Treatment

In some patients, medication known as channel blockers have been effective. This treatment relaxes and opens up the blood vessels (vasodilate). Another medication that may be used is a diuretic, which may help to reduce fluid in the patients with the right heart insufficiency. TRACLEER® (bosentan), an endothelin-receptor antagonist, has been in approved for use in Canada and presumably works by relaxing the blood vessels, reducing the high pressure in them and enabling the heart to pump blood more effectively. If PAH progresses, treatment with FLOLAN® (epoprostenol) has been shown to be effective. This is a continuous intravenous infusion into the heart. REMODULIN® (treprostenil) which is in the same class as FLOLAN® is also approved for use in Canada and can be administered subcutaneously. Patients who fail to respond to drugs may become a candidate for a lung transplant.

The Future

Many research projects are presently being conducted into understanding the causes and mechanisms of PAH. New therapies under investigation may soon be available.

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We would like to thank **Dr. Oleg Nadashkevich** for his advice with this information pamphlet.

Your Local Scleroderma Group

Scleroderma

Pulmonary Arterial Hypertension

Pulmonary Arterial Hypertension

Scleroderma Society of Canada

What is Pulmonary Hypertension?

Pulmonary hypertension is simply elevated blood pressure in the artery that carries the blood from the heart to the lungs. The disease is progressive in nature and may mean that patients become increasingly limited. This disease affects people of all ages, ethnic background and both sexes.

Pulmonary hypertension is now referred to as idiopathic pulmonary hypertension (IPAH - formerly known as primary pulmonary hypertension), when it occurs without any relation to other diseases. It is referred to as pulmonary arterial hypertension (PAH) when it is associated with the effects of other diseases such as rheumatoid arthritis, lupus, and scleroderma.

The World Health Organization has defined a patient as having pulmonary arterial hypertension when the pulmonary arterial pressure is higher than 25mmHg at rest and 30mmHg when exercising.

Symptoms

Patients may experience the following symptoms:

- no symptoms in early stages
- first symptom is shortness of breath especially when walking or climbing stairs
- coughing, which worsens upon exertion
- constant feelings of fatigue
- shortness of breath at higher elevations
- shortness of breath even at rest at more advanced stages
- unusual chest pains
- swelling of neck veins, belly and feet
- extreme limitations in daily activities
- dizziness or fainting.

Causes

The cause of PAH is not known, but it is known that it occurs commonly in scleroderma patients with either the diffuse or limited forms. In these patients, PAH can occur with or without lung fibrosis (interstitial lung disease). In PAH patients without the lung fibrosis, a dysfunction in the regulation of blood flow results in the vasoconstriction (narrowing) of the vessels that carry blood from the right side of the heart to the lungs. Such narrowing makes it more difficult for blood to get through to the lungs so the heart has to pump harder. The extra stress on the heart causes it to enlarge and become less flexible.

Endothelin, a potent vasoconstrictor, is elevated in the blood and lung tissue of scleroderma patients.

Over time, the walls of blood vessels get thicker and stiffer and finally become so narrowed that they may become completely blocked. The consequence of poor blood flow is tissue damage.

Lung fibrosis occurs in nearly all patients with systemic scleroderma. Significant lung fibrosis in scleroderma patients can lead to PAH because the scarring in the lungs causes the vessels to narrow.

Tests

Because of the high prevalence of PAH in scleroderma patients, the World Health Organization recommends that scleroderma patients be screened **annually** for PAH.

These tests may include

- echocardiogram
- pulmonary function tests (including DLCD)
- chest x-rays
- high resolution CT scans
- bronchoscopy.

Other tests to establish a diagnosis of PAH may include

- electrocardiogram
- tests to determine the presence of anti-centromere and anti-Scl-70 (topo I) autoantibodies
- six minute walk test
- assessment of function in tasks of daily living
- Doppler echocardiogram (This test determines the size of the heart chambers and how well the valves are working, and can estimate pulmonary arterial pressures.)
- right heart catheterization, which is usually done only if results will change the patient's therapy.

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