

## Medical Matters.

### HAEMOPHILIA.

Dr. Paul Fildes, in an interesting article in a recent issue of the *London Hospital Gazette*, describes this somewhat rare disease. The cardinal features are, he says, three in number, and may be boldly summed up in the following definition—an inherited and abnormal tendency in males to bleed.

*Hæmorrhage.*—The question of bleeding may be considered first. The essential feature in this respect is that the application of trauma, which would, in a normal person, pass unnoticed, in a bleeder determines an hæmorrhage, and when this hæmorrhage is established there is no tendency for it to stop in the usual manner. The blood is usually described as trickling or oozing away from a surface in spite of all treatment, and continuing till death ensues, or, which is very much more common, till spontaneous arrest occurs after the subject is reduced to a condition of profound anæmia.

An outstanding feature of hæmophilic bleeding is that in the majority of cases it can be definitely traced to *trauma*. The hæmorrhages are frequently stated to have been spontaneous in onset, but are usually of a nature or in a position commonly liable to slight knocks or pressure. The skin lesions vary from a normal bruise to an hæmatoma according to the severity of the injury.

The liability to hæmorrhage is always *chronic*. It is noticed in early childhood, and makes the existence of the sufferer wretched, throughout youth to manhood, or perhaps old age. It appears, however, to be established that, if adult old age is attained, a progressive decrease in the number and danger of the hæmorrhages may occur, so that in middle life the individual may be practically free.

*Variation in the intensity* of the liability to bleeding is frequently well marked, and has been clearly demonstrated over a period of years by Ripke. A boy may be seen literally covered with bruises and prostrated by some particular bleeding. Pressure with the thumb upon the skin will determine a bruise in five minutes, yet a week or two later the same boy may be free from bruises, react normally to pressure or a needle stab, and even undergo a cut without unusual hæmorrhage.

The *general condition* of the patient at the time of a bleeding is undoubtedly in the great majority of cases, apart from anæmia, good. Fever, malaise, and constitutional disturbance are slight or absent, the former being chiefly seen in cases where the blood is retained within the body. The boy lies quiet and expectant,

“as if,” as an author says, “he realised the danger of his position.” When the bleeding stops, recovery is usually not prolonged, there having been no further drain on the patient’s vitality than the loss of a quantity of blood. The boy regains his habit of gaiety and recklessness, which outlook on life is so constantly mentioned as to constitute a minor feature of the disease.

*Sex.*—With reference to the question of sex in hæmophilia, it is necessary to state that, after an exhaustive examination of the literature, no condition has been found in a *female* in any way closely resembling the disease known as hæmophilia. In the most elaborate and best-known cases, females are definitely stated to have been unaffected, but the literature is full of alleged female bleeders. . . . That the females in bleeder families are abnormal in some unknown particular must readily be admitted. They are the active propagators of the disease, and they differ from normal women in the great size of their families; but that they are liable to hæmophilia is quite unsupported by evidence.

It is, however, impossible to dismiss the subject of hæmophilia in women without reference to that rare and obscure condition, sometimes referred to as hæmatidrosis or sweating of blood. This disease, which is probably a complex of pathological states, constitutes a chronic “hæmorrhagic diathesis.” . . . The symptoms consist of hæmorrhage or the exudation of bloodstained fluid under the skin or from any area of the body surface. Of these hæmorrhages, the most remarkable, and that which was considered a marvel, is sweating of blood from the pores, hair follicles, or sweat glands. The attack is often preceded by some violent emotion or sometimes trauma. In any case, the subjects are usually of a neurotic temperament.

*Inheritance.*—It has always been considered one of the most remarkable features of hæmophilia that the disease runs in families, and the earliest reporters were so definite on this point that even in 1820 Nasse had sufficient material at his command to formulate his so-called law—that hæmophilia is propagated entirely by the unaffected females in bleeder families to their sons. This type of propagation is illustrated in every bleeder family, and must be accepted as the rule. It is, however, admitted that in a small number of cases the disease appears to have been propagated through a male, either a bleeder or a normal man. The number of instances, however, is so small that they may represent what might be described as “a descriptive error”: in fact, that the assumption of propagation through the male is erroneous.

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