

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.

Knowledge

GO TO THE
ADPKD PATIENT
ROUTE MAP

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 is possible.

ADPKD is a complex disease that can sometimes affect the 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 health 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 the families should report from a health service.

The aim is to help patients and families to:

- **manage their own health** as much as possible with their health care 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 to manage ADPKD. It also covers management and support, including:

- [signs and symptoms](#)
- [diagnosis](#)
- [treatment](#)
- [support services](#)

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.

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
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- **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 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.

Decisions 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.



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?

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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.

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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



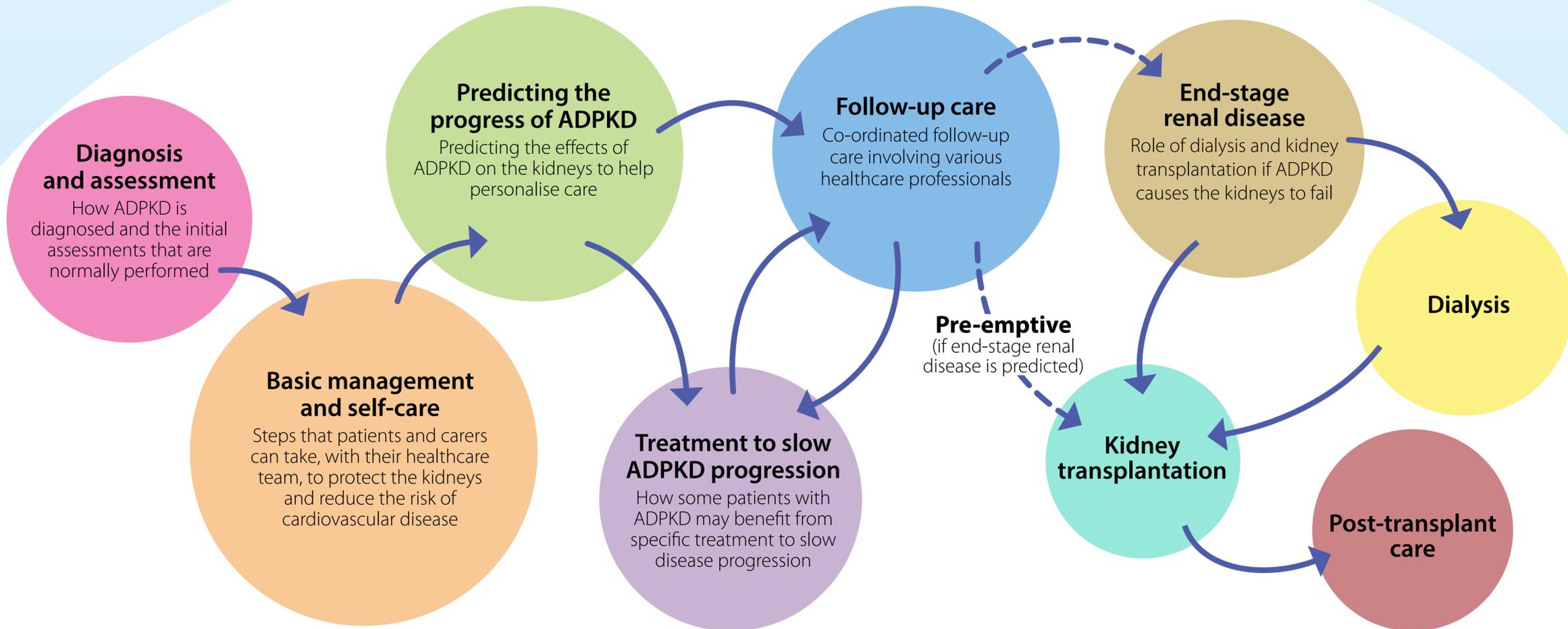
Scientific knowledge

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](#)).



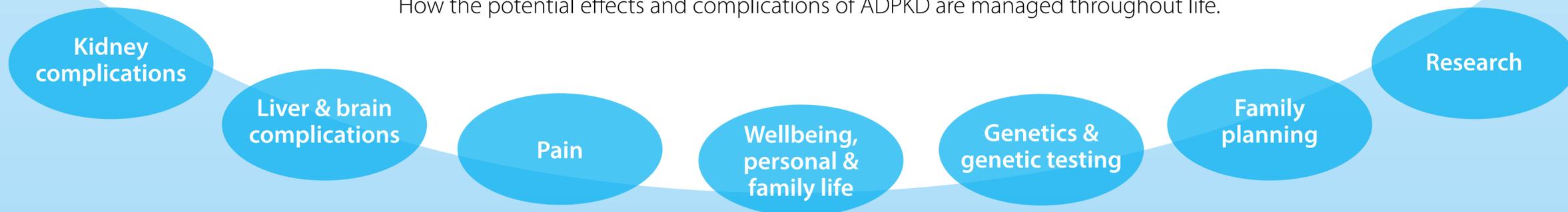
Please click on any bubble to move to that section.

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

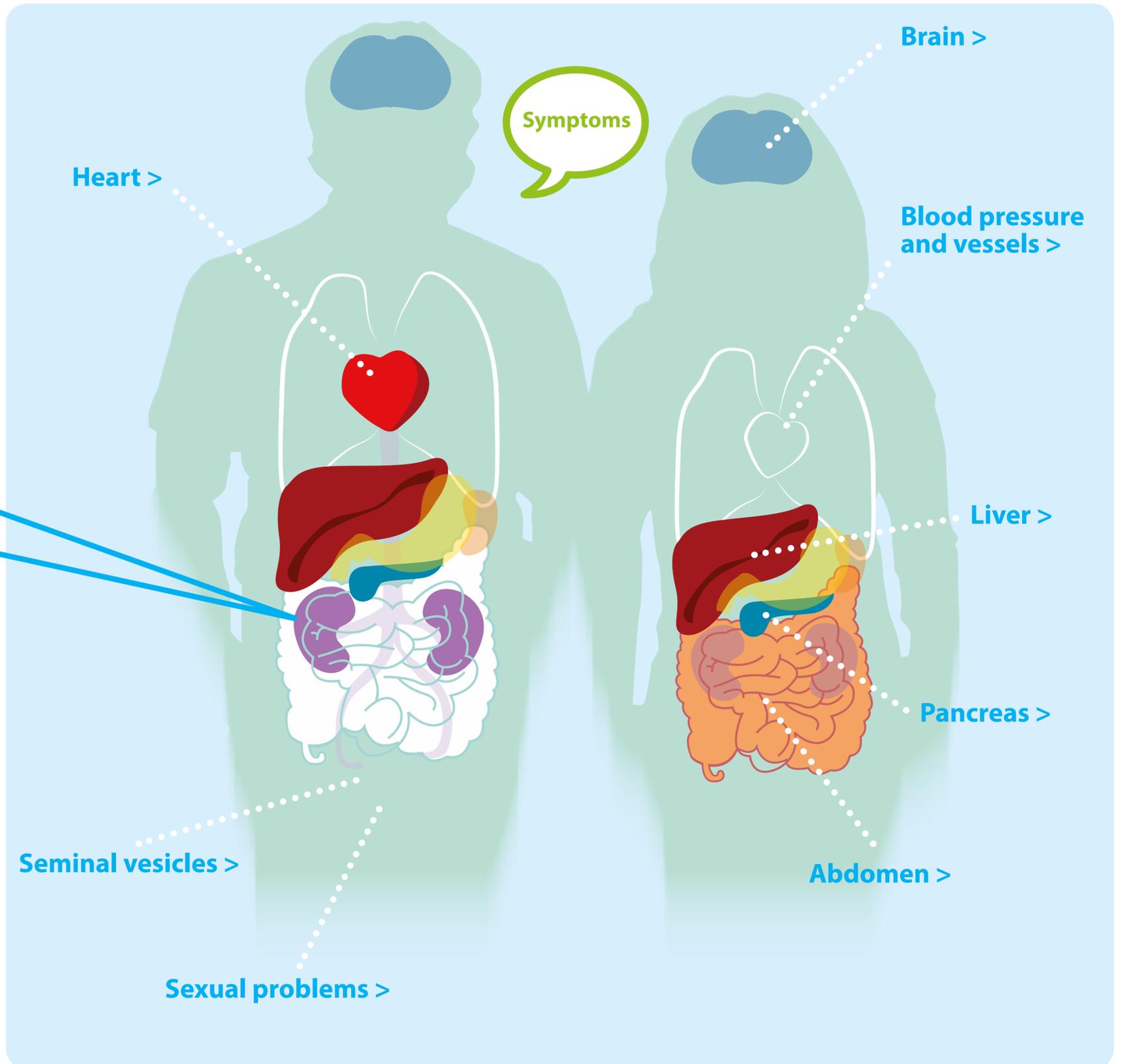
ADPKD is a type of [chronic kidney disease](#) > that causes [cysts](#) > to grow and multiply in the kidneys. If the cysts grow very large they can eventually cause the kidneys and the abdomen to swell. Cysts can eventually stop the kidneys working properly and sometimes this can lead to kidney failure, 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:

ADPKD affects different people in different ways. Some people experience the effects of the disease early. It is also important to remember that many people have no visible symptoms, but still have the effects of ADPKD. Although the disease can be asymptomatic, it is still a disease and needs to be managed. Please contact your doctor, being aware of the following:

More information is available on the website [www.adpkd.org.uk](#) about the effects of ADPKD.

Diagnosis

ADPKD is a type of [autosomal dominant disease](#). This means that you only need to have 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 but not have the disease. This is called being a [carrier](#). You can also have the disease but not have the faulty gene. This is called [genetic mosaicism](#).

From your ADPKD, you can help to understand your own health and the progression of the disease through your [genetic testing](#).

How can the disease be 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](#).

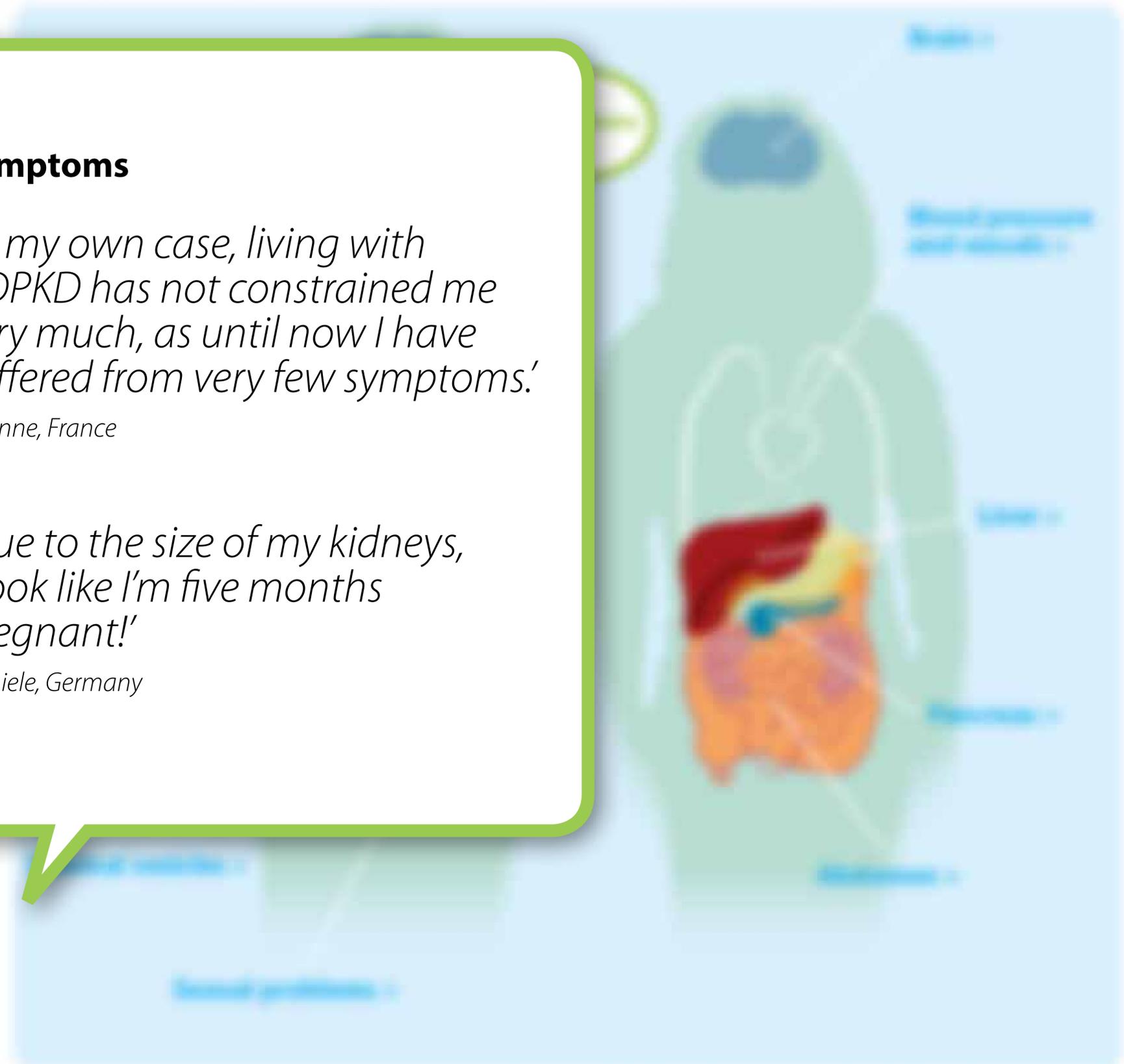
Symptoms

'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



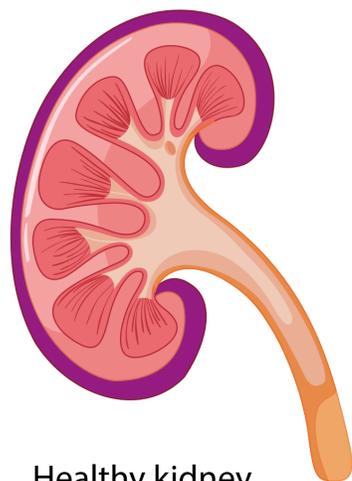
The various conditions from ADPKD can affect the health of patients.

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

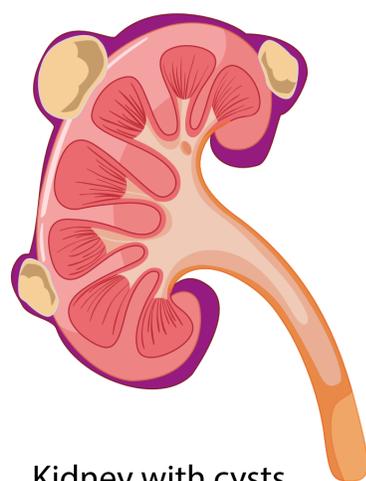
ADPKD affects different parts of the body in different ways. It can cause kidney failure, high blood pressure, brain aneurysms, and other conditions.

Cysts

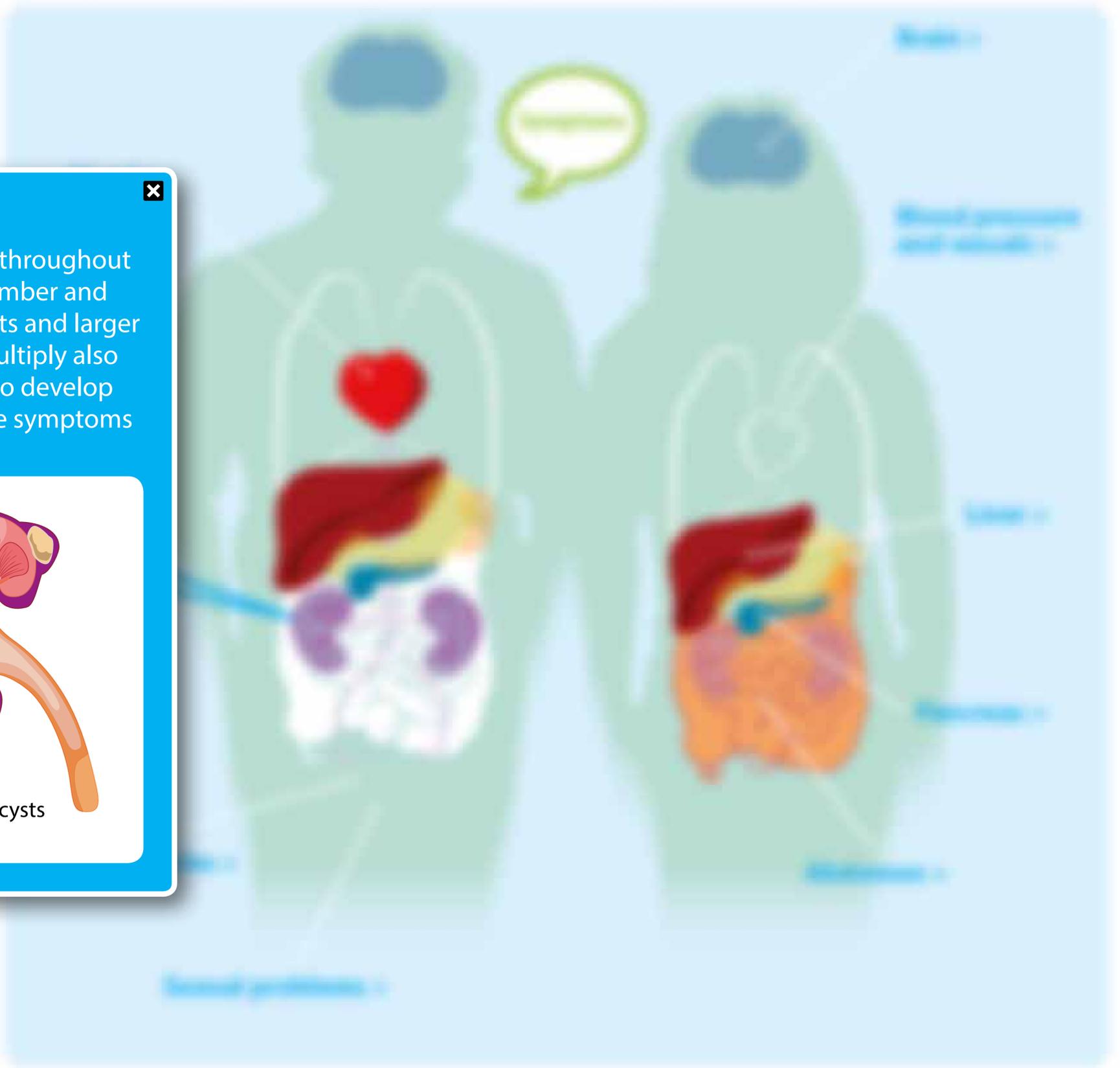
Cysts are fluid-filled sacs that grow and multiply throughout life in the kidneys of people with ADPKD. The number and size of cysts varies – some people have more cysts and larger cysts than others. How quickly they grow and multiply also differs between patients. The cysts usually start to develop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



Healthy kidney



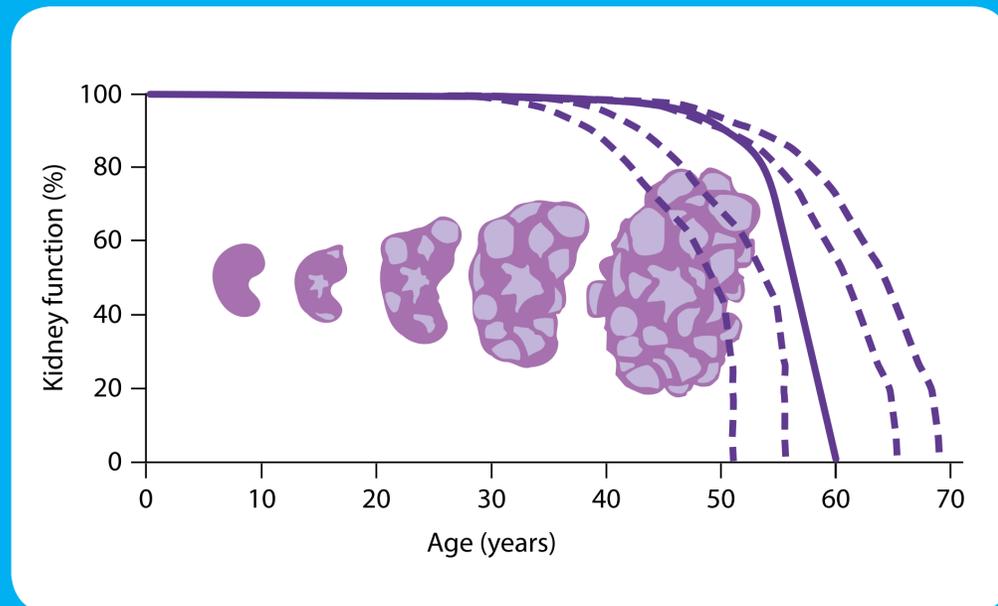
Kidney with cysts



Chronic kidney disease ✕

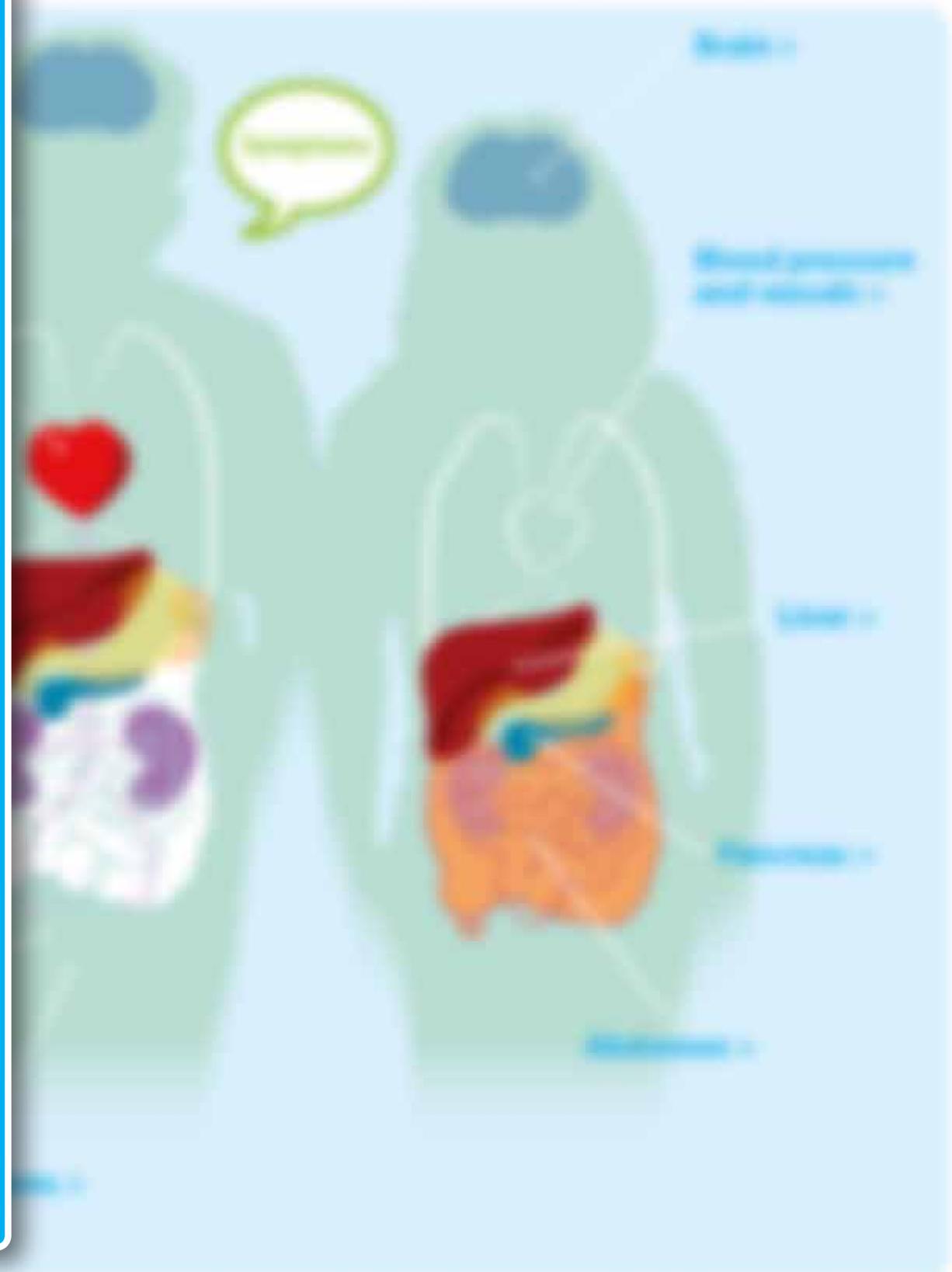
Chronic kidney disease (CKD) is a long-term condition in which the kidneys do not work normally or when they are damaged.

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.



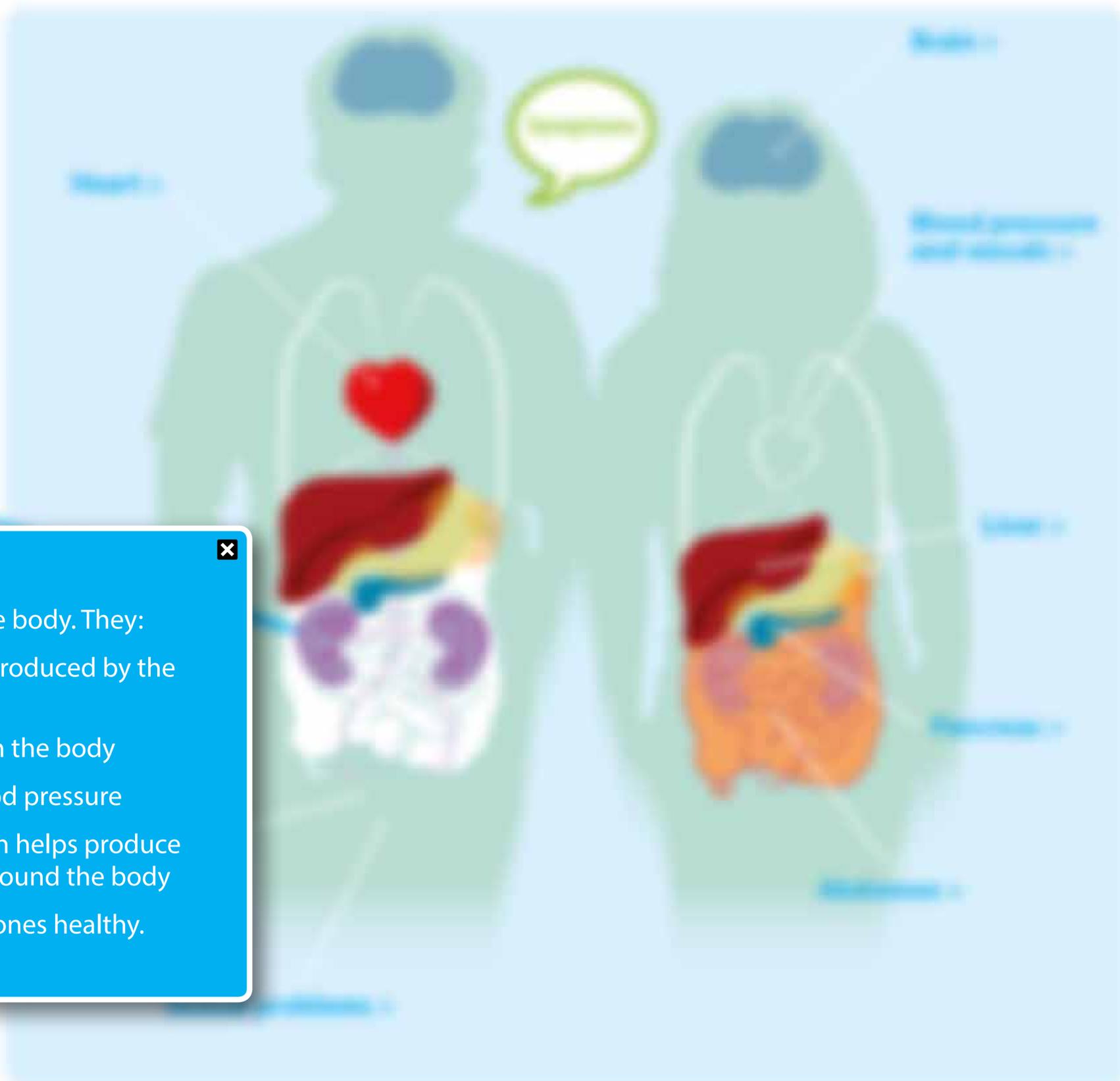
Understanding ADPKD

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

ADPKD affects the kidneys and sometimes other parts of the body, in an inherited way.

ADPKD affects different people in different ways. An important consideration for affected people is to also consider the possibility that their family may have other health conditions, such as high blood pressure, stroke and heart disease. 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 of the faulty gene, you will have a more severe form of the disease. You can also have the disease if you have one copy of the faulty gene and one copy of the normal gene.

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.



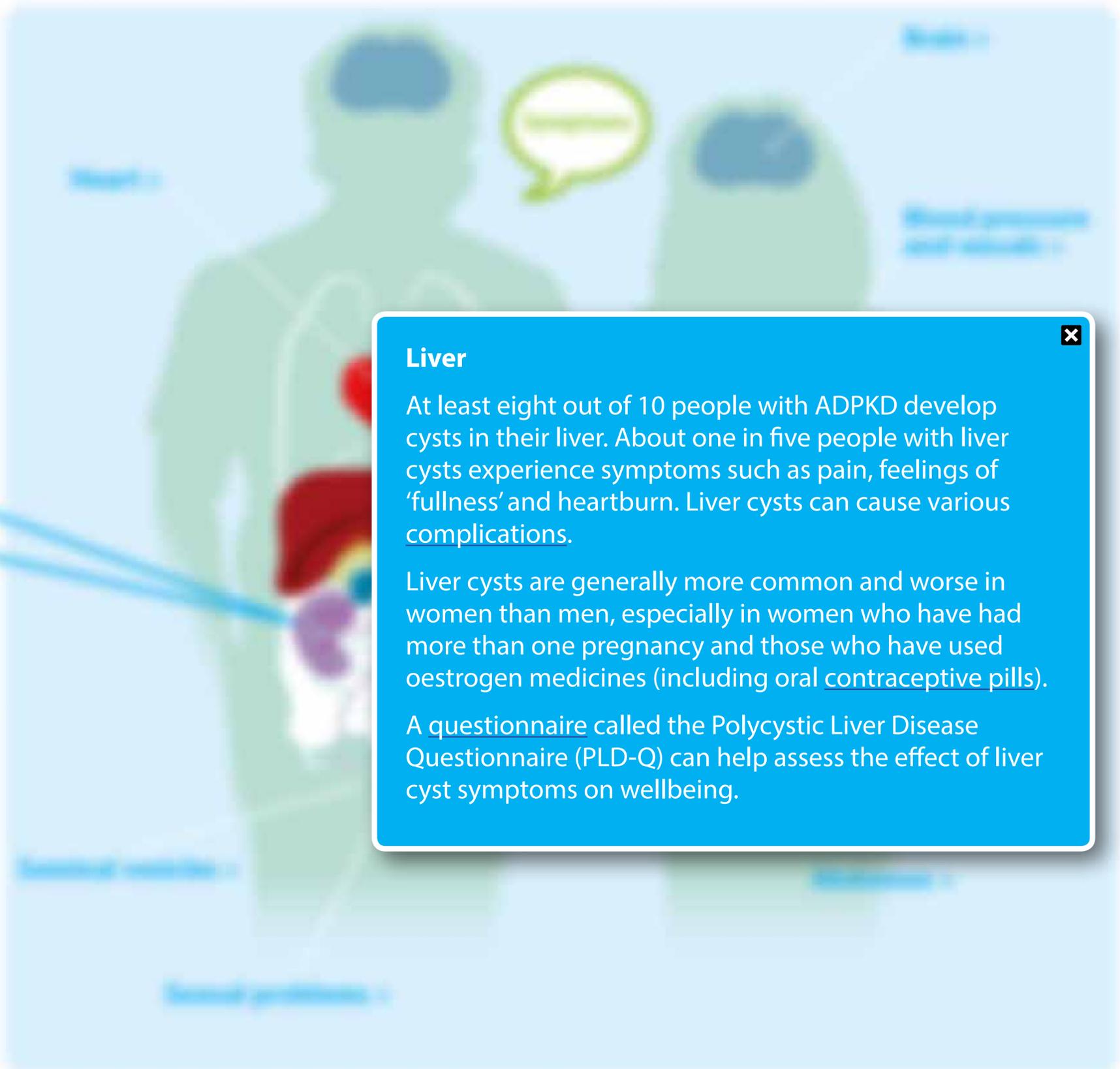
Understanding ADPKD

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 [patient 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. The disease is passed on from one parent to their children. The disease is passed on from one parent to their children.

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. It is important to understand that each person's disease is truly unique. 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.

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

Kidneys

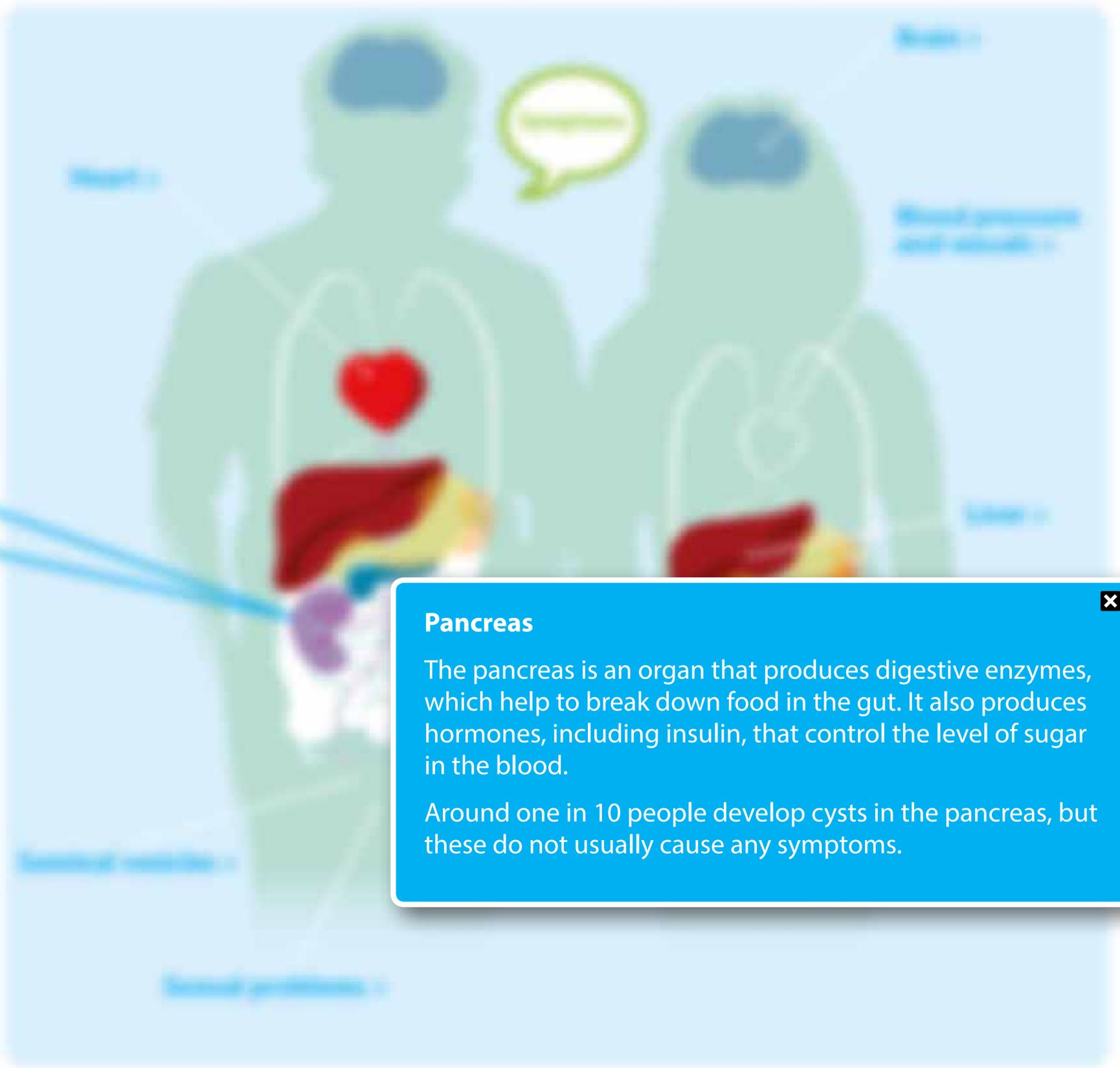
ADPKD is a type of [autosomal dominant disease](#) - this means that you only need one copy of the faulty gene to get the disease. If you have one faulty gene, you can eventually cause the kidneys and the pressure in your blood to gradually stop the kidneys working properly and sometimes this can lead to kidney failure. [Living with kidney failure](#) can be difficult and you will need other [treatments](#) to survive.

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

Living with the disease in pregnancy

Mediating 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](#).



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.



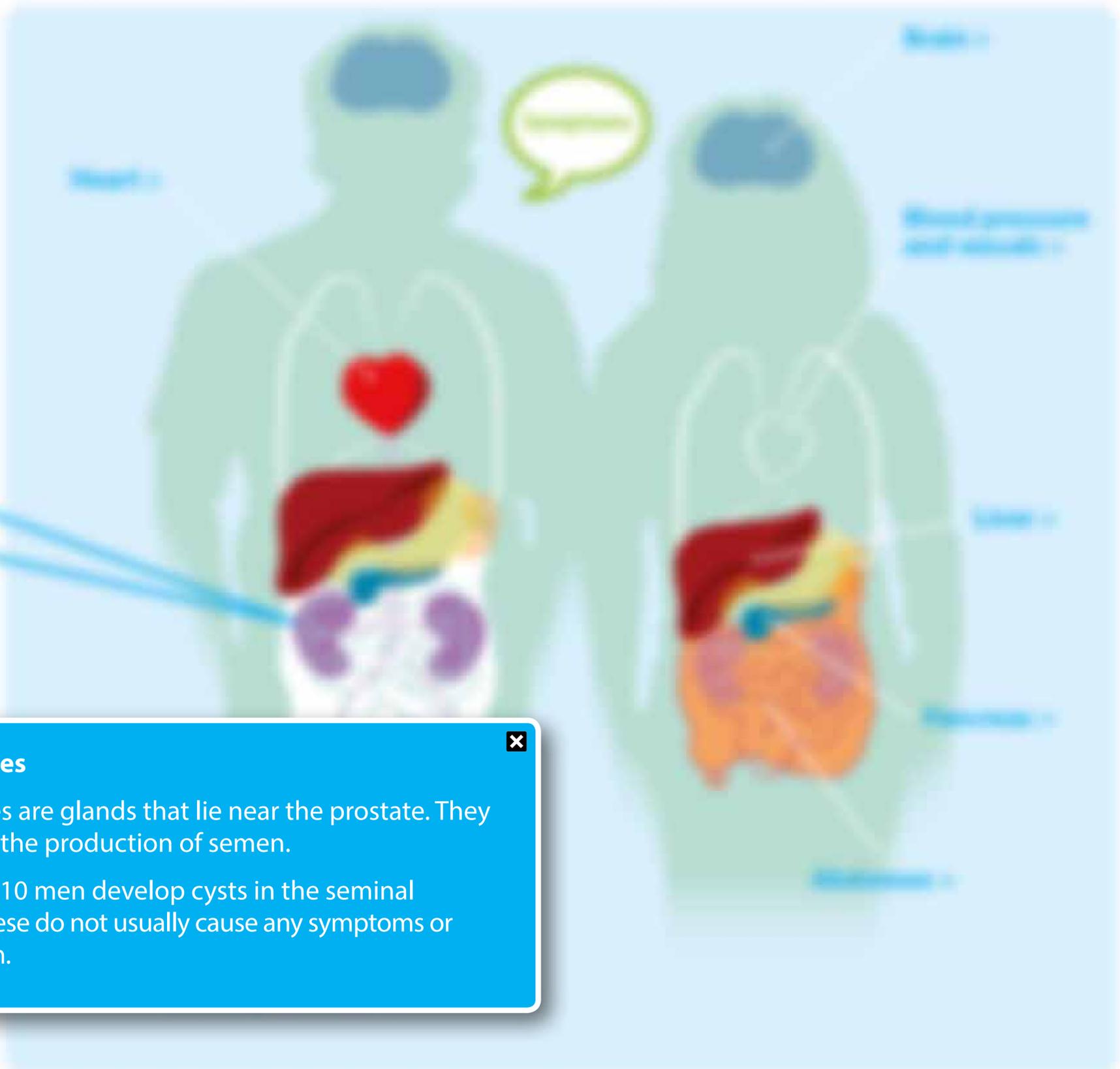
Understanding ADPKD

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

ADPKD affects the kidneys and sometimes other parts of the body. It can 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 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 our website [www.gerf.org.uk](#) or contact our helpline on 0800 012 3456.



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 bladder 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 symptoms of kidney failure.**

If you have ADPKD, you can help to reduce your risk from the progression of the disease through certain **lifestyle changes**.

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.



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 much can be done to help reduce, manage and slow the effects of ADPKD. Although the disease can cause a significant effect on life, it does not mean the people with the disease cannot live happy, long and productive lives.

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

Kidneys

ADPKD is a type of [autosomal dominant disease](#). This means that you only need to have one faulty gene to get the disease. If the other gene is healthy, you can usually pass the healthy one to your children. You can usually stop the faulty gene being passed on to your children by using [assisted reproductive techniques](#).

If you have ADPKD, you can help to control your blood pressure by managing it with [medicines](#).

How can the disease be managed?

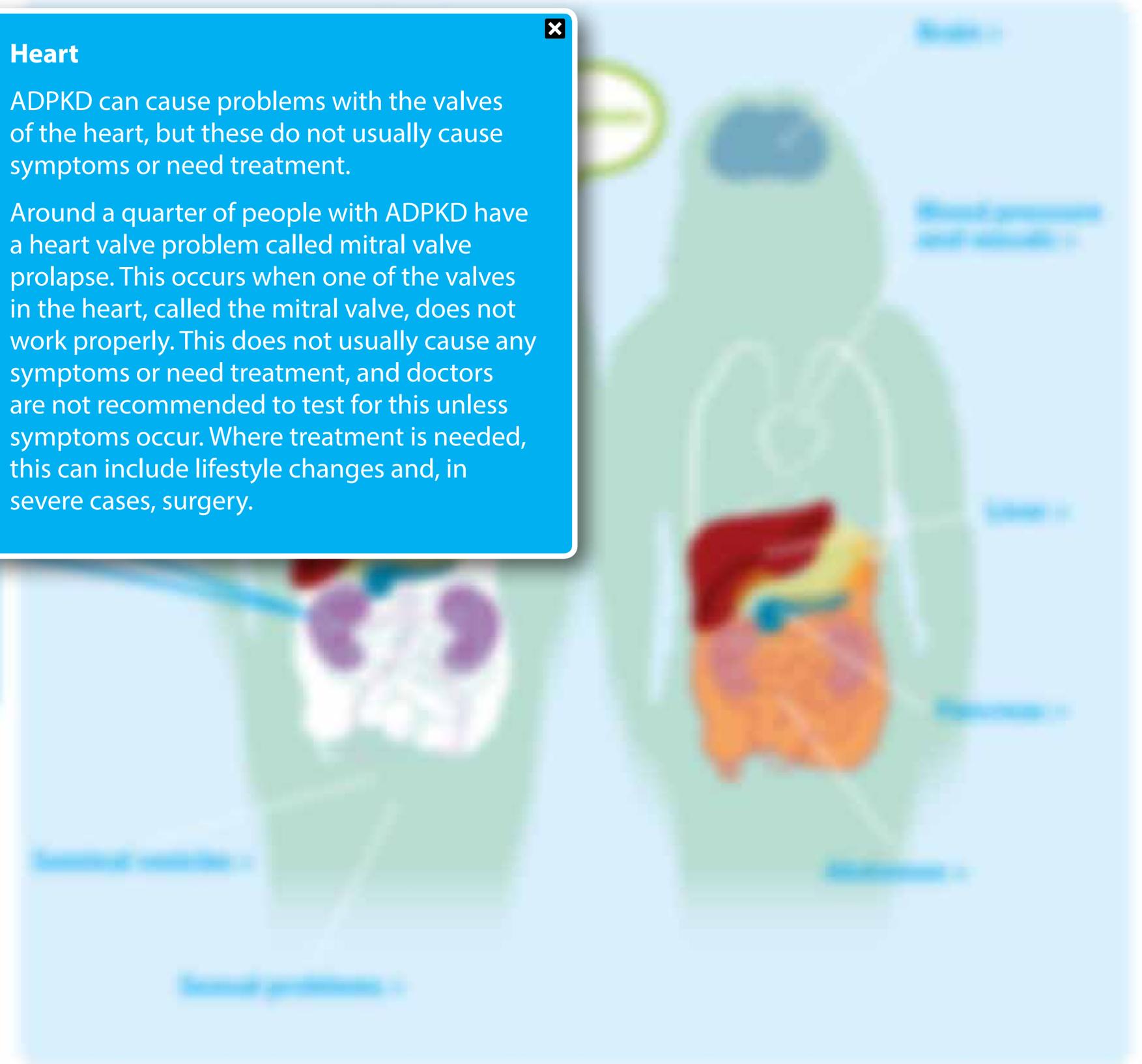
Reducing personal and family life

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

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.



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

ADPKD affects the structure and sometimes the function of the kidneys in both kidneys.

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

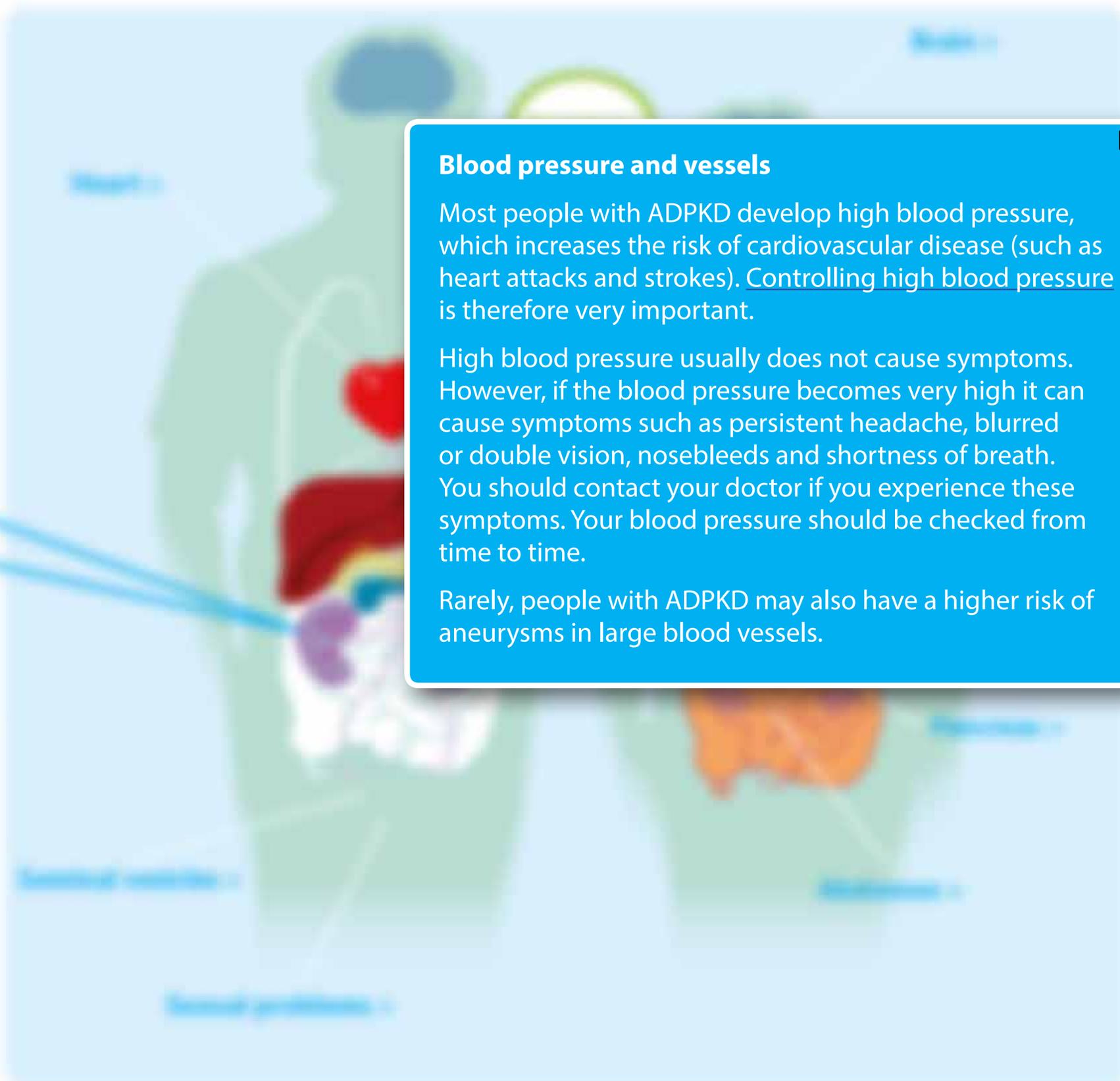
ADPKD is a type of [autosomal dominant disease](#) - this means that you only need to have one faulty gene to be affected. If the other gene is normal, you can usually pass the healthy and the abnormal to your children. You can usually stop the kidney cysts from growing and sometimes they can lead to kidney failure. [More information](#) website provides more information about the [genetics of ADPKD](#).

Even if you have ADPKD, you can help to prevent or reduce the progression of the disease through [certain lifestyle changes](#).

How can the kidneys be supported?

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.

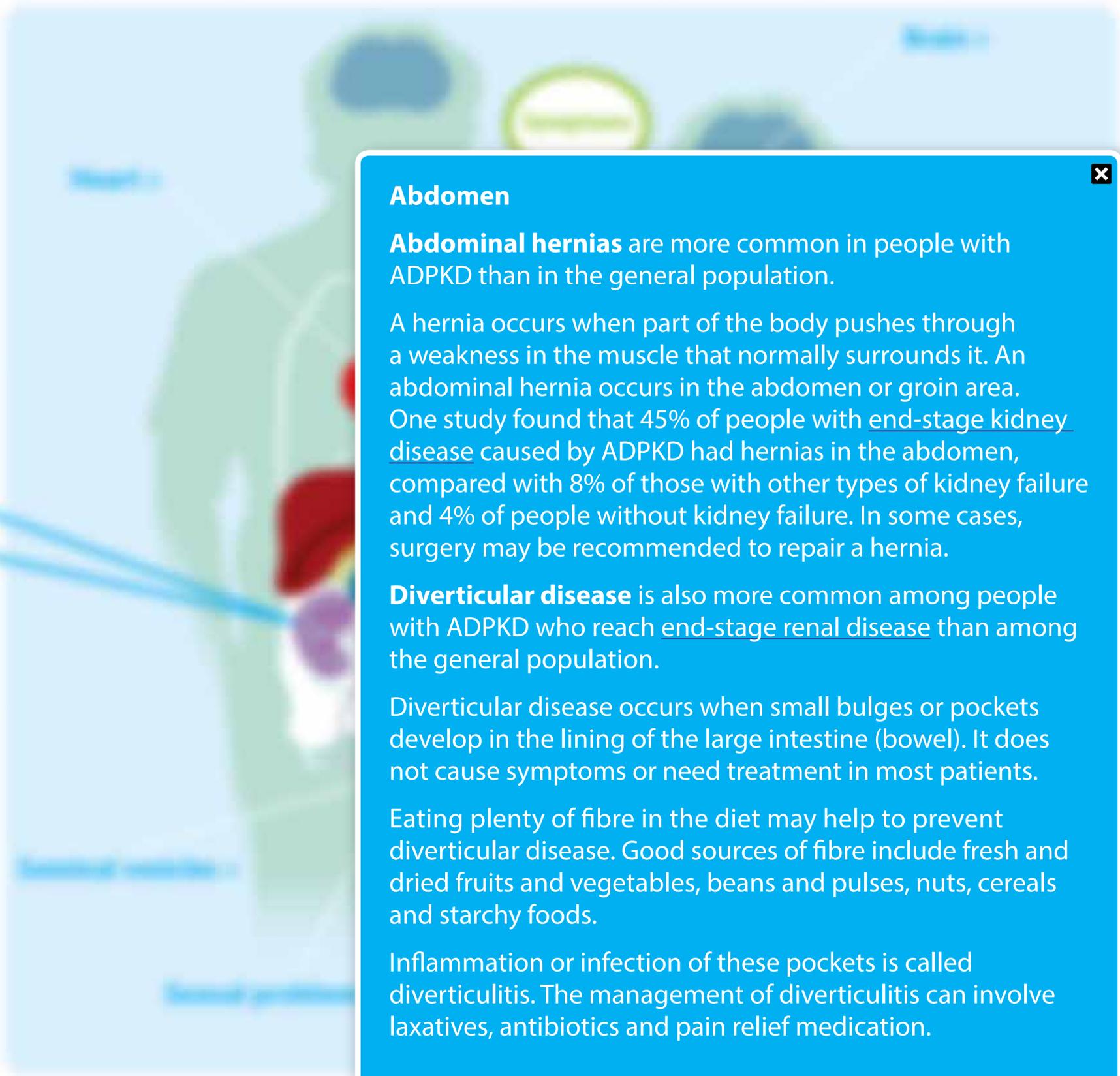
Kidneys

ADPKD causes **fluid-filled cysts** to develop in the kidneys. In some cases, the cysts can become very large and eventually cause the kidneys and the abdomen to swell. This can eventually stop the kidneys from working properly and sometimes lead to kidney failure. [More information](#) website provides more information about the effects of ADPKD.

For more ADPKD information, please visit [www.kidney.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 [ADPKD 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.



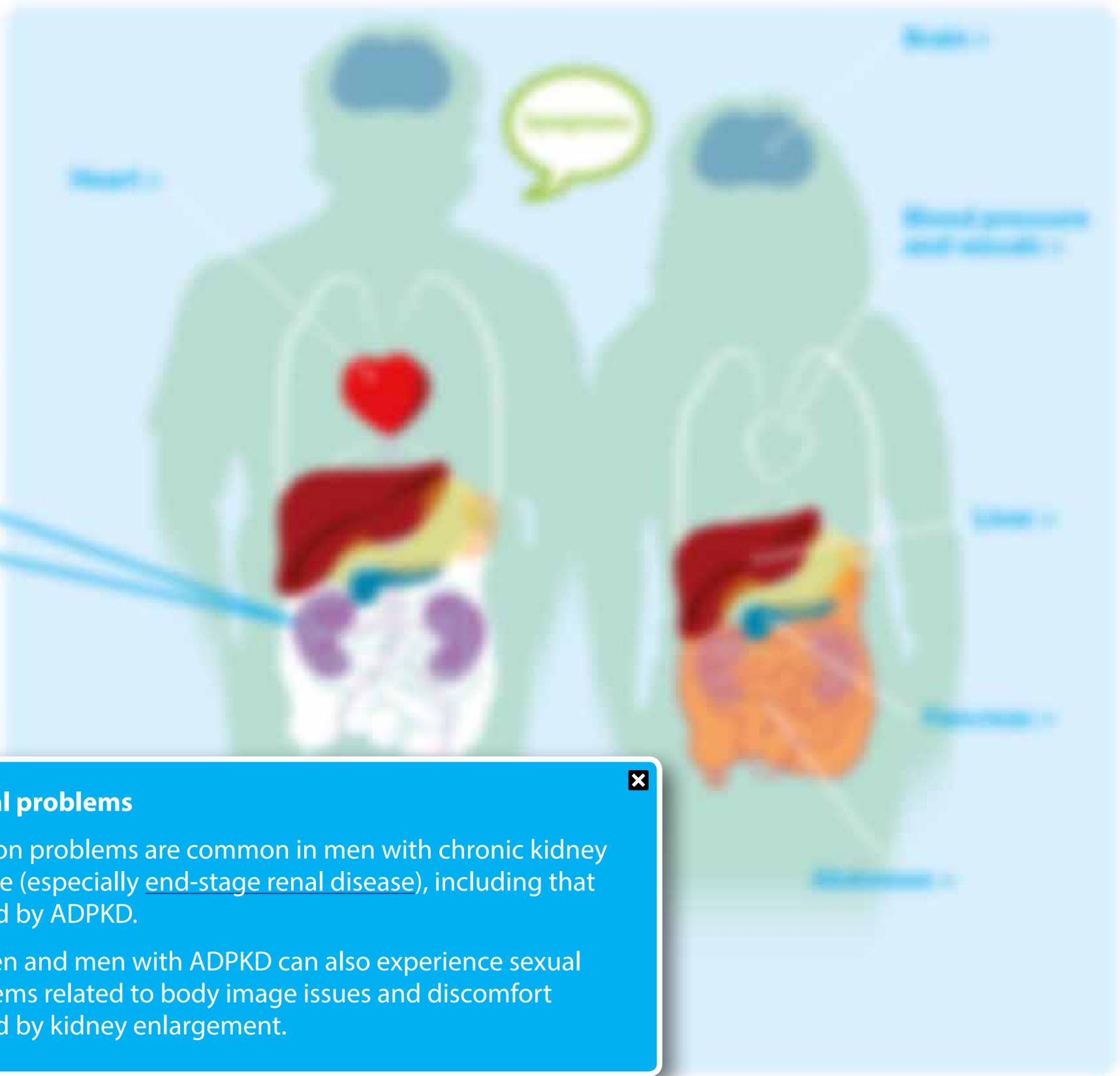
Understanding ADPKD

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](#) - this means you only need one copy of the faulty gene to get the disease. If you only have one copy, you can usually control the disease and the symptoms it causes. You can usually stop the disease getting worse and sometimes you can lead a healthy life. In some cases, you may need to have a kidney transplant or dialysis.
If you have ADPKD, you can help to control your disease from the progression of the disease through certain [lifestyle changes](#).

How can the kidneys be supported?

Maintaining personal and family life
ADPKD can interfere with normal activities, such as work, family life and sport. You can have a significant [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 informed 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?



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.

It should be designed as a clinically appropriate to address [complications](#), manage [comorbidities](#), preserve kidney function, ease the risk of [cardiovascular](#) disease, and maintain optimum quality of life.

Information and support to help patients and their families with [depression](#) and [self care](#) measures and to deal with the impact of the condition.

ADPKD can describe [quality related](#) outcomes ranging from 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">Genetic testingImagingRenal function	<ul style="list-style-type: none">Genetic testingImagingRenal functionCardiovascular riskCardiovascular health	<ul style="list-style-type: none">Genetic testingImagingRenal functionCardiovascular riskCardiovascular health
<ul style="list-style-type: none">Genetic testingImagingRenal function	<ul style="list-style-type: none">Genetic testingImagingRenal functionCardiovascular riskCardiovascular health	<ul style="list-style-type: none">Genetic testingImagingRenal functionCardiovascular riskCardiovascular health



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 the implications of this for you and your family.

It is important to understand the implications of ADPKD for your family. You may want to consider genetic testing for your family members.

It is important to understand the implications of ADPKD for your family. You may want to consider genetic testing for your family members.

ADPKD can be treated with medication. It is important to understand the implications of ADPKD for your family. You may want to consider genetic testing for your family members.

Who is involved in the healthcare team?

Patients with ADPKD may need to see a variety of healthcare professionals. It is important to understand the implications of ADPKD for your family. You may want to consider genetic testing for your family members.

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 involved in ADPKD care?

Specialist	Role
Nephrologist	Lead the care of the patient with ADPKD.
Geneticist	Provide genetic testing and counselling.
Neurologist	Manage headaches and other neurological symptoms.
Cardiologist	Manage high blood pressure and other cardiovascular symptoms.
Endocrinologist	Manage diabetes and other endocrine symptoms.
Psychologist	Provide psychological support and counselling.
Physiotherapist	Provide physical therapy and exercise advice.
Dietitian	Provide dietary advice and support.
Pharmacist	Provide medication advice and support.
Healthcare professional	Provide general support and advice.



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

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 family member with high blood pressure or polycystic ovaries
- 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.

Ultrasound

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

Examination under assessment

Examination under assessment will allow doctors to see if there are any cysts in the kidneys and also check if the kidneys are working properly.

Other investigations

In some cases, a doctor may suspect ADPKD has affected other parts of the body.

Examination investigations include:

- [Blood pressure](#)
- [Blood tests](#)
- [Kidney function tests](#)

Doctors may be able to identify other signs of polycystic ovaries, polycystic kidneys, or polycystic kidneys in other parts of the body.

- [Genetic testing](#)
- [Genetic testing](#)
- [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.



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 experiencing symptoms such as high blood pressure or kidney stones
- are having a routine ultrasound scan of their kidneys

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

Diagnosis and kidney assessment

ADPKD is usually diagnosed by a [general practitioner](#) or [nephrologist](#) who will take a history and examine you.

They will also measure the [blood pressure](#).

They will also check to see if there is any [protein](#) in the urine by doing the [function of the kidneys](#).

They will:

Check blood

ADPKD is usually first diagnosed in the UK in people who have a family history of the condition. They will check to see if there is any protein in the urine by doing the function of the kidneys.

- [Blood pressure](#)



Other investigations

In some cases a doctor will also check the [size of the kidneys](#) and the [position of the kidneys](#).

Other investigations include:

- [Ultrasound](#)
- [MRI](#)
- [Genetic testing](#)

Doctors may be able to identify other types of [genetic](#) conditions such as [polycystic ovary syndrome](#), [hemochromatosis](#) or [Marfan syndrome](#).

- [Genetic testing](#)
- [MRI](#)
- [Ultrasound](#)



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 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 identifiable people who:

- have a past [history of CKPD](#)
- are women in their 40s or 50s, with or without a history of pregnancy
- are related or married to a CKPD patient and someone else in their family has the disease

CKPD is also first suspected in general practitioners family doctors.



Diagnosis and kidney assessment

CKPD is usually diagnosed by a general practitioner (GP) or family doctor. The GP or family doctor will:

- [check blood tests](#)
- [check urine tests](#)
- [check blood pressure](#)

The GP or family doctor will also refer you to a specialist CKPD clinic for further assessment of the kidney.

[Blood tests](#)

CKPD is usually first diagnosed in the UK as a general practice finding in identifiable people. This is because the symptoms are so common and so long-lasting.

- [Blood tests](#)



Nephrologist ✕

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

Referral to a specialist

Referral to a specialist is usually made by a general practitioner (GP) or family doctor. Referral is then made:

- [to a specialist](#)
- [to a specialist](#)

Referral may be advised in other cases of people with CKPD and conditions associated with CKPD in their work.

- [to a specialist](#)
- [to a specialist](#)



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 or their partner, with or without a family history of ADPKD;
- are themselves or their partner, with or without a family history of ADPKD.

ADPKD is often first suspected in people with a family history of ADPKD.

Diagnosis and kidney assessment

ADPKD is usually diagnosed by a [genetic test](#).
Diagnosis and assessment include tests 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 other parts of the body.

Diagnosis and assessment include:

- [blood pressure](#) monitoring;
- [blood tests](#) to check for kidney function and other health problems;
- [kidney scans](#) to see and measure the cysts.

Diagnosis and assessment include:

- [blood pressure](#) monitoring;
- [blood tests](#) to check for kidney function and other health problems;
- [kidney scans](#) to see and measure the cysts.



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

Key and recent changes of CKPD are covered and discussed in this step to manage the disease. Typically, CKPD is diagnosed and assessed by following the steps:

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 ²)
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 end-stage renal disease	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 CKPD is diagnosed and the initial tests and examinations that are normally performed.

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

Identifying people with CKPD

CKPD is usually first suspected in identifiable people who:

- have a long [history of diabetes](#)
- are especially prone to kidney disease, such as high blood pressure in pregnancy
- are related or connected to CKPD because someone else in their family has the disease

CKPD is also first suspected in general practices family doctors.

Diagnosis and kidney assessment

CKPD is usually diagnosed by a [general practitioner](#). Regular kidney assessments include urine and blood tests.

Blood tests

Blood tests allow doctors to see how well the kidneys are working.

Urine tests

Urine is passed out of the blood and into the bladder. The amount of protein in the urine is a sign of CKPD. A high amount of protein in the urine is a sign of CKPD.

Health history

CKPD is usually diagnosed in the UK by a general practice doctor. It is usually first suspected in people who have a long history of diabetes, high blood pressure, or kidney disease.

Family history

CKPD is also first suspected in general practices family doctors.

Other investigations

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

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 in a step-by-step way.

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 ADPKD has had symptoms due to their family has the disease

ADPKD is also first suspected in general population 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

Renal function tests are blood and urine tests 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. Genetic testing can also be used to identify people who are at risk of developing ADPKD.

Genetic testing



Other investigations

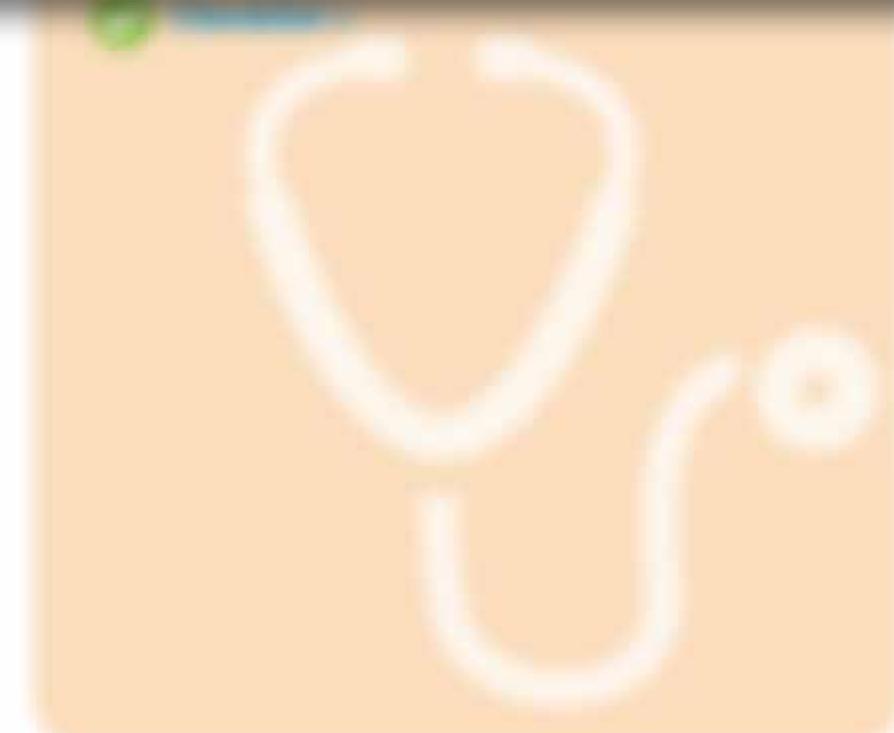
It is important to check for ADPKD-related complications of the [liver](#).

Investigations include:

Imaging

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 30 years of age.

- [see also: Genetics of ADPKD](#)
- ADPKD is usually first diagnosed in adulthood, usually after 30 years of age.
- ADPKD is usually first diagnosed in adulthood, usually after 30 years of age.

ADPKD is often first diagnosed in young people with family history.



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 ACPHD is diagnosed and the initial tests and examinations that are normally performed.

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

Identifying people with ACPHD

ACPHD 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 ACPHD because someone else in their family has the disease

ACPHD is also first suspected in people with a family history of the disease.

Diagnosis and kidney assessment

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

Blood tests

Blood tests allow doctors to see whether you have ACPHD.

Imaging tests

Some tests can be used to check whether you have ACPHD. Imaging tests include:

Imaging tests

ACPHD is usually diagnosed by the following tests. Some tests are usually done in a hospital. Some tests can be done at home. Some tests are done in a hospital. Some tests are done in a hospital.

Other investigations

It is important to check whether you have other health problems. Some of the tests include:

Some tests include:

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

Here are some of the most important things you can do to help protect your kidney function:

- Stop smoking
- Eat a healthy diet
- Exercise regularly
- Keep your blood pressure under control
- Keep your cholesterol under control
- Keep your diabetes under control

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 many other risk factors for cardiovascular disease that you can control.

How can I control my blood pressure?

Keeping it up

It's important to keep your blood pressure under control. This means taking your medicine as prescribed and checking your blood pressure regularly. Your healthcare team should be able to help you with this.

Living personal and family life

It's important to live a healthy lifestyle. This means eating a healthy diet, exercising regularly, and not smoking. Your healthcare team should be able to help you with this.

How can I control my cholesterol?

Smoking

How can blood pressure be controlled?

There are many ways to control your blood pressure. These include taking medicine, eating a healthy diet, exercising regularly, and not smoking.

There are many ways to control your blood pressure. These include taking medicine, eating a healthy diet, exercising regularly, and not smoking.

There are many ways to control your blood pressure. These include taking medicine, eating a healthy diet, exercising regularly, and not smoking.

How can blood pressure be controlled?



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 important to prevent or slow the development of conditions such as high blood pressure and diabetes. You can do many things that may help prevent and control these conditions. This includes:

• **Weight and diet** – see [Weight and diet](#) for more information.

• **Exercise** – see [Exercise](#) for more information.

Other risk factors

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

Smoking

Smoking is a major risk factor for cardiovascular disease. It is important to stop smoking if you are a smoker. For more information, see [Smoking](#).

Alcohol

Drinking alcohol can increase the risk of cardiovascular disease. It is important to drink alcohol in moderation. For more information, see [Alcohol](#).

Stress

Stress can increase the risk of cardiovascular disease. It is important to manage stress. For more information, see [Stress](#).

High cholesterol

High cholesterol can increase the risk of cardiovascular disease. It is important to manage cholesterol. For more information, see [High cholesterol](#).

High blood pressure

High blood pressure can increase the risk of cardiovascular disease. It is important to manage blood pressure. For more information, see [High blood pressure](#).

Diabetes

Diabetes can increase the risk of cardiovascular disease. It is important to manage diabetes. For more information, see [Diabetes](#).

Other risk factors

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

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

Smoking

Smoking is a major risk factor for cardiovascular disease. It is important to stop smoking if you are a smoker. For more information, see [Smoking](#).

Alcohol

Drinking alcohol can increase the risk of cardiovascular disease. It is important to drink alcohol in moderation. For more information, see [Alcohol](#).

Stress

Stress can increase the risk of cardiovascular disease. It is important to manage stress. For more information, see [Stress](#).

High cholesterol

High cholesterol can increase the risk of cardiovascular disease. It is important to manage cholesterol. For more information, see [High cholesterol](#).

High blood pressure

High blood pressure can increase the risk of cardiovascular disease. It is important to manage blood pressure. For more information, see [High blood pressure](#).

Diabetes

Diabetes can increase the risk of cardiovascular disease. It is important to manage diabetes. For more information, see [Diabetes](#).

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 m². 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 health things, because all cardiovascular disease is preventable. They include:

- Drinking water to help kidney function which may protect kidney function in ADPKD.

• [Not smoking](#)

- [Not drinking alcohol](#)
- [Not eating too much salt](#)
- [Not eating too much fat](#)

For people with [diabetes](#)

For people with [diabetes](#)

Research on the benefits of low sodium diets for people with ADPKD is limited. However, low sodium diets may help reduce the growth of kidney cysts in some people with ADPKD. In people with [diabetes](#), controlling high blood pressure and keeping sodium intake below the risk may be the most important.

How can blood pressure be controlled?

For people with high blood pressure, the doctor and the patient should do particular attention to reduce the risk of cardiovascular disease.

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

People [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!

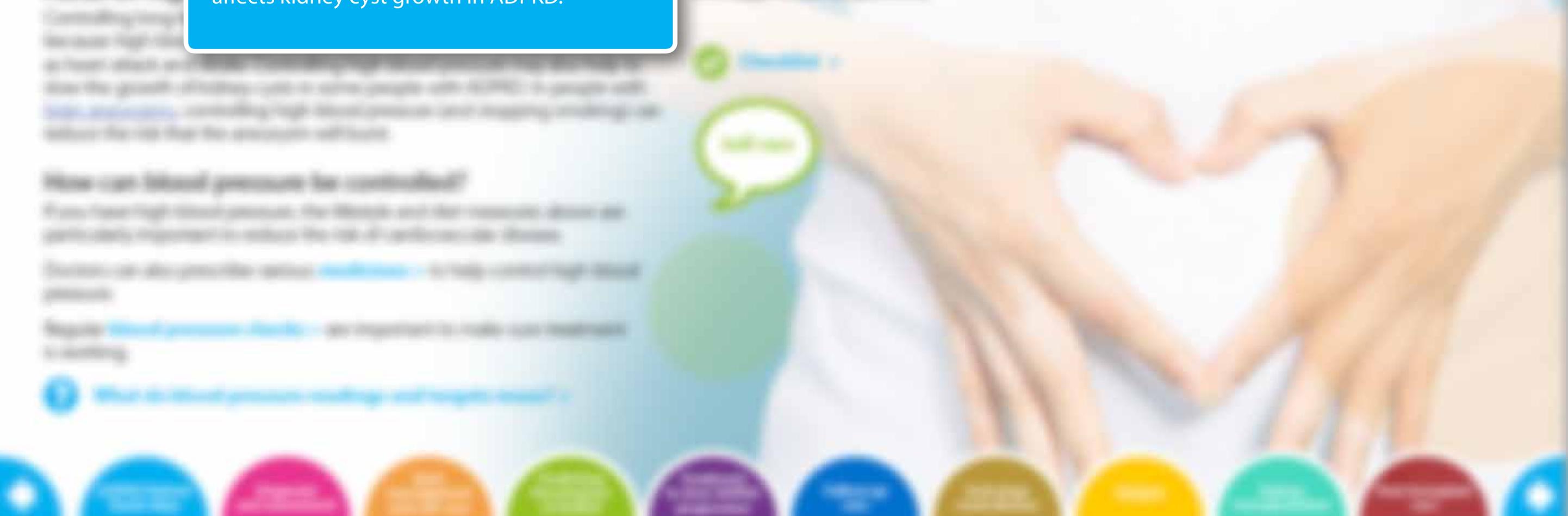
Living the way that can help to stay healthy is not easy. Staying healthy means making good choices and doing what you need to do every day. For some people, it can be difficult to manage your long-term health. Your healthcare team should be able to provide further resources of help and support locally and online. Family friends and [support groups](#) can also provide valuable help and advice.

Maintaining personal and family life

People with kidney disease can still live a full and active life with the right help. There are many [support groups](#) and [resources](#) available. If you have ADPKD, it is important to work with your healthcare team to find out what you can do to manage your health and to find out what resources are available to help you.

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 usually a slow process. ADPKD can be managed through diet and lifestyle changes to help protect kidney function and reduce the risk of high blood pressure and cardiovascular disease.

Most of these are general health advice, but some are specific to ADPKD. This includes:

- [Eating a healthy diet](#)
- [Drinking enough water](#)
- [Not smoking](#)
- [Drinking alcohol](#)
- [Not taking NSAIDs](#)
- [Not taking herbal supplements](#)

For more information on these topics, see the [ADPKD self care guide](#).

Focus on high blood pressure

Controlling your blood pressure is one of the most important things you can do to protect your kidney function. High blood pressure can lead to kidney damage and is a major risk factor for cardiovascular disease. It is important to keep your blood pressure under control to help protect your kidney function and reduce the risk of cardiovascular disease.

How can blood pressure be controlled?

There are several ways to control your blood pressure. These include lifestyle changes, such as eating a healthy diet, drinking enough water, not smoking, and not taking NSAIDs or herbal supplements. Your doctor may also prescribe medication to help control your blood pressure.

For more information on these topics, see the [ADPKD self care guide](#).

Other risk factors

There are several other risk factors for cardiovascular disease, including high cholesterol, diabetes, and smoking. It is important to manage these risk factors to help protect your kidney function and reduce the risk of cardiovascular disease.

For more information on these topics, see the [ADPKD self care guide](#).

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 chronic kidney disease, lifestyle changes can help to slow the progression of the disease. This includes eating a healthy diet, staying active, and not smoking.

More information on general health advice for people with CKD is available on the [CKD website](#).

For more information on lifestyle changes, visit the [CKD website](#).

How to monitor your blood pressure

It's important to check your blood pressure regularly. This can help you to spot any changes early and take action if needed.

How to take your blood pressure

There are several things you can do to ensure you get an accurate reading. These include sitting down, resting for a few minutes, and not talking or moving during the measurement.

For more information on how to take your blood pressure, visit the [CKD website](#).

How can high blood pressure be controlled?

Controlling your blood pressure is important to help slow the progression of CKD. There are several ways to do this, including taking medication, eating a healthy diet, and staying active. It's important to work with your doctor to find the best way for you to control your blood pressure.

How can blood pressure be controlled?

There are several ways to control your blood pressure, including taking medication, eating a healthy diet, and staying active.

For more information on how to control your blood pressure, visit the [CKD website](#).

It's important to work with your doctor to find the best way for you to control your blood pressure.

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 as it can help reduce the risk of long-term complications in ADPKD.

How healthy is...

...my kidney?

...my blood pressure?

...my cholesterol?

...my diabetes?

...my heart?

...my overall health?

...my quality of life?

...my mental health?

...my social life?

...my work?

...my family life?

...my relationships?

...my finances?

...my housing?

...my transport?

...my education?

...my leisure time?

...my hobbies?

...my interests?

...my values?

...my beliefs?

...my culture?

...my religion?

...my ethnicity?

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 good habits you have started. Staying healthy means you'll be able to manage your condition better. It's important to be able to manage your condition. Your healthcare team should be able to help you with this. They can provide advice and support. They can also provide advice on how to manage your condition.

Work and family life

It's important to have a good work-life balance. This means you can manage your condition better. It's important to have a good work-life balance. This means you can manage your condition better. It's important to have a good work-life balance. This means you can manage your condition better.

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 or at high risk of kidney disease, it is important to prevent the development of conditions such as high blood pressure and high cholesterol, as these can do more damage than kidney disease itself and increase the risk of heart disease and cardiovascular disease.

Many of these are preventable. There are several things you can do to help prevent and control these conditions. They include:

- [Stop smoking](#) - stop smoking as soon as you can, as it can damage your kidneys and increase the risk of heart disease and cardiovascular disease.
- [Stop drinking alcohol](#) - stop drinking alcohol as soon as you can, as it can damage your kidneys and increase the risk of heart disease and cardiovascular disease.
- [Lose weight](#) - if you are overweight, losing weight can help control your blood pressure and cholesterol, and reduce the risk of heart disease and cardiovascular disease.
- [Exercise](#) - regular exercise can help control your blood pressure and cholesterol, and reduce the risk of heart disease and cardiovascular disease.

For more information on these and other things you can do, 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 reduce the risk of heart disease and cardiovascular disease.

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 range?

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, or are worried about what you may have, you may wish to discuss any such problems with your healthcare team so that they can provide information, care and support as appropriate.

Links



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 follow the recommendations of your healthcare team. If you have kidney disease, you may be able to do things that may help protect your kidney function and reduce the risk of high blood pressure and cardiovascular disease.

There are several things you can do to help protect your kidney function. These include:

- [Stop smoking](#) - stop smoking as soon as you can
- [Stop drinking alcohol](#) - stop drinking alcohol as soon as you can
- [Stop taking NSAIDs](#) - stop taking NSAIDs as soon as you can
- [Stop taking herbal supplements](#) - stop taking herbal supplements as soon as you can

For more information on these things, see the [stop smoking](#) and [stop drinking alcohol](#) pages.

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 disease. Controlling high blood pressure may also help to slow the growth of kidney cysts in some people with ADPKD. To learn more, see the [high blood pressure](#) page.

How can blood pressure be controlled?

There are several ways to control blood pressure, including:

- [Lifestyle changes](#) - such as eating a healthy diet and exercising

- [Medicines](#) - such as ACE inhibitors and diuretics

- [Regular blood pressure checks](#) - so you can see if your blood pressure is under control

Other risk factors

There are other things you can do to help protect your kidney function and reduce the risk of cardiovascular disease.

Diabetes

Diabetes is a condition where your blood sugar is too high.

Diabetes can damage your kidneys and increase the risk of cardiovascular disease.

For more information on diabetes, see the [diabetes](#) page.

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

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

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

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 each patient's individual disease.

The rate that ADPKD progresses can be measured and even estimated in advance. The main factors are:

- identify patients with rapidly progressing disease who may be eligible for certain treatments or for [transplants](#)
- 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 [symptoms and signs](#) before the age of 30 years. It is recommended that ADPKD progression should be checked every 3-4 years in dependent patients with this family history.



Kidney Function Tests

The rate that ADPKD progresses can be measured and even estimated in advance. The main factors are:

- identify patients with rapidly progressing disease who may be eligible for certain treatments or for [transplants](#)
- 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.

[Blood pressure](#) is the force of the pushing of the blood against the walls of the arteries. High blood pressure can be treated with medicine to slow ADPKD progression.

Some of these factors in the [PROPKD](#) score are related to the [PROPKD](#) score. In addition, the rate of progression of [proteinuria](#) and the rate of progression of [proteinuria](#) are also related to the rate of progression of kidney function.



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

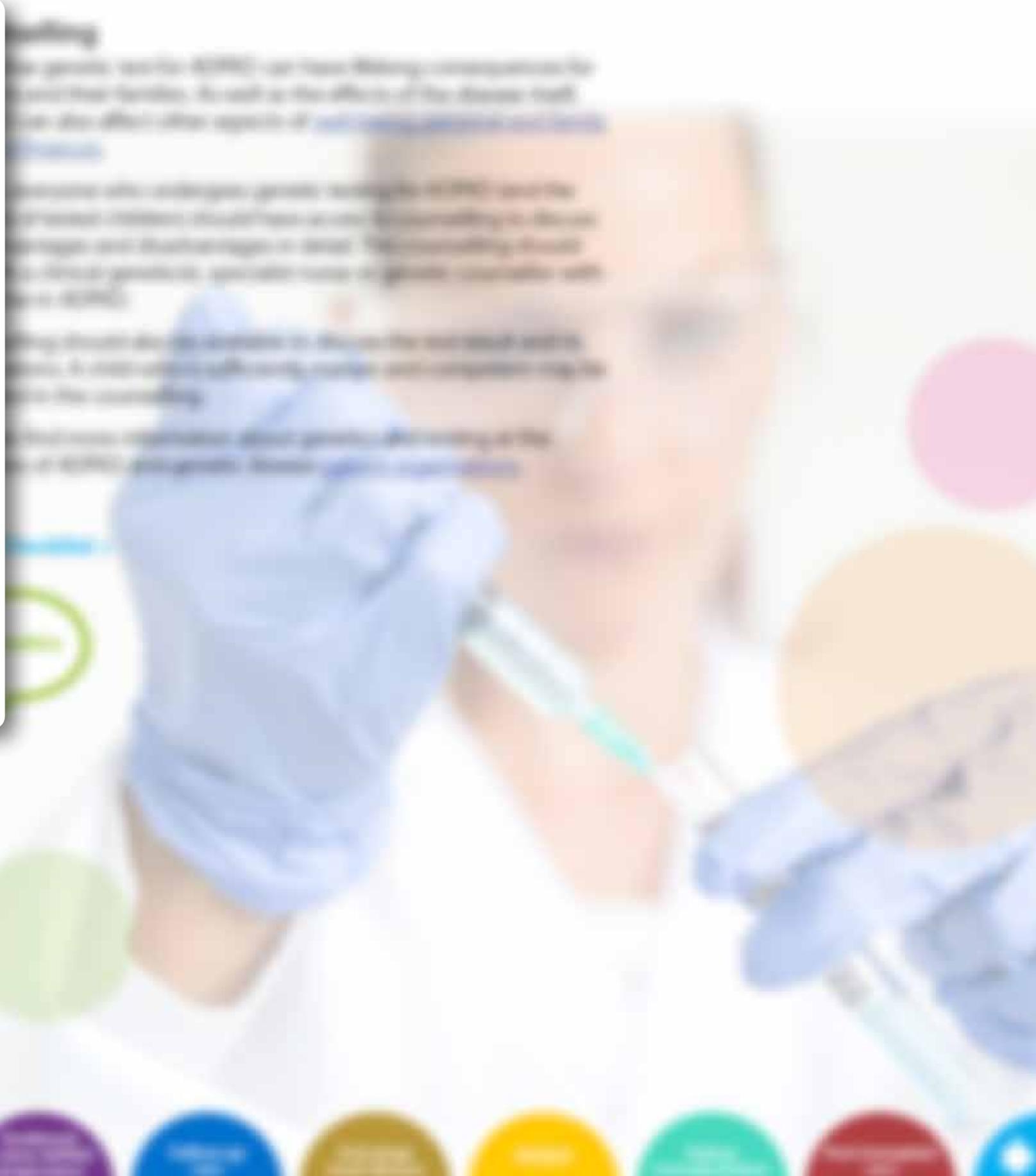
ADPKD is a genetic condition that can be passed from parents to their children. This section explains the basics of ADPKD genetics, inheritance and genetic testing.

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 if you are at risk of passing the gene to your children.



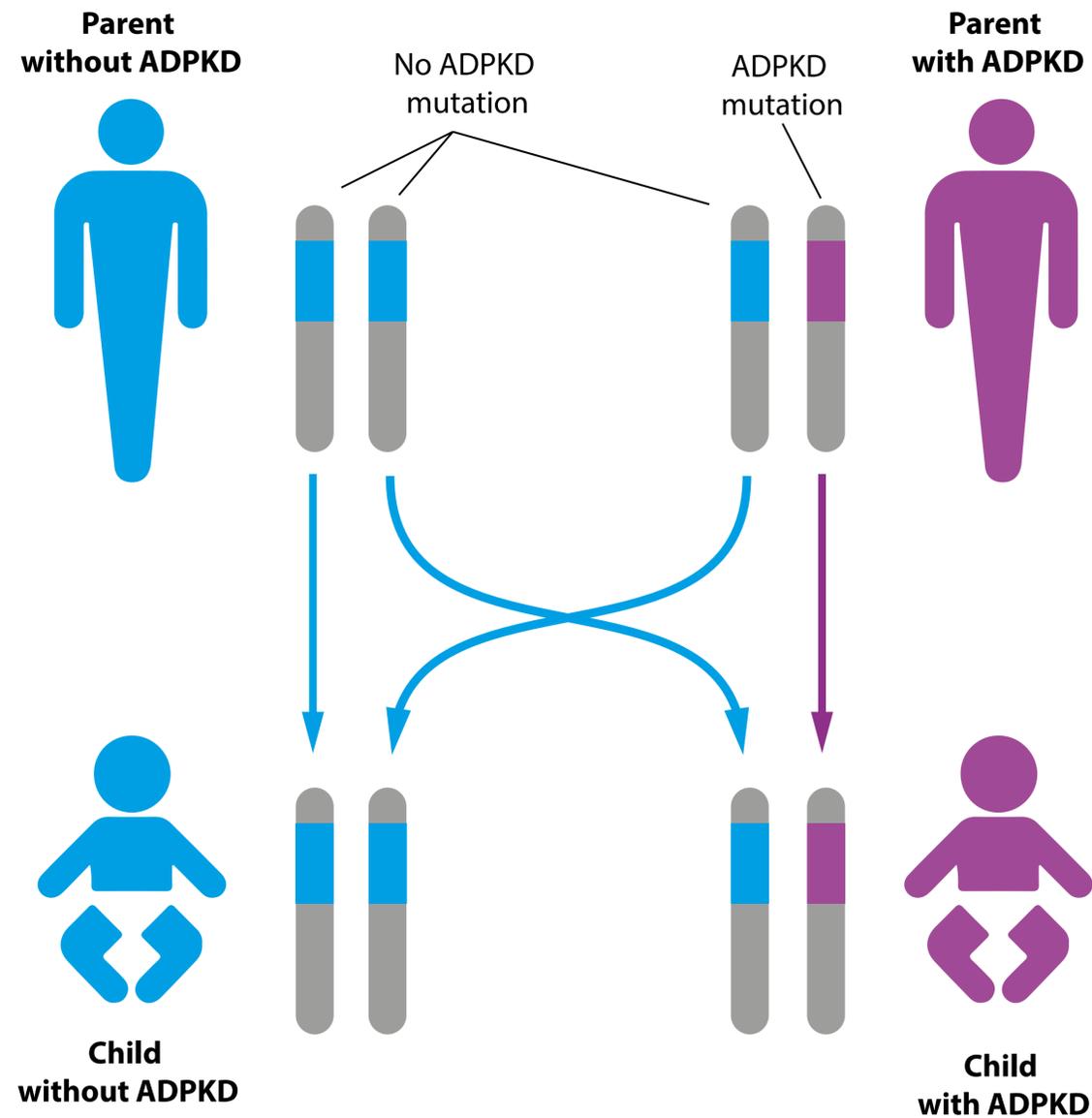
Dominant pattern

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 12 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 the mutation.

ADPKD is not the same as autosomal recessive kidney disease. Autosomal recessive kidney disease is caused by a different mutation.

Genetic testing

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

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

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

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.

Pre-implantation

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

Genetic testing can also be used to find out if you have a mutation in the PKD1 or PKD2 gene. This can help you decide if you want to have children.

Genetic testing can also be used to find out if you have a mutation in the PKD1 or PKD2 gene. This can help you decide if you want to have children.

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 showed that some patients with ADPKD may benefit from specific treatment to slow disease progression.

Who is suitable for this treatment?

The evidence that is currently available suggests that patients with ADPKD who have a certain level of kidney function and a certain number of kidney cysts may benefit from this treatment.

What is the benefit?

The evidence suggests that if the benefit shown in a clinical trial was maintained, the overall health of each patient would improve over the course of their life.

What are the main side effects?

The most common side effect of this treatment is an increase in uric acid levels. If you take this medicine, you should take [allopurinol](#) to prevent any side effects.

Are there any other treatments being developed for ADPKD?

Other medicines are being investigated to slow the progression of ADPKD and reduce the risk of kidney failure. These may include [sodium bicarbonate](#) and [acetazolamide](#).

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.



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. Currently, there are no treatments available that can slow down the disease and help reduce the risk of kidney failure.

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 be given some blood tests for the first 12 months of treatment and every 12 months after that.

What are the main side effects?

The most common side effect of this treatment is an increase in the number of cysts in the kidneys. You may also experience [weight gain](#) and [swelling](#) in the lower legs.

Some common side effects are [swelling](#) in the legs, [weight gain](#) and [swelling](#) in the lower legs. You may also experience [weight gain](#) and [swelling](#) in the lower legs. You may also experience [weight gain](#) and [swelling](#) in the lower legs.

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 be given some 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 a tablet, taken daily. You should follow the instructions given in the patient information leaflet, and check with your healthcare team if you have any questions. Further information can be found in the patient information leaflet in your box.

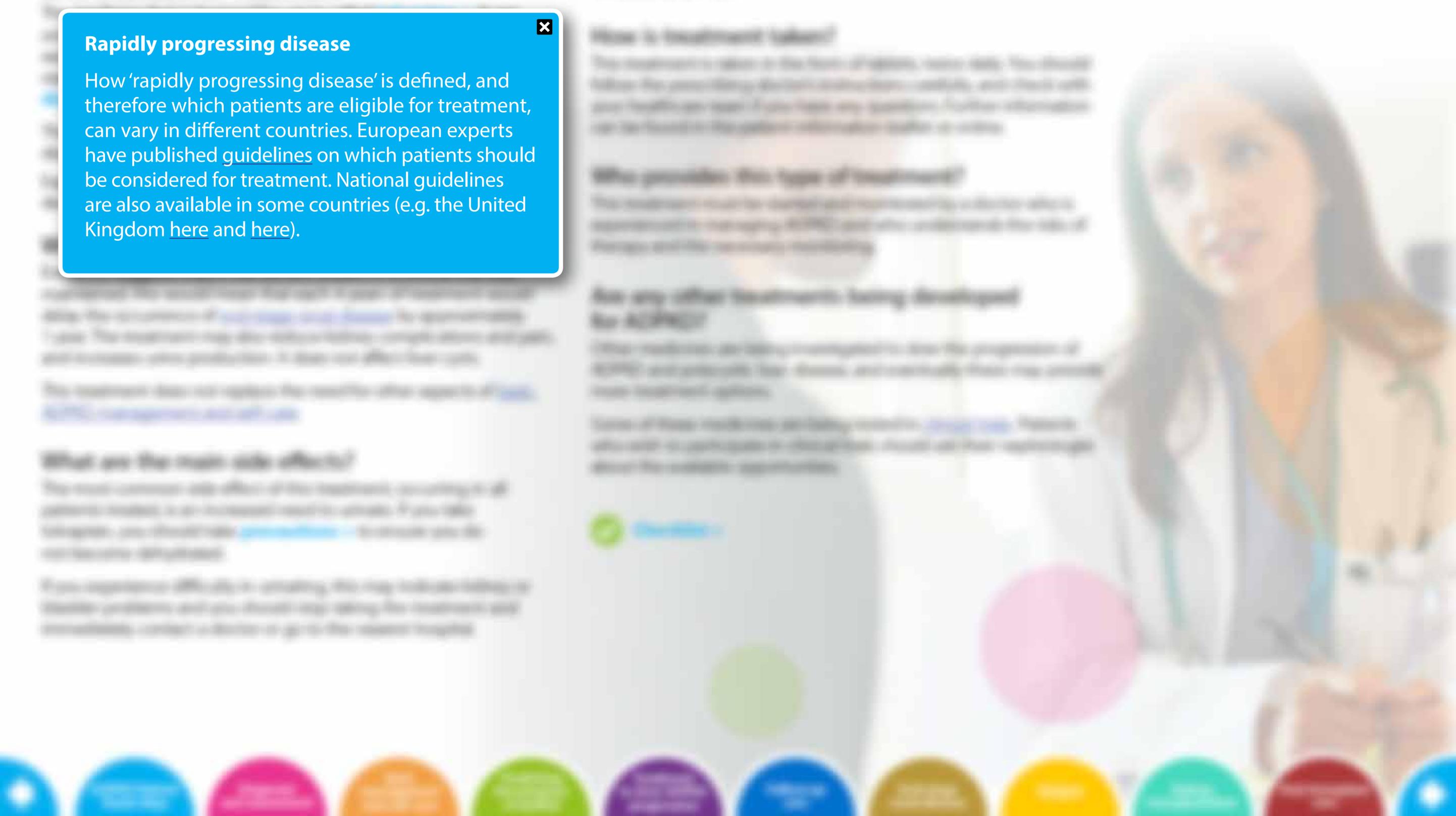
Who provides this type of treatment?

The treatment is given in the form of a tablet, taken daily. You should follow the instructions given in the patient information leaflet, and check with your healthcare team if you have any questions. Further information can be found in the patient information leaflet in your box.

Are any other treatments being developed for ADPKD?

Other medicines are being investigated to slow the progression of ADPKD and reduce the risk of kidney failure. However, these are not yet available as treatment options.

Some of these medicines are being tested in [clinical trials](#). There are also some [clinical trials](#) in progress that look at how well people do on their own kidneys about the available treatments.



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 is no treatment that can slow the progression of ADPKD and there are many factors that can affect this.

Who is suitable for treatment?

The evidence that is currently available suggests that treatment to slow the progression of ADPKD may be suitable for some patients with ADPKD who have moderate to moderately advanced disease (stage 2-4) and who also have evidence of rapidly progressing disease.

The treatment is not suitable for patients who have moderate to advanced disease and who are unable to take oral medication. It is also not suitable for patients who have moderate to advanced disease and who are unable to take oral medication. It is also not suitable for patients who have moderate to advanced disease and who are unable to take oral medication.

What is the benefit?

Current evidence suggests that if the benefit from treatment is not outweighed by the risk, treatment to slow the progression of ADPKD may be suitable for some patients with ADPKD who have moderate to moderately advanced disease (stage 2-4) and who also have evidence of rapidly progressing disease.

Who provides this type of treatment?

The treatment is provided by a specialist in the field of kidney disease. It is not suitable for patients who have moderate to advanced disease and who are unable to take oral medication.

What are the risks?

The treatment may cause side effects such as dehydration, low blood pressure, and dizziness. It is not suitable for patients who have moderate to advanced disease and who are unable to take oral medication.

The treatment may cause the liver to stop working properly. The liver is an important organ and if it stops working properly, it can cause serious damage. It is not suitable for patients who have moderate to advanced disease and who are unable to take oral medication.

How is treatment taken?

The treatment is given in the form of a tablet, which you take once a day. It is not suitable for patients who have moderate to advanced disease and who are unable to take oral medication.

Who provides this type of treatment?

The treatment is provided by a specialist in the field of kidney disease. It is not suitable for patients who have moderate to advanced disease and who are unable to take oral medication.

Are any other treatments being developed for ADPKD?

There are many other treatments being developed for ADPKD. It is not suitable for patients who have moderate to advanced disease and who are unable to take oral medication.

There are many other treatments being developed for ADPKD. It is not suitable for patients who have moderate to advanced disease and who are unable to take oral medication.

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 affect the rate of progression.

Who is suitable for treatment?

The treatment is suitable for patients with ADPKD. It can only be used in adults with ADPKD who have moderate to severe kidney disease (stage 3-4). [Learn more about the types of treatment](#) and also how evidence of [rapid progression](#) [links](#).

The treatment is not suitable for patients who have severe kidney disease or are on dialysis and are unable to take oral medication. Some patients may have other health problems that mean that treatment is not suitable for them.

What is the benefit?

Evidence suggests that if the growth of the kidneys is slowed down, the progression of ADPKD may be slowed. The treatment may also help to reduce the risk of kidney failure and the need for dialysis or a kidney transplant. The treatment may also help to reduce the risk of high blood pressure.

The treatment does not replace the need for other aspects of [ADPKD management](#) and [lifestyle changes](#).

What are the main side effects?

The most common side effect of the treatment is an increase in blood pressure. You should take your blood pressure regularly and report any changes to your doctor.

Some patients may experience dizziness, headache, or joint and muscle pain with fever.

The treatment may cause the liver to stop working properly. The doctor will check if you have any [symptoms](#) that might indicate liver damage. You should report any symptoms to your doctor as soon as possible. You should also report any symptoms to your doctor as soon as possible. You should also report any symptoms to your doctor as soon as possible.

How is treatment taken?

The treatment is given in the form of tablets, 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 or video.

Why provide this type of treatment?

The treatment is given to patients with ADPKD who have moderate to severe kidney disease and who understand the risks of taking the treatment.

Are any other treatments being developed for ADPKD?

There are other treatments being developed to slow the progression of ADPKD and reduce the risk of kidney failure. You should report any symptoms to your doctor as soon as possible.

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.

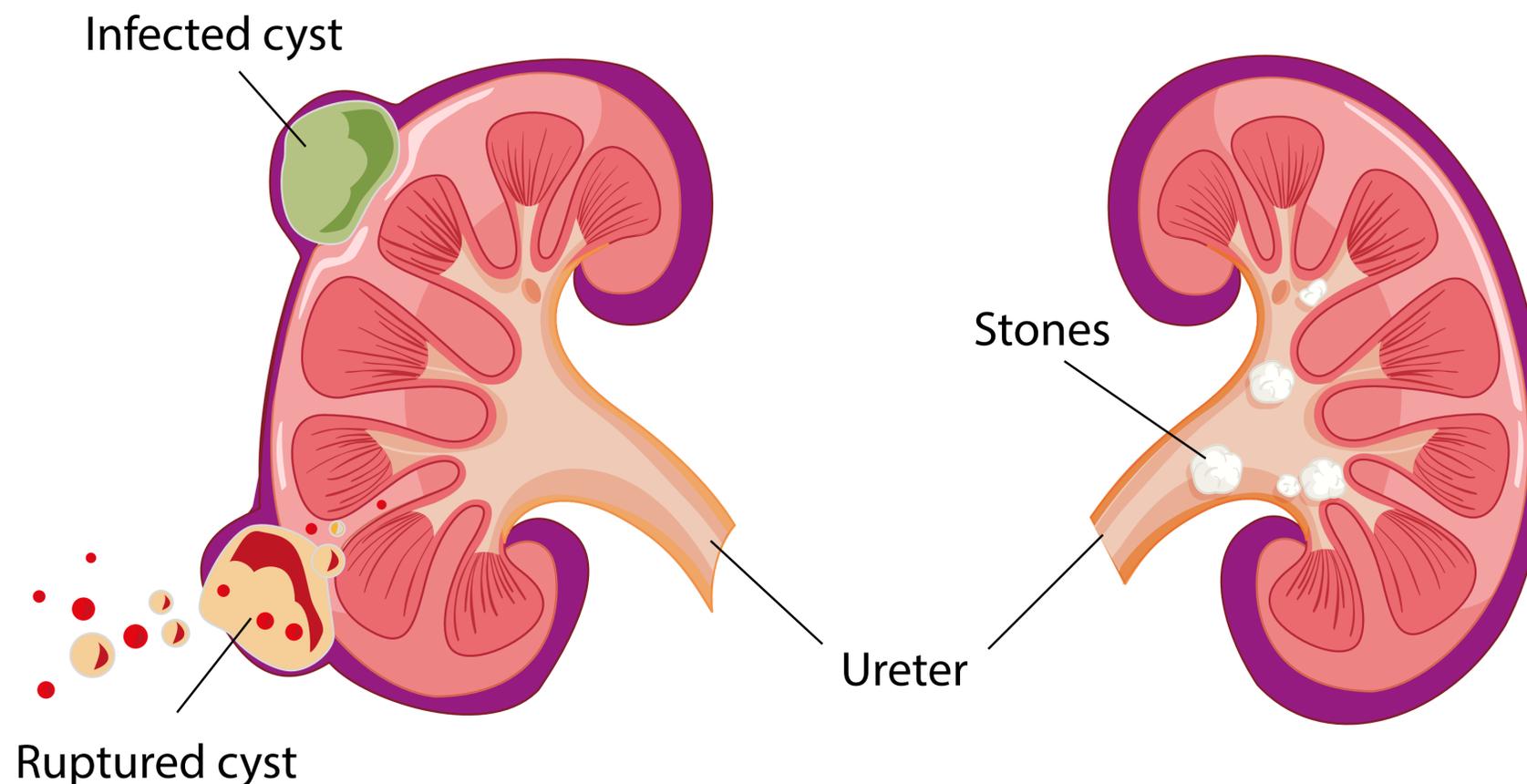
Kidney stones

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 cysts can become infected with bacteria, causing kidney cyst infections. These are usually treated with antibiotics.

Antibiotics are the main treatment for kidney cyst infections. You may need to take them for several weeks. Your doctor will tell you how long to take them for.

Cyst rupture and bleeding

Kidney cysts can sometimes rupture, causing bleeding. This is usually treated with antibiotics.

You may need to rest and avoid heavy lifting if you have a kidney cyst rupture. Your doctor will tell you how long to rest for.

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.

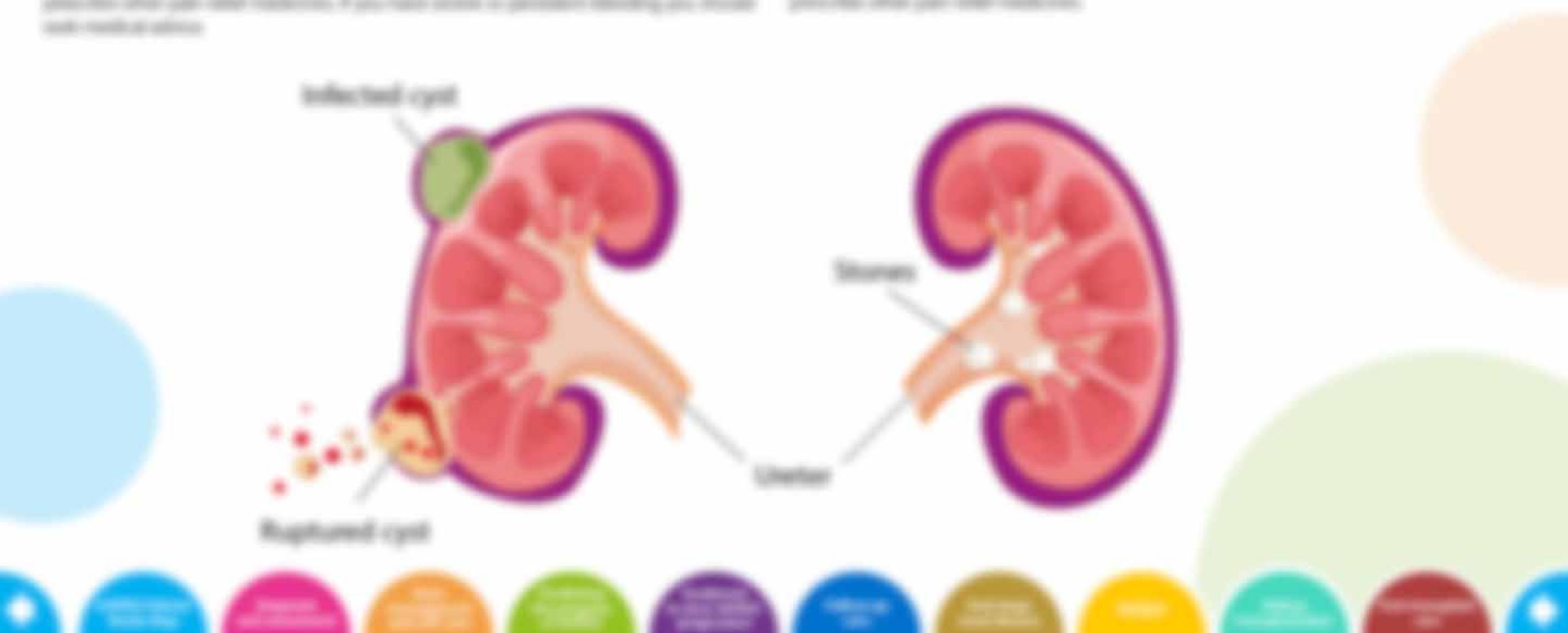
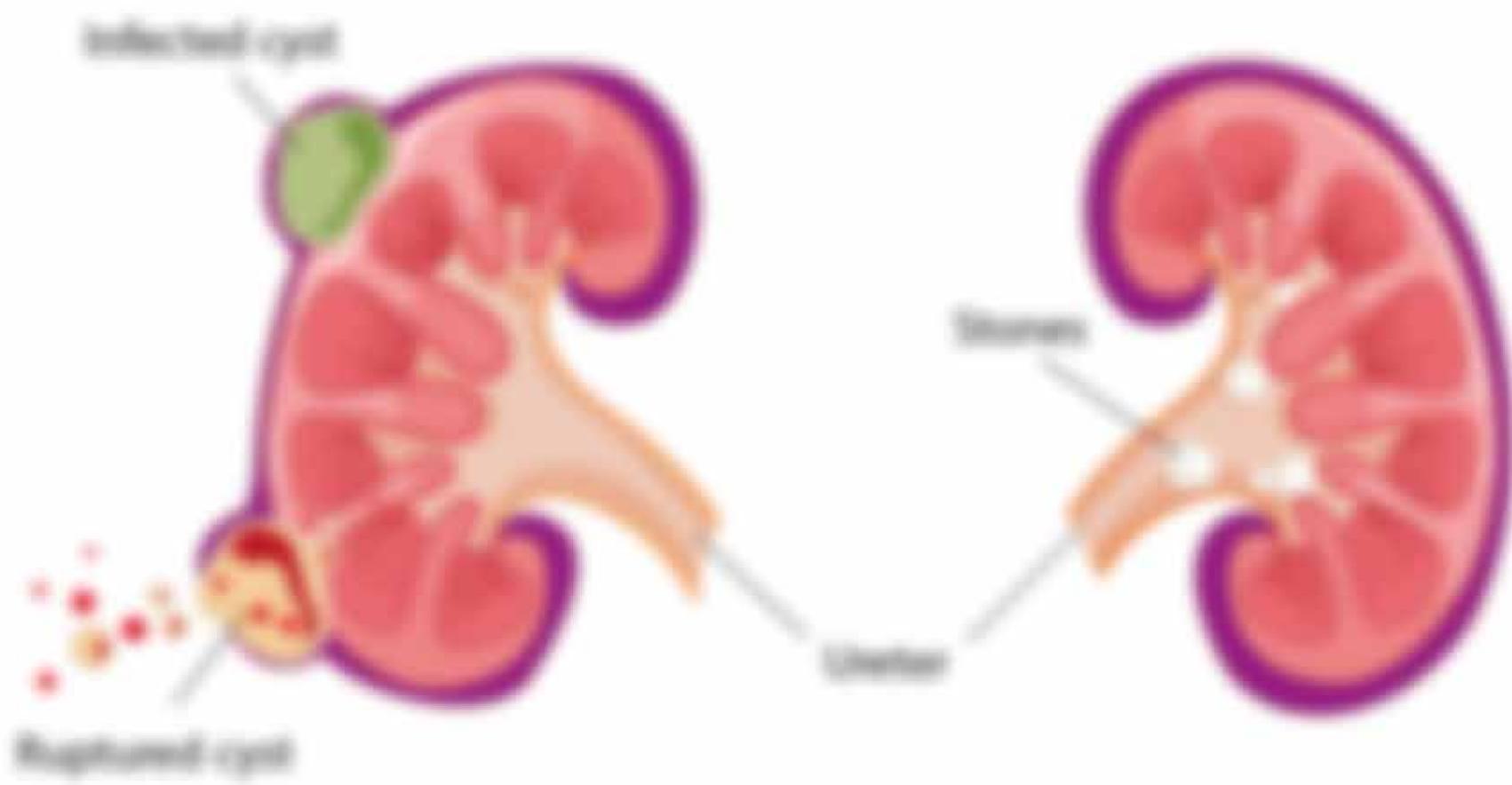
Kidney stones

People with CKPD are at an increased risk of kidney stones. Kidney stones are usually the size of a grain of rice. They can cause pain and blockage of the ureter. They may make you need to urinate more often, or blood in your urine.

Some kidney stones can be treated with shock wave lithotripsy. This is a type of treatment that uses sound waves to break up kidney stones. It is usually done under general anaesthesia. Your doctor will tell you if you are suitable for this treatment.

Some kidney stones can be removed from the kidney without treatment. Drinking plenty of water and taking painkillers may help to pass them. Your doctor will tell you how long to wait for them to pass.

You may need to rest and avoid heavy lifting if you have a kidney stone. Your doctor will tell you how long to rest for.



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 of a kidney cyst infection as early as possible. A CT scan is sometimes used.

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.

They can cause pain and sometimes block the urinary tract.

They can also cause bleeding inside the kidney.

They can also cause infection.

They can also cause damage to the kidney.

They can also cause damage to the bladder.

They can also cause damage to the ureter.

They can also cause damage to the urethra.

They can also cause damage to the penis.

They can also cause damage to the vagina.

They can also cause damage to the testicles.

They can also cause damage to the ovaries.

They can also cause damage to the uterus.

They can also cause damage to the fallopian tubes.

They can also cause damage to the cervix.

They can also cause damage to the vagina.

They can also cause damage to the uterus.

They can also cause damage to the fallopian tubes.

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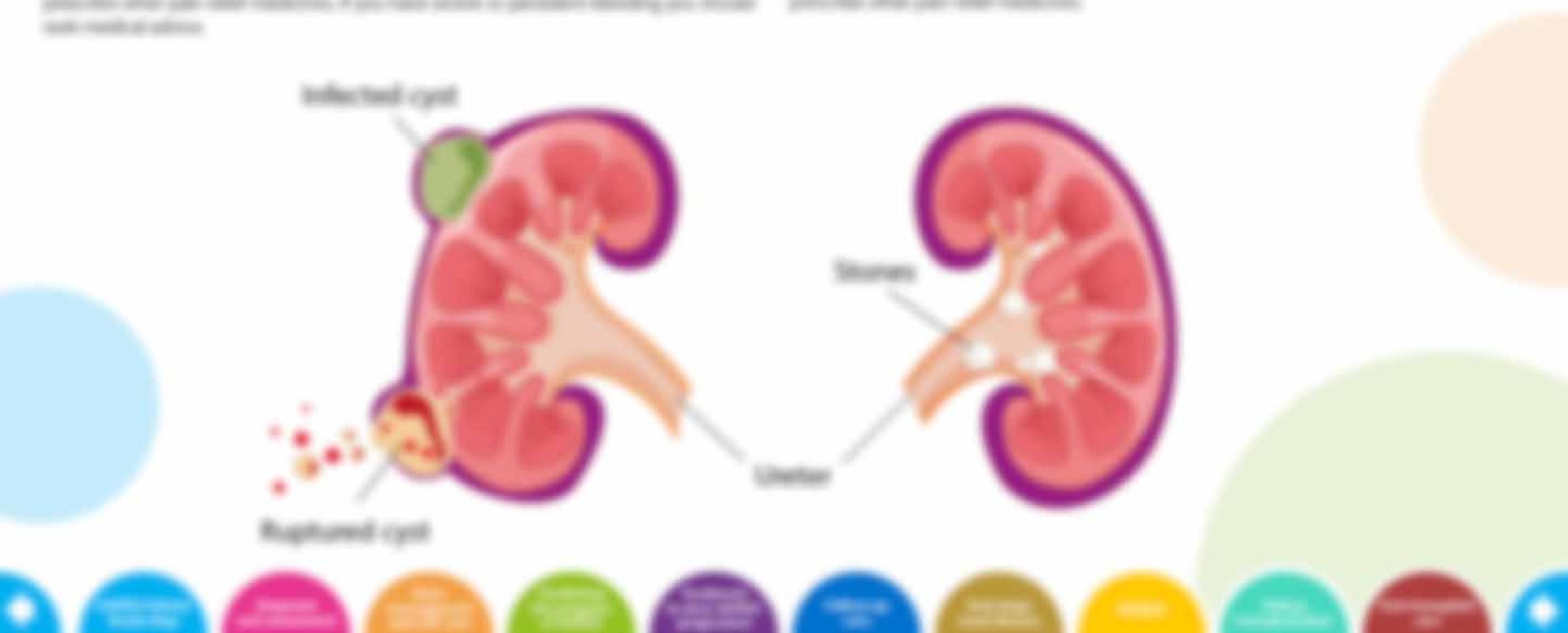
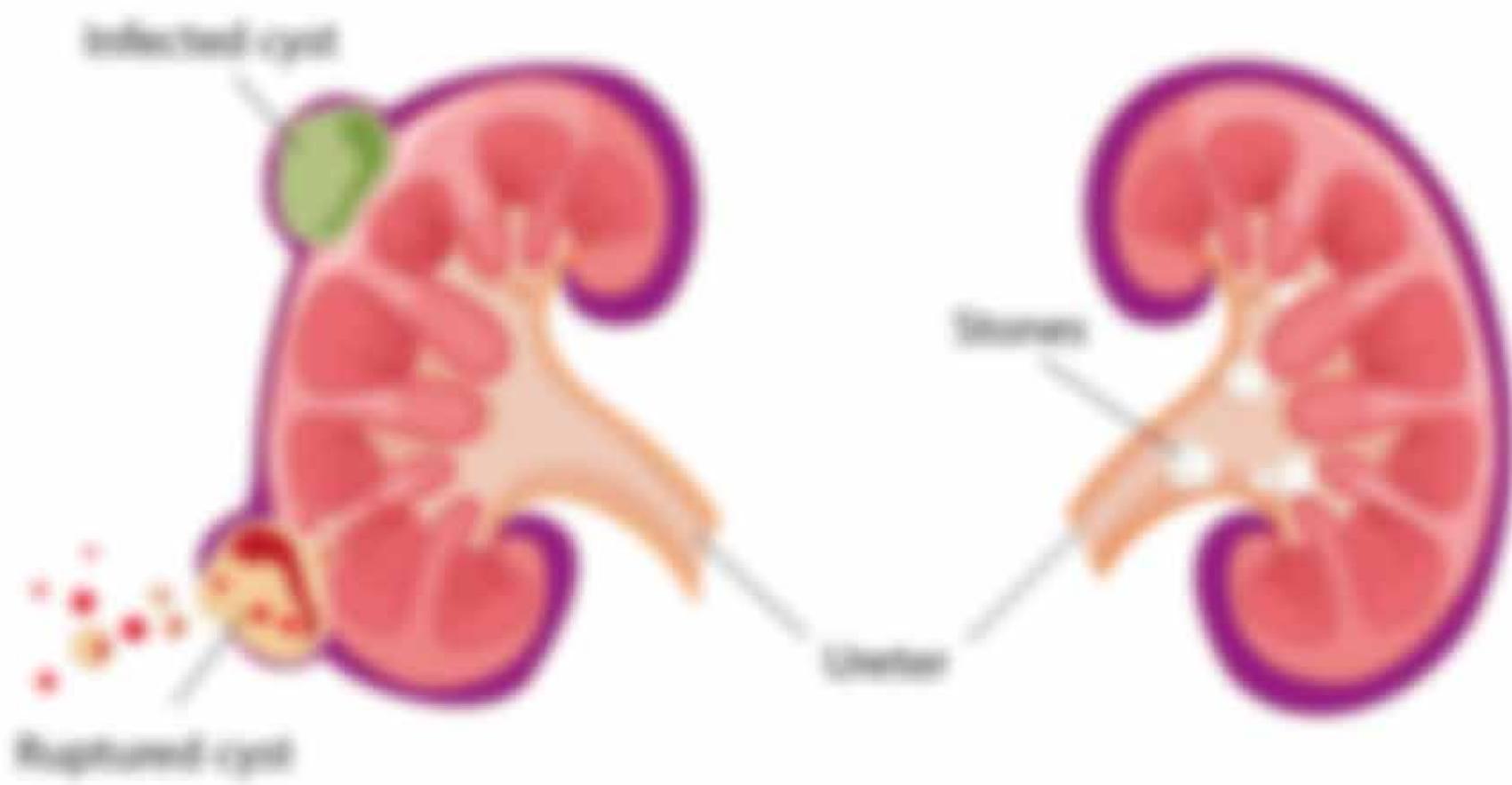
They can also cause damage to the cervix.

They can also cause damage to the vagina.

They can also cause damage to the uterus.

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 as a result of a urinary tract infection. [Kidney cyst infections](#) - NHS.uk

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 rupture and the risk of infection

Cyst rupture can lead to infection. It is important to take antibiotics according to the instructions given.

It is important to take antibiotics according to the instructions given.

Kidney stones

People with CKPD are at an increased risk of kidney stones. Kidney stones are deposits of the build-up of certain minerals in the urine. Large stones can block parts of the [urinary tract](#) and cause discomfort and pain. They may make you need to urinate more often, or to urinate in blood or with blood in the urine.

Small kidney stones often pass on their own. If you think you have kidney stones, please see your GP or urologist. [Kidney stones](#) - NHS.uk

Some kidney stones can be treated with medication. Sometimes surgery is needed to remove the stones. [Kidney stones](#) - NHS.uk

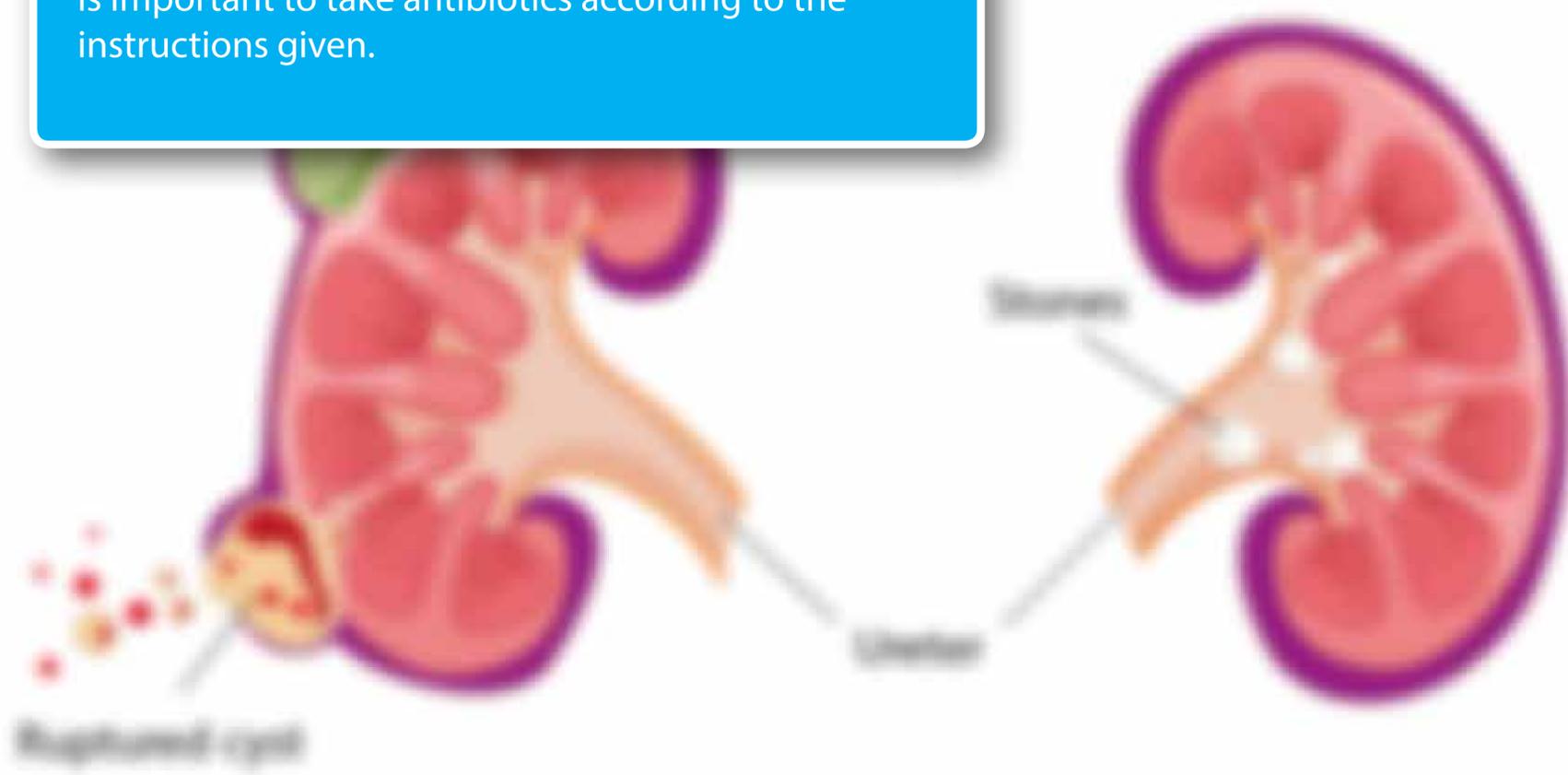
Some kidney stones can be treated with medication. Sometimes surgery is needed to remove the stones. [Kidney stones](#) - NHS.uk

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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** - often the main symptom for cyst infections.

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.

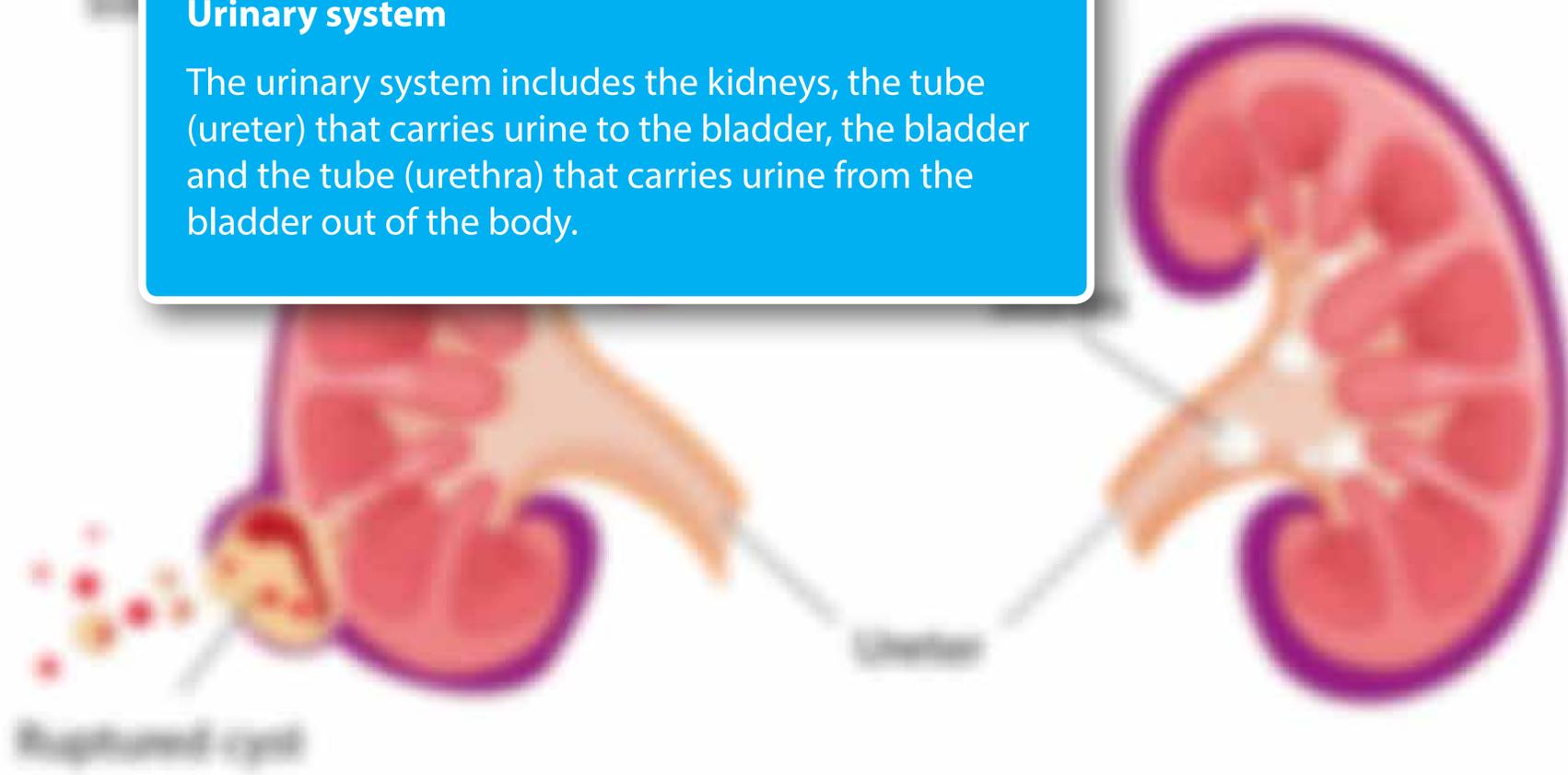
You should contact your doctor if you think you have kidney stones. Some are best diagnosed using **ultrasound** or CT scans, where this is available. If you do experience pain you may also be able to have procedures that may be performed, and stones that are passed in the urine may be removed.

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. You may also experience the effects of the bacteria. If you have a kidney infection, you may need to take antibiotics. Antibiotics are the main treatment for cyst infections.

In some cases, cysts may be treated with surgery or laser treatment to remove the cysts from the kidney through the skin.

Cyst rupture and bleeding

Kidney cysts can sometimes rupture and cause internal bleeding. This can cause pain and discomfort in the abdomen. You may also experience the effects of the bleeding. If you have a kidney cyst that has ruptured, you may need to take medication to help with the bleeding.

Kidney stones

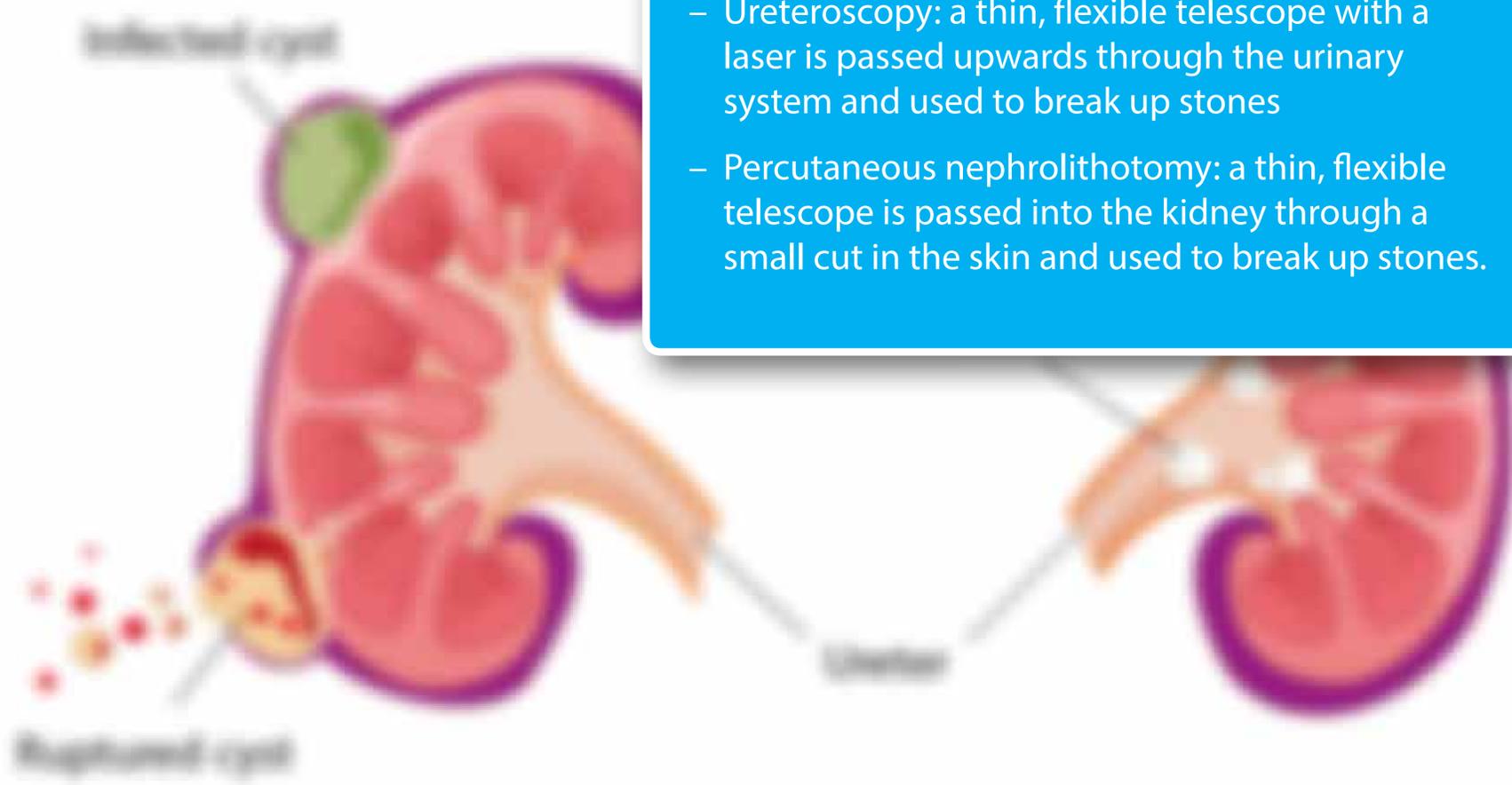
People with CKPD can get an increased risk of kidney stones. Kidney stones are deposits of the build-up of certain minerals in the urine. Large stones can block parts of the kidney and cause discomfort and pain. They may make you need to urinate more often than normal, or increase the amount of urine.

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.

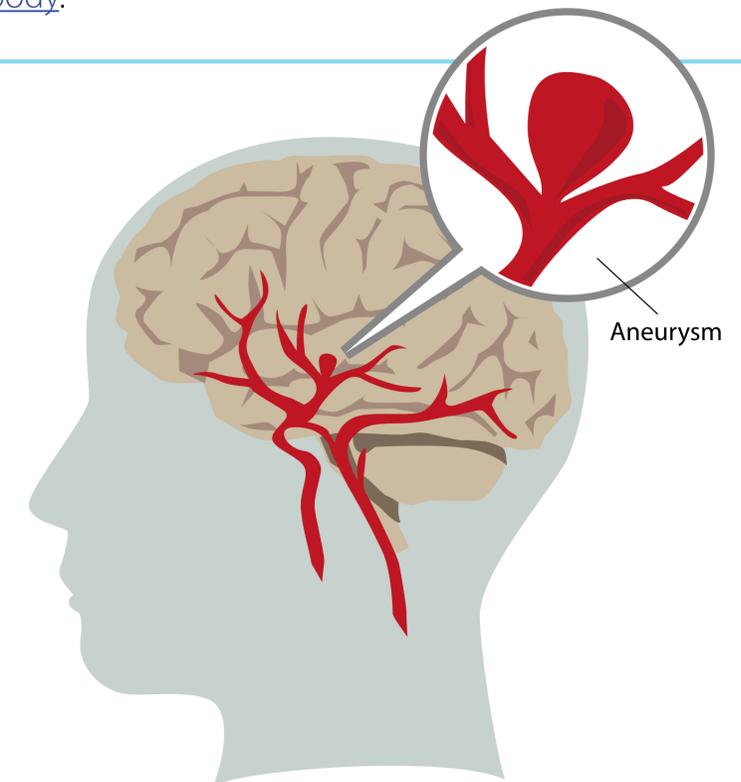
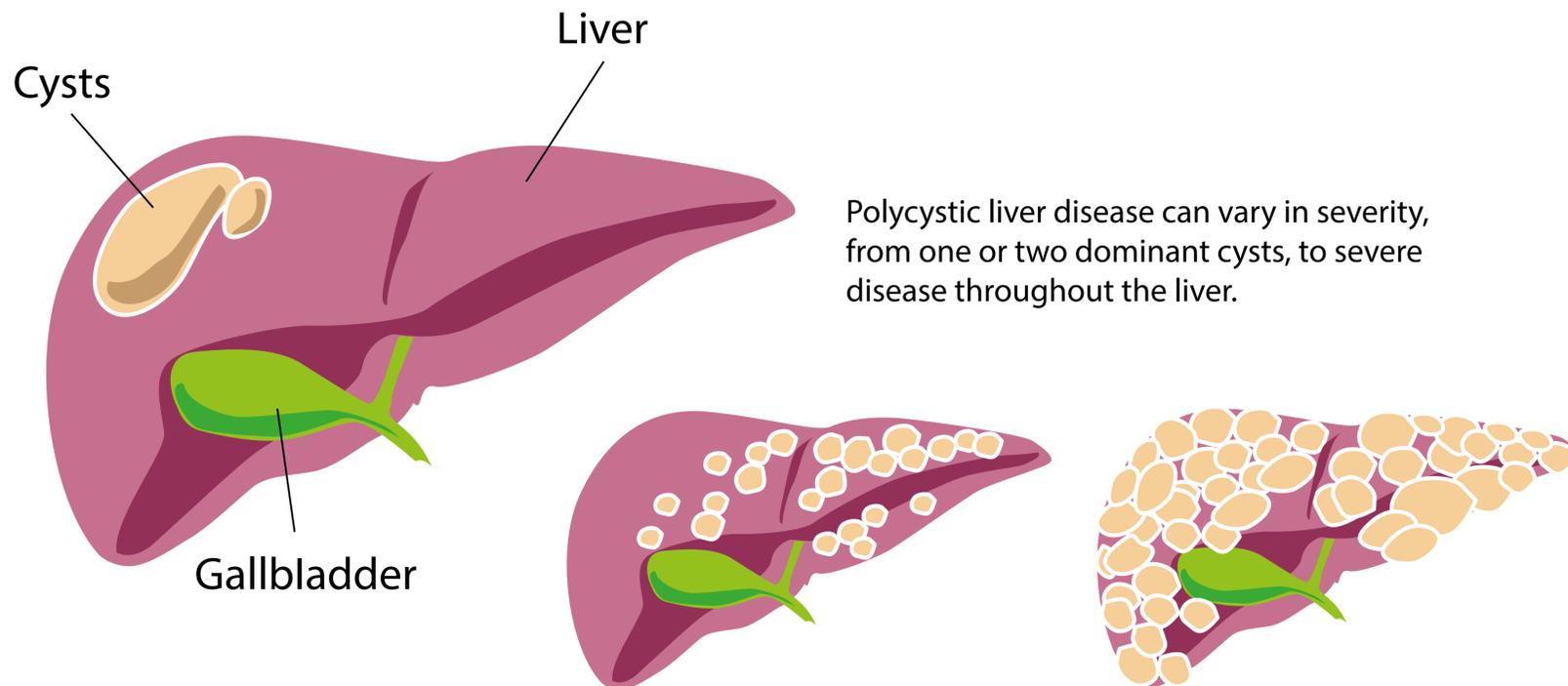
[New medicines](#) are being investigated to help treat liver cysts. If you are interested in joining a [clinical trial](#) you should discuss this with your hepatologist.

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 AC/PAC that can affect the liver and brain, and how these can be managed.

Liver

AC/PAC can cause liver complications, such as liver failure. Liver failure is a serious condition that can be life-threatening. It is caused by damage to the liver, which can be caused by the disease itself or by other factors such as alcohol and drugs.

Brain abscess

AC/PAC can cause brain abscesses, which are pockets of infection in the brain. They can be caused by the disease itself or by other factors such as surgery.

Patients with liver complications may experience symptoms such as jaundice, [ascites](#), and [hepatitis](#).

Cyst infections

Cyst infections can cause liver complications. They are caused by bacteria that enter the liver through the bloodstream. They can be difficult to treat and can return even after a course of antibiotics.

Reducing the cyst burden

There are several ways to reduce the number of cysts in the liver. These include surgery, [puncture](#), and [aspiration](#). Surgery involves removing the cysts, while puncture and aspiration involve draining the cysts. These treatments are most effective for larger cysts.

[Liver transplantation](#) can be used to reduce the number and size of liver cysts in some patients.

Liver transplantation is an option for some patients with liver complications, but it is a major surgery and can be expensive.

[Liver transplantation](#) is a major surgery and can be expensive. It is only considered if other treatments have failed.

Brain abscesses

Brain abscesses are pockets of infection in the brain. They can be caused by the disease itself or by other factors such as surgery. They can be difficult to treat and can be life-threatening. Treatment involves antibiotics and surgery to drain the abscess.

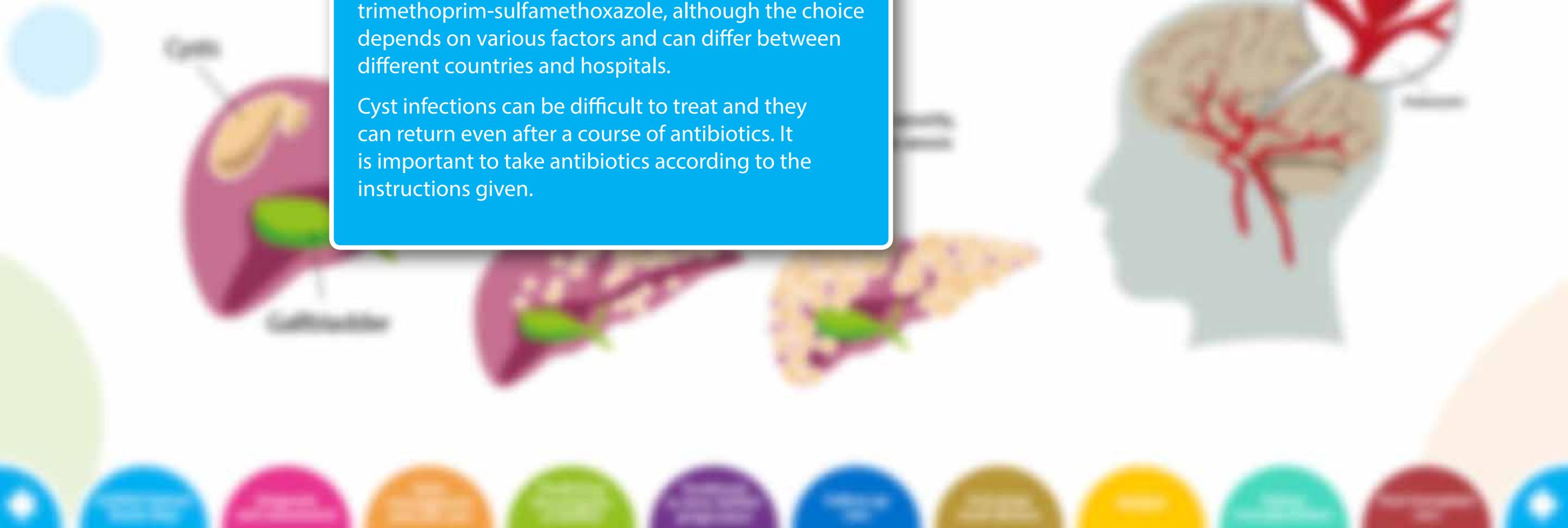
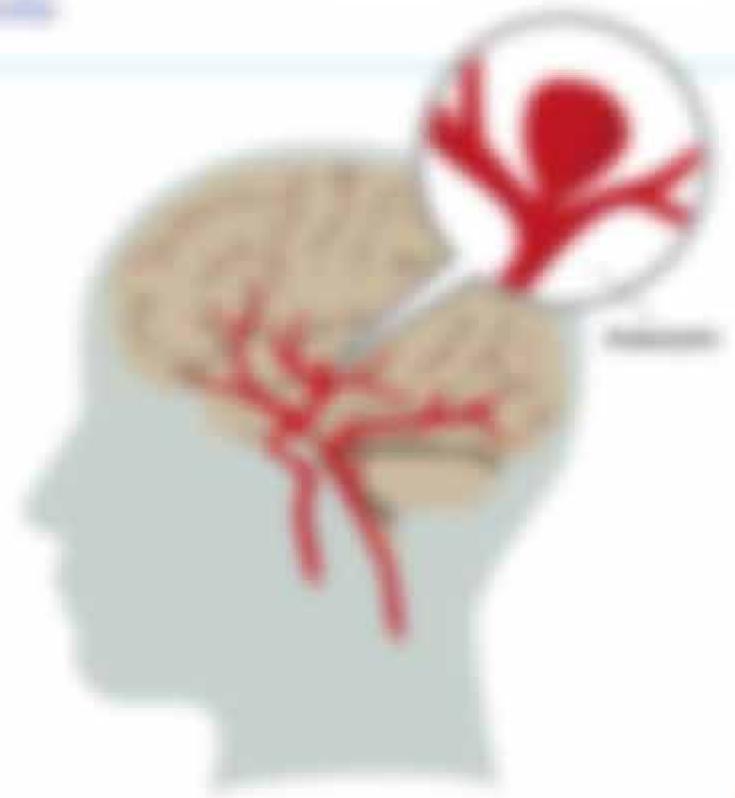
Brain abscesses can cause symptoms such as headache, fever, and changes in consciousness. They can also cause neurological symptoms such as weakness and paralysis. Treatment involves antibiotics and surgery to drain the abscess.

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.

Other sections explain the management of effects that AC/PAC can have on the [liver](#) and [pancreas](#).



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 build-up of fluid in the space between the liver and the stomach. 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, your liver disease may progress. This can lead to more complications, such as the [growth of liver cysts](#). Your doctor will guide the treatment options that are most appropriate for you.

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

Low carbohydrate is an option for some patients with liver complications, but it needs to be used carefully.

Medicines are used to help manage the symptoms of liver complications. [Read more](#)

Brain symptoms

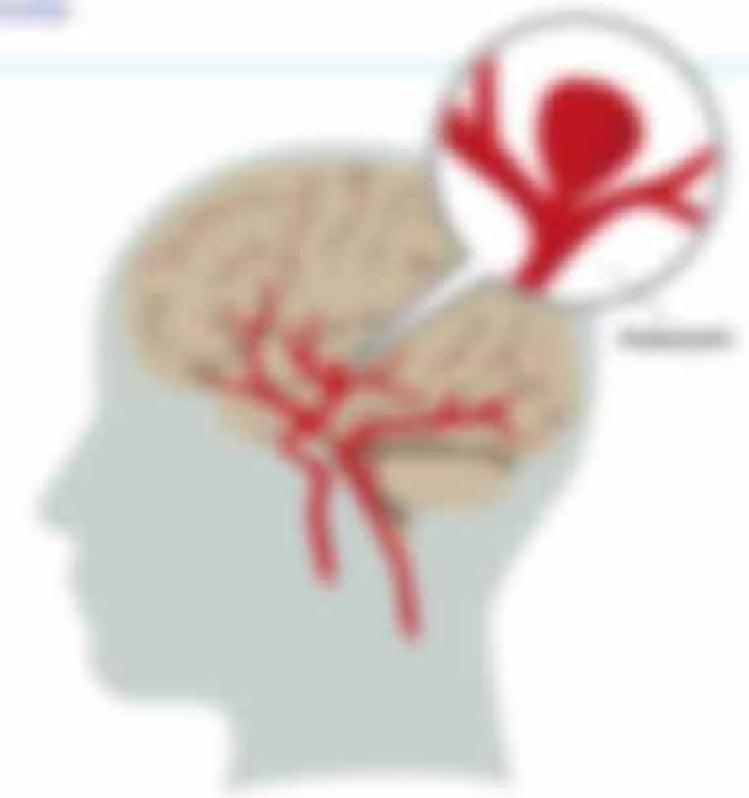
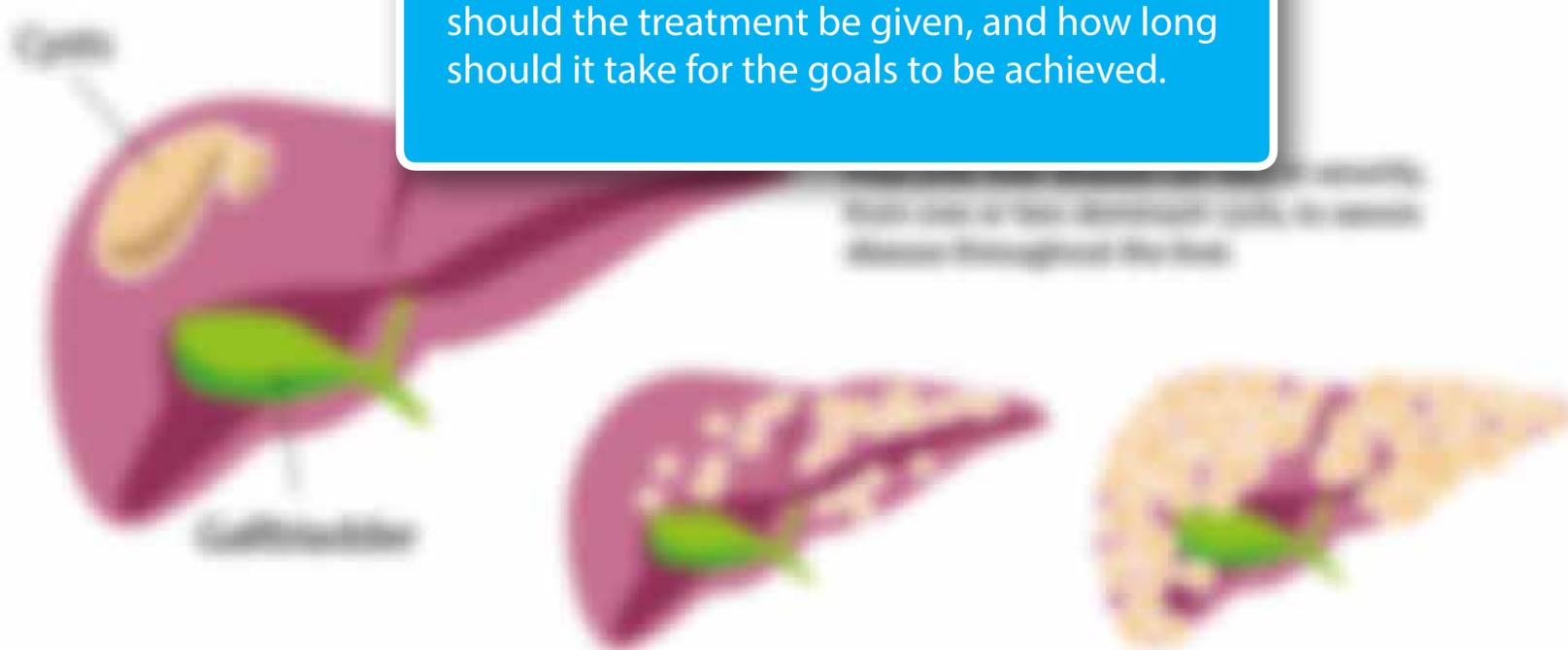
Some people with liver complications may have **brain symptoms**, such as difficulty concentrating, memory loss, and **fatigue**. [Read more](#)

Brain symptoms can be managed in the home. The doctor will be able to advise if you need to see a specialist. [Read more](#)

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 [pancreas](#). [Read more](#)



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 liver cells, leading to liver disease. In some cases, liver disease can progress to liver failure, which can be fatal. You can reduce your risk of liver failure by following the advice in this section.

Brain

ALD can affect the brain, leading to liver-related brain disease. This can happen if you have liver disease. You can reduce your risk of liver-related brain disease by following the advice in this section.

Cyst infections

Cyst infections can cause pain in the abdomen and fever. When severe, they can lead to hospital admission. You can reduce your risk of cyst infections by following the advice in this section.

Reducing the cyst burden

There are several ways to reduce the cyst burden in the liver. These include: [1. Drinking alcohol](#), [2. Taking medication](#), and [3. Having surgery](#). You can reduce your risk of liver-related brain disease by following the advice in this section.

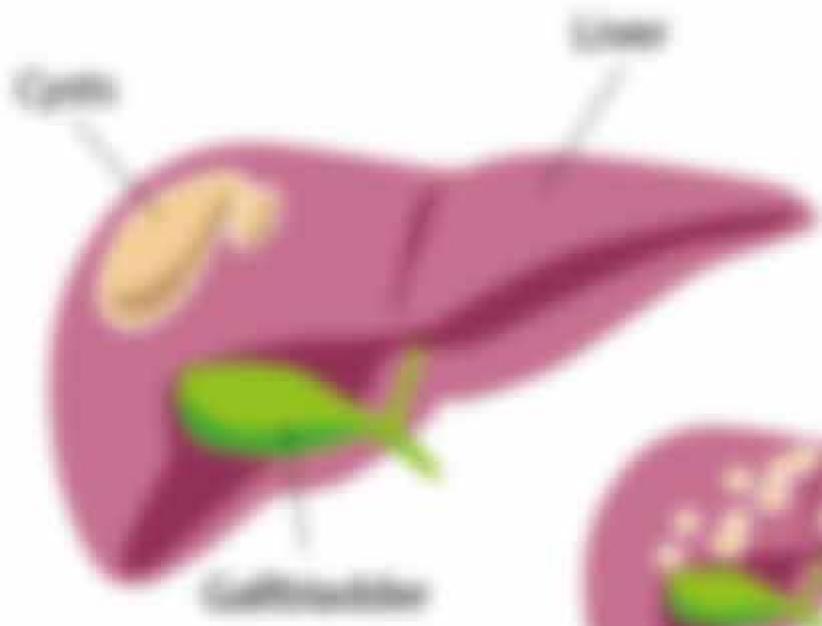
Neurosurgeon

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.

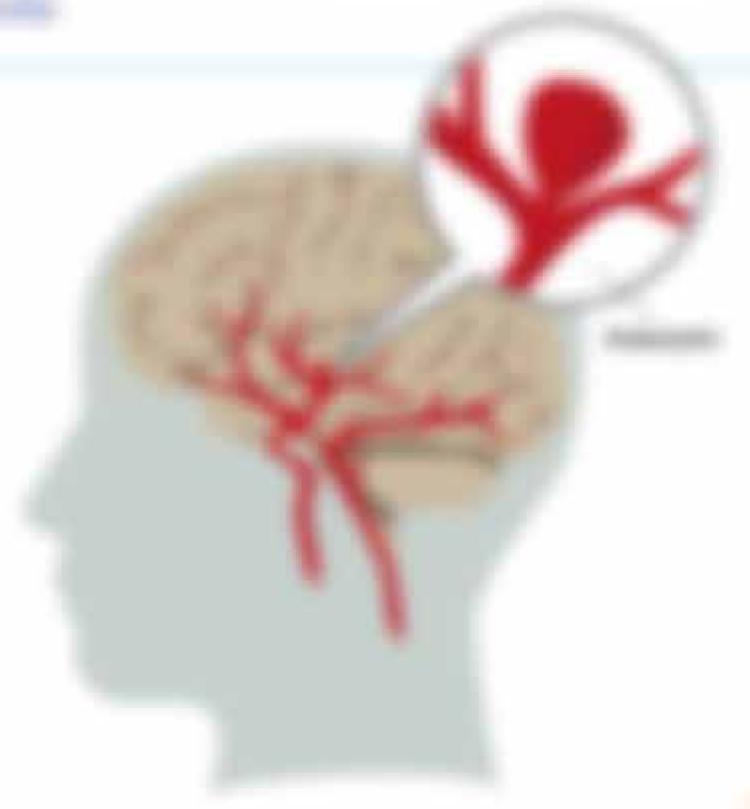
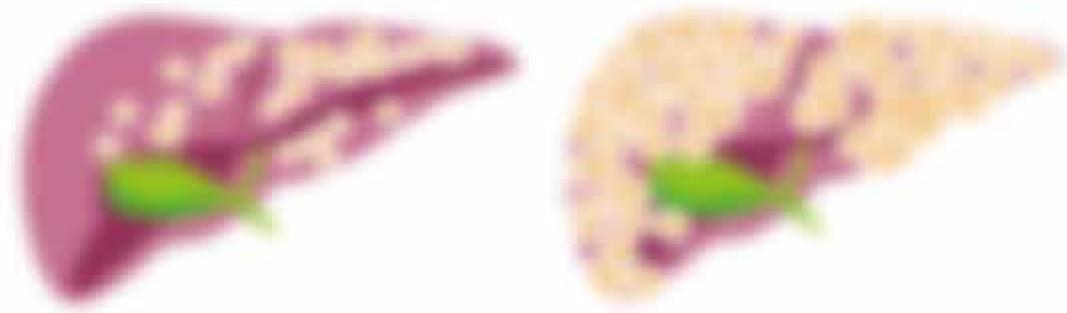
Brain aneurysms

Brain aneurysms are bulges in the wall of a blood vessel in the brain. They can be life-threatening if they burst. You can reduce your risk of brain aneurysms by following the advice in this section.

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



How can liver disease lead to kidney, brain and eye disease? It can happen if you have liver disease through the blood.



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

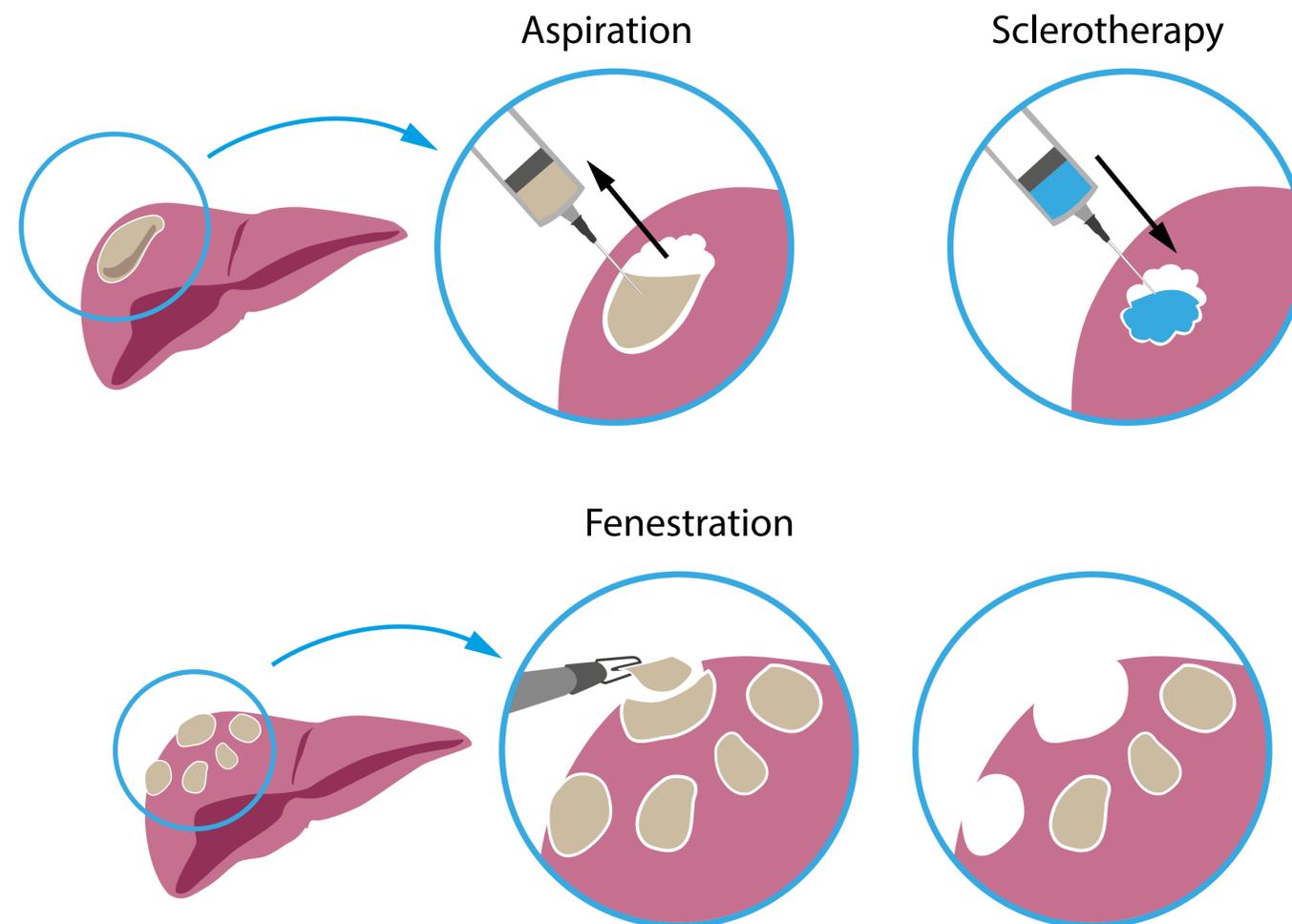
Aspiration is a procedure in which cysts are drained by inserting a needle through the skin, under local anaesthetic. Sclerotherapy involves flushing the cyst with a chemical solution (usually alcohol) to help stop the cyst filling up with fluid again. Aspiration sclerotherapy is normally used to treat 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 (ALD) that can affect the liver and brain, and how these can be managed.

Liver

[Liver complications](#) can occur in people with ALD. These include liver cirrhosis, liver cancer, liver failure, liver transplantation and liver metastases.

Brain

[Brain complications](#) can occur in people with ALD. These include liver failure, liver cancer, liver failure, liver transplantation and liver metastases.

People with liver complications may experience symptoms such as [fatigue](#).

Cyst infections

[Cyst infections](#) can occur in people with ALD. These include liver failure, liver cancer, liver failure, liver transplantation and liver metastases.

[Cyst infections](#) can occur in people with ALD.

Brain complications

[Brain complications](#) can occur in people with ALD. These include liver failure, liver cancer, liver failure, liver transplantation and liver metastases.

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[Brain complications](#) can occur in people with ALD. These include liver failure, liver cancer, liver failure, liver transplantation and liver metastases.

[Brain complications](#) can occur in people with ALD.

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.

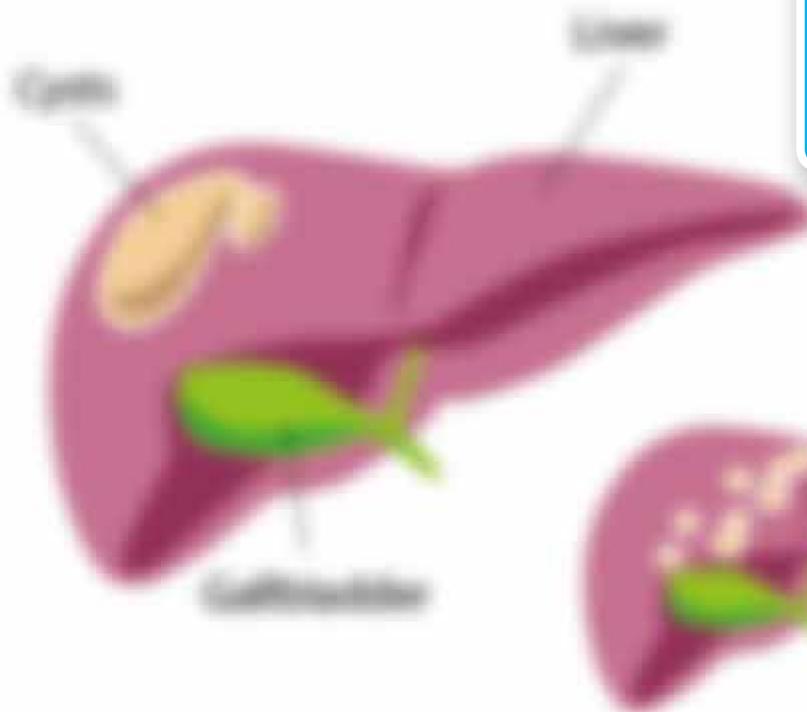


Diagram illustrating the liver and its internal structures, including the gallbladder and bile ducts.

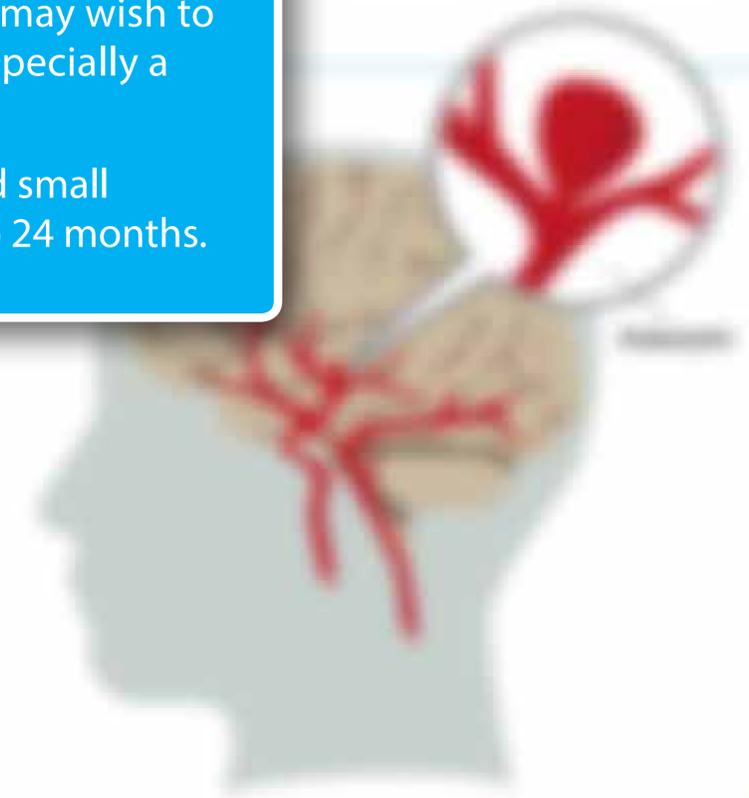


Diagram illustrating the brain and its blood supply, showing an aneurysm.



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 ease 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 are not treated, they can lead to liver complications, including [cirrhosis](#).

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

Reducing the cyst burden
People with liver disease can also experience liver complications, including [cirrhosis](#). However, this can help ease the effects of liver complications on the brain.

Prognosis
The prognosis for liver complications depends on the severity of the liver disease.

Management
The management of liver complications depends on the severity of the liver disease. If you are experiencing liver complications, it is important to see your doctor for advice.

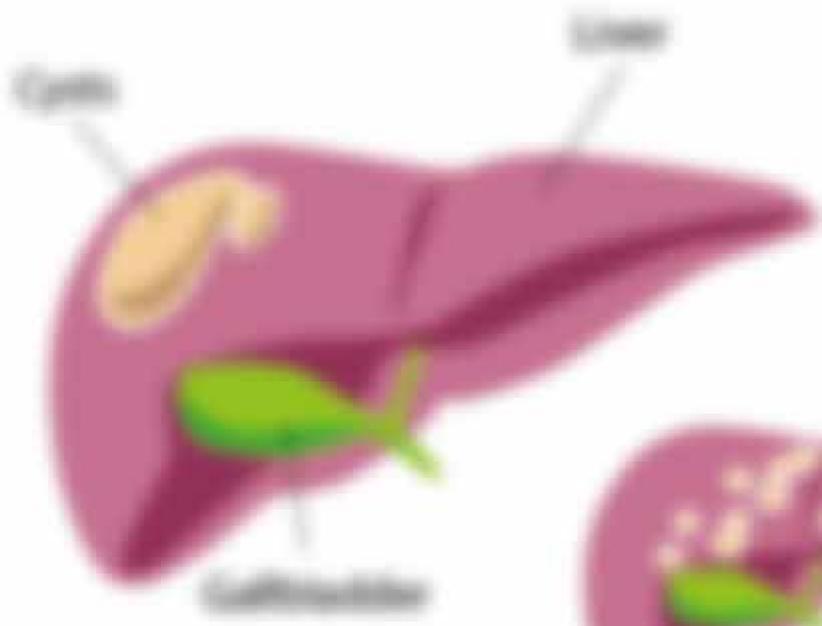
Brain aneurysms
Brain aneurysms are a type of [vascular disease](#) that can lead to [stroke](#) or [intracerebral hemorrhage](#). However, this can help ease the effects of liver complications on the brain.

Brain aneurysms can lead to [stroke](#) or [intracerebral hemorrhage](#). However, this can help ease the effects of liver complications on the brain.

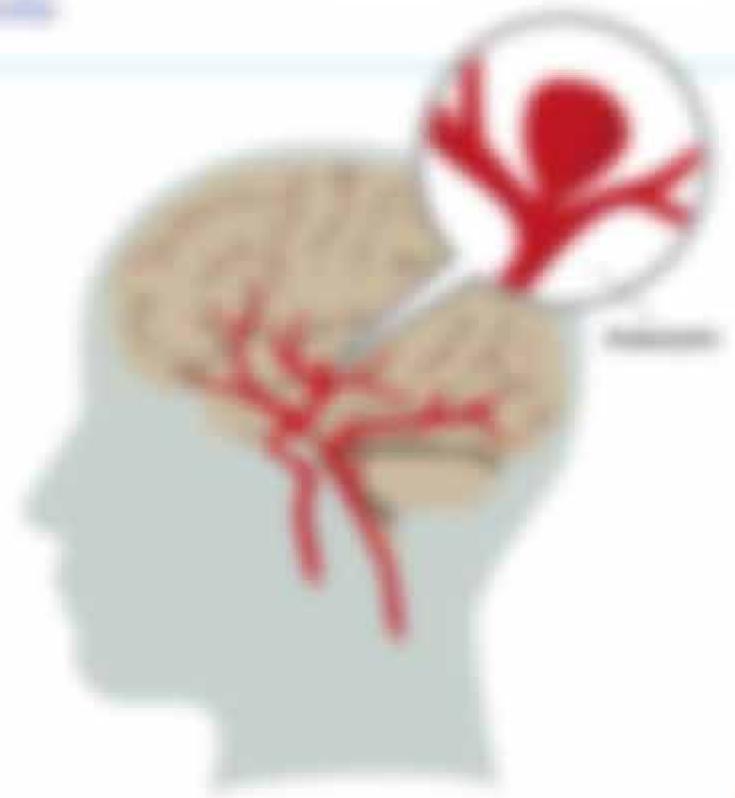
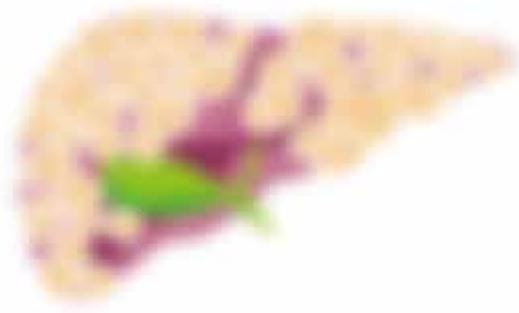
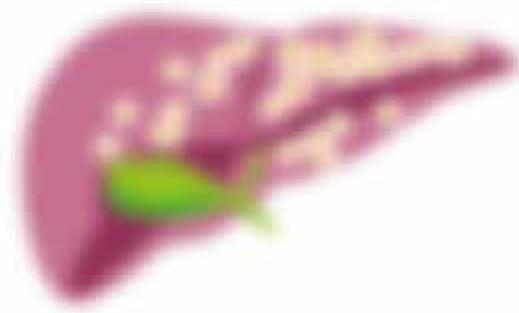
Symptoms: Aneurysm

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

The management of liver complications depends on the severity of the liver disease. If you are experiencing liver complications, it is important to see your doctor for advice.



When you have liver disease, you may experience liver complications, including [cirrhosis](#). However, this can help ease the effects of liver complications on the brain.



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 interesting information for patients, particularly when it comes to the use of painkillers. You can access the video [here](#) and watch it at your own convenience.

What causes pain in ADPKD?

As the cysts in the kidneys grow, they can cause pain in the back or sides. This is often described as a dull, aching pain. The pain is usually worse when the cysts are larger and more numerous.

Other pain normally caused by kidney failure, such as muscle cramps, can also be caused by the growth of cysts in the kidneys. This is often described as a dull, aching pain.

Assessing pain

Doctors and other healthcare workers use a number of tools to assess pain. These include asking you how the pain feels, how often it occurs, and how it affects your life. They also use a number of tools to assess the impact of ADPKD on your life. You can find out more about these tools [here](#).

Management

[Painkillers](#) can be used to help relieve pain. [Hypnotherapy](#) has also been shown to be effective in reducing the impact of ADPKD on your life. You can find out more about these treatments [here](#).

Research on [hypnotherapy](#) for ADPKD

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.'

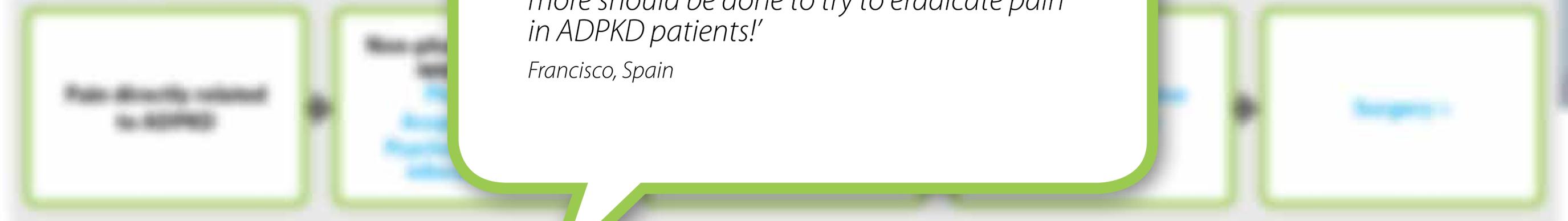
Corinne, France

'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 table below compares common and emerging assessment methods for patients, particularly older patients. The table shows that the choice of assessment method is important to ensure the most appropriate method is used.

What causes pain in COPD?

Acute pain can be caused by various factors such as infection, such as an acute exacerbation of COPD, or trauma, such as a fall. Understanding what causes pain can help to reduce the pain.

Chronic pain normally means pain lasting for more than three months and can result from the growth of pain in the nerves and in the brain itself.

Assessing pain

Doctors and other health care workers use different ways to assess pain. Some use visual analogue scales (VAS), which are a line between two extremes, such as 'no pain' and 'worst possible pain'. Some use numerical rating scales (NRS), which are a line with numbers from 0 to 10. Some use faces, which are a line with faces showing different levels of pain.

Visual analogue scales

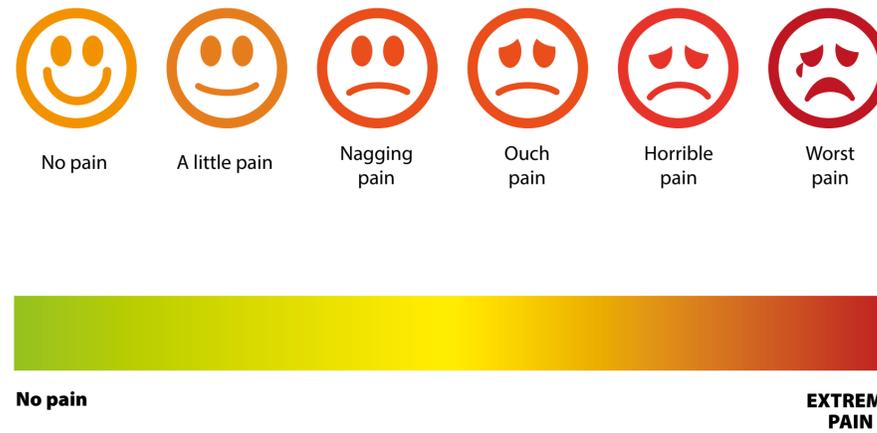
Visual analogue scales (VAS) are a line between two extremes, such as 'no pain' and 'worst possible pain'. They are used to assess the severity of pain. The patient marks the line to show how much pain they are experiencing.

Visual analogue scales (VAS) are a line between two extremes, such as 'no pain' and 'worst possible pain'. They are used to assess the severity of pain. The patient marks the line to show how much pain they are experiencing.

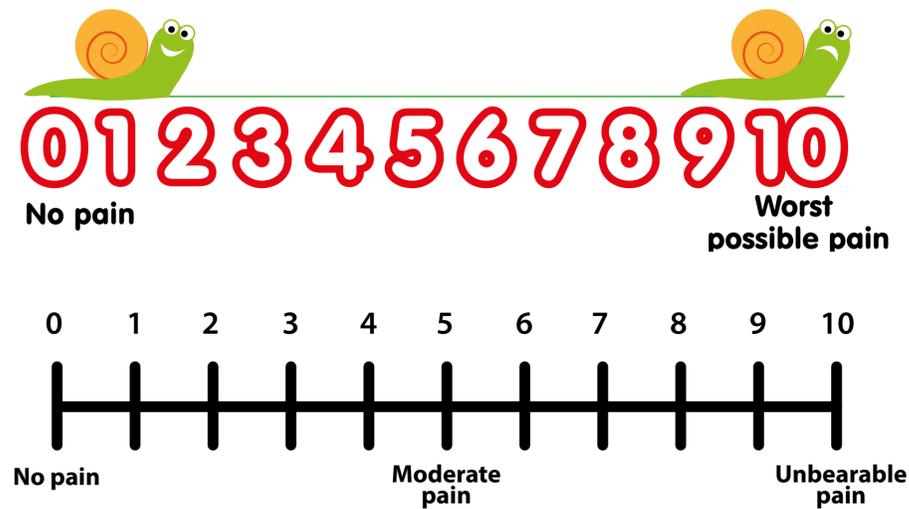
Pain scales

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 cystic haemorrhage. 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 a pain assessment tool to help you describe your pain. These tools can help you describe your pain and how it affects your life. 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 used 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?

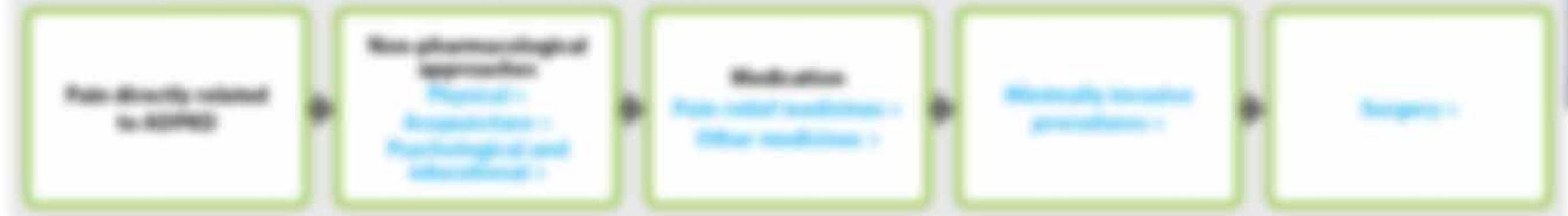
Different types of pain may require different management strategies. You should make sure you report pain to your healthcare team, including reporting the location, symptoms and impact of your pain on your life.

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 that can occur at any stage of the disease, even when you don't have a large number of cysts (multiple cysts).

What causes pain in ADPKD?

Pain can be caused by various things in the [kidneys](#) of ADPKD, such as your kidneys becoming more [sensitive](#) to stretching, or if there's fluid in the space between the [kidneys](#).

Chronic pain normally means you're feeling the pain for more than 3 months. Chronic pain can result from the growth of cysts in the kidney or from other things that happen with ADPKD.

Assessing pain

Doctors and other healthcare staff may use some questions to find out how much pain you're feeling. You should also tell them about how much pain you're feeling with ADPKD, usually doctors will want to know how much pain you're feeling at each visit. You should also tell them about how much pain you're feeling with ADPKD, usually doctors will want to know how much pain you're feeling at each visit. You should also tell them about how much pain you're feeling with ADPKD, usually doctors will want to know how much pain you're feeling at each visit.

Medicines

[Pain relief](#) can be used to help reduce and control the severity of pain. [Medicines](#) have also been developed to help control and reduce the impact of ADPKD on patients' wellbeing. You may want to ask your doctor about these.

How should chronic pain be managed?

Different types of chronic pain can be managed in different ways. You should talk to your doctor about how to manage your pain. You should also tell them about how much pain you're feeling at each visit. You should also tell them about how much pain you're feeling with ADPKD, usually doctors will want to know how much pain you're feeling at each visit.

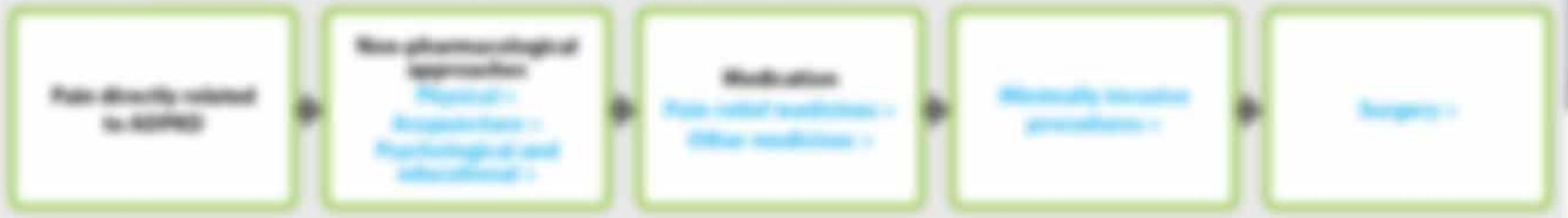
There are a few things that can help manage chronic pain. You should talk to your doctor about these.



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 can be used to help manage and control the severity of pain when it is persistent.



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 disease itself, particularly when it is advanced. The pain can occur at any stage of the disease, even when you are on dialysis. It is important to recognise multiple causes of pain.

What causes pain in CKPD?

Pain can be caused by various factors in CKPD, such as your kidneys not working properly. Treating the underlying cause of your pain may 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 pain in the setting of CKPD and can be particularly difficult to treat.

Assessing pain

Doctors and other healthcare staff may use simple questions to assess how much pain you are experiencing. CKPD usually doesn't cause sharp, sudden pain, but it can cause a dull ache or a constant pain. You should make sure you report pain to your healthcare team, carefully explaining the location, symptoms and impact of your pain on your life.

Medication

Pain relief: can be used to help relieve and manage the severity of pain. **Antidepressants:** have also been developed to help control pain and relieve the impact of CKPD on patients' wellbeing. You may wish to ask your doctor about these.

How should chronic pain be managed?

Different types of chronic pain need different approaches. It is important to work together to manage chronic pain. Depending on the type and severity of your pain, you may need:

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



Treatments can be used to help manage the underlying disease factors which cause pain when it is advanced.



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

Pain is the most common and challenging symptom of ACPND in many patients, particularly when persistent. You can assess at any stage of the disease, even early on, with a response to regular analgesic treatment.

What causes pain in ACPND?

Pain can be caused by various factors or be **secondary** to ACPND, such as pain related to underlying disease. 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 pain in the setting of liver and can be particularly difficult to treat.

Assessing pain

Doctors and other healthcare staff may use simple approaches to assess pain in people with ACPND, usually 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. **Antidepressants** have also been developed to help control and relieve the impact of ACPND on patients' wellbeing. 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 may be helpful to use a combination of different types of pain relief. The most common types of pain relief used are:

- Analgesics** - as with all medicines, these should be used as directed.
- Antidepressants** - used to help control and relieve the impact of ACPND on patients' wellbeing.

Doctors in the hospital may be able to help with the management of chronic pain.

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 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 in our [Kidney stones](#) section.

What causes pain in ADPKD?

There are several causes of pain in ADPKD, such as kidney stones, cysts, and infections. The most common cause of pain in ADPKD is kidney stones, which can be caused by a variety of factors, including a diet high in salt and protein.

Other causes of pain in ADPKD include cysts, which can grow in the kidneys and cause pain, and infections, which can also cause pain. If you are experiencing any of these symptoms, it is important to see your doctor.

Assessing pain

Doctors and other healthcare workers use a variety of tools to assess pain. These tools can help them understand how much pain you are in and what is causing it. They can also help them decide on the best treatment for your pain. It is important to talk to your doctor about your pain and how it is affecting your life.

Medication

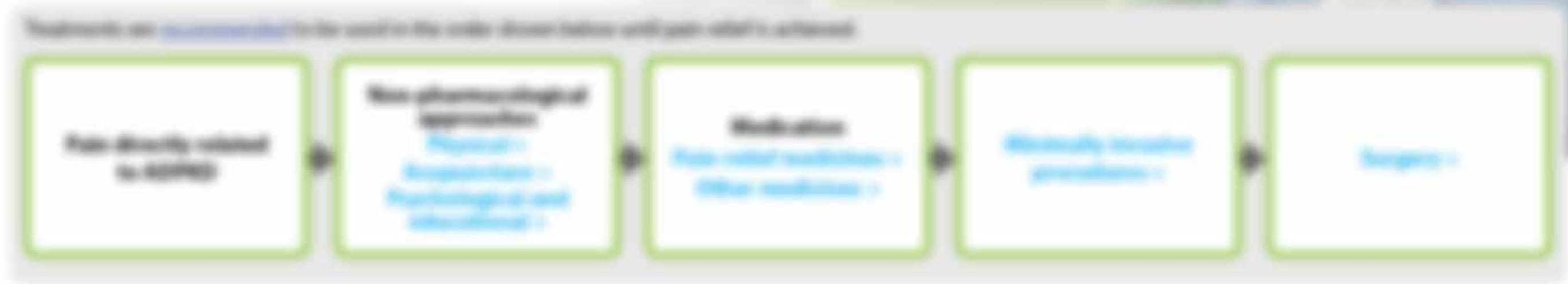
[Painkillers](#) can be used to help relieve pain. There are several different types of painkillers, and your doctor can help you choose the one that is best for you. It is important to use painkillers as directed and to avoid using them for a long time.

How should chronic pain be managed?

Chronic pain is a long-term condition that can be difficult to manage. It is important to work with your doctor to develop a plan for managing your pain. This may include a combination of medication, physical therapy, and other 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.



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?

Pain can be caused by various factors in ADPND, such as joint inflammation, nerve damage, and underlying issues of bone structure to 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 bone and can be particularly difficult to treat.

Assessing pain

Doctors and other healthcare staff may use simple questionnaires to assess pain in ADPND, usually 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

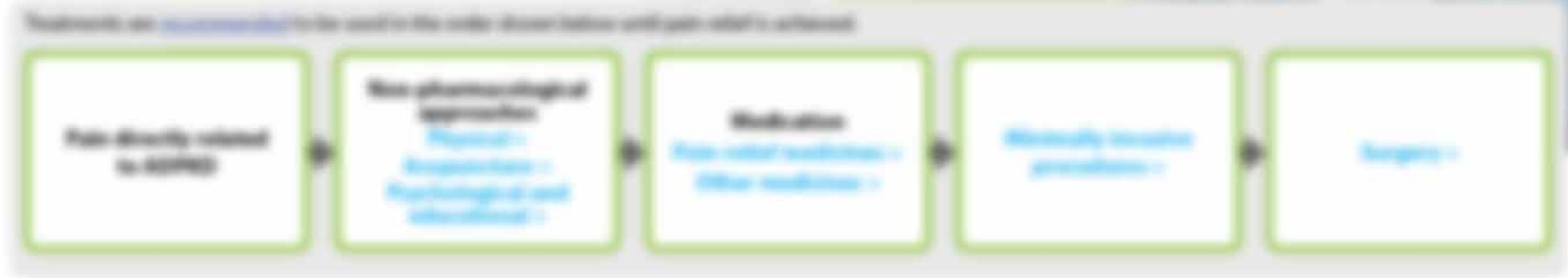
Paracetamol can be useful to help relieve pain and reduce the severity of pain. **Corticosteroids** have also been developed to help control pain and reduce the impact of ADPND on patients' wellbeing. You may wish to ask your doctor about these.

How should chronic pain be managed?

Different types of chronic pain and health care professionals often work together to help chronic pain. Depending on the cause and type of pain, the management may include:

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 when you are waiting for a transplant to manage it (multiple sclerosis).

What causes pain in ADPKD?

Pain can be caused by various things or by [cysts](#) in ADPKD, such as cyst infections and kidney stones. Treating the underlying cause of these things may 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 things that are difficult to treat.

Assessing pain

Doctors and other healthcare staff may use simple questionnaires to find out how much pain you are in. People with ADPKD usually describe pain using a visual analogue scale (VAS) or a numeric rating scale (NRS). You should make sure you report pain to your healthcare team, carefully explaining the location, symptoms and impact of your pain and experience.

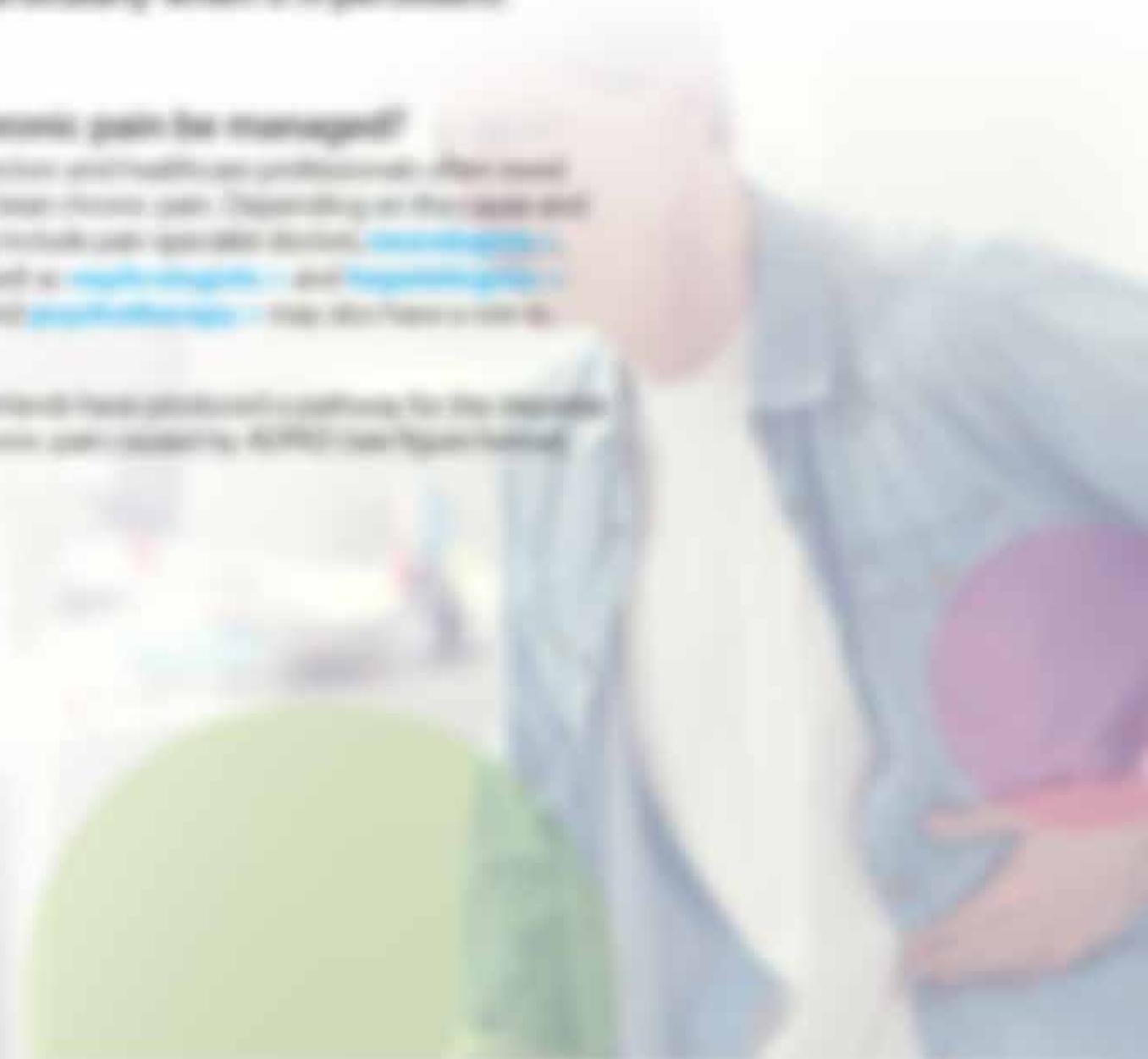
Medication

[Pain relief](#) can be useful to help relieve and manage the severity of pain. [Antibiotics](#) have also been developed to help control and reduce the impact of ADPKD on patients suffering. You may wish to ask your doctor about these.

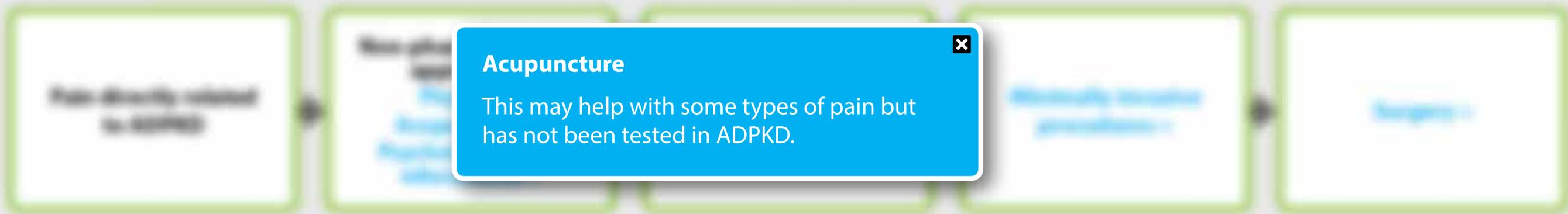
How should chronic pain be managed?

Different types of 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](#), [nurse](#), [dietitian](#), [occupational therapist](#) and [counsellor](#). You should have a care plan for your chronic pain.

There is also information on [pain management](#) for the overall management of chronic pain, including ADPKD, on [painmanagement.org.uk](#).



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 contains an overview of ADPKD for some patients, particularly when it comes to the pain that can occur at any stage of the disease, even when you don't have a large number of cysts.

What causes pain in ADPKD?

Pain can be caused by various things, such as [hypertension](#) or [kidney stones](#), which are common in ADPKD, as well as [cystic degeneration](#) or [cystic growth](#). Treating the underlying cause of these things may 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 things that are difficult to treat.

Assessing pain

Doctors and other healthcare staff may use [pain scales](#) to see how much pain you are in. People with ADPKD usually describe their pain as a dull ache or a sharp pain that comes and goes. You should make sure you report pain to your healthcare team, usually explaining the location, symptoms and impact of the pain on your life.

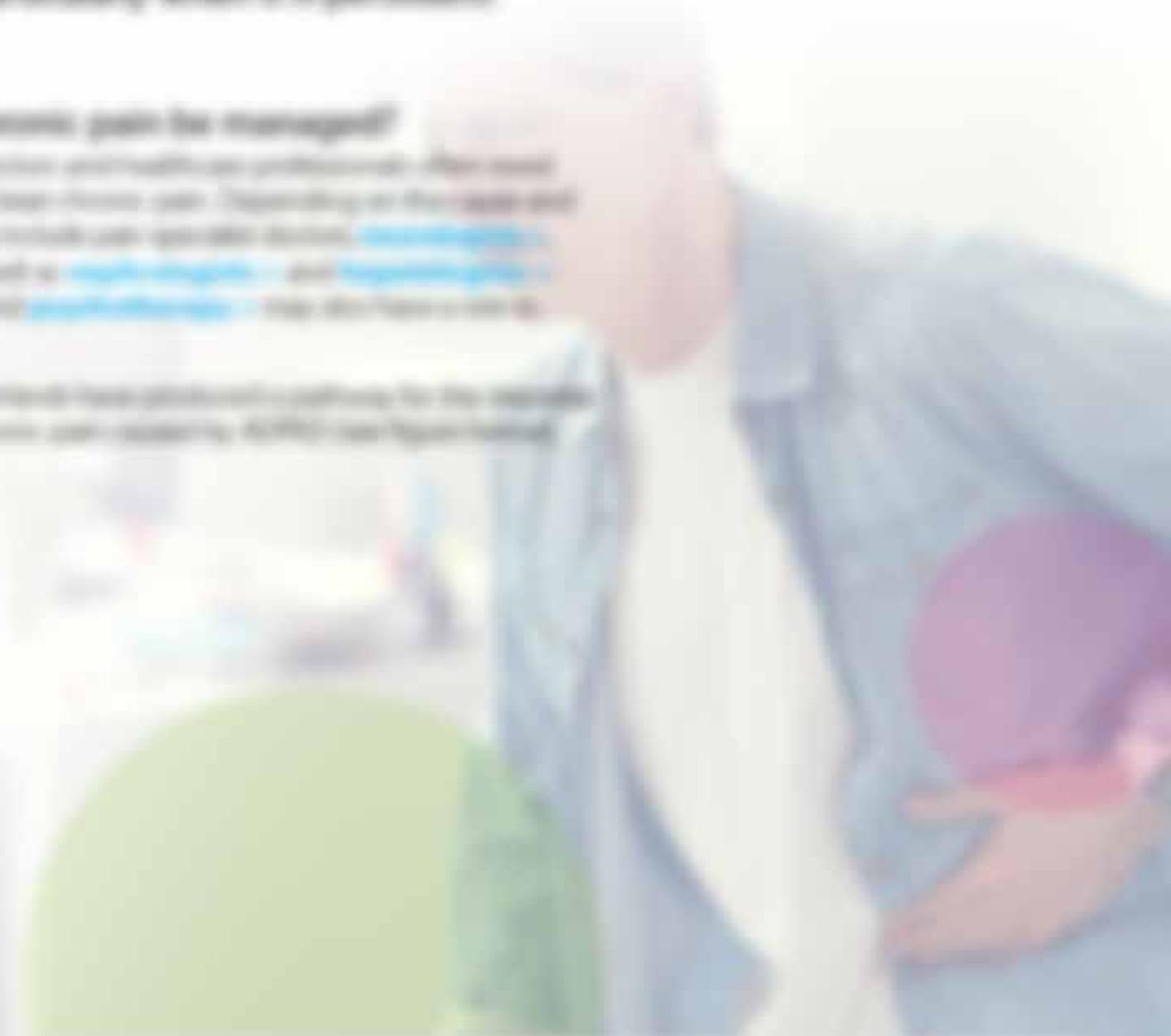
Medication

[Pain relief](#) can be useful to help reduce and manage the severity of pain. [Antidepressants](#) have also been developed to help control pain and to reduce the impact of ADPKD on patients' wellbeing. You may wish to ask your doctor about these.

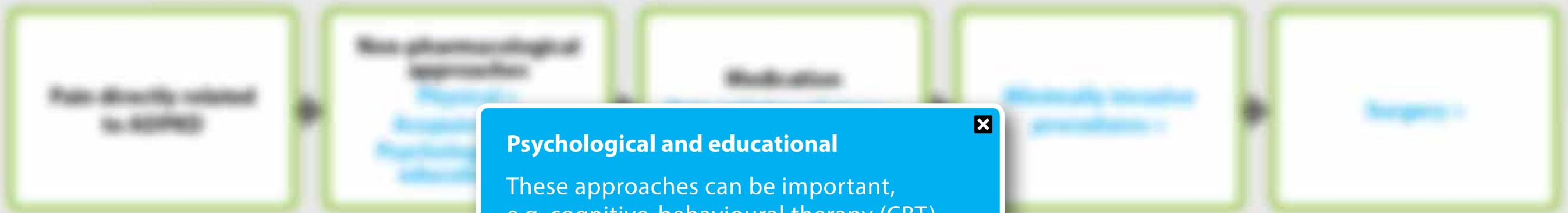
How should chronic pain be managed?

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](#), [counsellor](#), [nurse](#), [dietitian](#), [social worker](#) and [occupational therapist](#). You should have a plan to help you manage your pain.

There is also information on [pain management](#) for the overall management of chronic pain, including ADPKD, on [painmanagement.org.uk](#).



Treatments are [recommended](#) to be used in the order shown below with pain relief as a last resort.



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.

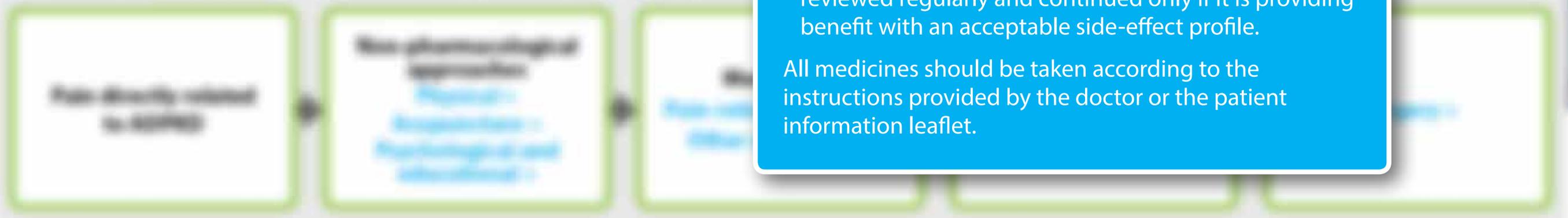
Assessing pain

Doctors will ask you about your pain, how often you get it, and how long it lasts. They will also ask you about any other symptoms you have, such as nausea, vomiting, or changes in your appetite. They will also ask you about any other medicines you are taking, including over-the-counter medicines.

Medicines

[Paracetamol](#) can be used to help relieve pain. It is usually taken every 4-6 hours. [NSAIDs](#) can also be used to help relieve pain, but they can be harmful to the kidneys. [Opioids](#) can be used to help relieve pain, but they can be addictive. You should only use them if you are prescribed them.

Medicines are [prescribed](#) to help relieve the pain. Some medicines can help with the pain, but they can also have side effects. You should only use them if you are prescribed them.



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 at any stage of the disorder, even when you are not a specialist in managing multiple symptoms.

What causes pain in AD/HD?

Pain can be caused by various factors or by [comorbidities](#) in AD/HD, such as an infection, an injury, or a condition. 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 simple questionnaires to assess pain in people with AD/HD. These questionnaires should normally be done 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 helpful to help relieve and manage the severity of pain. [Antidepressants](#) have also been developed to help control pain and relieve the impact of AD/HD in patients suffering the long-term or persistent pain.

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 pain and type of pain, this may include your general doctor, [physiotherapist](#), [psychologist](#), [counsellor](#), [social worker](#), [occupational therapist](#), [dietitian](#), [nurse](#) and [pharmacist](#).

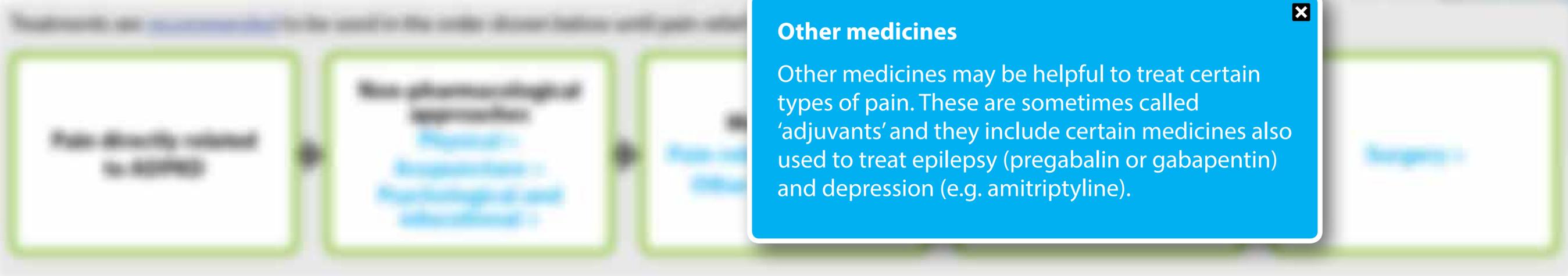
Doctors in the following areas provide support for the overall management of chronic pain in people with AD/HD: [general medicine](#), [neurology](#), [psychiatry](#), [physiotherapy](#), [occupational therapy](#), [dietetics](#), [social work](#), [counselling](#), [psychology](#), [nursing](#) and [pharmacy](#).

Doctors in the following areas provide support for the overall management of chronic pain in people with AD/HD: [general medicine](#), [neurology](#), [psychiatry](#), [physiotherapy](#), [occupational therapy](#), [dietetics](#), [social work](#), [counselling](#), [psychology](#), [nursing](#) and [pharmacy](#).



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).



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 the stage of the disease with help from your doctor to manage it better. Multiple options are available.

What causes pain in ADPKD?

Pain can be caused by various factors in ADPKD, such as cyst infections, stretching of the kidney capsule, and growth of cysts in the kidney or liver. Understanding what causes the pain can help you manage it better.

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 workers use different approaches to assess pain in ADPKD. Usually, doctors will use a visual analogue scale (VAS) to assess pain. You should make sure you report pain to your healthcare team, especially regarding the location, symptoms and impact of your pain and experience.

Medication

Non-steroidal anti-inflammatory drugs (NSAIDs) can be used to help reduce pain and reduce the severity of cysts. **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 doctors and healthcare professionals often work together to help manage pain. Depending on the cause and type of pain, the team might use various drugs, **physical therapy**, **acupuncture**, **hypnosis**, **and cognitive behavioral therapy** to help the pain to be better managed.

Doctors in the following areas provide support for the overall management of chronic pain in ADPKD: **neurologists**, **psychiatrists**, **and psychologists**.



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 **individualized** to be used in the most effective way with your doctor's guidance.



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 the severity of the disease with help from a specialist to manage multiple symptoms.

What causes pain in ADPKD?

Pain can be caused by various factors or by **hypertension** in ADPKD, such as cyst infections and kidney stones. 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 cysts in the kidney or from other conditions 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 during your next clinic visit. You should make sure you report pain to your healthcare team, carefully explaining the location, symptoms and impact of your pain on your life.

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 its impact. The impact of ADPKD on patients suffering the long-term effects of pain should be discussed.

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, the team could use specialist services, **analgesics**, **antidepressants**, **anticonvulsants**, **steroids** and **opioids** to help the pain to be better controlled.

Doctors in the following areas provide specialist services for the management of chronic pain: **general ADPKD care** and **specialist services**.



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.



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 to improve your quality of life and how to get help.

Living with ADPKD

ADPKD can affect your quality of life in many ways. You may experience physical symptoms such as pain, fatigue and difficulty concentrating. You may also experience emotional symptoms such as anxiety, depression and a sense of isolation. It is important to talk to your doctor about these symptoms and to seek support if you need it.

Living with ADPKD - How can you improve your quality of life? This section explains how to get help and how to improve your quality of life.

Living with ADPKD - How can you improve your quality of life? This section explains how to get help and how to improve your quality of life.

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

Seeking professional help

You're not alone



Wellbeing, personal and family life

This section explains how ADPKD can affect your wellbeing, personal 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 can be a source of stress, worry, and sadness. It can also affect your ability to work, study, and enjoy life. It is important to seek support and help when you need it.

ADPKD can also affect your relationships with family and friends. It can be a source of tension and conflict. It is important to communicate your feelings and needs to those around you.

There are many things that you can do to help manage your emotional wellbeing. You can talk to your doctor, a counsellor, or a support group. You can also try relaxation techniques, such as meditation and yoga.

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 you can do to help and what support patients can expect.

Emotional impact

ADPKD can have a significant impact on your wellbeing, and it can be difficult to cope with the emotional and physical challenges of the disease. It can also affect your family life and your relationships with others.

It is important to talk to your healthcare team about how you are feeling and what you need. They can help you to manage your emotions and provide you with the support you need.

There are many things you can do to help you cope with ADPKD, such as talking to your healthcare team, joining a support group, and taking part in physical and social activities.



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 life. The stress of an illness, such as a chronic and progressive one, can have a profound impact on the quality of life and on the ability to cope with ADPKD in the long term.

Work

Some patients with ADPKD may have to leave their jobs to work.

Financial issues

ADPKD can affect long-term health and the associated cost of care.

 **Questionnaires** have been developed specifically to help measure the impact of ADPKD on quality of life and wellbeing.

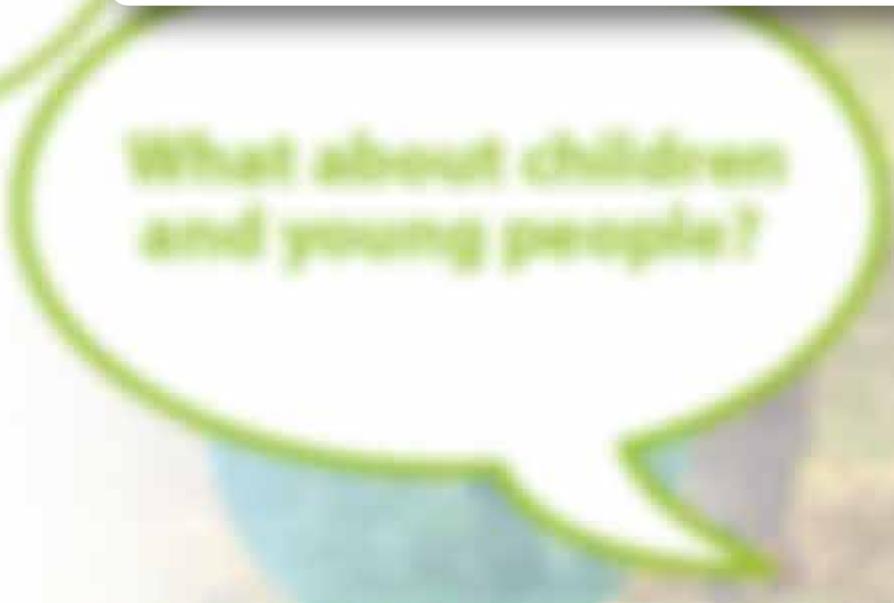
Polycystic Liver Disease Questionnaire (PLD-Q) can help assess the effect of liver cyst symptoms. This can be found [here](#) (p. 11–16).

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 contribute to stress, anxiety, depression and other mental health problems. It is important to seek support if you are experiencing any of these symptoms.

Support - You can find support groups and other resources to help you cope with the emotional impact of ADPKD on you and your family.

Work and employment

There are many things that patients and their 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 professional 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 community life. It can also have a significant impact on the lives of young people with ADPKD, and on the lives of their families.

Support: You can find support groups for people with ADPKD in your local area.

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.

Life

ADPKD can affect your quality of 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.

PGD 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 is because pregnancy can be associated with a higher risk of a woman with **CKPD** experiencing their mother's symptoms for the first time.

For all women with CKPD, it is important to have a family with CKPD. This is because pregnancy can be associated with a higher risk of a woman with CKPD experiencing their mother's symptoms for the first time.

Diagnosis and transplantation
There are good reasons to consider kidney transplantation during **CKPD** and the **CKPD** **transplantation** - you may want to discuss this with your nephrologist or kidney team.

The implementation and prenatal genetic diagnosis
CKPD is caused by a **genetic** mutation, which can be tested for in embryos created in vitro with **CKPD** before a woman is even able to become pregnant. This allows you to select embryos that do not have the mutation.

Genetic diagnosis
Prenatal **CKPD** is a disease that can be passed on to a child. It is caused by a **genetic** mutation, which can be tested for in embryos created in vitro with **CKPD** before a woman is even able to become pregnant. This allows you to select embryos that do not have the mutation.

Genetic diagnosis
Prenatal **CKPD** is a disease that can be passed on to a child. It is caused by a **genetic** mutation, which can be tested for in embryos created in vitro with **CKPD** before a woman is even able to become pregnant. This allows you to select embryos that do not have the mutation.

People with **CKPD** 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 **CKPD** experiencing their mother's symptoms for the first time.

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Genetic testing
It is possible to test embryos created in vitro with **CKPD** before a woman is even able to become pregnant. This allows you to select embryos that do not have the mutation.

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.



This section explains how ADPKD can affect family planning and pregnancy.

People with ADPKD can have different levels of kidney function. The severity of kidney disease can vary from mild to severe. People with severe kidney disease may need dialysis or a kidney transplant.

ADPKD is a genetic condition that causes the kidneys to grow larger than normal. This can lead to kidney failure over time. The severity of the condition varies from person to person.

Medical problems
ADPKD can cause a variety of medical problems, including high blood pressure, urinary tract infections, and kidney stones. These problems can be managed with medication and lifestyle changes.

Male fertility
ADPKD can affect the ability of men to have children. This is because the disease can cause the testes to produce fewer sperm.

Contraception
People with ADPKD should use contraception to prevent pregnancy. This is because pregnancy can be risky for people with kidney disease.

Pregnancy
Pregnancy can be risky for people with ADPKD. This is because the disease can cause complications during pregnancy, such as high blood pressure and pre-eclampsia.

People with ADPKD should talk to their healthcare provider about the risks of pregnancy. They should also discuss family planning options.

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 a nephrologist. This involves a long wait for an appointment.

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 team about ways of managing these sexual 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 avoid the use of oral contraceptives because of the risk of kidney complications. 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 women **developing** kidney complications during pregnancy and when delivering their babies, and there are considerations regarding the use of pain **relievers**.

Diagnosis and transplantation

There are good considerations regarding kidney donation being **done** and the **kidney transplantation**, you may wish to discuss this with your nephrologist or health care team.

The implementation and prenatal genetic diagnosis

ADPKD is usually passed **autosomal recessive**, inheritance requires a person with ADPKD to have a partner who only carries a copy of the disease gene with their other gene.

The implementation genetic diagnosis

The implementation genetic diagnosis (PGD) can allow the parents to know if the foetus has ADPKD or not before it is born. This is usually done by using a small sample of cells from the foetus to check for the presence of the ADPKD mutation, and whether it is passed from the mother or the father.

PGD can only be performed if the genetic genetic mutation causing ADPKD is the partner has been identified. This information about ADPKD is available **here**.

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 a nephrologist. This involves a long wait for an appointment.

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Researching

It is possible to research in the area of genetic testing and family planning with ADPKD.

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 ADHD can affect family planning and pregnancy.

People with ADHD can face difficult decisions when considering whether to have children. This section explains what you can expect with ADHD and how you can make the most of your options.

ADHD and family planning – people with ADHD may find it difficult to plan ahead. This section explains how you can make the most of your options.

Sexual problems – people with ADHD may have sexual problems. This section explains how you can make the most of your options.

Plan fertility – people with ADHD may find it difficult to plan ahead. This section explains how you can make the most of your options.

Contraception – people with ADHD may find it difficult to plan ahead. This section explains how you can make the most of your options.

Pregnancy – people with ADHD may find it difficult to plan ahead. This section explains how you can make the most of your options.

Supporting the use of ADHD medication – people with ADHD may find it difficult to plan ahead. This section explains how you can make the most of your options.

Diagnosis and transplantation – people with ADHD may find it difficult to plan ahead. This section explains how you can make the most of your options.

The implementation and prenatal genetic diagnosis – people with ADHD may find it difficult to plan ahead. This section explains how you can make the most of your options.

The implementation genetic diagnosis – people with ADHD may find it difficult to plan ahead. This section explains how you can make the most of your options.

ADHD and the use of genetic diagnosis – people with ADHD may find it difficult to plan ahead. This section explains how you can make the most of your options.

People with ADHD and genetic diagnosis – people with ADHD may find it difficult to plan ahead. This section explains how you can make the most of your options.

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.

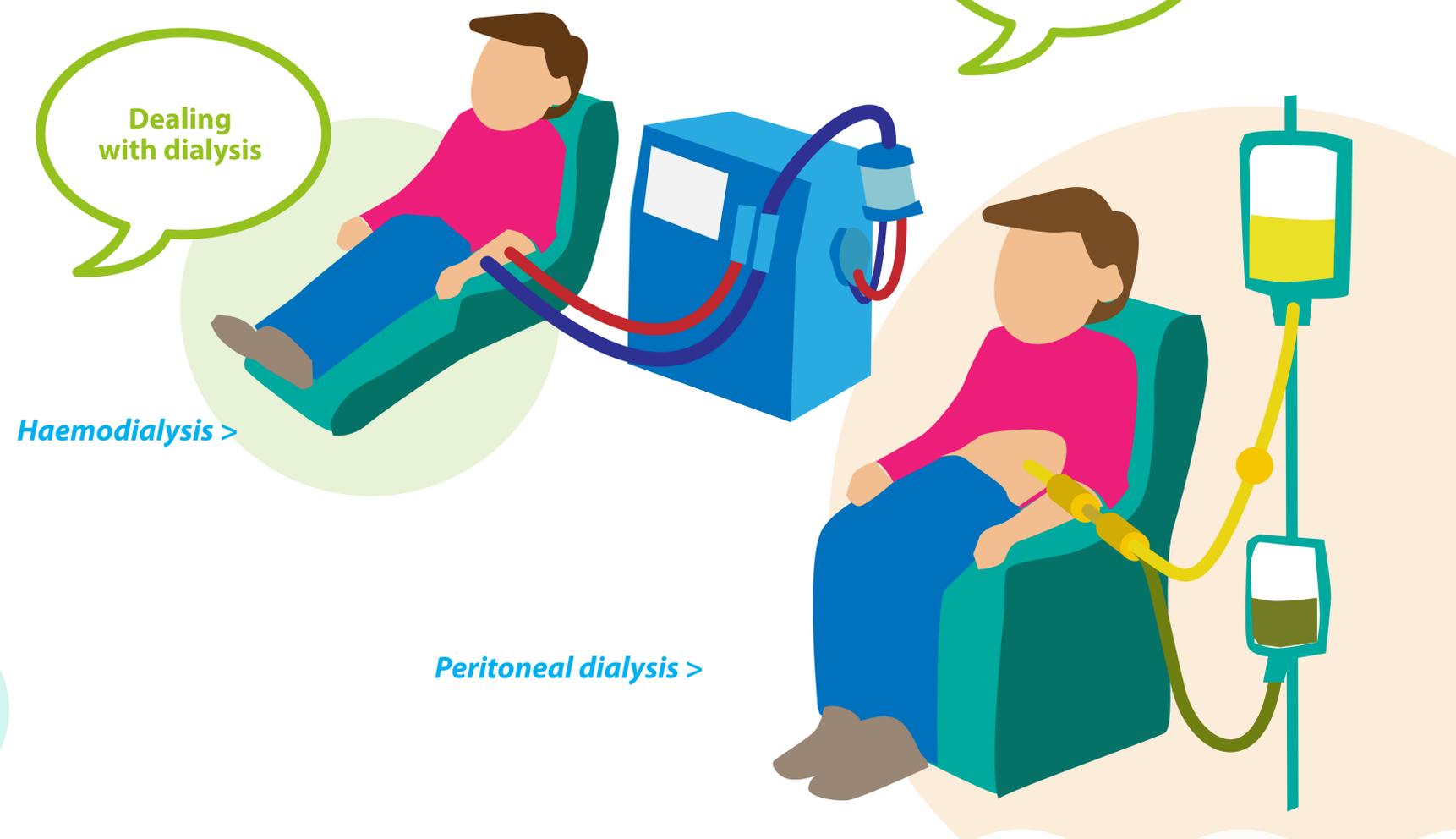
Local kidney and ADPKD [patient organisations](#) will have patients whom you can ask about ESRD and its treatment. Dialysis and transplantation are generally the same for all patients with ESRD and meeting fellow patients is usually a good and helpful experience.

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.

For more information on dialysis, visit www.nhs.uk or call the patient helpline on 0800 328 0000.

How can I get help with the costs of dialysis?

Most people with ESRD have a health insurance fund (Gesetzliche Krankenkasse) which covers the costs of dialysis.

If you are not covered by a health insurance fund, you can apply for a health insurance fund. You can also apply for a health insurance fund if you are self-employed or have a job.

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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. The lower the eGFR, the less well the kidneys are working.

ESRD is the final stage of CKD. It means the kidneys are no longer able to filter waste and extra fluid from the blood.

How is ESRD treated?

There are two main ways to treat ESRD: dialysis and kidney transplantation.

Dialysis is a process that filters the blood to remove waste and extra fluid. It can be done in a hospital or at home.

Kidney transplantation is a surgical procedure that replaces a failed kidney with a healthy one from a donor.

Both treatments help to control symptoms and prevent complications of ESRD.

It is important to work closely with your healthcare team to choose the best treatment for you.

Living with ESRD can be challenging, but there are many resources available to help you manage your condition.

For more information, visit our website or contact our patient support team.

Thank you for reading this information. We hope it helps you understand ESRD and your treatment options.

Diagnosis

Doctors will use blood and urine tests to check kidney function. They will also ask about your symptoms and medical history.

ESRD is usually diagnosed when the eGFR is below 15. This means the kidneys are not working well enough to filter the blood.

Other tests, such as imaging, may be used to look for the cause of kidney disease. This helps doctors decide on the best treatment for you.

It is important to see a nephrologist (kidney specialist) if you have CKD. They will help you understand your condition and manage your health.

Living with ESRD can be challenging, but there are many resources available to help you manage your condition.

For more information, visit our website or contact our patient support team.

Thank you for reading this information. We hope it helps you understand ESRD and your treatment options.

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 ²)
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 end-stage renal disease	Less than 15



End-stage renal disease

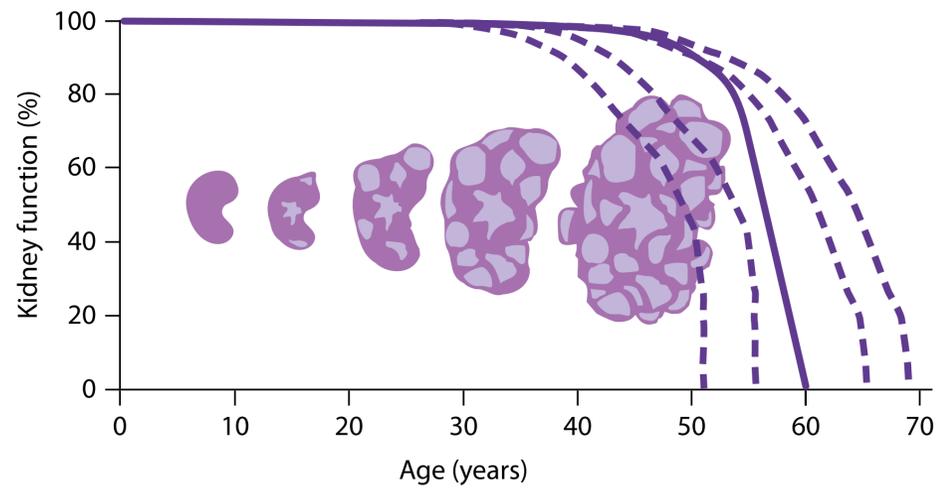
This section explains end stage renal disease (ESRD) and the treatment options for patients with ADPKD who reach this stage of kidney disease.

The information in this section is for people with ADPKD who are interested in learning more about ESRD and the treatment options for people with ADPKD who reach this stage of kidney disease.

Diagnosis
Doctors will check the blood to measure the waste products and electrolytes 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 discusses the importance of diet and fluid intake for people with ESRD.

Haemodialysis

Haemodialysis is a treatment for ESRD that uses a dialysis machine to filter the blood. It is performed in a clinic or hospital. There are two main types of haemodialysis: intermittent and continuous. Intermittent haemodialysis is performed three times a week for 4 hours. Continuous haemodialysis is performed every day for 24 hours. The choice of treatment depends on the patient's condition and preferences.

Haemodialysis



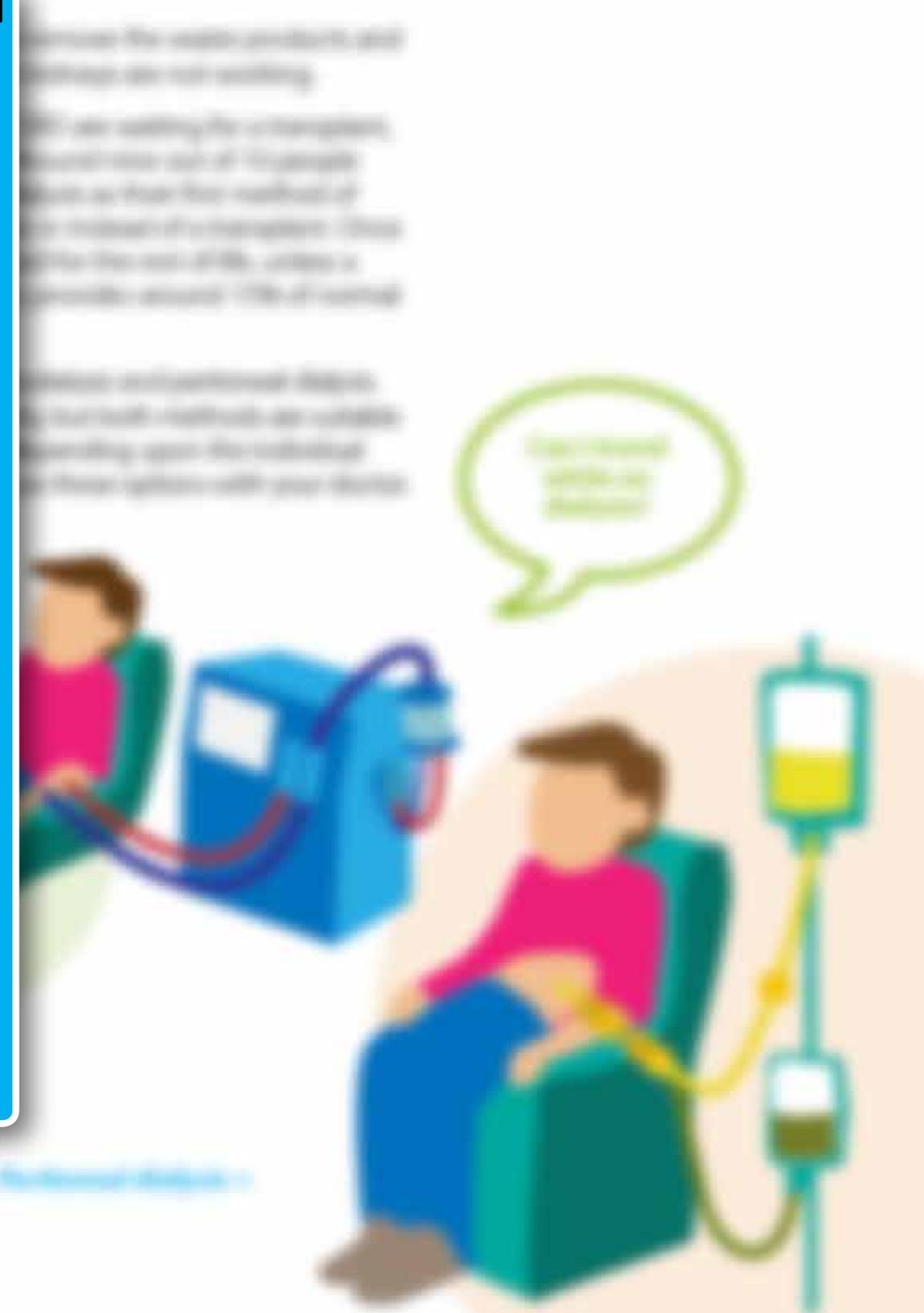
Haemodialysis uses a dialysis machine to filter the blood outside the body. Blood is removed via a needle in the arm and passed through a tube into the machine. Filtered blood is then returned to the arm via another tube.

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.



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 is not intended for people who have not yet reached this stage of kidney disease.

For more information on end stage kidney disease, visit [www.kidney.org](#).

How to choose a dialysis modality

When you have end stage kidney disease, you will need dialysis. There are two main types of dialysis: haemodialysis and peritoneal dialysis. Your healthcare team will help you choose the best option for you.

Haemodialysis is done in a dialysis centre. Peritoneal dialysis can be done at home. Both types of dialysis help to remove waste and extra fluid from your blood.

Peritoneal dialysis uses the lining of your abdomen to filter your blood. Haemodialysis uses a machine to filter your blood.

Both types of dialysis have pros and cons. Your healthcare team will help you decide which one is best for you.

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.

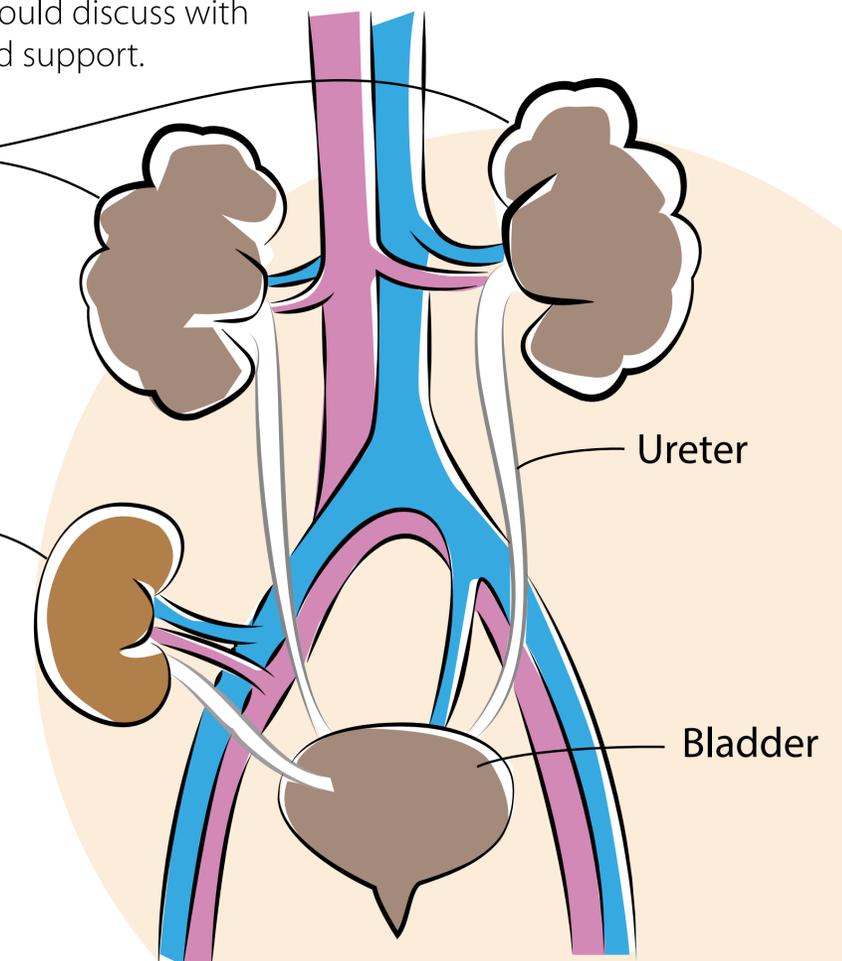
Remember, you will still have ADPKD even after a kidney transplant and so [complications](#) that occur elsewhere in the body may still need treatment.

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.



Diseased kidneys

Transplant kidney



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

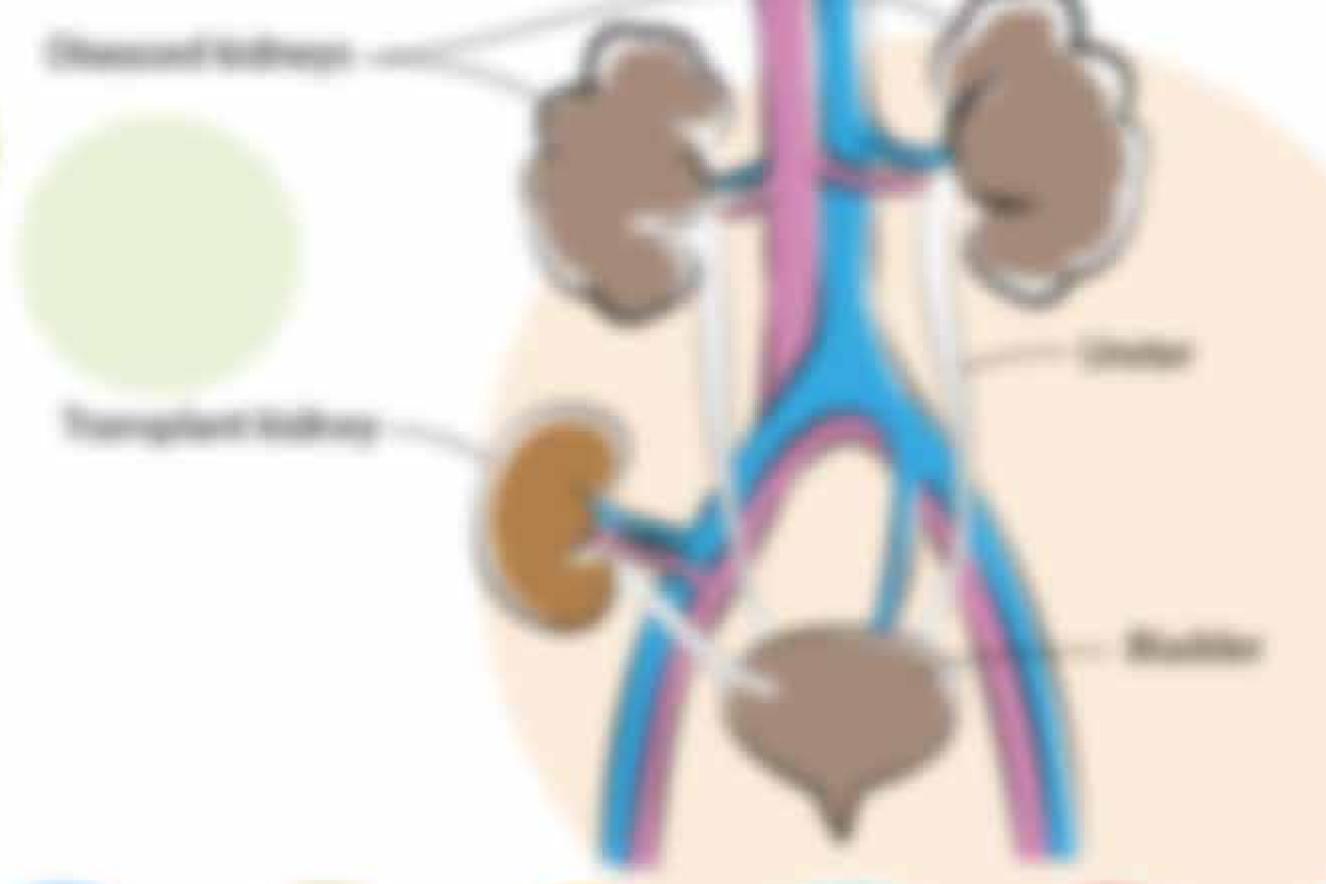
Where possible, kidney transplantation is the best treatment for ESRD. This is because a successful transplant restores kidney function without the need for dialysis and can offer patients a better quality of life. It is also far more cost-effective for the health system, as compared with dialysis.

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 certain substances in your blood. It is important to follow your doctor's advice and to take your medication exactly as prescribed. You will also need to avoid certain foods and drinks, and to avoid getting sick. It is also important to avoid getting pregnant for a certain period of time after the transplant. You will also need to avoid certain activities, such as contact sports, and to avoid driving for a certain period of time after the transplant. You will also need to avoid certain medications, such as over-the-counter painkillers, and to avoid certain supplements, such as herbal remedies. You will also need to avoid certain infections, such as chickenpox and measles. You will also need to avoid certain travel destinations, such as countries with a high risk of malaria. You will also need to avoid certain animals, such as cats and dogs, and to avoid certain plants, such as ivy and holly. You will also need to avoid certain people, such as people who have certain infections. You will also need to avoid certain situations, such as crowded places and public transport. You will also need to avoid certain events, such as large gatherings and festivals. You will also need to avoid certain activities, such as swimming in public pools and hot tubs. You will also need to avoid certain foods, such as raw vegetables and undercooked meat. You will also need to avoid certain drinks, such as alcohol and caffeine. You will also need to avoid certain supplements, such as vitamins and minerals. You will also need to avoid certain medications, such as antibiotics and antifungals. You will also need to avoid certain infections, such as urinary tract infections and skin infections. You will also need to avoid certain travel destinations, such as countries with a high risk of malaria. You will also need to avoid certain animals, such as cats and dogs, and to avoid certain plants, such as ivy and holly. You will also need to avoid certain people, such as people who have certain infections. You will also need to avoid certain situations, such as crowded places and public transport. You will also need to avoid certain events, such as large gatherings and festivals. You will also need to avoid certain activities, such as swimming in public pools and hot tubs. You will also need to avoid certain foods, such as raw vegetables and undercooked meat. You will also need to avoid certain drinks, such as alcohol and caffeine. You will also need to avoid certain supplements, such as vitamins and minerals. You will also need to avoid certain medications, such as antibiotics and antifungals. You will also need to avoid certain infections, such as urinary tract infections and skin infections.

It is important to follow your doctor's advice and to take your medication exactly as prescribed. You will also need to have regular blood tests to check your levels of certain substances in your blood. It is important to follow your doctor's advice and to take your medication exactly as prescribed. You will also need to have regular blood tests to check your levels of certain substances in your blood.



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 receive a kidney transplant?

Patients with CKD who are on dialysis and whose kidneys are no longer working well may be eligible for a kidney transplant.

Why donate a kidney?

The most common reason for donating a kidney is to help someone with CKD who is on dialysis.

Living donor: A living donor is someone who is healthy enough to donate a kidney and who is able to live a normal life after the surgery.

Deceased donor: The deceased donor program is a program in which living donors donate kidneys from someone who has recently died.

How can I find a living donor? You can find a living donor through a transplant center or a national living donor registry. You can also find a living donor through a national living donor registry.

What does transplant involve?

Living donor transplant: A living donor transplant involves a major surgery in which the donor's kidney is removed and the recipient's kidney is replaced.

When receiving a transplant, it's important to stay as healthy as possible through [pre-transplant care](#) and [post-transplant care](#). The transplant team should give you specific instructions about what to do before the time of the surgery.

Generally, transplantation is not a fully curative treatment for people with CKD, as well as any other form of kidney disease. As with any operation, there are [risks](#) and you should discuss these with the transplant team.

Post-transplant care - what happens afterwards?

After transplantation, you will need to take medication to prevent your body from rejecting the kidney. The amount of medication will vary depending on your situation. You will also need to have regular check-ups with your transplant team.

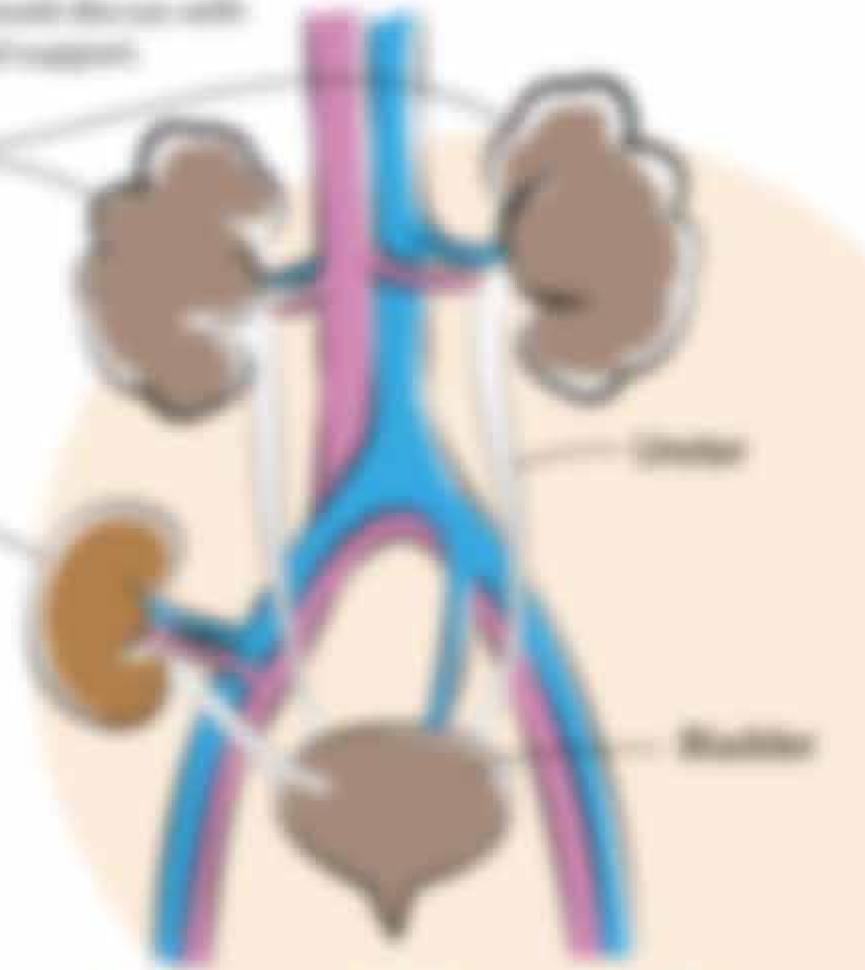
Living donor: A living donor transplant involves a major surgery in which the donor's kidney is removed and the recipient's kidney is replaced. The donor will need to have regular check-ups with their transplant team.

Deceased donor: A deceased donor transplant involves a major surgery in which a kidney from a deceased donor is used. The recipient will need to have regular check-ups with their transplant team.

Transplantation can have emotional and psychological effects on both the donor and the recipient. Support is available for those affected by these issues. You can find more information about the available options for advice, care and support.

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.



[How long does a kidney transplant last?](#)



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.

Best transplant case - what happens afterwards?

After a successful kidney transplant, the patient will need to take immunosuppressive drugs to prevent their body from rejecting the new kidney. The doctor will also monitor the patient's kidney function and overall health.

Living donor transplantation is generally considered to be the best option because it has a higher success rate and a lower risk of complications.

Patients who receive a kidney transplant from a living donor often have a better long-term outcome compared to those who receive a kidney from a deceased donor.

It is important for patients to follow their doctor's instructions and attend regular check-ups to ensure the transplant is working well.

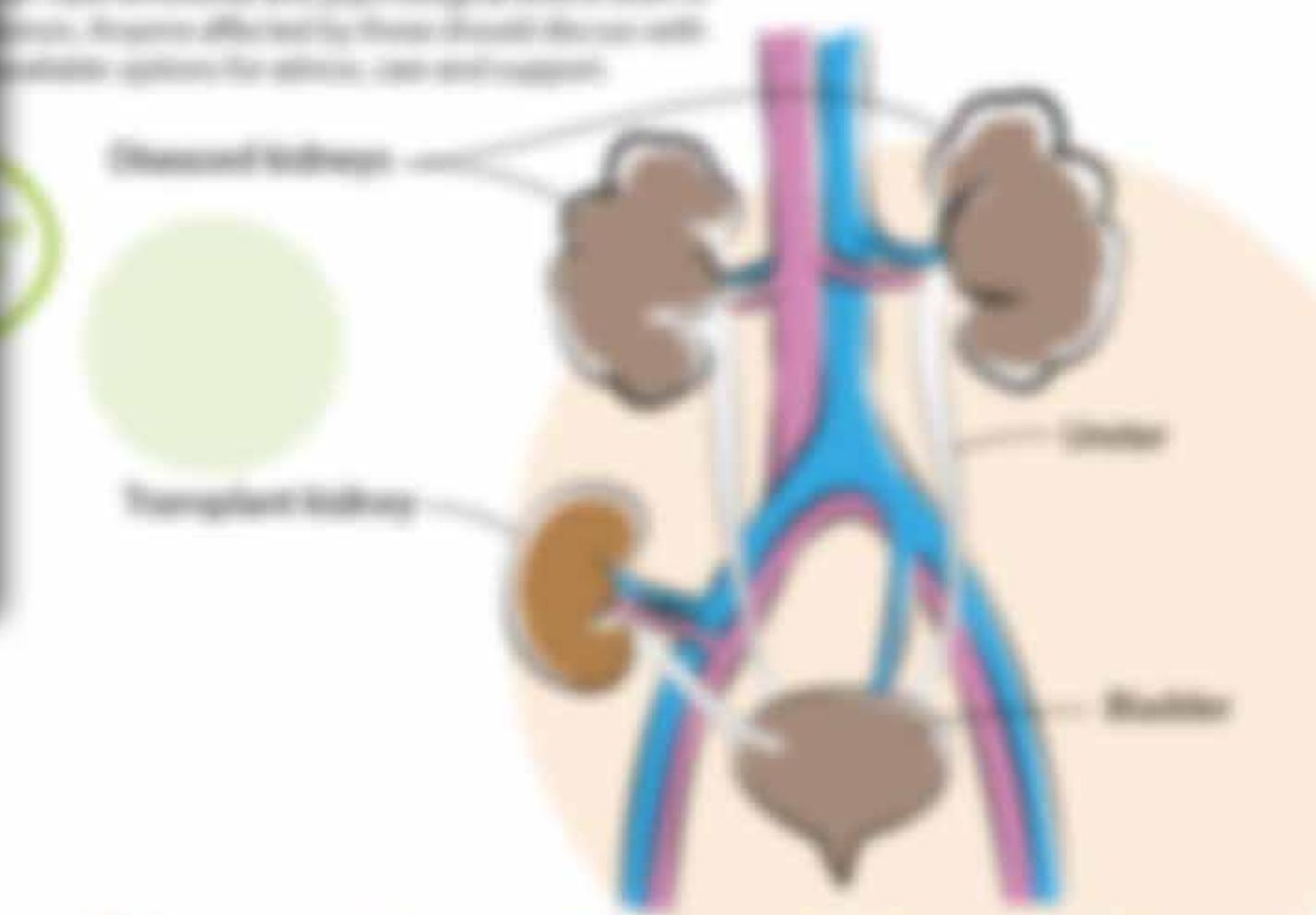


Living donor

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.



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 will only be successful if the donor and the recipient are compatible. It is usually not possible to transplant a kidney from one person to another.

It is not possible to transplant a kidney from one person to another. However, a kidney transplant can be performed if the donor and the recipient are compatible. It is usually not possible to transplant a kidney from one person to another.

Best transplant case - what happens afterwards?

After transplantation, you will need to take medicines to prevent your body from rejecting the kidney. The medicines will help your body to accept the kidney. The medicines will also help to prevent you from getting infections. You will need to take these medicines for the rest of your life.

Living donor [kidney transplant](#) is considered to be the best option for kidney transplantation. It is usually possible to transplant a kidney from a living donor. The donor will be a family member or a friend. The donor will be a healthy person. The donor will be a person who is not on dialysis. The donor will be a person who is not on a transplant 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.

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.

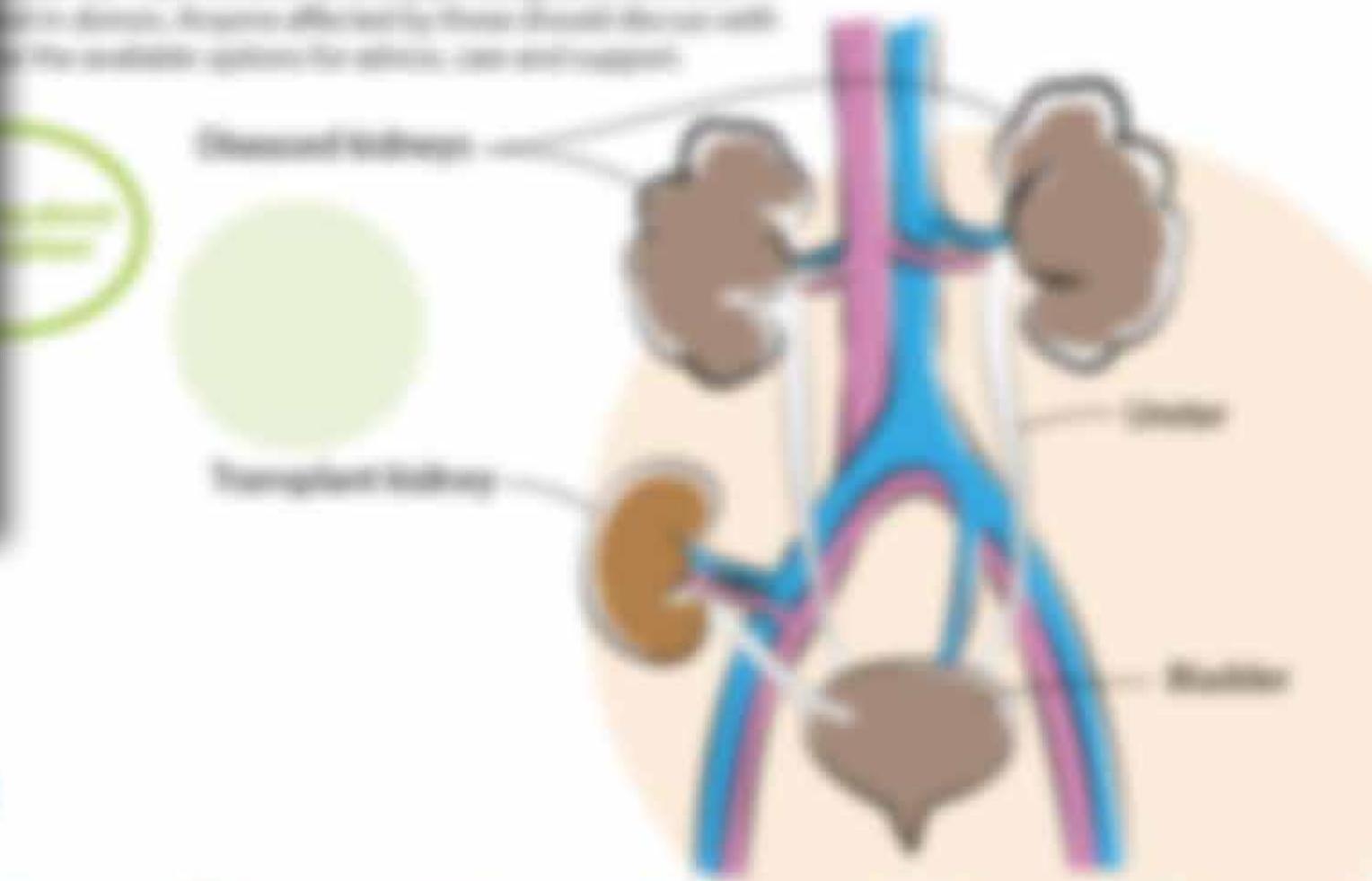
Patients should discuss the situation in their country. Once a deceased donor kidney becomes available, urgent transplant surgery is performed as soon as possible.



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](#). Sometimes, kidney transplantation is not suitable for someone with [cancer](#). It is usually not an option for people with [heart failure](#) or [liver disease](#).

It can take a long time to wait for a [deceased donor kidney](#) and people with CKD may be considered for a transplant from a living donor.

Why donate the kidney?

The most common reason for donating a kidney is to help someone with CKD. Donating a kidney does not affect the donor's health.

Living donor

A living donor is someone who gives one of their kidneys to a person with CKD.

Deceased donor

A deceased donor is someone who has died and their organs are donated to someone who needs them.

The donor

Anyone over the age of 18 can donate a kidney. The donor must be healthy and have a good understanding of the risks and benefits of donating a kidney.

What does the surgery involve?

The surgery is carried out under general anaesthetic. The donor's abdomen is opened up and the kidney is removed. The recipient's abdomen is also opened up and the donor kidney is placed in the body.

What happens after surgery?

The recipient will need to take medication to prevent their body from rejecting the donor kidney. They will also need to have regular check-ups to ensure the kidney is working properly.

Living donor

Living donors are usually family members or friends of the recipient. They are screened for health problems and must understand the risks and benefits of donating a kidney.

Deceased donor

Deceased donors are people who have died and their organs are donated to someone who needs them. The organs are usually donated to someone on the transplant list.

Waiting time

The waiting time for a deceased donor kidney can be long. It can take several years to get a transplant. Living donors are usually able to get a transplant more quickly.

Cost

Kidney transplantation is a major surgical operation and can be expensive. However, it is often worth the cost because it can improve the recipient's quality of life and reduce the need for dialysis.

Living donor

Living donors are usually able to get a transplant more quickly than people who are waiting for a deceased donor kidney. This is because there are fewer living donors than deceased donors.

Best transplant case - what happens afterwards?

After transplantation, the recipient will need to take medication to prevent their body from rejecting the donor kidney. They will also need to have regular check-ups to ensure the kidney is working properly. The donor will also need to have regular check-ups to ensure their own kidney is working properly.

Living donor [transplantation](#) is considered to be the best option for kidney transplantation because it usually results in a longer survival time for the recipient. However, it is not always possible to find a living donor and the [waiting time](#) for a deceased donor kidney can be long.

There are several [types of kidney transplantation](#) and each has its own risks and benefits. The recipient will need to take medication to prevent their body from rejecting the donor kidney. They will also need to have regular check-ups to ensure the kidney is working properly.

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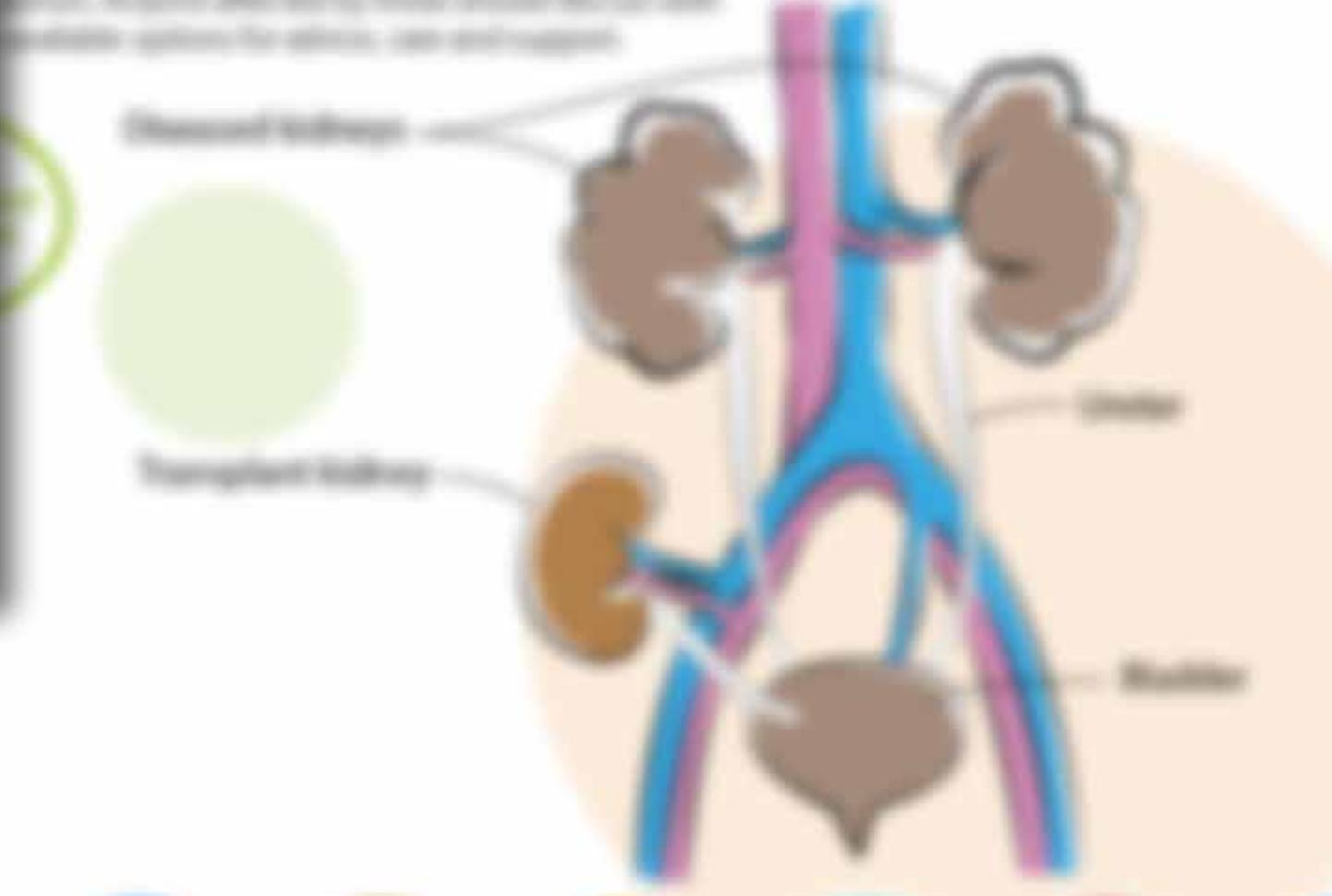


Kidney transplantation surgery

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 kidney transplants are performed on a living donor.

When possible, kidney transplantation is the best treatment for CKD. However, [not everyone is eligible for a transplant](#). A transplant is only possible if you have a suitable donor. Kidney transplantation is not suitable for everyone and the [risks](#) of a transplant can vary.

It can take a while to find a donor. [Living donors](#) together with CKD may be considered for a transplant and are being managed.

Why donate the kidney?

The transplant team will be able to advise you on the risks of donation. You may wish to discuss these options with your transplant team.

Living donor: A living donor is someone who gives a kidney to help someone with CKD. 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 kidneys after they have died.

How can I find a living donor? You can find a living donor through a transplant center or a living donor program. You can also find a living donor through a transplant center or a living donor program.

Post-transplant care - what happens afterwards?

After transplantation, you will need to take medicines to prevent your body from rejecting the kidney. You will also need to take medicines to prevent infection. You will also need to take medicines to prevent blood clots. You will also need to take medicines to prevent diabetes. You will also need to take medicines to prevent high blood pressure.

Living donor [transplants](#) are considered to have a better long-term outcome than deceased donor [transplants](#). However, you will still need to take medicines to prevent rejection and infection. You will also need to take medicines to prevent blood clots and diabetes.

After you receive a [living donor kidney transplant](#), you will need to take medicines to prevent rejection and infection. You will also need to take medicines to prevent blood clots and diabetes. You will also need to take medicines to prevent high blood pressure.

Remember you will still need CKD care after kidney transplant. [CKD](#) [medicines](#) may still be needed in the body may still need treatment.

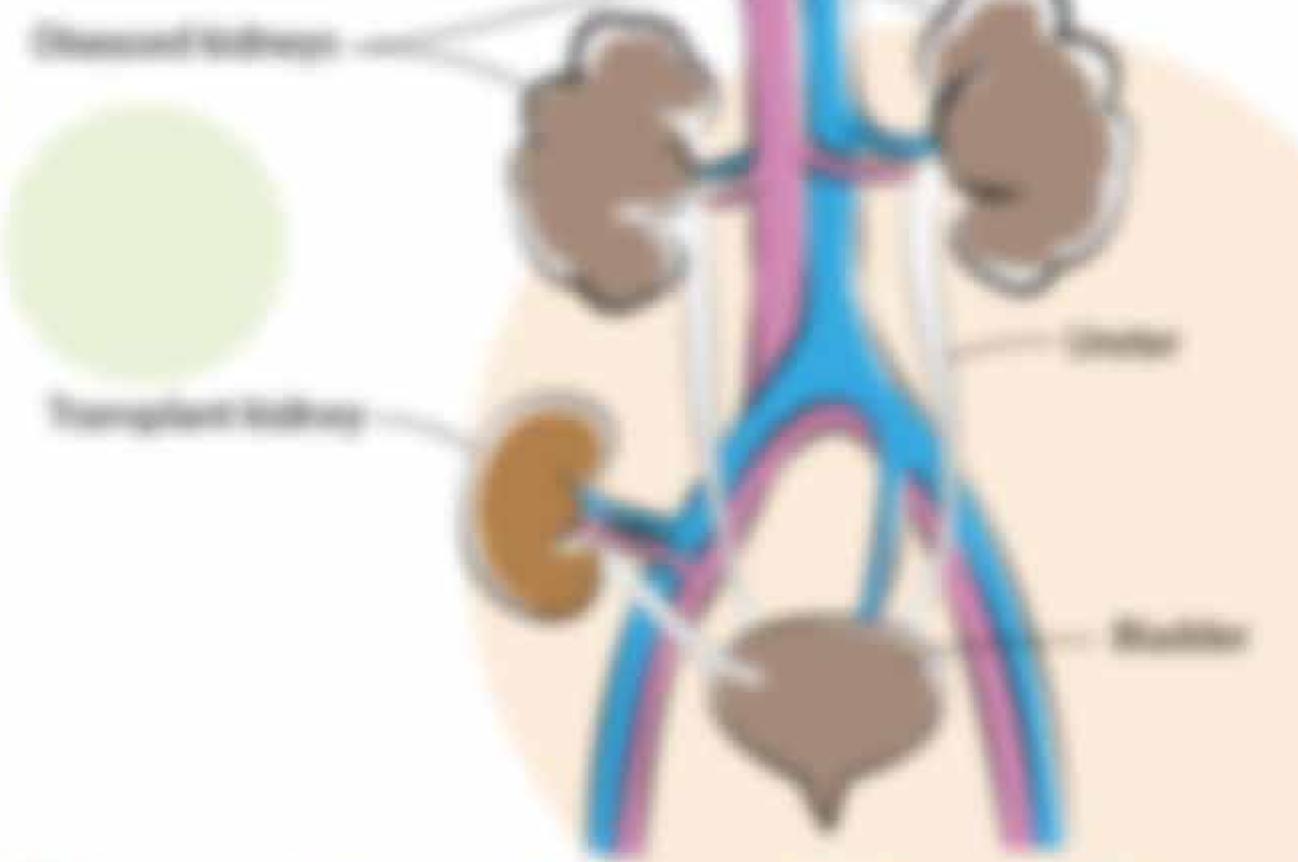
Transplantation can have emotional and psychological effects on both the donor and the recipient. Discuss this with your transplant team. You may also want to discuss the available options for after-care support.



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.



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 get a kidney: one from a donor kidney (which is the most common) or one from a deceased donor.

When possible, kidney transplantation is the best treatment for CKD. However, [you must have a transplant](#) to receive a kidney. There are several factors that can affect the success of a transplant, including the health of the donor and the recipient.

It is not clear whether patients with [ADPKD](#) together with CKD may be considered for a transplant and receiving a transplant.

Who donates the kidney?

The transplant can be provided by a living or deceased donor. Living donors can be family members or friends with good relationships.

Living donor: A living donor is a healthy person who gives a kidney to a patient with CKD. The donor's kidney will continue to work in their body.

Deceased donor: The donor is a person who has died recently. The kidney is taken from the donor's body and given to a patient with CKD.

How long does a kidney transplant last?
A kidney transplant can last for many years, but it is not guaranteed to last for the rest of your life. The length of time a transplant lasts depends on many factors, including the health of the donor and the recipient, the quality of the transplant, and the care you receive after the transplant.

What does transplant work for?

Living donor: A living donor is a healthy person who gives a kidney to a patient with CKD. The donor's kidney will continue to work in their body.

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What happens after a transplant?

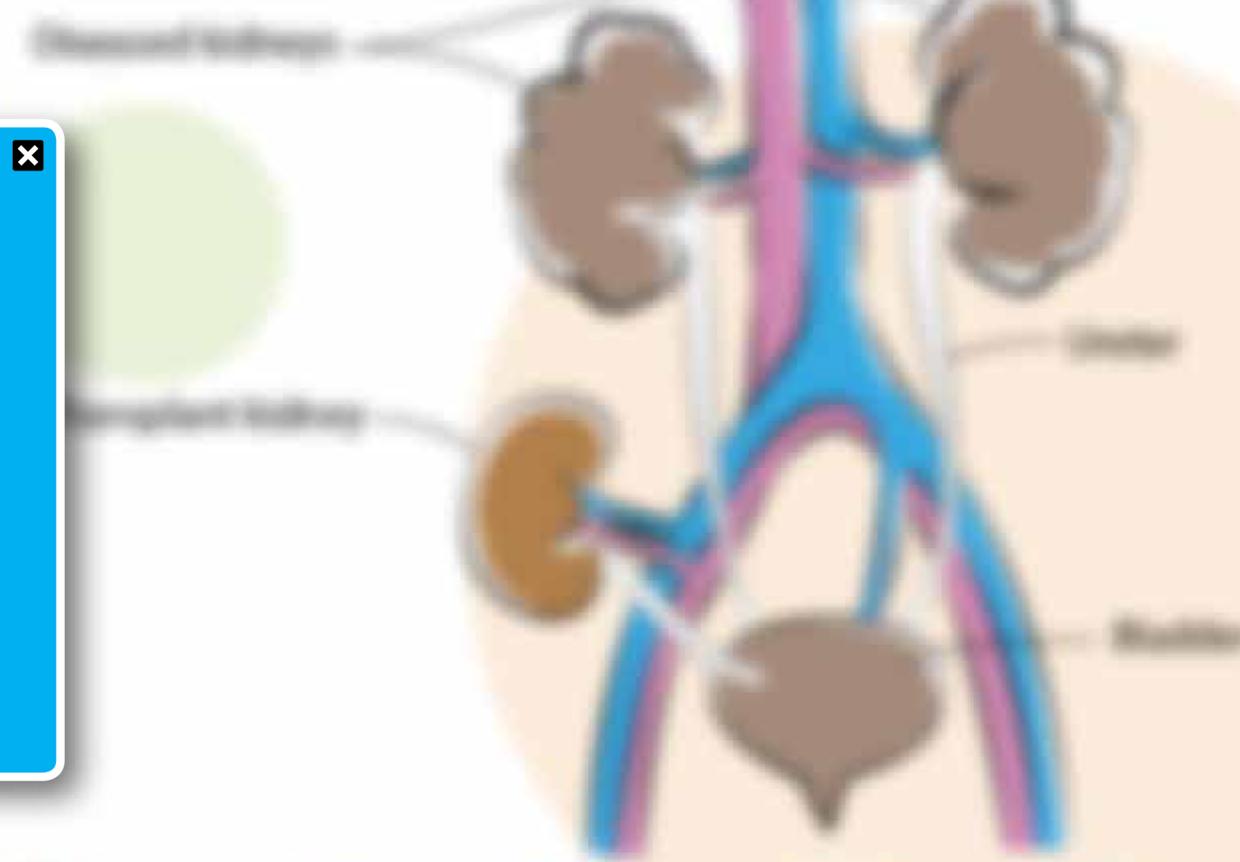
After a transplant, you will need to take medication to prevent your body from rejecting the kidney. You will also need to have regular check-ups with your doctor to monitor the health of your new kidney. It is important to follow your doctor's instructions carefully to ensure the best possible outcome for your transplant.

Living donor: A living donor is a healthy person who gives a kidney to a patient with CKD. The donor's kidney will continue to work in their body.

Deceased donor: The donor is a person who has died recently. The kidney is taken from the donor's body and given to a patient with CKD.

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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, [you should discuss the risks and benefits of kidney transplantation with your doctor](#) to see if it is right for you.

It can also be used with people [on dialysis](#) together with CKD to help to control their blood pressure and reduce the need for dialysis.

Why donate the kidney?

The most common reason for someone to donate a kidney is to help someone with CKD who is on dialysis.

Living donor: A living donor is someone who is healthy and gives one of their kidneys to someone with CKD.

Deceased donor: The donor is someone who has died and their kidney is given to someone with CKD.

How can I find out more about kidney transplantation?
You can find out more about kidney transplantation by visiting [our website](#) or by contacting [our helpline](#).

What does transplant involve?

Living donor transplant: A living donor transplant involves a living donor giving one of their kidneys to someone with CKD.

When receiving a transplant, it is important to stay as healthy as possible through [good diet and exercise](#) and [not smoking](#). The transplant team should give you specific instructions about what to do when the time of the operation.

Generally, transplantation is used as a way to help someone with CKD, as well as any other health problems. As with any operation, there are [risks](#) and you should discuss these with the transplant team.

[How long does kidney transplantation last?](#)

Post-transplant care - what happens afterwards?

The transplant team will discuss with you what to expect after the operation. This includes taking immunosuppressant drugs to prevent your body from rejecting the kidney. You will also need to have regular check-ups with your doctor to monitor the kidney and the side effects of the drugs.

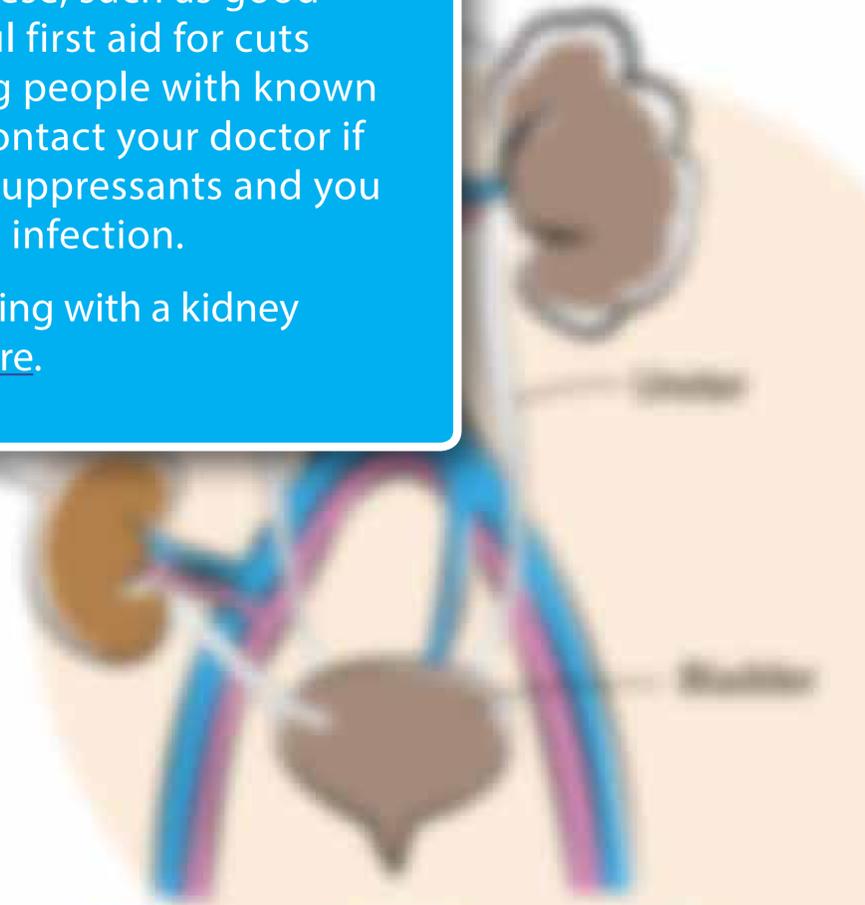


Follow-up care

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 used in the transplant.

There are two ways of getting a kidney transplant. The first is to get a kidney from a living donor. The second is to get a kidney from a deceased donor. The waiting time for a kidney transplant can be long.

It is important to talk to your doctor about the risks and benefits of kidney transplantation.

Why donate the kidney?

There are many reasons why someone might want to donate a kidney. Some people donate to help a friend or family member who has CKD. Other people donate to help someone who has a long waiting time for a kidney transplant.

Living donor: A living donor is someone who has a healthy kidney and is able to donate one of their kidneys to someone who has CKD.

Deceased donor: A deceased donor is someone who has died and has donated their kidney to someone who has CKD.

There are many reasons why someone might want to donate a kidney. Some people donate to help a friend or family member who has CKD. Other people donate to help someone who has a long waiting time for a kidney transplant.

What does transplant involve?

Living donor transplant: A living donor transplant involves a living donor donating one of their kidneys to someone who has CKD.

When receiving a transplant, it is important to stay as healthy as possible. This means eating a healthy diet, exercising regularly, and taking your medicines. The transplant team should give you specific instructions about what to do when the time of the operation.

Generally, transplantation is used as a way to help someone who has CKD. It is not a cure for CKD. It is important to talk to your doctor about the risks and benefits of kidney transplantation.

How long does a kidney transplant last?

What happens after a transplant?

After a kidney transplant, you will need to take immunosuppressant medicines for the rest of your life. These medicines help to prevent your immune system from attacking the new kidney. It is important to take your medicines exactly as your doctor tells you to.

It is important to stay as healthy as possible. This means eating a healthy diet, exercising regularly, and taking your medicines. The transplant team should give you specific instructions about what to do when the time of the operation.

It is important to talk to your doctor about the risks and benefits of kidney transplantation.

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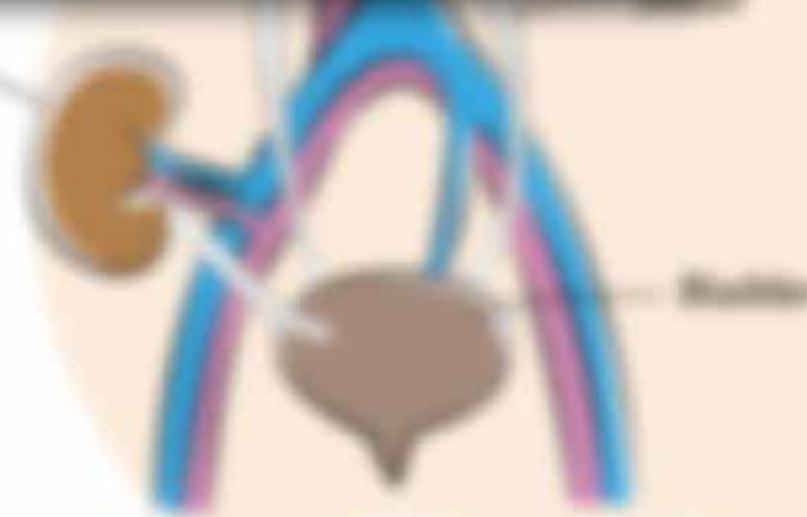
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Generally, transplantation is used as a way to help someone who has CKD. It is not a cure for CKD. It is important to talk to your doctor about the risks and benefits of kidney transplantation.

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 according to your individual circumstances. Depending on your individual circumstances, this may be a regular check-up or the need to adjust your treatment.

Remember that having your PKD [checked regularly](#) can be an important source of advice, support and information on these topics.

Transition care for adolescents

Adolescents with ADPKD may transition from paediatric health care services to adult services.

While these services are offered to adolescents, it is important for adolescents to transition from paediatric to adult services to help ensure continuity of care.

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.

What can patients do?

There are many ways that you can play a role in controlling your condition. These include:

-  **Get active**
-  **Stay hydrated**
-  **Reduce sodium**
-  **Monitoring and managing blood pressure**
-  **Planning for and staying on top of your care**



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 hospitals.

Regular care

People with ADPKD you will need regular care to help you manage your condition. This often involves regular appointments with your doctor. These include your kidney function, blood pressure and complications. The type of treatment you receive will depend on your doctor's experience and 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 other parts of the body.

Remember that you can also get support and information on these topics.

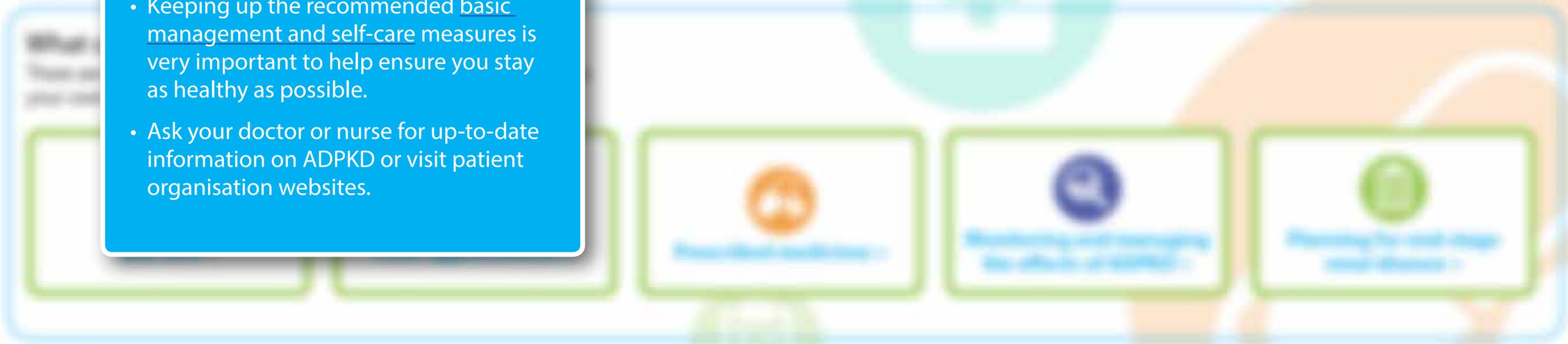
Transition care for adolescents

Adolescents with ADPKD may transition from paediatric health care services to adult services.

Read this document to understand the transition pathway to transfer the adolescents from paediatric 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 your GP or a specialist, and regular blood tests to check your kidney function.

Other specialists

Some people with ADPKD may also need to see other specialists, such as a dietitian, a psychologist, or a social worker. These specialists can help you manage your condition 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 the local GP. However, there are also some 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 health 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, you may need to see a specialist. Some people also need to see a specialist in general health care, such as a dietitian or a pharmacist, who can help you with your diet and your use of health, as well as ADPKD.

Some people may also need to see a specialist in their local region of health, support and information services.

Specialist care for adolescents

Adolescents with ADPKD may need to see a specialist health care service in their area.

Some people may also need to see a specialist in their local region of health, support and information services.

What can patients do?

There are many ways that you can help to manage your health. These include:

-  **Take all prescribed medicines**
-  **Follow your diet**
-  **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 from their healthcare team. This will include regular check-ups, blood tests and ultrasounds. The type of treatment you receive will depend on your stage of disease and other factors.

Other specialists

Your healthcare team will refer you to other specialists if you have [high blood pressure](#), [anemia](#) or [bone disease](#) in other parts of the body.

Some people with ADPKD may also have a genetic condition called [polycystic ovary syndrome](#) or [PCOS](#), which can affect the ovaries and lead to [infertility](#), as well as ADPKD.

What can patients do?

There are many things you can do to help 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 and requirements of each case.

Nephrology care

Typically, if you have ADPKD you will require regular care from 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 patients to other kinds of [specialist services](#) if necessary, for example if they experience [hypertension](#) 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 primary health care services to adult services.

Read [this document](#) to understand the transition pathway to transfer the adolescents from paediatric to adult services to help ensure continuity of care.

What can patients do?

There are many ways that you can play a role in contributing to your own care. These include:

- 1. [Self-care](#)
- 2. [Your symptoms](#)
- 3. [Healthcare decisions](#)
- 4. [Promoting and ensuring the safety of research](#)
- 5. [Planning for end-stage renal disease](#)

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.

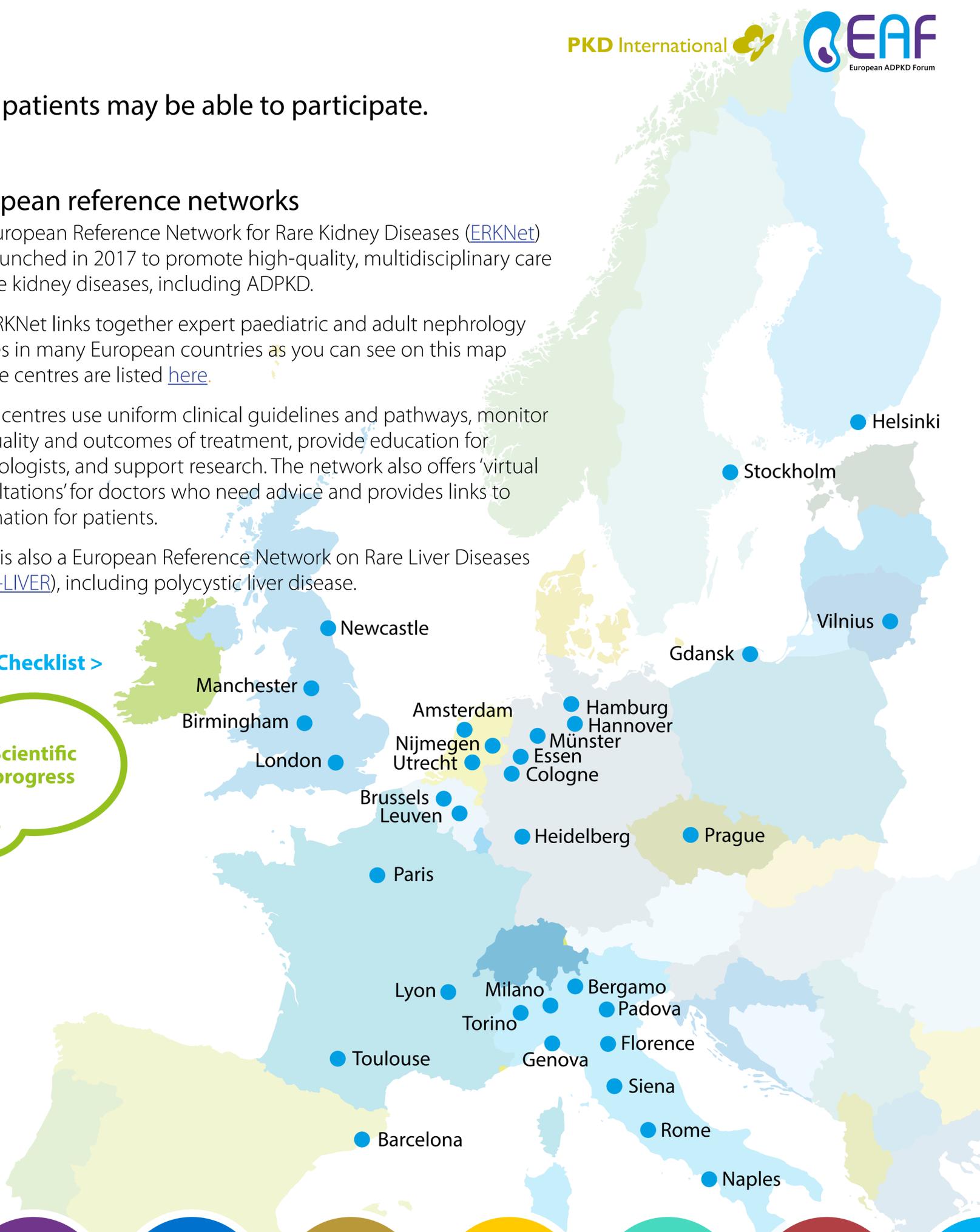
The ERKNet links together expert paediatric and adult nephrology centres in many European countries as you can see on this map – these centres are listed [here](#).

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.

 **Checklist >**

 **Scientific progress**



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 data collected is used to help understand the disease, such as how it affects people, how it is treated, and how often treatments are successful. Registers can also be used to help identify people who are at risk of developing the disease, or who are already affected by it.

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Registers can also be used to help identify people who are at risk of developing the disease, or who are already affected by it.

Clinical trials

Clinical trials are research studies that test the effectiveness of a new treatment or drug. They are usually carried out in a hospital or research centre. Clinical trials can help to find out if a new treatment is better than the current one, or if it has fewer side effects.

Clinical trials can help to find out if a new treatment is better than the current one, or if it has fewer side effects.

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 to understand the disease, such as how it affects people, and how effective treatments are. Registries also help researchers to identify areas of information that are missing with a disease.

ADPKD registries can be used to:

Provide information to help you understand your condition and how it affects you. You can also use the information to help you make decisions about your care.

Clinical trials

Clinical trials are research studies that test the effectiveness of the ability to produce or deliver a treatment, such as a drug, or other type 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

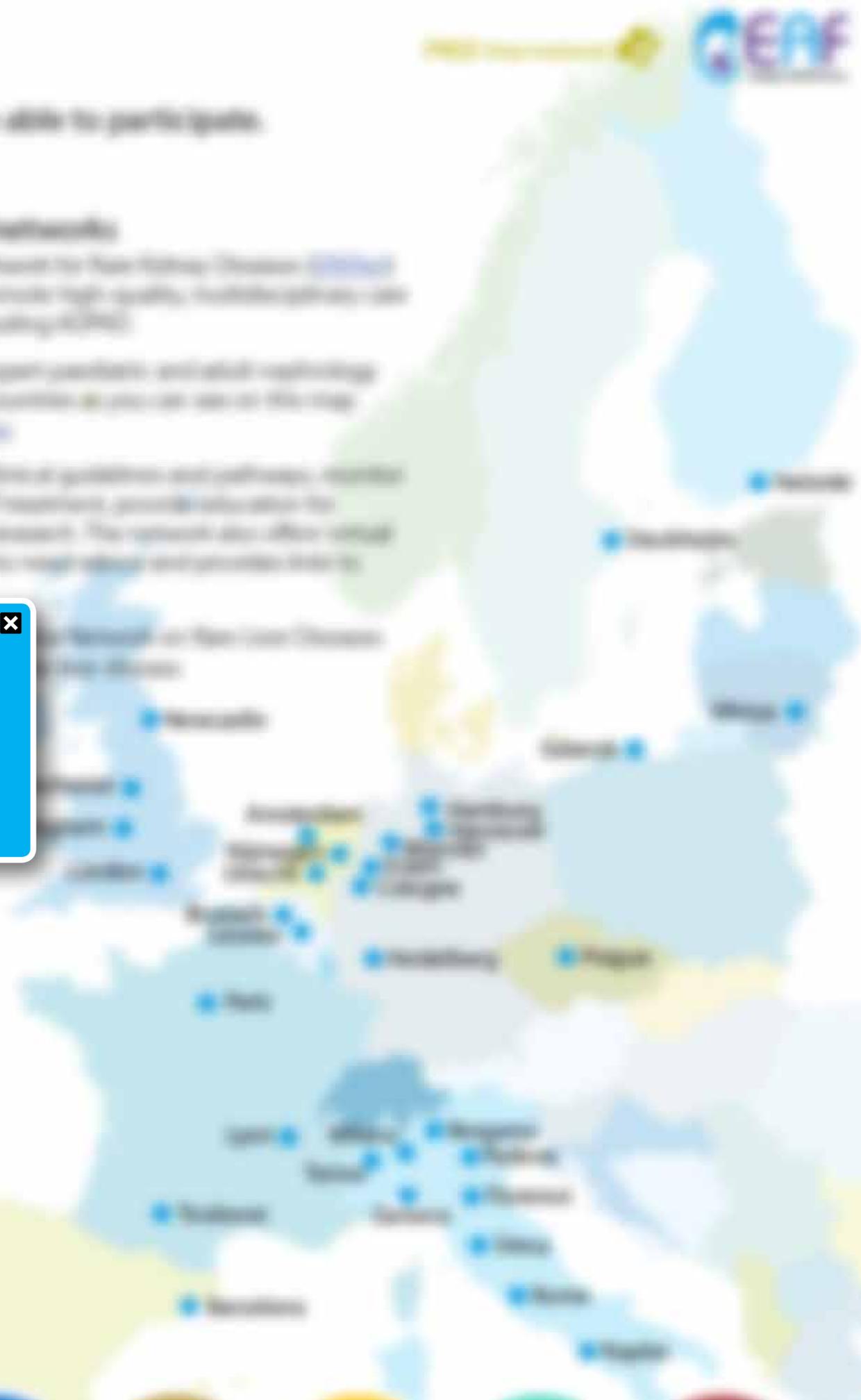
The European Reference Network for Rare Kidney Diseases ([ERN RD](#)) was established in 2017 to promote high quality, multidisciplinary care for rare kidney diseases, including ADPKD.

The ERN RD also organises regular meetings and other networking events in many European countries and you can see on this map - these events are listed [here](#).

These centres are within clinical guidelines and networks, provide the quality and consistency of care, provide education for healthcare professionals, and support research. The network also offers virtual consultations for those who may not be able to attend in person.

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 in your country or ADPKD care in the system, health care providers should consider each together to ensure resources across the system are used in the right way. It is also important to ensure that patients and providers in the system are aware of the system and its components, and that the system is designed to be user-friendly and accessible to all.

Current needs in ADPKD

ADPKD is a chronic, progressive condition that can lead to kidney failure. It is a leading cause of end-stage kidney disease (ESKD) in many countries. The [2019 Global Burden of Disease Study](#) found that ADPKD is a leading cause of disability-adjusted life years (DALYs) and deaths in many countries, particularly in high-income countries. The [2019 Global Burden of Disease Study](#) also found that ADPKD is a leading cause of kidney failure in many countries.

Policy context

ADPKD is a chronic, progressive condition that can lead to kidney failure. It is a leading cause of end-stage kidney disease (ESKD) in many countries. The [2019 Global Burden of Disease Study](#) found that ADPKD is a leading cause of disability-adjusted life years (DALYs) and deaths in many countries, particularly in high-income countries. The [2019 Global Burden of Disease Study](#) also found that ADPKD is a leading cause of kidney failure in many countries.

Multiple primary care

ADPKD is a chronic, progressive condition that can lead to kidney failure. It is a leading cause of end-stage kidney disease (ESKD) in many countries. The [2019 Global Burden of Disease Study](#) found that ADPKD is a leading cause of disability-adjusted life years (DALYs) and deaths in many countries, particularly in high-income countries. The [2019 Global Burden of Disease Study](#) also found that ADPKD is a leading cause of kidney failure in many countries.

Technology update

ADPKD is a chronic, progressive condition that can lead to kidney failure. It is a leading cause of end-stage kidney disease (ESKD) in many countries. The [2019 Global Burden of Disease Study](#) found that ADPKD is a leading cause of disability-adjusted life years (DALYs) and deaths in many countries, particularly in high-income countries. The [2019 Global Burden of Disease Study](#) also found that ADPKD is a leading cause of kidney failure in many countries.

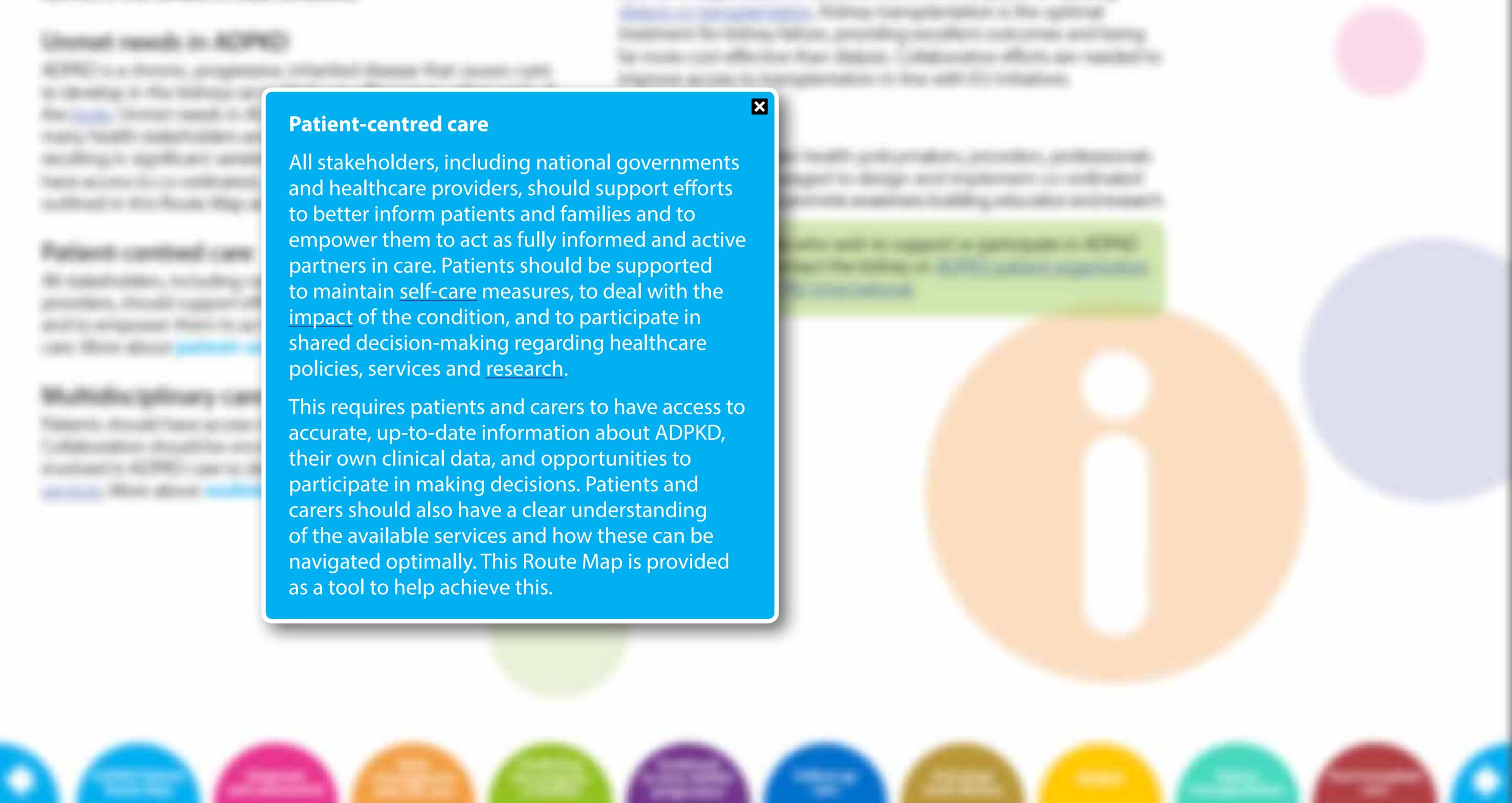
Implementation

ADPKD is a chronic, progressive condition that can lead to kidney failure. It is a leading cause of end-stage kidney disease (ESKD) in many countries. The [2019 Global Burden of Disease Study](#) found that ADPKD is a leading cause of disability-adjusted life years (DALYs) and deaths in many countries, particularly in high-income countries. The [2019 Global Burden of Disease Study](#) also found that ADPKD is a leading cause of kidney failure in many countries.

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 document covers the following areas:
- General recommendations
- Clinical care
- Research
- Education and training
- Health economics and financing
- Health equity and access
- Health systems and governance

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 system support they need at the right time. It is up to healthcare providers and patients to design, adapt or even to redesign 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 countries [where genetic health conditions are not addressed in the health system](#) may not be able to address [genetic health conditions](#) or address the needs of people with [genetic](#).

Remote medical care

In countries, including national governments and healthcare providers, should support efforts to better inform patients and health professionals about their own or their relatives' and genetic partners' care. More about [genetic medical care](#).

Multi-disciplinary care

There is a need for multi-disciplinary care for people with ADPKD. This includes:

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.

Technology update

Research [imaging - total kidney volume](#) and [automated total kidney volume](#) - 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 people with end-stage kidney disease, and many people with ADPKD need kidney transplantation. Transplantation offers an excellent option to improve quality of life with end-stage kidney disease.

Conclusion

Collaboration between health policymakers, providers, professional associations and managers to design and implement coordinated ADPKD services across primary, secondary, tertiary and quaternary care.

Health professionals who wish to support a person with ADPKD should discuss their care with a [genetic counsellor](#) or a [geneticist](#).



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 efficiently address current and emerging ADPKD care needs at local level.

Current needs in ADPKD

ADPKD is a chronic progressive condition where the number of cysts in the kidneys can increase over time, often leading to kidney failure. The [2019](#) survey results in ADPKD include a list of concerns among kidney health professionals and a list of recommendations for addressing kidney health professionals' concerns in care, including with ADPKD. These have been used to inform [ADPKD clinical guidelines](#) and [ADPKD clinical practice guidelines](#).

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](#) is essential to improve ADPKD care.

Transplantation

ADPKD is associated for several years with kidney failure, leading to kidney transplantation. Kidney transplantation is the optimal treatment for kidney failure, providing excellent outcomes and quality of life for many ADPKD patients. Transplantation offers an excellent option for ADPKD patients with kidney failure.

Conclusion

Continued research into genetic testing, pre-implantation genetic diagnosis and transplantation is key to improve ADPKD care and outcomes for patients with kidney failure.

Genetic testing and pre-implantation genetic diagnosis are 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 vision and goals of the health system, and that they are able to deliver the care that is needed for the population.

Global needs in NCDs

NCDs are a major cause of premature mortality and disability worldwide. The burden of NCDs is increasing rapidly, and is expected to continue to do so in the coming decades. This is due to a combination of factors, including changes in diet and physical activity, increasing tobacco and alcohol use, and an ageing population. NCDs are a leading cause of death and disability, and are a major burden on health systems and economies. It is important that health systems are able to deliver the care that is needed for the population.

Primary prevention

Primary prevention is the first line of defence against NCDs. It involves promoting healthy behaviours and reducing risk factors. This can be done through a variety of measures, including education, legislation, and environmental changes. It is important that health systems are able to deliver the care that is needed for the population.

Multidisciplinary care

Multidisciplinary care involves the collaboration of different healthcare professionals to provide comprehensive care for patients. This can be done through a variety of measures, including education, legislation, and environmental changes. It is important that health systems are able to deliver the care that is needed for the population.

Technology update

Healthcare providers should stay up to date on the latest technology and research. This can be done through a variety of measures, including education, legislation, and environmental changes. It is important that health systems are able to deliver the care that is needed for the population.

Implementation

Implementation of NCD prevention and control strategies requires a coordinated effort across different sectors. This can be done through a variety of measures, including education, legislation, and environmental changes. It is important that health systems are able to deliver the care that is needed for the population.

Conclusion

Prevention and control of NCDs requires a coordinated effort across different sectors. This can be done through a variety of measures, including education, legislation, and environmental changes. It is important that health systems are able to deliver the care that is needed for the population.

Health systems should be able to deliver the care that is needed for the population. This can be done through a variety of measures, including education, legislation, and environmental changes. It is important that health systems are able to deliver the care that is needed for the population.

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	Association pour l'Information et la Recherche sur les maladies Rénales Génétiques (AIRG) Belgique
Finland	Munuais- ja maksaliitto (Finnish Kidney and Liver Organization)
France	Association Polykystose France (APKF) Association pour l'Information et la Recherche sur les maladies Rénales Génétiques (AIRG) France
Germany	PKD Familiäre Zystennieren e.V.
Ireland	Irish Kidney Association
Italy	Associazione Italiana Rene Policistico (AIRP)
Netherlands	Nierpatienten Vereniging Nederland (NVN)
Spain	Asociación para la Información y la Investigación de las Enfermedades Renales Genéticas (AIRG) España Federación Nacional de asociaciones para la lucha contra las enfermedades del riñón (ALCER)
Switzerland	SwissPKD Association pour l'Information et la Recherche sur les maladies Rénales Génétiques (AIRG) Suisse
Turkey	Turkish Society of Nephrology Cystic Kidney Diseases Working Group
UK	PKD Charity Genetics Alliance UK

North America

Canada	PKD Foundation of Canada
USA	PKD Foundation

Asia

Japan	PKD Foundation
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Australasia

Australia	PKD Foundation Australia
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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

Basic
management
and self-care

Predicting
the progress
of ADPKD

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to slow ADPKD
progression

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care

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renal disease

Dialysis

Kidney
transplantation

Post-transplant
care