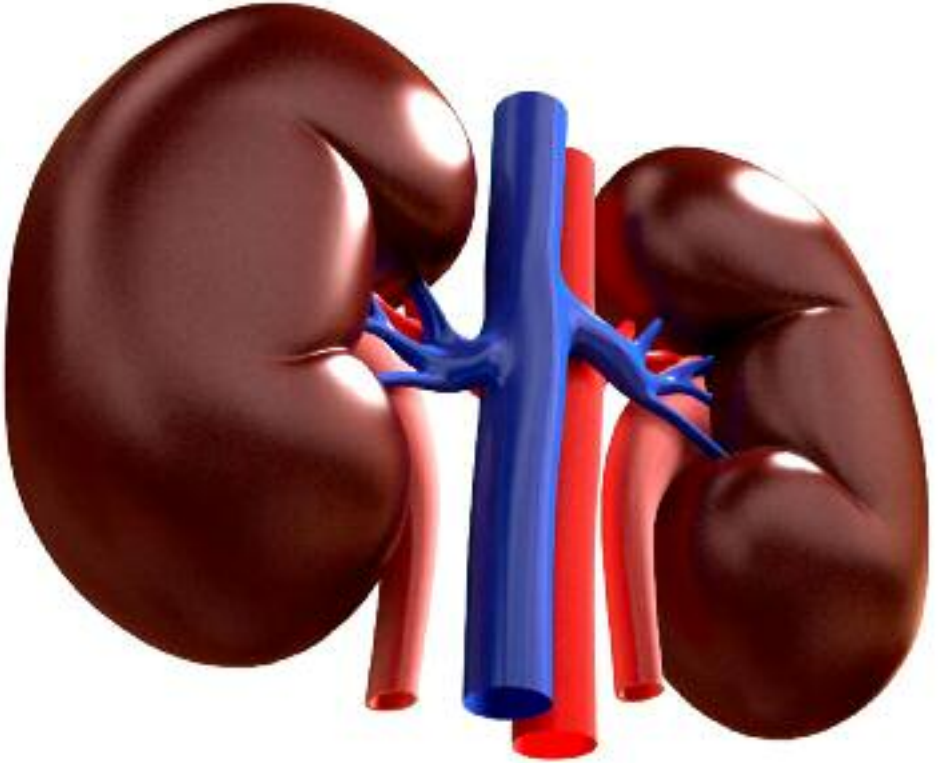


# LUPUS

and the Kidneys



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LUPUS UK is the national charity caring for those with systemic lupus erythematosus (SLE) and discoid lupus erythematosus (DLE) supporting our members through the Regional Groups and advising others as they develop the symptoms prior to diagnosis.

You can help by taking up membership  
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**[www.lupusuk.org.uk](http://www.lupusuk.org.uk)**

Reg. charity nos 1051610, SC039682

Please contact National Office should you require further information on the sources used in the production of this booklet or for further information about lupus. LUPUS UK will be pleased to provide a booklist and details of membership.

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NHS Foundation Trust

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## Introduction

This booklet gives general information about Systemic Lupus Erythematosus (SLE), what it is, common symptoms, treatments and how it can affect the kidneys.

**If you want any more information please ask your doctor.**

## What is lupus?

SLE, also known as 'lupus', is a long term condition that causes inflammation in various parts of the body. It is an autoimmune disease. Normally the body's immune system helps protect against infection but in lupus, the immune system does not work properly and produces autoantibodies that attack healthy cells and tissue.

Lupus can affect almost any part of the body including the skin, joints, blood vessels and organs such as the heart and kidneys.

People with lupus have symptoms that can range from mild to severe, and may come and go over time. Some of the most common symptoms include:

- Severe tiredness/fatigue
- Weight loss
- Fevers
- Skin changes (including a red 'butterfly' rash across the cheeks and nose or other sun exposed areas and scaly rashes that can appear on areas not exposed to sunlight)
- Painful, stiff or swollen joints (arthritis)
- Poor circulation in the fingers and toes which causes them to become pale (Raynaud's)

When lupus affects the kidneys, it is usually referred to as 'lupus nephritis'. Most often this causes no symptoms and is detected by testing the urine or blood for abnormalities. It is important to recognize since in severe cases, it can lead to permanent kidney damage. If lupus nephritis causes very large amounts of protein to leak from the kidneys, swelling in the legs or more generally ('oedema') may develop.

## Who gets lupus?

Anyone can develop lupus but it is much more common in young or middle aged women than men. It is also much more common in black and Asian people than in white people.



## What causes lupus?

The causes of lupus are not fully understood but are likely to be due to a combination of genetic and environmental factors. Lupus causes periods of illness ('flares') and periods of feeling well with no symptoms or signs of disease ('remission'). It is not infectious and cannot be caught from another person

## How is lupus diagnosed?

There is no single test for lupus. The diagnosis is usually made based on symptoms and a number of special blood tests which measure autoantibodies and levels of inflammation. These include:

- ANA (anti-nuclear antibodies)
- DNA antibodies (anti-double stranded DNA antibodies)
- Complement levels (C3 & C4)
- Blood count which looks at levels of different blood cells
- ESR - which looks at inflammation levels

## Who looks after patients with lupus?

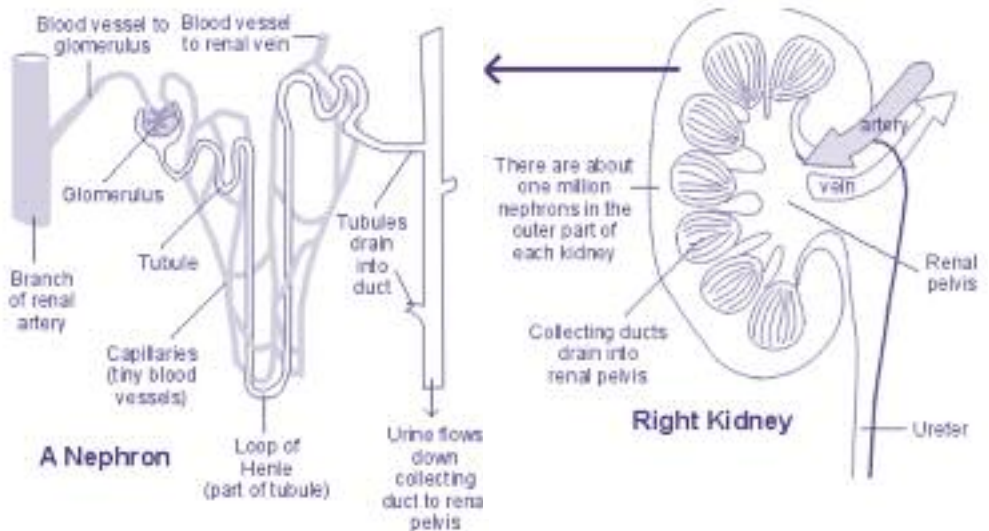
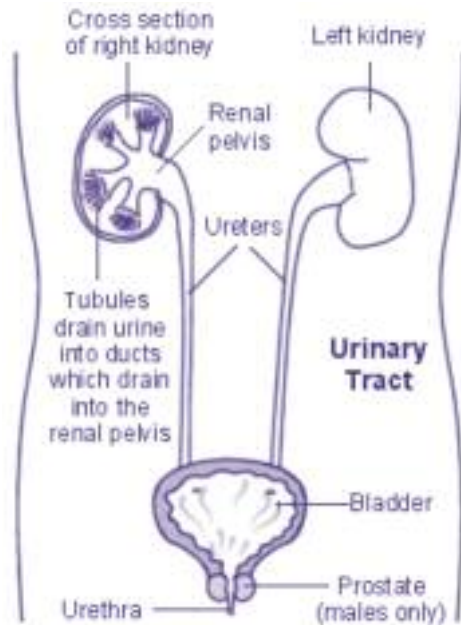
Many different doctors can be involved in looking after people with lupus but nephrologists (kidney doctors) and rheumatologists (arthritis specialists) are most often involved. Since lupus is a complicated condition, it is usually a good idea for you to see doctors who specialise in treating the disease. Your GP will also make an important contribution to looking after your lupus and your general health. You may see a specialist lupus nurse as well.

## About lupus and the kidneys

Lupus can affect the kidneys. You have two kidneys which are bean shaped and each about the size of a fist. They are below the ribcage, towards the back and sit to the left and right of the spine. Within each kidney there are about a million 'nephrons' which filter the blood. Blood is filtered to get rid of extra water and waste products. The filtered fluid forms urine. The kidneys are also involved in blood pressure control and in other aspects of health including producing vitamin D and maintaining a normal blood count (preventing anaemia).



The following diagrams show the position of your kidneys, the structure of the kidney and a nephron



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## What is lupus nephritis?

Lupus nephritis is a form of inflammation which affects the nephrons within the kidneys, occurring as they are attacked by the immune system. In most cases treatments are effective and kidney function is preserved. However for some patients, damage caused by scarring in the kidney means that less nephrons are able to work properly. This is referred to as 'chronic kidney disease or CKD'. After successful treatment of lupus nephritis, 'flares' or 'relapses' can still occur, sometimes years after the original diagnosis.

Inflammation in the kidneys from lupus can be difficult to identify as the person with lupus does not usually experience specific symptoms. However, it can be detected and monitored as described later in this booklet.

### **The following complications can occur with lupus nephritis:**

- 'Nephrotic syndrome'. If you suffer from nephrotic syndrome your kidneys leak very large quantities of protein (especially albumin) into your urine. When this happens fluid gathers in the tissues (often particularly in your legs) causing swelling (called oedema)
- Sudden deterioration of kidney function is rare in lupus but important to recognise
- Patients with lupus nephritis are more likely to have other organ threatening manifestations of lupus (such as, nervous system lupus) than those with lupus but without nephritis

## Who gets lupus nephritis?

Lupus nephritis is a complication of lupus. About one in three people with lupus develop kidney disease. Most often this happens within the first five years after diagnosis and commonly lupus nephritis is detected at the same time as the diagnosis of lupus is made but in other patients, nephritis develops later on. Younger patients and those from certain ethnic groups (including black and Asian patients) are more likely to develop lupus nephritis.

## How is lupus nephritis diagnosed?

Lupus nephritis may cause high blood pressure or swelling around the ankles, legs, fingers or face. However, often there are few or no symptoms and a diagnosis is made from various tests:

### **Urine tests**

Blood (haematuria) or protein (proteinuria) in the urine is a sign of kidney damage. These are the earlier and most important signs of lupus nephritis. Urine should be routinely tested when people with lupus are seen in clinic.

## **Blood tests**

The kidneys remove waste products from the blood. If the blood contains high levels of waste products, the kidneys are not working properly and kidney function is deteriorating.

Levels of creatinine in the blood are used to measure your kidney function and this is often converted into 'eGFR' which allows medical practitioners to calculate an approximate percentage of how well your kidneys are working.

Creatinine levels are often normal in people with active lupus nephritis and an abnormal result usually indicates that kidney damage is already severe. The goal is to investigate and start treatment before this happens.

## **A kidney biopsy**

A biopsy is a procedure to obtain a small piece of kidney tissue so it can be looked at under a microscope. This biopsy shows how much inflammation there is and whether there is any scarring to the kidneys.

# **What else do I need to know about lupus and the kidneys?**

A kidney biopsy allows your doctor to look at the kidney tissue and decide on the best treatment for you. Lupus nephritis is split into different categories from Class I (one) to Class V (five).

People with Class I or Class II lupus nephritis are unlikely to require powerful treatments. The types of nephritis that can potentially lead to permanent damage and CKD are 'class III' (three), 'class IV' (four) and 'class V' (five) nephritis. These are the conditions for which more powerful treatments are used.

CKD has important implications for your general health. Overall, only 10-20 people out of a hundred with lupus nephritis develop kidney failure requiring dialysis or transplantation and this generally happens over a number of years but less severe levels of CKD can still cause problems such as high blood pressure.

How well the treatment of lupus nephritis has worked is described in terms of:

- Complete remission - blood and protein disappear from the urine and kidney function keeps steady or improves,
- Partial remission - blood and protein in the urine reduce and kidney function keeps steady or improves
- Resistance - no real reduction in blood and protein in the urine and kidney function may get worse
- Relapse - nephritis flares (blood and protein reappear) after successful treatment has produced a remission

## Can the risk of worsening kidney disease be predicted?

Complete or partial remissions indicate a good chance of keeping kidney function steady in the long term whereas resistance or frequent relapses increase the risk of CKD.

Taking all the medication prescribed for you is very important and not doing so greatly increases risk of long term kidney damage.

## What treatments are used for lupus nephritis?

### The aim of treatment

The aim of treatment is to dampen down the inflammation and stabilize or improve kidney function. For lupus nephritis (especially class III, IV or V), 'immunosuppressive' drugs are used. Common immunosuppressive drugs include steroids, mycophenolate mofetil, rituximab and cyclophosphamide. Hydroxychloroquine is another important part of treatment for many patients with lupus. These drugs are also used to treat other features of lupus such as arthritis but often the diagnosis of lupus nephritis means that treatment needs to be increased to control the inflammation.

### Steroids

Steroids for lupus nephritis can either be given as intravenous injections directly into a vein over 1-3 days or as tablets (prednisolone) taken daily. Sometimes a combination of injected and tablet steroids is used.

### Mycophenolate

Mycophenolate is a tablet given on a daily basis. The dose of the mycophenolate is built up over a few weeks and then monitoring blood tests are needed to check that the drug is not causing any side effects.

### Rituximab

Rituximab is a newer biological drug which is given by intravenous injection into a vein. It works by removing B cells which are involved in producing autoantibodies which cause inflammation in lupus.

### Cyclophosphamide

Cyclophosphamide is a powerful medication used for reducing inflammation and is also usually given intravenously into the vein. Your doctor will explain how many doses of cyclophosphamide you are likely to need. These are usually given every fortnight over a period of three months. Antibiotics may be prescribed for





six months after the start of cyclophosphamide to reduce the risk of getting infections during and straight after the treatment.

### **How long does treatment last?**

Most treatment can be given as an out-patient and tablets will need to be continued for a period of years (often three years minimum and sometimes lifelong).

Immunosuppressive drugs, like any other drug, can cause side effects and your doctor and lupus nurse will talk to you about these.

### **Monitoring progress**

Regular blood tests, urine tests and blood pressure checks are carried out to look at kidney function, levels of inflammation, response to treatment and to monitor for side effects from treatments. These results will be discussed with you at clinic appointments.

A further kidney biopsy may be needed to look at the response to treatment or if the inflammation becomes active again.

## **What else can help?**

In addition to immunosuppressive drugs the following steps are also important:

- Keeping blood pressure under control (usually with tablets such as 'ACE inhibitors' or related drugs)
- Preventing infections - it is advisable to avoid sources of infection where possible and it is generally safe and recommended to have an annual influenza vaccination and a regular pneumococcal vaccination to reduce the risk of serious infections.
- Eating a well balanced diet and maintaining a healthy weight. This usually includes reducing the amount of salt in your diet.
- Not smoking - stopping smoking is something that can really improve your general health and well-being particularly when you have lupus. Help with stopping smoking is available from your family doctor's surgery and local chemists displaying - Smoking Cessation Available Here signs.
- Taking regular exercise
- People with lupus nephritis may be at greater risk of heart attacks and stroke and so it is important to reduce risk factors such as high cholesterol levels. Your cholesterol level should be checked on an annual basis either at your hospital appointment or through your family doctor.
- People with lupus nephritis, especially if taking steroid tablets can be at risk of developing osteoporosis (brittle bones) and so medication to prevent or treat this may also be needed.



## Lupus and chronic kidney disease (CKD)

For patients who develop permanent kidney damage due to lupus nephritis, this usually happens gradually over a period of years, but there may be a sudden deterioration if the nephritis flares up. When it becomes clear that someone has kidney damage that can't be improved with medication, help is given to plan ahead in the same way as with patients who have advanced chronic kidney disease due to other causes.

This usually involves choosing the type of treatment (e.g. dialysis) that a person wishes to have. A person's potential suitability to undergo a kidney transplant is also considered.

## Lupus and dialysis

In general, people with lupus do as well on dialysis as those with kidney failure due to other causes. There is some added risk of infection from immunosuppressive drugs when someone is on dialysis; however, this is carefully monitored. Usually, lupus becomes less active at this stage and it is often possible to gradually reduce and sometimes stop immunosuppressive drugs once someone with lupus is on regular dialysis treatment.

## Lupus and kidney transplant

Overall, people with lupus do very well after a kidney transplant. The risks of lupus recurring in a transplant kidney are very low (less than 5%) and in general, transplant medications also keep other lupus symptoms under control. Some patients with lupus also have an increased risk of clotting. Your kidney doctor will discuss with you whether this might affect a transplant kidney and what steps can be taken to help.

## Lupus family planning and pregnancy

### **Pregnancy**

Many women with lupus, including those with nephritis, have successful pregnancies. There are, however, increased risks of complications and it is very important to discuss this with your kidney doctor or rheumatologist before considering pregnancy. It is important that your lupus is in remission before pregnancy.

## **Medications**

Some immunosuppressive medications, in particular cyclophosphamide, mycophenolate and rituximab are unsafe in pregnancy and treatment should be altered before trying to conceive. Other medications including 'ACE inhibitors' such as Ramipril or angiotensin receptor blockers for blood pressure should be stopped before trying to conceive or as soon as a woman knows that she is pregnant. Steroids, azathioprine and hydroxychloroquine can be used in pregnancy.

## **Abnormal kidney function**

For women with abnormal kidney function or significant amounts of protein in the urine, the risks from pregnancy include a form of high blood pressure (called pre-eclampsia), pregnancy loss, very premature birth, low birth weight babies and worsening of the mother's kidney function. It is especially important that woman with kidney problems discuss this with their specialist before planning a pregnancy. It is likely that the pregnancy will be more carefully monitored to reduce the risks and be able to react quickly if a problem develops.

## **Anti-phospholipid syndrome**

Some women with lupus also have a condition called 'antiphospholipid syndrome' which leads to blood clots and can trigger miscarriage. A kidney doctor or rheumatologist can test you for this condition before you become pregnant.

## **Men with lupus**

Men with lupus should also discuss their medication with a specialist before trying to have a baby (in particular men recently treated with cyclophosphamide or mycophenolate should not conceive a baby) and with regards to contraception.

## **Contraception**

An appropriate and reliable form of contraception is important for both women and men with lupus who are taking immunosuppressive medications such as mycophenolate, rituximab, cyclophosphamide, azathioprine. (See the separate LUPUS UK booklet - Lupus A Guide to Pregnancy.)

## **What about the contraceptive pill?**

Standard contraceptive pills contain a hormone oestrogen which may not be recommended if you have lupus. Alternative contraceptive pills which contain progesterone only or low dose oestrogen contraceptive pills may be more acceptable. You should discuss this with your lupus/kidney doctor. Your family doctor and family planning clinics are able to give advice regarding other methods of contraception such as barrier methods i.e. condoms.



# Glossary

**Antiphospholipid Syndrome (APS)** - A disorder where the blood is sticky and tends to clot too quickly. This can affect veins or arteries anywhere in the body resulting in various symptoms. APS can be secondary to lupus or occur on its own (Primary APS).

**Autoantibodies** - Antibodies that attack the body's own tissue rather than viruses or bacteria.

**Autoimmune disease** - A condition where the body's defence (immune system) attacks the body's own tissues rather than attacking germs.

**Biopsy** - Removing a small amount of body tissue - this allows the tissue to be looked at under a microscope to diagnose whether the tissue is diseased.

**Cholesterol** - Fatty substance found in the bloodstream, high levels can increase the risk of heart attacks and strokes

**Dialysis** - This is a way of supporting or replacing kidney function. There are different types of dialysis available which allow waste products and unneeded water to be removed from the blood.

**Immune system** - Body tissues that help the body to fight infection.

**Immunosuppression** - Drugs used to dampen down the immune system in conditions where the body is attacking its own tissues.

**Inflammation** - Bodily reaction to infection or injury of living tissue.

**Kidney transplant** - An operation where a healthy kidney is given to someone whose kidneys are severely damaged

## Additional information

These websites provide useful information for patients with lupus and their families

- [www.lupusuk.org.uk](http://www.lupusuk.org.uk)
- [www.uklupus.co.uk](http://www.uklupus.co.uk)
- [www.westmidlandslupus.co.uk](http://www.westmidlandslupus.co.uk)
- [www.arthritisresearchuk.org](http://www.arthritisresearchuk.org)

## Other sources of information

- The National Kidney Federation website [www.kidney.org.uk](http://www.kidney.org.uk)
- Go online and view NHS Choices website for more information about a wide range of health topics [www.nhs.uk/Pages/HomePage.aspx](http://www.nhs.uk/Pages/HomePage.aspx)
- You will also find local information leaflets within the different renal departments.





Publicity materials, leaflets, posters, a dvd for the newly diagnosed, media releases and more are always available from the charity's National Office for better awareness about lupus in clinics, hospitals and public places.