Scleroderma Association B.C.

RAYNAUD'S PHENOMENON AND DIGITAL ULCERS

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WHAT IS RAYNAUD'S PHENOMENON?

The most common and earliest problem observed in systemic sclerosis is Raynaud's phenomenon (RP). This phenomenon is due to a narrowing of the blood vessels in the fingers caused by exposure to cold and strong emotions, and is manifested by a change in colour of the fingers, which turn white, then blue and finally red.

The white ("syncopal") phase represents a partial stoppage of blood flow and is characterized by a well-defined pallor of the affected fingers. It may also be associated with numbness in the fingers. This phase is always present in RP, unlike the other phases which may or may not be present. The blue ("cyanotic") phase is due to a lack of oxygen in the tissues. Finally, the red ("hyperemic") phase is due to the return of blood flow and may be accompanied by pain with a burning sensation. RP mainly affects the fingers, but can also affect the toes, nose or ears.

RP can be "primary", i.e., isolated and not associated with an underlying autoimmune disease. However, when it occurs after the age of 40 or is associated with finger ulcerations (open lesions in the skin that heal very slowly) or other symptoms and signs of systemic sclerosis, a diagnosis of underlying systemic sclerosis should be suspected and sought.

DIGITAL ULCERS

When RP is secondary to systemic sclerosis, RP is initially caused by a reversible hyperreactivity of the vessels to cold. Later, excessive proliferation of several cells causes a fixed narrowing of the opening of the vessels, such that circulation is reduced due to permanent structural changes. In this context, the loss of blood circulation can become severe to the point of causing ulcerations (open skin lesions) at the fingertips. These lesions can be slow to heal and can be complicated by local infection of the skin (cellulitis) or of the underlying bone (osteitis). In extreme cases, RP can be severe to the point of necrosis (death of part of the finger) or, rarely, self-amputation (loss of the fingertip).

WHAT CAN BE DONE TO AVOID AGGRAVATING RAYNAUD'S PHENOMENON?

In order to improve RP symptoms and prevent complications associated with RP, certain environmental factors that can aggravate RP should be avoided, including cold, stress, tobacco smoking, as well as medications and products with a vasoconstrictive effect (vessel narrowing). Here are some general measures suggested for patients with RP. **See the last page of this document for some general measures suggested to patients with RP.**

These simple measures may be sufficient to treat RP in the early stage (before structural damage has occurred). However, when there is structural damage to the vessels (fixed narrowing), these non-pharmacological measures become insufficient and the treating physician will often use medications to help dilate the vessels.

MEDICATIONS TO TREAT RAYNAUD'S PHENOMENON

Most medications for the treatment of RP work by relaxing the blood vessels, allowing better blood flow to the extremities.

The first line of drug treatment for uncomplicated RP (without ulceration, necrosis or risk of self-amputation) is a calcium channel blocker (e.g., Nifedipine XL or Amlodipine). The dose is gradually increased as tolerated until symptoms improve. If the medication is not tolerated (dizziness, headache, swelling of the feet) or is not effective at the highest tolerated dose, the medication is switched to another calcium channel blocker.

If there is still no efficacy or if there is intolerance, a medication of another class, alone or sometimes in combination, such as a phosphodiesterase type 5 (PDE5) inhibitor (e.g., Sildenafil or Tadalafil) may be used. The addition of an angiotensin II receptor antagonist (e.g., Losartan) or a selective serotonin reuptake inhibitor (e.g., Fluoxetine) may also be considered.

If RP does not respond to these treatments or is severe with impairment of function and quality of life, a prostacyclin (e.g., Flolan) or its analogues can be given intravenously on an outpatient basis.

WHAT IF THERE ARE DIGITAL ULCERS?

When digital ulcers are present, aggressive treatment is required to accelerate healing and prevent necrosis or infection of the finger. The dose of the calcium channel blocker already prescribed is increased to the maximum tolerated dose, with the addition of a PDE5 inhibitor. The addition of low-dose aspirin (81 mg daily) may also be useful to promote better local circulation if there is no contraindication (e.g., gastrointestinal bleeding). A statin (e.g., Atorvastatin) may also be added for its potential protective effects on the blood vessels. Ulcers can be extremely painful and the use of narcotic drugs may be necessary.

In more severe cases, daily intravenous treatment for five consecutive days with a prostacyclin (Flolan), followed by maintenance treatment every three weeks thereafter, may be given on an outpatient basis to accelerate ulcer healing.

Injecting an anesthetic product (lidocaine or bupivacaine) into the ulcerated fingers ("local chemical sympathectomy") may be effective in improving the blood flow locally in some cases, but with benefits that are usually temporary.

In extremely severe cases where the physician feels there is a risk of necrosis and self-amputation of fingers (fingers that remain permanently white or become purplish despite treatment), emergency treatment is required: the patient should be hospitalized and placed in a warm, quiet place for increased monitoring during treatment and further investigation (blood samples and assessment of vessel permeability in radiology after local injection of a dye, or "arteriography"). Treatment of ulcerations is sometimes maximized by the addition of an intravenous or subcutaneous anticoagulant (heparin), and Flolan and local chemical sympathectomy should be initiated promptly if necessary.

If ulcer episodes recur or are very severe despite the treatments listed above, Bosentan, an endothelin antagonist, may be added. This medication works on the blood vessels and can prevent the recurrence of digital ulcers.

Well-established digital ulcers can take up to several months to heal. They must be closely monitored by the doctor for the onset of infection or progression to necrosis. Signs that an infection may be present include pus draining from the ulcer, significant redness and swelling around the ulcer and on the affected finger, or rapid progression of the ulcer. There may also be fever and chills. The doctor will then check for germs (bacterial culture) and, depending on the severity, prescribe antibiotics by mouth or intravenously.

WHAT CAN BE DONE TO AVOID AGGRAVATING RAYNAUD'S PHENOMENON?

In order to improve RP symptoms and prevent complications associated with RP, certain environmental factors that can aggravate RP should be avoided, including cold, stress, tobacco smoking, as well as medications and products with a vasoconstrictive effect (vessel narrowing).
Here are some general measures suggested for patients with RP.

AGGRAVATING FACTOR	RECOMMENDATIONS
COLD	▶ Keep the whole body warm (e.g., wear thermal underwear, heat-retaining hat). ▶ Keep fingers warm (e.g., warm mittens or electric hand warmers). ▶ Avoid sudden exposure to cold. ▶ Avoid abrupt changes in temperature (e.g., from a heated room to an air-conditioned room), cold breezes and cold humid air. ▶ Quickly apply methods to end an RP attack: place hands in a warm place (e.g., warm water, under the armpits) or move your arms in rotation to promote circulation in the extremities.
STRESS	Reduce stress.
TOBACCO	Avoid smoking and/or being exposed to cigarette smoke (quitting smoking can significantly improve RP).
MEDICATIONS AND PRODUCTS WITH VASOCONSTRICTIVE EFFECTS	**Avoid or consult your pharmacist before using:** ▶ Nasal decongestants ▶ Amphetamines ▶ Some antihypertensive medications (e.g., clonidine, beta-blockers) ▶ Medications to treat attention deficit hyperactivity disorder, ADHD (e.g., methylphenidate or dextroamphetamine) ▶ Anti-migraine medications (e.g., ergotamine) ▶ Weight loss pills ▶ Natural products containing ephedra
CAFFEINATED BEVERAGES	Reduce consumption of caffeine-containing beverages (e.g., coffee, tea, colas).
SYMPATHOMIMETIC RECREATIONAL DRUGS	Avoid cocaine and amphetamines (e.g., speed).