

Patient Handout

Lupus occurs when the body's immune system starts attacking parts of the body it was designed to protect. This can lead to inflammation, swelling, and damage to body tissues and organs.

The lungs are one of the organs that can be involved. There are a wide range of problems that can arise in the lungs with lupus. Any part of the lung can be involved, such as the airways (tubes that carry air in and out of the body), the tissue itself, the pleura (lining around the lungs), the muscles that helps you breathe, or the blood vessels.

Screening & Diagnosis

Identifying when the lungs are involved in lupus is important. Sometimes lung issues are the first clue of a bigger issue going on throughout the whole body. Other times, lung problems arise later in the disease. Common symptoms, if present, include:

- Shortness of breath
- Cough
- Chest pain
- Reduced exercise ability

If not found early, your illness could get worse. For that reason, your doctor may recommend certain tests to be performed at your initial diagnosis, at certain time points for screening, or other times depending on your symptoms. Pulmonary function tests and pictures of the chest are the most common tests performed.

Common screening tests:

Chest CT – a test that combines an x-ray with a computer to create detailed pictures of the lungs. CT scans do not hurt. However, your child does need to be still for this test.

Pulmonary Function Tests – this test tells us how well you can get air in and out of the lungs.

Bronchoscopy – a camera passes from the mouth or nose and through the windpipe to the lungs. This allows the doctor to look at the upper airways and lungs. Bronchoscopies are performed under anesthesia.

Echocardiogram – a picture of the heart performed by ultrasound.

Six Minute Walk Test – A six-minute walk test measures the distance a person can walk over a six-minute period. The person can walk as fast or slow as they can and can rest if needed.

Based on your child's symptoms and certain patterns on these tests, a diagnosis can often be made.

Types of Lung Involvement & Management

The following are disorders that can occur in the lungs:

Pleuritic pain: The pleura is the sac surrounding the lungs. When this becomes inflamed it can be painful to breathe. Up to 60% of patients with lupus will have this type of chest pain at some point. At times, a small amount of fluid may gather outside of the lung.

This pain often responds well to medicines called NSAIDs, like ibuprofen or naproxen. If no response in several weeks with these medicines, steroids by mouth are often used.

Interstitial lung disease (ILD): ILD related to lupus often presents with slow onset of dry cough, shortness of breath, and reduced exercise ability. Pulmonary function tests (PFTs) and a chest CT are done to determine the degree of involvement and to monitor progression.

The most common finding on PFTs in this disease is a reduced ability for gas to transfer from the lungs to the red blood cells in the blood. This can lead to reduced oxygen saturations. Other findings that can be seen include lung restriction. This means that the lungs are limited in their ability to fill up or expand.

Some ILD needs treatment while other times if mild or stable symptoms, doctors may closely monitor you. If medicines are needed, often steroids like prednisone are first line therapy.

Lupus and Lung Disease (continued)

Acute Pneumonitis: Pneumonitis is a general name for lung inflammation. This is rare, and patients are often admitted to the hospital when this occurs. Symptoms with this disorder happen quickly within a few days and include fever, chest pain, shortness of breath, cough, and low oxygen levels in the blood. This is not caused by an infection like pneumonia, but infections are important to rule out if your child has these symptoms because the symptoms are similar. For this reason, you may be started on antibiotics while tests are being run. Treatment for the inflammation is often with steroids by mouth, and if needed intravenously (IV).

Pulmonary Hypertension (PH): PH occurs when there is high blood pressure in the blood vessels in your lungs. Your heart must work harder to pump blood through the lungs when this occurs, putting stress on the right side of your heart.

Symptoms can include shortness of breath, heart racing, tiredness, difficulty exercising, dizziness, or swelling in your arms and legs.

An echocardiogram is done to estimate the pressure in your lungs. At times a heart catheterization may be needed for diagnosis – this is done under anesthesia.

Pulmonary Hemorrhage: This is when there is bleeding in lungs. This is rare. Patients may present with coughing up blood, cough, or shortness of breath. Labs often show anemia, or less oxygen carrying cells in the blood.

Patients are often admitted to the hospital when this occurs. A bronchoscopy is done to make the diagnosis.

High dose steroids given through an IV and other medicines that affect the immune system are often used to stop and control bleeding.

Shrinking Lung Syndrome: This is not common but is seen in 1-5% of lupus patients. Symptoms include shortness of breath, pleuritic chest pain as described above, and a decline in lung function.

Steroids and medicines that suppress the immune system may improve symptoms and help regain lung function.

Other important notes:

Infections: Patients with lupus have a higher chance of getting certain infections. This is in part due to the medicines that are used to treat lupus. For this reason, staying up-to-date with shots, including a yearly flu vaccine, is very important. Your doctor may also recommend other shots or certain preventive medicines to help protect you.

Tobacco Smoke: Smoking can make lupus worse. Smoking cigarettes increases the risk of lung infections. This includes children who are exposed to second and thirdhand smoke. Smoking can also make medicines used to treat lupus less effective.

For this reason, we encourage patients and families to stop smoking. Quitting smoking is easier said than done. Talk to your doctor about the best way to begin this journey.

For Tennessee residents, the TN Quitline is a great resource. They provide support online or by phone – all services are confidential and free. You can call 1-800-QUIT-NOW to get started.

Ongoing monitoring:

Ongoing monitoring for these diseases with a lung doctor is essential. Tests such as PFTs and the six-minute walk test are helpful in following the progress of the disease and response to treatment. Repeat chest imaging may be needed as well.

Contact Us:

Vanderbilt Children's Pulmonary Team works closely with our Pediatric Rheumatology team to screen, diagnose, and manage kids with lupus and lung concerns.

The Rare Lung Diseases Program

Monroe Carell Jr. Children's Hospital at Vanderbilt

Phone: 615-875-6901

Fax: 615-322-7553

www.childrenshospitalvanderbilt.org