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## Systemic Lupus Erythematosus (SLE)

by Jennifer Pope

(Anatomy & Physiology 1551)

**A**n expansive range of symptoms appears and attacks without warning, has no known cause, and there is no known cure-Lupus. Systemic Lupus Erythematosus is called by the Lupus Foundation of America, Inc. (2016), "The Cruel Mystery". This paper will define Systemic Lupus Erythematosus (SLE or Lupus), explain the links thought to contribute to the etiology, list the common symptoms of SLE and health effects, and outline the current treatment options and research. The cruel mystery of lupus stems from its health effects ranging from a butterfly shaped rash, stroke, heart attack, organ inflammation, to hair loss ("What", 2016, p.1). The Lupus Foundation estimates that there are 1.5 million Americans who have a form of lupus and about 5 million people through the world ("Statistics", 2016, p.1). The symptoms and health effects of SLE are many, but the cure and cause are still a mystery.

SLE is an autoimmune disease where the immune system makes autoantibodies that attack and destroy healthy tissue, which can cause inflammation and damage. Daniel J. Wallace, MD in his book *The Lupus Book* (2013) says, "In simple terms, lupus erythematosus develops when the body becomes allergic to itself...In lupus, the body overreacts to an unknown stimulus and makes too many antibodies, or proteins directed against the body tissue. Thus, lupus is called an autoimmune disease (auto meaning self)" (p. 5). The immune system is the body system that produces proteins called antibodies that destroy foreign threats. However, the immune system of a person with SLE malfunctions and creates autoantibodies that attack some of the healthy cells in tissues and organs. These autoantibodies continue to destroy healthy tissue and other immune cells join; thus, leading to inflammation (Starkebaum, Zieve, Oglivie, & A.D.A.M. Editorial team, 2015, p.1). A more in depth look at the pathophysiology can be obtained from *The Lupus Book* (2013) by Daniel J. Wallace M.D. where he describes the enemy is in the cells.

Neutrophils (the white blood cells responsible for mediating acute inflammation) can increase inflammation in the body of lupus sufferers because of the way their blood plasma interacts with cytokins, complement, and adhesion molecules (the chemicals that draw cells closer to the site of inflammation). Lymphocytes, the white blood cells responsible for chronic inflammation, also have their function altered in lupus. The T-helper cells become more active, and the body becomes less responsive to T-suppressor cells. Natural killer lymphocytes promote inflammation and are not able to suppress or contain it. As a result, the body's system of tolerance is disrupted so much that B cells are signaled to make antibodies to the patient's own tissues, which are called autoantibodies. (p.27)

The normal immune system is changed in a person with lupus at the cellular level as the inflammatory responses and production of antibodies are at a higher rate, making an immunological abnormality. The autoantibodies are a key indicator of lupus. Daniel J. Wallace M.D. lists the antibodies in lupus patients in his book mentioned above.

These include antibodies that form against materials in the nucleus or center of the cell, such as antinuclear antibody, anti-DNA, anti-Sm, anti-RNP, and antihistone antibody; antibodies that form against cytoplasm or cell surface components, such as anti-Ro (SSA), anti-La (SSB), antiphospholipid, and antitribosomal blood cells, platelets, or nerve cells; and antibodies that form against circulating antigens, such as rheumatoid factors and circulating immune complexes. (p. 27)

The existence of these antibodies can be indicators to help diagnose patients, but the presence in blood tests is not enough diagnostic criteria to be fully conclusive. Antiphospholipid antibodies are made to destroy phospholipids and associated blood proteins. According to John Hopkins webpage "How lupus affects the body" (2016), fifty percent of lupus patients have these antibodies and may experience a blood clot. Antibodies can attack nerve cells and blood vessels impeding blood flow in the nervous system leading to cognitive issues, headaches, or vasculitis in the main nervous system (p.1). The inflammation in organs for lupus patients is due to the deposit of this immune complex (Wright & Bharadwaj, 2010, p.1).

Understanding the pathophysiology of lupus in the body brings forth the question of what the etiology is. According to the Center for Disease Control (CDC), lupus primarily occurs in women approximately four to twelve women for every man, and more frequently occurring in the childbearing years for females. The CDC states, "Blacks (and probably Hispanics, Asians, and Native Americans) are affected more than whites. People with a family history of SLE or other autoimmune diseases are at slightly higher risk for developing SLE" ("Systemic", 2015, p.1). Even with these concentrations of who is affected by lupus it truly can affect any gender, race, ethnicity, and age ("What", 2015, p.1). The CDC's webpage "systemic lupus erythematosus" says, "Among rheumatic conditions, SLE has a relatively high mortality (14.5% of all rheumatic disease mortality in 1997" (p.1). There has been improved diagnosis and management of the disease, but it is believed that approximately 10-15% of people with lupus will die due to complications ("Statistics", 2016, p.1). People with lupus can die from kidney disease, heart attacks, and related cardiovascular diseases. So, it is important for those with lupus to lead a healthy lifestyle lowering risk factors for heart disease ("What are the most", 2013, p.1). The research done thus far has found some trends or links to a possible cause yet inconclusive.

According to John Hopkins Lupus Center website (2016), the number one lupus center in America, there are links between lupus and genes, hormones, and environmental factors. Genes may have a role in lupus as the MHC class II and III are genes that make codes for proteins that interact with antigens and involve the inflammatory response. The malfunction of these genes is associated with lupus. The other genes linked to lupus are the genes that create the coding for opsonins, complement receptors, and antibody receptors ("Causes", 2016, p.1).

The second element in the etiology complexity of lupus are hormones. According to The John Hopkins Lupus Center (2016), women are nine times more likely to develop lupus than men. The higher levels of the female sex hormone estrogen enhance the function of the immune system. Women notice more symptoms around the menstrual cycle as well. "For this reason, the incidence of autoimmune diseases is generally higher in women than in men" ("Causes", 2016, p.1).

The third element in the etiology complexity of lupus is environmental. According to The John Hopkins Lupus Center, "These environmental contributors are difficult to isolate, but researchers have established links between lupus and a variety of toxins, such as cigarette smoke, silica, and mercury. Infectious disease agents such as the Epstein-Barr Virus (EBV, which causes mononucleosis or "mono"), herpes zoster virus (the virus that causes shingles), and cytomegalovirus have also been implicated" ("Causes", 2016, p.1). There has been links between

certain drugs springing the onset of lupus symptoms, and ultraviolet light has been shown to agitate lupus.

Though the cause of lupus is still a mystery, the symptoms and related conditions are vast and at times cruel. Symptoms vary in every patient and can come and go through periods where there is a flare up of symptoms and periods where there is dormancy. According to the CDC, "Patients with SLE have an increased frequency of other autoimmune problems, such as Sjogren's syndrome (i.e., dry eyes, dry mouth) and antiphospholipid syndrome (i.e., clotting problems, strokes, fetal loss) that require additional treatments" ("Systemic", 2015, p.1). Other common symptoms are fatigue, fever, malaise, mouth sores, hair loss, butterfly shape rash, ultraviolet light sensitivity, swollen joints, and dizziness. The nervous system can be affected and result in headaches, numbness, tingling, seizures, vision problems, and personality change. The digestive system can be affected causing abdominal pain and nausea. The cardiovascular system can be affected causing arrhythmias. The respiratory system can be affected causing difficulty breathing in the lungs (pleurisy). The integumentary system may be affected causing patchy skin color or sores known as discoid lupus. The vascular system and circulatory system can be affected if Raynaud phenomenon develops causing a narrowing of blood vessels. The kidneys are a common organ that can become inflamed as lupus attacks (Starkebaum, Zieve, Oglivie, & A.D.A.M. Editorial team, 2015, p.1). The American College of Rheumatology article "Guidelines for Referral and Management of Systemic Lupus Erythematosus in Adults" indicates the uncertainty of symptoms and severity that can ensue. "In addition to the persistent risk of disease flares, more than one-half of SLE patients may develop permanent organ system damage" (Hahn, B.H. et al., 2012, p.1). As the body is working against itself this can give rise to uncertainty and fear in patients.

The pain and fatigue can interfere with everyday life, and quality of life has been researched as well. In a qualitative study on uncertainties and opportunities in lupus patients by Mattsson, M., Moller, B., Stamm, T., Gard, G., and Bostrom, C., one participant said, " '...It's so inconsistent, so to speak, it's so variable. I often say SLE is cancer number two, because you don't know if you will be able to be so very lively today, but you'll be having a flare-up next week, just like that, it's so very unpredictable ...'(IB)" (Mattsson, Moller., Stamm, Gard., & Bostrom 2015). There is a need for more research on the occupational therapy and counseling aspects to help patients cope with the physical, cognitive, and emotional impacts. From Jill Buyon's Abstract for "Systemic lupus erythematosus" (2015), she describes the complexity of the disease in relation to symptoms and manifestation as follows.

Indeed, it is the diversity of presentation, accumulation of manifestations over time, and undulating disease course that challenge the most astute of clinicians. With rare exception, the unifying laboratory abnormality is the presence of circulating antinuclear antibodies (ANA). Acknowledging the complexity of this disease, its broad differential diagnosis, and the need to develop better and more specific therapies, the American College of Rheumatology (ACR) has designated 11 diagnostic criteria...The presence of four or more criteria is required for diagnosis. They need not necessarily present simultaneously: a single criterion such as arthritis or thrombocytopenia may recur over months or years before the diagnosis can be confirmed by the appearance of additional features. (Buyon, J, 2015, p.1)

With this being said, symptoms and severity vary from patient to patient and it is unpredictable the exact course the disease may take. The Systemic Lupus International Collaborating Clinics (SLICC) group revised and validated the ACR criteria for lupus. The

following summarizes the final criterion established from Table 3 in the article "Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus"(2012). Clinical criteria include the following major classifications which include acute cutaneous lupus, chronic cutaneous lupus, oral ulcers, nonscarring alopecia, synovitis involving two of more joints over thirty minutes, serositis, renal showing urine protein-to-creatinine ratio of 500mg protein or red blood cell casts, neurologic (seizures, psychosis, mononeuritis multiplex, peripheral or cranial neuropathy, acute confusional state), hemolytic anemia, leukopenia, thrombocytopenia. The immunological criteria list is ANA level above range, presence of Anti-dsDNA, presence of Anti-Sm, Antiphospholipid antibody positive, low complement (C3, C4, CH50), and Direct Coombs' test (Petri, M. et al., 2014, p.1).

The patient must present with four of the above criteria to be diagnosed with this disease. The common tests done are complete blood count, erythrocyte sedimentation rate, kidney and liver assessment, urinalysis, antinuclear antibody, chest X-ray, echocardiogram, and biopsy of kidney at times (MayoClinic Staff 2014). Once diagnosed, there are certain treatment depending on the symptoms.

There are treatment options for lupus patients depending on severity and activity such that the rheumatologist may have different treatments over the patient's lifetime that need to be followed. NSAIDS are anti-inflammatory medications that help with muscle, joint, and other tissue inflammation ("Lupus medications", 2016, p.1). Anti-Malarial drugs such as Plaquenil are used to control lupus over the lifetime helping suppress symptoms like fatigue, rashes, joint pain, mouth sores, and abnormal blood clotting. In certain cases, cylophosphamid (a chemotherapy drug) may be used to help tame the lupus activity. Steroids are synthetic cortisone medication that are effective at reducing the swelling, pain, and inflammation of lupus, but have many risks with the benefits. (Ginzler, E., Tayar, J., & American College of Rheumatology Committee, 2015, p.1). Immunosuppressive medications help to suppress the immune system from overworking. DHEA is a mild male hormone that helps with some symptoms such as hair loss, joint pain, fatigue, and cognitive dysfunctions (John Hopkins). When lupus starts inflaming and attacking the kidneys (nephritis) it can result in end-stage renal disease and there has not been a decline in numbers since 2004 even despite therapeutic treatments developed (Hahn, B. H. Overall, 2012, p.1). A newer treatment called Benlysta or belimumab approved in 2011 by the FDA for adult SLE sufferers, has been used to treat arthritis symptoms (Ginzler, E., Tayar, J., & American College of Rheumatology Committee, 2015, p.1). The reevaluation of the treatment plan, tests, and symptom monitoring must be through the patient's lifespan to ensure control of this disease.

In conclusion, SLE is an autoimmune disease that causes autoantibodies to generate and attack healthy tissues and creates inflammation by expanding blood vessels or complex deposits overtime. SLE can affect any organ and changes overtime with frequency of flares. SLE's exact cause is still unknown, but there are some links between a complex interplay of genes, hormones, and environmental factors currently being reviewed. There is a slew of symptoms involving multiple body systems, and they vary patient to patient. The diagnostic criteria require a person to have four of the criteria to be diagnosed. The Lupus Foundation of America is one of the largest organizations that is bringing more awareness for this disease and research. The Lupus Foundation of America has started a 2016 peer-reviewed research fund to study stem cells, environmental triggers of lupus in children and teens, and provide funding for research that has been delayed ("Lupus Foundation", 2016, p.1). Though lupus is deemed "The Cruel Mystery", there is research past and current that is helping to piece together the aspects of this mystery to unmask the cause and develop a cure.

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