HEMOPHILIC ARTHRITIS

(BLEEDER'S JOINTS)

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In spite of the fact that the majority of hemophilics who reach the age of puberty are subject to joint disturbances, hemophilic arthritis is a rather rare condition and most of the affected individuals know that they are bleeders and promptly inform the surgeon of this fact when any operative procedure is considered. .Consequently, it does not occur to the average surgeon that he may some day open a hemophilic joint under an erroneous diagnosis. This, in spite of the facts that a hemophilic arthritis may closely resemble conditions for which operative intervention is indicated and that the literature contains several reports of surgical tragedies which resulted from operations upon hemophilic joints under an erroneous diagnosis. Consequently, it is important that surgeons who operate upon joints should know thoroughly the clinical picture of hemophilic arthritis.

In this paper I wish to report an unhappy experience of my own in which a fatality was narrowly averted, and also wish to describe for the first time in English the clinical picture and pathology of hemophilic arthritis as well as to make certain additions to our knowledge of the subject which have resulted from the study of my operated case.

N. McG., aged thirteen, was referred to the St. Louis Children's Hospital from the dispensary for the removal of a painful tumor over the mesial condyle of the right femur. The dispensary diagnosis was old calcified hematoma. He was admitted to the hospital September 5, 1929.

His father was believed to be living and well, but had not been heard from for several years. His mother died following childbirth, but not from hæmorrhage. One sister was living and well at the age of twenty-six. Five sisters and brothers had died in infancy from pneumonia or diphtheria. No history of bleeders in the family.

The boy had had measles and pertussis in infancy. He has always bruised easily and large, hard, blue-black swellings followed the bruises and lasted several days. At the age of three he bled several days from a small cut in the tongue. He always had to bandage small cuts very tightly in order to stop the bleeding. He has had occasional nose bleeds, but they never lasted over thirty minutes. There was no especial bleeding following the removal of the deciduous teeth. From time to time he has been troubled with painful swellings of one or more joints. The swellings usually occurred spontaneously as he was not able to recall any injury. The involved joint became enlarged, tense, and painful. This condition persisted for from a few days to a week and then gradually subsided and the joint returned to normal. An exception to the above was the right knee, as will be mentioned below. The joints involved were the right knee, both ankles, both elbows, and various joints of both hands and fingers.

He is quite positive that the subcutaneous hematomata and joint swellings have been much less frequent during the past two years.

At the age of five years he fell and struck the right knee on the pavement. The knee became markedly swollen and painful and the patient remained in bed about a month. The pain gradually disappeared and the swelling decreased, but some swelling persisted over the internal condyle of the femur and the knee remained flexed. This swelling and flexion deformity have persisted until the present time and the swelling has always been quite tender. From time to time the knee has been markedly swollen and painful. These attacks have usually followed a slight injury and have caused him to remain in bed for from four to fourteen days. He does not know how many times the knee has been swollen, but states that the number is large. The last time was about six months ago.

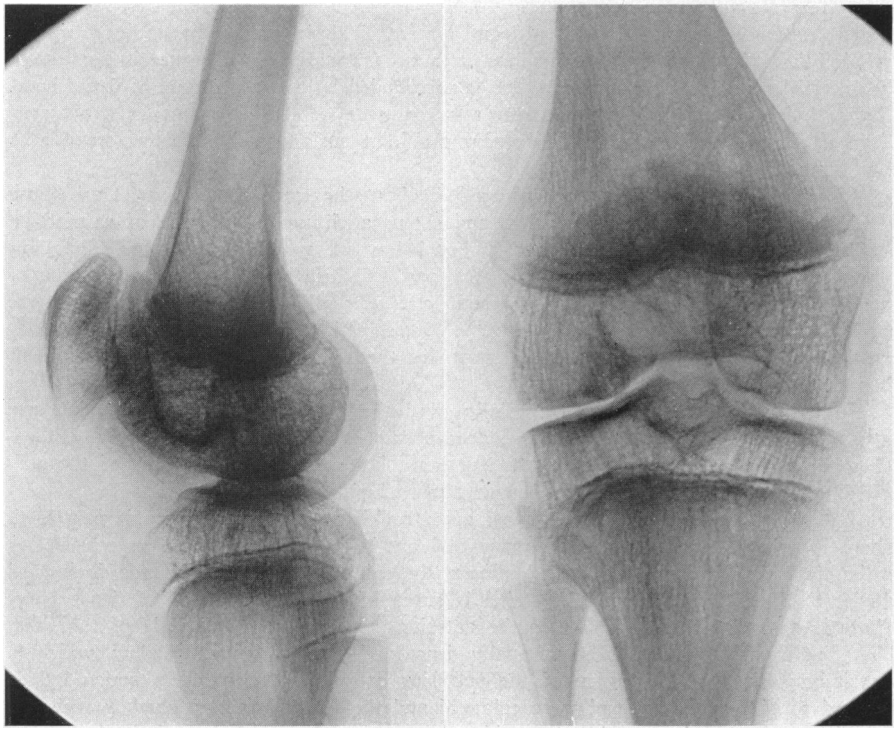

FIG. 1.—X-rays of knee of hemophilic arthritis before the operation. The characteristic dark shadows in the periarticular tissues do not show in the prints, but the larger rarefied areas may be seen in the femur and tibia.

He was a pale, slender boy, height sixty inches, weight seventy-nine pounds. General physical examination was negative excepting a soft blowing systolic murmur at the heart apex. All joints were normal except the right knee. There were 30 degrees of permanent flexion of the right knee and a valgus deformity of ten degrees. Flexion was limited to 90 degrees by pain and motion was accompanied by soft crepitus. There was no actual shortening of the extremity or atrophy of the calf, but there was 1.5 centimetres atrophy of the thigh. The right knee was slightly larger than the left and the internal condyle of the femur was unusually prominent. On palpation no excess fluid was demonstrable in the joint, but the joint capsule was felt to be thickened and constricted. Its margins had a rubber-like consistency and could be rolled beneath the fingers. The thickening was especially marked over the internal condyle of the femur and here a hard cartilage-like mass was palpable. There were no redness or

local heat, but this mass was quite tender and was continuous with the thickened capsule and could be moved slightly upon the underlying bone.

The above history was obtained after the operation. The case came under my care when I was asked to look after the orthopœdic service for a few days while the surgeon in charge was out of the city.

Laboratory findings.—Urine, normal. Wassermann, negative. Red blood-cells, 4,590,000. White blood-cells, 9400. Hb., 70 per cent., Blood clotting time, seven minutes (done in the dispensary).

X-Ray findings.—(Fig. 1.) Antero-posterior view, right knee: The X-ray shows moderate enlargement of the bones of the right leg, the lower end of the femur being nine centimetres broad on the right and eight centimetres on the left. The bones are quite atrophic. The epiphyseal line is not markedly irregular, but is doubly contoured, especially over the external condyle, and is broadened. In addition to the atrophy there is a rather large circular area of rarefaction in the mesial portion of the external condyle of the femur, which is surrounded by a narrow zone of dense bone. This area is one and one-half centimetres in diameter. There is a smaller ovoid area of rarefaction in the internal condyle near the joint surface and a similar small area near the border of the external condyle.

The condyles of the tibia are enlarged in about the same proportion as are those of the femur, and the bone is rarefied, and shows a circumscribed area of rarefaction similar to those in the femur and located just below the spines of the tibia. The joint space is narrowed, the articular surfaces are roughened and the small areas of rarefaction in the inner condyle of the femur are unusually prominent while the lateral borders of the condyles are serrated. The intercondylar notch in the femur is moderately broadened and deepened. The spines of the tibia are slightly more prominent than are those on the other side.

In the periarticular tissues, especially over the mesial portion of the joint, are rather dense shadows which are in places circumscribed and quite sharply defined. These do not appear to be attached to the bone, but merge into the mass of the surrounding capsule, which is more opaque than usual.

Lateral view, right knee: The bones are atrophic and the joint space is narrowed. There is a large area of rarefaction one and one-half centimetres by two centimetres in the epiphysis of the femur which begins at the epiphyseal line and extends downward almost to the end of the condyle. This is surrounded by a thin zone of dense bone. There is a similar smaller defect in the anterior portion of the epiphysis of the tibia. The joint capsule appears to be unusually dense and casts a definite shadow, which is especially marked in the region of the posterior capsule. The patellar tendon bulges forward as though the fat pad were enlarged and the fat pad casts a shadow which is more dense than is that on the other side. The articular surfaces of all the bones are very thin and slightly roughened.

As I had obtained only a history of an injury with resultant deformity and disability, it was my impression that this was a traumatic arthritis and that the tumor was a mass of fibrous tissue or cartilage which should be removed. This impression seemed to be confirmed by a casual glance at the X-ray which I first saw just before I made the incision.

Operation.—September 9, 1929. Under a tourniquet, a longitudinal incision about five inches long on the mesial side of the patella was carried down through the capsule of the joint. The skin and subcutaneous tissue and joint capsule were apparently normal. Beneath the fibrous capsule and separated from it by a small amount of areolar tissue a layer of dense fibrous tissue was encountered. This was incised in the line of the incision down to the synovial membrane without opening the synovial cavity. The synovial membrane was unusually dark in color. A small opening was made in it and about twenty cubic centimetres of blood flowed from the joint cavity. This blood was dark in color, though not the purplish color of venous blood, and it

contained no clots. At this point in the operation the diagnosis of hemophilic arthritis was made, but since the damage was done there seemed to be no reason why an attempt should not be made to relieve the patient of the tender swelling over the internal condyle provided he survived the effects of the operation. Consequently, the operation was continued.

The incision in the synovial membrane was enlarged and the joint was inspected. The synovial membrane was dark chocolate in color and the joint cavity was filled with hypertrophied synovial folds and villi. The articular cavity was smaller than normal and this was particularly true of the quadriceps bursa, which extended only about half an inch above the patella and was crossed by several bands of synovial tissue.

The fat pad was unusually dense and fibrous in character and a thick fold of dense synovial tissue extended along the mesial border of the patella and tended to overhang the articular margin of that bone. The articular cartilage on the femur, tibia, and patella was yellowish-brown in color and contained many areas in which the cartilage was either thinned or had entirely disappeared and in the eroded areas the underlying bone was covered by a delicate chocolate-colored connective tissue.

These areas of thinning and erosion in the articular cartilage were irregular in size and contour and suggested the sharply demarcated maplike appearance as described by König.[1] There was also some marginal erosion of the articular cartilage and a very thin narrow marginal zone where the cartilage appeared to be invaded by the hyperplastic synovial tissue, but no definite pannus was present and there was no tendency to the formation of marginal osteophytes or ecchondroses. The erosion and thinning of the cartilage seemed to be from the underlying bone as the eroded areas did not correspond to the areas of greatest pressure in the joint and in the thinned areas the surface of the cartilage was smooth. The semilunar cartilages were pale, reddish-brown in color, but were normal in other respects.

The patella was not displaced outward as it was not deemed advisable to enlarge the incision sufficiently to permit this. The thickened mass of synovial tissue and dense fibrous subsynovial tissue which covered the anteromesial aspect of the mesial condyle of the femur was excised *en bloc*, as was the thick band which extended along the mesial border of the patella. Then with a knife a small bit of the margin of the articular cartilage of the femur approximately one-half by one centimetre was excised. It was found that the underlying bone was very atrophic and could be cut easily with a knife.

All visible vessels were then ligated and the wound was carefully closed in layers and a pressure dressing applied before the tourniquet was removed.

The patient was returned to the ward in good condition and orders were left that he be kept quiet with morphine, the limb elevated and that he be matched for transfusion and transfused immediately if any unusual amount of bleeding occurred.

During the afternoon the leg became cyanotic and covered with small purpuric spots (subcutaneous hæmorrhages). Consequently, the dressing was loosened.

September 10.—The day after the operation, fresh blood appeared on the dressing at 11:00 A.M. The dressing was tightened and the bleeding stopped, but reappeared in about an hour and continued until four o'clock in the afternoon, when it seemed to cease spontaneously. He was transfused at 5:00 P.M., receiving 300 cubic centimetres of whole blood. After the transfusion the bleeding began again and I saw him for the first time since the early morning, at which time his condition had been good. I removed the large blood-soaked pressure dressing and found that there was no general ooze but that a small artery was loose near the upper end of the wound. The bleeding could be stopped at will by compressing this against the femur. This was ligated by passing a curved needle into the incision and under the artery and out through the skin. It was not necessary to open the wound. Bleeding stopped.

September 11.—Second transfusion, 500 cubic centimetres whole blood.

September 16.—The knee began to swell and was quite painful.

September 19.—Temperature was 39.8° by mouth. The knee was very tense and painful and a small amount of blood was oozing from the wound. The skin edges were necrotic in places from the tension. Knee aspirated and 120 cubic centimetres of old blood were removed. Smears and cultures of this were negative for bacteria.

September 20.—Knee again very tense and skin edges beginning to separate. Eighty cubic centimetres of old blood were removed by aspiration. A good deal of blood remained in the knee, but this was apparently clotted. Smears and culture of the aspirated blood were negative for bacteria.

September 22.—The wound continued to look bad and a plaster bandage was applied and the extremity elevated.

September 23.—A window was cut in the dressing and all sutures were removed. As the sutures were removed the edges of the wound separated, revealing a large blood clot (about eight by five centimetres) which completely filled the anterior portion of the joint. This was firmly adherent to the front and inner side of the femur.

September 25.—Third transfusion, 500 cubic centimetres whole blood.

October 1.—The clot was lifted out, leaving a necrotic base.

October 14.—Plaster bandage removed and Thomas splint and adhesive traction applied to correct the flexion deformity, which was now about 60 degrees. The

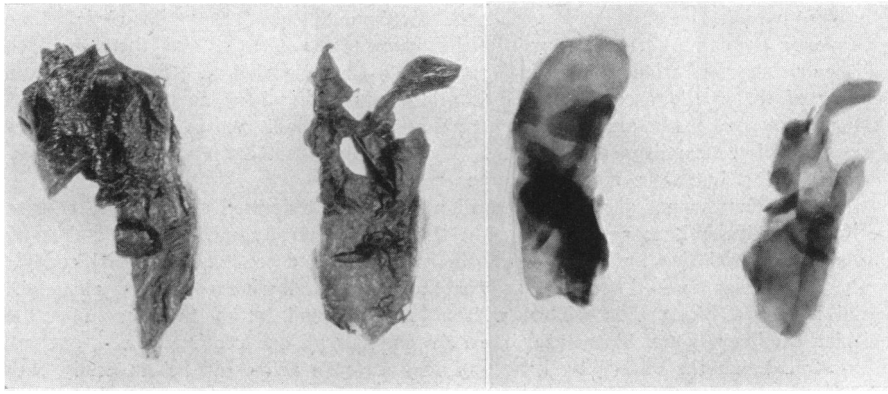

FIG. 2.—Photograph on the left and X-ray on right of part of synovial membrane removed at operation. The dense shadow cast by this tissue in the X-ray is due to the large amount of iron in it.

necrotic tissue had by this time separated and the wound was comparatively clean and its edges were being pulled together by adhesive strips. From this time on the convalescence was uneventful and the patient left the hospital on crutches.

The blood-platelet count was first done September 11, after the first transfusion, and daily thereafter until September 26. On the eleventh the platelet count was 284,000 and it rose slowly to 386,000 on the eighteenth. At this time the patient was started on twenty minims of irradiated ergosterol (acterol) daily and the platelet count rose rapidly to 745,000 on the twenty-second and remained at about this level during the period of observation.

The red blood-cells numbered 4,100,000 on admission and 3,810,000 after the second transfusion. Although there was no further bleeding the red blood-cells gradually dropped to 2,380,000 on September 25. This occurred while the platelet count was steadily rising to 745,000, under the influence of acterol. After the third transfusion the patient was put on a liver diet and on October 18 the red blood-cells were 4,100,000, hæmoglobin 70 per cent., white blood-cells 11,400, and clotting time eight and one-half minutes (one-half minute longer than it was before the operation).

The temperature rose after the operation and reached 37.6° on the fourth day. On the eleventh day the temperature reached 39.8°. At this time the knee was dis-

tended with blood and infection was suspected, but no growth was obtained from the aspirated material.

Pathological Examination of Material Removed.—Gross Description.—(Fig. 2.) The specimen consists of a mass of synovial membrane five by four centimetres in diameter and two smaller pieces. The synovial membrane is dark cholocate color. Much of the surface is covered by numerous large and small rounded villi, folds and synovial bands. Some of the villi are pedunculated and are attached to the surface by long slender pedicles while others are sessile in character. Many of the folds and villi branch extensively and in some areas they form an intricate moss-like structure. On section these large villi are quite friable, contain no visible fibrous tissue, have the appearance of raw calves' liver and some of them contain deep clefts or cavities which are filled with unclotted blood. The subsynovial tissue contains a layer of dense white fibrous tissue which in some places is four millimetres in thickness. The delicate synovial membrane can be stripped from this with comparative ease.

In addition to the above, there is a small fold of dense fibrous tissue about one

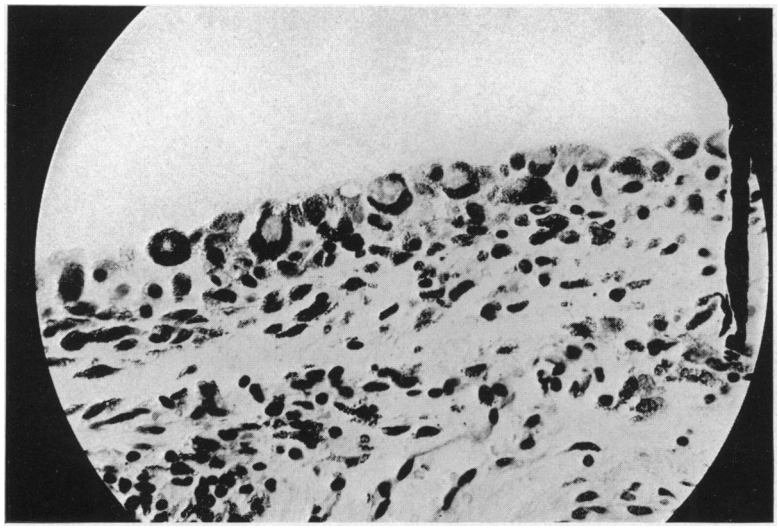

FIG. 3.—Synovial surface. The large cells along the surface are monocytes and the large granules are pigment granules. Four millimetres objective.

centimetre in width, one-half centimetre in thickness and three centimetres in length. The synovial membrane over this is only slightly hypertrophied and is less dark in color than are the villi.

The third specimen consists of a small block of cartilage two millimetres in thickness and five millimetres square with a bit of the attached synovial membrane and underlying bone.

Microscopical Examination.—Projecting from the synovial surface are many folds and villi. These are large and very numerous in the loose or areolar areas of the joint and are small and few in number or even absent in the dense fibrous areas.

The most striking feature in the sections is that the tissues, and especially those near the synovial surface, are loaded with granules and masses of yellowish-brown pigment.

Synovial Surface Layer.—Over most of the surface the synovial lining cells are moderately increased in size and number, forming a surface layer which is from four to ten cells in thickness. The majority of these cells are the moderately enlarged synovial lining or fixed connective-tissue cells. They range from spindle to stellate

or ovoid in shape and possess a faintly basophilic protoplasm and a rather deeply staining nucleus which is larger than that of normal synovial lining cells. Many of these cells contain a few or moderate number of the pigment granules. (Fig. 3.)

Scattered through the surface layer are great numbers of large phagocytic cells (macrophages), many of which are loaded with the yellowish-brown pigment. Many of these cells contain a large pale nucleus with one or more large neucleoli and are characterized by a sharply defined clear zone in the protoplasm on one side of the nucleus. This clear zone is roughly spherical in shape, like ground glass in appearance, and free from the pigment with which the rest of the cytoplasm may be loaded. It corresponds to the rosette of the monocyte, and these cells are believed to be monocytes. A few of the monocytes contain two nuclei and occasional mitotic figures are found in the connective-tissue lining cells.

In the dense fibrous areas of the joint the surface is composed of a layer of collogenic tissue and the cells are for the most part imprisoned in lacunæ and buried

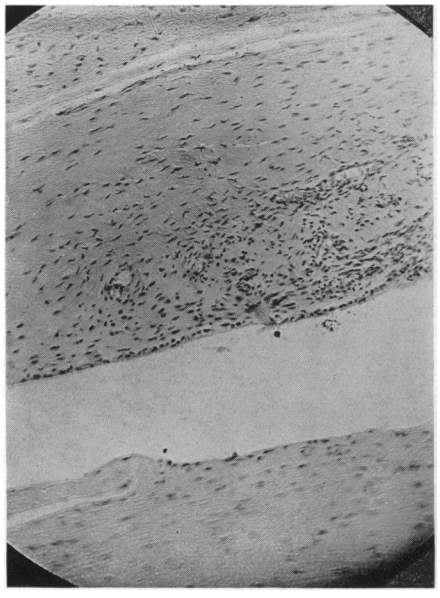

FIG. 4.

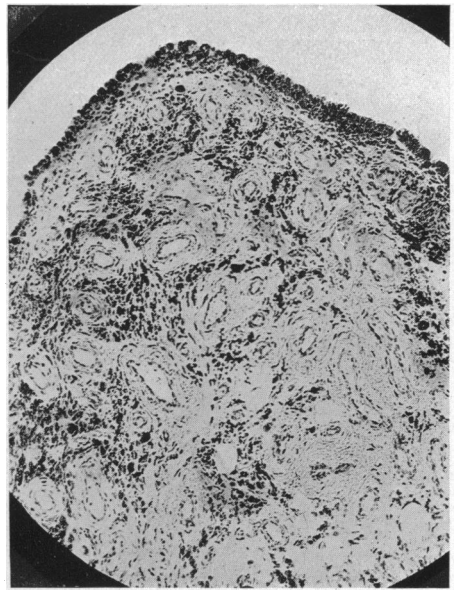

FIG. 5.

FIG. 4.—Fibrous areas of the synovial surface. Note that in the upper surface the cells are imbedded in lacunæ. Eight millimetres objective.

FIG. 5.—Vascular area of synovial surface. The dark material is blood pigment. Sixteen millimetres objective.

beneath the surface. (Fig. 4.) In spite of the fact that these cells are enclosed in lacunæ many of them are about twice as large as are synovial cells in similar situations in normal joints and are not only enlarged, but possess long processes, many of which extend up to the articular surface. None of these cells contains any of the yellowish pigment which is so abundant in the cells covering the areolar areas of the joint surface, nor is this dense tissue invaded by macrophages. In the transition zones between the fibrous and the areolar areas a few of the synovial cells contain minute granules of pigment.

Subsynovial Tissue.—In the loose or areolar areas of the joint surface the subsynovial tissue is extremely vascular and infiltrated with macrophages and contains large amounts of yellow pigment. (Fig. 5.) The ground work of this tissue is composed of rather coarse bundles of collagenic fibrous tissue separated by fat and areolar tissue. The macrophages are scattered everywhere through the tissue and

are in many places collected into masses. A large percentage of these macrophages are loaded with the yellow pigment. In addition to the macrophages there are a considerable number of small round cells or lymphocytes scattered through the subsynovial tissue and occasionally these are collected into small nodules similar to those seen in chronic arthritis, and sometimes these are perivascular in arrangement. The blood-vessels vary greatly in size and in the thickness of their walls. In many places the tissue is extremely vascular and wide blood spaces with very thin walls lie just beneath the layer of synovial lining cells. In other places the tissue contains large numbers of small thick walled arteries.

In addition to the blood-vessels the subsynovial tissue contains a large number of spaces which are lined by synovial lining cells. In the sections these spaces appear to be cysts in the subsynovial tissue, but examination of the gross specimen indicates that most, if not all, of the apparent cysts are really deep clefts in the subsynovial tissue

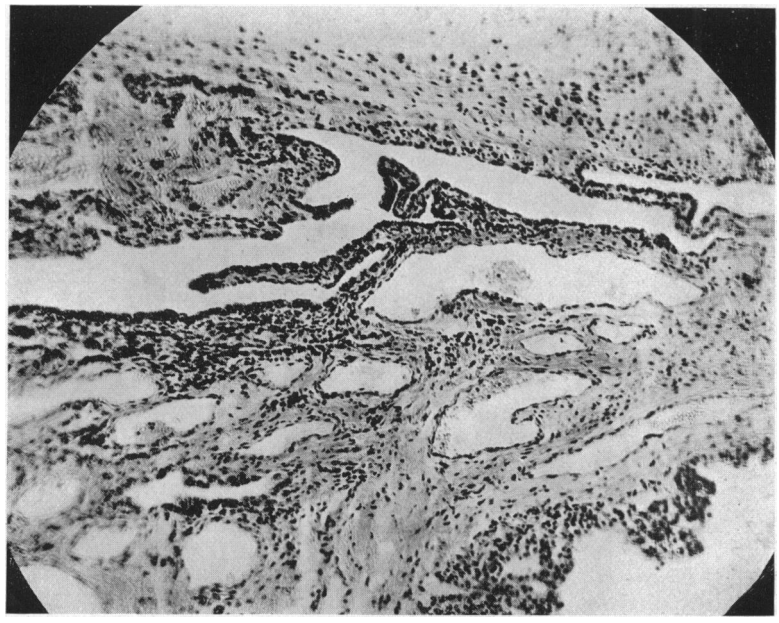

FIG. 6.—Cyst-like depressions in the synovial surface lined by synovial lining cells. These are believed to communicate with the joint cavity. Sixteen millimetres objective.

and that they communicate with the joint cavity and are lined by the same type cells as is the rest of the joint. (Fig. 6.)

The thickness of the layer of areolar subsynovial tissue varies greatly in different sections. In some places there is only a thin zone beneath the layer of lining cells (Fig. 7) while in others there is a wide vascular subsynovial layer. (Fig. 8.)

The subsynovial tissue rests on a layer of very dense collagenic fibrous tissue made up of bundles of fibres which run in various directions. The greater part of this dense fibrous tissue is not infiltrated by round cells or macrophages, but it contains occasional circumscribed areas into which the macrophages have penetrated and a few areas are seen in which the tissue is infiltrated by red blood-cells apparently the result of a recent hæmorrhage, and these are mixed with a moderate number of macrophages which are engorged with yellowish pigment.

Occasional clefts or possibly cysts lined by cells loaded with pigment granules penetrate deeply into the subsynovial fibrous tissue lying between collagenic bundles

and in other areas the tissue is made up of a mass of collagenic bundles which are separated by small blood-vessels and islets of engorged macrophages.

In the dense fibrous areas of the joint the collagenic fibrous tissue approaches or may even comprise the synovial surface and in these areas it is not infiltrated with macrophages.

The villi vary greatly in size and number and tend to resemble in structure the type of tissue from which they spring. They are most numerous in the loose areolar areas of the joint surface and here the villi are very complex in structure, containing many indentations and folds, and are extremely vascular and markedly infiltrated with macrophages. (Fig. 9.) In the areas where the subsynovial layer is very thin, the villi are short and slender and relatively simple in structure (Fig. 7), and in some parts of the dense fibrous area the villi are composed of dense fibrous tissue and contain no blood-vessels or macrophages.

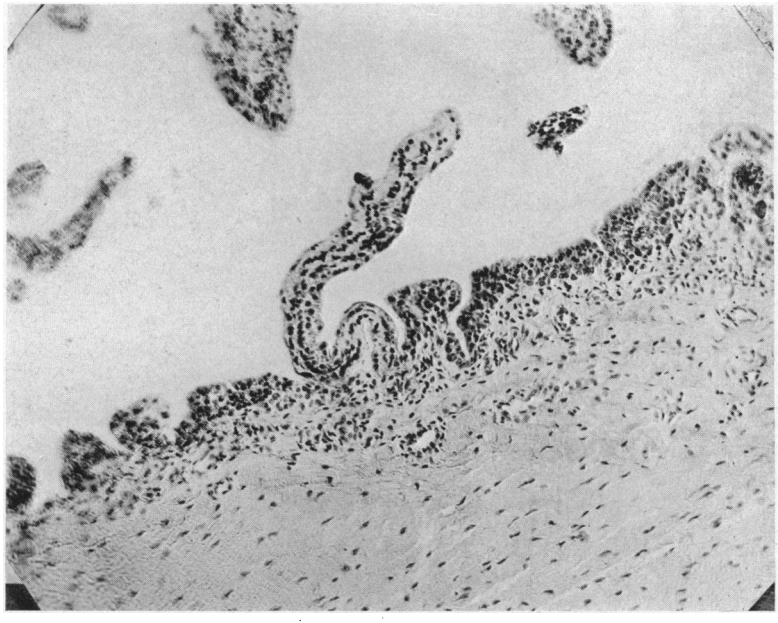

FIG. 7.—Transition zone between areolar and fibrous type of synovial surface. Eight millimetres objective.

The yellowish-brown pigment which is scattered through the tissues is obviously blood pigment and gives a strong iron reaction. The greater part of this pigment is found in the bodies of the macrophages but a considerable amount is lying free in the tissue in masses varying from minute granules to conglomerates fifteen or more microns in diameter. In addition to the pigment in the macrophages minute granules of pigment are present in the fixed connective-tissue cells and in the connective-tissue cells making up the walls of the blood-vessels and even in the endothelial lining cells of both artries and veins in areas where the tissue contains large amounts of pigment. In other words, in such areas practically every type of cell present contains some of the old blood pigment.

Cartilage Mœrgin.—The margin of the articular cartilage is overlapped by a layer of rather vascular synovial and subsynovial tissue. (Fig. 10.) This has invaded the cartilage for a short distance. The surface of the cartilage has undergone fibrosis and is invaded by vascular connective tissue. The synovial layer contains numerous small branching villi which are quite vascular and contain large numbers of macrophages

which are loaded with pigment. There is very little evidence of hyperplasia in the cartilage and the underlying bone is extremely atrophic so that in many places the cartilage practically rests upon the bone marrow and the cancellous bone of the epiphysis.

Later Condition.—June 10, 1931, the boy came in for an examination by request. At the present time he is fifteen years old, weighs 105 pounds and has just finished his first year of high school. He states that he has had no pain in the knee since leaving the hospital and that the operation has completely relieved his symptoms, with the exception of limitation of motion in the right knee. He walks three miles to school and during the past year he has missed only one-half day of school and this was because of a cold. He swims, plays ball, and does practically everything but skate. He still has a tendency to bruise after an injury, but this is not as severe as it used to be. Occasionally he hurts his fingers playing ball and they become swollen and painful for a few days, but the symptoms clear up spontaneously. From time to time he

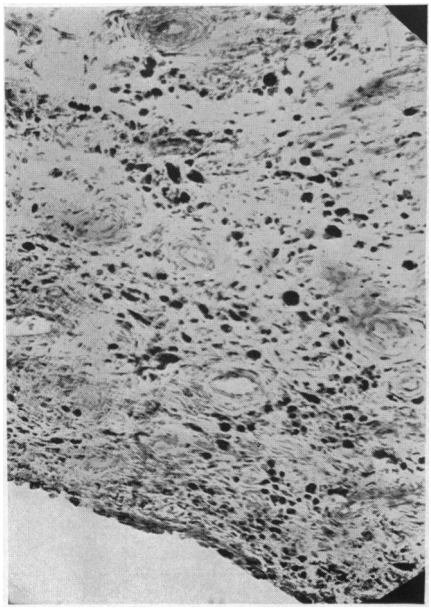

FIG. 8.

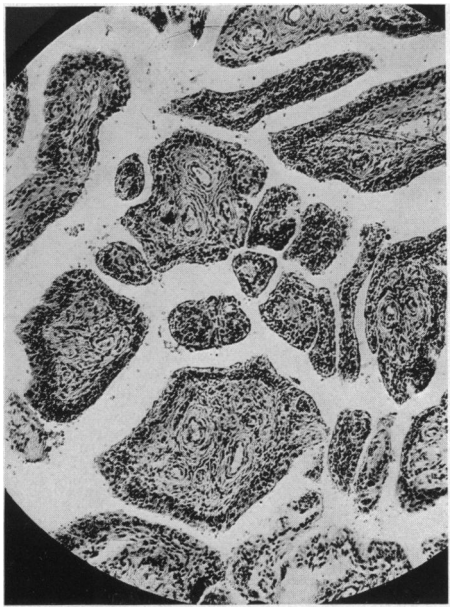

FIG. 9.

FIG. 8.—Areolar type of synovial surface. The large dark masses in the tissue are blood pigment and much of this is lying free in the tissue. Sixteen millimetres objective.
FIG. 9.—Cross-sections of multiple small vascular villi. Sixteen millimetres objective.

falls on his hands, and when this occurs the wrist, especially the left wrist, is apt to be swollen and painful for a few days, but this also clears up spontaneously. Six days ago he fell and injured his right hip. Two days later the hip became sore, stiff and painful and the hip is now in a position of flexion and he walks with considerable limp. He states that this is the first time that this hip has ever been injured and that up until this recent injury he was able to walk with scarcely a limp.

The patient is a tall, slender boy and apparently normal except for the right hip and right knee. (Fig. 11.) The right hip is flexed and moderately sensitive on movement. There is no swelling or external evidence of injury. The hip is slightly tender on deep palpation. There is permanent flexion of 60 degrees and flexion is limited to 90 degrees. Rotation is limited about 50 per cent.

The Right Knee.—There is permanent flexion of 25 degrees and the flexion is

limited to 90 degrees. There is a large, slightly tender scar over the anteromesial aspect of the knee and there is slight crepitus on motion. There is no pain in the knee with motion or weight bearing. There is some discoloration over the tibial tubercle which the patient states is due to a recent fall.

Blood-clotting time eight and one-half minutes by test-tube method (anything under fifteen minutes normal).

X-rays taken June 10, 1931, as compared with those taken before the operation, show decrease in the bone atrophy, disappearance of the thickened tissue around the internal condyles which was removed at the operation, increase in the size of the bones and slight increase in the irregularity of the articular surfaces. (Fig. 12.) An X-ray of the hip taken at this time is negative.

Historical.—The American literature on hemophilic arthritis is very meagre. With the exception of descriptions in text-books the only articles which I have found on the subject are those of Doub and Davidson,[2] Youmans,[3] and Wilson.[4]

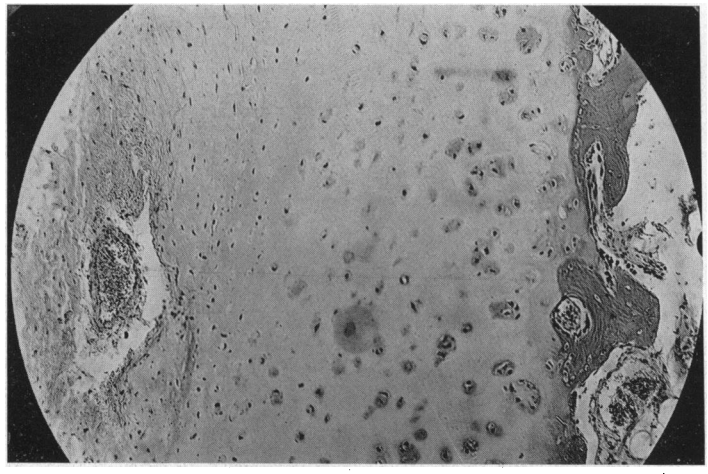

FIG. 10.—Articular cartilage near the margin. The surface on the left is being invaded by vascular connective tissue while the subchondral bone on the right shows marked atrophy. Eight millimetres objective.

As the German and French literature has been reviewed by König,[1] Linser,[5] Zesas,[6] Du Pan,[7] Freund,[8] and Reineke and Wohlwill,[9] it will be considered only briefly in this paper. The older authors considered the joint affections in bleeders as being due to rheumatism and Volkmann is given credit for first differentiating the joint disturbances in hemophilics from rheumatism. In his text-book on diseases of the bones and joints published in 1868 he stated that spontaneous hemarthroses occurred in scurvy and hemophilia and that they might also result from trauma. The next year Reinert and Gasses expressed the opinions that bleeders' joints were due to intra-articular hæmorrhage.

The modern conception of the hemophilic arthritis dates from König's paper which appeared in the Klinische Vorträge in 1892. He divided the condition into three stages: (1) Hemarthrosis, (2) panarthritis, and (3) regressive stage, and warned against operation under a mistaken diagnosis.

Pathology of Hemophilic Arthritis.—The gross changes in the joints were accurately described by König and relatively little has been added to his original description. The microscopical changes in the synovial membrane are most completely described in the pathological description of the material in my case, which is included in this paper, and the microscopical changes in the cartilage and bone are most completely described in the papers by

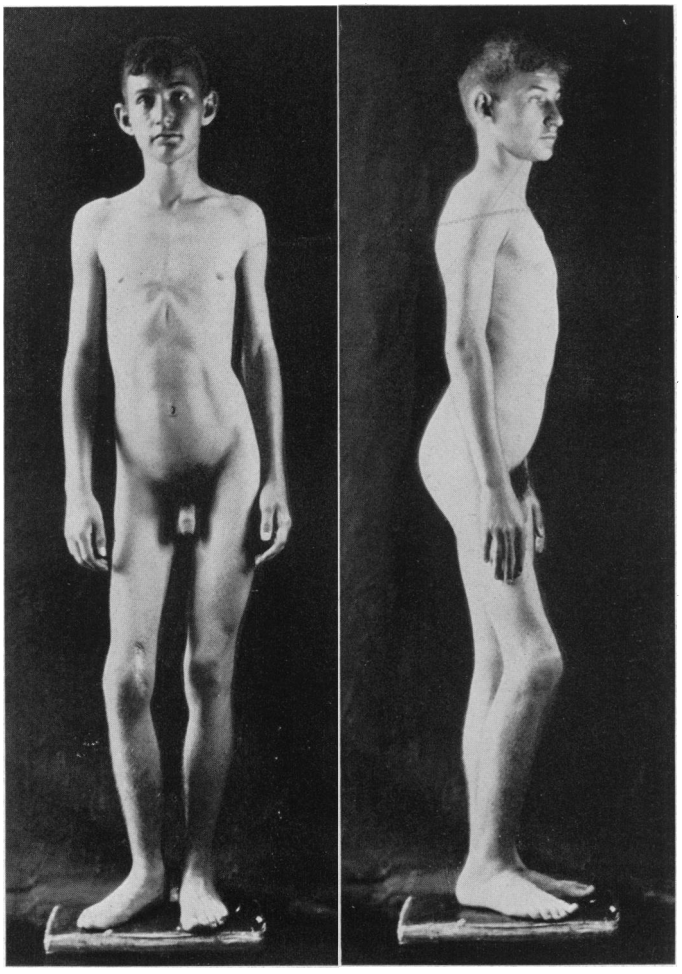

FIG. 11.—Photograph of patient one and one-half years after the operation. Note the flexion deformity of the right knee and a large scar over the internal condyle. When this photograph was taken there was an acute hemarthrosis in the right hip.

Freund[8] and Reineke and Wohlwill.[9] From the literature and my own observations the pathological changes can be described briefly as follows:

Changes in the Soft Tissues.—The hemophilic starts out with apparently normal joints. Due to some injury, or even without any known injury, bleeding into a joint may occur. This may begin early in life as in the case

illustrated in Fig. 13, where the first bleeding into the knee occurred at the age of two years and eight months. The joint becomes distended with blood under pressure which may be equal to the systolic pressure of the blood as the blood in the joint does not clot, at least not for a long time, and is in communication with that in the blood-stream.

In addition to its mechanical effect the blood in the joint acts as an irritant and hyperplasia of the synovial membrane occurs. As I have shown elsewhere,[10] a somewhat similar hyperplasia may be produced experimentally in animals by repeated injections of blood into normal joints. With this

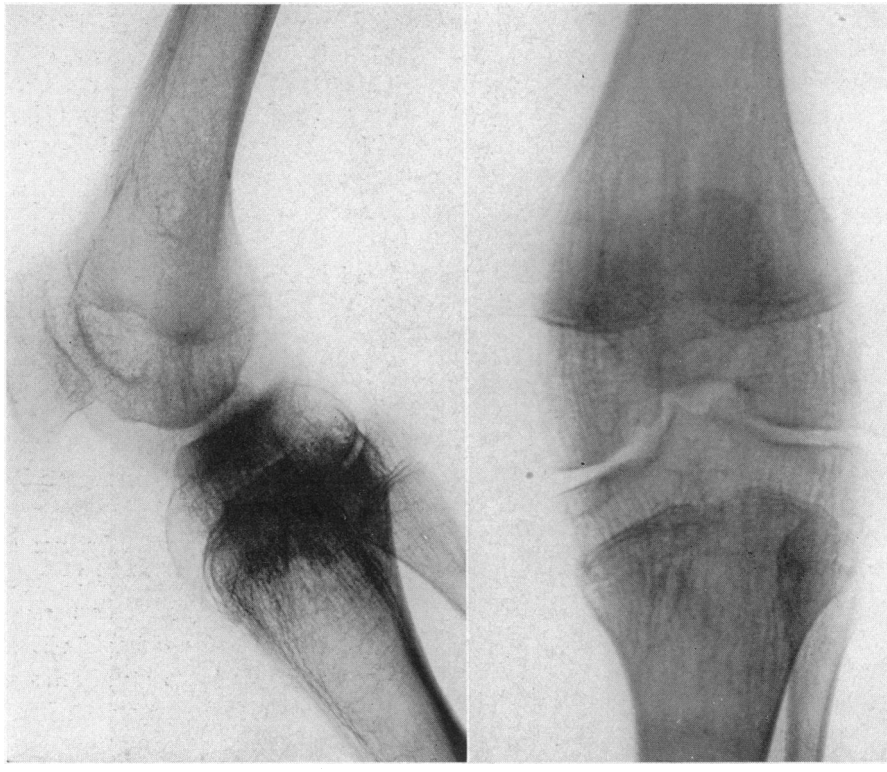

FIG. 12.—X-rays of operated case one and one-half years after operation. Note the increased roughening in the joint and the decrease in the bone atrophy. The large rarefied area in the femur persists and the opaque synovial tissue which was removed at the operation is absent in the film.

synovial hyperplasia there is an accumulation of macrophages in the subsynovial tissues and these are constantly wandering back and forth between the joint cavity and subsynovial tissues.[11] These macrophages phagocytize the red blood-cells, become engorged with them, and as the red cells disintegrate the blood pigment remains in the body of the macrophages. These engorged macrophages settle down in the subsynovial tissues beneath the fibrous capsule of the joint and eventually die, thus setting free the blood pigment. This blood pigment may remain *in situ* or may be picked up by other macrophages. Eventually the bleeding ceases, the blood in the joint

is absorbed and the joint tends to return to normal, but in a hemophilic the bleeding recurs from time to time and with each recurrence there is an addition to the amount of blood pigment which is laid down by the macrophages in the subsynovial tissues.

Each new hæmorrhage is an added irritation to the synovial membrane and this hypertrophies with the formation of folds and villi. Not only is the blood pigment taken up by the macrophages, but, in the advanced cases, as I have shown in this paper, practically all of the cells in the vicinity contain some of the fine pigment granules. In other words, the area around the joint is saturated with blood pigment. While some of this enters the lymphatics and general circulation much of it remains permanently in the tissues around the joint just as does India ink when it is injected into joints.[11] An idea of the large amount of pigment which may be deposited in some of these joints is conveyed by the fact that in one of Freund's[8] cases in which the synovial membrane was analyzed for iron it was found that 71 per cent. of the ash was iron. Eventually, if the tendency to bleed ceases, the synovial membrane tends to return to a more nearly normal state, the synovial cells decrease in size and number, and the villi tend to atrophy.

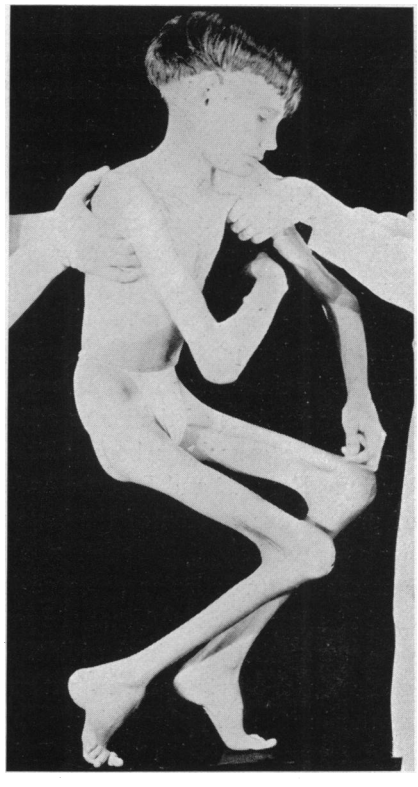

FIG. 13.—Photograph of a severe hemophilic seven years of age with very severe contractures of both knees.

The accumulation of blood pigment in the subsynovial tissues and probably the repeated occurrence of bleeding into a joint under pressure serves as an irritant which results in the formation of a layer of dense fibrous connective tissue in the subsynovial tissue. In the case on which I operated this resulted in a marked decrease in the size of the synovial cavity and the subsynovial fibrous tissue envelope around the joint was distinct from the fibrous capsule of the joint and separated from it by thin areolar tissue. In such a case a tendency to fibrous ankylosis is present and this fibrous ankylosis is partly due to thickening and shortening of the fibrous capsule and ligaments, and partly to the production of new fibrous tissue in an abnormal situation. Not only does the new fibrous layer limit motion in the joint, but it also inhibits the resorption of blood from the joint cavity.

Changes in the Cartilage.—The cartilage probably remains normal for

a long time, but eventually it becomes eroded around its margins by the encroachment of the hyperplastic synovial membrane just as occurs in any infectious or atrophic arthritis. In addition to the marginal destruction there is a variable amount of spotty destruction of the cartilage over the articular surface of the bones. These areas of destruction are preceded by death of the cartilage cells and fibrosis and degeneration of the matrix. Whether this is the result of the blood in the joint or whether it is the result of subchondral hæmorrhages is not known. However, the cartilage destruction does not coincide with pressure areas in the joint and the destroyed areas are irregular in contour and have been described by König[1] as map-like in character. This map-like appearance is, we believe, characteristic of a hemophilic joint. Where the cartilage has been destroyed the underlying bone becomes covered with a layer of connective tissue and there may result a depression or cavitation in the bone which may be filled with a blood clot or organized hematoma or lined by connective tissue. There is little tendency for the production of connective tissue between the articular surfaces of adjacent bones and so far as we know bony ankylosis has not occurred in any hemophilic joint.

Changes in the Bone.—The characteristic feature in the bone is that the intra-articular portions of the bone may contain cavities. These cavities may be in the nature of simple depressions either on the sides of the intra-articular portions of the bones or in the articular surfaces, or they may occur deep in the cancellous bone. The origin of these cavities is not definitely known. Freund[8] and Reineke and Wohlwill[9] believe that they are the result of progressive erosion of the bone which is brought about in the same manner as is the bone destruction by an aneurism. That is, they believe that the increased intra-articular pressure is a very important factor in the production of the cavities and that the underlying bone is gradually eroded by the pressure of the blood in the joint. In addition to this they believe that hemophilic blood exerts some as yet unknown chemical effect upon bone which results in its rapid absorption.

In support of this theory Reineke and Wohlwill[9] describe the marked erosion of the cortex of the femur which occurred beneath a subperiosteal hematoma from which the patient died and in which the autopsy specimen was carefully studied and described. I do not know whether either or both of these theories are correct. It seems to me, however, that the most plausible explanation of the bone destruction with cavitatian is that it results from intra-osseous hæmorrhage. As we know, the bone destruction does not occur until late in the disease. At some stage of the disease the bones become markedly atrophic as a result of disuse. It seems quite probable that intra-osseous hæmorrhage may be a fairly common phenomenon in atrophic bone and may result from ordinary use without definite trauma. In a hemophilic such an intra-osseous hæmorrhage would tend to progress and result in the production of an area of aseptic inflammation in the involved area, and this

in turn would tend to result in the absorption of the bone by the inflammatory tissue. Whether or not this theory is true we do not know.

The areas of bone destruction are frequently so extensive that they are clearly visible in the X-ray and when present are characteristic of advanced hemophilic arthritis. Not only are they present in the interior of the bone, but there is a subperiosteal erosion of the bone which results in a pseudo-lipping due to the undermining of the articular margins. Occasionally, as a result of loss of cartilage on the articular surface,[12] true marginal osteophytes or exostoses may occur in these joints, but these are relatively rare. If, as the patient grows older, the tendency to bleed into the joint becomes less or ceases there may result a joint which closely resembles that of the ordinary hypertrophic arthritis. On the other hand, with repeated bleedings and progressive destruction of the articular surface marked deformities may occur.

Clinical Picture.—The clinical picture varies with the stage of the process and most authors follow the classification of König, who recognized three stages: (1) Hemarthrosis, (2) panarthritis resembling tuberculosis, and (3) regressive stage with erosion of the joint borders. I am not able to separate the panarthritic from the regressive stage and do not think that this has been done successfully by König or any other author whom I have consulted. Consequently, only two types of bleeders' joints will be recognized: (1) Acute hemarthrosis and (2) chronic arthritis.

(1) *Stage of Acute Hemarthrosis.*—This is the stage of acute hæmorrhage into a joint which in other respects may be practically normal or may be the site of a chronic hemophilic arthritis. The joint disturbances in hemophilia nearly always begin in childhood and the surgeon may see the patient during the first attack, but it is usually possible to obtain a history of similar attacks in the involved joint or in other joints, and it will be found that these previous disturbances either cleared up spontaneously or after a few days' rest and that the involved joints returned to normal or a chronic arthritis developed. The joint disturbance may have occurred spontaneously, even while the patient was asleep in bed, but in most instances it will have followed a minor injury such as a contusion or unusual strain.

It is characteristic for the effusion to appear rapidly and marked swelling may occur within a few minutes so that the patient may even see the joint swell, as sometimes occurs in acute Charcot joints. In some instances the swelling is a very gradual process and may progress slowly over a period of several hours or days. The degree of swelling ranges from a joint containing a slight amount of excess fluid to one which is extremely tense and distended with fluid under considerable pressure. (Fig. 14.) In a case cited by Reineke and Wohlwill[9] the intra-articular pressure was so great that the blood spurted over two neighboring beds when a needle was inserted into the knee.

Pain may be slight or intense and tends to vary directly with the degree of swelling and pressure within the joint. The same is true of the func-

tional disability. These joints vary from those with slight swelling and no limitation of motion or pain on motion or even on weight bearing to those which are greatly swollen, very tense and painful, and in which weight bearing is prohibited by pain and practically all motion is prevented by pain and muscle spasm.

On physical examination the patient is usually a slender, pale, anæmic-looking male and presents nothing remarkable except the involved joint. The findings in the involved joint vary with the amount of the intra-articular hæmorrhage and the tension on the capsule. If the amount of excess fluid is not great the joint may show only slight swelling and in the case of a superficial joint the signs of excess fluid in the joint are present. If the

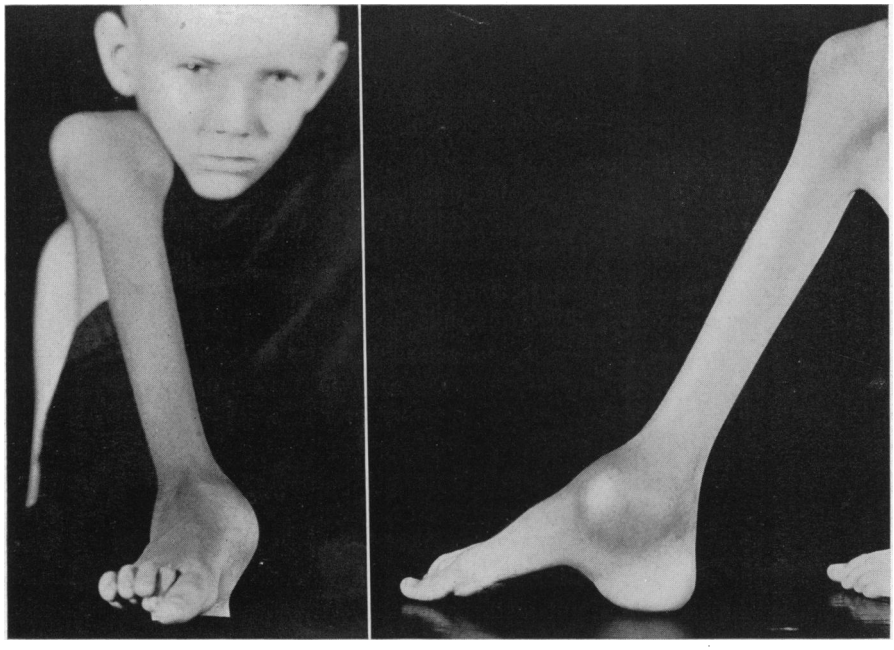

FIG. 14.—Acute hemarthrosis in right ankle. Same case as preceding.

blood is present under pressure the joint may be markedly swollen and acutely tender and the tenderness may extend up and down the limb. (Fig. 14.) Local heat and redness are not present, but a joint which is covered by a relatively thin layer of tissue such as a finger or an ankle-joint may be definitely blue (cyanotic) in color.

In the severe cases the joint is fixed in a position of moderate flexion by muscle spasm and both active and passive motions are limited. There may be some local heat, but redness and œdema are not present. As a rule the bleeding is into the joint and the swelling follows the outline of the synovial cavity, but there may be more or less extra-synovial hæmorrhage and in such cases dark bluish areas may appear in the subcutaneous tissues near the joint. These subcutaneous discolorations resemble the bruises which are so

frequent in hemophilics and are most evident three or four days after the beginning of the joint bleeding. Unless the joint is involved in a chronic arthritis there is no muscle atrophy or deformity other than that incident to the muscle spasm in the severe cases.

The temperature tends to be slightly elevated and in exceptional cases may reach 39 or 40 degrees C. and there is usually a moderate leucocytosis, but the prostration found in acute infectious processes is absent.

In the röntgenogram the bones tend to be normal, but the joint cavity is distended with fluid and this, being blood, casts a shadow which is rather more dense than that usually seen in acute synovitis.

(2) *Chronic Arthritic Stage.*—This may be defined as the stage in which the involved joint fails to return to an apparently normal condition after the hæmorrhage. It may follow the first hæmorrhage into the joint, as occurred in the knee of the operated case reported in this paper, but this is unusual, and permanent changes in the joint do not ordinarily occur until after several such attacks. A rather extreme case is that reported by Gocht,[13] in which hemarthrosis of one knee had occurred forty-five times, but the blood had always been resorbed and normal function restored.

In the arthritic stage it will be found that after an acute hemarthrosis the joint remained swollen, tender, sore, and painful for several weeks or months and that there is more or less permanent disturbance of function with a tendency to repeated attacks of acute swelling and pain which are usually the result of minor injuries. It should be pointed out that this stage resembles ordinary chronic arthritis in that it tends to progress by remissions. That is, the joint quiets down after an acute attack and there is more or less restoration of function, but the joint is not so good as it was before the attack and with recurring attacks there is an increased permanent disability.

During an acute attack in the arthritic stage the joint is swollen, painful, tender, and disabled as in the stage of hemarthrosis. The difference between the two stages is that in the arthritic stage the swelling tends to be less because it is limited by the subsynovial fibrous tissue and the blood in the joint is not resorbed promptly, and contracture, deformity, peri-articular thickening and muscle atrophy are present and tend to increase after each fresh hæmorrhage.

After the acute hæmorrhage has subsided, the muscle atrophy, deformity, limited motion, and disability persist. The joint is more or less swollen and this is partly due to a small amount of excess fluid (blood) which is almost constantly present in these joints, but largely to thickening of the periarticular tissues. This thickening follows the outline of the synovial cavity and on palpation the indurated subsynovial tissue may be felt. In addition to the above there may be more or less actual thickening of the bones entering into the joint and this may be general or local from the ossification of old subperiosteal hematomas.

In the regressive stage of König the tenderness and pain on motion have

largely disappeared, but there is soft crepitus on motion and the amount of motion is more or less limited. In severe cases there may be fibrous ankylosis with practically no motion in the joint, but I know of no case in which true bony ankylosis has occurred.

The most severe deformities which I have seen in hemophilic arthritis are illustrated in Figs. 13 and 15. This seven-year-old boy is a severe familial hemophilic and trouble with the knees began at the age of two years. On admission there was permanent flexion of 90 degrees in one knee and of 110 degrees in the other. On account of the deformities the patient has not walked for over five years. The parents did not wish us to attempt to

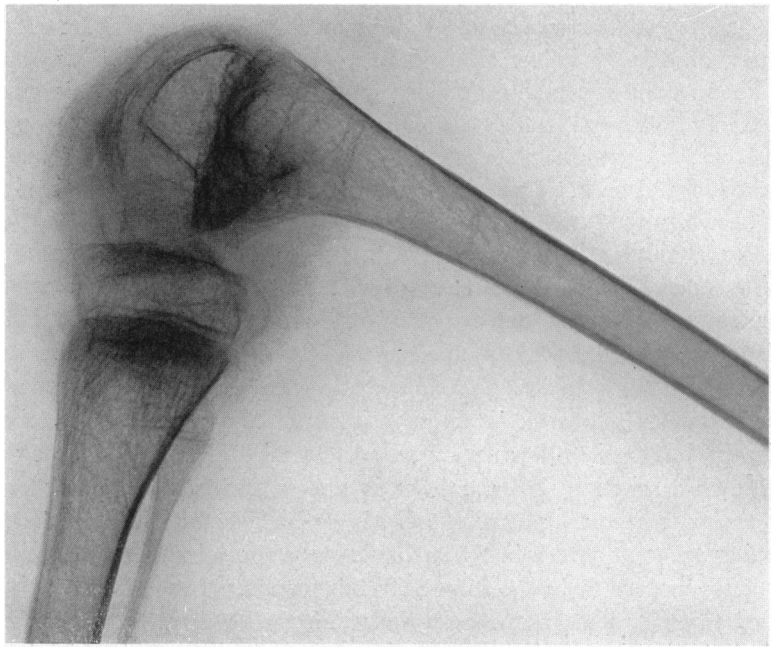

FIG. 15.—X-ray of knee-joint of preceding case. Note the marked atrophy in the bones, roughening of the joint surfaces and cloudiness of the joint capsule.

straighten the knees, but brought the boy to the Shriners' Hospital because of the acute hemarthrosis of the ankle. (Fig. 14.) A few days after his admission, while lying in bed the patient developed acute hemarthrosis in one elbow, and one finger and a moderate hematoma behind one ear and a very large hematoma (about five by ten inches) on the back and right shoulder. All of these were present at one time.

The knees of the above case resembled those of a typical case of Still's disease (atrophic, rheumatoid, or chronic infectious arthritis). The knee of my operated case resembled a traumatic arthritis and the knees of the man whose X-rays are shown in Figs. 16 and 17 resembled a hypertrophic arthritis. During the intervals between attacks this man had relatively little disability and on physical examination the knees presented only moderate

periarticular thickening and soft crepitus on motion and slight limitation of motion.

It is thus evident that the chronic hemophilic joint may present a very variable clinical picture and that these chronic joints are subject to acute hæmorrhages which result in periods of marked disability (panarthritis of König) which may be prolonged or clear up within a week or two and that after these acute symptoms have cleared up the joint returns to the original relatively painless chronic arthritic stage (regressive stage of König). But the acute symptoms may return at any time. As the patients grow older the hæmorrhages tend to become less frequent. For instance, the man whose knee is shown in Figs. 16 and 17 now goes two years or more without trouble, whereas during childhood he had several acute hemarthroses a year, and the same is true of my operated case.

The temperature and blood picture tend to be normal except during an acute hæmorrhage into the joint when there may be more or less fever and a moderate leucocytosis. The blood-clotting time tends to be prolonged to a variable degree.

Röntgenographical Appearance.—The X-ray findings have been described by Gocht,[13] Mermingas,[14] Hubscher,[15] Neumann,[16] Mankiewicz,[17] Rhonheimer,[18] Montonari,[19] Engels,[20] Dun Pan,[7] Petersen,[21] Carry,[22] Freund,[8] Doub and Davidson,[2] and Reineke and Wohlwill.[9] In the acute hæmorrhagic stage there are no changes in the bones, but the blood in the joint casts a shadow which is slightly more dense than that of the usual synovial effusion.

In the arthritic stage the X-rays reveal abnormalities in the soft parts and bones which have been variously described as resembling the changes produced by tuberculosis, hypertrophic arthritis, atrophic arthritis, or traumatic arthritis. As a matter of fact, while they may present some of the changes found in any of the above conditions the röntgenograms of a typical case of hemophilic arthritis may present characteristic features which make it possible to make the diagnosis from the X-ray alone. These features are a markedly increased density of the synovial tissues and crater-like depressions or punched-out defects in the intra-articular portions of the bones. However, many cases do not present these characteristic features and these cannot be diagnosed by the X-ray alone.

The increase in density in the synovial tissues often takes the form of definite shadows resembling areas of calcification. These shadows are present in the recesses of the joint and follow the outline of the synovial cavity. A careful examination of the affected knee-joints in our operated case gives one the impression that the synovial membrane is thickened and more or less calcified. These shadows are much more definite and sharply outlined than is the cloudiness of the joint space in tuberculosis and are more regular in outline and general in distribution than are the extra-articular calcified masses in Charcot joints. In addition to the above the patella may float on account of effusion into the joint and the patellar tendon may be bowed forward by the fluid or hypertrophied fat pad beneath it.

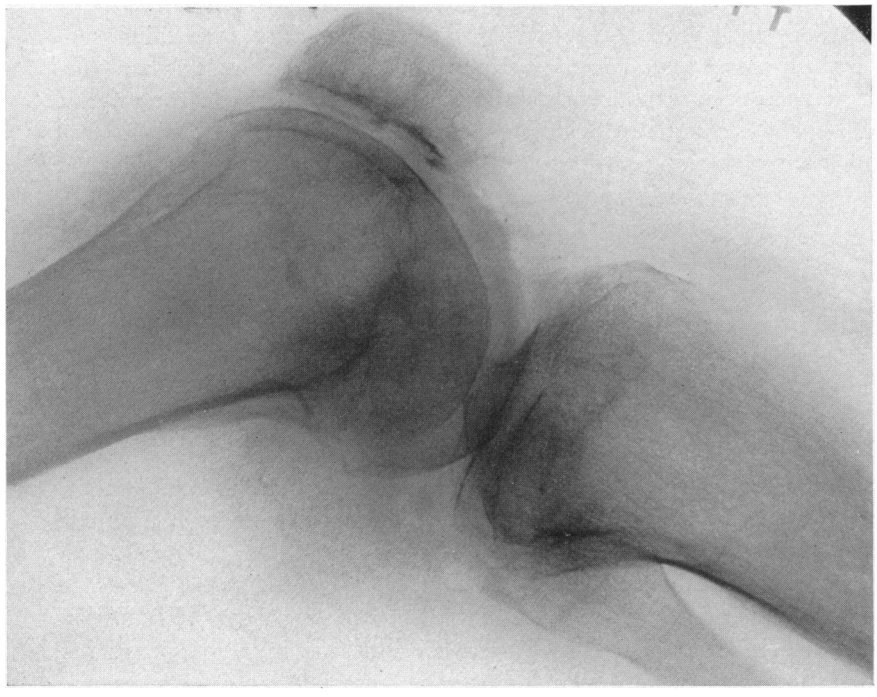

FIG. 16.

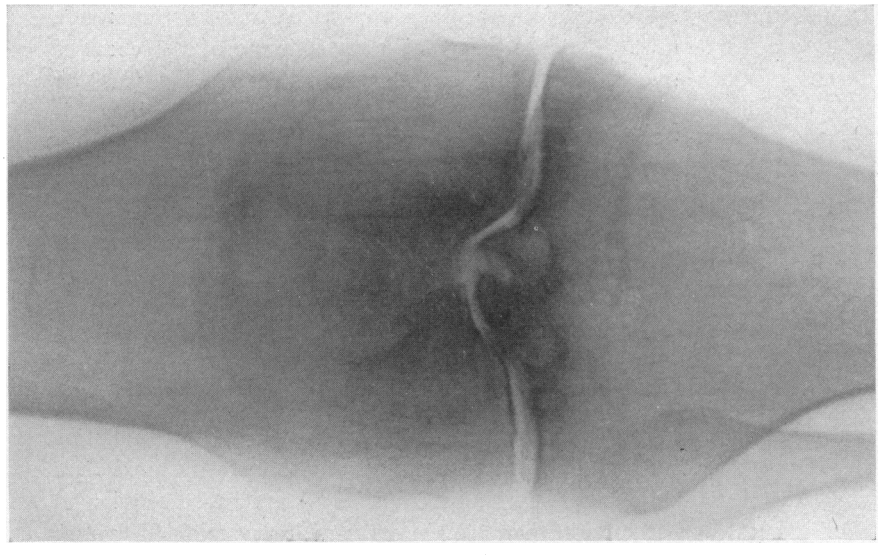

FIG. 17.

FIG. 16.—X-ray of knee-joint in a man forty-one years old. Chronic hemophilic arthritis. Note the resemblance to chronic hypertrophic arthritis.

FIG. 17.—Lateral view of same case as preceding. Note the cloudiness in the joint capsule and the punched-out area in the articular surface of the patella

The joint space is narrowed to a variable degree and may be obliterated, but bony ankylosis apparently does not occur. The articular surfaces of the bones are more or less irregular and indented and the ends of the bones entering into the joint are more or less deformed. In the knee the condyles, especially of the femur, may be broadened or less broad than normal. A rather characteristic picture is one in which the sides of the condyles of the femur and tibia present definite hollowed-out defects just above or below the articular margin giving one the impression of border osteophytes. And border osteophytes have been described in these joints, but they are quite unusual. In knees presenting these lateral defects the notch between the femoral condyles is usually broader and deeper than normal and the spines of the tibia are apt to be deformed. There may be considerable new bone formation from the calcification of a subperiosteal hematoma and this is especially apt to occur in the elbow. In the hip the head of the femur may be deformed in such a manner that the X-ray picture resembles that of Legg-Calve-Perthes' disease (osteochondritis deformans juvenilis).

Many authors have described atrophy of the bones, and this is generally present (Fig. 15), but if the arthritis is quiescent and the patient has been using the extremity for some time the bones may be of approximately normal density. (Figs. 16 and 17.)

The characteristic rarefied areas are described by Engels[20] as opening into the joint with crater-like formations, but they may be deep in the bone and well removed from the joint space, and here they resemble the punched-out areas such as are produced by myeloma or metastatic carcinoma. In my cases they have been surrounded by a shell of bone which is slightly more dense than is that in the vicinity. They may be present in elbows and other joints, but are most marked in the knees.

Diagnosis.—It is not especially difficult to arrive at a correct diagnosis if the surgeon thinks at all of the condition. The great difficulty is that the possibility of encountering a hemophilic joint does not occur to the surgeon and he may not learn that the patient is a bleeder until after the joint has been opened. At least this has been the state of affairs in most of the cases in the literature which were operated upon and died from hæmorrhage following the operation (König,[1] Tillmann,[23] Zielewicz,[24] Froelich,[25] Zesas,[6] and others). As a rule, these patients were orphan boys.

In my case the operation was the result of operating upon a patient after a casual examination, the diagnosis having been made by others. However, unless I had obtained a history of hemophilia I do not believe that I would have suspected a hemophilic arthritis in my case and would have operated just the same, as my knowledge of the subject was very casual until my error caused me to study it.

In the Stage of Acute Hemarthrosis.—This is to be differentiated from traumatic synovitis, acute rheumatic fever, acute pyogenic arthritis, gonorrhœal arthritis and osteomyelitis.

The most characteristic feature of an acute hemophilic hemarthrosis is the

sudden onset and rapid progress of the condition either spontaneously or after a mild injury. If the injury has been severe enough to lead the surgeon to suspect a traumatic synovitis, the swelling and effusion into the joint will occur more rapidly and will be greater than one would expect to find after such an injury and the joint capsule will be unusually tense and the patient will complain of pain even when the joint is at rest.

In acute rheumatic fever, acute pyogenic arthritis and acute osteomyelitis, the onset is more gradual and the pain is the first symptom while the swelling comes later. Likewise, in these conditions the temperature is apt to be higher, 103° or more, and the leucocyte count is usually elevated above 15,000. In hemophilic hemarthrosis the temperature and leucocyte count are as a rule only moderately elevated (100° to 101° and 10,000 to 12,000 white blood-cells).

In all of the above conditions there is usually some increase in the local temperature, but in hemophilic hemarthrosis the skin over the involved joint is normal in temperature and color unless it has been injured, but may appear cyanotic if the joint is close to the surface, while in the other conditions there is usually more or less redness of the skin.

Gonorrhœal arthritis resembles an acute pyogenic arthritis except that the onset is usually more gradual and the general and local symptoms are more mild in character. The gradual onset will usually distinguish it from a hemophilic hemarthrosis and in addition one may obtain either a history or physical evidence of gonorrhœa.

From what has been said above, the diagnosis may seem comparatively easy and yet if one be confronted with a flexed hip in which all motions are limited by muscle spasm and which is acutely painful, tender, and sensitive and the patient has a temperature of 102.5° F. and a white blood-cell of 15,000 and gives a history of having been perfectly well the day before one is not apt to think of hemophilic hemarthrosis, which may be the condition present, and such hips have been operated upon and fatal hæmorrhages have resulted.

The most important diagnostic feature is the history, and most hemophilics know that they are bleeders and will tell the surgeon so, or their families know it and will volunteer the information and it is thus that many surgical catastrophies have been averted. As was stated above, most of the operated cases have been young orphan boys.

In any joint condition it is always advisable to inquire whether or not a similar condition ever occurred in the affected joint or in other joints. And in the hemophilic this question will usually be answered in the affirmative because most of these patients have had hemarthroses in various joints from time to time and will usually tell the surgeon that the symptoms tend to clear up after a few days' rest. Such a statement made by a pale, slender boy should always lead the surgeon to suspect and rule out hemophilia by a careful history and accurate determination of the coagulation time of the blood.

Finally, the diagnosis must be made by the history and a determination of the coagulation time of blood, and this is most accurately done by the test-tube method. However, it is to be emphasized that a normal coagulation time does not rule out hemophilia as the coagulation time undoubtedly varies from time to time in hemophilics. For instance, by the test-tube method under fifteen minutes is considered normal. In my operated case the coagulation time varied from seven and one-half to eight and one-half minutes.

In suspected cases the joint should be aspirated with a small needle and if blood is obtained and acute traumatic synovitis can be ruled out, the diagnosis of hemophilic hemarthrosis is fairly certain.

In the Stage of Chronic Arthritis.—The symptoms may be severe or relatively mild depending upon whether or not an acute hæmorrhage has recently occurred in the joint. In the relatively quiescent state during the interim between acute attacks the joint is moderately swollen and the swelling is found to be largely the result of periarticular thickening, but the joint may also contain a small amount of excess fluid. There is no local heat or redness and the joint is not, as a rule, especially tender. There is a variable degree of flexion deformity and motion is limited, but ankylosis is not present. Function of the joint may or may not be painful and there is moderate atrophy of the muscles of the limb.

With an acute hemarthrosis the symptoms resemble those of that stage as described above. After a period of time which may last for several weeks these acute symptoms gradually subside and the joint returns to the relatively quiescent state as described above and this may last for months or years and the patient be relatively free from symptoms until a fresh hæmorrhage occurs in the joint.

The X-ray shows narrowing of the joint space, a variable amount of erosion around the articular margins and a variable amount of bone atrophy with cloudiness of the joint area due to the shadow cast by the non-bony contents of the articular capsule.

The above descriptions might serve equally well for chronic tuberculosis of the given joint and almost as well for certain low-grade monarticular arthritic conditions of traumatic or unknown etiology. As a matter of fact, most of the cases which have been operated upon were operated upon with the erroneous diagnosis of tuberculosis.

How, then, is one to differentiate the chronic hemophilic joint? An accurate history of the onset and course of the disease is the most important factor in the diagnosis. In the hemophilic joint the onset of the trouble can always be traced back to an acute swelling of the joint which may or may not have followed an injury and there will be a history of many such episodes. Usually there will have been several attacks of hemarthrosis before the true chronic arthritis began, the joint having returned to a normal condition after the earlier attacks, but occasionally, as in my operated case, the arthritis may date from the first attack. In such cases one can obtain a history of repeated acute attacks since the onset of the arthritis and the patient will state that the

acute symptoms subsided after a few days' rest. And in addition to the chronic arthritis, which may be monarticular or involve two or three joints, there is a history of milder disturbances in other joints.

Such a history should lead the surgeon to suspect a hemophilic joint. Of course, if the surgeon should learn that the patient is a bleeder, the diagnosis is clear. If there is a history of abscess formation in the joint, hemophilia may be ruled out as these joints do not suppurate.

An important point in the X-ray diagnosis is that the shadow cast by the thickened synovial tissues and capsule is more dense and sharply defined than is the case with any other type of arthritis with which I am familiar. However, I have seen X-rays of hemophilic arthritis of many years' standing in which the X-ray picture closely resembled that of chronic hypertrophic arthritis, and Petersen[21] reports cases in the hip which closely resembled old Legg-Calve-Perthes' disease (osteochondritis deformans juvenilis). The areas of rarefaction in the depths of the cancellous bone which are regarded as pathognomonic by Engels[20] are not always present and it should also be noted that such areas may occasionally be seen in tuberculosis or in arthritis from other causes. Consequently, one must conclude that in many, and perhaps in the majority of cases the diagnosis cannot be made from the X-ray alone. In my operated case the röntgenologist's diagnosis was chronic arthritis and a calcified hematoma.

In case a hemophilic arthritis is suspected, the clotting time of the blood should be ascertained in order to confirm the diagnosis; but, as was stated above, this may be misleading.

Treatment of Acute Hemarthrosis.—With acute hæmorrhage into a large joint rest to the involved joint is indicated and if necessary the patient should not only be put to bed, but the joint should be splinted or even immobilized in a plaster-of-Paris bandage until pain disappears and the blood in the joint cavity is largely absorbed. There is also the question as to whether or not these joints should be aspirated. At times they are intensely painful and in such cases aspiration has been carried out without ill effects. However, this is not a procedure without a certain amount of danger, as illustrated by the case of Petersen[21] in which the joint was aspirated and then continued to bleed from the puncture wound until it eventually became infected and the patient died with a streptococcus infection. Consequently, if aspiration is decided upon it should be done with the full understanding on the part of the patient or the patient's parents that it is not without a certain amount of risk and the aspirating needle should be of very small bore. On the other hand, in my case after operation, aspiration with a twenty-two-gauge needle was done on three separate occasions with no ill effects, but at that time in my case the blood-clotting time was within normal limits.

When the swelling disappears from the joint, function may be resumed, but the patient should be cautioned against indulging in activities which would tend to result in traumatism of any sort.

Chronic Hemophilic Arthritis.—In the chronic stage of the disease the

treatment may be divided into two phases: (1) The correction of deformities, and (2) support of the involved joint.

Correction of Deformities.—Most of these knee-joints, and occasionally other joints, develop severe deformities as in the case illustrated in Fig. 13 in which the flexion contracture of both knees is beyond 90 degrees. In the majority of cases, however, the deformities are not so severe. No attempt should be made to correct these deformities unless the deformity is a definite handicap to the patient. If this is true the deformity should be corrected by conservative means and operations upon the joints should not be done. In other words, the deformities may be corrected by traction or by wedging plasters or by mechanical appliances in which by means of pressure and counter-pressure with or without traction the contracted tissues are gradually stretched and the joints are straightened. After the deformities have been corrected the limb must be maintained in the corrected position over sufficient time to prevent recurrence.

Support to the Involved Joints.—As a rule these joints are not painful except during the stage of acute disability, and no support is necessary. Occasionally, however, in the case of a knee or an ankle, an elastic bandage may give the patient considerable comfort. Very rarely is a supporting splint such as a Thomas walking caliper indicated.

Severe Hæmorrhage.—In the great majority of instances the bleeding into a joint is not sufficient to materially deplete the blood volume of the patient and tends to cease spontaneously with rest. However, alarming or even fatal hæmorrhages may result from accidents or operations and subcutaneous hematoma of great size may occur spontaneously. In such cases it is imperative that the blood coagulation time be reduced to normal. There are various preparations on the market which tend to hasten the coagulation of the blood, but in a surgical emergency reliance is to be placed on a transfusion of whole blood from a matched donor. This is the method of choice as it not only reduces the coagulation time of the blood, but also restores the blood lost by the hæmorrhage.

The Hemophilia.—At the present time we have no acceptable treatment for this condition. In a recent communication by Birch[26] it is shown that the failure of the blood to clot is due to an abnormal toughness of the blood platelets in that they are highly resistant to hypertonic salt solution and fail to disintegrate when the blood is shed. Since hemophilia does not occur in the female, Birch[26] has treated two cases with ovarian extract and has reported that the tendency to bleed has been controlled by this method for periods of eleven and five months respectively.

This is not a new idea as some years ago, while working at the Boston Children's Hospital, I administered ovarian extract to two hemophilics under the direction of the late Dr. James S. Stone, who was then Chief of the Surgical Service. So far as we were able to determine in our cases the ovarian extract made no difference in the condition. It is possible that we did not use the right product.

On the other hand, it should be pointed out that the degree of hemophilia varies from time to time in given patients and while the report of Birch is important and the method should be tried we reserve our opinion until further cases have been reported and these cases have been followed over a longer time. In our operated case, as stated, we were able to obtain a large increase in the number of blood platelets by the administration of irradiated ergosterol. Whether or not this increase in the number of blood platelets will result in materially shortening the clotting time has yet to be determined.

BIBLIOGRAPHY

[1] König, Franz: Die Gelenkerkrankungen bei Blutern mit besonderer Berucksichtigung der Diagnose. Klinische Vortrage, N. F. Nr. xxxvi (Chirurgie, Nr. xi), pp. 233–243, 1892.

[2] Doub, and Davidson: Röntgen-Ray Examination of the Joints of Hemophilics. Radiology, vol. vi, p. 217, 1926.

[3] Youmans, J. B.: Report of a Case of Hemophilia with Joint Involvement. Wisconsin Medical Journal, vol. xviii, p. 257, 1919–1920.

[4] Wilson, M. G.: Hemophilic Arthritis. Amer. Jour. Dis. Child., vol. xviii, p. 127, 1919.

[5] Linser, P.: Beitrag zur Kasuistik der Blutergelenke. Beitrage zur Klinische Chirurgie. Bruns, vol. xvii, pp. 105–123, 1896.

[6] Zesas, D. G.: Über Gelenkerkrankungen bei Blutern. Fortschr. d. Med., vol. xxiii, p. 293, 1905.

[7] Du Pan, Martin: De l'arthropathie hemophilique. Revue Medicale de las suisse Romande, vol. xxxv, p. 547, 1915.

[8] Freund, E.: Die Gelenkerkrankungen der Bluter. Archiv. fur pathologische anatomie und physiologie, vol. cclvi, pp. 158–189, 1925.

[9] Reinecke, and Wohlwill: Hemophilic Arthritis. Arch. f. klin. Chir., vol. cliv, p. 425, 1929.

[10] Key, J. A.: Experimental Arthritis: The Reaction of Joints to Mild Irritants. Jour. Bone and Joint Surgery, vol. xi, p. 705, 1929.

[11] Key, J. A.: The Mechanisms Involved in the Removal of Colloidal and Particulate Carbon from Joint Cavities. Jour. Bone and Joint Surgery, vol. viii, No. 3, pp. 666, 1926.

[12a] Key, J. A.: Pathologic and Experimental Observations on Hypertrophic Arthritis. American Medicine, vol. xxv, No. 10, pp. 610–621, 1930.

[12b] Key, J. A.: Traumatic Arthritis and the Mechanical Factors in Hypertrophic Arthritis. Jour. of Lab. and Clin. Med., vol. xv, No. 11, p. 1145, 1930.

[12c] Key, J. A.: Experimental Arthritis: The Changes in Joints Produced by Creating Defects in the Articular Cartilage. Jour. Bone and Joint Surgery. In Press.

[13] Gocht, Hermann: Über Blutergelenke und ihre Behandlung. Archiv. fur Klinische Chirurgie, vol. lix, p. 482, 1899.

[14] Mermingas, K.: Beitrag zur Kenntniss der Blutergelenke. Archiv. fur Klinische Chirurgie, vol. lxviii, p. 188, 1902.

[15] Hubscher: Zur Behandlung des kontrakten Blutergelenkes. Korresp.-Bl. Schweiz. Arzte., vol. xl, No. 12, pp. 353–360, 1910.

[16] Neumann: Über Blutergelenke. Inaug.-Diss. Berline, 1913. (Quoted from Freund.)

[17] Mankiewicz: Berlin Klin. Wochenschr., 1913.

[18] Rhonheimer: Arthritis Deformans Juvenilis und Blutergelenke. Munch. med. Wochenschr., vol. lxiv, No. 2, pp. 1392–1395, 1917.

[19] Montonari, U.: Delle artrite emofiliache. Chir. degli org. mov., vol. vi, No. 2, p. 212, 1922.

[20] Engels, Hermann: Über das Blutergelenk und sein Röntgenbild. Fortschritte auf dem gebiete Röntgenstrahlen, vol. xxv, p. 197, 1917–1918.
[21] Petersen, O. H.: Das Blutergelenk und seine Beziehungen zu den deformierenden Gelenkerkrankungen. Archiv. fur. Klinische Chirurgie, vol. cxxvi, p. 456, 1923.
[22] Carry, M.: Arthrites Hemophiliques Chroniques. Presentation de Malade. Lyon Medical, vol. cxxxix, p. 631–633, 1927.
[23] Tillmann: Zur Frage der Blutergelenke. Deutsche Aerzte-Zeitung, vol. ii, pp. 421–424, 1900.
[24] Zielewicz: Traumatischer Hämarthros des Kniegelenke bei einem Bluter. Zentralble f. Chirurgie, vol. vii, p. 243, 1880.
[25] Froelich: Abnorme Formen von Blutergelenken. Zeitschrift fur orthopedische chirurgie, vol. xiv, pp. 600–610, 1905.
[26] Birch, C. L.: Hemophilia. Proc. Soc. Exper. Biol. and Med., vol. xxviii, p. 753, April, 1931.