Report of a case of hemophilia / by G.S. Towne.

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X-ray picture showed no evidence of clinical tuberculosis. In spite of temperature slightly above normal she was allowed to return to work and has been at work ever since. Has been apparently well and has gained in weight.

CASE XI. Miss ——, age 14. First seen May 13, 1909. Complained of slight cough. On physical examination a few scattered rales heard opposite the 7th to 10th v. s. Temperature 100.2°, pulse 128. On May 15th they were practically the same. During the summer while the child was resting most of the time her temperature was normal though the pulse remained very rapid. Since September 3d it has nearly always been above 100°, once reaching 101 1-5°. The pulse has varied from 122 to 140. Examination of the chest on two occasions has shown no abnormal signs referable to the lungs except one occasion during an acute cold scattered rhonchi were heard on both sides of the chest December 4, 1909, at the bases. On auscultation of the heart a systolic (?) murmur has been heard in the pulmonary area and occasionally at the apex not transmitted. Leucocyte count January 1910, 9000. Von Pirquet test positive.

(NOTE.—May, 1910. Since the above report physical signs of mitral stenosis have developed in this case and a diagnosis of chronic endocarditis has been made.)

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Clinical and Pathological Notes

Report of a Case of Hemophilia. By G. S. TOWNE, M. D.,

Read before the Medical Society of the County of Saratoga, March 29, 1910.

Our knowledge of hemophilia is modern. Its historians have been able to find the records of but few cases or families prior to those of the last century. The characteristics of this disease were first classified by American physicians in the early part of the nineteenth century, and again later, during the past thirty years, Hughes, Gould, Hutchinson, Harris, Holton and Dunn have added much to the literature upon this subject.

It is my purpose in this paper to summarize some of the characteristics of this disease, which have been observed by the above named authors and upon which all are practically agreed, and later to report a case which recently came under my care and incidentally gave me much anxiety.

In a majority of cases the disposition is hereditary, The fault may be acquired, however, but nothing is known of the con-

ditions under which the disease may thus arise in healthy stock. The hereditary transmission of this disease is truly remarkable. Osler mentions the Appleton-Swain family of Reading, Mass., in which there have been cases for the past two centuries, covering a series of seven generations. The usual mode of transmission is through the mother, who is not herself a bleeder, but the daughter of one. The reversion to the ancestral stock through the mother alone is almost the rule, and the daughters of a bleeder though healthy and free from any tendencies are almost certain to transmit the disposition to the male offspring. A peculiar fact also is the ratio of cases which occur between the sexes. One author places the ratio at eleven males to one female, another at thirteen to one. As a rule the disease makes its appearance during the first two years of the patient's life. It is rare for the manifestations to be delayed until the tenth or twelfth year. Again, all grades of social life are equally affected. Such families of bleeders are usually large, the members very healthy in appearance with fine soft skins.

The pathology of this most interesting disease is obscure. While a congenital fragility of the vessels is said to exist, it has never been proven. It has been observed that bleeders always suffer from prodromic symptoms which are due to an increase in the volume of blood. Hence variability in the volume has been invoked as causal. Disturbed innervation diminishing from time to time the vascular tone is thought by Musser, Pierce and others to be the pathological factor. It is mere hypothesis, as have been all suggestions thus far put forward, regarding the pathology of hemophilia. As suggested by Eicharst it may be possible that the changes in the blood are chemical and that our present laboratory methods are not adequate to discover them.

Attention is commonly called to a bleeder by the occurrence of a hemorrhage difficult to control though induced by some trifling cause. The extraction of a tooth is one of the most frequent of these events. It may be the prick of a pin or a scratch or a slight cut as in vaccination or nothing at all may be discovered. The tendency may manifest itself at the umbilical cord at birth or in Jewish children at the circumcision. On the other hand the same accidents which are without results early in life may induce it later. Uncontrollable epistaxis is one of the most frequent manifestations, occurring in 169 out of

334 cases collected by Grandidier. It may be induced simply by blowing the nose. Other situations are the mouth, stomach, ear and eyelids. On the other hand hemorrhages rarely occur in the interstices of organs and though interstitial hemorrhages do occur it is the result of some trifling blow, when the well known "black and blue" appearance is produced. According to Tyson the absence of interstitial hemorrhages, except as the result of some cause, however trifling, may be said to distinguish hemophilia from the acquired tendency. The external hemorrhages including those of the mouth and nose may be profuse and even fatal. They often last 24 hours and longer. When checked reaction from them is rapid and the victims rapidly resume their natural appearance, though repeated hemorrhages may engender a permanent anaemia.

The comparative rarity of hemophilia excuses in part the surgeon who, when in the midst of or after the completion of an operation, suddenly realizes that the patient is a "bleeder." At least this is a very consoling thought to me and without it I should hardly have the courage to write this paper for the County Medical Society. Unlike most other morbid conditions, when, during the physical examination or history taking, a lead may be gained and followed up from general interrogation, this portentous state of body will remain unguessed until active work is commenced unless the direct thought is in the clinician's mind and the questions asked and developed: "Is the patient a bleeder?"

A case of hemophilia came under my care last January in which fortunately the outcome was satisfactory, though I was remiss in forgetting to ask in regard to the possibility of hemophilia before beginning active work. It concerned L. W., a male adult, age 24, a blond, with an unusually fair, soft skin, suffering from a left sided indirect inguinal hernia. He was admitted to the Saratoga hospital January 19, 1910, and gave the following history: Father dead, cause unknown; mother living, but not well. Has profound anaemia, which is a family trait. One sister living and well. Personal history: Has always been well with the exception of diseases of childhood; has a tendency to constipation; had one attack of appendicitis two years ago, from which he recovered without an operation; has had a left sided indirect inguinal hernia for seven years which has been fairly well controlled by means of a truss. Recently the truss has

failed to keep the hernia reduced and as a consequence it has occasioned much distress and inconvenience. Physical examination showed a normal condition of heart, lungs, liver, spleen and bladder. Urine 1028, acid, no alb. and no sugar. After a proper preparation and etherization, the operation was begun. From the first it was noticed that the hemorrhage was very profuse, complicating the technique greatly. So hampered were we by the hemorrhage that both Dr. Ressiguie and myself commented upon the occurrence, and the possibility of his being a bleeder was discussed. We resorted to every known means at our command to control the oozing, but failed. To apply hot compresses produced but little temporary effect; to tie all the bleeding points was hopeless and to suture a portion of tissue which was oozing freely simply added two more bleeding points at the point of entrance and exit of needle, which I am bound to say had a very curious but subduing effect upon one's self-confidence. The wound was dressed with a firm pad of gauze bound tightly over the wound. The dressings were watched closely for blood stains and the patient for signs of hemorrhage. Everything seemed to go well for three days, when the patient began to complain of pain through his abdomen. He was restless, his temperature and pulse considerably accelerated and there was marked tympanites which salines and enemas failed to relieve. The wound was examined carefully and it had every appearance of healing by first intention. During the next two days, all the symptoms were exaggerated with a fetid breath, badly coated tongue and a peculiar greenish yellow tint to the skin added. The wound was again examined with great care and a fluctuation detected. A small opening was made through the original incision and about four ounces of dark fluid blood emitted. The opening was immediately opened down to the fascia to the full size of the first incision and packed tightly with gauze. The accumulated blood seemed to come from the interior of the abdomen, welling up through the sutured fascia. The cut surfaces began to ooze immediately. During the next six hours the dressing had to be reinforced twice on account of the extensive hemorrhage, at which time I again packed the wound with gauze saturated with adrenalin chloride 1-1000. I placed the patient upon calcium chloride, gr. x v, q, IVh, and gelatine, one cup full t, i, d, with his nourishment. Further inquiry into his history, following his recovery from ether, revealed the fact that

every cut, scratch or abrasion always bled very profusely, and this trait had also been present in his maternal grandmother. He had no knowledge of any other ancestor having the tendency.

The oozing of the blood continued in spite of the adrenalin, but showed a tendency to abate at each dressing while the old blood that had extravasated the tissues, and doubtless dissected up the peritoneum, still continued to escape through the wound. At the time the wound was opened his temperature had reached 103 3-5°, with the pulse at 110 and leucocyte count 12,900. In four days the temperature and pulse were normal, but there was still marked oozing each time the wound was packed. An interesting feature of the wound was its tendency to granulate. I never saw granulations accumulate so rapidly. The patient left the hospital just twenty days after the operation with all the annoying symptoms incident to the extravasation of blood abated and the wound nearly closed; but with granulations that required attention daily. In another week the wound was closed. I fully expected to have the operation result in a failure, but up to the present time (Saturday, March 26), he feels perfectly well and is following his occupation of knitter with no signs or symptoms of a return of the hernia.

My experience with hemophilia has been limited to three cases. One was in a case of typhoid fever which occurred during my interneship in Hartford Hospital nine years ago. In this case the family history was very striking in its hereditary transmission from one generation to another. The patient died from intestinal hemorrhage early in the course of the disease. The second one was a case of pregnancy which occurred in my practice 2½ years ago. In this instance I was familiar with the family history of hemophilia, in which there had been one death from hemorrhage, and fortified myself and the patient with calcium chloride for two weeks previous to the expected confinement. Everything worked well; which might have been the case without treatment, for it is the rule that hemophilia is no barrier to the development of large families.

I have no desire to encounter another case of hemophilia complicating a surgical procedure. The attendant risks to the patient are too great, and the anxiety which such cases occasion the operator, are too exasperating to warrant the attempt.

In case of subsequent legal action by the parents or relatives after an unfortunate ending to some cutting operations, it would

be fair to suppose that a jury in relieving the attending surgeon of the responsibility would be bound to consider the shortcoming of failure to learn of the contra-indication to the use of the knife, along with the manifest peculiarity of the disease itself. Certainly some weight in favor of the operator would be the degree of necessity which prompted the operation.

Editorial

> But Lady Penelope went on—"If you knew, my lord, how I lament my limited means on those occasions! but I have gathered something among the good people at the Well. I asked that selfish wretch, Winterblossom, to walk down with me to view her distress, and the heartless beast told me he was afraid of infection—infection from a puer— puerperal fever! I should not perhaps pronounce the word, but science is of no sex. However, I have always used thieves' vinegar essence and never have gone farther than the threshold.
>
> SIR WALTER SCOTT, BART.
>
> *St. Ronan's Well.*

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Postural Treatment and Lavage of the Renal Pelvis

In *Surgery, Gynecology and Obstetrics* for February, 1910, Dr. Paul M. Pilcher urges the advantage of postural treatment and lavage for the relief of pyelitis in pregnancy. Dr. Pilcher states that the true pyelitis of pregnancy is an acute catarrhal inflammation of the pelvis of the kidney which occurs suddenly during the course of a normal pregnancy. It is acute in its onset, usually unilateral, affecting more frequently the right than the left kidney, runs an acute course and tends to spontaneous recovery without permanent injury to the kidney.

He reports eight cases. In seven of these, cystoscopic examinations were made and the ureters catheterized. In only one case (bilateral infection) was it found necessary to terminate the pregnancy. The other seven cases were successfully treated by lavage of the renal pelvis, the employment of the elevated trunk posture and diuretics.

From his own observations he believes that the conditions may result from the following causes: *first,* that a few cases are due to toxic influences and hematogeneous infection in a kidney whose

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