

VASOSPASTIC CRISIS

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Vasospasm of the lower extremity is a relatively uncommon finding usually observed in patients with Raynaud's syndrome. However, many varying clinical conditions are associated with a concurrent vasospastic episode affecting both the lower and upper extremities. To date, several disease states are well established to include "vasospasm". (Table 1) This text will focus on Raynaud's Syndrome and its potential complications, as it is undoubtedly the most prevalent underlying cause of vasospasm of the digits in the foot. Bear in mind that systemic illness as well as the untoward effects of medication need to be included in a differential diagnosis for anyone presenting with signs and/or symptoms related to vasospasm.

Raynaud's Syndrome is defined as episodic digital vasospasm occurring in response to cold or emotional stimuli and is associated with blanching, cyanosis or both. Classically, this condition presents as pallor (white), followed by cyanosis (blue) and lastly as digital flow is restored in response to ischemia (red). The digits are usually of normal color between attacks.

The term Raynaud's Syndrome was coined by Maurice Raynaud (1834-1881), when he described this phenomenon in a patient with systemic sclerosis (PSS) in 1863. It was Jonathan Hutchinson in 1899 who pointed out the consistent association of Raynaud's phenomena with sero-negative disease. Worldwide, especially in geographic areas with cool, damp climates such as Denmark, England, and the Northwest portion of the United States, up to 20% of the population is affected. Another specific group of individuals also predisposed to developing Raynaud's Syndrome include individuals who utilize vibratory equipment and tools in their occupation.

TABLE 1

DISEASES WITH VASOSPASTIC COMPONENTS

Acrocyanosis
Cryoglobulinemia
Erythromelalgia
Livedo Reticularis
Polymyositis/Dermatomyositis
Progressive Systemic Sclerosis
Reflex Sympathetic Dystrophy
Sjorgen's Syndrome
Systemic Lupus Erythematosus

The great majority of patients affected by Raynaud's present with an occasional nuisance of cold toes or fingers. However, in a small subset of patients, Raynaud's Syndrome signals the presence of a potentially serious ailment, commonly resulting in severe ischemic changes and intractable ulcers resistant to conventional therapy.

The literature contains data suggesting that the mechanism of Raynaud's phenomenon falls into two categories which must be differentiated in the individual patient. The first category consists of a forceful digital artery contraction in response to cold or emotion stimuli. This contraction is sufficiently strong enough to overcome the distending luminal pressure in the digital artery causing digital artery closure. The other mechanism occurs in patients with digital artery obstructive disease already present. These patients have fixed digital artery obstruction(s) with associated diminished digital artery pressure. Hence, a presumably normal digital artery contraction in response to cold or emotional stress may be suffi-

cient to overcome the diminished intraluminal pressure and thereby cause digital artery closure. (Fig. 1)

Once the differential diagnosis of Raynaud's Syndrome has been entertained, several laboratory tests are available that allow the accurate diagnosis of this condition although none are perfect. Currently, there is no test that permits quantitation of the severity of Raynaud's phenomenon. Thus, there are accurate qualitative tests available, but no quantitative tests. Unfortunately, without a reliable test to rate the severity of the disease, it is difficult to group similar patients for randomization, or to assess the response to treatment in an objective manner. This difficulty has hampered research into the pathophysiology of this disease state, as well as treatment protocols.

Photoplethysmography (PPG) records the analog wave forms, and is considered to be the most accurate test for diagnosing lower extremity disease. A PPG measurement of the patient's toe at normal temperature is recorded. The measurement is recorded again after the foot has been immersed in ice water for 10 seconds. A normal response shows only a modest increase in peak amplitude whereas an abnormal or vasospastic response results in a total or near total cessation of detectable digital flow. This test has proven to be very useful for the lower extremity, however, interestingly has not been validated for disease of the hand. (Fig. 2A - 2C) Several other tests are available including many sophisticated modalities with most utilizing a form of cold challenge to confirm a diagnosis.

Treatment of this disorder is multifactorial and should be approached in logical fashion. A

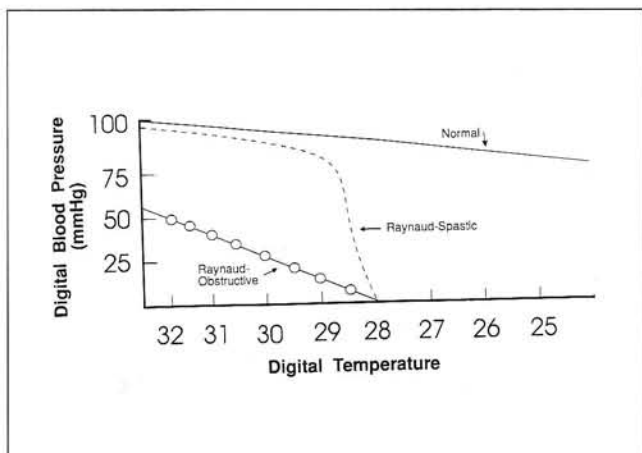


Fig. 1. Representation of response to cold in vasospastic patients.

lack of definitive quantitative measurements has made the patient's subjective evaluation the most sensitive marker to treatment response. Conservative measures are numerous and they should be undertaken in all individuals with Raynaud's Syndrome. Education regarding the disease patho-

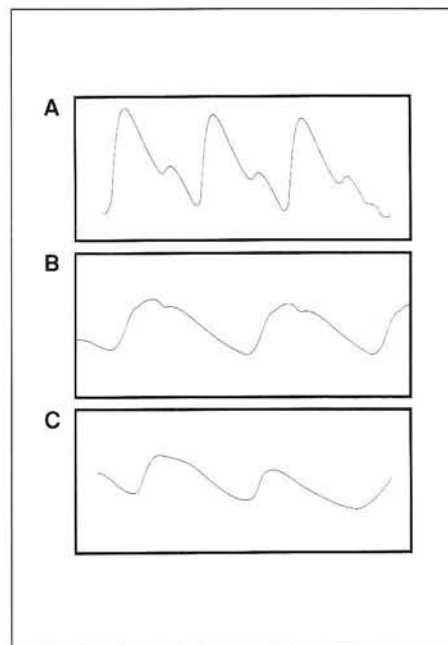


Fig. 2 A. Normal digital photoplethysmography waveform. B. Peaked digital photoplethysmography waveform. C. Obstructive digital photoplethysmography waveform.

physiology should be clearly described to the patient to allay fears and rid false impressions of this disorder. Many patients are afraid that circulation problems will ultimately lead to digital gangrene or amputation. It is vital to explain to the patient that the pallor and cyanosis in the great majority of cases is an exaggeration of a normal response.

All patients must be strongly advised to stop smoking, as smoking dramatically increases the tendency of arterio-spasm. Precautions should also be taken to protect the extremities from the noxious stimulus which produces the symptoms. Protection from cold exposure, as well as control of emotional stress should be stressed. Medications which have been associated with vasospasticity should be discontinued if possible, including ergotamine preparations, beta-adrenergic blockers, and oral contraceptives.

Most patients will do well if conservative measures are offered and compliance is ensured.

Those patients that do not gain substantial relief from the measures outlined in Table 2 require additional treatment. Medication, as a general rule should be reserved for patients with frequent or severe attacks. The drugs utilized in the treatment of Raynaud's Syndrome frequently cause undesirable side effects.

Reserpine has traditionally been the drug of choice prescribed for the treatment of Raynaud's phenomena recalcitrant to conservative treatments. Reserpine is no longer recommended as a treatment because of the severe side effects including lethargy, depression, impotence, and increased gastric acid production. Other medications have also been prescribed including alpha-adrenergic blocking agents such as methyldopa, guanethidine, prazosin, and tolazline. These preparations have many side-effects which restrict their effectiveness.

Direct vasodilators, specifically Nifedipine (Procardia) are now frequently prescribed with high patient acceptability and response. This agent works as a calcium channel blocker and actively dilates the arterial wall. The correct dosage depends upon the severity of symptoms and should be tailored individually with a usual response to 10 mg once or twice daily. If this dosage is not appropriate, titrating the drug according to relief of symptomatology and diminishment of side effects is recommended. Other calcium channel blockers including verapamil and diltiazem are not as effective as nifedipine, and are not usually recommended in the treatment of Raynaud's Syndrome. Trental (pentoxifylline) does not seem to be effective in treating Raynaud's, although it may be of benefit in patients with obstructive disease as it may help in the rheology of the red blood cell. Other treatments have included Griseofulvin, which has a vasodilatory effect as well as its well-known anti-fungal properties. However, this has not had more success than other less-toxic drugs. Ketanserin, a serotonin antagonist has recently been shown to be effective as well. Many newly developing prostaglandins show promise although more data is needed before these agents will be recommended.

Minimal value has been seen in treating patients with unresolved symptomatology from the newer operative techniques aimed at microsurgical bypass. Temperature biofeedback,

Pavlovian conditioning, and plasmapheresis are also mentioned in the literature as a "last ditch" effort to overcome Raynaud's Syndrome. Some improvements have been documented, although few studies are available.

SUMMARY

The vast majority of patients with this disease receive symptomatic relief from conservative and medical approaches. Disease associated with systemic illness generally improves with adequate management of the underlying disorder. All patients presenting with vasospastic disease should be thoroughly educated and monitored for effective relief of this disease state.

RISK MANAGEMENT CONCERNS

Care should be taken with patients who relate an accurate history of Raynaud's-like signs and symptoms. Potentially disastrous results can be avoided when surgically treating foot pathology in patients suspected of exhibiting Raynaud's phenomenon.

TABLE 2

OPTIONAL TREATMENTS

- I. General
 - A. Education
 - B. Avoid Cold
 - C. Avoid medications that aggravate symptoms
 - D. Stop Smoking
 - II. Medical
 - A. Direct smooth muscle relaxants
 - B. Sympathetic blocking agents
 - C. Calcium channel blockers
 - III. Operation
 - A. Digital artery sympathectomy
 - B. Microvascular reconstruction
 - C. Cervicothoracic sympathectomy
 - IV. Other
 - A. Behavior modification
 - B. Biofeedback
 - C. Treatment of associated disease
 - D. Pavlovian vasodilatation conditioning
 - E. Plasmapheresis
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