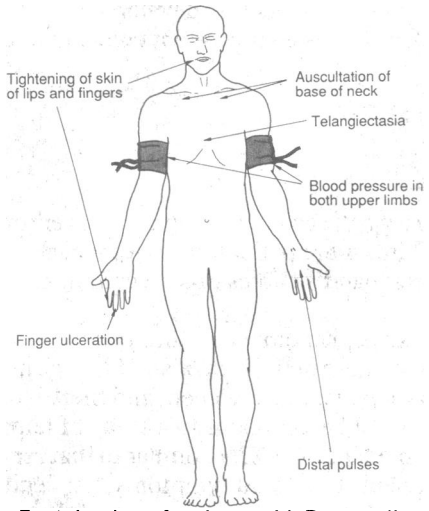
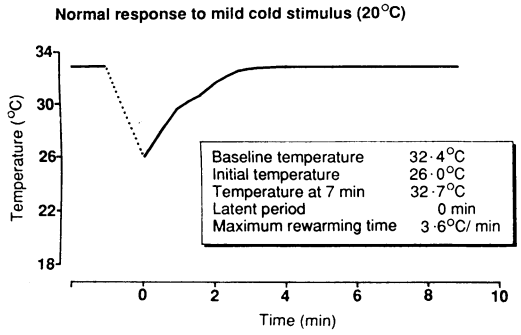
RAYNAUD'S SYNDROME AND SIMILAR CONDITIONS

M H Grigg, John H N Wolfe

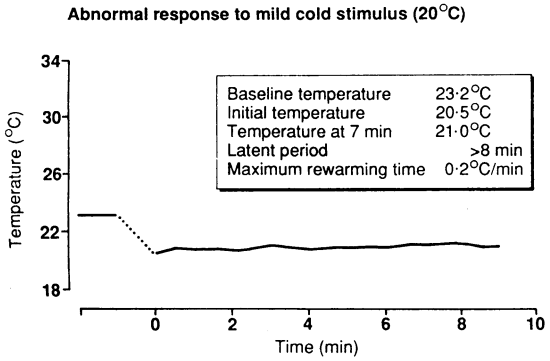
Raynaud's phenomenon



Examination of patients with Raynaud's phenomenon.



Digital skin temperature before and after immersion of hand in water at 20°C for one minute by a normal volunteer. Rewarming occurred within three minutes. Zero time is when the hand was removed from the water.



Digital skin temperature before and after immersion of hand in water at 20°C for one minute by a patient with Raynaud's phenomenon. Rewarming to baseline temperature had not occurred within nine minutes.

The circulation of the extremities can be regulated by temperature—for example, exposure to cold in a normal person will cause vasoconstriction and a decrease in blood flow to the skin. Abnormalities of this apparently simple response have been of interest since Maurice Raynaud described the syndrome that bears his name in 1862. He attributed the clinical signs that he observed to overactivity of the sympathetic nervous system, and so began a series of controversies that persist today.

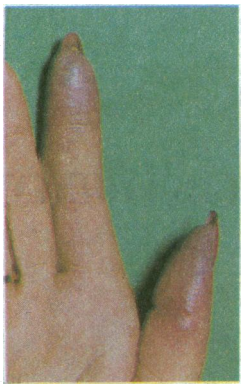
Diagnosis

Underlying many of the controversies surrounding Raynaud's phenomenon is the subjectivity of the diagnosis. This depends on a history of the characteristic colour changes in the fingers that are provoked by exposure to cold, and—less reliably—by emotion; cold, however, is the only one of the factors that is universally accepted. These colour changes may be accompanied by parasthesia and other sensations, but pain is not a prominent feature.

Many tests have been devised to try to find an objective method of diagnosis. These include plethysmography (usually mercury in a silicone rubber strain gauge, which can monitor changes in the volume of the digit by arterial pulsation); Doppler ultrasonography (the digital arteries can be insonated to see if they are patent, and even to measure pressure); laser Doppler flowmetry and direct capillaroscopy (which can be used to assess the velocity of red blood cells in the microcirculation); and thermal entrainment (which measures changes in blood flow in one extremity while the other is exposed repeatedly to opposing thermal stimuli). Thermal entrainment is non-invasive, quick to do, and reproducible, and may be of most benefit in assessing the effects of drugs.

The simplest way of assessing skin blood flow is by measuring its temperature, as there is a linear relation between temperature and blood flow up to digital skin temperatures of 34°C. The absolute skin temperature is not, however, a good discriminator, because it is influenced by various factors (both internal and external) that are difficult to control. To separate normal from abnormal values it is necessary to apply a cold stimulus and then monitor rates of rewarming. Normal subjects rewarm rapidly after cessation of a mild cold stimulus (water at 20°C), whereas patients with Raynaud's phenomenon go through a latent period. In addition, once rewarming has begun patients with vaso-obstruction as well as vasospasm do not seem to rewarm as fast as normal subjects.

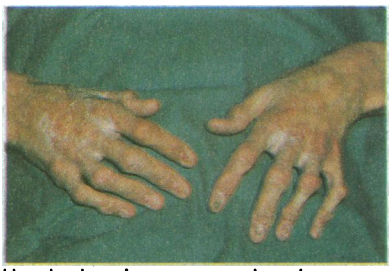
The idea that vessels "hyper-respond" to cold is attractive, but there has been a growing realisation that the pathophysiological defect is failure to recover from a cold stimulus rather than an initial over-response to it.



Arteriovenous malformation masquerading as Raynaud's phenomenon.

In 1893 Hutchinson drew attention to the association between Raynaud's phenomenon and scleroderma, and since that time the number of possible underlying disorders has increased. Although there is a need to separate the so called "primary" and "secondary" types, the distinction becomes more blurred as our understanding of the condition increases and "Raynaud's phenomenon" is seen as the final common pathway of a number of abnormalities. Initially secondary Raynaud's was thought to be uncommon, but with the search for an underlying disorder becoming more thorough this may not be the case.

Both vasospasm and fixed obstruction of the lumen of the digital vessels occur to a greater or lesser degree in most patients. More importantly there is concern that patients may progress from vasospasm to develop digital and microvascular obstruction. Undoubtedly this may be the result of progression of the underlying disease, but it could also be the result of continued, unprotected exposure to cold. The experience of patients who have had frostbite in the past and who subsequently develop hypersensitivity to cold in the affected part confirms that cold alone can cause severe damage.

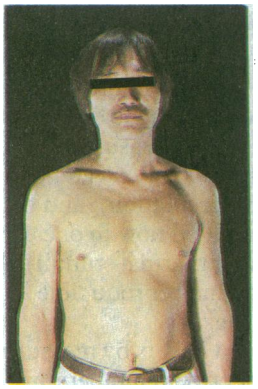


Hands showing severe scleroderma.

Incidence

The relative incidence of primary and secondary Raynaud's is entirely dependent on referral patterns. Thus more patients with secondary Raynaud's are seen in specialist Raynaud's clinics than in community practice.

The overall incidence of Raynaud's phenomenon, both primary and secondary, is difficult to estimate because of the lack of an objective method of diagnosis. Certainly about 90% of patients are women, and there is often a family history. Hypersensitivity to cold is common. In a study of female factory workers in Denmark the incidence was 22%, similar to that reported from the United States. Many patients have mild symptoms that begin when they are teenagers and gradually abate around the time of the menopause. Nevertheless, between 5% and 15% of patients with Raynaud's phenomenon will develop overt scleroderma (about 90% of patients with scleroderma also have Raynaud's phenomenon). Of most concern is the (fortunately) small group—less than 1%—who go on to develop digital gangrene and require amputation. Patients with poor prognoses cannot reliably be predicted in the early stages of their disease, but they do seem to develop vaso-obstruction in addition to vasospasm.



Patient with left cervical rib.

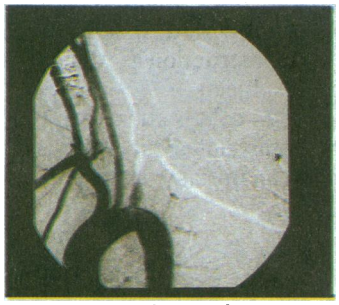
Clinical assessment

Having made the initial clinical diagnosis of Raynaud's phenomenon, the assessment should be directed at: (*a*) identifying any underlying disorders, and (*b*) assessing the effect of the disease on the patient, as this will indicate the treatment.

The patient should be questioned about family history, drug taking, and occupation—especially handling ice (as with some workers in the food industry) and the use of vibrating machinery. In addition, a history of arthralgia, dysphagia, or xerostomia should be sought as these suggest an underlying collagen disorder.

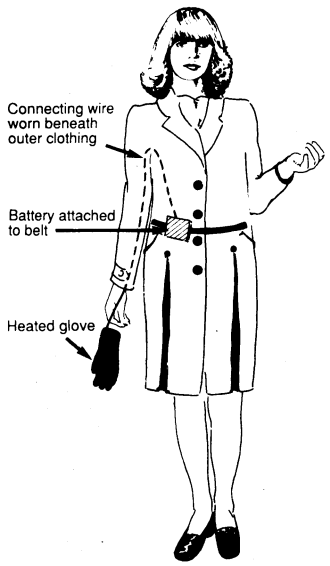
Physical examination should include careful assessment of upper limb pulses by both palpation and auscultation; the latter should be done over both the supraclavicular fossas and the deltopectoral triangle. These results, together with the measurements of blood pressure in both upper limbs, may show an anatomical distortion of the axillary or subclavian arteries—for example, as a result of cervical rib or band. Asymmetrical Raynaud's raises the possibility of a "mechanical," and therefore surgically correctable, arterial lesion. This is particularly true in the older age groups when atherosclerosis of the main upper limb arteries may be causing symptoms. A search should also be made for skin tightening, particularly of the fingers and around the mouth, for telangiectasia, and for carpal tunnel syndrome.

Investigations are to some extent determined by the suspicions aroused during clinical assessment, but should include full blood count, biochemical and urine analysis, and radiographs of the hands.



Digital subtraction angiogram showing subclavian "steal" syndrome.

Vascutherm battery powered gloves.



The glove is powered by a rechargable battery, and the connecting wire is inconspicuous beneath the clothing.

Treatment

There is no cure for Raynaud's phenomenon. The palliative treatment must depend on the severity of the symptoms. For patients with mild symptoms explanation and reassurance may be all that is required. From the patient's point of view the problem is not only the "poor circulation," but also the anxiety that this provokes. If appropriate, community support may be given through the Raynaud's Association Trust, 112 Crewe Road, Alsager, Cheshire ST7 25A, telephone 0270-872776.

General advice is of paramount importance: stop smoking and avoid cold. Electrically heated gloves (Vascutherm) have been of great benefit to some patients. Skiers with this problem have found that the centrifugal force resulting from a windmill action of the arms can stave off the effects and maintain some perfusion of the finger tips.

For patients with moderate symptoms drugs may be needed. Thymoxamine (Opilon) is a mild α_1 and α_2 adrenergic receptor blocker that increases blood flow to the skin. In contrast with some other agents it has few side effects and is well tolerated in a dose of 40 mg four times a day. The calcium channel blocking agent nifedipine is also useful (maximum dose 20 mg twice a day); the patient should not be started on the full dose—an initial dose of 10 mg twice a day should be increased after a week to prevent side effects. For more resistant cases a combination of agents (to reduce the incidence of side effects)—for example, guanethidine 10 mg daily and prazosin 1 mg twice a day—may be considered.

The severe cases, particularly those in which there is ulceration or gangrene, are a challenge to the clinician. Admission to hospital may be required. Prostaglandin infusions, reserpine given intra-arterially, and plasmapheresis are all useful in some patients. Prostacyclin usually has a pleasing immediate result and in some patients the improvement may be maintained for several months. Two or three admissions to hospital for infusions can tide a patient over the bleak winter months. The patient is started on 6 ng/kg/hour and the dose increased to 15 ng/kg/hour depending on the effect and the side effects. Side effects are dose related, and almost immediate relief will be obtained by reducing the infusion.

The indications for cervicothoracic sympathectomy are few. Beneficial effects, especially in patients with connective tissue disorders, are difficult to achieve and are short lived.

Other vasospastic conditions

Conditions with which Raynaud's phenomenon is associated

- Connective tissue disorders:
  - Scleroderma
  - Systemic lupus erythematosus
  - Rheumatoid arthritis
  - Other
- Obstructive arterial disease:
  - Thoracic outlet syndrome
  - Atherosclerosis
  - Thromboangiitis obliterans
- Occupations:
  - Vibration
  - Cold
- Drugs:
  - Ergotamine
  - β-Blocking agents
  - Cytotoxic agents
  - Oral contraceptives
- Miscellaneous:
  - Neoplasia
  - Neurological disorders
  - Endocrinological disorders
  - Arterial trauma
  - Arteritis
  - Other

Acrocyanosis may be difficult to distinguish from Raynaud's phenomenon, but as the management is similar the distinction may not be of critical importance. The absence of temporal fluctuation of symptoms together with the presence of oedema may assist the diagnosis.

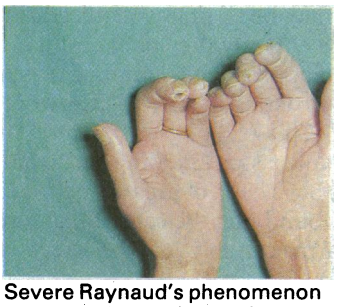
White finger syndrome is an occupational disease. Sustained exposure to vibration at given frequencies results in permanent neurological and musculoskeletal changes.

Livedo reticularis is primarily a cosmetic problem confined to the legs. The cause seems to be spasm of the cutaneous arterioles, which become more prominent when they are cold. The inadequacy of drug treatment has in the past led to treatment with lumbar sympathectomy, with mixed results.

Erythromelalgia and causalgia—Whereas pain is not a feature of Raynaud's disease, burning pain of the hands or feet suggests erythromelalgia. In many respects this is the opposite of Raynaud's, because heat provokes an attack and the patient seeks relief with cold water or by standing barefoot on a cold floor. β Blocking agents have been used to obtain symptomatic relief; carbamazepine 200 mg twice a day may also be beneficial. Burning pain is also a feature of causalgia, in which the association with previous sensory nerve damage is well established. There is often a functional element as well.

Chilblains—Patients prone to chilblains complain bitterly of the cold. This is an inflammatory condition—in chronic cases there is angiitis with intimal proliferation and a perivascular infiltrate of neutrophils and lymphocytes. Protection from the cold, anti-Raynaud's drugs, and anti-inflammatory ointments may be helpful.

Other causes of digital gangrene



Severe Raynaud's phenomenon can result in ulceration of the finger tips.

Digital ulceration or gangrene may also be caused by non-vasospastic conditions. Emboli may be discharged from the fibrillating atrium, the ventricle after a myocardial infarction, or from an aneurysm of the subclavian artery. Arteriovenous malformations in the hand can cause enough shunting of blood to result in distal ischaemia. Occasionally, thrombotic disorders such as polycythaemia or thrombocythaemia may cause digital ischaemia, but this is most unusual in the hand.

Vascutherm gloves are distributed by Camp Ltd, Worcester. We acknowledge with thanks the assistance of the audiovisual department, St Mary's Hospital, London, in the preparation of the illustrations.

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The *ABC of Vascular Diseases* has been edited by Mr John H N Wolfe.

Junior Doctors. The New Deal

Manpower and hours of work agreements are complementary

Paul D Miller

This is the second in a series of articles which explore the new deal on junior doctors' hours of work and explain how it will be implemented.

In the mid-1980s *Achieving a Balance* and *Plan for Action* aimed to address the problems of the hospital career structure.[1,2] The ratio of junior doctors to consultants was and has remained approximately three to two. This has caused severe bottlenecks. For a medical graduate to spend no more than 10 to 11 years in training followed by 25 to 30 years as a consultant the ratio needs to drop to one to two and a half.

In 1988-9 3467 doctors graduated from British medical schools; fewer than half (1577 born in the United Kingdom or Ireland) entered general practice. But between 1981 and 1989 an average of only 331 new consultant posts were created each year. The consultant retirement rate is under 600 a year, and although consultant expansion increased towards the end of the 1980s, total opportunities in 1987-8 and 1988-9 were still only approximately 1000 posts a year.

Achieving a Balance introduced the concept of career and visiting registrars. A career registrar is a United Kingdom or European Communities doctor who is entitled to pursue a career in Britain. A visiting registrar is an overseas doctor who is expected to return home after training. It was agreed that the number of career registrars and senior registrars in all specialties would be adjusted to the number required to fill the expected number of consultant opportunities. If the career registrar quota was lower than the current number of registrars any shortfall could be made up by visiting registrars. So *Achieving a Balance* does not necessarily require any reduction in the total number of registrars.

Implicit in the agreement is the acceptance that many registrar posts are not training posts but fulfil a service need which should be performed by consultants or staff grade doctors; other registrars would be more appropriately replaced by senior house officers. The quotas issued by the Joint Planning Advisory Committee (JPAC) do not hinder career prospects, but make career prospects clearer by removing the false promise of dead end posts which might previously have been designated as registrar posts. If there are only 100 consultant opportunities a year in a specialty it is wasteful to train 200 registrars each year.

Under *Achieving a Balance* appointment to a career registrar post should normally be a good indicator that there would be a consultant post available when the doctor has completed training. Just as importantly, doctors who cannot obtain career registrar posts in their chosen specialty should be able to reconsider their career options in their mid-20s when they are senior house officers and not in their late 30s when they are already senior registrars with little chance of achieving a consultant post. *Achieving a Balance* introduced 100 extra consultant posts, an early retirement scheme for consultants (which has since been extended to associate specialists), and mechanisms for converting registrar and senior registrar posts surplus to training requirements to consultant posts. The Department of Health has now issued quotas for senior registrar and registrar posts, based on the best available manpower data, to be achieved by 1992-4 for senior registrars and 2000 for registrars.[3]

On to the next step

Once agreement had been reached on *Achieving a Balance* and the necessary mechanisms for implementation had been established attention turned to the problem of the excessive hours being worked by junior doctors and the difficulties and inconveniences caused by the gradual erosion of their conditions of service, such as the deterioration of doctors' messes and on call facilities.

Despite being told that their hours had markedly improved and that onerous rotas were being phased out most junior doctors knew that this was not true. At a time when the average working week was about 38 hours it was not difficult to find juniors on one in two rotas or on one in three rotas with prospective cover working 136 hours a week. These anecdotes were supported by the Dowie report which demonstrated an increase in hours of duty between 1985 and 1987-8, with the average junior doctor being on duty for 90 hours a week.[4] These doctors are also expected to study for postgraduate qualifications and research projects which are necessary for advancing their careers. This leaves little time for families, social life, or recreational interests.

After a long campaign by the BMA's Hospital Junior Staff Committee (HJSC) and other groups including

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