SCLERODERMA & RAYNAUD'S UK

Pulmonary Hypertension in Systemic Sclerosis (Systemic Scleroderma)

"Pulmonary (arterial) hypertension" literally means "high blood pressure in the pulmonary artery". This is a completely different condition to the common "high blood pressure" which can affect large numbers of people. Pulmonary hypertension is often abbreviated to PH or PAH. Pulmonary hypertension is a rare lung disorder in which the blood pressure in the pulmonary artery rises above normal. This abnormally high pressure affects the small blood vessels in the lungs, resulting in less blood getting to the lungs and less oxygen being carried into the blood stream. As a result the right side of the heart has to work harder to try and compensate for the difficulty with the blood vessels in the lungs.

1 in 10 people with systemic scleroderma develop **pulmonary hypertension**

What are the symptoms of pulmonary hypertension?

- Shortness of breath with minimal exertion
- Tiredness
- Fatigue
- Dizzy spells
- Chest pain
- Swelling in ankles or legs

Symptoms of pulmonary hypertension are caused either by the low levels of oxygen in the blood stream or by the heart not working as efficiently as it should due to the extra work it is having to do.

Why have I got pulmonary hypertension?

About 1 in 10 people with systemic sclerosis (systemic scleroderma) can go on to develop pulmonary hypertension. It is thought that this is because systemic sclerosis thickens and stiffens the walls of the pulmonary artery so that it cannot open up easily to relieve the pressure inside it. The pressure inside the pulmonary artery is therefore higher than it should be resulting in the extra strain on the heart described above.

Although pulmonary hypertension occurs as a result of a person's systemic sclerosis it often does not start to develop until many years after the first symptoms of systemic sclerosis appear. When pulmonary hypertension has occurred as a result of systemic sclerosis, it is called "connective tissue disease-associated pulmonary hypertension". It is possible to get pulmonary hypertension as a result of many different conditions including lupus, blood clots in the lungs, liver disease and HIV infection, but also for no known cause.

How do we test for pulmonary hypertension?

There are many tests which may be carried out if your doctor suspects you may have pulmonary hypertension including an echocardiogram, a chest x-ray and lung function tests. However the only test which can accurately measure the pressure in your pulmonary artery and therefore definitely confirm pulmonary hypertension is a right heart catheter.
Most patients with systemic sclerosis will undergo regular testing to try and identify early on if pulmonary hypertension or any other problems are developing.

Treatments

Although at present there is no cure for pulmonary hypertension there are lots of different treatments available. These are aimed at reducing symptoms and improving quality of life.

Anti-coagulants (e.g. warfarin)
This prevents blood clots forming and allows the blood to flow more freely.

Diuretics (e.g. frusemide, spironolactone)
These decrease the amount of fluid in the body, which reduces the amount of work the heart has to do.

Oxygen
For those with low oxygen levels in the blood, this helps you to breathe more easily and should be used for approximately 16 hours a day.

Oral tablets
These can help to reduce the pressure in the pulmonary artery.

Inhaled treatments
These can help to open up the pulmonary artery thereby reducing the pressure.

Subcutaneous or intravenous infusions
These continuous infusions either just below the skin or into a vein provide a steady supply of strong medication to open up the pulmonary artery and reduce the pressure.

PROCEDURES AND OPERATIONS
There are surgical procedures which may help although not all are effective for patients who have systemic sclerosis-associated pulmonary hypertension.

All medications for the treatment of pulmonary hypertension need to be taken for the rest of the patient's life.

Your Care

Both systemic sclerosis and pulmonary hypertension are very rare conditions. To ensure access to specialist treatments it is important that patients are managed by a designated centre. There are eight designated pulmonary hypertension centres in the UK based in Newcastle, Sheffield, Cambridge and London (four hospitals).

The designated centre for systemic sclerosis-associated pulmonary hypertension or pulmonary hypertension associated with other connective tissue disorders is The Royal Free Hospital, Pond Street, London NW3 2QG. Tel: 020 7472 6354.

THE FUTURE
The range of options for treating pulmonary hypertension has expanded in the last few years resulting in a much improved outlook for those with systemic sclerosis-associated pulmonary hypertension. Trials into new treatments are ongoing constantly and will hopefully produce even more effective treatments.

PULMONARY HYPERTENSION SPECIALIST CENTRES IN THE UK AND IRELAND

Western Infirmary, Glasgow
Tel: 0141 211 1836
Freeemans Hospital, Newcastle
Tel: 0191 233 6161
Royal Hallamshire Hospital, Sheffield
Tel: 0114 271 1719
Papworth Hospital, Cambridgeshire
Tel: 01480 830541

Mater Miserlcordiae Hospital, Republic of Ireland
Tel: 00 3531 8032000

Great Ormond Street Hospital, London
Tel: 020 7405 9200 (ext 1005 / 1007)
Hammersmith Hospital, London
Tel: 020 8383 2330
Royal Brompton Hospital, London
Tel: 020 7351 8121
Royal Free Hospital, London
Tel: 020 7794 0500

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