Raynaud’s syndrome, scleroderma and associated disorders

The aim of this information sheet is to help answer some of the questions you may have about Raynaud’s syndrome, scleroderma and associated disorders. If you have any further questions or concerns, please do not hesitate to speak to a doctor or nurse caring for you.

What is Raynaud’s syndrome?
Raynaud’s syndrome is a common condition where the blood supply to the extremities is interrupted or reduced. This usually affects the fingers and toes, but occasionally the nose or ears. Attacks are usually provoked by cold or a sudden change in temperature. During an attack the affected part first becomes white, then turns blue as the tissues use up the oxygen and finally bright red as the arteries relax and fresh blood rushes in.

Raynaud’s can vary in form, from very mild to severe and requiring treatment. Anyone of any age can suffer from Raynaud’s but younger women are affected more commonly. It seems to be a change in temperature rather than just cold exposure that precipitates an attack, so although Raynaud’s is worse in winter, attacks can occur in summer.

Stress or anxiety can also provoke a Raynaud’s attack. Some cases of Raynaud’s are associated with some other diseases, then it is called ‘secondary Raynaud’s’.

What is scleroderma?
Although over 95% of patients with scleroderma have Raynaud’s, the chances of someone with Raynaud’s developing scleroderma are small – less than 2% in women and 6% in men.

The word scleroderma means ‘hardening of the skin’, although the condition is not limited to the skin. It is a disease of the connective tissue, which is the tissue that holds our bodies together. Therefore the internal organs can also be affected. The majority of sufferers have the mild form where there is limited skin involvement, usually of the hands and feet, with the skin here becoming stiff and shiny. The gullet (oesophagus) may also be affected, making eating and swallowing difficult. In some people tiny deposits of calcium can form under the skin, called ‘calcinosis’, which can cause ulcers (sores). In its more severe form, called ‘diffuse scleroderma’, wide areas of skin and internal organs such as the lungs, bowel, heart and kidneys are affected.
Localised scleroderma (the milder form) can be divided into two types:

- Morphea scleroderma – this only affects the skin and there is no spread internally. The changes to the skin may last for many years before they resolve.
- Linear scleroderma – these band-like lesions may cause skin disfigurement and may affect the tissue and joint underneath the lesion.

**How are these conditions diagnosed?**
Your medical history is the most important consideration. Blood tests may help, as can examining the small blood vessels at the base of the nail – this test is called a ‘nail fold capillaroscopy’.

**Are they hereditary?**
There is no evidence at present that either Raynaud’s or scleroderma are directly inherited. There is however a genetic predisposition, so that the chances of being affected are greater if a relative has the problem.

**How are they treated?**
Your GP or specialist may prescribe a vasodilator, which is a drug that relaxes the blood vessels. Occasionally, your specialist may feel an operation called a ‘sympathectomy’ may be of benefit. This involves either cutting or destroying the nerves that cause the arteries to constrict. This operation is more successful for Raynaud’s of the feet, however it is not recommended for the majority of people with Raynaud’s, as it does not usually produce longer-term benefits and there is the possibility that the Raynaud’s could return.

People who develop Raynaud’s as teenagers often have a form that is benign (harmless and not associated with any other condition) and will disappear with age. Unfortunately, this is not true in all cases and sometimes this form of Raynaud’s does persist.

There is no cure for scleroderma at present but there are many effective treatments available to alleviate specific symptoms. As everyone is different and your Raynaud’s will be individual to you, you should discuss these issues with your doctor.

**How can I help myself?**
There are several things you can do which may help. The most important are to stop smoking, take regular exercise and keep warm.

**Smoking**
If you are a smoker you must make a sincere and determined effort to give up completely. Tobacco is harmful as it causes the blood vessels to constrict, decreasing the blood flow to the fingertips. Your GP should be able to discuss strategies for you to give up smoking or arrange for you to see a smoking cessation counsello. Nicotine replacement may also help and you should discuss this with your doctor or pharmacist. If you would like to give up smoking, please speak to your nurse or call the Trust stop smoking service on **020 7188 0995**, or call the NHS Smoking Helpline on **0300 123 1044**.
**Eating for warmth**
Eating and drinking can help you keep warm. Try to eat lots of small meals to maintain your energy. High protein foods, milk, meat, fish and fresh vegetables are best. Hot meals and plenty of hot drinks are essential.

**Exercise**
Gentle exercise will help your circulation. Try to avoid sitting for long periods. Get up and walk around the room, moving arms and legs to maintain the circulation. Do not, however, let your fingers or toes get cold. In cold weather take exercise indoors.

**Clothing**
Tight clothing should be avoided as this may restrict blood flow. Hands and feet should always be adequately covered. A scarf should be used to keep the face warm in cold weather, and a hat and several layers of clothing should be used to keep the head and body warm. Feet are especially prone to cooling; therefore a good thick pair of socks is essential. Wet shoes and clothes should be changed as soon as possible.

**Associated conditions**

**Vibration white finger (VWF)**
Those who work with vibrating tools have a tendency to develop Raynaud’s, especially if the vibration is coarse and low of frequency. This can become permanent even after the work has stopped. VWF is an industrial disease, and you may be eligible for compensation.

**Chilblains**
These usually appear on the extremities – fingers, toes and ears. The skin may first become itchy, then red, swollen and very tender to the touch. Chilblains occur as a result of defective circulation on exposure to cold. Clothing that rubs should be avoided.

**Rheumatoid arthritis**
Arthritis affects the lining of the joints. This lining produces a fluid that lubricates the joint, and when affected by rheumatoid arthritis it becomes inflamed. More fluid is produced causing a red, painful, swollen joint. About 10% of people with rheumatoid arthritis have secondary Raynaud’s.

**Systemic lupus erythematosus (SLE)**
This is characterised by a rash sometimes seen on both cheeks and the bridge of the nose, and chronic inflammation of the blood vessels and connective tissues of the body. There is associated tiredness, joint pain, mouth ulcers, hair loss and Raynaud’s.

**Erythromyalgia**
This is a chronic disorder characterised by persistent warmth, pain and redness, mainly affecting the feet and lower legs. The majority of people with erythromyalgia also experience Raynaud’s symptoms.

**Chemical or drug induced**
Some chemicals at work (such as vinyl chloride) or medicines such as beta blockers, migraine tablets or the oral contraceptive pill may aggravate Raynaud’s. If you are prescribed any medicines and you experience Raynaud’s-type symptoms, check with your GP who may be able to alter your medication.
Useful sources of information

Contact us
If you have any questions or concerns before or after you have left hospital, please contact the vascular specialist nurses, t: 07825 503902 (Monday to Friday, 8am–4pm).

You can also call Luke Ward, t: 020 7188 3566 or Sarah Swift Ward, t: 020 718 8842 (24 hours) and speak to the ward sister or nurse in charge.

The above contacts can put you in touch with a vascular consultant if you require this.

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit www.guysandstthomas.nhs.uk/leaflets

Pharmacy Medicines Helpline
If you have any questions or concerns about your medicines, please speak to the staff caring for you or call our helpline.
t: 020 7188 8748 9am to 5pm, Monday to Friday

Your comments and concerns
For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS). To make a complaint, contact the complaints department.
t: 020 7188 8801 (PALS)  e: pals@gstt.nhs.uk
t: 020 7188 3514 (complaints)  e: complaints2@gstt.nhs.uk

Language and accessible support services
If you need an interpreter or information about your care in a different language or format, please get in touch.
t: 020 7188 8815  e: languagesupport@gstt.nhs.uk

NHS 111
Offers medical help and advice from fully trained advisers supported by experienced nurses and paramedics. Available over the phone 24 hours a day.
t: 111

NHS website
Provides online information and guidance on all aspects of health and healthcare, to help you make choices about your health.
w: www.nhs.uk

Get involved and have your say: become a member of the Trust
Members of Guy’s and St Thomas’ NHS Foundation Trust contribute to the organisation on a voluntary basis. We count on them for feedback, local knowledge and support. Membership is free and it is up to you how much you get involved. To find out more, please get in touch.
t: 0800 731 0319  e: members@gstt.nhs.uk  w: www.guysandstthomas.nhs.uk/membership