Three cases of Raynaud's disease / by Thomas Barlow.

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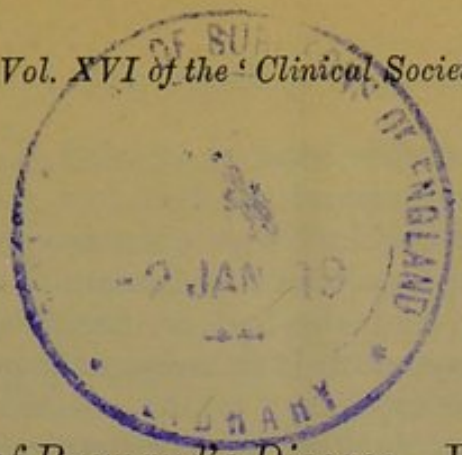

Three Cases of Raynaud's Disease. By Thomas Barlow, M.D. *Read April* 27, 1883.

I HAVE watched three cases which came within the category of Raynaud's disease, but none of them so severe as that which forms the text of Dr. Southey's paper.

I will narrate them in the order of their severity, taking the mildest case first. I must apologise for the imperfection of the notes. When I saw the first and the third cases I was in the dark as to the nature of the affection.

I. In October, 1882, my colleague, Dr. Poore, was good enough to show me, in the out-patient room, a little girl, æt. 5, who presented an appearance at that time quite new to both of us. The whole of the right foot and ankle, and the leg for a distance of three or four inches upwards, had become within a very short time cold and of a nearly uniform greyish-blue colour.

The child whimpered a little with pain in the foot, and did not like it to be handled much. Just below both elbows on the dorsal surface of the forearm there was an area of perhaps three inches in length of ill-defined blueness and coldness. This slight blue area was more marked on the right than the left side.

The first thought on looking at the foot was that there had been some arrest of arterial blood supply with subsequent venous stasis. But the child, although pale, had no cardiac disease. There was no reason for suspecting embolism into or disease of the large arterial trunks, and the smaller degree of the affection of the skin below the elbows made one willing to suppose that the condition might be temporary, and perhaps due to spasmodic contraction of vessels. The child was admitted into the ward under Dr. Fox, and I learnt that the blueness passed away within an hour or two, and in the evening and next day the child seemed perfectly well.

The day but one afterwards, about 3.30 P.M., whilst sitting

in the ward, she got another attack, which I saw. There was slight blueness and coldness of the left foot and ankle. The child whimpered a little, but was able to walk across the ward with a slight limp, and the foot became natural again in less than half an hour, and she had no further trouble. She was sent out next day, and I subsequently got a complete history of her from her mother.

The child had been suckled nearly two years; had been late in teething, and probably rickety to some extent, to which her chest, which was somewhat pigeon breasted, bore witness. She had not suffered from any of the acute specific diseases, and there had been nothing special in her history till February, 1881, when she was about three years and a half old. It was very cold weather, and the child was out of doors, but she was carried in somebody's arms, and the mother remembers that she wore worsted stockings and boots at the time. When brought into the house she complained of cold feet, and vomited some watery stuff. Her feet were quite blue, and the blueness extended for a short distance above the ankles. She was put into a warm bath, but without benefit; the blueness remained from 5 o'clock till midnight. Next day she was all right, and played as usual.

Seven months passed before she had another attack, and this was in the month of September. She was outside at the time, and as before was wearing worsted stockings. The blueness lasted two hours. She had another attack in a week's time, and several during the winter following.

Since that time her attacks have been confined to the cold weather, but they come on occasionally indoors, especially if the child sits about much. The mother has a strong belief in the value of exercise in warding off attacks. The attacks occur mostly in the afternoons, never in the nights whilst in bed. The longest duration observed has been seven hours, but many attacks pass off in less than an hour. Latterly the elbows have been affected, but the ears and nose have never suffered. Some of the attacks commence with gaping and complaint of sickness. She has never vomited except in the initial attack. She is sometimes thirsty and hungry when the attack commences. The mother has never seen anything wrong with the child's urine.

II. The second case was also that of a little girl, æt. 5, sent to me by my friend, Mr. Alford, of Haverstock Hill, who had carefully noted all her symptoms.

She was of healthy parentage; her father was said to suffer from rheumatism in the knees, and his mother from the history had had some deforming joint disease of knees and hands. This child was the third of the family of four children, the others being healthy.

She had been suckled for twelve months, and with the exception of what was called "congestion of brain from teething" at sixteen months, had been a healthy child. She had had whooping-cough and measles when one year old, and had got through them well. She had never had ague, nor had her parents suffered from it.

In September, 1881, being then about three years and a half old, she had her first attack of coldness and blueness with pain, affecting one foot and lasting for several hours. Very soon after this attack commenced the child passed some very dark urine. She had another similar attack in a few days, and then repeated attacks during the winter until the month of April, 1882. There were sometimes two or three during the week. The duration was not generally more than three hours. The attacks were much more liable to appear on going outside, but occurred indoors also. They never occurred in the early morning nor during the night, but were most common about midday. The pain always preceded the coldness and blueness. The child did not pass dark urine with every attack, and never more than once with each attack.

During the first winter, along with the above symptoms, she complained on some occasions of pain in her stomach, and in one attack the left hand became cold and blue up to the wrist. Her last attack of the first winter was on May 6. She was then free till September, 1882. I saw her in February, 1883. Up to that time she had sometimes gone fourteen days without any attack, and at other times she had had two in the day.

On February 15, when I saw her, the day after an attack, her feet were cooler than the rest of the body, but there was nothing else noteworthy. She was a sensitive, intelligent child, not robust looking, rather thin, but not unduly pale, and without any sign whatever of visceral disease. Some of the dark urine which she had passed the day before was brought for examination. It was acid in reaction, and gave on boiling about one tenth of albumen, which came down in granular form and was unaltered by the addition of nitric acid. There was a deep blue reaction with tincture of guaia-

cum and turpentine. The deposit showed, however, under the microscope, no blood-corpuscles, but fine brown granular stuff and crystals of oxalate of lime.

During the week which followed she had two attacks, one of which lasted two hours, affecting the left foot, and one an hour and a half, affecting both feet. After the first attack the urine was black but not after the second.

March 15.—For about eight days she had had an attack daily mostly in the left foot, but on some days in both. She was suffering from an attack when I saw her. Both her feet were blue and cold as high as the ankles. On this occasion her urine had a copious deposit of lithates, but gave no reaction with the guaiacum test and was free from albumen. Her temperature was 99·6° in the axilla. The skin, excepting that of the feet, felt natural. The heart sounds were natural; the spleen could not be felt; her tongue was clean. Some subsequent specimens of urine gave characters similar to the first. On one occasion there was a deposit ef phosphates. In some attacks I learnt that the stomachache before referred to sometimes preceded and sometimes succeeded by an interval of two hours the pain and coldness of the feet. The dark urine did not always follow attacks in which stomachache had been complained of, and sometimes appeared after attacks in which there was no stomachache. I was not able to elicit that the child had any other referred visceral pain, as, for example, in the loin. Her stomachache she referred to below the tip of the ensiform cartilage. The mother had come to the conclusion that the more widely diffused attacks, viz. those affecting both feet and a hand, preceded or succeeded by stomachache, were less severe in duration and pain than those which affected one foot only; and I think it comes out from my notes that these latter attacks were more frequently followed by dark urine than the former, but this requires further investigation.

The last attack of the present season was on May 10.

III. John P., a lock maker, æt. 42, came to me at the end of January, 1883, complaining of pains in the feet. He stated that he had been a healthy man before his present complaint. There was nothing in his previous history to suggest either rheumatism, gout, syphilis, or ague.

In the month of April, 1880, whilst at Darlington, he began to suffer from a series of attacks of sharp pain in the

heel of the left foot which he found extremely cold. The pain and coldness would last for twenty minutes or more and then pass off, and afterwards the feet would burn and feel hot to the touch. Subsequently the toes and the other foot became affected. The instep was much less affected than the toes and heels.

He was away from work for three weeks and less liable to the attacks when in bed. The doctor told him at first it was rheumatism and subsequently neuralgia. Neither then nor at any time since has he had any joint affection along with the attacks.

After about three weeks he was able to go to his work again, but found himself often taken with these attacks in the street. He became quite helpless in them sometimes, so that he was obliged to sit down until his feet "came to" again. He sometimes found that he could keep off the attacks by sharp walking, but directly he stood still or sat down, an attack would come on, and when fairly established he was quite unable to walk it off. It is quite clear that when the warm weather came the attacks diminished in frequency and severity, but according to his statement he has been very few days entirely free from attacks since April, 1880.

During the second winter (that is 1880-81) the attacks affected the right foot more than the left, and there was much more blueness of the toes, and he began to have blue patches on his thighs. He suffered most in the month of January, and this has been the case during the present winter. He has been many days unable to go to his work, and although he is decidedly better in bed, he is not absolutely free even there from slight attacks. Besides the patches on the thighs he has also had similar patches on the buttocks, and his fingers are nearly always cold but do not become blue in the same way as the toes. One month ago he found that there was a sore place at the end of the second and third toes of the left foot.

He has had no trouble with his eyesight and never seen anything wrong with his urine. The patient was rather a spare, ill-nourished man. He had no sign of visceral disease that I could make out and he was not specially anæmic. His toes were all greyish-blue and cold, and at the extreme tip of the second and third toes of the left foot there was a small circumscribed sore from which there had evidently been superficial loss of substance. These sores were not like ordinary chilblains, the skin round was not swollen or shiny.

On the outer side of each foot the skin was bluish and on the outer side of the middle third of the right thigh there was a bluish area about the size of two crown pieces. At a subsequent period a similar patch was seen on the left thigh and others on each buttock. These patches it is difficult to describe. The skin was not raised over them, and at the time when I felt them, they were not colder than the adjacent parts of the limb. They were uniform in colour and the tint was intermediate between that of a patch of fading erythema and that of one of the large ecchymoses sometimes seen along with purpura. The appearance was such as could be imagined to be obtained by lightly staining the deeper layers of the skin with some hæmatoxylon. The patches were permanent for some weeks and were the situations where the patient explained to me that he felt at various times cold and tingling.

The only other point about this patient was that his left calf was a little smaller and perhaps more flabby than his right. This made one think of the possibility of some obscure spinal cord mischief, but there was nothing else to suggest it, and the patient assured me that this difference in the calves had been noticed by other doctors for a considerable time, but that he had not suffered inconvenience from it.

I examined the patient's urine on two occasions and found it normal. He attended for about six weeks, I fear with little benefit. When the warm weather came his attacks again declined in severity.

Remarks.—The general subject has been thoroughly discussed by Dr. Southey in the *St. Bartholomew's Hospital Reports,* and a remarkable case was also shown by him at a recent meeting of the Pathological Society. Moreover, an example of what was probably a severe manifestation of this disease, was recorded by Mr. Thomas Smith in the *Clinical Society's Transactions* for 1880, under the title of "Spontaneous Gangrene of Thumb and Finger. Also Mr. Hutchinson has shown and described cases of the severe form in which limited gangrene occurred. It would, therefore, be unseemly for me to enter upon any general observations on the subject. But there are a few points to which I should like to refer in regard to what may be called the mechanism of the disease, its alliances, its nomenclature, and its therapeutics.

(1) As to what may be called the mechanism of the disease, it is of course impossible to dogmatise; but the first case in

Raynaud's last papers* seems to establish that what really takes place is a spasm of arterioles. In this case, after characteristic attacks involving the extremities had recurred during two or three months, the patient began to suffer from ocular troubles. During the attacks the eyesight was good, but on their subsidence his vision became, as he expressed it, "troubled and confused," clearing up again with the next attack. This was more marked in the left than the right eye. During such an interval between two attacks it was found on ophthalmoscopic examination that there was considerable narrowing of the lumen of the central artery of the retina and its primary branches, and that partial momentary "strangulations" could be seen at times. There was very marked pulsation of the retinal veins. During the occurrence of an attack of local asphyxia of the limbs, ophthalmoscopic examination did not reveal complete subsidence of the above phenomena, but they were very much lessened in amount. The ultimate recovery of the patient from the limb affection was attended by complete disappearance of the ocular troubles. It seems quite clear that the eye affection and the local asphyxia of the extremities were allied in their nature and mode of production. Raynaud half regrets in his comments that in this case the affections of limbs and of the eyes were not simultaneous, but this appears to me a matter of little importance. It is, indeed, most interesting to note, in the history of the second case that I have recorded, suggestions of the progressive march of spasmodic arteriole contraction to different tracts of the body.

It is also, I think, important to note the varying intensity and distribution of different attacks in the same individual, and that although the bilateral symmetry on which Raynaud has insisted seems often to obtain, it is by no means invariable, and when it occurs is not necessarily equal in amount.

The last development of Raynaud's doctrine, if I may summarise his own words, is that there is a peripheral excitation, most commonly consisting in an impression produced by change of temperature on the cutaneous nerves, and that whilst in the normal state either very low temperatures or exposure for a long period are necessary for the production of more or less analogous effects, in these individuals an insignificant difference is sufficient; further, that the peripheral stimulus affects that part of the grey matter of the cord which

* "Local Asphyxia of Extremities," *Archives Générales de Médecine*, 1874, vol. i, pp. 5, 189.

presides over the vaso-motor innervation, and that a great exaggeration of the irritability of that part of the cord must be assumed.

Now given the initial slight peripheral stimulus there seems no reason why the central disturbance should not radiate and become manifest in several different regions successively instead of simultaneously, and this brings me to consider (2) the alliances of the disease.

I think everybody who has watched these cases, and also those cases of paroxysmal hæmatinuria in which no evidence of ague can be obtained, must have been struck with certain points of resemblance. They are not in a true sense periodic, but they are both paroxysmal. Attacks in both affections have a remarkable relation to changes of temperature. By far the greater number of cases of both are exclusively winter or cold-weather affections, and if not exclusively they are primarily so, and if the attacks do not vanish they notably diminish when the warm weather appears. It is worthy of note that the paroxysms in Cases I and II never started in the night when the patients were in bed, and the same obtains with regard to paroxysmal hæmatinuria.

In both the paroxysms may begin with yawning or with vomiting, and the extremities, as I can testify, may in the onset of an attack of paroxysmal hæmatinuria become extremely cold and blue. It is also, I think, important when comparing the latter affection to an ague fit to remember that there is nothing corresponding to the sweating stage, and although I can recall one marked example of hot stage, I believe that this is not at all a constant feature.

I am not so foolish as to say that Raynaud's disease and paroxysmal hæmatinuria are the same disease, but only that they are allied diseases, and it is of great interest to note that in my second case many of the attacks combined the features of the two diseases.

I believe that Dr. Wilks has referred orally to his having a case under his care with a similar combination, and Dr. Southey has also referred to it.

It will be worthy of investigation in cases of Raynaud's disease whether any other visceral paroxysmal affection can be ascertained, like the splenic enlargement which often occurs with the hæmatinuria cases, and whether also some of the cases of temporary ocular ischæmia, which are known to ophthalmologists, may not be found to present local asphyxia elsewhere.

With respect to antecedent diseases, in none of the three cases which I have described could ague be supposed to have anything to do with the affection. There was in these cases no question of syphilis, but in a remarkable case which was under the care of my friend Dr. Henry Humphreys, of St. Leonards, and which progressed to the stage of extensive symmetrical superficial gangrene of both lower limbs, there were unquestionable signs of inherited syphilis. Dr. Humphreys has told me that he had no reason to correlate the two affections.

The President has asked whether in these cases there were any arthritic signs. In my cases there was nothing definite. I have an adult patient under observation who suffers from cold extremities, and whose fingers, toes, ears, and nose are always a little bluish, and he also suffers from slowly progressive symmetrical contraction of most of his finger-joints; but as he has never had any paroxysmal attacks his case ought not to be included in this group.

Raynaud in one of his cases refers to plastic thickening along some of the tendons of the hands, which thickening cleared up entirely when the attacks of local asphyxia ceased.

Mr. Hutchinson has asked the question whether some of the cases of end-joint arthritis which he figures in his *Illustrations of Clinical Surgery* may not be allied to those of local asphyxia. This will doubtless receive further investigation.

I think it would be productive of confusion if Raynaud's cases were mixed up with cases of chilblains. The subjects of Raynaud's disease are not, I believe, the subjects of chilblains in the ordinary sense. When they get sores at the extremities the sores are, if one may so say, definitive in character, and due to actual gangrene of greater or less extent, sometimes, indeed, going to the length of a digit.

The essential clinical note of Raynaud's disease, at all events primarily, is the paroxysmal character of the circulatory disturbance, and this I apprehend does not apply to those who are the subjects of ordinary chilblains.

(3) With regard to nomenclature, it appears to me that the great difficulty is to get a term which will include cases which differ much in their distribution and intensity, but which are essentially the same. Raynaud admits that his original term, "local asphyxia," is not a good one, and there are obvious objections to the term symmetrical gangrene in the fact that many of the cases do not go on to gangrene, and also, that when gangrene occurs it is not always symmetrical. I

doubt whether any better term than Raynaud's disease (which has already obtained a partial currency) will be found.

(4) As to therapeutics, it would be presumptious for me to offer more than suggestions. I will only mention that I found that a patient of mine, who was the subject of paroxysmal hæmatinuria, and who had been from childhood always washed with very hot water, was greatly improved by using first hot then cold water, and ultimately only cold water. At all events, she became able, even on a snowy day in winter to walk a considerable distance without getting an attack.

Applying this to my second case of Raynaud's disease, during an attack I used friction with cold water, and her mother was willing to admit that the attack sooner came to an end than had formerly been the case.

Raynaud has, in his last paper, recorded some cases which seem to show the benefit of the descending constant current down the spine, and this, I think, one certainly ought to test in any future opportunity, as well as local galvanism to the extremities in which the asphyxia first starts.

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