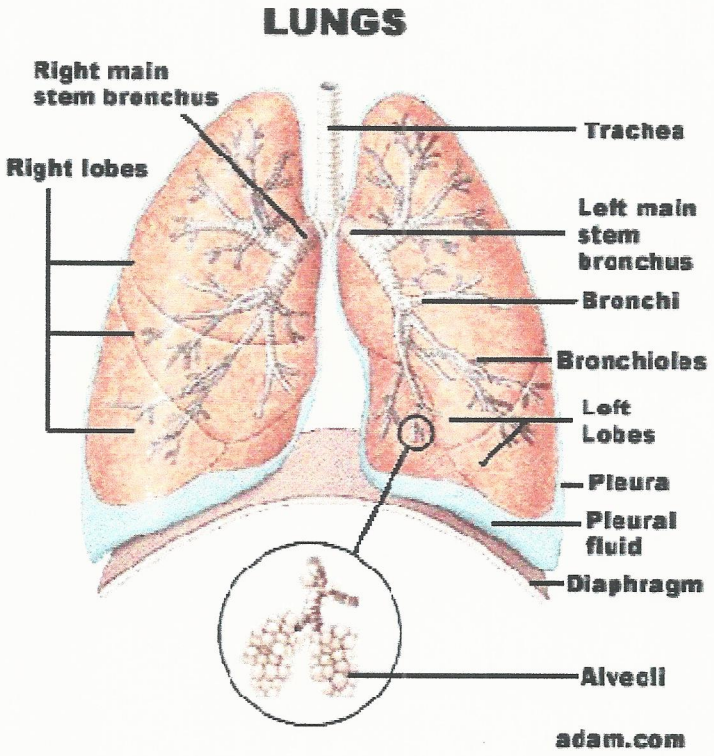
Lung Diseases Associated with Lung Transplantation

Lung transplantation is a treatment for those with end-stage lung disease. It is used with a variety of disorders. there are over 80 lung diseases that are treated with lung transplantation; however, many are extremely rare. The following four procedure are available: single lung, bilateral lung, heart-lung transplants, and transplantation of the lobes from a living-related donor. Improved survival rates are noted secondary to improved client selection, better immunosupression methods, and technical advances. It does not result in a cure and does require a lifetime of medications, monitoring, and famly support.



Alpha-1 Antitrypsin (AAT) Deficiency is an autosomal recessive (genetic) disorder that may affect the lungs and liver. AAT is a protein that normally is found in the lungs and protects the lungs from a natural enzyme (neutrophil elastase) that protects the lungs during inflammation from bacterial infections and smoking. A severe deficiency of ATT results in premature destruction of the alveoli (air sacs) which can be seen on a chest X-ray. More information can be found at: https://www.alpha1.org/what-is-alpha1/

Bronchiectasis is a chronic disease that is characterized by permanent, abnormal dilation of the medium bronchi as a result of inflammation. This damages the elastic and muscular structures supporting the bronchial wall and results in airway obstrction. The most comon causes are bacterial infections, mucus plugs (as seen in CF) and other inflammatory disease processes.

Bronchiolitis Obliterans (BOS) is inflammation of the bronchioles and airflow obstruction that progresses over time. It is a major manifestation of chronic rejection in the lung tramsplant recipient.

Chronic Obstructive Pulmonary Disease (COPD) is a disease characterized by persistent airflow limitation and chronic inflammation of the airways that is usually progressive. Inability to expire air is a major characterisitic of COPD. As the air is trapped during expiration in the alveoli, the volume of residual air is trapped and the small airways are destroyed. There are gas exchange abnormalities resulting in increased carbon dixide levels and low oxygen levels.

Chronic bronchitis is an independent disease that may preceed airflow limitations. Emphysema is the destruction of the alveoli (air sacs) and explains only one of the structureal abnormalitiesi in people with COPD.

Cystic Fibrosis (CF) is a genetic disease characterized by a defect in the transport of sodium and chloride within the epithial cells to the cells outer surfaces. CF primariy affects the lungs, gastrointestinal tract, and reporductive system. As a result of the mutation cells that line the lungs, pancrease, intestines and other organs produce secretions that are low in sodium content making mucus abnormally thick and sticky. Resulting mucus plugs cause organ scarring and eventually organ failure.The sweat chloride test is considered the gold standard for diagnosis.More informatin can be found at: https://www.cff.org/.

Eisenmenger's Syndrome is a congential heart defect which includes a ventricular septal defect (a hole between the 2 venticles), pulmonary hypertension with pulmonary artery enlargement and increased size of the right ventricle. The increased pressure of blood flowing through the shunt increases the pressure in the pulmonary artery. Over time, this increased pressure damages the smaller blood vessels in the lungs. This is treated with a combined heart-lung transplantation. More information can be found at: https://rarediseases.org/rare-diseases/eisenmenger-syndrome/.

Interstial Lung Disease (ILD) is also called diffuse parenchymal lung disease. It refers to more than 200 disorders in which the tissues between the lung's air sacs (alveoli) are inflammed. Scarring (fibrosis) can result and is not reversible. Causes can include inhalation of occupational & environmental toxins, certain medications, radiation therapy, infections, and connective tissue disease. Often, the cause is unknown.

Idiopathic Pulmonary Fibrosis (IPF) is a chronic, progressive disorder characterized by persistent inflammation and scar tissue formation in the lungs, Chest X-ray findings are often non-specific. Pulmonary function tests show restriction and poor gas exchange. It is often diagnosed by bronchoscopy and a CT scan but open lung biopsy is considered the standard for diagnosis. More information can be found at: https://ipffoundation.org/about-us/.

Lymphangioleiomyomatosis (LAM) is a lung disease where the muscle cells that normally line the airways and blood vesels invade the lung tissues and grow into the walls of the airways causing obstruction. The air sacs also swell and form cysts which impact oxygenation and gas exchange. It typically affects women and the cause is unknown. More information can be found at https://www.thelamfoundation.org/.

Primary Pulmonary Hypertension (PPH) is characterized by elevated pulmonary arterial pressure caused by constriction of the pulmonary vessels in response to low oxygen levels in the air sacs (alveoli hypoxia). Normally, the pulmonary presures are low. Pressures on the right side of the heart must increase to force blood into the lungs but the individual eventually developes symptoms of right-sided heart failure. PPH is often seen in people with severe COPD but it can be found in those with no known cause. More information can be found at: https://phassociation.org/.

Sarcoidosis is a chronic, multi-system disease wher areas of inflammation occur in different organs. Very small growths, granulomas, are associated with it and appear in the lungs, lymph nodes, eyes, skin, spleen, liver, kidney, etc. Treatment is based on the pulmonary function and disease progression. Some people will experience a spontaneous remission. More information can be found at https://www.lung.org/lung-health-diseases/lung-disease-lookup/sarcoidosis.