Haemophilia: read before the New Orleans Medical and Surgical Association, May 2d, 1884 / by J.H. Bemiss.

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Publication/Creation

[New Orleans]: [publisher not identified], 1884.

Persistent URL

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MEDICAL AND SURGICAL JOURNAL.

SEPTEMBER, 1884.

PRIGINAL PAPERS.

Hæmophilia.

Read before the New Orleans Medical and Surgical Association, May 2d, 1884.

By J. H. Bemiss, M. D.

It is hardly possible to present anything very startling as to the disease itself, but each case usually possesses some individual peculiarities which add something to the general history of this affection.

The first history that I shall read is that of a patient of Dr. W. H. Watkins, and it is by the doctor's permission that I am enabled to present these notes.

I was kindly asked by Dr. W. H. Watkins to see Wm. M., who was, at the time the doctor met me, suffering from a severe attack of bleeding.

William is a well built young man, 18 years old; he has a rather dark complexion, brown eyes and auburn hair. The capillary circulation of the surface does not seem more than ordinarily conspicuous. He was born in Summit, Miss., and lived in that State during his early childhood. Had always been a healthy boy, save an occasional paroxysm of malarial fever, which presented nothing unusual.

So far as can be recalled, he had had no hemorrhages during his early childhood, unless there may have occurred a few attacks of *epistaxis*, such as boys often have, and which in his case attracted no attention.

When about 13 or 14 years old, however, his mother remembers scolding him for not sleeping under his mosquito bar, as his legs were covered from his knees down with little spots like mosquito bites. She got him another bar, and was accustomed thereafter to see that he was protected at night, but these spots continued to appear, and were very slow in leaving.

From this time on these petechiæ, if so they may be called, began to grow larger and to have the appearance of bruises. His mother remembers his coming home one evening with a large bluish spot which covered nearly the whole of his cheek and part of his neck. She charged him with having been in a fight, but he did not even know that his face was so marked. These patches began to come so frequently and to be so long in disappearing—some three or four weeks—that his father took him to Dr. W. for advice.

Shortly after this, in the spring of 1880, he had his first, serious, open hemorrhage. It was from the nose, and continued, with short intermissions, for nearly six (6) weeks, and until William was extremely prostrated. A point to be remembered is, that no pain attended these bleedings, and no assignable cause was mentioned.

As soon as strong enough, Dr. W. sent William to the country to recuperate, but he had another severe attack of epistaxis after getting there.

About a year later, William had the measles, and during the febrile stage he bled severely from the same surface.

Thus for two years he had hemorrhages, but at irregular intervals, and with no evidences of periodicity. A few spots of extravasation occasionally showed themselves, though not as many as before the open hemorrhages began. Throughout this period the main source of the blood was the nose, though he had passed a small quantity from his rectum on one or two occasions.

In the spring of 1883 he began to have intermittent fever, the paroxysms appearing on the 7th, 14th and 21st days. In these attacks he lost more or less blood. It was on an

occasion of this kind that I saw him. While in Carrollton he had a chill late in the evening, followed by a high fever which lasted all night, and which was attended by loss of blood from nose, stomach, rectum. He managed to reach his home on Josephine street about 12 M. the following day and Dr. W. saw him at 2 P. M. He then had a temperature of 103°, his pulse was very weak and rapid; he was greatly excited, and was passing blood from his nose and in large quantities from his rectum. He had also vomited some blood. I saw him at 3 P. M. His general condition was much the same as at 2 P. M., but epistaxis had ceased, and his temperature was 104°. While Dr. W. and I were standing near his bed talking the patient called for the vessel and passed from his rectum not less than 24 fl. 3 of dark, fluid blood which seemed to have no disposition to coagulate. I was told by Dr. W. that this non-coagulability characterized all of his hemorrhages.

William had been given ergot both by mouth and hypodermically, and also 1/4 gr. morphine hypodermically.

With the exception of a few small discharges of blood from rectum, which may have been retained, and not fresh blood, the hemorrhages ceased. It would be impossible to say how much blood he had lost, but he remembered that he was bleeding more or less all the night and morning previous to the Doctor's seeing him, and at that time, 2 P. M., he was losing large quantities from his nose, stomach and rectum, and moreover this did not entirely cease until sometime after our joint visit at 3 P. M.

He makes blood rapidly, a feature of all his attacks, and so was soon up and at his occupation of engineer on Carrollton Railroad.

Dr. W. has frequently examined William's heart and lungs and reports them apparently healthy. The blood was examined microscopically and appeared normal, with a due proportion of white and red corpuscles.

The treatment has extended over such a long period that Dr. W. was only able to give me an outline of the main parts. All the mineral and vegetable hemostatics were

used during the attacks, including lead acetate, gallic acid, ergot, digitalis and ipecac.

Tinct. iron was used in very large doses, 20 to 30 drops between attacks for its superior action in increasing nutrition. Bichloride mercury proved of temporary service, though its action was not clearly understood.

Williams' grand-mother at 65 years of age had a very severe attack of hematemesis, which her Doctor said was due to the change of life, but her daughter said her mother had passed the climacteric years before. She vomited at that time an alarming amount of blood, and was very much prostrated by it. A peculiarity which attracted her daughter's attention at that time, and to which she attached much importance, was the fact that an enlarged spleen of the old lady's was reduced to just a little above normal size by this hemorrhage, but afterwards regained its large dimensions.

This old lady frequently had attacks of epistaxis which were very severe. She never complained of profuse menstruation, but she was peculiar in never alluding to such matters either in herself or others. Nor can Mrs. B. recall that her mother was wont to flood after delivery. But she does know that her mother always had large bluish spots on her body and limbs, and in the latter part of her life she was subject to vomiting blood. All these attacks were painless and attended by no fever or other bad symptoms.

She died at quite an advanced age, over 70, and quite suddenly. She was taken sick at II A. M., and died at IO P. M., the same day. The cause was some form of hemorrhage, in which unconsciousness was a symptom, but Mrs. B. was not present and can not tell its exact nature.

This old lady was a stout, well-formed, active woman. Her skin was beautifully clear, her eyes blue and her hair dark. In other words, but for the color of the hair, was a typical blonde.

Mrs. B., William's mother, is a large, fine looking woman,

with a complexion and features exactly like her mother's. She has for years had large spots on her limbs and body, some as large as, or even larger than, her hand. A slight blow is sufficient to cause an extravasation which is very slow in leaving. Others come without any apparent cause. The first one occurred some six (6) months after her marriage and without any cause that she could recall. As a young girl she suffered from epistaxis occasionally, and now her menses are very profuse, and in her confinements she has flooded severely. She has had some miscarriages and at those times her flooding was alarming. Dr. W. anticipates them as much as possible by rapid delivery and the giving of ergot.

Mrs. B. is at the present time in excellent health, but is near the menopause, and it will be interesting to know if she suffers from this tendency during that period.

Mrs. B. had a brother, who was a mild bleeder, if I may use the term. His severest attack was during the early part of the war, and it took the form of epistaxis and hematemesis. Otherwise he appeared healthy. Like his mother, he died very suddenly in June, 1883, and likewise of hemorrhage in some form.

One brother died of consumption.

Mrs. B.'s children, other than William, seem healthy enough, unless it be her little girl 8 years old who frequently bleeds at the nose and always has bruises on her limbs. Her mother thinks these are the result of her active wild disposition.

The same little girl, however, had acute inflammatory rheumatism, for which Dr. W. treated her, and in which her heart became affected.

The main points in this case are:

1st. The absence of the blonde type or any charactertics of thin-skinned or leuco-phlegmatic people.

2d. The strong history of heredity, and the lack of any other factor, unless malaria appears as an exciting cause.

3d. The lateness of announcement of the diathesis.

4th. The commencement as purpura hemorrhage, and

its disappearance when the hemorrhage becomes external.

5th. The dark grumous blood so very slowly coagulable.

6th. The thin clear complexion of the grandmother, a partial sufferer all her life.

7th. Her suffering after the climacteric and its continuance until death.

8th. The mother's trouble at her menses; the flooding after delivery and the abortions especially.

9th. Females seem, in this history at least, to be as liable

as males.

10. The good affect of morphine.

11. The rheumatism in the little girl.

Some may question the propriety of placing the following case here; but there are certainly some points favoring the diagnosis of hæmophilia, and no other was

apparent.

James Ryan, aged 20, a native of New Orleans, came to my father's office about 11 A. M., May 19, 1883, complaining of a sore throat. He had some fever, his tongue was coated, and an examination of his throat showed bot of his tonsils red and swollen, but the right by far the more so. It extended across the median line, pushing the uvula to the left and almost occluding the passage. The finger seemed to detect fluctuation without the least doubt. His throat trouble had begun some five or six days ago, but for a month previous he had had diarrhæa and dysentery; he also had a cough, with purulent expectoration. He had lost flesh rapidly, and had night sweats. Was never a stout boy.

There was no history of any diathesis in his parents, but his mother is an ignorant Irish woman, and her state. ments are not definite. His father died suddenly in 1878

from what the doctor called heart disease.

With a sharp-pointed curved bistoury, guarded to within an eighth of an inch of the point, the inner convex side of the tonsil was punctured; the left was not lanced. A small quantity, perhaps a half teaspoonful, of blood oozed from the wound, but no pus. After remaining in the office some ten minutes and no pus appearing, he was given general directions and told to go home. About one o'clock, p. M., his brother came in great haste, to say that James was bleeding to death. The patient lived quite a distance from any line of cars, and from the point where he left the car he could easily be traced some five or six squares to his home by the mouthfuls of blood which he had spat up at every ten or twelve paces.

I found him sitting on the edge of a chair in a meanly furnished, badly ventilated and dark room. He was in a cold perspiration, pallid, almost pulseless, and greatly excited: His shirt was bloody, there was blood on the floor, and a wash bowl in front of him contained probably a pint of half clotted blood. His mother told me she had just emptied a bowl containing a like quantity. In attempting to clear his throat of some of the clots, he became nauseated and vomited quite a large amount of half clotted, partially discolored blood. An examination of his throat was attended with much difficulty because of the darkness of the room and the clots of blood which adhered to his teeth and sides of his throat, thereby obstructing the view Moreover, any attempt to depress his tongue was attended by an effort at vomiting. However, the result of such examination as I did make seemed to be as follows: the tonsil was reduced in size and there seemed to be a slight wound on the inner side from which bright arterial looking blood was oozing. I immediately gave him 3ii fluid extract of ergot, and by means of a mop applied some Mousel's solution of iron to the wound. This seemed to have some good effect, but unfortunately, about five minutes later, he vomited again, and this apparently started the wound afresh; for in addition to the blood which he vomited, he spat up some fresh blood.

Supposing that the ergot which I had previously given him had been vomited, at least the greater portion of it, I again administered ergot with some whiskey and tincture of digitalis. I had previously put him to bed on his back with his head slightly elevated. In addition I applied the Monsel solution occasionally, but about its only effect seemed to be to discolor the parts so as to make examination all the more difficult, and to increase the nausea.

After this he spat up two mouthfuls more of blood, but save a little oozing apparently from the wound, the hemorrhage ceased. Ergot, brandy and digitalis were given in small and frequently repeated doses, and milk and beef tea were ordered to be prepared at once. As soon as I could do so I took Dr. T. G. Richardson and my father, Dr. S. M. Bemiss, to see the case. After a careful examination they were convinced that the hemorrhage was not due to a wound of any large vessel, but was either pulmonary or due to a hemorrhagic tendency. An examination of the lungs, however, was negative. At Dr. R.'s suggestion I administered gr. 1/4 morphia, hypodermically, puncturing with a needle the biceps muscle. The general treatment above indicated was continued. The patient was seen again at about seven that evening, and was comparatively easy and taking nourishment. Next morning the patient was better, but complained of pain in his arm where the puncture had been made. It was somewhat red in that locality and looked as if an abscess was forming. In the evening his arm was very painful and much swollen, having an erysipelatous appearance. Warm applications were made to the arm and morphia was given internally. General treatment, except ergot, continued. Next morning the arm was swollen throughout, the shoulder being involved. It was red and very painful. The patient died suddenly at 10 A. M. When examined some two hours before death his pulse had been found to be weak, irregular and intermittent. His heart when listened to at the same time was very irregular and laboring greatly.

I must add here before proceeding to the next case, Dr. Brewer's statement that he had known the boy for some time, and had to attend him on several occasions. One in particular being a hemorrhage for which there was no as-

signable cause.

The points to be especially noted are—

- 1. The absence of any hereditary history. If a case of hæmophilia, it is one de novo.
- 2. The dysentery, and the progressive emaciation and cough.
 - 3. The nature of the affection, tonsillitis.
 - 4. The character of the puncture.
- 5. Length of time after puncture before the hemorrhage begun.
- 6. Dr. Brewer's statement that he knew the boy to be a bleeder.
- 7. The cause of death, i. e., the swelling of the arm and the cardiac failure.
- 8. The possibility that by the suppurative action in the gland a small vessel was eroded, which gave way when the supporting pus was removed.

The third case is also one to which some doubt attaches, because experienced physicians had made a different diagnosis; but a careful statement of the symptoms seems to point to the affection which we are discussing.

F. G., age 18, a native of Germany, was sent to the Hawaian Island by his physicians as a consumptive. The young man was wonderfully developed for his age. He was six feet one inch tall, with broad shoulders and strong muscles. He was a typical blonde, clear complexion, light hair, full set of light whiskers and blue eyes. On several occasions, both before leaving home and after his arrival on the Island, he had had severe hemorrhages which he said were always from his lungs Frequently repeated examinations of his lungs failed to reveal a cavity or any indications of tubercular deposits. He, however, had a cough, aud after one of these hemorrhages, which were undoubtedly pulmonary, there were subcrepitant rales to be heard over both lungs. One peculiarity was always present, namely: a very rapidly acting heart.

In excitement, especially such as attended his hemorrhages, it was not only rapid, but irregular and intermittent, but there were no morbid sounds. He became very fond of digitalis, because of its steadying effect upon the heart. After his attacks he would pick up very rapidly. He had no purulent expectorations, though he had the very dry cough alluded to above; had no night sweats, was not emaciated, and was, all in all, the picture of health.

The hemorrhages, at least those coming under my observation, were always from the lungs, and did not come up in gushes as in cases of rupture of very large vessels in phthisis, but in mouthfuls at intervals of five to ten minutes. In other words, the blood seemed to ooze as from a sponge. I saw him one night lose a large quantity, about two pints, but he was ten hours doing so. Opium and acetate of lead pills, however, had an immediate hæmostatic effect.

In this case we note:

1st. Rapidity of growth, favoring origination de novo of hæmophilia.

2d. Absence of family history.

3d. Absence of pulmonary symptoms.

4th. Character of heart's action, giving rise to irregular congestions and capillary rupture.

5th. Character of bleeding; sudden onset, slow but steady continuance and happy effect of such remedies as

opium and digitalis.

Hæmophilia, or hemorrhagic diathesis, or bleeders' disease, is an affection around which much mystery has hung because no one has been able to formulate the underlying pathology. Some contend that it is an alteration of the blood in the direction of a diminution, or any variation in quality of its fibrin, which stands as the cause of the disease. Others assert that the capillaries are deficient in the strength of their walls. Another set maintain that the pressure of the blood due to disturbed action of the heart brings about the extravasations. Still a fourth class say that the nerve supply to the vessels is in some way disordered. Any one or more of these factors may be at the bottom of the diathesis, and it is easily seen that it would

be a difficult matter indeed to demonstrate satisfactorily any one of the causes mentioned. It may be that in course of time those who will have a sufficient number of cases to study the disease properly, will be able to group them by the symptoms into classes which will be characterized by one or the other of the causes stated or yet to be discovered.

Leaving these disputed points of pathology, there may be stated under the head of etiology the following:

1st. Heredity. This seems to be well proven by facts and generally accepted, though Ritter terms the hæmophilia of infancy hæmophilia acquisita, and denies that it is the same as the affection of later life. He admits that heredity may be proven, but the true cause is some other diseased condition, e.g., fungoid growths that are found under some circumstances in the blood vessels. Most modern writers, however, say that when the tendency appears in early life, it may be gradually overcome as the child reaches puberty, provided of course it was not so severe as to have caused death quickly. On the other hand, if the affection should lie dormant until adult life, it is apt to be obstinate and to continue. This latter point is illustrated by the grandmother mentioned in case No. 1, and perhaps William will prove to be another instance. Under this head may be mentioned a statement by a late writer that a mild bleeder is not liable to transmit his diathesis to his children, but through his daughters to his grandchildren, but a female bleeder may transmit immediately to her children. This is illustrated in our first case.

This brings us to the influence of:

2d. Sex. The sexes are unequally affected, by far the greater number of bleeders being males—about 1-12 or 15. A peculiarity to be noted is that a female born of a family with this diathesis may show no signs of it herself, but her sons are almost sure to be bleeders. When the female is affected it is usually the case that the disease attaches itself to her special functions, i. e., her menses will be profuse, she may flood after delivery, abortions may occur. Boerner states that a girl may not evince the tendeney until the beginning of menstruation or marriage produces a crisis which will excite it.

- 3d. Malaria often appears as the exciting cause, precipitating an attack because of the internal congestion of the cold stage, though the underlying pathology be weak vascular structure, deficient innervation, or something else as yet unknown.
- 4. Among the other causes stated is a defibrinated state of the blood as evidenced by its non-coagulability. This point has been denied by some, and, very possibly it does fail to exist in many instances. But such men as Flint and others contend that this is often the cause. This was probably partly the cause in William's case.
- 5. Finally there are undoubtedly cases which arise de novo, and by the term it is not meant to include those cases which are the result of leukæmia, or chlorosis, or such allied diseases. In such diseases as these last it is only a symptom, important though it may be.

The clinical history embraces the symptoms detailed above—namely, a hemorrhage which begins without any known cause in most cases, in others, appearing as an exaggerated function and continuing either steadily or intermittently until a state of acute anæmia or death results.

In infancy the umbilicus is a frequent source of the hemorrhage. In male adult life the bleeding is usually from the nasal mucous membrane and stomach, but may be from any surface or organ: e. g., lungs, kidney, bladder, rectum, skin. In females, the genital organs frequently furnish the bleeding surfaces.

Wounds, especially lacerated wounds, and blows are very liable to result in serious loss of blood. Local inflammations also, and abscesses are very apt to give rise to severe hemorrhage. A woman in child-birth is in great danger of flooding, and during her menses, and particularly at the climacteric, is almost sure to have trouble. Among other symptoms a very common one is the appearance of large spots of subcutaneous extravasation.

Those who have attacks of hematemis or hemorrhage from the lungs, frequently have premonition of the same in a sensation of weight, oppression, or even pain in the locality from which the loss is about to occur, but this is by no means general, on the contrary the opposite is the rule.

An important symptom, and one which is conspicuous by its absence in the cases which I have read, is rheumatic affection of the joints, they are not of the nature of acute inflammatory rheumatism, but more on the order of chronic infiltrations or effusions. The little girl spoken of in the first history was a sufferer on two occasions from rheumatism, one attack being especially severe, and that it was of the acute inflammatory form is shown by the fact that her heart became affected.

The prognosis is uncertain. In infancy it is bad, especially when the umbilicus is the seat of hemorrhage. In those children who gradually improve as they approach puberty the outlook for entire *cure* is good. But hæmophilia appearing in old age is especially prone to last and to hasten, if not actually to cause death.

Suggestions as to treatment are especially meagre, and this largely because the pathology is so little understood. All use ergot, and the various vegetable and mineral astringents. Some use iron (Fe Cl) in order that they may build up the blood and thereby improve nutrition. Others still trust largely to the so-called neurotics under the belief in the nervous origin of hæmophilia.

Cholera Discussion.

ROOMS NEW ORLEANS MEDICAL & SURGICAL ASS'N,
NEW ORLEANS, August 9th, 1884.

In conformity with the resolution of August 2, 1884, the special meeting for the discussion of cholera, was called to order at 8 p. m., August 9, 1884, the President, D. E. P. Shepard, in the chair.

The President stated what the object of the meeting was, and called upon Dr. S. M. Bemiss to open the discussion. Dr. Bemiss stated that not knowing the exact limit to which he was expected to go, he had prepared some remarks on the whole subject.

The following is an abstract of Dr. Bemiss' paper:

It was not an easy matter to condense the subject into the compass of a paper, therefore for the sake of brevity and system he would consider the subject under three heads:

1. Cholera as an epidemic disorder, including the nature of its special cause, and its mode of multiplication, and spread.

2. The disease from a sanitary stand point involving the

means of prevention.

3. Pathology and Treatment.

There are several definitions of cholera, but the one according best with present doctrines is that "cholera consists in the sum of the changes and symptomatic phenomena produced by the presence of the cholera poison in the human system."

He sums up the first division thus:

1. Cholera poison is material and particulate.

2. It so clearly possesses the power of reproduction that we are forced to regard it as an organism. It may be termed a "contagium vivum."

3. In this country it has never been domiciled. India

is the home and birth-place of cholera.

4. It finds access to the human system principally by means of drinking water, or other fluids and foods.







