

SYSTEMIC LUPUS ERYTHEMATOSUS

SLE



NEW ZEALAND
PAEDIATRIC RHEUMATOLOGY SERVICE



What is Systemic Lupus Erythematosus(SLE)?

SLE is a disease that causes inflammation in many different parts of the body. Inflammation is a process that causes parts of the body to become painful, swollen, lose function and sometimes become hot or red. It can affect the skin, causing a rash, or the joints causing arthritis. It can also cause inflammation to organs you cannot see, such as kidneys, heart, lungs and the nervous system; you may not even get symptoms. If the inflammation is not treated properly, it can

sometimes cause permanent damage.

The most common body parts involved include the:

Skin, kidneys, blood, joints, heart, brain and lungs.

Lupus is a chronic disease. Chronic means that the disease can go on for many years, perhaps life long. This does not mean that you will be on medications or feel ill forever. There will be times when the disease is active (flare) and times when it is quiet (remission). Sometimes it goes away completely.

What causes SLE?

The cause of lupus is unknown. We do know the symptoms of lupus are caused by an abnormal reaction of the immune system. The immune system protects our bodies against infections caused by foreign invaders such as germs and viruses.

In someone with lupus, instead of the immune system only attacking the invader, it attacks the body's own cells. The end result is different areas of the body becoming inflamed.

Why Me?



Nothing you did, or did not do, caused you to get lupus. SLE is not contagious which means you didn't catch it from someone nor can they catch it from you. We do know that Lupus is more common in girls, usually over the age of six.

In some families there may be more than one person with lupus or other people in the family may have arthritis or other autoimmune diseases.



Did you know that "lupus" comes from the latin "wolf" and erythematous means "red"?

How is SLE diagnosed?

SLE is not always easy to recognize. The disease can begin with fever, tiredness and lack of energy, poor appetite and general aches and pains. To make the diagnosis, the doctor will:

- take a full history of all of the symptoms
- do a physical examination
- collect blood and urine samples

The doctor may order other tests to check specific area of the body if she/he suspects a problem.

What are the signs and symptoms of SLE?

The signs and symptoms of lupus vary from person to person. This can be very mild or much more severe. Specific signs and symptoms can be related to different parts of the body:

Skin

- Malar rash or 'butterfly' shaped rash over the cheeks and across the bridge of the nose.
- Rash that comes out when there is exposure to the ultraviolet light from the sun (photosensitivity)

Joints

- Pain, stiffness, warmth and swelling in the joints (arthritis)

Kidneys

- Inflammation can cause damage to the kidneys and (if untreated or very severe) may result in kidney failure
- Damage to the kidneys can cause high blood pressure or when too much protein is lost, swelling of the face & legs

Heart and Lungs

- Inflammation of the lining around the heart and lungs may cause pain or fluid to accumulate in those areas

Central Nervous System

- Inflammation in the brain can cause headaches, memory loss, mood swings and sometimes more serious problems such as seizures or thinking problems (psychosis)

Other symptoms of Lupus

- Extreme fatigue
- Sore muscles
- Hair loss
- Colour changes and/or sores on the fingers and toes, especially in cold weather (Raynaud's phenomenon)
- Weight loss
- Blood clots
- Mouth sores on the tongue and the inside of the mouth (that don't usually hurt)

What should I expect when I come to clinic?

There are a number of people that make up the Rheumatology team. Your child may be seen by the Rheumatologist (specialist), Rheumatology fellow (a doctor training in Rheumatology), the paediatric registrar, Physiotherapist, Occupational Therapist and the Clinical Nurse Specialist.

There are times when you will have many appointments. We will try to make sure you miss as little of school as possible. The clinic visits are very important as it lets the team monitor your disease activity and make adjustments to your medications. It is also a good time for you and your parents to ask questions.



What investigations are done in lupus?

Blood tests

Blood tests help to diagnose SLE and to monitor the activity of the disease. These are also done to check for side effects of your medication. Usually a blood and urine test will be done at every clinic visit. Although there is no one test that can definitely say whether a person has lupus or not, there are lab tests which are indicators of lupus. These include:

- Anti-nuclear antibody test (ANA)
- Anti-double stranded DNA antibodies (anti-DNA)
- Serum complement
- Erythrocyte Sedimentation Rate (ESR)
- Antiphospholipid antibodies
- Full Blood Count (FBC)

Urine Test

This is an important test to monitor the kidney. If the kidneys are inflamed, red blood cells, and/or protein may be found in the urine.

Kidney Biopsy

A biopsy is where a small amount of tissue is taken from the kidney with a long needle and examined under a microscope to determine the extent of inflammation. This test is usually done if other kidney tests are abnormal.

Pulmonary Function Test

This is a breathing test that looks at the functioning of the lungs. It is a routine test that is usually done regularly to help look for inflammation in the lungs.

Echocardiogram

This is an ultrasound that looks at the functioning of the heart.

Bone Density Scan

This is a special x-ray that looks at how strong your bones are. One of the medicines (prednisone) can cause bones to be more brittle.

Ultrasound, MRI and CT scans are also commonly used and will be explained in more detail if these tests are recommended.

Other important things to think about.

Sun Protection

People with SLE need to pay particular attention as the rash can become more active with sun exposure and trigger a flare in your lupus.

Slip, Slop, Slap!!

Exercise

Exercise will help strengthen your muscles and bones, keep joints flexible and controls your weight. The physiotherapist and occupational therapist may recommend specific exercises.

Rest

Fatigue is often a symptom of lupus when the disease is active. This will improve but it is important to pace yourself and not over do it.

Contraception/Pregnancy

If you fall pregnant while your lupus is not stable you can have a severe flare.

Contraception and barrier methods are therefore recommended in sexually active patients. Please discuss this with your doctor who can give you more information.

Smoking/Alcohol/Drugs

Smoking and other drugs/alcohol should be avoided as they can make your lupus worse and can interact with your medications.

Diet

There is no special diet which will cure your child's illness. A well balanced diet that includes a variety of foods is the key to healthy nutrition. Your rheumatology team may make recommendations about Calcium and Vitamin D if you are on prednisone.

What about infections?

Chicken Pox

Children who develop chicken pox when taking prednisone or other immunosuppressants can become very ill. If your child is in direct contact with a child with chicken pox they may need to have Varicella Immune Globulin (VZIG) within 72 hours of contact. If your child develops chicken pox they may need antiviral medication to lessen the severity of the illness.

Measles

It is preferable that children are fully immunized. However, if they are not (or they are taking prednisone and immunosuppressants), they may need to have Measles Immunoglobulin within 6 days of exposure to measles.

Influenza

It is recommended that you have a flu injection each year.

What medications will make the lupus better?

The treatment is aimed at decreasing inflammation in the joints, skin, kidneys, or other organs and avoiding a lupus flare. The treatment regime will change from time to time depending on how the lupus is affecting the body. Sometimes when the disease is quiet, there may not be a need for any medications.

Corticosteroids (Prednisone and Methylprednisolone)

These drugs are used for severe disease. They act by suppressing the immune system and decreasing inflammation. If they need to work fast they can do so. Unfortunately they do have side effects, which is why your doctors try to control the inflammation using other medication in the long term. The side effects are related to the dose and duration you are on the medication.

You must never stop your steroids suddenly or change the dose without discussing it with the Rheumatology team.

Hydroxychloroquine (Plaquenil)

This is a drug that most young people with lupus will be on long term (for many years). It is helpful for treating the rash in lupus and also helpful in improving some abnormalities in the blood. It is often the only medication needed. It has relatively few side effects, but does require you to have your eyes checked by an eye doctor from time to time.

Non-steroidal anti-inflammatory drugs (NSAIDS)

These help control inflammation

Immunosuppressive Medicines.

There are several different immunosuppressive drugs which may be prescribed to help control lupus. These help settle down the immune system. These include, azathioprine, mycophenolate mofetil (MMF), methotrexate, cyclophosphamide and cyclosporin. These will be discussed in detail if recommended by the rheumatology doctors.

Living with Lupus

The most important aspect of managing your lupus is remembering to take your medicines and attending the clinics for regular check ups and blood tests. You may feel frustrated at times and this is normal. Remember to discuss your frustrations and worries with your family and your rheumatology team.

Remember that you are not alone. There are a number of useful websites to help you learn more about lupus as well as support you in dealing with a chronic illness. Please see below.



Useful Websites:
www.uklupus.co.uk
www.mylupus.org
www.arthritisresearchuk.org
www.lupus.org.nz
www.lupussupport.org.nz

