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# Systemic Lupus Erythematosus

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## 1. WHAT IS SYSTEMIC LUPUS ERYTHEMATOSUS

### 1.1 What is it?

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that can affect various organs of the body, especially the skin, joints, blood, kidneys and central nervous system. "Chronic" means that it can last for a long time. "Autoimmune" means that there is a disorder of the immune system, which, instead of protecting the body from bacteria and viruses, attacks the patient's own tissues.

The name "systemic lupus erythematosus" dates back to the early 20th century. "Systemic" means that it affects many organs of the body. The word "lupus" is derived from the Latin word for "wolf" and it refers to the characteristic butterfly-like rash on the face, which is similar to the white markings on a wolf's face. "Erythematosus" in Greek means "red" and it refers to the redness of the skin rash.

### 1.2 How common is it?

SLE is recognized worldwide. The disease appears to be more common in people of African American, Hispanic, Asian and Native American origin. In Europe about 1:2,500 persons are diagnosed with SLE and about 15% of all lupus patients are diagnosed prior to the age of 18 years. Onset of SLE is rare before the age of 5 and uncommon before adolescence. When SLE appears before the age of 18, physicians use different names: paediatric SLE, juvenile SLE and childhood-onset SLE. Females of child-bearing years (15 to 45) are most often affected and, in that particular age group, the ratio of affected females to males is 9 to 1. Before puberty, the proportion of affected males is higher and

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about 1 of every 5 children with SLE is male.

### **1.3 What are the causes of the disease?**

SLE is not contagious; it is an autoimmune disease, where the immune system loses its ability to distinguish between a foreign substance and an individual's own tissues and cells. The immune system makes a mistake and produces, amongst other substances, autoantibodies that identify the person's own normal cells as foreign and attack them. The result is an autoimmune reaction, which causes inflammation of specific organs (joints, kidneys, skin, etc). Inflamed means that affected body parts become hot, red, swollen and sometimes tender. If the signs of inflammation are long-lasting, as they may be in SLE, then damage to the tissues may occur and normal function is impaired. This is why treatment of SLE is aimed at reducing the inflammation.

Multiple inherited risk factors combined with random environmental factors are considered responsible for this abnormal immune response. It is known that SLE can be triggered by various factors, including hormonal imbalance in puberty, stress, and environmental factors such as sun exposure, viral infections and medications (e.g. isoniazid, hydralazine, procainamide, anti-seizure medications).

### **1.4 Is it inherited?**

SLE can run in families. Children inherit some as yet unknown genetic factors from their parents that may predispose them to develop SLE. Even if they are not necessarily pre-destined to develop SLE, they may be more likely to get the disease. For example, an identical twin has no more than a 50% risk of getting SLE if the other twin is diagnosed with SLE. There is no genetic testing or a prenatal diagnosis available for SLE.

### **1.5 Can it be prevented?**

SLE cannot be prevented; however, the affected child should avoid contact with certain situations that may trigger the onset of the disease or cause the disease to flare (e.g. sun exposure without using sunscreens, some viral infections, stress, hormones and certain medications).

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## **1.6 Is it infectious?**

SLE is not infectious. This means that it cannot be passed from person to person.

## **1.7 What are the main symptoms?**

The disease may begin slowly with new symptoms appearing over a period of several weeks, months or even years. Non-specific complaints of fatigue and malaise are the most common initial symptoms of SLE in children. Many children with SLE have intermittent or sustained fever, loss of weight and appetite.

With time, many children develop specific symptoms that are caused by involvement of one or several organs of the body. The skin and mucosal involvement are very common and may include a variety of different skin rashes, photosensitivity (where exposure to sunlight triggers a rash) or ulcers inside the nose or mouth. The typical 'butterfly' rash across the nose and cheeks occurs in one-third to one half of affected children. Increased hair loss (alopecia) may sometimes be noticed. The hands turn red, white and blue when exposed to the cold (Raynaud's phenomenon). The symptoms can also include swollen and stiff joints, muscle pain, anaemia, easy bruising, headaches, seizures and chest pain. Kidney involvement is present to some degree in most children with SLE and it is a major determinant of the long-term outcome of this disease.

The most common symptoms of major kidney involvement are high blood pressure, protein and blood in urine and swelling, particularly in the feet, legs and eyelids.

## **1.8 Is the disease the same in every child?**

Symptoms of SLE vary widely between individual cases so that each child's profile or list of symptoms is different. All of the symptoms described above can occur either at the beginning of SLE or at any time during the course of the disease with different severity. Taking medications that were prescribed by your lupus doctor will help control SLE symptoms.

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## **1.9 Is the disease in children different from the disease in adults?**

SLE in children and adolescents has manifestations as SLE in adults. However, in children, SLE has a more severe course in that children more often show several features of inflammation due to SLE at any given time. Children also have kidney and brain disease with SLE more often than adults.