RAYNAUD'S DISEASE, WITH SPECIAL REFERENCE TO THE NATURE OF THE MALADY*

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Raynaud's disease properly so-called is a condition affecting those parts of the body that are particularly influenced by exposure to cold; it is most usually a symmetrical affection of the hands, less commonly of the feet, and much less commonly of the ears and nose. In the hands, where it is most conspicuously displayed, the affection consists of attacks, usually in response to external cold, in which the skin becomes discoloured and numb. The temperature of the hands of these people is always near to that of the surrounding atmosphere when this is low.

DESCRIPTION OF TYPICAL CASE

In what may be described as typical attacks the events, as recent investigations have shown, are as follows in the hands while at rest. The digital arteries, already constricting in response to cold, become closed, and the circulation to the fingers ceases. This event passes unnoticed by the patient, and because the fingers are cold cyanosis appears very gradually and is usually but slight. When the fingers are in use they may be at this stage either faintly violaceous, or pale from passive emptying of the vessels. The arterial spasm continuing, the small vessels of the skin, which give the skin colour, also enter into actual spasm, and the fingers became completely blanched. This secondary spasm leading to blanching of the skin is not an integral part of the attack, as has been thought, but is a delayed and simple consequence of loss of circulation to the fingers; a similar spasm and blanching occurs in the minute vessels of the skin of the body generally when the circulation ceases and the body cools at death. Another delayed event is numbness of the fingers. Like the waxy pallor of the skin, it is a simple sequence to loss of circulation, the tactile nerves of the fingers losing their function after a period of half to one hour. It is the pallor, or the numbness, or both, which usually first attracts the patient's attention; and these events are usually described by them as forming, though they do not actually constitute, the first events of the attack. As the fingers begin to recover—and in other cases throughout most of the attack—they are of a deep violet tint. When blood begins to circulate again a state of reactive hyperaemia is displayed, and the fingers are bright red in colour and tingle, as during recovery from a long period of local circulatory arrest, however induced. They may or may not become warm. It is notable that in the attack the fingers show discoloration of their tips first of all, the discoloration spreading to their bases and sometimes invading the whole hand. That is so because the digital arteries at first contract to obliteration near their terminations, and the obliterative spasm spreads gradually up the vessels of the finger, subsequently to involve vessels of a corresponding order supplying the skin on the dorsum of the hand.

The attack is here described as careful recent studies have shown it usually to occur; the description differs a little from that originally given by Raynaud.

* Abstract of a lecture given at St. John's Hospital for Diseases of the Skin.

NUTRITIONAL CHANGES IN FINGERS

In patients in whom the attacks are very easily and frequently provoked small areas of necrosis frequently appear at the tips of the fingers; these separate painfully, leaving little scars—a phenomenon that is usually repeated. Massive necrosis is quite unusual. Sclerodactyly is common in the severe cases. The nutritional changes in the fingers are described quite briefly, because they concern us only in so far as they help to identify advanced cases of what is called Raynaud's disease. In the mildest cases the disease consists of vascular changes only and is but an inconvenience. In the severer cases nutrition of the tissues suffers, and the malady becomes very painful, intractable, and crippling.

RAYNAUD'S ORIGINAL THEORY

Raynaud published his thesis in 1862.¹ As is now recognized, he contributed to knowledge by isolating the form of circulatory disturbance that bears his name, and by separating a spasmodic affection of the vessels of the extremities from those due to gross obstructive disease of the arteries. Raynaud, however, also advanced a theory of causation—namely, that the loss of circulation in the affected parts happens through sympathetic nervous impulses and is due to irritability of vasomotor centres. Difficulties have appeared in the way of accepting this theory, which has been held ever since Raynaud put it forward, and recent work² has led to the belief that the essential trouble lies, not in the nervous system, but in the vessels of the part showing the disturbance of circulation. It has been proved² that, while other vessels may also become narrowed, it is spasm of the small artery of the size of the digital vessels that is responsible for the loss of circulation. The evidence for this last statement will not be given in this lecture, which will confine itself to a statement of the reasons why the vasomotor theory must be abandoned and why the theory of local vascular change must take its place.

EVIDENCE AGAINST RAYNAUD'S THEORY

The first reason for doubting Raynaud's theory was the order in which the vessels are involved. The vascular spasm begins in the tips of the fingers, as we have seen, to spread up the fingers and involve them eventually in their length, and subsequently those of the hand. When vaso-constriction is brought about through the sympathetic nervous system in animals or man, the vessels appear to contract simultaneously in all the territory involved; there is no evidence of an orderly succession—a spread from the periphery to the centre. Recent observations have shown that the vessels of the fingers in Raynaud's disease can be brought to a state of spasm by purely local cooling, and that the digital arteries can be brought into complete spasm quite locally over any short stretch of their course through the finger. The orderly manner in which the finger is involved when a patient is exposed to a cold atmosphere is due merely to the fact that the fingers cool first at their tips and later at their bases.

LOCAL AND CENTRAL ACTION

Now when a subject is exposed to a cold atmosphere the cold affects the vessels of the fingers in two ways. It acts upon them locally, and reflexly through the central nervous system. There are these two actions, and it is quite clear that, when a patient who has Raynaud's disease develops an attack on being exposed to a cold atmosphere, both these factors help to close down the vessels of the fingers. Both factors in similar circum-

stances come into action in normal people and cause arterial constriction; but in these the closure is incomplete. It is necessary for us to ascertain, if we are to understand Raynaud's disease, wherein lies the abnormality. It is not abnormal for digital vessels to contract in response to exposure of the body to a cold atmosphere; but it is abnormal for them to close completely. Theoretically we can place the fault in the nervous system or locally; but we may not assume it to be in both places in one and the same case, or even in two different cases of what we regard as the same disease. We cannot blame the nervous system in any patient unless we can show that in Raynaud's attacks the vasomotor impulses are unusually powerful. Now there is no evidence that this is so; our evidence goes to show that the vasomotor impulses do not constitute the abnormal factor. Consider the usual case of Raynaud's disease in which an attack is regularly and quickly brought on by exposure to a cold atmosphere. If the right hand of such a subject is guarded from cold by immersion in water at 30° C. the exposure of the rest of the body to suitable cold, including the left hand, produces spasm in the vessels of the left hand only. Spasm in the hand kept in water at 30° C. has never been seen; this is a strong argument in favour of the view that the nervous factor is not the abnormal one.

On the other hand, in such a case as has been described, it is almost always possible at a moderately warm room temperature to produce spasm in the vessels of one hand, and sometimes even of one finger, by plunging the corresponding part, and that only, into water at 20° C. or a little less.

ANAESTHETIZATION OF ULNAR NERVE AND SYMPATHECTOMY

These observations quite clearly suggest that Raynaud's vasomotor theory is untenable. It is proved to be untenable by observations in which the sympathetic impulses are removed. If a case of Raynaud's disease is chosen in which spasm is easily produced and is intense, then anaesthetization of the mixed nerve (for example, the ulnar) does not relieve the spasm, when this is already present, in the manner in which it undoubtedly would relieve the spasm were this due to abnormal vasomotor impulses. Anaesthetization of the ulnar nerve produces an intense and immediate vaso-dilatation of the fifth finger in normal subjects, even when that finger is cold at the time of anaesthetization. When the finger of a Raynaud's case is cold and its vessels are in spasm, anaesthetization of the nerve either produces no perceptible effect on the vessels, merely reddens the finger without warming it, or warms it after a great delay. This delayed warming is due to a slight relaxation of the spasm owing to removal of normal vaso-constrictor tone. This may be sufficient to warm the finger a little, which will itself cause further relaxation and further warming.

It is quite true that sympathectomy, by which is meant removal of the inferior cervical ganglion, relieves Raynaud's phenomenon on the corresponding side; but, as might be expected from what has been said, it does not cure it.^{2,3} In cases in which vascular spasm was originally easy to induce it can again be induced after the hand has been robbed of all sympathetic nerve supply. While these facts actually dispose of the vasomotor theory, it requires careful and precise thought before we can arrive at a proper conception of the disease and the relation of vasomotor impulses to the attacks. A very similar set of circumstances was discussed many years ago in the instance of the action of digitalis in cases of auricular fibrillation. It was found that if the ventricle is slowed by using digitalis it is quickened again by sub-

sequently using atropine. From this it was at first concluded, and incorrectly concluded, that the digitalis slowing results purely from increased vagal tone. The correct conclusion is that quickening under atropine is due, in large part or wholly, to release of the normal vagal tone—the tone present before digitalis is administered. That is the correct conclusion because the release is incomplete; atropine does not bring the rate nearly up to the same level as it does when no digitalis has been administered. When the attacks of vascular spasm in Raynaud's disease are relieved by sympathectomy the relief is also incomplete; it is due, not to removal of abnormal vasomotor impulses, but to removal of normal ones. Such relief as is seen is precisely what is to be expected if we hold—what I have no doubt is the correct view—that a local fault and not increased vasomotor tone is responsible for the disease.

STRUCTURAL CHANGES IN ARTERIES

When there is general exposure of the normal body to a cold atmosphere there is, as has been said, a double reaction: there is the direct response of the vessels of the fingers to cold, and there is a response through the central nervous system. In normal subjects these two influences together combine to produce great narrowing of the vessels of the fingers; the narrowing is so great that only a very sluggish stream of blood passes through them. Different normal subjects vary in the degree to which the vessels narrow, and the transition from normal subjects to those in whom the obstruction becomes complete—and that are properly called cases of Raynaud's disease—is not abrupt; it is quite gradual. A great many subjects whom we regard as normal narrowly escape the malady.

It is here to be pointed out that the local fault may be theoretically of either one of two kinds. It may be due to structural change in the arteries involved, or it may be due to these vessels reacting more powerfully to such a local stimulus as cold. There is evidence for believing that in the patients who present spasm that is easily induced the vessels are often actually abnormal structurally. They will not open up as widely as will normal vessels in response to heat, for example; and such anatomical evidence as we possess, though admittedly meagre, points to the presence of intimal thickening. If exposure of the body of a normal subject to a cold atmosphere produces an increase in the tone of the digital vessel almost sufficient to close its lumen, it is easy to understand that the presence of even a little intimal thickening might make just the difference between incomplete and complete closure. It is going to be a very difficult matter to separate this factor of intimal thickening from the factor of increased response to local stimuli, and to assess the relative parts played by the structural and functional element in bringing the vessel to complete obliteration in individual cases—though there is reason to believe that each factor may play its part. Given that the two factors are present, there remains another problem. The intimal thickening may be regarded as consecutive to the spasm and to diminished blood flow; or, on the other hand, early structural change in the vessel may be regarded as responsible for, or associated with, change in the reactivity of this vessel to local stimuli. These are questions of much theoretical interest, but not for the moment of practical importance.

RAYNAUD'S DISEASE A LOCAL AFFECTION OF THE ARTERIES

The important matter at the moment is to recognize that, whatever views we take of these questions, we are still left with the conclusion that Raynaud's disease is

a local affection of the arteries, and is not caused by a disturbance of the vasomotor nervous system. That is a conclusion which sooner or later must influence our management of these cases.

A curious affection appearing in recent years amongst workers with tools or machines that vibrate is of much interest to this discussion.⁴ It occurs in a number of different trades, but may be illustrated by stone-masons who use a chisel driven by compressed air. These workers develop a condition in the fingers indistinguishable from Raynaud's disease; the circulation to them is lost on ordinary exposure to cold. But what is most noteworthy is that the fingers most exposed to the vibration are those that develop this abnormality first. The example is cited as an instance of Raynaud's malady arising locally and asymmetrically out of an obvious local cause.

We may safely conclude that in Raynaud's disease the fault is purely a local one and that vasomotor tone is normal during the attack. But to explain the many interesting reactions of these patients we may not neglect the factor of vasomotor tone.

ILLUSTRATIVE REACTIONS EXPLAINED BY NEW THEORY

Perhaps a clear understanding of what is in fact not a very simple matter may be aided by citing a series of examples briefly illustrating the many reactions that may take place in different circumstances that can be defined, and by showing that all these reactions are readily explained if we assume that the fault is local and the vasomotor system normal. This view of the pathology of the disease is adequate, and it is alone adequate, in interpreting the examples given. The following instances all apply to the susceptible fingers of a case of Raynaud's malady.

1. The first example is the most familiar; it is that of the usual spontaneous attack. An attack develops on simple exposure to a room temperature of 15° C. It is in part due to increase in vasomotor tone, and in part to a local response to cold. The abnormality, however, is a local abnormality, and may consist of increased susceptibility of the vessels to cold or a facilitation of closure owing to local intimal thickening.

2. The subject's body and hands are warm in a warm room (22° C.). Vasomotor tone is equal and apparently normal, and remains so in the two hands. One hand is now placed in water at 20° C.; in this, and in this only, an attack develops. Such an attack is to be ascribed to a local fault.

3. The subject while warm is taken into a cold room (15° C.) and exposed with the right hand in water at 30° C. An attack develops in the left hand only. In this instance, although vasomotor tone is raised in both hands in response to the cooling of the body, the hand that is protected from local cold is prevented from developing an attack. The increased vasomotor tone is not abnormal, and therefore fails by itself to close the vessels.

4. The subject is exposed in a cool room and the hands are protected, being kept at a temperature (about 25° C.) that is just insufficiently low to close the vessels and to induce an attack. The subject now experiences an emotional disturbance or pain; there is a rise of vasomotor tone and an attack supervenes. In this case the vessels are already abnormally narrowed, though not occluded, and an increase of vasomotor tone (a normal increase) is now sufficient, acting on these abnormally narrowed vessels, to complete the closure.

5. It is quite possible that a normal vasomotor impulse might close a digital vessel that is at a higher temperature (say 30° C.) if that digital vessel had much intimal thickening. Here again, though an increase of vasomotor tone might be the only apparent factor, the structural

alteration of the vessel would be the real cause of the attack, because it alone would be abnormal.

6. A patient is presenting an attack in a cool room, and anaesthetization of the ulnar nerve fails to abolish it. In this case removal of normal vasomotor tone is insufficient to release spasm in vessels that are closed owing to a local abnormality.

7. A patient who has lost the sympathetic nerve supply to one arm dips both hands into water at 20° C. while in a cool room. An attack develops in both hands, but is more severe on the non-sympathectomized side. In this instance both hands present an abnormal local response to cold, but on the sympathectomized side this response is counteracted to a slight extent by loss of normal sympathetic tone. If the malady is less severe the loss of sympathetic supply (by sympathectomy) on one side may even suffice to prevent the attack developing in that hand, and ulnar anaesthesia may likewise prevent the corresponding (anaesthetized) finger from becoming cyanotic. But these facts are compatible with the theory of local causation and form quite inconclusive arguments for the vasomotor view.

8. If the body of a patient upon whom cervical sympathectomy has long since been performed on one side is kept very warm while the two hands are cooled by immersion in cold water, then the attack may be confined to the sympathectomized side.⁵ This apparent paradox is due to the fact that after sympathectomy full vasodilatation is not maintained; and so, when there is a conspicuous vaso-dilatation, as from heating the body, the vessels on the intact side become wider than those on the sympathectomized side. It leads curiously to the attack, which is clearly the result of a local cause, being confined to the limb from which the sympathetic nerves have been removed.

REFERENCES

- ¹ Raynaud: *L'Asphyxie locale*, etc., Paris, 1862.
- ² Lewis: *Heart*, 1929-31, xv, 7.
- ³ Lewis and Landis: *Ibid.*, 1929-31, xv, 329.
- ⁴ Papers by Hamilton and by Seyring in *Arch. f. Gewebepathologie und Gewerbehygiene*, 1930, i, 348, 359; and personal unpublished observations.
- ⁵ Lewis and Pickering: *Heart*, 1931, xvi, 33.

A CONSIDERATION OF BACKACHE FROM THE ORTHOPAEDIC STANDPOINT*

BY

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When a patient complains of low back pain, with or without sciatica, an eliminative diagnosis leaves us with a group of conditions which may be definitely regarded as orthopaedic in nature, the source of the difficulty lying usually in the lumbo-sacral or sacro-iliac joints. One is able to say this when physical examination reveals localized joint tenderness, muscle spasm, stiffness, and deformity referable to one or other of these articulations. In affections of the more superficial joints these features are readily discernible, but in the low back the joints are deeply seated, and a superficial examination entirely fails to bring out the salient points on which a diagnosis based upon anatomical facts and a knowledge of the statics of the body can be made.

This whole field has been a mysterious one in the past, but owing to the efforts of the Boston school of orthopaedic surgeons, headed by Dr. Joel Goldthwait, a step-by-step method of physical examination and the interpretation to be built up upon the facts elicited has

* Paper read at a clinical meeting of the Fife Branch of the British Medical Association, April 14th, 1932.