

T H E S I S

NOTES OF A CURIOUS CASE

showing

SIMILARITY OF PURPURA AND REYNAUD'S DISEASE

by

JOHN LITHGOW, M.B., C.M.

Glasgow,

September 1898.

ProQuest Number:27552948

All rights reserved

INFORMATION TO ALL USERS

The quality of this reproduction is dependent upon the quality of the copy submitted.

In the unlikely event that the author did not send a complete manuscript and there are missing pages, these will be noted. Also, if material had to be removed, a note will indicate the deletion.



ProQuest 27552948

Published by ProQuest LLC (2019). Copyright of the Dissertation is held by the Author.

All rights reserved.

This work is protected against unauthorized copying under Title 17, United States Code
Microform Edition © ProQuest LLC.

ProQuest LLC.
789 East Eisenhower Parkway
P.O. Box 1346
Ann Arbor, MI 48106 – 1346

Mrs. McI. aged 26, residing at Omoa Square, married had one child now aged 18 months which is still being nursed. She came from Ireland 6 months ago.

On Monday 1st February 1897 she felt sick and drowsy, complained of a sore throat and felt so unwell that she was compelled to keep her bed for two days. Feeling a little better, but still complaining she rose on the third day and washed. She was menstruating at this time. She was very much fatigued after washing, and her attention was drawn to her legs by feeling pain in them. On examination she found numerous spots about the size of a pin's head on her legs, which during the next two days increased in size, and she became very ill with vomiting and purging.

A doctor was called and some medicine prescribed. For the next two or three days vomiting and purging continued with increasing weakness. The doctor of the Works was called, and he, to check these symptoms, painted a blister over her stomach. Next day she

observed a large red spot on her left cheek, this she attributed to a drop of the blistering fluid falling on it. Two days after this she observed spots on her arms similar to those on her legs.

I was called to see her on Saturday evening, 13th February, and the thirteenth day of her illness. I found her in a very low state due to weakness of the heart, the sounds being almost inaudible, her condition being such as to make it unadvisable for me to do more than make a very superficial examination. I immediately prescribed:-

R.

Ferri et Ammon Citrat. *grs viii*
 Spt. Ammon. Aromat. *ʒj*

every 3 or 4 hours.

Next day she was stronger, but it was not till Monday the 15th that she was in a state to allow a thorough examination to be made. The state of matters I then found was:-

Patient was fairly well nourished but sallow and ill looking. Pulse 120, Temperature normal.

Tongue red and raw looking, slightly pimpled. Over the left cheek was a deep red round mark about half an inch in diameter. On the left eyelid there was a similar mark. On the nose and forehead were a number of smaller ones. Both shoulders were thickly covered with these spots which were here of much smaller size, few of them being larger than a split pea, but very numerous, almost touching each other. This description also applies to the outer surface of the arms, down to the elbows. On the forearms there were a few similar spots. The dorsal aspect of both hands was pretty thickly spotted with them.

The body was almost entirely free of them.

On both buttocks and outer aspects of the hips they were very numerous, fading away as they neared the knee.

In the popliteal space they were larger in size. From the knee downwards they were about the size of a split pea and very numerous. The soles of the feet were one large patch.

The bowels had moved on the previous day and were described as being very loose and black in colour.

Previous History: Five years ago patient was confined to bed for 14 weeks with rheumatism. There was no history of syphilis, nor appearance from the state of her glands, etc., of her having suffered from it.

For some time previous her husband had been out of employment, consequently she had suffered great privations, her diet being largely composed of potatoes.

A closer inspection of these spots showed them to be blood stains underneath, or into the skin. The margins were abrupt, only very few running into each other, as a rule the margins were free and unconnected, though where they were most numerous there was scarcely a hair's breadth between them. In colour they were dark red. They did not disappear on pressure, nor were they painful on pressure. They were not raised above the surface. In size the majority of them were about the size of a split pea. The larger ones were similar to the smaller except as regards their size. The one on the left cheek showed a distinct inflammatory areola round it.

The soles of the feet appeared as if they had been beaten with a stick and no trace of individual circles could be seen, it was a general ecchymosis.

On 16th I observed the pillow stained with blood which had come from the mouth, and examination showed a little blood still oozing from the inside of the cheeks, and lips. The gums were red and swollen. On this day also a pretty profuse watery exudation appeared from the feet. So profuse was this exudation that the bed was quite wet all round them. The patient was now prescribed Lime Juice Cordial ad. lib., and a lotion of Plumbi Sub Acet. as an astringent wash for the feet, this lotion in a few days checked the exudation.

From this till the 21st there was no new symptom to report. Temperature keeping normal, pulse still extremely feeble, the spots, especially the larger ones, had gradually been turning darker in colour, while the smaller stains had almost faded away. Bowels had once or twice been loose and still black (probably now from the Fe. in the mixture), but were readily controlled with Pulv. Doveri. She had vomited on several

occasions, and no blood was observed in the vomited matter.

She felt so well on the 22nd that she sat up for a few minutes. On the 23rd I found her worse, her temperature had gone up to 100.5° , and there now appeared in certain places where the spots were largest, and the staining remained, indications of ulcerations commencing. There also appeared a new crop of smaller spots similar to the first crop, this second crop was more marked on the legs, which also were a little oedematous.

Next day, 24th, I again made a thorough examination and found temperature normal, pulse very feeble, bowels had been loose the previous night, the motions still being dark in colour. She was coughing a good deal but nothing abnormal could be heard in the lungs. She was unable to move hands or feet and these were painful to touch or move. All the spots on the face and eyelids had disappeared, a raised scab was over the site of the largest spot on the cheek. A few small darkish shot like stains were on the shoulder

and outer surfaces of both upper arms. On the left arm, near the elbow, on the outer aspect, was a deep circular sore about one inch in circumference with a black dry shrivelled slough in the centre; also a similar sore on the point of the elbow. On most of the fingers, and knuckles of left hand were small abrasions as if there had been small blisters underneath the epidermis, and these blisters had been rubbed and broken, there were a few stains, similar to those on the shoulder on the outer aspect of the fore arm. On the point of the right elbow there was a deep sore about one and a half inches in circumference. At the end of the first phalynx on the little finger there was a blister containing dark fluid, while at the end of the metacarpal bone of the fore-finger there was a white blister. There were a few dark red shot like stains all down the right arm similar to those on the left arm.

There was nothing on the body.

Both hips had three or four small sores on them, none of these were large or deep.

Both popliteal spaces contained each a large,

deep sore, with internal slough as in the elbows.

On both legs were stains which were brighter in colour than those of the arms, and appeared to be of more recent date; these stains were also larger in size than those on the arm. Over both external malleoli were sores, which had not yet broken down. Both legs and feet were oedematous, slightly pitting on pressure. On the soles of both feet were several large, white, irregular blisters.

A more minute examination of these sores showed each (with the exception of the small ones on the hips), to be about one quarter of an inch deep, with abrupt and clean cut circular margins. No inflammatory areola round them; each containing a small dry round slough. Save the absence of the inflammatory areola the sores were very much the same as I have seen in severe small burns before the dead tissue was thrown off.

The sores on the malleoli showed how these ulcers were formed. Surrounding these sores (on malleoli) there was a very slight inflammatory blush which enclosed a zone containing white fluid inside which

was a black spot of dead tissue.

Treatment now was:-

R.

Ferri et Amm. Cit. *grs VIII*

Tr. Digit. *mxx*

Spt. Chlorof. *mxxx*

On the 25th temperature was 101° , there was slight bleeding from the nose. The urine was carefully examined for albumen, and blood, with negative results. On the 26th specimens of her blood was drawn from her arm into an apparatus sent by The Clinical Research Association Limited, London, and forwarded to them for examination, also on this day treatment was altered to:-

Tabloids of Supra Renal Substance. Grs. V.
One three times a day.

Temperature registered 101° . On 27th a new and profuse crop of spots appeared, principally on the front of both thighs and legs. She complained of having had diarrhoea during the night. Both legs

and feet were still a little painful to touch and oedematous.

She was not seen by me on 28th. On 1st March there were dark stains where the last crop of spots had appeared. The fluid had now escaped from the sores on the malleoli, and these were now similar to the others - sharp, clean-cut margin, with dry, shrivelled black slough in the centre. Temperature was 100° and reported a little diarrhoea during the night.

On 2nd March temperature was 99.5° and save that the swelling had now disappeared from the legs there was nothing else to report.

On 3rd March temperature was 99° , she complained of some diarrhoea during the night.

On 4th March temperature had risen to 100° . She complained today of abdominal pain and flatulence; also of vomiting bilious matter.

On 5th temperature had fallen to 98.8° . She still complained of great abdominal pain. The tongue is coated.

On 6th March temperature had again risen to 101° .

The tongue was very much furred and dryish. She still complained of pain in the abdomen, to control which I gave her some Tr. Opii. The sores on the elbows had now thrown off their slough and appeared deep, clean, and healing. The small ones on the hips were also clean and healing like. That on the left popliteal space still held its slough, while that on the right had thrown it off.

Of the sores on the malleoli, that over the right still contained its slough while the left was clean and fresh. On the feet the blisters were still unbroken and now appeared as if filled with water tinged with blood. On the inside of the left great toe there is a white blister while on the inside of the right great toe there is a red patch. There is no oedema nor spots. She complains of diarrhoea and passing slime with the motions.

On 7th March temperature registered 102° . Diarrhoea had now ceased, also abdominal pain. There is a little oedema of the feet. The heart sounds are much stronger and she feels much better altogether. The tongue has also cleaned. On 8th March temperature

was 101^o, has had no more diarrhoea nor abdominal pain, tongue now quite clean. There is marked oedema of hands and face and very slight oedema of feet. Examined her urine today with negative results.

On 9th there is still a little oedema on hands and feet. Temperature this day was 101^o. From this date till the 17th the temperature registered normal (see temperature chart at end).

On the 18th there was a slight rise in temperature and patient complained of abdominal pain and slight diarrhoea.

On the 19th March my attention was for the first time drawn to her back, there I found a very deep ulcer in the centre of her back just inside the nates; this ulcer was fully an inch in depth, the bone being seen, in size it was little more than the size of a quill, like the others there was complete absence of inflammatory areola. This ulcer may have been formed for some time, and from the stage it was in I am of opinion it was formed at the same time as the others.

On 22nd she again complained of abdominal pain

and temperature stood at 99°.

On 27th March pains which were of a rheumatic nature were complained of in all her limbs, and temperature was again slightly raised.

On 31st the temperature was up to 100.8°, the rheumatic pains were now principally confined to the shoulders.

On 1st April the abdominal pain returned and temperature kept up for a few days.

On 17th April she reported having menstruated (this function having been in abeyance during the whole course of the illness except as was reported on the first day she complained - see page 7 of report), and complained of severe abdominal pain.

On 18th a new and extensive crop of spots almost similar to first crop appeared (see pages 3 and of report). On inquiry I found she had, unknown to me, neglected taking the tabloids for four days. These were immediately commenced again and the spots disappeared in a few days.

There was nothing now of importance to report about the case. She had no more abdominal pain,

diarrhoea nor spots, and slowly regained her strength.

The ulcers were a long time before they were closed over, they were dressed daily with carbolic lotion, when healed over the cicatrices were all adherent indicating the sores to have penetrated near to the bone.

She was kept on the tabloids for some weeks, and ultimately became as strong as ever she had been, after which she returned to Ireland.

Perhaps it would be as well before making any remarks about the case to give a copy of the report received from T. Gouland Hopkins of the Clinical Research Association Limited London.

" At the same time I think
 "they (the Association) ought to have impressed
 "upon you more fully the fact that the quantity
 "was altogether too small for any investigation
 "worthy of the name. The quantity used for the
 "counting of corpuscles is available for this
 "purpose and really for nothing else. Even as
 "regards a microscopic examination of the in-
 "dividual corpuscles the solution sent with the

"Gowar's apparatus while efficient as a pre-
"servative for the purpose of examination is
"not sufficient to maintain any of the minute
"changes which may occur in disease nor to dif-
"ferentiate the varieties of leucocytes.

"The following, however, was my reason
"for so long a delay. In the specimen sub-
"mitted the red corpuscles while present in
"normal or nearly normal numbers (4,500,000
"per C.M.M.) showed a peculiar tendency to a
"granulation of the haemoglobin. I thought
"I had seen this before in specimens sent by
"post as yours was, and believed it to be ac-
"cidental. But as I expected just then to be
"able to get at a case of Purpura I thought it
"well to make some comparative observations be-
"fore reporting. This case did not become
"available, however, and I waited some weeks for
"another.

"I found no kindred appearance, however,
"even after prolonged standing in the fluid.
"On the other hand normal blood of which several

"specimens were tried, always gave negative results.

"Whether the appearance seen in your case was really special to it or only accidental I am unable to say.

"What one found was that in five out of every six of the corpuscles the pigment seemed broken up within the stroma, so that a network of fine lines was seen dividing the separate particles.

"The white corpuscles were in quite normal proportion to the red and so far as can be seen in the Gowar's fluid were of normal forms.

"For anything to be done towards a research into the pathology of purpura from a clinical point of view at least 5 or 6 ounces of blood would be necessary."

Synopsis of Report

Woman aged 26, who has suffered some privations, living mostly on potatoes, complains of sick-

ness, sore throat, drowsiness, commences to menstruate, then haemorrhagic spots appear, almost entirely confined to the extensor surface of the body. Has vomiting and purging motions being black in colour, probably mixed with blood, bleeding from the mouth and gums with extreme weakness of heart, no blood in urine, with relapses or recrudescence of spots, first, three weeks after first symptom, second, four days after first relapse. Also about three weeks after first symptom there appears round sloughs symmetrically situated in places where there is little or no muscle, viz. on elbows, inside of knees, malleoli, fingers, and centre of back. The local gangrenous spot being thrown off in a few days leaving a deep ulcer slow to heal. A slightly raised temperature; severe abdominal pain complained of during the whole course of the illness possibly worse after appearance of ulcers. No relapses after taking Supra-Renal Substance tabloids till these were omitted for a few days, when the spots immediately re-appeared and at the same time re-appearance of the menstrual flow. The spots soon disappear on

patient again taking the tabloids. Appearance of granulation of haemoglobin, 5 out of every 6 of the corpuscles having the pigment broken up within the stroma a network of fine lines dividing each separate particle. Complete recovery of patient.

Remarks on Symptoms

The dominant features in this case are (a) The spots, (b) The Sloughs, (c) Abdominal Pain, (d) Changes in the blood.

(a) The spots were undoubtedly due to the escape of a very small quantity of blood from the capillaries, judging from the appearance of each spot in that it was small, round, and distinct. I am inclined to think that they were due to rupture of the capillaries rather than to diapedesis. Each spot underwent the changes we expect in subcutaneous hemorrhages during the process of absorption, being bright red at first, gradually darkening till at last they appear as a stain. They seemed to be almost

directly under the epidermis, this is borne out by the fact of the blood escaping from the mouth where the membrane is tender and easily ruptured.

The distribution of the haemorrhagic spots requires special notice. They were not distributed all over the surface of the body in a haphazard manner, but showed a pretty regular, symmetrical distribution, the extensor surfaces of the limbs being the parts principally affected e.g. the shoulders, the back of the arms, the hips, and front of legs. Had the patient folded arms then assumed a crouching posture nearly all the spots were on the parts of the body which would now be visible; while the parts flexed (or not exposed), would have few, if any, spots. They were about equally distributed on each side of the body and showed affinity for the extensor surfaces.

(b) The sloughs were, to my mind, the most remarkable feature in the whole course of the illness. The sore on the face was, no doubt, due to a drop of the blistering fluid as it had no slough and healed up in a few days by scab.

It was not till the third week of the illness and immediately after the second crop of haemorrhagic spots that these sloughs were observed; when first observed the fluid portion of most of them had escaped, leaving the dry gangrene spot in the centre. These sloughs did not seem to be a continuation of the haemorrhagic spots, they were very much larger than these spots. The largest of the spots being considerably less than the size of a threepenny piece while the sloughs were about the size of a one shilling piece. The sloughs were circular with no irregularity or ragged edges as would indicate the coalescence of two or more of the haemorrhagic spots to form one slough; also the sloughs were very much deeper than the spots.

On the whole appearances indicated that these sloughs were not a further stage of development of the haemorrhagic spots that these later appeared and disappeared as such.

The sloughs were over joints, and principally over those joints which were covered by skin, and subcutaneous tissue, with little or no muscle clothing

them, they penetrated to the bone, which could be readily felt with a probe. Though the sloughs were over the joints they were not caused by pressure, and were absent from the positions where bed sores form; even the one on the back was quite free from pressure, it being inside of and kept off the bed by the nates. Besides not being in the positions where pressure is applied by persons lying in bed they were much less in size than generally met with in bed sores; there was complete absence of the inflammatory areola which accompanies bed sores, and bed sores are never so faultlessly round as these sloughs were.

Before the necrosed parts were thrown off, as a slough they were hard, black, and shrivelled, indicating that the cause of the death of the part had been dry gangrene; these round spots had in some manner been deprived of their proper blood supply, had become necrosed without any fluid in them and so shrivelled up and were sloughed off.

Another peculiarity of the gangrene was its symmetrical arrangement, we have the same parts on both

sides of the body similarly affected, with one in the middle line of the back.

The ulcers left were very slow in healing and the cicatrices are now adherent.

The sloughs were, to sum up, round, deep, symmetrical, due to dry necrosis and over joints or in the neighbourhood of joints.

(c) Abdominal pain was complained of more or less from the first and after some time was the most urgent symptom. What the cause of it at first was would be difficult to say, but taking into account the fact that the illness commenced with haemorrhage from the vagina (which may have been pathological or physiological), then haemorrhage into or underneath the skin, subsequently from the mouth, the probability is that the abdominal pain was partly due to haemorrhage into the mucous membrane of the intestinal tract, or into some other internal cavity or organ. About the time that the sloughs were being thrown off we find from the report indication of gastric disturbances viz. diarrhoea, flatulence, abdominal pain, pain

on pressure, bilious vomiting and coated tongue, also increased temperature which continues for two or three days, while further on in the history of the case she complains of passing slime with her motions.

Allowing for errors of observation or what else might have been seen had she been constantly under skilled observation I think these symptoms are clear indications of some serious abdominal lesions being present, possibly about the same time as the sloughs were thrown off on the external surface of the body, a similar process was taking place internally. It will be pointed out at the end that this abdominal pain was also partly due to a neurosis.

(d) Unfortunately there can be little more said about the changes in the blood than is comprised in the report from the Clinical Research Association; whether the granular appearance was accidental, due to the process of collecting and forwarding, or a specialty of this case, it seems impossible to say. No doubt too little blood had been collected, but owing to the weak state of the patient it was impos-

sible to collect more. Had I been able to collect from 5 to 6 ounces it would have enabled a thorough chemical investigation to have been made.

It must also be borne in mind that it was not till the 26th day of her illness that the blood was collected.

Remarks on Treatment

Owing to the debilitated condition of the patient treatment was at first stimulant. The nursing was of the poorest. The folks were extremely poor and in a poor locality. Early in her trouble the neighbours diagnosed her disease to be of a terrible and foul nature which they would be sure to catch if they went about her. Consequently the nursing was left to her husband, who had to remain at home from his work to do so.

After she had rallied a little her symptoms not now being so urgent, and when the symmetrical dis-

tribution of the spots became so conspicuous not only in the first attack but in the following one I thought of trying something else. The report from the Clinical Research Association was not received till some time after the patient had quite recovered, consequently I had nothing to divert my attention from the conclusions I had arrived at, viz. that this was due to some disturbance in some central organ and that central organ had control over the blood vessels. This being so and knowing that the sympathetic nerves are the nerves which supply the blood vessels I thought the Supra-Renal Body Substance might be tried and some of Burroughs Wellcome & Co's Tabloids of that gland was obtained. The result and degree of success has been shown. The third crop was making its appearance when first administered, this crop was the third in 27 days. No more crops appeared till she left off taking the tabloids about three weeks afterwards when immediately a new crop with symptoms similar to the first (except the ulcers), made its appearance.

The result would at any rate justify the Supra Renal Body Substance being tried in a similar case.

Discussion on Similarity to other Diseases and Diagnoses.

The diseases which this case simulates are I think comprised in the following list (a) Syphilis, (b) Scurvy, (c) ~~Raynaud's~~ Disease, (d) Purpura Haemorrhagica.

(a) Syphilis may simulate almost any organic disease and consequently in almost all organic diseases we have first to satisfy ourselves as to the presence or absence of the syphilitic element. When we have a case such as this one which shows a few of the characteristics peculiar to syphilis, then we must make a more careful study of the case in order to exclude entirely the possibility of there being a syphilitic tint. This resembles syphilis in that it is a skin eruption which does not itch, comes out

in successive crops, is symmetrical, has ulcers which are round and symmetrical, the lesions are polymorphous (stains and ulcers), there are also joint pains.

On the other hand there are many symptoms awaiting, the strictest enquiries could elicit no syphilitic history, there was no history of contagion, no sore throat, no falling out of the hair, the cervical glands were not enlarged, and a healthy child had been born. She had had no miscarriages.

There being thus altogether an entire absence of any of the positive symptoms of syphilis we are justified in saying this case is undoubtedly not syphilitic.

(b) Scurvy - Of all the diseases mentioned scurvy is perhaps the one which this disease least simulated in its whole course, still amongst the earlier symptoms there was much as is described by Bristowe page 615 under Symptoms of Scurvy. There was the sallow complexion, the rheumatic pains, the petechial spots appearing first on the

lower extremities, the extravasations as seen on the soles of the feet, the soft and bleeding gums, dropsy of lower extremities and extreme feebleness of the heart. These symptoms were enough when diagnosing the case to direct the attention to scurvy and it looked very much like scurvy developing; so much so to my mind that I immediately put her on lime juice.

The following paragraph quoted from Bristowe 619 on treatment of scurvy at once shelves this idea. "The provision enforced in emigrant ships, "and which has been found effectual in preventing "the occurrence of scurvy is that each person must "have weekly, at least eight ounces of preserved "potatoes, and three ounces of other preserved "vegetables (carrots, onions, turnips, celery or "mint) besides pickles and three ounces of Lime "Juice."

According to page 4 of this report the patient's diet had largely been composed of potatoes. This, I consider, enough of itself to exclude scurvy without discussing the other symptoms.

(c) In the year 1862 Dr. Maurice Reynaud published 25 cases under the name of Local Asphyxia and Symmetrical gangrene of the extremities. Similar affections are now known by the name of Reynaud's disease, and as the case reported by me has a conspicuous symptom in common with Reynaud's disease as described by Bristowe I will give short extracts from that author more directly bearing on that symptom. He says (page 614) "Sometimes portions of the area, in which the arterial circulation is in abeyance, actually die. The tips of one or more fingers or toes, portions of the nose or ears, small area of the general surface, or even whole of certain digits, or the hand, or foot, blacken, shrivel, and become gangrenous and after a longer or shorter period are thrown off. Not unfrequently the rash becomes the seat of an eruption which has a close resemblance to chilblains. This is apt to occur about the knuckles, and elsewhere on the hands or feet, and on the ears and nose. It may also occur elsewhere. . . . Sometimes blebs

"containing a sanguinolent fluid rise upon the affected parts and are followed by intractable sores."

In Reynaud's description there is no mention made of what has now become to be recognised as an essential symptom of the disease viz. Paroxysmal Haematuria.

"Indeed there is strong reason for believing that Paroxysmal Haematuria, even when existing apart from the usual superficial phenomena of arterial spasms, is due to spasmodic contraction of the renal arteries, and is, therefore, a variety of Reynaud's Disease." (Bristowe page 616).

In comparing, we have, in a moderately severe case of Reynaud's disease gangrene of the extremities - gangrene of those parts where bone, cartilage or tendon is covered by skin, and devoid of muscular tissue, with little or no fat; for example fingers, toes, ears and nose. In the note of my case the gangrene occurs over joints and principally over those joints which are covered by skin, and subcutaneous tissue with little or no muscle clothing them

(page 20) also blebs containing sanguinolent fluid (page //).

There is no other symptom in the two cases which can be compared. In the one case there is absence of haematuria, while in the other there are no purpuric spots, still the presence in both cases of symmetrical gangrene in anatomically similar positions warrant us in placing them under the same category, this similarity will be further referred to when I discuss the pathology.

Purpura - This case resembles and up to a certain point is undoubtedly purpura. Purpura is a disease which seems recently to have attracted the attention of several workers.

The term conveys an indefinite meaning, we have "purpura" in fevers, in debilitating diseases, liver diseases etc.

The symptoms are various and according as a symptom is described a new subdivision is formed to include cases having this newly described symptom; this subdivision bearing the name of the discoverer,

or the symptom discovered, so giving rise to a long list of different forms of purpura, thus we have:-

- 1 Purpura Simplex.
- 2 Purpura Haemorrhagica or Morbus Maculosus - Werlhofii.
- 3 Henoch's Disease.
- 4 Peliosis or Purpura Rheumatica
- 5 Willans Purpura Urticans
- 6 Purpura Fulminans

all of which, though having the principal symptom, have individual peculiarities, which according to their discoverer warrant a special term.

The pathology is obscure, opinion seems to be divided whether the blood or the blood vessels are at fault.

With Purpura included by all the above terms, save the most malignant, my case has much in common.

It is my intention at present to accept these varieties above enumerated, give a short description of the characteristics features of each with report

of cases as far as available.

1. Purpura Simplex, the simplest form, is perhaps the most common and include those cases, whose principal and almost only symptom is subcutaneous extravasations of blood in the form of petechia, independent of any general specific disease or local mischief.

For a good example of this the following may be cited.

Tom F. aged 6, bright and fairly well nourished, parents in poor circumstances. Had measles and chicken-pox. While undressing him his mother observed a few small purple spots on upper part of thighs and abdomen. Next day the spots were more numerous, none of them were larger than a split pea, they were not elevated above the level of the skin, nor did they disappear on pressure, and they were pretty sharply defined at their margins. They were fairly evenly distributed about the thighs, abdomen and chest, and there were a few about the ankles. Temperature was normal, pulse about 78, no vomiting, bowels slightly loose. The child was put to bed much against his will as he did not feel ill, and

ordered Parrish's Syrup and a milk diet. In three or four days the petechia had all disappeared and the patient was allowed to get up. Next day a fresh crop of spots had appeared, rather less numerous than the first. This also quickly disappeared and there was no return. At no time did the child complain of pain and there was no epistaxis, melaena, haematemesis, nor haematuria, nor was any cardiac murmur audible (Dr. Angus, Bingley).

For my purpose I would ask that the even distribution of the petechia be noted.

2. *Purpura Haemorrhagica* is the term by which the more severe form is known. In it there are the *Purpura* spots with haemorrhage from one or other of the mucous surfaces, and more or less constitutional disturbance. There is always a tendency in these cases (more pronounced in proportion to their severity), for haemorrhages to take place from the mucous surfaces. Thus there may be epistaxis, bleeding from the gums, or other parts within the mouth, haemoptysis, or bleeding from the stomach, or bowels, kidneys, or

other parts of urinary tract, uterus, or vagina (Bristowe 612).

The case for illustration I saw several years ago and as I kept no notes of it I quote it briefly from memory.

A lad about 10 years old when first seen by me had a purpuric rash principally on the limbs; he had also pretty profuse epistaxis and bleeding from the gums. He was suffering from constitutional disturbance, and was rather weak when I saw him, though not bad enough to take to his bed of his own accord. He had no heart murmur. The ultimate result of the case I do not know.

3. Henoch's Disease is characterised by abdominal pain, being the urgent symptom. There are the purpuric spots, the haemorrhages from the mucous membranes and severe pain in the abdomen, and usually pain in the limbs.

The following is a description by Henoch. "A boy 15 years old had a slight attack of jaundice, the result of gastro-duodenal catarrh. A few

"days afterwards there developed pain in the joints
"of the fingers of both hands without any swelling.
"After a few days an extensive purpura came on affect-
"ing the skin of the abdomen and of the upper parts of
"the thighs, and very soon this was accompanied by
"violent intestinal symptoms, intense colic, vomit-
"ing and black motions. The pain became very severe
"preventing sleep, and was accompanied by great tender-
"ness of the abdomen in the region of the transverse
"colon, which was also distended and on percussion gave
"a loud tympanic note, moderate fever was present never
"exceeding 101° Fah. After five days these symptoms
"disappeared but after a three days' interval a re-
"lapse followed which ran a similar course to the first
"attack, with extensive purpura, green vomiting, severe
"colic, fever, and after a week convalescence. The
"symptoms returned a third time in the next week: The
"stools during the attack always contained blood, being
"either quite black, as in melaena, or orange yellow
"mixed with more or less clots."

4. Peliosis or Purpura Rheumatica includes those cases where a rheumatic element is either a symptom

or a coincidence. Fresh developments of articular pains and of haemorrhagic spots appear together and there may be haemorrhages elsewhere as well, it is regarded by some as a mere coincidence of purpura with a rheumatic attack (Finlayson's Manuel 165). I saw a case recently which would be classed under this heading. A little girl who previously had had rheumatics, but had been quite well for some time, complained of pains in the joints, principally elbows, knees, and ankles; on the following day a purpuric rash had developed. There was some constitutional disturbance, the temperature being slightly raised for a few days. The rash which was of short duration was almost confined to the front of the legs below the knee and the outside of arms. She was confined to bed and the rash disappeared in about three days, the rheumatic pains being complained of for about ten days. There was no loss of blood from mucous surfaces, nor heart murmur. She made a good recovery.

5. Willan's Purpura Urticans is an affection which Willans included under Diseases of the Skin as

he did purpura and other rashes, - Scarlet fever, Measles, etc. under the term exanthema. It bears a strong resemblance to a form of purpura the characteristic feature of which is round oval wheals and is described by Bristowe (page 612, 613) as follows. "A form of purpura is occasionally met with in which "the eruption (occupying mainly the extremities and "coming out in crops at irregular intervals), con- "sist in the first instance of round or oval rosy "wheals, the colour of which does not fade on pressure, "and remains after the subsidence of the wheals until "the matter on which it depends is removed by absorp- "tion. The spots rarely present the dark almost "black colour of ordinary petechiae. The symptoms "are apt to be continued for several weeks or even "months, and in some cases which have been under our "care comprised occasional haematuria. The affec- "tion agrees in some respect with Willans' Purpura "Urticans."

6. Purpura Fulminans is the most malignant form in which purpura can appear. The malignancy is so striking that I would be inclined to accept the

view expressed by Dr. Angus viz., that "we have to do with an intense septicaemia." The illustrating case is from Starr's Disease of Children p. 401.

L.M. female aged 19. Had always lived in most affluent circumstances, had never been sick except from slight anaemia during the past two years. Father when a boy would bleed from slight causes, no further haemophilic history. March 7th, 1 a.m. slight chill without rise in temperature, very nervous and anxious, 12.45 p.m. marked chill, fever rising to 103.5° and epistaxis becoming more and more profuse in spite of every effort to check it.

March 8th, 1 p.m. temperature 98.4° , pulse 130 irregular and weak, marked pallor of skin, prostration profound. Complete mental apathy though her mind was clear when she was aroused: new purpuric spots appearing. Gums normal no evidence of endocarditis nor of any other appreciable disease. Spleen enlarged, epistaxis still continuing, the blood being dark and not coagulating. Profuse uterine haemorrhage.

Haemorrhage was checked by plugging posterior and anterior nares with cotton dipped in collodion and firm tamponing. 8 p.m. temperature 102.8° , pulse 130-180 weak and irregular, semi-coma alternating with periods of restlessness and mild delirium. Still slight haemorrhage from nose and uterus in spite of former treatment. 10 p.m. about 1 pint of warm sterilized saline injection with slight but temporary improvement. Cardiac tonics, whiskey and digitalis were frequently administered.

March 9th. 9 a.m. temperature 104.8° , pulse 148, Respiration 32. Large and offensive tarry stools of altered blood - injection of saline continued. 6 p.m. complete coma, temperature 106.2° pulse weak and flickering.

March 10th. 2 a.m. She died two and a half days after the onset of the disease. No autopsy was permitted and bacterial examination could not then be made.

When we compare the case as reported by me with the descriptions and reports of these different forms of purpura we find symptoms in each

which are included in my case. Because of the haemorrhages from the mouth, bowels and uterus it could be placed in the division purpura haemorrhagica. Because of the rheumatic-like pains it could be included in the purpura rheumatica. Because of the urgent abdominal symptoms it could be included under Henoch's Disease.

There is still the symmetrical gangrene in my case which is not included in the descriptions given nor in illustrations of any published case.

Dr. Angus reports a case in which one of the extravasations near the knee joint formed a superficial abscess which was opened and healed rapidly (British Medical Journal May 28th 1898 p. 1393); but this abscess I think cannot be looked upon as of the same nature as the sloughs described by me.

The comparison then of this case with Syphilis, Scurvy, Reynaud's Disease and Purpura shows that with Syphilis and Scurvy the resemblance is only slight and cannot stand minute investigation while with Reynaud's Disease and Purpura it has symptoms

in common with each and were we to inflict one patient with each of those diseases at one time the symptoms would be much as I report. It, however, bears the strongest resemblance to purpura, but has a symptom which as far as I am aware has not yet been reported and therefore cannot be included in any of the known sub-divisions of purpura; following the prevailing custom a new sub-division would be required to include those cases (providing we say it is purpura), of purpura which have symmetrical sloughs on the limbs especially in the region of the joints. I, however, intend making use of this case for another purpose than to add to the already long list of the different forms of purpura.

If we refer to the different cases of purpura reported and if we include the case reported by me it will be seen that there is a marked symmetrical distribution of the signs and symptoms which would indicate that it is due to some central lesion, this I shall now attempt to prove, after which it will be seen that the difference between ~~Raynaud's~~ Disease and

Purpura is only one of degree of the same central lesion.

Haemorrhages occur in two ways, viz. I by Diapedesis, and II by Rupture of the Blood Vessels.

I. Diapedesis may not be considered.

II. Rupture of the blood vessel may be caused by (A) Violence from without, as wound: (B) Alteration or Disease of the Wall of the Blood Vessel.

A. Violence from without need not be considered.

B. Alteration or Disease causing weakening of blood vessels may be due to:-

1. The blood vessels may be altered and weakened by changes of, or abnormal ingredients in the blood deteriorating the walls and so allowing the contents to escape.
2. The blood vessels may be altered and weakened by changes of, or abnormal ingredients in the blood, irritating the sympathetic nerve fibres which by reflex action through the vaso-motor act on the blood vessels.
3. The blood vessels may be altered and weakened directly through the vaso-motors by irritation of the central nervous system.

There is no doubt that the haemorrhage of purpura is due to rupture of the wall of the blood vessel and I will now discuss what the cause of the rupture is. Coats' Pathology (page 45) says, "In Purpura Haemorrhagica there is also haemorrhage traceable to weakening of the walls of the vessels from a change in the constitution of the blood." Bristowe page 613 says, "The blood seems to present no constant departure from the normal condition. It seems very probable, however, that the primary morbid condition is in the capillary and other small vessels than in the blood and that the latter escapes into the tissues in consequence of their rupture."

Carter's Elements of Medicine (page 93) says, "Purpura is supposed to be due to one or both of the following conditions (a) Malnutrition of the walls of the smaller blood vessels: (b) Alterations in the quality of the blood." The blood has been examined frequently in Purpura, and as authorities do not agree, the blood evidently does not present any constant change from the normal.

It is essential that there be an escape of

blood from the vessels in purpura, and as far as we know there is no necessity for a given change in the blood; we must conclude that there is no change in the constitution of, or abnormal ingredient in the blood acting either (1) directly on the walls, or (2) reflexly through the nerves of the blood vessels causing them to deteriorate, weaken and rupture, so allowing haemorrhage to occur.

The only other possible manner in which capillaries may rupture is (3) by direct irritation of the vaso-motor centre in the nervous system and this I believe to be the cause of Purpura.

I will now consider the cause of Reynaud's Disease.

Reynaud says that "this disease was caused by "a more or less persistent spasm commencing in the "capillary arteries and extending thence to arteries "of larger size. He showed that the affection tended "to have a symmetrical distribution, and inferred, "therefore, that the vascular contractions were "determined by influences emitted from the vaso- "motor centres." (Bristowe 614.)

That this is the cause of Reynaud's disease may be accepted as, in one case Reynaud observed actual contraction of the retinal arteries attended with impairment of vision occurring in alternation with symptoms presumably referable to spasm of the arteries of some of the peripheral parts of the limbs. That the spasm at any rate in many cases is of central nervous origin is strongly suggested by the marked tendency to symmetrical distribution of the phenomena of the disease. (Bristowe 616.) The conclusion we have now arrived at is that Purpura and Reynaud's Disease are both of central nervous origin and in order to localise the lesion we must briefly touch on the anatomy and physiology of the parts connected with the vaso-motor nervous system.

It is a well known fact that each function is localised in a centre in the cerebro-spinal system. The vaso-motor nerves form an important part of the sympathetic nervous system which (the sympathetic nervous system) has its centre in the medulla oblongata on the floor of the fourth ventricle. It is the case when a centre is irritated it depends entirely on the

amount of irritability whether the function controlled by that centre is excited or paralysed. Therefore it depends on the amount of irritation of the vaso-motor centre on the floor of the fourth ventricle whether the vaso-motor action is excited or paralysed, but we have two sets of nerves connected with the arteriols, the action of which are directly opposed to each other viz. the vaso constrictors and the vaso dilators, and as these sets may be irritated separately we will have the following different actions to consider.

(a) Slight irritation of the vaso constrictor centre would cause increased action of the vaso constrictors, which would overcome the dilators resulting in constriction of the capillaries.

(b) Severe irritation of the vaso constrictor centre would cause paralysis of constrictors action, the dilators would prevail and so result in dilatation and weakness of the capillaries causing proneness to rupture.

(c) Slight irritation of the vaso dilator centre would cause increased action of the vaso

dilators and result would be overcoming the constrictors, dilating and weakening the wall of the capillaries, so causing proneness to rupture.

(d) Severe irritation of the vaso dilator centre would cause paralysis of the vaso dilators so allowing the constrictors to prevail.

It will be seen that there are two possibilities by which rupture may occur viz., Paralysis of vaso constrictors and Stimulation of vaso dilators. Also two possibilities by which constriction may occur viz. Stimulation of vaso constrictors and Paralysis of vaso dilators, and it will be very much a matter of conjecture what the clinical appearance would be in a central lesion causing the foregoing stimuli or paralysis.

To make it complete I would advance the following theory:-

(a) Stimulation (due to irritation) of the vaso constrictor centre would cause constriction of the capillaries resulting in symmetrical anaemia and gangrene, it may be of the extremities or other parts for which there may be some affinity.

(b) Paralysis (due to irritation) of the vaso constrictors, the vaso dilators act, but owing to some unknown inherent force to resist change (which is present in all cells) the vaso dilators are able to prevail only at points it may be certain individual muscle cells so causing dilatation, weakness and rupture at these points - the result would be symmetrical small haemorrhages.

(c) Stimulation (due to irritation) of the vaso dilators, the vaso dilators act causing dilatation, weakness and rupture of the capillaries, this would be more severe than in last case, also symmetrical and cause larger bleeding points and more profuse haemorrhages which may take the form of severe haemorrhages from the mucous surfaces.

(d) Paralysis (due to irritation) of the vaso dilators, the vaso constrictors act but owing to some inherent force to resist change (which is present in all cells) the vaso constrictors are able to prevail only at points, it may be certain individual muscle cells, so causing contraction of that point. It will be evident that there will be no appreciable

change observed, as contraction of one small point in the wall of the vessel will not cause constriction of whole wall.

I am aware that different conclusions may be arrived at as to the exact phenomena which would be observed according as we stimulate or paralyse the different vaso-motors, more particularly as to whether the inherent force to resist change would be greater when the function is paralysed or when the opposing one stimulated, which could be argued quite as well as the theory I have advanced.

It is enough for my purpose if I have shown that irritation of the vaso-motor centre will cause symmetrical gangrene, purpuric spots, ecchymosis, haemorrhages from mucous surfaces, haematuria.

What the irritation is I shall not attempt to suggest. Why the haemorrhages should appear underneath the skin and from the mucous surfaces, while there are seldom haemorrhages (scanty or profuse) into internal cavities, or into the paranchyma of internal organs must be explained in the same manner

as we would explain why eczema attacks the surface of the body, viz. it has an affinity to attack these parts.

The abdominal and rheumatic pains I would look upon as partly due to irritation of the sympathetic nerves. In connection with the kidney symptom which is so common in both Reynaud's Disease and Purpura, I shall quote from Dr. Coat's Manual page 19, where he reports a case of injury to the medulla oblongata which had apparently destroyed the vaso-motor centres of the kidneys.

Dr. Angus reports a patient of his, after having purpura, becoming distinctly "nervous;" he also reports loss of patellar reflexes in another, thus no doubt going a long way to prove the disease as being due to a neurosis.

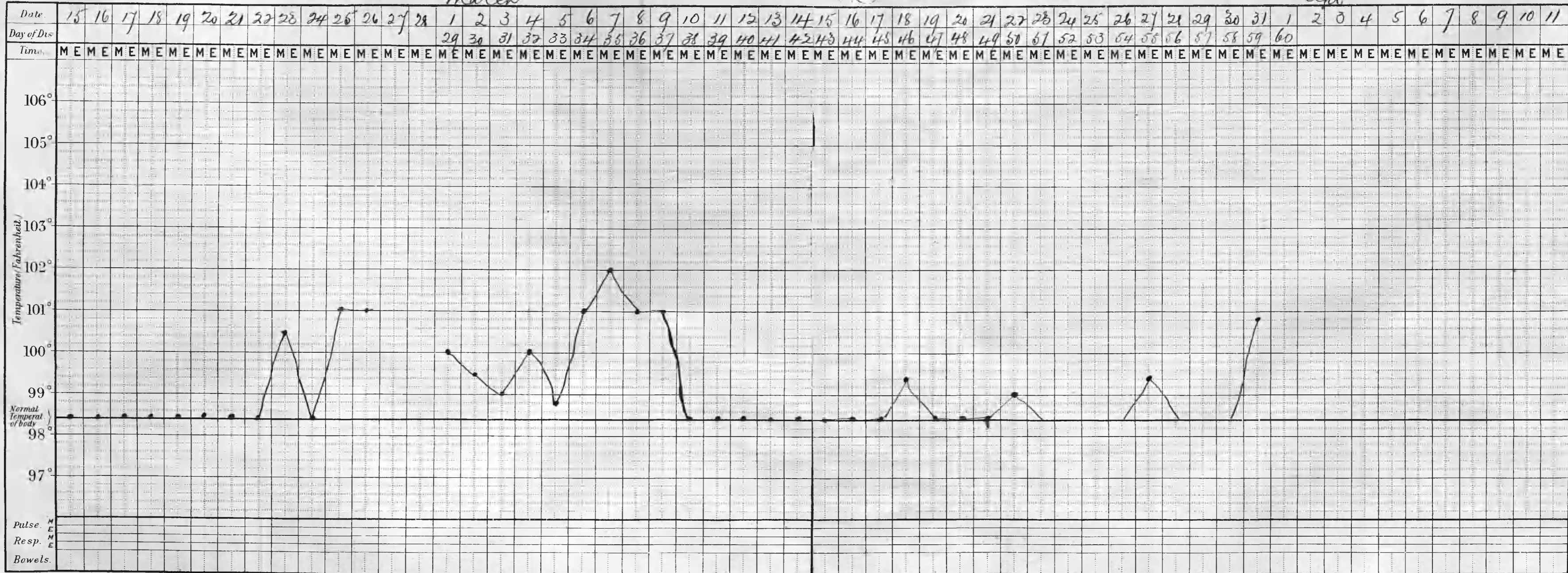
The conclusions which we are now justified in coming to are that Reynaud's disease is due to contraction of blood vessels in extremities and possibly in kidneys caused by irritation of the vaso-motor centre in the medulla. Purpura is due to irrita-

tion of the vaso-motor centre in the medulla the amount of irritation determining the lesion whether petechia, haemorrhages or gangrene, with or without nervous symptoms.

We have then two diseases the symptoms of which overlap (as evidenced by reported cases) both due to irritation of the one part. Therefore there is no difficulty in coming to the conclusion that Reynaud's Disease and Purpura are one and the same disease; that they are merely different degrees of severity of the same lesion in the same manner as spasmodic movements and paralysis may both be caused by cerebral haemorrhage. That they should be removed from diseases of the vascular organs and placed amongst the diseases of the nervous system. That the cause of the disease is an affection of the vaso-motor centre situated in the medulla oblongata.

Name Mr Mc J Age 26 Disease measles Admitted Mr Mc J Age 26 Disease chol. Admitted Mr Mc J Age 26 Disease chol.

Feb



(COPYRIGHT) A VERTICAL LINE MAY BE DRAWN AT THE END OF EACH WEEK OF DISEASE . FOR NOTES OF CASE SEE BACK OF CHART. (RIDGEN'S CLINICAL CHART.) Printed & Published by H K Lewis, 136, Gower Street, W.C.