

# ADPKD Patient Route Map

Helping patients and families through their healthcare journey



## Using the ADPKD Patient Route Map

To use all of the features of this Route Map, you must open it using the Adobe Acrobat Reader, available to download [here](#).

It's best to view the Route Map in 'Single Page' mode. Use the interactive features below to find information and to move around the Route Map as you wish.



Use these arrows to move to the next or previous page.

**Pop-ups** > Click on the blue text to get further information on this topic as a 'pop-up' without moving pages.

These include:

 **Questions** >

 **Checklists** >

**Hyperlinks** Click on the blue, underlined text to move straight to another section with more information on this topic. In most cases these links are to other parts of the document. In some cases, the links are to websites. Hyperlinks inside pop-ups are white, but still underlined.

 **Patient quotes** Click on these to hear from other people affected by ADPKD across Europe. Click again on the quote bubble to close it.

**Navigation panel** Use this panel to see where you are in the Route Map and to click to move to any other section.



## Development of this Route Map

The Autosomal Dominant Polycystic Kidney Disease (ADPKD) Patient Route Map was developed jointly by the **European ADPKD Forum** (EAF), an independent, international group of expert doctors and patient advocates, and **PKD International**, the international ADPKD patient support group alliance.

The idea for this Route Map came from a Round Table meeting of expert doctors, patient representatives and organisations involved in caring for people with ADPKD. The resulting [EAF multidisciplinary position statement on ADPKD care](#) explains the basis for the Route Map.

People with ADPKD, and representatives of various ADPKD and kidney health patient organisations, provided input during the development of this Route Map.

All authors and reviewers are listed [here](#).

## Sponsorship

Otsuka Pharmaceutical Europe Ltd initiated and facilitated the EAF and funded its activities. The ADPKD Patient Route Map and the EAF Multidisciplinary Position Statement were funded by Otsuka Pharmaceutical Europe Ltd and Ipsen Farmaceutica BV.

No authors or reviewers received fees in respect of this project. This Route Map represents the opinions of the authors and not necessarily those of the sponsors.

## Copyright

This Route Map is free to download from the [PKD International website](#).

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## What is ADPKD?

Autosomal dominant polycystic kidney disease (or 'ADPKD') is a chronic condition that causes [cysts](#) > to develop in the kidneys and sometimes elsewhere in the [body](#). These cysts can cause various symptoms and they may cause the kidney to stop working properly. ADPKD is an inherited, [genetic](#) disease usually passed from parents to their children at birth. It is typically diagnosed in adulthood, but can also be diagnosed in children and even before birth.

ADPKD is a complex disease that can sometimes be overwhelming for patients and their families. In addition to the physical effects of ADPKD, the diagnosis can also have important emotional and psychological effects, including feelings of fear and anger.

Although ADPKD cannot be cured, patients, families and healthcare professionals can take various steps to help look after the kidneys and manage the effects of the disease.

Knowledge is power – and so learning about ADPKD can empower patients and carers to cope with the disease and to get the care and support they need.

## What is the aim of the ADPKD Patient Route Map?

This Route Map has been developed by healthcare experts and patients to explain the types of care and support that patients and families should expect from a health service.

The aim is to help patients and families to:

- **manage their own health** as much as possible, with their healthcare team
- **talk about ADPKD** with their [healthcare team](#) and to participate in making decisions about their own care, when required
- **make the best use of available services** so that everyone affected by ADPKD gets the care, support and information they need, at the right time.

The Route Map can also help [patient organisations](#) and [healthcare policymakers and providers](#) to improve services for people affected by ADPKD.

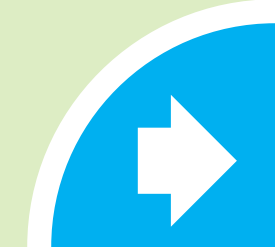
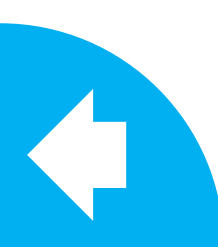
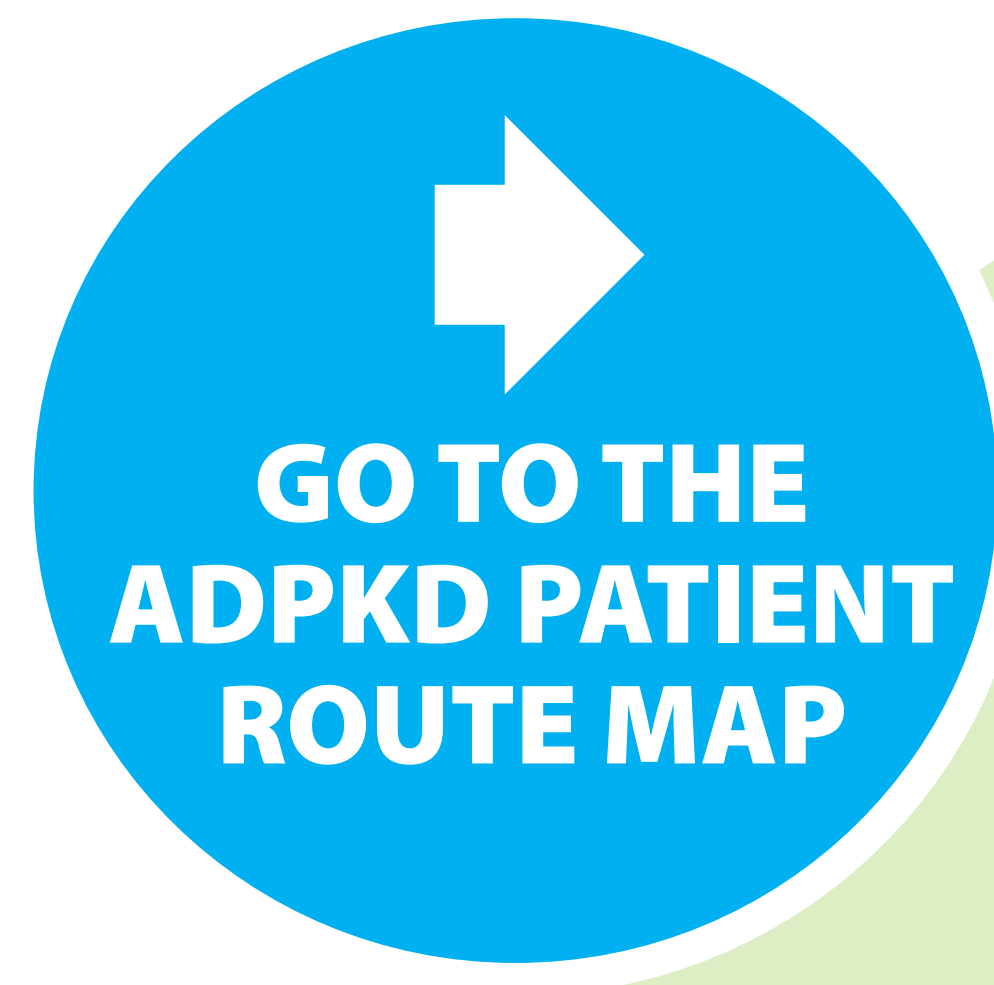
## What does the Route Map cover?

The Route Map explains what ADPKD is and how it affects patients and families. It explains how ADPKD is diagnosed, investigated and managed at each stage during the lifelong patient care pathway. It also provides advice on issues such as genetics, family planning, emotional wellbeing and finances.

The Route Map is based on the latest [scientific knowledge](#) > about ADPKD and insights from experts and patients from across Europe.

Checklists are provided to help patients and families get the most out of consultations, and to help healthcare teams ensure that patients are always at the centre of their care pathway.

A list of [patient organisations](#) is provided – these are a vital source of information and support.



## What is ADPKD?

Autosomal dominant polycystic kidney disease (ADPKD) is a chronic condition that causes [cysts](#) to develop in the kidneys and sometimes elsewhere in the [body](#). These cysts can grow and they may cause the kidneys to stop working. ADPKD is an inherited [genetic](#) disease usually passed from children's parents. It is typically diagnosed in adulthood but diagnosis in children and even babies can occur.

ADPKD is a complex disease that can sometimes affect patients and their families. In addition to the physical effects of ADPKD, the diagnosis can also have important psychological effects, including feelings of fear and uncertainty.

Although ADPKD cannot be cured, patients, family members and healthcare professionals can take various steps to help look after the effects of the disease.

Knowledge is power – and so learning about ADPKD can help patients and their families to cope with the disease and the support they need.

## What is the aim of the ADPKD Patient Guide?

The Patient Guide has been developed by health professionals to explain the signs of cysts and support family members should require from a health service.

The aim is to help patients and families to:

- **manage their own health** as much as possible with their own team
- **talk about ADPKD** with their [healthcare team](#) and make decisions about their own care, when needed
- **make the best use of available services** to help manage the effects of ADPKD, get the care, support and information they need, at the right time

The Patient Guide can also help [patients, organisations and healthcare professionals and carers](#) to improve services for people affected by ADPKD.

## What does the Patient Guide cover?

The Patient Guide explains what ADPKD is, how it affects patients, and how it is diagnosed. It also covers ADPKD's symptoms, management and treatment, and how to get help with living with ADPKD.

### Knowledge

*'It's a great help to better understand her illness.'*

*Peter (husband of a patient with ADPKD), Germany*

*'With more knowledge and patient education, you can assume more control of the disease.'*

*Juan, Spain*

*'Knowledge is so important – almost as important as medicine. The more you understand, the more likely you are to adhere to your treatment plan.'*

*Cathriona, Ireland*

## What is ADPKD?

Autosomal dominant polycystic kidney disease (ADPKD) is a complex disease that can sometimes be asymptomatic for patients and their families. In addition to the physical effects of ADPKD, the disease can also have important emotional and psychological effects, including feelings of fear and anger.

Although ADPKD cannot be cured, patients, families and healthcare professionals can take various steps to help look after the kidneys and manage the effects of the disease.

Knowledge is power – and so learning about ADPKD can empower patients and cars to cope with the disease and to get the care and support they need.

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The Route Map can also help **patient organisations** and **healthcare professionals and carers** to improve services for people affected by ADPKD.

## What does the Route Map cover?

The Route Map explains what ADPKD is and how it affects patients. It explains how ADPKD is diagnosed, investigated and treated, and what steps during the illness patients can follow. It also covers an issue such as genetics, family planning, fertility and pregnancy.

The Route Map is based on the latest **scientific knowledge** about ADPKD and insights from experts and patients from across Europe.

**Guidance** is provided to help patients and families get the most out of consultations, and to help healthcare teams ensure that patients are always at the centre of their care pathway.

A list of **patient organisations** is provided – these are a vital source of information and support.



**Cysts** are fluid-filled sacs that grow and multiply in the kidneys and often in other parts of the body (especially the liver) in people with ADPKD.

## What is ADPKD?

Autosomal dominant polycystic kidney disease (ADPKD) is a chronic condition that causes [cysts](#) to develop in the kidneys and sometimes elsewhere in the [body](#). These cysts can cause kidney symptoms and they may cause the kidneys to stop working properly. ADPKD is an inherited [genetic](#) disease usually passed from parents to their children at birth. It is typically diagnosed in adulthood but can also be diagnosed in children and even before birth.

ADPKD is a complex disease that can sometimes be overwhelming for patients and their families. In addition to the physical effects of ADPKD, the diagnosis can also have important emotional and psychological effects, including feelings of fear and anger.

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The Route Map can also help [patient organisations](#) and [healthcare professionals and carers](#) to improve services for people affected by ADPKD.

## What does the Route Map cover?

The Route Map explains what ADPKD is, how it affects patients and families. It explains how ADPKD is diagnosed, investigated and managed at various stages of the disease, and what care and support is available.

It also provides information on:

The Route Map covers ADPKD and the

[diagnosis](#) and of [investigation](#) steps at the

of [management](#) of information



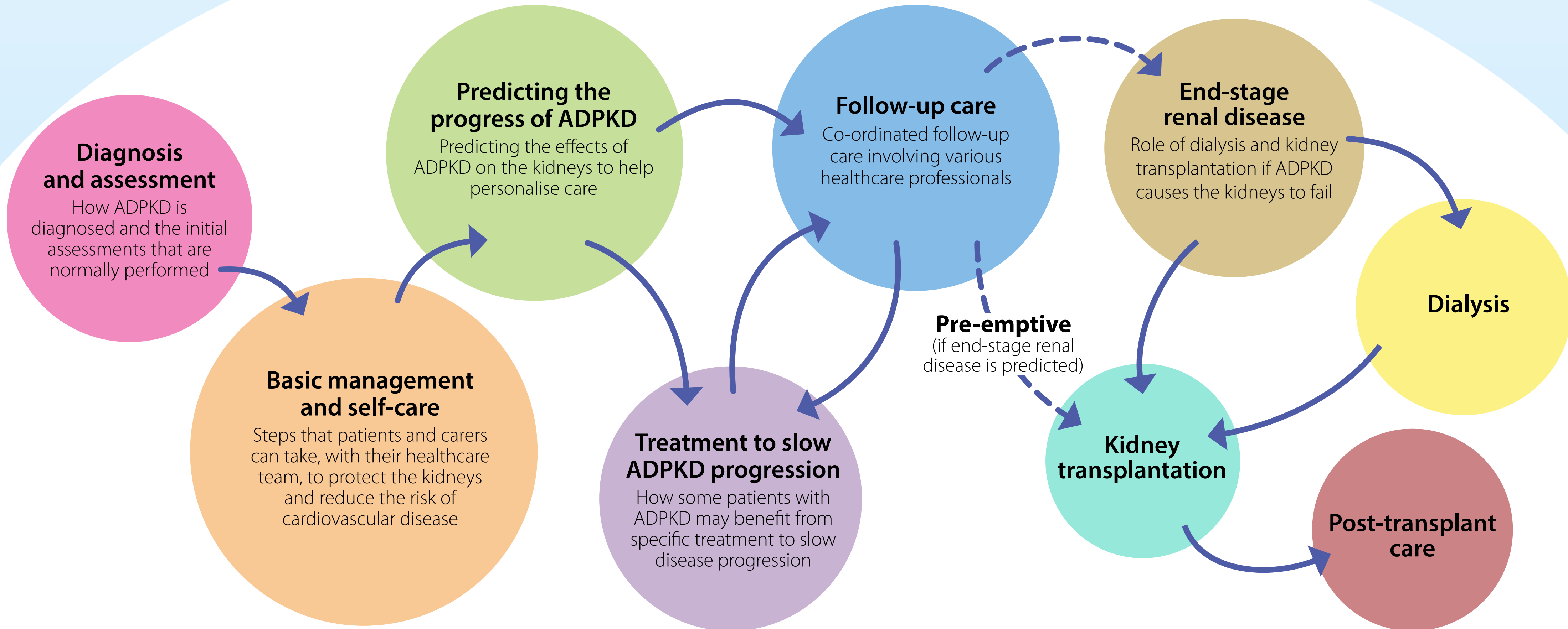
### Scientific knowledge

The Route Map draws on the [ADPKD Consensus Conference Report](#) published by the Kidney Disease – Improving Global Outcomes (KDIGO) initiative, the [European ADPKD Forum \(EAF\) Report](#), the [EAF Multidisciplinary Position Statement on ADPKD care](#), and other recent guidance (see [Further reading](#)).



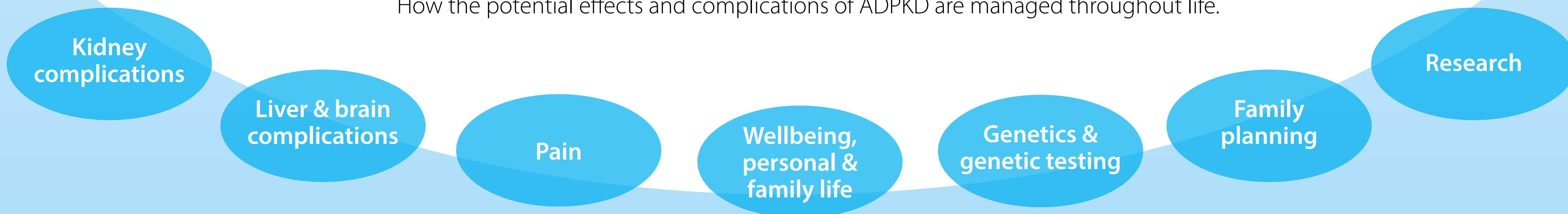
Please click on any bubble to move to that section.

## Lifetime ADPKD care pathway



## Ongoing care and support

How the potential effects and complications of ADPKD are managed throughout life.



## General information

Understanding ADPKD

Principles of ADPKD care

Information for health policymakers and providers

Patient organisations

Further reading

Authors and acknowledgements

# Understanding ADPKD

This section outlines how ADPKD can affect the health of patients.

ADPKD affects the kidneys and sometimes other parts of the body, as you can see here.

ADPKD affects different people in different ways, so not everyone will experience the effects shown here. It is also important to remember that much can be done to help reduce, manage and treat the effects of ADPKD. Although the disease can have a significant effect on life, it does not mean that people with the disease cannot live happy, long and productive lives.

Many [patient organisation](#) websites provide more information about the effects of ADPKD.

## Kidneys

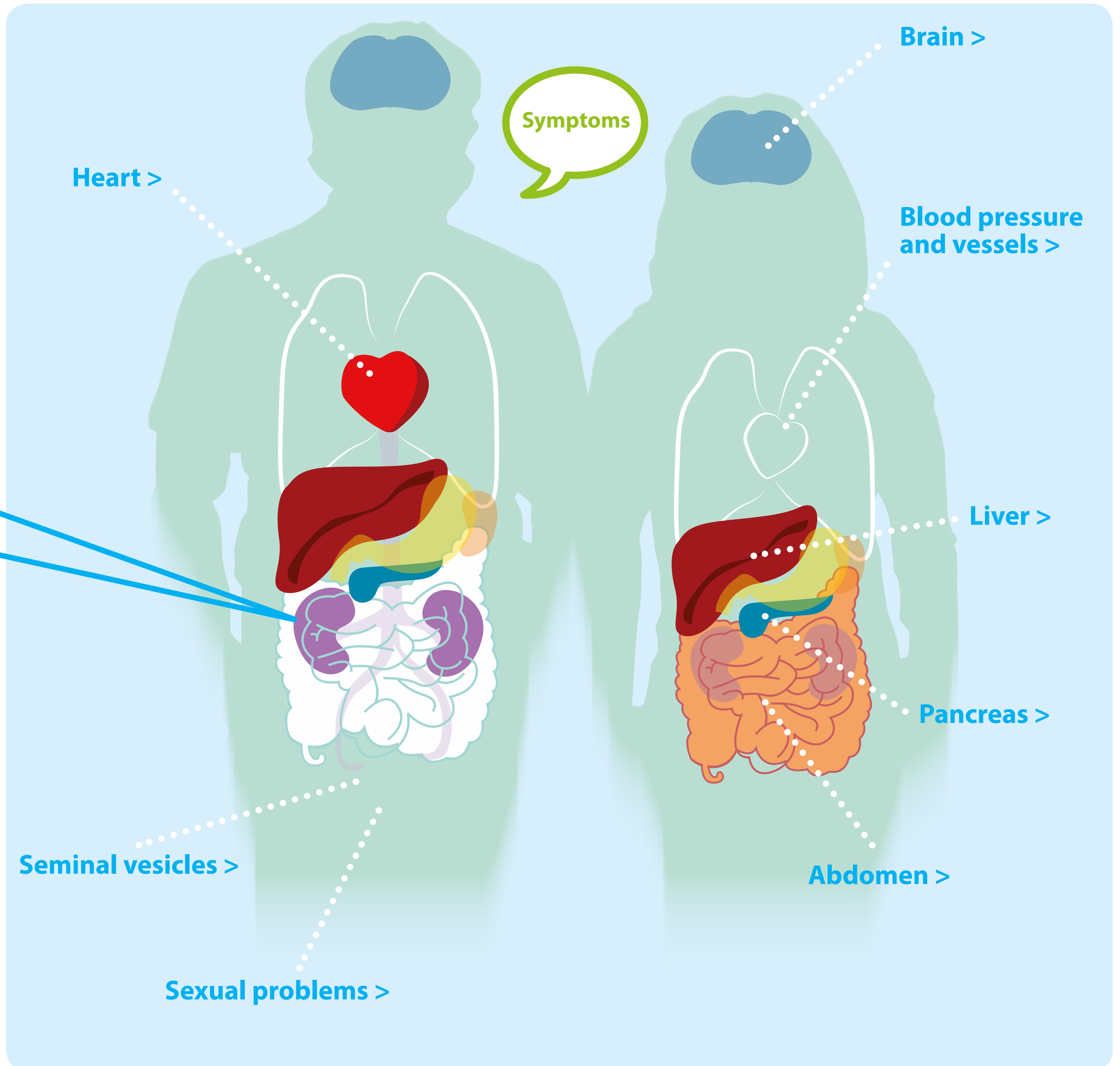
ADPKD is a type of [chronic kidney disease](#) > that causes [cysts](#) > to grow and multiply in the kidneys. If the cysts grow very large they can eventually cause the kidneys and the abdomen to swell. Cysts can eventually stop the kidneys working properly and sometimes this can lead to kidney failure, or '[end-stage renal disease](#)'. Cysts can also cause [pain](#) and other [complications in the kidney](#).

If you have ADPKD, you can help to protect your kidneys from the progression of the disease through certain [diet and lifestyle](#) measures.

## ? Why are the kidneys so important? >

## Wellbeing, personal and family life

ADPKD can interfere with normal activities, such as socialising, family life and work. This can have a significant [emotional and psychological impact](#).





This section outlines how ADPKD can affect the health of patients.

ADPKD affects the kidneys and sometimes other parts of the body. It can also affect the brain.

ADPKD affects different people in different ways. Some people experience the effects of ADPKD early in life, while others experience the effects of ADPKD later in life. Some people experience the effects of ADPKD through the growth of the kidneys, while others experience the effects of ADPKD through the growth of other parts of the body.

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**Diagnosis**  
ADPKD is a type of [autosomal dominant](#) genetic disease. This means that you only need one copy of the faulty gene to get the disease. You can inherit the faulty gene from one or both of your parents. You can also have the faulty gene without knowing it. You can also have the faulty gene without knowing it. You can also have the faulty gene without knowing it.

## How can ADPKD be diagnosed?

**Monitoring personal and family life**  
ADPKD can be diagnosed with several different tests, such as imaging tests, blood tests and genetic tests. You can also have a [genetic test](#) to see if you have the faulty gene.

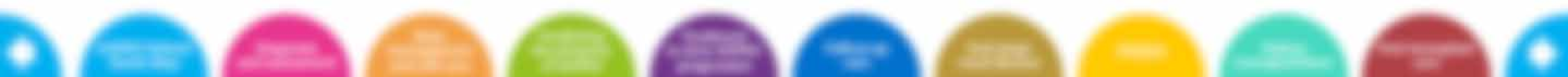
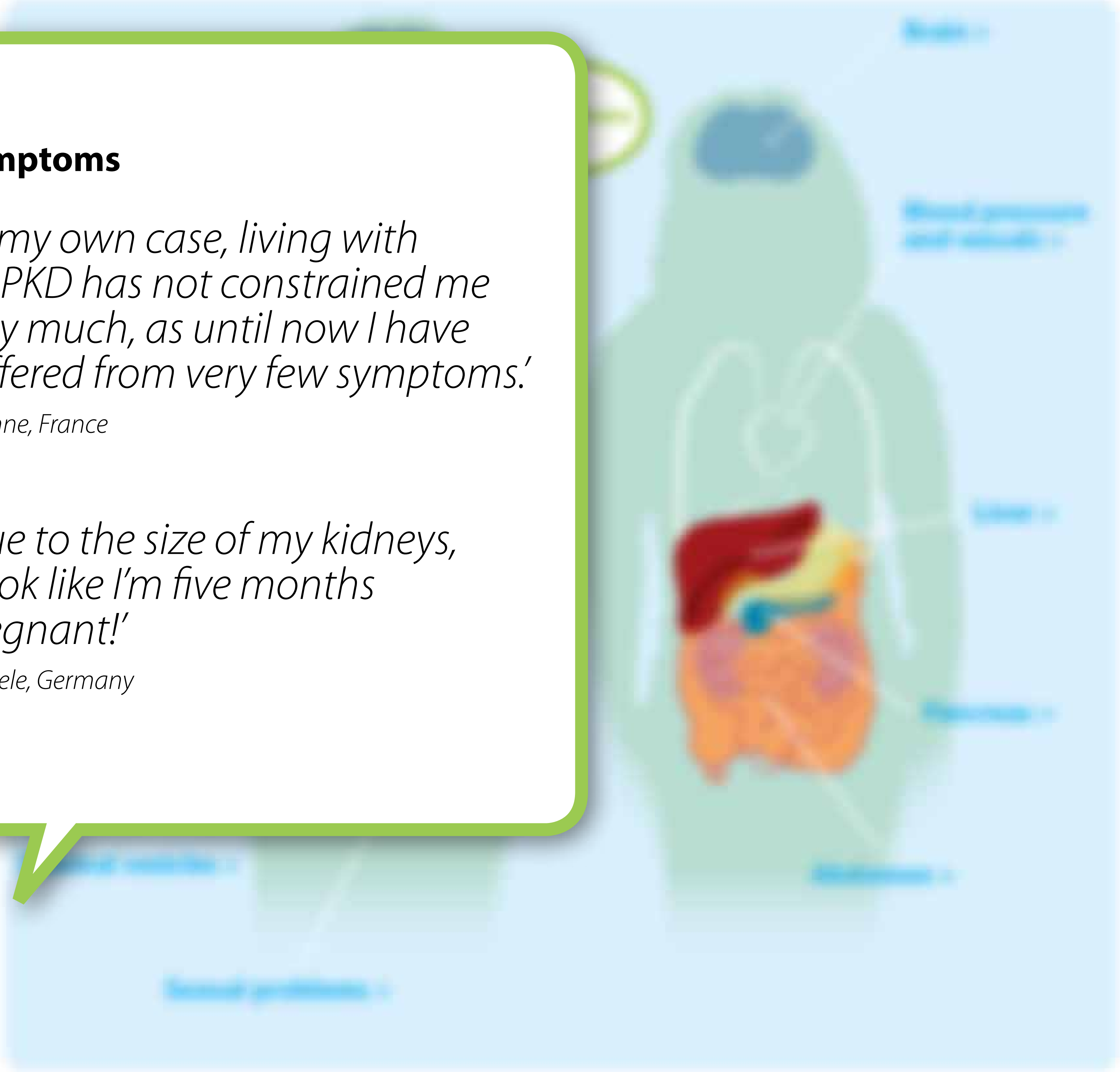
## Symptoms

*'In my own case, living with ADPKD has not constrained me very much, as until now I have suffered from very few symptoms.'*

Corinne, France

*'Due to the size of my kidneys, I look like I'm five months pregnant!'*

Daniele, Germany



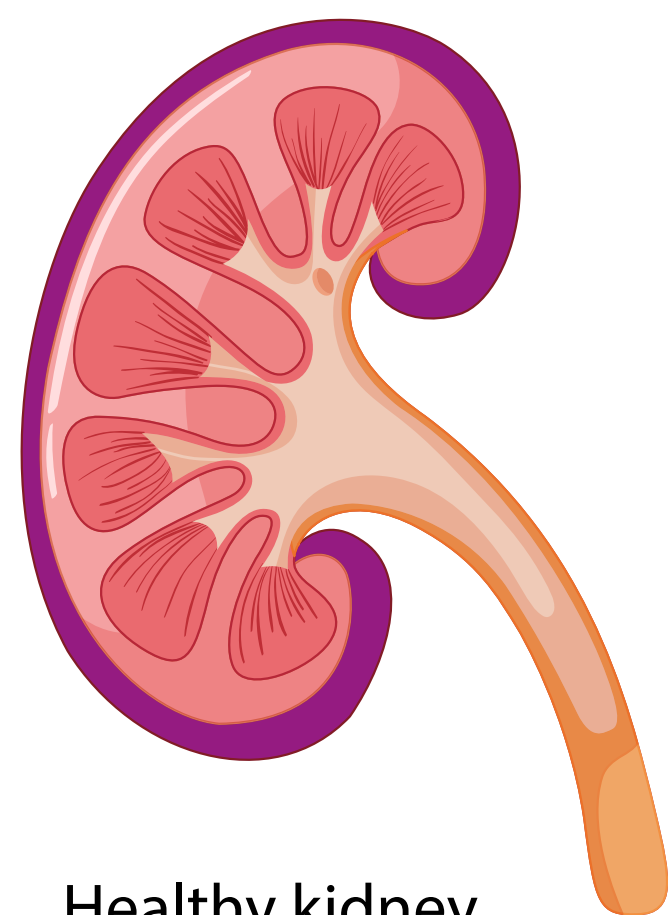
The various conditions from ADPKD can affect the health of patients.

ADPKD affects the number and location of the cysts in the body.

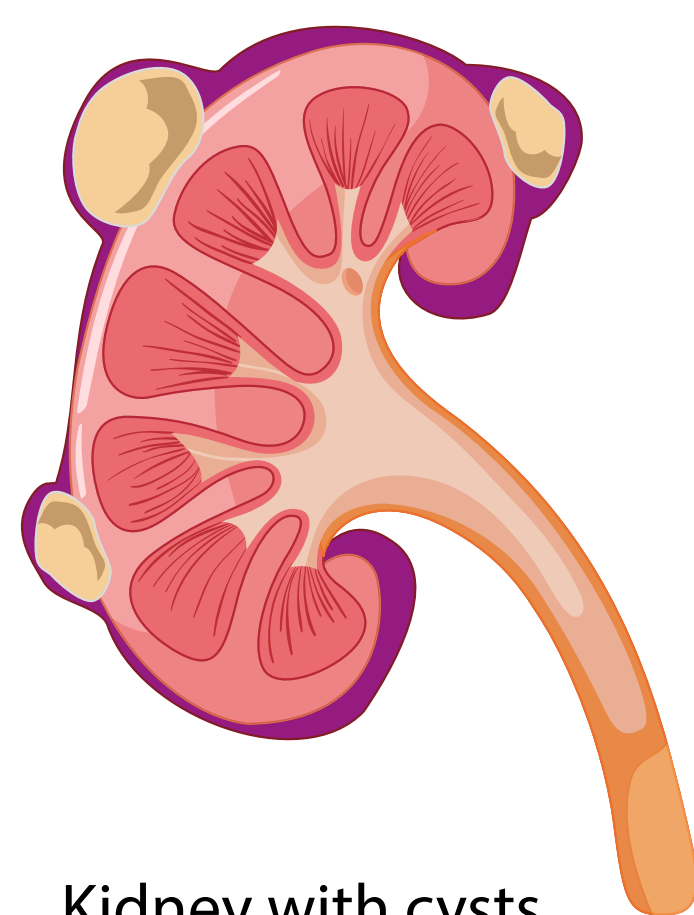
ADPKD affects different parts of the body in different ways. It can cause kidney failure, high blood pressure, liver disease, brain aneurysms, and other conditions.

## Cysts

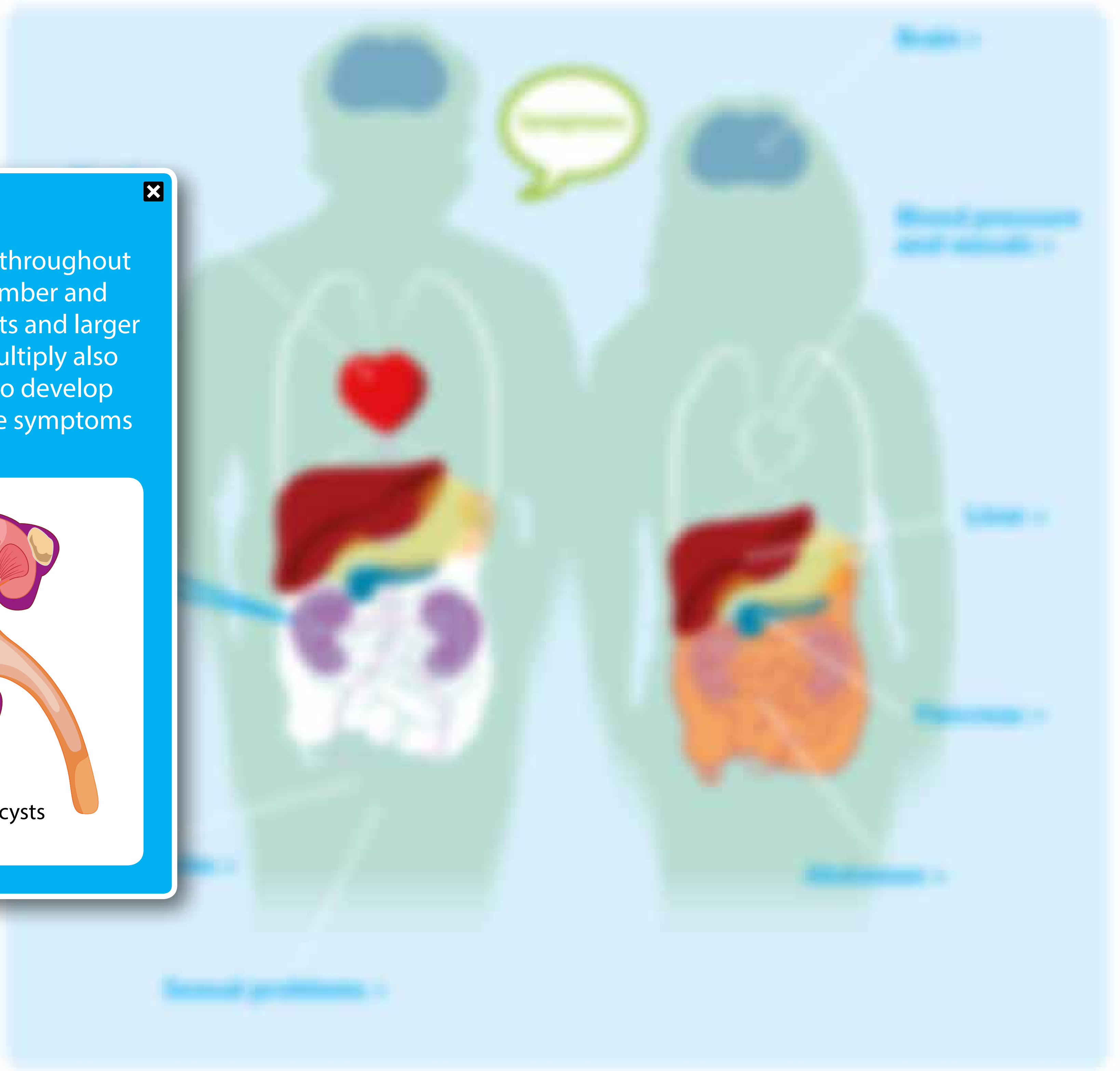
Cysts are fluid-filled sacs that grow and multiply throughout life in the kidneys of people with ADPKD. The number and size of cysts varies – some people have more cysts and larger cysts than others. How quickly they grow and multiply also differs between patients. The cysts usually start to develop even before birth, but most people do not notice symptoms until adulthood.



Healthy kidney



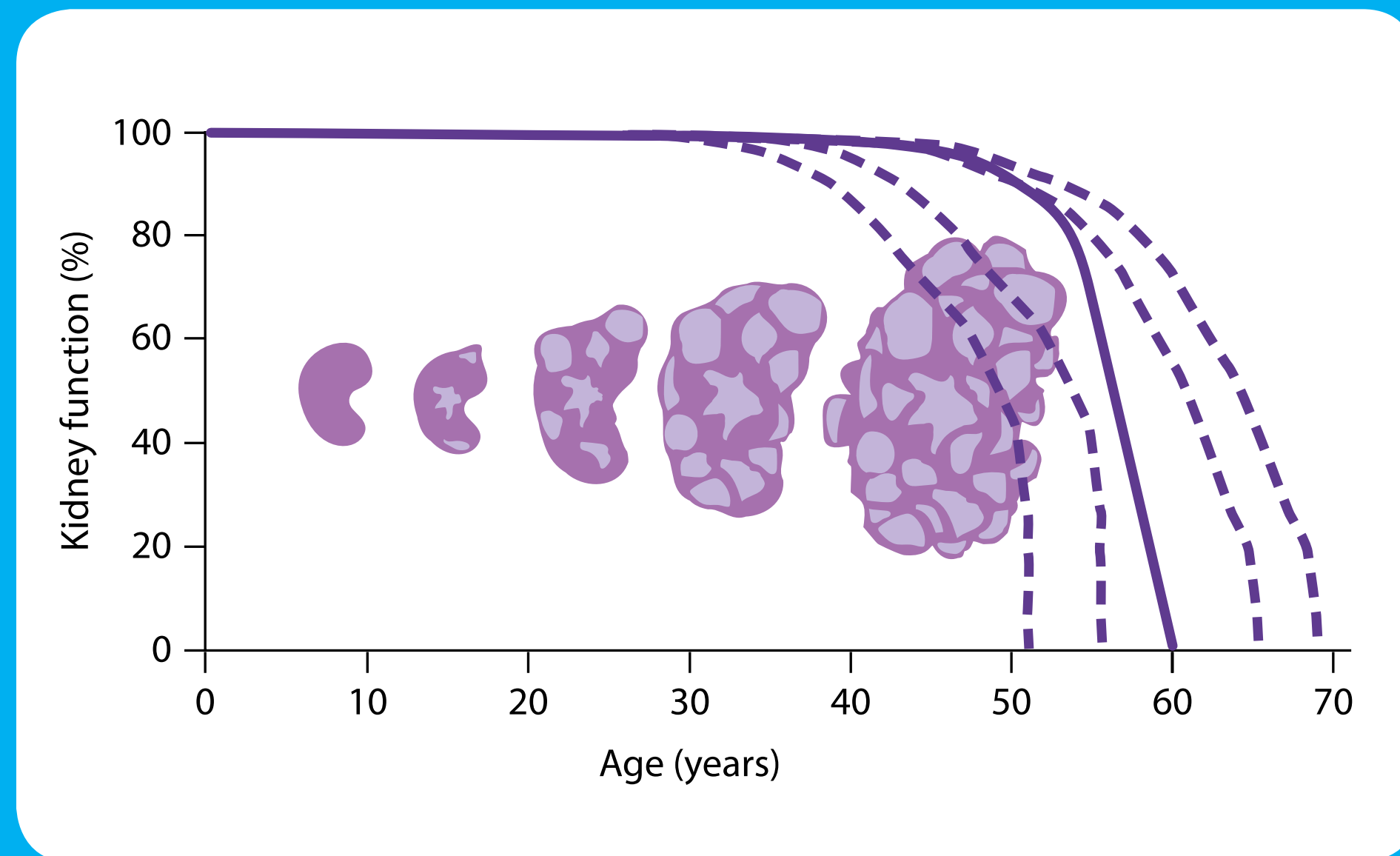
Kidney with cysts



## Chronic kidney disease ✕

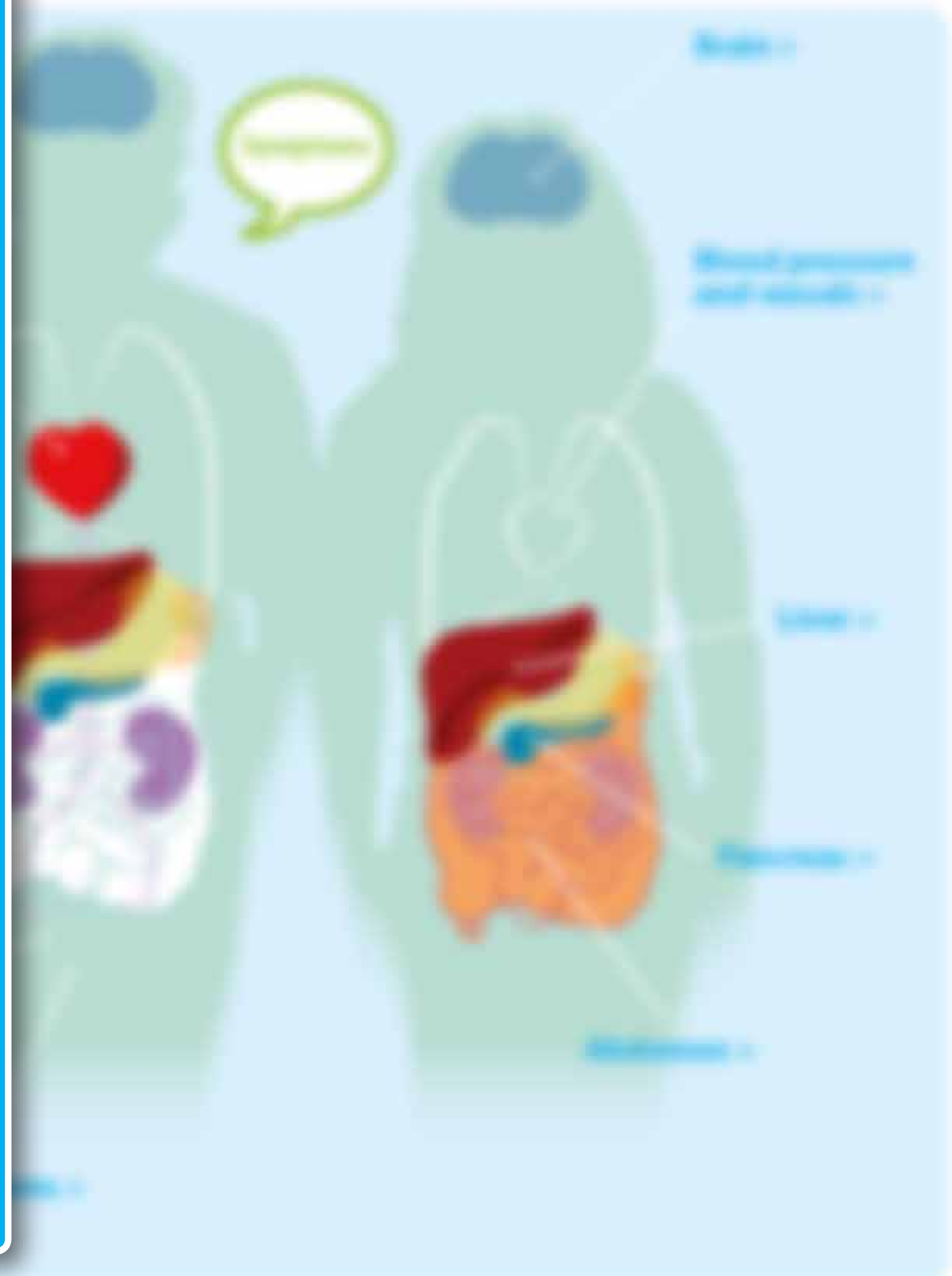
Chronic kidney disease (CKD) is a long-term condition in which the kidneys do not work normally or when they are damaged.

The kidneys can continue to work normally during the early stages of ADPKD, when there are fewer, small cysts. However, as the cysts grow and multiply they can stop the kidneys working properly, causing CKD (see illustration). The rate at which kidney disease progresses varies between people, as shown by the different dotted lines.



The severity of CKD is normally monitored by measuring how well the kidneys filter fluid, as explained in more detail in the [Diagnosis and assessment](#) section.

Eventually, ADPKD can cause [end-stage renal disease \(ESRD\)](#), the most severe stage of CKD also known as kidney failure.

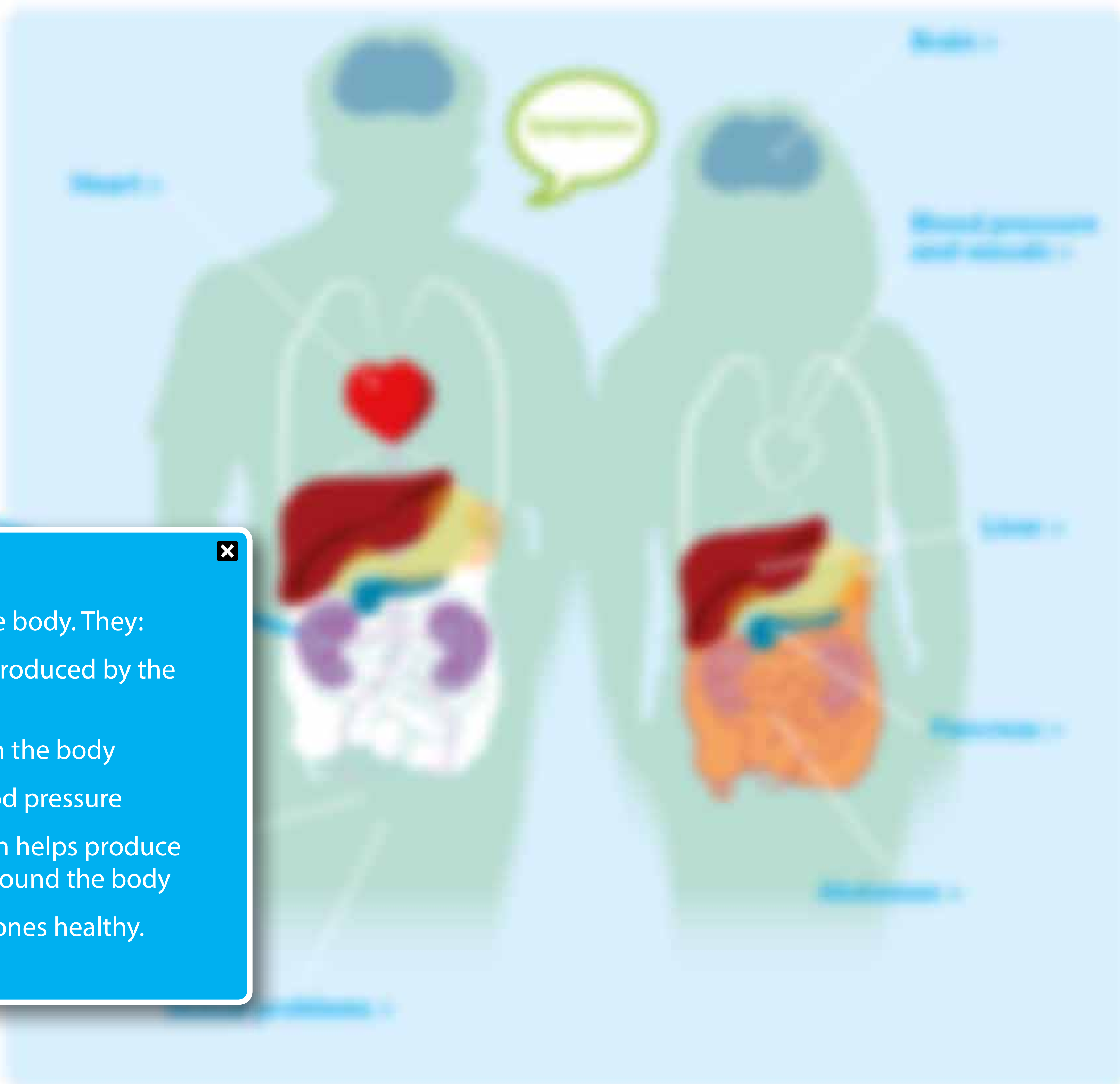


This section outlines how ADPKD can affect the health of patients.

ADPKD affects the kidneys and sometimes other parts of the body, in a similar way.

ADPKD affects different people in different ways. An important consideration for all is blood pressure. It is also important to remember that much can be done to help reduce, manage and slow the effects of ADPKD. Although the disease can have a significant effect on life, it does not mean that people with the disease cannot live happy, long and productive lives.

For more information, visit [www.kidney.org](#) website, please read more information about the effects of ADPKD.



## Kidneys

ADPKD is a type of [autosomal dominant disease](#). This means that you only need one copy of the faulty gene to have the disease. If you have two copies, you will have a more severe form of the disease and the disease will be passed on to your children.

### Why are the kidneys so important?

The kidneys have many important roles in the body. They:

- filter the blood to remove waste products produced by the body – these are excreted in the urine
- adjust the balance of water, salts and acid in the body
- release renin, which helps regulate the blood pressure
- produce the hormone erythropoietin, which helps produce the red blood cells that transport oxygen around the body
- produce vitamin D, which helps keep the bones healthy.



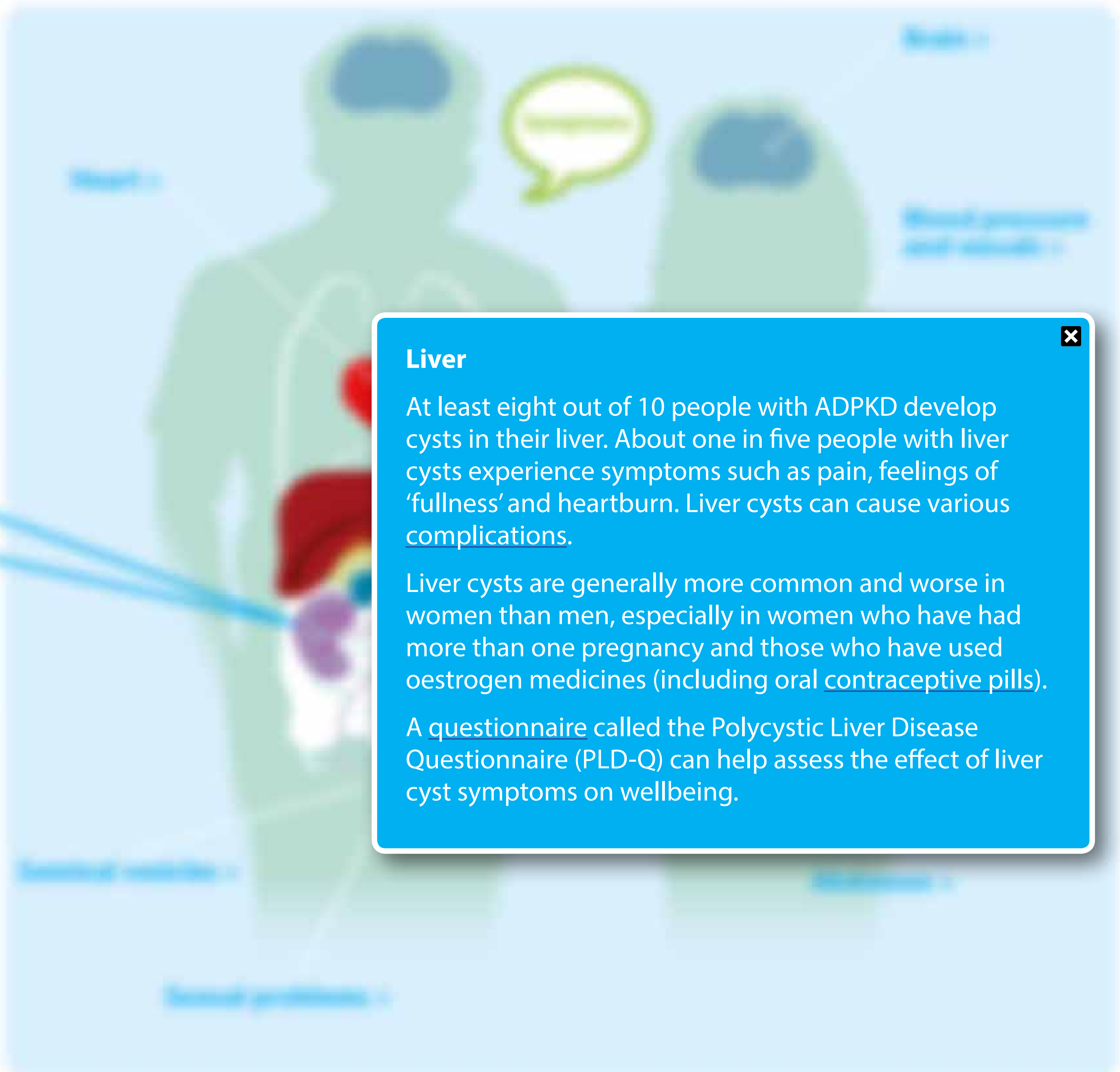
# Understanding ADPKD

This section outlines how ADPKD can affect the health of patients.

ADPKD affects the kidneys and sometimes other parts of the body. It can also affect:

ADPKD affects different people in different ways. An important consideration for adults is how to manage the effects of ADPKD. It is also important to remember that much can be done to help reduce, manage and ease the effects of ADPKD. Although the disease can have a significant effect on life, it does not mean that people with the disease cannot live happy, long and productive lives.

For more information, visit our [resources](#) page for information about the effects of ADPKD.



**Kidneys**  
ADPKD is a type of [autosomal dominant disease](#). This means that you only need one copy of the faulty gene to get the disease. If you have two copies, you will get the disease. The disease is passed on from one parent to their children. If you have ADPKD, you can help to reduce the risk of passing on the disease through [genetic testing](#).

## How can the disease be managed?

**Reducing personal and family life**  
ADPKD can interfere with normal activities, such as working, family life and work. You can have a significant [impact on your life](#).

**Liver**

At least eight out of 10 people with ADPKD develop cysts in their liver. About one in five people with liver cysts experience symptoms such as pain, feelings of 'fullness' and heartburn. Liver cysts can cause various complications.

Liver cysts are generally more common and worse in women than men, especially in women who have had more than one pregnancy and those who have used oestrogen medicines (including oral [contraceptive pills](#)).

A [questionnaire](#) called the Polycystic Liver Disease Questionnaire (PLD-Q) can help assess the effect of liver cyst symptoms on wellbeing.



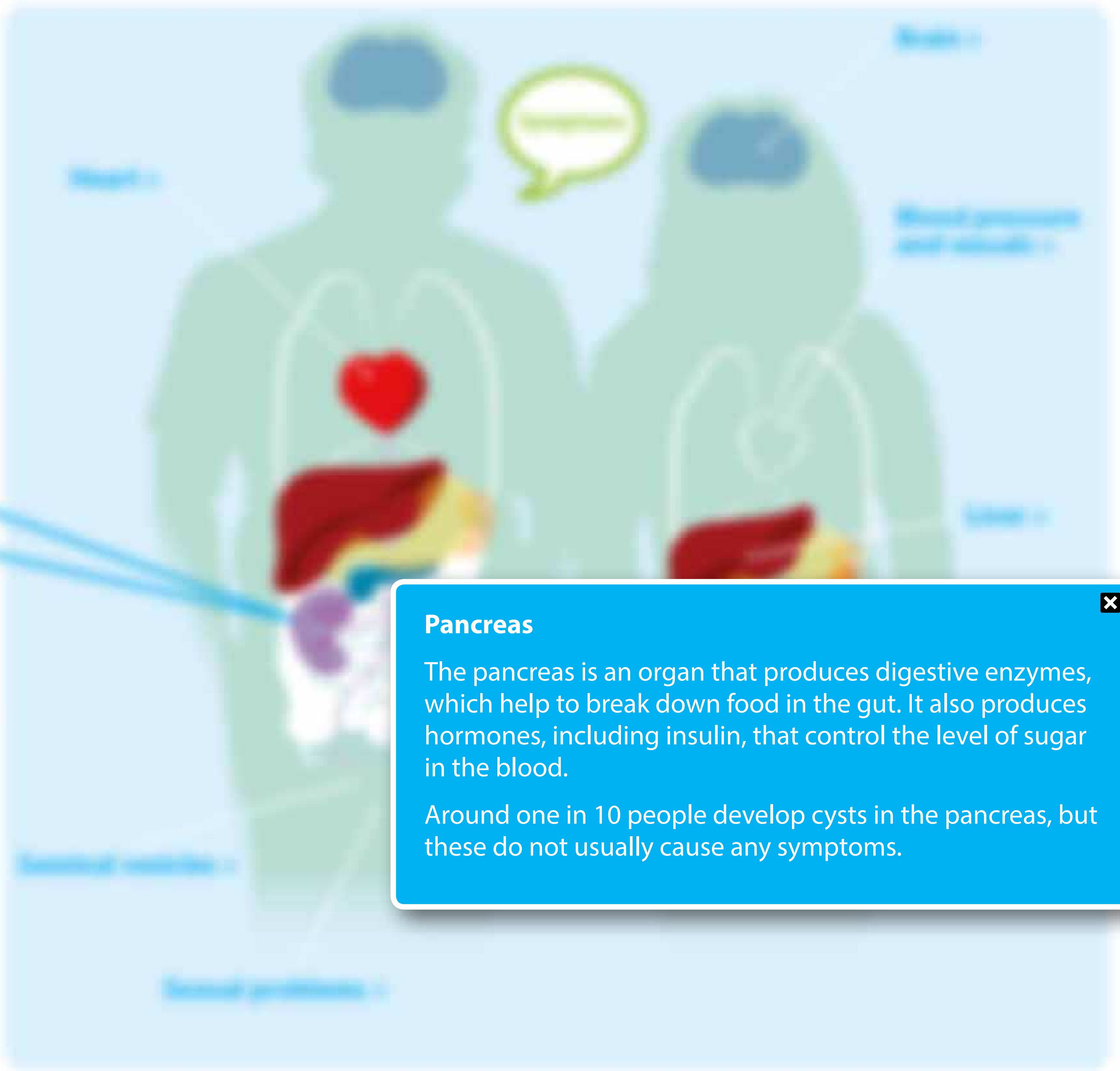
# Understanding ADPKD

This section outlines how ADPKD can affect the health of patients.

ADPKD affects the kidneys and sometimes other parts of the body. It can also affect:

ADPKD affects different people in different ways. An important consideration for all those affected is the importance of ensuring that you can live a full and active life. It is also important to understand that much can be done to help reduce, manage and ease the effects of ADPKD. Although the disease can have a significant effect on life, it does not mean that people with the disease cannot live happy, long and productive lives.

For more information, visit our [patient resources](#) page for information about the effects of ADPKD.



**Kidneys**  
ADPKD is a type of [cystic kidney disease](#) that causes [cysts](#) to grow and multiply in the kidneys. If the cysts grow very large they can eventually cause the kidneys and the abdomen to swell. Cysts can eventually stop the kidneys working properly and sometimes they can lead to kidney failure. [Living with kidney failure](#) can be difficult and you may need dialysis or a kidney transplant.

If you have ADPKD, you can help to prevent complications from the progression of the disease through [certain lifestyle changes](#).

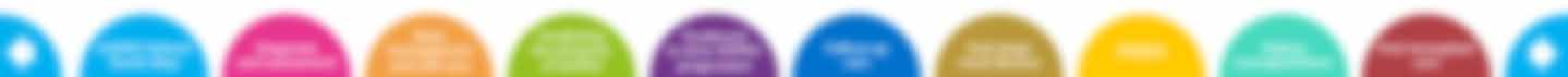
**Pancreas**

The pancreas is an organ that produces digestive enzymes, which help to break down food in the gut. It also produces hormones, including insulin, that control the level of sugar in the blood.

Around one in 10 people develop cysts in the pancreas, but these do not usually cause any symptoms.

## Living with ADPKD

**Maximising personal and family life**  
ADPKD can interfere with normal activities, such as working, family life and work. You can have a significant [impact on your life](#).



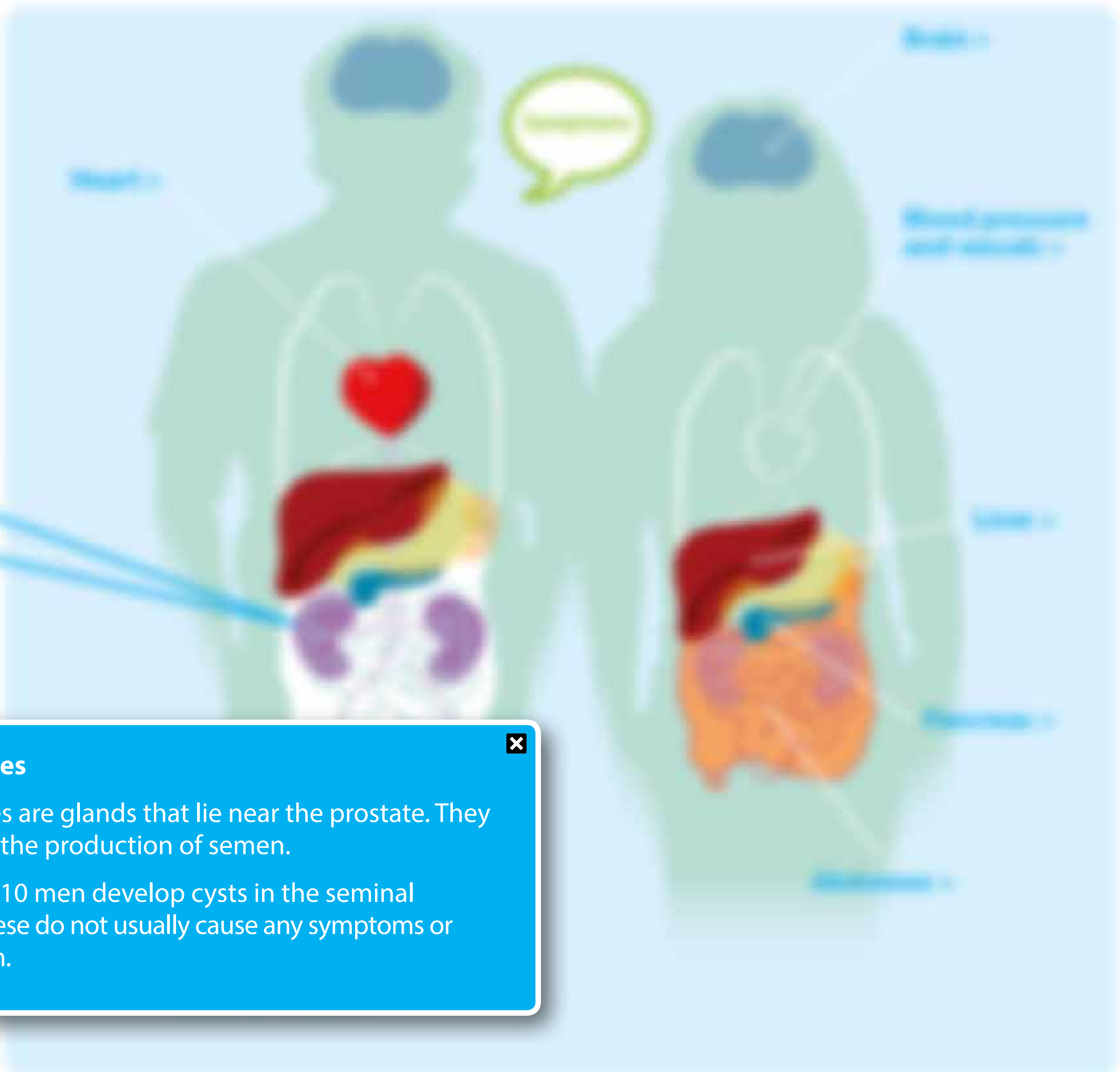
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ADPKD affects the kidneys and sometimes other parts of the body. It can run in families.

ADPKD affects different people in different ways. An important consideration for all those affected is to also remember to consider the health of the brain. It is also important to remember that much can be done to help reduce, manage and ease the effects of ADPKD. Although the disease can have a significant effect on life, it does not mean that people with the disease cannot live happy, long and productive lives.

For more information, visit our website [www.geff.org.uk](#) where you can find more information about the effects of ADPKD.



**Kidneys**  
ADPKD is a type of **fluid-filled cysts** that can grow in the kidneys. If the cysts grow very large they can eventually cause the kidneys and the abdomen to swell. Cysts can eventually stop the kidneys working properly and sometimes this can lead to kidney failure. **It is important to see your doctor** if you have any of the following symptoms:

- Pain in the back or sides
- Blood in your urine
- Frequent urination
- High blood pressure
- Swelling in your legs

If you have ADPKD, you can help to reduce your risk of kidney failure by following the advice of your doctor. You can also help to reduce your risk of kidney failure by following the advice of your doctor.

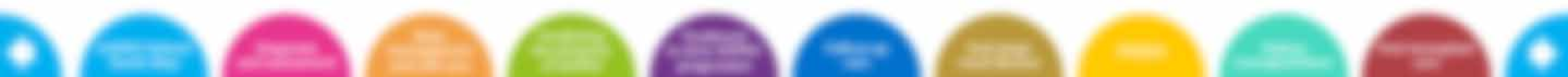
**Seminal vesicles**

Seminal vesicles are glands that lie near the prostate. They are involved in the production of semen.

Around four in 10 men develop cysts in the seminal vesicles, but these do not usually cause any symptoms or affect the sperm.

## How can the kidneys be supported?

Reducing potassium and family life... ADPKD can sometimes affect the heart, and this can be serious. You can help to reduce your risk of heart disease by following the advice of your doctor.



This section outlines how ADPKD can affect the health of patients.

ADPKD affects the kidneys and sometimes other parts of the body, such as the brain.

ADPKD affects different people in different ways. An individual's experience of the effects of ADPKD varies. It is also important to remember that most people have a healthy kidney, manage well and live for decades with ADPKD. Although the disease can have a significant effect on life, it does not mean the people with the disease cannot live happy, long and productive lives.

[More information](#) website provides more information about the effects of ADPKD.

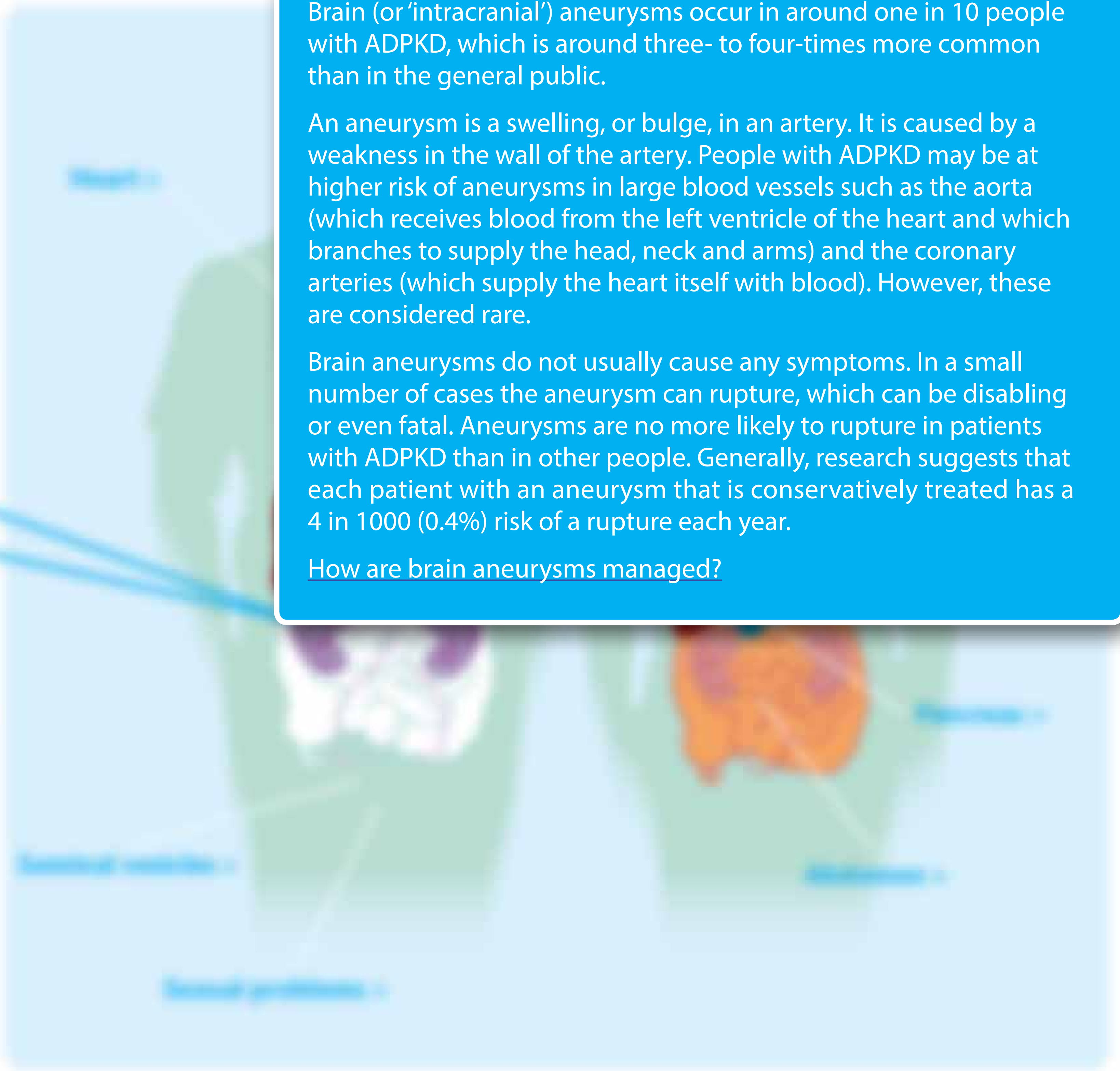
**Outcomes**

ADPKD causes **brain aneurysms**. The brain is a very sensitive organ and a rupture of the aneurysm can cause a stroke, which can be fatal. The aneurysm can also cause a headache, which can be severe. The aneurysm can also cause a seizure, which can be fatal. The aneurysm can also cause a loss of consciousness, which can be fatal. The aneurysm can also cause a loss of vision, which can be fatal. The aneurysm can also cause a loss of hearing, which can be fatal. The aneurysm can also cause a loss of taste, which can be fatal. The aneurysm can also cause a loss of smell, which can be fatal. The aneurysm can also cause a loss of touch, which can be fatal. The aneurysm can also cause a loss of pain, which can be fatal. The aneurysm can also cause a loss of temperature, which can be fatal. The aneurysm can also cause a loss of pressure, which can be fatal. The aneurysm can also cause a loss of vibration, which can be fatal. The aneurysm can also cause a loss of position, which can be fatal. The aneurysm can also cause a loss of movement, which can be fatal. The aneurysm can also cause a loss of coordination, which can be fatal. The aneurysm can also cause a loss of balance, which can be fatal. The aneurysm can also cause a loss of posture, which can be fatal. The aneurysm can also cause a loss of gait, which can be fatal. The aneurysm can also cause a loss of speech, which can be fatal. The aneurysm can also cause a loss of writing, which can be fatal. The aneurysm can also cause a loss of drawing, which can be fatal. The aneurysm can also cause a loss of reading, which can be fatal. The aneurysm can also cause a loss of calculation, which can be fatal. The aneurysm can also cause a loss of memory, which can be fatal. The aneurysm can also cause a loss of attention, which can be fatal. The aneurysm can also cause a loss of concentration, which can be fatal. The aneurysm can also cause a loss of motivation, which can be fatal. The aneurysm can also cause a loss of energy, which can be fatal. The aneurysm can also cause a loss of endurance, which can be fatal. The aneurysm can also cause a loss of strength, which can be fatal. The aneurysm can also cause a loss of speed, which can be fatal. The aneurysm can also cause a loss of accuracy, which can be fatal. The aneurysm can also cause a loss of precision, which can be fatal. The aneurysm can also cause a loss of consistency, which can be fatal. The aneurysm can also cause a loss of reliability, which can be fatal. The aneurysm can also cause a loss of trustworthiness, which can be fatal. The aneurysm can also cause a loss of integrity, which can be fatal. The aneurysm can also cause a loss of honesty, which can be fatal. The aneurysm can also cause a loss of fairness, which can be fatal. The aneurysm can also cause a loss of justice, which can be fatal. The aneurysm can also cause a loss of equity, which can be fatal. The aneurysm can also cause a loss of balance, which can be fatal. The aneurysm can also cause a loss of harmony, which can be fatal. The aneurysm can also cause a loss of peace, which can be fatal. The aneurysm can also cause a loss of tranquility, which can be fatal. The aneurysm can also cause a loss of calmness, which can be fatal. The aneurysm can also cause a loss of serenity, which can be fatal. The aneurysm can also cause a loss of contentment, which can be fatal. The aneurysm can also cause a loss of satisfaction, which can be fatal. The aneurysm can also cause a loss of fulfillment, which can be fatal. The aneurysm can also cause a loss of happiness, which can be fatal. The aneurysm can also cause a loss of joy, which can be fatal. The aneurysm can also cause a loss of pleasure, which can be fatal. The aneurysm can also cause a loss of delight, which can be fatal. The aneurysm can also cause a loss of ecstasy, which can be fatal. The aneurysm can also cause a loss of bliss, which can be fatal. The aneurysm can also cause a loss of happiness, which can be fatal. The aneurysm can also cause a loss of joy, which can be fatal. The aneurysm can also cause a loss of pleasure, which can be fatal. The aneurysm can also cause a loss of delight, which can be fatal. The aneurysm can also cause a loss of ecstasy, which can be fatal. The aneurysm can also cause a loss of bliss, which can be fatal.

## How are brain aneurysms managed?

**Managing personal and family life**

ADPKD can interfere with normal activities, such as working, family life and work. You can have a significant impact on your life.



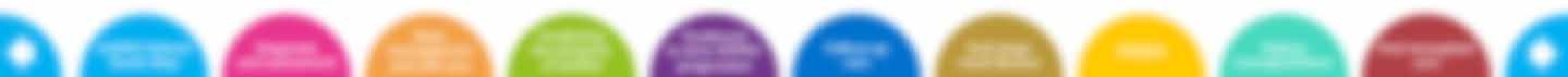
### Brain

Brain (or 'intracranial') aneurysms occur in around one in 10 people with ADPKD, which is around three- to four-times more common than in the general public.

An aneurysm is a swelling, or bulge, in an artery. It is caused by a weakness in the wall of the artery. People with ADPKD may be at higher risk of aneurysms in large blood vessels such as the aorta (which receives blood from the left ventricle of the heart and which branches to supply the head, neck and arms) and the coronary arteries (which supply the heart itself with blood). However, these are considered rare.

Brain aneurysms do not usually cause any symptoms. In a small number of cases the aneurysm can rupture, which can be disabling or even fatal. Aneurysms are no more likely to rupture in patients with ADPKD than in other people. Generally, research suggests that each patient with an aneurysm that is conservatively treated has a 4 in 1000 (0.4%) risk of a rupture each year.

[How are brain aneurysms managed?](#)





This section outlines how ADPKD can affect the health of patients.

ADPKD affects the structure and function of the parts of the body that filter and clean the blood.

ADPKD affects different people in different ways. An important consideration for all is blood pressure. It is also important to remember that most people have to take medicine, manage diet and look for other ways to help. Although the disease can cause a significant effect on the body, you can still enjoy the things you love and live a long and healthy life.

For more information, visit our website [www.adpkd.org.uk](#) or call our helpline on 0800 011 2222.

## Kidneys

ADPKD is a type of [autosomal dominant disease](#). This means that you only need one copy of the faulty gene to get the disease. If you have one faulty gene, you can pass it on to your children and the chance is 50% that you will pass it on. If you have two faulty genes, you will have the disease and you will pass it on to your children. You can also have the disease and not know it. This is called [carrier status](#).

If you have ADPKD, you can help to control your blood pressure by taking medicine. You can also help to control your blood pressure by taking medicine.

## Living with the disease in everyday life

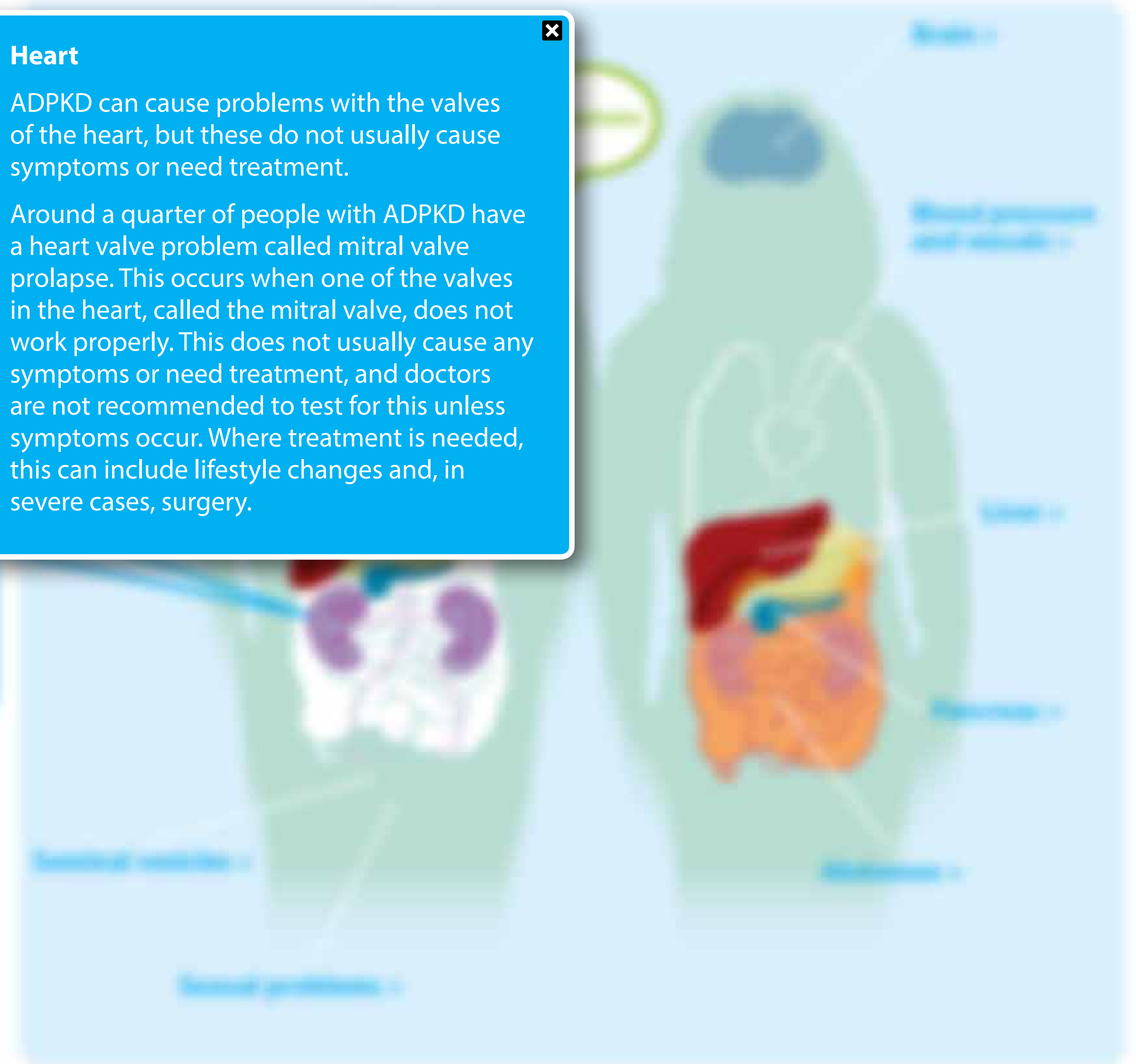
### Managing personal and family life

ADPKD can interfere with normal activities, such as working, family life and work. You can have a good quality of life if you manage your condition well.

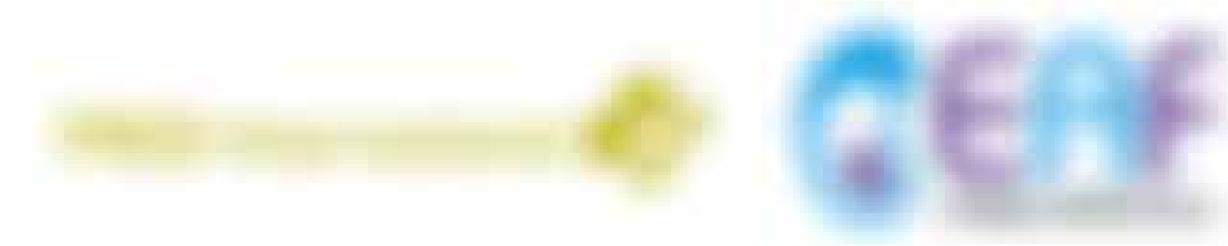
## Heart

ADPKD can cause problems with the valves of the heart, but these do not usually cause symptoms or need treatment.

Around a quarter of people with ADPKD have a heart valve problem called mitral valve prolapse. This occurs when one of the valves in the heart, called the mitral valve, does not work properly. This does not usually cause any symptoms or need treatment, and doctors are not recommended to test for this unless symptoms occur. Where treatment is needed, this can include lifestyle changes and, in severe cases, surgery.



# Understanding ADPKD



This section outlines how ADPKD can affect the health of patients.

ADPKD affects the kidneys and sometimes other parts of the body, such as the liver.

ADPKD affects different people in different ways. An important consideration for all is blood pressure. It is also important to remember that much can be done to help reduce, manage and slow the effects of ADPKD, although the disease can have a significant effect on life. It does not mean that people with the disease cannot live happy, long and productive lives.

[More information](#) website provides more information about the effects of ADPKD.

## Kidneys

ADPKD is a type of [autosomal dominant disease](#) - this means that you only need one copy of the faulty gene to develop the disease. If you have ADPKD, you will usually have two copies of the faulty gene, one from each parent. You can usually only get the disease if both your parents have a copy of the faulty gene. However, in some cases, you can develop the disease if only one of your parents has a copy of the faulty gene.

If you have ADPKD, you can help to reduce the risk of kidney failure by following the advice of your doctor through [regular blood pressure checks](#).

## Living with the disease in pregnancy

### Reducing personal and family life

ADPKD can interfere with normal activities, such as working, family life and work. You can have a significant [impact on your life](#).



## Blood pressure and vessels

Most people with ADPKD develop high blood pressure, which increases the risk of cardiovascular disease (such as heart attacks and strokes). Controlling high blood pressure is therefore very important.

High blood pressure usually does not cause symptoms. However, if the blood pressure becomes very high it can cause symptoms such as persistent headache, blurred or double vision, nosebleeds and shortness of breath. You should contact your doctor if you experience these symptoms. Your blood pressure should be checked from time to time.

Rarely, people with ADPKD may also have a higher risk of aneurysms in large blood vessels.



This section outlines how ADPKD can affect the health of patients.

ADPKD affects the kidneys and sometimes other parts of the body, including the brain.

ADPKD affects different people in different ways. An individual's experience of the effects of ADPKD varies. It is also important to remember that most people have to take action, manage and live with the effects of ADPKD. Although the disease can have a significant effect on life, it does not mean that people with the disease cannot live happy, long and productive lives.

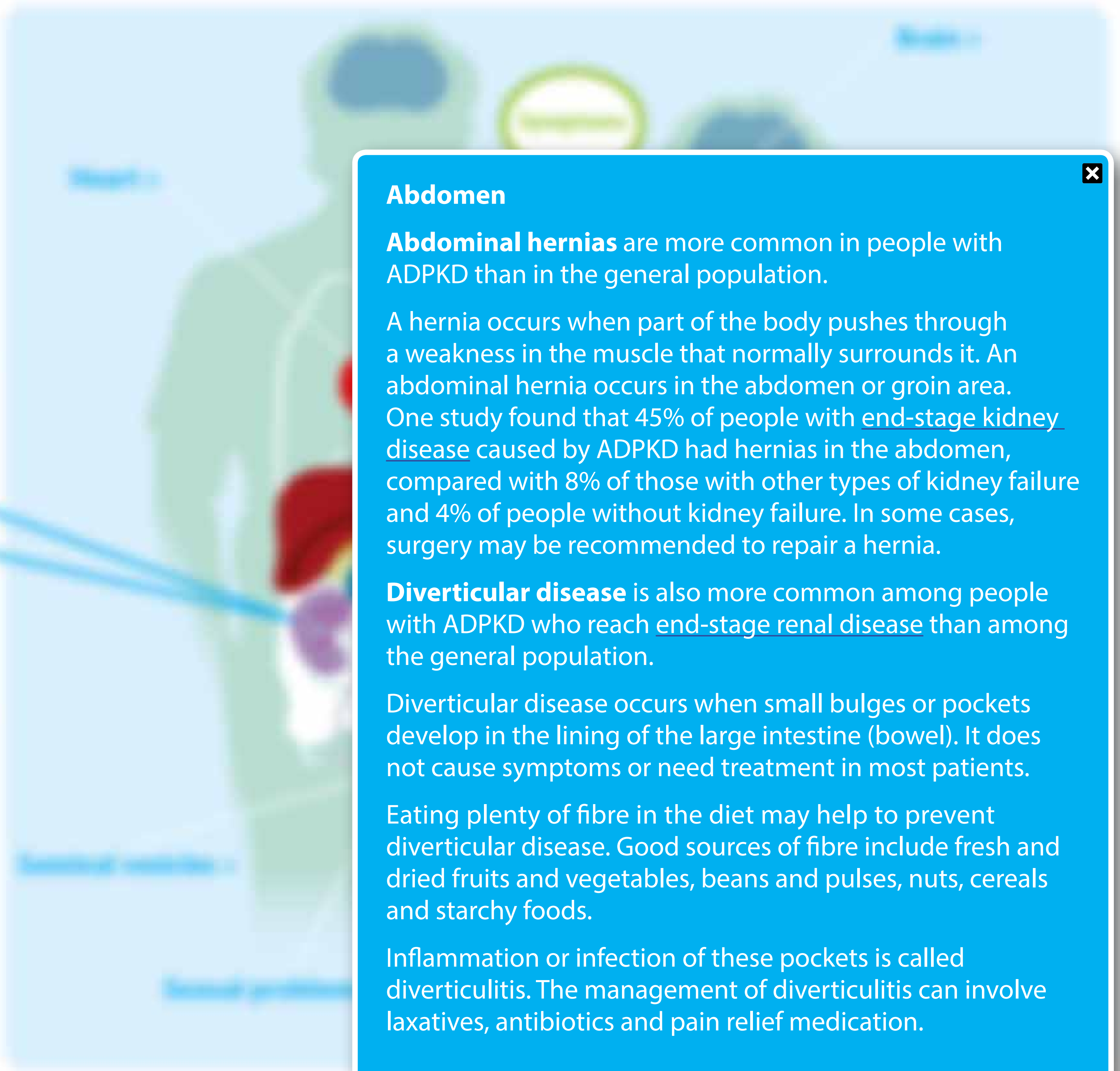
[More information](#) website provides more information about the effects of ADPKD.

**Kidneys**  
ADPKD causes **fluid-filled cysts** to develop in the kidneys. These cysts can grow and multiply over time. If the cysts grow too large, they can eventually cause the kidneys and the abdomen to swell. Cysts can eventually stop the kidneys from working properly and sometimes they can lead to kidney failure. [More information](#) website provides more information about the effects of ADPKD.

For more ADPKD information, please visit [www.geerf.org.uk](#) for the management of the disease through [renal replacement therapy](#).

## Managing personal and family life

ADPKD can sometimes affect personal and family life. It is important to talk to your doctor about the effects of ADPKD. You can find [more information](#) on the website.



**Abdomen**

**Abdominal hernias** are more common in people with ADPKD than in the general population.

A hernia occurs when part of the body pushes through a weakness in the muscle that normally surrounds it. An abdominal hernia occurs in the abdomen or groin area. One study found that 45% of people with end-stage kidney disease caused by ADPKD had hernias in the abdomen, compared with 8% of those with other types of kidney failure and 4% of people without kidney failure. In some cases, surgery may be recommended to repair a hernia.

**Diverticular disease** is also more common among people with ADPKD who reach end-stage renal disease than among the general population.

Diverticular disease occurs when small bulges or pockets develop in the lining of the large intestine (bowel). It does not cause symptoms or need treatment in most patients.

Eating plenty of fibre in the diet may help to prevent diverticular disease. Good sources of fibre include fresh and dried fruits and vegetables, beans and pulses, nuts, cereals and starchy foods.

Inflammation or infection of these pockets is called diverticulitis. The management of diverticulitis can involve laxatives, antibiotics and pain relief medication.



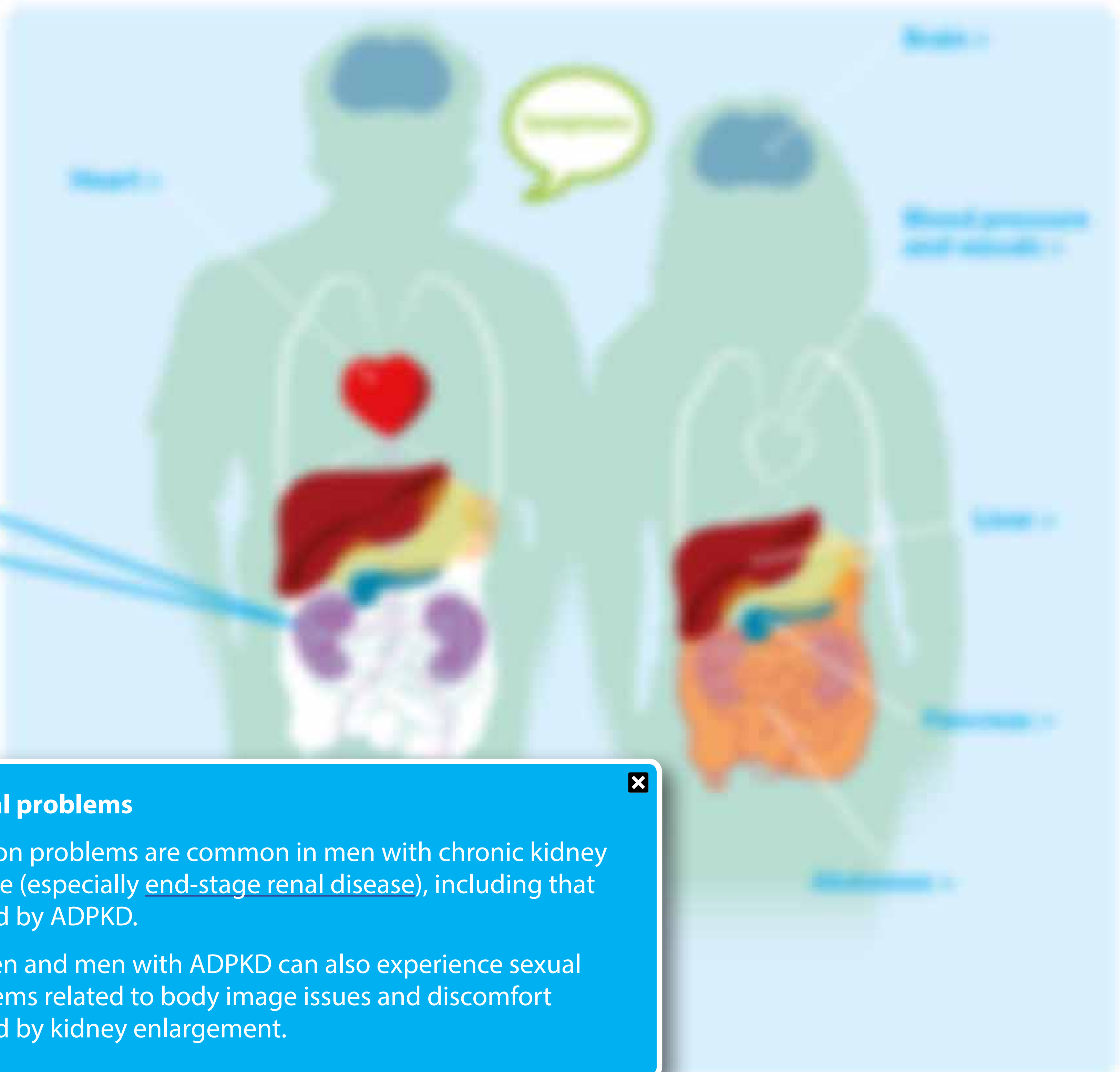
# Understanding ADPKD

This section outlines how ADPKD can affect the health of patients.

ADPKD affects the kidneys and sometimes other parts of the body. It can also affect:

ADPKD affects different people in different ways. It is important to understand that much can be done to help reduce, manage and ease the effects of ADPKD. Although the disease can have a significant effect on life, it does not mean that people with the disease cannot live happy, long and productive lives.

For more information, visit our [patient resources](#) page for information about the effects of ADPKD.



**Kidneys**  
ADPKD is a type of [autosomal dominant disease](#) - that means you only need one copy of the gene to get the disease. If the gene you have gets very large then you can eventually cause the kidneys and the abdomen to swell. You can eventually stop the kidney swelling, improve and sometimes stop or delay kidney failure, or [live longer with kidney failure](#) with the right care and other [combinations of therapies](#).  
If you have ADPKD, you can help to control your kidney health by managing the disease through [certain lifestyle changes](#).

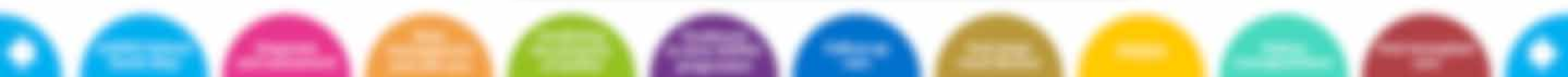
## How can the kidneys be supported?

**Maximising personal and family life**  
ADPKD can interfere with normal activities, such as work, family life and sport. You can have a significant [positive impact](#) on your life.

**Sexual problems**

Erection problems are common in men with chronic kidney disease (especially end-stage renal disease), including that caused by ADPKD.

Women and men with ADPKD can also experience sexual problems related to body image issues and discomfort caused by kidney enlargement.



This section explains some principles of good ADPKD care that patients and carers should expect.

ADPKD is a chronic disease which currently cannot be cured. To help limit its effects, patients should have access to lifelong care that involves:

- 1) A comprehensive assessment that takes into account the effects and [complications](#) that ADPKD can cause throughout the body, the likely course of the disease (i.e. [prognosis](#)) and the impact of the disease on the patient and the family.
- 2) Access to treatment (as clinically appropriate) to relieve [symptoms](#), manage [complications](#), preserve kidney function, lower the risk of [cardiovascular](#) disease, and maintain patients' quality of life.
- 3) Information and support to help patients and their families with recommended [self-care](#) measures and to deal with the impact of the condition.

ADPKD care should be [patient-centred](#) > and involve a range of specialist healthcare professionals, according to each patient's individual needs. Specialist care should start as soon as possible after diagnosis.

 [What does 'patient-centred' mean? >](#)



## Who is involved in the healthcare team?

All patients with ADPKD should have access to a [nephrologist](#) > who understands ADPKD and the various ways it affects patients and families. This may be a nephrologist in a hospital or clinic.

In certain situations, referral to a nephrologist who specialises in ADPKD may be helpful to provide particular types of care, such as predicting the disease [prognosis](#) and helping to slow this where possible, managing [complications](#) and prescribing specific medication to slow ADPKD progression, and to provide opportunities for patients to participate in [research](#).

Patients may also need access to various types of doctors and healthcare professionals with expertise in ADPKD. This is sometimes called 'multidisciplinary' care – the figure shows the healthcare professionals who may be involved.

 [How is ADPKD care organised? >](#)

 [What is the European Reference Network for Rare Kidney Diseases? >](#)

## Who's who in ADPKD care

### Diagnosis and assessment

- Geneticist**
  - Genetic testing and counselling
- Radiologist**
  - Detailed imaging (kidney, liver, etc.)
  - Brain aneurysm

### Nephrology

- Nephrologist**
  - Adult or paediatric
  - Initial assessment
  - Follow-up care
  - End stage renal disease management
- Prognosis
  - Complications
  - Medication to slow ADPKD progression
  - Research
- ADPKD specialist**
  - Potential role in some aspects / complications and research co-ordination
  - According to services

### General and follow-up care

- Primary care physician** (GP or family doctor)
  - Follow-up / shared care
  - Family planning
- Paediatrician**
  - Care of infants, children and young people
- Psychologist / psychiatrist**
  - Psychological effects (e.g. depression, anxiety)
- Dietician**
  - Dietary education
- Social services**
  - Impact on daily life, finances, family, etc

### Manifestations / complications

- Cardiologist**
  - Heart complications
- Hepatologist**
  - Liver cyst complications
- Liver surgeon**
  - Severe liver complications
- Transplant surgeon**
  - Kidney transplantation
  - Liver transplantation
- Pain team**
  - Chronic pain
  - Anaesthetist / pain specialist
  - Physiotherapist
  - Psychologist
- Urologist**
  - Kidney stones
- Neurosurgeon**
  - Aneurysms
- Obstetrics / gynaecology**
  - Prenatal advice
  - Pregnancy complications

This section explains some principles of good ADPKD care that patients and carers should expect.

ADPKD is a chronic disease which currently cannot be cured. It may lead to kidney failure, which may require dialysis or kidney transplantation.

It is important to understand that ADPKD can progress differently and unpredictably. Some people may have mild symptoms, while others may have severe symptoms. The rate of progression varies between individuals and is influenced by factors such as the number of kidneys affected and the presence of other conditions.

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ADPKD can be treated with medication to help control blood pressure and reduce the risk of kidney failure. Some people may also benefit from surgery to remove cysts or to reduce the size of the kidneys.

## Who is involved in the healthcare team?

Patients with ADPKD should have access to a multidisciplinary healthcare team. This team should include a nephrologist, a dietitian, a social worker, a psychologist, and a patient education specialist. The team should work together to provide comprehensive care.

A patient education specialist is a healthcare professional who provides information and support to patients and their families. They help patients understand their condition, manage their symptoms, and make decisions about their care.

**Deciding about me**

*'No decision about me, without me!'*

*Daniel, Spain*

## What is the best patient-centred care?

## Who is who in ADPKD care?



This section explains some principles of good ADPKD care that patients and carers should expect.

ADPKD is a chronic disease which currently cannot be cured. It may lead to kidney failure, which may require dialysis or kidney transplantation.

It is important to understand that ADPKD is a progressive disease and **genetics** play a role in ADPKD. It can be passed through the family. The rate of progression of the disease can **improve** with the support of the patient and the family.

It is important to understand that ADPKD is a chronic **condition** which **management** involves a long-term, multi-disciplinary approach. It is important to understand that ADPKD is a chronic condition which **management** involves a long-term, multi-disciplinary approach.

Information and support is available for patients and their families with ADPKD. **ADPKD** resources are available with the support of the patient.

## Who is involved in the healthcare team?

Patients with ADPKD should have access to **genetics** and other specialists. ADPKD is a chronic condition which affects patients and families. The care team is made up of a range of professionals.

A range of specialists should be involved in the care of patients with ADPKD. It is important to understand that ADPKD is a chronic condition which **management** involves a long-term, multi-disciplinary approach. It is important to understand that ADPKD is a chronic condition which **management** involves a long-term, multi-disciplinary approach.

Patients may also need to work with a range of other professionals. It is important to understand that ADPKD is a chronic condition which **management** involves a long-term, multi-disciplinary approach. It is important to understand that ADPKD is a chronic condition which **management** involves a long-term, multi-disciplinary approach.

**What does 'patient-centred' mean?** ✕

Patients should be placed at the centre of their care and treatment journey. Patients have crucial roles in managing their own ADPKD throughout their lives. Patients and families are given the knowledge and opportunity to act as fully informed partners in making decisions about their own care and about healthcare policies, services and research related to ADPKD.



# Principles of ADPKD care

This section explains some principles of good ADPKD care

ADPKD is a chronic disease which currently cannot be cured. To help limit its effects, patients should have access to lifelong specialist medical care.

UK comprehensive assessment that takes into account the effects and complications that ADPKD can cause throughout the body, the likely course of the disease for [prognosis](#) and the impact of the disease on the patient and the family.

Efforts to improve an already compromised or at-risk [cardiovascular](#) health, [cardiovascular](#) disease and other factors, such as the risk of [hypertension](#), diabetes, and maintain optimum quality of life.

Information and support to help patients and their families with [depression](#) and [anxiety](#) related to the disease and with the impact of the condition.

ADPKD can describe [quality of life](#) - a wide range of general health and well-being, according to each patient's individual needs. Specific care should not be seen as possible after diagnosis.

## What does patient-centred mean?



- 1. How is ADPKD care organised?
- 2. What is the impact of ADPKD on the patient's quality of life?

**Nephrologist** ✕

A nephrologist is a doctor who specialises in diseases of the kidney.

## Who's who in ADPKD care

Diagnosis and assessment	Monitoring	Medical and lifestyle advice
<ul style="list-style-type: none"><li>Primary care</li><li>Specialist care</li></ul>	<ul style="list-style-type: none"><li>Primary care</li><li>Specialist care</li><li>Specialist care</li><li>Specialist care</li></ul>	<ul style="list-style-type: none"><li>Primary care</li><li>Specialist care</li><li>Specialist care</li></ul>
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This section explains some principles of good ADPKD care that patients and carers should expect.

ADPKD is a chronic disease which currently cannot be cured. It may lead to kidney failure, which may require dialysis or kidney transplantation.

It is important to understand that ADPKD is a genetic disease and you can pass it on to your children. It is important to understand that ADPKD is a genetic disease and you can pass it on to your children.

ADPKD can be treated with various medicines. It is important to understand that ADPKD is a genetic disease and you can pass it on to your children.

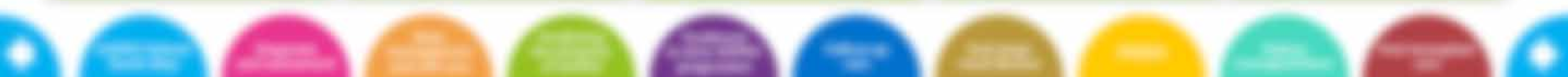
Who is involved in the healthcare team?  
Patients with ADPKD should have a multidisciplinary team approach to their care. This may include a nephrologist, dietitian, nurse, pharmacist, and other healthcare professionals.

### How is ADPKD care organised?

ADPKD care can involve various medical specialists and healthcare professionals. The organisation and co-ordination of these services varies between countries and regions.

ADPKD care is normally led by a nephrologist. In some situations, referral to a nephrologist with specialist expertise in ADPKD may be helpful. A team approach where all specialists in ADPKD care work in the same centre or clinic is considered beneficial. This is often not available in practice, although most university hospitals should be able to provide most of the services that may be needed. Where a local nephrologist or hospital cannot offer all services necessary, patients may be referred to other specialist ADPKD services.

### Who is who in ADPKD care



This section explains some principles of good ADPKD care that patients and carers should expect.

ADPKD is a chronic disease which currently cannot be cured. It may lead to kidney failure, which may require dialysis or kidney transplantation.

It is important to understand that ADPKD can progress differently and at different rates. Some people may have symptoms for many years before the kidneys are affected. Some people may have symptoms for many years before the kidneys are affected.

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ADPKD can be treated with [medicines](#) which can help to slow down the growth of the kidneys. Some people may also need to take medicines to help with the symptoms of the disease.

## What does a good ADPKD care mean?

## Who is who in ADPKD care

Diagnosis and assessment		Monitoring		Treatment		Support and education	
Genetic testing	Ultrasound	Blood tests	Imaging	Medicines	Transplantation	Education	Support groups
Genetic counselling	Genetic testing	Blood pressure	Genetic testing	Medicines	Transplantation	Education	Support groups
Genetic testing	Genetic testing	Blood pressure	Genetic testing	Medicines	Transplantation	Education	Support groups

### What is the European Reference Network for Rare Kidney Diseases?

The European Reference Network for Rare Kidney Diseases (ERKNet) was launched in 2017 to promote high-quality, multidisciplinary care for rare kidney diseases, including ADPKD.

The ERKNet links together expert paediatric and adult nephrology centres in many European countries. A full list of the centres is [here](#). These centres use uniform clinical guidelines and pathways, monitor the quality and outcomes of treatment, provide education for nephrologists, and support research. The network also offers 'virtual consultations' for doctors who need advice and provides links to information for patients.

There is also a European Reference Network on Rare Liver Diseases (RARE-LIVER), including polycystic liver disease.



This section explains how ADPKD is diagnosed and the initial tests and examinations that are normally performed.

Early and accurate diagnosis of ADPKD allows patients and doctors to take steps to manage the disease. Typically, ADPKD is diagnosed and assessed via a pathway like this.

## Identifying people with ADPKD

ADPKD is usually first suspected or identified in people who:

- have typical [symptoms of ADPKD](#) >
- are examined for another reason, such as high blood pressure or pregnancy
- are tested (or 'screened') for ADPKD because someone else in their family has the disease.

ADPKD is often first suspected by general practitioners (family doctors).

## Diagnosis and kidney assessment

ADPKD is normally diagnosed by a [nephrologist](#) >. Important initial assessments include scans and kidney function tests.

### [Kidney scans](#) >

Kidney scans allow doctors to see and measure the cysts caused by ADPKD.

### [Kidney function tests](#) >

Tests carried out on the blood and urine show to what extent ADPKD is affecting the function of the kidneys.

### [Genetic testing](#)

ADPKD is caused by mutations in the DNA, or genes. Genetic testing to identify these mutations is not necessary for most patients, but is sometimes used in certain situations.

### [? Are there special issues for children?](#) >

Diagnosis

## Other investigations

It is important to find out if ADPKD has affected other parts of the [body](#).

Important investigations include:

- [Blood pressure](#) >
- [Liver](#) >
- [Brain aneurysm](#) >

Patients may be referred to [other types of specialist doctors and healthcare professionals](#), depending on their needs.

### [? Should other family members be checked?](#) >

### [✓ Checklist](#) >



This section explains how PKD is diagnosed and the initial tests and examinations that are normally performed.

## Diagnosis

*'The PKD Association has helped me in many ways to re-orientate after my unexpected diagnosis. Most importantly it has helped to take away my fears about the disease.'*

*Stefan, Germany*

*'I did not know I was a polycystic patient. I had a scan because at the age of 36 I had hepatitis A. The doctor told me that I had cysts on both kidneys. When I was diagnosed, I was not surprised. I suspected that there was a genetic disease in my family because all the aunts on my father's side of the family had ended up on dialysis. At the time of the diagnosis, the doctor asked me how many children I had. "I have two children" I replied. He was very surprised and said: "... and you are not in dialysis yet?" I touched wood. It's been 20 years since that day, and every day I am thankful because whatever happens I have been luckier than any expectation.'*

*Tina, Italy*

# Diagnosis and assessment

This section explains how ADPKD is diagnosed and the initial tests and examinations that are normally performed.

Key and accurate diagnosis of ADPKD allows patients and doctors to take steps to manage the disease. Typically, ADPKD is diagnosed and assessed as follows:

## Identifying people with ADPKD

ADPKD is usually first suspected in identifiable people who:

- have a family history of ADPKD;
- are themselves another person with a high blood pressure or pregnancy
- are themselves suspected to have ADPKD because someone else in their family has the disease.

ADPKD is often first suspected in people who:

## Diagnosis and kidney assessment

ADPKD is normally diagnosed by [ultrasound](#).  
Examination under assessment include cysts and kidney function tests.

### Genetic testing

Genetic tests allow doctors to see and measure the gene causing ADPKD.

### Imaging techniques

Genetic testing can be used to see if you have ADPKD or if you have a family history of ADPKD.

## Other investigations

In response to a diagnosis of ADPKD you should also have other tests of the body.

Investigation investigations include:

### Genetic testing

### Ultrasound

### Genetic testing

Doctors may be able to identify other signs of genetic testing and conditions associated with ADPKD.

### Genetic testing

### Ultrasound

### Genetic testing

### Are there special issues for children?

ADPKD is typically identified in adults, but it may also be diagnosed in children. Infants and children with kidney cysts should be referred to a paediatric nephrologist. ADPKD can be difficult to diagnose in children using imaging alone. A genetic test is sometimes used to confirm the diagnosis if imaging results are unclear.



# Diagnosis and assessment

This section explains how ADPKD is diagnosed and the initial tests and examinations that are normally performed.

Key and accurate diagnosis of ADPKD allows patients and doctors to take steps to manage the disease. Typically, ADPKD is diagnosed and assessed as follows:

## Identifying people with ADPKD

ADPKD is usually first diagnosed in people who:

- have a family history of the condition
- are themselves part of a family with a history of the condition
- are themselves affected by the condition

ADPKD is often first diagnosed in people who have a family history of the condition.

### Symptoms of ADPKD

In adults these include pain in the abdomen, side or back, or blood in their urine. In children these include bedwetting and urinary tract infections.

## Diagnosis and kidney assessment

ADPKD is usually diagnosed by a [general practitioner](#) or [nephrologist](#).

- A [general practitioner](#) or [nephrologist](#) will usually ask you about your family history and any symptoms you have.
- They will also ask you about your blood pressure and whether you have any other health problems.
- They will also ask you about your diet and whether you smoke or drink alcohol.

**Family history**  
ADPKD is usually first diagnosed in people who have a family history of the condition. A general practitioner or nephrologist will usually ask you about your family history and whether you have any other health problems.

**General practitioner or nephrologist**



## Other investigations

In addition to asking you about your family history and any symptoms you have, a general practitioner or nephrologist will usually ask you about your blood pressure and whether you have any other health problems.

- [Blood pressure](#)
- [Blood tests](#)
- [Ultrasound](#)

There may be other tests that you will need to have, such as a [renal biopsy](#) or [kidney scan](#), depending on your case.

**Other tests**



# Diagnosis and assessment

This section explains how CKPD is diagnosed and the initial tests and examinations that are normally performed.

Key and accurate diagnosis of CKPD allows patients and doctors to take steps to manage the disease. Typically, CKPD is diagnosed and assessed as follows:

**Identifying people with CKPD**

CKPD is usually first suspected in individuals who:

- have a family [history of CKPD](#)
- are experiencing another issue, such as high blood pressure or pregnancy
- are related or connected to CKPD but with someone else in their family has the disease

CKPD is also first suspected in general practitioners family history.

**Diagnosis and kidney assessment**

CKPD is normally diagnosed through a series of tests and examinations which include:

- [Blood tests](#)
- [Urine tests](#)
- [Kidney scans](#)

**Managing CKPD**

CKPD is usually managed through a series of tests and examinations which include:

- [Blood tests](#)
- [Urine tests](#)
- [Kidney scans](#)

**Nephrologist** ✕

A nephrologist is a doctor who specialises in diseases of the kidney.



This section explains how ADPKD is diagnosed and the initial tests and examinations that are normally performed.

Key and accurate diagnosis of ADPKD allows patients and doctors to take steps to manage the disease. Typically, ADPKD is diagnosed and assessed as follows:

## Identifying people with ADPKD

ADPKD is usually first suspected in identifiable people who:

- have a family history of ADPKD;
- are themselves another person with a high blood pressure or pregnancy;
- are themselves suspected to have ADPKD because someone else in their family has the disease.

ADPKD is also first suspected in people with enlarged kidneys.

## Diagnosis and kidney assessment

ADPKD is usually diagnosed by [ultrasound](#).  
Further kidney assessment includes renal and kidney function tests.

### Kidney scans

Kidney scans allow doctors to see and measure the cysts caused by ADPKD.

An ultrasound scan is usually used first. This is a simple imaging method that most clinics can do routinely.

Magnetic resonance imaging (MRI) may also be used. This is a more detailed, accurate and expensive scan performed in a hospital or clinic radiology department.

## Other investigations

It is important to check for ADPKD in affected other parts of the body.

Further investigations include:

- [blood pressure](#) monitoring;
- [renal function](#) tests;
- [renal ultrasound](#) scans;
- [magnetic resonance imaging \(MRI\)](#) scans.

For further information:





This section explains how CKPD is diagnosed and the initial tests and examinations that are normally performed.

Key and recent changes of CKPD are shown in the table below. For more information on the latest changes, please refer to the CKPD diagnosis and assessment in updating the site.

## Kidney function tests

Kidney function tests include the measurement of the level of creatinine in a sample of blood. Creatinine is a substance produced naturally by the body. Doctors use the creatinine level to estimate the glomerular filtration rate (eGFR). eGFR is an important measure of how well the kidneys are doing their job in removing waste products and excess fluid from the blood. The part of the kidney that does this filtering job is the glomerulus, hence the name 'glomerular filtration rate'. The eGFR falls as kidney function gets worse.

There are five stages of chronic kidney disease, defined by the eGFR.

Stage	Description of kidney function change	eGFR level (ml/min/1.73m <sup>2</sup> )
1	Normal kidney function	90 or higher
2	Mild loss	60–89
3a	Mild to moderate loss	45–59
3b	Moderate to severe loss	30–44
4	Severe loss	15–29
5	Kidney failure or <a href="#">end-stage renal disease</a>	Less than 15

The urine is also tested for the presence of blood (called haematuria) or protein (proteinuria), both of which can indicate kidney damage.

# Diagnosis and assessment

This section explains how KCPND is diagnosed and the initial tests and examinations that are normally performed.

Early and accurate diagnosis of KCPND allows patients and doctors to take steps to manage the disease. Typically, KCPND is diagnosed and assessed in a step-by-step way.

### Identifying people with KCPND

KCPND is usually first suspected in individuals who:

- have a family history of KCPND
- are members of another ethnic group with high prevalence of KCPND
- are related to someone with KCPND but who does not have the disease

KCPND is often first suspected in people with proteinuria.

### Diagnosis and kidney assessment

KCPND is normally diagnosed by a **general practitioner**.  
Diagnosis and assessment include tests and kidney function tests.

#### Blood tests

Blood tests allow doctors to see whether there is proteinuria (KCPND).

#### Urine tests

Urine is passed out of the bladder and some of it is analysed. KCPND is often first detected by finding protein in the urine.

#### Health history

KCPND is usually the result of the DM, a genetic disease, leading to kidney disease. It can also be caused by other conditions, but is usually not hereditary.

- Proteinuria**

### Other investigations

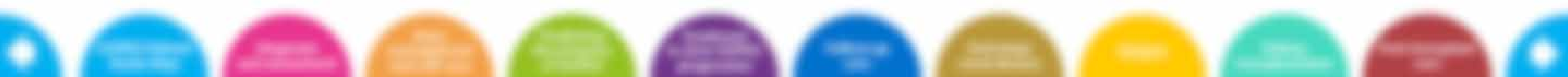
It is important to find out if KCPND has affected other parts of the body.

#### Other tests

- Health check**
- Health check**
- Health check**

**Blood pressure**

It is very important to identify and control high blood pressure, to reduce the risk of cardiovascular disease (such as heart attacks and strokes).



# Diagnosis and assessment

This section explains how ADPKD is diagnosed and the initial tests and examinations that are normally performed.

Help and advice regarding ADPKD shows patients and doctors in clear steps to manage the disease. Typically, ADPKD is diagnosed and assessed as follows:

## Identifying people with ADPKD

ADPKD is usually first suspected in identifiable people who:

- have a family [history of ADPKD](#)
- are themselves another person with a high blood pressure or pregnancy
- are related to someone for whom there are some signs in their family that the disease

ADPKD is often first suspected in general practitioners family history.

## Diagnosis and kidney assessment

ADPKD is normally diagnosed by [ultrasound](#).  
Diagnosis and assessment include tests and kidney function tests.

### Ultrasound

Ultrasound allows doctors to see and measure the size of the kidneys.

### Renal function tests

These are carried out on the blood and urine to check kidney function. ADPKD is affecting the function of the kidney.

### Genetic testing

ADPKD is caused by mutations in the PKD1 or PKD2 genes. Genetic testing is usually done to confirm a diagnosis of ADPKD. It is usually done in people with a family history of ADPKD.

### Genetic testing

ADPKD is caused by mutations in the PKD1 or PKD2 genes. Genetic testing is usually done to confirm a diagnosis of ADPKD. It is usually done in people with a family history of ADPKD.

## Other investigations

It is important to check for ADPKD-related complications of the [liver](#).

Investigations include:

### Ultrasound

### Liver

All patients with ADPKD should be assessed for liver cysts using imaging (usually by ultrasound at first). People with liver cyst complications should be referred to a hepatologist (a doctor who specialises in diseases of the liver), as necessary.



This section explains how ADPKD is diagnosed and the initial tests and examinations that are normally performed.

Key and recent changes to ADPKD clinical practice and research in this topic are covered in the document 'Specialty ADPKD in Diagnosis and Assessment in Supporting the NHS'.

## Identifying people with ADPKD

ADPKD is usually first diagnosed in adulthood, usually after:

- first signs of [hypertension](#)
- an increasingly swollen kidney, with or without discomfort or pain
- an ultrasound scan for ADPKD because someone else in their family has the disease

ADPKD is also first diagnosed in young children with kidney stones.



## Should other family members be checked?

ADPKD is an inherited, genetic disease. People diagnosed with ADPKD should be offered counselling about the benefits and risks of informing other family members and offering them the opportunity to be checked for the disease. This screening can be done by a general practitioner, with referral to a nephrologist to confirm the diagnosis.

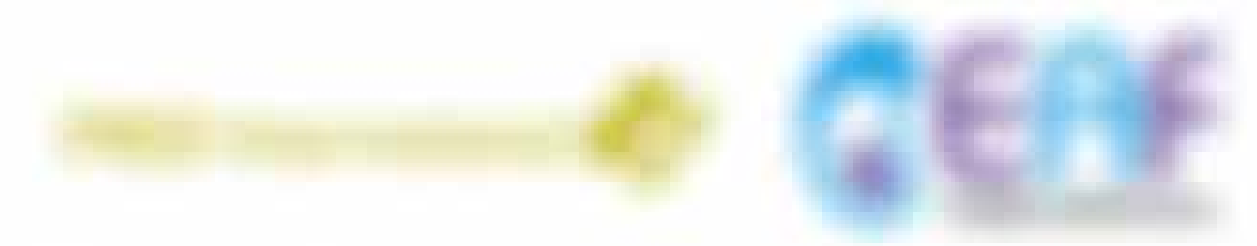
Checking for ADPKD allows family members with the disease to get the advice, treatment and support they may need, as early as possible. However, a diagnosis of ADPKD can have important lifelong effects, including psychological and financial consequences. Counselling can help in balancing these advantages and disadvantages when making decisions.

Routine screening for ADPKD is not recommended for children (under 18 years) who do not yet have any signs or symptoms of the disease, even if one or both parents has ADPKD. This is because of the negative psychological and financial consequences that the diagnosis may have. Children who do have signs and symptoms of ADPKD should of course be checked and referred to a paediatric nephrologist if they have cysts.

Pre-implantation genetic diagnosis (PGD) can be used to test for genetic mutations linked with ADPKD in embryos created by in vitro fertilisation. This is discussed more in the section on Family planning.



# Diagnosis and assessment



This section explains how ACPH is diagnosed and the initial tests and examinations that are normally performed.

Key and accurate diagnosis of ACPH allows patients and doctors to take steps to manage the disease. Typically, ACPH is diagnosed and assessed as follows:

### Identifying people with ACPH

ACPH is usually first suspected in identified people who:

- have a high [blood pressure](#)
- are members of another ethnic group with a high blood pressure prevalence
- are members of a family with ACPH (see also [genetics](#))

ACPH is often first suspected in people with a family history of the disease.

### Diagnosis and kidney assessment

ACPH is usually diagnosed by [blood pressure](#) measurements. Blood pressure measurements include systolic and diastolic blood pressure.

#### Initial tests

Initial tests often include blood tests to check kidney function and cholesterol levels.

#### Further tests

Further tests often include kidney function tests and kidney ultrasound.

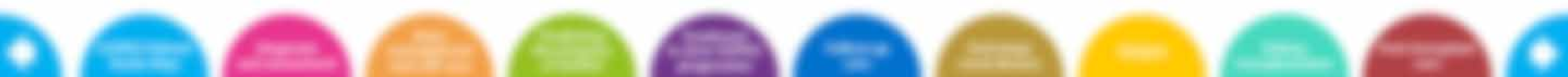
### Other investigations

It is important to check whether ACPH has affected other parts of the body.

### Brain aneurysm

Routine screening of all patients for [brain aneurysms](#) is not recommended because most aneurysms have a low risk of rupture and because [surgery](#) to prevent aneurysms from rupturing carries risks.

However, screening is recommended in patients with long life expectancy who 1) have a family history of aneurysms or bleeding in the brain, 2) have had a previous rupture, 3) are members of high-risk professions (e.g. airline pilots) and 4) are anxious about aneurysms even after receiving adequate information.



## Checklist: Diagnosis and assessment



**Kidney scan**

- Ultrasound performed and results explained
- MRI performed (where available and necessary) and results explained

**Kidney function tests**

- Kidney function tests (blood and urine) performed and result explained
- Chronic kidney disease stage explained

**Other investigations and issues**

- Blood pressure: tested and results discussed
- Blood lipid (cholesterol) tests
- Review and advice on diet, smoking and lifestyle
- Liver: liver scan performed (when appropriate) and results discussed
- Brain: possible need for screening discussed
- Genetic testing: genetic testing discussed where relevant

**Family screening**

- Counselling provided on suitability and implications of screening other family members and available options for this service

Notes and questions you would like to ask your healthcare team

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# Basic management and self care

This section explains the steps patients can take, together with their healthcare team, to help protect their kidney function and reduce the risk of cardiovascular disease.

## Lifestyle and diet

Few specific diet or lifestyle measures have yet been proven to prevent or slow the development of cysts in people with ADPKD. However, if you have ADPKD you can do many things that may help protect your kidney function and lower the risk of high blood pressure and cardiovascular disease.

Many of these are general healthy lifestyle measures also recommended for everyone else. They include:

- Drink more water to stay fully hydrated, which may protect kidney function in ADPKD.
- **Stop smoking.** >
- Maintain a healthy **body weight and do some form of regular exercise.** >
- Eat a healthy **diet.** >
- Drink less **caffeine.** > (e.g. in coffee or cola drinks) and less alcohol.

You might be advised to make other lifestyle and diet changes if you reach [end-stage renal disease](#).

## Focus on high blood pressure

Controlling long-term high blood pressure (hypertension) is very important because high blood pressure increases the risk of cardiovascular disease, such as heart attack and stroke. Controlling high blood pressure may also help to slow the growth of kidney cysts in some people with ADPKD. In people with [brain aneurysms](#), controlling high blood pressure (and stopping smoking) can reduce the risk that the aneurysm will burst.

## How can blood pressure be controlled?

If you have high blood pressure, the lifestyle and diet measures above are particularly important to reduce the risk of cardiovascular disease.

Doctors can also prescribe various [medicines](#) > to help control high blood pressure.

Regular [blood pressure checks](#) > are important to make sure treatment is working.

 [What do blood pressure readings and targets mean? >](#)

## Other risk factors

Your doctor may recommend you take other medicines to control [other risk factors](#) > for cardiovascular disease.

 [What about complementary or alternative therapies? >](#)

## Keeping it up!

Caring for your own health is very important. Maintaining a healthy lifestyle and diet, and taking prescribed medicines according to the instructions, can be difficult to maintain over long periods. Your healthcare team should be able to provide further sources of help and support locally and online. Family, friends and [patient organisations](#) can also provide valuable help and advice.

## Wellbeing, personal and family life

Patients and families can take steps to limit and deal with the effects that ADPKD can have on [wellbeing, personal and family life](#). If you have ADPKD, or are a parent of a child with ADPKD, you may wish to discuss any such problems with your healthcare team so that any necessary information, care and support can be provided.

 [Checklist >](#)

 Self care

## Self care

*'Every time I go to my doctor, I realise I'm not alone and that there are many other people in my situation coping with ADPKD. If they are managing to overcome it, so will I.'*

*Claudia, Spain*

*'I accepted it and said, "All right, let's get married. I want to have children. I'm going to start a low sodium diet. I want to lose a bit of weight. And that will benefit me because it is beneficial for my kidneys." And these plans helped me to have a sense of control in my life.'*

*Brenda, the Netherlands*

*'Now my husband with ADPKD knows he will need to drink more and he has learned to calibrate the salt intake a bit better before competitions and he is very happy with the result. He doesn't even notice the difference much, because he always used to drink a lot, and he doesn't have any problems needing the toilet at night.'*

*Flavia, Switzerland*

*'In principle, live as normally as possible! Research is likely to provide more possibilities to deal with the disease and perhaps prevent kidney failure one day.'*

*Alexander, Austria*





# Basic management and self care

This section explains the steps patients can take, together with their healthcare team, to help protect their kidney function and reduce the risk of cardiovascular disease.

## Lifestyle and diet

For people with a chronic kidney disease, there are some things you can do to help protect your kidney function and reduce the risk of cardiovascular disease.

There are some general health advice that can help reduce the risk of cardiovascular disease. This includes:

- [Lifestyle and diet](#)
- [Smoking](#)
- [Alcohol](#)
- [Exercise](#)
- [Stress](#)

### Smoking

Smoking cessation helps to reduce the risk of cardiovascular disease (i.e. coronary heart disease and stroke) and cancer. Practical help and support to stop smoking may be available.

## Other risk factors

There are other risk factors you can take other measures to control [other risk factors](#) for cardiovascular disease.

### How do I know if my blood pressure is abnormal/high?

## Keeping it up

It's important to keep up with your treatment. Stopping or changing your treatment without talking to your healthcare team can increase the risk of complications. Your healthcare team should be able to provide further advice if you are struggling to take your medicine. Family members can also provide valuable help and advice.

## Living personal and family life

It's important to live your life as normally as possible. You should talk to your healthcare team about any concerns you have. If you have any questions or need more information, call your healthcare team or the NHS helpline on 111.

### Smoking



## How can blood pressure be controlled?

There are some things you can do to help control your blood pressure. This includes:

There are some things you can do to help control your blood pressure. This includes:

There are some things you can do to help control your blood pressure. This includes:

### How do I know if my blood pressure is abnormal/high?



# Basic management and self care

This section explains the steps patients can take, together with their healthcare team, to help protect their kidney function and reduce the risk of cardiovascular disease.

## Lifestyle and diet

For people with a chronic kidney disease, it is very important to control diet, the development of salt in people with CKD. However, if you have CKD you can do many things that may help protect your kidney function and reduce the risk of high blood pressure and cardiovascular disease.

Most of these are general health advice, which means all recommendations apply to you. They include:

- Stop smoking and avoid second-hand smoke
- [Body weight and exercise](#)
- [Blood pressure](#)
- [Diet](#)
- [Diabetes](#)
- [Lifestyle advice](#)

For people with CKD, it is very important to control diet, the development of salt in people with CKD. However, if you have CKD you can do many things that may help protect your kidney function and reduce the risk of high blood pressure and cardiovascular disease.

## Focus on high blood pressure

Controlling high blood pressure is very important for people with CKD. High blood pressure increases the risk of cardiovascular disease, such as heart attack and stroke. Controlling high blood pressure may also help to slow the growth of kidney disease in some people with CKD. In people with CKD, controlling high blood pressure can help reduce the risk of cardiovascular disease.

## How can blood pressure be controlled?

Controlling blood pressure, the doctor and the patient, there are several ways to reduce the risk of cardiovascular disease.

Doctors can also prescribe some [medicines](#) to help control high blood pressure.

Some [blood pressure checks](#) are important to make sure treatment is working.

## What is blood pressure reading and target level?

## Other risk factors

There are many other risk factors that can increase the risk of cardiovascular disease.

## What is blood pressure reading and target level?

## Keeping it up

Controlling your blood pressure is an ongoing process. Keeping healthy blood pressure under control is a long-term goal. It is important to work with your healthcare team to make sure you are taking the right steps to keep your blood pressure under control.

## Personal and family life

It is important to work with your healthcare team to make sure you are taking the right steps to keep your blood pressure under control. It is also important to work with your healthcare team to make sure you are taking the right steps to keep your blood pressure under control.

**Body weight and exercise**

Maintaining a healthy body weight and regularly exercising are recommended to help prevent and control high blood pressure.

Exercise: this can include walking, gardening, dancing and all kinds of sports – although it may be sensible to avoid high contact sports to avoid trauma to the kidneys.



## Diet

### *Salt reduction*

Recent [research](#) showed that higher dietary salt intake caused greater kidney growth in patients with ADPKD. The researchers studied data from 'HALT-PKD', a clinical trial of the effect of certain blood pressure medicines on the progression of ADPKD. They concluded that moderate salt restriction (to no more than 6 g a day) is beneficial in ADPKD, but you should not remove salt from your diet entirely. You may be referred to a dietician to provide a diet plan.

The recommended salt levels are lower for children.

Age	Salt per day (sodium equivalent)
1–3 years	2 g (0.8 g)
4–6 years	3 g (1.2 g)
7–10 years	5 g (2 g)
11 years and over	5–6 g (2–2.4 g)

### *Moderate protein*

There is no good evidence that low-protein diets slow the progression of ADPKD. Adults with ADPKD are [advised](#) to eat the same, moderate amount of protein (0.75–1.0 g per kg of body weight per day) recommended for the general population. Guidelines for general chronic kidney disease care recommend that adults eat no more than 0.8 g of protein per kg of body weight daily when their [estimated glomerular filtration rate \(eGFR; see kidney function tests\)](#) falls below 30 ml/min/1.73 m<sup>2</sup>. People at risk of CKD progression are [recommended](#) to avoid a high protein intake (>1.3 g/kg/day). Any restriction on dietary protein should preferably involve education by a renal dietician and monitoring to avoid malnutrition.

### *Fibre*

Eating plenty of fibre in the diet may help to prevent [diverticular disease](#).

### *Sources*

See [Further reading](#). Some patient organisation websites provide further information about diet and ADPKD.

# Basic management and self care

This section explains the steps patients can take, together with their healthcare team, to help protect their kidney function and reduce the risk of cardiovascular disease.

## Lifestyle and diet

For people who already have kidney disease, it is important to prevent or slow the development of cysts in people with ADPKD. However, if you have ADPKD you can do many things that may help protect your kidney function and reduce the risk of high blood pressure and cardiovascular disease.

Many of these are general healthy lifestyle measures that can benefit everyone. They include:

- Drinking enough water to stay hydrated, which may protect kidney function in ADPKD.

### • [Not smoking](#)

- Not drinking alcohol
- Not smoking
- Not taking drugs

For people with high blood pressure, it is important to:

## Focus on the

Controlling your blood pressure can help to slow down the growth of kidney cysts in some people with ADPKD. In people with ADPKD, controlling high blood pressure can help to reduce the risk of cardiovascular disease.

## How can blood pressure be controlled?

For lowering blood pressure, the doctor and the patient should do particularly important to reduce the risk of cardiovascular disease.

Doctors can also prescribe special [medicines](#) to help control high blood pressure.

Regular [blood pressure checks](#) are important to make sure treatment is working.

### • [What is blood pressure reading and target level?](#)

## Other risk factors

There are many other risk factors for cardiovascular disease.

### • [What does compliance in dialysis treatment?](#)

## Keeping it up!

Keeping your own health in any condition, following healthy lifestyle advice, and taking prescribed medicines as advised, can be difficult to maintain over long periods. Your healthcare team should be able to provide further resources of help and support locally and online. Family friends and [support groups](#) can also provide valuable help and advice.

## Maintaining personal and family life

People with kidney disease often have to deal with the effects of dialysis. It can be hard to manage over long periods. Your healthcare team should be able to provide further resources of help and support locally and online. Family friends and [support groups](#) can also provide valuable help and advice.

### Caffeine

Keeping caffeine intake to a moderate level (2 cups of coffee or 4 cups of tea, per day) may be advisable for general cardiovascular health, although there is no evidence that it affects kidney cyst growth in ADPKD.



# Basic management and self care

This section explains the steps patients can take, together with their healthcare team, to help protect their kidney function and reduce the risk of cardiovascular disease.

## Lifestyle and diet

For people with chronic kidney disease, lifestyle changes can help to protect kidney function. The development of kidney disease with ADPKD is caused by your genes. ADPKD can be managed through diet and lifestyle changes to help reduce the risk of high blood pressure and cardiovascular disease.

How often you go to the toilet depends on how much you drink. This means:

- Drink water when you are thirsty
- Don't drink too much alcohol
- Don't smoke
- Don't take too much salt
- Don't take too much sugar
- Don't take too much fat
- Don't take too much protein

For more information on diet and lifestyle changes, see [Diet and lifestyle changes](#).

## Focus on high blood pressure

Controlling your blood pressure is important for people with ADPKD. High blood pressure can lead to kidney damage and cardiovascular disease. It can also lead to other health problems. Your doctor will check your blood pressure regularly and prescribe medication if necessary.

## How can blood pressure be controlled?

There are several ways to control blood pressure. These include lifestyle changes, such as eating a healthy diet and exercising regularly. Medication is also available to help control blood pressure.

For more information on blood pressure, see [Blood pressure](#).

## Other risk factors

There are other risk factors for cardiovascular disease, such as smoking, high cholesterol and diabetes. These can be managed through lifestyle changes and medication.

For more information on other risk factors, see [Other risk factors](#).

**Medicines to control high blood pressure**

Many different types of medicines (sometimes called 'antihypertensive' drugs) can be used to treat high blood pressure. Doctors consider various factors when choosing a blood pressure medicine for an individual, including the presence of other diseases.

Usually, medicines called angiotensin converting enzyme (ACE) inhibitors and angiotensin receptor blockers (ARBs) are the recommended first choices for people with ADPKD. ACE inhibitors have names ending in 'pril', such as enalapril, lisinopril, perindopril and ramipril. ARBs have names ending in 'sartan', such as candesartan, irbesartan, losartan and telmistartan.

If these are not appropriate, or if additional medicines are necessary, then other medicines called beta-blockers, diuretics or calcium channel blockers may be considered depending on the individual circumstances. Your doctor might prescribe a combination of medications to control your blood pressure.



This section explains the steps patients can take to protect their kidney function and reduce the risk of complications.

## Lifestyle and diet

For people with ADPKD, lifestyle and diet changes can help to slow down the development of cysts in people with ADPKD. You can also help to reduce the risk of complications and reduce the risk of high blood pressure and kidney disease.

There are several ways to help reduce the risk of complications. These include:

- [Lifestyle changes](#) - such as eating a healthy diet and staying active
- [Blood pressure](#) - keeping your blood pressure under control
- [Weight management](#) - staying at a healthy weight
- [Alcohol](#) - limiting your alcohol intake
- [Smoking](#) - stopping smoking

## Focus on high blood pressure

Controlling your high blood pressure is important to help slow down the development of cysts in people with ADPKD. High blood pressure can also lead to kidney damage and stroke. Controlling high blood pressure may also help to slow the growth of kidney cysts in some people with ADPKD. For more information, see [high blood pressure](#). Controlling high blood pressure can help to reduce the risk of complications.

## How can blood pressure be controlled?

Controlling high blood pressure can help to slow down the development of cysts in people with ADPKD. There are several ways to help control high blood pressure.

There are several ways to help control high blood pressure. These include:

There are several ways to help control high blood pressure. These include:

## What is blood pressure reading and target range?

### Blood pressure checks

The standard blood pressure target for people with ADPKD is a reading no higher than 140/90 mmHg. However, the target should be personalised, taking into account age and other diseases. It may help if you know your own blood pressure target and to monitor it at home. You should discuss with your doctor what action you should take if your readings are higher than your target.

You can measure your own blood pressure at home using simple electronic devices. In certain situations, you might be given a special device to continuously monitor your blood pressure for a period at home.

This can help to provide further information about your blood pressure at different times of the day. It is important to learn how to take these measurements properly and to provide your doctor with the measurements at your clinic visit.



# Basic management and self care

This section explains the steps patients can take, together with their healthcare team, to help protect their kidney function and reduce the risk of cardiovascular disease.

## Lifestyle and diet

For people with chronic kidney disease, lifestyle changes can help to protect the development of complications such as ADPKD. However, if you have ADPKD you can do more things that may help protect your kidney function and reduce the risk of long-term complications and cardiovascular disease.

Most of these are general health advice, which means they are recommended for everyone. They include:

- Stop smoking or quit if you already smoke, which may protect your heart and kidneys.

## How healthy is...

- How healthy is...
- How healthy is...
- How healthy is...
- How healthy is...

For more information on...

How can I help...

How can I help...

How can I help...

How can I help...

How can I help...

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How can I help...

## Other risk factors

There are many other risk factors that can lead to cardiovascular disease.

## What does compliance in medication management mean?

### Keeping it up!

Keeping the doctor's advice up to date is important. Stopping or changing medication without advice can be dangerous. It is important to talk to your healthcare team if you are having difficulty with your medication. Your healthcare team should be able to help you with any problems you have with your medication. They can also provide advice on how to take your medication.

## Work and family life

It is important to talk to your healthcare team if you are having difficulty with your work or family life. They can provide advice on how to manage your condition and help you to stay healthy and active.

**What do blood pressure readings and targets mean?**

Blood pressure readings are given as two numbers, followed by the units by which blood pressure is measured, known as 'mmHg' (millimetres of mercury).

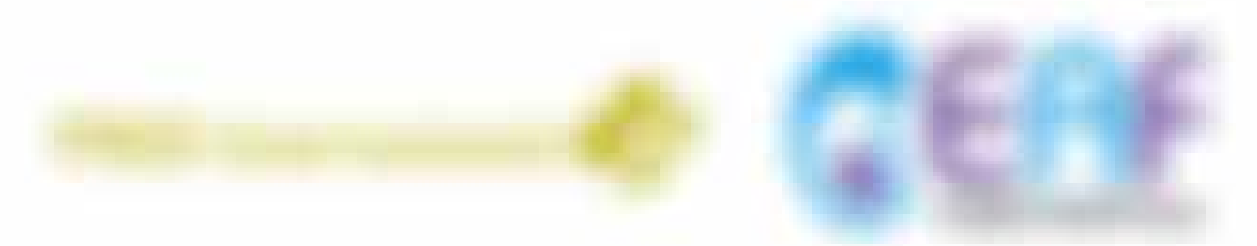
The higher of the two numbers (called the systolic blood pressure) is the pressure measured when the heart beats. The lower number (the diastolic blood pressure) is the pressure when the heart rests between beats.

Blood pressure targets indicate the readings that each number should not exceed. The typical target of 140/90 mmHg means that the systolic blood pressure should be no higher than 140 mmHg and the diastolic no higher than 90 mmHg.

Blood pressure targets should be personalised for individual patients, taking into account age and other diseases. For example, a lower target may benefit younger people with ADPKD (i.e. those aged 15–49 years) who have good kidney function and cardiovascular disease or diabetes (a disease that causes the level of sugar in the blood to be too high).



# Basic management and self care



This section explains the steps patients can take, together with their healthcare team, to help protect their kidney function and reduce the risk of cardiovascular disease.

## Lifestyle and diet

For people with kidney disease, certain lifestyle changes can help to protect kidney function and reduce the risk of cardiovascular disease. These include:

• **Not smoking** - stop or reduce or quit smoking as soon as possible.

• **Drinking alcohol** - limit alcohol intake to no more than 14 units per week.

• **Not drinking** - avoid drinking alcohol.

• **Not drinking** - avoid drinking alcohol.

• **Not drinking** - avoid drinking alcohol.

• **Not drinking** - avoid drinking alcohol.

For more information on these and other lifestyle changes, see our [lifestyle and diet](#) page.

## Focus on high blood pressure

Controlling your high blood pressure (hypertension) is very important because high blood pressure increases the risk of cardiovascular disease, such as heart attack and stroke. Controlling high blood pressure may also help to slow the growth of kidney cysts in some people with ADPKD. In people with [ADPKD](#), controlling high blood pressure can help to reduce the risk of kidney failure.

## How can blood pressure be controlled?

For people with high blood pressure, the doctor and the pharmacist should be particularly important to reduce the risk of cardiovascular disease.

Doctors can also prescribe certain [medicines](#) to help control high blood pressure.

Some [blood pressure checks](#) are important to make sure treatment is working.

• [What is blood pressure reading and target level?](#)

## Other risk factors

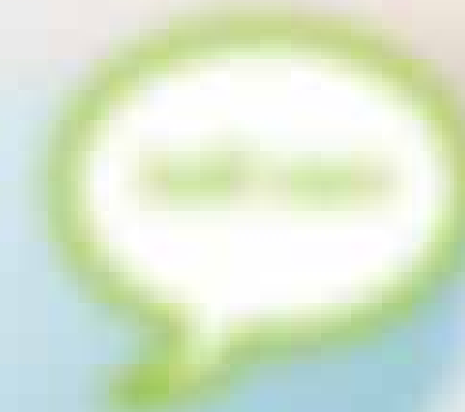
Other medicines that may be prescribed to help control risk factors for cardiovascular disease (depending on individual circumstances) may include:

- medicines to control high cholesterol levels (such as statins)
- low-dose aspirin to help stop the blood from clotting.

## Following personal and family life

People with kidney disease can still enjoy a full and active life with the right help. ADPKD can have a [psychological impact](#). If you have ADPKD, it is important to talk with your doctor about any health problems you may have. Your healthcare team can help you manage information and get support and advice.

• [Lifestyle](#)





# Basic management and self care

This section explains the steps patients can take, together with their healthcare team, to help protect their kidney function and reduce the risk of cardiovascular disease.

## Lifestyle and diet

For people with chronic kidney disease, there are many things you can do to help protect your kidney function. These include:

• **Not smoking** - stop smoking if you are a smoker. This will help reduce the risk of heart disease and cardiovascular disease.

• **Not drinking alcohol** - stop drinking alcohol if you are a drinker. This will help reduce the risk of heart disease and cardiovascular disease.

### Not smoking

• **Not drinking alcohol** - stop drinking alcohol if you are a drinker. This will help reduce the risk of heart disease and cardiovascular disease.

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## Focus on high blood pressure

Controlling your high blood pressure (hypertension) is very important because high blood pressure increases the risk of cardiovascular disease and kidney damage. Controlling high blood pressure may also help to slow the growth of kidney cysts in some people with ADPKD. To learn more, see [high blood pressure](#). Controlling high blood pressure can help reduce the risk of heart disease and cardiovascular disease.

## How can blood pressure be controlled?

Controlling high blood pressure, the doctor and the patient have an important role to play in reducing the risk of cardiovascular disease.

Doctors can also prescribe certain [medicines](#) to help control high blood pressure.

Some [blood pressure checks](#) are important to make sure treatment is working.

## What is blood pressure reading and target level?

## Other risk factors

There are many other risk factors that can increase the risk of heart disease and cardiovascular disease.

## Diabetes

Diabetes is a condition where the body does not produce enough insulin to control the amount of sugar in the blood.

Diabetes can increase the risk of heart disease and cardiovascular disease.

Diabetes can also increase the risk of kidney damage.

Diabetes can also increase the risk of heart disease and cardiovascular disease.

Diabetes can also increase the risk of kidney damage.

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Diabetes can also increase the risk of kidney damage.

## What about complementary or alternative therapies?

Complementary (or 'alternative') therapies include various treatments and practices that are not part of mainstream or conventional medicine. There is no good evidence that any complementary or alternative therapy helps protect the kidneys or slows the progression of ADPKD. Some herbs used in traditional herbal medicines may damage the kidneys.

Certain types of complementary non-medical therapies may help you to cope with the effects of ADPKD and are sometimes used in pain relief.

You are advised to ask your doctor before using any complementary therapy and not to stop a treatment prescribed by a doctor on the advice of a complementary practitioner without discussing it with your doctor.

Usually you will be required to pay for complementary therapies yourself.



## Checklist: Basic management and self-care

For details on these aspects please refer to the Route Map itself.

**Lifestyle advice provided on the following topics, with support as necessary**

- Staying hydrated
- Stopping smoking
- Maintaining a healthy body weight
- Getting enough exercise
- Eating a healthy diet, including lowering salt (sodium) intake
- Reducing alcohol to recommended limits

**Blood pressure and other cardiovascular risk factors**

- Blood pressure tested and discussed
- Lifestyle measures discussed and agreed
- Choice of prescription medicine explained and agreed, where necessary
- Target explained and agreed
- Appropriateness and options for home monitoring discussed

**Other cardiovascular risk factors**

- Need for cholesterol-lowering treatment, or other therapy, discussed

**Wellbeing, personal and family life**

- Impact of ADPKD on personal, wellbeing and family life discussed, where necessary, and action taken

**Family planning issues discussed and addressed**

**Information about patient organisations and other forms of support**

Notes and questions you would like to ask your healthcare team

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This section explains how the effects of ADPKD on the kidneys can be measured and the future progression estimated, to help personalise the care provided.

Cysts grow and multiply throughout life in people with ADPKD. The prognosis – that is, the rate at which the disease progresses and the effect it has on the kidneys – varies between patients. In some people, cysts grow and multiply so slowly that serious kidney disease (including [end-stage renal disease](#)) may only happen very late in life, or not at all. However, in other people the disease progresses more rapidly. This may be affected by ADPKD management, as well as each patient's individual disease.

The rate that ADPKD progresses can be measured, and even estimated in advance. This can be useful to:

- identify patients with rapidly progressing disease who may be suitable for certain treatments or for [clinical trials](#)
- evaluate whether treatment is effective
- plan for kidney [transplantation or dialysis](#), later in the disease course.

The progression of kidney function can be assessed in several ways.

## Family history

Patients are at increased risk of rapid ADPKD progression if other members of their family with the disease reached [end-stage renal disease](#) before the age of 58 years. It is recommended that ADPKD progression should be checked every 3–5 years in diagnosed patients with this family history.

 [Checklist >](#)

 Prognosis

## Kidney function tests

The most important factor in the prognosis of ADPKD is [kidney function](#). Good kidney function suggests a better prognosis than bad kidney function regardless of cyst growth. Doctors can predict how kidney function will change based on repeated measurements of creatinine levels in the blood and urine (see [kidney function tests](#)).

However, people with ADPKD can have normal kidney function for many years, even though their cysts continue to grow and multiply. For this reason, doctors also use other tests to monitor and predict ADPKD progression. Two methods that may be used are: the total kidney volume (TKV) and the Predicting Renal Outcomes (PRO) PKD score.

### Total kidney volume

**Total kidney volume >** (or TKV) is a measure of the swelling of the kidneys caused by cysts. It is calculated from kidney scans and can be used to assess ADPKD progression.

### PROPKD score

The **PROPKD score >** is a research tool that predicts the risk of progression according to four factors. One of these factors is the [genetic](#) mutation present and the use of the PROPKD score is limited by the cost and availability of [genetic testing](#). This score is currently used only for research and not for the routine management of patients.

## Prognosis

*'I found out I had a polycystic kidney at the age of 35, I already had two children and I was told I would soon be on dialysis. As a matter of fact, I am now 68 years old, and my overall condition is fairly good.'*

*Antonia, Italy*

*'When I was informed about my ADPKD it was a brutal shock and difficult news to accept. Luckily, soon after, I met a nephrologist who still follows me today and who managed to instil hope and to turn the gloomy picture into something more positive. It is crucial that doctors take time with their patients when the pathology is first announced in order to further bring hope and support.'*

*Corinne, France*

*'Although kidney failure was predictable in my case, it was not clear when it would actually happen. In principle, the process from the first notable restrictions to kidney failure took about 10 years. The diagnosis was clear, there were no secrets. For years there were no treatment options other than blood pressure adjustment and diet, followed then by the "radical cure" of dialysis and transplantation. The output of the latter is certainly not predictable. In my case, the additional cyst growth in my liver – which was initially not prognosticated – was an aggravating circumstance. This was even more dramatic than kidney disease, because of its size and the associated massive complaints.'*

*Elisabeth, Austria*

This section explains how the effects of ADPKD on the kidneys can be measured and the future progression estimated, to help personalise the care provided.

Use your doctor's advice to help you decide if you should have a kidney scan. There are several types of kidney scan and your doctor will advise you on the best one for you. You may also want to discuss the availability of scans in your area. Some scans are more expensive than others and some are not available in all areas. You may also want to discuss the availability of scans in your area. Some scans are more expensive than others and some are not available in all areas.



## Total kidney volume

The total kidney volume (TKV) is calculated based on imaging from kidney scans.

- Ultrasound scans are inexpensive, widely available and can be used to estimate TKV in some situations.
- Magnetic resonance imaging (MRI) scan is the ideal method for measuring TKV, as this is more accurate than ultrasound. However, some patients may have limited access to MRI scans, especially for repeated TKV measurement. Faster and simpler MRI techniques are now increasingly available.

Doctors take into account the height and age of the patient when assessing TKV. This is because the size of the kidney means different things in a small person compared with a larger person, and in a young person compared with an older person.

Ideally, TKV should first be measured around the time of diagnosis and then at a later point.

TKV can be used in two ways to assess ADPKD progression:

### *Risk prediction*

A single TKV measurement can be used to predict how quickly ADPKD will progress in the future. This may be used to make decisions about treatment. Most patients with ADPKD can be classified into one of five risk categories for disease progression (called 1A–1E) based on their TKV, adjusted for their height. In general, patients in Classes 1C–1E will have rapid disease progression.

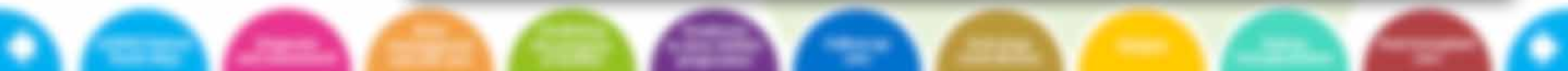
### *Repeated measurement*

Repeating the TKV measurement several times allows doctors and patients to see how quickly the kidney volume is actually changing and progressing. The availability of repeated TKV measurement varies so you may wish to discuss this with your nephrologist.

It is recommended that patients whose TKV increases more than 5% per year should be considered to have rapidly progressing ADPKD. This assessment should preferably be based on three or more measurements using MRI scans, each at least 6 months apart.

The prediction of ADPKD progression is normally conducted in centres that have a special expertise in ADPKD. A list of expert centres within the [European Reference Network for Rare Kidney Diseases](#) can be found [here](#).

The length of the kidney (rather than the volume) can also be measured by ultrasound and used to predict whether ADPKD is likely to progress rapidly.



# Predicting the progress of ADPKD

This section explains how the effects of ADPKD on the kidneys can be measured and the future progression estimated, to help personalise the care provided.

Kidney function declines throughout life in people with ADPKD. The progress – that is, the rate at which the disease progresses and the effects on the kidneys – varies between patients. In some people, kidney function declines so slowly that serious kidney disease (including [end-stage kidney disease](#)) may only happen very late in life, or not at all. However, in other people, the disease progresses more quickly. This may be affected by ADPKD management, as well as other genetic-related factors.

The rate that ADPKD progresses can be measured and then estimated in advance. This can be helpful:

- identify patients with rapidly progressing disease who may be eligible for certain treatments or for [transplant](#)
- evaluate whether treatment is effective
- plan for kidney [replacement or dialysis](#) late in the disease course.

The progression of kidney function can be assessed in several ways:

## Family history

Patients are at an increased risk of rapid ADPKD progression if other members of their family with the disease started [haematuria](#) and [proteinuria](#) before the age of 30 years. It is recommended that ADPKD progression should be checked every 3-4 years in high-risk patients with this family history.



### Kidney Function Tests

The rate that ADPKD progresses can be measured and then estimated in advance. This can be helpful:

- identify patients with rapidly progressing disease who may be eligible for certain treatments or for [transplant](#)
- evaluate whether treatment is effective
- plan for kidney [replacement or dialysis](#) late in the disease course.

The progression of kidney function can be assessed in several ways:

#### PROPKD score

The PROPKD score predicts the risk of ADPKD progression according to whether the patient:

- 1) is male or female
- 2) needed treatment for high blood pressure before the age of 35 years
- 3) had any of the main kidney complications of ADPKD before the of 35 years and
- 4) has ADPKD caused by a genetic mutation called PKD1.

[Haematuria](#) is the presence of blood in the urine. It is a common complication of ADPKD. It can be a sign of kidney damage and may lead to kidney failure. It is important to report any blood in the urine to your doctor.

[Proteinuria](#) is the presence of protein in the urine. It is a common complication of ADPKD. It can be a sign of kidney damage and may lead to kidney failure. It is important to report any protein in the urine to your doctor.





ADPKD is a genetic condition that can be passed from parents to their children. This section explains the basics of ADPKD genetics, inheritance and genetic testing.

## How is ADPKD inherited?

ADPKD is usually caused by abnormalities (or mutations) in one of two **genes** > : these are known as *PKD1* and *PKD2*. In fact, ADPKD is the most common genetic kidney disease.

ADPKD is inherited in a **dominant pattern** >. This means that the ADPKD mutation need only to be present in the inherited genes from one parent for it to cause ADPKD.

ADPKD is not the same as autosomal *recessive* polycystic kidney disease (**ARPKD**), which is a separate, rare disease with a different inheritance pattern.

## Genetic testing

Genetic testing can identify if either of the *PKD1* or *PKD2* mutations are present. It is done by testing a small sample of blood or saliva.

Usually doctors do not need to use genetic testing to diagnose ADPKD, but it can help the diagnosis in some **situations** >.

Pre-implantation and prenatal genetic testing are also possible for the purpose of **family planning**, although the availability of these tests varies.

Genetic testing for ADPKD is complex and expensive and is normally performed by geneticists in centres with appropriate experience. However, genetic tests are becoming quicker and less expensive, thanks to advances in technology.

The EAF and PKD International believe that, where possible, all patients with ADPKD should have access to testing where it is appropriate. However, at present the availability of genetic testing for ADPKD varies widely between countries and regions.

## Counselling

A positive genetic test for ADPKD can have lifelong consequences for patients and their families. As well as the effects of the disease itself, ADPKD can also affect other aspects of **well-being, personal and family life, and finances**.

Ideally, everyone who undergoes genetic testing for ADPKD (and the parents of tested children) should have access to counselling to discuss the advantages and disadvantages in detail. This counselling should be with a clinical geneticist, specialist nurse or genetic counsellor with expertise in ADPKD.

Counselling should also be available to discuss the test result and its implications. A child who is sufficiently mature and competent may be included in the counselling.

You can find more information about genetics and testing at the websites of ADPKD and genetic disease **patient organisations**.

✔ **Checklist** >

Genetics



## Genetics

*'I was diagnosed by pure chance when I was aged 23. I have a "de novo" ADPKD mutation – the first case in my family. I didn't think much about it when I was young, but now that I'm 45, and after having two kids, I can't stop thinking about it. My daughter is only 10 and was diagnosed when she was barely 2. I have huge feelings of guilt for having transmitted this disease. The mental stress out of it is so persistent and so strong that sometimes I think I am the disease . . . I am scared that my son could also be affected. I wish there was more psychological support for patients.'*

*Silvia, Italy*

*'In my family of origin, four out of six siblings are affected. The disease was passed on to us by our father; it's not known whether he inherited the disease or whether a new mutation occurred in his case. My daughter is also affected by the disease. She is already aware of this, since she has learned from experiencing my decline, and my dialysis and transplantation. It is my wish, however, not to make the disease a permanent topic in our house. It is important that she can live as carefree as possible regardless of the disease and, above all, that she chooses her career aspiration freely, regardless of the disease.'*

*Miriam, Austria*

*'In my family it is difficult to talk about the disease because everyone is highly anxious about the risk of transmitting it to the next generation. With my kids I decided to talk openly and in full transparency but without undue dramatisation.'*

*Corinne, France*

*'I was diagnosed when I was 16. One day after a competition I went to the toilet and there was a lot of blood in my urine. I was so scared! They took me to hospital and did a lot of checks. In the beginning the doctor thought I had kidney cancer, then we found out it was something else. After that, my father and older brother were diagnosed with the same disease, but nobody told me that I had a genetic disease and I never understood that! It was only many years later when a doctor told me "What? You are having children. Are you crazy? Don't you know you have a genetic disease?" I was shocked. . . It took me weeks to come to terms with this fact. I felt so guilty. Fortunately, my wife had a very positive outlook towards this thing. I would like to tell everyone that I am now 47, we have a very normal life and except for that very first trip to the hospital I've never had any issues with PKD!'*

*Rolf, Switzerland*

ADPKD is a genetic condition that can be passed from parents to their children. This section explains the basics of ADPKD genetics, inheritance and genetic testing.

## Genes

Genes are sections of DNA contained inside almost all the body's cells. They act as a set of instructions, telling the cells how to make the proteins that build and regulate the body. Most genes are inherited and so determine the characteristics that are passed from parents to their children. Mutations in the genes can cause a wide variety of inherited diseases. ADPKD is usually caused by one of two genetic mutations – these are known as *PKD1* and *PKD2*. *PKD1* mutations are most common, causing around three-quarters (75%) of cases. ADPKD caused by *PKD1* mutations is generally more severe and rapidly progressing than that caused by *PKD2* mutations.

Current genetic tests fail to identify a *PKD1* or *PKD2* mutation in around one in 10 people with ADPKD. In a few of these cases, PKD may be caused by other genes. Recent studies have shown that mutations in several other genes can cause ADPKD. The PKD in these cases is usually mild.

Genetic testing can identify the presence of a mutation in a gene. This can help to determine if you have ADPKD or if you are a carrier of the gene. Genetic testing can also help to determine the severity of your condition.



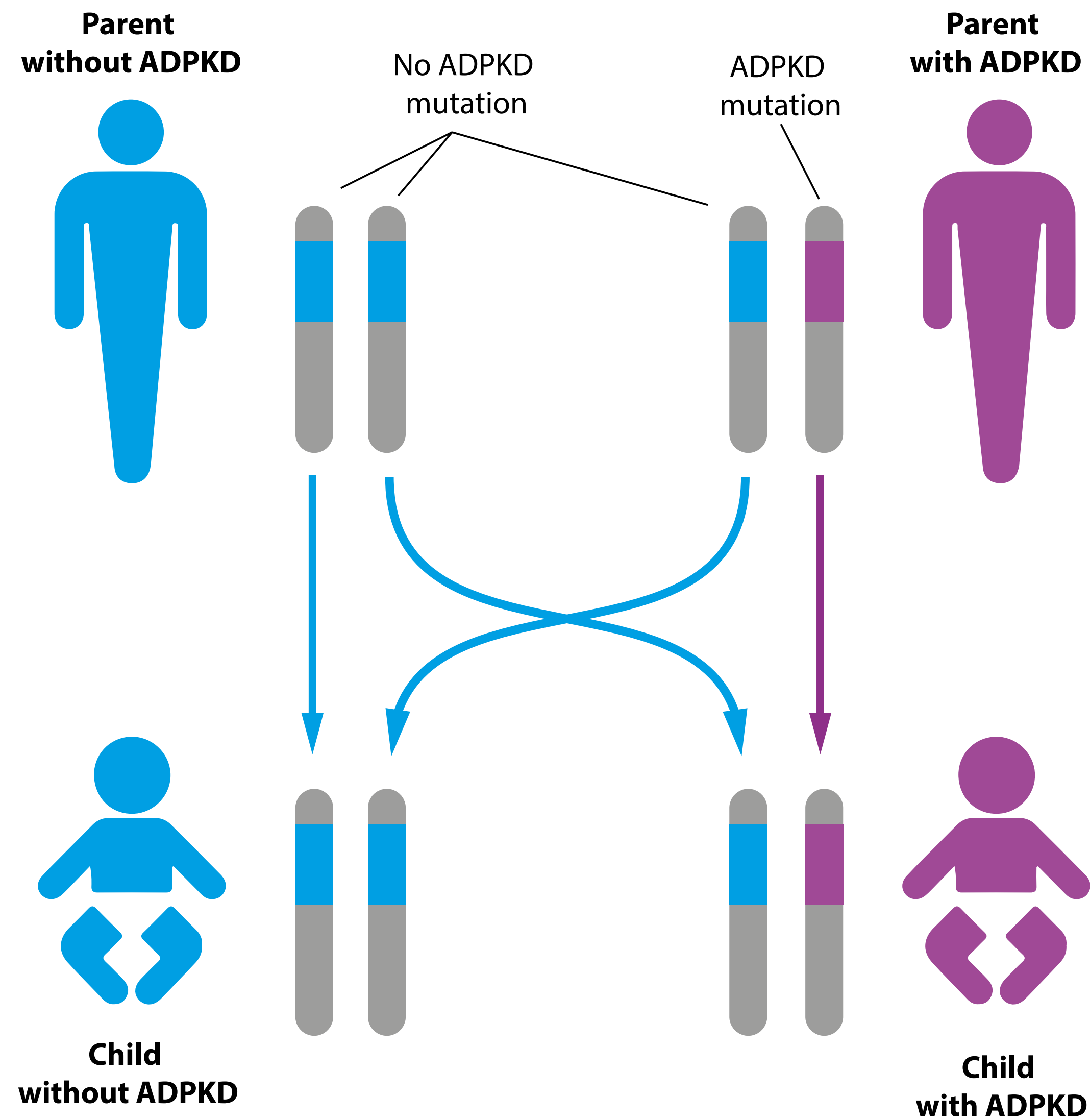
### Dominant pattern

ADPKD is inherited in a 'dominant' pattern, meaning that the ADPKD mutation need only be present in the inherited genes from one parent for it to cause the disease. This means that:

- If either of your parents has ADPKD, there is a one in two (50%) chance that you will have inherited the disease.
- If you have ADPKD, there is also a one in two (50%) chance that each child you have will inherit the disease.

Children who do not inherit the abnormal gene causing ADPKD will not develop the disease or pass it to their children. ADPKD cannot 'skip' a generation, but sometimes people with ADPKD can go through life without being diagnosed and still pass the disease to a child.

In a small number of cases (fewer than one in 10), genetic mutations causing ADPKD occur spontaneously in patients for the first time, i.e. without there being any previous history of the disease in the family. Any children of these patients will still have a 50% chance of inheriting the disease.



We all have two copies of most genes – one copy inherited from each parent

Each child with one parent with an ADPKD mutation has a 50% (1 in 2) chance of inheriting it



ADPKD is a genetic condition that can be passed from parents to their children. This section explains the basics of ADPKD genetics, inheritance and genetic testing.

## How is ADPKD inherited?

ADPKD is usually caused by mutations in two genes called [PKD1](#) and [PKD2](#). These genes are located on chromosomes 16 and 4 respectively. ADPKD is the most common genetic kidney disease.

ADPKD is inherited in an [autosomal dominant](#) way. This means that the ADPKD mutation can be passed on to your children if you have just one copy of the gene that is mutated.

ADPKD is not the same as autosomal recessive kidney disease. Autosomal recessive kidney disease is caused by two copies of a mutated gene.

## Genetic testing

Genetic testing can be used to find out if you have a mutation in the PKD1 or PKD2 gene.

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## Counselling

Genetic testing for ADPKD can have many consequences for you and your family. As well as the effects of the disease itself, ADPKD can also affect other aspects of your life.

ADPKD can also affect other aspects of your life. It can affect your work, your relationships and your health. It can also affect your ability to have children.

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### Situations

Situations where genetic testing can be helpful include:

- where the diagnosis is unclear, especially in infants and children or if there is no family history of ADPKD
- to help predict the prognosis of ADPKD (e.g. using the [PROPDK](#) score)
- to make sure the abnormality is not present in a potential living kidney donor
- [pre-implantation genetic diagnosis](#), which is used to test whether embryos have ADPKD genes prior to in vitro fertilisation.



# Treatment to slow ADPKD progression

This section explains how some patients with ADPKD may benefit from specific treatment to slow disease progression.

In recent years, research has been performed into medicines to slow the progression of ADPKD. Currently, one medicine is licensed for use in Europe and others are being studied in clinical trials.

## Who is suitable for treatment?

The medicine that is licensed for use is called [tolvaptan >](#). It can only be used by adults with ADPKD who have normal or moderately reduced kidney function (stage 1–3 [chronic kidney disease](#)) at the start of treatment, and who have evidence of [rapidly progressing disease >](#).

This treatment is not available everywhere. Patients may wish to discuss its availability, and your suitability, with your nephrologist.

Experts recommend that patients should be fully involved in deciding on whether treatment is appropriate for them.

## What is the benefit?

Evidence suggests that, if the benefit shown in a clinical trial was maintained, this would mean that each 4 years of treatment would delay the occurrence of [end-stage renal disease](#) by approximately 1 year. The treatment may also reduce kidney complications and pain, and increases urine production. It does not affect liver cysts.

This treatment does not replace the need for other aspects of [basic ADPKD management and self-care](#).

## What are the main side effects?

The most common side effect of this treatment, occurring in all patients treated, is an increased need to urinate. If you take tolvaptan, you should take [precautions >](#) to ensure you do not become dehydrated.

If you experience difficulty in urinating, this may indicate kidney or bladder problems and you should stop taking the treatment and immediately contact a doctor or go to the nearest hospital.

This treatment may cause the liver to stop working properly. You should tell your doctor if you have any [symptoms >](#) that might indicate liver damage. Blood tests to check the liver function need to be done each month for the first 18 months of treatment and every 3 months after that.

## How is treatment taken?

This treatment is taken in the form of tablets, twice daily. You should follow the prescribing doctor's instructions carefully, and check with your healthcare team if you have any questions. Further information can be found in the patient information leaflet or online.

## Who provides this type of treatment?

This treatment must be started and monitored by a doctor who is experienced in managing ADPKD and who understands the risks of therapy and the necessary monitoring.

## Are any other treatments being developed for ADPKD?

Other medicines are being investigated to slow the progression of ADPKD and polycystic liver disease, and eventually these may provide more treatment options.

Some of these medicines are being tested in [clinical trials](#). Patients who wish to participate in clinical trials should ask their nephrologist about the available opportunities.



# Treatment to slow ADPKD progression

This section explains how some patients with ADPKD may benefit from specific treatment to slow disease progression.

A recent study suggests that some patients with ADPKD may benefit from treatment to slow disease progression.

## Who is suitable for this treatment?

The evidence that is currently available suggests that treatment to slow disease progression may be suitable for some patients with ADPKD.

## What is the benefit?

The evidence suggests that treatment to slow disease progression may be suitable for some patients with ADPKD.

## What are the main side effects?

The most common side effect of this treatment is an increase in uric acid levels.

## Are any other treatments being developed for ADPKD?

Other medicines are being investigated to slow the progression of ADPKD.

## Tolvaptan

Tolvaptan is a type of medicine called a vasopressin-2 antagonist. It works by blocking the action of the hormone vasopressin, which is involved in the development of kidney cysts in ADPKD.

The evidence suggests that treatment to slow disease progression may be suitable for some patients with ADPKD.

## Who is suitable for this treatment?

The evidence that is currently available suggests that treatment to slow disease progression may be suitable for some patients with ADPKD.

## What is the benefit?

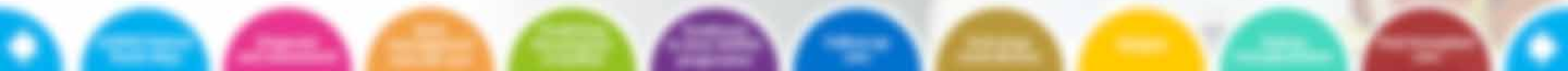
The evidence suggests that treatment to slow disease progression may be suitable for some patients with ADPKD.

## What are the main side effects?

The most common side effect of this treatment is an increase in uric acid levels.

## Are any other treatments being developed for ADPKD?

Other medicines are being investigated to slow the progression of ADPKD.



# Treatment to slow ADPKD progression

This section explains how some patients with ADPKD may benefit from specific treatment to slow disease progression.

A recent study showed that some patients with ADPKD may benefit from treatment to slow disease progression. Currently, there are no other treatments available to slow disease progression in ADPKD.

## Who is suitable for treatment?

### Rapidly progressing disease ✕

How 'rapidly progressing disease' is defined, and therefore which patients are eligible for treatment, can vary in different countries. European experts have published [guidelines](#) on which patients should be considered for treatment. National guidelines are also available in some countries (e.g. the United Kingdom [here](#) and [here](#)).

The treatment may cause the liver to stop working properly. The liver will stop working if you have any [liver disease](#). You might not get liver damage. However, to check the liver function, you'll need to have blood tests for the first 12 months of treatment and every 12 months after that.

## How is treatment taken?

The treatment is given in the form of tablets, taken daily. You should follow the instructions on the packaging carefully, and check with your healthcare team if you have any questions. Further information on the treatment is available in the patient information leaflet.

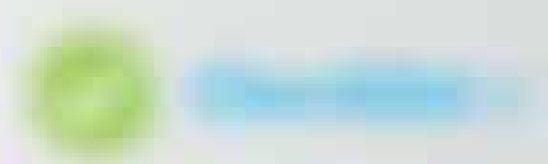
## Who provides this type of treatment?

The treatment is given by a specialist nurse or pharmacist. You should have regular appointments with your healthcare team to check the side effects of the treatment and to monitor your kidney function.

## Are any other treatments being developed for ADPKD?

Other medicines are being investigated to slow the progression of ADPKD and reduce the risk of stroke. Unfortunately, there may not be any other treatment options.

Some of these medicines are being tested in [clinical trials](#). There are also some medicines in development that may be used to treat the side effects of the treatment.





# Treatment to slow ADPKD progression

This section explains how some patients with ADPKD may benefit from specific treatment to slow disease progression.

A recent study showed that some patients with ADPKD may benefit from specific treatment to slow disease progression. This section explains how some patients with ADPKD may benefit from specific treatment to slow disease progression.

## Who is suitable for treatment?

The treatment is suitable for patients with ADPKD who have moderate to severe kidney disease (stage 3-4) and who also have evidence of rapid progression.

The treatment is not suitable for patients who have moderate to severe kidney disease but who do not have evidence of rapid progression. It is also not suitable for patients who have moderate to severe kidney disease but who do not have evidence of rapid progression.

## What is the benefit?

Research suggests that if the growth of the kidneys is slowed, the progression of kidney disease may be delayed. This means that some patients with ADPKD may benefit from specific treatment to slow disease progression.

## How is treatment taken?

The treatment is taken in the form of a tablet, once daily. You should follow the prescribing doctor's instructions carefully and check with your healthcare team if you have any questions. Further information can be found in the patient information leaflet or video.

## Why provide this type of treatment?

The treatment is provided to help slow the progression of kidney disease in patients with ADPKD who have moderate to severe kidney disease and who also have evidence of rapid progression.

The treatment may cause the liver to stop working properly. You should not use alcohol if you have any liver problems. You should also avoid liver damage. You should also check the liver function regularly. You should also check the liver function regularly for the first 12 months of treatment and every 12 months after that.

## How is treatment taken?

The treatment is taken in the form of a tablet, once daily. You should follow the prescribing doctor's instructions carefully and check with your healthcare team if you have any questions. Further information can be found in the patient information leaflet or video.

## Who provides this type of treatment?

The treatment is provided by a specialist nurse or pharmacist who is experienced in managing ADPKD and who understands the risks of drugs and the monitoring process.

## Are any other treatments being developed for ADPKD?

Other medicines are being investigated to slow the progression of ADPKD and to help with the symptoms. You should check with your healthcare team if you have any questions.

Some medicines are being investigated to help with the symptoms of ADPKD. You should check with your healthcare team if you have any questions.

**Precautions** ✕

If you are taking tolvaptan, to avoid dehydration during treatment you should always have access to water and be able to drink enough when you feel thirsty and to prevent thirst (e.g. by always having water to hand).



# Treatment to slow ADPKD progression

This section explains how some patients with ADPKD may benefit from specific treatment to slow disease progression.

A recent study showed that some patients with ADPKD may benefit from treatment to slow the progression of ADPKD. Currently, there are no treatments available to slow the progression of ADPKD, and there are many factors that can affect the progression of ADPKD.

## Who is suitable for treatment?

The treatment is suitable for patients with ADPKD who have moderate to severe kidney disease (stage 3-4) and who have evidence of rapid progression (stage 3-4).

The treatment is not suitable for patients who have moderate to severe kidney disease (stage 3-4) and who have evidence of slow progression (stage 3-4).

Some patients may have other medical conditions that may affect the progression of ADPKD, and your doctor will advise you if treatment is appropriate for them.

## What is the benefit?

Current evidence suggests that if the growth of ADPKD is slowed, the progression of ADPKD may be delayed. The treatment may also help to slow the progression of ADPKD, and there are many factors that can affect the progression of ADPKD.

The treatment does not replace the need for other aspects of ADPKD management, such as blood pressure control.

## What are the main side effects?

The most common side effects of the treatment are dizziness, headache, and fatigue. These side effects are usually mild and go away on their own.

Some patients may experience other side effects, such as nausea, vomiting, and loss of appetite. If you experience any of these side effects, you should contact your doctor.

The treatment may cause the liver to enlarge, and this may lead to liver damage. You should inform your doctor if you have any signs of liver damage, such as nausea, vomiting, fever, tiredness, loss of appetite, pain in the abdomen, dark urine, jaundice (yellowing of skin or eyes), itching, or joint and muscle pain with fever.

## How is treatment taken?

The treatment is taken in the form of a tablet, once daily. You should follow the instructions on the packaging carefully, and check with your healthcare team if you have any questions. Further information can be found in the patient information leaflet (PIL).

## Why provide this type of treatment?

The treatment is provided to help slow the progression of ADPKD, and there are many factors that can affect the progression of ADPKD.

## Are any other treatments being developed for ADPKD?

There are many other treatments being developed for ADPKD, and your doctor will advise you if any of these treatments are suitable for you.

**Symptoms that might indicate liver damage** ✕

You should inform your doctor immediately if you have signs that could indicate potential liver problems such as nausea, vomiting, fever, tiredness, loss of appetite, pain in the abdomen, dark urine, jaundice (yellowing of skin or eyes), itching, or joint and muscle pain with fever.





# Managing kidney complications

This section explains the main complications of ADPKD that can affect the kidneys, and how these can be managed.

## Kidney cyst infections

Kidney cysts can become infected by bacteria. This can cause fever and pain in the abdomen. Cyst infections can be difficult to diagnose. A type of scan called **positron emission tomography >** (PET) is sometimes used.

**Antibiotics >** are the main treatment for cyst infections.

In some cases cysts may be drained, either by surgery or a procedure in which a needle is passed into the kidney through the skin.

## Cyst rupture and bleeding

Kidney cysts can sometimes burst (or 'rupture') and bleed, causing blood to appear in the urine.

In most cases bleeding resolves without treatment within 7 days.

You may self-treat pain using over-the-counter painkillers. If necessary, doctors may prescribe other pain relief medicines. If you have severe or persistent bleeding you should seek medical advice.

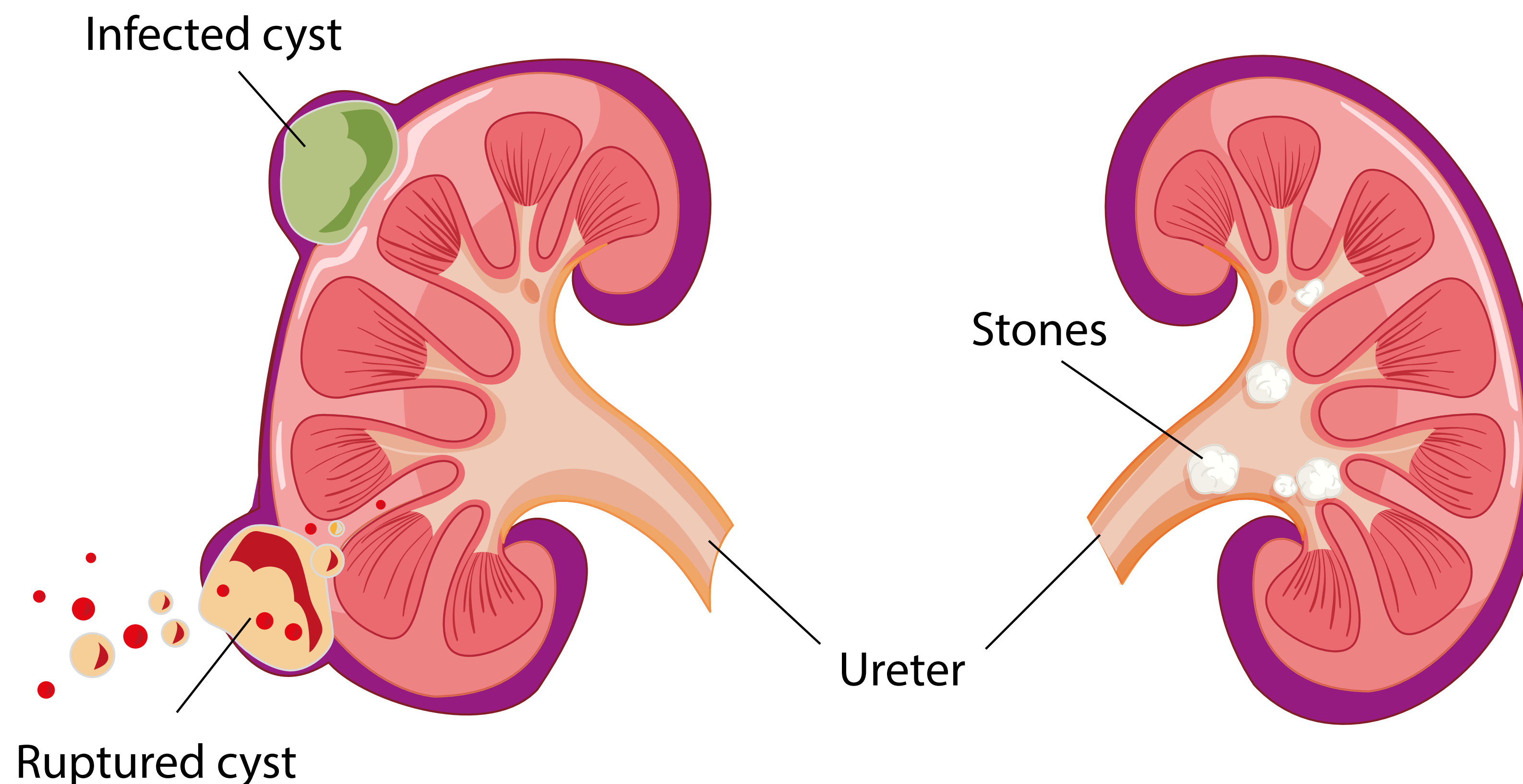
## Kidney stones

People with ADPKD are at increased risk of kidney stones. Kidney stones are caused by the build-up of certain chemicals in the urine. Larger stones can block parts of the **urinary system >** and cause discomfort and pain. They may make you need to urinate more often than normal, or blood to appear in the urine.

You should contact your doctor if you think you have a kidney stone. Stones are best diagnosed using a **computed tomography >** (CT) scan, where this is available. X-ray or ultrasound scans may also be used. Urine and blood tests may be performed, and stones that are passed in the urine may be examined.

Small stones may be passed in the urine, without treatment. Drinking extra water to increase the urine flow can help to flush out stones. For **larger stones >**, medical help is necessary.

You may self-treat any pain using over-the-counter painkillers. If necessary, doctors may prescribe other pain relief medicines.



# Managing kidney complications

This section explains the main complications of CKPD that can affect the kidneys, and how these can be managed.

## Kidney cyst infections

Kidney cyst infections are caused by bacteria that enter the kidney through the bloodstream or from the urinary tract.

Antibiotics are the main treatment for kidney cyst infections. You may also need to take painkillers to help with any pain.

## Cyst rupture and abscess

A kidney cyst can sometimes rupture, causing pain and bleeding. This is usually treated with antibiotics.

You may also need pain relief over the counter paracetamol. If necessary, doctors may prescribe other pain relief medicines. If you have severe or persistent bleeding you should seek medical advice.

## Kidney stones

People with CKPD are at an increased risk of kidney stones. Kidney stones are usually the size of a grain of rice, but some can be as large as a golf ball. They are made up of crystals of calcium and other minerals. They can cause pain and may need to be removed. This is usually done by surgery.

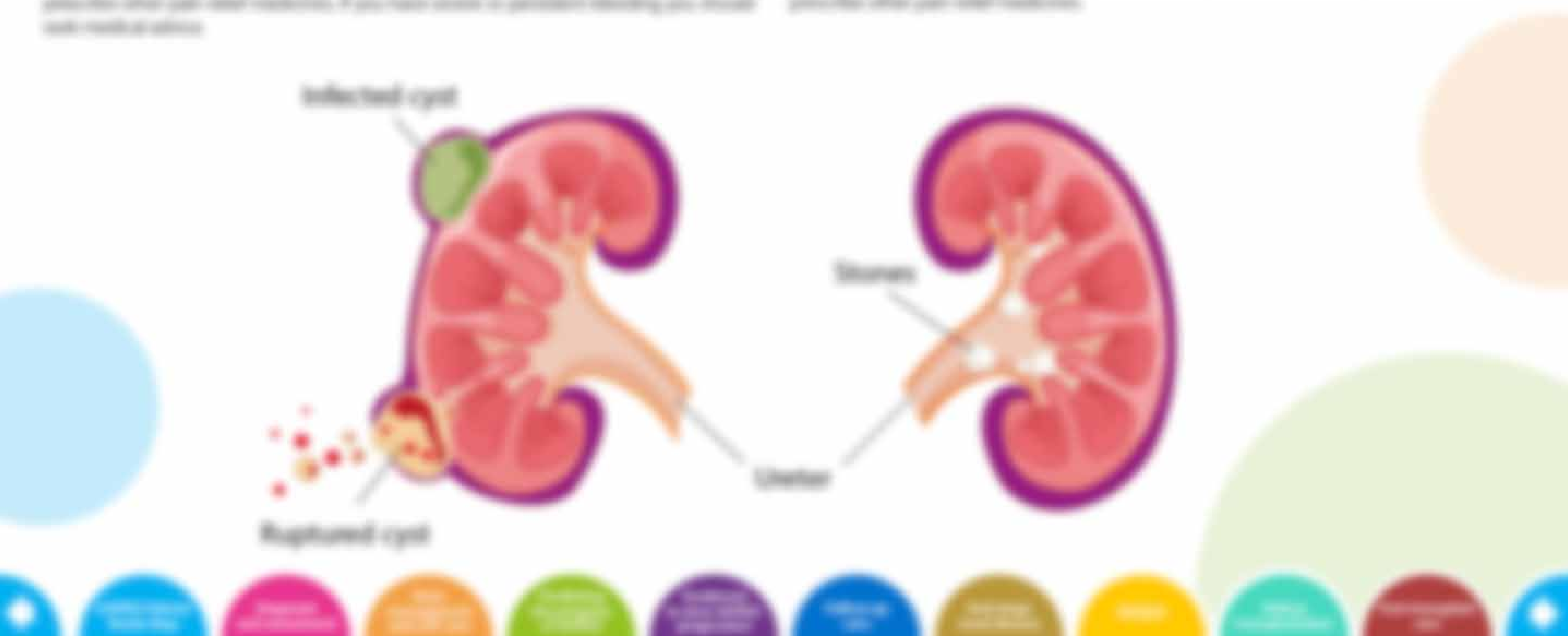
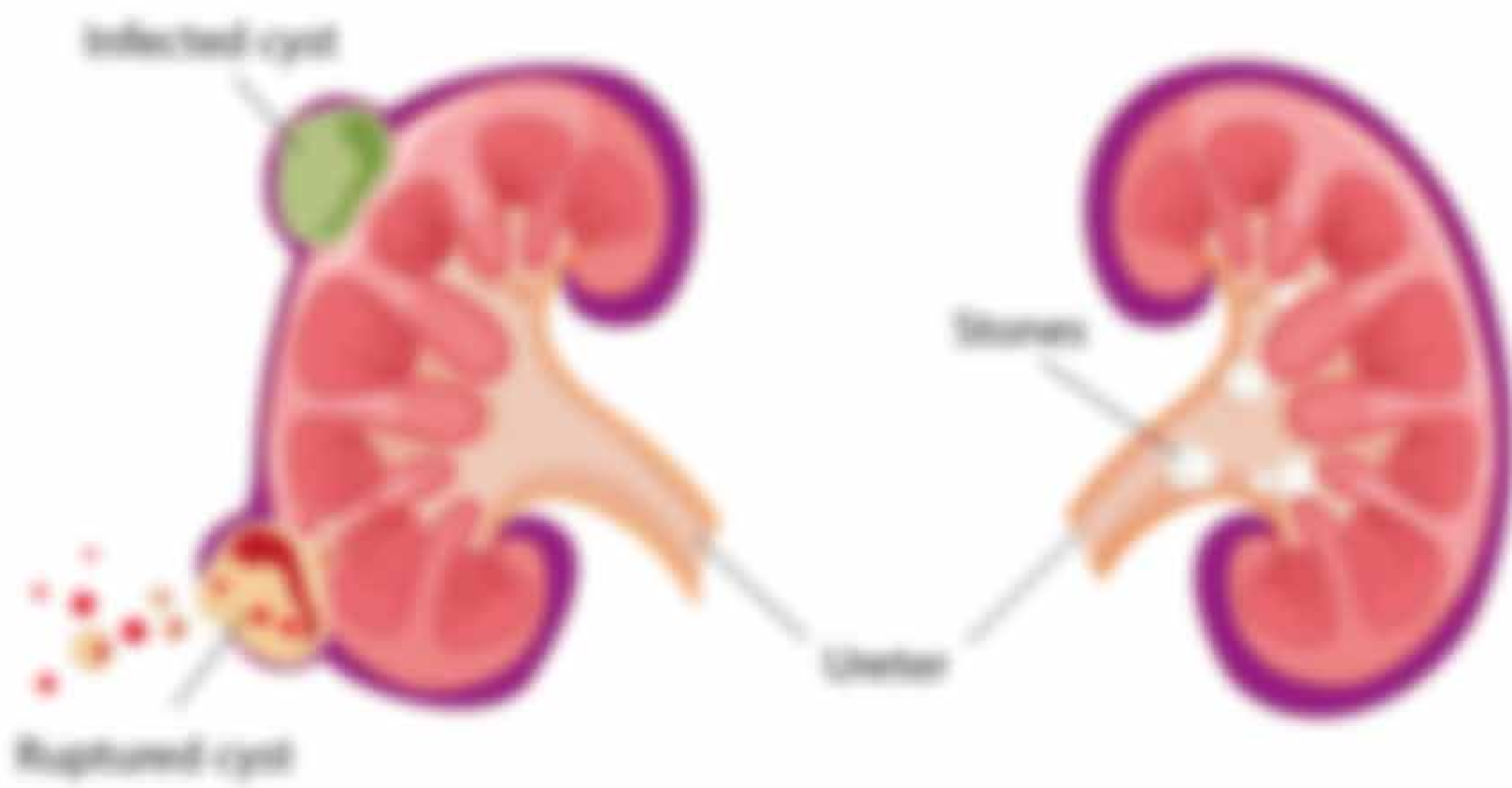
Some kidney stones can be treated with shock wave lithotripsy. This is a non-invasive treatment that uses sound waves to break up the stones. Other treatments may be performed, and some removed by the other way to surgery.

Stones may be passed in the urine without treatment. Drinking plenty of water to flush the stones from the body is the first step. For larger stones, medical help is necessary.

You may also need pain relief over the counter paracetamol. If necessary, doctors may prescribe other pain relief medicines.

**Positron emission tomography**

Positron emission tomography (PET) is a type of scan that uses small amounts of radioactive materials, a special camera and a computer to provide detailed images of the organs and tissues inside the body.



# Managing kidney complications

This section explains the main complications of CKPD that can affect the kidneys, and how these can be managed.

## Kidney cyst infections

Kidney cysts can become infected by bacteria. This can sometimes happen in the presence of an infection in the bladder. It is important to get a diagnosis as early as possible. [Antibiotics](#) are the main treatment for cyst infections.

[Antibiotics](#) are the main treatment for cyst infections.

In some cases cysts may be drained either by surgery or by a procedure in which a needle is inserted into the kidney through the skin.

## Cyst rupture and bleeding

Kidney cysts can sometimes burst or rupture and cause bleeding inside the cyst.

In most cases bleeding resolves without treatment within 7 days.

You may still need pain relief over the course of several days. If necessary, doctors may give you other pain relief medicines. If you have severe or persistent bleeding you should seek medical advice.

## Kidney stones

Kidney stones can form in the kidney. They are made of minerals and salts that have crystallised out of the urine.

[Painkillers](#) can help to relieve the pain.

The stones may pass on their own or you may need surgery to remove them.

## High blood pressure

High blood pressure can sometimes be caused by kidney disease.

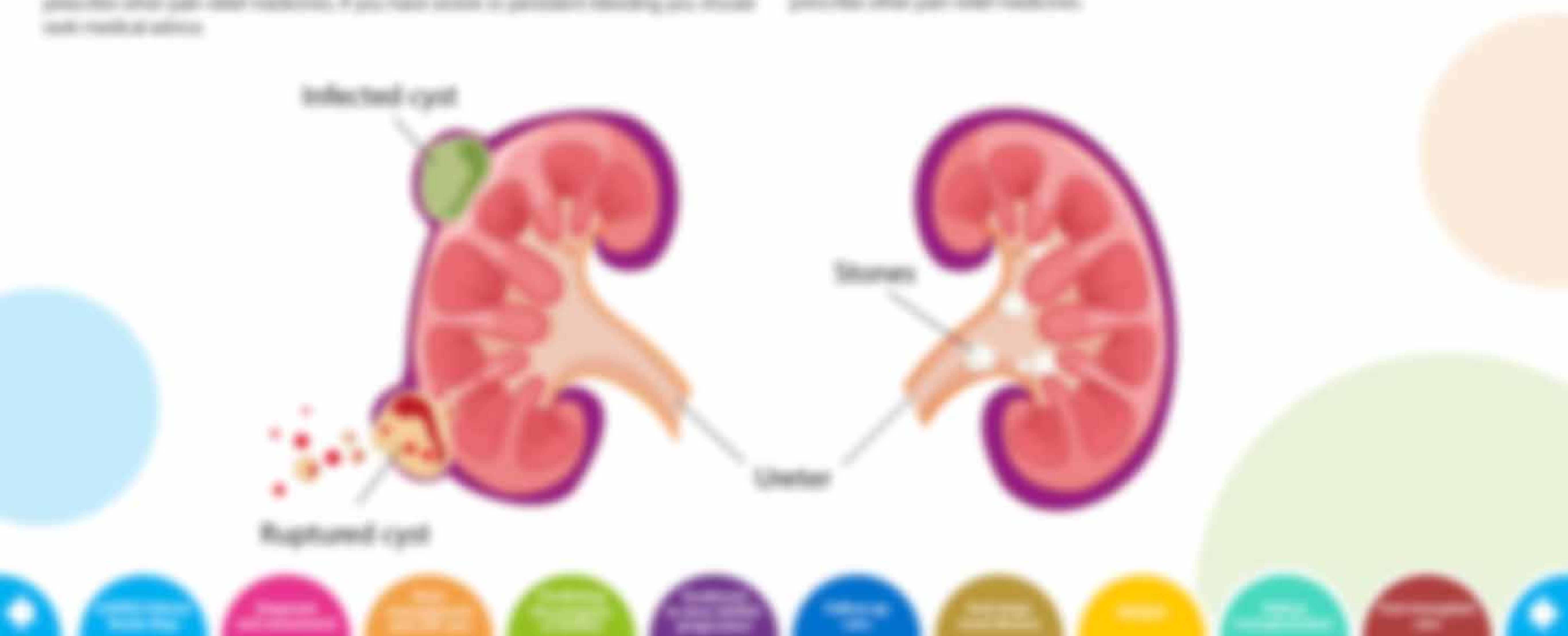
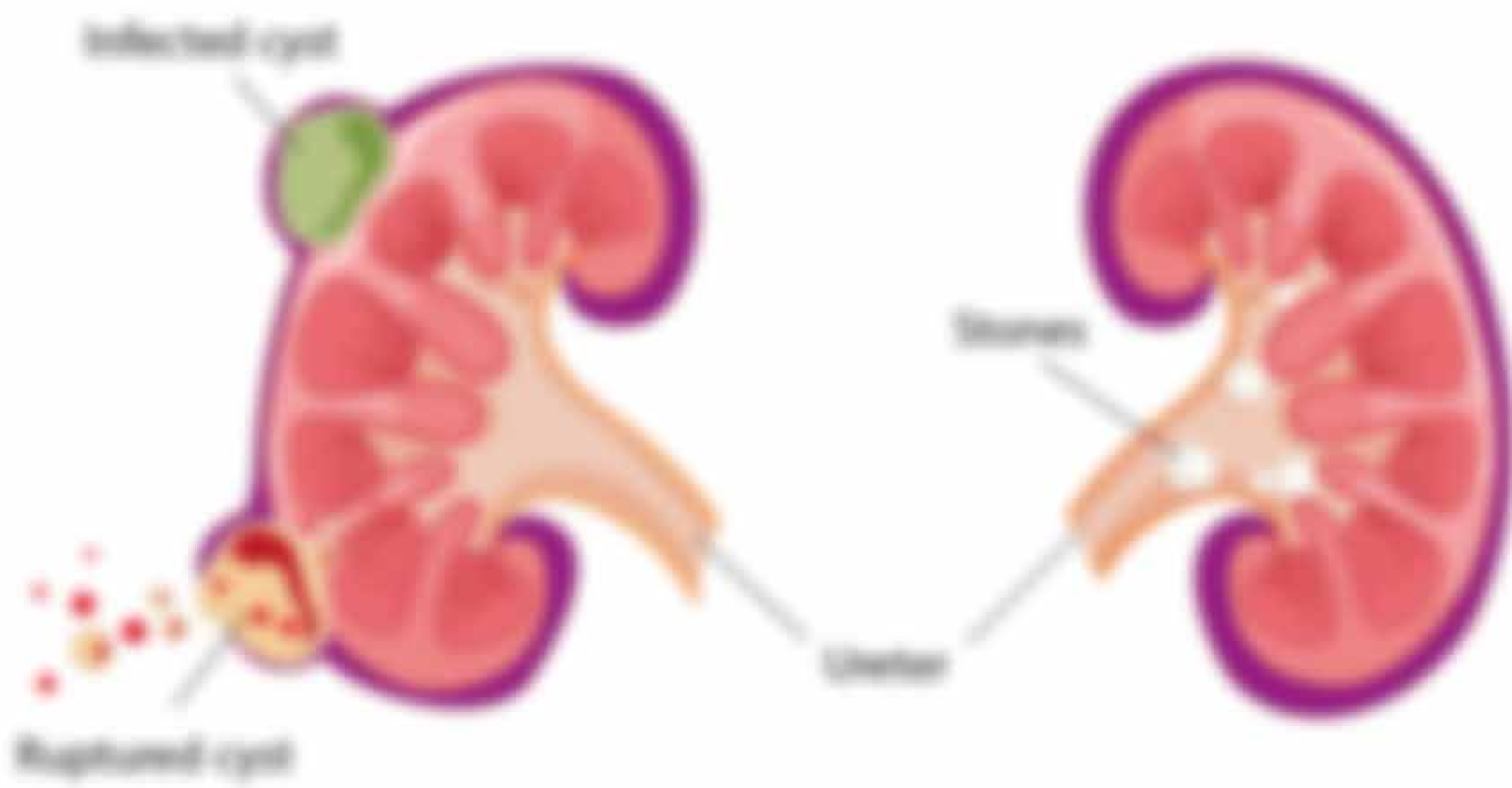
[Antihypertensive](#) medicines can help to lower blood pressure.

You may still need other pain relief over the course of several days.

If necessary, doctors may give you other pain relief medicines.

**Computed tomography** ✕

A computed tomography (CT, or CAT) scan uses x-rays and a computer to make images of the organs and tissues inside the body. These images are more detailed than those from standard x-rays. A substance known as a contrast dye is sometimes given by mouth or injection to help make the images clearer.



# Managing kidney complications

This section explains the main complications of CKPD that can affect the kidneys, and how these can be managed.

## Kidney cyst infections

Kidney cysts can become infected with bacteria. This can sometimes occur in the presence of kidney stones or if the cyst is large. It is important to see your doctor if you have a kidney cyst and you are experiencing any of the following symptoms:

**Symptoms:** ...

...

## Cyst rupture and infection

...

## Kidney stones

People with CKPD are at an increased risk of kidney stones. Kidney stones are deposits of the salts in urine. They can form in the kidney or in the ureter. They can cause pain and may lead to kidney failure. They may make you need to urinate more often. It is important to see your doctor if you have any of the following symptoms:

**Symptoms:** ...

...

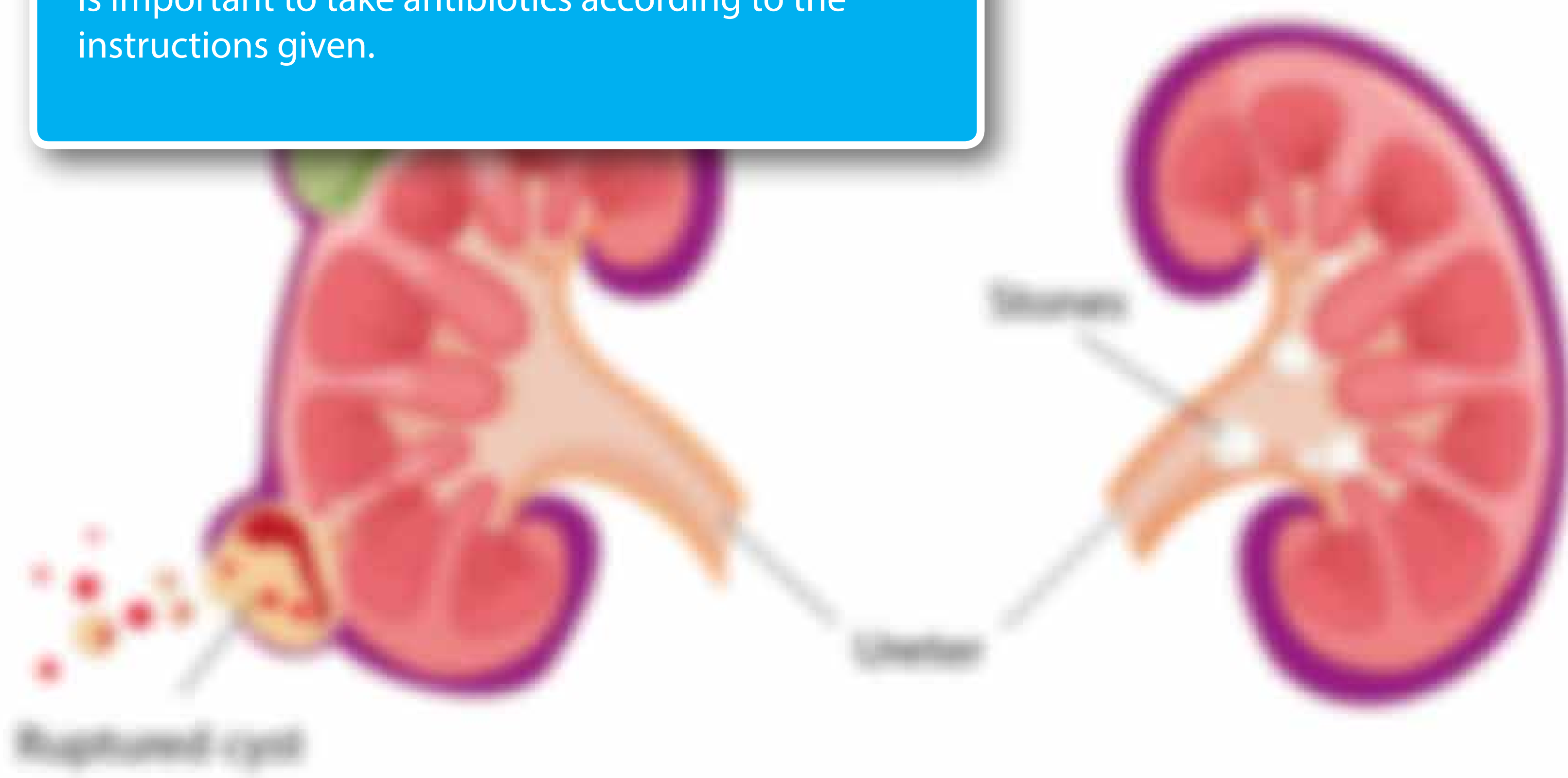
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**Antibiotics**

Antibiotics are medicines used to treat infections caused by bacteria. First-line antibiotics often used for cyst infections include members of the fluoroquinolone class (e.g. levofloxacin) and trimethoprim-sulfamethoxazole, although the choice depends on various factors and can differ between different countries and hospitals.

Cyst infections can be difficult to treat and they can return even after a course of antibiotics. It is important to take antibiotics according to the instructions given.



# Managing kidney complications

This section explains the main complications of CKPD that can affect the kidneys, and how these can be managed.

## Kidney cyst infections

Kidney cysts can become infected by bacteria. This can cause pain and fever in the abdomen. Cyst infections can be difficult to diagnose. Signs of a cyst infection include:

- Abdominal pain** - on the right side of the abdomen.

In some cases cysts may be treated either by surgery or medication in which a needle is inserted into the kidney through the skin.

## Cyst rupture and bleeding

Kidney cysts can sometimes burst or rupture and cause complications such as bleeding in the urine.

In most cases bleeding resolves without treatment within 7 days.

You may still need pain relief over the course of several days. If necessary, doctors may prescribe other pain relief medicines. If you have severe or persistent bleeding you should seek medical advice.

## Kidney stones

People with CKPD can get increased risk of kidney stones. Kidney stones are caused by the build up of certain chemicals in the urine. Large stones can block parts of the **ureter** and cause discomfort and pain. They may make you need to urinate more often than normal, or blood to appear in the urine.

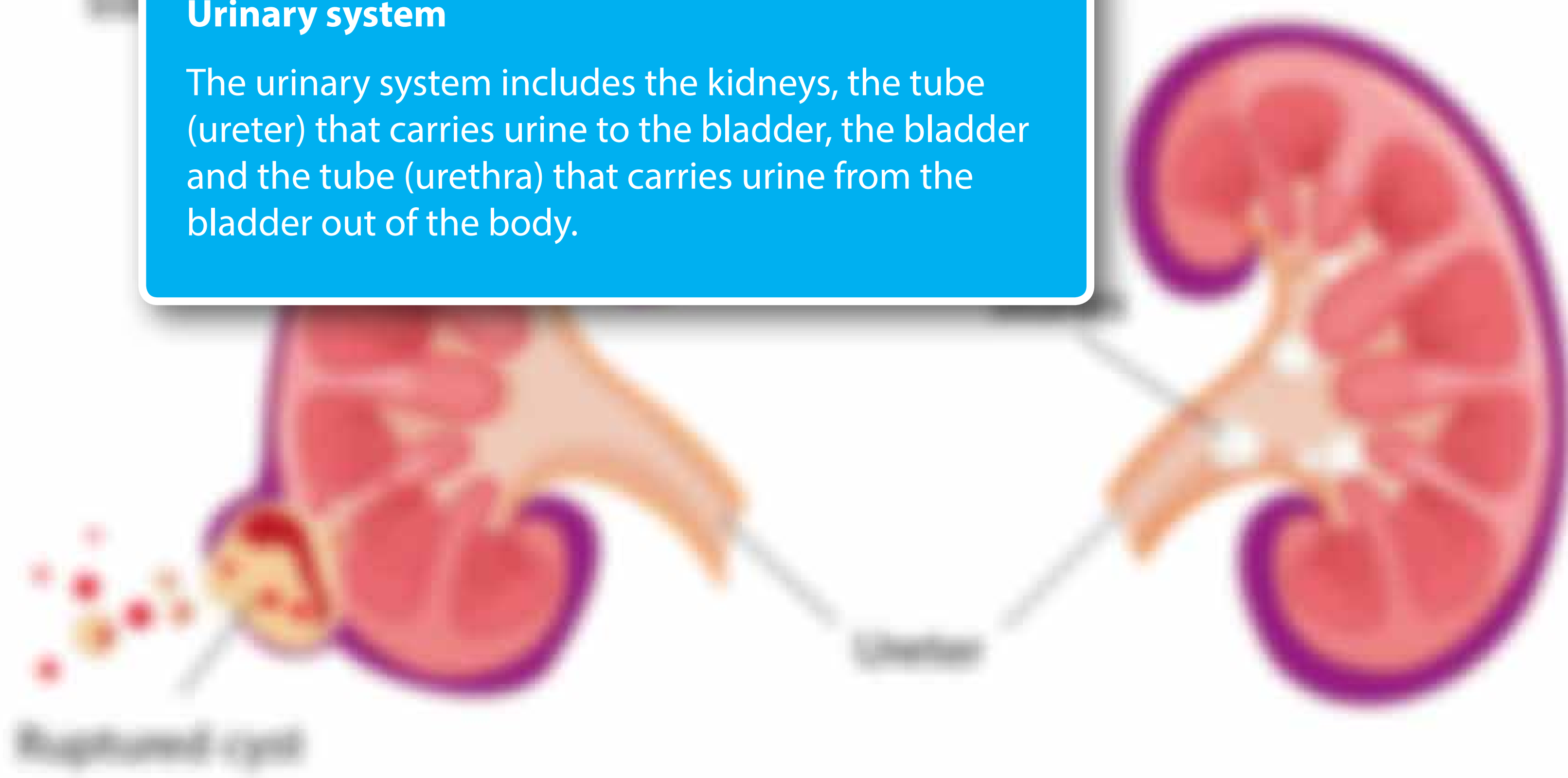
You should contact your doctor if you think you have kidney stones. Some smaller stones may pass naturally within 2-3 weeks, where this is possible. If you do experience pain you may also be able to have procedures that may be performed, and stones that are passed in the urine may be examined.

Small stones may be passed in the urine without treatment. Drinking plenty water to increase the urine flow can help to flush out stones. For **large stones**, medical help is necessary.

You may still need pain relief over the course of several days. If necessary, doctors may prescribe other pain relief medicines.

**Urinary system** ✕

The urinary system includes the kidneys, the tube (ureter) that carries urine to the bladder, the bladder and the tube (urethra) that carries urine from the bladder out of the body.





# Managing kidney complications

This section explains the main complications of CKPD that can affect the kidneys, and how these can be managed.

## Kidney cyst infections

Kidney cysts can become infected by bacteria. This can cause pain and discomfort in the abdomen. Cyst infections can be difficult to diagnose. Signs of a cyst infection include:

- Pain in the back or side
- Fever
- Blood in the urine

It is important to get treatment for cyst infections as soon as possible to prevent the infection from spreading to the rest of the kidney.

## Cyst rupture and bleeding

Kidney cysts can sometimes rupture and cause internal bleeding. This can cause pain and discomfort in the abdomen. Signs of a ruptured cyst include:

- Pain in the back or side
- Blood in the urine
- Blood in the stool

## Kidney stones

People with CKPD can get an increased risk of kidney stones. Kidney stones are deposits of the salts in urine. They can be very painful. Signs of kidney stones include:

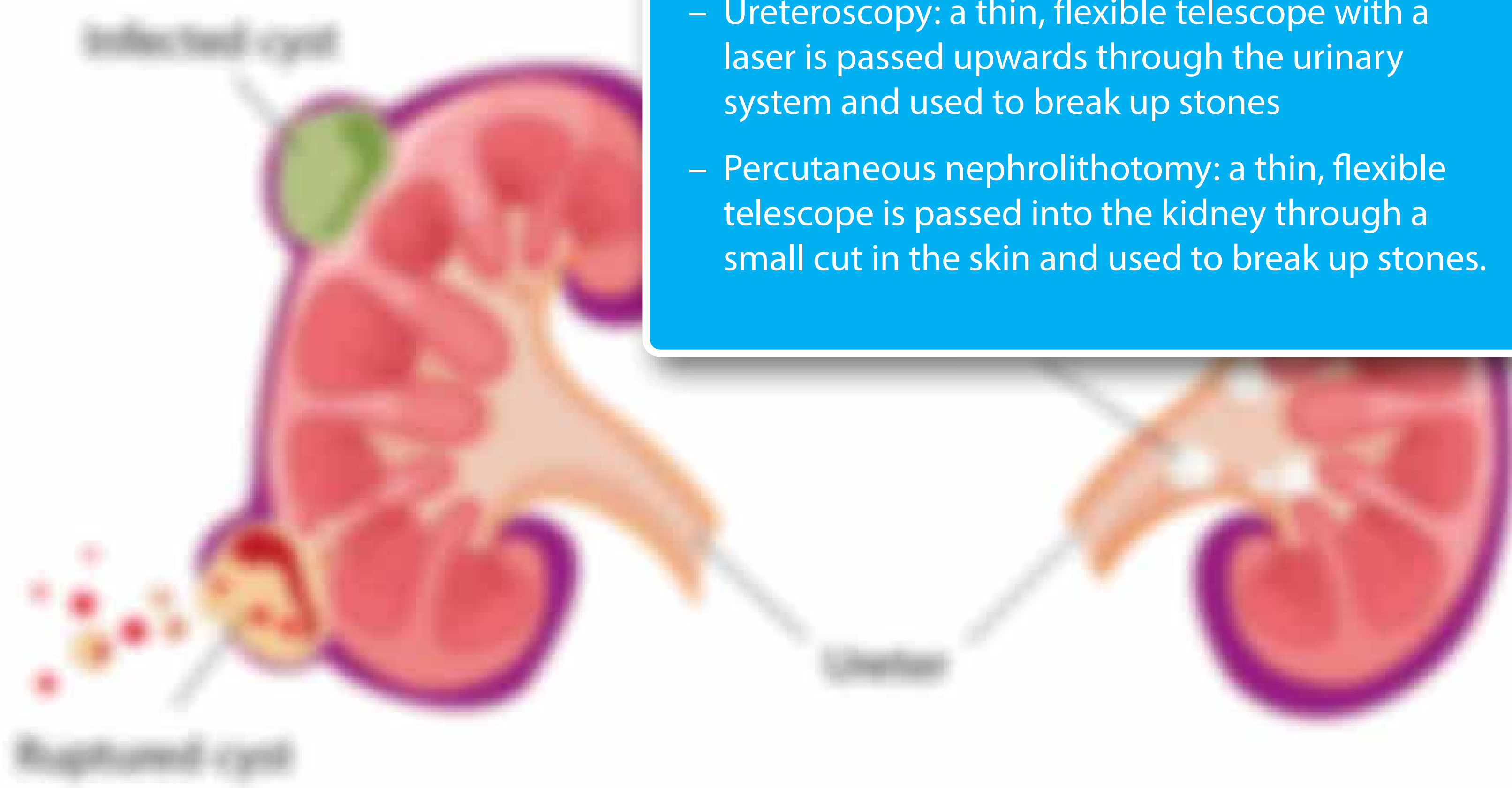
- Pain in the back or side
- Blood in the urine
- Nausea and vomiting

### Larger stones

Larger kidney stones need medical care, and you may need to be referred to a **urologist** (a doctor who specialises in diseases of the urinary system).

Possible treatments for larger kidney stones include:

- Extracorporeal shockwave lithotripsy: this uses ultrasound to break up stones so that they can be passed in the urine
- Ureteroscopy: a thin, flexible telescope with a laser is passed upwards through the urinary system and used to break up stones
- Percutaneous nephrolithotomy: a thin, flexible telescope is passed into the kidney through a small cut in the skin and used to break up stones.



# Managing liver and brain complications

This section explains the complications of ADPKD that can affect the liver and brain, and how these can be managed.

## Liver

[Liver cysts](#) do not cause symptoms, or need treatment, in most patients. However, they can become infected and, when large, can cause significant pain and discomfort.

### Did you know?

A [questionnaire](#) called the Polycystic Liver Disease Questionnaire (PLD-Q) can help assess the effect of liver cyst symptoms on wellbeing.

Patients with liver cysts causing symptoms should be referred to a [hepatologist](#) >.

## Cyst infections

Liver cyst infections can cause pain in the abdomen, and fever. When severe, they are best diagnosed using a scan called positron emission tomography ([PET](#)) >.

Cyst infections are treated mainly with [antibiotics](#) >.

## Reducing the cyst 'burden'

If you have polycystic liver disease, it is important that you and your hepatologist agree on the [goal of treatment](#) >, because this will guide the treatment options that are most appropriate for you.

[Surgery](#) > can be used to reduce the number and size of liver cysts causing severe symptoms.

Liver transplantation is an option for some patients with very severe liver cysts, but is rarely needed.

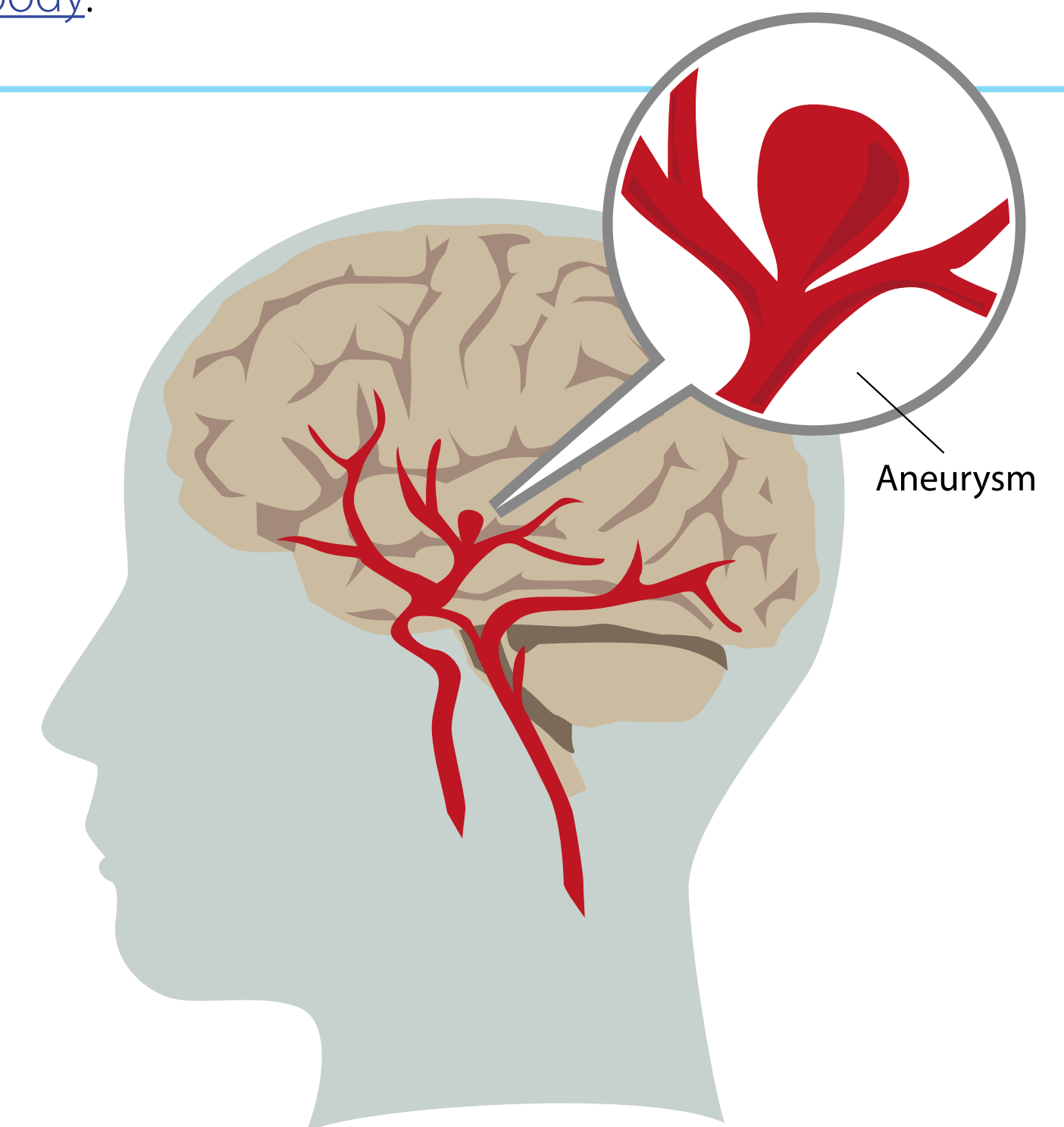
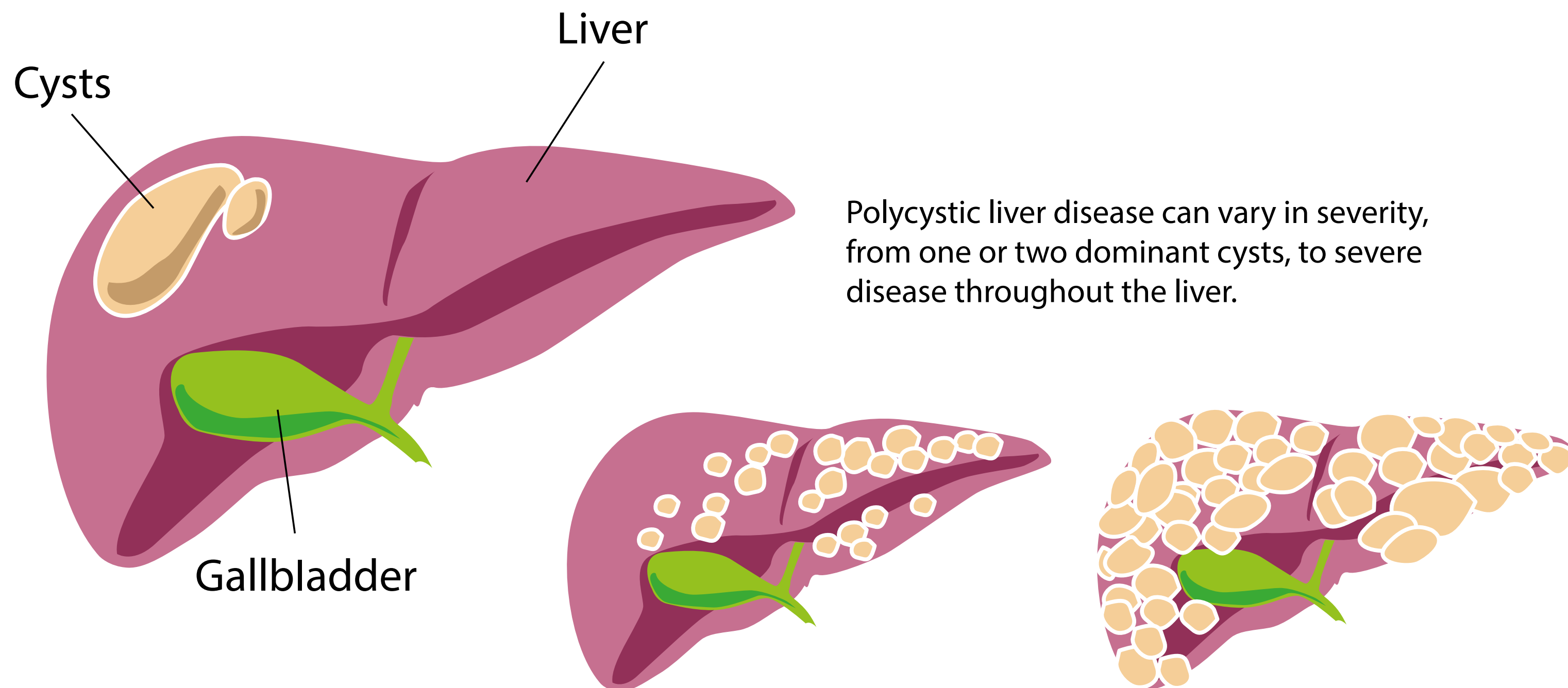
[New medicines](#) are being investigated to help treat liver cysts. If you are interested in joining a [clinical trial](#) you should discuss this with your hepatologist.

## Brain aneurysm

Patients found to have intact [brain aneurysms](#) should ideally be treated by a multidisciplinary team, including a [neurosurgeon](#) > and [neurovascular radiologist](#). [Surgery](#) > is sometimes performed to help prevent an aneurysm from bursting (rupturing) by stopping its blood supply.

A burst aneurysm causes bleeding in the brain. You should call an ambulance if you or someone you know with ADPKD has [symptoms](#) > that you think might be caused by a ruptured brain aneurysm.

Other sections explain the management of effects that ADPKD can have in the [kidneys](#) and [elsewhere in the body](#).



# Managing liver and brain complications

This section explains the complications of ALD (ALD) that can affect the liver and brain, and how these can be managed.

**Liver**  
ALD can lead to liver complications, such as liver disease and liver failure. However, they can be managed with medication and lifestyle changes. For more information on liver complications, see the [Liver complications](#) page.

**Brain disease**  
ALD can lead to brain complications, such as liver disease. However, they can be managed with medication and lifestyle changes. For more information on brain complications, see the [Brain complications](#) page.

People with liver complications may experience health problems such as [ascites](#).

**Cyst infections**  
Liver complications can lead to cyst infections. However, they can be managed with medication and lifestyle changes. For more information on cyst infections, see the [Cyst infections](#) page.

**Reducing the cyst burden**  
There are ways to reduce the number of cysts in the liver. This can be done with medication and lifestyle changes. For more information on reducing the cyst burden, see the [Reducing the cyst burden](#) page.

**Diagnosis**  
Diagnosis can be used to reduce the number and size of liver cysts, leading to better symptoms.

Liver transplantation is an option for some patients with liver complications, but it is only needed in some cases.

**Prognosis**  
Prognosis is the chance of recovery from liver disease. It can be managed with medication and lifestyle changes. For more information on prognosis, see the [Prognosis](#) page.

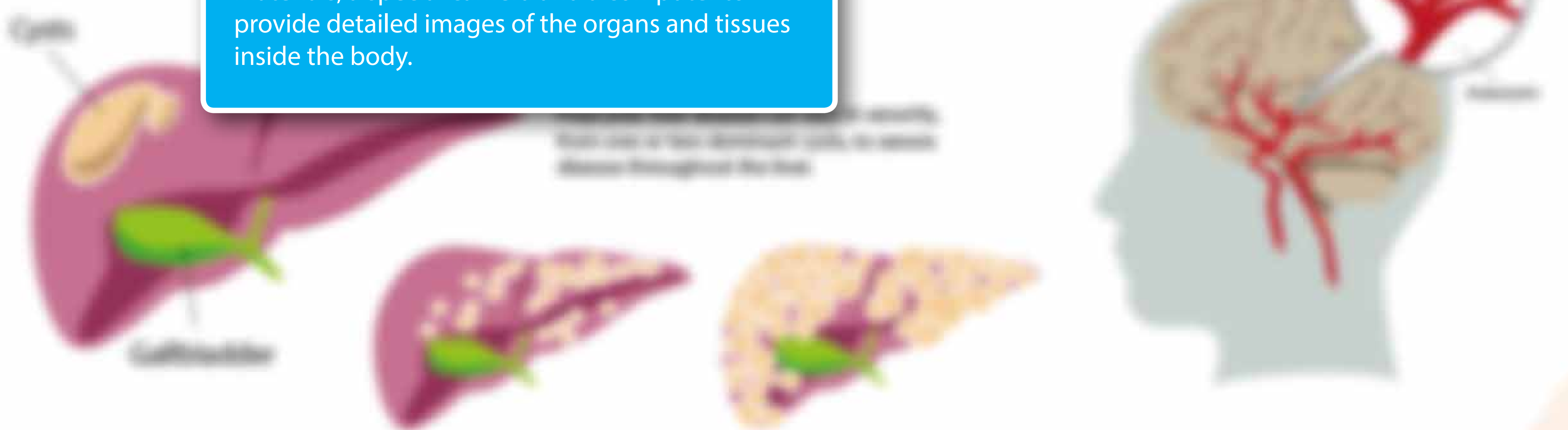
**Brain symptoms**  
Brain symptoms can be managed with medication and lifestyle changes. For more information on brain symptoms, see the [Brain symptoms](#) page.

Brain symptoms can be managed with medication and lifestyle changes. For more information on brain symptoms, see the [Brain symptoms](#) page.

Other sections explain the management of effects from ALD on the [liver](#) and [pancreas](#).

**Positron emission tomography**

Positron emission tomography (PET) is a type of scan that uses small amounts of radioactive materials, a special camera and a computer to provide detailed images of the organs and tissues inside the body.



# Managing liver and brain complications

This section explains the complications of ALD (ALD) that can affect the liver and brain, and how these can be managed.

## Liver

ALD can lead to liver complications, such as liver disease, cirrhosis, liver cancer, liver failure and liver transplantation and liver failure.

## Brain disease

ALD can lead to brain complications, such as liver disease, cirrhosis, liver failure and liver transplantation and liver failure.

ALD can lead to brain complications, such as liver disease, cirrhosis, liver failure and liver transplantation and liver failure.

## Cyst infections

Cyst infections can be difficult to treat and they can return even after a course of antibiotics. It is important to take antibiotics according to the instructions given.

## Reducing the cyst burden

ALD can lead to liver complications, such as liver disease, cirrhosis, liver failure and liver transplantation and liver failure.

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## Brain symptoms

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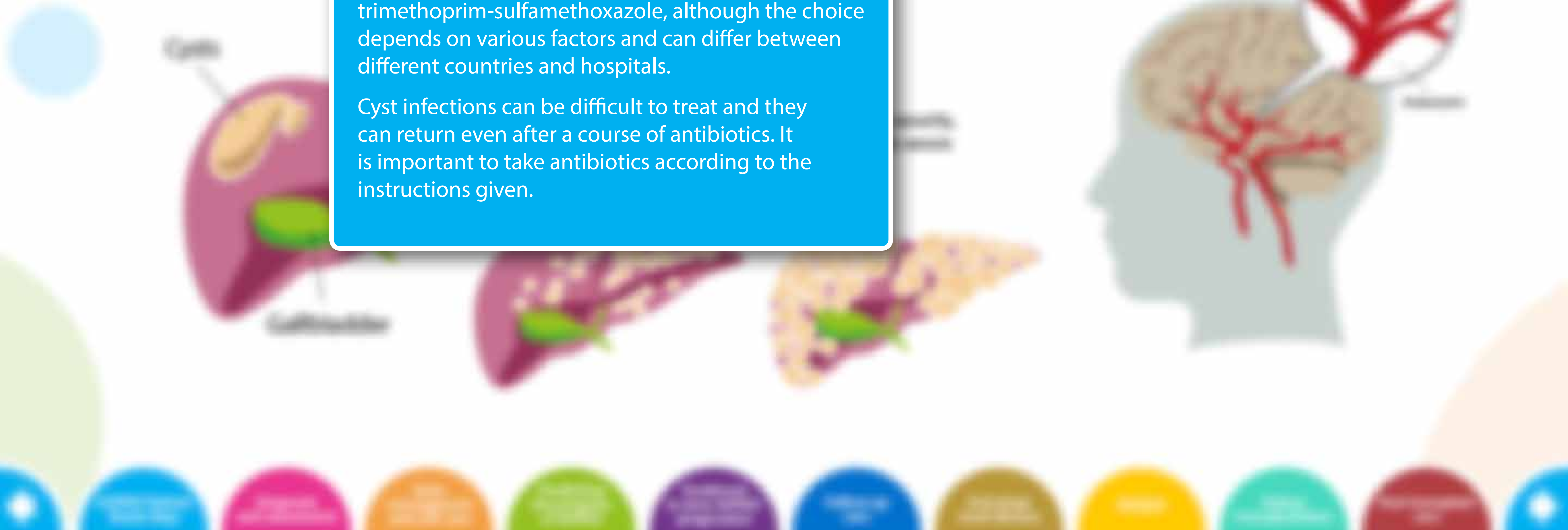
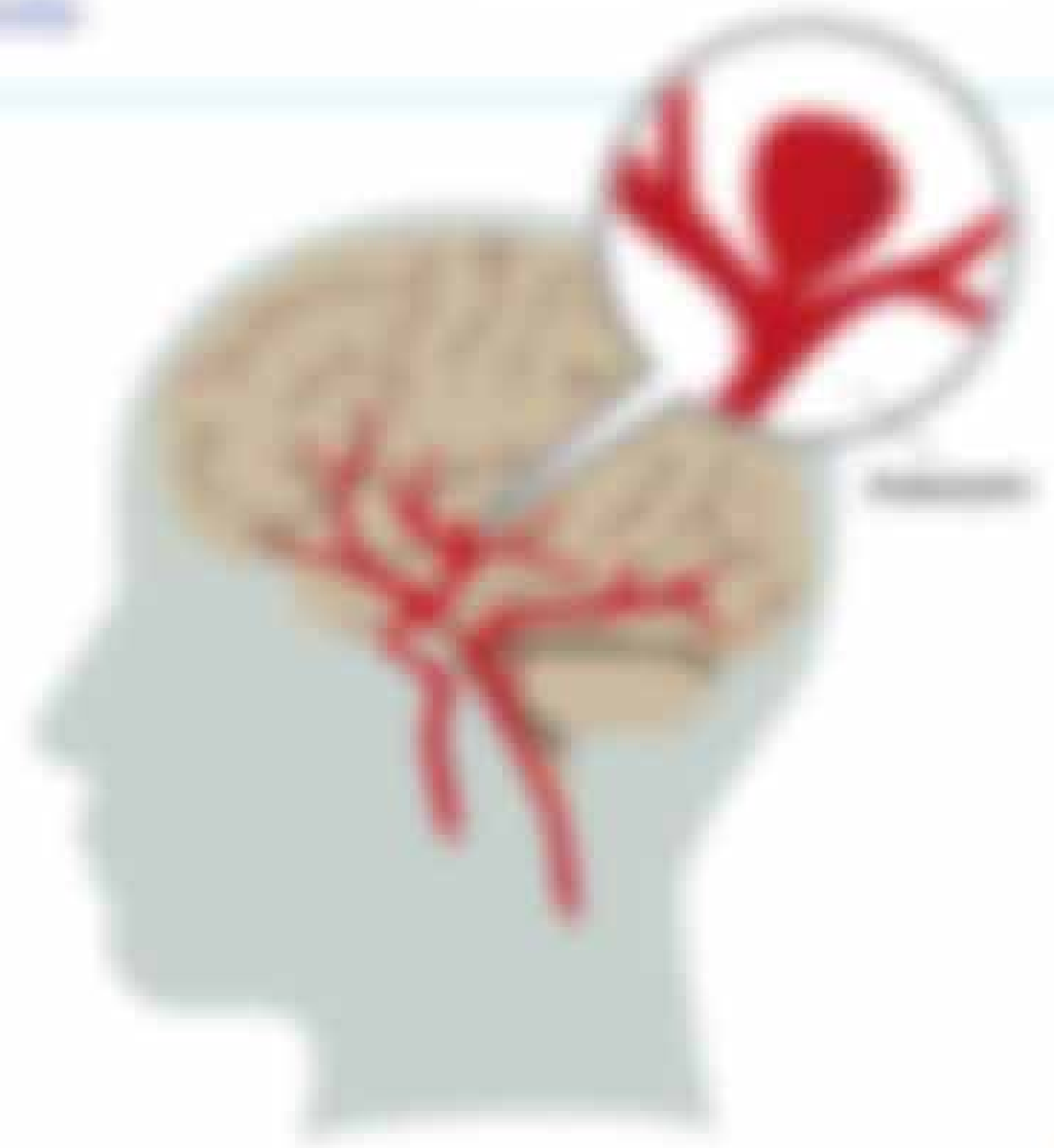
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**Antibiotics**

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Cyst infections can be difficult to treat and they can return even after a course of antibiotics. It is important to take antibiotics according to the instructions given.

Other sections explain the management of effects from ALD on the liver and brain.



# Managing liver and brain complications

This section explains the complications of ALD (ALD) that can affect the liver and brain, and how these can be managed.

## Liver

[Liver](#) - ALD can cause symptoms, as well as treatment. It can cause cirrhosis, liver failure, liver cancer, liver metastases and liver transplantation. See also [liver](#) and [liver](#).

## Brain

[Brain](#) - ALD can cause symptoms, as well as treatment. It can cause cirrhosis, liver failure, liver cancer, liver metastases and liver transplantation. See also [liver](#) and [liver](#).

## Cyst infections

[Cyst infections](#) - ALD can cause symptoms, as well as treatment. It can cause cirrhosis, liver failure, liver cancer, liver metastases and liver transplantation. See also [liver](#) and [liver](#).

## Reducing the cyst burden

[Reducing the cyst burden](#) - ALD can cause symptoms, as well as treatment. It can cause cirrhosis, liver failure, liver cancer, liver metastases and liver transplantation. See also [liver](#) and [liver](#).

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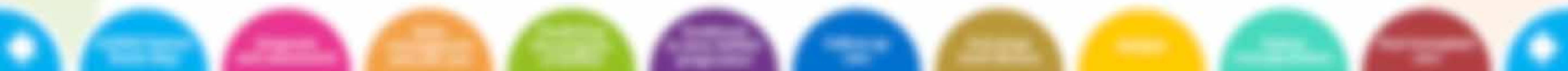
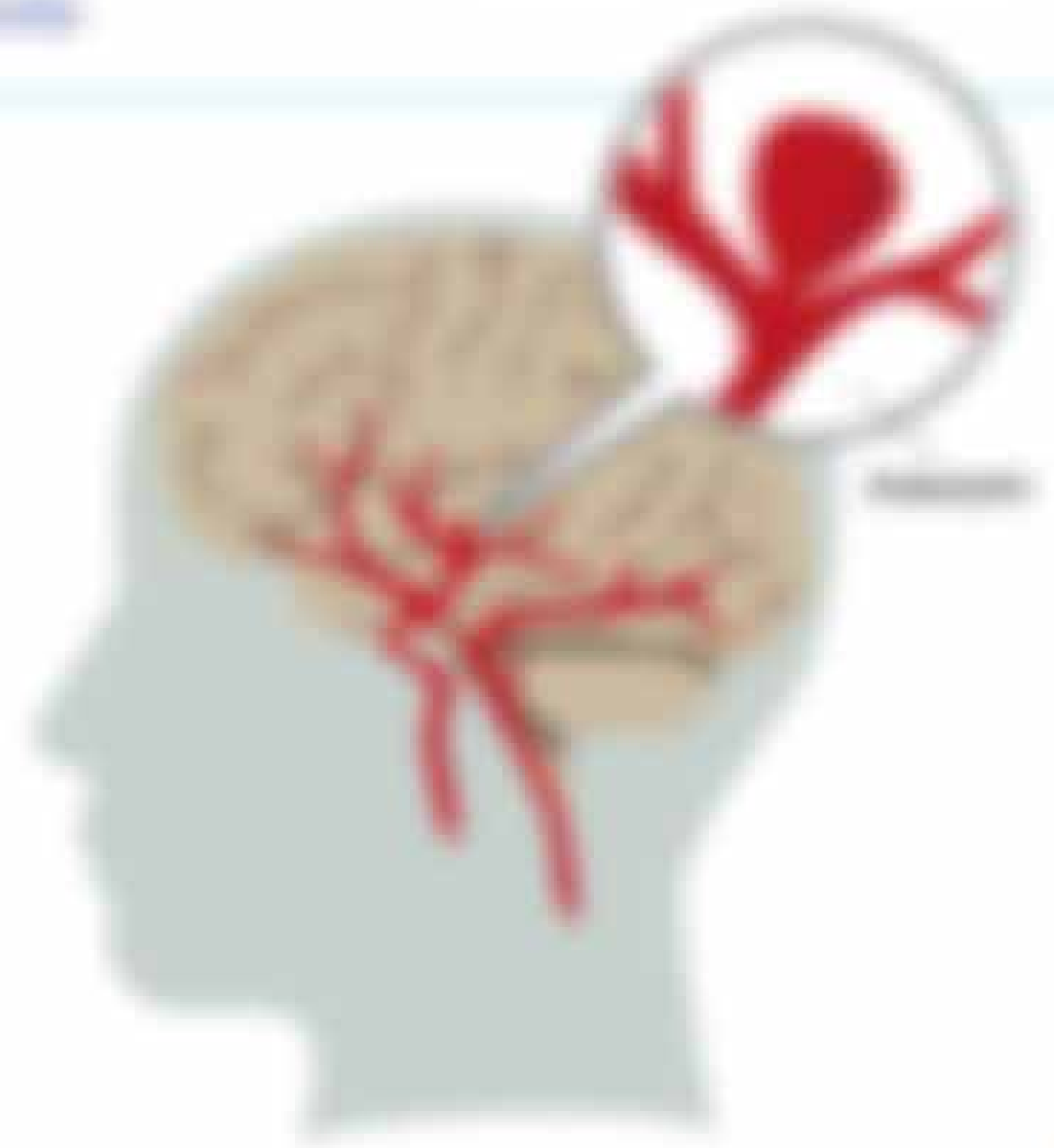
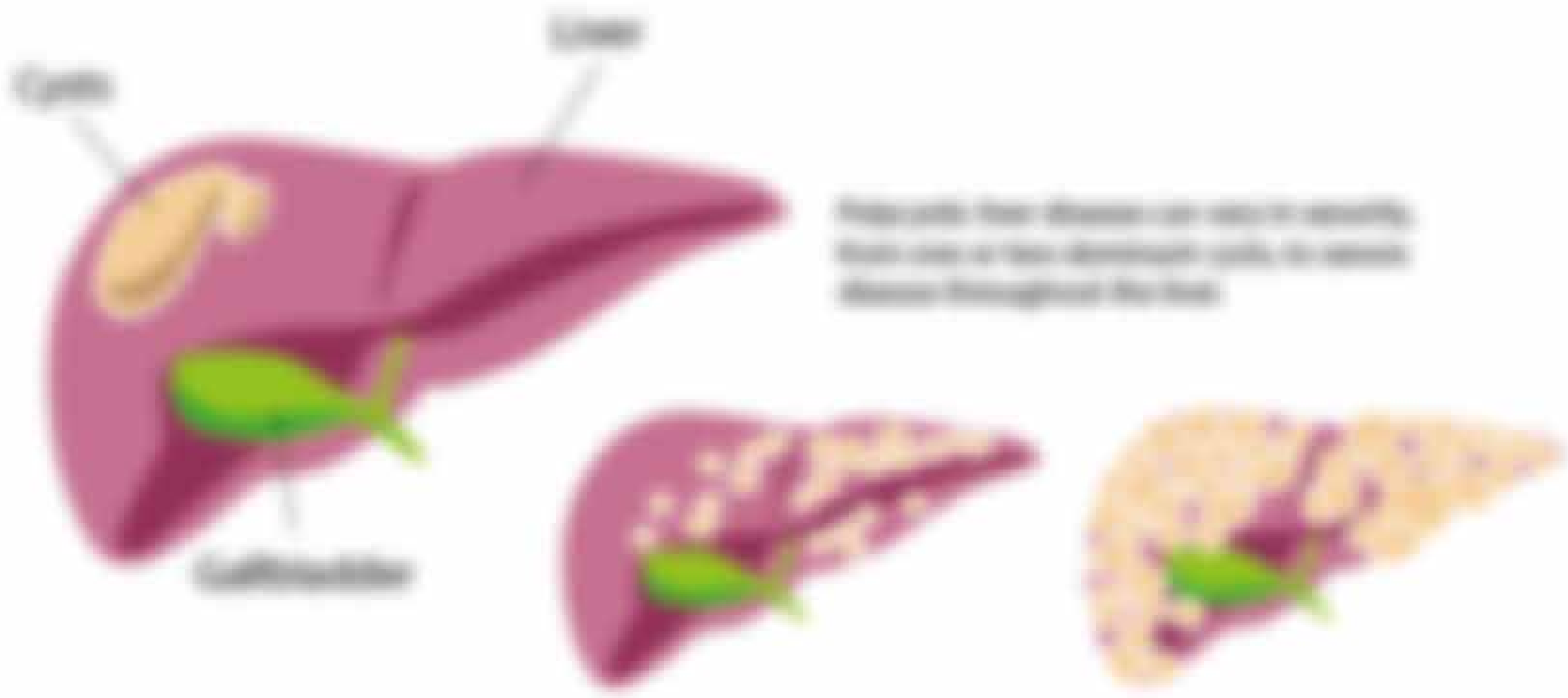
## Brain aneurysms

[Brain aneurysms](#) - ALD can cause symptoms, as well as treatment. It can cause cirrhosis, liver failure, liver cancer, liver metastases and liver transplantation. See also [liver](#) and [liver](#).

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**Hepatologist**  
A hepatologist is a doctor who specialises in diseases of the liver.

Other sections explain the management of effects that ALD can have on the [liver](#) and [brain](#).



# Managing liver and brain complications

This section explains the complications of ACPHD that can affect the liver and brain, and how these can be managed.

## Liver

**Enlarged liver** - An enlarged liver (hepatomegaly) is a common complication of ACPHD. It can cause discomfort, bloating, and can sometimes affect your ability to eat and drink.

## Ascites

**Ascites** is a condition where fluid builds up in the abdomen. It can help ease the effects of liver complications on walking.

People with liver complications may experience weight loss. [Read more](#)

## Cyst infections

Cyst infections can cause pain in the liver. When severe, they can lead to abscesses. [Read more](#)

## Reducing the cyst burden

From time to time, liver disease can progress. The aim of your treatment is to reduce the **cyst burden** - the number and size of the cysts in your liver.

**Diuretics** can be used to reduce the number and size of liver cysts causing severe symptoms.

Some medications can be used for some patients with liver complications, but it's not always possible.

**Transcatheter arterial chemoembolisation (TACE)** is a treatment option for some patients with liver complications. [Read more](#)

## Brain complications

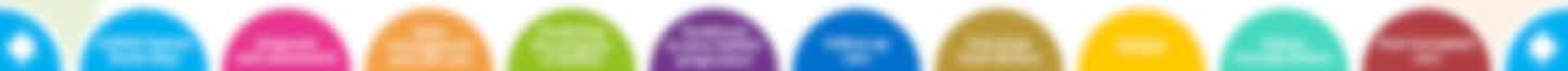
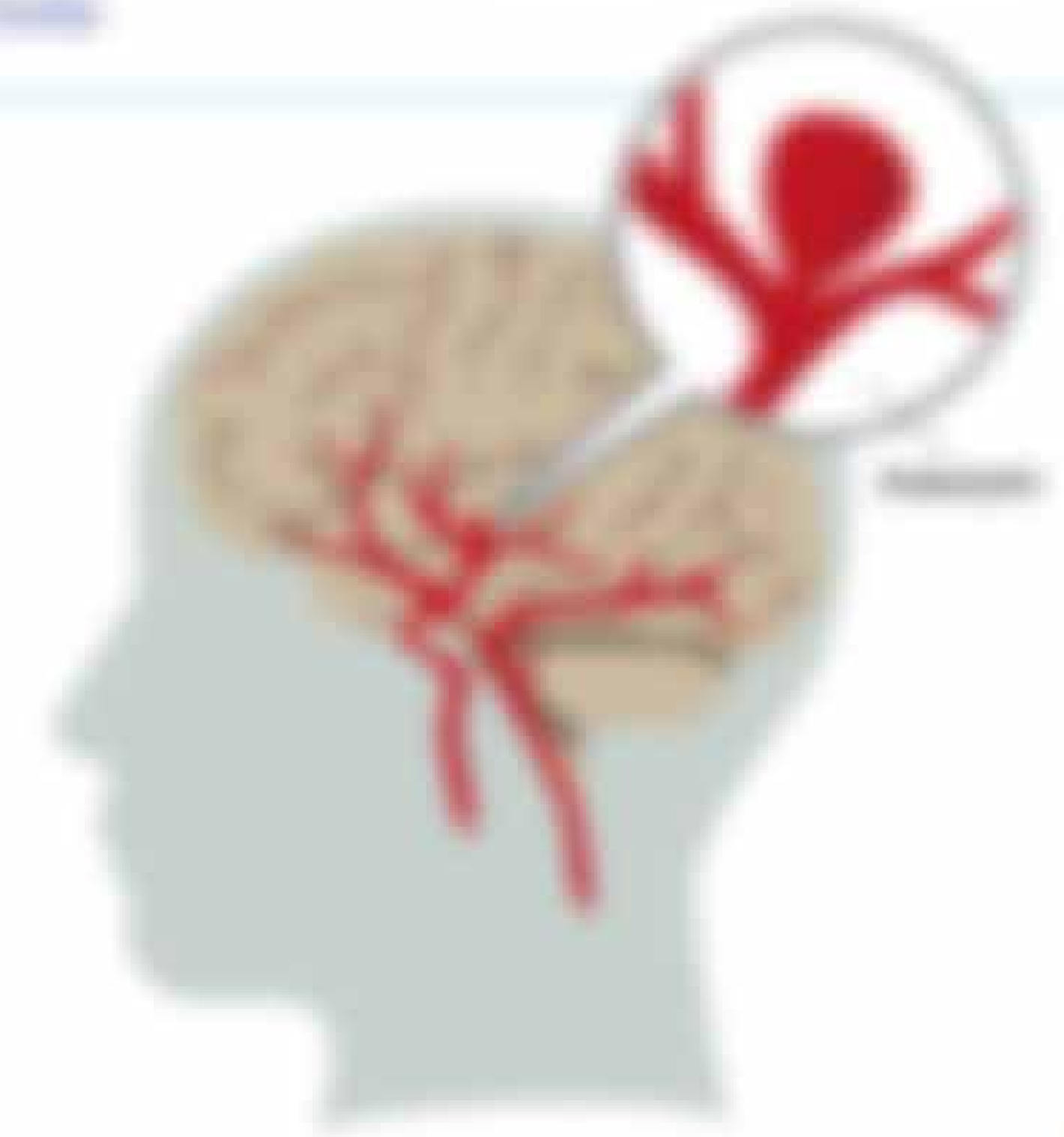
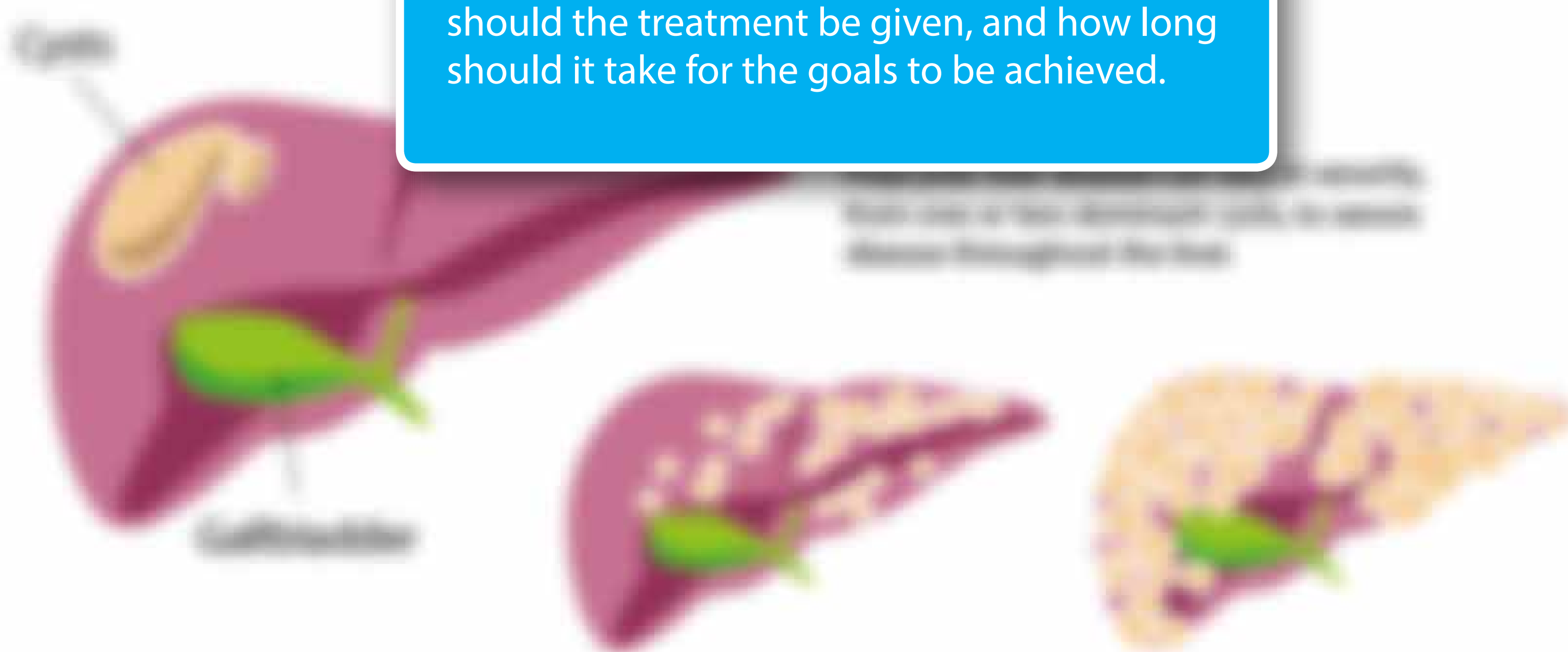
Some patients may experience **brain complications** due to the effects of liver disease, including **hypertension** and **intracerebral haemorrhage**. **Hydrocephalus** is a condition where fluid builds up in the brain, which can lead to swelling and pressure on the brain.

Brain complications can be managed in the brain. The goals of an **anticoagulant** are to prevent you from getting **ACPHD complications** that can affect your quality of life or health.

**Goal of treatment**

The goals of treatment may include reducing the size of the enlarged liver, but also improving your quality of life and/or reducing the symptoms caused by the liver cysts. Topics for discussion include: how long should the treatment be given, and how long should it take for the goals to be achieved.

Other sections explain the management of effects that ACPHD can have on the [liver](#) and [kidneys](#).



# Managing liver and brain complications

This section explains the complications of ALD (ALD) that can affect the liver and brain, and how these can be managed.

### Liver

ALD can damage your liver, which is your body's main filter. It can also cause other complications, such as:

- Ascites** - a build-up of fluid in the abdomen
- Encephalopathy** - a condition that affects the brain
- Hypertension** - high blood pressure
- Portal hypertension** - high blood pressure in the liver
- Varicose veins** - swollen veins in the legs
- Yellowing of the skin** - jaundice

### Cyst infections

Cyst infections can cause pain in the abdomen and fever. When severe, they can lead to hospital admission. Cyst infections are treated with **antibiotics**.

### Reducing the cyst burden

There are several ways to reduce the number of cysts in your liver. These include:

- Medication** - taking medicine to help reduce the number of cysts.
- Procedures** - using procedures to help reduce the number of cysts.

### Brain complications

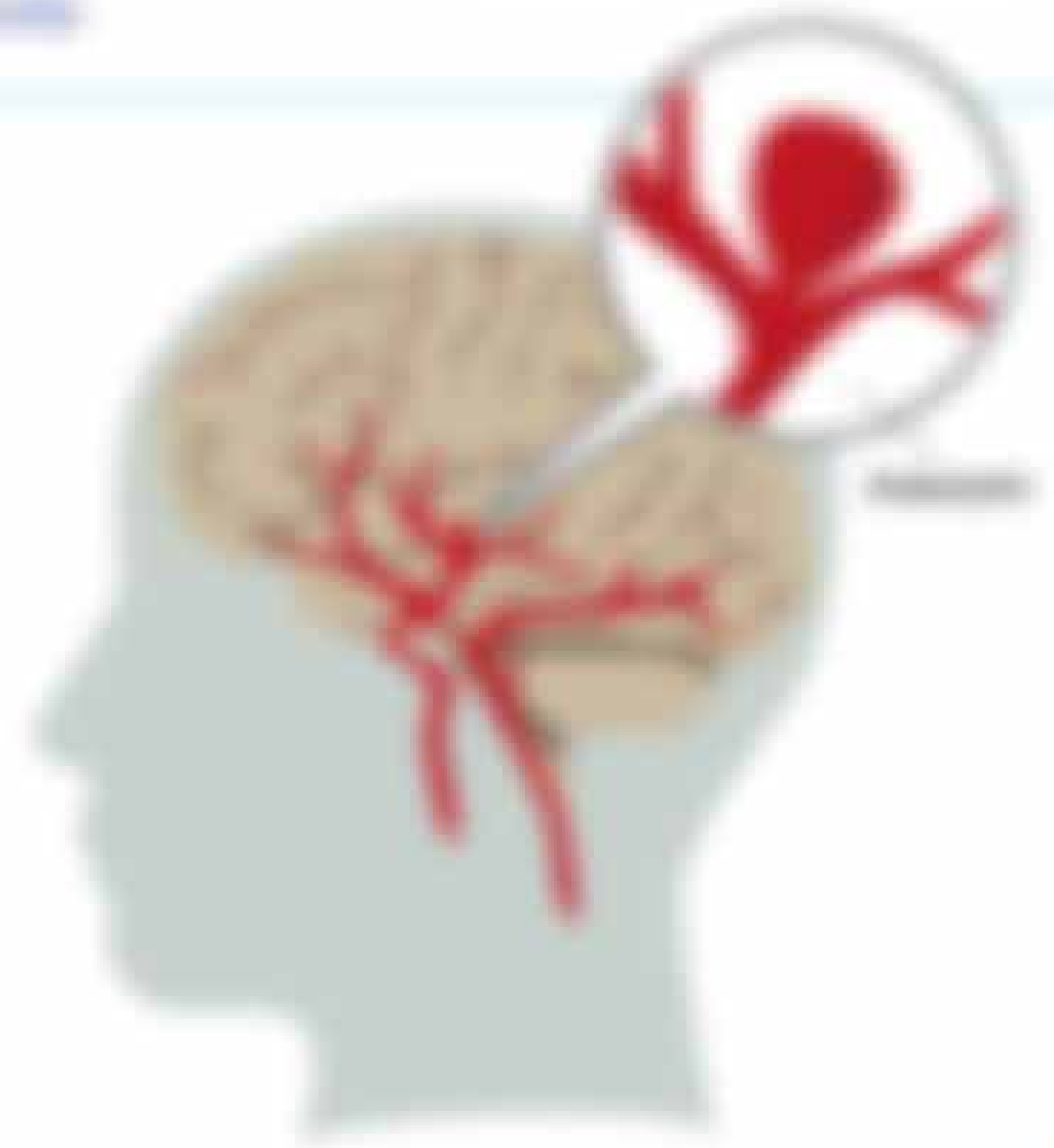
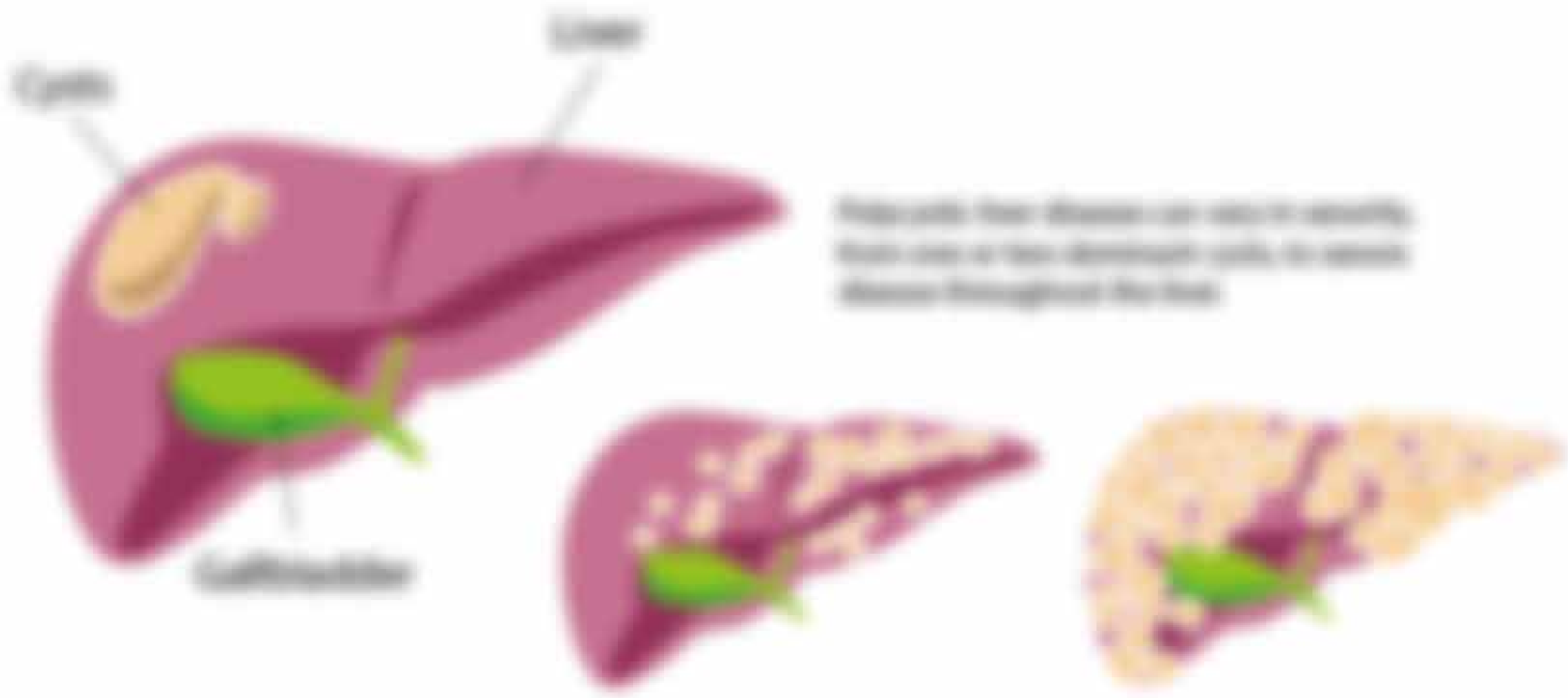
ALD can cause complications in the brain, such as:

- Encephalopathy** - a condition that affects the brain.
- Hypertension** - high blood pressure.
- Portal hypertension** - high blood pressure in the liver.

**Neurosurgeon**

A neurosurgeon specialises in the diagnosis and surgical treatment of disorders of the nervous system, including the brain, spinal cord and other nerves throughout the body.

This section explains the management of effects that ALD can have on the **liver** and **brain**.



# Managing liver and brain complications

This section explains the complications of ALD (ALD) that can affect the liver and brain, and how these can be managed.

### Liver

[Liver](#) - liver disease symptoms, signs and treatment. Liver disease symptoms, signs and treatment. Liver disease symptoms, signs and treatment. Liver disease symptoms, signs and treatment.

### Brain

[Brain](#) - brain disease symptoms, signs and treatment. Brain disease symptoms, signs and treatment. Brain disease symptoms, signs and treatment. Brain disease symptoms, signs and treatment.

### Cyst infections

[Cyst infections](#) - cyst infections can occur in the liver and brain. Cyst infections can occur in the liver and brain. Cyst infections can occur in the liver and brain. Cyst infections can occur in the liver and brain.

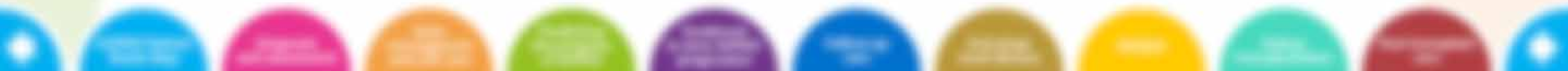
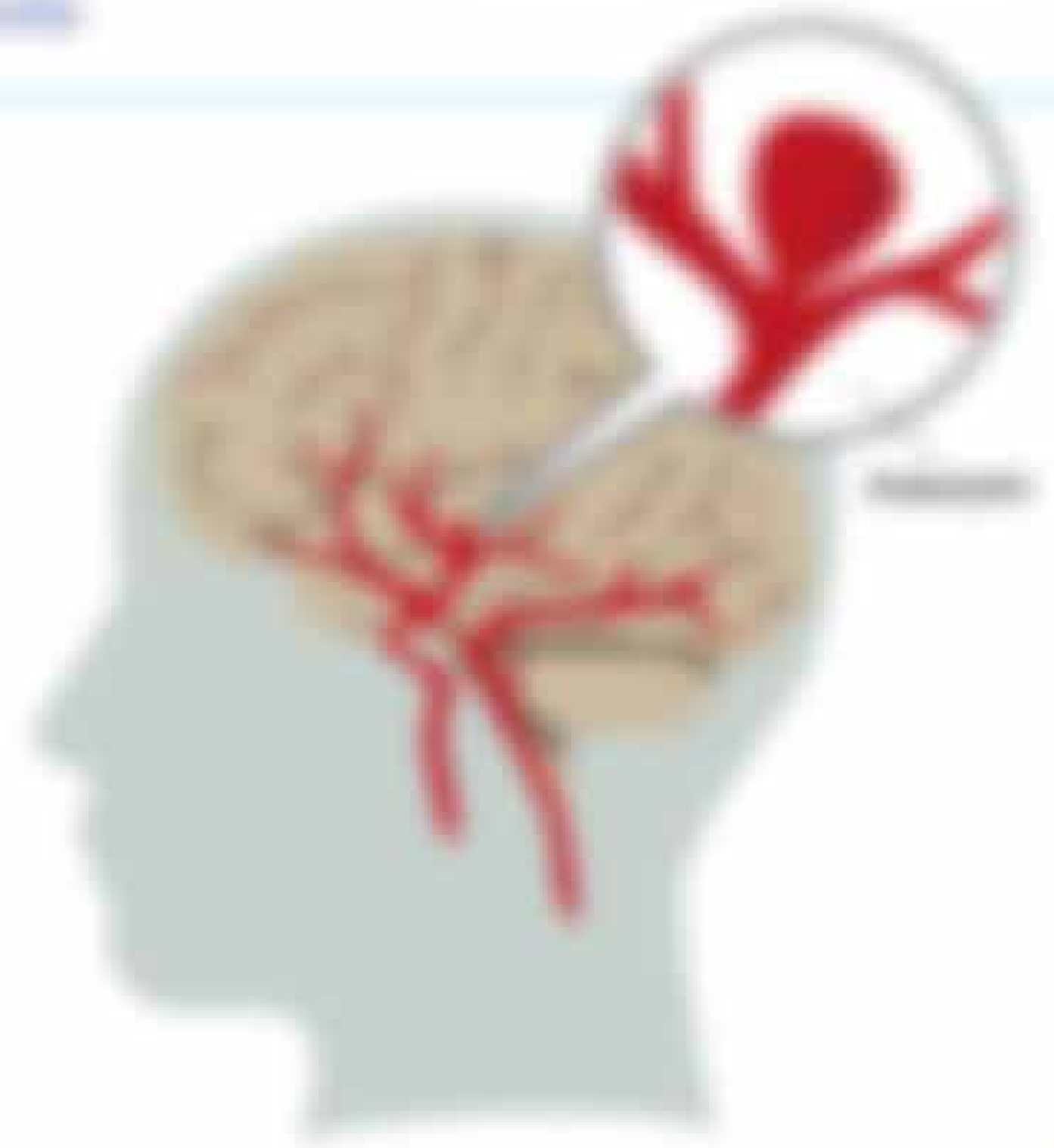
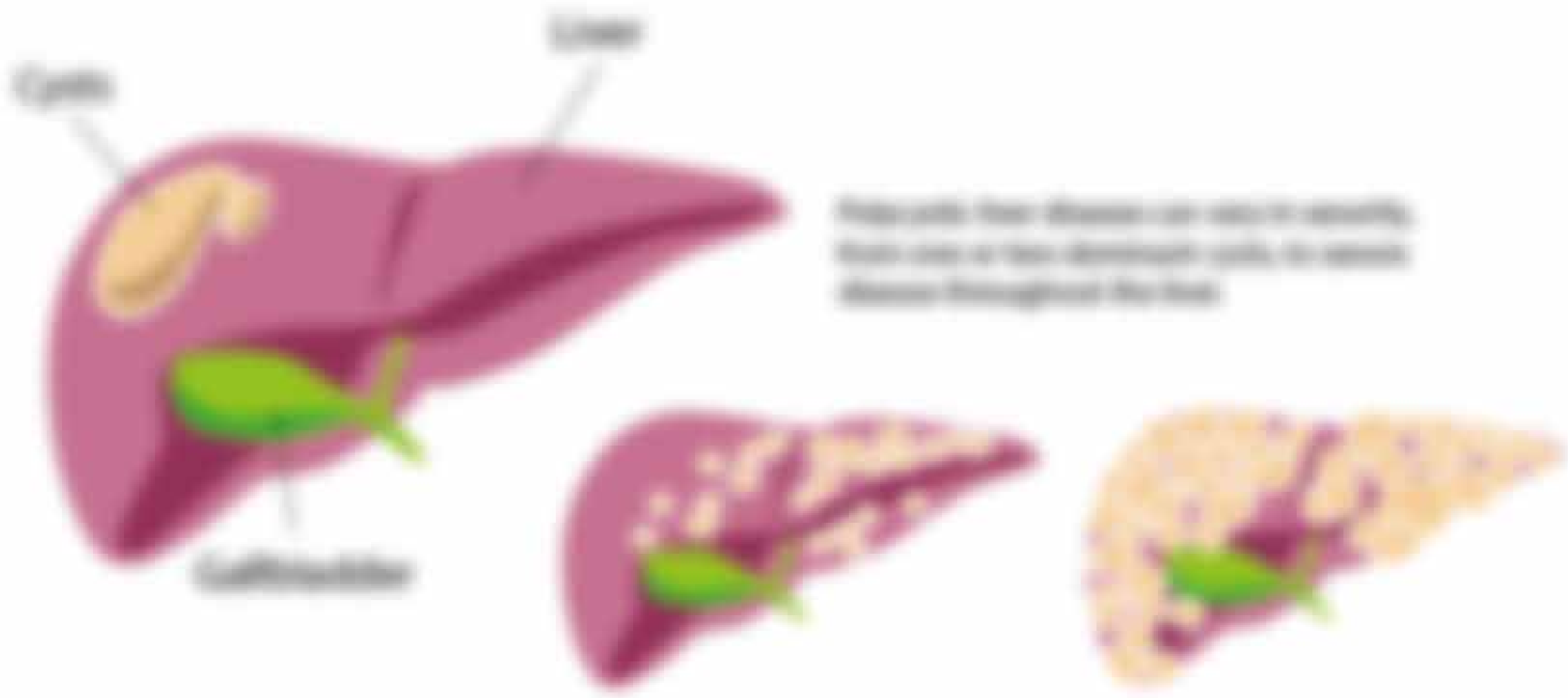
### Reducing the cyst burden

[Reducing the cyst burden](#) - reducing the cyst burden. Reducing the cyst burden. Reducing the cyst burden. Reducing the cyst burden.

**Neurovascular radiologist**

An interventional neurovascular radiologist specialises in 'minimally invasive' image-based surgical procedures (or 'interventions') used in the diagnosis and treatment of diseases of the head, neck and spine.

This section explains the management of effects of ALD (ALD) on the [liver](#) and [brain](#).





## Surgery: Liver cysts

Several procedures may be used to treat liver cysts, depending on the situation. It is recommended that these procedures should only be carried out by surgeons with specific expertise in polycystic liver disease.

### *Aspiration and sclerotherapy*

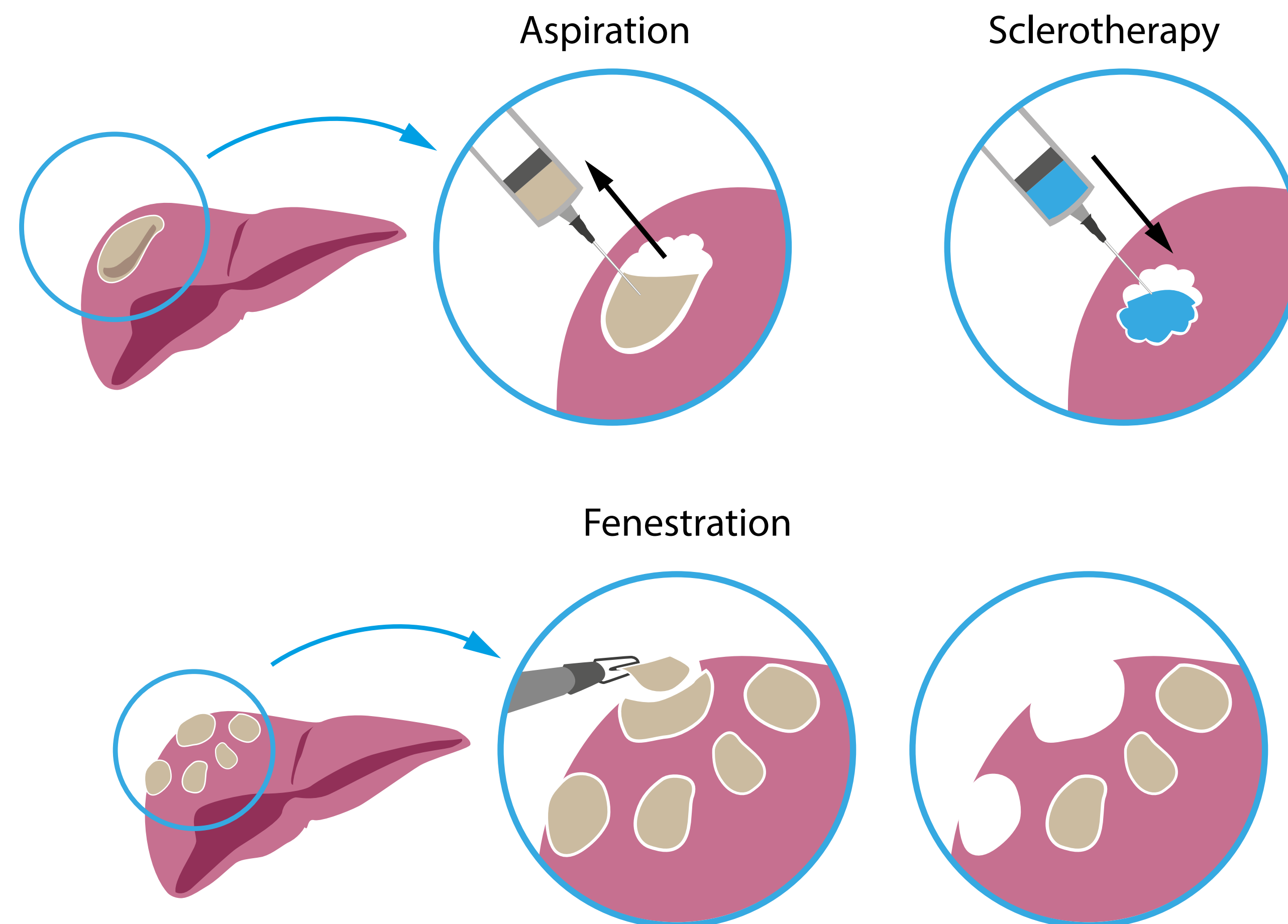
Aspiration is a procedure in which cysts are drained by inserting a needle through the skin, under local anaesthetic. Sclerotherapy involves flushing the cyst with a chemical solution (usually alcohol) to help stop the cyst filling up with fluid again. Aspiration sclerotherapy is normally used to treat patients with symptoms caused by a large dominant cyst (typically around 5 cm in diameter, or larger).

### *Fenestration*

After a cyst is cut open, part of its wall is removed so that it does not swell up again. This can usually be done using 'keyhole' (or laparoscopic) surgery, needing only a small cut in the skin. Having fenestration can prevent a liver transplant being needed in the future.

### *Segmental liver resection*

Surgery can be performed to remove parts of the liver where severe cysts are localised. This is normally done only if other treatments cannot be used or do not work.



# Managing liver and brain complications

This section explains the complications of ALD that can affect the liver and brain, and how these can be managed.

## Liver

[Liver cirrhosis](#) is a long-term condition where the liver becomes damaged and scarred. This can lead to liver failure and other complications such as [portal hypertension](#) and [ascites](#).

## Brain

[Liver encephalopathy](#) is a condition where toxins from the liver build up in the blood and affect the brain. This can lead to confusion, drowsiness and other symptoms of [liver encephalopathy](#).

People with liver cirrhosis may experience [bleeding](#) due to [portal hypertension](#).

## Cyst infections

[Liver cysts](#) are fluid-filled sacs that can form in the liver and brain. When present, they can cause symptoms such as [pain](#) and [pressure](#).

[Liver cysts](#) are treated with [medication](#).

## Brain complications

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## Cyst infections

[Liver cysts](#) are fluid-filled sacs that can form in the liver and brain. When present, they can cause symptoms such as [pain](#) and [pressure](#).

[Liver cysts](#) are treated with [medication](#).

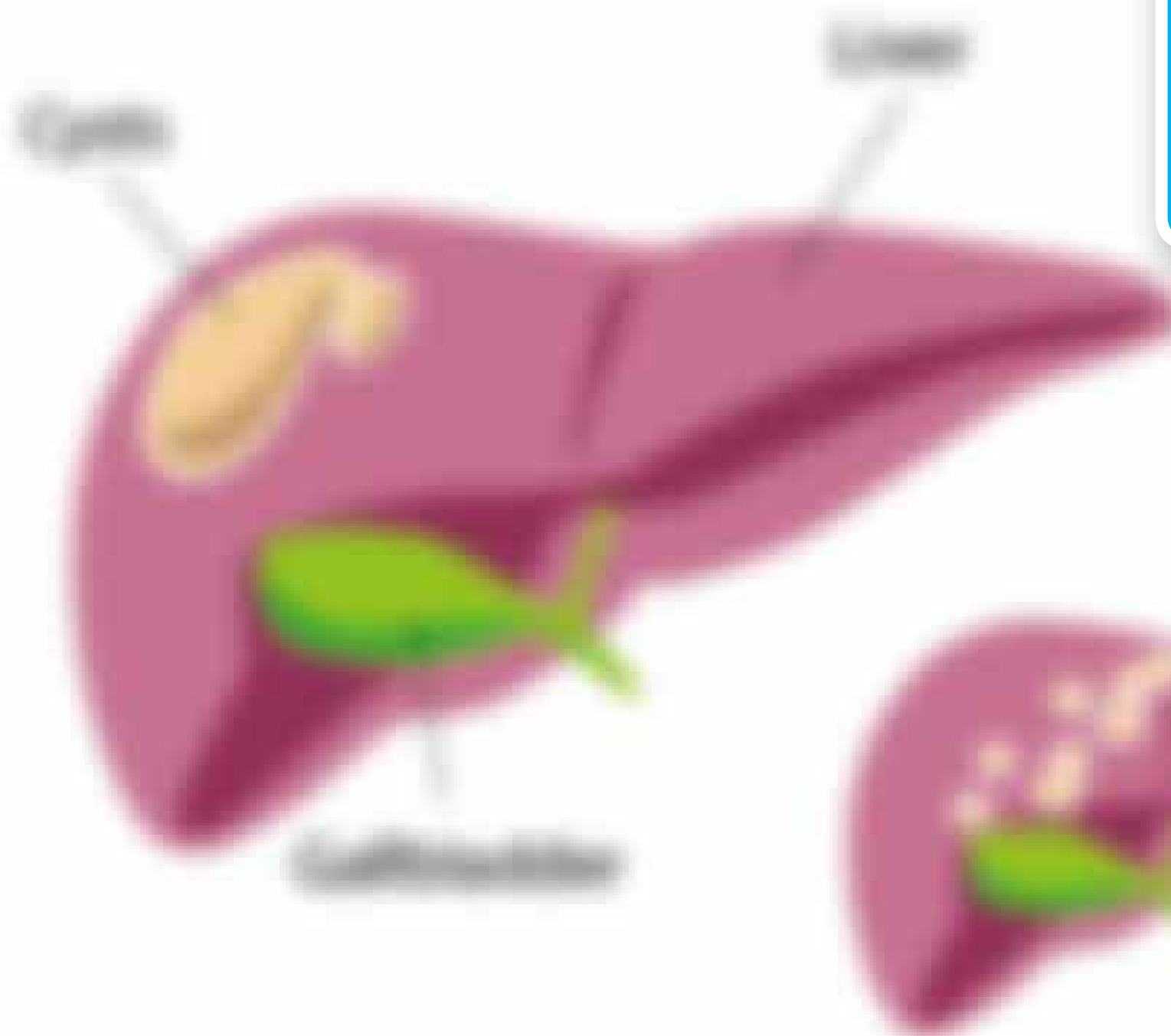
**Surgery: Aneurysms**

Surgery is sometimes performed to help prevent an aneurysm from bursting by stopping its blood supply. The two main methods are:

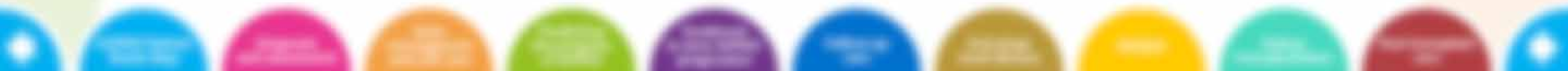
- Clipping: a metal clip is passed through a small opening in the skull and used to seal off the aneurysm
- Coiling: a tube is inserted into an artery in the leg or groin, passed to the aneurysm and used to block it with metal coils.

These procedures carry risks and so need careful consideration. The risks depend on the individual situation. If you are diagnosed with an unruptured aneurysm you may wish to discuss the risks with your healthcare team, especially a neurosurgeon or neurovascular radiologist.

It is recommended that patients with untreated small aneurysms should be assessed again every 6 to 24 months.



Flow chart showing the flow of blood and bile through the liver.



# Managing liver and brain complications

This section explains the complications of ALD (ALD) that can affect the liver and brain, and how these can be managed.

**Liver**  
ALD can lead to liver complications, as liver treatment is most effective if started early. However, this can become difficult and often requires liver transplantation and dialysis.

**Brain health**  
ALD can lead to brain complications, such as liver disease. However, this can help reduce the effects of liver complications on the brain.

People with liver complications may experience double vision or [blurred vision](#).

**Cyst infections**  
Liver complications can lead to cysts in the liver and brain. When these cysts are not treated, they can lead to liver complications or [brain infections](#).

Cyst infections are treated with [antibiotics](#).

**Reducing the cyst burden**  
People with liver disease can also have cysts. This can lead to liver complications, such as [portal hypertension](#). However, this can be managed with [liver transplantation](#) and [dialysis](#).

**Diagnosis**  
ALD can be diagnosed with a blood test and liver biopsy.

ALD can be managed with [liver transplantation](#) and [dialysis](#).

**Prognosis**  
ALD can lead to liver complications, such as [portal hypertension](#). However, this can be managed with [liver transplantation](#) and [dialysis](#).

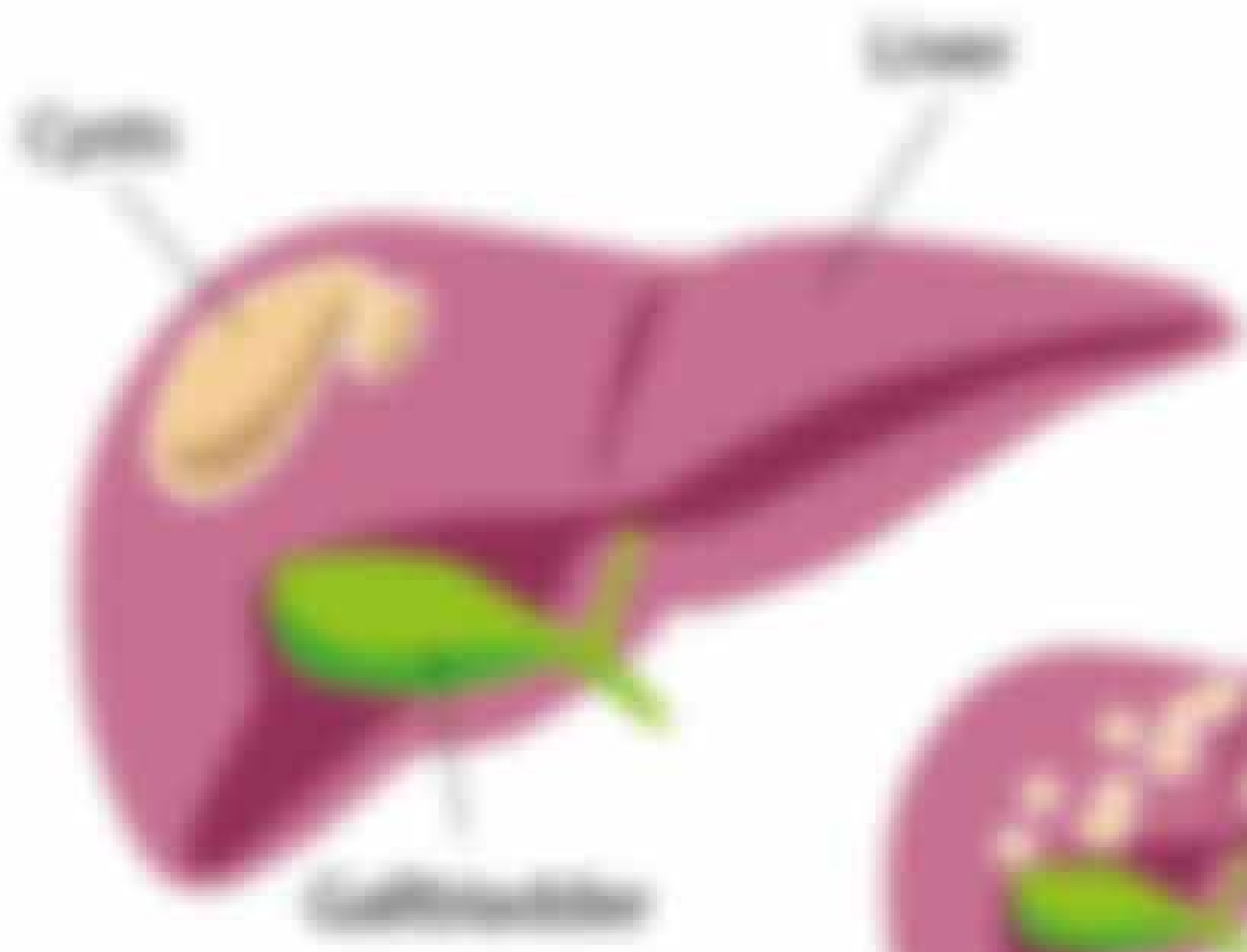
**Brain aneurysms**  
People with liver disease may have [brain aneurysms](#). These can lead to [stroke](#) or [brain aneurysms](#). However, this can be managed with [liver transplantation](#) and [dialysis](#).

People with liver disease may experience double vision or [blurred vision](#). This can be managed with [liver transplantation](#) and [dialysis](#).

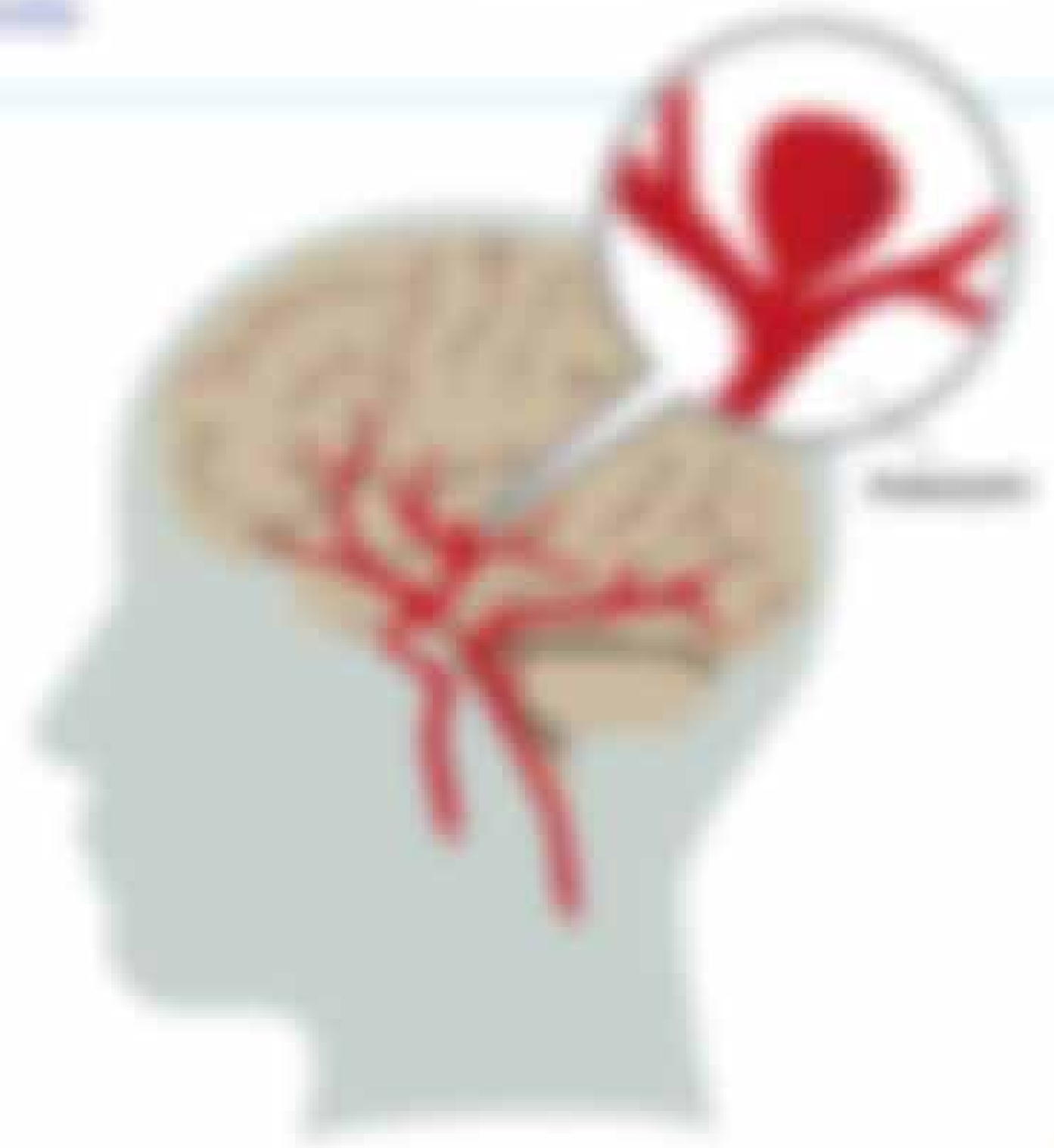
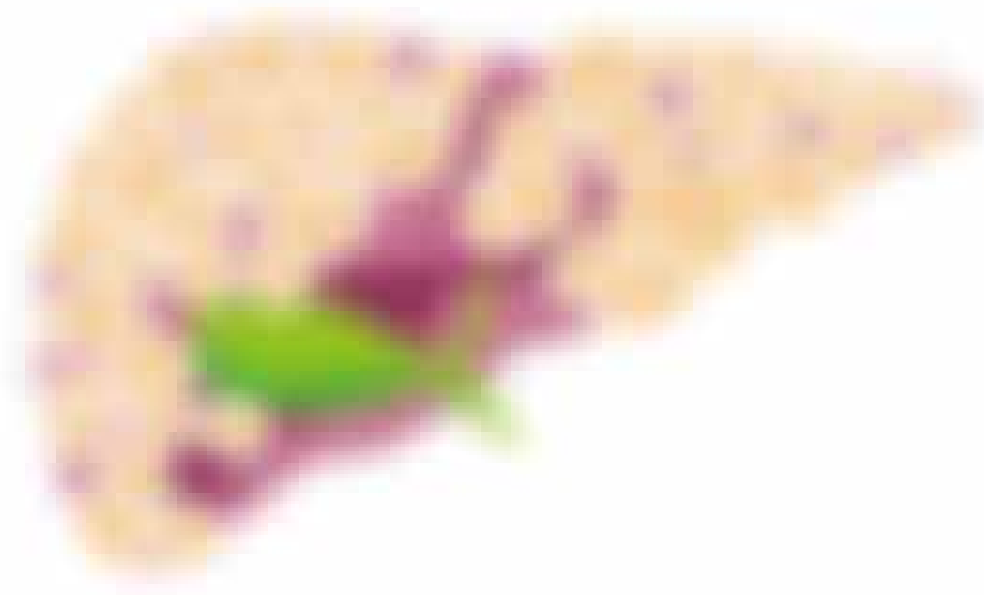
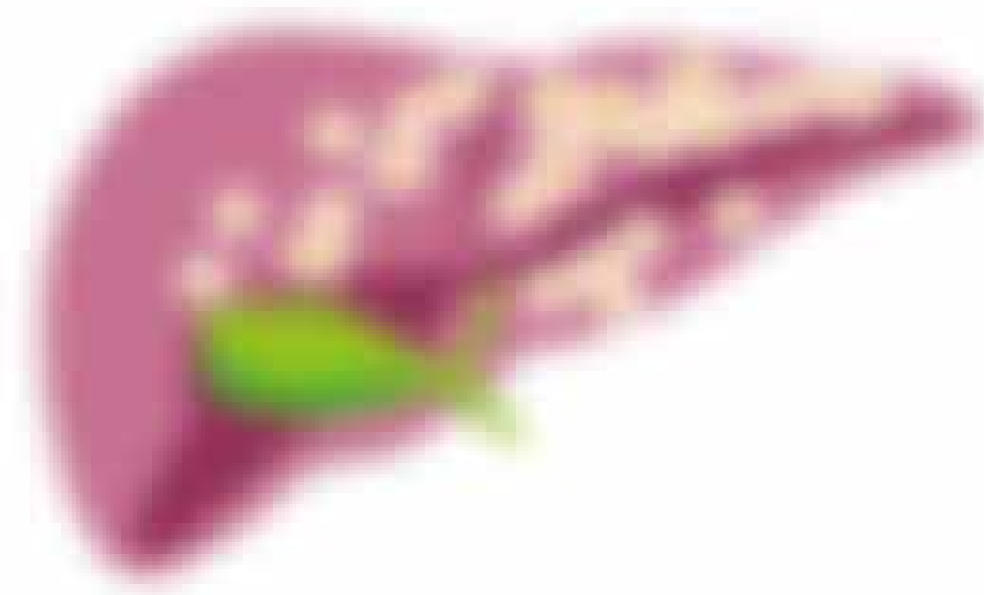
People with liver disease may experience double vision or [blurred vision](#). This can be managed with [liver transplantation](#) and [dialysis](#).

**Symptoms: Aneurysm**

The symptoms of a ruptured aneurysm can include severe headache, sickness, blurred double vision and loss of consciousness.



People with liver disease can also have cysts. This can lead to liver complications, such as portal hypertension. However, this can be managed with liver transplantation and dialysis.



This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

Pain is the most common and important symptom of ADPKD for many patients, particularly when it persists. Pain can occur at any stage of the disease, even early on, and it is important to recognise, investigate and treat it.

## What causes pain in ADPKD?

Acute pain can be caused by various kidney or liver [complications](#) of ADPKD, such as cyst infections and kidney stones. Treating the underlying cause of these should help to relieve the associated pain.

Chronic pain normally means pain lasting for more than 3 months. Chronic pain can result from the growth of cysts in the kidney or liver, and can be particularly difficult to treat.

## Assessing pain

Doctors and other healthcare staff may not always appreciate how much pain can affect people with ADPKD. Ideally, doctors and nurses should routinely ask about pain at each clinic visit. You should make sure you report pain to your healthcare team, carefully explaining the location, symptoms and impact of any pain you experience.

### Did you know?

[Pain scales](#) > can be useful to help measure and monitor the severity of pain. [Questionnaires](#) have also been developed to help patients and doctors to assess the impact of ADPKD on patients' wellbeing. You may wish to ask your doctor about these.

## How should chronic pain be managed?

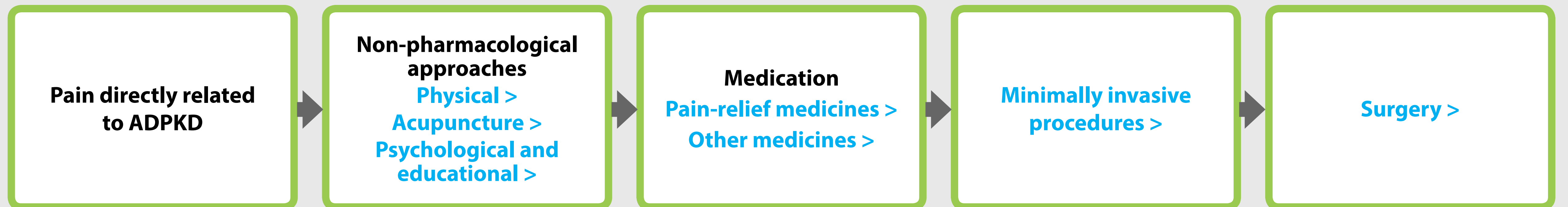
Different types of doctors and healthcare professionals often need to work together to treat chronic pain. Depending on the cause and type of pain, this may include pain specialist doctors, [neurologists](#) >, [radiologists](#) >, as well as [nephrologists](#) > and [hepatologists](#) >. [Physiotherapy](#) > and [psychotherapy](#) > may also have a role in some situations.

Experts in the Netherlands have produced a pathway for the stepwise management of chronic pain caused by ADPKD (see figure below).

 [Checklist](#) >



Treatments are [recommended](#) to be used in the order shown below until pain relief is achieved.



This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

The video below contains some key messages and highlights common patient concerns, particularly when it comes to the use of painkillers. You can access the video [here](#) and also find a transcript of the video [here](#).

## What causes pain in ADPKD?

Pain can be caused by various factors in ADPKD, such as cyst enlargement, kidney stones, urinary tract infections, and haematuria. It is important to discuss any pain you experience with your healthcare professional.

Chronic pain is usually caused by cysts pressing on nerves. Chronic pain can lead to the growth of cysts and may be particularly difficult to treat.

## Assessing pain

Doctors and other healthcare workers use a variety of tools to assess pain in ADPKD patients. These include visual analogue scales (VAS) and the Brief Pain Inventory (BPI). It is important to discuss your pain with your healthcare professional so they can assess the impact of ADPKD on your pain and help you to manage it.

## Management

[Painkillers](#) can be used to help manage pain in ADPKD. [Non-steroidal anti-inflammatory drugs \(NSAIDs\)](#) can also be used, but it is important to discuss the risks of NSAIDs with your healthcare professional as they may lead to kidney damage.

## Non-pharmacological management

### Pain

*'I find that my renal consultant has a hard time understanding the chronic and acute pain I get due to ADPKD. It is often brushed aside.'*

*Anonymous, UK*

*'I am lucky not to have encountered renal or back pain since I have been diagnosed, but I do believe that practising sport regularly helps in managing the pain.'*

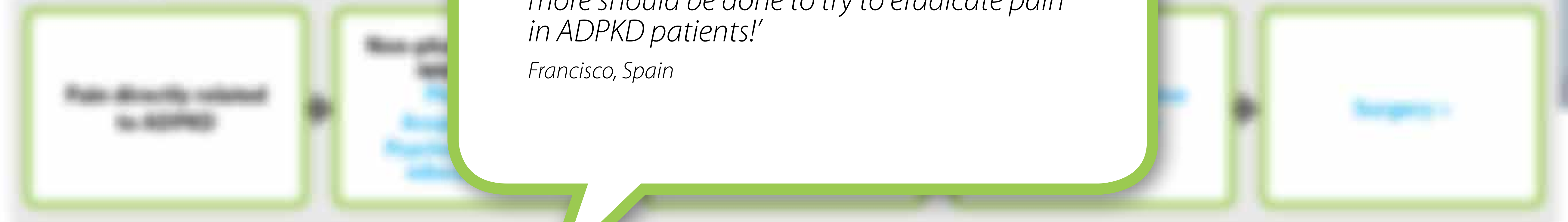
*Corinne, France*

*'In my own experience I have found that hypnotherapy can help to reduce anxiety and pain.'*

*Brenda, the Netherlands*

*'No one should have pain in their life – more should be done to try to eradicate pain in ADPKD patients!'*

*Francisco, Spain*



This section explains how pain is assessed

The scales used to assess and measure pain are used to help patients, particularly older patients, to rate their pain. The scales are used to help the doctor to understand the patient's pain.

## What causes pain in COPD?

Acute pain can be caused by various factors such as infection, such as an acute infection, or trauma. The underlying cause of pain should be identified to allow the pain to be treated.

Chronic pain normally means pain lasting for more than three months. Chronic pain can result from the growth of pain in the body and can be particularly difficult to treat.

## Assessing pain

Doctors and other health care workers use different pain scales to assess pain. The scales are used to help the doctor to understand the patient's pain. The scales are used to help the doctor to understand the patient's pain.

## Visual analogue scales

Visual analogue scales are used to help patients to rate their pain. The scales are used to help the doctor to understand the patient's pain. The scales are used to help the doctor to understand the patient's pain.

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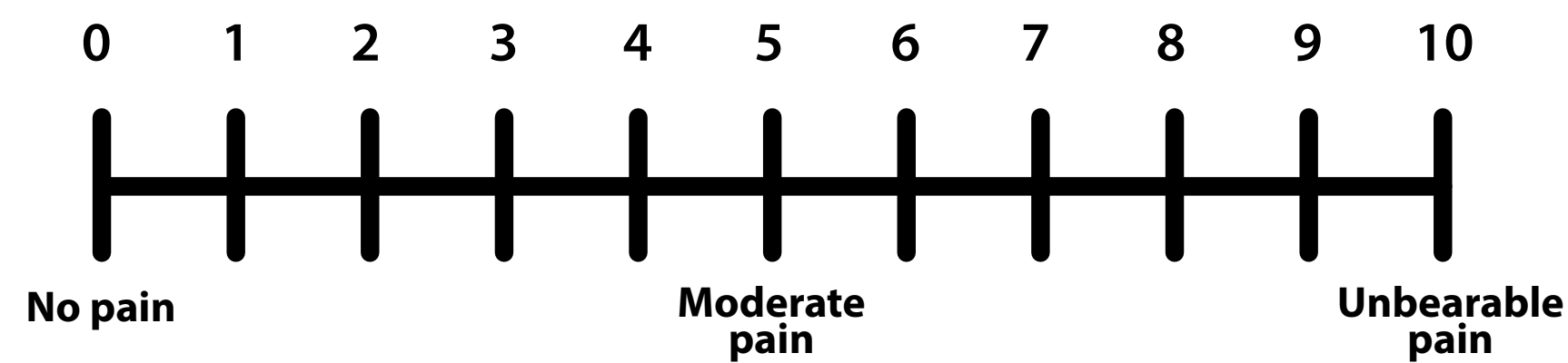
## Pain scales

Common ways that patients can rate their own pain include:

- Visual analogue scales: you use these to indicate how severe your pain is on a line between two extremes, such as 'no pain' and 'pain as bad as it could be'.



- Numerical rating scales: these are like visual analogue scales, but use numbers to rate the severity of pain, usually between 0 and 10.



This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

The most common cause of pain in ADPKD is kidney stones, particularly when they are large. You can learn more about the signs and symptoms of kidney stones and how to manage them in the section on kidney stones.

## What causes pain in ADPKD?

Pain can be caused by kidney stones or by [cysts](#) in ADPKD, such as cyst infections and kidney stones. Treating the underlying cause of pain should help to reduce the associated pain.

Chronic pain normally means pain lasting for more than 12 weeks. Chronic pain can result from the growth of cysts in the kidney or from other conditions affecting the kidney.

## Assessing pain

Doctors and other healthcare staff may use simple questionnaires to assess how much pain you are experiencing with ADPKD. These questionnaires should normally be done once or twice a week. You should make sure you report pain to your healthcare team, including reporting the location, symptoms and impact of your pain on your life.

## Medication

[Pain relief](#) can be useful to help reduce and manage the severity of pain. [Antibiotics](#) have also been developed to help control and reduce the impact of ADPKD on patients suffering the most with kidney stones.

## How should chronic pain be managed?

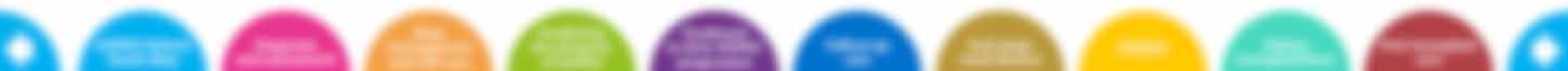
Different types of pain may require different treatments. You should make sure you report pain to your healthcare team, including reporting the location, symptoms and impact of your pain on your life.

**Neurologist** ✕

A neurologist is a doctor who specialises in treating diseases of the nervous system, which includes the brain, spinal cord and the nerves throughout the body.



Treatments are [recommended](#) to be used in the order shown below until pain relief is achieved.



This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

The video below contains an overview of ADPKD for some patients, particularly when it comes to the pain. You can access this video at the end of the document with the link to the relevant page.

## What causes pain in ADPKD?

Pain can be caused by various factors in ADPKD, such as cyst infections, kidney stones, haematuria, and stretching of the renal capsule to relieve the associated pain.

Chronic pain normally means pain lasting for more than 3 months. Chronic pain can result from the growth of cysts in the kidney or from other conditions such as arthritis.

## Assessing pain

Doctors and other healthcare staff may use simple questionnaires to assess how much pain you are experiencing with ADPKD. These questionnaires should normally be done once or twice a week. You should make sure you report pain to your healthcare team, usually reporting the location, symptoms and impact of the pain on your life.

## Medication

**Paracetamol** can be useful to help relieve pain and reduce the severity of cysts. **Codeine** has also been developed to help control pain and reduce the impact of ADPKD on patients' wellbeing. You may wish to ask your doctor about these.

## How should chronic pain be managed?

Different types of chronic pain can be managed together to help control the overall pain. This may include using **paracetamol** and **codeine** with **opioids** and **antidepressants**.

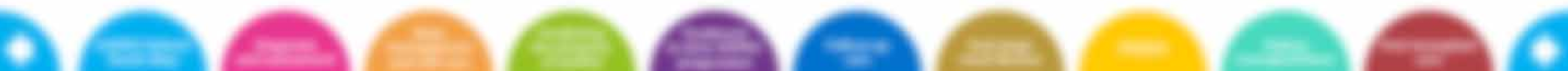
There is also information on the management of chronic pain.

**Radiologist** ✕

A radiologist is a doctor who specialises in diagnosing and treating disease and injury using medical imaging techniques such as x-rays, computed tomography (CT), magnetic resonance imaging (MRI), positron emission tomography (PET) and ultrasound.



Treatments are **recommended** to be used in the order shown below until pain relief is achieved.





This section explains how pain is assessed and managed in CKPD, particularly when it is persistent.

The most common cause of pain in CKPD is the underlying disease, particularly when it is advanced. The pain can occur at any stage of the disease, even when there is no obvious reason for it. Multiple medicines may be needed to manage the pain.

## What causes pain in CKPD?

Pain can be caused by various factors in CKPD, such as bone infections, osteoarthritis, kidney stones, and underlying causes of bone disease. It is important to identify the cause of the pain.

Chronic pain normally means pain lasting for more than 12 weeks. Chronic pain can result from the growth of pain in the body or from an acute pain that has become chronic.

## Assessing pain

Doctors and other healthcare staff may use simple pain scales to assess how much pain you are in. CKPD needs to be managed with a range of medicines and other treatments. You should make sure you report pain to your healthcare team, including reporting the location, symptoms and impact of your pain on your life.

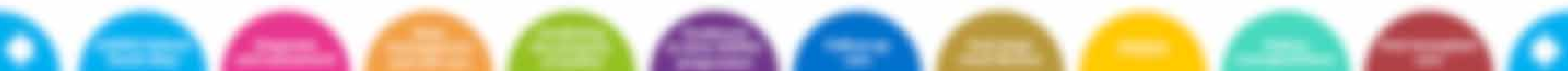
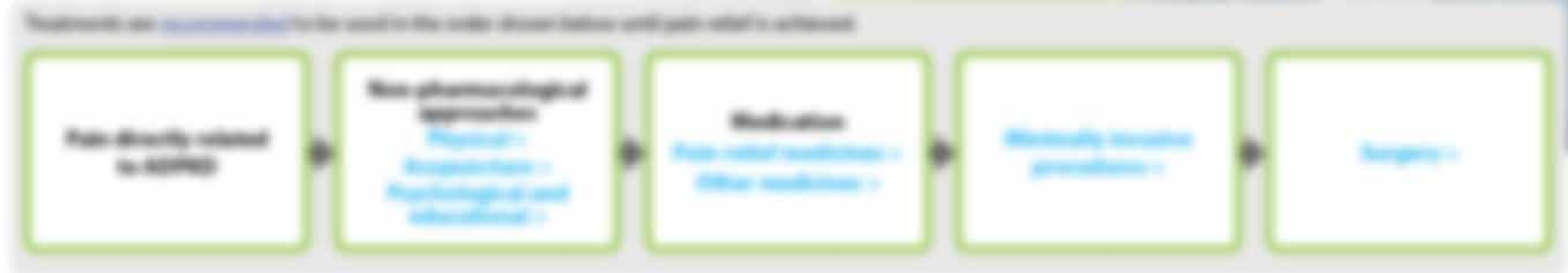
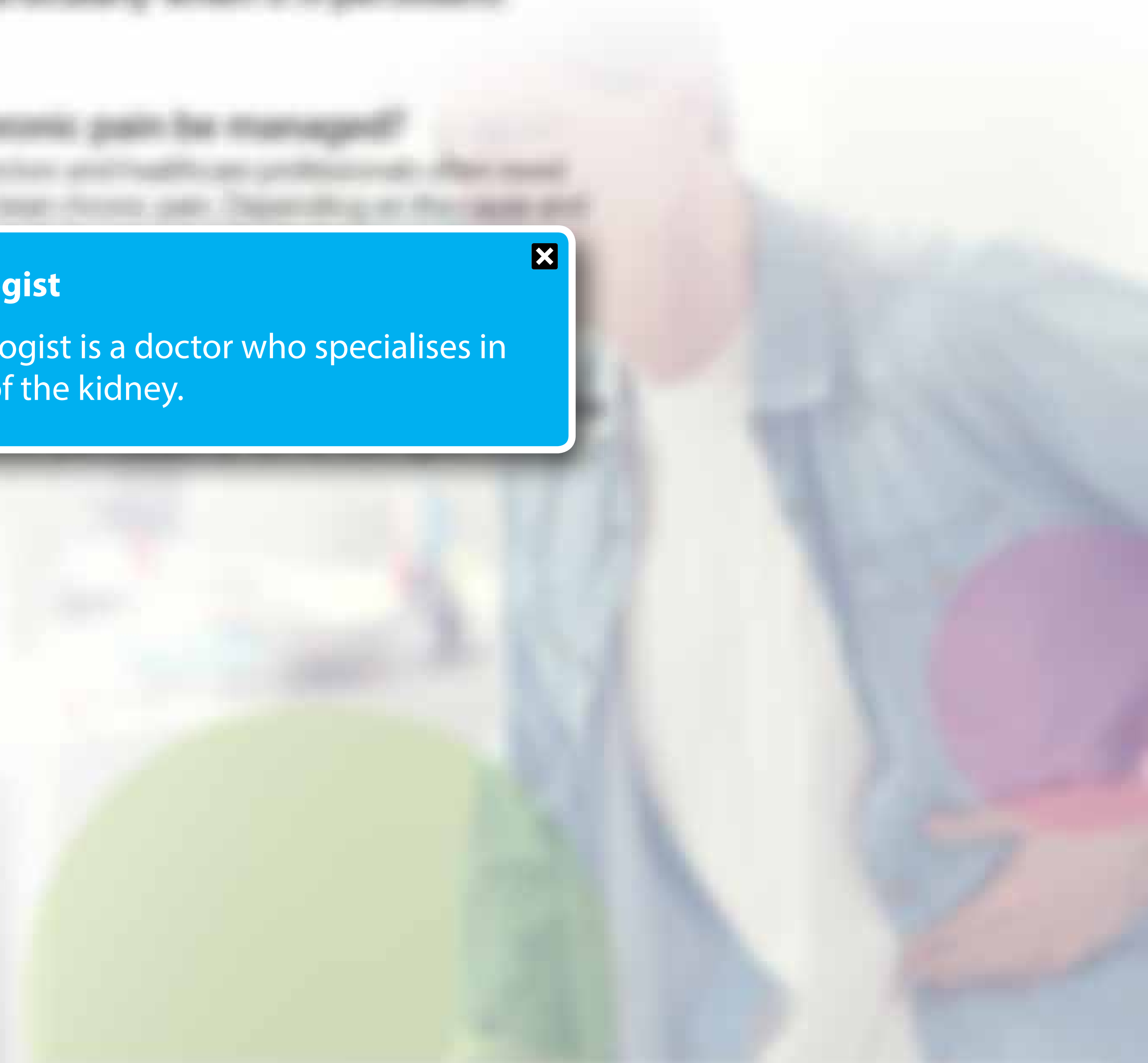
## Medicines

**Paracetamol** can be used to help relieve pain. However, the severity of pain in CKPD may be too severe for paracetamol to help. Other medicines, such as opioids, may be needed to manage the pain. It is important to discuss the impact of CKPD on your pain with your doctor.

## How should chronic pain be managed?

Chronic pain in CKPD should be managed with a range of medicines and other treatments. It is important to discuss the impact of CKPD on your pain with your doctor.

**Nephrologist** ✕  
A nephrologist is a doctor who specialises in diseases of the kidney.



This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

Pain is the most common and challenging symptom of ADPKD for many patients, particularly when it is persistent. You can access all the steps of the disease management plan on [ADPKD.org](#) or [ADPKD.org.uk](#) (multiple versions).

## What causes pain in ADPKD?

Pain can be caused by various factors in ADPKD, such as cyst infections, stretching of the kidney capsule, or the growth of cysts in the kidney or liver.

Chronic pain normally means pain lasting for more than 3 months. Chronic pain can result from the growth of cysts in the kidney or liver and can be particularly difficult to treat.

## Assessing pain

Doctors and other healthcare staff may use simple questionnaires to assess how much pain you are experiencing with ADPKD. These doctors and nurses should routinely ask about pain at each clinic visit. You should make sure you report pain to your healthcare team, carefully explaining the location, symptoms and impact of your pain and experience.

## Medication

[Pain relief](#) can be useful to help relieve and manage the severity of pain. [Antibiotics](#) have also been developed to help control and decrease the impact of ADPKD on patients suffering. You may wish to ask your doctor about these.

## How should chronic pain be managed?

Different types of chronic pain may be managed in different ways. It is important to work together to create a management plan that suits your needs and preferences.

- [Pain relief](#) - as with other types of pain, the most effective approach is to use a combination of different types of pain relief.
- [Antibiotics](#) - ADPKD-related kidney infections can cause pain.
- [Surgery](#) - to remove the source of pain.

Doctors in the UK have developed a management plan for chronic pain in ADPKD.

**Hepatologist** ✕

A hepatologist is a doctor who specialises in diseases of the liver.



Treatments are [recommended](#) to be used in the order shown below until pain relief is achieved.



This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

The video below contains and explains a number of ADPKD-related terms, particularly when it comes to pain. You can access any stage of the video, with only an audio transcript to support it. Multiple screens.

## What causes pain in ADPKD?

Pain can be caused by various factors in ADPKD, such as cysts, infections, and kidney stones. Understanding the underlying cause of your pain is key to relieving the associated pain.

Chronic pain normally means pain lasting for more than 12 weeks. Chronic pain can sometimes be a sign of a more serious condition, so it's important to talk to your doctor about it.

## Assessing pain

Doctors and other healthcare workers use a range of tools to assess how much pain you are in. These tools can help you understand your pain and how to manage it. You should make sure you report your pain to your healthcare team, so they can help you manage it better.

## Medication

Medication can be used to help relieve pain and reduce the severity of pain. Medication has also been developed to help control and manage the impact of ADPKD on your health. You may want to talk to your doctor about this.

## How should chronic pain be managed?

Different types of chronic pain need to be managed in different ways. It's important to work together to manage chronic pain. Depending on the type and severity of your pain, you may need to use a range of treatments.

### Physiotherapy

Physiotherapy is a physical form of treatment used to help people affected by injury, illness or disability through movement and exercise, manual therapy, education and advice.



Treatments can be used to help manage pain in ADPKD. The video below explains how to manage pain when it is persistent.



# Managing pain

This section explains how pain is assessed and managed in ADPND, particularly when it is persistent.

The video will discuss common and emerging symptoms of ADPND in your patients, particularly when persistent. You will learn about the steps of the disease, with early on with a diagnosis to manage. Multiple sources.

## What causes pain in ADPND?

Acute pain can be caused by various factors in the [pathophysiology of ADPND](#), such as joint inflammation and muscle spasms. Treating the underlying cause of these symptoms is often the most effective way to manage pain.

Chronic pain normally means pain lasting for more than 12 weeks. Chronic pain can result from the growth of pain in the setting of long-term or the persistence of acute pain.

## Assessing pain

Doctors and other healthcare staff may use simple questions to assess how much pain can affect people with ADPND. Needs, desires and needs should routinely be discussed at each visit with you. You should make sure you report pain to your healthcare team, carefully explaining the location, symptoms and impact of your pain and experience.

## Medication

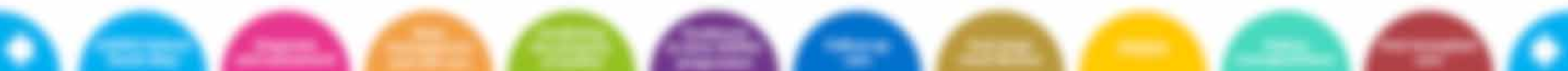
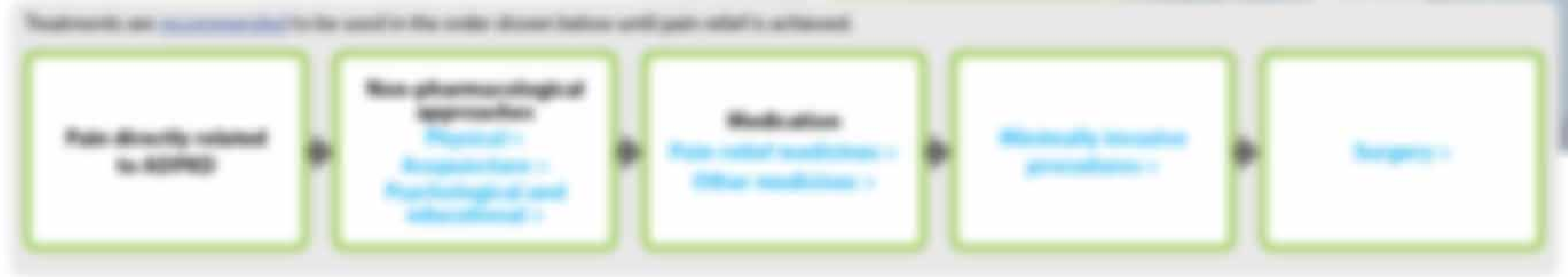
[Pain relief](#) can be useful to help improve your quality of life. [Medications](#) have also been developed to help control and manage the impact of ADPND on patients' wellbeing. You may wish to ask your doctor about these.

## How should chronic pain be managed?

Different types of chronic pain and health conditions often need to work together to help manage pain. Depending on the cause and type of pain, the most suitable approach is often:

### Physical

These include physiotherapy, massage, ice and heat pads, and Alexander technique (which focuses on body posture and movement).



This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

Pain is the most common and bothersome symptom of ADPKD in many patients, particularly when it is persistent. You can assess at any stage of the disease, even early on, and it is important to recognise multiple potential causes.

## What causes pain in ADPKD?

Pain can be caused by various factors or have **multiple causes** in ADPKD, such as cyst infections, overactive nerves, kidney size, underlying cause of liver disease, or to reduce the associated pain.

Chronic pain normally means pain lasting for more than 12 weeks. Chronic pain can result from the growth of cysts in the kidney or from other factors, particularly difficult to treat.

## Assessing pain

Doctors and other healthcare staff may use simple questionnaires to assess how much pain you often experience with ADPKD, usually doctors will want to know how often you have pain at each other visit. You should make sure you report pain to your healthcare team, carefully explaining the location, symptoms and impact of your pain and experience.

## Medication

**Pain relief** can be useful to help relieve and manage the severity of pain. **Analgesics** have also been developed to help control pain and reduce the impact of ADPKD on patients' wellbeing. You may wish to ask your doctor about these.

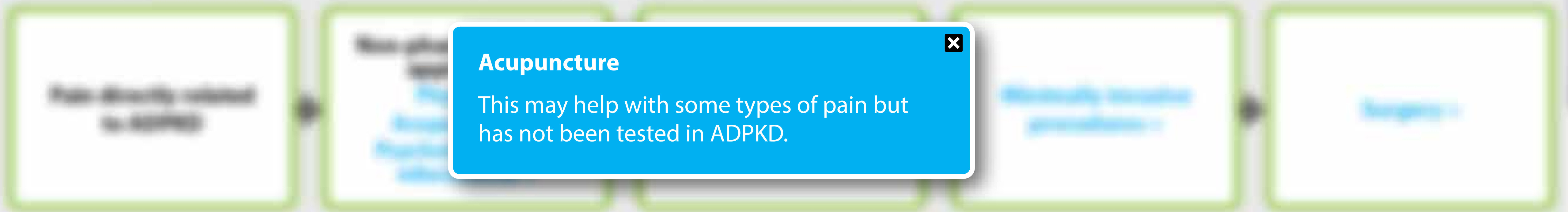
## How should chronic pain be managed?

Different types of doctors and healthcare professionals often need to work together to help chronic pain. Depending on the cause and type of pain, this may include your general doctor, **physiotherapist**, **psychologist**, **social worker**, **occupational therapist**, **dietitian**, **acupuncturist** and **complementary medicine** practitioners.

Doctors in the Netherlands have produced a pathway for the management of chronic pain supporting ADPKD care (see below).



Treatments are **recommended** to be used in the order shown below until pain relief is achieved.



**Acupuncture**  
This may help with some types of pain but has not been tested in ADPKD.



This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

The video below reviews and compares common causes of ADPKD-related pain, particularly when persistent. You can access any stage of the disease with only an initial diagnosis to suggest multiple causes.

## What causes pain in ADPKD?

Acute pain can be caused by various factors or by [cystitis](#) in ADPKD, such as cyst infections and kidney stones. Treating the underlying cause of these should help to reduce the associated pain.

Chronic pain normally means pain lasting for more than 12 weeks. Chronic pain can result from the growth of cysts in the kidney or from other factors which are difficult to treat.

## Assessing pain

Doctors and other healthcare staff may use simple questionnaires to assess pain in people with ADPKD. These doctors and nurses should routinely ask about pain at each clinic visit. You should make sure you report pain to your healthcare team, carefully explaining the location, symptoms and impact of your pain and experience.

## Medication

[Pain relief](#) can be useful to help relieve and manage the severity of pain. [Antibiotics](#) have also been developed to help control and decrease the impact of ADPKD-related cystitis. You may wish to ask your doctor about these.

## How should chronic pain be managed?

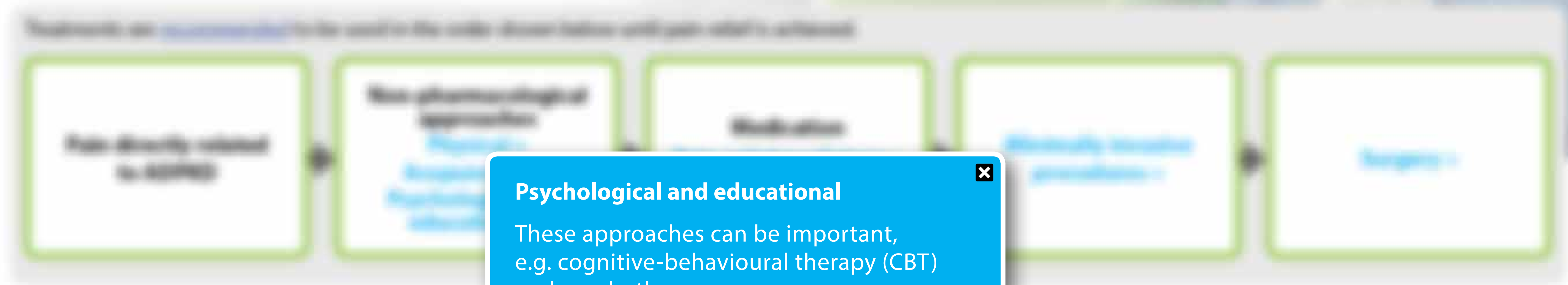
Different types of doctors and healthcare professionals often need to work together to help chronic pain. Depending on the cause and type of pain, this may include your general doctor, [physiotherapist](#), [dietitian](#), [psychologist](#) and [nurse](#). You should have a care plan for your chronic pain.

Doctors in the following areas have produced guidelines for the management of chronic pain relating to ADPKD: [see here](#).



**Psychological and educational** ✕

These approaches can be important, e.g. cognitive-behavioural therapy (CBT) and psychotherapy.



This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

The video below explains how pain is assessed and managed in ADPKD. It covers persistent pain, particularly when it is chronic. You can access a copy of the video [here](#) or watch it on YouTube [here](#).

## What causes pain in ADPKD?

Pain can be caused by several things, including kidney stones, cysts, and infections. It can also be caused by the growth of the kidneys, which can lead to the stretching of the kidney capsule.

Chronic pain usually means you're getting the same pain over and over again. It can be caused by the growth of the kidneys or by the stretching of the kidney capsule.

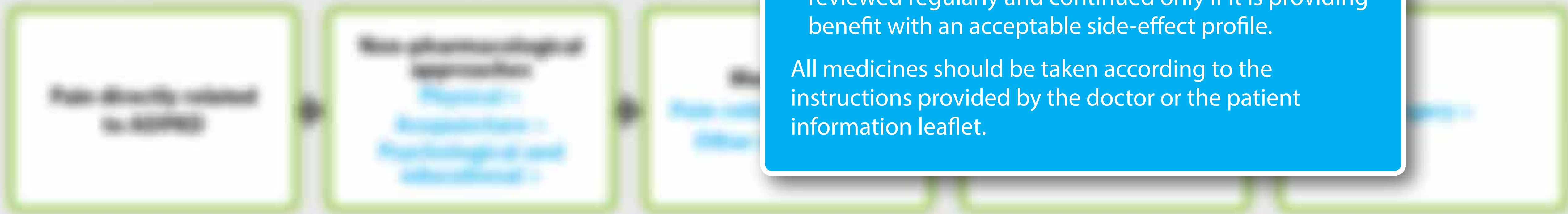
## Assessing pain

Doctors will often use a rating system to help them understand how much pain you're feeling. This is usually a scale from 0 to 10, where 0 is no pain at all and 10 is the worst pain you can imagine. You should make sure you're able to do this at home as well, so you can tell your doctor how much pain you're having.

## Medicines

**Paracetamol** can be used to help relieve pain and reduce the severity of inflammation. It can also be used to help with fever. **NSAIDs** can also help with pain, but they can be harmful to the kidneys, so you should only use them if your doctor says it's safe. You should also avoid alcohol and other medicines that can be harmful to the kidneys.

Medicines can be used to help relieve pain and reduce the severity of inflammation.



## Pain-relief medicines

Pain-relief medicines (analgesics) are often used in a 'ladder', starting with milder medicines and working up to stronger ones if necessary, as follows:

- Acetaminophen (paracetamol) is often used first.
- Non-steroidal anti-inflammatory drugs (NSAIDs; e.g. ibuprofen and diclofenac) or mild opioids may be used if pain relief is insufficient. However, NSAIDs can damage the kidneys and they are not recommended for use by patients whose kidney function is impaired – you can find more information about their use in ADPKD [here](#). If NSAIDs are suitable, they may be combined with paracetamol.
- Strong opioids (e.g. morphine) may be used when other medicines do not provide enough pain relief. Possible side effects of opioids include constipation, nausea, vomiting, sedation and mental changes. These medicines can also lead to psychological dependence (addiction). However, this is rare when opioids are prescribed appropriately for suitable patients with chronic pain, and when treatment is properly monitored. Opioid treatment should be reviewed regularly and continued only if it is providing benefit with an acceptable side-effect profile.

All medicines should be taken according to the instructions provided by the doctor or the patient information leaflet.

This section explains how pain is assessed and managed in AD/HD, particularly when it is persistent.

This is the most common and important symptom of AD/HD in many patients, particularly when it is persistent. You can assess the severity of the chronic pain with an [assessment](#) to measure the impact of the pain.

## What causes pain in AD/HD?

Chronic pain can be caused by various factors or [conditions](#) in AD/HD, such as [painful conditions](#) or [injury](#). Treating the underlying cause of these conditions may reduce the associated pain.

Chronic pain normally means pain lasting for more than 12 weeks. Chronic pain can result from the growth of pain in the setting of long-term or persistent physical illness.

## Assessing pain

Doctors and other healthcare staff may use [assessment](#) tools to assess how much pain can affect people with AD/HD. [Pain assessment](#) tools should normally ask about pain at each visit. You should make sure you report pain to your healthcare team, carefully explaining the location, symptoms and impact of your pain and experience.

## Medication

[Pain relief](#) can be helpful to help relieve and manage the severity of pain. [Medication](#) has also been developed to help control and manage the impact of AD/HD on patients suffering the long-term or persistent pain.

## How should chronic pain be managed?

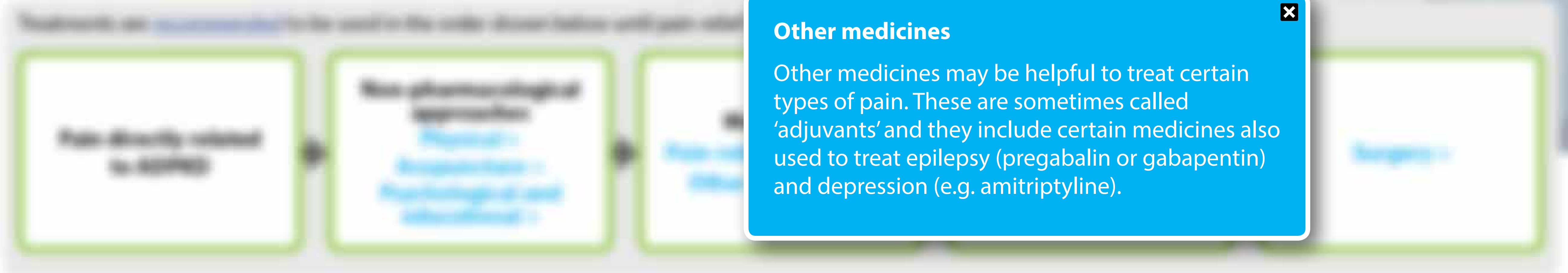
Different types of chronic pain and healthcare professionals often need to work together to help chronic pain. Depending on the type and type of pain, the management may include [medication](#), [cognitive behavioural therapy](#), [physical therapy](#) and [self-help](#). You should make sure you report pain to your healthcare team, carefully explaining the location, symptoms and impact of your pain and experience.

There is a [link](#) between chronic pain and conditions for the management of chronic pain including AD/HD and hypermobility.



**Other medicines** ✕

Other medicines may be helpful to treat certain types of pain. These are sometimes called 'adjuvants' and they include certain medicines also used to treat epilepsy (pregabalin or gabapentin) and depression (e.g. amitriptyline).





# Managing pain

This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

Pain is the most common and distressing symptom of ADPKD in many patients, particularly when it is persistent. You can assess the severity of the disease with help from a specialist in managing multiple cysts.

## What causes pain in ADPKD?

Pain can be caused by various factors, such as **hypertension** or **ADPKD**, such as cyst infections and kidney stones. Treating the underlying cause of pain should help to reduce the associated pain.

Chronic pain normally means pain lasting for more than 12 weeks. Chronic pain can result from the growth of cysts in the kidney or from other factors, such as arthritis.

## Assessing pain

Doctors and other healthcare staff may use simple questionnaires to assess how much pain you suffer. People with ADPKD usually describe pain using simple words, such as sharp, dull or aching. You should make sure you report pain to your healthcare team, especially regarding the location, symptoms and impact of your pain on your life.

## Medication

**Paracetamol** can be useful to help relieve pain, but remember the severity of your **kidney disease** may also have developed to help control pain. Always discuss the impact of ADPKD on your condition with your healthcare team about these.

## How should chronic pain be managed?

Different types of doctors and healthcare professionals often need to work together to help manage pain. Depending on the cause and type of pain, this may include your general doctor, **physiotherapist**, **psychologist** and **specialist**. You should make sure you report pain to your healthcare team, especially regarding the location, symptoms and impact of your pain on your life.

Doctors in the following areas provide support for the management of chronic pain in people with ADPKD: **general medicine**

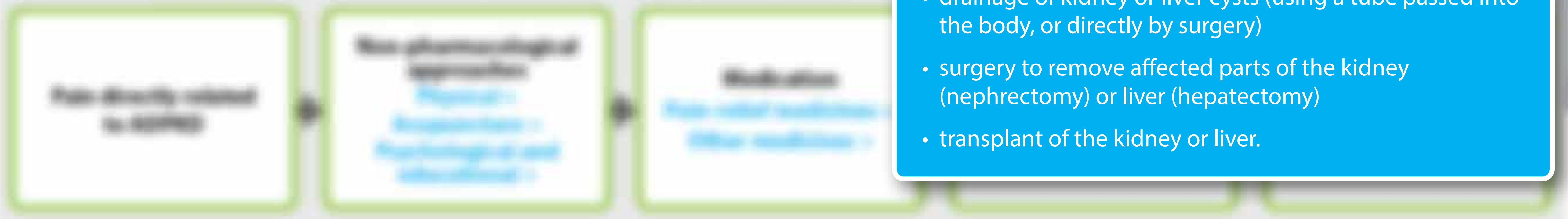


**Surgery**

Surgical measures are used only as a last resort. They include:

- drainage of kidney or liver cysts (using a tube passed into the body, or directly by surgery)
- surgery to remove affected parts of the kidney (nephrectomy) or liver (hepatectomy)
- transplant of the kidney or liver.

Treatments are **recommended** to be used in the order shown below, with pain relief as a priority.



This section explains how pain is assessed and managed in ADPKD, particularly when it is persistent.

The most common cause of pain in ADPKD is kidney stones, particularly when they are large. You can learn more about this type of stone and what you can do to prevent them from coming back in the next section.

## What causes pain in ADPKD?

There are several reasons why you may experience pain in ADPKD, such as kidney stones, cysts, or infections. Finding the underlying cause of your pain is the first step to relieving it.

Chronic pain normally means you're feeling the same pain more than 12 weeks. Chronic pain can result from the growth of cysts in the kidney or from other conditions that are difficult to treat.

## Assessing pain

Doctors and other healthcare workers use different approaches to assess pain. Some use a scale to rate your pain, while others use a visual analogue scale. You should make sure you report your pain to your healthcare team, especially if it's severe, persistent, or impacts your quality of life.

## Medication

Medication can be used to help relieve pain, but it's important to use it correctly. Your healthcare team can help you understand how to use pain medication safely and effectively, and they can also help you understand the risks of using pain medication long-term.

## How should chronic pain be managed?

Different types of doctors and healthcare professionals often work together to help manage pain. Depending on the cause and type of pain, this may include your general doctor, a pain specialist, a psychologist, and a physical therapist. You should also consider lifestyle changes.

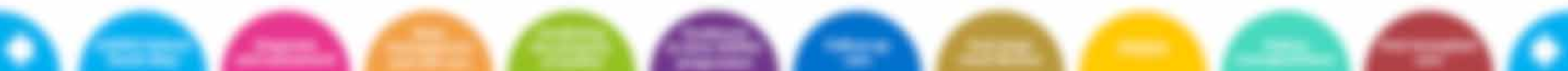
There is also information on how to manage pain for the overall management of chronic pain in ADPKD.



**Minimally invasive therapies** ✕

These are procedures in which the nerve supply to the body part causing pain is blocked by injections or removed by surgery. These procedures need specialist expertise.

Treatments are recommended to be used in the order shown below until pain relief is achieved.



### Checklist: Managing pain



- Pain discussed and assessed (at each clinic visit)
- Advice given on self-care for pain
- Chronic pain investigated by referral to other specialists, as appropriate
- Options for pain management discussed and agreed
- Information about patient organisations and other forms of support

Notes and questions you would like to ask your healthcare team

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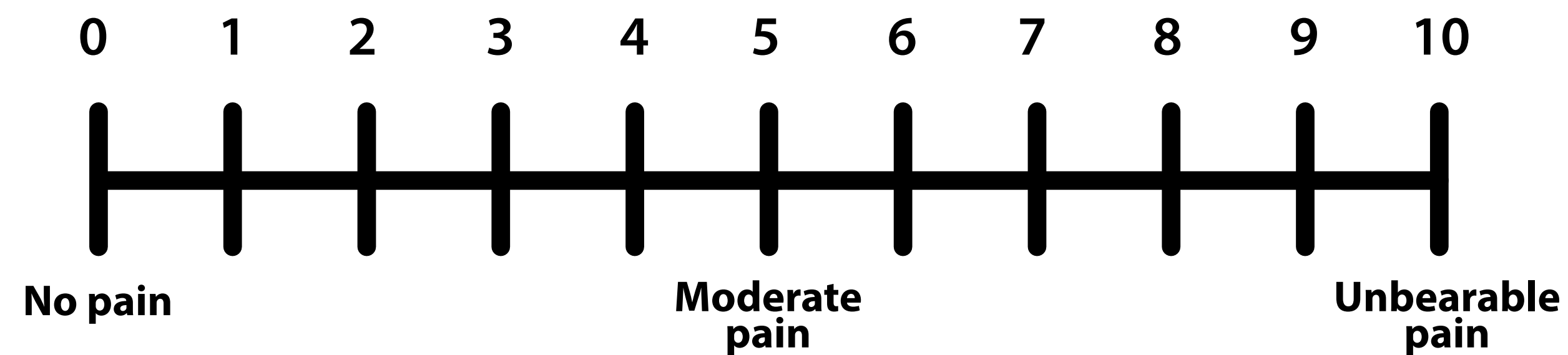
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Here is a scale that you can use to record the severity of your pain.



# Wellbeing, personal and family life

This section explains how ADPKD can affect the wellbeing, personal and family lives of patients and their families, what can be done to help cope with these effects, and what support patients can expect.

## Emotional impact >

ADPKD can have a significant emotional and psychological impact on patients and their families, and yet communicating about the disease can be difficult. Some healthcare professionals may not fully understand the impact ADPKD can have, both on physical and mental health.



**Questionnaires >** have been developed specifically to help measure the impact of ADPKD on quality of life and wellbeing.

## What can patients and families do? >

There are many things that patients and families can do to help cope with ADPKD.

## Work >

Some patients with ADPKD find that the disease affects their ability to work.

## Financial issues >

ADPKD can affect things like health and life insurance and mortgages.

## Checklist >

**Seeking professional help**

**Coming to terms with ADPKD**

**You're not alone!**

**What about children and young people?**

## **You're not alone!**

*'Contacting the self-help group was good for me. It was good to meet other affected people and relatives and exchange thoughts. It helped me to think positively again and not only see the negative things. As the wife of a man with ADPKD, I might have been more worried than my husband is himself. We can handle it better now – even if it is still a challenge every day.'*

*Isabel, Austria*

*'It would be too much to say that the National Patient Day in Berlin saved my life, but at least it really changed the way I look at many things. Thank you so much!'*

*Sven, Germany*

*'I was at a PKD meeting in Freiburg at the weekend. It helped me to see how many people are affected and to know that I am not alone.'*

*Patricia, Germany*

*'Peer support is so important, be it from family, friends, support groups or medical staff. If you feel you need it, make sure to find it.'*

*Cathriona, Ireland*

*'I am a transplant patient and my "personal psychologist" – Luisa Sternfeld Pavia, the Chairperson of the Associazione Italiana Rene Policistico (AIRP) patient organisation – has played a decisive role during the course of my disease. I thank her wholeheartedly!'*

*Roberto, Italy*

*'It is truly important to feel sheltered by peers in patient's organisations. In addition, by the doctors and nurses.'*

*José, Spain*

*'If talking about ADPKD helps you, talk about it!'*

*Carmen, Spain*

*'Often just hearing about other people's experiences with PKD can make a difference. By joining the patient organisation on Facebook I now know there are people out there experiencing the same things as me, and learning from them really helps.'*

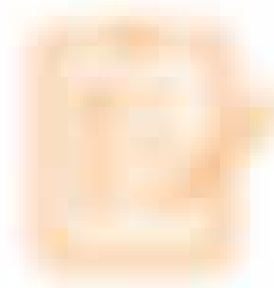
*Polly, UK*

## Wellbeing, personal and family life

This section explains how ADPKD affects you and your family, what can be done...

### Living with ADPKD

ADPKD can affect your quality of life and your family. You may feel worried about the future or the effect of the disease on your life. It is important to talk to your doctor about your concerns and to get support if you need it.



**Testimonial:** How does ADPKD affect your life? How do you cope with it? How do you support your family? How do you support your doctor?

### Living with ADPKD

There are many things that you can do to help you cope with ADPKD.

### What about children and young people?

*'Announcing the disease to my daughter was difficult, as one is bound to feel guilty for being responsible for transmitting a genetic disease to your child. Luckily, she seems to be coping with limited anxiety. Also, the fact that she is working in the health field will certainly provide her with further tools to confront her disease constructively.'*

Corinne, France

*'I found out I had a polycystic kidney when I was 40. I had three children, only one of whom is affected by the disease... At the beginning we chose not to say anything about the disease until they were 18 years old.'*

Roberto, Italy

*'Avoid overprotection in teenagers, they are normal and healthy people even with ADPKD. They need information and education for the future, but it takes time to accept things and go to the doctor, it's a personal decision the right time to cope with it.'*

Ricardo, Spain



## Wellbeing, personal and family life

This section explains how ADPKD can affect your wellbeing and family life, and what can be done to help.

### Emotional impact

ADPKD can have a significant emotional impact on you, your family, and your community. It's important to talk to your doctor about your feelings and to seek support if you need it.

**Support groups** can help you to share your experiences with others who understand what you're going through. They can provide a safe space to talk and to get advice.

**Self-help resources** can help you to manage your feelings and to find ways to cope with ADPKD.

### Seeking professional help

*'I was desperate after several years of dialysis and a donor kidney that did not work. Every day was a rainy day – sad and worrying. I've been trying to get professional support from a mental health clinic. Unfortunately, this has turned out to be very difficult.'*

*Claus, Austria*

*'I have been getting psychological care for about 3 years, I already began to prepare for it before dialysis with regular meetings once a month. I think that was and is a very good "investment". Neither your family nor your friends can "absorb" certain topics. It is important to discuss some things outside that context. The ADPKD self-help group in Nuremberg, Germany, is very supportive.'*

*Phillipp, Austria*

You're not alone

Seeking professional help

## Coming to terms with ADPKD

*'Once I realised how lucky I am to live these days, with the kind of healthcare and options available for dialysis and transplantation, I really get the feeling to be "happy" with my disease... I already lost friends and family to much more painful diseases or fateful incidents... At times I have had pain from surgery and other procedures, but compared with patients with liver, heart or lung diseases I have come to know via patient groups, I am really happy to "only" suffer from kidney disease. It has changed me as a person, also in a positive way. In ways, it has made me a more caring person – it taught me to have patience and appreciate some different values compared with other people my age.'*

*Claus, Austria*

*'Don't worry unduly about not being able to do your usual things. It's a huge life event – create new usuals!'*

*Cathriona, Ireland*

*'The motto of my life has always been "always positive"! Positivity helps to live life in the best possible way and reduces the perception of negative factors. Positivity reduces stress and improves life!'*

*Giovanni, Italy*

*'It's a choice. When you have a condition it's a choice how you want to live with it. You can shut yourself off, be angry, or you can choose the other way.'*

*Brenda, Netherlands*

*'You can worry about a lot of things and let worries overwhelm you, but it really only serves to worry about the things that you can really change or control yourself.'*

*Pedro, Spain*

*'As patients with ADPKD, we learn to enjoy life despite our health.'*

*Salvador, Spain*

*'It's your body and you live with it. It was hard for me to accept that I was sick, but when you accept it, you have to respect it. Simply, I kept living my life.'*

*Anna, Sweden*



This section explains how ADPKD can affect the wellbeing, personal and family lives of patients and their families, and what support patients can expect.

## Emotional impact ✕

Worry and fear are common, following a diagnosis of ADPKD. Some people have difficulty coming to terms with the diagnosis and may wish to avoid thinking about it. Others may have a sense of relief that a diagnosis has finally been made, meaning that they can access care and support.

Some people have feelings of anger or resentment at inheriting ADPKD from a parent. It is important to remember that it is not the fault of the parents. The parents of people with ADPKD often experience feelings of guilt.

In the longer term, patients sometimes have feelings of helplessness and frustration, for example because of pain, body image issues, issues resulting from sexual problems, the effect of the disease on work and finances, the potential future need for dialysis, or concerns following a diagnosis of intracranial aneurysm. Some patients may experience depression related to their ADPKD.

Adolescent patients can face difficulties in coping with ADPKD while dealing with other pressures associated with physical and hormonal changes, education, relationships and family life.

The family members of patients can be affected by these issues, together with the impact of the disease on work and finances. Clearly, the diagnosis of ADPKD in a child has significant emotional and psychological effects on parents. Some parents may have difficulty deciding when, how and what to tell their children about ADPKD.



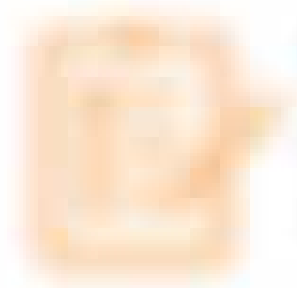
## Wellbeing, personal and family life

This section explains how ADPKD can affect the wellbeing, personal and family lives of patients and their families, what support is available and what support patients can request.

**Emotional impact**  
ADPKD can have a significant emotional impact on patients and their families. It is important to talk to your healthcare team about how you are feeling and what support is available. You may also want to talk to a counsellor or other healthcare professional for help.

**Peer support**  
Peer support from other patients and carers may be available locally in person, or online. Patient organisations, mental health charities and other support organisations can provide this kind of contact and information.

**Talking to children**  
It can be difficult to know what to say to children about ADPKD, and when it is right to do so. The UK PKD Charity offers specific advice to parents about talking to children about the disease. Genetic counsellors may also be able to help here.



### What can patients and families do?



Patients and families deal with the impact of ADPKD in many ways. General suggestions that may help include:

- Find a way to talk about problems – this may be done with a family member or friend, or professional help may be sought from a counsellor or other healthcare professional. It helps if you can fully explain how the disease affects you and your family.
- Some patients and carers may need care and support at times because of stress, depression or anxiety. You may wish to ask your healthcare team about the available support services, including counselling and mental health specialists. Early referral to this kind of support may help prevent more serious problems later.
- It can help to find out how other people affected by ADPKD feel about the disease and cope with it. Peer support from other patients and carers may be available locally in person, or online. Patient organisations, mental health charities and other support organisations can provide this kind of contact and information.
- It can be difficult to know what to say to children about ADPKD, and when it is right to do so. The UK PKD Charity offers specific advice to parents about talking to children about the disease. Genetic counsellors may also be able to help here.

Exercise and other types of physical and social activity may help to relieve stress.



# Wellbeing, personal and family life

This section explains how ADPKD can affect the wellbeing, personal and family lives of patients and their families, what can be done to help cope with these effects, and what support patients can expect.

## Emotional impact

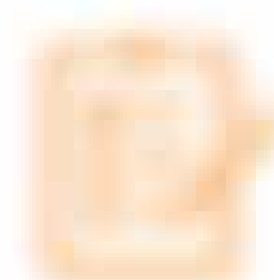
ADPKD can have a significant emotional and psychological impact on patients and their families, and on community and the wider society. It can affect the health and wellbeing of patients and their families, and can lead to a range of health and social issues.

## Work

Patients with ADPKD may face difficulties in their work, and may need to take time off work.

## Financial issues

ADPKD can affect long-term health and the associated costs of care.

 **Questionnaires** have been developed specifically to help measure the impact of ADPKD on quality of life and wellbeing.

**Polycystic Liver Disease Questionnaire (PLD-Q)** can help assess the effect of liver cyst symptoms. This can be found [here](#) (p. 11–16).

**Questionnaires** ✕

Questionnaires have been developed specifically to help measure the impact of ADPKD on quality of life and wellbeing. These include the ADPKD Genetic Psychosocial Risk Instrument (GPRI-ADPKD) (available in the 'Supplementary data' file [here](#)) and the ADPKD Impact Scale (ADPKD-IS).

In addition, a questionnaire called the Polycystic Liver Disease Questionnaire (PLD-Q) can help assess the effect of liver cyst symptoms. This can be found [here](#) (p. 11–16).

If you are interested you may wish to ask your healthcare team about these questionnaires.



# Wellbeing, personal and family life

This section explains how ADPKD can affect the wellbeing, personal and family lives of patients and their families, what can be done to help cope with these effects, and what support patients can expect.

## Emotional impact

ADPKD can have a profound emotional and psychological impact on patients and their families, and can be particularly difficult to cope with if you have to leave employment or limit your work to look after affected family members (e.g. spouses and children). Some people also experience difficulties at work because ADPKD limits the activities they can do or requires them to take time off for clinic appointments. Over time, many patients report a loss of personal and family earnings.

 **Support:** You may be eligible for financial support if you are unable to work because of ADPKD or unable to do your usual work.

## Financial implications

There are many things that patients and families can do to help cope with ADPKD.

## Work

In common with many other chronic and progressive diseases, ADPKD can affect employment for patients or carers who leave employment or limit their work to look after affected family members (e.g. spouses and children). Some people also experience difficulties at work because ADPKD limits the activities they can do or requires them to take time off for clinic appointments. Over time, many patients report a loss of personal and family earnings.

If you are medically assessed for employment your diagnosis of ADPKD might arise, especially if you give permission for the assessing company to access your medical records.

If ADPKD is causing problems with work, you might be able to:

- get advice on employment issues from social services (or adult social care) authority, citizen's advice services and trade unions
- apply for social benefit payments to help you cope with financial difficulties – patients should discuss this with their social services authority
- get further advice on these matters from patient organisations in your country.

Having ADPKD can affect certain career choices – for example it may prevent you from joining the armed services. You may wish to seek advice from a careers advice service about this, especially if you are a young person with ADPKD.

You're not alone

What about and your

Seeking personal help

# Wellbeing, personal and family life

This section explains how ADPKD can affect the wellbeing, personal and family lives of patients and their families, what can be done to help cope with these effects, and what support patients can expect.

## Emotional impact

ADPKD can have a profound emotional and psychological impact on patients and their families, and on the community. It can be a source of stress, worry, and sadness, and can affect the ability to work, study, and enjoy life. It can also lead to feelings of isolation and loneliness.

**Support:** You can find support groups and resources to help you cope with the emotional impact of ADPKD on you and your family.

## Financial services

There are many things that patients and families can do to help cope with ADPKD.

## Work

ADPKD can affect your ability to work.

## Travel

ADPKD can affect your ability to travel.

## Family

ADPKD can affect your family life.

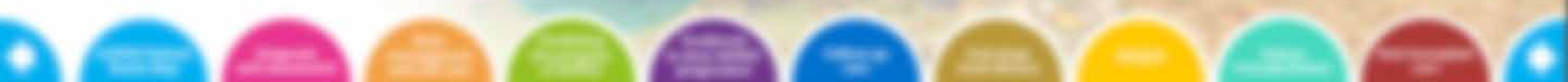


### Financial services

Your diagnosis of ADPKD is likely to arise when you are assessed for health or life insurance, which can be a requirement for substantial financial loans such as mortgages. You may be asked to undergo a medical examination and you may also be asked about your family's medical history, to check for inherited diseases. If you do not answer all questions honestly, your policy may be invalid when a claim is made. You should be sure to specify that you have ADPKD, as the prognosis of other forms of polycystic kidney disease may differ.

Certain types of insurance policies or mortgages may not be available for people with ADPKD, and available policies are usually more expensive than for people without the disease.

- Patient organisations, social services and consumer advice services may provide further advice on the local situation regarding financial issues and actions that patients and carers can take.
- Your healthcare team and patient organisations may also be able to provide information to help you communicate with financial services companies.





This section explains how ADPKD can affect family planning and pregnancy.

People with ADPKD can face difficult decisions when considering whether to have children. This is because pregnancy can be associated with certain risks in women with ADPKD and because their children might inherit the disease.

The EAF and PKD International believe that all patients with ADPKD should have access to family planning services, including counselling and advice on pregnancy, pre-implantation genetic diagnosis, and contraception. This includes young people and adolescents.

## Sexual problems

**Erection problems >** are common in men with chronic kidney disease. Women and men with ADPKD can also experience sexual problems related to body image issues and discomfort caused by kidney growth. You may wish to seek advice from your family doctor or nephrology team if you experience these problems.

## Male fertility

Men with ADPKD may develop cysts in the [seminal vesicles](#), but these do not usually affect the sperm or fertility.

## Contraception

The female sex hormones, oestrogen and progesterone, may worsen liver cysts. Women with moderate-to-severe polycystic liver disease are generally advised to avoid the use of oral contraceptives containing these hormones. Other types of contraceptives are normally available.

## Pregnancy

The majority of women with ADPKD have successful pregnancies. Pregnancy generally progresses normally in women who have ADPKD and whose blood pressure and kidney function are normal.

There is a higher risk of certain [complications >](#) related to high blood pressure and reduced kidney function, and there are considerations regarding the use of some [medicines >](#).

## Dialysis and transplantation

There are special considerations relating to family planning during [dialysis](#) and after [kidney transplantation](#) – you may wish to discuss these with your nephrology or dialysis team.

## Pre-implantation and prenatal genetic diagnosis

ADPKD is caused by specific [genetic](#) mutations, which are usually inherited from a parent. A person with ADPKD faces a one in two (50%) chance of passing the disease onto each child they have.

### **Pre-implantation genetic diagnosis**

Pre-implantation diagnosis (PGD) can show if an embryo created via in vitro fertilisation (IVF) has a genetic mutation linked to ADPKD. This allows people with ADPKD who want to have children to choose an embryo that does not have the ADPKD mutation, and therefore to prevent their children from having the disease.

PDG can only be performed if the specific genetic mutation causing ADPKD in the parent has been identified. More information about PGD is available [here](#).

The EAF and PKD International believe that PGD should be available to all people with ADPKD, as recommended by the [KDIGO Controversies Conference](#).

Results from one [survey >](#) suggest that most people with ADPKD agree with this. However, access to PGD varies across Europe because of regulatory, ethical, legal and funding policies. Differing attitudes towards PGD can also affect access to this method.

### **Prenatal testing**

It is possible to use prenatal testing to check whether an unborn baby in the womb has a genetic mutation linked to ADPKD. This usually involves testing small samples of tissue from the placenta, together with ultrasound scans.

This section explains how **CKPD** can affect family planning and pregnancy.

People with **CKPD** can face difficult decisions when considering whether to have children. This decision depends on the extent of the kidney disease in women with **CKPD** and how well their health is expected to be.

**For all women with kidney disease, it is important to discuss with their doctor the risks of pregnancy and the benefits of dialysis and transplantation. The risks and benefits are different for each woman.**

**Sexual problems**  
Men with **CKPD** often have problems with sexual function. These problems can affect their ability to have children.

**Erection problems** ✕

Problems in gaining or maintaining erections (often known as **erectile dysfunction** or **impotence**) are most common in **end-stage renal disease** and have several possible causes. Kidney transplantation may not improve erectile problems in some men.

**Male fertility**  
Men with **CKPD** often have problems with sexual function. These problems can affect their ability to have children.

**Contraception**  
The risks of pregnancy are higher when a woman has kidney disease. Women with kidney disease should discuss with their doctor the risks of pregnancy and the use of different types of contraception. Other types of contraception are available.

**Pregnancy**  
The majority of women with **CKPD** have successful pregnancies. However, pregnancy is generally considered to be a high-risk pregnancy for women with **CKPD**, and women should discuss with their doctor the risks of pregnancy.

There is a higher risk of complications if a woman with **CKPD** becomes pregnant. Women with **CKPD** should discuss with their doctor the risks of pregnancy and the use of different types of contraception.

**Diagnosis and transplantation**  
There are good reasons to consider kidney transplantation before dialysis and the **benefits** of transplantation. You may want to discuss this with your nephrologist or doctor.

**The implementation and prenatal genetic diagnosis**  
**CKPD** is usually passed **genetically**. However, it is not always possible to predict whether a woman with **CKPD** has a child with **CKPD** before pregnancy. The risks of this are discussed with your doctor.

**Genetic diagnosis**  
Prenatal genetic diagnosis is available for women with **CKPD** who are considering pregnancy. This allows you to find out if your child has **CKPD** before you have the child. However, this is not always possible.

**Transplantation**  
Kidney transplantation is a good option for women with **CKPD** who are considering pregnancy. However, it is not always possible to predict whether a woman with **CKPD** has a child with **CKPD** before pregnancy.

**Sexual problems**  
Men with **CKPD** often have problems with sexual function. These problems can affect their ability to have children.

**Male fertility**  
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This section explains how ADPKD can affect family planning and pregnancy.

People with ADPKD can have different levels of kidney function. The severity of the disease can vary from mild to severe. Some people with ADPKD may have high blood pressure.

The risk of complications during pregnancy is higher for women with ADPKD. This is because the disease can affect the kidneys and the urinary tract. It can also affect the heart and the lungs.

**Medical problems**  
Women with ADPKD may have other medical problems. These include high blood pressure, heart disease, and liver disease. These problems can affect pregnancy and the health of the baby.

**High fertility**  
Women with ADPKD may have a higher chance of getting pregnant. This is because the disease can affect the ovaries and the uterus.

**Complications**  
Pregnancy can be complicated for women with ADPKD. This is because the disease can affect the kidneys and the urinary tract. It can also affect the heart and the lungs. Complications can include high blood pressure, pre-eclampsia, and kidney failure.

**Pregnancy**  
The majority of women with ADPKD have mild to moderate disease. These women can usually have a healthy pregnancy. However, they may need to be monitored closely during pregnancy.

Some women with ADPKD may have severe disease. These women may not be able to have a healthy pregnancy. They may need to be monitored closely during pregnancy.

## Complications



Women with ADPKD are more likely than women without the disease to develop high blood pressure and pre-eclampsia, a potentially serious complication involving high blood pressure and blood in the urine (proteinuria). This means that monitoring during pregnancy is particularly important for women with ADPKD.

Hormones released during pregnancy may increase the growth of liver cysts, but this is not likely to affect the liver function or the pregnancy.

The risk of infections of the kidneys and urinary tract are more common during pregnancy – these can be treated as necessary.

The growth of kidney cysts is not usually affected by pregnancy. However, having multiple pregnancies may increase the risk that kidney function declines over time.

Some experts recommend against pregnancy in women with moderate to severe chronic kidney disease (stage 3–5) caused by ADPKD because of risks to the mother and child.

Women with ADPKD may be advised to give birth in hospitals, rather than elsewhere, so that the midwife and obstetrician (a doctor who specialises in pregnancy, childbirth and the period after childbirth) can closely monitor the mother and baby.

The healthcare team, or a specialist obstetrician, will be able to explain these issues further and develop a monitoring and care plan for patients who wish to go ahead with a pregnancy.

This section explains how ADPKD can affect family planning and pregnancy.

People with ADPKD can face difficult decisions when considering whether to have children. This is because pregnancy can be associated with a higher risk of a woman with ADPKD developing their kidney complications for their babies.

**For all people considering whether to have children with ADPKD, it is important to have a genetic counselling session, usually involving a specialist in pregnancy and reproductive genetic diagnosis, and sometimes a counsellor. This is to help you understand your options.**

## Sexual problems

**Sexual problems** are common in people with kidney failure. Women with ADPKD can also experience sexual problems related to their kidney disease and associated hormonal changes. You may wish to seek advice from your health care professional about how to manage these problems.

## Male fertility

Men with ADPKD may develop problems with the **seminal vesicles**, the glands that usually affect the sperm in fertility.

## Contraception

The choice of contraceptive methods and combinations may differ from usual. Women with kidney failure should discuss their options with their health care professional. Other types of contraception are available.

## Pregnancy

The majority of women with ADPKD have successful pregnancies. However, pregnancy progression normally in women with ADPKD, and other kidney problems associated with kidney disease.

There is a higher risk of complications **associated with kidney disease** and other related health problems, and there are considerations regarding the use of some **medications**.

## Diagnosis and transplantation

There are good considerations regarding kidney donation being **safe** and the **benefits** you may wish to discuss with your nephrologist or health care team.

## The implementation and prenatal genetic diagnosis

ADPKD is usually passed **autosomal recessive** inheritance pattern. A person with ADPKD has a 50% chance of passing the disease onto each child they have.

## The implementation genetic diagnosis

The implementation genetic diagnosis (PGD) can allow you to determine whether or not the foetus will have a genetic disease before you start a pregnancy. Women with ADPKD who want to have children can choose a genetic test that can detect the ADPKD mutation, and therefore predict their children from having the disease.

PGD can only be performed if the genetic genetic mutation causing ADPKD is the same as the mutation that information about PGD is available **now**.

For all people considering whether to have children with ADPKD, it is important to have a genetic counselling session, usually involving a specialist in pregnancy and reproductive genetic diagnosis, and sometimes a counsellor.

**For all people considering whether to have children with ADPKD, it is important to have a genetic counselling session, usually involving a specialist in pregnancy and reproductive genetic diagnosis, and sometimes a counsellor. This is to help you understand your options.**

## Researching

It is possible to research in the area of genetic testing and other related health problems, and there are considerations regarding the use of some **medications**.

**Survey** ✕

According to a survey in the UK, just over half of 96 participating patients with ADPKD would have tried PGD (or might consider it in the future) if it were available on the National Health Service. The majority (69%) of patients believed that PGD should be offered to patients with ADPKD.



This section explains how **ACE** can affect family planning and pregnancy.

Some ACE inhibitors can affect decisions about conceiving children or having a child. This is because pregnancy can be associated with a higher risk of a woman with ACEI and her baby than women not taking the drug.

**ACEI** can affect pregnancy outcomes for a woman with ACEI. This is because pregnancy can be associated with a higher risk of a woman with ACEI and her baby than women not taking the drug.

**Sexual problems**  
Some women who take ACEI can also experience sexual problems such as a lower sex drive and decreased sexual desire. This may be due to side effects from your blood pressure medicine or your experience with the drug.

**Male fertility**  
The use of ACEI may affect sperm in the **male**. This may be due to side effects from your blood pressure medicine.

**Contraception**  
The use of ACEI may affect the effectiveness of some forms of birth control. This is because pregnancy can be associated with a higher risk of a woman with ACEI and her baby than women not taking the drug.

**Pregnancy**  
The use of ACEI may affect pregnancy outcomes for a woman with ACEI. This is because pregnancy can be associated with a higher risk of a woman with ACEI and her baby than women not taking the drug.

**Other things to know**  
Some ACEI can affect decisions about conceiving children or having a child. This is because pregnancy can be associated with a higher risk of a woman with ACEI and her baby than women not taking the drug.

**Diagnosis and transplantation**  
There are special considerations regarding heart disease being diagnosed and the **transplantation**. You may want to discuss this with your nephrologist or doctor.

**The implementation and prenatal genetic diagnosis**  
ACEI can affect genetic testing. This is because pregnancy can be associated with a higher risk of a woman with ACEI and her baby than women not taking the drug.

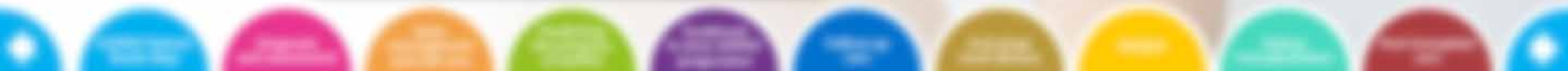
**The implementation genetic diagnosis**  
The use of ACEI may affect genetic testing. This is because pregnancy can be associated with a higher risk of a woman with ACEI and her baby than women not taking the drug.

ACEI can also be performed if the genetic testing results show ACEI in the person's blood. This information about ACEI is useful.

Some ACEI can affect decisions about conceiving children or having a child. This is because pregnancy can be associated with a higher risk of a woman with ACEI and her baby than women not taking the drug.

**Medicines** ✕

Some medicines are not recommended for use in pregnant or breastfeeding women. For example, medicines called angiotensin converting enzyme (ACE) inhibitors or angiotensin receptor blockers (ARBs), used to control high blood pressure are not recommended for women who are pregnant or who wish to try for a pregnancy – your doctor may prescribe an alternative, necessary.



# End-stage renal disease

This section explains end-stage renal disease (ESRD) and the treatment options for patients with ADPKD who reach this stage of kidney disease.

The kidneys have [important roles](#) that are essential to life. The kidneys of people with ADPKD often continue to work normally for many years. However, over time the growth and multiplication of cysts can interfere with the kidney function.

Eventually this can cause the kidneys to fail, meaning that they are not working well enough to support life. This is known as end-stage renal disease (ESRD), or stage 5 [chronic kidney disease \(CKD\)](#) >.

## ? How common is ESRD in people with ADPKD?

Most people with ESRD need 'renal replacement therapy' with either dialysis or [kidney transplantation](#). Kidney failure is life-threatening unless it is treated by one of these approaches.

Doctors can [predict the prognosis](#) of ADPKD and estimate when a patient is likely to reach ESRD. This allows doctors and patients to discuss the available treatment options in advance and plan what to do if ESRD occurs. This is particularly important if a kidney transplant from a living donor is possible. Most hospitals have a special clinic to inform patients on the choices when they are approaching ESRD.

There are many diseases that can lead to ESRD. ADPKD is the most common inherited disease that can lead to dialysis and transplantation. Overall, around one in 10 patients with ESRD have ADPKD.

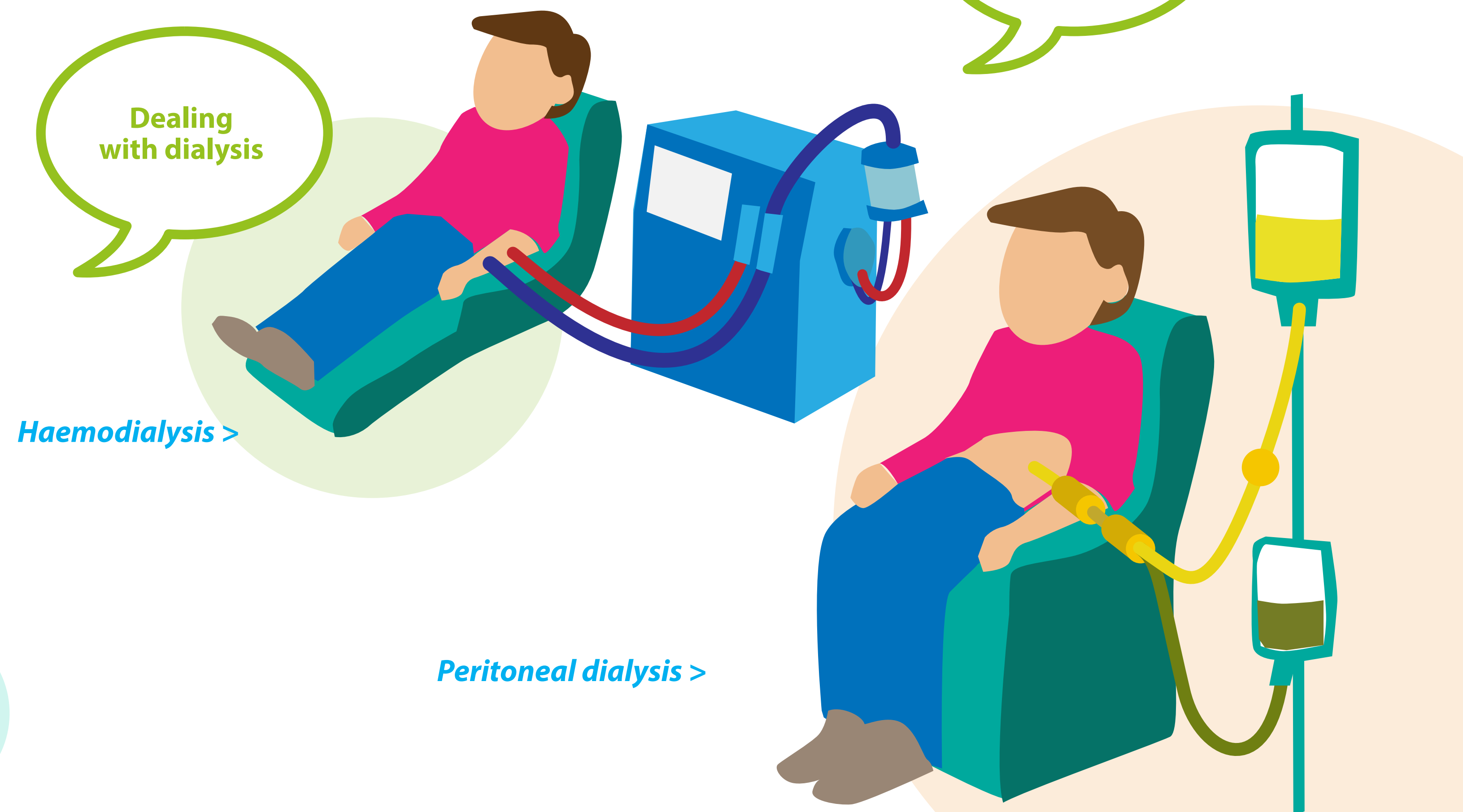
Local kidney and ADPKD [patient organisations](#) will have patients whom you can ask about ESRD and its treatment. Dialysis and transplantation are generally the same for all patients with ESRD and meeting fellow patients is usually a good and helpful experience.

## Dialysis

Dialysis artificially filters the blood to remove the waste products and excess water that build up when the kidneys are not working.

Dialysis is used while patients with ESRD are waiting for a transplant, or if transplantation is not possible. Around nine out of 10 people with ESRD caused by ADPKD have dialysis as their first method of renal replacement therapy, i.e. before or instead of a transplant. Once dialysis is started, it must be continued for the rest of life, unless a transplant is performed. Dialysis only provides around 15% of normal kidney function.

There are two forms of dialysis: haemodialysis and peritoneal dialysis. Haemodialysis is used most commonly, but both methods are suitable for use in most people with ADPKD depending upon the individual circumstances. You may wish to discuss these options with your doctor.



This section explains end-stage renal disease (ESRD) and how it affects people with kidney failure.

The section also explains how people with ESRD can get dialysis and how they can get help with the costs of dialysis.

Eventually, you will need dialysis to help your kidneys work properly. The section explains how to get dialysis and how to get help with the costs of dialysis.

### How can I get help with the costs of dialysis?

There are several ways to get help with the costs of dialysis. You can get help with the costs of dialysis through your health insurance fund.

You can also get help with the costs of dialysis through your local council. You can get help with the costs of dialysis through your local council if you are on a low income.

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### Can I travel while on dialysis?

*'As far as I know, there are great dialysis holidays.'*

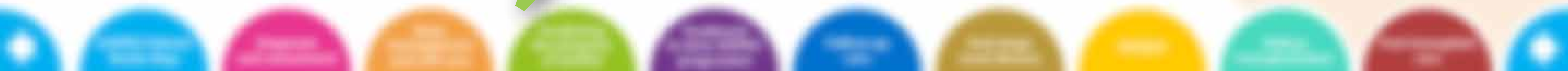
*Nora, Austria*

*'We go on holiday every year and we have always had the opportunity to get dialysis. Of course, we always look in good time for a suitable centre. We have booked through dialysis holiday providers, but mostly we do it ourselves. We then send the doctor's letter to the centre and go on vacation. When my husband still did peritoneal dialysis, we used to send the order so that the dialysis bags and accessories were delivered to our hotel before our arrival, without any problem. For us, dialysis is no reason not to enjoy our vacation.'*

*Valerie, Austria*

*'Thankfully, it was possible to travel despite dialysis. In part, I was assisted by the dialysis team in my ward, but mostly I had to take care of getting in touch with dialysis centres myself. The payment was dealt with by the health insurance fund. I had to take care of the costs of travelling from the hotel to the dialysis centre and back.'*

*Claus, Austria*



## Dealing with dialysis

*'I would recommend everyone having dialysis to take something to do, read or play, it also helps to chat with your "neighbours" there, if they feel like it. If possible, you should try to keep busy, then time will pass by faster.'*

*Nora, Austria*

*'For me it came suddenly "out of the blue" that I had to undergo dialysis. Thankfully, I had the opportunity to talk with some other dialysis patients and hear why they chose one or the other treatment method. I think that made it easier for me to accept my fate.'*

*Phillipp, Austria*

*'When my partner started his dialysis treatment I felt relieved because until then his health continuously declined. The course of the disease turned an athletic and active father and partner into a tired, exhausted person. It was really painful to watch this without being able to help.'*

*Gabriele, Germany*

*'We love dialysis, we don't hate it. The treatment allows us to survive waiting for the transplant to improve our lives. It is true that we depend on a machine to survive, but at least you can find these machines everywhere around the world.'*

*Daniel, Spain*

*'Being on dialysis was sometimes an emotional strain – and probably on my loved ones too. Waiting for a donor with no set date rather than having a live donor planned is just that – a "waiting game" – and it can be tortuous. But, however hard dialysis could get, I remembered it was keeping me alive.'*

*Andy, UK*



# End-stage renal disease

This section explains end-stage renal disease (ESRD) and the treatment options for patients with CKD who reach this stage of kidney disease.

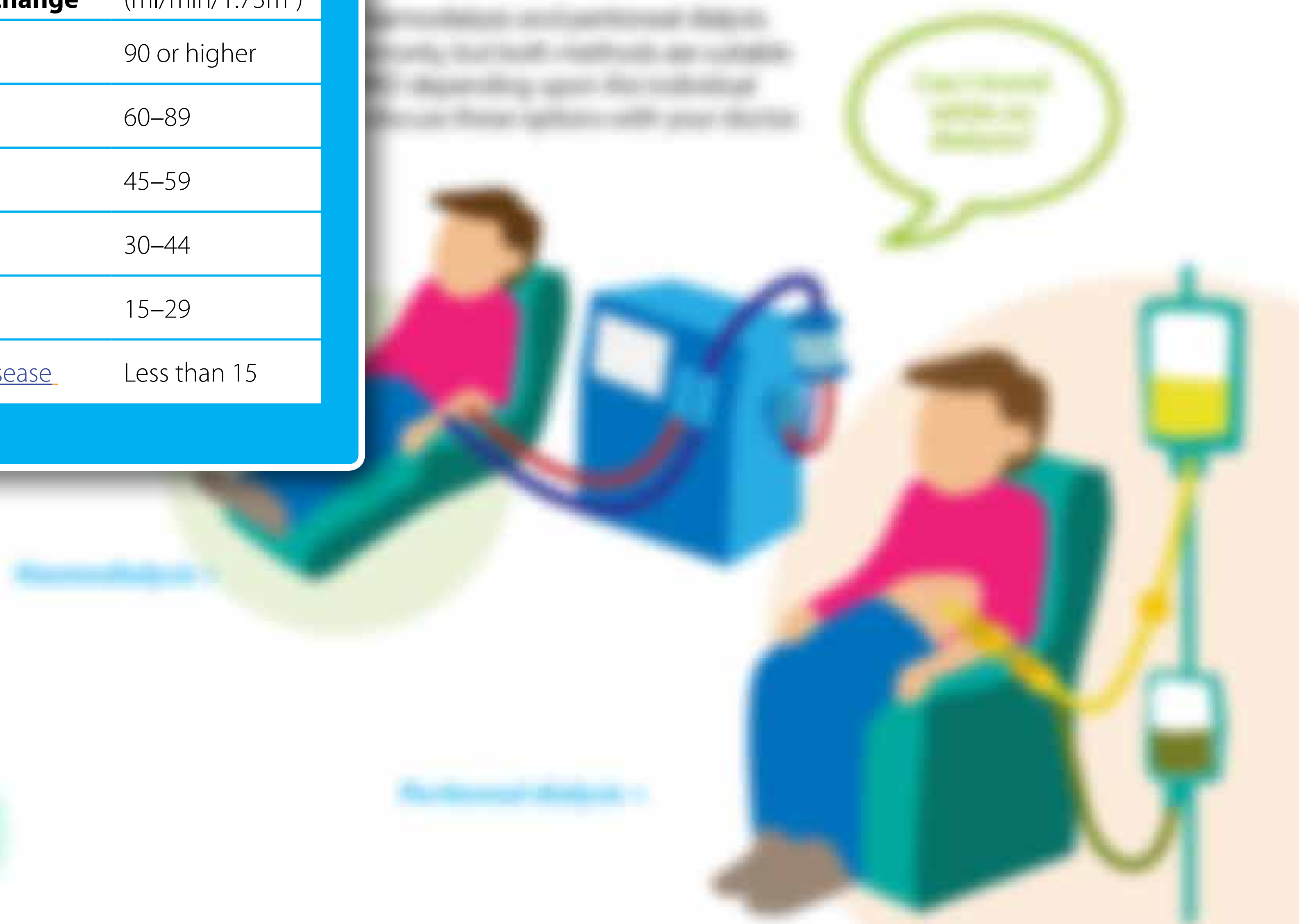
The estimated glomerular filtration rate (eGFR) is an important measure of kidney function. It is calculated from a blood test and a person's age, sex, and body size.

Diagnosis  
Doctors will use blood tests to check the levels of waste products and electrolytes in the blood to see if you have CKD.

## Stages of chronic kidney disease

There are five stages of chronic kidney disease. These are defined by a person's estimated glomerular filtration rate (eGFR), a measure based on a blood test.

Stage	Description of kidney function change	eGFR level (ml/min/1.73m <sup>2</sup> )
1	Normal kidney function	90 or higher
2	Mild loss	60–89
3a	Mild to moderate loss	45–59
3b	Moderate to severe loss	30–44
4	Severe loss	15–29
5	Kidney failure or <a href="#">end-stage renal disease</a>	Less than 15



# End-stage renal disease

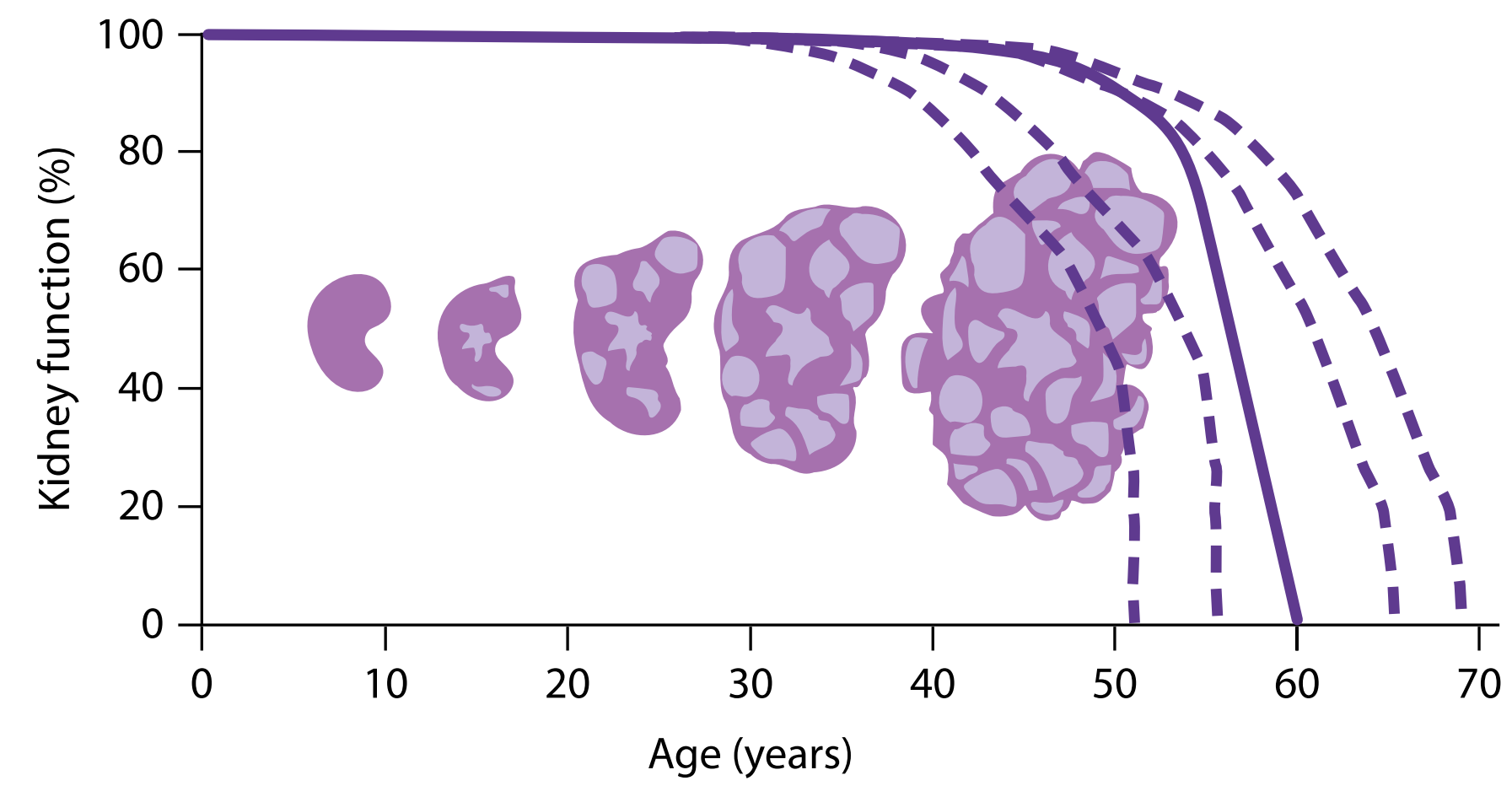
This section explains end stage renal disease (ESRD) and the treatment options for patients with ADPKD who reach this stage of kidney disease.

The kidneys filter waste and extra water from the blood. In people with ADPKD, the kidneys are overgrown and contain many cysts. This can lead to the gradual loss of kidney function over time.

**Diagnosis**  
Doctors will check the blood to measure the waste products and extra water that build up when the kidneys are not working.

## How common is ESRD in people with ADPKD?

Most people with ADPKD will reach ESRD at some point, but the age at which this occurs varies between patients. Around half of patients reach ESRD by the age of 60 years. Around two or three out of 10 people will never reach ESRD.





# End-stage renal disease

This section explains end stage renal disease (ESRD) and the treatment options for patients with ESRD who reach this stage of kidney disease.

The section also covers the different types of dialysis and how to choose the right one for you.

It also explains how to prepare for dialysis and what to expect during a session.

## Haemodialysis

Haemodialysis is a type of dialysis that uses a dialysis machine to filter the blood outside the body.

It is normally performed three times a week, with each session lasting around 4 hours.

Haemodialysis is performed at a nephrology clinic but in some cases it can be done at home.

You can discuss your options with your clinic.

### Haemodialysis



Haemodialysis uses a dialysis machine to filter the blood outside the body. Blood is removed via a needle in the arm and passed through a tube into the machine. Filtered blood is then returned to the arm via another tube.

*Preparation:* A small operation is performed to create a special blood vessel (called a 'fistula') in the arm, through which blood is passed to and from the dialysis machine. If there are problems with creating this fistula, a tube can be inserted into a large vein in the neck instead. This is called a 'neck line' and is used as a temporary measure.

*Procedure:* Haemodialysis is normally performed three times a week, with each session lasting around 4 hours. However, this can vary. Haemodialysis is performed at a nephrology clinic but in some cases it can be done at home. You can discuss your options with your clinic.

*Side effects and precautions:* People who have dialysis are instructed to limit the amount of fluid they drink. Special dietary requirements (normally advised by a dietician) are needed to reduce the intake of salt and other minerals – this helps reduce the build-up of fluid and minerals between dialysis sessions.

The main side effects include: tiredness, low blood pressure, sepsis (infection in the blood), muscle cramps and itchy skin. You can ask your healthcare team for further advice about these.

...the water outside and ...

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...the water outside and ...



# End-stage renal disease

This section explains end stage renal disease (ESRD) and the treatment options for patients with ADPKD who reach this stage of kidney disease.

The information in this section is for people with ADPKD who have reached end stage kidney disease. It does not cover the general advice for people with kidney disease.

For more information on end stage kidney disease, see the section on [End-stage renal disease \(ESRD\)](#).

## How to choose a dialysis treatment

When you reach end stage kidney disease, you will need to choose a dialysis treatment. There are two main options: haemodialysis and peritoneal dialysis.

Haemodialysis is done in a dialysis centre. It involves a machine that filters your blood. Peritoneal dialysis is done at home. It involves a catheter that filters your blood through your abdomen.

Both treatments have pros and cons. You should talk to your healthcare team to decide which one is best for you.

Peritoneal dialysis is often preferred for people with ADPKD because it is less restrictive than haemodialysis. However, it may not be suitable for everyone.

### Peritoneal dialysis



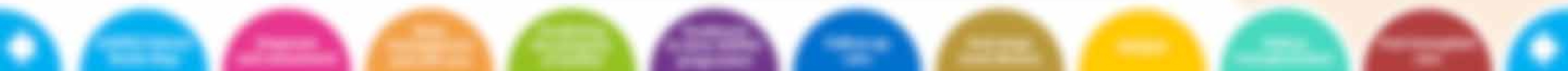
Peritoneal dialysis filters the blood through the lining of the patient's own abdomen (called the peritoneum). A special dialysis fluid is instilled into the space in the abdomen (the peritoneal cavity). This fluid draws waste products and excess water from the blood as it passes through the peritoneum, and is then removed.

Peritoneal dialysis can be used instead of haemodialysis in many people with ADPKD. However, when the kidneys are very large or there are frequent cyst infections, peritoneal dialysis might not be a good option.

*Preparation:* An operation is performed to insert a permanent tube (called a catheter) through the skin into the abdomen.

*Procedure:* Dialysis fluid is pumped into the peritoneal cavity, left there for several hours and then drained into a bag. The process is then repeated, using fresh fluid, several times a day. If available, special machines can do all this overnight while you sleep. Peritoneal dialysis can be done by patients at home, but it must be done every day. CAPD allows you to do other things during fluid exchanges, e.g. use a computer, sew or watch television. A type of peritoneal dialysis called automated peritoneal dialysis uses a machine to do the exchanges overnight while you sleep.

The main side effects include: tiredness, peritonitis (infection of the peritoneum), hernia and weight gain. Some people find the catheter troublesome. You can ask your healthcare team for further advice on these issues.



## What is kidney transplantation?

Kidney transplantation involves removing a kidney from a donor and transferring it to a person with ESRD. People can live with just one kidney so only one donor kidney needs to be transplanted.

Where possible, kidney transplantation is the best treatment for ESRD. Preferably, [pre-emptive kidney transplantation](#) > is performed before ESRD occurs. However, kidney transplantation is not suitable for everyone and the [criteria](#) > for suitability can vary.

In rare cases, patients with severe [liver cysts](#) together with ESRD may be considered for a combined liver and kidney transplant.

## Who donates the kidney?

The transplanted kidney can be provided by a living or deceased donor. You may wish to discuss these options with your nephrologist.

**Living donor** > : A kidney donation from a suitable living person tends to work best and can be planned in advance as an 'elective' operation.

**Deceased donor** > : The alternative is for patients to go onto a waiting list to receive a kidney from a person who has recently died.

The EAF and PKD International believe that patients with ESRD should be offered the opportunity to join a kidney transplant waiting list, if they are medically suitable.

## What does transplant involve?

**Kidney transplantation surgery** > involves a major operation organised by a transplant team and performed by a transplant surgeon.

While awaiting a transplant, it is important to stay as healthy as possible through [basic ADPKD management and self-care measures](#). The transplant team should give specific instructions about what to do nearer the time of the operation.

Generally, transplantation is just as likely to be successful in people with ADPKD as with any other kind of kidney disease. As with any operation, there are [risks](#) > and you should discuss these with the nephrologist or transplant surgeon.

 **How long does a kidney transplant work for?** >

## Post-transplant care – what happens afterwards?

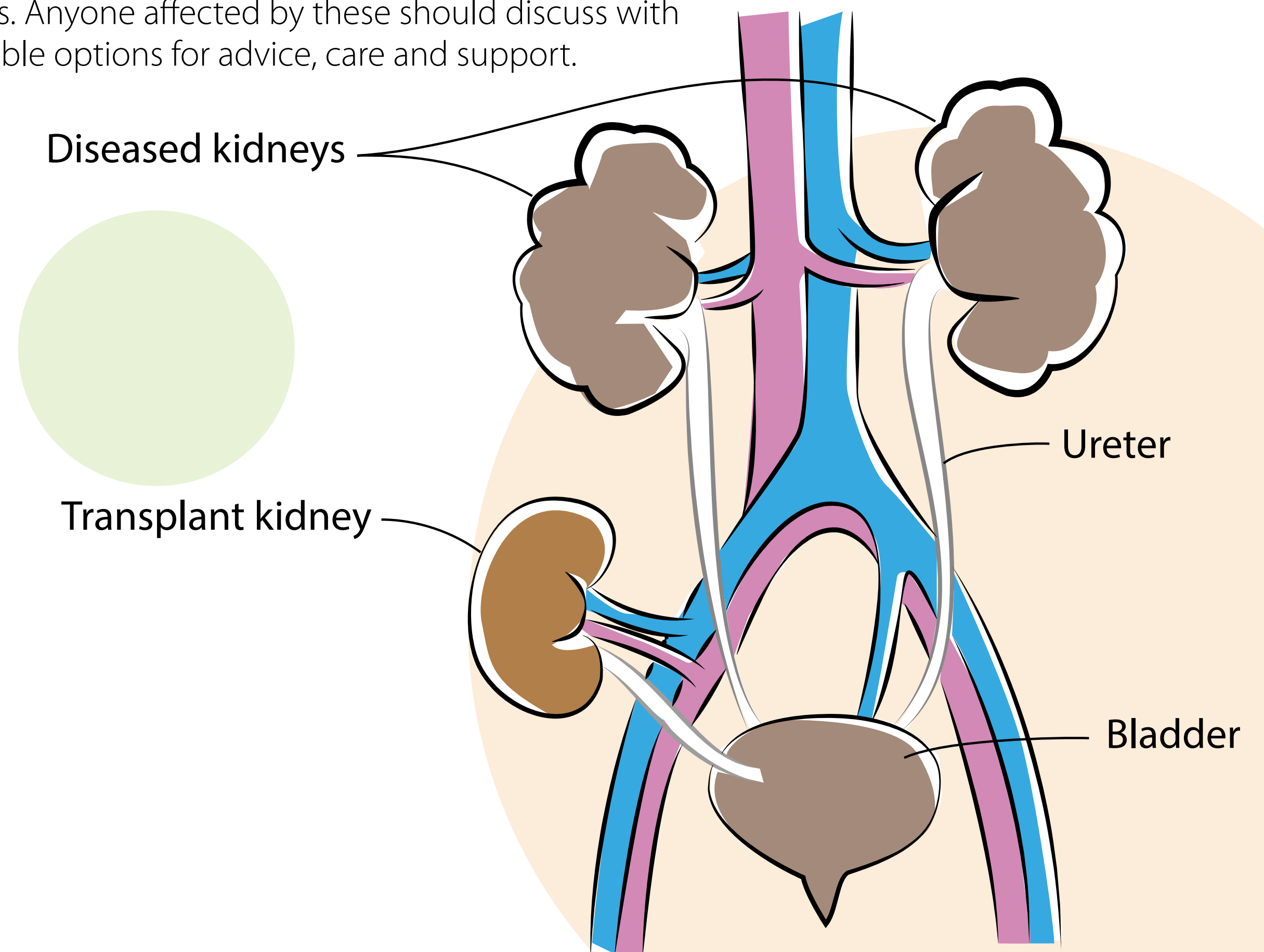
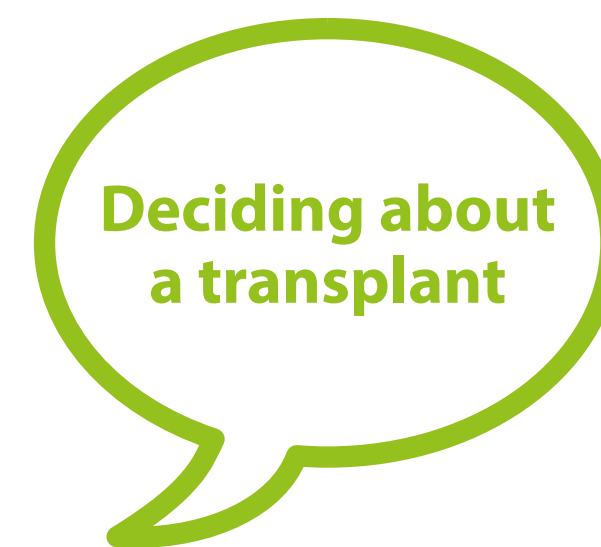
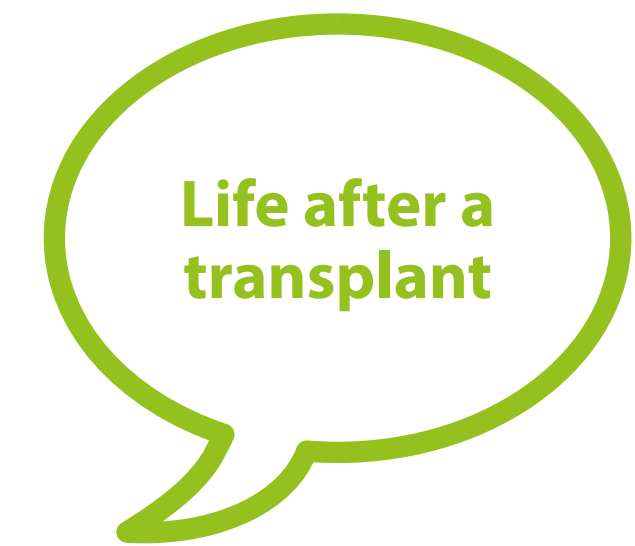
Most transplanted kidneys start to work immediately. Sometimes a transplant can take a few days or weeks to work properly, in which case dialysis may be needed temporarily. The time taken for recovery varies, but most patients should be able to leave hospital in around 1 week and to return to work and normal activities within a few months.

Long-term [follow-up care](#) > is essential to make sure the transplanted kidney is working, to manage [immunosuppressant medication](#) > necessary to prevent the body rejecting the new kidney, and to check for any complications.

Many core aspects of [basic ADPKD management and self-care](#) are still important, such as a healthy diet, weight control, smoking cessation, limiting alcohol intake and treatment for high blood pressure.

Remember, you will still have ADPKD even after a kidney transplant and so [complications](#) that occur elsewhere in the body may still need treatment.

Transplantation can have emotional and psychological effects both in patients and in donors. Anyone affected by these should discuss with their doctor the available options for advice, care and support.



## Deciding about a transplant

*'I simply couldn't ask anyone from my social circle to donate a kidney – I couldn't have accepted it, although some friends did offer.'*

*Claus, Austria*

*'Since I needed both a liver and kidney transplant, the decision for or against was very difficult. I decided to take the step only after a thorough preparation, which focused mainly on ethical and psychological concerns. My family was also an issue of course. My basic idea was first of all that the surgery could be fatal and second, that the transplantation and the possible lifetime thereafter would certainly be more successful if I made my decision independently of other people. Meanwhile, I've already experienced my first "transplant birthday", with relatively few complications. That said, I still feel fragile sometimes.'*

*Selina, Austria*

*'It was upsetting when strangers thought I was pregnant because of my enlarged liver... In a sense I feel emotionally more comfortable after the transplant. Previously I felt the "duty" of having to keep working and functioning until final kidney failure occurred – emotionally and mentally it was incredibly difficult and*

*distressing. Carefree living was not possible, and apart from that, the responsibility towards my daughter, and the fear and panic not to be able to accompany her to adulthood in the worst-case scenario.'*

*Helena, Austria*

*'After the doctors told me I needed a new kidney I was speechless. And that's really rare for me!'*

*Petra, Germany*

*'When my estimated glomerular filtration rate (eGFR) dropped below 20, it was suggested that I start thinking about live transplant. My great niece volunteered and was a really good match. From the time she agreed to donate to transplant was about 2¾ years. I had the transplant at eGFR 9, in October 2016. The transplant team told me that mine was a 'textbook' pre-emptive live donation. My niece sailed through it – she wanted food as soon as she came back from theatre! The transplant was on the Thursday and she went home on the Monday. I was in the full week. Once out, I was very well looked after by my wife.'*

*Martin, UK*

## Life after a transplant

*'Even though I'm glad that dialysis treatment exists, I'm happy I don't have to do it three times a week anymore – since I'm successfully transplanted now. Dialysis had taken away too much from my quality of life, though I managed to get it back a little by little. After many years with a transplant, I realise that one becomes careless and sloppy when it comes to taking immunosuppressant medicines. You know very well that you have to take them, but the "inner laziness" is sometimes bigger. Since you do not feel any pain or immediate reaction if you do something wrong, your attention decreases.'*

*Benjamin, Austria*

*'The transplant operation went smoothly and I woke in recovery as if nothing had happened, apart from all the tubes. However, the following day I did feel nauseous, and was sick, then Day 2 I started to go into full recovery, and Day 3 the kidney started working and all tubes were removed. On Day 5 I was able to come home. Initially it went well. I have had a hiccup with a viral infection, but my lifestyle is getting back to normal. I'm feeling healthier, and about to start up physical fitness again. Even with the hurdles I've encountered, my GFR has been up to 65%, and I am looking forward to the future.'*

*Ian, UK*

*'Dialysis hardly worked for me at all. The doctors had said that my need for a transplant was urgent. Around three months after an operation to remove my PKD kidneys, the consultant felt I could risk going on the transplant list. Just one week later, I was at home and I received a call to go immediately to the hospital. I had no reservations about having the transplant even though I realised I*

*was vulnerable to infection and still had pain from recent surgery. My creatinine level went from 1,400 to 100 overnight and I woke up feeling like a brand-new woman. I was well cared for and a future felt possible once again. I have been on quite a journey over the last 13 years. Certainly, this donation prolonged my life span and increased my quality of life immensely – I have been able to travel, gain a Masters degree, continue to work for some time and create a home for myself. I have also had to deal with the traumatic impact ADPKD has had on my emotional and physical health, and have had some unfortunate experiences of employers and colleagues not understanding what this has been like. Even so I wouldn't have had it any other way. To share life with another person is a great privilege in both directions.'*

*Nicki, UK*

*'Immediately after the transplant, my skin lost its yellow tinge and I gained a normal complexion. I felt normal again, just like I used to. The biggest change was no longer being tied to dialysis three times a week, and once again being able to do all the things that I used to without having to take regular breaks. One thing I will never forget. How lucky I have been.'*

*Rob, UK*

*'I have no dialysis and no dietary restrictions and my fitness is such that I play golf, have been back pedalling on the bike and I competed at this year's Transplant Games. Life will never entirely return to what it was before, but every day feels like a massive bonus. I am immeasurably grateful to the medics and of course to my donor and his family.'*

*Stephen, UK*

## Pre-emptive kidney transplantation

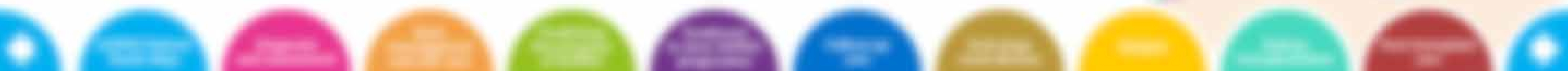
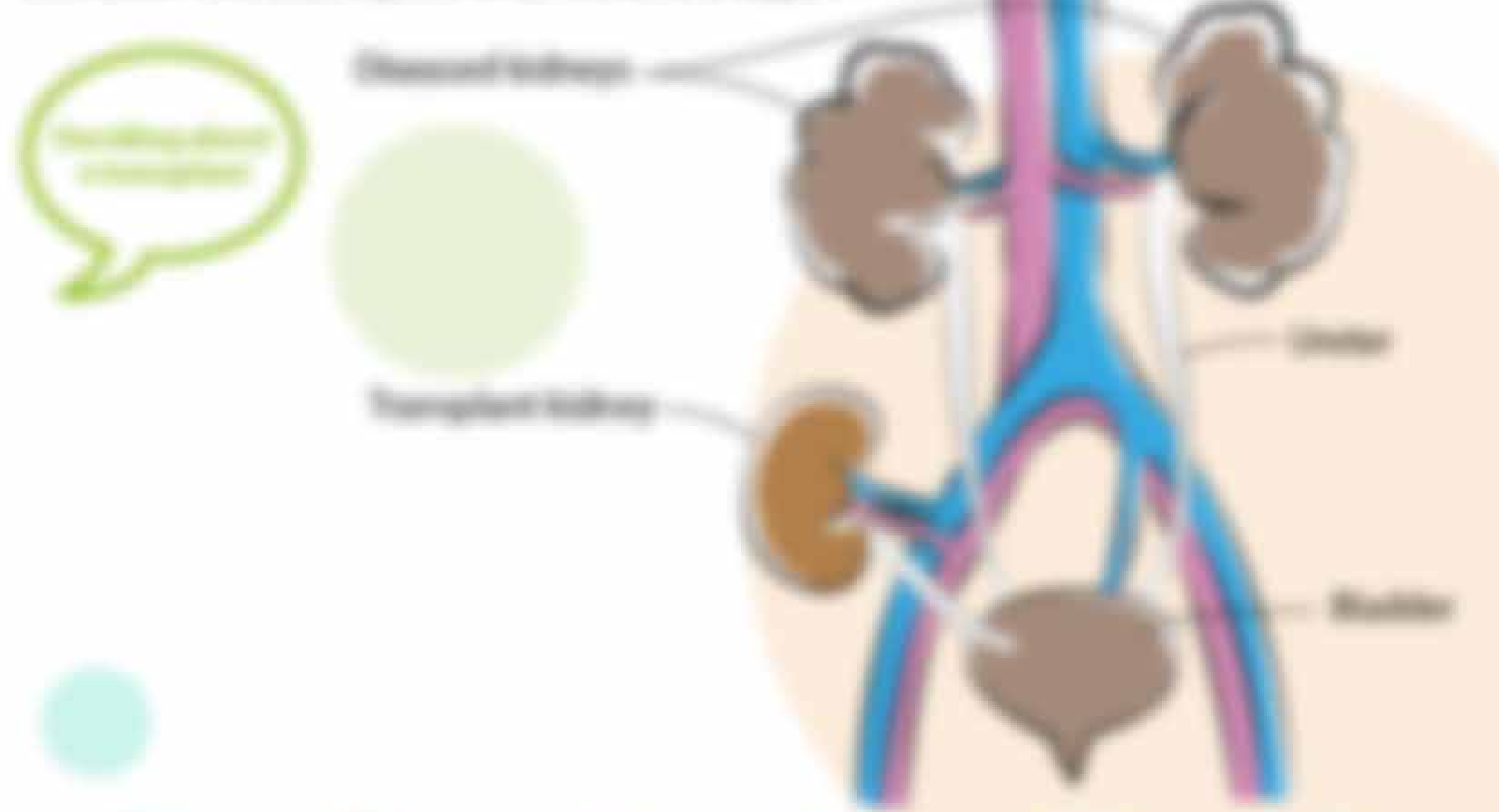
Where possible, kidney transplantation is the best treatment for ESRD. This is because a successful transplant restores kidney function without the need for dialysis and can offer patients a better quality of life. It is also far more cost-effective for the health system, as compared with dialysis.

Preferably, kidney transplantation is best done 'pre-emptively' before ESRD occurs because this is most likely to be successful. A pre-emptive transplant allows patients to choose to receive a kidney either from a living donor or a deceased donor.

## Post-transplant care - what happens afterwards?

After a kidney transplant, you will need to take medication to prevent your body from rejecting the new kidney. This is called immunosuppression. You will also need to have regular blood tests to check your levels of immunosuppressants and to monitor your kidney function. It is important to follow your doctor's advice carefully. You will also need to have regular check-ups with your transplant team. You will also need to have regular check-ups with your transplant team. You will also need to have regular check-ups with your transplant team.

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# Kidney transplantation

## What is kidney transplantation?

Kidney transplantation involves removing a kidney from a donor and transferring it to a patient with CKD. There are two types of kidney transplantation: living donor and deceased donor.

## Who can donate a kidney?

Living donors can be family members or friends. Deceased donors are people who have died and their organs are donated to someone in need.

## Why donate a kidney?

Living donors can help someone with CKD who is not yet on dialysis. Deceased donors can help someone who has been on dialysis for a long time.

**Living donor:** A living donor is someone who gives one of their kidneys to someone with CKD. This is usually a family member or friend.

**Deceased donor:** A deceased donor is someone who has died and their organs are donated to someone in need. This is usually a family member or friend.

**How long does a kidney transplant last?** A kidney transplant can last for 10 to 15 years, but some people have had theirs last for 20 years or more.

## What does transplant involve?

**Living donor transplant:** This involves a living donor giving one of their kidneys to a patient with CKD. The surgery is usually done laparoscopically.

**Deceased donor transplant:** This involves a deceased donor's kidney being donated to a patient with CKD. The surgery is usually done open.

Generally, transplantation is used as a life-saving treatment for people with CKD, as well as for those with other kidney diseases. It is not a cure for CKD, but it can help you live longer and better with the help of a transplant surgeon.

[How long does a kidney transplant last?](#)

## Post-transplant care - what happens afterwards?

After transplantation, you will need to take immunosuppressant drugs to prevent your body from rejecting the kidney. You will also need to have regular blood tests to check your kidney function and to monitor for any side effects of the drugs.

**Living donor transplant:** You will usually be able to go home the day after surgery. You will need to take immunosuppressant drugs for the rest of your life.

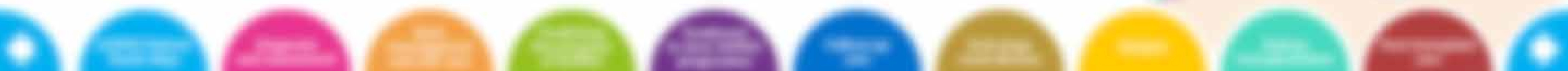
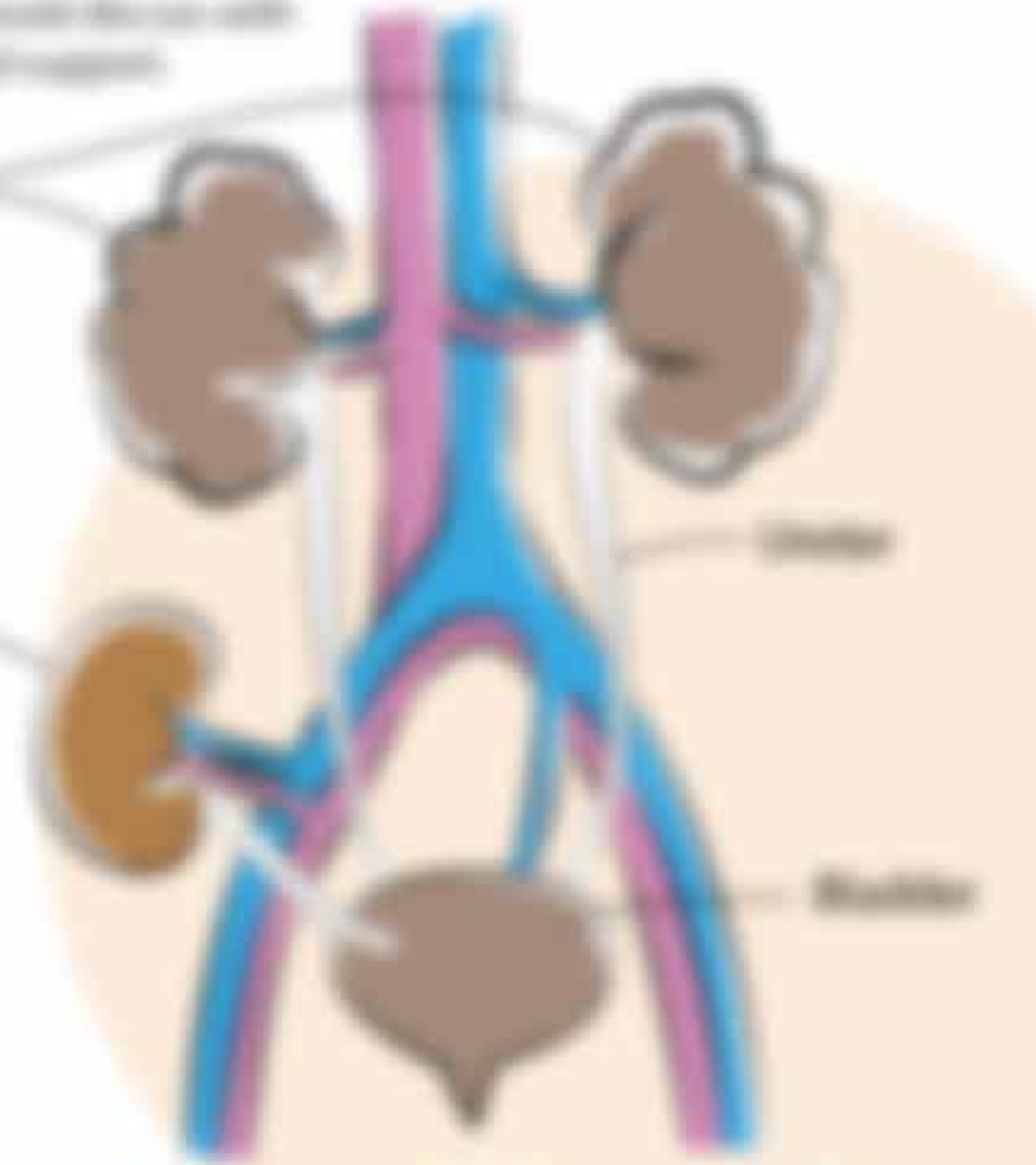
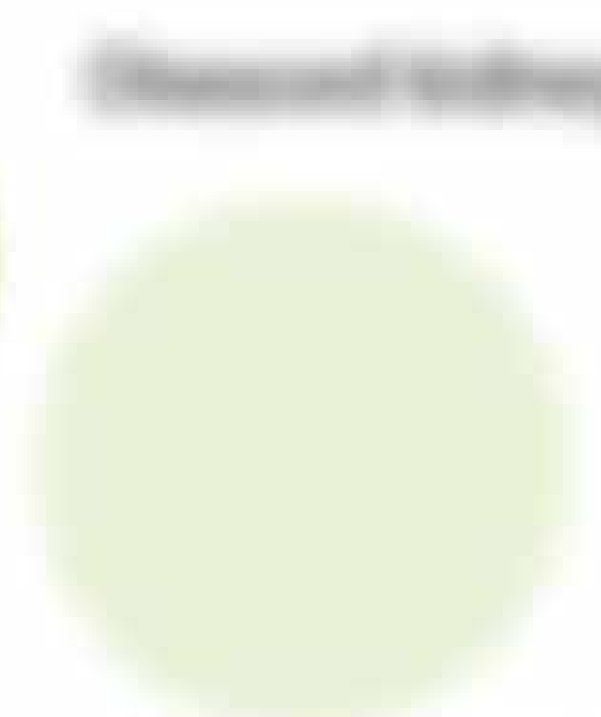
**Deceased donor transplant:** You will usually be in hospital for 1 to 2 weeks after surgery. You will need to take immunosuppressant drugs for the rest of your life.

Immunosuppressant drugs can have side effects, such as increasing your risk of infections and cancer. You will need to have regular blood tests to check your kidney function and to monitor for any side effects of the drugs.

Transplantation can have emotional and psychological effects, such as stress and depression. It is important to talk to your doctor about these issues and to seek support if you need it.

**Criteria**

Generally, patients must be healthy enough for surgery, have a good chance of a successful transplantation, and be willing and able to take certain treatments that are necessary after the transplant.



# Kidney transplantation

## What is kidney transplantation?

Kidney transplantation involves removing a diseased kidney and replacing it with a healthy one. This can be done using a kidney from a deceased donor or a living donor.

There are two types of kidney transplantation: living donor and deceased donor. Living donor transplantation involves a healthy person donating one of their kidneys to a patient with kidney failure.

## Living donor

Living donor transplantation requires a suitable donor who is willing and able to undergo surgery to remove one of their kidneys. The donor is often a close relative as this reduces the risk that the patient's immune system will attack (or 'reject') the donated kidney.

Potential donors are carefully checked to make sure they do not have ADPKD themselves – this may include genetic testing. Many other assessments are also recommended for potential donors, including tests for HIV, hepatitis B and C and cytomegalovirus (CMV) to reduce the risk that these infections are transmitted to the patient. Potential donors should also be counselled about the risks and requirements associated with donation.

Attitudes and policies regarding living donor transplantation vary across Europe. Living donor kidney transplantation is used most commonly in the Netherlands, followed by Turkey, Cyprus, Denmark and the United Kingdom. It is far less common in some other European countries.

## Best transplant case - what happens afterwards?

After transplantation, the patient will need to take medication to prevent their immune system from attacking the new kidney. This is called immunosuppression. The doctor will also check the kidney function regularly to make sure it is working properly.

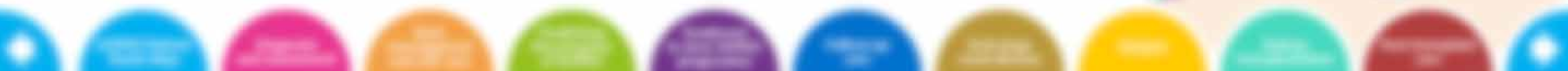
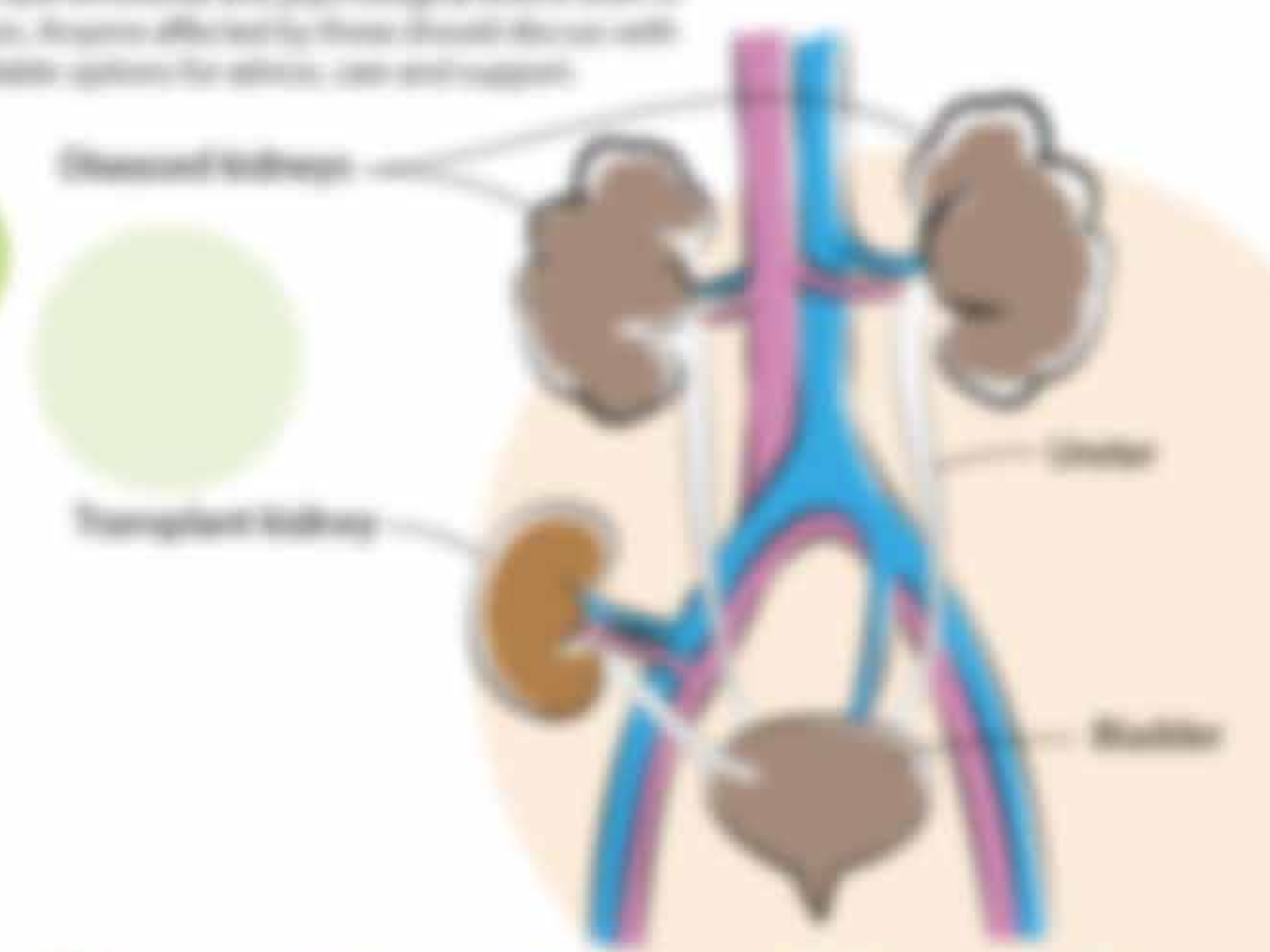
Living donor transplantation is considered to be the best option for many people with kidney failure.

Living donor transplantation is a complex process and it is important to talk to a specialist about the risks and requirements associated with donation.

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# Kidney transplantation

## What is kidney transplantation?

Kidney transplantation involves removing a kidney from a donor and transferring it to a patient with CKD. There are two types of kidney transplantation: living donor kidney transplant and deceased donor kidney transplant.

When possible, kidney transplantation is the best treatment for CKD. However, [not everyone is eligible for a transplant](#). A transplant will only be successful if the donor and recipient are compatible. The [compatibility test](#) is used to check this.

It is important to discuss with your [healthcare professional](#) whether you are suitable for a transplant and what the risks are.

## Best transplant case - what happens afterwards?

After transplantation, you will need to take medicines to prevent your immune system from rejecting the kidney. The medicines will also help to prevent infection. The medicines will also help to prevent rejection of the kidney. The medicines will also help to prevent rejection of the kidney.

Living donor [kidney transplant](#) is considered to be the best option for kidney transplantation. It is considered to be the best option for kidney transplantation. It is considered to be the best option for kidney transplantation.

The [National Kidney Foundation](#) (NKF) estimates that there are 100,000 people on the kidney transplant waiting list in the US. In the UK, there are approximately 10,000 people on the kidney transplant waiting list.

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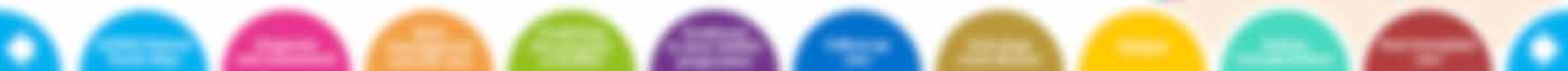
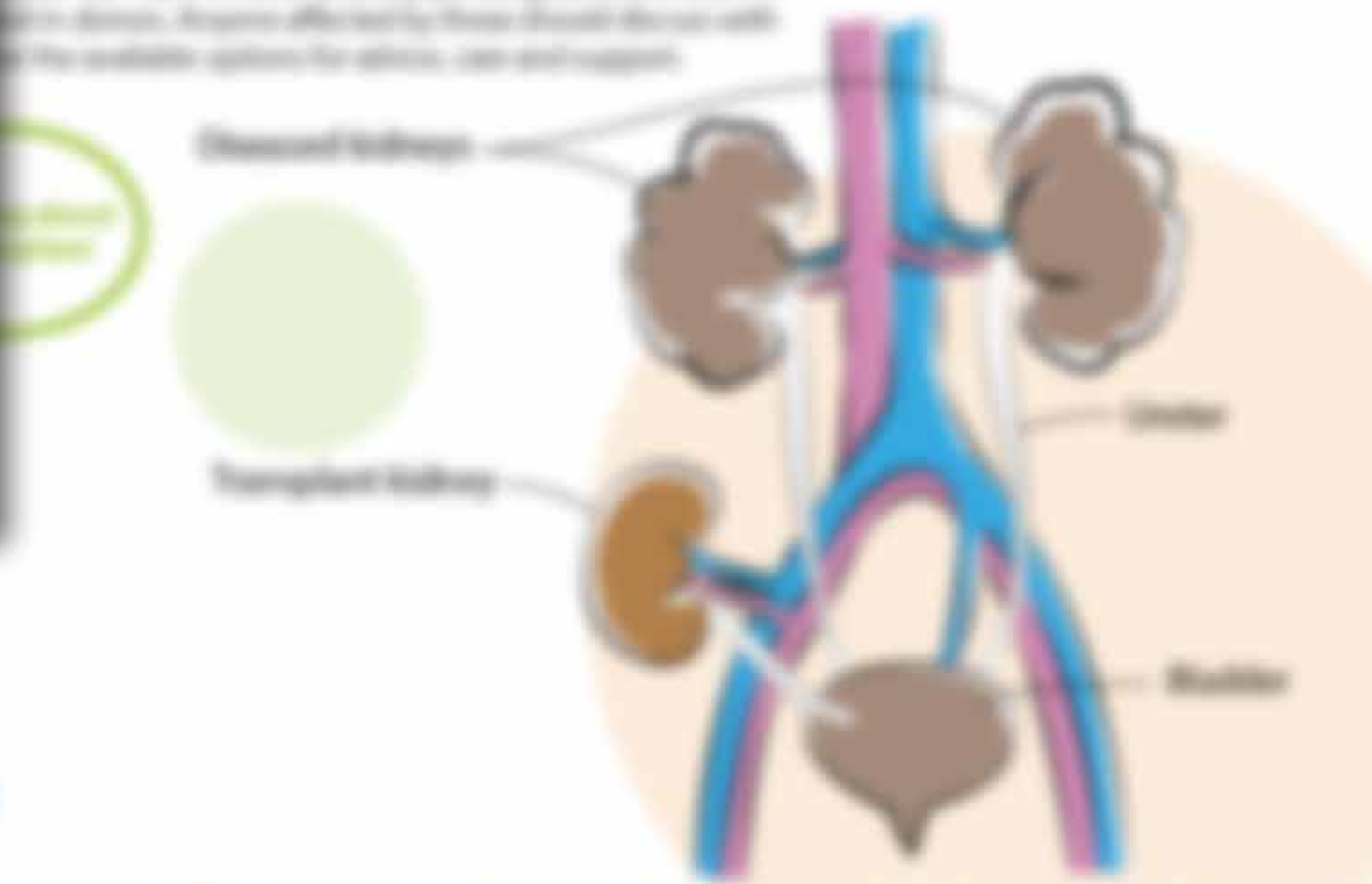
The [National Kidney Foundation](#) (NKF) estimates that there are 100,000 people on the kidney transplant waiting list in the US. In the UK, there are approximately 10,000 people on the kidney transplant waiting list.



**Deceased donor** ✕

For a deceased donor transplant, patients need to go onto a waiting list to receive a kidney from a person who has recently died. Once a kidney is available, the transplantation operation is performed at short notice, as quickly as possible.

The length of waiting lists varies between countries. Patients may have to wait many months, or even several years, for a transplant. In the UK, for example, patients spend an average of 2–3 years on a waiting list. The main problem is a shortage of available kidneys. Patients should discuss the situation in their country. Once a deceased donor kidney becomes available, urgent transplant surgery is performed as soon as possible.



# Kidney transplantation

## What is kidney transplantation?

Kidney transplantation involves removing a kidney from a donor and transferring it to a patient with CKD. There are two types of kidney transplantation: living donor kidney transplantation and deceased donor kidney transplantation.

When possible, kidney transplantation is the best treatment for CKD. However, [not everyone is eligible for a transplant](#). A transplant can help you feel better and live longer. However, kidney transplantation is not a cure for CKD. You will still need to take medicines and follow a diet to help your kidneys.

It can take a while to get a transplant. You may need to wait for a while before you can get a transplant. You may also need to wait for a while before you can get a transplant.

## Why donate the kidney?

The most common reason for donating a kidney is to help someone with CKD. You can donate a kidney to a living donor or a deceased donor.

## Living donor

A living donor is someone who gives a kidney to a patient with CKD.

## Deceased donor

A deceased donor is someone who gives a kidney to a patient with CKD after they have died.

## How long does it take to get a transplant?

It can take a while to get a transplant. You may need to wait for a while before you can get a transplant.

## What does the surgery involve?

The surgery involves removing a kidney from a donor and transferring it to a patient with CKD.

## What does the patient experience?

The patient will experience pain and discomfort during the surgery. They will also need to take medicines and follow a diet to help their kidneys.

## What are the risks of kidney transplantation?

There are several risks of kidney transplantation, including infection, rejection, and complications from the surgery.

## How long does a kidney transplant last?

A kidney transplant can last for several years, but it is not a cure for CKD. You will still need to take medicines and follow a diet to help your kidneys.

## What should I expect after the surgery?

After the surgery, you will need to take medicines and follow a diet to help your kidneys. You will also need to have regular check-ups with your doctor.

## How long does kidney transplantation wait list?

The wait list for kidney transplantation can be long. It can take several years to get a transplant.

## Best transplant case - what happens afterwards?

After a successful kidney transplant, you will need to take medicines and follow a diet to help your kidneys. You will also need to have regular check-ups with your doctor. The most common complication is rejection, which can happen if your body sees the new kidney as a threat and attacks it. This can happen within a few weeks.

Living donor [transplantation](#) is considered to result in better long-term kidney survival compared to [deceased donor transplantation](#). However, it is important to consider the risks of rejection, which can happen if your body sees the new kidney as a threat and attacks it.

There are several risks of [living donor transplantation](#), including infection, rejection, and complications from the surgery. However, living donor transplantation is considered to result in better long-term kidney survival compared to deceased donor transplantation.

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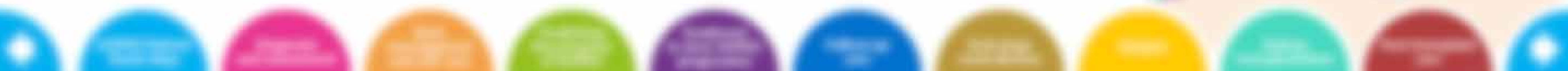
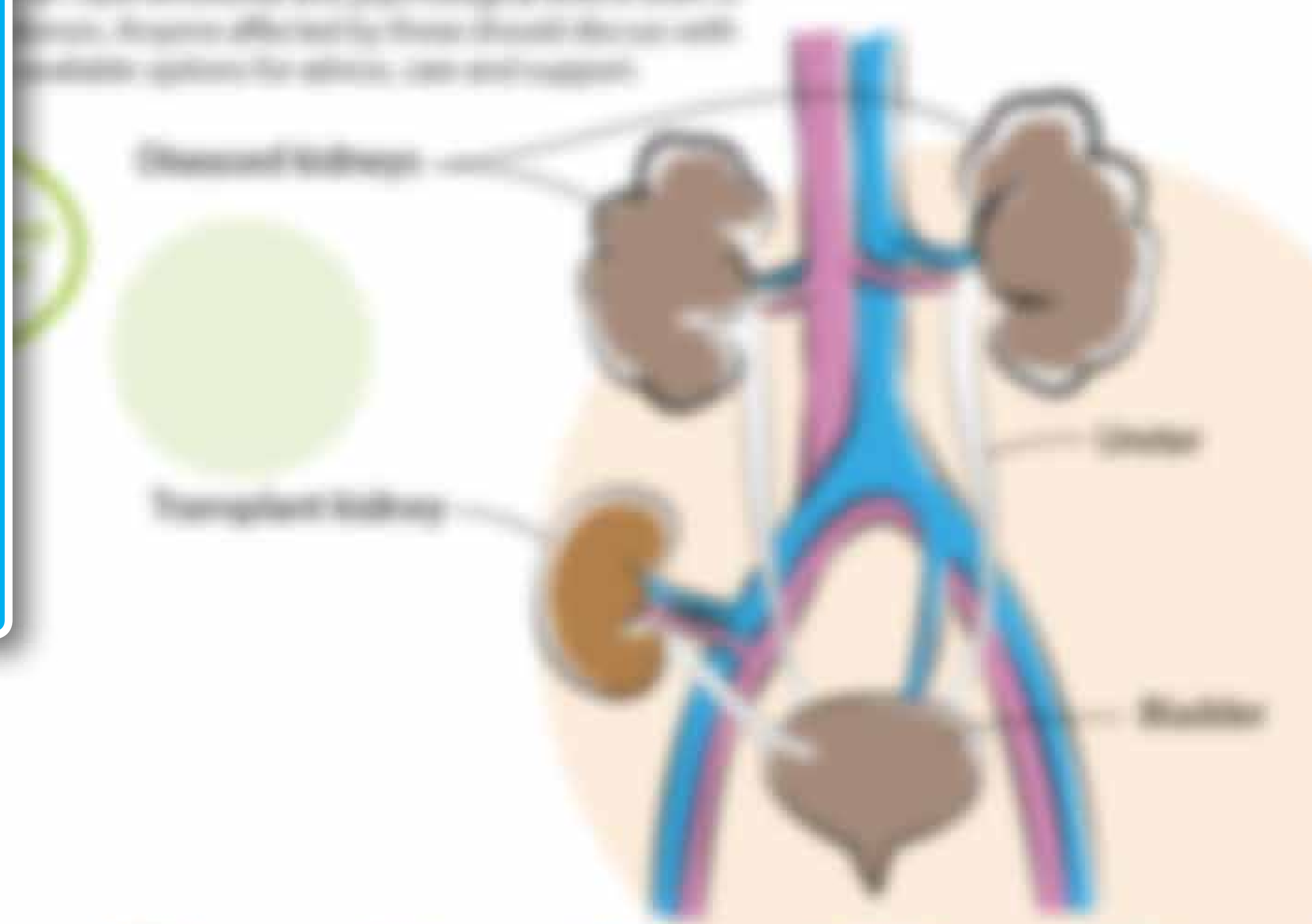
After a successful kidney transplant, you will need to take medicines and follow a diet to help your kidneys. You will also need to have regular check-ups with your doctor. The most common complication is rejection, which can happen if your body sees the new kidney as a threat and attacks it.

### Kidney transplantation surgery

Kidney transplantation is a major surgical operation carried out under general anaesthetic. It may be done at a specialist transplant centre.

The abdomen is opened up and the donor kidney is inserted. The kidney is connected to the normal blood vessels and to the tubes that take urine to the bladder.

The patient's own kidneys are usually left in place and normally these shrink after transplant surgery. However, one or both may be removed in certain situations, for example if they are very large and taking up space needed for the transplanted kidney, or if they are causing complications.



# Kidney transplantation

## What is kidney transplantation?

Kidney transplantation involves removing a kidney from a donor and transferring it to a person with CKD. A kidney can be used with or without the donor's liver. Some donors have kidney disease in the transplanted kidney.

When possible, kidney transplantation is the best treatment for CKD. However, [not everyone is eligible for a transplant](#). A transplant can help you live longer, but it does not cure CKD. However, kidney transplantation is not suitable for everyone and the [risks](#) of a transplant can vary.

It can cause problems with your [blood sugar](#) together with CKD may be considered for a combination of dialysis and transplant.

## Why donate the kidney?

The transplant team can be considered for living or deceased donor. You may wish to discuss these options with your transplant team.

**Living donor:** A living donor is someone who voluntarily gives a kidney to help someone else. A living donor can be a family member or a friend.

**Deceased donor:** The deceased donor program is a program that allows people to donate their organs after they have died.

**How can I find a living donor?** You can find a living donor through a transplant center or a national living donor registry.

### Risks

The risks of transplantation surgery include urinary tract infections, diverticulitis, blood clots and diabetes. There is also the risk that your body's immune system might reject the new kidney. This risk is reduced using medicines that suppress the immune system, known as immunosuppressants.

Living kidney donors should also discuss the potential risks associated with donation, which include hypertension and increased costs of health insurance.

## Post-transplant care - what happens afterwards?

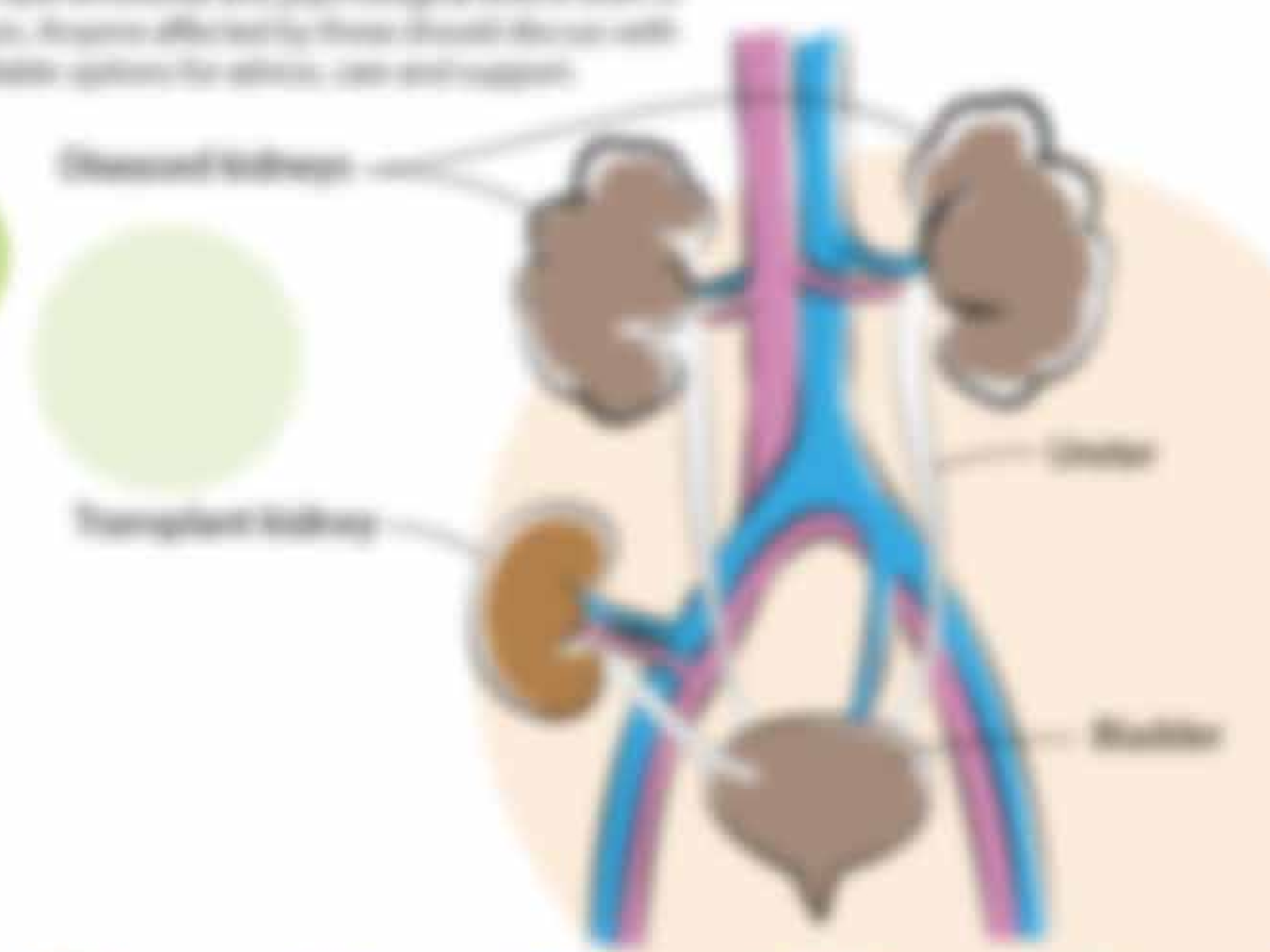
After transplantation, you will need to take medicines to prevent your body from rejecting the kidney. The medicines you take will also help you stay healthy. You will need to have regular check-ups with your transplant team. The first year after the transplant is the most important. You will need to have regular check-ups with your transplant team to make sure the kidney is working properly.

Living with a [transplanted kidney](#) is similar to living with the transplanted kidney in a living donor. [Living with a transplanted kidney](#) is similar to living with a transplanted kidney in a living donor. You will need to have regular check-ups with your transplant team to make sure the kidney is working properly.

After you receive a [transplanted kidney](#), you will need to take medicines to prevent your body from rejecting the kidney. The medicines you take will also help you stay healthy. You will need to have regular check-ups with your transplant team to make sure the kidney is working properly.

Remember you will still have CKD even after kidney transplant. [Living with a transplanted kidney](#) is similar to living with a transplanted kidney in a living donor. You will need to have regular check-ups with your transplant team to make sure the kidney is working properly.

Transplantation can have emotional and psychological effects on you. It is important to discuss these with your transplant team. You will need to have regular check-ups with your transplant team to make sure the kidney is working properly.



# Kidney transplantation

## What is kidney transplantation?

Kidney transplantation involves removing a kidney from a donor and transferring it to a patient with CKD. There can be two ways to do this: either as a living donor kidney transplant or as a deceased donor kidney transplant.

When possible, kidney transplantation is the best treatment for CKD. However, [you must have a compatible recipient](#). Kidney transplantation is not suitable for everyone and the [waiting list](#) for a kidney can be long.

In rare cases, patients with severe [hypertension](#) together with CKD may be considered for a combined heart and kidney transplant.

## Who donates the kidney?

The transplantation can be provided by either a deceased donor, but may well be done from a patient with your background.

**Living donor:** A kidney donation from a suitable living person will usually last and can be obtained in advance of an intended recipient.

**Deceased donor:** The donation is for patients on a waiting list for a kidney donation from someone who has recently died.

**How long does a kidney transplant last?**  
A kidney transplant usually lasts for 10-15 years, but some can last for 20 years or more.

## What does transplant surgery involve?

**Living donor transplant:** The donor's kidney is removed through a small incision in the back. The recipient's kidney is removed through a larger incision in the back.

**Deceased donor transplant:** The donor's kidney is removed through a larger incision in the back. The recipient's kidney is removed through a larger incision in the back.

**Combined heart and kidney transplant:** The donor's heart and kidney are removed through a larger incision in the back. The recipient's heart and kidney are removed through a larger incision in the back.

**How long does a kidney transplant last?**  
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## What happens after a kidney transplant?

After transplantation, you will need to take immunosuppressant medication to stop your body from rejecting the kidney. The dose will be adjusted over time. You will also need to take other medicines to prevent infection and to control blood pressure. You will need to have regular blood tests to check your kidney function and to monitor the levels of your immunosuppressants.

**Living donor transplant:** You will usually be able to go home the day after surgery. You will need to take immunosuppressant medication and other medicines to prevent infection and to control blood pressure. You will need to have regular blood tests to check your kidney function and to monitor the levels of your immunosuppressants.

**Deceased donor transplant:** You will usually be able to go home the day after surgery. You will need to take immunosuppressant medication and other medicines to prevent infection and to control blood pressure. You will need to have regular blood tests to check your kidney function and to monitor the levels of your immunosuppressants.

**Combined heart and kidney transplant:** You will usually be able to go home the day after surgery. You will need to take immunosuppressant medication and other medicines to prevent infection and to control blood pressure. You will need to have regular blood tests to check your kidney function and to monitor the levels of your immunosuppressants.

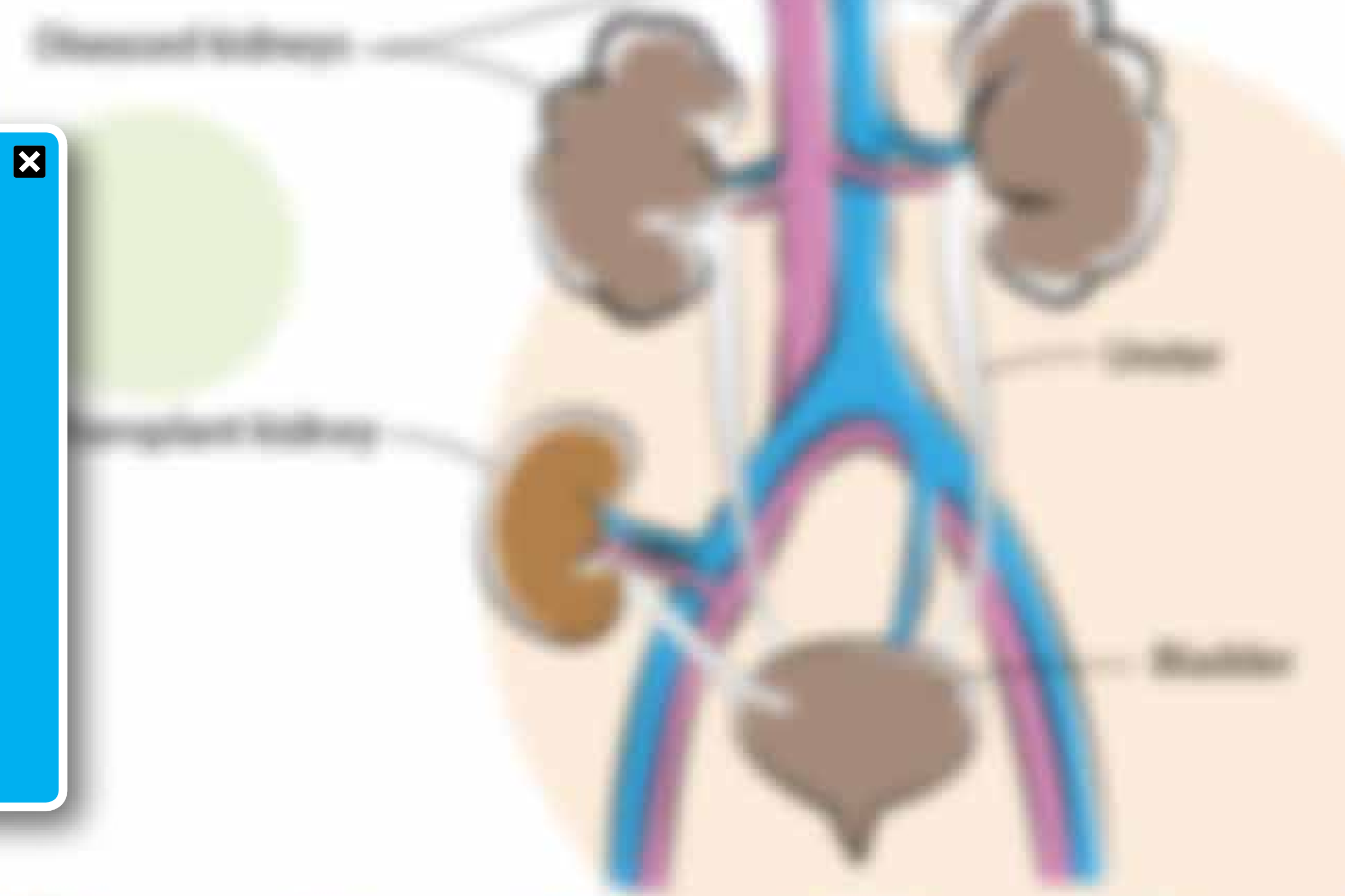
**How long does a kidney transplant last?**  
A kidney transplant usually lasts for 10-15 years, but some can last for 20 years or more.



**How long does a kidney transplant work for?**

Many factors affect how long a transplanted kidney will continue to work and so this varies between patients. A large national study of patients with ADPKD in France showed that 93% of transplants were working after 5 years, 87% after 10 years and 79% after 15 years.

Patients whose transplant does stop working properly can normally go back onto a transplant waiting list.



# Kidney transplantation

## What is kidney transplantation?

Kidney transplantation involves removing a kidney from a donor and transferring it to a recipient with CKD. There are two types of kidney transplant: living donor kidney transplant and deceased donor kidney transplant.

When possible, kidney transplantation is the best treatment for CKD. However, [not everyone is eligible for a kidney transplant](#). You should discuss with your doctor whether kidney transplantation is right for you and the [risks](#) of kidney transplant.

It can take a while to wait for a [living donor kidney](#) or a [deceased donor kidney](#). You may be able to wait for a kidney transplant while you are on dialysis.

## Why donate the kidney?

The most common reason for someone to donate a kidney is to help someone with CKD. You may also donate a kidney to help someone with other health problems.

**Living donor:** A living donor is someone who is healthy and gives one of their kidneys to someone with CKD. Living donor kidney transplants have a higher success rate than deceased donor kidney transplants.

**Deceased donor:** The deceased donor program is a program that allows people to donate their kidneys after they have died. Deceased donor kidney transplants have a lower success rate than living donor kidney transplants.

**How long does a kidney transplant last?**  
A kidney transplant can last for many years. However, it is important to take good care of your kidney transplant. You should discuss with your doctor how long your kidney transplant is likely to last.

## What does transplant involve?

**Living donor kidney transplant:** A living donor kidney transplant involves a living donor giving one of their kidneys to someone with CKD. The recipient will have their own kidney removed and the donor kidney will be placed in their body.

When receiving a transplant, it is important to take good care of your kidney transplant. You should discuss with your doctor how long your kidney transplant is likely to last. The recipient will have their own kidney removed and the donor kidney will be placed in their body.

Generally, transplantation is used as a way to help someone with CKD. It is used for people with CKD who are not responding to dialysis. It is used for people with CKD who are not responding to dialysis. You should discuss with your doctor how long your kidney transplant is likely to last.

## How long does a kidney transplant last?

## What happens after a kidney transplant?

After a kidney transplant, you will need to take immunosuppressant drugs to prevent your body from rejecting the kidney. You will also need to have regular check-ups with your doctor to monitor your kidney function and the side effects of the drugs. You should also take good care of your kidney transplant by taking good care of your health.

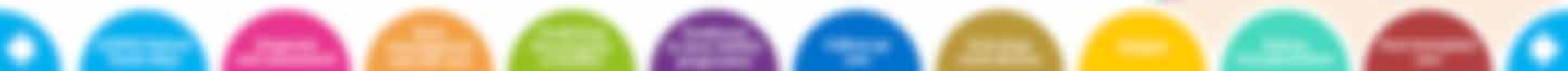
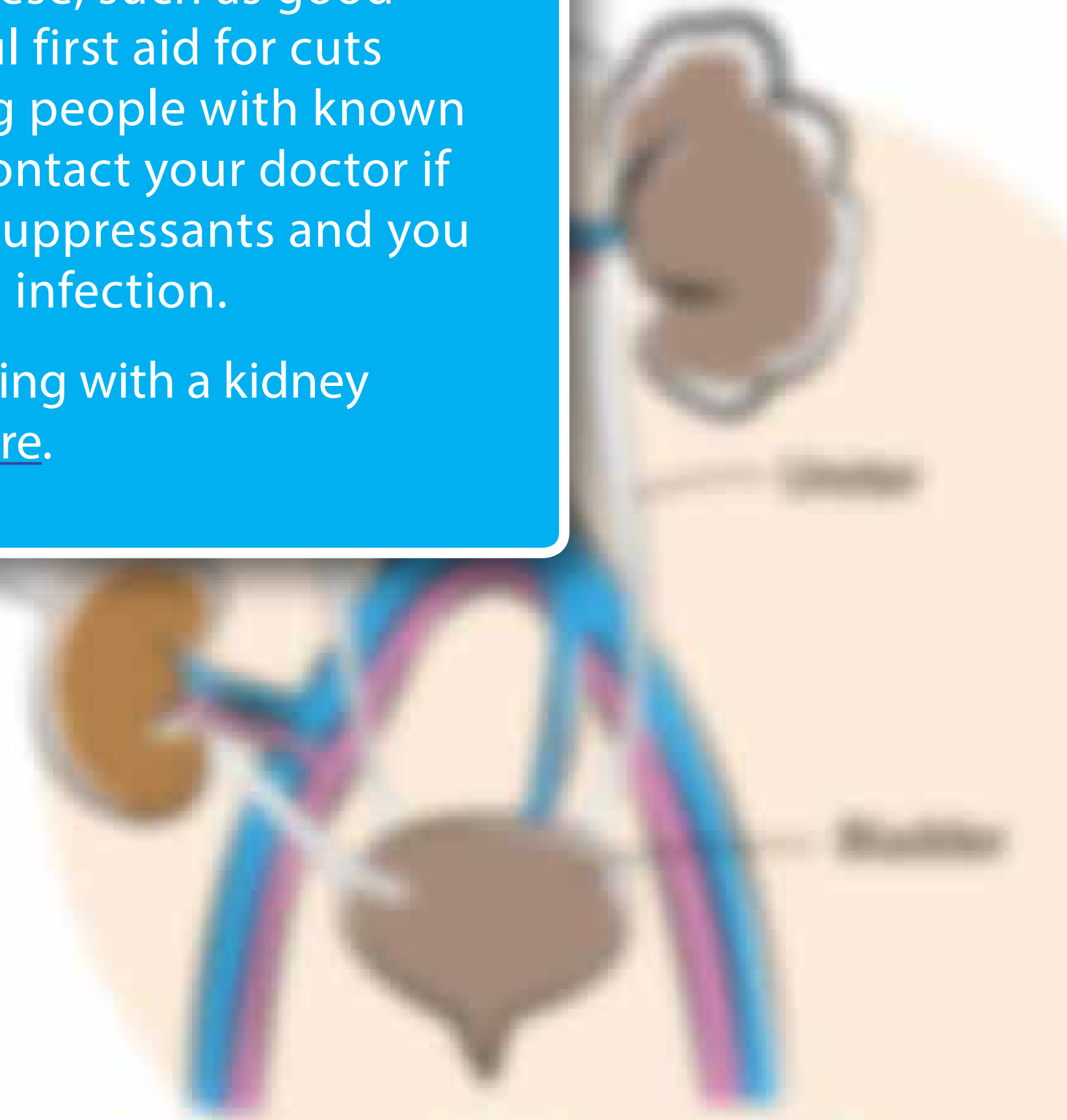


### Follow-up care

After a kidney transplant, long-term follow-up care is essential, for the following reasons:

- To check that the transplanted kidney is working and has not been rejected by the body. In the longer term, it is important to make sure that the kidney continues to work properly.
- To monitor immunosuppressant therapy and manage any side effects (depending on the specific drugs used); you should discuss these with your doctor.
- Importantly, by suppressing the immune system immunosuppressants leave you susceptible to infections. Simple precautions can be taken to avoid these, such as good personal hygiene, careful first aid for cuts and grazes, and avoiding people with known infections. You should contact your doctor if you are taking immunosuppressants and you think you might have an infection.

More information about living with a kidney transplant can be found [here](#).



# Kidney transplantation

## What is kidney transplantation?

Kidney transplantation involves removing a kidney from a donor and transferring it to a person with CKD. There can be two ways of getting an organ from a donor kidney: either it is transplanted...

There are two ways of getting a kidney transplant: either you get a kidney from a living donor or you get a kidney from a deceased donor. The living donor can be...

It is important to understand that you will need to take immunosuppressant medication for the rest of your life to prevent your immune system from attacking the new kidney.

## Why donate the kidney?

The most common reason for someone to donate a kidney is to help someone with CKD. There are many reasons why someone might want to donate a kidney...

**Living donor:** A living donor is someone who has a healthy kidney and donates one of their kidneys to someone with CKD.

**Deceased donor:** A deceased donor is someone who has died and their organs are donated to someone with CKD.

There are many reasons why someone might want to donate a kidney. Some people donate to help someone they know, while others donate to help someone they don't know.

## What does transplant involve?

**Living donor transplant:** A living donor transplant involves a living donor donating one of their kidneys to someone with CKD.

**Deceased donor transplant:** A deceased donor transplant involves a deceased donor's kidney being donated to someone with CKD.

The transplant team will discuss the risks and benefits of the transplant with you before you decide whether to have the transplant.

Generally, transplantation is a good option for people with CKD, as well as for people with other health problems. It can help you live longer and feel better.

There are many things you should think about before you decide whether to have a kidney transplant.

## Post-transplant care - what happens afterwards?

After a kidney transplant, you will need to take immunosuppressant medication for the rest of your life. You will also need to have regular check-ups with your healthcare team to monitor your kidney function and overall health.

It is important to understand that you will need to take immunosuppressant medication for the rest of your life to prevent your immune system from attacking the new kidney.

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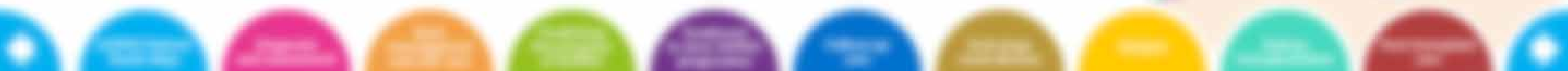
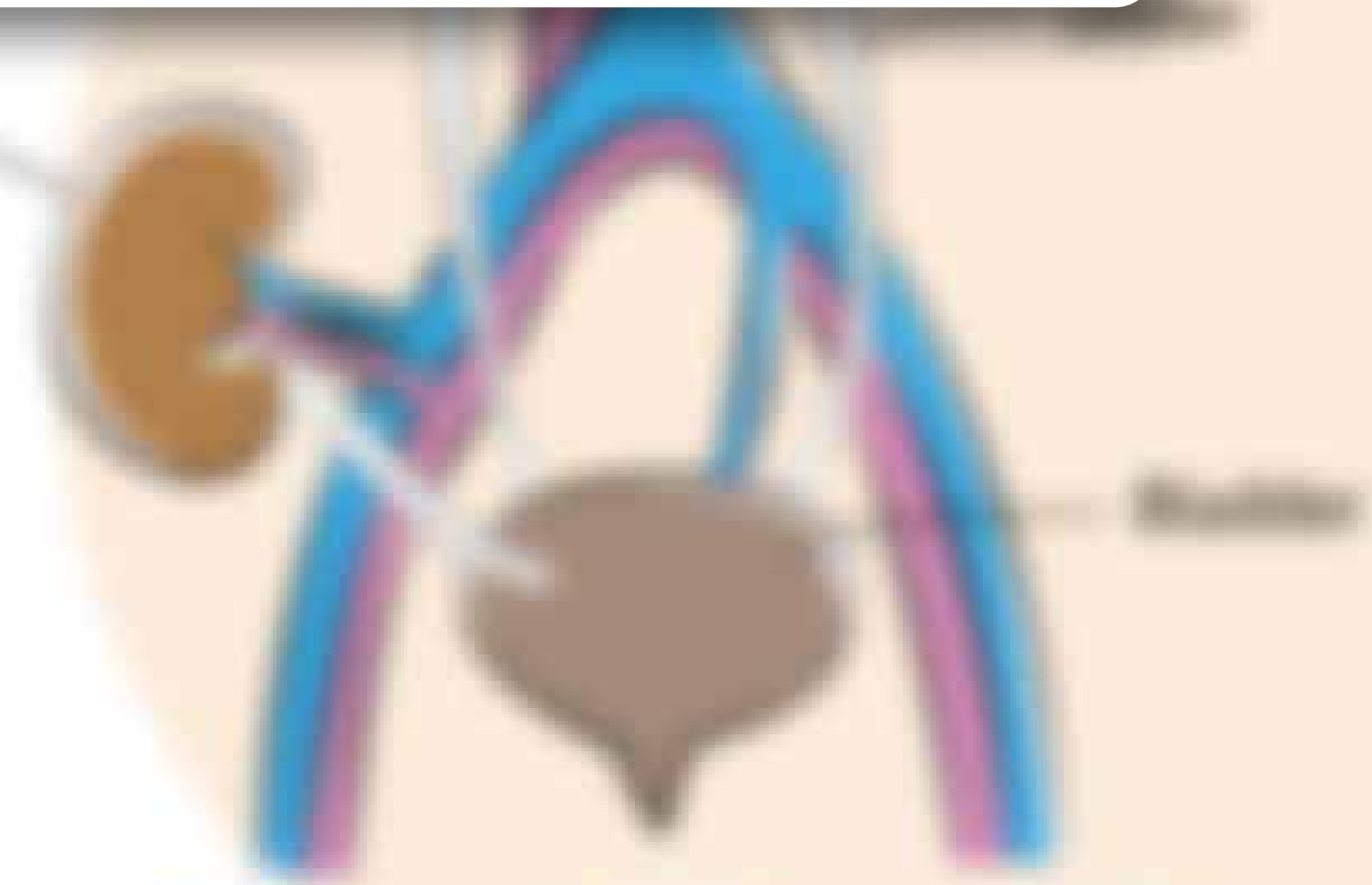
There are many things you should think about before you decide whether to have a kidney transplant.

The transplant team will discuss the risks and benefits of the transplant with you before you decide whether to have the transplant.

**Immunosuppressant medication**

People who receive a kidney transplant usually need to take immunosuppressant medications for the rest of their life to prevent their immune system from attacking the new kidney. Commonly used immunosuppressants include tacrolimus, ciclosporin, azathioprine, mycophenolate, prednisolone and sirolimus.

Possible side effects of immunosuppressants include: diabetes, high blood pressure, weakening of the bones (osteopenia), changes in the cholesterol levels in the blood, and kidney problems. Importantly, these medicines can make patients vulnerable to infections. This means that precautions are necessary to reduce the risk of infections (such as vaccinations and good hygiene), and patients need to contact their healthcare team if they think they might have an infection. Symptoms of infection can include high temperature (fever 38°C/100.4°F or above), headache, aching muscles, diarrhoea and vomiting.



This section explains the type of long-term follow-up care required by people with ADPKD.

If you have ADPKD you will need lifelong follow-up care involving a multidisciplinary care team, depending on your individual circumstances. How this care is co-ordinated depends on the local organisation of healthcare.

## Nephrology care

Typically, if you have ADPKD you will remain under the care of a nephrologist. How often you see your nephrologist depends on many **factors** >. These include your kidney function, your symptoms and complications, the type of treatment you receive and how quickly your disease is expected to progress.

## Other specialists

The nephrologist will refer patients to other kinds of [specialist doctors](#) and [healthcare staff](#) if necessary, for example if they experience [complications](#) in other parts of the body.

Family doctors (also called primary care doctors or general practitioners) play an important role in providing and co-ordinating care for other diseases and aspects of health, as well as ADPKD.

Remember that kidney and PKD [patient organisations](#) can be an important source of advice, support and information on these topics.

## Transition care for adolescents

Adolescents with ADPKD face a transition from paediatric healthcare services to adult services.

Ideally, there should be a defined and co-ordinated pathway to transition for adolescents from paediatric to adult services, to help maintain continuity of care.



## What can patients do?

There are many ways that you can play a vital role in contributing to your own care. These include:



# Follow-up care

This section explains the type of long-term follow-up care required by people with ADPKD.

How often ADPKD care will need to be followed up will vary depending on your individual circumstances. This will also be influenced by the rate of progression of the disease.

## Key messages

### Factors

These include the level of kidney function, your symptoms and complications, and how quickly your disease is expected to progress. Clinic visits will also be needed more often for patients who start specific treatment to slow ADPKD progression.



Remember that having ADPKD [doesn't guarantee](#) you'll be able to support yourself with the help of family and friends.

### Transition care for adolescents

Adolescents with ADPKD may transition from paediatric health care services to adult services.

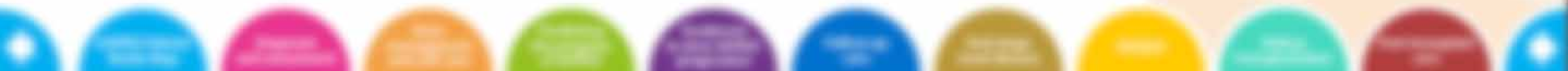
Health care providers will discuss with adolescents whether to transfer the adolescents from paediatric to adult services to help ensure continuity of care.

Health care providers will discuss with adolescents in general, and adolescents with ADPKD in particular, the importance of providing and understanding care for other chronic conditions as well as ADPKD.

## What can patients do?

There are many ways that you can play a role in managing your condition. These include:

-  **Get active**
-  **Stay hydrated**
-  **Manage stress**
-  **Monitoring and managing blood pressure**
-  **Monitoring and managing blood sugar**





# Follow-up care

This section explains the type of long-term follow-up care required by people with ADPKD.

If you have ADPKD you will need long-term follow-up care involving a number of different care teams. Depending on your individual circumstances, this may also include decisions on the best options for treatment.

## Regular care

Typically if you have ADPKD you will need regular care from a number of different care teams. This often includes your general practitioner (GP) and a number of other specialists. These include your kidney specialist, your dietitian and other specialists. The type of treatment you receive will also vary over time as you progress.

## Other specialists

The specialists will also address any other health issues you may have. For example, you may need to see a specialist for high blood pressure or other health issues.

Remember that you can also get support and information on these topics from patient organisations.

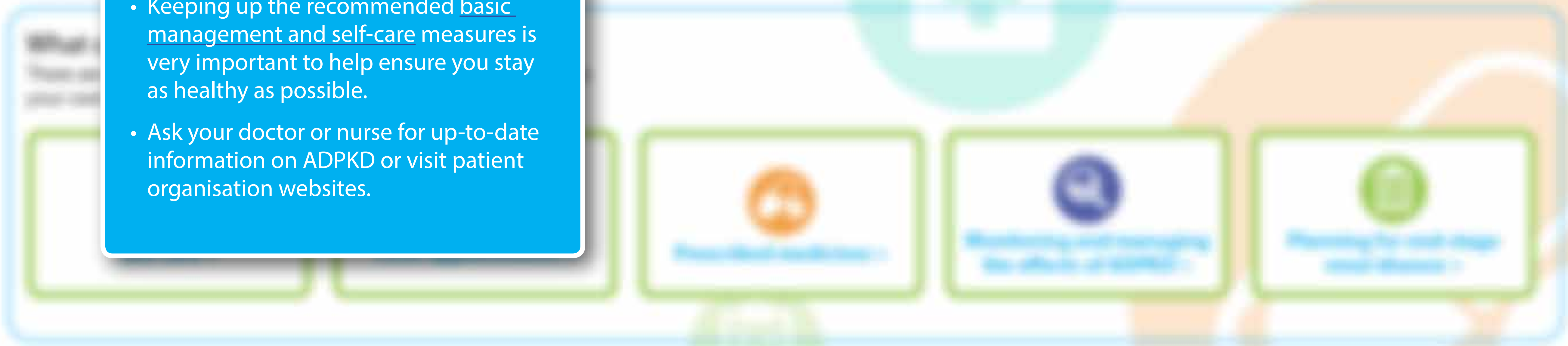
## Transition care for adolescents

Adolescents with ADPKD may experience their general health care services to adult services.

Read this document to understand how to transition from your current care to adult services to help ensure continuity of care.

**Self-care** ✕

- Keeping up the recommended basic management and self-care measures is very important to help ensure you stay as healthy as possible.
- Ask your doctor or nurse for up-to-date information on ADPKD or visit patient organisation websites.



This section explains the type of long-term follow-up care required by people with ADPKD.

For more information on all the things you can do to manage your ADPKD, see our [ADPKD guide](#). This guide is a collection of information on all the things you can do to manage your ADPKD.

## Regular care

People with ADPKD need regular care from their healthcare team. This includes regular check-ups with their doctor or specialist. You should also have regular blood tests to check your kidney function.

## Other specialists

You may need to see other specialists, such as a dietitian, a physiotherapist, or a counsellor. These specialists can help you manage your ADPKD and improve your quality of life.

## What can patients do?

There are many things you can do to help manage your ADPKD. These include:



## Clinic appointments

- Try to keep all appointments or reschedule in advance, as necessary.
- It will help your healthcare team if you give them as much information as possible about your health. For example, you should tell them about any changes to your health that have occurred, any specific symptoms or pain, any new medicines you are taking (prescribed or bought over the counter, including complementary therapies), and any side effects of treatment you have experienced.
- It might help to write down things you wish to tell the team in advance and take these notes to the appointment – perhaps using the checklists in this Route Map.
- Feel free to explain any effects of ADPKD on your wellbeing, personal and family life.
- Always ask questions if you are unsure about anything. It may help to prepare questions in advance. For example, three important questions you might like to ask are:
  - What are my options?
  - What are the possible benefits and risks of those options?
  - What help do I need to make my decision?

Some patient organisations provide help with asking questions.

# Follow-up care

This section explains the type of long-term follow-up care required by people with ADPKD.

If you have ADPKD you will need long-term follow-up care involving specialists who can help. Depending on your individual circumstances, this may be in your local hospital or the local region of healthcare.

## Regular care

Normally if you have ADPKD you will receive care from your GP. However, there are other specialists who can help. These include your kidney specialist, your dietitian and your pharmacist. The type of treatment you receive will depend on your disease and your progress.

## Other specialists

The specialists will also address any other health issues you have. For example, if you experience **hypertension** in other parts of the body.

Some doctors also offer services such as genetic counselling or counselling on providing information on the other disease and signs of health, as well as ADPKD.

Some people may also need **specialist care** for an important aspect of their support and information on their health.

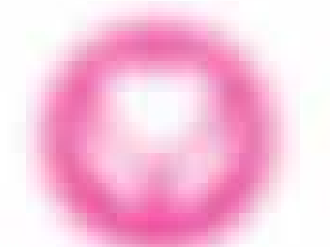
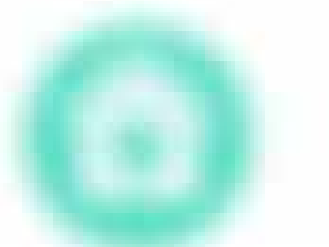
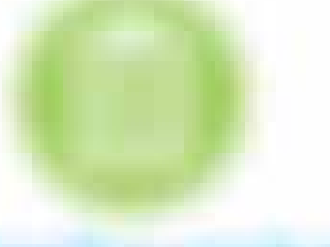
## Specialist care for adolescents

Adolescents with ADPKD may experience their parents health care services in adult services.

Some people may also need **specialist care** for an important aspect of their support and information on their health.

## What can patients do?

There are many ways that you can help in your long-term care. These include:

-  **Take all prescribed medicines**
-  **Take all prescribed medicines**
-  **Take all prescribed medicines**

**Prescribed medicines** ✕

- Take all prescribed medicines according to the instructions given. Make sure you understand their possible side effects, any actions you can take to reduce the risk of these, and what to do if you experience side effects. You can ask your healthcare team about these issues.
- Tell your healthcare team about any changes to your health or the medicines you are taking. Contact your doctor, nurse or pharmacist if you have any questions or concerns about your medicines.

This section explains the type of long-term follow-up care required by people with ADPKD.

For more ADPKD care information, please visit our website [www.kidney.org](#) or call our helpline on 0800 368 8600. You can also visit our website on the web page of the National Kidney Foundation.

## Regular care

People with ADPKD will need regular care to monitor the size of their kidneys, how well they are working, and to manage any complications. The type of treatment you receive will depend on your stage of disease and symptoms.

## Other specialists

The nephrologist will refer you to other kinds of [healthcare professionals](#) to manage other symptoms or complications in other parts of the body.

Some people also need to see a dietitian to get advice on what to eat. You may also need to see a psychologist or counsellor to help you deal with the stress of ADPKD.

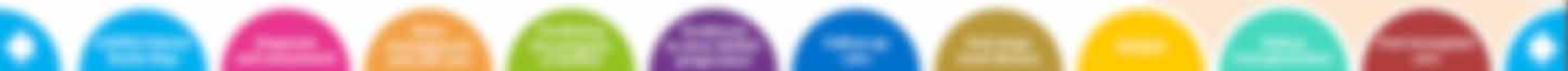
## What can patients do?

There are many ways that you can help to manage your ADPKD. These include:



## Monitoring and managing the effects of ADPKD

- It will help your healthcare team if you keep a record of your medicines, symptoms and general health.
- It can help to understand what stage of chronic kidney disease you have and what symptoms could indicate that this is getting worse. Patients in some countries have access to their own test results to help them monitor and manage their disease. You may wish to ask your doctor about these aspects.
- Make sure you understand the possible complications of ADPKD, what symptoms these can cause, what action to take and when.
- Agree with your healthcare team how you can best manage pain at home, and when you should contact the team.
- It can help to understand the roles and responsibilities of the various healthcare professionals involved in your care. Also, make sure everyone who treats you knows that you have ADPKD.
- Ask your healthcare team about the services available to help you deal with the impact of ADPKD on wellbeing, personal and family life.
- You can contact your healthcare team if you have any questions with respect to family planning and aspects such as the screening of family members for ADPKD.



# Follow-up care

This section explains the type of long-term follow-up care required by people with ADPKD.

If you have ADPKD you will need lifelong follow-up care involving a multidisciplinary care team. Depending on your individual circumstances, this care can be tailored to meet the needs of your specific situation.

## Nephrology care

Typically if you have ADPKD you will require regular visits with a nephrologist. How often you see your nephrologist depends on many factors. These include your kidney function, your symptoms and complications, the type of treatment you receive and how quickly your disease is expected to progress.

## Other specialists

Your nephrologist will refer you to other kinds of specialists if necessary, for example if they experience complications in other parts of the body.

Some doctors also offer primary care services or general practitioners may be responsible for providing and coordinating care for other chronic and acute conditions, as well as ADPKD.

Remember that having ADPKD [doesn't guarantee](#) you'll be an expert source of advice, support and information on these topics.

## Transition care for adolescents

Adolescents with ADPKD may experience their parents' health care services to adult services.

Read this document to understand the transition pathway to transfer the adolescents from parents to adult services to help ensure continuity of care.

## What can patients do?

There are many ways that you can play a role in controlling your own care. These include:

- 1. **Self-care**
- 2. **Take responsibility**
- 3. **Partner with your doctor**
- 4. **Plan your care and manage any other conditions**
- 5. **Plan for your future and share it**

**Planning for end-stage renal disease**

- If your ADPKD eventually progresses towards end-stage renal disease, it will be important to discuss with your nephrologist the available options for treatment, and to agree a plan.

This section explains the types of ADPKD research in which patients may be able to participate.

Research is underway to help improve the scientific understanding of ADPKD and to improve care for patients. You may be able to participate in this research if you wish, depending on your circumstances and where you live.

Patients interested in participating in research should discuss this with their healthcare team, or contact their nearest kidney and ADPKD [patient organisation](#) or other specific research groups below.

## Registries

Patient registries are databases that collect information about patients with specific diseases. This allows researchers to study various aspects of the disease, such as how it affects people, how it progresses over time, and how effective treatments are in practice. Registries are a valuable source of information, especially for rare or uncommon diseases such as ADPKD.

ADPKD registries exist in several [European countries](#) >. There is also an international registry for children with ADPKD, called [ADPedKD](#).

If you would like to join an ADPKD registry you should discuss this with your nephrologist. If you join, you will be asked to sign a consent form for the use of your information to be included in the registry.

## Clinical trials

Clinical trials are research studies that test the efficacy (i.e. the ability to produce a desired or intended result) and safety of medicines or other types of treatment. Some clinical trials involve healthy volunteers, while others involve patients with specific diseases such as ADPKD.

There are several types of clinical trial. If you are interested in participating in this type of research, ask your nephrologist or contact one of the nephrologists on the list shown.

You can also find out more about clinical trials at the [EU Clinical Trials Register](#), [ClinicalTrials.gov](#) and the [PKD Foundation](#).

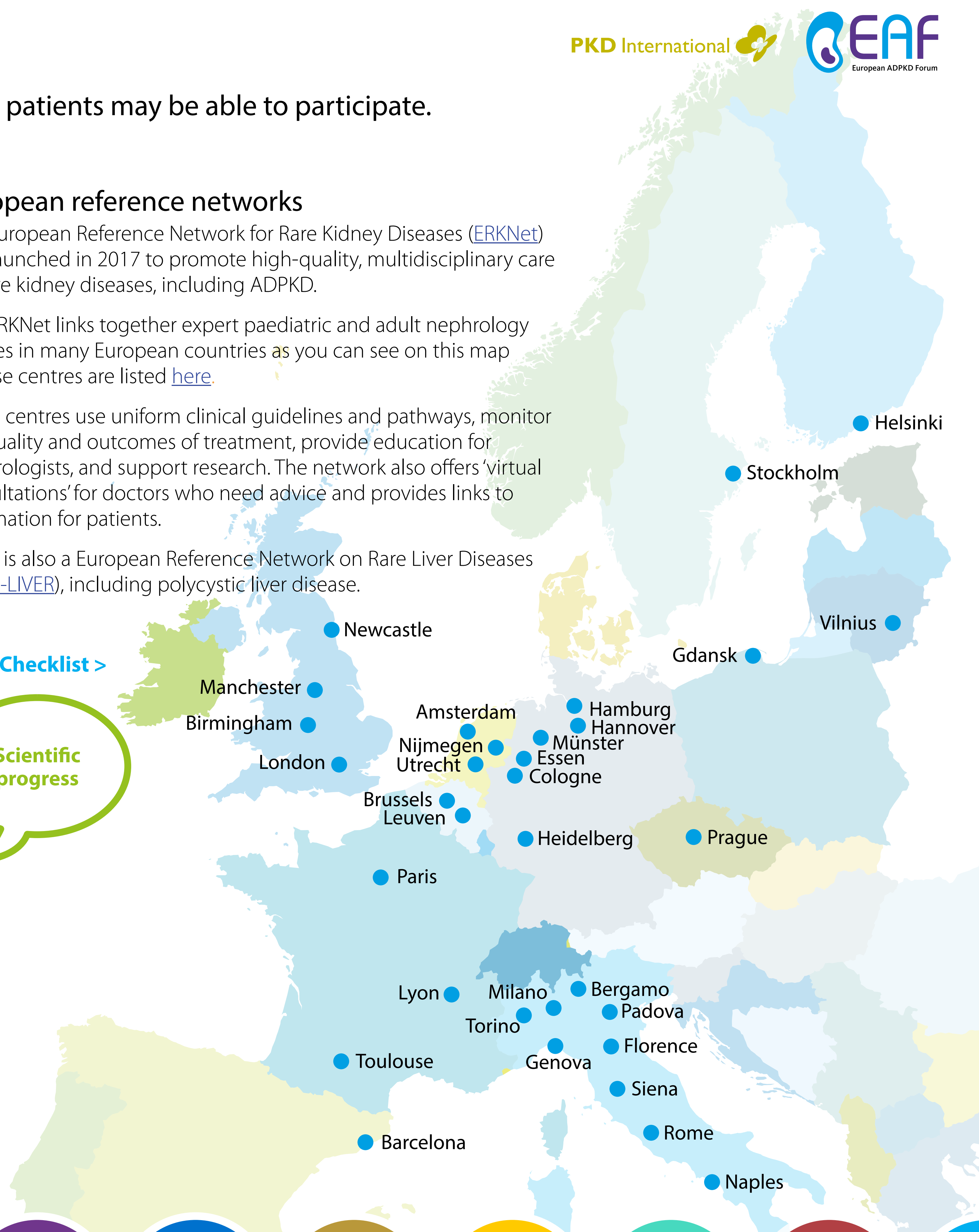
## European reference networks

The European Reference Network for Rare Kidney Diseases ([ERKNet](#)) was launched in 2017 to promote high-quality, multidisciplinary care for rare kidney diseases, including ADPKD.

The ERKNet links together expert paediatric and adult nephrology centres in many European countries as you can see on this map – these centres are listed [here](#).

These centres use uniform clinical guidelines and pathways, monitor the quality and outcomes of treatment, provide education for nephrologists, and support research. The network also offers 'virtual consultations' for doctors who need advice and provides links to information for patients.

There is also a European Reference Network on Rare Liver Diseases ([RARE-LIVER](#)), including polycystic liver disease.



## Research

This section explains the types of (CKPD) research in which patients may be able to participate.

Research is undertaken to help improve the overall understanding of European reference networks

Research is undertaken to help improve the overall understanding of CKPD and to improve care for patients. You may be able to take part in this research if you wish. Depending on your circumstances, you may be able to:

participate in research that is aimed at understanding the condition better  
participate in research that is aimed at understanding the condition better  
participate in research that is aimed at understanding the condition better

### Registers

Registers are databases that collect information about people with specific diseases. The information collected is used to help researchers understand the disease better, to help them develop new treatments and to help them provide better care for people with the disease. Registers are often used to help researchers understand the disease better, to help them develop new treatments and to help them provide better care for people with the disease.

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### Clinical trials

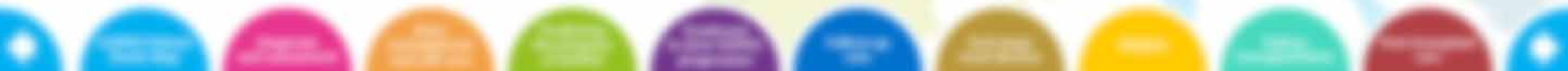
Clinical trials are research studies that test the effectiveness of a new treatment or to compare one treatment with another. Clinical trials are often used to help researchers understand the disease better, to help them develop new treatments and to help them provide better care for people with the disease.

Clinical trials are often used to help researchers understand the disease better, to help them develop new treatments and to help them provide better care for people with the disease.

### Scientific progress

*'I hope that scientific progress can improve both life expectancy and the course of treatment of all patients with polycystic kidney disease.'*

*Teresa, Italy*



This section explains the types of ADPKD research in which patients may be able to participate.

Research is undertaken to help improve the overall understanding of ADPKD and to improve care for patients. You may be able to participate in this research if you wish. Depending on your circumstances and preferences:

Patients interested in participating in research should discuss this with their healthcare team, or contact their nearest ADPKD [ADPKD Association](#) or other specific research group below.

## Registries

Registries register and monitor data on other information about patients with specific diseases. The data registered in these registries helps doctors, scientists and researchers to learn more about the condition, how it affects people, and what causes it. Some registries also collect samples of information about patients with a disease.

ADPKD registries can be used to:

Plan and deliver the best care for ADPKD with your healthcare team, and to help for the use of your information.

## Clinical trials

Clinical trials are research studies that test the effectiveness of the ability to produce or deliver a particular result and safety of medicines or other types of treatment. Some clinical trials involve healthy volunteers, while others involve patients with specific diseases such as ADPKD.

There are several types of clinical trial. You can sometimes participate in this type of research, and your healthcare team can advise you of the opportunities in the local area.

## European reference networks

The European Reference Network for Rare Kidney Diseases ([ERN RD](#)) was established in 2017 to promote high quality, multidisciplinary care for rare kidney diseases, including ADPKD.

The ERN RD also organises expert panels, and other networking events in many European countries. You can see on this map - these events are listed [here](#).

These centres are within ERN RD guidelines and networks, provide the quality and expertise of researchers, provide education for healthcare professionals, and research support. The network also offers virtual consultations for doctors who may have limited experience with ADPKD.

**European countries** ✕

Countries in which ADPKD registries exist include Austria, Denmark, Finland, France, Germany, Greece, Italy, Netherlands, Romania, Spain, Sweden, Turkey and the United Kingdom.







# Information for health policymakers and providers

This section provides recommendations to authorities responsible for healthcare policy, planning and provision at national and local levels.

The ADPKD Patient Route Map explains the main elements of good practice in ADPKD care to help patients, families, healthcare providers and policymakers work together to ensure everyone receives the care and support they need, at the right time. It can help healthcare providers and policymakers to design, adapt or assess co-ordinated services to efficiently address unmet needs among people affected by ADPKD, in the context of local conditions.

## Unmet needs in ADPKD

ADPKD is a chronic, progressive, inherited disease that causes cysts to develop in the kidneys and which can affect many other parts of the [body](#). Unmet needs in ADPKD include a lack of awareness among many health stakeholders and a lack of co-ordinated care pathways, resulting in significant variations in care. Individuals with ADPKD should have access to co-ordinated, [patient-centred, multi-specialist care](#), as outlined in this Route Map and [elsewhere](#).

## Patient-centred care

All stakeholders, including national governments and healthcare providers, should support efforts to better inform patients and families and to empower them to act as fully informed and active partners in care. More about [patient-centred care](#) >.

## Multidisciplinary care

Patients should have access to a nephrologist with expertise in ADPKD. Collaboration should be encouraged between the various specialists involved in ADPKD care to design and implement [co-ordinated services](#). More about [multidisciplinary care](#) >.

## Technology uptake

Advances in [imaging](#) >, [genetic testing](#) > and [communications and information technology](#) > could help to improve ADPKD care.

## Transplantation

ADPKD is responsible for around one in 10 of all patients needing [dialysis or transplantation](#). Kidney transplantation is the optimal treatment for kidney failure, providing excellent outcomes and being far more cost-effective than dialysis. Collaborative efforts are needed to improve access to transplantation in line with EU initiatives.

## Conclusion

Collaboration between health policymakers, providers, professionals and patients is encouraged to design and implement co-ordinated ADPKD services and to promote awareness building, education and research.

Patients and families who wish to support or participate in ADPKD advocacy should contact the kidney or [ADPKD patient organisation](#) in their country, or [PKD International](#).



This section provides recommendations to authorities responsible for healthcare policy, planning and provision at national and local levels.

The ADPKD system from the system for care delivery is a good example of ADPKD care in the system. It is a good example of a system that is designed to be patient-centred and to be able to support the needs of the system. It is a good example of a system that is designed to be patient-centred and to be able to support the needs of the system.

## Technology update

Access to [healthcare services](#) and [healthcare services](#) is essential to improve ADPKD care.

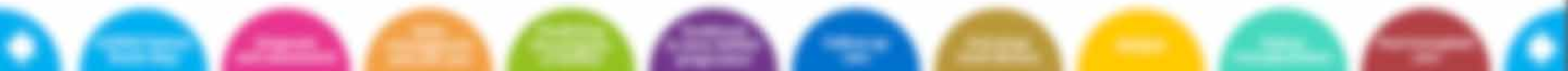
## Implementation

ADPKD is a complex condition that requires a multidisciplinary approach. It is a good example of a system that is designed to be patient-centred and to be able to support the needs of the system.

### Patient-centred care

All stakeholders, including national governments and healthcare providers, should support efforts to better inform patients and families and to empower them to act as fully informed and active partners in care. Patients should be supported to maintain self-care measures, to deal with the impact of the condition, and to participate in shared decision-making regarding healthcare policies, services and research.

This requires patients and carers to have access to accurate, up-to-date information about ADPKD, their own clinical data, and opportunities to participate in making decisions. Patients and carers should also have a clear understanding of the available services and how these can be navigated optimally. This Route Map is provided as a tool to help achieve this.



This section provides recommendations to authorities responsible for healthcare policy, planning and provision at national and local levels.

The following recommendations are based on the evidence from the systematic review and the expert panel. They are intended to provide a high level of guidance to inform policy and practice.

### Technology update

Review [technology update](#) and [technology update](#) to improve ADPKD care.

## Multidisciplinary care ✕

Patients should have access to a nephrologist with expertise in ADPKD. Collaboration should be encouraged between the various specialists involved in ADPKD care to design and implement co-ordinated services.

Where possible, a team approach with all specialties provided in one centre or clinic would be expected to benefit research, expert and patient networking, efficiency and patient outcomes. Where this is not possible, managed co-ordination and networking of local or national specialist services is important to optimise patient care. Managed co-ordination would be expected to facilitate prompt, accurate diagnosis, avoidance of duplication of tests, better management of disease complications and manifestations, evidence-based access to treatment to slow disease progression and ultimately to improve patient outcomes.

We encourage policymakers and providers to support the European Reference Network on Kidney Diseases (ERKnet) and European Reference Network on Rare Hepatological Diseases (ERN RARE-LIVER). These networks will facilitate the sharing of knowledge, experience, medical research, teaching, training and resources.



# Information for health policymakers and providers

This section provides recommendations to authorities responsible for healthcare policy, planning and provision at national and local levels.

The ADPKD system from the system for care elements of your practice or ADPKD care in the system, besides, healthcare providers and patients work together to ensure resources across the care system support the needs of the right time. It is an important healthcare provider and patient to design, adapt or even to deliver services to efficiently address their needs, ensuring people effectively ADPKD in the context of local conditions.

## Global needs in ADPKD

ADPKD is a chronic, progressive inherited disease that causes cysts to develop in the kidneys and which can affect many other parts of the body. Common needs in ADPKD include a lack of awareness among many health professionals and a lack of coordinated care pathways, including a significant number of care individuals with ADPKD. Global health systems in a number of countries are still struggling to address [ADPKD patient needs](#) and to address the needs of the right time.

## Remote medical care

In countries, including national governments and healthcare providers, should support efforts to better inform patients and health professionals about their condition and to help them to access care when they need [remote medical care](#).

## Multidisciplinary care

There is a need for multidisciplinary care for ADPKD patients, including nephrologists, radiologists, geneticists, dietitians, and other healthcare professionals.

## Technology update

Research [imaging](#), [genetic testing](#) and [remote medical care](#) [technology](#) to improve ADPKD care.

## Transplantation

ADPKD is responsible for around 10% of all patients needing [kidney transplantation](#). Kidney transplantation is the optimal treatment for end-stage kidney disease, providing excellent outcomes and quality of life compared to dialysis. Transplantation offers an excellent option to improve quality of life and health-related quality of life for patients.

## Conclusion

Collaboration between health policymakers, providers, professional associations and managers to design and implement coordinated ADPKD care across primary, secondary, tertiary and quaternary care.

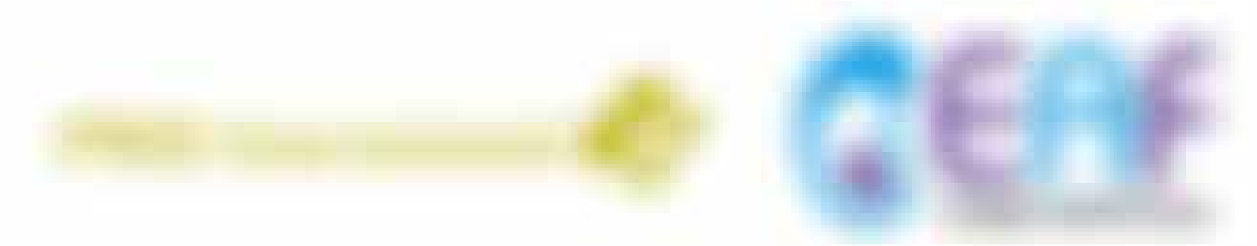
**Health professionals who wish to support a patient with ADPKD should consider the value of [ADPKD patient education](#) in their country or [transplantation](#).**

**Imaging** ✕

New automated imaging methods can reduce the costs and labour needed to measure total kidney volume (the most important marker of ADPKD progression). Increased uptake of these techniques would greatly benefit care.



# Information for health policymakers and providers



This section provides recommendations to authorities responsible for healthcare policy, planning and provision at national and local levels.

The ADPKD system from the system for care delivery at your service or ADPKD care in the system, for example, healthcare providers and patients work together to ensure resources across the system support the care of the right people in the right medical setting and at the right time. It is an essential part of the system to ensure that the right people are in the right setting to receive the right care at the right time.

## Current needs in ADPKD

ADPKD is a chronic, progressive condition that causes cysts to develop in the kidneys and other organs. Other parts of the body, such as the liver, pancreas and spleen, may also be affected. The condition is most commonly inherited, but can also be caused by a mutation in the PKD1 or PKD2 genes. The condition is most commonly diagnosed in young adults, but can also be diagnosed in children. The condition is most commonly diagnosed in young adults, but can also be diagnosed in children. The condition is most commonly diagnosed in young adults, but can also be diagnosed in children.

## Genetic testing

Access to genetic testing varies across Europe with key barriers including the cost of tests, resourcing of services, diverse reimbursement policies, and a lack of clear, reliable information in some countries. The EAF and PKD International believe that genetic testing should be available to patients for whom it is clinically indicated, and that all patients should have access to pre-implantation genetic diagnosis (PGD).

The uptake of faster and cheaper genetic tests could herald a greater role for genetic testing in the diagnosis of ADPKD and in predicting the disease prognosis.

## Technology update

Access to [genetic testing](#) and [pre-implantation genetic diagnosis \(PGD\)](#) is essential to improve ADPKD care.

## Transplantation

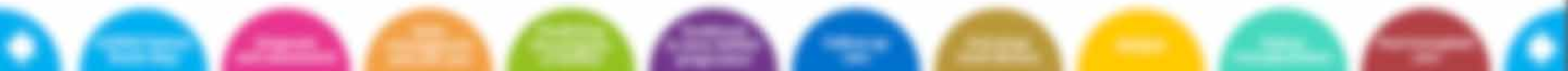
ADPKD is associated for several reasons with a higher need for [transplantation](#). Early transplantation is the optimal treatment for patients with end-stage renal disease, and early transplantation is associated with a higher survival rate than late transplantation. Transplantation offers an excellent option for patients with end-stage renal disease.

## Conclusion

Transplantation remains the best option for patients with end-stage renal disease. Early transplantation is the optimal treatment for patients with end-stage renal disease, and early transplantation is associated with a higher survival rate than late transplantation. ADPKD is associated for several reasons with a higher need for transplantation.



Access to genetic testing and pre-implantation genetic diagnosis (PGD) is essential to improve ADPKD care.



# Information for health policymakers and providers

This section provides recommendations to authorities responsible for healthcare policy, planning and provision at national and local levels.

The WHO notes that the system for care delivery is just one of the key components of a health system. Health systems are complex and multi-faceted, and together they determine the health outcomes that a country achieves. It is important that health systems and providers are designed, shaped and supported in ways that are consistent with the overall health system strategy and the health system's goals.

## Global needs in NCDs

NCDs are a major, progressive, preventable cause of death and disability in the world and account for almost 70% of all deaths. The WHO notes that NCDs include a wide range of diseases, including heart disease, cancer, chronic respiratory diseases, diabetes, and stroke. NCDs are preventable and many of the risk factors for NCDs, such as tobacco use, alcohol consumption, and poor diet, are modifiable.

## Primary prevention

Primary prevention, including national governments and health systems, should support efforts to reduce tobacco, alcohol, and unhealthy diet consumption. This is an essential and effective approach to reduce the burden of NCDs.

## Multisectoral approach

Health systems should take a multisectoral approach to address the social, economic, and environmental determinants of NCDs.

## Technology update

Investment in digital health and information technology is essential to improve NCD care.

## Implementation

NCDs require a coordinated, multi-sectoral approach to address the underlying causes of the disease. This includes strengthening health systems, promoting healthy behaviors, and addressing social and economic determinants of health.

## Conclusion

Addressing NCDs requires a multisectoral, multiscale approach that involves national governments, health systems, and communities. This approach is essential to reduce the burden of NCDs and improve health outcomes.

WHO provides guidance on the implementation of NCD prevention and control strategies. This includes information on the role of health systems, communities, and individuals in reducing the burden of NCDs.

**Communications and information technology** ✕

New telecommunication and information technologies can facilitate multi-specialist networking, avoiding patients having to travel to access expert care that does not exist in their country. This technology can also promote patient empowerment and self-care.



## Europe

Belgium	<a href="#">Association pour l'Information et la Recherche sur les maladies Rénales Génétiques (AIRG) Belgique</a>
Finland	<a href="#">Munuais- ja maksaliitto (Finnish Kidney and Liver Organization)</a>
France	<a href="#">Association Polykystose France (APKF)</a> <a href="#">Association pour l'Information et la Recherche sur les maladies Rénales Génétiques (AIRG) France</a>
Germany	<a href="#">PKD Familiäre Zystennieren e.V.</a>
Ireland	<a href="#">Irish Kidney Association</a>
Italy	<a href="#">Associazione Italiana Rene Policistico (AIRP)</a>
Netherlands	<a href="#">Nierpatienten Vereniging Nederland (NVN)</a>
Spain	<a href="#">Asociación para la Información y la Investigación de las Enfermedades Renales Genéticas (AIRG) España</a> <a href="#">Federación Nacional de asociaciones para la lucha contra las enfermedades del riñón (ALCER)</a>
Switzerland	<a href="#">SwissPKD</a> <a href="#">Association pour l'Information et la Recherche sur les maladies Rénales Génétiques (AIRG) Suisse</a>
Turkey	<a href="#">Turkish Society of Nephrology Cystic Kidney Diseases Working Group</a>
UK	<a href="#">PKD Charity</a> <a href="#">Genetics Alliance UK</a>

## North America

Canada	<a href="#">PKD Foundation of Canada</a>
USA	<a href="#">PKD Foundation</a>

## Asia

Japan	<a href="#">PKD Foundation</a>
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## Australasia

Australia	<a href="#">PKD Foundation Australia</a>
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## International

[Federation of European associations of patients affected by Renal Genetic diseases \(FEDERG\)](#)

[PKD International](#)



## Many of these articles are freely available online.

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ADPKD Patient  
Route Map

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and assessment

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management  
and self-care

Predicting  
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of ADPKD

Treatment  
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progression

Follow-up  
care

End-stage  
renal disease

Dialysis

Kidney  
transplantation

Post-transplant  
care