SCLERODERMA & RAYNAUD'S UK

The Heart in Systemic Sclerosis (Systemic Scleroderma)

Heart disease becomes more common as we get older and systemic sclerosis (systemic scleroderma), usually starts in the 50s and 60s so it is not surprising that heart disease is commonly found in people with systemic sclerosis. Systemic sclerosis can also directly affect the heart, so people with systemic sclerosis are at greater risk of heart disease than others.

For most people with systemic sclerosis, any involvement of the heart is so mild that it is very difficult to be sure whether it is present or not. Very minor involvement of the heart causes an inability to reach high levels of exercise, but because of lung disease and musculoskeletal problems, reaching very high levels of exercise is usually not possible in people with systemic sclerosis anyway.

When heart disease is severe then symptoms are the same as for everyone else, with breathlessness, chest discomfort, swollen ankles and palpitations. The same treatments we use in everyone else with heart disease also work in heart disease in people with systemic sclerosis.

Types of heart disease in systemic sclerosis

Ordinary heart disease
Coronary artery disease, due to fat being laid down in the walls of the vessels that supply the heart with blood, is the most common single cause of death in the western world, even in women. Systemic sclerosis provides no protection against coronary artery disease, and may slightly increase the likelihood of developing this condition. Coronary disease causes angina (chest discomfort on exercise), heart attacks (severe prolonged chest discomfort associated with heart muscle death) and heart failure (breathlessness and swelling).

Valvular heart disease, especially aortic stenosis (narrowing of the main valve that controls blood flow leaving the heart for the rest of the body), which is common in elderly people, appears to be just as common in systemic sclerosis as every one else. Symptoms are similar to coronary disease (left) although blackouts are more likely.

Hypertensive heart disease occurs when the work of the heart is increased to deal with high pressure. The muscle of the heart is thickened which can lead to rhythm problems (palpitations) and heart failure. High blood pressure affects nearly 50% of middle aged people and is also increased in systemic sclerosis because of kidney involvement, which in turn causes increased blood pressure (the kidney is very important in setting blood pressure levels in the body).

Small vessel disease
Coronary artery disease affects the large vessels on the surface of the heart; in systemic sclerosis, the tiny vessels within the heart muscle also show abnormalities being more prone to spasm and failure to relax properly. This can cause very tiny areas of damage widely spread throughout the heart, which we see as slightly reduced power of the heart and slowed relaxation.

Rhythm problems

Atrial fibrillation (a completely irregular heart rhythm) is one of the commonest rhythm disorders of elderly people; it is much more common if high blood pressure, coronary artery disease or heart failure is present. Atrial fibrillation tends to develop where there is scarring (fibrosis) of the heart, and systemic sclerosis is strongly associated with scarring (fibrosis) of organs. It is not however clear that atrial fibrillation is much more common than expected in systemic sclerosis. Other causes of palpitations, such as extra beats, do seem to be frequent in systemic sclerosis.

Myocarditis

Inflammation can occasionally be a marked feature of systemic sclerosis, and rarely this can occur in the heart. Patients with myocarditis have symptoms of heart failure and rhythm problems; we can now identify this fairly easily with blood tests and scans and treat it with immunosupression.

Systemic sclerosis heart disease

The most common type of heart disease directly caused by systemic sclerosis is increased strain of the right heart due to pulmonary hypertension. Around one in ten patients with systemic sclerosis have high blood pressure in the lung arteries. This increases the work of the right heart, causing it to thicken and eventually fail. For more details about pulmonary hypertension please see the leaflet on pulmonary hypertension.

Systemic sclerosis also causes pericardial effusions (collection of fluid in the sac that the heart sits in). This rarely causes any problems. It can be due to inflammation of the lining of the pericardial sac or increased pressure within the heart (for example when the heart is responding to high blood pressure in the lungs).

Cardiac fibrosis or scarring of the heart muscle can be found in around one in five people with systemic sclerosis. Usually this is just something noticed on scans, but rarely can become very severe and can make the heart unable to relax and can cause heart failure.

INVESTIGATING HEART DISEASE IN SYSTEMIC SCLEROSIS

Investigations can either take the form of risk assessment, screening or diagnostic. Risk assessment is now very common in heart disease: cholesterol assessment, smoking history, presence of diabetes, high blood pressure and family history allows us to predict the likelihood of any individual developing coronary disease, and to start treatment before the disease occurs to prevent problems in the future. Antibody profiling also helps us to identify those at increased risk of pulmonary hypertension, which we can then treat early to prevent progression to heart disease.

Screening is undertaken to identify heart disease before symptoms develop. ECGs (an electrical tracing of the heart done with some sticky pads attached to the chest and limbs) can show rhythm problems, evidence of thickened muscle or scarring, slowing the contraction of the heart. This is a very simple test which should be done every year and compared to previous years to see where changes occur. An echocardiogram (an ultrasound of the heart) is a more sophisticated test of the heart, but provides different information. It is used to assess the contraction and relaxation of the heart, the function of the heart valves, the thickness of the heart muscle and the pressures the heart is facing. We recommend that this test is performed every year and that particular attention is paid to the right heart during the test.

Diagnostic tests are used when symptoms suggesting heart disease are present to find out the actual cause or exclude heart disease as the cause. These range from simple tests like the ECG and ECHO above, through blood tests to very specialised tests like cardiac catheterisation (passing tubes from the leg to the heart) and magnetic resonance scanning (a very powerful magnetic scanner to look at the tissue of the heart). Patients with systemic sclerosis should undergo very detailed investigation where any symptoms of heart disease exist (breathlessness, chest discomfort, altered consciousness, palpitations, ankle swelling) because heart disease is common in systemic sclerosis and may be difficult to diagnose.

The only special precautions required when investigating heart disease are to avoid doing catheters of the heart using the wrist artery (as these are smaller and more prone to spasm and injury in patients with systemic sclerosis) and to use contrast (chemicals used in catheter tests and MRI scanning) carefully if the kidneys have been affected by systemic sclerosis.

Treating heart disease in systemic sclerosis

Heart disease is poorly tolerated in systemic sclerosis, however, because systemic sclerosis affects most organs, doctors are often over-cautious about recommending sufficient treatment for the heart. The correct balance is to recognise that symptoms are not necessarily due to heart disease and the risks of operation are higher, so more thorough investigation is required, however, the benefits of correct treatment are greater, so treatment where appropriate should be comprehensive.

There is no reason in people with systemic sclerosis not to do heart operations including bypass grafting or valve replacement where this is appropriate, just that the team performing the operation should take greater care to fully understand the effect of lung, kidney and gut involvement on the recovery from the operation.

Medical treatments for heart disease are no different in systemic sclerosis to other patients with heart disease, with some exceptions. Beta-blockers should be used very cautiously as they can aggravate Raynaud's. Anticoagulants and antiplatelet therapies can usually be used so long as gut bleeding is not a problem. Statins can be used to control cholesterol, but for patients with muscle involvement (myositis) it is important to keep an extra close eye on the muscle enzyme levels. A number of drugs are cleared from the body by the kidneys (e.g. digoxin), so such agents should be used carefully in patients who have kidney involvement. Most drugs can upset the gut, so being aware of any significant change in gut symptoms in the weeks after starting a new treatment is particularly helpful.

As a general principle, heart disease should be actively sought and managed in people with systemic sclerosis, but it should be managed in co-operation with someone who fully understands systemic sclerosis; in general this requires that your cardiologist and rheumatologist work together.

Bride House
18-20 Bride Lane
London
EC4Y 8EE
T: 020 7000 1925
E: info@sruk.co.uk

112 Crewe Road
Alsager
Cheshire
ST7 2JA
T: 01270 872776
E: info@sruk.co.uk

Helpline: 0800 311 2756

www.sruk.co.uk

@WeAreSRUK **/WeAreSRUK**

Registered Charity England and Wales No 1161828

Reprinted March 2016