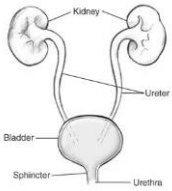


Multicystic Dysplastic Kidney

Monash Children's Hospital

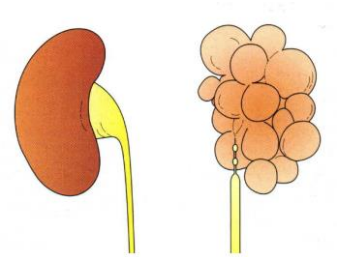
What is normal?

Most people are born with two fist-sized kidneys that each drain via a small muscle-lined tube (ureter) into the bladder.

The kidneys' job is to filter the blood, with millions of microscopic filters (roughly one million per kidney) known as glomeruli. The filters lose some function with time and normal ageing processes. There is usually a good safety margin, such that you have more than enough kidney function for a normal lifespan.

What is multicystic dysplastic kidney?

In some people, a kidney is entirely replaced by fluid-filled sacs or cysts. This is a 'multicystic dysplastic kidney' or MCDK. The 'kidney' has no function.



How does this happen?

Normal kidney development is complex and has many steps. If an error occurs in any step, an abnormal kidney may develop. MCDK is thought to develop when the drainage of urine from the kidney into the ureter is completely blocked. MCDK develops during the foetal period, from 5 weeks gestation.

Usually only one kidney is affected and the other kidney is able to take over the function for both. Very rarely, both kidneys are affected. This situation usually proves fatal.

Who gets it?

MCDK affects approximately 1 in 4000 people. It is more common in boys than in girls.

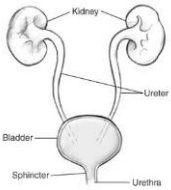
In most cases, the cause of the MCDK cannot be determined. In rare cases, genetic causes or exposure to medication during pregnancy may contribute to MCDK. If there is a family history of abnormal kidney development, genetic tests may be appropriate. If this is not the case, the chances of finding a genetic abnormality on testing are small.

Why is it a problem?

In most people with MCDK, the other kidney works normally. It will also usually compensate with extra growth and thus take over the work of both kidneys. The MCDK will usually shrink and disappear.

If someone has only one kidney, they have half the number of working filters. The have even fewer filters if the single remaining kidney is abnormal as well. These filters adapt, up to a point, and most of the time you can live a perfectly healthy life with one functioning kidney (with a few small modifications). In fact, this is exactly what happens when someone donates a kidney for a kidney transplant. However it is vitally important to protect the remaining filters in the kidney from further damage and strain.

It can be difficult to predict how much kidney tissue is needed before someone's health becomes affected. Kidney disease is typically silent and slow, so it important to have regular check ups (see below). Even with a healthy single kidney, high blood pressure (hypertension) and proteinuria (protein in the urine) may develop, so annual check ups should continue for life.



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Other issues

Sometimes there are other abnormalities in the urinary tract that place the remaining kidney at risk of ongoing damage and these may need to be dealt with.

MCDK may be associated with urinary tract infection or with hypertension.

In some cases, there may be structural abnormalities of the normal kidney, including:

- Vesicoureteric reflux – 'backwash' of urine from the bladder to the upper urinary tract
- Pelviureteric junction obstruction (PUJO) – blockage at the junction of the pelvis (collecting system of the kidney) and ureter
- Vesicoureteric junction obstruction (VUJO) – blockage between the ureter and the bladder

The ultrasound appearance of the normal kidney over the first years of life can be helpful in determining if these conditions might be present. Please see information sheets on vesicoureteric reflux and hydronephrosis, for details of supplemental tests that are sometimes recommended.

How is it diagnosed?

MCDK does not usually cause symptoms. It is commonly discovered on antenatal ultrasound.

After birth, further tests may be necessary, to confirm the diagnosis and to check that the other kidney is normal. This will usually involve an ultrasound scan, checking the size and shape of the kidneys. Repeated ultrasound scans through childhood may be advised, to monitor the growth of the other kidney.

Where there are concerns with the functioning kidney, other test may be needed to investigate specific concerns.

What can we do?

The MCDK requires no treatment, as the abnormal kidney will usually shrink and disappear with time. Children with MCDK are observed periodically and repeated ultrasounds performed to ensure that the healthy kidney is growing well.

In some cases, surgical removal of the MCDK can be performed, if it fails to disappear or if complications develop.

Regular check ups are important to ensure the good kidney remains healthy; usually once per year for blood pressure, blood tests, kidney ultrasound and urine test. If the kidney filters are becoming strained there may be high blood pressure or protein leak through the filters into the urine.

Minimising chances of kidney damage due to diabetes or hypertension is also important, through:

- Maintaining a healthy body weight
- Avoid smoking/alcohol
- Regular exercise
- Avoid high blood pressure

Minimising chance of injury to single kidney by avoiding potentially traumatic situations such as very physical contact sports, and Bungy jumping.

It is possible to have a kidney shield made to fit under clothing to protect the kidney whilst playing sport. This costs between $150-200.

More information?

Please speak to your General Practitioner or Paediatric Nephrologist.