A case of haemophilia with fibrous thickening in the appendix region / by F. Parkes Weber.

Contributors

Weber, Frederick Parkes, 1863-1962.
Bulloch, William, 1868-1941
Royal College of Surgeons of England

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CLINICAL EVENINGS.

28

November 9th, 1908.

A CASE OF HÆMOPHILIA WITH FIBROUS THICKENING IN THE APPENDIX REGION.

By Dr. F. PARKES WEBER.

HERMANN Z., aged 23 years, a rather delicate-looking man, a German waiter in London.

During the last two years he has suffered from spontaneous hæmatomata, or hæmatomata from slight traumatism, in both lower extremities and in both inguinal regions of the abdomen, and has also had attacks of hæmophilic swelling in both knee-joints and in the left shoulder-joint. In 1907 he had severe hæmorrhage after the extraction of some teeth. His hæmophilic swellings are accompanied by fever, sometimes considerable, up to 103° F., or even slightly higher, in the evenings. A hæmophilic swelling in the right inguinal region of the abdomen, such as he suffered from last summer, might therefore be mistaken for acute appendicitis. Some of his hæmophilic swellings have been apparently completely reabsorbed, though others have left chronic fibrous thickenings. His last trouble was a large hæmatoma, which gradually developed in the left thigh after a slight fall on September 9th. At one time the skin over it was so tense and discoloured that it looked as if it might slough. This hæmatoma has very much diminished in size, but still constitutes a fluctuating swelling in the upper-inner part of the left thigh. There is also still a firm mass (retroperitoneal ?) to be felt in the right inguinal region of the abdomen, and there is slight limitation of movement in both knee-joints. A thickening can likewise be felt over the front of the lower part of the right femur, just above the knee-joint.

There has been no fever since September 22nd. The patient has been treated with calcium lactate. No examination of the blood has been made.

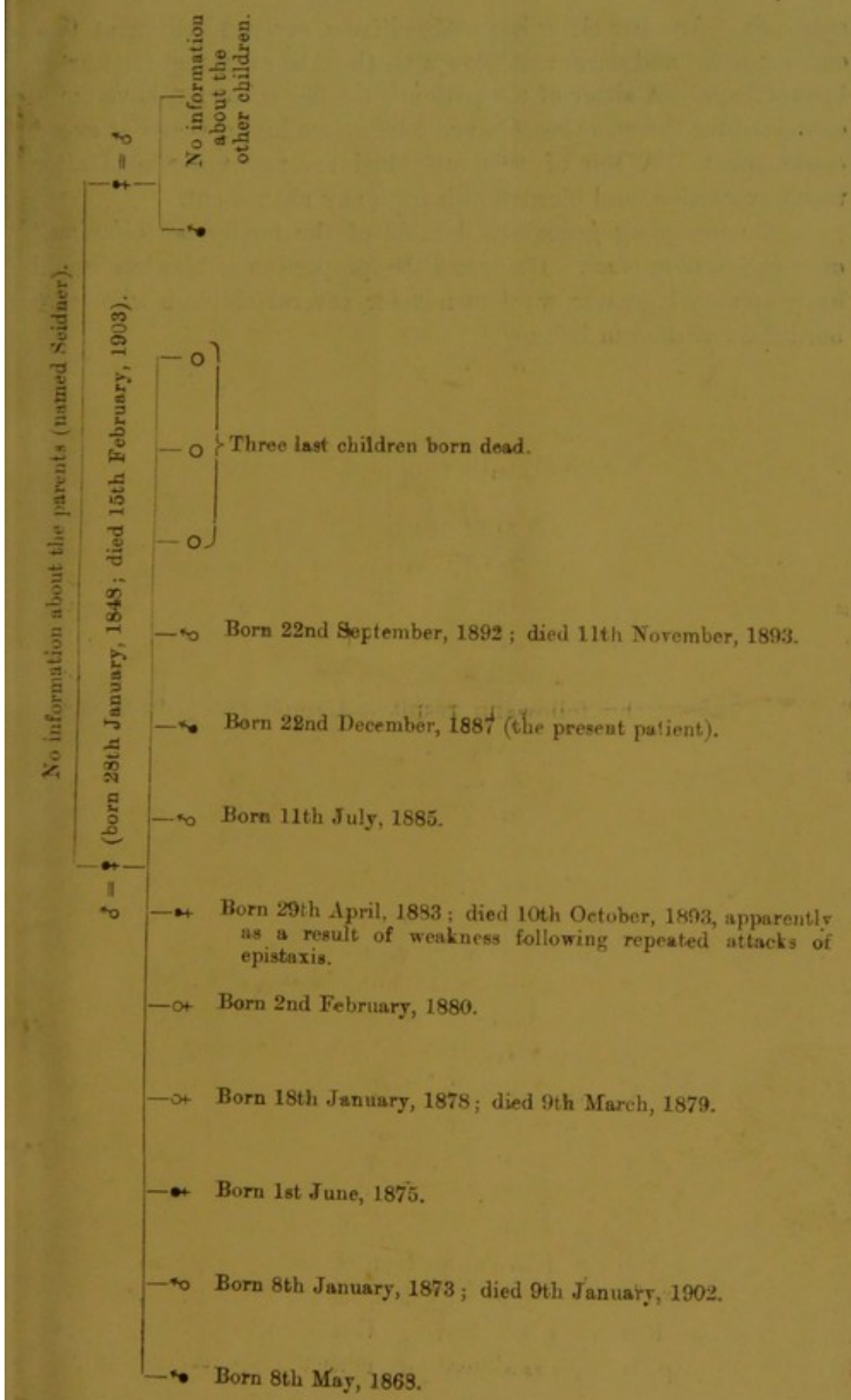
As a child the patient often suffered from epistaxis and bled easily from the gums. He remembers at one time passing urine for some days "looking light red wine." When 10 years of age he had a swelling near the right knee (doubtless a traumatic hæmatoma), and he then heard his mother tell the doctor that surgical interference was dangerous as he was a "bleeder." He does not remember ever having had hæmorrhage from the lungs, stomach, or bowels. Apart from the hæmophilic trouble the patient presents no evidence of any disease. He is the youngest of a family of 12 children, but knows very little of the family history. A son of a maternal aunt tended to bleed excessively from his gums.

Dr. Weber had to thank Dr. Michels, under whose care the patient was at first, for kindly allowing him to show the case.

Addendum.

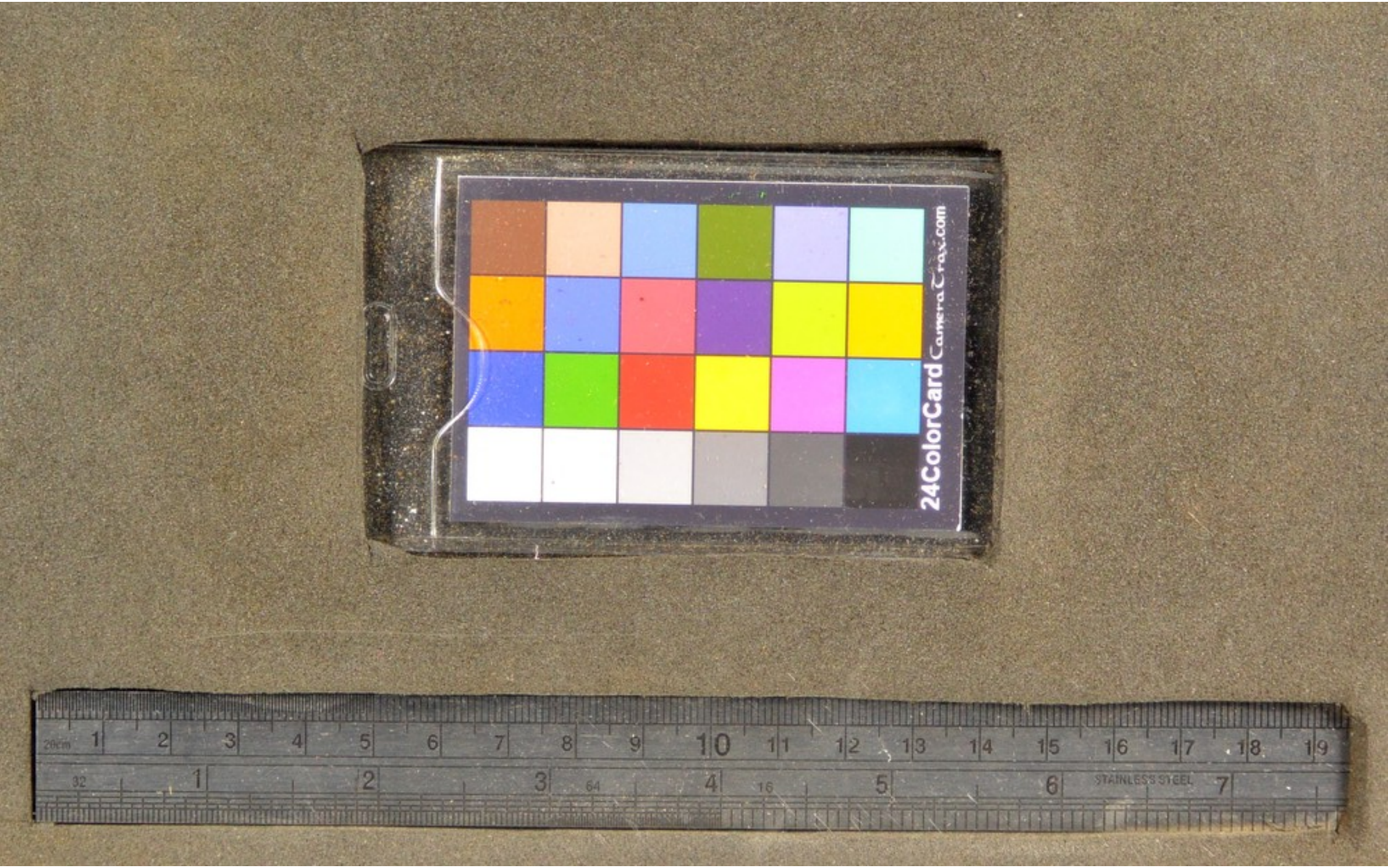
Blood Examination (*November* 13*th*, 1908).—Hæmoglobin (by Haldane's method), 105 per cent. of the normal standard; red cells, 6,400,000 in the cubic millimetre of blood; white cells, 4,500. Dr. A. E. Boycott kindly made a differential count of 500 white cells, which gave the following result:—Lymphocytes, 24·2 per cent.; intermediates, 4·8 per cent.; large hyalines, 9·8 per cent.; neutrophile polymorphonuclears, 58·2 per cent.; eosinophiles, 2·4 per cent.; mast cells, 0·6 per cent. Dr. Boycott thought the definite increase in large hyaline cells deserved notice. The resistance of the red cells to hæmolysis (as tested by the method given by Ribierre) seemed to be normal. The coagulation time (tested by Sir A. E. Wright's coagulometer) seemed to be considerably increased, but estimated again a week later it seemed to be very little, if at all, above the normal.

Some further information has been obtained from the patient's father (who lives in Germany). The father remembers that his son (the patient) had very severe hæmorrhage from biting his tongue at the age of 3 or 4 years, and from a wound on his forehead at 5 years of age. The patient's mother, who died at the age of 55 years, suffered from severe bleeding at her confinements, and would not let a tooth be drawn for fear of hæmorrhage. One of the patient's brothers, now aged 40 years, had severe and persistent bleeding when he had a tooth drawn. One of his sisters died at 10½ years of age from weakness in consequence of continual


No information about the parents (named Seidner).
♀ = ♂
No information about the other children.
♂ = ♀ (born 28th January, 1848; died 15th February, 1903).
Three last children born dead.
Born 22nd September, 1892; died 11th November, 1893.
Born 22nd December, 1887 (the present patient).
Born 11th July, 1885.
Born 29th April, 1883; died 10th October, 1893, apparently as a result of weakness following repeated attacks of epistaxis.
Born 2nd February, 1880.
Born 18th January, 1878; died 9th March, 1879.
Born 1st June, 1875.
Born 8th January, 1873; died 9th January, 1902.
Born 8th May, 1868.

epistaxis. Another sister has suffered from severe bleeding at two confinements, and dares not allow a tooth to be extracted for fear of hæmorrhage. A sister of the patient's mother could not have teeth extracted for the same reason, and had severe hæmorrhage at every confinement. A son of this aunt had severe hæmorrhage after tooth extraction, and bleeds much from any chance cut.

In the foregoing genealogical table of the family *the "bleeders" are represented in black.* Doubtless the proportion of bleeders would have appeared greater were it not for several children who were born dead or died in infancy.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