

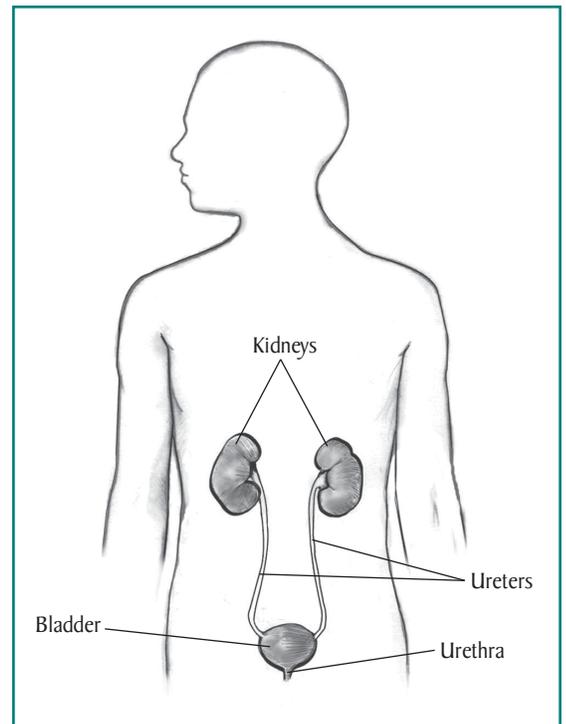
Amyloidosis and Kidney Disease

What is amyloidosis?

Amyloidosis is a rare disease that occurs when amyloid proteins are deposited in tissues and organs. Amyloid proteins are abnormal proteins that the body cannot break down and recycle, as it does with normal proteins. When amyloid proteins clump together, they form amyloid deposits. The buildup of these deposits damages a person's organs and tissues. Amyloidosis can affect different organs and tissues in different people and can affect more than one organ at the same time. Amyloidosis most frequently affects the kidneys, heart, nervous system, liver, and digestive tract. The symptoms and severity of amyloidosis depend on the organs and tissues affected.

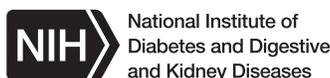
What are the kidneys and what do they do?

The kidneys are two bean-shaped organs, each about the size of a fist. They are located just below the rib cage, one on each side of the spine. Every day, the two kidneys filter about 120 to 150 quarts of blood to produce about 1 to 2 quarts of urine, composed of wastes and extra fluid. The urine flows from the kidneys to the



The kidneys are two bean-shaped organs, each about the size of a fist.

bladder through tubes called ureters. The bladder stores urine. When the bladder empties, urine flows out of the body through a tube called the urethra, located at the bottom of the bladder. In men, the urethra is long, while in women it is short.



What types of amyloidosis affect the kidneys?

Primary amyloidosis and dialysis-related amyloidosis are the types of amyloidosis that can affect the kidneys.

Primary Amyloidosis of the Kidneys

The kidneys are the organs most commonly affected by primary amyloidosis. Amyloid deposits damage the kidneys and make it harder for them to filter wastes and break down proteins. When the kidneys become too damaged, they may no longer be able to function well enough to maintain health, resulting in kidney failure. Kidney failure can lead to problems such as high blood pressure, bone disease, and anemia—a condition in which the body has fewer red blood cells than normal.

Dialysis-related Amyloidosis

People who suffer from kidney failure and have been on long-term dialysis may develop dialysis-related amyloidosis. This type of amyloidosis occurs when a certain protein, called beta-2 microglobulin, builds up in the blood because dialysis does not remove it completely. The two types of dialysis are

- **hemodialysis.** Hemodialysis uses a special filter called a dialyzer to remove wastes and extra fluid from the blood.
- **peritoneal dialysis.** Peritoneal dialysis uses the lining of the abdominal cavity—the space in the body that holds organs such as the stomach, intestines, and liver—to filter the blood.

Dialysis-related amyloidosis is a complication of kidney failure because neither hemodialysis nor peritoneal dialysis effectively filters beta-2 microglobulin from the blood. As a result, elevated

amounts of beta-2 microglobulin remain in the blood. Dialysis-related amyloidosis is relatively common in people with kidney failure, especially adults older than 60 years of age, who have been on dialysis for more than 5 years.¹

Read more in these publications at www.kidney.niddk.nih.gov:

- *Treatment Methods for Kidney Failure: Hemodialysis*
- *Treatment Methods for Kidney Failure: Peritoneal Dialysis*

What are the signs and symptoms of primary amyloidosis of the kidneys?

The most common sign of primary amyloidosis of the kidneys is nephrotic syndrome—a collection of signs that indicate kidney damage. The signs of nephrotic syndrome include

- albuminuria—an increased amount of albumin, a protein, in the urine. A person with nephrotic syndrome excretes more than half a teaspoon of albumin per day.
- hyperlipidemia—a condition in which a person's blood has more-than-normal amounts of fats and cholesterol.
- edema—swelling, typically in a person's legs, feet, or ankles and less often in the hands or face.
- hypoalbuminemia—a condition in which a person's blood has less-than-normal amounts of albumin.

Read more in *Nephrotic Syndrome in Adults* at www.kidney.niddk.nih.gov.

¹Amyloidosis. Mayo Clinic website. www.mayoclinic.com/health/amyloidosis/DS00431. Updated August 6, 2011. Accessed July 25, 2014.

Other signs and symptoms of primary amyloidosis may include

- fatigue, or feeling tired
- shortness of breath
- low blood pressure
- numbness, tingling, or a burning sensation in the hands or feet
- weight loss

What are the symptoms of dialysis-related amyloidosis?

The symptoms of dialysis-related amyloidosis may include

- pain, stiffness, and fluid in the joints.
- abnormal, fluid-containing sacs, called cysts, in some bones.
- carpal tunnel syndrome, caused by unusual buildup of amyloid proteins in the wrists. The symptoms of carpal tunnel syndrome include numbness or tingling, sometimes associated with muscle weakness, in the fingers and hands.

Dialysis-related amyloidosis most often affects bones, joints, and the tissues that connect muscle to bone, called tendons. The disease may also affect the digestive tract and organs such as the heart and lungs. Bone cysts caused by dialysis-related amyloidosis can lead to bone fractures. Dialysis-related amyloidosis can also cause tears in tendons and ligaments. Ligaments are tissues that connect bones to other bones.

How is primary amyloidosis of the kidneys diagnosed?

A health care provider diagnoses primary amyloidosis of the kidneys with

- a medical and family history
- a physical exam
- urinalysis
- blood tests
- a kidney biopsy

Medical and Family History

Taking a medical and family history may help a health care provider diagnose amyloidosis of the kidneys. He or she will ask the patient to provide a medical and family history.

Physical Exam

A physical exam may help diagnose primary amyloidosis of the kidneys. During a physical exam, a health care provider usually

- examines a patient's body to check for swelling
- uses a stethoscope to listen to the lungs
- taps on specific areas of the patient's body

Urinalysis

A health care provider may use urinalysis—the testing of a urine sample—to check for albumin and amyloid proteins in urine. The patient provides a urine sample in a special container at a health care provider's office or a commercial facility. A nurse or technician can test the sample in the same location or send it to a lab for analysis. More-than-normal amounts of albumin in urine may indicate kidney damage due to primary amyloidosis. Amyloid proteins in urine may indicate amyloidosis.

Blood Tests

The health care provider may use blood tests to see how well the kidneys are working and to check for amyloid proteins and hyperlipidemia. A blood test involves drawing a patient's blood at a health care provider's office or a commercial facility and sending the sample to a lab for analysis. Blood tests for kidney function measure the waste products in the blood that healthy kidneys normally filter out. Hyperlipidemia may indicate nephrotic syndrome. Amyloid proteins in blood may indicate amyloidosis.

Kidney Biopsy

Only a biopsy can show the amyloid protein deposits in the kidneys. A health care provider may recommend a kidney biopsy if other tests show kidney damage. A kidney biopsy is a procedure that involves taking a piece of kidney tissue for examination with a microscope. A health care provider performs a kidney biopsy in a hospital with light sedation and local anesthetic. The health care provider uses imaging techniques such as ultrasound or a computerized tomography (CT) scan to guide the biopsy needle into the kidney and take the tissue sample. A pathologist—a doctor who specializes in diagnosing diseases—examines the tissue in a lab for amyloid proteins and kidney damage.

The biopsy results can help the health care provider determine the best course of treatment. Read more in *Kidney Biopsy* at www.kidney.niddk.nih.gov.

How is dialysis-related amyloidosis diagnosed?

A health care provider diagnoses dialysis-related amyloidosis with

- urinalysis
- blood tests
- imaging tests

A health care provider can use urinalysis and blood tests to detect the amount of amyloid proteins in urine and blood. Imaging tests, such as x-rays and CT scans, can provide pictures of bone cysts and amyloid deposits in bones, joints, tendons, and ligaments. An x-ray technician performs imaging tests in a health care provider's office, an outpatient center, or a hospital. A radiologist—a doctor who specializes in medical imaging—interprets the images. A patient does not require anesthesia.



X-ray image showing amyloid deposits in the wrist

How is primary amyloidosis of the kidneys treated?

A health care provider treats primary amyloidosis of the kidneys with the following:

- medication therapy, including chemotherapy
- a stem cell transplant
- treating other conditions

Medication therapy. The goal of medication therapy, including chemotherapy, is to reduce amyloid protein levels in the blood. Many health care providers recommend combination medication therapy such as

- melphalan (Alkeran), a type of chemotherapy
- dexamethasone (Decadron), an anti-inflammatory steroid medication

These medications can stop the growth of the cells that make amyloid proteins. These medications may cause hair loss and serious side effects, such as nausea, vomiting, and fatigue.

Stem cell transplant. A stem cell transplant is a procedure that replaces a patient's damaged stem cells with healthy ones. Stem cells are found in the bone marrow and develop into three types of blood cells the body needs. To prepare for a stem cell transplant, the patient receives high doses of chemotherapy. The actual transplant is like a blood transfusion. The transplanted stem cells travel to the bone marrow to make healthy new blood cells. The chemotherapy a patient receives to prepare for the transplant can have serious side effects, so it is important to talk with the health care provider about the risks of this procedure.

Read more in *What Is a Blood and Marrow Stem Cell Transplant?* at www.nhlbi.nih.gov/health/health-topics/topics/bmsct.

Treating other conditions. Primary amyloidosis has no cure, so treating some of the side effects and other conditions seen with the disease is essential. Other conditions may include

- anemia—treatment may include medications
- depression—treatment may include talking with a mental health counselor and taking medications
- fatigue—treatment may include changes in diet and activity level
- kidney disease—treatment may include medications to help maintain kidney function or slow the progression of kidney disease

A patient and his or her family should talk with the health care provider about resources for support and treatment options.

Read more about kidney disease at www.kidney.niddk.nih.gov.

How is dialysis-related amyloidosis treated?

A health care provider treats dialysis-related amyloidosis with

- medication therapy
- newer, more effective hemodialysis filters
- surgery
- a kidney transplant

The goal of medication therapy and the use of newer, more effective hemodialysis filters is to reduce amyloid protein levels in the blood. Medication therapy can help reduce symptoms such as pain and inflammation. A health care provider may treat a person with dialysis-related amyloidosis who has bone, joint, and tendon problems, such as bone cysts and carpal tunnel syndrome, using surgery.

Dialysis-related amyloidosis has no cure; however, a successful kidney transplant may stop the disease from progressing.

Read more in *Treatment Methods for Kidney Failure: Transplantation* at www.kidney.niddk.nih.gov.

Eating, Diet, and Nutrition

Researchers have not found that eating, diet, and nutrition play a role in causing or preventing primary amyloidosis of the kidneys or dialysis-related amyloidosis. People with nephrotic syndrome may make dietary changes such as

- limiting dietary sodium, often from salt, to help reduce edema and lower blood pressure
- decreasing liquid intake to help reduce edema and lower blood pressure
- eating a diet low in saturated fat and cholesterol to help control more-than-normal amounts of fats and cholesterol in the blood

Health care providers may recommend that people with kidney disease eat moderate or reduced amounts of protein. Proteins break down into waste products that the kidneys filter from the blood. Eating more protein than the body needs may burden the kidneys and cause kidney function to decline faster. However, protein intake that is too low may lead to malnutrition, a condition that occurs when the body does not get enough nutrients.

People with kidney disease on a restricted protein diet should receive blood tests that can show low nutrient levels. People with primary amyloidosis of the kidneys or dialysis-related amyloidosis should talk with a health care provider about dietary restrictions to best manage their individual needs.

Points to Remember

- Amyloidosis is a rare disease that occurs when amyloid proteins are deposited in tissues and organs.
- Primary amyloidosis and dialysis-related amyloidosis are the types of amyloidosis that can affect the kidneys.
- The most common sign of primary amyloidosis of the kidneys is nephrotic syndrome.
- The signs of nephrotic syndrome include
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 - a kidney transplant

Hope through Research

The National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) conducts and supports a variety of research in kidney diseases and urinary tract disorders. The knowledge gained from these studies is advancing scientific understanding of why kidney diseases and urinary tract disorders develop, leading to improved methods of diagnosing, treating, and preventing them.

Clinical trials are research studies involving people. Clinical trials look at safe and effective new ways to prevent, detect, or treat disease. Researchers also use clinical trials to look at other aspects of care, such as improving the quality of life for people with chronic illnesses. To learn more about clinical trials, why they matter, and how to participate, visit the NIH Clinical Research Trials and You website at www.nih.gov/health/clinicaltrials. For information about current studies, visit www.ClinicalTrials.gov.

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About the Kidney Failure Series

The NIDDK Kidney Failure Series includes booklets and fact sheets that can help the reader learn more about treatment methods for kidney failure, complications of dialysis, financial help for the treatment of kidney failure, and eating right on hemodialysis. Free single printed copies of this series can be obtained by contacting the National Kidney and Urologic Diseases Information Clearinghouse.

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The National Kidney Disease Education Program (NKDEP) is an initiative of the National Institute of Diabetes and Digestive and Kidney Diseases, National Institutes of Health, U.S. Department of Health and Human Services. The NKDEP aims to raise awareness of the seriousness of kidney disease, the importance of testing those at high risk, and the availability of treatment to prevent or slow kidney disease.

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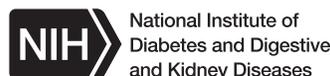
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You may also find additional information about this topic by visiting MedlinePlus at www.medlineplus.gov.

This publication may contain information about medications and, when taken as prescribed, the conditions they treat. When prepared, this publication included the most current information available. For updates or for questions about any medications, contact the U.S. Food and Drug Administration toll-free at 1-888-INFO-FDA (1-888-463-6332) or visit www.fda.gov. Consult your health care provider for more information.



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