Patient Advice and Information

Vasoconstrictive Disorders (Raynaud's, Scleroderma, Acrocyanosis & Associated Conditions)

1. What is Raynaud's phenomenon?

The circulation of the extremities [especially the skin] can be **regulated by temperature**. Thus, exposure to cold, in a normal person, will cause **vasoconstriction** [spasm of the small skin blood vessels] and a decrease in blood flow to the skin. This is caused by intense activity of the nerves that supply skin and its blood vessels. In normal people the recovery time when temperature returns to normal levels is quick [2min]; however, in some there is either an **over-response to cold** or [more likely] **failure to recover from a cold stimulus**. The incidence of such an abnormal response to cold may reach up to 20% of young women in the whole population.

- **Raynaud's phenomenon** is a condition where the blood supply to the extremities, usually the fingers and the toes but occasionally the nose and the ears, is interrupted. This is usually provoked by exposure to cold. During an attack the affected part becomes **white and dead looking**, then turns **blue** as the tissues use up the oxygen and **finally bright red** as the arteries relax and fresh blood rushes in. These changes in colour may be accompanied by paraesthesia and other sensations but pain is not a prominent feature.

- **Primary Raynaud** [when it is not associated with other diseases] can vary from a mild form, being little more than a nuisance, to a severe form requiring treatment. Anyone of any age can suffer from Raynaud's, but **teenage women** are affected more commonly. Certainly 90% of the patients are women and there is often a family history. The symptoms may decline in severity after the menopause. It seems to be an abrupt change in temperature rather than just cold exposure that precipitates an attack, so although worse in winter, it can occur in summer. Stress, anxiety or emotional stimuli may also provoke an attack.

- **Secondary Raynaud's** is the same condition but this time it is associated with some other diseases [i.e. scleroderma, rheumatoid arthritis] or external influence [i.e. drugs like oral contraceptives].

Conditions with which Raynaud's phenomenon may be associated

- **Connective tissue disorders**

{scleroderma, systemic lupus, rheumatoid arthritis etc.}

- **Drugs**

{ergotamine, b-blockers, cytotoxic agents, oral contraceptives}

- **Obstructive arterial disease**

{thoracic outlet syndrome, thromboangitis obliterans}

- **Occupational hazards**

{vibration, cold}

- **Miscellaneous**

{arteritis, arterial trauma, endocrinal disorders, hypothyroidism, neurological disorders like multiple sclerosis, neoplasia, etc.}

2. What is Scleroderma?

The word scleroderma means hardening of the skin. It is a **disease of the connective tissue,** which, as the name implies, holds our body together. Therefore not only the skin, but the internal organs can be also affected. The majority of sufferers have a mild form of the disease where there is **limited skin involvement**, usually of the hands and feet, becoming **stiff and shiny**. The **gullet often becomes affected** making eating and swallowing difficult. Some patients also form tiny deposits of calcium under the skin [calcinosis], which can cause ulceration.

In the more severe form called **diffuse scleroderma**, wide areas of the skin and the internal organ such as the lungs, bowel, heart and kidneys are affected.

Localized scleroderma may be divided into two types: morphea and linear scleroderma. **Morphea** is the name given to localized patches of hardening of the skin. **Linear scleroderma** develops in childhood and may affect the growth of a limb and is usually limited to one area. Unlike morphea, linear scleroderma tends to involve deeper layers of the skin and can affect the mobility of the underlying joints.

About 90% of patients with scleroderma will also suffer from Raynaud's phenomenon but only in 1% of these the symptoms are severe. Between 5% and 15% of patients with Raynaud's phenomenon will sometime develop overt scleroderma.

3. Which are the other vasoconstrictive disorders?

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

- **Chiblains.** Patients with chiblains complain bitterly of the cold. As a result of defective skin circulation on exposure to cold, the skin may become first itchy, then red, swollen and very tender to touch. This rash appears on the extremities, fingers toes and ears, due to **inflammation that causes angiitis**. Clothing that rubs should be avoided.

Typical appearance of chiblains

- **Acrocyanosis.** Is similar to Raynaud's and is extremely difficult to differentiate from it. There is usually some oedema, there isn't such a fixed temporary fluctuation of symptoms and in most of cases the initial change in colour is blue and not white.

- **Erythromelalgia and causalgia.** This is a chronic disorder characterized by persistent warmth, pain and redness, mainly affecting the feet and lower legs. It may seem opposite to Raynaud's as heat often provokes the symptoms [different drugs are also used for treatment [b-blockers and carbamazepine 200mg bd].

- **Livedo reticularis**. Is primarily a cosmetic problem affecting the legs of young women. Is caused by spasm of the skin vessels on cold resulting in light bluish patches.

- **Rheumatoid arthritis.** Arthritis affects the lining of the joints. This lining produces the fluid that lubricates the joint and when affected by rheumatoid arthritis it becomes inflamed and swollen. About 105 of rheumatoid arthritis sufferers experience Raynaud's phenomenon.

- **Systemic Lupus Erythematosus**. Lupus is characterized by a rash, which is usually seen on both cheeks and the bridge of the nose, as well as chronic inflammation of the blood vessels and connective tissue of the body. There is associated tiredness, joint pain, mouth ulcers, hair loss and Raynaud's.

- **Chemical or drug induced Raynaud's**. Some chemicals at work [vinyl chloride] or drugs as beta-blockers, migraine tablets and oral contraceptives 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.

4. How are these vasoconstrictive disorders diagnosed?

The **history of the disease** is most important. Measurement of skin temperature or examination of the small blood vessels at the nail bed [**capillaroscopy**] may be also helpful. A variety of tests may follow initial diagnosis as assessment should be directed at identifying any underlying disorders [if applicable] and assessing the effect of the disease on the patient's quality of life as this will indicate treatment. **Investigations** are to some extent determined by the suspicions aroused on clinical examination but should include full blood count, thyroxine levels, biochemical and urine analysis, Ra-test, ESR, ANA, Lupus anticoagulant and radiographs of the neck/chest and hands.

5. Is Raynaud's 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.

6. 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 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 finger tips. The best way to give up is to choose a day when you are going to stop completely than trying to cut down gradually. If you do have trouble giving it up, please ask your doctor who can put you in touch with a support group.

- **Eating**. 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, especially before retiring at night, 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 to the extremities. 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 trunk warm. Feet are especially prone to cooling, therefore a good thick pair of socks and proper shoes are essential. Wet shoes and clothes should be changed as soon as possible.

7. What about treatment?

There is no cure for most of vasoconstrictive disorders. Treatment depends on the severity of the symptoms. Try to push away the anxiety that the problem provokes.

- For mild symptoms the **conservative treatment** described above usually suffices. Electrically heated gloves [Vascutherm] and special shoes made from Goretex may be of benefit.

- For moderate symptoms **some drugs may be needed**. Your specialist may prescribe thymoxamine [Opilon] a mild a_1 and a_2 adrenergic receptor blocker that increases blood flow to the skin. It has the least side effects and is well tolerated in a dose of 40mg four times a day. For more resistant cases vasodilators may be used [nifedipine] or combinations of drugs [i.e. guanethidine 10mg daily plus prazocin 1mg twice a day]

- Occasionally, and for severe symptoms, your specialist may feel an operation called **sympathectomy** may be of benefit. This involves either cutting or destroying the nerves that cause the arteries to constrict. The operation is more successful for raynaud's of the feet.

- For severe cases, particularly those with ulceration and fingertip gangrene **admission to the hospital and drug infusions** [prostacyclin 6-15ng/kgr/hour for 12 hours per day] may be of benefit.

8. What is the prognosis?

people who develop Raynaud's as teenagers often have a form that is benign and will disappear with age. Unfortunately, this is not true in all cases as sometimes Raynaud's may persist.

For secondary Raynaud's, prognosis is good for only some of the cases. There is no cure for scleroderma at present, but there are many effective treatments available to alleviate specific symptoms.