lest, as in this case, the outcome proves the cause to have been, though not organic disease of the valves or vessels, something more definite and dangerous than a mere nerve-storm.

In the treatment of aortic disease the primary object to achieve is the establishment and maintenance of sufficient compensation. In the case of our first patient, who suffered recently from rheumatic fever, the chances are that his compensation never became firmly established—that is to say, that the length of time over which he rested and the amount of care which was taken of him otherwise after his acute illness was not enough to build up his myocardium against the strain of his subsequent life. In addition to this, he has been careless of himself and has indulged in alcohol freely and unwisely. Consequently he comes to us with all the evidences of broken compensation; fortunately for him, it has not gone beyond moderate pulmonary congestion. Already since his admission the improvement in his condition is marked; the mitral murmurs which he then had have almost disappeared; the cardiac rhythm is regular; the locomotive pulsation in the vessels has diminished, and there is a distinct though feeble second aortic sound to be heard in the carotid arteries. There is now no cough and no physical signs in the lungs; the liver dullness has become normal; he is taking food well, and is sleeping comfortably in a normal position the night through. This amelioration has taken place under the influence of complete rest in bed, a simple and moderate diet, regulation of the bowels by means of salines and an occasional small dose of calomel. At first he took a mixture of sodium salicylate, digitalis, strychnine, and ammonium carbonate; this was followed later on by another, which he is still taking, containing dilute phosphoric acid, iron perchloride, nux vomica, and strophanthus.

The only point worthy of comment in the medicinal treatment of a case of this type is the employment of digitalis. The old tradition that digitalis is a dangerous drug in aortic disease clings to many of us so pertinaciously that we hesitate to avail ourselves of its valuable assistance. This is not, I venture to think, good practice. The true action of digitalis is that of a tonic, the best of all we possess, to the myocardium. When compensation is well maintained, its exhibition is never indicated, but whenever this fails, it should be called to the rescue, irrespective of the nature or situation of the valvular lesion. Not only so, but at the outset of its administration the drug may be given advantageously in bolder doses than are customarily employed. Fifteen minims of the tincture every four hours may be ordered with perfect safety, if we are careful to watch its effects and curtail the quantity when we get evidence of saturation. That evidence is forthcoming in an abnormally slow cardiac rhythm, diminished urinary secretion, and disturbance of the digestive organs. In many instances the good effects of the drug are evidenced only after these physiological effects are produced. No harm ever results if, at this stage, the drug is either temporarily withheld or its quantity diminished to 7 or 10 minims every eight hours. Strophanthus and convallaria are recommended as valuable alternatives which are better suited to aortic conditions; but though they are useful remedies, their good effect upon the heart falls far short of digitalis, and it is by no means uncommon to find a speedy improvement under the influence of digitalis, when the others have failed to bring about the least response.

Another point about the administration of digitalis which is not sufficiently recognized is the safety that may be secured by its combination with nitro-glycerine. In all cases where vascular tension is high the 1 per cent. solution of nitro-glycerine given along with digitalis prevents increase of peripheral resistance, and so robs the digitalis of that influence upon the arterioles on account of which its administration is supposed to be contraindicated.

In the case of our second patient, who has likewise improved decidedly since his admission to hospital, the treatment has been somewhat different. His chief complaint, when first brought here, was of pain in the chest on exertion; shortness of breath, which prevented his lying down in bed at night; and recurring anginal attacks of a severe type. This is a combination of symptoms with which we are very familiar here, and which is enough of itself to suggest aortic disease. You have satisfied your-

selves, I hope, that his case is one belonging to the degenerative type of this ailment, but, in addition to the knowledge you have gained of his present condition from the physical examination you have made, I must remind you that, when first seen, his pulse was one of very high tension. Here we found a leading indication for treatment, and, in accordance with our usual plan in cases of this description, he was at once put on a mixture of potassium iodide and nitro-glycerine. Both these drugs, and especially the latter, are valuable vaso-dilators, and, under their influence, this man rapidly experienced relief to his breathing and diminution in his precordial discomfort. He was kept closely confined to bed, his diet was simple and mostly sloppy, and every night for a week he was given one grain of calomel, followed in the morning by a moderate dose of ordinary white mixture. After ten days he was so much better that we were able to put him on iodide of iron, strychnine, and strophanthus, and to allow him a greater amount of freedom in regard both to diet and exertion. He has had no recurrence of his anginal attacks since a few days after his admission; he is now able to sleep comfortably in the ordinary recumbent position, and a moderate amount of walking exercise can be indulged in daily without producing either cardiac discomfort or undue physical fatigue. As regards the dosage of the iodide and nitro-glycerine in cases of this description, I am persuaded we often err in making the quantities too small—15 gr. of sodium iodide, protected by 2 minims of Fowler's solution, and combined with 2 minims of the 1 per cent. solution of nitro-glycerine may with perfect safety be given every four hours. Whenever the pulse becomes soft and begins to quicken, longer intervals should be allowed between the doses, and eventually the nitro-glycerine may be wholly withdrawn and the iodide continued in smaller amount, say 5 gr., along with strychnine and one or other of the milder preparations of iron. In regard to calomel, its value is not confined to its aperient effect; it is a useful intestinal antiseptic as well, and also assists in diminishing the tension in the arteries. Obviously the nature of these patients' occupation is inimical to the maintenance of their circulatory balance when they return to work. We always urge them to find a new field for their labours, in which exposure, muscular strain, and dietetic irregularity will enter less into their daily experience, but it is difficult for most men after middle life to embark upon a new calling, and many of our cases ultimately succumb to the circumstances of an environment which is unusually trying and hard even for those who are in the possession of full vigour and healthy organs.

A Clinical Lecture

UPON

THE SURGICAL ASPECTS OF HAEMOPHILIA,

WITH ESPECIAL REFERENCE TO

TWO CASES OF VOLKMANN'S CONTRACTURE RESULTING FROM THIS DISEASE.

BY

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I WISH to show you three patients to-day who illustrate very well some of the common surgical symptoms of haemophilia, and belong to two families which exhibit in a remarkable way the peculiar hereditary characteristics of this disease.

W. W. is a well-grown, intelligent lad, who has had almost every variety of bleeding. But first let me speak of his family, and for those to whom a tabular representation appeals more than a description I give a diagram of their genealogical tree (Table I).

This boy's great-grandmother had one daughter and one son. The son was a "bruise and bleeder," and died of bleeding from the extraction of a tooth; the daughter, who was well and healthy, had six children. One son, who is now 36, has six healthy children; one son died at 17 from epistaxis, one son died at 6 from the bleeding of a finger crushed in a door; one daughter (Mrs. C.), is married, and has two children, a daughter of 3 who is normal, and a son of 5 who is a bleeder. The second

TABLE I.

Female.					
Female.			Male. Bled to death after extraction of tooth.		
Male, 35. Well.	Male. Died at 17, bleeding from nose.	Male. Died at 6, bleeding from finger crushed in door.	Female (Mrs. C.)	Female (Mrs. W.)	Female. Well. Unmarried.
6 healthy children.			Male, 5. Bleeder.	Male, 3. Bled to death at 4 months.	Male. Bleeder.
					Male. Well.

daughter (Mrs. W.) has had three sons, one of whom you see here and one who is quite normal. You will notice that we have considered four generations, and that in each case the females are free from the disease, but that they bear children of whom the great majority of the males suffer from haemophilia. The only male who is without the disease, and married, has begotten healthy children. In other words, the females transmit the disease without suffering from it, and the males are liable to suffer from it, but not so liable to transmit it. It is stated, however, that if a man with haemophilia begets children the boys suffer from a severe form of the disease.

I have only had the opportunity of seeing and examining one of the haemophilic relations of this boy, Harry C., who is now 5 years old, and is the boy's cousin. When three days old he had severe bleeding from the umbilicus, which continued more or less for three weeks. In this respect he is remarkable, because it has been noted by Legge that bleeding from the navel is not found in haemophilia. When he was 2 years old he fell and cut his lip, and the bleeding from this continued for three days; when he was 5 this accident was repeated, with the result that much more severe haemorrhage was caused. Local styptics and the actual cautery applied to the part only made matters worse, as is generally the case in haemophilia, and he was taken to the infirmary and kept in bed for ten days, one deep stitch being used to bring together the wound. A fortnight ago his nose bled for three days without stopping, and at the same time, without any known cause he had pain and swelling in the right knee and ankle-joints.

And now to come back from his relatives to William W. himself. In childhood his bleedings and bruising were so many and varied that I need not detain you with an account of them. When he was 11 he fell and cut his lip and this continued to bleed for about a week; at or about the same time he hit his right elbow and a very large swollen bruise appeared in this region and spread down the arm. A few days later he noticed that all his fingers became "clawed," that is to say, bent up into the palm, and at the same time they were all blistered, and you will see presently that his hand retains to this day evidence of the truth of these statements. He consulted a doctor, who bound his arm up in splints for four weeks. At the end of this period he found his forearm was weak and shrivelled, and that he was unable to supinate it at all. He has since regained the power of extending the fingers almost fully, though the index finger still remains somewhat flexed, but he has never been able to supinate the forearm, and this has been a serious matter to him, as he was thereby forced to abandon a position as tram conductor because he could not put out his hand, palm upwards, for the fares, and it caused his rejection as a candidate for service on a railway, because when he was asked to cover his right eye, he did so with his knuckles, and hence revealed the condition which, together with his haemophilia, had otherwise been overlooked. When I first heard this history I was inclined

to think that the pressure of the splint had been accountable for the "paralysis" of his hand, and, indeed, the patient himself was not slow to suggest this explanation. But I have found another instance of this rare combination of two rare conditions, and I will defer any further remarks about the arm until I have shown you the other patient. When this lad was 13 years old he was hit by a stone on the left knee. It became much swollen, and from that time to the present it has never completely recovered itself, but is often, without apparent cause, swollen and painful, and, indeed, the opposite knee has not been free from the same condition. When he was 15, and again when he was 16, he had bad attacks of haematuria, lasting for three weeks and one week at a time. The urine was bright red and acid, the blood being intimately mixed with the urine the whole time. This bleeding stopped spontaneously, or perhaps, I ought to say, as the result of taking drugs, without producing much constitutional disturbance. It is a remarkable fact

that visceral haemorrhages in haemophilia are not very frequent and are seldom fatal; it is common enough to hear of haemophilics bleeding to death from the nose, lips, or teeth, but very rare to hear of one who dies from haemoptysis, haematuria, or melaena. The day before yesterday

Fig. 1.—W. W.'s right arm. Note the wasted muscles, the slight flexion of the wrist and index finger, and the fixed pronation. He cannot supinate it any further than this. The bones of the forearm are 2 in. shorter than those of the opposite side.

he slipped off his bicycle and produced certain recent evidences of his bleeding capacity, which I am able to show you. He did not actually fall prostrate, but he simply alighted on his feet in such a manner as to give them a slight twist or strain. Over the right thigh in front is a large hard swelling, tender to the touch and of indefinite outline. There is no discoloration of the skin, and I think we may conclude that it represents a haematoma deep in or under the quadriceps extensor muscle. This must have resulted from a violent muscular contraction made in the effort of saving himself from falling, which ruptured some of the fibres of the muscle together with small blood vessels. Both knee-joints can be felt to grate when moved, and in the left is a considerable quantity of fluid, but neither knee is at present at all painful; this condition is, of course, of old standing. In both ankles there is considerable stiffness and pain, and by palpating the joints on either side of the tendo Achillis and in front fluid fluctuation is evident. This appears to be the first complaint that he has ever made about the ankle-joints, and you will notice that they are much more painful than the knees; it is usually the case that the first effusion into a haemophilic joint causes much more pain than the subsequent ones. This may be due to the fact that the first effusion is pure blood, and the later are chronic serous effusions set up by chronic synovitis; or it may be the result of a blunting of sensibility of the joint structures that results from repeated effusions.

We now come to the consideration of the other two patients, who are brothers, and who belong to a very large family of bleeders.

Again we can go back four generations—to these boys'

TABLE II.

Female.				Female.			
				Male. Died from bleeding from slight wound of gum at 24.			
4 males. All well.				7 females. All well.			
Male. Died at 6, bleeding at nose.				Female (Mrs. H.)			
Male. Died at 10, bleeding at nose.				6 females. Well.			
Male. 28. Well.				Male. 6. Bleeder.			
Male. (S. H.) 18. Bleeder.				Male. (C. H.) 14. Bleeder.			

great-grandmother. (Table II.) This woman had two daughters only. One of them had one son, who was a bleeder, and he died only a few weeks ago in the Royal Infirmary. He was a shoemaker, and, as is the habit of these workers, he kept the nails which he was about to drive into the shoe, in his mouth. The point of one of them penetrated his gum, and, after twenty days' bleeding, he died of anaemia at the age of 24. He had often had haemorrhages before, but they had not been so severe; and it is very noteworthy that the severity of the bleeding, even in the same patient, varies very much at different ages and at different times in life without any apparent reason. As a general rule, the severity of the bleeding diminishes with advancing years, but then, as in this case, some trivial wound causes a fatal issue. But to return to the family history. The boys' grandmother is alive and well to-day, and is the mother of fourteen children—six sons and eight daughters. Four sons are well, and seven daughters are well and married. One son died at the age of 6 from epistaxis, and one died at 10 from the same cause. The one married daughter, Mrs. H., is the mother of these boys. She has ten children—six healthy daughters, none of whom are married, and four sons, only one of whom is free from haemophilia. The one brother, aged 6

Fig. 2.—Left arm of C. H. Note the muscular wasting and the fixed pronation. He cannot supinate it any further than this. The radius is $\frac{1}{2}$ in. shorter than on the other side.

years, who is a bleeder, I have not shown you, because to-day his disease is quiescent; but his mother gives a long account of bleeding from cuts, large and frequent bruises, and swollen knee-joints.

The elder of these two lads, Samuel H., is 18 years old, and he has the usual history of inordinate loss of blood from trivial cuts and wounds, together with frequent joint troubles in his knees. I show him because he illustrates very well the condition that results in the joints from repeated haemorrhages. Both knees are normal on inspection and measurement, but round the edges of the articular cartilages some nodular thickening can be felt, and, on moving the joints, cracking can be heard and grating felt. There is nothing in the physical signs in the boy different from an ordinary case of osteo-arthritis, and the condition is, in fact, nothing more or less. If we could examine the inside of his knee-joints we should find the cartilage fibrillated and worn away in channelled grooves, where the bone would be exposed as a dense polished surface. The edges of the cartilage would be thrown out into nodular excrescences, and the whole articular surfaces stained a deep rust colour, which would be the only evidence of the origin of this osteo-arthritis.

And it may not be out of place to warn you that if ever you meet with a patient in the neighbourhood of the Victoria Embankment who has effusion into a joint or marked grating, not to commit yourself to a diagnosis of chronic arthritis or to urge operative treatment without first giving a thought to the possibility of such a condition being due to haemophilia. The apparent spontaneity of the condition, together with the youth of the patient, should be enough to put you on your guard, and a very few questions about bleeding will generally reveal the nature of the case,

as these unfortunate people are usually fully aware of the nature of their condition.

The third patient I am showing to-day is Cleophas H., he is 14 years old, and among his many juvenile bleedings and bruising one only deserves mention. At the age of 9 he bit his lip, and was taken to the Children's Hospital, where, in his own words, he was "strapped down in bed" for three weeks. But it is to his joints that I wish to draw your attention. When he was 7 he became involved in a crowd at the time of the general election. His father, in order to extricate him, pulled him by the left arm. This caused great bruising, swelling, and stiffness of the elbow and wrist, and when the bruise disappeared he was left with a somewhat wasted limb, which he has never been able to supinate. To-day if you examine his joints you will find that there is an effusion of fluid into both elbow-joints, which is most conspicuous on either side of the triceps tendon where distinct fluctuation can be obtained. The left knee gives a distinct grating sensation on move-

ment, and the right knee presents a large tense fluid swelling which apparently came on spontaneously a few days ago. It measures $14\frac{1}{2}$ in. in circumference, as compared with $12\frac{1}{2}$ in. on the left side. The fluctuating enlargement upon which the patella can be felt to ride

extends 4 in. above the joint and 2 in. below it. Probably this contains almost pure blood, but, as he has had such a swelling several times before, there is little or no tenderness accompanying the present condition. You can readily understand how such a large and tense effusion of blood into the joint, if often repeated, will stretch and weaken the ligaments; and this event often occurs, and contributes in no small measure to the ultimate weakness of the joint, in addition to the disability caused by osteo-arthritis.

Now let me devote a few moments to the consideration of the condition of the left arm, together with that of the right arm of W. W., the first case I showed you. In both cases the hand is held in a position of almost complete pronation. And yet slight movements, which are free and painless, can be obtained in the radio-ulnar joints, showing that there is no ankylosis. In both cases, also, there is decided muscular wasting. In the first case there is slight flexion of the index finger, but this can be extended when the wrist is flexed. In neither case is there any anaesthesia, nor is the electrical reaction of muscles or nerves altered at all. In the case of the bigger boy the affected right arm is not only $\frac{1}{2}$ in. smaller in circumference than the left, owing to muscular wasting, but the length of the bones is less, the radius measuring only 9 in. as compared with $9\frac{1}{2}$ in. of the opposite side. In the other boy there is also both wasting and actual shortening of the bones to the extent of $\frac{1}{2}$ in.

These are all symptoms of a somewhat rare condition termed Volkmann's contracture, after the surgeon who first described it in 1875, and it is now generally described as ischaemic paralysis.

A very complete account of this condition, together with

notes of 15 cases, has been given by Dudgeon.¹ In the great majority of cases (13 out of the 15 in the series quoted by Dudgeon) the affection comes on after a fracture of the arm or forearm has been treated by splints and bandages. But in a case of Mr. Wallis it arose after the use of an Esmarch's bandage applied for one hour to the arm; and in a case of Professor Schloffer it resulted from thrombosis of the axillary artery. This contracture is caused by pressure on the muscles of the forearm, depriving them of blood. Some of the fibres of the delicate and highly organized striated muscle then die or undergo rigor mortis, and are replaced by fibrous tissue or, in rare instances, by bone (myositis ossificans). This causes contracture, wasting, and pseudo-paralysis. The feature in which both my cases differ from most of the recorded cases of ischaemic paralysis is that pronation is much more marked than flexion of the fingers and wrist. But this is accounted for by the fact that there has been a constant effort to move the fingers, which has probably overcome the flexor contraction, of which the first case gives a distinct history. The pronation, on the other hand, is the position of rest assumed by the hand when held in a sling, and also is that used in all ordinary acts, such as writing or using a knife or fork. So that there was probably no voluntary or systematic effort made to stretch the contracted pronators, and now you can feel in both these boys a tense, fibrous cord in the position of the pronator radii teres when supination is attempted.

In 6 out of Dudgeon's series of 15 partial or complete pronation of the arm was left as a permanent condition in Volkmann's contracture.

The actual shortening of the bones is difficult to account for. It is present in almost all cases of Volkmann's contracture occurring in children, and therefore its presence in these two cases confirms the idea of the condition being one of this contracture. The most likely explanation seems to be that when the muscles become atrophied and partially functionless, their blood supply is much lessened; and as all the blood vessels which supply the growing ends of the bones are branches of muscular vessels, it is easy to understand how a lessened blood supply will lead to diminished growth.

It seems at first sight a contradiction in terms to speak of ischaemic paralysis as occurring in haemophilia. But this contradiction is only one of words and not of ideas. In both the cases I have described there was a great haemorrhage into the soft tissues of the forearm, and this, confined under the tense skin or fascia, compressed the muscles and brought their circulation to a standstill. There was too much blood outside the muscles and too little in them.

In the case of the younger boy no splints were used at all, and in that of the elder his description leaves no doubt that the typical contracture had developed before the splints were applied. As far as I am aware, there is no other instance recorded of the occurrence of this peculiar contracture in haemophilia, and it is very remarkable that I should have come across these two cases at the same time who happen to live in neighbouring streets.

In conclusion, I must say a few words about the treatment of haemophilia. From a surgical point of view it may be summarized as—rest and pressure. When the joints become swollen they should be firmly bandaged, and kept at rest until the effusion subsides. It is by neglect of this treatment that the ligaments become stretched and weakened, and the changes of osteo-arthritis hastened. For the external bleeding do nothing but keep the patient in bed. One of the fatal cases I have referred to was bleeding from a scratched gum for fourteen days, during which he walked about from doctor to doctor, trying each new remedy, and bleeding steadily in spite of all, until at last he found refuge in the infirmary, only to die from the resulting anaemia. No remedies, local or general, have any effect on the haemorrhage, and therefore leave that alone and turn all your attention to treating the patient's general condition. If this rule was always followed from the first onset of the bleeding probably many lives would be saved.

REFERENCE.

¹ *Lancet*, January 11th, 1902, p. 78.

COR TRILOCULARE BIATRIATUM.

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THE following case borders on the unique:

History.

On December 22nd, 1905, I saw a man, aged 35, in consultation with Dr. Webb of Gorton. The patient was propped up in bed, and presented the usual appearance of one suffering from chronic heart disease. His breathing was difficult, but he was not cyanosed nor were the fingers clubbed; he had a cough with bronchial expectoration. There was some oedema about the ankles. The area of cardiac dullness was considerably increased; the cardiac impulse was more extensive than usual, the apex beat being difficult to locate. The centre of the impulse was about the fifth interspace, a little within the nipple line. There was no thrill. On auscultation, the first sound was found to be replaced by a loud, rushing, systolic murmur, which extended over the whole cardiac area. No other murmur could be distinguished. The second sound was muffled. Some amount of oedema was present at the base of the lungs. The liver dullness was slightly increased. There was no ascites.

Subsequently I learned that the patient improved for a time; he then relapsed, and expressed a strong wish to go into hospital. He was so determined that (whilst recognizing his critical condition) Dr. Webb reluctantly consented, and he was admitted under my care on May 28th, 1906, into the Salford Royal Hospital. His condition was most distressing. He was deeply and universally cyanosed and was gasping for breath. The heart beats were tumultuous and irregular; the veins were overfilled, and those in the neck pulsated. There was general oedema, with effusion into the pleural and peritoneal cavities; the lungs were oedematous. The urine was almost suppressed. The surface of the body was cold and clammy. The patient was fully conscious, and earnestly pleaded for something to relieve the breathing; but in spite of all treatment it became worse, and he died on the second day after admission.

Necropsy.

An examination of the body was made by Dr. Mair, pathologist to the hospital, on May 31st, who reported as follows:

General oedema of the body, especially marked in the lower extremities, penis, and scrotum. Cyanosis of face, neck, and arms. There is considerable serous exudate in the pleurae on both sides. The lungs show passive congestion and oedema.

The Heart: The ventricular portion is much enlarged, and there is no difference in thickness of wall indicative of right and left ventricle. On incising the ventricular wall, it is found that there is complete absence of the septum between the two ventricles. Into the common ventricle open the mitral and tricuspid orifices in their normal positions. Two arterial trunks emerge from the ventricle in the relative positions of aorta and pulmonary artery. The anterior of these gives off the two coronary arteries; its wall is thicker, while its lumen is smaller, than that of the posterior trunk. The latter is thin-walled, and the valves at its commencement are extensively sclerosed and calcified. Unfortunately, these trunks were not followed to their distribution; but it seems likely that the anterior is the aorta and the posterior the pulmonary artery, there being thus transposition of these vessels. The auricular portion of the heart presents nothing abnormal, except that the foramen ovale is patent. The heart weighs 1½ lb.

There is a large amount of serous exudate in the peritoneal cavity. The liver shows advanced "nutmeg condition." The spleen is enlarged and firm. The kidneys show passive congestion.

The illustration is reproduced from a photograph, taken after Professor Lorrain Smith had arranged the incised wall of the ventricle, so that the four openings of the arteries and auricles may be seen communicating with the one cavity. The mouth of the aorta, slit open, is seen immediately above a glass rod, there placed to hold the ventricular wall apart. Behind the left end of the rod a cavity is seen, an inch and a quarter in depth and five-eighths of an inch in diameter; this appears to represent a rudimentary right ventricle.

Professor A. H. Young, to whom I am indebted for the photograph from which the illustration is taken, exhibited the heart and described the developmental anomalies causative of the malformation at a recent meeting of the Anatomical Society of Great Britain and Ireland. A description will appear in the *Journal of Anatomy*.

Malformation of the heart as above described is exceedingly rare, and when encountered in adult life it is phenomenally so. W. T. not only lived to the age of 35 years, but, with the exception of the last six months, he enjoyed average good health throughout the whole of his life. I am indebted to Dr. Webb for the following

THE Hungarian Chamber of Deputies has passed a Bill providing that insurance against sickness and accidents shall be compulsory.