

Raynaud's Disease and Phenomenon

Spasm of arterioles, usually in the digits and occasionally in other acral parts (eg, nose, tongue), with intermittent pallor or cyanosis.

Raynaud's disease, most common in young women (60 to 90% of reported cases), is idiopathic. **Raynaud's phenomenon** is secondary to other conditions, such as connective tissue disorders (scleroderma, RA, SLE), obstructive arterial diseases (arteriosclerosis obliterans, thromboangiitis obliterans, thoracic outlet syndrome), neurogenic lesions, drug intoxications (ergot, methysergide), dysproteinemias, myxedema, primary pulmonary hypertension, and trauma.

Pathology and Pathophysiology

Although the pathogenesis of Raynaud's disease remains uncertain, research into prostaglandin metabolism, microcirculation, and the role of the endothelial cell is yielding promising results. Raynaud's phenomenon is associated clinically with migraine headaches, variant angina, and pulmonary hypertension, suggesting that these disorders may share a common vasospastic mechanism.

In **Raynaud's disease**, the threshold for the vasospastic response is lowered by local cold or anything that activates sympathetic outflow or releases catecholamines (eg, emotion). Vessels are histologically normal in early stages, but in advanced cases the arterial intima may thicken and thromboses may form in small arteries.

Symptoms and Signs

Intermittent blanching or cyanosis of the digits is precipitated by exposure to cold or by emotional upset. Color changes may be triphasic (pallor, cyanosis, redness--reactive hyperemia) or biphasic (cyanosis, redness). They do not occur above the metacarpophalangeal joints and rarely involve the thumb. Pain is uncommon, but paresthesias are frequent during the attack. Vasospasm of the digital arteries and arterioles may last minutes to hours but is rarely severe enough to cause gross tissue loss. Rewarming the hands restores normal color and sensation.

Diagnosis

Raynaud's disease is differentiated from Raynaud's phenomenon by bilateral involvement and no evidence of an underlying cause. In Raynaud's disease, trophic skin changes and gangrene are absent or affect minimal areas, and symptoms do not worsen despite their presence for many years.

In **Raynaud's phenomenon**, there is a recognizable underlying cause. For example, with scleroderma, there may be tightness or thickening of the skin and telangiectases of the hands, arms, or face; difficulty swallowing; painful trophic ulcers on the fingertips; and

symptoms referable to other systems. Wrist pulses are usually present, but **Allen's test**, which is usually negative in Raynaud's disease, frequently shows occlusion of the radial or ulnar arterial branches distal to the wrist. In this test, the examiner faces the patient and places the thumbs over the radial and ulnar pulsations of one hand. After the patient clenches the fist to expel the blood from the hand, the examiner compresses the arteries. When the patient opens the fist, the hand is pale. The examiner then releases pressure from the radial artery but not the ulnar artery. If the radial artery distal to the wrist is patent, the hand will rapidly turn pink. If the artery is occluded, the hand will remain pale. This maneuver is repeated with the pressure released from the ulnar artery but not the radial artery. Noninvasive testing of the affected digits with plethysmography before and after exposure to cold can differentiate occlusive from vasospastic disease.

Treatment

Raynaud's disease, when mild, may be controlled by protecting the body and extremities from cold. The patient must stop smoking because nicotine is a vasoconstrictor. In a few patients, relaxation techniques (eg, biofeedback) may reduce vasospastic episodes. Prazosin 1 to 2 mg po at bedtime (repeated in the morning if necessary) and the Ca blocker nifedipine 10 to 30 mg po tid may be helpful. Pentoxifylline 400 mg bid or tid with meals is reportedly effective. Phenoxylbenzamine 10 mg po daily to tid and guanethidine 10 mg po daily to tid have had occasional success.

Therapy for **Raynaud's phenomenon** depends on recognition and treatment of the underlying disorder. Phenoxylbenzamine 10 mg po daily to tid may be useful. Antibiotics, analgesics, and, occasionally, surgical debridement are essential for patients with extremely painful and infected fingertip ulcers, especially those with scleroderma.

Research on the use of prostaglandins (thromboxane) is encouraging. Regional sympathectomy is reserved for patients with progressive disability; it often abolishes the symptoms, but relief may last only 1 to 2 yr. Results from sympathectomy are generally better in patients with Raynaud's disease than in those with Raynaud's phenomenon. In Raynaud's disease and Raynaud's phenomenon, β -blockers, clonidine, and ergot preparations are contraindicated, because they cause vasoconstriction and may induce or worsen symptoms.