

Juvenile SLE

Juvenile SLE (systemic lupus erythematosus) also known as lupus, is an autoimmune disorder affecting children and young people under the age of 18. Any part of the body can be affected by lupus, but commonly joints are involved making SLE a form of autoimmune arthritis.

Signs and symptoms

Often early warning signs of lupus are very similar to other diseases and so can be easily missed, or misdiagnosed. Even once diagnosed, symptoms can come and go – new symptoms may appear while others resolve. Symptoms often vary from person to person as well.

Common Symptoms

Fatigue

Loss of appetite/weight loss

Swollen/aching joints

Muscle aches

Fevers (over 100 degrees F)

Skin rashes – in particular a rash over the cheeks in a “butterfly” pattern and /or rashes that develop with sun-exposure

Hair loss and/or brittle hair

Mouth and/or nose ulcers

Color changes (blue and/or white) in fingers when exposed to cold or stress (Raynaud’s Phenomenon)

Children with SLE are more likely than adults to have problems with organs such as their kidneys and brain. Signs are symptoms of this may include:

Dark urine

Swelling in feet, legs and eyelids

Shortness of breath and/or chest pain

Headaches, memory problems, seizures

Causes

Like most other autoimmune disorders, the exact causes of Juvenile SLE are unknown. It is thought to be a combination of genetic and environmental factors. A parent with Lupus has only a 5% chance of passing on lupus to their child, and often if one identical twin has lupus the other does not, therefore it is thought that something other than genetics triggers SLE. As there is such as extreme female predominance, it is thought hormones play a role too.

It is not contagious.

Diagnosis

Diagnosing Juvenile SLE can be a complicated process. There is no single test or symptom used to make a diagnosis, but a number of factors have to be taken into account.

A number of blood tests may be used:

Complete blood count (CBC)

Antibody (ANA)

Anti-DNA
Complement (C3 and C4)
C-reactive protein (CRP)
Erythrocyte sedimentation rate (ESR or sed rate)

These tests can also be used later, after diagnosis is made, to monitor the progression of the disease.

Other lab tests, X-rays or biopsies can be used to test for inflammation, particularly around the vital organs.

The 11 criteria for lupus

Lab results may not give a clear picture. Generally, **at least 4** of the following list of symptoms must be present to make a diagnosis.

Malar rash (butterfly-shaped rash over the cheeks and nose)
Discoid Rash (raised scaly patches of skin)
Photosensitivity (skin rash from sun exposure)
Oral ulcers (small sores in the mouth)
Arthritis (swelling and pain in at least two joints)
Cardiopulmonary problems (inflammation around the heart and/or lungs)
Neurological Problems (seizures and/or psychosis)
Kidney problems (for example blood in the urine)
Hematologic (blood) problems (low levels of red or white blood cells or platelets)
Positive ANA blood test
Other positive blood tests

Average Age of Onset

The most common age of onset for Juvenile SLE is 15-18 years. In this age group the disease behaves more or less like adult onset lupus. It can range in severity from mild symptoms, to a life-threatening disease.

It is less common for onset to be before the age of 15, however symptoms can be more severe for this age group and there is a higher chance of organ involvement.

It is rare for under 5 years old to develop SLE. Under 5 year olds who do develop lupus are often the sickest of lupus patients, because their immune systems are underdeveloped anyway, because of their age.

There is a female predominance in SLE, with women and girls making up 90% of all cases.

Treatment

Medications for the treatment of SLE generally fall into two categories: Nonimmunosuppressants and Immunosuppressants. Nonimmunosuppressants are milder drugs that fight inflammation and aim to ease discomfort. Immunosuppressants are more powerful drugs which aim to bring the malfunctioning immune system under control. Some of these have significant side effects, as they suppress the immune system increasing the risk of infection.

Nonimmunosuppressants

Antimalarial drugs (plaquenil etc.) These help to reduce flare ups of lupus.

Nonsteroidal anti-inflammatory drugs (NSAIDs) These help to ease swelling and stiffness, particularly in children with lupus arthritis.

Immunosuppressants

Corticosteroids (most often prednisone)

Disease-modifying anti-rheumatic drugs (DMARDs) Methotrexate etc.

Biologics

Cytotoxins

Physical and/or occupational therapies may be needed to help increase mobility and muscle strength. This can also help children learn to do daily activities in ways that are easier on their bodies.

Because chronic illnesses can be mentally and emotionally difficult to deal with, children with SLE may find psychotherapy or counseling a useful addition to their treatment.

Lifestyle changes at home: a healthy diet, staying active, getting plenty of rest and being careful with sun-exposure, can also help children with SLE.