

reduced in chronic interstitial lung disease. High-resolution CT scans are used to look at the lung and chest structures. Chronic lupus interstitial lung disease is primarily treated with corticosteroids, although other immunosuppressive drugs may be necessary. In general, the lung function can be stabilized with treatment.

PULMONARY HYPERTENSION

Very rarely, people with lupus develop pulmonary hypertension or high blood pressure in the blood vessels within the lung. If severe, this can be life-threatening, and there tends to be little chance for improvement. Patients with severe pulmonary hypertension should be on blood thinners. Although newer agents such as Tracleer (bosentan) or prostacyclins may improve symptoms such as shortness of breath, there are no uniformly successful medical treatments. Heart-lung transplants may be an option for some people with pulmonary hypertension caused by SLE.

PULMONARY EMBOLI

Pulmonary emboli are blood clots that block the pulmonary arteries. At first, they cause pleuritic (lung) pain and shortness of breath. These clots can lead to abnormal oxygen exchange in the lung and even death. Tests for the presence of pulmonary emboli include ventilation-perfusion (breathing and blood flow) scans of the lung, and angiography (dye injected into an artery). There may also be an evaluation for thrombophlebitis (inflammation of a vein due to a blood clot). Risk factors for pulmonary emboli in lupus patients include antiphospholipid antibodies, decreased blood levels of protein S, possible vascular damage, and prolonged immobility. If risk factors are present, patients are treated with anti-coagulants (blood thinners). Changes in corticosteroid dose or immunosuppressive medications are not indicated.

PULMONARY HEMORRHAGE

Pulmonary hemorrhage, or bleeding into the lung, is a rare but potentially fatal complication of SLE. Typically, individuals have fever, shortness of breath, and cough. They may have blood-tinged sputum. This is usually seen in the setting of multi-organ system involvement from SLE and a rapidly falling red blood cell count. Treatment usually includes high-dose corticosteroids with immunosuppressive agents. Aggressive supportive care is also crucial to maximize chance for recovery.

SHRINKING LUNG SYNDROME

Occasionally, a lupus patient will have shortness of breath and sharp chest pains without signs of pleuritis or of interstitial lung disease. This rare patient may have the shrinking lung syndrome. In this condition, there is a loss of lung volume. The diaphragm is elevated on the chest X-ray. The shrinking lung syndrome in lupus may be from involvement of the diaphragm. Treatment with corticosteroids and immunosuppressive drugs usually results in improvement of the symptoms and of lung function.

Conclusion

The broad array of cardiopulmonary problems associated with SLE requires a close working relationship between patient and physician. Preventive measures to reduce the number of flares and rapid evaluation of new or changing symptoms are crucial to minimize long-term problems. Treatment is always individualized to the type of heart and/or lung involvement. Ongoing medical supervision is essential to optimize therapy and prevent long-term side effects and complications.

The Lupus Foundation of America

The Lupus Foundation of America (LFA) was established in 1977 to educate and support those affected by lupus and find the cure. The LFA supports research, education, awareness, patient services, and advocacy.

The Lupus Foundation of America is the only nationwide organization exclusively serving individuals, families and friends affected by lupus. The LFA has hundreds of local chapters and support groups throughout the United States, as well as international affiliates around the world.

The LFA is a grassroots, volunteer-driven organization. Contact the LFA or the chapter that serves your area to find out how you can become involved in our mission.

For information about lupus or to locate the chapter nearest you, visit our website at www.lupus.org or call toll-free 1-800-558-0121.

Sign up for our mailing list on the LFA website home page and help pass federal legislation that will benefit people with lupus. You'll receive periodic advocacy updates and other breaking lupus news and information.



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Cardiopulmonary Disease and Lupus

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The heart and lungs are frequently affected in people with systemic lupus erythematosus (SLE) and can cause a variety of problems, ranging from mild to serious or even life-threatening. It is very important to know the differences between cardiopulmonary (heart and lung) complications and non-lupus related problems. When investigating a person with symptoms suggesting a cardiopulmonary problem, a number of possible causes must be considered. Some of these, such as pericarditis, myocarditis, coronary vasculitis, pleuritis, pneumonitis, or pulmonary emboli can occur in SLE. Others, such as infectious pneumonia, esophageal spasm, reflux esophagitis, and costochondritis are not necessarily lupus-related. The medical examination and laboratory and other tests will help to determine the cause of the problem so appropriate therapy can be used.

Cardiac (Heart) Involvement

Lupus can involve all parts of the heart: the pericardium (sac surrounding the heart); the myocardium (muscle layer); the endocardium (lining of the inside of the heart and heart valves); and the coronary arteries.

PERICARDITIS

Pericarditis, or inflammation of the pericardium, the sac around the heart, is the most common heart problem in people with lupus. This condition occurs when antigen-antibody, (or immune) complexes cause inflammation within the pericardium. The usual symptoms include sharp chest pain and occasionally, shortness of breath. The pain can change with changes in position. Frequently, it is relieved by leaning forward slightly. Some cases of pericarditis are asymptomatic, meaning individuals may not experience any physical symptoms.

Blood tests, chest x-rays, an EKG, and an echocardiogram are usually ordered to help diagnose pericarditis. The echocardiogram is an ultrasound of the heart that will tell the physician if there is fluid around the heart. Pericarditis can occur in conditions other than lupus; therefore the cause must be determined before treatment begins. If pericarditis is due to infection or kidney failure, the treatment is different than if it is due to lupus. Lupus pericarditis can be treated with anti-inflammatory agents. If this form of therapy is unsuccessful, corticosteroid treatment will usually result in a beneficial response.

MYOCARDITIS

When lupus causes inflammation of the myocardium, myocarditis occurs. However, serious heart muscle disease is not common in SLE. The symptoms of myocarditis include: unexplained rapid heart beat; abnormal electrocardiogram; irregular heart beat; and heart failure. Myocarditis is often seen with inflammation of other muscles in the body. Treatment of lupus myocarditis usually includes high doses of corticosteroids. Immunosuppressive drugs such as Cyclosporin (cyclophosphamide), Imuran (azathioprine), or intravenous immunoglobulin (IVIg) may be added if the inflammation is not completely controlled with corticosteroids. Myocarditis can lead to tissue damage when functional heart tissue is replaced by scar tissue.

ENDOCARDITIS

When lupus causes inflammation of the endocardium (endocarditis), the heart valves can be damaged, but this condition rarely affects the pumping efficiency of the heart. Valvular dysfunction may be detected by auscultation (listening with a stethoscope) or an echocardiogram. The surface of the affected valves may become thickened or develop wart-like growths called Libman-Sacks lesions. Although these growths may cause heart murmurs, it is uncommon for them to seriously affect the function of the valves. If bacteria lodge in the growths, infection (bacterial endocarditis) can occur. This too is uncommon, but is potentially very serious and requires hospitalization. Rarely, the inflammation and scarring of valves leads to a deformity requiring surgical valve replacement.

CORONARY ARTERY DISEASE

The most common abnormality of the coronary arteries in SLE is of premature narrowing caused by atherosclerosis. These arteries deliver blood and oxygen to the heart muscle and are vital to the heart's pumping function. Narrowing or blockage of an artery (coronary artery disease) can lead to angina (chest pain) and even a heart attack. The narrowing of the coronary arteries in people with lupus may occasionally be due to inflammation of the blood vessel wall (vasculitis), arterial spasm, or blood clots.

Atherosclerosis is the most common cause of coronary artery disease in lupus. Lupus patients may have more cardiac risk factors than other people. They may have hypertension (from kidney disease or corticosteroid use), elevated cholesterol or diabetes from corticosteroids and be inactive because of joint problems or from general lupus activity. However, even after taking into account the increased prevalence of these risk factors for coronary artery disease lupus patients are still more likely to develop atherosclerosis than other people. Since prevention is the primary treatment of coronary artery disease, controlling modifiable cardiac risk factors and lupus disease activity and carefully monitoring corticosteroid use are all helpful in reducing heart attacks in people with lupus.

Damage of the heart can develop from inflammation due to active lupus or from secondary effects of medications. Treatment of cardiac problems must be individualized for each person and for each problem. Early and accurate diagnosis, combined with aggressive therapy to reduce organ damage, is crucial in order to minimize permanent heart damage. Typical tests include a chest X-ray, EKG, echocardiogram, and blood tests to evaluate lupus activity.

Pulmonary (Lung) Involvement

Lupus can affect the lungs in many ways. It can involve the membrane lining the lung, the lung itself, the blood vessels within the lung, and even the diaphragm. Pleuritis (pleurisy) is the most common pulmonary manifestation of SLE. The pleura is a membrane that covers the outside of the lung and the inside of the chest cavity. It produces a small amount of fluid that lubricates the space between the lung and the chest wall. Inflammation of this membrane, or pleuritis, may occur in lupus. Sometimes an abnormal amount of fluid builds up in the pleural space between the lung and the chest wall. This is called a pleural effusion and occurs less often than pleuritis. If the effusion is large enough, it can be seen on a

chest X-ray. Since infection or conditions other than lupus can cause pleural effusions, the physician may need to take a sample of the fluid and perform tests to help determine its cause.

PLEURAL DISEASE

Symptoms of pleuritis include severe, often sharp, stabbing pain that may be pinpointed to a specific area or areas of the chest. The pain is made worse by taking a deep breath, coughing, sneezing, or laughing. Patients with pleuritis may also experience shortness of breath. Analgesics (pain-relieving medications), non-steroidal anti-inflammatory drugs (NSAIDs), and/or corticosteroids may be used to treat pleuritis. Pleural effusions will usually respond to these medications or clear by themselves with time.

LUPUS PNEUMONITIS

Pneumonitis is inflammation within the lung tissue. Infection is the most common cause of pneumonitis in people with lupus. Bacteria, virus, or fungi are organisms that can cause infection in the lung. Sometimes pneumonitis may occur without infection and is then called non-infectious pneumonitis. Lupus pneumonitis is an uncommon feature of SLE. Since both forms of pneumonitis have the same symptoms (e.g., fever, chest pain, shortness of breath, and cough), a person is assumed to have an infection until proven otherwise. The diagnosis of pneumonitis requires blood tests, sputum tests, and X-rays. A high resolution CT scan (HRCT) may be helpful. Bronchoscopy (a visual inspection of the inside of the lungs) and/or lung biopsy may also be necessary to determine if infection is the cause of the pneumonitis.

Treatment of pneumonitis initially includes a course of antibiotics to cover the possibility of infection. If laboratory and other diagnostic tests show no proof of infection, then the diagnosis is likely lupus pneumonitis. This non-infectious pneumonitis is treated with high doses of corticosteroids. Immunosuppressive drugs may be considered if the inflammation cannot be controlled with corticosteroids.

CHRONIC DIFFUSE INTERSTITIAL LUNG DISEASE

Chronic diffuse (widespread) interstitial lung disease is a relatively uncommon manifestation in people with SLE. Chronic interstitial lung disease causes scarring of the lung. This scarred tissue acts as a barrier to the oxygen that normally moves easily from the lung into the blood. The symptoms include: gradual onset of a chronic, dry cough; pleuritic-like chest pains; and difficulty breathing during physical activity. Besides lupus, there are other reasons for this condition. A high resolution CT and/or nuclear scan are often helpful in determining the cause. Special procedures, such as bronchoscopy, bronchoalveolar lavage, and/or lung biopsy may be required. Correct identification of the cause is necessary in order to choose the proper treatment.

Chronic interstitial lung disease may progress. This progression can be measured. Changes in pulmonary function tests, oxygen saturation and HRCT may be used to assess disease activity and response to therapy. The pulmonary function test assesses the ability of the lungs to receive, hold, and use air. The oxygen saturation test measures how readily oxygen moves through the lung and into the blood stream. Oxygen saturation is usually