

Understanding and Treating Lupus



So little is known about this autoimmune disease, which affects more than 1.5 million Americans. But, improved testing and increased research are resulting in better treatment for lupus patients.

By Ronale Tucker Rhodes, MS

In 2006, Diane Sawyer aired a segment on “Good Morning America” to spread awareness about lupus.¹ In the segment, she interviewed actress and model Kelly Jean Drury, who eight years earlier, at age 23, was diagnosed with the disease. Kelly is one of the 1.5 million Americans who have lupus. And, while this disease is widespread, occurring in one in 2,000 people, very few are even aware of it, and those who are rarely understand what it is.

Chances are, most people know someone who has lupus. In fact, there is a 20 percent chance of having a parent or sibling with lupus. Ninety percent of those with lupus are women, and the disease is two to three times more prevalent among people of color. While diagnosis typically occurs between the ages of 15 and 45, the disease can strike at any age, and many go undiagnosed. Those who do get diagnosed typically wait, on average, four years and have to see three different doctors to get diagnosed.

What Is Lupus?

Lupus is a chronic autoimmune disease that can damage any part of the body — skin, joints and/or organs. Lupus occurs when the immune system cannot tell the difference between viruses, bacteria and germs (foreign invaders) and the body's healthy tissues, and it creates autoantibodies that attack and destroy the healthy tissues. Those autoantibodies cause inflammation, pain and damage in various parts of the body. The disease can range from mild to life-threatening. It is not contagious, and it is not like or related to cancer or HIV.²

There are five types of lupus. The most common, and the one most people are referring to when they say lupus, is systemic lupus erythematosus (SLE). This type affects many different organ systems, and is marked by chronic inflammation, especially of the kidneys, joints and skin. The cardiovascular and nervous systems also can be affected.

When lupus is limited to the skin, it is called chronic cutaneous lupus erythematosus (CCLE, also known as discoid lupus erythematosus [DLE]), subacute cutaneous lupus erythematosus (SCLE) or tumid lupus. While this type of lupus can exist in people who do not have systemic lupus, 5 percent or more of people with this type of lupus may develop SLE later in life.

Drug-induced lupus erythematosus is caused by certain drugs that cause lupus-like symptoms in people who do not have SLE. This form of lupus is temporary, and it usually subsides within months after the medication is stopped. Medications known to cause this type of lupus include blood pressure medications hydralazine and methyldopa; a heart medication called procainamide; a drug called D-penicillamine used to treat metal poisoning; minocycline, used to treat acne; and anti-TNF, used to treat rheumatoid arthritis. Many physicians are choosing not to prescribe these medications.

Neonatal lupus erythematosus may affect the babies of women who possess the autoantibodies anti-Ro, anti-La and anti-RNP. Only 40 percent of women bearing children with neonatal lupus actually have lupus, but lupus or Sjogren's syndrome may occur later in life. While neonatal lupus typically involves only the baby's skin and subsides on its own without treatment, 1 percent to 2 percent of these infants experience congenital heart block, which can be treated with the implantation of a pacemaker.

Childhood lupus affects the body in the same manner as adult lupus, yet boys are more likely to get childhood lupus than men are likely to get adult lupus. Childhood lupus also affects certain organs such as the kidneys to a greater

degree, and the incidence of kidney disease is two times greater. It also requires more aggressive treatment than adult lupus.³

Symptoms of Lupus

On the "Good Morning America" segment, Dr. Susan Manzi, co-director of the Lupus Center of Excellence at the University of Pittsburgh Medical Center, said: "Lupus patients are like snowflakes. No two present the same; no two look the same." In fact, signs and symptoms may come on suddenly or develop slowly, they may be mild or severe, and they may be temporary or permanent. Most patients have episodes, called flares, when signs and symptoms get worse for a while and then improve or even disappear completely for some time.⁴

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The symptoms of lupus depend upon what body organs are affected and how seriously they are affected. Those with SLE typically are extremely tired and have skin rashes or joint pain. In fact, nearly all people with lupus have mild to extreme fatigue, and increased fatigue is a sign that a flare is about to occur. Most people with lupus have joint pain at some time, and about 70 percent of lupus patients report that joint and muscle pain was their first sign of the disease. Most lupus patients also develop skin rashes, which are often an important clue to the diagnosis. Typically, a butterfly rash appears over the cheeks and bridge of the nose. Other common skin symptoms include skin sores or flaky red spots on the arms, hands, face, neck or back; mouth or lip sores; and a scaly, red or purple raised rash on the face, neck, scalp, ears, arms and chest.

Other symptoms of lupus include sensitivity to light, nervous system symptoms (most commonly headaches, but also difficulty with memory or concentration, or numbness or weakness of the arms or legs), heart problems, lung problems, mental health problems (anxiety and depression), low-grade fever, changes in weight, swollen glands, inflammation of blood vessels in the skin, swelling of the hands and feet, and anemia. Some people with

lupus also have Raynaud's phenomenon, which affects the small vessels that supply blood to the skin and soft tissues under the skin of the fingers and toes, causing them to turn white and/or blue or red. With Raynaud's, the skin affected will feel numb, tingly and cold to the touch.⁵

There also are other conditions that cause symptoms similar to lupus. These conditions include rheumatoid arthritis, glomerulonephritis (inflammation of the kidneys), scleroderma (a disease of the body's connective tissue), Sjogren's syndrome (a condition that causes dry mouth and dry eyes), chronic fatigue syndrome and vasculitis (inflammation of the blood vessels).⁶

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Causes of Lupus

Dr. Manzi says that lupus patients often think they may have somehow "caught" the disease. But, lupus isn't contagious. Nor is there a single moment that may have triggered the onset of the disease. In fact, doctors and researchers are unsure what exactly causes lupus, but they do believe that it is a combination of elements. The first is genetics. While there is no scientific proof, it is believed that heredity is at least one factor in determining an individual's propensity for developing lupus. However, if a person has a family history of the disease, he or she may be more susceptible to it, but it doesn't mean he or she will get it.

The second element is the environment. It's possible that certain environmental factors may trigger lupus. Those factors include exposure to ultraviolet light, smoking and stress, and exposure to toxins such as trichloroethylene in well water and silica dust. In the past, it was believed that certain hair products and topicals caused lupus, but that's no longer true.

While research concerning the link between lupus and hormones is in its infancy, the findings are still nebulous, with some studies finding a link and others failing to find a link. However, abnormal estrogen metabolism is considered a risk factor. Viruses and bacteria also are considered

risk factors. Individuals with viruses like cytomegalovirus, parvovirus and hepatitis C may develop lupus, but no causal link has been established. It is known that the Epstein-Barr virus in children is linked to childhood lupus. Last, long-term use of some medicines can trigger lupus and lupus flares.⁷

Diagnosing Lupus

Because no two people present with the same signs and symptoms, lupus can be difficult to diagnose. Plus, there is no one test that can diagnose lupus. The disease is typically diagnosed through a combination of laboratory tests, signs and symptoms, and physical examination findings.

Laboratory tests include both urine and blood tests. A complete blood count to measure the number of red and white blood cells and platelets, as well as the amount of hemoglobin (a protein in red blood cells), could show anemia, which commonly occurs in lupus. A low white blood cell or platelet count also may occur in lupus. A blood test to determine the erythrocyte sedimentation rate (the rate at which red blood cells settle to the bottom of a tube in an hour) can be conducted to see if there is a faster than normal rate, which may be a sign of a systemic disease such as lupus. Since lupus can affect organs, a blood test can assess how well an individual's kidneys and liver are functioning. And, a urinalysis may show an increased protein level or red blood cells in the urine, which may occur if lupus has affected the kidneys. Another lab test, the antinuclear antibody (ANA) test, looks for the presence of these antibodies, which may indicate a stimulated immune system. However, while most people with lupus have a positive ANA test, most people with a positive ANA test do not have lupus. So, more specific antibody testing may be needed.

Imaging tests also may be needed if it is suspected that lupus is affecting the lungs or heart. A chest X-ray can reveal abnormal shadows that suggest fluid or inflammation in the lungs. And, an echocardiogram, which uses sound waves to produce real-time images of the heart, can check for problems with valves and other portions of the heart. Because lupus can harm the kidneys in different ways and treatments can vary, in some cases, it may be necessary to take a biopsy of kidney tissue. The biopsy is obtained with a needle or through a small incision.⁸

Beyond lab tests, there are criteria developed for diagnosis of lupus. In 1997, the American College of Rheumatology (ACR) updated its criteria for the third time. According to the ACR, a patient may have lupus if four or

more of 11 criteria are present without any time restriction. For a full list of the ACR's criteria, go to www.rheumatology.org/practice/clinical/classification/SLE/1997_update_of_the_1982_acr_revised_criteria_for_classification_of_sle.pdf.

Treatment for Lupus

How lupus is treated also depends upon the disease signs and symptoms. Fortunately, treatments have increased dramatically in recent decades. Which medications are used to treat the disease is based on the patient's age, sex, health, symptoms and lifestyle. And, as signs and symptoms flare and subside, these medications and their dosages may need to be changed.⁹

Medications most commonly used to control lupus include nonsteroidal anti-inflammatory drugs (NSAIDs — either over-the-counter or those available by prescription), antimalarial drugs (such as hydroxychloroquine [Plaquenil]), corticosteroids (such as prednisone) and immune suppressants (such as cyclophosphamide [Cytoxan], azathioprine [Imuran, Azasan], mycophenolate [Cellcept], leflunomide [Arava], and methotrexate [Trexall]).¹⁰

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NSAIDs decrease inflammation, and they may be used alone or in combination with other types of drugs to control pain, swelling and fever. Antimalarials were originally used to treat malaria, but clinical studies have found that continuous treatment with antimalarials may prevent flares of lupus from recurring. Corticosteroids, closely related to a natural anti-inflammatory hormone, work by rapidly suppressing inflammation, and because they are so potent, doctors seek the lowest dose with the greatest benefit. Immunosuppressives restrain the overactive immune system by blocking the production of immune cells.

A few other therapies also are available. Belimumab (Benlysta), a B-lymphocyte stimulator (BLys) protein



inhibitor, was approved by the U.S. Food and Drug Administration in March 2011 for patients with lupus who are receiving other standard therapies. It is given by infusion to reduce the number of abnormal B cells thought to be a problem in lupus patients. However, studies to date have shown that African-American patients and patients of African heritage do not respond to belimumab. Methotrexate (Folex, Mexate, Rheumatrex), a disease-modifying antirheumatic drug, may help control the disease in some patients. And, dehydroepiandrosterone (DHEA) and intravenous immunoglobulin (IVIG) may be used for controlling lupus when other treatments haven't worked.⁹

Managing Day to Day

Lupus can significantly impact an individual's quality of life. A study on work loss associated with lupus estimated that almost three-quarters of the study's 982 participants would stop working before the usual age of retirement, and half of those who had jobs when they were diagnosed (during their mid-30s on average) would no longer be working by age 50.⁹

Most patients, like Kelly Jean Drury, live their lives day by day, never knowing what the future holds. Yet, lupus patients can maintain a high-quality lifestyle. First, it's important that they learn the warning signs of a flare and take steps to reduce its intensity. Periodic increases in disease activity can usually be managed by varying medications. And, during flares, lupus patients should avoid ultraviolet light.¹¹

They also must receive regular healthcare, rather than seeking help only when symptoms worsen. Women should receive regular preventive healthcare, such as gynecological and breast examinations. Men should have the prostate-specific antigen test. And, both men and women should have their blood pressure and cholesterol checked on a regular basis. If taking corticosteroids or antimalarial medications, an eye exam should be performed at least yearly. And, patients should be aware of their increased risk of premature cardiovascular disease.⁹

Most people with lupus will live a normal life span due to improved diagnosis and treatment.

Women with lupus are considered high risk. They have an increased risk of miscarriages and can have flares during pregnancy. Therefore, they require close observation during pregnancy, delivery and the postpartum period, including fetal monitoring by the obstetrician during later pregnancy. Those who are at risk for miscarriages can be identified with a test for the presence of phospholipid antibodies in the blood. If these antibodies are present in a woman with lupus, blood-thinning medications and IVIG for selected people can be used.¹¹

Future Outlook

Researchers are working hard to answer several questions about lupus: Why are women more likely than men to have the disease? Why are there more cases of lupus in some racial and ethnic groups, and why are cases in these groups often more severe? What goes wrong in the immune system and why? How can immune system function be corrected once something goes wrong? What treatment approaches will work best to lessen lupus symptoms? And, how can lupus be cured?

The National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), a component of the U.S. Department of Health and Human Services' National Institutes of Health, is funding researchers across the U.S.

who are developing new and better ways to study the disease. These include laboratory studies that compare aspects of the immune systems of people with lupus with those of others both with and without lupus, as well as using mice with disorders resembling lupus to better understand the abnormalities of the immune system that occur in lupus and to identify possible new therapies.⁹

The Lupus Foundation of America is also on the forefront of fighting this disease. But, as Dr. Manzi explains in the "Good Morning America" segment, the foundation needs a Michael J. Fox for lupus; it has no spokesperson to be its advocate. "This is a very devastating disease," says Dr. Manzi. "It can be fatal. Kelly's very lucky that her disease is under control, but many people die of this disease. So, we need more research dollars; we need more help."

The overall outlook for people with lupus is improving every decade. It is unknown whether the number of deaths attributed to the disease has been on the rise in the last 20 years due to the actual increase in mortality or just better identification and reporting of the disease. But, it is known that most people with lupus will live a normal life span due to improved diagnosis and treatment.⁷ ■

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