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Registered Non-Profit Organisation
No 002-847 NPO

LUPUS (SLE)

Systemic Lupus Erythematosus

GET IN TOUCH

Questions and requests for information from members
and non-members are welcome.

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This information leaflet is published by the
Arthritis Foundation as part of its continuing education
programme for all people with arthritis.

About this booklet

This booklet is written for people with lupus and their families and friends. It explains how lupus develops, how it affects different parts of the body, how it can affect people in different ways and how doctors diagnose it. We then explain how doctors diagnose it. We then explain how it can be treated and offer hints and advice on living with it more easily. Near the end of the booklets you will find information on how to contact the Arthritis Foundation and a brief glossary of medical words (like cartilage). We have put these in italics when they are first used in the booklets.

Introduction

Lupus is a form of arthritis that mainly affects women during their childbearing Years. It is one of the autoimmune rheumatic diseases, caused by a fault in the body's immune system. The immune system normally produces antibodies to fight infections. In people with autoimmune diseases, antibodies are produced which against certain body tissues and cause inflammation.

When we produced an earlier edition of this booklet, around half of those who wrote to Arthritis Foundation about it complained that the booklet did not emphasise the serious nature of the disease. The other half complained, equally bitterly, that it made lupus appear to be worse than it actually is. Both points of view are valid. For many people lupus represents not much more than a nuisance condition, but for some the disease is very troublesome, even life threatening.

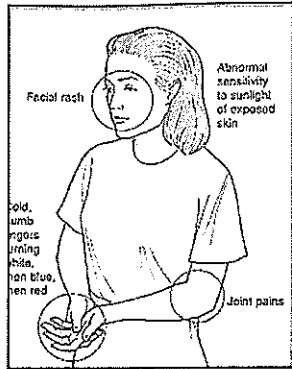
This uncertainty is a challenge to both the individual and the doctors who are trying to help.

Lupus (Latin for wolf) takes its name from the fact that it can cause serious rashes across the cheeks and nose (rather fancifully resembling the face of a wolf). However, thanks to modern treatment we rarely see these very severe rashes nowadays.

There are two main forms of lupus. One, discoid lupus, affects only the skin whereas the other, systemic lupus, involves the joints and also involve the internal organs as well. The full name of this is systemic lupus erythematosus – 'erythematosus' simply meaning red'. This booklet deals only with systemic lupus erythematosus (sometimes shortened to 'SLE').

What are the common symptoms of lupus?

Lupus can mimic many different diseases. However, it usually starts with joint pains, especially in the small joints of the hands and feet, which may flit from one set of joints to another quite quickly. Skin rashes are also common and these are often made worse when exposed to strong sunlight. You may develop groups of mouth ulcers (not just the one or two that everyone gets from time to time) and these can come back again and again. You may also feel completely



exhausted and this can be one of the most common and difficult features of systemic lupus. For many people the disease does not cause symptoms or complications other than tiredness, joint pains and skin rashes. However, in some people it may cause one or more other problems. The challenge facing the doctor looking after people with lupus is to try and identify those who might develop other problems and treat them accordingly.

The remainder of this booklet explains in some detail the nature of lupus and how it might affect some people, mainly with the more severe form of the disease.

Who gets lupus?

Lupus affects mainly young women. It is much commoner among black women living in Europe and America (though strangely it is rare in Africa) and among Chinese and Asian women. Around one in 250-500 women of Afro-Caribbean origin develop lupus, compared to one in China and one in 4000 Caucasians in New Zealand. It is about nine times as common in women as in men. About one in ten cases begin after the age of 50.

What causes lupus?

There is no one cause of systemic lupus erythematosus. It seems to be due to a combination of factors, including environmental factors (such as previous infections), the balance of hormones in the body, and genetic factors. However, lupus is not an inherited disease it is very rarely passed on in the family. There are many basic problems affecting the immune system in people with this disease. The immune system is your body's self-defence system, made up of cells that patrol throughout the body, in the blood stream, lymphatic system and organs.

These cells search out and destroy foreign invaders (such as viruses, bacteria). Unfortunately, when you have systemic lupus erythematosus your immune system cannot distinguish between your own tissue and foreign invaders. It begins to attack your own body.

One of the results of this is that proteins called antibodies are formed. Because the body is attacking itself; these are also known as auto antibodies. They can bind to various parts of your body tissues and in severe forms of lupus can cause damage to various organs including the kidneys, heart, lungs, skin and brain.

Which parts of the body might be affected?

The skin

It is common for a rash to develop over parts of your body which are exposed to the sun; the face, wrists and hands. However, sometimes these rashes can affect other parts of the body. Poor circulation can cause the hands to change colour, with the skin going white, blue and red in the cold. This common problem is known as Raynaud's phenomenon, after a famous French physician, Maurice Raynaud.

The hair

Loss of hair (alopecia) is common with lupus. This is a minor problem for most people but occasionally you may have quite serious hair loss. As with the other features of this disease hair loss may 'come and go'. You will probably find that your hair grows back. You may prefer to wear a wig if you have serious hair loss (though this is rare).

The joints

Unlike rheumatoid arthritis, lupus does not usually cause damage and deformity in the joints although pain and swelling can be the main symptoms of the disease. Perhaps one in every 20 people with lupus develop more severe joint problems which may mimic those of people with rheumatoid arthritis.

The Kidneys

Estimates vary but up to one third of people with lupus may develop some inflammation of the kidneys. This does not usually cause problems unless the disease is very severe. Your urine can be tested with a special plastic stick which is coated with various chemicals (a dipstick). This can show if there is protein or blood in the urine. Severe lupus may seriously damage the kidneys but this is rare. This can be treated by modern forms of kidney treatment including dialysis or transplantation.

Blood pressure

Lupus can cause high blood pressure, particularly if the kidneys are involved. Steroid tablets in high dose may also cause an increase in blood pressure. Treatment with drugs to suppress the immune system (see later) can often control and occasionally cure high blood pressure caused by kidney involvement. Also, the various drugs available specifically to control blood pressure are very effective.

The brain and nervous system

You may feel depressed and anxious from time to time. It can be hard to tell whether this is part of the disease itself or simply the normal reaction to having a serious illness.

Many people have problems in coming to terms or coping with the disease. It is important to share these feelings with the doctor or other medical professionals looking after you.

The medical team can also help you with counselling if you need it. Migraine affects as many as one in three people with lupus. Although lupus can sometimes mimic other neurological conditions, this is rare. However, if lupus does affect the nervous system, it can cause fits or other problems. Often, treatment will control or cure the symptoms so, it is important to ask your doctor about any problems you have.



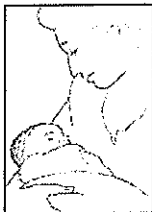
The heart and lungs

Occasionally lupus may affect these organs directly. More often, it inflames the lining tissues around the heart and lungs (the pericardium and pleura). This causes pericarditis or pleurisy with breathlessness and sharp pains in the chest which are worse when taking a deep breath.

Rarely, large amounts of fluid may develop in these lining layers, causing severe shortness of breath. This symptom can be treated successfully by removing the fluid.

Other organ involvement

Lupus rarely affects the gut, pancreas, liver, or spleen. Lupus may affect the bone marrow and cause anaemia, or a reduction in the number of platelets (which help the blood to clot) or white blood cells. It may, on occasion, cause swelling of the lymph glands, for example in the neck, which may cause discomfort. Some people with lupus have tendency to develop blood clots in veins or arteries. This is usually caused by antiphospholipid and anticardiolipin antibodies. These can also affect pregnancy (see below)



How do doctors diagnose lupus?

Doctors use a range of tests to check if you have lupus, including:

- A wide variety of blood tests mainly looking for types of autoantibodies. One of the classic tests looks for antinuclear antibodies and antibodies to double-stranded DNA (which makes up genes and chromosomes). Antibodies structures known as Ro and La (combinations of proteins in cell nucleus) are associated with the neonatal (new-born) lupus syndrome or an accompanying Sjogren's syndrome. Antibodies to Ro alone are more commonly associated with skin rashes. People with antiphospholipid antibodies have increased risk of miscarriage (see later) and of developing blood clots.
- Measurements of certain enzymes which make up of the immune system known as 'complement'. These are measured frequently to assess the activity of the disease. The most common of these measurements are those known as C3 and C4, which are low when the disease is active.
- Regular blood tests to check bone marrow and kidney function so that any problems can be swiftly treated. Depending which organ, the doctor suspects of being involved, a wide variety of tests are available to check the function of the hearts, lungs, liver, and spleen. These include x-rays, ultrasound, scans computer tomographic (CT) scans and magnetic resonance imaging (MRI) scans. The kidney can also be checked with further tests such as kidney filtration tests.

How can lupus be treated?

Lupus can not be cured but it can be controlled.

If you have joint pains and skin rashes, you can often be treated with just non-steroidal anti-inflammation drug (NSAIDs) and/ or an antimalarial drug, usually hydroxychloroquine (Plaquenil), which is also effective at treating the tiredness or exhaustion which can be caused by the disease. Non-steroidal anti-inflammatory drugs can occasionally irritate the lining of the stomach, so it is important for your doctor to arrange regular monitoring of your need for these drugs. There is a considerable debate among doctors as to whether eye tests need to be carried out on a regular basis for people taking Plaquenil. Most doctors continue to ask for these eye tests as a precaution, although very few have ever seen any serious problems due to this drug. If you have more serious complications, such as pericarditis, you will be given corticosteroids (often called 'steroids'). These drugs were introduced in the late 1940s and are remarkably effective, sometimes lifesaving. The dose prescribed will be kept as low as possible to minimise the possible side-effects. These include lowered resistance to infections, osteoporosis, high blood pressure and diabetes.

When lupus is particularly active, it can cause inflamed kidneys, severe anaemia or a very low platelet count. In these cases, you will usually be given very high doses of steroids together with other immunosuppressive drugs including aza-thioprine, cyclophosphamide and possibly cyclosporin (Neoral). These are powerful drugs which need to be carefully monitored with blood and urine tests.

The doctor will usually reduce them as quickly as possible as the activity of the disease subsides.

For the most difficult cases of lupus other forms of therapy can be used, including the use of intravenous gamma globulin injections and plasma exchange. However the occasions on which these are needed are rather few and far between.

What are the long-term prospects if I have lupus?

For many people, lupus is often no more than a nuisance. However, for some people it can be a very troublesome and unpredictable condition. If you have one of the more serious forms of lupus, you should be treated by a rheumatologist with an interest in this condition, sometimes in a 'lupus clinic'. Only careful monitoring over many years will make sure the disease is neither over- nor under-treated, so it is essential for all people with lupus to be assessed regularly.

What can I do to help myself?

There are number of things you can do to help yourself:

Stay out of the sun

Avoid sunbathing. Too much ultra-violet radiation can flare both the skin rash and sometimes the lupus in the internal organs. If you must go to a hot climate, use a sun-blocking cream, factor 25 or greater. This is available on prescription for people with lupus.

Avoid infections

If you have lupus, then you are more susceptible to infection. Take sensible Precautions. For example, if you are already on immunosuppressive drugs, avoid contact with friends and relatives who have infectious disease.

Watch what you eat

People are very interested in whether diet therapy can help control lupus. Unfortunately, the evidence is very limited. There is some evidence to suggest that a diet low in saturated fat and supplemented by fish oil may be helpful.

Avoid stress

Try to avoid situations which you know will cause stress

Lupus in special situations

Conception and pregnancy

You should use contraceptive pills which contain only progesterone or low dose oestrogen, or consider physical methods of contraception (such as the condom or the cap). If you are given oestrogen after the menopause as a treatment for thin bones (osteoporosis) the risk of a flare developing is slightly increased to about one in seven.

There is conflicting evidence about whether pregnancy itself is more likely to make lupus flares up. However, if your disease is well under control at the time the baby is conceived and do not have kidney disease you will probably go through pregnancy with little trouble. If you do have kidney disease, then you may well have increased protein in your urine during the later stages of pregnancy. If you have high levels of an antibody, known as antiphospholipid antibodies, then you are more likely to suffer a miscarriage.

Pregnancy is normally safe, but seek your doctor's advice before starting a baby Only about one in twenty mothers with lupus have antibodies to structures known as Ro and La. If you have these antibodies, then your baby may have a brief skin rash or a form of heart disease which may require a pacemaker to be fitted shortly after birth.

Doctors are naturally worried about using drugs during pregnancy. It is usually safe to use corticosteroids and there are now many examples of safe use of hydroxychloroquine and azathioprine throughout pregnancy. However, your doctor will try to avoid using these drugs if possible.

Lupus in childhood

Lupus is rather rare in children. It does not usually occur before the age of five, although It has been described in children as young as two. The symptoms and type of organ involvement is somewhat different from those seen in adults -one large recent study showed, for example, that haematological (blood-related) involvement is more common in children. Broadly speaking, though, children with lupus will be treated in much the same way as adults.

Lupus after 50

Relatively few cases (about 10%) begin in this age group and it is often thought that lupus activity begins to decrease after the menopause. However, there is one extra problem for older people with lupus if they are treated with corticosteroids. As mentioned earlier, one of the side-effects of corticosteroids is to cause thinning of the bones(osteoporosis). This can increase the risk of bone fractures as older people are already more likely to have osteoporosis because of their age. Overall, in people over 50, lupus tends to be less severe, but the side-effects of treatment can be more of a problem than in younger people.

What other diseases can affect me if I have lupus?

Around one in five people with lupus also suffer from Sjogren's syndrome. This causes dryness of the eyes and mouth, and is named after a Swedish ophthalmologist, Henrik Sjogren.

A little under one in ten people with lupus suffer from an autoimmune disease which affects the thyroid gland. This may occasionally make the gland overactive but more usually it makes it underactive. An underactive thyroid gland is often swollen. It causes dry skin, weight gain, and can make you 'slow down' mentally. However, all these symptoms are cured by taking thyroid hormone tablets. Much less often, people with lupus may develop rheumatoid arthritis or an inflammation of the muscle. Very rarely, other autoimmune disease may also develop in people with lupus. Most of these additional complications respond well to drug treatment.

Questions and answers**Is this disease hereditary?**

No. Though genetic factors are involved in the development of the disease, many other factors are also involved so it is extremely rare for children of people with lupus to develop the condition.

How much should I rest?

There is no simple answer to this question. When the disease is active, you may not feel like doing very much. When the disease is inactive, you will not gain anything from 'excessive' resting. Activity will help to keep your mind off the disease. Many people with lupus do however complain that tiredness is a significant problem. Your body will dictate how much sleep and rest are needed, though some drugs, particularly hydroxychloroquine and steroids, will help to balance the situation.

What about other forms of treatment?

Acupuncture may relieve pain. Like many forms of alternative pain relief, it seems to depend very much on whether you believe it will work.

My hands and feet go blue in the cold weather.**Can I do anything about this?**

This problem is almost certainly due to Raynaud's phenomenon which is caused by spasm of the small blood vessels of the hands and feet. Make sure you keep warm and wear thick gloves and socks/tights.

Does stress affect the disease?

Worrying will probably not affect how your lupus develops. However, it will obviously make you feel much worse. Your doctor may be able to help by arranging for you to see a psychologist who will suggest ways of coping designed for you as an individual. This is something which many people find very helpful.

Does the weather make any difference?

The climate will not alter the course of the disease. Some of the worst cases are found in people who live in tropical climates. Although cold, damp weather may make the problem seem worse, it does not really do any specific damage.

Are immunisations safe?

In the main, yes. However, we do not recommend 'live virus vaccines' if you are on immuno-suppressive drug. These vaccines include yellow fever, 'live' typhoid and oral poliomyelitis. Make sure that you check with your doctor before being immunised.

Summary

Lupus causes troublesome symptoms rather than serious problems in most people. However, effective treatment is available for the vast majority of people with lupus.

Anaemia- a shortage of haemoglobin (oxygen carrying pigment) in the blood resulting in a decrease in the ability of the blood carry oxygen around the body. Anaemia can be caused by a shortage of iron in the diet.

Antibodies- blood proteins which are usually formed in response to germs, Viruses or any other substances which the body sees as foreign or dangerous. The role of antibodies is to attack these foreign substances and make them harmless.

Anticardiolipin antibodies - antibodies which form against a certain type of phospholipid, cardiolipin.

Antiphospholipid antibodies-antibodies which form against phospholipids (a type of fat) which often make up the surface of cells.

Autoantibodies- antibodies which, instead of reacting with germs or viruses, react with the body's own tissues.

Autoimmune disease- a disorder of the body's defence mechanism (immune system), in which antibodies and other components of the immune system attack the body's own tissues.

Cartilage - strong material on bone ends that act as a shock absorber. Its slippery surface allows smooth movement between bones.

Computer tomographic (CT) scan - a type of scan which records images of sections or 'slices' of the body using x-rays. These images are then transformed by a computer into cross-sectional pictures.

Corticosteroids - drugs which have a very powerful effect on inflammation. Often called 'steroids' for short. The adrenal glands in the body produce a 'natural' supply but much larger doses are used to treat autoimmune diseases.

Diabetes - the most common form is properly called diabetes mellitus. This is a condition in which the body does not deal with sugars properly due to damage the pancreas gland causing a lack of the hormone insulin.

Dialysis - a method of separating particles in a liquid using a semi-permeable membrane. In kidney dialysis the patient's blood is circulated through a special machine which uses this method to remove waste materials or poisons from the blood.

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Immune system- the tissues which enable the body to resist infection. They include the thymus, the bone marrow, and the lymph nodes (see also 'lymphatic system' below).

Lymphatic system - a network of vessels in the body which transport lymph fluid from the tissues to the bloodstream. It plays an important part in the body's immune system and includes the lymph nodes which filter out foreign substances (antigens).

Magnetic resonance imaging (MRI) - a type of scan which uses high-frequency radio waves to build up pictures of the inside of the body. It works by detecting water molecules in the body's tissue. **NSAIDs** - non-steroidal anti-inflammatory drugs. A large family of drugs, prescribed for different kinds of arthritis, which reduce inflammation and control pain, swelling and stiffness.

Oestrogen - one of a group of hormones in the body which controls female sexual development. Synthetic oestrogen is used in the most common oral contraceptive pill known as the 'combined' pill. The combined pill contains both oestrogen and progesterone.

Osteoporosis - a condition where bones become less dense and more fragile, and more fragile, and therefore easier to break or fracture.

Pericarditis- inflammation of the membranes around the heart. The more acute (short - lasting) form causes fever and chest pains, and fluid may build up in between the membranes, causing shortness of breath.

Pericardium- the membranous 'sac' around the heart (see 'pericarditis' above).

Platelets - disc-shaped cells in the blood which help the blood clot when there is bleeding.

Pleura - the linings of the lungs and of the inside of the chest wall (see 'pleurisy' below).

Pleurisy - inflammation of the pleura (the lining of the lungs and of the inside surface of the chest wall), causing pain on deep breathing.

Progesterone - one of a group of hormones in the body. In women it is responsible for preparing the lining of the uterus for pregnancy. Synthetic progesterone is used in the oral contraceptive pill (both in the 'combined' pill, which contains oestrogen and progesterone, and in the progesterone-only pill).

Raynaud's phenomenon- a circulatory problem which causes the blood supply to certain parts of the body to be greatly reduced. It can make the fingers turn white, then blue, then red. Raynaud's phenomenon can also occur with the condition scleroderma (see arc booklets Raynaud's Phenomenon and Scleroderma).

Rheumatoid arthritis – a common inflammatory disease affecting the joint, mainly starting in the smaller joints in a symmetrical pattern (that is, for example, both hands or both wrists at one). See arc booklet Rheumatoid Arthritis.

Ultrasound scanning - a type of scan which uses high-frequency sound waves to examine and build up pictures of the inside of the body.