Raynaud’s syndrome, scleroderma & 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 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 worse in winter, it 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 (called secondary Raynaud’s).

What is scleroderma?
Although over 95% of patients with scleroderma have Raynauds, the chances of someone with Raynauds developing scleroderma are small – it is 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, not only the skin can be affected, but also internal organs. The majority of sufferers have the mild form where there is limited skin involvement, usually of the hands and feet, becoming stiff and shiny. The gullet may also be affected making eating and swallowing difficult. Some patients also form tiny deposits of calcium under the skin, called calcinosis, which can cause ulcers. In 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 can be divided into two types:
- Morphea scleroderma – this 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?
The history of the disease is most important. Blood tests may help, as can examining the small blood vessels at the base of the nail, which is called nail fold capillaroscopy.

Is it 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 cases as it does not usually produce longer term benefits.

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

There is no cure for scleroderma at present but there are many effective treatments available to alleviate specific symptoms. As each case is different 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 is to stop smoking, take regular exercise and keep warm.

Smoking
If you area 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 counsellor. Nicotine replacement may also help and you should discuss this with your doctor or pharmacist. You can contact the free NHS smoking helpline on 0800 022 4 322 or the Knowledge & Information Centre (KIC) (details below) who will give you details of our support services.

Eating for warmth
Eating and drinking can help you keep warm. Try to eats 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, which 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 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 and swollen. More fluid is produced causing a red, painful swollen joint. About 10% of rheumatoid arthritis sufferers have secondary Raynaud’s.

Systemic Lupus Erythematosi
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 sufferers also experience Raynaud’s symptoms.

Chemical or drug induced
Some chemicals at work (vinyl chloride) or drugs such as Beta Blockers, migraine tablets or the oral contraceptive may aggravate Raynaud’s, therefore 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.

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

You can also contact Luke ward on 020 7188 3566 or Evan Jones ward 020 7188 2262 (24 hours) and speak to the ward sister or nurse in charge.

The above contacts can put you in touch with the following vascular consultants should you wish to do so: Miss Rachel Bell, Mr Stephen Black, Mr Tom Carrell, Mr Michael Dialynas, Mr Tommaso Donati, Mr Bijan Modarai, Mr Morad Sallam, Mr Mark Tyrell, Mr Hany Zayed, Mr Said Abisi, Mr Andrew McIrvine.

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

Patient Advice and Liaison Service (PALS)
To make comments or raise concerns about the Trust’s services, please contact PALS. Ask a member of staff to direct you to the PALS office or:
e: 020 7188 8801 at St Thomas’  t: 020 7188 8803 at Guy’s  e: pals@gstt.nhs.uk

Language Support Services
If you need an interpreter or information about your care in a different language or format, please get in touch using the following contact details.
t: 020 7188 8815  fax: 020 7188 5953

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

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

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