



Autoimmune Resource and Research Centre

Information Sheet

Systemic Lupus Erythematosus Defining Lupus

Lupus is a disorder of the immune system known as an autoimmune disease. In autoimmune diseases, the body harms its own healthy cells and tissues. This leads to inflammation and damage to various body tissues. Lupus can affect many parts of the body, including the joints, skin, kidneys, heart, lungs, blood vessels, and brain. Although people with the disease may have many different symptoms, some of the most common ones include extreme fatigue, painful or swollen joints (arthritis), unexplained fever, skin rashes, and kidney problems.

At present, there is no cure for lupus. However, lupus can be very successfully treated with appropriate drugs, and most people have a mild form of the disease and can lead active, healthy lives. Lupus is characterised by periods of illness, called flares, and periods of wellness, or remission. Understanding how to prevent flares and how to treat them when they do occur helps people with lupus maintain better health. Intense research is underway and scientists are continuing to make great strides in understanding the disease, which may ultimately lead to a cure.

Two of the questions researchers are studying are who gets lupus and why. We know that many more women than men have lupus. Lupus can run in families, but the risk that a child or brother or sister of a patient will also have lupus is still quite low.

Although "lupus" is used as a broad term, there actually are several kinds of lupus:

- Systemic lupus erythematosus (SLE) is the form of the disease that most people are referring to when they say "lupus". The word "systemic" means the disease can affect many parts of the body. The symptoms of SLE may be mild or serious. Although SLE usually first affects people between the ages of 15 and 45 years, it can occur in childhood or later in life as well. This booklet focuses on SLE.
- Discoid lupus erythematosus refers to a skin disorder in which a red, raised rash appears on the face, scalp, or elsewhere. The raised areas may become thick and scaly and may cause scarring. The rash may last for days or years and may recur. A small percentage of people with discoid lupus have or develop SLE.
- Drug-induced lupus refers to a form of lupus caused by specific medications. Symptoms are similar to those of SLE (arthritis, rash, fever, and chest pain) that typically go away when the drug is stopped.
- Neonatal lupus is a rare form of lupus affecting newborn babies of women with SLE or certain other immune system disorders. At birth, the babies have a skin rash, liver abnormalities, or low blood counts, which entirely go away over several months. However, babies with neonatal lupus may have a serious heart defect. Physicians can now identify most at-risk mothers, allowing for prompt treatment of the infant at or before birth. Neonatal lupus is very rare, and most infants of mothers with SLE are entirely healthy.

Understanding What Causes Lupus

Lupus is a complex disease whose cause is unknown. It is likely that there is no single cause but rather a combination of genetic, environmental, and possibly hormonal factors that work together to cause the disease. The exact cause may differ from one person to another. Scientists are making progress in understanding the processes leading to lupus. Research suggests that genetics plays an important role; however, no specific "lupus gene" has been identified. Instead, it appears that several genes may increase a person's susceptibility to the disease.

The fact that lupus can run in families indicates that its development has a genetic basis. However, scientists think that genes alone cannot account for who gets lupus. Other factors must also play a role. Some of the factors that scientists

are studying include sunlight, stress, certain drugs, and infectious agents such as viruses. Even though a virus might trigger the disease in susceptible individuals, a person cannot "catch" lupus from someone else.

In lupus, the body's immune system does not work as it should. A healthy immune system produces substances called antibodies that help fight and destroy viruses, bacteria, and other foreign substances that invade the body. In lupus, the immune system produces antibodies against the body's healthy cells and tissues. These antibodies, called autoantibodies ("auto," means self), contribute to the inflammation of various parts of the body, causing damage and altering the function of organs and tissues.

Symptoms of Lupus

Each person's experience with lupus is different, although there are patterns that permit accurate diagnosis. Symptoms can range from mild to severe and may come and go over time. Common symptoms of lupus include painful or swollen joints, unexplained fever, and skin rashes, along with extreme fatigue. A characteristic skin rash may appear across the nose and cheeks, the so-called butterfly or malar rash. Other rashes occur elsewhere on the face and ears, upper arms, shoulders, chest, and hands.

Other symptoms of lupus include chest pain, hair loss, sensitivity to the sun, anaemia (a decrease in red blood cells), and pale or purple fingers and toes from cold and stress. Some people also experience headaches, dizziness, depression, or seizures. New symptoms may continue to appear years after the initial diagnosis, and different symptoms can occur at different times.

Common Symptoms of Lupus

- Painful or swollen joints and muscle pain
- Unexplained fever
- Red rashes, most commonly on the face
- Chest pain upon deep breathing
- Unusual loss of hair
- Pale or purple fingers or toes from cold or stress (Raynaud's Phenomenon)
- Sensitivity to the sun
- Swelling (edema) in legs or around eyes
- Swollen glands
- Extreme fatigue

In some people with lupus, only one system of the body such as the skin or joints is affected. Other people experience symptoms in many parts of their body. Just how seriously a body system is affected also varies from person to person. Most commonly, joints and muscles are affected, causing arthritis and muscle pain. Skin rashes are quite common. The following systems in the body also can be affected by lupus:

- **Kidneys** - Inflammation of the kidneys (nephritis) can impair their ability to get rid of waste products and other toxins from the body effectively. Because the kidneys are so important to overall health, lupus affecting the kidneys generally requires intensive drug treatment to prevent permanent damage. There is usually no pain associated with kidney involvement, although some patients may notice that their ankles swell. Most often the only indication of kidney disease is an abnormal urine or blood test.
- **Lungs** - Some people with lupus develop Pleuritis, an inflammation of the lining of the chest cavity that causes chest pain, particularly with breathing. Patients with lupus also may get pneumonia.
- **Central nervous system** - In some patients, lupus affects the brain or central nervous system. This can cause headaches, dizziness, memory disturbances, vision problems, stroke, or changes in behaviour.

- Blood vessels - Blood vessels may become inflamed (vasculitis), affecting the way blood circulates through the body. The inflammation may be mild and may not require treatment or may be severe and require immediate attention.
- Blood - People with lupus may develop anaemia, leucopenia (a decreased number of white blood cells), or a decrease in the number of platelets (thrombocytopenia). Some people with lupus may have abnormalities that cause an increased risk for blood clots.
- Heart - In some people with lupus, inflammation can occur in the heart itself (myocarditis and endocarditis) or the membrane that surrounds it (pericarditis), causing chest pains or other symptoms. Lupus can also increase the risk of atherosclerosis.

Diagnosing Lupus

Diagnosing lupus can be difficult. It may take months or even years for doctors to piece together the symptoms to diagnose this complex disease accurately. Making a correct diagnosis of lupus requires knowledge and awareness on the part of the doctor and good communication on the part of the patient. Giving the doctor a complete, accurate medical history (for example, what health problems you have had and for how long) is critical to the process of diagnosis. This information, along with a physical examination and the results of laboratory tests, helps the doctor consider other diseases that may mimic lupus, or determine if the patient truly has the disease. Reaching a diagnosis may take time and occur gradually as new symptoms appear.

No single test can determine whether a person has lupus, but several laboratory tests may help the doctor to make a diagnosis. The most useful tests identify certain autoantibodies often present in the blood of people with lupus. For example, the antinuclear antibody (ANA) test is commonly used to look for autoantibodies that react against components of the nucleus, or "command centre", of the patient's own cells. Most people with lupus test positive for ANA; however, there are a number of other causes of a positive ANA besides lupus, including infections, other rheumatic or immune diseases, and occasionally as a finding in normal healthy adults. The ANA test simply provides another clue for the doctor to consider in making a diagnosis. In addition, there are blood tests for individual types of autoantibodies that are more specific to people with lupus, although not all people with lupus test positive for these and not all people with these antibodies have lupus. In other words, no single blood test makes the diagnosis of Lupus.

Some tests are used less frequently but may be helpful if the cause of a person's symptoms remains unclear. The doctor may order a biopsy of the skin or kidneys if those body systems are affected. Again, all these tests merely serve as tools to give the doctor clues and information in making a diagnosis. The doctor will look at the entire picture - medical history, symptoms, and test results, to determine if a person has lupus.

Other laboratory tests are used to monitor the progress of the disease once it has been diagnosed. A complete blood count, urinalysis, blood chemistries, and erythrocyte sedimentation rate (ESR) test can provide valuable information.

Treating Lupus

Diagnosing and treating lupus is often a team effort between the patient and several types of health care professionals. A person with lupus can go to his or her family doctor, or can visit a rheumatologist. A rheumatologist is a doctor who specialises in rheumatic diseases (arthritis and other diseases of the joints, bones, and muscles). Clinical immunologists (doctors specialising in immune system disorders) also treat people with lupus. As treatment progresses, other professionals often help. These may include nurses, psychologists, social workers, and specialists such as nephrologists (doctors who treat kidney disease), haematologists (doctors specialising in blood disorders), dermatologists (doctors who treat skin disease), and neurologists (doctors specialising in disorders of the nervous system). If the disease is limited to a specific body organ (ie kidney) then that specialist may be the main doctor involved in treating the person.

The range and effectiveness of treatments for lupus have increased dramatically, giving doctors more choices in how to treat the disease. It is important for the patient to work closely with the doctor and take an active role in treatment. Once lupus has been diagnosed, the doctor will develop a treatment plan based on the patient's age, sex, health, symptoms, and lifestyle. Treatment plans are tailored to the individual's needs and may change over time. In developing a treatment plan, the doctor has several goals: to prevent flares, to treat them when they do occur, and to minimise organ damage and complications. The doctor and patient should re-evaluate the plan regularly to ensure that it is as effective as possible.

Several types of drugs are used to treat lupus. The treatment the doctor chooses is based on the patient's individual symptoms and needs. For people with joint or chest pain or, drugs that decrease inflammation, referred to as nonsteroidal anti-inflammatory drugs (NSAIDs), are often used. While some NSAIDs are available over the counter, a doctor's prescription is necessary for others. NSAIDs may be used alone or in combination with other types of drugs to control pain, swelling, and fever. Even though some NSAIDs may be purchased without a prescription, it is important that they be taken under a doctor's direction. Common side effects of NSAIDs, including those available over the counter, can include stomach upset, heartburn, diarrhoea, and fluid retention. Some patients with lupus also develop liver and kidney inflammation while taking NSAIDs, making it especially important to stay in close contact with the doctor while taking these medications.

Antimalarials are another type of drug commonly used to treat lupus. These drugs were originally used to treat malaria, but doctors have found that they also are useful for lupus. Exactly how antimalarials work in lupus is unclear, but scientists think that they may work by suppressing parts of the immune system. A common antimalarial used to treat lupus is hydroxychloroquine (Plaquenil). It may be used alone or in combination with other drugs and generally is used to treat fatigue, joint pain, skin rashes, and inflammation of the lungs.

Clinical studies have found that continuous treatment with antimalarials may prevent flares from recurring. Side effects of antimalarials can include stomach upset and, extremely rarely, damage to the retina of the eye. Regular eye checks must be carried out.

The mainstay of lupus treatment involves the use of corticosteroid hormones, such as prednisone. Corticosteroids are related to cortisol, which is a natural anti-inflammatory hormone. They work by rapidly suppressing inflammation. Corticosteroids can be given by mouth, in creams applied to the skin, or by injection. Because they are potent drugs, the doctor will seek the lowest dose with the greatest benefit. Short-term side effects of corticosteroids include swelling, increased appetite, weight gain, and emotional ups and downs. These side effects generally stop when the drug is stopped. It can be dangerous to stop taking corticosteroids suddenly, so it is very important that the doctor and patient work together in changing the corticosteroid dose. Sometimes doctors give very large amounts of corticosteroid by vein over a brief period of time (days) ("bolus" or "pulse" therapy). With this treatment, the typical side effects are less likely and slow withdrawal is unnecessary.

Long-term side effects of corticosteroids can include stretch marks on the skin, excessive hair growth, weakened or damaged bones (osteoporosis), high blood pressure, damage to the arteries, high blood sugar, infections, and cataracts. Typically, the higher the dose of prolonged corticosteroids, the more severe the side effects. Also, the longer they are taken, the greater the risk of side effects. Researchers are working to develop alternative strategies to limit or offset the use of corticosteroids. For example, corticosteroids may be used in combination with other, less potent drugs, or the doctor may try to slowly decrease the dose once the disease is under control. People with lupus who are using corticosteroids should talk to their doctors about taking supplemental calcium and vitamin D or other drugs to reduce the risk of osteoporosis (weakened, fragile bones). Sometimes stronger drugs such as methotrexate (Folex) may be used to help control the disease.

For patients whose kidneys or central nervous systems are affected by lupus, a type of drug called an immunosuppressive may be used. Immunosuppressives, such as azathioprine (Imuran) and Cyclophosphamide (Cytoxan), restrain the overactive immune system by blocking the production of some immune cells and curbing the action of others. These drugs may be given by mouth or by infusion (dripping the drug into the vein through a small tube). Side effects may include nausea, vomiting, hair loss, bladder problems, decreased fertility, and increased risk of cancer and infection. The risk for side effects increases with the length of treatment. As with other treatments for lupus, there is a risk of relapse after the immunosuppressives have been stopped.

Working closely with the doctor helps ensure that treatments for lupus are as successful as possible. Because some treatments may cause harmful side effects, it is important to report any new symptoms to the doctor promptly. It is also important not to stop or change treatments without talking to the doctor first.

Because of the nature and cost of the medications used to treat lupus, their potentially serious side effects, and the lack of a cure, many patients seek other ways of treating the disease. Some alternative approaches that have been suggested include special diets, nutritional supplements, fish oils, ointments and creams, chiropractic treatment, and homoeopathy. Although these methods may not be harmful in and of themselves, and they may be associated with symptomatic or psychosocial benefit, no research to date shows that they affect the disease process or prevent organ damage. Some alternative or complementary approaches may help the patient cope or reduce some of the stress associated with living with a chronic illness. If the doctor feels the approach has value and will not be harmful, it can be incorporated into the patient's treatment plan. However, it is important not to neglect regular health care or

treatment of serious symptoms. An open dialogue between the patient and the physician about the relative values of complementary and alternative and more traditional therapy is essential in permitting the patient to make an informed choice about treatment options.

Lupus and Quality of Life

Despite the symptoms of lupus and the potential side effects of treatment, people with lupus can maintain a high quality of life overall. One key to managing lupus is to understand the disease and its impact. Learning to recognise the warning signs of a flare can help the patient take steps to ward it off or reduce its intensity. Many people with lupus experience increased fatigue, pain, a rash, fever, abdominal discomfort, headache, or dizziness just before a flare. Developing strategies to prevent flares can also be helpful, such as learning to recognise your warning signals and maintaining good communication with your doctor.

It is also important for people with lupus to receive regular health care, instead of seeking help only when symptoms worsen. Having a medical exam and laboratory work on a regular basis allows the doctor to note any changes and may help predict flares. The treatment plan, which is tailored to the individual's specific needs and circumstances, can be adjusted accordingly. If new symptoms are identified early, treatments may be more effective. Other concerns also can be addressed at regular check-ups. The doctor can provide guidance about such issues as the use of sunscreens, stress reduction, and the importance of structured exercise and rest, as well as birth control and family planning. Because people with lupus can be more susceptible to infections, the doctor may recommend yearly influenza vaccinations for some patients.

<p style="text-align: center;">Warning Signs of a Flare</p> <ul style="list-style-type: none">• Increased fatigue• Pain• Rash• Fever• Abdominal discomfort• Headache• Dizziness
<p style="text-align: center;">Preventing a Flare</p> <ul style="list-style-type: none">• Learn to recognise your warning signals• Maintain good communication with your doctor

People with lupus should receive regular preventive health care, such as gynaecological and breast examinations. Regular dental care will help avoid potentially dangerous infections. If a person is taking corticosteroids or antimalarial medications, a yearly eye exam should be done to screen for and treat eye problems.

Staying healthy requires extra effort and care for people with lupus, so it becomes especially important to develop strategies for maintaining wellness. Wellness involves close attention to the body, mind and spirit. One of the primary goals of wellness for people with lupus is coping with the stress of having a chronic disorder. Effective stress management varies from person to person. Some approaches that may help include exercise, relaxation techniques such as meditation, and setting priorities for spending time and energy.

Developing and maintaining a good support system is also important. A support system may include family, friends, medical professionals, community organisations, and organised support groups. Participating in a support group can provide emotional help, boost self-esteem and morale, and help develop or improve coping skills.

Learning more about lupus may also help. Studies have shown that patients who are well informed and participate actively in their own care experience less pain, make fewer visits to the doctor, build self-confidence, and remain more active.

Tips for Working with Your Doctor

- Seek a health care provider who will listen to and address your concerns.
- Provide complete, accurate medical information.
- Make a list of your questions and concerns in advance.
- Be honest and share your point of view with the health care provider.
- Ask for clarification or further explanation if you need it.
- Talk to other members of the health care team, such as nurses, therapists, or pharmacists.
- Do not hesitate to discuss sensitive subjects (for example, birth control, intimacy) with your doctor.
- Discuss any treatment changes with your doctor before making them.

Pregnancy for Women with Lupus

Although a lupus pregnancy is considered high risk, most women with lupus carry their babies safely to the end of their pregnancy. Experts disagree on the exact numbers, but 20 to 25 percent of lupus pregnancies end in miscarriage, compared to 10 to 15 percent of pregnancies in women without the disease. Pregnancy counselling and planning before pregnancy are important. Ideally, a woman should have no signs or symptoms of lupus and be taking no medications for at least 6 months before she becomes pregnant.

Some women may experience a mild to moderate flare during or after their pregnancy; others do not. Pregnant women with lupus, especially those taking corticosteroids, also are more likely to develop high blood pressure, diabetes, hyperglycaemia (high blood sugar), and kidney complications, so regular care and good nutrition during pregnancy are essential. It is also advisable to have access to a neonatal (newborn) intensive care unit at the time of delivery in case the baby requires special medical attention. About 25 percent (one in four) of babies of women with lupus are born prematurely, but do not suffer from birth defects.

*Taken from NIAMS - National Institute of Arthritis and Musculoskeletal and Skin Diseases
Publication Date: September 1997 Revised: October 2002*

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ARRC provides education, support and research services for people living with a range of systemic and organ-specific autoimmune diseases.

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ARRC information for patients, carers & Health Professionals

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Content last updated August 2016

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