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|  | **What is Lupus**?  **-The Mechanics:**  What is Lupus? This is a question that has been puzzling scientists and doctors since the onset of this disease but many people have succeeded in compiling broad and ever changing definitions of the disease. According to The Lupus Foundation of America, (2000), Lupus is defined as a chronic, autoimmune disease which causes inflammation of various parts of the body, especially the skin, joints, blood, and kindneys. Lupus, of systematic lupus erythematosus, has been such a challenge to the medical world because it is complex and the problems mimic many diseases grouped together, so it is very hard to understand. The immune system plays a major role in Lupus. The immune system usually protects the body against viruses, bacteria, and other foreign materials called antigens. In Lupus, the autoantigens of the immune system that trigger the immune response cannot distinguish these antigens from itself and so it attacks its own healthy tissue and organs. The immune system has three cells that play an active roll. They are the B-lymphocytes (B cells), the T lymphocytes (T cells), and the phagocytes. The major roll of the lymphocytes is that they are part of the white blood cells that produce lymphoid tissue, the main cells in the immune system that make antibodies, often called "Killer B and T cells" (Lahita). The T cells recognize the foreign antigens and they either kill it, or give it the the B cells which then make an antibody to kill it through a process known as autoimmune response. There are five proteins in that make-up the immune system. They are classified into five classes of immunoglobulin molecules (that kill foreign substance) and that are made by the B cells. The way these proteins work is that antibodies attach themselves to foreign material, called antigens, and form an immune complex where the foreign material is engulfed by a scavenger cell (phagocyte) which then destroys the immune complex. The waste material then travels to the spleen, which serves as a sort of garbage can for immune complexes. This process is called the antigen-antibody reaction. Inflammation is also common in Lupus and usually occurs in joints where white blood cells and immune complexes are present. Inflammation is the reaction of tissues in response to infection, injury, or invasion. It can take the form of swelling, pain, heat, redness, and impaired function as a result of an immune system reaction. The body uses inflammation to protect the damaged are from its surroundings, by conducting a series of chemical reactions with molecules called mediators. Lupus patients often experience swelling or inflammation in healthy joints due to the unnecessary immune reactions and immune complexes that the body is producing. In conclusion, Lupus is a result of the malfunction of the immune system, where it can not distinguish foreign antigens from its own healthy tissue, resulting in a person�s immune system to attack its own healthy body.  **-The Types:**  Lupus can take on three major forms: discoid lupus, systematic lupus, and drug-induced lupus. A fourth type is also known, neonatal LE, but it is not as common. All of these types of Lupus are the same in that they are all chronic diseases with similar symptoms, but they can have very different effects.  ***1. Discoid Lupus:*** The first form, discoid lupus, is not as serious as systematic LE as it mainly involves only the skin, not internal organs. It results in a negative ANA test and begins with patches of skin becoming red, scaly, and itchy, but painless, usually due to sun exposure. These are called "discoid lesions" (Saywell). They usually occur on the cheeks, back of hands, and nose and eventually heal but usually leave scars or discoloration of the skin, called reticulate telangectasia. When these lesions occur on the scalp, hair loss, called alopecia, is usually experienced as they heal. The lesions can also be aggravated by more sun exposure, shampoo, or perfume. Some diseases that are common among Discoid LE patients are chilblains, Raynaud�s Phenomenon, where the hand and feet turn blue and white due to exposure to cold temperatures and temperature changes or stress, and anemia (low hemoglobin). Also, Doctors Christina and Laura Saywell report that joint pain is also common and around 55% of discoed LE patients have at least one abnormal blood test such as anemia or positive ANA (anti-nuclear antibodies) (Saywell 2). Discoid LE can be diagnosed with a skin biopsy where it is observed under a light microscope and in discoid patients, the dermal junction lights up. To treat the lesions associated with Discoid LE, topical and intralesional steroid treatments are usually very effective, however, antimalarial drugs may also be needed. Medakate, a Lupus support and educational information group, reports that Discoid LE is considered a mild and occasionally severe disease that only rarely progresses to systematic LE. This may be true; however, there has been extensive controversy of whether Discoid and Systematic LE are related, if one leads to the other, or if they are two separate diseases all together. There is substantive evidence to support both sides. Here it is as presented by Dr. Christina Saywell of the Skin and Cancer Foundation and Dr. Laura Saywell of Sydney:  -***Evidence supporting that DLE is linked to SLE:***  1. "Discoid" lesions occur in both diseases and 5% of patients who initially experience discoid lesion eventually progress to SLE. Also, up to 20% of SLE patients get a discoed lesion at some point during their disease.  2. There have been very similar microscopic observations of skin biopsies and blood test abnormalities between Discoid and systematic patients.  3. In both Discoid LE and Systematic LE, Lupus Profundus may occur, which is involvement of the fatty tissues below the skin.  ***-Evidence Supporting that DLE and SLE are totally separate diseases:***  1. There is a very acute risk of only 5% that DLE patients will progress to SLE  2. There have been different immunoflorecence findings, which is a specialized pathology test, on the skin biopsy between SLE and DLE  3. DLE and SLE patients have genetic differences where they are genetically programmed to develop one form but not the other, and this can not be altered as a person� s genetic make-up is imprinted for life.  The evidence shows that the two diseases are definitely very closely related, but doctors must be careful in associating the two directly due to the differences in genetic compositions. Also, statistics show that the risk of a DLE patient progressing to SLE is very low. There are some factors that suggest a DLE patient is more likely to develop SLE. Some of these include: widespread lesions, not responding to treatment, kidney disease, repetitive abnormal blood tests showing positive SS-DNA and DS-DNA, a very accurate antibody test for diagnosing SLE. Again, Discoid LE is not as serious as Systematic LE, can usually be successfully treated, and is almost never fatal.  **2. Drug-Induced Lupus:** A second form of Lupus is called Drug-Induced Lupus and it occurs when people take certain drugs that aggravate symptoms that mimic the disease. The Lupus Book reports that 15,000-20,000 new cases of DILE are diagnosed each year in the US. Ninety percent of these cases derive from the top three drugs that aggravate the disease, according to Dr. Daniel J. Wallace, are Pronestyl (procainamide), Apresoline (hydralazine), and methyldopa (Aldonet). Wallace also adds that when isoniazid (INH), chlorpromazine (thorazine), and D-penicillamine are also considered, that 99% of all clinically relevant cases can be accounted for. This has been a problem because these drugs are prevalently used today to treat heart irregularities called arrythmias, high blood pressure, and tuberculosis. Other drugs that have been known to cause the disease are beta-blockers, tricyclic antidepressants, penicillin, birth control pills, antibiotics, NSAIDS, and sulfa drugs. No one knows exactly how these drugs cause Lupus, but surprisingly, these drugs do not affect patients who already have the disease. Also, this kind of Lupus has no involvement with genetics, but instead there are certain cell markers called the transplantation antigens or histocompatibility markers that predispose people to diseases. DR4 is a marker that has been associated with patient of Drug-Induced Lupus, however the disease can affect virtually anyone who takes the drugs and is biochemicaly and genetically susceptible to the disease. This is reflected in the statistics of SLE vs. DILE. In SLE, women compose 90% of the patients, where as in DILE, the disease is equally distributed among men and women, according to The Lupus Book. DILE is also very rare among African-Americans, which is the most prevalent race of SLE patients. And finally, the average age of diagnosis for SLE is from 20 to 40 years of age, where DILE is most common at age 60 (Wallace). Symptoms of DILE are very similar to those of SLE including joint pain, rash, fever, and pain in breathing, however, DILE patients rarely experience symptoms indicating organ involvement. The first step in treating DILE is to stop the use of the problem drug immediately. If this is done, no further therapy may be needed. If the case is severe, steroids may be prescribed. The good news about drug-induced lupus is that unlike the other forms of Lupus, the disease is completely reversible with proper treatment.  **3. Neonatal Lupus:** Neonatal Lupus is a very rare condition which occurs when the baby is diagnosed with Lupus at birth. Usually there is no risk of a mother with Lupus transmitting the disease to her baby, but an exception is made for a small 10% of women who have the anti-Ro antibody and can pass on the disease, says Dr. and Dr. Saywell. This occurs when the placenta is crossed with the mother�s anti-Ro antibody and causes Lupus in the growing fetus. The baby is usually born with or quickly develops lesions around the eyes that are lost within twelve months when the mother�s antibodies circulate through and leave the baby�s body. The babies do not actually have Lupus themselves, but can develop heart problems, which require a pacemaker for the rest of the baby�s life. Usually, there is no sign of Lupus in the mother during pregnancy, so she is not usually tested until signs of the baby�s rash develop. Unfortunately, most of these "healthy" women usually go on to develop connective tissue diseases such as Lupus several years down the road. Surprisingly, this form of Lupus usually indicated more problems for the mother than for the child.  **4. Systematic Lupus:** The last form of Lupus, the one that my survey was focused on, is called systematic lupus, or systematic lupus erythematosus. Systematic means "all over" and erythematosus means "red." It got its name from the red skin lesions that occur in 80% of patients with the disease, similar to those in Discoid LE (Saywell 4). Even though DLE and SLE share common symptoms, SLE is a much more serious and life-threatening disease as it affects both the skin and internal organs. In addition to skin lesions, ulcers and weals also appear, indicating damage to the blood vessels. The most distinctive external symptom of SLE is the malar butterfly rash that occurs over the cheeks and bridge of the nose, though it is only present in one third of patients (Excite health home). Some other symptoms of SLE include fatigue; fever; rash that is raised with white bumps; red fingertips, toes, fingernails, palms, or soles of the feet; vasculetis (inflamed small blood vessels and ulcers); a fishnet pattern on the skin; pockets of inflamed shin; hair loss; sun sensitivity; ulcers in the mouth or nose; joint pain; nodules on tendons (usually elbow or ankle); muscle inflammation; swollen lymph nodes; enlargement of the partoid glands (in cheeks); inflammation of the lining of heart (perdicarditis) or lungs (pleurisy); low platelet count; low white blood cell count; high blood pressure; weight loss; joint pain; seizures; psychosis; numbness and tingling; blood in urine; nausea; and vomiting. There are so many symptoms for the disease because every patient is different, experiencing different symptoms and new ones seem to arise in every new case. Besides causing overall weakness, fatigue, and malaise (general discomfort), SLE also attacks organ systems. According to Dr. Robert G. Lahita and Mr. Robert H. Philips, here is a general overview of how the disease affects each system:  ***-Skin:***  rashes, lesions, red inflammations, hair loss, ulcers, discoloration, and sun sensitivity  ***-Chest:*** pain from pleurisy (inflammation of the lining of the lungs) and perdicarditis (inflammation of the lining of the heart), difficulty breathing, pain, shortness of breath, and rapid heartbeat.  ***-Muscular:*** weakness, aches, pains, and inflammation  ***-Joints:*** arthritis-like pain, swelling, redness, and stiffness  ***-Blood:*** low red blood cell count (anemia), low white blood cell count (easy infection), raised gamma globulin, false positive syphilis test, high ANA, high DS-DNA  ***-Cardiac or Circulatory:*** swelling of extremities, fluid in sack around the heart, Raynaud�s Phenomenon  ***-Eye Problems:*** hemorrhages, cotton wool spots, clotted blood vessels  Systematic Lupus is a chronic disease because there is no cure, but it is also acute because it does not show itself all the time, but instead occurs in flares, or bursts of disease activity. The remainder of the research is devoted to the diagnosis, treatment, and possible causes of this complex and growing disease know as Systematic Lupus Erythematosus.  **-The Criteria Set By the American College of Rheumatology (ACR):**  The American College of Rheumatology (ACR) is a professional association to which almost all of the United State�s Rheumatologists belong. The ACR developed a criterion for defining the disease in 1971, and then revised it in 1982. These criteria are specific to Systematic Lupus and do not apply to Discoid or Drug-Induced Lupus. Four of the eleven criteria must be met for a patient to be diagnosed with SLE. The first four deal with the skin, the second four deal with specific organ areas, and the remaining three deal with abnormalities in lab tests. There are many other symptoms of SLE that are not included in the eleven criteria, such as Raynaud�s Phenomenon, because they are not present only in SLE.  -**ACR (1982) Revised Criteria for the Classification of Systematic Lupus Erythematosus as published by The Lupus Book (1995) is:**  **Skin Criteria:**  1. Butterfly rash (lupus rash over cheeks and nose)  2. Discoid rash (a thick, disk-like rash that scars, usually on sun-exposure)  3. Sun sensitivity (rash after being exposed to ultraviolet A and B light)  4. Oral ulcerations (recurrent sores in mouth or nose)  **Systematic Criteria:**  5. Arthritis (inflammation of two peripheral joints with tenderness, swelling, or fluid  6. Serositis (inflammation of lining of the lung-also called pleura-or the heart-also called pericardium)  7. Kidney disorder (protein in urine samples or abnormal sediment in urine seen under the microscope)  8. Neurologic disorder (seizures or psychosis with no other explanation)  **Laboratory Criteria:**  9. Blood abnormalities (hemolytic anemia, low white blood cell counts, low platelet counts)  10. Immunologic disorder (blood testing indicating either a positive LE cell preparation, anti-DNA, false-positive syphilis test, or a positive anti-Sm)  11. Positive ANA blood test  I will now discuss the processes and tests that involve the analysis of symptoms and lab tests and all of the factors that contribute to the diagnosis of Systematic Lupus Erythematosus.  ([Next)](http://docs.google.com/intro5.html) | |
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