Renal hypoplasia



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 hypoplasia (or kidney hypoplasia) means that part of a kidney does not fully develop in the womb. The kidney may be only slightly smaller than usual or it may be tiny. Because of its size, it may not work as well as a normal-sized kidney.

Renal hypoplasia 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. It is quite a common – it is estimated that one baby in a few hundred will be born with one hypoplastic (small) kidney.







Overview

About the urinary system and urine

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

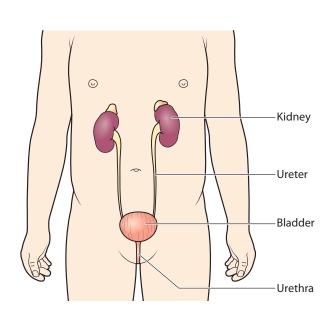
The **kidneys** are bean-shaped organs. They filter blood to remove extra water and waste in urine (wee). Most of us have two kidneys. They are on either side of our spine (backbone), near the bottom edge of our ribs at the back.

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





Causes

Renal hypoplasia is relatively common – it is estimated that one baby in a few hundred is born with a small kidney.

It is not always possible to know why renal hypoplasia 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 hypoplasia 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 hypoplasia may be suspected if one or both of the kidneys look smaller than usual but otherwise normal.

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 hypoplasia 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 a **urinary tract infection (UTI)** or after an accident.

Other conditions that look like renal hypoplasia

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

- Renal dysplasia one or both kidneys are smaller than usual, but have also not developed properly and may have cysts.
- 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 hypoplasia

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

- Antenatal hydronephrosis one or both kidneys do not drain urine properly. In renal hypoplasia, 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 hypoplastic kidney.

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 hypoplasia and look for any complications.

Unilateral renal hypoplasia

In **unilateral renal hypoplasia**, one kidney is smaller than usual. ('Unilateral' means one side.) Most babies born with one small kidney have no complications and do not need special treatment. However, they may be at risk of **urinary tract infections (UTIs)** and/or **hypertension** (high blood pressure) later in life.

Sometimes the other kidney grows larger than normal to make up for the one small kidney.

More about unilateral renal hypoplasia

Bilateral renal hypoplasia

In **bilateral renal hypoplasia**, both kidneys are smaller than usual. ('Bilateral' means two sides.) Some babies born with two small kidneys have no immediate complications. Others need more support at birth, including ventilation to help them breathe.

All children with bilateral renal hypoplasia 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** and/or a **kidney transplant** may be needed.

» More about bilateral renal hypoplasia



Causes

Doctors understand that there are some possible causes of renal hypoplasia, 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 hypoplasia can be broken down:

- renal: to do with one or both kidneys
- hypo: fewer than usual
- 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 hypoplasia is relatively common. It is estimated that one baby in a few hundred is born with a kidney that is smaller than usual.

How does it happen?

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

Causes

It is not always possible to explain why renal hypoplasia happens, However, there are rare causes that can be identified.

Genetic mutations (inherited)

Most cases of renal hypoplasia 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 hypoplasia 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 hypoplasia 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 angiotensin-converting 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 affect other family members?

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

Causes: Read more about how renal hypoplasia happens

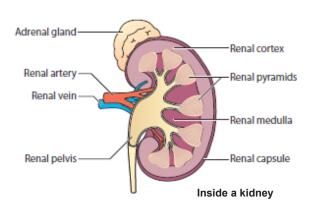
How renal hypoplasia happens

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 hypoplasia, this process does not work, so that the kidney does not grow normally. The kidney is smaller than usual and has fewer nephrons.

How renal hypoplasia affects the kidneys and how they work

A hypoplastic kidney may not be able to remove as much waste, water and salts from the body as a normal kidney. If one kidney is hypoplastic and the other kidney is normal, together they can usually work well enough for there to be no problems.





Unilateral renal hypoplasia (one kidney)

In unilateral renal hypoplasia, one kidney is smaller than usual because it has not fully developed while a baby is growing in the womb. ('Unilateral' means one side.) The other kidney usually looks and works as normal, and may grow larger to help do the work of two kidneys. Most people have no long-term problems with unilateral renal hypoplasia, and some do not know that they have one kidney that is smaller than usual.

Before birth

Unilateral renal hypoplasia 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 hypoplasia 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 pictures of the inside of the body.

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 ones below.

- 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 hypoplastic 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 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.

Chronic kidney disease

In many people with unilateral renal hypoplasia, 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 hypoplasia 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.



About the future

Most people live normal lives with unilateral renal hypoplasia. The other kidney usually works normally, and may grow larger to help do the work of two kidneys.

Your child should be able to do all the things 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 hypoplasia. 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 hypoplasia (both kidneys)

In bilateral renal hypoplasia, both kidneys are smaller than usual because they have not fully developed in the womb. ('Bilateral' means two sides.)

Before birth

Bilateral renal hypoplasia may be suspected at the 20 week antenatal ultrasound scan, which looks at your baby growing in the womb.

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 hypoplasia is affecting your baby.

Referral

If bilateral renal hypoplasia 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 hypoplasia 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.

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Complications and treatment at birth

A small number of babies have symptoms and complications if the hypoplasia 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.

Kidney function

Usually, even if the hypoplastic kidneys are not working as normal, they are working well enough that there are no symptoms at birth. Because hypoplastic 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.

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)

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.

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

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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.

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.
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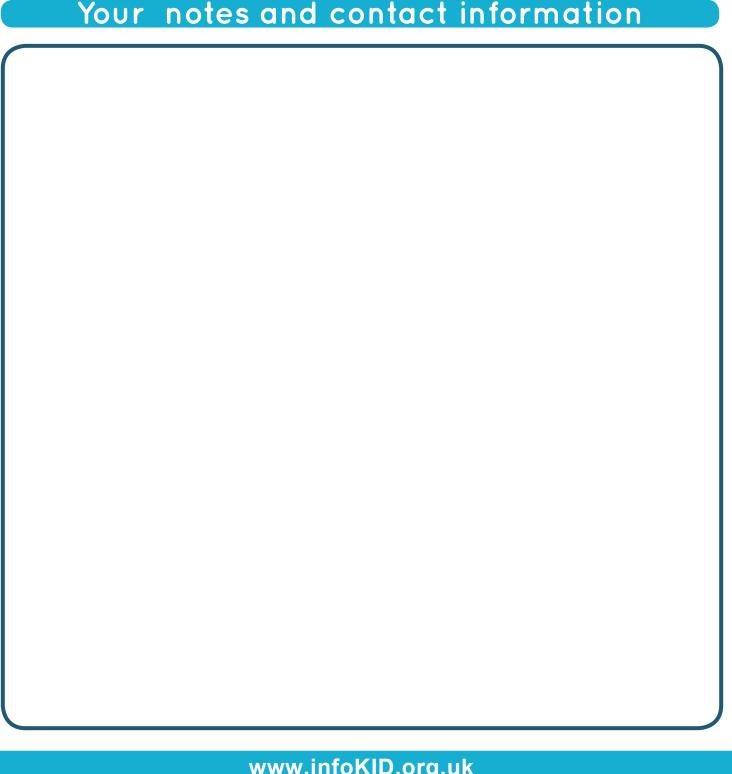
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Further information

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www.infoKID.org.uk







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