What is kidney dysplasia?

Kidney dysplasia is a condition in which the internal structures of one or both of a fetus' kidneys do not develop normally while in the womb. During normal development, two thin tubes of muscle called ureters grow into the kidneys and branch out to form a network of tiny structures called tubules. The tubules collect urine as the fetus grows in the womb. In kidney dysplasia, the tubules fail to branch out completely. Urine that would normally flow through the tubules has nowhere to go. Urine collects inside the affected kidney and forms fluid-filled sacs called cysts. The cysts replace normal kidney tissue and prevent the kidney from functioning.

Kidney dysplasia can affect one kidney or both kidneys. Babies with severe kidney dysplasia affecting both kidneys generally do not survive birth. Those who do survive may need the following early in life:

- blood-filtering treatments called dialysis
- a kidney transplant

Children with dysplasia in only one kidney have normal kidney function if the other kidney is unaffected. Those with mild dysplasia of both kidneys may not need dialysis or a kidney transplant for several years.

Kidney dysplasia is also called renal dysplasia or multicystic dysplastic kidney.
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, which is composed of wastes and extra fluid. Children produce less urine than adults—the amount they produce depends on their age. The urine flows from the kidneys to the bladder through the two ureters, one on each side of the bladder. The bladder stores urine. The muscles of the bladder wall remain relaxed while the bladder fills with urine. As the bladder fills to capacity, signals sent to the brain tell a person to find a toilet soon. When the bladder empties, urine flows out of the body through a tube called the urethra, located at the bottom of the bladder.
The kidneys, ureters, bladder, and urethra are parts of the urinary tract. Read more about the kidneys and the urinary tract at www.kidney.niddk.nih.gov.

What causes kidney dysplasia?
Genetic factors can cause kidney dysplasia. Genes pass information from both parents to the child and determine the child’s traits. Sometimes, parents may pass a gene that has changed, or mutated, causing kidney dysplasia.

Genetic syndromes that affect multiple body systems can also cause kidney dysplasia. A syndrome is a group of symptoms or conditions that may seem unrelated yet are thought to have the same genetic cause. A baby with kidney dysplasia due to a genetic syndrome might also have problems of the digestive tract, nervous system, heart and blood vessels, muscles and skeleton, or other parts of the urinary tract.

A baby may also develop kidney dysplasia if his or her mother takes certain prescription medications during pregnancy, such as some used to treat seizures and high blood pressure. A mother’s use of illegal drugs, such as cocaine, during pregnancy may also cause kidney dysplasia in her unborn child.

How common is kidney dysplasia?
Kidney dysplasia is a common condition. Scientists estimate that kidney dysplasia affects about one in 4,000 babies.1 This estimate may be low because some people with kidney dysplasia are never diagnosed with the condition. About half of the babies diagnosed with this condition have other urinary tract defects.2

Who is more likely to develop kidney dysplasia?
Babies who are more likely to develop kidney dysplasia include those

• whose parents have the genetic traits for the condition
• with certain genetic syndromes affecting multiple body systems
• whose mothers used certain prescription medications or illegal drugs during pregnancy

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What are the signs of kidney dysplasia?

Many babies with kidney dysplasia in only one kidney have no signs of the condition. In some cases, the affected kidney may be enlarged at birth and may cause pain.

What are the complications of kidney dysplasia?

The complications of kidney dysplasia can include:

- hydronephrosis of the working kidney. A baby with kidney dysplasia in only one kidney might have other urinary tract defects. When other defects in the urinary tract block the flow of urine, the urine backs up and causes the kidneys and ureters to swell, a condition called hydronephrosis. If left untreated, hydronephrosis can damage the working kidney and reduce its ability to filter blood. Kidney damage may lead to chronic kidney disease (CKD) and kidney failure.

- a urinary tract infection (UTI). A urine blockage may increase a baby’s chance of developing a UTI. Recurring UTIs can also lead to kidney damage.

- high blood pressure.

- a slightly increased chance of developing kidney cancer.

How is kidney dysplasia diagnosed?

Health care providers may be able to diagnose kidney dysplasia during a woman’s pregnancy using a fetal ultrasound, also called a fetal sonogram. Ultrasound uses a device, called a transducer, that bounces safe, painless sound waves off organs to create an image of their structure. Fetal ultrasound is a test done during pregnancy to create images of the fetus in the womb. A specially trained technician performs the procedure in a health care provider’s office, an outpatient center, or a hospital, and an obstetrician or a radiologist interprets the images. An obstetrician is a doctor who specializes in pregnancy and childbirth. A radiologist is a doctor who specializes in medical imaging. The patient—in this case, the fetus’ mother—does not need anesthesia for this procedure. The images can show defects in the fetus’ kidneys and other parts of the urinary tract.

Health care providers do not always diagnose kidney dysplasia before a baby is born. After birth, health care providers often diagnose kidney dysplasia during an evaluation of the child for a UTI or another medical condition. A health care provider uses ultrasound to diagnose kidney dysplasia after the baby is born.

How is kidney dysplasia treated?

If the condition is limited to one kidney and the baby has no signs of kidney dysplasia, no treatment may be necessary. However, the baby should have regular checkups that include:

- checking blood pressure.

- testing blood to measure kidney function.

- testing urine for albumin, a protein most often found in blood. Albumin in the urine may be a sign of kidney damage.

- performing periodic ultrasounds to monitor the damaged kidney and to make sure the functioning kidney continues to grow and remains healthy.
How can kidney dysplasia be prevented?
Researchers have not found a way to prevent kidney dysplasia caused by genetic factors or certain genetic syndromes. Pregnant women can prevent kidney dysplasia by avoiding the use of certain prescription medications or illegal drugs during pregnancy. Pregnant women should talk with their health care provider before taking any medications during pregnancy.

What is the long-term outlook for a child with kidney dysplasia in only one kidney?
The long-term outlook for a child with kidney dysplasia in only one kidney is generally good. A person with one working kidney, a condition called solitary kidney, can grow normally and may have few, if any, health problems.

The affected kidney may shrink as the child grows. By age 10, the affected kidney may no longer be visible on x-ray or ultrasound. Children and adults with only one working kidney should have regular checkups to test for high blood pressure and kidney damage. A child with urinary tract problems that lead to failure of the working kidney may eventually need dialysis or a kidney transplant.


What is the long-term outlook for a child with kidney dysplasia in both kidneys?
The long-term outlook for a child with kidney dysplasia in both kidneys is different from the long-term outlook for a child with one dysplastic kidney. A child with kidney dysplasia in both kidneys

• is more likely to develop CKD.

• needs close follow-up with a pediatric nephrologist—a doctor who specializes in caring for children with kidney disease. Children who live in areas that don’t have a pediatric nephrologist available can see a nephrologist who cares for both children and adults.

• may eventually need dialysis or a kidney transplant.

Eating, Diet, and Nutrition
Researchers have not found that eating, diet, and nutrition play a role in causing or preventing kidney dysplasia.

Kidney Dysplasia

Points to Remember

• Kidney dysplasia is a condition in which the internal structures of one or both of a fetus’ kidneys do not develop normally while in the womb.

• Genetic factors can cause kidney dysplasia.

• Genetic syndromes that affect multiple body systems can also cause kidney dysplasia.

• A baby may also develop kidney dysplasia if his or her mother takes certain prescription medications during pregnancy, such as some used to treat seizures and high blood pressure.

• Many babies with kidney dysplasia in only one kidney have no signs of the condition.

• Health care providers may be able to diagnose kidney dysplasia during a woman’s pregnancy using a fetal ultrasound, also called a fetal sonogram.

• Health care providers do not always diagnose kidney dysplasia before a baby is born.

• If the condition is limited to one kidney and the baby has no signs of kidney dysplasia, no treatment may be necessary.

• Researchers have not found a way to prevent kidney dysplasia caused by genetic factors or certain genetic syndromes.

• Pregnant women can prevent kidney dysplasia by avoiding the use of certain prescription medications or illegal drugs during pregnancy.

• The long-term outlook for a child with kidney dysplasia in only one kidney is generally good.

• The long-term outlook for a child with kidney dysplasia in both kidneys is different from the long-term outlook for a child with one dysplastic kidney. A child with kidney dysplasia in both kidneys:
  – is more likely to develop chronic kidney disease (CKD)
  – needs close follow-up with a pediatric nephrologist
  – may eventually need dialysis or a kidney transplant

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  – needs close follow-up with a pediatric nephrologist
  – may eventually need dialysis or a kidney transplant
Hope through Research

The National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) conducts and supports basic and clinical research aimed at understanding kidney and urologic disorders, including urinary tract abnormalities, UTIs, CKD, and kidney failure. Through its Pediatric Urology Program, the NIDDK funds research into bladder and urinary tract development. For example, NIDDK-funded researchers are conducting studies on the genetics of kidney and urologic development and defects. The NIDDK also funds the GenitoUrinary Development Molecular Anatomy Project (GUDMAP), a scientific and medical consortium working to develop a public database to facilitate urologic research. The database’s key feature is a molecular atlas of gene expression in developing organs in the genitourinary tract.

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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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.
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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