



Lupus and Overlap Raynaud's Phenomenon

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Raynaud's phenomenon

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Raynaud's phenomenon (pronounced /re??no?z/) (r?y-N?Z), in medicine, is a vasospastic disorder causing discoloration of the fingers, toes, and occasionally other extremities. This condition can also cause nails to become brittle with longitudinal ridges. Named for French physician Maurice Raynaud (1834 - 1881), the cause of the phenomenon is believed to be the result of vasospasms that decrease blood supply to the respective regions.

Emotional stress and cold are classic triggers of the phenomenon and the discoloration follows a characteristic pattern in time: white, blue and red.

It comprises both Raynaud's disease (primary Raynaud's), where the phenomenon is idiopathic, and Raynaud's syndrome (secondary Raynaud's), where it is caused by some other instigating factor. Measurement of hand-temperature gradients is one tool used to distinguish between the primary and secondary forms.

It is possible for the primary form to progress to the secondary form.

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Incidence

The phenomenon is more common in women than men, with the Framingham Study finding that 5.8% of men and 9.6% of women suffered from it.



Epidemiology

There is a familial component to primary Raynaud's, and presentation is typically before two. Smoking worsens frequency and intensity of attacks, and there is a hormonal component. Sufferers are more likely to have migraine and angina than controls.

Secondary Raynaud's has a number of associations:

Connective tissue disorders:

- scleroderma
- systemic lupus erythematosus
- rheumatoid arthritis
- Sjögren's syndrome
- dermatomyositis
- polymyositis
- Eating disorders
- Anorexia Nervosa
- Obstructive disorders
- atherosclerosis
- Buerger's disease
- subclavian aneurysms
- thoracic outlet syndrome
- Drugs
- Beta-blockers
- cytotoxic drugs - particularly
- chemotherapeutics and most especially bleomycin
- cyclosporine
- ergotamine
- sulfasalazine
- Occupation
- jobs involving vibration, particularly drilling
- exposure to vinyl chloride, mercury
- exposure to the cold (e.g. by working packing frozen food)
- Others
- hypothyroidism
- cryoglobulinemia
- malignancy
- reflex sympathetic dystrophy

It is important to realize that Raynaud's can herald these diseases by periods of more than 20 years in some cases, making it effectively their first presenting symptom. This can be the case in the CREST syndrome, of which Raynaud's is a part.

Symptoms

The condition causes painful, pale, cold extremities. This can often be distressing to those who are not diagnosed, and sometimes it can be obstructive. If someone with Raynaud's is placed in too cold a climate, it could potentially become dangerous.

Unilateral Raynaud's, or that which is present only in the hands or feet, is almost certainly secondary, as primary Raynaud's is a systemic condition.

However, a patient's feet may be affected without him or her realizing it.

In pregnancy, this sign normally disappears due to increased surface blood flow.

Investigations

A careful history will often reveal whether the condition is primary or secondary. Once this has been established, investigations are largely to identify or exclude possible secondary causes.

Digital artery pressure: pressures are measured in the digital arteries before and after cooling the hands. A drop of 15mmHg or more is diagnostic.

Doppler ultrasound: to assess flow

Full blood count: this can reveal a normocytic anemia suggesting the anemia of chronic disease or renal failure

Urea & Electrolytes: this can reveal renal impairment

Thyroid function tests: this can reveal hypothyroidism An autoantibody screen, tests for rheumatoid factor, Erythrocyte sedimentation rate and C-reactive protein, which may reveal specific causative illnesses or a generalized inflammatory process

Nail fold vasculature: this can be examined under the microscope

Pathophysiology

Primary Raynaud phenomenon, stemming from Raynaud disease, is an exaggeration of vasomotor responses to cold or emotional stress.

More specifically, it is a hyper activation of the sympathetic system causing extreme vasoconstriction of the peripheral blood vessels, leading to tissue hypoxia. Chronic, recurrent cases of Raynaud phenomenon can result in atrophy of the skin, subcutaneous tissues, and muscle. It can also rarely cause ulceration and ischemic gangrene.

Treatment

Treatment options are dependent on the type of Raynaud's present. Raynaud's syndrome is treated primarily by addressing the underlying cause, but includes all options for Raynaud's disease as well.

Treatment of primary

Raynaud's focuses on avoiding triggers:

General measures

Avoidance of any environmental triggers, e.g. cold, vibration, etc. (although emotional stress is a recognized trigger, it tends to be impossible to consciously avoid).

Warm clothing for the extremities such as mittens or headbands.

Hormone regulation and assessment of the type of hormonal contraception used, if any. Contraception which is low in estrogen is preferable, and the progesterone only pill is often prescribed.

Smoking cessation.

Emergency measures

If white finger (Raynaud's), occurs unexpectedly and a source of warm water is available allow tepid to slightly warm water to run over the affected digits while gently massaging the area. Continue this process until the white area turns pink or a normal healthy color.

If triggered by exposure in a cold environment, and no warm water is available, place the affected digits in a warm body cavity - arm pit, crotch, or even in the mouth. Keep the affected area warm at least until the whiteness returns to pink or a healthy color, avoid continued exposure to the cold.

