Henry (F. P.)

Clinical Report of Two Cases of Raynaud's Disease.

BY

FREDERICK P. HENRY, M.D.,

OF PHILADELPHIA.

presented by the author

FROM

THE AMERICAN JOURNAL OF THE MEDICAL SCIENCES.

JULY, 1894.

Reprinted from The American Journal of the Medical Sciences, July, 1894.

CLINICAL REPORT OF TWO CASES OF RAYNAUD'S DISEASE.[1]

By Frederick P. Henry, M.D.,
of Philadelphia.

Case I.[2]—Patrick M., aged fifty-five years, native of Ireland, laborer, was admitted to the Philadelphia Hospital, November 11, 1893.

Family history. The patient's father died at sixty-five, of asthma, and mother at seventy-five, of "old age." Two brothers have died—one at thirty-five, of unknown cause, and the other at twenty-two, of smallpox. Three sisters died in childhood, of measles and smallpox. Two brothers still living and healthy. He knows of no hereditary disease in his family.

Previous history. He had smallpox and measles in infancy, malarial fever at eight years of age, doubtful left-sided pleurisy twelve years ago. Has used alcoholic liquors in moderate amount, averaging from two to three glasses of beer daily, but would occasionally drink to excess. On the other hand, months would sometimes pass without any indulgence in either beer or spirits. The patient has never had any kind of venereal disease. For the past six winters he has been subject to a severe cough, which has regularly caused the loss of a few pounds in weight. During the spring and summer the cough disappeared and the loss of weight was regained. The summer of 1893 was an exception to this rule. In January, 1893, he acquired a "cold," which has been with him ever since. The cough occurs in paroxysms most severe in the morning, and is attended with a yellowish, scanty, and tenacious expectoration. There has been neither hæmoptysis nor night-sweats. In August, 1893, the patient noticed that his urine was of a deep-red color during the forenoon. This bloody appearance of the urinary secretion was present almost continuously for about one month, but varied somewhat in intensity. One morning in October, 1893, after the patient had been much exposed to cold, he noticed that the helix of both ears was of a deep-blue color. This discoloration continued about two hours, during which the ears felt cold and were painful. Since the last-mentioned date these attacks of local asphyxia have recurred on each exposure to cold. At first the signs of circulatory disturbance were more marked in the right ear, but later the left ear became the more involved, and about December 15th a small portion of the border of its helix became gradually converted into a dry, black sphacelus.

December 25, 1893. The tip of the nose became cyanotic early in the morning on exposure to cold, and has presented the same appearance nearly every morning since. On January 20, 1894, the second and third fingers of the right hand as far as the metacarpo-phalangeal joints

[1] Read at the meeting of the Association of American Physicians, Washington, May 29, 1894.

[2] For the notes of this case I am indebted to Dr. Louis Préfontaine, resident physician.

became deeply cyanosed on exposure to cold in the morning. This "local asphyxia" has extended to the remaining fingers and to the first phalanx of the thumb of the same hand, but did not at first involve the left hand.

The blood was examined by the resident physician, Dr. Préfontaine, on February 9, 1894. Number of red corpuscles, 2,900,000; hæmoglobin, 43 per cent; white corpuscles not increased in number. No tubercle bacilli could be detected in the sputum.

February 10, 1894. The patient was exposed for a few minutes to the morning air in order to make him more fully illustrative of a clinical lecture on Raynaud's disease which I delivered at the Philadelphia Hospital at 10 o'clock. The experiment was highly successful, the helices of both ears, the tip of the nose, the fingers and pulp of thumb of the right hand, and the three first fingers of the left hand becoming deeply cyanotic and remaining in the same condition for nearly an hour. At 3.30 the same afternoon he passed four ounces of urine of a deep cherry-red or reddish-brown color, which reacted imperfectly to the guaiacum test for blood pigment.

11*th*. Cyanosis of ears and nose.

12*th*. Cyanosis of ears and nose. At 9.30 voided three ounces of reddish-brown urine, which did not respond to Gmelin's bile test, but gave the reaction to Mahomed's test, and contained a few red blood-corpuscles. In the afternoon the urine was of normal appearance, but contained about 7 per cent. of albumin, roughly estimated. Its specific gravity was 1021.

13*th*. Urine still darker than that of yesterday morning, reacting similarly, and containing a very few red corpuscles. It also contained a few short, rather broad hyaline casts stained with pigment. Albumin, 35 per cent. by volume. The afternoon urine was perfectly normal in appearance and gave similar reaction; it was free from casts and contained about 9 per cent. of albumin; specific gravity, 1023. The cyanosis this morning was comparatively slight.

14*th*, 9 A.M. Urine brownish-red, but not so dark as yesterday, acid, no bile reaction, no blood-cells or casts; contains a few large, flat epithelial cells; albumin abundant, apparently 30 per cent. by volume; quantity voided too small for specific gravity test; free from sugar. A specimen sent to Prof. Marshall, of the University of Pennsylvania, at 12 o'clock was of a faint brownish-red hue; much lighter than in the morning; no blood-cells in specimen; did not respond to bile test; free from casts; specific gravity, 1030; reaction acid; albumin, 15 per cent. by volume; free from sugar. 2.30 P.M. Urine amber colored; no sediment; specific gravity, 1027; acid; a trace of albumin; no bile or sugar present; no casts found under the microscope.

15*th*. Cyanosis slight and limited to ears and nose. The urine passed at 7.30 A.M. was of normal appearance, free from sediment, of specific gravity 1020, and contained no albumin, casts, or blood corpuscles. It was acid and free from sugar. 11 A.M. Urine brownish-red, precisely like that passed at 12 o'clock the day before; sediment slight; specific gravity, 1026; acid; contains 15 per cent. by volume of albumin; free from casts, blood-cells, and sugar. A specimen sent to Dr. Marshall. 3.30 P.M. Urine of a cherry-red tint; acid; specific gravity, 1025; distinct ring with Heller's test; no cells or casts. 7.45 P.M. Urine normal in all respects; specific gravity, 1021.

16*th*, 9.30 A.M. Urine of same color as specimen voided yesterday at 11 A.M., and similar in all other respects. 12.30 P.M. Urine darker than at 9.30; acid; specific gravity, 1027; free from blood-cells and casts; contains about 12 per cent. by volume of albumin. 3.40 P.M. Urine entirely normal in appearance; free from albumin, casts, and cells; specific gravity, 1022. This morning the cyanosis was slightly marked on the nose and ears and absent from the fingers. It persisted, however, longer than usual, being as well marked at 5 P.M. as at any time during the morning. The weather was quite cold. To day Dr. Marshall sent the following report: "The spectroscope fails to show the presence of blood-coloring matter in Patrick M.'s urine."

17*th*, 7.30 A.M. Urine normal. 10 A.M. Urine of light-brownish tint; acid; specific gravity, 1022; no reaction with Mahomed's test; no casts; trace of albumin. Cyanosis is very slight to-day, affecting mostly the left ear, point of nose, and right ear, and absent from the hands. 4 P.M. Urine normal in all respects.

18*th*. Urine shows no change to-day; cyanosis slightly less than yesterday and in same areas.

19*th*. Urine normal in color and free from casts, cells, and albumin, although the cyanosis was more pronounced than yesterday in the ears and nose, and was perceptible in the fingers for a short period in the morning between 8.30 and 8.50. On the patient's rubbing his hands briskly it disappeared.

21*st*. The patient was allowed to go out on a pass for forty-eight hours. On his return he reported that on the morning of the 22d, which was very cold, his toes up to the metatarso-phalangeal joints were deeply cyanotic, as well as his nose and ears. His urine also was red during the two mornings of his absence from the hospital.

24*th*. A cold morning, but the patient keeps himself warm near the heater, so that the cyanosis is but moderate and limited to the nose and ears. The urine is normal in color, acid, of specific gravity 1023, and contains neither albumin, casts, nor blood-cells.

The urine was examined almost daily up to March 16th, when the patient left the hospital. It is unnecessary to give the details of these examinations, since they are precisely similar to those above recorded. The cyanosis was of daily recurrence, but varied greatly in severity and extent; on the other hand, the albuminuria was markedly intermittent, being invariably present when the urine was of a deep brownish red hue, and greatly diminished or absent when it approached or reached the normal color. The contradictory results of the examination of the urine for blood-coloring matter are inexplicable. The gross appearances of the secretion during the paroxysms of "local asphyxia" were precisely those of hæmaturia or hæmoglobinuria, and yet no absorption bands were perceived on spectroscopic examination. On the other hand, on several occasions the reaction of blood was obtained with the guaiacum test. As is well known, the late Dr. Mahomed, of London, claimed that this test would show the presence of hæmoglobin when the spectroscope failed to detect it. He stated, however, that it was not applicable to urine which responded to the ordinary tests for albumin. In the case of Patrick M. the spectroscope failed to detect blood-coloring matter, and yet in the same specimen, which was highly albuminous, the guaiacum reaction for blood was distinct. On February 23d I took a

specimen of the urine to Prof. Henry Leffmann, who kindly examined it and sent me the following report:

"I made several experiments to-day with that sample, and got the guaiacum reaction for blood-coloring matter very plainly. I rendered the liquid acid by acetic acid, which, of course, caused much effervescence and produced a flocculent precipitate. This precipitate gave the blood reaction very strongly. I found that neither reagent alone nor when mixed, without adding some of the sample, gave any color."

Dr. Leffmann's examination of the urine with the spectroscope gave negative results.

A specimen was also examined by Dr. Cattell, assistant pathologist to the Philadelphia Hospital, who reported that it reacted in thirty seconds to Almen's test for blood-coloring matter.

When I went on duty at the Philadelphia Hospital on February 1st I found Patrick M. in a ward assigned to phthisical patients. It has already been mentioned that tubercle bacilli were not found in his sputum. I examined his chest with the utmost care, and found neither dulness on percussion nor any auscultatory sign of consolidation or cavity. Sonorous râles were audible all over the chest, and were more marked during expiration, which was prolonged. The thoracic expansion amounted to only one inch and a half, the chest measuring at the height of a forced inspiration thirty-six inches, and at the end of a forced expiration thirty-four and one-half inches. There was a distinct history of attacks of paroxysmal dyspnœa for several years past, and taking this into consideration with the physical signs just mentioned, I concluded that the condition of the lung was one of moderate emphysema, dependent upon asthma and associated with chronic bronchitis. This history of previous asthmatic attacks which, I repeat, is confirmed by existing signs of emphysema, is extremely interesting, inasmuch as it demonstrates a tendency to spasm of unstriated muscle in other tissues besides the arterial.

The heart, it should be stated, showed no signs of disease.

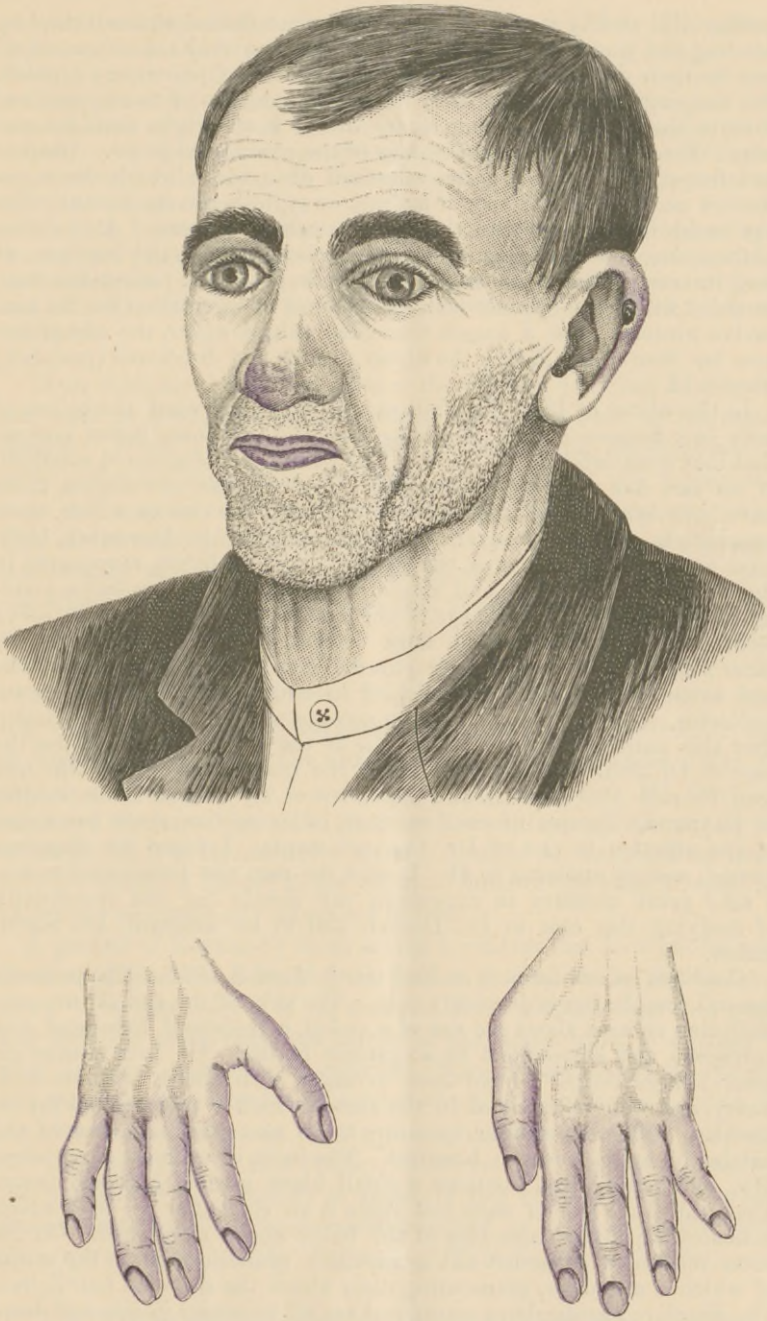
On March 16th the patient left the hospital and gave a false address, so that my efforts to keep him under observation were thwarted. A few days before he left, the necrosed portion of tissue dropped from his ear, leaving a clean-cut semilunar loss of substance in the helix.

On April 13th, he came to my office and reported that he had been very poorly since leaving the hospital, especially during the recent cold and damp weather. The left ear was of a dark-purple color, but contained no gangrenous patches. The nose, right ear, and hands were of normal appearance. I examined his blood with the following result:

Number of red corpuscles per cubic millimetre, 3,166,666; hæmoglobin, 60 per cent; white corpuscles not increased in number. The patient passed about two ounces of turbid, deep reddish-yellow urine, which was acid, of specific gravity 1030, and contained a trace of albumin (Heller's test). It was free from sugar. This sample was examined for blood-coloring matter by Dr. Leffmann, but with negative results.

Case II.[1]—Elizabeth R., aged seventy-seven years, born in Maryland, a widow, formerly occupied with indoor and outdoor farm work. Her

[1] From notes taken by Dr. Lucas, resident physician.



CASE I.—Patrick M., showing cyanosis (local asphyxia) of hands, nose, and ears, and gangrenous patch in left ear.

*

mother died at fifty years of age of some acute disease characterized by flooding and vomiting. Two sisters died from the results of miscarriages; two brothers died of pleurisy, and two children of pulmonary phthisis. She had excellent health in early life. Since the age of twenty has had chronic rheumatism, recurring every winter in the right shoulder and arm. There are nodosities in certain of the phalangeal joints. She has had frequent attacks of gastro-intestinal disturbance which she terms cholera morbus. No history of malaria or syphilis can be elicited. She has never been addicted to the use of alcoholic liquors. At the time of the menopause she had a severe uterine hemorrhage, and has since, at long intervals, been troubled with metrorrhagia. The patient has been troubled with local symmetrical disturbances of circulation for the past twelve winters. For a longer time than this, however, the end of her nose has been rough during the winter months and discolored (purplish) when cold.

In the winter of 1881–82, while exposed to a cold wind, the helices of both ears became painful, and when she reached home it was noticed that they were swollen and black. This painful and discolored condition of the ears has recurred each winter. For the past two winters there have been symmetrical areas of cyanosis upon the cheeks, which, upon exposure to cold, become very blue and painful. In December, 1893, after an attack of influenza, the signs of local asphyxia reappeared in the old areas of invasion, and similar disturbances of circulation manifested themselves on the backs of the hands, the wrists, and the lower forearms. At the same time large elongated bullæ appeared on the inner side of each foot, running parallel with the first metatarsal bone and extending the greater portion of its length, and one on the outer malleolus of the left foot. It was on account of the last-mentioned lesion that this patient was admitted to one of the surgical wards under the care of Dr. John B. Deaver. One of the resident physicians who had seen Patrick M. (Case I.), and who knew of my interest in the subject of Raynaud's disease, informed me that, in his opinion, there was a case of the affection in one of Dr. Deaver's wards. I found his diagnosis correct, and on applying to Dr. Deaver the case was transferred to me. I take great pleasure in expressing my thanks for the opportunity of studying this case to Dr. Deaver, and to his assistant, Dr. Kleinstuber.

Condition on admission to medical ward, March 2, 1894. The patient's general complexion is decidedly pale. The skin of the end of the nose, including an area about the size of a nickel, is thickened, indurated, and verrucous, and surrounded by a cyanotic border. On both cheeks are several large copper-colored spots arranged with almost absolute symmetry, and well represented in the sketch which I pass around for inspection. Precisely similar spots are to be seen on the dorsum of the hands, the wrists, and the forearms. The helix of each ear is swollen, blue, and tender, and contains a small black area of necrosed tissue. Upon the inner side of each foot there is an elongated red spot which indicates the former situation of the bullæ above mentioned. On the outer malleolus of the left ankle there is a cyanotic area, in the centre of which is a shallow, granulating ulcer about the size of a half-dollar. The discolored areas above mentioned are all increased in size and deepened in color in the early morning when the temperature of the ward is comparatively low. The urine has shown nothing abnormal, although

repeatedly examined for albumin, sugar, casts, and blood. The patient, however, claims to have passed urine at times resembling milk, at other times as dark as coffee. Careful physical examination has revealed nothing abnormal with lungs, heart, liver, or other internal organ. An examination of the blood showed the number of red corpuscles to be normal, and their percentage of hæmoglobin to be 75. They presented no noticeable alterations of size or shape. The white cells were not increased in number.

May 17. During the last month fresh gangrenous spots have developed in the ears (there are now three in each) and there are complaints of pain when the temperature falls but a few degrees. Both ears are more cyanotic than formerly. In other respects the patient's condition is unchanged.

These cases are typical examples of the affection described by Maurice Raynaud in 1862,[1] and since classified under the head of Raynaud's disease. When clinically complete, the affection is characterized by great pallor and coldness of peripheral parts, such as the fingers, toes, nose, and ears, on exposure to cold, followed by intense venous congestion of the same parts, and eventually, if the circulatory disturbances continue, by well-defined areas of dry gangrene. These are the essential clinical features of the disease, but, in addition, hæmoglobinuria, as in Case I., and scleroderma, as in Case II., have been observed in a few cases. Raynaud's thesis contains a report of twenty-five cases, of which twenty were of the female sex. The disease is most prevalent in early life, the great majority of Raynaud's cases occurring between the ages of eighteen and thirty. The same observer noted five cases of the disease in children between three and nine years of age. He, therefore, suggested the term "juvenile gangrene" as an appropriate one for the affection, but it is objectionable because it might be supposed to imply that the disease is limited to the young. Both of the cases I report are of individuals decidedly advanced in life.

A perusal of Raynaud's thesis makes it evident that he regarded gangrene as an essential feature of the disease, at least in its full development. The first chapter of his treatise, which contains forty-four pages and occupies more than one-fourth of the whole work, is devoted to what he terms spontaneous gangrene.[2] Nevertheless, among the twenty-five cases which it contains there are several in which the process did not eventuate in gangrene, but stopped at the stage of "local syncope," or "local asphyxia." It is evident, therefore, that the minor grades of the disease, the *petit mal*, so to speak, of this arterial spasm, to which renewed attention has been recently called by S. Solis-Cohen,[3] were thoroughly recognized by Raynaud. This is shown, besides, more

[1] De l'Asphyxie locale et de la Gangrène symmétrique des Extrémités. Paris, 1862.
[2] It is headed "Quelques Considérations sur la Gangrène spontanée."
[3] "Vasomotor Ataxia," AMER. JOURN. OF THE MEDICAL SCIENCES, February, 1894.

explicitly in the introductory remarks of his thesis, in which occurs the following:

"My ambition would be rather to demonstrate that certain facts of gangrene of the extremities which one meets at long intervals in practice, and of which the strange appearance is apt to disconcert the most skilful, are in reality much less singular than one would be tempted to believe, and can be connected by intermediate steps with other facts much more common, and which only escape attention by their every-day occurrence."[1]

The chief theories of the cause of Raynaud's disease are: (1) that it is due to an endarteritis obliterans; (2) that it is due to peripheral neuritis; (3) that it is the result of vascular spasm. The last is the theory of Raynaud himself.

The early age of many of the patients, the intermittent character of the symptoms, and the great rarity of the disease as compared with endarteritis, suffice to exclude the latter as a causative factor in its production. There can, I think, be no doubt that cases of symmetrical gangrene occasionally reported as examples of Raynaud's disease have an anatomical basis in endarteritis obliterans, but they are in my opinion spurious. A case of the kind referred to is reported by George W. Jacoby,[2] of New York, and is highly interesting from several points of view. The patient, a male, aged forty-two years, first applied for treatment on account of numbness and coldness of the fingers (local syncope). This was followed by local asphyxia of the same parts, and subsequently by gangrene, the entire third phalanx of the left medius eventually sloughing away. Later, the signs of interstitial nephritis (albuminuria, urine of low specific gravity, casts, hyaline and granular, and hypertrophy of the left ventricle) appeared, and about four years after he was first attacked the patient died in an apoplectic seizure. In this interesting case it is probable, as Jacoby states, that the primary lesion was an arterio-capillary fibrosis, which gradually invaded the vascular system, attacking the vessels of the kidney later than those of the hands.

Jacoby gives the notes of another case in which the asphyxia was limited to the hands, and in which a cure was effected by a prolonged course of anti-syphilitic treatment. The cause of the condition was, presumably, a syphilitic arteritis.

There is little to support the theory that peripheral neuritis is the cause of Raynaud's disease. In the first place, the rarity of the latter and the frequency of the former militate strongly against it. Secondly, the cases in which neuritis has been found post-mortem may be examples of mere coincidence, or, more probably, the neuritis was second-

[1] Barlow's translation of Raynaud's Thesis. New Sydenham Soc., 1888, vol. cxxi.
[2] New York Medical Journal, Feb. 7, 1891.

ary to the local circulatory disturbance. A degree of asphyxia sufficient in its severe forms to produce gangrene might, in its lighter grades, readily lead to degenerative and inflammatory changes in the nerves of the affected part.

The theory of arteriole spasm is certainly the one that is most in accordance with the clinical phenomena. The disease is most prevalent in females and in the young—*i. e.*, in those whose vasomotor system is most impressible. It occurs in paroxysms which are caused by the surest exciter of vascular spasm—cold. Finally, in several cases during the paroxysm there has been dimness of vision, which was shown by the ophthalmoscope to depend upon a contraction of the central artery of the retina and its branches, and in one recently reported by H. M. Thomas[1] the attacks of local syncope were followed by a chill, loss of consciousness, and convulsions. Such facts are in the highest degree corroborative of Raynaud's view, that the disease known by his name is due to an "enormous exaggeration of the excito-motor energy of the gray parts of the spinal cord which control the vasomotor innervation."

The symptom, hæmoglobinuria, occasionally observed is best explained by the theory of vascular spasm. The origin of the hæmoglobinuria may be twofold: it may be due to excretion of hæmoglobin that has been separated from the red corpuscles in the peripheral asphyxiated parts—nose, ears, and fingers—or it may be due to an asphyxia of the renal or other internal vessels. I incline to the latter view as most applicable to Case I., and for the reason that the hæmoglobinuria was the first symptom in the case. It will be recalled that the deep-red color of the urine attracted the patient's attention in the *summer* of 1893, about two months before the peripheral asphyxia was observed.

I freely acknowledge, however, that the facts are in favor of the view that in the majority of cases in which hæmoglobinuria is observed this symptom is dependent upon an excretion of hæmoglobin which has been separated from the red corpuscles in the peripheral asphyxiated parts. Hæmoglobinuria coinciding with hæmoglobinæmia of the blood of the asphyxiated fingers has been frequently observed.[2] On the other hand, hæmoglobinæmia may exist, and probably often does, without hæmoglobinuria. An instance of the latter kind is reported by Taylor and Colman.[3] The patient was a girl, ten years of age, who was subject to attacks of dead finger (local syncope) affecting the distal phalangeal joints. "Local asphyxia" was never observed. In blood withdrawn

[1] Johns Hopkins Hospital Reports, vol. ii.

[2] For example, by A. T. Myers: Trans. Clin. Soc. London, 1885. Boas: Deutsch. Archiv für klin. Med., 1883. Bristowe and Copeman: Trans. Med. Soc. London, 1889.

[3] Trans. Clin. Soc. London, 1890.

from the finger during an attack the liquor sanguinis was distinctly colored, while the red corpuscles were shrivelled and irregular in shape and many of them devoid of hæmoglobin. Neither hæmoglobinuria nor albuminuria was ever present. I regret that a more thorough examination of the blood taken from the fingers during the stage of asphyxia was not made in the first of my cases. It was examined but upon one occasion and nothing abnormal was detected. It is probable that hæmoglobinæmia is frequently present without hæmoglobinuria. The spleen and liver are capable of storing up large quantities of hæmoglobin; in fact, according to Ponfick, it is not until the separated hæmoglobin amounts to one-sixtieth of the total amount in the blood that hæmoglobinuria occurs. Until the hæmoglobinæmia has reached this degree, the attack, so far as the urine is concerned, is only manifested by albuminuria. It seems to me probable that paroxysmal albuminuria in other affections than Raynaud's disease may be but a symptom of a comparatively mild grade of hæmoglobinæmia.

The relation between Raynaud's disease and chilblains has been recently studied by Legroux,[1] and is worthy of further investigation. As this writer tersely remarks, chilblains do not come at will,[2] a predisposition is essential to their production, and he raises the question whether chilblains, local asphyxia, and symmetrical gangrene may not represent different degrees of a necropathic dystrophy, of which the most striking example is furnished by syringomyelia.

To those who have seen at least a single instance of Weir Mitchell's disease (erythomelalgia) it seems scarcely credible that it should have been confounded with the affection of Raynaud. Nevertheless, this has been done. I will content myself with stating here that the differences between them are radical.[3]

I have said nothing about the treatment of these cases because no systematic treatment was pursued in either of them. With the exception mentioned in the report of Case I., they were protected from cold and nourished as well as circumstances permitted. It was expected that with the approach of milder weather their condition would improve, and this expectation was fulfilled. It was my intention to test in one of the cases the effect of the electrical bath as recommended by Barlow and R. Glasgow Patteson,[4] and in the other that of the continuous administration of glonoin in small oft-repeated doses.

My report is nothing more than it pretends to be—a clinical one. I have made no attempt to analyze the numerous recorded cases, still less

[1] Annales de Dermatologie et de Syphiligraphie, 1892, tome iii.

[2] "N'a pas des engelures qui voudra."

[3] For the differential diagnosis between these diseases the reader is referred to Dr. Weir Mitchell's article on "Erythromelalgia," in the Medical News for August 19, 1893.

[4] Dublin Journ. Med. Sci., vol. xciii.

to separate the spurious among them from the genuine. Raynaud's disease is a paroxysmal affection, and, therefore, cases in which peripheral gangrene has followed injury of the part affected or has gradually supervened upon an obliterating endarteritis, as well as those in which the stages of local syncope and local asphyxia have been absent, are, to say the least, to be regarded with suspicion.