RAYNAUD PHENOMENON

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The phasic colour reaction of—white, blue and red—which occurs in the digits following their exposure to cold is known as the Raynaud Phenomenon (R.P.). Its production is not difficult to understand. The pallor is due to the complete cessation of finger blood flow resulting from spasm of the digital arteries, together with the evacuation of the blood from the small skin venules by the massaging effect of movement. During the recovery phase the first trickle of blood through the relaxing vessels is avidly deoxygenated by the anoxic tissues and the finger skin becomes 'dusky' or blue. Finally, a reactive hyperaemia occurs subsequent to the tissue anoxia and causes the pink or red stage of the phenomenon. In the normal finger it is not possible to obliterate intermittently the

lumen of the digital artery by a cold-induced spasm, and the occurrence of a R.P. in a digit indicates a local or general vascular abnormality.

It is convenient for purposes of classification to divide the conditions leading to such intermittent colour changes into primary and secondary groups.

Primary (or Idiopathic) Group

This is the common form in which the inborn and often familial abnormality is limited to an unusual responsiveness of the digital artery muscle to cold. This Lewis (1932) most aptly named the 'local fault.' It is a more frequent and more severe condition in women than in men. The attacks may commence in infancy although their maximal incidence is in adolescence. The

FIG. 1.—Upper figures represent the subclavian artery. Middle and lower figures represent the digital arteries in relaxation and contraction.

tips or whole fingers of both hands are usually involved symmetrically. The attacks can be produced by placing the hands in water at 15°C. for five minutes while occluding the arterial supply to the fingers by a pneumatic tourniquet. The fingers rapidly cool to water temperature which induces a spasm of the digital vessels. On release of the tourniquet the fingers are found to be numb and white.

Although the ease with which attacks may be provoked in many individuals may cause considerable discomfort and inconvenience, nutritional lesions are limited to minor nail change, pulp wasting or recurrent finger infection. Finger gangrene, calcinosis or scleroderma probably never result from this type of phenomenon, although there is no reason why a person with such a common complaint as idiopathic R.P. cannot develop a separate disease entity.

On clinical examination there is no evidence of systemic disease and arteriography, or the reactive hyperaemia test, demonstrate patent digital vessels.

Secondary Raynaud Phenomenon

There are two essentially distinct mechanisms at play in this group. Digital colour changes may be induced by an obliterative vascular lesion reducing the blood flow to the digits to such a degree that the normal response of the digital vessels to cold is sufficient to complete the occlusion. Alternatively, changes in the arterial muscle may occur through disease or physical agents, such as high-frequency vibrations, which cause it to over-react to cold. It should be remembered, however, that these two mechanisms are not mutually exclusive and both may be present in varying proportions in any particular case (Fig. 1).

Raynaud Phenomenon due to Obliterative Vascular Disease

There are many types of occlusive arterial lesions occurring in the major and in the digital vessels. Although it is often tacitly assumed that thrombo-angitis obliterans can be readily separated from atherosclerosis by its histological and clinical characteristics, in fact the differentiation is by no means clear cut and many indeterminate cases occur. Atherosclerosis may involve major or minor arteries in either sex and its clinical appearance in the upper limb is usually preceded by evidence of chronic ischaemia in the legs or by anginal symptoms (Fig. 2). In the main it involves the middle and older age groups. In the third and fourth decades in men the obliterative arterial lesions tend to begin in the smaller peripheral vessels and ascend proximally, being frequently accompanied by episodes of superficial or deep phlebitis. Digital or limb gangrene often

FIG. 2.—Atherosclerosis arteriogram showing digital artery thromboses, sinuosity of the radial and obliteration of the ulnar arteries.

ensues. Spontaneous digital gangrene (Lewis and Pickering, 1934), external trauma as from a crutch or internal trauma from a cervical rib and embolism are less common etiological factors in this group of R.P.

In patients suffering from obliterative vascular lesions the affected extremities become markedly cyanosed on exposure to cold and the white phase of the phenomenon may be transient or absent. The digits are involved asymmetrically and evidence is constantly present of advanced nutritional lesions, such as osteoporosis, skin-ulcers or

FIG. 3.—Case of vibration tool syndrome. Note adequate filling of large and small vessels.

loss of the subcutaneous tissue particularly in the pulp spaces. Signs of chronic ischaemia will be present in the lower limb in many of the patients or there will be evidence of a local or cardiac cause for the vascular lesions.

Raynaud Phenomenon Following the Development of a 'Local Fault'

The change in the degree of response of an artery and the critical temperature at which arterial spasm occurs can be induced in a finger by trauma, prolonged cooling, exposure to vibrations or by vascular disease. The latter two causes are the most prevalent and important.

The list of occupations by which a worker may

become exposed to high-frequency vibrations has extended enormously in the last two decades. These include heavy tools, such as pneumatic drills, with a large amplitude vibration and relatively slow frequency, and the lighter high-velocity flex-driven tools used in polishing and buffing. Although the amplitude of the vibrations and their wave-form are probably important, the maximum incidence of the complicating R.P. develops in those handling tools of about 3,000 r.p.m. The duration of exposure required for the onset of symptoms may vary from months to years, and the fingers first affected are those which guide or press most against the tool. The less exposed digits follow in sequence and once the tendency to R.P. has developed little retrogression in the severity or frequency of attacks appears to take place after the patient ceases to use vibration tools. Nutritional lesions do not occur in vibration tool R.P. and the reactive hyperaemia test demonstrates normal patency in the digital arteries (Fig. 3).

The diseases which bring about a change in vessel response to cold may be conveniently grouped under the heading of 'collagen diseases.' These present a difficult clinical problem. The patient of either sex may, on first examination, appear to suffer from idiopathic R.P. with a symmetrical bilateral involvement and careful clinical examination may fail to detect any joint or connective tissue lesion although the E.S.R. may be raised and the peripheral vascular tone tends to remain high between the attacks of R.P. The reactive hyperaemia test may show an abnormal delay or patchy flush. Only after an interval of months or years does a further manifestation of the systemic disease appear with joint, tendon or skin lesions, and at this stage it may be possible to diagnose rheumatoid arthritis, scleroderma or acrosclerosis. Although digital artery thromboses cannot be demonstrated initially, these frequently appear with the progression of the disease and nutritional or ulcerative finger lesions become evident. The importance of this group lies in the necessity for instituting treatment for the collagen disease rather than the R.P., which does not respond satisfactorily to sympathectomy. In addition the stress of a surgical operation may flare-up the systemic disease and such complications as a persistent pyrexia or pericardial effusion have resulted in some of our cases.

Clinical Differentiation

It is apparent that the symptom of R.P. is one common to many diverse clinical conditions and must never be accepted as idiopathic unless a complete investigation has failed to reveal systemic disease. Idiopathic R.P. invariably involves both hands in an approximately symmetrical fashion

and its onset is in adolescence or early adult life, often associated with a history of similar attacks in the siblings. A reactive hyperaemia test should always be performed. The hand is immersed to the wrist in water at 40°C. and vascular occlusion maintained for five minutes by a pneumatic cuff at a suprasystolic pressure. At the end of this time the hand is removed from the water, dried and the fingers massaged white. On release of the cuff the pink flush should spread evenly to the tips of the digits within three to four seconds. When digital artery thromboses or narrowing has occurred or should the digital arteries fail to relax, then a delay or asymmetry of return of the flush will be evident, implying that the case is not one of the idiopathic group. An E.S.R. and blood count are routinely performed together with a radiological examination of the wrists and hands for joint change and of the shoulder girdle for the presence of a cervical rib element.

Treatment

To cure or alleviate R.P. it would be necessary either to change the response of the digital arteries to cold (the local fault) or, in cases with occlusive vascular disease, to increase the resting blood flow to the digits. We know of no way of influencing the local fault although the effects of cortisone are being explored experimentally. Some increase in the resting blood flow to the fingers, and therefore rather slower cooling, can be obtained by a sympathetic denervation or by the administration of vasodilator drugs. The hand blood flow following sympathectomy is only significantly increased for a few weeks (Walker, Lynn and Barcroft, 1950) and the digital vessels quickly regain their pre-operative tone and, in accordance with Cannon and Rosenbleuth's 'law of denervation,' may, in fact, become over-sensitive to circulating adrenaline and nor-adrenaline. Results of the operation in severe cases or where a marked local fault is present are therefore disappointing. It is not unusual to see a 'relapse' within a week of operation in the collagen group of cases. In patients with obliterative vascular disease the clinical response is more satisfactory and healing of the nutritional lesions often results. The type of sympathectomy

preferred is the anterior (Telford) approach through which the second and third thoracic ganglia are removed on both sides at the same operative session. The removal of the stellate ganglion does not improve the operative results and causes considerable distress from the resultant ptosis and nasal congestion. Removal of a cervical rib where this is the causative agent will relieve the R.P. Workers who develop vibration R.P. should abandon the use of the vibration tools before the attacks become severe.

The vasodilator drugs, many of which are of great experimental and pharmacological interest, have proved very disappointing in clinical practice. None of them act selectively on the digits or limbs and their ganglionic or adrenergic blocking properties have general effects on the cardiovascular system, the gastrointestinal tract, the pupils and secretory glands. In addition many of the drugs have multiple pharmacological properties; thus Tolazoline B.P. (Priscoline) has pilocarpine and histamine-like actions. In general, although the mild cases may benefit from Tolazoline, its administration is frequently accompanied by unpleasant side effects such as dizziness, nausea, flushing and palpitations.

Summary

1. The production of the colour changes occurring in Raynaud Phenomenon has been discussed.
2. Clinically the patients have been grouped into (a) primary or idiopathic; (b) secondary to obliterative vascular disease; (c) secondary to the development of a 'local fault.'
3. The differentiation of the above cases has been described.
4. The limitations of sympathectomy and vasodilator drugs in the treatment of Raynaud Phenomenon have been discussed.

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