Pulmonary Hypertension & SJÖGREN’S SYNDROME
ABOUT PULMONARY HYPERTENSION

Pulmonary hypertension (PH), which means high blood pressure of the lungs, is a complex, often-misunderstood disease.

In regular hypertension — known as high blood pressure or systemic hypertension — the pressure is higher than it should be in arteries throughout the body. Systemic hypertension can be measured with a blood pressure cuff.

PH refers only to the blood vessels in the lungs. Normal pulmonary artery pressure averages between 8 and 20 mmHg at rest. PH, or high blood pressure in the lungs, frequently results from a narrowing of the small blood vessels in the lungs. As the vessels can become stiff and narrow, the right side of the heart enlarges and must work harder to pump blood through the vessels. The average resting pulmonary artery pressure for PH is above 20 mmHg.

There are five groups of PH based on different causes. Each form of PH is different, so it is important to find a PH specialist to accurately pinpoint the cause. Your PH specialist will develop a treatment plan for your type as soon as possible after a confirmed diagnosis.

People with PH can live many years, thanks to ongoing research advancements, PH-specific health care and customized therapy.
Sjögren’s syndrome is an autoimmune disease characterized by inflammation of the salivary, lacrimal (tear) and respiratory glands. Dry eyes, mouth and nose are the most common symptoms.

Sjögren’s is the third most common autoimmune disease. It can develop at any age but is most common in women between 30 to 50 years old. While Sjögren’s can develop on its own, it also is associated with other connective tissue diseases such systemic lupus or scleroderma.

Arthritis, rash, gastroenteritis and respiratory symptoms are commonly found among people with connective tissue disorders. Sjögren’s can affect many organs, most commonly the lungs, but also the kidney, thyroid and liver.

Nearly half of Sjögren’s patients have lung issues. About half of those with lung issues develop coughing, and 40% have shortness of breath. Based on published research, the frequency of respiratory involvement is variable.
People with Sjögren’s syndrome can develop PH, chronic cough, blood clots in the lungs, fluid around the lungs, vasculitis and other lung disorders, including interstitial lung disease.

PH rarely develops in Sjögren’s. When it does, it can result from damage to the lung tissue, as a reaction to low oxygen levels, or blood-vessel abnormalities in the lungs. Common signs of Sjögren’s-related PH include cough, shortness of breath, leg swelling, chest pains and fainting. Symptoms that suddenly arise, rather than develop over a period of time, could indicate pulmonary blood clots.

**DIAGNOSING PH**

Sjogren’s patients with respiratory symptoms should undergo pulmonary function (breathing) tests, and chest CTs and x-rays to evaluate the cause. Those with shortness of breath should be evaluated for interstitial lung disease and PH. Although interstitial lung disease (inflammation of the lung tissue) is more common than pulmonary hypertension, the two diseases can overlap.

*If your doctor suspects PH, consider consulting a specialist experienced in assessing and treating PH to develop your care plan and start your treatment. Visit PHAssociation.org/phcarecenters to find a PHA-accredited Pulmonary Hypertension Care Center.*

Pulmonary hypertension should move to the top of the possible causes for people with shortness of breath, swelling of the legs, chest pain and fainting and normal pulmonary function tests.

Sjögren’s patients with suspected PH often need blood tests, chest CTs, perfusion-ventilation scans and echocardiograms (ultrasound of the heart) to evaluate heart function and estimate blood pressure in the lungs. Blood work such as the pro-NT-BNP may hint to the presence of pulmonary hypertension (PH) as it often increases in patients with PH.
If tests show signs of PH, such as an enlarged right ventricle, fluid around the heart or elevated pulmonary artery pressure, the patient will need a right heart catheterization. A right heart catheterization can confirm PH and identify the type of PH.

Your doctor will review all of the information and determine whether your PH is secondary to interstitial lung disease or Pulmonary Arterial Hypertension (PAH). If your PH is secondary to PAH, elevated pressures in the lungs would be a result of a disorder primarily affecting small blood vessels in the lungs.

**TREATMENT**

A right heart catheterization helps determine how PH should be treated. Treatment depends on one of the many underlying causes of PH in Sjögren’s.

If Sjögren’s interstitial lung disease is the primary cause, then the primary treatment would be for the lung disorder and auto-immune disease. Treatment for underlying autoimmune disease includes immunosuppressant medications and steroids such as prednisone, azathioprine or mycophenolate, based on the patient’s individual medical issues. This form of PH may also respond to inhaled pulmonary vasodilator therapy.

Oxygen therapy is used to improve overall lung function for those with low-oxygen levels. Diuretics (water pills) can decrease swelling and fluid retention, as well as shortness of breath.
Treatment for people with PAH isn’t well-defined because of its rarity. It follows the approach used for other autoimmune diseases, such as scleroderma or lupus. It’s generally treated with multiple PAH-specific therapies, including:

- Phosphodiesterase inhibitors, such as sildenafil or tadalafil.
- Endothelin antagonists, such as bosentan, ambrisentan or macitentan.
- Oral quanylate cyclase inhibitor (riociguat)
- Prostacyclin pathway medications (treprostonil in several forms iloprost, selexipag and epoprostenol).

Treatment is customized according to patients’ medical issues, preferences and PAH severity. Some might need prescription medication to treat dry mouth and relieve other Sjögren’s-related symptoms.

**HEALTH AND WELLNESS**

All people with PH should eat a healthful diet and avoid excessive salt intake. Exercise is important to maintain strength, flexibility and conditioning. Exercise can be supervised, such as pulmonary rehabilitation, or individual practice in consultation with your care team. Avoid exercising to exhaustion or if you experience severe symptoms.
ABOUT THE PULMONARY HYPERTENSION ASSOCIATION

Headquartered in Silver Spring, Maryland, the Pulmonary Hypertension Association (PHA) is the oldest and largest nonprofit patient association in the United States dedicated to the pulmonary hypertension (PH) community. PHA’s mission is to extend and improve the lives of those affected by PH.

To achieve its mission, PHA engages people with PH and their families, caregivers, health care providers and researchers worldwide who work together to advocate for the PH community; provide support to patients, caregivers and families; offer up-to-date education and information on PH; improve quality patient care; and fund and promote research.

PHA would like to thank Victor J. Test, M.D., FCCP, Texas Tech School of Medicine and Physicians, Lubbock, Texas, in the writing, review and development of this brochure.

References