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Systemic sclerosis

Systemic sclerosis is a type of scleroderma, which means hard skin. If you have scleroderma, you have areas of hardening of the skin. In systemic sclerosis, you can also get hardening of some of your internal organs. This stops them working normally.

There are many possible symptoms and problems that may develop with systemic sclerosis because the extent of the disease can vary greatly from person to person. People with systemic sclerosis should have regular monitoring of blood pressure, blood and urine tests, and lung function, and should report any unexplained symptoms promptly to a doctor. Systemic sclerosis cannot be cured but there are various treatments which can help. Newer treatments have improved the outlook for this condition.

What is scleroderma?

The word scleroderma means hard skin. There are two kinds of scleroderma:

- **Localised scleroderma (also known as morphoea)**. This only affects the skin. It causes some areas of the skin to become harder than usual. This type of scleroderma is more common in children. See the separate leaflet called Localised Scleroderma (Morphoea) for more details.
- **Systemic sclerosis**. This causes some areas of the skin to become harder than usual and it may involve one or more of the body's internal organs. There are two main forms of systemic sclerosis:
    - Limited cutaneous systemic sclerosis (lcSSc).
    - Diffuse cutaneous systemic sclerosis (dcSSc).

The rest of this leaflet is only about systemic sclerosis.

What is systemic sclerosis?

Systemic sclerosis is an autoimmune disease. This means that the immune system causes damage to some of the body's own cells. Normally, our body makes proteins called antibodies to fight infections - for example, when we catch a cold or have a sore throat. These antibodies help to kill the germs causing the infection. In autoimmune diseases the body makes similar antibodies (autoantibodies) that attack its normal cells. In systemic sclerosis, the immune system damages the connective tissue cells, which are found in many parts of the body. Your connective tissue supports, binds or separates other tissues and organs within your body and can also be a kind of packing tissue in your body. So, systemic sclerosis is also classed as a connective tissue disease.

What causes systemic sclerosis?

It is not known what triggers the immune system to cause systemic sclerosis. There is probably an inherited tendency to develop it. Systemic sclerosis is more common in people who have a family member with systemic sclerosis. Systemic sclerosis may be triggered by certain germs (such as viruses), medicines, street drugs and chemicals. A number of different influences seem to be involved.

What is known is that cells called fibroblasts are affected by the immune system in some way and make too much of a protein called collagen. The collagen gets deposited in skin, blood vessels and organs. This causes scarring and thickening (fibrosis). Blood vessels are also damaged.

Who develops systemic sclerosis?

Systemic sclerosis is very uncommon. It affects about 3-9 of every 100,000 people in the UK. Women are four or five times more likely than men to develop it. It is more common in North America and Australia than it is in Europe.

Systemic sclerosis usually begins between the ages of 40-50 years but can affect any age group. It is rare in children.

What are the symptoms of systemic sclerosis?

There are lots of possible symptoms of systemic sclerosis. Which ones are present will vary between different people.

Raynaud's phenomenon

The earliest symptom of systemic sclerosis is a problem called Raynaud's phenomenon. In this condition there are temporary changes to the skin in response to cold or stress - usually in the fingers, toes and nose. The skin turns very pale, blue and then red. It returns to normal when warm or no longer stressed. This is caused by a narrowing of the small blood vessels in situations where there is cold or stress. **Note**: if you have Raynaud's phenomenon it does NOT necessarily mean that you will develop systemic sclerosis. Raynaud's phenomenon on its own is a common condition and most people who have Raynaud's phenomenon will not develop systemic sclerosis. See the separate leaflet called Raynaud's Phenomenon for details.

Other skin symptoms of systemic sclerosis

- Patches of hard or thickened skin. In limited cutaneous systemic sclerosis (lcSSc) this affects the face, forearms and lower legs up to the knee. It tends to start and progress slowly. In diffuse cutaneous systemic sclerosis (dcSSc) skin changes also can involve the upper arms, thighs or trunk. This usually comes on quickly, over a few months, but tends to settle and improve after 3-5 years. If this affects your mouth it can be difficult to open it widely.
- Swelling of fingers and toes - a common early sign. Fingers may look sausage-like; hand movement may be painful or difficult.
- Ulcers occurring on fingertips.
- Chalky hard swellings (called calcinosis) forming in the skin.
- Dry or itchy skin, with less hair growth.

General symptoms

- Feeling tired.
- Weight loss.

Muscle and joint symptoms

- Muscle pains.
- Joint pain and swelling.
- Difficulty in moving joints because of the tightness of the skin.

Gut symptoms

- Symptoms in the gullet (oesophagus), such as heartburn (acid reflux). Also, the muscles in the gullet may work less well, causing difficulty in swallowing lumpy foods.
- Feeling full after meals because the stomach does not empty properly.
- Bloating of the tummy.
- Diarrhoea or constipation.

Lung symptoms

- Scarring of the lung tissue, called interstitial fibrosis. This can give you a cough or make you feel breathless as the lungs can't work as well as usual.
- High blood pressure in the arteries of the lungs, called pulmonary arterial hypertension. This can make you feel faint, dizzy or breathless. It can be very serious.

How is systemic sclerosis diagnosed?

Systemic sclerosis can be difficult to diagnose as it can develop gradually and can take different forms.

There is no single test or feature which gives a yes/no answer to systemic sclerosis. Instead, the diagnosis is made after looking at the whole person, taking into account their symptoms, physical examination and blood tests. This may involve seeing a specialist - usually a rheumatologist, who is a doctor specialising in joint and connective tissue diseases.

Tests which can be helpful are:

- Examination of the fingernails, using a skin microscope: this looks at the tiny blood vessels (capillaries) in the nail area, which can show changes linked to systemic sclerosis.
- Blood tests.
- Blood can be tested for particular autoantibodies which are linked to systemic sclerosis.
- An X-ray of the hand may show chalk deposits (calcinosis).

Are there any complications of systemic sclerosis?

Systemic sclerosis can cause complications and may affect almost any organ in the body. This happens because it is a connective tissue disease, and connective tissue is found throughout the body. The condition affects different people in different ways. Complications can vary from very mild with no symptoms, to more severe.

The most common complications of systemic sclerosis are:

- Skin may become tight (contracted), or may develop ulcers or nodules.
- Problems with teeth if there is difficulty opening the mouth enough to brush them properly.
- Reduced saliva production can cause dryness of the mouth.
- Bleeding from the gut.
- Blockage (obstruction) of the bowels.
- Incontinence of the bowels.
- Erection problems (impotence) in men.
- Heart complications. Several different problems can occur if the muscle of the heart is affected by scar tissue.
- Lung problems. There are two types of lung problems which may develop, mentioned in the section about symptoms.
- The thyroid gland can become underactive.

- Depression.
- 'Thinning' of the bones (osteoporosis).
- Blood pressure can become high.
- Kidneys may become less efficient.
- Sometimes, kidney problems worsen quickly and the blood pressure becomes very high. This is called scleroderma renal crisis and accelerated hypertension. It needs urgent treatment. Symptoms are headaches, blurred vision, seizures, breathlessness, leg and foot swelling, or reduced urine production. If you have any of these symptoms see a doctor immediately.

What is the treatment for systemic sclerosis?

At present, there is no cure for systemic sclerosis. However, much can be done to help. The aims of treatment are:

- To relieve symptoms.
- To prevent the condition from progressing, as much as possible.
- To detect and treat complications early.
- To minimise any disability.

Treatment aim 1 - to relieve symptoms of systemic sclerosis

- For the skin, moisturisers and stretching exercises help with dry or tight skin.

- Raynaud's phenomenon symptoms are reduced by keeping the hands warm - for example, by using heated gloves. Medicines which open up the arteries to bring more blood to the fingers can be used to improve the symptoms. In the UK currently nifedipine is the only pill licensed for this. However, studies show other medicines can be effective, and some are used in the UK and elsewhere. These are medicines usually used for other conditions. They cause the blood vessels to widen (dilate) and this helps get blood to your fingers if you have Raynaud's phenomenon. These include:
    - Losartan.
    - Sildenafil.
    - A type of antidepressant medication called selective serotonin reuptake inhibitors (SSRIs).
    - Iloprost.
- Stomach and gut problems can be relieved by medicines such as:
    - Omeprazole to reduce acid secretion.
    - Domperidone to help with stomach action.
    - Laxatives for constipation.
    - Medicines which help treat diarrhoea, such as loperamide.
- If swallowing lumpy foods is difficult then it may help to have lots to drink with meals. Surgery may be required in difficult cases, particularly if partial blockage or bowel incontinence develops.

There are various other treatments which can help, depending on individual symptoms.

Treatment aim 2 - to prevent systemic sclerosis from progressing, as much as possible

Treatments are used which suppress the body's immune system (because it is the immune system which is overactive in systemic sclerosis). Examples of these treatments are steroids, methotrexate, cyclophosphamide, azathioprine, and mycophenolate mofetil. The medicines that are used will depend on the individual situation.

You should not smoke - because this is healthier for the blood vessels and lungs.

Treatment aim 3 - to detect and treat systemic sclerosis complications early

Regular monitoring is needed to check blood pressure, kidney function (with blood and urine tests) and lungs (which may need a scan). These tests may detect changes before any symptoms are noticed - and early detection allows early treatment.

Various treatments are available for different complications. These include:

- Skin: dressings and antibiotic medicines are used for skin ulcers. Surgery may help with tight skin, nodules or ulcers.
- Lungs: new medicines such as bosentan, sildenafil, ambrisentan, iloprost and epoprostenol have improved the treatment of pulmonary hypertension.
- High blood pressure and kidney problems are treated with angiotensin-converting enzyme (ACE) inhibitors.
- Thyroid replacement tablets are given for an underactive thyroid gland.

- Tablets such as sildenafil or tadalafil for problems with erections (impotence). These may need to be taken regularly rather than as needed.
- You may be more susceptible to infections, and so should receive antibiotics promptly for any infective illness such as a chest infection.

Treatment aim 4 - to minimise disability

- If required, physiotherapists can advise on exercises to keep joints mobile and muscles strong.
- If required, occupational therapists can advise on various aids (such as splints to support the joints) and can help with daily living tasks.
- Regular dental checks are important if you have dry mouth symptoms.

- Patient support groups, such as Scleroderma and Raynaud's UK (SRUK) can provide information and support.
- Patient education/self-management programmes can help people to understand their condition and be more in control of their lives. For example, the Bath Scleroderma Education Programme run by the Royal National Hospital for Rheumatic Diseases in Bath, or the Expert Patient Programmes run by local health organisations.
- You may be entitled to benefits if your daily activities are affected, or if you require extra help.

How does systemic sclerosis affect pregnancy?

It is possible to have a baby if you have systemic sclerosis. You should try to plan your pregnancy at a time when the disease is stable. Nevertheless, the pregnancy would be classed as a high-risk pregnancy because there is an increased risk of pregnancy complications compared with women who do not have systemic sclerosis. Specialised care from your rheumatologist and obstetrician is advisable. A rheumatologist specialises in joint and connective tissue diseases and an obstetrician in pregnancy and childbirth.

If you are planning a pregnancy, it is important to discuss your medication with a doctor **before** you start trying for a baby - because some of the medicines used to treat systemic sclerosis are not suitable during pregnancy.

What is the outlook) for people with systemic sclerosis?

For most people with systemic sclerosis, the disease is either mild or moderate. For a small number (minority), the disease can be severe or life-threatening. The outlook depends very much on which form of the disease you have and on how it progresses. It is usually possible to make some prediction for each individual; this would need to be done by a specialist.

As a general rule, people with the limited skin (cutaneous) type of systemic sclerosis have a milder form of the condition which progresses slowly. People with the diffuse cutaneous type have a faster onset of the condition and its complications, but then it will often stabilise and the skin often improves with time. Internal organs can be affected in either type. The amount of skin involvement does not correspond with the amount of organ damage.

Although no cure has been found yet for systemic sclerosis, treatments have advanced in recent years and the outlook has improved. Research in this area is continuing.

Further reading

- Systemic Sclerosis; DermNet NZ
- BSR and BHPR guideline for the treatment of systemic sclerosis; British Society for Rheumatology (2016)
- Nemeth A, Szamosi S, Horvath A, et al; Systemic sclerosis and pregnancy. A review of the current literature. Z Rheumatol. 2014 Mar;73(2):175-9. doi: 10.1007/s00393-013-1267-x.
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