys are working.
* **Kidney biopsy.** A small section of kidney tissue is removed and sent to a lab. This test diagnoses lupus nephritis. It also can help show how bad the disease is. There can be more than one biopsy over time.

**TREATMENT**

There's no cure for lupus nephritis. Treatment aims to:

* Reduce symptoms or make symptoms go away, called remission.
* Keep the disease from getting worse.
* Keep symptoms from coming back.
* Keep kidneys working well enough to not need a machine to filter waste from blood, called dialysis, or a kidney transplant.

### **Supportive treatments**

In general, these treatments might help people with kidney disease:

* **Diet changes.** Limiting the amount of protein and salt in the diet can help the kidneys work better.
* **Blood pressure medicines.** Medicines called angiotensin-converting enzyme (ACE) inhibitors and angiotensin II receptor blockers (ARBs) can help control blood pressure. These medicines also keep protein from leaking from the kidneys into the urine. Medicines called diuretics help get rid of fluid.

### **Medicines**

Treating severe lupus nephritis might need medicines that slow or stop the immune system from attacking healthy cells. Medicines often are used together. Sometimes certain medicines used at first are switched to prevent toxic effects.

Medicines to treat lupus nephritis might include:

* Steroids, such as prednisone (Rayos).
* Cyclosporine (Gengraf, Neoral, Sandimmune).
* Voclosporin (Lupkynis).
* Tacrolimus (Astagraf, Envarsus, Prograf).
* Cyclophosphamide (Cytoxan).
* Azathioprine (Azasan, Imuran).
* Mycophenolate (CellCept).
* Rituximab (Rituxan).
* Belimumab (Benlysta).

Ongoing clinical trials are testing new treatments for lupus nephritis.

### **Treatment options for kidney failure**

For people who progress to kidney failure, treatment options include:

* **Dialysis.** Dialysis helps remove fluid and waste from the body, maintain the right balance of minerals in the blood, and manage blood pressure by filtering the blood through a machine.
* **Kidney transplant.** If kidneys stop working, a kidney from a donor, called a transplant, might be needed.

**COMPLICATION**

Lupus nephritis can cause:

* Hypertension.
* Kidney failure.
* A higher risk of getting cancer, especially one that starts in the cells of the immune system, called B-cell lymphoma.
* A higher risk of heart and blood vessel problems.

## **Prevention**

### **How can I prevent lupus nephritis?**

If you have lupus, there’s no clear way to prevent lupus nephritis. Some medications (i.e., hydroxychloroquine) might prevent it, so it’s important to follow-up with your hematologist and be treated for lupus if needed.

## **Outlook / Prognosis**

### **What’s the prognosis (outlook) for people with lupus nephritis?**

People who receive timely treatment for lupus nephritis have a positive outlook. People with lupus nephritis who receive medication, dialysis or a kidney transplant tend to do as well as people with other kidney diseases who receive these treatments. But most people need to manage the disease with medication or dialysis for the rest of their lives.

### **What are the long-term complications of lupus nephritis?**

In addition to kidney failure, other long-term complications of lupus nephritis include:

* Higher risk of certain cancers, including B-cell lymphoma.
* Heart and blood vessel problems.

## **Living With**

### **When should I call my doctor about lupus nephritis?**

Contact your healthcare provider right away if you experience any of the following symptoms, as they could be signs of sudden kidney failure:

* Abdominal pain.
* Difficulty urinating, or not producing much urine.
* Fatigue or drowsiness.
* Foamy or bloody urine.
* Itchy skin.
* Increased blood pressure.
* Loss of appetite.
* Nausea and vomiting.
* Shortness of breath.
* Swelling.

## **Differential Diagnoses**

* Chronic Glomerulonephritis
* Diffuse Proliferative Glomerulonephritis
* Granulomatosis with Polyangiitis (GPA, formerly Wegener Granulomatosis)
* Membranous Glomerulonephritis
* Polyarteritis Nodosa
* Rapidly Progressive Glomerulonephritis

## **Epidemiology**

### Frequency

In a multi-ethnic international cohort of patients enrolled within 15 months (mean, 6 months) after SLE diagnosis and assessed annually, lupus nephritis occurred in 700 of 1827 patients (38.3%). Lupus nephritis was frequently the initial presentation of SLE; it was identified at enrollment in 80.9% of cases. Patients with nephritis were younger, more frequently men and of African, Asian, and Hispanic race/ethnicity.

In a study of a large Spanish registry, lupus nephritis was histologically confirmed in 1092 of 3575 patients with SLE (30.5%). The mean age at lupus nephritis diagnosis was 28.4 years. The risk for lupus nephritis development was significantly higher in men, in younger individuals, and in Hispanics. Patients receiving antimalarials had a significantly lower risk of developing lupus nephritis.

### Age-related demographics

Most patients with SLE develop lupus nephritis early in their disease course. SLE is more common among women in the third decade of life, and lupus nephritis typically occurs in patients aged 20-40 years.Children with SLE are at a higher risk of renal disease than adults and tend to sustain more disease damage secondary to more aggressive disease and treatment-associated toxicity.

### Sex-related demographics

Because the overall prevalence of SLE is higher in females (ie, female-to-male ratio of 9:1), lupus nephritis is also more common in females; however, clinical renal disease has a worse prognosis and is more common in males with SLE.

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## **minimal change disease**

**DEFINITION AND DESCRIPTION**

Many diseases can affect your kidney function by attacking and damaging the glomeruli, the tiny filtering units inside your kidney where blood is cleaned. The conditions that affect your glomeruli are called glomerular diseases. One of these conditions is *minimal change disease (*MCD). Minimal change disease is a disorder where there is damage to your glomeruli. The disease gets its name because the damage cannot be seen under a regular microscope. It can only be seen under a very powerful microscope called an *electron microscope.* Minimal change disease is the most common cause of [nephrotic syndrome](https://www.nlm.nih.gov/medlineplus/ency/article/000490.htm) in children. It is also seen in adults with nephrotic syndrome, but is less common. Those with MCD experience the signs and symptoms of nephrotic syndrome much quicker than they would with other glomerular diseases.

## **causes**

## In adults, the disease is usually secondary (it is caused by another disease or drug). In children, MCD is usually primary (or idiopathic, which means the exact cause is not known). If you have secondary causes for MCD, the disease may occur or be related to:

* Allergic reactions
* Use of certain painkillers called non-steroidal anti-inflammatory drugs (NSAIDs)
* Tumors
* Infections caused by a virus

## **signs and symptoms**

You may notice the following signs and symptoms of MCD:

Foamy urine due to large amounts of protein leaking into your urine, called [proteinuria](https://www.kidney.org/atoz/content/proteinuriawyska)

* Swelling in body parts, like your ankles and around your eyes, due to fluid building up in your body, called edema
* Weight gain due to the fluid your body is not able to get rid of
* [Nephrotic Syndrome**:**](https://www.kidney.org/atoz/content/nephrotic) A set of symptoms that happen together and affect your kidneys. These include:
  + Swelling in body parts like your legs, ankles, or around your eyes (edema)
  + Large amounts of protein in your urine ([proteinuria](https://www.kidney.org/atoz/content/proteinuriawyska))
  + Loss of protein in your blood
  + High levels of fat or lipids in your blood (high cholesterol)

Always speak with your doctor if you experience any of these signs and symptoms.

## **How is Minimal Change Disease Diagnosed?**

The first clues are the signs and symptoms. Your healthcare provider may run tests to help understand the cause of your symptoms and find the proper treatment for you.

These tests are:

* **Urine test:** A urine test will help find protein and blood in your urine.
* **Blood test:** A blood test will help find levels of protein, cholesterol, and wastes in your blood.
* **Glomerular filtration rate (GFR):** A blood test will be done to know how well your kidneys are filtering the wastes from your body.
* **Kidney biopsy:** In this test, a tiny piece of your kidney is removed with a special needle, and looked at under a microscope.

If a kidney biopsy shows little or no damage under a regular microscope, then a diagnosis of MCD may be made if other symptoms, such as protein in the urine and swelling, are noticed. Because MCD is the most common cause of nephrotic syndrome in children, they first get treated for MCD before getting a biopsy. Most people will have a response in fewer than 8 weeks. If the protein in the urine disappears, the doctors may call the disease steroid-sensitive nephrotic syndrome instead of MCD. If treatment does not improve their symptoms over the course of several months a biopsy is done to see if there is another cause for their symptoms.

## **How is minimal change disease treated in children?**

MCD is usually easier to treat than other glomerular diseases. The treatment plan for nephrotic syndrome in children with MCD is usually with a type of drug called a corticosteroid, often called steroids. It is very important to not stop treatment suddenly. By sticking to the full treatment plan, your child will be less likely to relapse (experience the signs and symptoms again).

For children who do not respond to traditional treatment they have what is called steroid-resistant nephrotic syndrome or SRNS. Treatment for SRNS includes other combinations of drugs. It is recommended that children with SRNS take a blood pressure medication (ACE inhibitor or ARB). These two drugs control high blood pressure and reduce the amount of protein in the urine.

## **How is minimal change disease treated in adults?**

The treatment for nephrotic syndrome in adults with MCD is usually with a type of drug called a corticosteroid, often called steroids. You may notice that you start getting better within weeks, or less, although it may take an adult longer to respond than a child. It is important to stick with your treatment plan until all medications are finished; even if your symptoms go away sooner

If you are a woman and want to have children, you should speak with your healthcare provider to see how the medicines you are given affect this process.

For symptoms of swelling (edema), your healthcare provider may give you:

* ACE inhibitor or ARB medicines
* Diuretics (water pills)
* Limit sodium (salt) in your diet

## **Will minimal change disease cause kidney failure?**

Kidney failure is rare if you have minimal change disease. Almost all children and adults recover from MCD and avoid relapses over the long term. However, some may experience relapses of the protein in the urine, which can often be treated in the same way as the first episode.

**EPIDEMIOLOGY**

Minimal change disease has an incidence of 2 to 7 new cases per 100,000 children. The exact prevalence is unknown; however, it is estimated to be about 10 to 50 cases per 100,000 children. A male predominance (2 to 1) is noted during childhood, which disappears during the adolescent years. Minimal change disease is not common in adults, and the exact incidence is not known

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis includes:

* Cardiac: Heart failure
* Hepatic: Liver failure, hepatocellular cirrhosis
* Digestive: Protein-losing enteropathy, malnutrition
* Renal: acute Glomerulonephritis, renal failure
* Immune: Angioedema, anaphylaxis
* Lymphatic: Primary/secondary lymphedema, congenital lymphedema

These diagnoses present with edema secondary to decreased intravascular oncotic pressure.

**PROGNOSIS**

Minimal change disease has very good prognosis for all ages if there is a response to corticosteroid therapy. The primary morbidity is related to the adverse effects of the medications.

**COMPLICATION**

These complications are mainly secondary to the extensive proteinuria that is seen with this disease.

Children and adults with MCD are at risk for developing complications such as:

* **Hypovolemia**
* **Infections (pneumonia, peritonitis, sepsis):** Secondary to the loss of immunoglobulins (IgG) and complement factors via the urine. Also, there is decreased opsonization of capsulated organisms.
* **Venous thromboembolism:** due to a decrease in antithrombin III, protein S, plasminogen, and increase in prothrombotic factors.
* **Hyperlipidemia**
* **Acute kidney injury:** From the intravascular volume depletion.

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# **Interstitial nephritis**

Interstitial nephritis is a kidney disorder in which the spaces between the kidney tubules become swollen (inflamed). This can cause problems with the way your kidneys work.

## **Causes**

Interstitial nephritis may be temporary ([acute](https://medlineplus.gov/ency/article/002215.htm)), or it may be long-lasting ([chronic](https://medlineplus.gov/ency/article/002312.htm)) and get worse over time.

The acute form of interstitial nephritis is most often a side effect of certain drugs.

The following can cause interstitial nephritis:

* Allergic reaction to a drug (acute interstitial allergic nephritis).
* Autoimmune disorders, such as anti-tubular basement membrane disease or Kawasaki disease.
* Infections.
* Long-term use of medicines such as acetaminophen (Tylenol), aspirin, and nonsteroidal anti-inflammatory drugs (NSAIDs). This is called analgesic nephropathy.
* Side effects of certain antibiotics such as penicillin, ampicillin, methicillin, and sulfonamide medicines.
* Side effects of other medicines such as furosemide, thiazide diuretics, omeprazole, triamterene, and allopurinol.
* Too little potassium in your blood.
* Too much calcium or uric acid in your blood.

## **Symptoms**

Interstitial nephritis can cause mild to severe kidney problems, including acute kidney failure. In about half of cases, people will have decreased urine output and other signs of acute kidney failure.

Symptoms of this condition may include:

* Blood in the urine
* Fever
* Increased or decreased urine output
* Mental status changes (drowsiness, confusion, coma)
* Nausea, [vomiting](https://medlineplus.gov/ency/article/003117.htm)
* Rash
* Swelling of any area of body
* Weight gain (from retaining fluid)

## **Exams and Tests**

Your health care provider will perform a physical exam. This may reveal:

* Abnormal lung or heart sounds
* High blood pressure
* Fluid in the lungs (pulmonary edema)

Common tests include:

* Blood chemistry
* BUN and blood creatinine levels
* Complete blood count
* Kidney biopsy
* Kidney ultrasound
* Urinalysis

## **Treatment**

Treatment depends on the cause of the problem. Avoiding medicines that lead to this condition may quickly relieve symptoms.

Limiting salt and fluid in the diet can improve swelling and high blood pressure. Limiting protein in the diet can help control the buildup of waste products in the blood (azotemia), which can lead to symptoms of acute kidney failure.

If dialysis is necessary, it usually is required for only a short time.

Corticosteroids or stronger anti-inflammatory medicines such as cyclophosphamide can sometimes be helpful.

## **Outlook (Prognosis)**

Most often, interstitial nephritis is a short-term disorder. In rare cases, it can cause permanent damage, including long-term (chronic) kidney failure.

Acute interstitial nephritis may be more severe and more likely to lead to long-term or permanent kidney damage in older people.

## **Possible Complications**

Metabolic acidosis can occur because the kidneys aren't able to remove enough acid. The disorder can lead to acute or chronic kidney failure or end-stage kidney disease.

## **When to Contact a Medical Professional**

Contact your provider if you have symptoms of interstitial nephritis.

If you have interstitial nephritis, contact your provider if you get new symptoms, especially if you are less alert or have a decrease in urine output.

## **Prevention**

Often, the disorder can't be prevented. Avoiding or reducing your use of medicines that can cause this condition can help reduce your risk. If needed, your provider will tell you which medicines to stop or reduce.

## **Alternative Names**

**Tubulointerstitial nephritis; Nephritis - interstitial; Acute interstitial (allergic) nephritis**

**DIFFERENTIAL DIAGNOSIS**

Clinical presentations and laboratory results of TIN are not specific but overlap with most kidney diseases that cause AKI and renal insufficiency. When assessing suspected TIN in patients with renal insufficiency, the following problems should be considered:

**Acute Tubular Necrosis:** ATN is the most common cause of acute renal failure characterized by tubular cell necrosis for various reasons, such as ischemia, nephrotoxins such as aminoglycosides, heavy metals, urate, radiocontrast dye, or other toxic agents. In addition, manifestations like oliguria, metabolic acidosis, elevated BUN and creatinine, and electrolyte imbalances are similar to TIN. Muddy brown or granular casts are more likely with ATN.

**Atheroembolism:** Atheroembolism (cholesterol crystal emboli) should be considered in patients with a predominance of urinary WBC and RBC casts. Atheroemboli may also present with skin rashes, eosinophiluria, and eosinophilia. Skin changes usually include livedo reticularis and digital infarcts rather than the diffuse maculopapular rash of TIN. A history of endovascular diseases, older age, and obesity point towards atheroemboli.

**Glomerulonephritis:** A wide range of glomerulopathies ultimately can lead to renal impairment and mimic TIN to some degree. Some presentations of glomerulonephritis are similar to TIN, such as proteinuria and oliguria. WBC casts and dysmorphic RBCs are suggestive of glomerulonephritis rather than TIN.

**Obstructive Uropathy:** Urinary tract obstruction is a postrenal cause of acute renal failure. It is usually attributed to renal stones, tumors, and strictures and may cause anuria. In such patients, imaging helps to distinguish urinary obstruction from other causes of AKI. Obstruction can also cause urinary infections and must be treated to treat the infection effectively.

**Vascular Injury:** Various cardiovascular insults, such as renal artery stenosis, cardiac failure, vasculitis, reduced blood flow due to afferent arteriolar constriction in NSAID users, and reduced efferent arteriolar tone by ACE inhibitors, are common causes of AKI that clinically simulates TIN. The rash in vasculitis is typically purpura, while an allergic-type maculopapular rash would be more likely with drug-induced TIN.

**PROGNOSIS**

The prognosis depends on the cause of TIN, the timing of therapy, baseline renal function, prior offending agents, and exposure time to the underlying trigger. Chronicity, such as extensive fibrosis or tubular atrophy, portends worse outcomes. Early identification and removal of the cause improve renal outcomes.

Infectious causes of TIN are usually self-limited and respond well to antimicrobial treatment. Autoimmune-related TIN often relapses depending on the activity of the underlying disease, and renal function should continue to be followed. TINU, in particular, requires close ongoing follow-up with ophthalmology due to its recurrent nature and young cohort. Patients with kidney transplants who experience TIN due to viral causes should have closely monitored immunosuppressant and viral load levels, as each TIN episode can potentially worsen allograft function and precipitate a rejection episode.

**COMPLICATION**

Older patients are more vulnerable to complications. Renal insufficiency is a common manifestation that ultimately progresses to ESRD due to fibrosis of the interstitial and degeneration of tubular epithelial cells. In addition to renal insufficiency, inflammation or infection of proximal tubular cells can result in either decreased synthesis or hyporesponsiveness to erythropoietin by injured cells, leading to reduced RBC production by bone marrow and complications of anemia. TIN also increases angiotensin II activity, which leads to arterial hypertension due to sodium and fluid retention, vasoconstriction, and increased oxidative stress. Increased angiotensin concentration changes the hemodynamic status and oxidative stress, causing vasoconstriction.

**EPIDEMIOLOGY**

The global prevalence of acute AIN due to any cause is 1% to 3% among all renal biopsies. The overall prevalence of acute AIN increased to 15%-27% when the analysis was restricted to only AKI cases. Developed countries have a much higher proportion of drug-induced AIN than infection or autoimmune, while in developing countries, infectious causes are the most prevalent. Older patients are much more likely to have drug-induced AIN than systemic or autoimmune disease due both to the increased use of drugs and less active immune systems in this population. Each of the categories above has different demographic predominances. For example, TINU is a disease primarily of children and teenagers younger than 20, while anti-TBM disease is found in all age groups. Women tend to have much higher rates of SLE and Sjogren syndrome, while the distribution of sarcoidosis is about equal between the sexes.

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### **Henoch-Schonlein purpura**

**DEFINITION AND DESCRIPTION**

Henoch-Schonlein purpura is an inflammation of the small blood vessels of the skin, joints, bowels and kidneys. When blood vessels get inflamed, they can bleed into the skin, causing a reddish-purple rash (purpura).

The most striking feature of this form of vasculitis is a purplish rash, typically on the lower legs and buttocks. Henoch-Schonlein purpura can also cause abdominal pain and aching joints. Rarely, serious kidney damage can occur.

Henoch-Schonlein purpura can affect anyone, but it's most common in children under 10. The condition usually improves on its own. Medical care is generally needed if the disorder affects the kidneys.

**CAUSES**

In Henoch-Schonlein purpura, some of the body's small blood vessels become inflamed, which can cause bleeding in the skin, abdomen and kidneys. It's not clear why this initial inflammation develops. It may be the result of the immune system responding inappropriately to certain triggers.

Nearly half the people who have Henoch-Schonlein purpura developed it after an upper respiratory infection, such as a cold. Other triggers include chickenpox, strep throat, measles, hepatitis, certain medications, food, insect bites and exposure to cold weather.

**RISK FACTOR**

Factors that increase the risk of developing Henoch-Schonlein purpura include:

* **Age.** The disease mainly affects children younger than 10.
* **Sex.** Henoch-Schonlein purpura is slightly more common in males than in females.
* **Race.** White and Asian children are more likely to develop Henoch-Schonlein purpura than are black children.

**SYMPTOMS**

The four main characteristics of Henoch-Schonlein purpura include:

* **Rash (purpura).** Reddish-purple spots that look like bruises develop on the buttocks, legs and feet. The rash can also appear on the arms, face and trunk and may be worse in areas of pressure, such as the sock line and waistline.
* **Swollen, sore joints (arthritis).** People with Henoch-Schonlein purpura often have pain and swelling around the joints — mainly in the knees and ankles. Joint pain sometimes precedes the classical rash by one or two weeks. These symptoms subside when the disease clears and leave no lasting damage.
* **Digestive tract symptoms.** Many children with Henoch-Schonlein purpura develop belly pain, nausea, vomiting and bloody stools. These symptoms sometimes occur before the rash appears.
* **Kidney involvement.** Henoch-Schonlein purpura can also affect the kidneys. In most cases, this shows up as protein or blood in the urine, which you may not even know is there unless you have a urine test done. Usually this goes away once the illness passes, but some people develop persistent kidney disease.

**DIAGNOSIS**

Your doctor will be able to diagnose the condition as Henoch-Schonlein purpura if the classic rash, joint pain and digestive tract symptoms are present. If one of these signs and symptoms is missing, your doctor may suggest one or more of the following tests.

### **Lab tests**

No single laboratory test can confirm Henoch-Schonlein purpura, but certain tests can help rule out other diseases and make a diagnosis of Henoch-Schonlein seem likely. They may include:

* **Blood tests.** Your blood may be tested if your diagnosis isn't clear based on your signs and symptoms.
* **Urine tests.** Your urine may be tested for evidence of blood, protein or other abnormalities to determine if your kidneys are still working properly.

### **Biopsies**

People who have Henoch-Schonlein purpura often have deposits of a certain protein, IgA (immunoglobulin A), on the affected organ. Your doctor may take a small sample of skin so that it can be tested in a lab. In cases of severe kidney involvement, your doctor may suggest a kidney biopsy to help guide treatment decisions.

### **Imaging tests**

Your doctor may recommend an ultrasound to rule out other causes of abdominal pain and to check for possible complications, such as a bowel obstruction.

**TREATMENT**

Henoch-Schonlein purpura usually goes away on its own within a month with no lasting ill effects. Rest, plenty of fluids and over-the-counter pain relievers may help with symptoms.

### **Medications**

Corticosteroids, such as prednisone, may help shorten the time and intensity of joint and abdominal pain. Because these drugs can have serious side effects, discuss the risks and benefits of using them with your doctor.

### **Surgery**

If a section of the bowel has folded in on itself or ruptured, surgery may be needed.

**COMPLICATION**

For most people, symptoms improve within a month, leaving no lasting problems. But recurrences are fairly common.

Complications associated with Henoch-Schonlein purpura include:

* **Kidney damage.** The most serious complication of Henoch-Schonlein purpura is kidney damage. This risk is greater in adults than in children. Occasionally the damage is severe enough that dialysis or a kidney transplant is needed.
* **Bowel obstruction.** In rare cases, Henoch-Schonlein purpura can cause intussusception — a condition in which a section of the bowel folds into itself like a telescope, which prevents matter from moving through the bowel.

**EPIDEMIOLOGY**

IgAV is a rare disorder that typically affects children; however, the condition can also be seen in adults and adolescents. The majority of children are aged younger than 10. It is often more severe and likely to cause long-term renal disease in adults. It is the most common vasculitis among children, affecting 10 to 20 per 100,000 per year. IgAV is slightly more common among boys than girls but with about equal predilection in adults

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis of IgAV includes the following:

* IgA nephropathy
* Acute renal failure
* Acute glomerulonephritis
* Idiopathic thrombocytopenic purpura
* Disseminated intravascular coagulation
* Thrombotic thrombocytopenic purpura
* Hemolytic uremic syndrome
* Meningococcal meningitis
* Hypersensitivity vasculitis
* Systemic lupus erythematosus
* Polyarteritis nodosa
* Bacterial endocarditis
* Inflammatory bowel disease
* Wegener granulomatosis
* Rocky Mountain spotted fever

IgA Nephropathy (Berger Disease): A kidney disease caused by deposition of immunoglobulin A (IgA) in the glomeruli, leading to inflammation, blood and protein in urine, and potentially progressive kidney damage or failure.

Acute Renal Failure (Acute Kidney Injury): Sudden loss of kidney function due to injury, infection, or toxins, causing waste accumulation and fluid imbalance.

Acute Glomerulonephritis: Rapid inflammation of the glomeruli, often post-infectious, causing hematuria, proteinuria, edema, and impaired kidney function.

Idiopathic Thrombocytopenic Purpura (ITP): An autoimmune disorder leading to low platelet counts and increased bleeding risk without an identifiable cause.

Disseminated Intravascular Coagulation (DIC): A systemic activation of coagulation causing widespread clotting and bleeding due to consumption of clotting factors.

Thrombotic Thrombocytopenic Purpura (TTP): A rare disorder with small vessel clots, low platelets, hemolytic anemia, neurological symptoms, and kidney involvement.

Hemolytic Uremic Syndrome (HUS): Characterized by hemolytic anemia, thrombocytopenia, and acute kidney injury, often triggered by bacterial infections.

Meningococcal Meningitis: Severe bacterial infection of the meninges causing fever, headache, neck stiffness, and risk of septicemia.

Hypersensitivity Vasculitis: Immune complex-mediated inflammation of small blood vessels causing palpable purpura and systemic symptoms.

Systemic Lupus Erythematosus (SLE): A systemic autoimmune disease affecting multiple organs including kidneys, skin, joints, and blood vessels.

Polyarteritis Nodosa: A necrotizing vasculitis of medium-sized arteries causing organ ischemia and systemic symptoms.

Bacterial Endocarditis: Infection of heart valves causing fever, embolic phenomena, and immune complex-mediated symptoms.

Inflammatory Bowel Disease (IBD): Chronic inflammation of the gastrointestinal tract, including Crohn’s disease and ulcerative colitis, sometimes with systemic manifestations.

Wegener Granulomatosis (Granulomatosis with Polyangiitis): A vasculitis affecting small to medium vessels with granuloma formation, involving respiratory tract and kidneys.

Rocky Mountain Spotted Fever: A tick-borne rickettsial infection causing fever, rash, and vasculitis.

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### **cystic kidney disease**

**DEFINITION AND DESCRIPTION**

Cystic kidney disease describes a group of conditions that cause fluid-filled sacs (cysts) to form in or around your kidneys. Kidney cysts can prevent your kidneys from filtering wastes and excess water out of your blood. Cystic kidney disease can lead to kidney failure.

Another name for cystic kidney disease is renal cystic disease.

#### **How common is cystic kidney disease?**

It depends. Some cystic kidney diseases are very common. For example, simple kidney cysts occur in about 1 out of every 10 people. But other forms of cystic kidney disease are rare.

## **Symptoms and Causes**

### **What are the symptoms of cystic kidney disease?**

The various cystic kidney diseases have different symptoms. But some of the most common symptoms include:

* Back pain
* Flank pain
* Blood in your pee (hematuria)
* Trouble peeing, including not peeing as much as you usually do
* Enlarged kidneys or kidney masses
* Headaches
* High blood pressure
* Kidney infections
* Kidney stones

### **What causes cystic kidney disease?**

Cystic kidney diseases have different causes. Some may result from genetic variations. Others might develop over time due to diseases, birth defects or age.

Cysts occur when renal tubule pieces detach from a larger parent tube. Your kidneys have thousands of tiny tubes that clean your blood and release pee into your bladder.

#### **Who does cystic kidney disease affect?**

Risk factors for cystic kidney disease vary widely across the different types. But in general, you’re more likely to get cystic kidney disease if you’re:

* 50 or older
* Have chronic kidney disease (CKD) or kidney failure
* Have an abnormal gene

### **What are the complications of cystic kidney disease?**

Some of the more common complications of the various cystic kidney diseases include:

* Kidney failure
* Heart valve problems (more common if you have polycystic kidney disease)
* Liver cysts and pancreatic cysts (more common if you have polycystic kidney disease)
* Problems with growth and development delays in infants

## **Diagnosis and Tests**

### **How is cystic kidney disease diagnosed?**

A healthcare provider will ask you about your symptoms and review your medical history. They’ll also order one or more of the following imaging tests to check for kidney cysts:

* Prenatal ultrasound or kidney ultrasound
* CT scan
* MRI

A provider will also likely order blood tests and a pee test (urinalysis) to see how well your kidneys are filtering your blood.

## **Management and Treatment**

### **Is cystic kidney disease curable?**

Simple kidney cysts that don’t cause any symptoms may not need treatment. A healthcare provider may monitor the cysts and perform annual ultrasounds to make sure they don’t grow. If the cysts are painful or cause other symptoms, they may perform fine-needle aspiration to drain the cyst or laparoscopic surgery to cut or burn away the cyst tissue.

If you develop kidney failure from cystic kidney disease, a provider may recommend:

* Dialysis to clean your blood when your kidneys no longer work
* Kidney transplant (you may get a kidney from a living or deceased organ donor)
* Blood pressure medications (antihypertensives) to manage high blood pressure
* Lifestyle changes to manage high blood pressure, including at least 30 minutes of activity per day, maintaining a healthy weight for you and avoiding tobacco products, including smoking

#### **Should a polycystic kidney be removed?**

It depends. Unless you’re in a lot of pain or have other symptoms, healthcare providers usually don’t recommend removing your kidneys, even if they stop filtering wastes. They may still filter excess water from your body. But a provider may recommend a nephrectomy to remove your damaged kidneys if they cause a lot of pain or other symptoms.

## **Prevention**

### **How can I prevent cystic kidney disease?**

There’s no way to prevent cystic kidney disease. But talking to a healthcare provider when you first notice symptoms and working with a nephrologist may help slow the progression of some forms of cystic kidney disease.

## **Outlook / Prognosis**

### **What can I expect if I have cystic kidney disease?**

There’s no cure for cystic kidney disease. But there are many treatment options to slow the progression of polycystic kidney disease. You may need dialysis or a kidney transplant.

#### **At what age do people with PKD go into kidney failure?**

This can vary. It depends on the specific genetic variation. But it often has a similar pattern in a given family. PKD (polycystic kidney disease) involves two major genes — *PKD1* and *PKD2*. People with the *PKD1* gene variation tend to go on to kidney failure sooner (around mid-50s) than people with the *PKD2* gene variation (early 70s).

Your healthcare team will give you a better idea of what to expect according to your unique situation.

#### **What is the life expectancy of someone with polycystic kidney disease?**

Nearly 80% of people with autosomal dominant polycystic kidney disease have preserved kidney function at 50. Over 50% have preserved kidney function into their early 70s.

About one-third of babies who have autosomal recessive polycystic kidney disease at birth don’t survive. Babies who do survive need treatment for the rest of their lives.

## **Living With**

### **What foods should I avoid with kidney cysts?**

You may need to work with a renal dietitian to develop kidney-friendly eating patterns if you have chronic kidney disease or reduced kidney function due to cystic kidney disease. This may include:

* Avoiding foods high in sodium (salt)
* Limiting the amount of protein you eat
* Eating heart-healthy foods
* Avoiding foods that have high amounts of the electrolytes phosphorus and potassium, including dairy, beans, bananas, oranges and potatoes

### **When should I see a healthcare provider?**

Call a healthcare provider or get to the nearest emergency room if you have the following signs of sudden kidney failure, including:

* Abdominal pain
* Trouble peeing, including not peeing as much as you typically do
* Swelling (edema), especially around your hands, ankles or face
* Feeling very tired (fatigue) or drowsy
* Itchy skin
* Loss of appetite
* Nausea and vomiting
* Shortness of breath

# Medullary cystic kidney disease

ADTKD; Medullary cystic kidney disease; Renin associated kidney disease; Familial juvenile hyperuricemic nephropathy; Uromodulin associated kidney disease

Autosomal dominant tubulointerstitial kidney disease (ADTKD) is a group of inherited conditions that affect the tubules of the kidneys, causing the kidneys to gradually lose their ability to work.

Medullary cystic kidney disease type 1 (MCKD1) is an inherited condition that affects the kidneys. It leads to scarring (fibrosis) and impaired function of the kidneys, usually beginning in adulthood. The kidneys filter fluid and waste products from the body. They also reabsorb needed nutrients and release them back into the blood. As MCKD1 progresses, the kidneys are less able to function, resulting in kidney failure.

Declining kidney function in people with MCKD1 leads to the signs and symptoms of the condition. The features are variable, even among members of the same family. Many individuals with MCKD1 develop high blood pressure (hypertension), especially as kidney function worsens. Some develop high levels of a waste product called uric acid in the blood (hyperuricemia) because the damaged kidneys are unable to remove uric acid effectively. In a small number of affected individuals, the buildup of this waste product can cause gout, which is a form of arthritis resulting from uric acid crystals in the joints.

Although the condition is named medullary cystic kidney disease, only about 40 percent of affected individuals have medullary cysts, which are fluid filled pockets found in a particular region of the kidney. When present, the cysts are usually found in the inner part of the kidney (the medullary region) or the border between the inner and outer parts (corticomedullary region). These cysts are visible by tests such as ultrasound or CT scan.

## **Causes**

ADTKD is caused by mutations in certain genes. These gene problems are passed down through families (inherited) in an autosomal dominant pattern. This means the abnormal gene is needed from only one parent in order to inherit the disease. Often, many family members have the disease.

With all forms of ADTKD, as the disease progresses, the kidney tubules are damaged. These are the structures in the kidneys that allow most fluid in the blood to be filtered and returned to the blood.

The abnormal genes that cause the different forms of ADTKD are:

* *UMOD* gene -- causes ADTKD-*UMOD*, or uromodulin kidney disease
* *MUC1* gene -- causes ADTKD-*MUC1*, or mucin-1 kidney disease
* *REN* gene -- causes ADTKD-*REN*, or familial juvenile hyperuricemic nephropathy type 2 (FJHN2)
* *HNF1B* gene -- causes ADTKD-*HNF1B*, or maturity-onset diabetes mellitus of the young type 5 (MODY5)

When the cause of ADTKD is not known or a genetic test has not been done, it is called ADTKD-NOS.

## **Symptoms**

Early in the disease, depending on the form of ADTKD, symptoms may include:

* Excessive urination (polyuria)
* Gout
* Salt cravings
* Urination at night (nocturia)
* Weakness

As the disease worsens, symptoms of kidney failure may develop, which include:

* Easy bruising or bleeding
* Fatigue, weakness
* Frequent hiccups
* Headache
* Increased skin color (skin may appear yellow or brown)
* Itching
* Malaise (general ill feeling)
* Muscle twitching or cramps
* Nausea
* Pale skin
* Reduced sensation in the hands, feet, or other areas
* Vomiting blood or blood in the stool
* Weight loss
* Seizures
* Confusion, decreased alertness, coma

## Exams and Tests

Your health care provider will examine you and ask about your symptoms. You'll likely be asked if other family members have ADTKD or kidney disease.

Tests that may be done include:

* 24-hour urine volume and [electrolytes](https://www.mountsinai.org/health-library/special-topic/electrolytes)
* Blood urea nitrogen (BUN)
* Complete blood count (CBC)
* Creatinine blood test
* Creatinine clearance -- blood and urine
* Uric acid blood test
* Urine specific gravity (will be low)

The following tests can help diagnose this condition:

* Abdominal CT scan
* Abdominal ultrasound
* Kidney biopsy
* Kidney ultrasound

## **Treatment**

There is no cure for ADTKD. At first, treatment focuses on controlling symptoms, reducing complications, and slowing the progression of the disease. Because so much water and salt may be lost, you may need to follow instructions on drinking plenty of fluids and taking salt supplements to avoid dehydration.

As the disease progresses, kidney failure develops. Treatment may involve taking medicines and diet changes, limiting foods containing phosphorus and potassium. You may need dialysis and a kidney transplant.

## **Outlook (Prognosis)**

The age at which people with ADTKD reach end-stage kidney disease varies, depending on the form of the disease. It can be as young as in the teens or in older adulthood. Lifelong treatment may control the symptoms of chronic kidney disease.

## **Possible Complications**

Kidney damage due to ADTKD may lead to the following health problems:

* Anemia
* Bone weakening and fractures
* Cardiac tamponade
* Changes in glucose metabolism
* Congestive heart failure
* End-stage kidney disease
* High blood pressure
* Hyponatremia (low blood sodium level)
* Hyperkalemia (too much potassium in the blood), especially with end-stage kidney disease
* Hypokalemia (too little potassium in the blood)
* Infertility
* Menstrual problems
* Miscarriage
* Pericarditis
* Peripheral neuropathy
* Platelet dysfunction with easy bruising
* Skin color changes

## **When to Contact a Medical Professional**

Contact your provider if you have any symptoms of urinary or kidney problems.

## **Prevention**

Autosomal dominant tubulointerstitial kidney disease is an inherited disorder. It may not be preventable.

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**Hemolytic uremic syndrome (HUS)**

Hemolytic uremic syndrome (HUS) is a condition that can occur when small blood vessels become damaged and inflamed. This damage can cause clots to form in the vessels all through the body. The clots can damage the kidneys and other organs. Hemolytic uremic syndrome can lead to kidney failure, which can be life-threatening.

Anyone can get hemolytic uremic syndrome. But it's most common in young children. Most often, infection with certain strains of Escherichia coli (E. coli) bacteria is the cause.

Other infections, certain medicines or conditions such as pregnancy, cancer or autoimmune diseases can cause hemolytic uremic syndrome. It also can be the result of certain gene changes.

Hemolytic uremic syndrome is serious. But treating it in time leads to a full recovery for most people, especially young children.

CAUSES

The most common cause of hemolytic uremic syndrome is infection with certain strains of E. coli bacteria. This is especially true for children under age 5. Some of the E. coli strains make a toxin called Shiga toxin. These strains are called Shiga toxin-producing E. coli (STEC).

Most of the hundreds of types of E. coli are typical and harmless. But some strains of E. coli can lead to hemolytic uremic syndrome.

Other causes of hemolytic uremic syndrome can include:

* **Other infections.** This can include infection with pneumococcal bacteria, human immunodeficiency virus (HIV) or a flu virus.
* **Certain medicines.** These can include some of the medicines used to treat cancer and some medicines used to keep people who receive donor organs from rejecting the organs.
* **Complications of other conditions.** Rarely, these conditions can include pregnancy or conditions such as autoimmune disease or cancer.

An uncommon type of hemolytic uremic syndrome, called atypical, can be passed down through families. People who inherit the gene that causes this form of hemolytic uremic syndrome don't always get the condition. But an infection, the use of certain medicines or ongoing health conditions can start hemolytic uremic syndrome in people with the gene.

**RISK FACTOR**

Hemolytic uremic syndrome caused by E.coli can occur if you:

* Eat meat, fruit or vegetables with the bacteria.
* Swim in pools or lakes that have feces with the bacteria.
* Have close contact with an infected person.

The risk of getting hemolytic uremic syndrome is highest for:

* Children 5 or younger.
* People who have weakened immune systems.
* People with certain gene changes.

**SYMPTOMS**

The symptoms of hemolytic uremic syndrome vary, depending on the cause. The first symptoms of hemolytic uremic syndrome caused by E. coli bacteria might include:

* Diarrhea, which is often bloody.
* Pain, cramping or bloating in the stomach area.
* Fever.
* Vomiting.

All forms of hemolytic uremic syndrome damage blood vessels. This damage causes red blood cells to break down, called anemia. The condition also causes blood clots to form in the blood vessels and, in turn, damage the kidneys.

Symptoms of these changes include:

* Loss of color in the skin.
* Extreme tiredness.
* Easy bruising.
* Unusual bleeding, such as bleeding from the nose and mouth.
* Decreased urinating or blood in the urine.
* Swelling, called edema, of the legs, feet or ankles. Swelling occurs less often in the face, hands, feet or entire body.
* Confusion, seizures or stroke.
* High blood pressure.

**DIAGNOSIS**

A physical exam and lab tests can confirm a diagnosis of hemolytic uremic syndrome. Lab tests might include:

* **Blood tests.** These tests can show if the red blood cells are damaged. Blood tests also can show a low platelet count, low red blood cell count or a higher than usual level of a waste product usually removed by the kidneys, called creatinine.
* **Urine test.** This test can find unusual levels of protein and blood and signs of infection in urine.
* **Stool sample.** This test might find E. coli and other bacteria in stool.

If the cause of hemolytic uremic syndrome isn't clear, other tests might help find the cause.

**Treatment**

Hemolytic uremic syndrome needs treatment in the hospital. Treatment involves replacing lost fluids and minerals to make up for the kidneys not removing fluids and waste as well as usual. It also might involve getting nutrition through a vein.

### **Transfusions**

In the hospital, you might receive red blood cells or platelets through a vein, a process called a transfusion.

* Red blood cells can help reverse symptoms of anemia.
* Platelets can help blood clot better in people who are bleeding or bruising easily.

### **Medicines**

Lasting kidney damage from hemolytic uremic syndrome might be treated with a medicine to lower blood pressure. This medicine might prevent or slow more kidney damage.

For complications or the atypical form of hemolytic uremic syndrome, treatment might include a medicine called eculizumab (Soliris) to help prevent more damage to the blood vessels.

Anyone taking eculizumab needs to have a vaccination to prevent meningitis, a possible serious side effect of the medicine.

### **Surgery and other procedures**

Depending on the symptoms, the cause of hemolytic uremic syndrome and whether there are complications, treatment might include:

* **Kidney dialysis.** Dialysis removes waste and extra fluid from the blood. Dialysis is often done only until the kidneys begin working well again. But people with a lot of kidney damage might need long-term dialysis.
* **Plasma exchange.** Plasma is the fluid part of blood that helps blood cells and platelets circulate. Sometimes a machine is used to clear the blood of its own plasma and replace it with fresh or frozen donor plasma.
* **Kidney transplant.** Some people who have severe kidney damage from hemolytic uremic syndrome need a kidney transplant.

**COMPLICATION**

Hemolytic uremic syndrome can cause life-threatening complications, including:

* Kidney failure, which can be sudden, called acute, or happen over time, called chronic.
* High blood pressure.
* Stroke or seizures.
* Coma.
* Clotting problems, which can lead to bleeding.
* Heart problems.
* Digestive tract problems, such as problems with the intestines, gallbladder or pancreas.

**PREVENTION**

Meat or produce that has E. coli won't always look, feel or smell bad. To protect against E. coli infection and other illnesses from foods:

* Don't drink milk, juice or cider that isn't processed to make it safe to drink, called pasteurized.
* Wash hands well before eating and after using the restroom and changing diapers.
* Clean utensils and food surfaces often.
* Cook meat to an inside temperature of at least 160 degrees Fahrenheit (71 degrees Celsius).
* Defrost meat in the microwave or refrigerator, not on the counter.
* Keep raw foods separate from other foods. Don't put cooked meat on plates that had raw meat on them.
* Store meat below produce in the refrigerator to cut the risk of liquids such as blood dripping on produce.
* Avoid unclean swimming areas. Don't swim if you have diarrhea.

**WHEN TO SEE A DOCTOR**

See a member of your health care team right away if you or your child has bloody diarrhea or several days of diarrhea followed by:

* Urinating less.
* Swelling.
* Bruising.
* Unusual bleeding.
* Extreme tiredness.

Seek emergency care if you or your child doesn't urinate for 12 hours or more.

**EPIDEMIOLOGY**

HUS and aHUS are most often associated with children younger than 10, with most cases in those younger than 5 Globally, STEC causes 43 acute illnesses per 100,000 person-years and 3890 cases of HUS. STEC-HUS is one of the most common causes of pediatric renal replacement therapy. A large retrospective study showed that 15% of children (younger than 18) who presented to the emergency department with bloody diarrhea developed STEC-HUS.

STEC-HUS incidence is estimated at 0.57 cases per 100,000 children, and in the highest risk group—children aged 6 months to 2 years—the incidence is as high as 3 per 100,000 children. Incidence directly correlates with environmental exposure and agricultural practices, such as cattle raising, and most patients are diagnosed between April and September when cattle show higher colonization of STEC.

In contrast to HUS, aHUS is notably less frequent; nevertheless, aHUS exhibits substantially elevated morbidity and mortality rates. Like typical HUS, atypical HUS affects young children, predominantly those younger than 5, and prevalence is estimated at 2 to 9 cases per million people aged 20 years or younger. Cases due to *S Pneumonia* usually occur in the winter during cold season

**DIFFERENTIAL DIAGNOSIS**

Initially HUS may present similarly to other TMAs such as TTP, DIC, HELLP, and systemic vasculitis. Often clinical presentations and laboratory testing will rule out other causes.

**Thrombotic Thrombocytopenic Purpura (TTP)**

TTP is thrombotic microangiopathy characterized by a pentad of hemolytic anemia, thrombocytopenia, renal dysfunction, fever, and neurological dysfunction. TTP is due to a deficiency or mutation in "a disintegrin and metalloproteinase with a thrombospondin type 1 motif member 13" (ADAMTS13) and usually has adult-onset symptoms.

**Disseminated Intravascular Coagulation (DIC)**

DIC is the systemic activation of the coagulation cascade and is characterized by abnormal coagulation studies, including prolonged prothrombin time and activated partial thromboplastin time, elevated D dimer, and elevated fibrin degradation products, which are usually normal in HUS. Patients with DIC usually have serious underlying illnesses such as septic shock, trauma, or malignancy.

**HELLP Syndrome**

HELLP syndrome is observed in women pregnant in the third trimester or immediately postpartum, and it is characterized by hemolysis of red blood cells, elevated liver enzymes, and a low platelet count usually occurring with preeclampsia.

**Systemic Vasculitis**

Patients with systemic vasculitis typically present with inflammatory signs like fever, rash, and arthralgia, and lack prodromal diarrhea. Patients generally have a markedly elevated erythrocyte sedimentation rate.

**PROGNOSIS**

The prognosis of typical HUS is generally good with mortality estimated at 5% overall. However, up to 25% of patients with HUS develop long-term renal insufficiency with a glomerular filtration rate <80 mL/min/1.73 m2, hypertension, or proteinuria, which could predispose patients to increased renal insufficiency as they get older. The most significant prognostic indicator of ongoing renal dysfunction is the length of time on dialysis, with long-term complications evident after 2 to 3 weeks of dialysis dependency. One exception to the low mortality rate of typical HUS is in adults older than 60 who comprise most fatalities.

The course of aHUS has traditionally been much less benign than typical HUS; however, the advent of eculizumab treatment has decreased progression to ESRD or death in children from 30% to 50% down to 9% and in adults from 60% to 6% down to 15%

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**Nephrogenic systemic fibrosis**

**DEFINITION AND DESCRIPTION**

Nephrogenic systemic fibrosis is a rare disease that occurs mainly in people with advanced kidney failure with or without dialysis. Nephrogenic systemic fibrosis may resemble skin diseases, such as scleroderma and scleromyxedema, with thickening and darkening developing on large areas of the skin.

Nephrogenic systemic fibrosis can also affect internal organs, such as the heart and lungs, and it can cause a disabling shortening of muscles and tendons in the joints (joint contracture).

For some people with advanced kidney disease, being exposed to older gadolinium-based contrast agents (group 1) during magnetic resonance imaging (MRI) and other imaging studies has been identified as a trigger for development of this disease. Recognition of this link has dramatically reduced the incidence of nephrogenic systemic fibrosis. Newer gadolinium-based contrast agents (group 2) are not associated with an increased risk of systemic nephrogenic fibrosis.

**SYMPTOMS**

Nephrogenic systemic fibrosis can begin days to months, and even years, after exposure to an older gadolinium-based contrast agent (group 1). Some signs and symptoms of nephrogenic systemic fibrosis may include:

* Swelling and tightening of the skin
* Reddened or darkened patches on the skin
* Thickening and hardening of the skin, typically on the arms and legs and sometimes on the body, but almost never on the face or head
* Skin that may feel "woody" and develop an orange-peel appearance
* Burning, itching or severe sharp pains in areas of involvement
* Skin thickening that inhibits movement, resulting in loss of joint flexibility
* Rarely, blisters or ulcers

In some people, involvement of muscles and body organs may cause:

* Muscle weakness
* Limitation of joint motion caused by muscle tightening (contractures) in arms, hands, legs and feet
* Bone pain, particularly in the hip bones or ribs
* Reduced internal organ function, including heart, lung, diaphragm, gastrointestinal tract or liver
* Yellow plaques on the white surface (sclera) of the eyes

The condition is generally long term (chronic), but some people may improve. In a few people, it can cause severe disability, even death.

**CAUSES**

The exact cause of nephrogenic systemic fibrosis isn't fully understood. Fibrous connective tissue forms in the skin and connective tissues, resulting in scarring of tissue throughout the body, most commonly the skin and subcutaneous tissues.

Exposure to older gadolinium-based contrast agents (group 1) during magnetic resonance imaging (MRI) has been identified as a trigger for development of this disease in people with kidney disease. This increased risk is thought to be related to the kidneys' reduced ability to remove the contrast agent from the bloodstream.

The Food and Drug Administration (FDA) recommends avoiding older gadolinium-based contrast agents (group 1) in people with acute kidney injury or chronic kidney disease.

Other conditions may increase the risk of nephrogenic systemic fibrosis when combined with existing kidney disease and exposure to older gadolinium-based contrast agents (group 1), but the link is uncertain. These include:

* Use of high-dose erythropoietin (EPO), a hormone that promotes the production of red blood cells, often used to treat anemia
* Recent vascular surgery
* Blood-clotting problems
* Severe infection

**RISK FACTOR**

The highest risk of nephrogenic systemic fibrosis after exposure to older gadolinium-based contrast agents (group 1) occurs in people who:

* Have moderate to severe kidney disease
* Have had a kidney transplant but have compromised renal function
* Are receiving hemodialysis or peritoneal dialysis
* Have acute kidney injury

**DIAGNOSIS**

Diagnosis of nephrogenic systemic fibrosis is made with:

* **Physical exam** for signs and symptoms of the disease, and evaluation for a possible history of MRI using a gadolinium-based contrast agent when advanced kidney disease is present
* **A sample of tissue (biopsy)** taken from the skin and muscle
* **Other tests as needed** that may indicate involvement of muscles and internal organs

**TREATMENT**

There is no cure for nephrogenic systemic fibrosis, and no treatment is consistently successful in halting or reversing the progression of the disease. Nephrogenic systemic fibrosis only occurs rarely, making it difficult to conduct large studies.

Certain treatments have shown limited success in some people with nephrogenic systemic fibrosis, but more research is needed to determine if these treatments help:

* **Hemodialysis.** In people with advanced chronic kidney disease who are receiving hemodialysis, performing hemodialysis immediately after receiving a gadolinium-based contrast agent may decrease the possibility of nephrogenic systemic fibrosis.
* **Physical therapy.** Physical therapy that helps stretch the involved limbs may help slow the progression of joint contractures and preserve movement.
* **Kidney transplant.** For people who are appropriate candidates, improvement in renal function because of a kidney transplant may help improve nephrogenic systemic fibrosis over time.
* **Extracorporeal photopheresis with ultraviolet A.** This treatment involves drawing the blood outside the body and treating the blood with a drug that sensitizes it to ultraviolet light. The blood is then exposed to ultraviolet light and returned to the body. Some people have shown improvement after receiving this therapy.

These medications are experimental, but not currently in use. They have been shown to help some people, but side effects limit their use:

* **Imatinib (Gleevec).** Although this treatment shows some promise in reducing skin thickening and tightening, more research is needed.
* **Pentoxifylline (Pentoxil).** There is limited success with this medication, which theoretically decreases the thickness and stickiness (viscosity) of blood, aiding circulation. More research is needed.
* **Sodium thiosulfate.** Possible benefit has been shown using this medication, but more research is needed.
* **High-dose intravenous immune globulin.** Possible benefit has been shown using this medication, but more research is needed.

**PREVENTION**

Avoidance of older gadolinium-based contrast agents (group 1) is key to preventing nephrogenic systemic fibrosis, as newer gadolinium-based contrast agents (group 2) are safer and are not associated with increased risk.

**PROGNOSIS**

Nephrogenic systemic fibrosis is considered a debilitating, progressive disease. This condition causes visceral and cutaneous fibrosis in patients with severe renal insufficiency exposed to GBCAs. Although skin changes associated with NSF may be improved after the restoration of kidney function, especially after recovery from acute kidney injury, this condition does not usually regress spontaneously and almost always progresses relentlessly. The high mortality rate does not come from the cutaneous lesions but rather from visceral fibrosis, particularly in the cardiac and respiratory systems.

One group observed that 24-month mortality following examination was 48% in patients with skin changes and 20% in the cases where skin changes were absent. Within a few weeks of disease onset, most patients became wheelchair-bound due to contractures; patients can also experience flexion contractures if the condition involves a joint. Mortality in many patients has been attributed to falls and other complications due to limited mobility. In addition, many patients have reported causalgia and maddening pruritus.

EPIDEMIOLOGY

Nephrogenic systemic fibrosis is an iatrogenic disease first identified in 2000. According to a report by the FDA, about 4.5 million Americans are exposed to GBCAs annually. Until 2019, the FDA reported 3094 cases of NSF, including 742 deaths and 2922 serious cases. Of note, the US has the second-highest MRI utilization rate after Germany. As noted above, 32 cases were reported in each of 2019 and 2020.

NSF affects any ethnicity, sex, or age. The risk of developing NSF depends on the amount of residual renal function and the type of GBCA used. The amount of the initial dose and the cumulative dose of GBCA can also increase the risk. When high-risk group 1 GBCAs were used, the estimated incidence was 36.5 cases per 100,000 gadolinium-enhanced MRI examinations. Systematic reviews have found that no patients developed biopsy-proven NSF among patients receiving group 2 or 3 GBCAs; the estimated risk of NSF when using group 2 or 3 GBCAs is less than 0.07%

**DIFFERENTIAL DIAGNOSIS**

Nephrogenic systemic fibrosis diagnosis relies on excluding the other differential diagnoses; thus, healthcare professionals should be aware of these differentials. The following diseases encompass the most likely differential diagnosis:

* β-microglobulin amyloidosis
* Borreliosis
* Calciphylaxis
* Chronic graft versus host disease
* Drug-induced fibrosis
* Dermatofibrosarcoma protuberans
* Early cellulitis
* Early panniculitis
* Eosinophilic fasciitis
* Fibroblastic rheumatism
* Lipodermatosclerosis
* Radiation-induced fibrosis
* Scleroderma
* Scleromyxedema
* Phenylketonuria
* Porphyria cutanea tarda
* Superficial fibromatosis

## β-Microglobulin Amyloidosis (Dialysis-Related Amyloidosis)

A complication seen in patients undergoing long-term hemodialysis, caused by accumulation of β2-microglobulin protein that cannot be adequately cleared by failing kidneys or dialysis membranes. It leads to amyloid deposits mainly in osteoarticular structures such as joints, tendons, and bones, causing symptoms like carpal tunnel syndrome, shoulder pain, flexor tendon issues, and destructive bone lesions.

Other Conditions (Brief Descriptions)

* Borreliosis: Infection caused by Borrelia bacteria, transmitted by ticks; causes Lyme disease with skin rash, arthritis, and neurological symptoms.
* Calciphylaxis: Rare, serious condition in patients with kidney failure, characterized by calcification and thrombosis of small blood vessels causing painful skin necrosis.
* Chronic Graft Versus Host Disease: Immune-mediated complication after allogeneic stem cell transplant, causing fibrosis and inflammation in skin and other organs.
* Drug-Induced Fibrosis: Fibrotic tissue changes resulting from adverse drug reactions affecting skin or internal organs.
* Dermatofibrosarcoma Protuberans: A rare, slow-growing malignant skin tumor arising from dermal fibroblasts.
* Early Cellulitis: Acute bacterial skin infection causing redness, swelling, warmth, and pain.
* Early Panniculitis: Inflammation of subcutaneous fat presenting as tender nodules or plaques.
* Eosinophilic Fasciitis: Rare inflammatory disorder causing thickening and fibrosis of fascia with eosinophil infiltration.
* Fibroblastic Rheumatism: Rare rheumatologic condition with skin nodules and joint contractures due to fibroblast proliferation.
* Lipodermatosclerosis: Chronic inflammation and fibrosis of the skin and fat of the lower legs, often related to venous insufficiency.
* Radiation-Induced Fibrosis: Fibrotic tissue changes following radiation therapy, causing skin and organ stiffness.
* Scleroderma: Autoimmune connective tissue disease causing skin thickening and fibrosis, with possible internal organ involvement.
* Scleromyxedema: Rare disorder with widespread mucin deposition in skin causing papular eruptions and systemic symptoms.
* Phenylketonuria: Genetic metabolic disorder causing phenylalanine accumulation leading to intellectual disability if untreated.
* Porphyria Cutanea Tarda: Disorder of heme metabolism causing photosensitivity and blistering skin lesions.
* Superficial Fibromatosis: Benign fibrous tissue proliferation in superficial fascia, causing nodules or plaques.

Noting the history of GBCA exposure during the MRI examination will favor NSF among the differential diagnosis. Among the differential diagnoses, only β2-microglobulin amyloidosis occurs exclusively in individuals with advanced renal disease; however, it usually affects the shoulders, volar wrists, and tongue. As noted above, in contrast to NSF, scleroderma usually starts at the trunk and moves peripherally, while scleromyxedema usually involves the head and neck (the face is not involved in NSF)

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## **Abderhalden-Kaufmann-Lignac Syndrome**

Abderhalden-Kaufmann-Lignac Syndrome (AKL Syndrome) is a rare genetic disorder that presents itself during infancy and early childhood. AKL Syndrome is associated with widespread deposits of cystine crystals in various parts of the body, especially the conjunctiva and cornea. The excessive build up of cystine crystals can damage cells. AKL syndrome is also known as Nephropathic Cystinosis and is the most severe type of cystinosis. AKL Syndrome creates kidney damage and inhibits normal growth. Poor kidney function leads to the loss of salts and minerals in the body that are crucial to normal growth patterns.

#### **symptoms of Abderhalden-Kaufmann-Lignac Syndrome**

AKL Syndrome symptoms may include slow growth, excess thirst, dehydration, increased urination, and occasionally light sensitivity. The following are also symptoms to watch for:

* Developmental delay
* Dwarfism
* Rickets (soft bowed bones)
* Osteoporosis
* Renal tubular disease
* Aminoaciduria
* Glucose in the urine
* Low blood potassium levels
* Cysteine deposits in conjunctiva of eye
* Cysteine deposits in cornea

Cysteine deposition is most evident in the conjunctiva and cornea.

#### **What causes Abderhalden-Kaufmann-Lignac Syndrome?**

AKL Syndrome is caused by mutations in the CTNS gene on chromosome 17p13 and is inherited in autosomal recessive inheritance pattern. According to an article on the Mayo Clinic website, if a disease has an autosomal recessive inheritance pattern, both of the patient’s parents carry the gene but are not affected by the disease themselves. They have one mutated gene (recessive gene) and one normal gene (dominant gene) for the condition.

#### **DIAGNOSIS**

Diagnosis of AKL Syndrome is confirmed by measuring cystine levels in blood tests. If there is a high likelihood that a fetus may have it, the cystine levels can be taken from amniotic fluid.

## **Tests to Diagnose Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

### **Urine Examination**

* Urine Red Blood Cells are Absent
* Proteinuria- Higher concentration of protein found in urine
* Glycosuria- Higher concentration of carbohydrate found in urine.
* Low Osmolality
* Urine Electrolyte- Sodium, potassium, calcium and chloride excretion are measured.

### **Blood Examination**

* Mass Spectrometry- Mass spectrometry test is used to measure the white blood cells cysteine levels.
* Blood Electrolyte Study-Following electrolytes show abnormal levels.
  + Hypokalemia level – Low blood potassium level
  + Hypophosphatemia- Low serum phosphate level
  + Hypernatremia- Low sodium level is observed
  + Bicarbonate- Low bicarbonate level is observed.
  + pH- acidosis
* Blood Gas Studies-
  + Metabolic acidosis is observed.

### **Examination of Crystal-**

* Cystine crystals are examined in alkaline urine. Crystals are hexagonal in shape and colorless. Since crystals are colorless often crystals are examined under polarized background and polarized color inference.

### **Microbiology Studies for Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

* White Blood Cells- Cystine levels are examined within polymorphonuclear leukocytes.
* Fibroblast Culture- Cystine levels are measured in cultured fibroblast.

### **Kidney Biopsy for Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

* Histological examinations of kidney cells are performed to observe intracellular cystine.

### **Radiological Studies for Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

* Renal Ultrasound Examination- To evaluate kidney stone and size.
* CAT Scan- To evaluate kidney stone and size.
* MRI of the kidney- To evaluate kidney stone and size.

## **Treatment for Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

### **Symptomatic Treatment For Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

* Rehydration
* High dosage of vitamin D
* Electrolyte Supplements- Sodium citrate to treat metabolic acidosis
* Electrolyte Replacement- Potassium, bicarbonate and phosphate abnormalities treated with oral or intravenous supplements.

### **Therapeutic Treatment For Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

* **Cysteamine For Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**
* Cysteamine is a cystine depleting agent.4
* Cysteamine is the only medication, which has the potential to slow down the progression of cystinosis by removing cystine from cells.
* Cysteamine is a cystine-lowering agent, which depletes cystine levels in cells.

**NSAIDs To Treat Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

* Indomethacin- Reduces diuresis and water loss.
* May cause gastric (stomach) ulcer.

**Thyroid Hormone Treatment for Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

* Replace thyroid hormone

**Growth HormoneTreatment for Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

* Growth hormone is tried for improvement of growth.

**Cysteamine Eye Drops for Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

* Removes the cystine crystals in the cornea. Symptomatic relief of photophobia may be observed in most of the cases following treatment.

**Renal Transplantation for Abderhalden–Kaufmann–Lignac Syndrome or Nephropathic Cystinosis**

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Cysteamine is a cystine depleting drug that lowers cystine levels in AKL Syndrome patients. It is the only drug indicated for treating AKL Syndrome. It’s important that patients receive a diagnosis early and are started on treatment right away. By following these two steps, providers and patients can slow the development of AKL Syndrome and slow the progression of symptoms. Cystagon™, a capsule form of cysteamine, has been on the market since 1994. Unfortunately, it has to be taken every six hours of every day which can be quite inconvenient with most medications. However, Cystagon™ smells and tastes horrible. That makes patient compliance with dosing much more difficult. Even patients who’ve been treated will eventually require a kidney transplant.

[Abderhalden-Kaufmann-Lignac Syndrome or Nephropathic Cystinosis | Causes | Symptoms | Treatment](https://www.epainassist.com/abdominal-pain/kidney/abderhalden-kaufmann-lignac-syndrome-or-nephropathic-cystinosis)

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#### **IGM Nephropathy**

**DEFINITION AND DESCRIPTION**

IgM is short for Immunoglobulin M, one of the types of antibody our body produces to fight infection. This circulates in the blood. Nephropathy is a scientific term for kidney disease.

In this condition, IgM settles in the kidney and causes scarring and inflammation within the kidney, which can only be seen clearly under the microscope. Therefore, it is normally only diagnosed after a biopsy test of the kidney. What doctors see under the microscope is that the “glomeruli”, which are the tiny structures which filter the blood to make urine, are damaged by deposits of IgM.

#### **What are the symptoms of IGM Nephropathy?**

These are variable from case to case. In most cases there are no symptoms, but the damage to the glomeruli causes some blood and/or protein to appear in the urine. This blood is often invisible, and only detected on routine medical check-ups. In other cases the blood may be visible, coming in attacks every so often. Normally this condition is quite painless. However, in some cases there may be some pain over the kidneys, often occurring in attacks after a viral infection.

#### **What causes IGM Nephropathy?**

Basically no-one knows fully. IgM is part of the body’s defence against infection. As the antibody travels around in the blood and passes through the kidney it can get deposited in the filters (glomeruli) and then can cause an inflammatory reaction. Doctors do not know why this happens, and unfortunately cannot stop it.

#### **What will happen if I have IGM Nephropathy?**

The outcome is very variable, and you will need to ask your specialist how things are likely to be in your case. The possibilities are:-

1. It may continue unchanged for many years, requiring only regular check-ups with blood tests. This is probably the case in a majority of patients.
2. It may go away on its own in some cases.
3. In occasional cases kidney failure develops, leading to the question of dialysis or transplantation.

#### **What are the complications of IGM Nephropathy?**

1. High blood pressure may develop. This damages the kidneys and puts a strain on the heart and the rest of the circulation. Therefore high blood pressure should be treated vigorously.
2. As noted above, kidney failure may sometimes occur. If so, it develops slowly, so you need not be concerned about a sudden change in your condition overnight.
3. There may be pain over the kidneys in a minority of cases.

#### **Are there any treatments?**

This is not an easy condition to treat, and usually doctors rely on treatment of the blood pressure. Because the condition is caused by antibodies, treatments to reduce antibody production might be logical. However the drugs used for this have many side effects, and such treatment is very experimental.

#### **Can I lead a normal life with IGM Nephropathy?**

In most cases this condition does not affect normal life in any way. There is no special diet that will make the disease go away or get worse. You can continue with physical exercise and sports quite safely. The condition does not generally run in families, so you need not worry about having children. However, if you are planning a pregnancy you should discuss this with a doctor familiar with looking after pregnant women with kidney problems.

There will be some queries if you apply for mortgages or life insurance, so plan ahead and be prepared to have your doctors asked to supply a medical report.

**Differential diagnosis** of IgM nephropathy includes other kidney diseases that present with similar clinical and pathological features, particularly those involving immune complex deposition or mesangial proliferation. Key conditions to consider are:

* Minimal Change Disease (MCD): Often presents with nephrotic syndrome and minimal changes on light microscopy but lacks IgM deposits in the mesangium.
* Focal Segmental Glomerulosclerosis (FSGS): Shares clinical features such as proteinuria and nephrotic syndrome; some IgM nephropathy cases may evolve into FSGS, but FSGS shows segmental sclerosis on biopsy.
* IgA Nephropathy: Characterized by IgA deposits in the mesangium rather than IgM; presents with hematuria and proteinuria.
* Systemic Lupus Erythematosus (SLE) Nephritis: Can show mesangial immune deposits including IgM, but systemic symptoms and serology help differentiate.
* Rheumatoid Arthritis-associated Nephropathy: May have mesangial IgM deposits as part of systemic disease.
* Paraproteinemia-related Glomerulonephritis: Immune complex deposition including IgM can occur in monoclonal gammopathies.
* Alport Syndrome: Genetic disease with hematuria but no immune deposits; important to exclude.
* Other Secondary Causes of Mesangial IgM Deposits: Various systemic illnesses can cause mesangial IgM deposition, so clinical correlation and serological tests are essential to exclude secondary causes.

**Epidemiology of IgM Nephropathy (IgMN):**

* The reported prevalence of IgM nephropathy in renal biopsies varies widely, ranging from 2% to 18.5% in native kidney biopsies across different studies. This variability is influenced by differences in biopsy indications, diagnostic criteria, and populations studied.
* IgMN has been identified in both children and adults, with some studies suggesting a peak incidence in childhood or adolescence, while others report cases in older adults. There appears to be no significant gender or racial predilection.
* Most data come from biopsy series rather than population-based studies, so true incidence and prevalence in the general population remain unknown
* IgMN is recognized as an important cause of renal morbidity, especially in developing countries, and is often diagnosed in patients presenting with proteinuria or nephrotic syndrome
* The prevalence of hypertension among IgMN patients at diagnosis is reported to be around 33%, consistent with other glomerular diseases
* Long-term outcomes vary, with end-stage kidney disease (ESKD) developing in 6% to 25% of patients over extended follow-up periods

REFERENCES

<https://www.kidney.org.uk/igm-nephropathy>

IgA nephropathy

**IgA nephropathy (IgAN)**

is a rare kidney condition, one of a group of conditions called glomerulonephritis, where the body’s immune system damages the kidney.

IgA is short for Immunoglobulin A, one of the types of antibody our body produces to fight infection. It is designed to protect the gut against infection, and is generated by the gut, but also circulates in the blood. Nephropathy is a scientific term for kidney disease. In patients with IgA Nephropathy, the form of IgA molecules that circulate in the blood are abnormally sticky, and get trapped in the tiny filters in the kidney (called glomeruli) causing inflammation and damage.

#### **What are the symptoms of IgA Nephropathy?**

These are variable from case to case. In many cases there are no symptoms, but the damage to the glomeruli causes some blood to appear in the urine. This blood is often invisible, and only detected on routine medical check-ups. In other cases the blood may be visible, coming in attacks every so often. Sometimes an influenza type illness may spark off an attack of blood in the urine, which then clears after a few days.

Normally this condition is quite painless. However, in some patients who have acute attacks after the ’flu, there may be some pain over the kidneys and a feeling of sickness for a couple of days.

#### **What are the signs and symptoms of IgA Nephropathy?**

In many cases there are no symptoms, but the damage to the glomeruli can cause some blood to appear in the urine, sometimes making the urine look red or dark brown. Often, however, the blood is invisible, and only detected on routine medical check-ups. People sometimes have flare-ups of IgAN, often triggered by a cough or cold, where they see urine turn red or “coca-cola coloured”. Normally this condition is quite painless. However, in some patients who have acute attacks after the ’flu, there may be some pain over the kidneys and a feeling of sickness for a couple of days.

Some people also may have

* protein in the urine (proteinuria) – this is likely to only be seen under a microscope but if there a lot of protein, the urine may look frothy
* swelling or puffiness in parts of the body, especially in the legs
* High blood pressure which can damage the structure of the kidney if not treated

#### **What causes IgA Nephropathy?**

The exact cause of the disease is still unknown. IgA is known to play a role. In this condition, increased production of an abnormal IgA means that IgA gets trapped in the kidney and causes scarring and inflammation within the kidney, which can only be seen clearly under the microscope.

#### **How is IgA diagnosed?**

IgAN is often diagnosed after a routine urine test shows signs of blood and/or protein. A kidney biopsy may be conducted to confirm the diagnosis. What is seen under the microscope is that the “glomeruli”, which are the tiny structures which filter the blood to make urine, are damaged by deposits of IgA

#### **What will happen if I have IgA Nephropathy?**

The outcome is very variable, and you will need to ask your specialist how things are likely to be in your case. The possibilities are:-

1. It may continue unchanged for many years, requiring only regular check-ups with blood tests. This is probably the case in a majority of patients.
2. It may go away on its own in some cases.
3. In some cases kidney failure develops, leading to the question of dialysis and/or transplantation.

If treatment is needed, the initial aim is to protect kidney function by lowering blood pressure via lifestyle measures such as losing weight, stopping smoking and reducing salt in the diet.

#### **What are the complications of IgA Nephropathy?**

1. High blood pressure may develop. This damages the kidneys and puts a strain on the heart and the rest of the circulation. Therefore high blood pressure should be treated vigorously.
2. Kidney failure may sometimes occur. If so, it usually develops slowly, and your doctor will be able to give you an idea of how it is affecting you.
3. 3 There may be protein leakage from the kidneys. This may be slight and only detectable on urine tests. Occasionally, there are high levels of protein leakage leading to swollen ankles and high levels of cholesterol in the blood. This is called nephrotic syndrome and requires specialist assessment and treatment.
4. 4 There is a variant of IgA nephropathy called Henoch-Schönlein purpura. In this, the IgA antibodies affect not only the kidneys but also other parts of the body. A blotchy red rash may appear on the legs and buttocks. However, if you have had IgA nephropathy confined to the kidneys for some time, it would be very unusual for this to convert to the more serious Henoch-Schönlein purpura.

#### **Is there any treatment?**

This is not an easy condition to treat, and the aim is to protect the kidneys from further damage. Usually doctors rely on treatment of the blood pressure, using medications and lifestyle changes such as losing weight and reducing salt in the diet. If the cholesterol is very high, treatment to reduce the levels may help. If protein leaking in the urine, then medications (eg. ACE-inhibitors and SGLT2-inhibitors) can have protective benefits on the heart and kidney function over time, as shown by recent research. Some specialists also use long term treatment with Maxepa (fish oil) tablets.

Targeted-release budesonide can help to slow the decline of kidney function by reducing the amount of the IgA protein that builds up in the kidneys. There is also lots of research happening to test other drugs to see if we can improve the treatment of IgA Nephropathy and your doctor may ask you if you are interested in taking part in a clinical trial which may allow you to access some of the newer IgA Nephropathy treatments.

For Henoch-Schonlein purpura, doctors often use steroids (prednisolone), sometimes together with another drug, either cyclophosphamide, azathioprine, or mycophenolate.

#### **Can I lead a normal life with IgA Nephropathy?**

In most cases this condition does not affect normal life. There is no special diet that will make the disease go away or get worse. You can continue with physical exercise and sports quite safely. The condition does not generally run in families, so you need not worry about passing it on to your children. However, if you are planning a pregnancy you should discuss this with a doctor familiar with looking after pregnant women with kidney problems.

There will be some queries if you apply for mortgages or life insurance, so plan ahead and be prepared to have your doctors asked to supply a medical report.

There are currently a number of clinical trials taking place to research further treatment options for IgAN. Talk to your kidney team for more information.

**Epidemiology of IgA Nephropathy (IgAN):**

* Global Incidence: Approximately 2.5 cases per 100,000 people per year worldwide, with incidence increasing over time
* Geographic Variation:
  + Most common in East Asia (e.g., Japan incidence up to 4.2 per 100,000), followed by Europe, and least common in Africa (e.g., 0.06 per 100,000 in South Africa)
  + In Asia, IgAN may account for up to 40% of all glomerular diseases diagnosed by biopsy, compared to about 20% in Europe and 10% in North America[5](https://emedicine.medscape.com/article/239927-overview).
  + Higher prevalence in countries with routine urine screening programs (Japan, Hong Kong, Korea, Singapore) leads to earlier and more frequent diagnosis
* Ethnicity and Race:
  + Incidence is higher in Asian populations than in non-Asians
  + In the United States, incidence varies by region and race, but data are inconsistent regarding differences between Black and White populations
* Age and Gender:
  + Most commonly diagnosed in late teens to 30s.
  + Males are affected about twice as often as females
* Other Factors:
  + Socioeconomic status may influence diagnosis rates; some studies show higher diagnosis in more deprived areas, possibly due to healthcare access or biopsy practices
  + Differences in biopsy practices, healthcare access, and screening programs contribute to variability in reported incidence

**The differential diagnosis of IgA nephropathy (IgAN**) includes several conditions that share clinical, histological, or immunopathological features. Key differential diagnoses are:

* IgA Vasculitis (Henoch-Schönlein Purpura, HSP): A systemic small-vessel vasculitis with IgA deposition, presenting with purpuric rash, arthritis, abdominal pain, and renal involvement similar to IgAN. HSP is more common in children and generally has a more benign renal prognosis.
* IgA-Dominant Post-Infectious Glomerulonephritis: Occurs after infections, especially staphylococcal, with dominant IgA deposits but usually accompanied by clinical signs of infection and different histological features
* Alport Syndrome: A hereditary nephritis characterized by hematuria, hearing loss, and ocular abnormalities. Unlike IgAN, it lacks immune complex deposits and shows basement membrane abnormalities on electron microscopy
* Lupus Nephritis: Systemic lupus erythematosus can cause immune complex glomerulonephritis with multiple immunoglobulin deposits including IgA, but usually accompanied by systemic symptoms and positive serologies.
* Acute Poststreptococcal Glomerulonephritis: Presents with nephritic syndrome after streptococcal infection, with different immunofluorescence patterns (IgG and C3 dominant) and clinical course
* Other Small Vessel Vasculitis (e.g., ANCA-associated vasculitis): May cause hematuria and renal impairment but differ in serology and histopathology
* Thin Basement Membrane Disease: Causes isolated hematuria without immune deposits or significant proteinuria.
* Other Immune Complex Glomerulonephritides: Membranoproliferative GN and others may present with overlapping features but differ in immunofluorescence and clinical context

<https://emedicine.medscape.com/article/981516-differential>

<https://www.kidney.org.uk/iga-nephropathy>

<https://www.ncbi.nlm.nih.gov/books/NBK538214/>

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#### **Alport’s syndrome**

Alport’s syndrome is an inherited kidney disease. This means it is caused by a genetic mutation and runs in families. Men are more severely affected than women. It can lead to deafness as well as kidney failure.

#### **Why it is called Alport’s syndrome?**

Alport’s syndrome is named after Professor Arthur Cecil Alport, who recognised families with the syndrome in 1927. He was born in 1880 in South Africa, and graduated as a doctor in Edinburgh. First, he worked in Johannesburg (owning a small gold mine). After the First World War he moved to St Mary's Hospital, Paddington, London, where he was working when he described the syndrome of hereditary renal failure and deafness. Later he worked in Cairo, and died in 1959.

#### **What causes Alport’s syndrome**

Alport’s syndrome is caused by an abnormality in collagen in the body. Collagen is a ‘building block’, giving shape and strength to parts of the body not supported by bone. Cells, tiny packages of tissue that work to make energy and do all the 'work' inside our bodies, have no strength themselves. If cells were not held together, the human body would be like a lump of jelly. Bones are important in giving the human body shape and rigidity, but even soft tissues need holding together. This is the job of collagen.

There are several types of collagen, each of which are found in different parts of the body. Type 4 collagen is important in the kidney, ears and, to a lesser extent, in the eye. It is this type of collagen which is abnormal in Alport’s syndrome, leading to kidney failure, deafness, and abnormalities of the eyes. This abnormality occurs because the genetic code, which tells the body how to make collagen, is abnormal. Every gene has a specific name and the gene most commonly affected in Alport’s syndrome is called the COL4A5 gene. Other genes include COL4A3 and COL4A4. In around 15% of patients with the diagnosis, the genetic mutation arose in them rather than being passed from a parent.

The type 4 collagen gene is found on the ‘X’ chromosome, which determines the sex of a person. The human body has 23 pairs of chromosomes, one set from each parent. Hence, there are 46 chromosomes in total. One pair determines whether a person is male or female. This pair is made up of ‘X’ and ‘Y’ chromosomes. If someone has 2 X chromosomes ‘XX’, they are female. If someone has one X chromosome and one Y chromosome, ‘XY’, they are male. The Y chromosome is smaller than the ‘X’ chromosome, and it so happens that the COL4A5 gene is missing from the ‘Y’ chromosome. Therefore, if a male has a defective COL4A5 gene, there is no normal copy to help produce normal type 4 collagen. A woman has two ‘X’ chromosomes, hence the normal gene on the second chromosome helps produce normal type 4 collagen. Therefore, women with Alport’s syndrome are able to produce more normal collagen than men, and usually have mild disease. However they can pass Alport’s syndrome with severe disease, such as kidney failure to their sons. This type of inheritance, which is called 'sex linked' is important when family screening is considered.

#### **How is Alport’s syndrome diagnosed?**

Alport’s syndrome can be diagnosed in several ways.

· Evidence of blood or protein leak in the urine along with, a history of hearing loss or Alport’s syndrome in the family.

· A kidney biopsy (sample of kidney removed with a needle to look under a microscope). Alport’s syndrome has a particular appearance in the kidney when examined under the microscope.

· Testing DNA for genetic abnormalities, eg. COL4A5 gene.

#### **Family screening**

An explanation of which family members should be screened is given later. If someone is to be screened because of a family history, it may only be necessary to check the urine for blood. This can be done by taking a small sample of urine and dipping into it on a small plastic stick. Alport’s syndrome causes small amounts of blood to appear in the urine from a very early age. This test is not completely foolproof - a negative urine test may need to be repeated several times to be sure there is no blood, and blood can be present for other reasons, such as a urine infection.

#### **Kidney biopsy**

Alport’s syndrome causes a unique appearance when the kidney is examined under the microscope. Due to abnormal collagen the membrane that filters blood to make urine is split into several layers. Splitting is generally seen in men and in some women. Sometimes the membrane may just be very thin, which can make it more difficult to diagnose Alport’s syndrome.

If someone with a known family history of Alport’s syndrome has blood in the urine, it is usually not necessary to perform a [kidney biopsy](https://www.kidney.org.uk/kidney-biopsy). However, in some cases where the diagnosis is not proven, it may be necessary.

#### **Genetic testing**

The problem is that many different (well over 100) genetic abnormalities can cause Alport’s, and these differ from family to family. Genetic tests looking for a single genetic abnormality are relatively easy to perform. If it is possible to determine which genetic abnormality is present in a particular family, testing is possible.

Technology is available to perform genetic tests on human embryos in the laboratory (pre-implantation genetic diagnosis), so that the production of a child without Alport’s syndrome can be guaranteed. However, these techniques mean that the human egg has to be fertilised ‘in vitro’, in other words a ‘test tube baby’.

If you want to know more about genetic testing, you should consult a kidney specialist and a genetic specialist for up-to-date information.

#### **Diagnosis if typical eye abnormalities are found**

Sometimes Alport’s syndrome is diagnosed after an eye specialist has noticed the unusual eye problems that Alport’s can cause. Also, high tone deafness in someone with a family history of kidney trouble may alert a doctor to the possibility of Alport’s syndrome.

#### **Is it the same as Thin Basement Membrane disease?**

You may come across the term ‘Thin basement membrane disease’ which some may consider as a milder version of Alport’s syndrome. This is caused by mild mutations in the gene coding for Type 4 collagen, causing thinning of the filter membrane in the kidneys. Hence the affected individuals commonly have near non-visible amount of blood in the urine, almost never have kidney disease and do not have abnormalities of the ears or eyes.

#### **Is Alport’s syndrome common?**

Alport’s syndrome is not common. One or two out of 100 people starting dialysis have Alport’s syndrome, with a frequency in the general population of about 1 in 5,000, to 1 in 10,000. However, a greater number of women are likely to be carrying the mutated gene without symptoms.

#### **Is there any treatment to prevent kidney failure in Alport’s syndrome?**

At present, the abnormal gene cannot be replaced with a normal one hence no cure for Alport’s syndrome. However, as in all types of kidney disease, much can be done to try and reduce the rate of kidney damage, and to make someone feel as normal as possible as kidney failure develops.

[High blood pressure](https://www.kidney.org.uk/high-blood-pressure) is the most important factor that can speed up the decline in kidney function. Damaged kidneys cause high blood pressure, because one of the jobs of the kidneys is to control the level of blood pressure. High blood pressure damages the kidneys further, and so a vicious cycle develops. Strict control of the blood pressure can break this cycle, delaying the need for dialysis by years in some cases. There is some evidence that high blood pressure can start in childhood, so that starting treatment for high blood pressure even before the age of 10 can be beneficial.

Treatment for high blood pressure consists of a healthy, salt free diet with exercise. Any excess weight should be lost. Stop smoking and drink no alcohol, or have only a moderate intake. In most people with significant kidney disease, it is also necessary to use medications to reduce the blood pressure. Up to 4 or 5 different types of medications may be necessary in some people.

As kidney failure becomes advanced, anaemia may develop, which can be managed effectively with a drug called EPO.

#### **What about ear and eye damage - is this serious?**

Someone with Alport’s syndrome should also have their ears and eyes tested to detect any problems.

The inner ear contains type 4 collagen and is affected by Alport’s syndrome. Men are affected more than women. Some men may develop bilateral partial deafness, especially for high tones, and may require a hearing aid by the time they are in their late teens. However, this is not universal in Alport’s. Kidney failure may make the deafness worse, with some improvement after a kidney transplant. Women may have some hearing loss that could be detected by special tests done in hospital, but this does not often cause problems with normal conversation.

The medical term for the most common problem in the eye is ‘bilateral anterior lenticonus’ This means that, in both eyes, the front surface of the lens, the part of the eye over the pupil, bulges forwards. This change may be slight, and not visible to the naked eye. It may cause short sightedness, and it may be necessary to wear glasses. There may also be some dots and flecks on the back of the eye, but these are only visible to an eye specialist and should not affect vision.

#### **Does everyone with Alport’s syndrome develop kidney failure?**

Nearly all the men and about 1 in 10 women with Alport’s syndrome develop kidney failure. However, Alport’s syndrome can vary slightly from family to family. There is also a rare subtype of Alport’s in which kidney failure occurs in childhood.

It is rare for men with Alport’s to develop kidney failure before the age of 10 years. Most men develop kidney failure between the ages of 15 and 30, though in some families this is delayed to 50-70 years. Kidney failure develops slowly over a period of years, so that the need for dialysis can be planned; or in some cases, a kidney transplant might be carried out before dialysis was necessary.

Women with Alport’s syndrome have tiny amounts of blood in their urine (haematuria), sometimes with some [protein](https://www.kidney.org.uk/proteinuria). About 6 out of 10 women may develop protein in the urine. Although protein in the urine suggests a risk for progression to kidney failure, in many cases the kidney function remains normal and only about 1 in 10 women with Alport’s syndrome ever need dialysis or a kidney transplant.

#### **Can people with Alport’s syndrome have dialysis or a kidney transplant?**

Alport’s syndrome does not cause particular problems with dialysis. Nearly all people with Alport’s syndrome and kidney failure can have a kidney transplant, so long as they are generally fit. If a healthy family member is being considered as a donor they would have to undergo meticulous investigation. In general, kidney transplantation in Alport’s syndrome has a small risk of rejection due to Anti-Glomerular Basement Membrane Glomerulonephritis (Anti-GBM disease). This risk could be predicted by genetic testing of the recipient but no completed avoided.

#### **Should members of the family have tests to look for Alport’s syndrome?**

Yes. Both men and women with Alport’s syndrome have small amounts of blood in the urine from a very early age, so it is easy to test for Alport’s syndrome in relatives of someone known to be affected. However, it is not necessary to test every single family member. Specialist advice is necessary given the complex genetic inheritance of the disease. The above description of the inheritance of Alport’s syndrome applies to the 9 out of 10 families who have the commoner genetic problem. Some families are more complicated, and advice should be taken from a specialist in genetics.

The need for testing family members will be discussed from the point of view of a man with Alport’s syndrome, and then from the point of view of a woman with Alport’s syndrome.

#### **Who to test if a man has Alport’s syndrome**

#### **His parents**

Alport’s syndrome should have been inherited from his mother, though occasionally the genetic abnormality has occurred for the first time in the affected person. His mother should have urine tests for blood. If there is blood in the urine, kidney function and blood pressure should be tested, and a kidney specialist consulted. If the mother is completely clear, the father should be checked, in case there is a rarer variant of Alport’s syndrome.

#### **His brothers**

There is a 50:50 chance that a brother will have Alport’s syndrome. Urine should be tested for blood. If he has blood in the urine, kidney function and blood pressure should be measured, and a kidney specialist consulted. A brother might have Alport’s syndrome, and could pass this onto his daughters. If there is no blood in the urine on several tests, he should not have the Alport’s syndrome gene, and so cannot pass the condition onto his children.

#### **His sisters**

There is a 50/50 chance that each sister will have Alport’s syndrome, though remember that this is less serious in women than in men. Urine should be tested for blood. If she has blood in the urine, kidney function and blood pressure should be measured, and a kidney specialist consulted. If she has Alport’s syndrome, she could pass this onto her children. If there is no blood in the urine on several tests, she should not have Alport’s syndrome, and so cannot pass the condition onto her children.

#### **His sons**

A man with Alport’s syndrome cannot pass the condition onto his sons (unless he has one of the rarer variants of the disease). This is because the abnormal gene is on the ‘X’ chromosome, and in order to have a son, a man has to pass on his ‘Y’ chromosome to the child, and this does not carry Alport’s syndrome.

#### **His daughters**

Each daughter will have Alport’s syndrome, though remember that this is less serious in women than in men. Urine should be tested for blood to confirm the diagnosis. Kidney function and blood pressure should be measured, and a kidney specialist consulted. She could pass Alport’s syndrome onto her children (see below).

#### **Who to test if a woman has Alport’s syndrome**

#### **Her parents**

Alport’s syndrome should have been inherited from her mother or father. Her parents should have urine tests for blood. If there is blood in the urine, kidney function and blood pressure should be tested, and a kidney specialist consulted.

#### **Her brothers**

If Alport’s syndrome is inherited from the father, the male children should not have the syndrome. If Alport’s is inherited from the mother’s side, there is a 50:50 chance that he will have Alport’s syndrome. Urine should be tested for blood. If there is blood in the urine, kidney function and blood pressure should be measured, and a kidney specialist consulted. He might have Alport’s syndrome, and could pass this onto his daughters. If there is no blood in the urine on several tests, he should not have Alport’s syndrome, and so cannot pass the condition onto his children.

#### **Her sisters**

If Alport’s syndrome is inherited from the father, the female children should all have Alport’s syndrome. If Alport’s syndrome is inherited from the mother’s side, there is a 50:50 chance that the sister will have Alport’s syndrome. Urine should be tested for blood. If there is blood in the urine, kidney function and blood pressure should be measured, and a kidney specialist consulted. She might have Alport’s syndrome, and could pass this onto her children. If there is no blood in the urine on several tests, she should not have Alport’s syndrome, and so cannot pass the condition onto her children.

#### **Her sons**

There is a 50:50 chance that each son would have Alport’s syndrome. Urine should be tested for blood. If there is blood in the urine, kidney function and blood pressure should be measured, and a kidney specialist consulted. He might have Alport’s syndrome, and could pass this onto his daughters. If there is no blood in the urine on several tests, he should not have Alport’s syndrome, and so cannot pass the condition onto his children.

#### **Her daughters**

There is a 50:50 chance that a daughter will have Alport’s syndrome. Urine should be tested for blood. If there is blood in the urine, kidney function and blood pressure should be measured, and a kidney specialist consulted. She might have Alport’s syndrome, and could pass this onto her children. If there is no blood in the urine on several tests, she should not have Alport’s syndrome, and so cannot pass the condition onto her children

**EPIDEMIOLOGY**

Alport syndrome affects about 1 in 50,000 newborns, and males are more likely to be symptomatic than females. It is estimated that approximately 30,000 to 60,000 people in the United States (US) have this disorder. In the US, the overall incidence of end-stage renal disease (ESRD) in children is about 3% and 0.2% in the adult population.

Alport syndrome accounts for approximately 2.2% of children and 0.2% of adults with ESRD in the United States. In Europe, Alport syndrome accounts for 0.6% of patients with ESRD.

The common X-linked type of Alport syndrome resulting in ESRD predominantly affects males. However, the X-linked form of Alport syndrome affects almost as many females. Affected women are generally undiagnosed, but 15%–30% end up having renal failure by 60 years and hearing loss by middle age.

Alport syndrome is a significant cause of chronic kidney disease (CKD), leading to ESRD in adolescents and young adults, accounting for 1.5% to 3.0% of children on renal replacement therapies in Europe and the US.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnoses of Alport syndrome include the following:

* Immunoglobulin A nephropathy
* Thin GBM disease
* Acute post-streptococcal glomerulonephritis
* Medullary cystic disease
* Multicystic renal dysplasia
* Polycystic kidney disease

The most important diagnostic consideration in patients with Alport syndrome is thin basement membrane (TBM) disease, a collagen 4-related nephropathy closely related to Alport syndrome. The same genes appear to be involved in many individuals with the disorder. Unlike those with Alport syndrome, few extra-renal findings are present, symptoms are less severe, with progression to renal impairment is rarely found. Differentiating these disease processes is challenging, particularly in younger or female patients who are less likely to have other associated symptoms.

**PROGNOSIS**

In the X-linked disease form, the most common type of Alport syndrome, about 50% of males require dialysis or kidney transplantation by 30 years, and approximately 90% develop ESRD before 40. Female patients with X-linked Alport syndrome have a better prognosis, with about 12% developing end-stage renal disease (ESRD) by age 40. However, studies indicate significant renal morbidity in females with proteinuria and hearing impairment.

By age 60, this rate increases to about 30%, and by 60 years of age, the rate of ESRD approaches 40%. In the female population, proteinuria and hearing loss are risk factors for the progression to ESRD. In comparison, the autosomal recessive form of Alport syndrome can cause kidney failure by age 20. In contrast, the autosomal dominant form of the disease typically has a delay in ESRD until middle age.

**COMPLICATION**

Alport syndrome affects multiple organ systems. It can lead to the following complications:

* End-stage renal disease (ESRD)
* Hearing loss
* Visual defects
* Leiomyomatosis (smooth muscle overgrowth in the respiratory and gastrointestinal tract)
* Aneurysms of the thoracic and abdominal aorta
* Mental retardation

<https://www.ncbi.nlm.nih.gov/books/NBK470419/>

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## **Nephropathy**

The kidneys are made up of small filtering units called the nephrons. The deterioration of the functions of the nephrons is called nephropathy. Acute kidney infection is caused by different Plasmodium species like falciparum and ovale. The kidney condition can worsen due to low hydration and fluid loss caused by vomiting, sweating, and dehydration. Chronic kidney disease is generally associated with malaria in patients suffering from repeated episodes of infection. Acute kidney infection is the most common complication of malaria and can occur in patients with a severe disease, P.falciparum.

## **How Does Malaria Cause Renal Failure?**

Amongst all the Plasmodium species, P. falciparum causes the most severe form of malaria and is mainly responsible for acute kidney infections. The parasite in falciparum infection attacks the red blood cells (erythrocytes). These parasitized erythrocytes start the three mechanisms:

Hemodynamic:

Hemodynamic disturbances include parasitized erythrocytes, which adhere to adjacent healthy red blood cells, blood platelets, and the capillary endothelium. This results in intravascular knots and obstructs the microcirculation inside the kidneys.

Immunologic:

Immunological dysfunction happens when the parasite interacts with the body's innate immune system, and activation of B cells occurs, which causes the production of cytokines leading to immunosuppression. This mechanism produces antibodies leading to the formation of an immune complex, which is further related to injury to the glomerulus.

Metabolic Disturbances:

Metabolic disturbances include hemolysis of the red blood cells, causing anemia, jaundice, bleeding, and acute renal failure.

## **What Are the Symptoms of Malarial Nephropathy?**

* Proteinuria - It is the presence of protein in increased levels in the urine.
* Microalbuminuria - A condition wherein the albumin level in the urine is in the range of 30 to 300 mg (less than 30 mg is considered the normal level).
* Jaundice - Increase in the amount of bilirubin due to excessive destruction of red blood cells.
* Anemia - Decrease in the hemoglobin concentration.
* Hypoxia - Decrease in oxygen supply to the tissues.
* Chills, Fever, and Sweating - These symptoms return to normal temperature.
* Hyponatremia - Decrease in the sodium content in the blood.
* Hyperkalemia - Increase in potassium content in the blood.
* Oliguria or Anuria - Decrease in or complete cessation of urine.
* Electrolyte Disturbance - Imbalance in electrolyte concentration of the blood.
* Malarial Glomerulonephritis - P. falciparum infection leading to glomerulonephritis (disease that injures the glomeruli, the part of the kidney that filters blood) is uncommon. Children are more affected by this complication. The correct incidence of the malaria complication is unknown, but it is estimated to be around 18 percent.
* Nephrotic Syndrome Associated With Malaria - P. falciparum infection resulting in nephrotic syndrome (a kidney disorder characterized by the release of excessive protein in urine) is uncommon.

## **What Are the Risk Factors of Malarial Nephropathy?**

Risk factors for high mortality include acute illness, advanced age, increased parasitemia, hyperkalemia, oliguria, hypotension, severe anemia, jaundice, and altered consciousness level. Patients with poor prognosis may suffer from multiple organ involvement, severe diarrhea, and respiratory distress.

## **How to Diagnose Malarial Nephropathy?**

Signs and symptoms of malaria include fever, sweat, flu-like illness, chills, malaise, headache, muscle aches, tiredness, nausea, vomiting, and diarrhea. Malaria may cause anemia (decrease in red blood cell count) and jaundice (yellowish discoloration of the skin and eyes) because of the loss of red blood cells. If untreated, the infection becomes severe and may cause kidney failure, coma, and death.

* The precise way to diagnose malaria is through blood examination for the presence of the malarial parasites.
* P. falciparum malaria-causing kidney injury can be diagnosed by urine analysis containing the kidney injury biomarkers, neutrophil gelatinase-associated lipocalin (NGAL), and kidney injury molecule-1 (KIM-1), which have the advantage of detecting AKI earlier than creatinine.

## **How to Treat Malarial Nephropathy?**

Treatment of malarial nephropathy includes antimalarial drugs and measures that require correction of water and electrolyte disturbances, fluid replacement, and dialysis. The drug of choice for the treatment of malaria is Chloroquine. P. falciparum has resistance to Chloroquine.

Therefore, therapy should include:

* Primaquine.
* Quinine.
* Benflumetol.
* Mefloquine.

Primaquine is used in P. vivax and P. ovale to prevent relapse. Intravenous Quinine is widely used to treat cerebral and other serious complications of falciparum malariae. Antimalarial drug doses should be based on the patient's weight to provide good efficacy. For severe malaria treatment, intravenous or intramuscular antimalarial drugs are given.

The most effective drug in treating severe malaria is Artesunate, which is given for at least 24 hours. The solution of Artesunate is diluted in 5 ml of 5 percent dextrose and given intravenously or intramuscularly.

Dialysis should be considered in acute kidney injury treatment for more effectiveness. Antimalarial drugs are not filtered in hemofiltration dialysis, so dialysis does not interfere with the specific treatment of malaria. Further increase in the development of vaccines is required to prevent severe cases leading to renal failure. Malaria can be more severe in pregnant women than in women who are not. Malaria can increase the risk of pregnancy problems like prematurity and miscarriage.

Conclusion

Taking antimalarial drugs to kill the parasites as soon as diagnosed can prevent the illness. Prevent oneself from mosquito bites, especially at night, which can be done by using insect repellent and wearing long-sleeved clothing. A person who becomes ill with a fever, malaria, or flu-like illness during travel should seek medical care immediately. If one is traveling to a country where malaria transmission occurs, precautions should be taken against contracting malaria. Treatment of malaria-associated nephropathy should include antimalarial drugs and measures to control acute kidney injury. Malaria should be treated sooner as it causes severe damage to multiple organs.

The **differential diagnosis** of malarial nephropathy includes other causes of acute kidney injury (AKI), glomerulonephritis, and systemic infections that can involve the kidneys, especially in malaria-endemic regions. Key conditions to consider are:

* Acute Tubular Necrosis (ATN) from other causes: Such as sepsis, ischemia, toxins, or rhabdomyolysis, which can mimic malarial AKI clinically and histologically.
* Other Infectious Causes of AKI:
  + Leptospirosis: Can cause fever, jaundice, and AKI similar to malaria.
  + Viral infections: Such as hepatitis or dengue may cause renal impairment.
  + Bacterial sepsis: Leading to septic acute kidney injury.
* Glomerulonephritis of Other Etiologies:
  + Immune-complex mediated glomerulonephritis from infections other than malaria.
  + Post-infectious glomerulonephritis, which can present with hematuria and proteinuria.
* Hemolytic Uremic Syndrome (HUS): Can occur secondary to infections including malaria, presenting with AKI, thrombocytopenia, and hemolytic anemia.
* Disseminated Intravascular Coagulation (DIC): May complicate severe malaria and cause renal microvascular thrombosis.
* Other Tropical Diseases Causing Renal Failure:
  + Visceral leishmaniasis
  + Rickettsial infections
  + Trypanosomiasis
* Drug-Induced Nephrotoxicity: From antimalarial drugs or other medications.
* Traditional Herbal Medicine Toxicity: Common in endemic areas and can cause AKI.
* Obstructive Uropathy: May coexist or mimic clinical features.

**Epidemiology of Malarial Nephropathy:**

* Malarial nephropathy primarily occurs in Plasmodium falciparum and Plasmodium malariae infections, which are prevalent in tropical and subtropical regions such as Sub-Saharan Africa, Southeast Asia, and India[1](https://pubmed.ncbi.nlm.nih.gov/12563598/)[2](https://pmc.ncbi.nlm.nih.gov/articles/PMC5626226/)[5](https://en.wikipedia.org/wiki/Malarial_nephropathy).
* Malaria remains one of the most common endemic infectious diseases worldwide, with an estimated 200 to 300 million new cases annually and 200,000 to 600,000 deaths
* Kidney involvement in malaria is seen in a small but significant proportion of cases. Acute kidney injury (AKI) develops in approximately 4.8% or fewer of all malaria cases, but among severe falciparum malaria cases, the incidence of AKI can be as high as 26% at 48 hours and 18% at 7 days post-admission in some studies[2](https://pmc.ncbi.nlm.nih.gov/articles/PMC5626226/)[6](https://onlinelibrary.wiley.com/doi/10.1155/2019/4396108).
* Malarial nephropathy carries a high mortality risk (15-45%) in affected patients, especially when diagnosis or treatment is delayed.
* Risk factors for malarial nephropathy include high parasitemia, severe anemia, oliguria, low blood pressure, jaundice, and coexisting conditions such as diarrhea, hepatitis, or respiratory distress
* In endemic regions like Brazil, malaria incidence has decreased over recent years, but kidney complications remain a concern in severe cases
* Children are particularly affected by nephrotic syndrome secondary to malaria, especially with Plasmodium malariae infection, which can lead to chronic glomerulonephritis and progressive kidney disease

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## **APOL1-Mediated Kidney Disease (AMKD)**

AMKD is a type of kidney disease caused by variants (changes) in the *APOL1* (apolipoprotein L1) gene. Certain *APOL1* variants have been linked with different types of kidney disease, including a higher risk of high blood pressure-related chronic kidney disease (CKD) or [kidney failure](https://www.kidney.org/atoz/content/KidneyFailure), focal segmental glomerulosclerosis (FSGS), and HIV-associated nephropathy (HIVAN). Variants linked with disease are known as risk variants, such as *APOL1* risk variants.

Certain APOL1 risk variants have been linked with a higher risk of kidney disease in people of Western and Central African descent. Other [factors](https://www.kidney.org/atoz/content/social-determinants-health-and-chronic-kidney-disease) can also increase kidney disease risk.

## **Causes**

We each inherit two copies of the *APOL1* gene, one from our mother and one from our father. The gene codes (or contains instructions) for making a protein involved in the immune system.

It is believed that *APOL1* variants arose as protection from certain parasites in Western and Central Africa thousands of years ago, but some variants can also raise the risk of kidney disease. A similar example are variants related to the sickle cell trait and sickle cell disease; these variants protect from malarial infection.

People more likely to have an *APOL1* risk variant can include people who identify as Black/African American, Afro-Caribbean, and/or Hispanic/Latino. People with two copies of an *APOL1* risk variant have about a 15-20% chance of developing kidney disease in their lifetime. About 80% of people with two copies of an *APOL1* risk variant do not get kidney disease. Therefore, not everyone with two copies of an *APOL1* risk variant will get kidney disease. Also, people without *APOL1* risk variants can still get other types of kidney disease.

Kidney disease can be a result of multiple [physical](https://www.kidney.org/atoz/content/about-chronic-kidney-disease#causes), [environmental, and social factors](https://www.kidney.org/atoz/content/social-determinants-health-and-chronic-kidney-disease).

## **Signs and symptoms**

Many people living with AMKD do not have any symptoms until the more advanced stages of kidney disease and/or complications develop. Also, the symptoms you may have will depend on the type of kidney disease present.

Symptoms related to CKD may include:

* Foamy urine
* Urinating (peeing) more often or less often than usual
* Itchy and/or dry skin
* Feeling tired
* Nausea
* Loss of appetite
* Weight loss without trying to lose weight

Symptoms related to FSGS or HIVAN can include:

* Swelling in your legs, ankles and/or around your eyes (called edema)
* Weight gain due to extra fluid (water) build-up in your body
* Foamy urine
* High cholesterol (fat) levels in the blood
* Low levels of albumin (protein) in the blood

## **Tests**

### **Genetic Testing**

Genetic testing is the only way to find out if you have *APOL1* risk variants (G1, G2) that are linked with a higher risk for AMKD. *APOL1* gene testing can be ordered by a physician or genetic counselor. A genetic counselor is a healthcare professional with special training in genetics and genetic diseases. They can help answer questions about the test and its results.

The decision to have a genetic test is made in consultation with a healthcare professional. Possible reasons to have a genetic test include helping diagnose AMKD, checking if a person is at higher risk for AMKD, or to find out if family members are at risk for AMKD. Genetic testing can be considered for living kidney donor candidates depending on the individual situation and any decisions should include a discussion with your healthcare team.

### **General Tests for Kidney Disease**

Urine and blood tests can check for signs of kidney disease. Estimated glomerular filtration rate (eGFR) is a blood test that checks how well the kidneys are filtering your blood.

A urinary albumin-to-creatinine ratio (uACR) is a urine test that checks for high protein (albumin) in the urine, which is a sign of kidney damage.

Both tests are needed to have a clear picture of your kidney health. Having an eGFR under 60 and/or a uACR over 30 for three months or more is a sign you may have kidney disease.

A urine protein-to-creatinine ratio (uPCR) may also be used for certain kidney diseases. This test is similar to the uACR test, which measures albumin. Instead of measuring only the amount of albumin in your urine, it measures all the different proteins that may be present. A uPCR level of 150 mg/g or more can be a sign of proteinuria.

Other [tests](https://www.kidney.org/atoz/content/tests-to-check-your-kidney-health) (such as a kidney biopsy, ultrasound, or CT scan) may be used if more information is needed for a diagnosis.

You may also need additional [tests](https://www.kidney.org/atoz/content/understanding-your-lab-values) to monitor other conditions related to kidney disease.

## **Treatment**

Your healthcare team will work with you to create a treatment plan to help prevent kidney disease or keep it from getting worse. Certain nutrition and lifestyle recommendations and medicines may be involved. Be sure to keep up with medical visits.

There are no treatments specifically targeting AMKD. Most treatments are either for general health or are standard care for [chronic kidney disease](https://www.kidney.org/atoz/content/about-chronic-kidney-disease).

Studies on new treatments for AMKD and related kidney diseases are ongoing. You can speak with your healthcare team for more information on clinical trials and if it would be right for you.

### **Medications**

Your healthcare professional may prescribe one or more medicines to help slow down your kidney disease. Depending on the type of kidney disease, these medicines can include an [ACE inhibitor/ARB](https://www.kidney.org/atoz/content/angiotensin-converting-enzyme-ace-inhibitors-angiotensin-receptor-blockers-arbs), diuretic (water pill), an SGLT2 inhibitor and/or an [nsMRA](https://www.kidney.org/atoz/content/non-steroidal-mineralocorticoid-receptor-antagonists-nsmras). Your healthcare professional may also prescribe a statin (cholesterol medicine).

You may also need to take additional medications or supplements to manage any CKD complications you might have (if applicable).

### **Nutrition**

### Nutrition and eating healthy are important parts of your health. Eating healthy generally includes having more fruits and vegetables, and eating foods that are less processed and as close to fresh as possible. A person with kidney disease may have their diet change over time.

You may need to limit your sodium ([salt](https://www.kidney.org/sites/default/files/02-10-0412_EBB_Sodium.pdf)) intake, especially if you have high blood pressure. You may also be asked to limit protein. Nutrition and eating healthy can be a challenge for anyone, especially if you have kidney disease. A dietitian can help with a meal plan that’s right for you.

### **Exercise**

### Exercise and physical activity, along with nutrition, are important parts of your health. The Centers for Disease Control and Prevention (CDC) recommends 150 minutes of physical activity per week, which can be done in any interval – spread it out. Don't confuse physical activity with vigorous exercise. Any type of body movement helps including walking, gardening, dancing or doing chores. The key is to find something that you enjoy and works best for you.

## **Complications**

AMKD can lead to other health problems or complications. AMKD can lead to rapidly progressive kidney disease. As AMKD worsens, the risk of getting complications goes up. Complications related to AMKD can include:

* Cardiovascular disease (heart disease and/or stroke)
* High blood pressure
* Kidney failure

It is important to get regular check-ups to monitor these complications.

**PROGNOSIS**

The prognosis for patients with APOL1-mediated kidney disease is generally poor, with the APOL1 risk variants associated with faster progression to ESKD. The histological pattern of kidney injury is characterized by scarring and damage to the glomeruli. Clinically, it presents as proteinuria, which can be mild or severe and may or may not be associated with nephrotic syndrome and progressive loss of kidney function.

**Epidemiology**

APOL1-mediated kidney disease disproportionately affects people of African ancestry due to the higher prevalence of APOL1 risk variants (G1 and G2) in this population. These variants are rare or virtually absent in individuals of non-African descent. It is estimated that about 13% of African Americans carry two APOL1 risk alleles, putting them at a significantly higher risk for developing kidney disease compared to those without these variants. Global data suggests a much higher frequency of the high-risk APOL1 variants in West Africa (mainly Ghana and Nigeria), where approximately 25% of the population carries two APOL1 risk alleles. Other geographic regions on the African continent do not reflect a similarly high prevalence. Similar disparities have been found in Hispanic, Latino, and Afro-Caribbean communities.

The discovery of the APOL1 risk alleles has provided insight into the increased incidence of kidney disease in African American populations, who experience kidney failure at rates three to four times higher than white Americans. This disparity underscores the need for targeted research and tailored therapeutic approaches.

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Glomerulosclerosis Diabetic

**Nodular Glomerulosclerosis** is a kidney condition characterized by the formation of nodules in the glomeruli, which are tiny filtering units within the kidneys. This condition is often associated with diabetes and can lead to kidney dysfunction over time. The nodules are made up of extracellular matrix material, which can interfere with the kidney's ability to filter blood effectively.

Patients with Nodular Glomerulosclerosis may present with symptoms related to kidney dysfunction. Common symptoms include swelling in the legs and feet (edema), high blood pressure, and proteinuria, which is the presence of excess protein in the urine. In some cases, patients may not exhibit noticeable symptoms until the disease has progressed significantly.

## **DIAGNOSIS**

The diagnosis of Nodular Glomerulosclerosis typically involves a combination of clinical evaluation, laboratory tests, and imaging studies. Blood tests may reveal elevated levels of creatinine and urea, indicating impaired kidney function. Urinalysis can show proteinuria and sometimes hematuria (blood in the urine). A kidney biopsy is often necessary to confirm the diagnosis, as it allows for direct examination of the glomeruli under a microscope.

## **Treatment**

Treatment for Nodular Glomerulosclerosis focuses on managing symptoms and slowing disease progression. This often includes controlling blood pressure with medications such as ACE inhibitors or angiotensin receptor blockers. Managing blood sugar levels is crucial for patients with diabetes. In advanced cases, dialysis or kidney transplantation may be necessary.

## **Prognosis**

The prognosis for patients with Nodular Glomerulosclerosis varies depending on the underlying cause and the stage at which the disease is diagnosed. Early detection and management can slow the progression of kidney damage. However, if left untreated, the condition can lead to chronic kidney disease and eventually end-stage renal disease, requiring dialysis or transplantation.

## **CAUSES**

Nodular Glomerulosclerosis is most commonly associated with diabetes mellitus, particularly long-standing or poorly controlled diabetes. It can also occur in association with other conditions such as hypertension and certain genetic disorders. The exact mechanism by which these conditions lead to nodular formation in the glomeruli is not fully understood.

## **Epidemiology**

The prevalence of Nodular Glomerulosclerosis is closely linked to the prevalence of diabetes, as it is a common complication of diabetic nephropathy. It is more frequently observed in individuals with type 2 diabetes and is a significant cause of kidney-related morbidity and mortality in this population.

## **Pathophysiology**

The pathophysiology of Nodular Glomerulosclerosis involves the accumulation of extracellular matrix material in the glomeruli, leading to the formation of nodules. This process is thought to be driven by chronic hyperglycemia (high blood sugar levels) and hypertension, which cause damage to the glomerular capillaries and stimulate the production of matrix proteins.

## **Prevention**

Preventing Nodular Glomerulosclerosis primarily involves managing risk factors such as diabetes and hypertension. This includes maintaining good blood sugar control, adhering to a healthy diet, engaging in regular physical activity, and taking prescribed medications to control blood pressure. Regular monitoring of kidney function in at-risk individuals is also important.

**DIFFERENTIAL DIAGNOSIS**

* Chronic MPGN
* Dysproteinemias such as amyloidosis or monoclonal Ig deposition disease
* Organized glomerular deposition diseases such as immunotactoid GN, fibrillary GN, or fibronectin glomerulopathy
* Idiopathic nodular glomerulosclerosis (smoking and hypertension-related)
* Glomerular changes secondary to gout (somewhat controversial)