



SYSTEMIC LUPUS ERYTHEMATOSUS

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Systemic lupus erythematosus (SLE) is a disease that primarily affects the skin, but also affects many other organs and systems. SLE dates back to the 1800s, and since then has been progressively more and more described. The disease tends to be more common in women than men with a male to female ratio of 1:9. People of African descent have a predilection for the disease, especially compared to Caucasians, with a prevalence of 200 per 100,000 compared to 40 per 100,000, respectively. Patients typically start to develop symptoms of the disease in their 30s or 40s.

The cause of SLE is still not clear, but it is thought that it is an auto-immune disease characterized by a complex interplay between genetics and environment. Associations have been made with a family history, ultraviolet radiation (sunlight), and even certain medications used for various diseases. Skin manifestations can be present for weeks to months before diagnosis is made. The symptoms of SLE traverse a wide range of possibilities, but patients commonly report itching and burning of skin rashes, abdominal pain, tiredness, muscle pain, joint pain, fever, and weight changes.

The skin changes involved in SLE are varied with most being nonspecific for SLE. The “butterfly rash” is commonly described in SLE, and is described as a red, raised (macular) rash in the shape of a butterfly on the face with the “wings” spanning the cheeks and the “thorax and abdomen” on the nose. The butterfly rash can also have some crusty spots with scaly skin, and other spots consisting of eroded away skin. Patients can also have other kinds of rashes in other parts of the body. For instance, red raised bumps or blisters can occur on the face, hands, arms, scalp, or upper chest and back. Mostly, rashes tend to appear in areas that are exposed to the sun. Spots on the head may have hair loss, and people may also have ulcers in the mouth. Patients may also have a slew of other skin findings that are not specific to SLE, and in those cases a high sense of suspicion is warranted to facilitate an accurate diagnosis.

As the name of the disease eludes, other sites throughout the whole body besides the skin are involved in the disease as well. Regularly, people report issues with their muscles and joints primarily described as pain. Kidneys may decline in function, and inflammation of the lungs, gastrointestinal tract, and protective sac of the heart may occur. The spleen and liver may become enlarged, and central nervous system issues such as seizures and neuropathy may also occur. All of these problems may cause the affected organ or other organs to stop functioning completely and possibly can lead to death or severe impairment. For example, extensive involvement of the eye can occur and lead to blindness if not properly managed. While this list is not exhaustive and virtually any organ system can be involved, the prognosis of the disease is generally good with 93% of people still living 5 years after being diagnosed.

Diagnosis of SLE is essentially a diagnosis of exclusion; the diagnosis of SLE occurs after other diseases have been ruled out. The diagnosis is supported with clinical suspicion, blood tests, biopsy, and specific criteria devised by the American College of Rheumatology in 1982. **Biopsy** specimens can be examined under a microscope and processed in various ways to aid in the diagnosis. There are 11 described criteria by the American College of Rheumatology, and diagnosis can be made with 4 or more of the criteria being met at any time during evaluation. The criteria highlight the systemic and multi-organ nature of the disease and are as follows: malar rash, discoid rash, photosensitivity, oral ulcers, arthritis, serositis, renal disorder, neurologic disorder, hematologic disorder, immunologic disorder, and antinuclear antibody.

Treatment of SLE depends on the specific patient and the extent of the disease. Primarily, treatment is aimed at alleviating the symptoms of the disease, decreasing the amount of complications, and prolonging the survival of the patient. Treatment may be orchestrated by multiple physicians in many different specialties. Avoidance of exacerbating factors like certain drugs and sunlight protection is paramount. Commonly, **prednisone** and other immunosuppressive drugs like **azathioprine**,

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methotrexate, and **mycophenolate mofetil**. Topical, intra-lesional, and systemic therapy are frequently used alone or in combination. **Antimalarial** medications have been a mainstay of treatment for decades. Many other medications and treatments are used in the management of SLE ranging from exercise and smoking cessation to **retinoids** and **cyclosporine**. Research to continue for other therapies and biologic treatment has already transpired.

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