Renal dysplasia



This infoKID topic is for parents and carers about children's kidney conditions. Visit www.infoKID.org.uk to find more topics about conditions, tests & diagnosis, treatments and supporting information.

Each topic starts with an overview followed by several sections with more information. Links to sections in topic | Other topics available on website

Renal dysplasia (or kidney dysplasia) means that a kidney does not fully develop in the womb. The affected kidney does not have normal function – which means that it does not work as well as a normal kidney. It is usually smaller than usual, and may have some cysts, which are like sacs filled with liquid.

Renal dysplasia may be picked up before birth on the 20 week antenatal ultrasound scan, or soon after birth. It may also be picked up in an older child who has some symptoms.

An affected kidney is called a dysplastic kidney. Renal dysplasia rarely causes any problems during the pregnancy or in childbirth.

- If it happens in one kidney, it is called unilateral renal dysplasia. Although one of their kidneys will not work properly, most children have no serious problems, but will need monitoring for possible long-term effects.
- If it happens in both kidneys, it is called bilateral renal dysplasia. This is more serious, and will need follow-up during the pregnancy to find out how the baby is affected.



Overview

About the urinary system

The **urinary system** gets rid of things that the body no longer needs, so that we can grow and stay healthy.

The **kidneys** are bean-shaped organs. They filter blood to remove extra water and waste in urine (wee). Most of us have two kidneys. They are on either side of our spine (backbone), near the bottom edge of our ribs at the back.

The two **ureters** are long tubes that carry urine from the kidneys to the bladder.

The **bladder** is a bag that stores urine until we are ready to urinate. It sits low down in the pelvis.

The urethra is a tube that carries urine from the bladder to the **outside** of the body.

» More about the urinary system and kidneys





Renal dysplasia | Page 1

Causes

Renal dysplasia happens when part of the kidney does not develop properly in the womb. It is relatively common

It is not always possible to know why renal dysplasia happens. In the majority of cases, it is not caused by anything that the mother does during her pregnancy, and it is unlikely that a future pregnancy will result in renal dysplasia or other problems with the kidneys.

Occasionally a specific cause is found.

» More about causes

Test and diagnosis

Antenatal ultrasound scan

The 20 week **antenatal ultrasound scan** looks at your baby growing in the womb. Renal dysplasia may be suspected if one or both of the kidneys look smaller than usual or otherwise look abnormal.

The scan *cannot* always **diagnose** (identify) the problem. Although your doctor will not always know how your baby will be affected at birth, he or she is less likely to have significant problems if:

- he or she is growing well in the womb
- no other problems have been found, and
- there is a normal amount of **amniotic fluid** (or liquor).

You may need to go back to the hospital for more ultrasound scans during pregnancy.

Diagnosis later in childhood

Sometimes, renal dysplasia is only picked up after birth or when a child is older. It is usually found during a scan that a child is having for another reason, such as after a urinary tract infection (UTI) or after an accident.

Other conditions that look like renal dysplasia

Sometimes, renal dysplasia is thought to be another type of problem with the kidney(s).

- Renal hypoplasia part of the kidney does not develop properly, and the kidney is smaller than normal.
- Multicystic dysplastic kidney (MCDK) a more severe form of renal dysplasia. The whole of the affected kidney is a bundle of many cysts and does not work.
- **Reflux nephropathy** scars on the kidney.

Other conditions associated with renal dysplasia

Sometimes, renal dysplasia can be seen with other conditions that happen in the womb.

- Antenatal hydronephrosis one or both kidneys do not drain urine properly. In renal dysplasia, this is because the urine drainage system is 'baggy' and does not empty properly. The affected kidney becomes stretched and swollen. Antenatal hydronephrosis may get better at a later stage in the pregnancy, but your doctor will check how your baby is affected.
- Vesicoureteral reflux (VUR) when babies with VUR pass urine in the womb, some urine refluxes (goes back up) towards, and sometimes into, the kidneys. This can affect the fully working kidney and/ or the dysplastic kidney.
- Posterior urethral valves (PUV) a blockage in the back part of the urethra near the bladder in some boys.

Tests after birth

After your baby is born, he or she may need some **imaging tests** (scans). These use special equipment to get images (pictures) of the inside of the body. They are used to confirm that your child has renal dysplasia and look for any complications.

Unilateral renal dysplasia

In **unilateral renal dysplasia**, one kidney does not develop properly in the womb. ('Unilateral' means one side.) Most children born with one dysplastic kidney have no complications and do not need special treatment. Sometimes the other kidney grows larger than normal to make up for the dysplastic kidney. However, they may be at risk of or **urinary tract infections (UTIs)** and/or **hypertension** (high blood pressure) later in life.

» More about unilateral renal dysplasia

Bilateral renal dysplasia

In **bilateral renal dysplasia**, both kidneys do not develop normally. ('Bilateral' means two sides.) Some babies born with two dysplastic kidneys have no immediate complications. Others need more support at birth, including ventilation to help them breathe.

All children with bilateral renal dysplasia need monitoring, as some may go on to develop **kidney failure**. This occasionally happens while a baby or child is young, but is more likely to occur later in life, especially during puberty when children's bodies grow quickly. Children need to go back to the hospital or clinic throughout childhood to check how well their kidneys are working. Treatment can be started as soon as it is needed, to help your child grow and remain healthy. Eventually the kidneys may stop working and **dialysis** may be needed.

» More about bilateral renal dysplasia



Causes

Doctors understand that there are some possible causes of renal dysplasia, though it may not always be possible to identify the cause in your baby. It is not usually caused by anything that the mother does during her pregnancy.

About the name

Renal dysplasia can be broken down:

- renal: to do with one or both kidneys
- dys: not working properly
- plasia: to do with development.

It is one type of **congenital anomaly of the kidneys and urinary tract.** 'Congenital' means that the problem is present at birth and 'anomaly' means different than normal.

How common is it?

Renal dysplasia is relatively common. It is estimated that one baby in a few hundred may be affected.

How does it happen?

Renal dysplasia happens when part of the kidney does not develop properly in the womb.

Causes

It is not always possible to explain why renal dysplasia happens, but there are a few causes.

Genetic mutations (inherited)

Most cases of renal dysplasia are *not* inherited from the baby's mother or father. However, some cases are caused by **genetic mutations**. These are problems in the **genes** (which are in each of our body's living cells), which are passed on by the parents.

If your doctor thinks your baby has a problem that is caused by genetic mutations, you may be referred for **genetic testing** and counselling. Genetic testing usually involves getting a sample of blood or body tissue, which can be checked for a specific gene. **Genetic counselling** is a service that can give you information and guidance about conditions caused by genetic mutations.

Occasionally, renal dysplasia is part of a **syndrome**, which is a collection of symptoms and signs. These children may also have other problems, such as with their digestive system, nervous system, heart and blood vessels, muscles and skeleton, or other parts of their urinary system. These may be caused by genetic mutations.

Medicines and other drugs

A few cases of renal dysplasia are caused by some medicines taken by the pregnant woman – including prescribed medicines for seizures (also called convulsions or fits) or high blood pressure (such as angiotensinconverting enzyme (ACE) inhibitors and angiotensin receptor blockers (ARBs)). If you are pregnant, speak with your doctor about the medicines that you are taking.

It may also be caused by some illicit drugs, such as cocaine.

Will it happen in future pregnancies?

Unless you have been told that your baby's renal dysplasia was caused by a genetic mutation, it is unlikely that a future pregnancy will result in renal dysplasia, or other problems with the kidneys. If one of your children has renal dysplasia, it is unlikely another one of your children or family members will get it. Speak with your doctor for more information.

Causes: Read more about how renal dysplasia happens

How renal dysplasia happens

The kidneys form while a baby is growing in the womb. Normally, two tubes, which become the ureters, grow from the bladder into tissue, which becomes the kidneys. These tubes form the **renal pelvis**, the part of the kidney that collects urine. These tubes also form long tubes that link to the **nephrons**, the many tiny parts that filter blood and make urine.

In renal dysplasia, this process does not work, so that the kidney does not grow normally. The kidney is smaller than usual and has fewer nephrons. It may also develop with small **cysts**, which are like sacs filled with fluid (liquid).

How renal dysplasia affects the kidneys and how they work

A dysplastic kidney may not be able to remove as much waste, water and salts from the body as a normal kidney. If one kidney is dysplastic and the other kidney is normal, together they can usually work well enough to keep your child healthy.





Unilateral renal dysplasia (one kidney)

In unilateral renal dysplasia, part or all of one kidney does not fully develop while a baby is growing in the womb. ('Unilateral' means one side.) It may be smaller than a normal kidney and may have cysts, which are like sacs filled with liquid.

The other kidney usually looks and works normally. Most people have no long-term problems with unilateral renal dysplasia, and some do not know that they have one kidney that is smaller than usual.

Before birth

Unilateral renal dysplasia may be suspected on the 20 week **antenatal ultrasound scan**, which looks at your baby growing in the womb. It may be suspected if one or both of the kidneys look smaller than usual or otherwise look abnormal.

The scan also measures the amount of **amniotic fluid** (or **liquor**), the fluid that your baby floats in. The baby's kidneys start making urine and pass this out into the amniotic fluid. This fluid protects your baby from getting hurt from the outside and helps his or her lungs mature so he or she is ready to breathe after birth.

If there is not enough amniotic fluid (**oligohydramnios**), this may be a sign that the kidneys are not working well, and that there may be problems breathing after birth.

You may need more ultrasound scans during the pregnancy to find out how the dysplasia is affecting your baby. It does not usually have an impact on how your baby is delivered.

Tests after birth

After your baby is born, he or she may need some **imaging tests** (scans) to confirm the condition and look for any complications. These use special equipment to get images of the inside of the body.

- Ultrasound scan looks at the shape and size of your baby's kidneys and other parts of the urinary system. A small handheld device is moved around your child's skin and uses sound waves to create an image on a screen.
- DMSA scan checks for any damage on the kidneys. A chemical that gives out a small amount of radiation is injected into one of your child's blood vessels. This chemical is taken up by healthy parts of the kidney and a special camera takes pictures.
- MAG3 scan for babies who also have antenatal hydronephrosis, this shows how much blood is going into and out of their kidneys, and whether they are passing urine normally. As in the DMSA test, a chemical that gives out a small amount of radiation is injected into a blood vessel, and a special camera takes pictures.
- Cystourethrogram or MCUG (sometimes called a VCUG) usually for babies and children who are suspected of having vesicoureteral reflux (VUR). In this condition, some urine refluxes (goes back up) the wrong way up the ureters, towards, and sometimes into, the kidney. This test can check how your baby is passing urine. A special X-ray machine takes a series of images of the bladder while your baby is passing urine.

Complications and treatment

In most cases, babies do not need treatment. A small number of children have symptoms or complications, which may not happen until later in life. These may need follow up or treatment, such as medicines.

High blood pressure

Some children develop **hypertension**, blood pressure that is too high.

If your child has hypertension, he or she will need to reduce his or blood pressure so it is in the healthy range. Your child will probably need to eat a no-added salt diet, and may need to take medicines, to control his or her blood pressure. It is also recommended that all children, especially those with hypertension, keep to a healthy body weight and exercise regularly.

Occasionally, children who have a dysplastic kidney that is not working well and have blood pressure that cannot be controlled, will need the kidney removed in an operation called a **nephrectomy**. You may be referred to a **paediatric nephrologist**, a surgeon who treats children with problems in their urinary system, to discuss the operation.

Urinary tract infections

Some children get **urinary tract infections (UTIs)**, when germs get into the urine and travel up the urinary tract (or system) and cause an infection, usually in the bladder. Babies and children with UTIs may become irritable, have a fever, have pain when they wee, feel sick or be sick.

UTIs that keep coming back are more likely in children who also have **vesicoureteral reflux (VUR)**, when some urine refluxes (goes back up) towards, and sometimes into, the kidneys.

If your child has a UTI, he or she will need to take **antibiotics**, medicines that kill the germs.

➔ If you think your child has a UTI, seek medical advice.

Chronic kidney disease

In most people with unilateral renal dysplasia, the other kidney works normally. The normal kidney can work harder to compensate and do the work of two kidneys.

Sometimes children with unilateral renal dysplasia have an abnormality in the other kidney. If the other kidney does not work normally, your child may have reduced kidney function. He or she may be at greater risk of progressing to later stages of **chronic kidney disease (CKD)**, and will need more monitoring.



Renal dysplasia | Page 4

About the future

Many children will have no long-term problems. The other kidney usually grows larger to do the work of two kidneys.

Your child should be able to do all the things that other children their age do. He or she can go to nursery and school, play with other children and stay active.

Follow up

In the first few years of his or her life, your child may need to go back to the hospital for some tests. Later, he or she may need to see your family doctor about once a year. It is important to go to these appointments even if your child seems well. You will also have the opportunity to ask any questions. At these appointments your child may have:

- his or her blood pressure measured, to check for hypertension
- urine tests to check for protein in his or her urine (proteinuria), which may be a sign of problems in the kidney. You or a nurse collect some of your child's urine in a small, clean container. A dipstick will be dipped into the urine – this is a strip with chemical pads that change colour if there is protein in the urine
- **blood tests** to measure his or her kidney function
- his or her height and weight checked in childhood, to measure how much he or she is growing.

Living healthily

Your child can help protect his or her kidneys, and reduce the risk of hypertension later in life, by leading a healthy lifestyle through their child and adult years. This includes:

- eating a healthy diet with at least five servings of fruit and vegetables a day, taking care not to eat too much salt, sugar and fats (especially saturated fats)
- getting plenty of exercise
- not smoking.

Further support

This can be a difficult and stressful experience for you and your family.

➔ If you have any concerns or need additional support, speak with your doctor or nurse.

Further information

This is the end of the information about unilateral renal dysplasia. If you would like to read more about tests and diagnosis, treatment or supporting information, you can find a list of topics covered on the infoKID website at www.infoKID.org.uk.

Notes



Bilateral renal dysplasia (both kidneys)

In bilateral renal dysplasia, both kidneys do not develop properly while a baby is growing in the womb. They are smaller than usual and may have cysts, which are like sacs filled with liquid. ("Bilateral" means two sides.)

Before birth

Bilateral renal dysplasia may be suspected on the 20 week antenatal ultrasound scan, which looks at your baby growing in the womb. It may be suspected if one or both of the kidneys look smaller than usual or otherwise look abnormal

The scan also measures the amount of **amniotic fluid** (or liquor), the liquid that surrounds your baby, and that is partly made up of the baby's urine. This fluid protects your baby from getting hurt from the outside and helps the lungs mature so he or she is ready to breathe after birth.

If there is not enough amniotic fluid (oligohydramnios), this may be a sign that the kidneys are not working well, and that the baby may have problems breathing after birth.

You may need more ultrasound scans during the pregnancy to find out how the dysplasia is affecting your baby.

Referral

If bilateral renal dysplasia is suspected, you will be referred to specialist healthcare professionals as early as possible in the pregnancy. They may include a:

- foetal medicine specialist a doctor who specialises in the health of unborn babies (foetuses)
- obstetrician a doctor who specialises in pregnancy, delivering babies and the care of women after childbirth
- paediatrician a doctor who treats babies, children and young people
- paediatric nephrologist a doctor who treats babies, children and young people with kidney problems
- neonatologist a doctor who specialises in newborn babies.

Treatment

In most cases, there is no treatment needed before birth.

Options

Very rarely, there is concern that the problems with the baby's kidneys and/or lungs are very severe, and may mean that he or she will not be able to live a normal life.

In the most severe cases these problems can be lifethreatening and the baby may have kidney failure from birth. A team of healthcare professionals experienced in delivering babies and treating newborn babies with these problems will give you support. They will discuss with you all the options, and agree a plan for the remainder of the pregnancy and the delivery.

Tests after birth

After your baby is born, he or she may need some **imaging tests** (scans). These use special equipment to get images of the inside of the body. They are used to confirm that your child has bilateral renal dysplasia and look for any complications.

• An ultrasound scan is usually done first. This looks at the shape and size of your baby's kidneys and other parts of the urinary system. A small handheld device is moved around your child's skin and uses sound waves to create an image on a screen.

If problems are found on the ultrasound scan, your doctor may recommend other tests, which may include the below.

- DMSA scan looks at the kidneys to check for any damage. A chemical that gives out a small amount of radiation is injected into one of your child's blood vessels. This chemical is taken up by healthy parts of the kidney and a special camera takes pictures.
- MAG3 scan for babies who also have antenatal hydronephrosis, this shows how much blood is going into and of their kidneys, and whether they are passing urine normally. As in the DMSA test, a chemical that gives out a small amount of radiation is injected into a blood vessel, and a special camera takes pictures.
- Cystourethrogram or MCUG (sometimes called a VCUG) usually for babies and children who are suspected of having vesicoureteral reflux (VUR). In this condition, some urine refluxes (goes back up) the wrong way up the ureters, towards, and sometimes into, the kidneys. This test can check how your baby is passing urine. A special X-ray machine takes a series of images of the bladder while your baby is passing urine.

Complications and treatment at birth

A small number of babies have symptoms and complications if the dysplasia is severe.

Support for newborn babies

If your doctors think that your baby might have problems from birth, he or she will be reviewed by healthcare professionals who are experienced in looking after newborn babies. Your baby may need to be admitted into a **neonatal unit**, a special ward of the hospital for newborn babies. The team of healthcare professionals will measure how much urine your baby is passing, and how much he or she is growing.

In rare cases, babies are not able to breathe properly after birth because their lungs have not developed properly.



This is more likely in babies who had very little **amniotic fluid** (or **liquor**) when growing in the womb.

If your baby needs help breathing, he or she can be given oxygen or ventilation, using a machine that moves breathable air in and out of his or her lungs. Many of these babies improve as they begin to grow but occasionally in very severe cases this is not the case.

Long-term complications and treatment

High blood pressure

Some children develop **hypertension**, blood pressure that is too high.

If your child has hypertension, he or she will need to reduce their blood pressure so it is in the healthy range. Your child will probably need to eat a no-added salt diet, and may need to take medicines, to control his or her blood pressure. It is also recommended that all children, especially those with hypertension, keep to a healthy body weight and exercise regularly.

Urinary tract infections

Some children get **urinary tract infections (UTIs)**, when germs get into the urine and travel up the urinary tract (or system) and cause an infection, usually in the bladder. Babies and children with UTIs may become irritable, have a fever, have pain on weeing, feel sick or be sick.

If your child has a UTI, he or she will need to take **antibiotics**, medicines that kill the germs.

➔ If you think your child has a UTI, seek medical advice.

Kidney function

Usually, even if the dysplastic kidneys are not working as normal, they are working well enough that there are no symptoms at birth. Because dysplastic kidneys may not grow normally, children will need blood tests throughout their life to measure their **kidney function** (how well their kidneys are working).

Children who have long-term problems with how their kidneys work are said to have **chronic kidney disease** (CKD). A team of healthcare professionals will make sure your child gets the right tests, treatments and monitoring as they grow, according to how well his or her kidneys are working.

Progressive CKD and kidney failure

Your child's kidney function may get worse as the kidneys cannot grow as the child's body grows. In some children, this progresses to kidney failure, when the kidneys cannot support the body.

It is very rare for a baby to be born with kidney failure. Occasionally, a young baby or child has kidney failure. It is more likely during **puberty** or the start of the teenage years, when children go through a growth spurt and the smaller kidneys are no longer able to work for the bigger body.

Babies and children in kidney failure need specialist treatment. This may include **dialysis**, which uses special equipment or a machine to remove waste products and extra water from the body, and/or a **kidney transplant**, in which a healthy kidney from another person is transplanted into a patient's body.

» Read more in the infoKID topic on chronic kidney disease (CKD)

About the future

Your child should be able to do all the things that other children their age do. He or she can go to nursery and school, play with other children and stay active.

Follow up

Your child will need to go back to the hospital or clinic for follow-up appointments throughout his or her life. It is important to go to these appointments even if your child seems well. You will also have the opportunity to ask any questions. At these appointments your child may have:

- his or her blood pressure measured, to check for hypertension
- urine tests to check for protein in his or her urine (proteinuria), which may be a sign of problems in the kidney. You or a nurse collect some of your child's urine in a small, clean container. A dipstick will be dipped into the urine – this is a strip with chemical pads that change colour if there is protein in the urine.
- blood tests to measure his or her kidney function
- his or her height and weight checked in childhood, to measure how much he or she is growing.

Further support

This can be a difficult and stressful experience for you and your family.

➔ If you have any concerns or need additional support, speak with your doctor or nurse.

Further information

This is the end of the information about bilateral renal dysplasia. If you would like to read more about tests and diagnosis, treatment or supporting information, you can find a list of topics covered on the infoKID website at www.infoKID.org.uk.



<u>Your notes and contact information</u>

www.infoKID.org.uk





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For details on any sources of information used in this topic, please contact us through our website www.infoKID.org.uk.

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