

Lupus Remains Incurable, but Research Offers Hope

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Course Objectives

The goal of this program is to provide nurses with information about the manifestations of systemic lupus erythematosus and management of the person with the disease. After studying the information presented here, you will be able to —

- Describe the three types of lupus.
 - Identify the symptoms of systemic lupus.
 - Discuss the importance of patient communication as an integral part of nursing intervention.
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You've just been assigned a patient with systemic lupus erythematosus (SLE), and if you're straining your memory to recall exactly what lupus is, you're in good company. Although lupus is more prevalent in this country than AIDS, sickle cell anemia, cerebral palsy, multiple sclerosis, and cystic fibrosis,¹ some nursing schools don't cover it extensively. Lupus can affect any part of the body, is difficult to manage, and can be life-threatening. But if you have a clear understanding of the manifestations of the disease, and its often unpredictable course, caring for the person with lupus can be a challenging but rewarding experience.²

Lupus is an incurable disease that is not easy to diagnose. It is difficult to manage and can be life-threatening. Nurses knowledgeable about the manifestations of lupus, patient education resources, and current treatment options can be crucial in helping patients handle the often unpredictable course of their disease.²

Lupus is a chronic, inflammatory disease. It develops when the body's immune system, which normally protects against foreign antigens such as viruses and bacteria, becomes confused and mistakenly begins producing antibodies against a person's own cells and tissues. These [autoantibodies](#) react with the antigens to form immune complexes that can build up in various parts of the body, causing inflammation, pain, and injury to tissues.

Although researchers are making progress in understanding the causes of this complex disease, the precise etiology is not known. Many experts believe that genetics, environment, and hormones act together to trigger the disease.

Strong evidence indicates a hereditary component to lupus, with genes playing a major role in the disease process. As many as 100 genes may contribute to the genetic predisposition and development of lupus.³ The familial association of the disease and the fact that certain racial groups are more likely to develop lupus are two indicators of genetic predisposition. Studies of families show that a person with lupus is 10 to 12 times more likely to have a close relative with lupus than is a person without the disease.³ Research has also shown that African-American women in the U.S. have a three to four times higher incidence of the disease than Caucasian women, and among American Indians, certain tribes — the Sioux, Crow, and Arapaho — have a higher incidence of lupus than others.³

Although heredity appears to be the precursor for lupus, something else triggers the disease. Because identical twins share genetic makeup, studying them demonstrates a relationship between heredity and environment. If one twin has lupus, a 26% to 70% chance exists that the other will develop the disease.⁴ However, the fact that lupus does not consistently occur in the other suggests other factors in its development.

Environmental factors thought to trigger lupus include UV light and certain medications. Other factors may include stress, certain foods, and toxic chemicals. However, no research definitively links these factors with the onset of the disease.⁵ Infections, extreme stress, and surgery may trigger the symptoms of lupus in someone who already has the disease.

Lupus can occur in either sex and at any age. But women in their childbearing years are 10 to 15 times more likely to develop lupus than men. For that reason, hormones are thought to play a role in triggering the disease.⁶ But the effect of hormones in lupus is not clear. Abnormal metabolism of the female sex hormones, particularly estrogen, may influence the immune system, making women more susceptible to lupus. The development or increase in lupus activity with menstruation and pregnancy supports this concept.⁷

What type of lupus?

Lupus occurs in three main types: [cutaneous lupus erythematosus](#), [drug-induced lupus erythematosus](#), and systemic lupus erythematosus.

Cutaneous lupus affects the skin and may also involve the hair and mucous membranes. Cutaneous lupus itself has variants that manifest in different-looking rashes and symptoms. Cutaneous lupus does not affect the internal organs. However, in about 10% of patients it can evolve into the systemic form of the disease.⁸

Certain medications can create a syndrome similar to systemic lupus. The symptoms of drug-induced lupus occur after the use of certain prescribed drugs but disappear when the medication is discontinued. The medications most commonly connected with drug-induced lupus are hydralazine (Apresoline), procainamide (Pronestyl), and quinidine.⁹ Drug-induced lupus is more common in men over the age of 50 because they take these drugs more often. About 5% to 20% of the people who take procainamide or hydralazine will develop antibodies suggestive of lupus. With most of the other drugs, the risk is less than 1% that those taking the medications will develop drug-induced lupus.⁹ The symptoms of drug-induced lupus usually disappear within six months of discontinuing the medication.

Systemic lupus is usually more serious than cutaneous lupus and can affect any organ or organ system in the body, including the kidneys, heart, lungs, brain, blood, skin, and nervous system. The disease can follow an irregular pattern of remissions and flares.

Symptoms of systemic lupus range from minor to life-threatening. A person may experience very little pain — or extreme pain, especially in the joints. There may be no skin involvement — or disfiguring rashes. The person with systemic lupus may have no organ involvement — or extreme organ damage. Generally, no two people will have the same symptoms. The following are the most common symptoms and the percent of people with systemic lupus who have them: achy joints (95%), fever (more than 100 F) (90%), swollen joints (90%), prolonged or extreme fatigue (81%), skin rashes (74%), anemia (71%), kidney involvement (50%), pain in the chest on deep breathing (pleurisy) (45%), [butterfly-shaped rash](#) across the cheeks and nose (42%), light sensitivity (30%), hair loss (27%), problems with blood clotting (20%), fingers turning white or blue in the cold (Raynaud's phenomenon) (17%), seizures (15%), and mouth or nose ulcers (12%).⁶

Diagnosis is tricky

Because lupus symptoms can come and go, are usually vague, and often mimic those of other illnesses, it can be a difficult disease to diagnose. Diagnosis may take years. No one single test exists that can tell if a person has lupus. Diagnosis is usually made by a careful review of the entire medical history (including family history), a complete physical exam, and laboratory tests. A biopsy of the skin or kidney may also help with the diagnosis. Some symptoms the patient describes may appear to be totally unrelated; therefore, good communication is necessary when assessing and interviewing the patient.

The American College of Rheumatology developed ¹¹ criteria to help distinguish between lupus and other diseases. A person should meet four or more of the criteria to suspect lupus. (See sidebar.)

Medical management of a person with lupus can be a challenge. The goals are to manage symptoms, avoid complications of medical therapy, and prevent flares. Lupus treatment requires a team approach, and the patient should work closely with all health care providers to take an active role.

Depending on the type and severity of the symptoms — and the patient's medical history — the health care provider will try to treat the disease with the most conservative medications and minimum amount of medication possible. Effective pharmacological treatment can minimize symptoms, reduce inflammation, and maintain normal body functions. The medications prescribed usually depend on which organs are involved and the severity of the involvement. The choice of drugs is highly individualized and typically changes during the course of the disease.

Nonsteroidal antiinflammatory drugs are often the first line of defense for lupus and are effective in controlling mild inflammation and symptoms such as muscle or joint pain. NSAIDs are not effective for severe lupus or when lupus affects major organs. The adverse effects of long-term, high-dose therapy include stomach upset, ulcers, easy bruising, high blood pressure, and fluid retention.

Antimalarial medications suppress parts of the immune response and reduce inflammation, and are helpful for fatigue and moderate skin and joint problems.⁷ Patients need to understand that weeks or even months may pass before the drugs' beneficial effects are evident. Antimalarials are usually well-tolerated. High-dose therapy may damage the retina, causing vision problems. All patients taking antimalarials should see an ophthalmologist before starting treatment and every six to 12 months thereafter. Clinical studies show that continuous treatment with antimalarials may also prevent lupus flares.¹⁰

Corticosteroids have very potent antiinflammatory and immunoregulatory properties and are extremely effective for severe inflammation and organ involvement that are not responding to other treatments. A variety of adverse effects are possible when taking steroids, and the patient should be made aware that long-term adverse effects — such as avascular necrosis, osteoporosis, arteriosclerosis, weight gain, moon face, and increased appetite — may occur when steroids are taken at high doses over long periods of time. Steroids must not be discontinued abruptly, especially if they have been taken for more than four weeks or at high dosages. After that time, shrinking of the adrenal glands may occur and lead to a decreased natural production of cortisone. A slow tapering in the dosage allows the adrenal glands to regain their ability to manufacture natural cortisone.

More aggressive therapy, such as immunosuppressive or cytotoxic drugs, is used for the most serious manifestations of lupus, such as kidney, lung, central nervous system, or heart involvement, or for times when high-dose steroids are ineffective. Patients receiving these medications are especially vulnerable to severe adverse reactions, such as anemia, malignancy, infection, and decreased WBCs, and must be monitored during and after drug administration.

Research offers the best hope for finding the cause of lupus, making safe and more effective medications with fewer adverse effects, and eventually curing this debilitating disease. Current therapies for lupus focus on suppressing the entire immune system and reducing inflammation. The goal of future research will be to determine the specific cell mechanisms of inflammation and autoimmunity to develop

therapies that better target exactly what is going wrong without suppressing the entire immune system. Scientists are working to develop drugs that are directed at the disease itself, not just the symptoms, while taking into account the effects of lupus on organ systems.

To care for a person with lupus, nurses must have up-to-date knowledge about the disease, its many manifestations, and its often unpredictable course.² Because patients are most familiar with their symptoms, an important role for the nurse is listening to patients. Patients can provide valuable information about therapies that are helpful.

Providing validation and emotional support for a person with lupus is an integral part of patient care. Often a person with lupus has endured years of agonizing symptoms with either an incorrect diagnosis or no diagnosis at all. Because many lupus patients don't look sick, they are often discounted and cast aside. Often people with lupus feel frustrated, discouraged, and hopeless. Allowing patients to communicate their needs and adjusting nursing interventions to include individual routines recognizes the value of patients as authorities on their illness, creating a sense of dignity and control.

Nurses should assess and expand patients' knowledge of the disease, including through referral to community resources. A wealth of information is available in libraries, on the Internet, and from lupus support organizations. Encourage patients to learn as much as they can about their illness and remind them to discuss the effects of lupus on their personal lives.

Nurses should educate patients about their medications and not only include information about the expected response and adverse effects, but also stress the importance of taking medications as directed. Patients should never adjust a dose or stop taking any drug on their own. Education should also emphasize the importance of frequent follow-up and ongoing evaluation so that the provider can monitor symptoms and modify treatments to ensure that they remain as effective as possible.

Teach patients about nonpharmacological methods that may help prevent or reduce the risk of flares. People with lupus should avoid excessive exposure to UV light, apply sunscreen, and wear sun-protective clothing even on cold or cloudy days. Adequate rest and regular low-impact exercise may help prevent muscle weakness and fatigue, while immunizations protect against specific infections. Support groups, counseling, and talks with family members, friends, and health care providers may help alleviate the effects of stress. Negative habits such as smoking, consuming excessive amounts of alcohol, taking too much or too little of prescribed medication, or postponing regular medical checkups can be hazardous to people with lupus and can worsen the effects of the disease.¹⁰ Remind patients to see their providers on a routine basis instead of only if symptoms get worse or new symptoms appear.

Frequent assessments and accurate documentation will communicate patient needs to all members of the health care team. Documenting variations in the patient's symptoms will alert the team to changes in the patient's status that may be transient, but may hold significance.

Remember that each case of lupus is different, and the diverse needs of people with lupus make developing a "one-size-fits-all" plan of care impossible. A cooperative approach, a flexible plan of care, and careful listening skills will provide patients with support and the reassurance that their needs are being met.²

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