Summary

- Cystic kidney disease is a group of diseases that cause abnormal pockets of fluid (cysts) to form in the kidneys.
- Cystic kidney diseases include polycystic kidney disease, medullary cystic disease and medullary sponge kidney.
- There is no cure for cystic kidney disease - medical treatment aims to manage the symptoms and reduce the risk of complications.

Cystic kidney disease is a group of diseases that cause abnormal pockets of clear, watery fluid (cysts) to form in the kidneys. Cystic kidney diseases include polycystic kidney disease, medullary cystic disease and medullary sponge kidney. The cysts interfere with normal kidney function and may cause kidney failure.

Having a cyst on your kidney does not automatically mean that you have cystic kidney disease. Having one or more cysts on your kidney is common in older people and may not require treatment.

People with long-term kidney problems sometimes develop kidney cysts, particularly if they have kidney failure and have been on dialysis for a long time. This is called ‘acquired cystic kidney disease’ (ACKD) and does not usually require treatment.

Functions of kidneys

The human body normally has two kidneys, one on either side of the middle back, just under the ribs. A kidney is about the size of a fist. Some of the many functions of the kidneys include:

- controlling the amount of fluid and salts in the blood
- filtering out waste products and producing urine
- producing hormones that help to control blood pressure, red blood cell production and calcium levels
- controlling body chemistry to maintain the mineral, salt and acid balance in the blood
- activating vitamin D to keep the bones strong.

Kidney cysts

Cysts are abnormal blisters that may contain fluid or other matter. Many kinds of cysts can affect the kidney. Kidney cysts are classified by:

- cause – may include inherited disorders, acquired kidney disease or advancing age
- features – such as the number of cysts (one or more) and whether the cysts are simple or complicated. Simple cysts are non-cancerous pouches containing clear liquid, and can range in size from tiny blisters to large sacs
- location – outer (cortex) or inner (medulla) part of the kidney.

Types of cystic kidney disease

The three main types of cystic kidney disease are:

- **polycystic kidney disease (PKD)** – usually an inherited condition. Defective genes cause cysts to grow in the kidneys. Both kidneys are affected, but one may develop cysts earlier than the other. PKD is a common cause of kidney failure in Australia and equally affects men and women of all ethnic backgrounds. There is currently no cure for PKD, but research into preventing cyst growths and slowing the decline of kidney function continues
• **medullary cystic kidney disease (MCKD)** – an inherited condition. Cysts develop in the inner part (medulla) of the kidney. The kidneys shrink as the outer section thins. MCKD often causes kidney failure in people between the ages of 20 and 50 years. In a few cases, there is no family history, which may indicate a genetic mutation. The childhood disease ‘juvenile nephronophthisis’ is similar to MCKD

• **medullary sponge kidney** – cysts develop in the urine-collecting ducts and tubules of one or both kidneys. The condition may be congenital (present at birth) and, in some cases, seems to run in families. It is not clear if this is because of genetic mutation or if the condition is inherited. The exact cause of medullary sponge kidney is not known and there is no cure. Kidney failure is rare, but can develop as a result of the repeated infections or kidney stones that can occur with this condition.

**Diagnosis of cystic kidney disease**

The tests recommended by your doctor depend on which cystic kidney disease is suspected. Generally, diagnostic tests may include:

- physical examination – to detect high blood pressure or enlarged kidneys
- urine tests – to look for blood or protein in the urine
- blood tests – to assess kidney function
- renal ultrasound – a simple test that uses sound waves to detect cysts in the kidneys. It is good at identifying even quite small cysts
- computed tomography (CT) and magnetic resonance imaging (MRI) scans – these can detect very small cysts. They may be required if the results from the ultrasound are inconclusive or if more information is needed
- genetic testing – generally used for family studies and not a routine test.

**Treatment for cystic kidney disease**

There is no cure for any of the three cystic kidney diseases. Medical treatment aims to manage symptoms and reduce the risk of complications.

Your doctor’s choice of treatment depends on a range of factors including:

- the type of cystic kidney disease you have
- the severity of the disease
- whether or not you are having complications
- the type and severity of complications
- whether or not your kidneys are failing
- your age and general health.

Treatment may include:

- lifestyle changes
- dietary changes
- medications
- surgery.

There are some developments in research into preventing cyst growth in polycystic kidney disease (PKD). Trials are being conducted in some states in Australia with medications that may stop the sacs from filling with fluid. A worldwide study published in 2012 showed promising results for a medication that inhibits cyst growth and slows the decline of kidney function.

**Where to get help**

- Your doctor
- **Kidney Health Australia** helpine Tel. 1800 454 363