in young women and causes the most profuse hæmorrhage, and I have often had such cases sent to me as piles. The sigmoidoscope of course at once clears up the diagnosis. Treatment of these cases is, however, difficult, as the bleeding, though easily stopped, is very liable to recur at some later time.

AN ABSTRACT OF
REMARKS ON CASES SEEN
 AT THE EAST SUSSEX HOSPITAL, HASTINGS,
 ON OCTOBER 18, 1927, AND
**A LECTURE ON SPLENO-
 MEGALY.**

*(Delivered under the Auspices of The Fellowship
 of Medicine),*

By BERNARD MYERS,
 C.M.G., M.D.

THREE cases were seen, the first being one of congenital hypertrophic stenosis, which had just been successfully operated upon; the second, a child suffering from enuresis, and the third a moderate case of hemichorea.

The points relating to diagnosis, prognosis and treatment were gone into, and in the case of hypertrophic stenosis, the after-treatment, and of enuresis, the causations were dealt with in detail; the latter case occurred in a neurotic child, and called for the treatment of a child of that type.

Questions were asked about the feeding of infants. The chemistry of human milk, the food requirements of the child, the physiology of digestion and caloric values were elaborated upon.

The following lecture was then delivered :—

THE SIGNIFICANCE OF ENLARGEMENT OF THE SPLEEN IN CHILDREN.

We all meet with cases of enlargement of the spleen from time to time, and it is

necessary to assess its significance in each individual case. For this purpose we must have a definite procedure to help the elucidation of the case in order that diagnosis may be established as soon as possible, and suitable treatment commenced. However, in order to do this we require a working knowledge of the anatomy, physiology and pathology of the spleen with a sufficiency of information concerning the clinical conditions in which the spleen is enlarged.

[After a preliminary account of the anatomy and physiology of the spleen the lecturer proceeded as follows :—]

FREQUENCY OF ENLARGEMENT OF THE SPLEEN.

Carpenter,¹ who analysed 348 cases of enlarged spleens in children, found that there were 48 during the first three months of life, 46 during the second, 21 during the third, and 48 between nine and twelve months, which makes 163 cases during the first year. There were 117 cases during the second year, 19 during the first half of the third year, and 16 during the second half. The number of cases then diminished greatly up to the age of 12 years, and only equalled 27 from 5 to 12 years. Some occurring in rickets may only have been palpable owing to deformity of the ribs, and did not necessarily denote enlargement of the spleen; probably, therefore, a number of the above cases came under that category.

During the first six months of life enlargement of the spleen is probably chiefly due to syphilis and tuberculosis. Up to two years rickets is not infrequent, and von Jaksch's anæmia may occur. Then there are the ordinary fevers of childhood—measles, whooping-cough, &c. Later, essential thrombocytopenic purpura hæmorrhagica, lymphadenoma and Banti's disease &c., have to be kept in mind. Acholuria, jaundice, septicæmia, malaria, and kala-azar

¹ *British Medical Journal*, 1903, ii, p. 463.

are also met with in children from time to time.

CLASSIFICATION.

Rolleston suggested that for practical purposes splenomegaly in children might be conveniently placed under the following headings, but it is obvious that some diseases would go under more than one heading :—

Familial.—This includes chronic splenomegalic hæmolytic jaundice, Gaucher's disease, congenital syphilis.

Those associated with Changes in the Blood-picture, such as acute leukaemia, pernicious anæmia, aplastic anæmia, thrombocytopenic purpura hæmorrhagica, infantile splenic anæmia (von Jaksch's), chronic splenic anæmia, Banti's disease, erythræmia (Vaquez's primary leucocythæmia with chronic cyanosis and splenomegaly and polycythæmia ; it does not occur in the first decade of life).

Infective.—Thus, acute septicæmia, enteric fever, malaria, pneumonia, scarlet, measles, influenza, &c., and also in some intestinal infections. The enlargement is due to congestion chiefly, but acute hyperplasia may be present, and sometimes small hæmorrhages. Chronic : Tuberculosis, lymphadenoma, congenital syphilis, Still's disease, &c.

Those of Tropical Origin.—Kala-azar, malaria.

Mechanical, as primary new growth, hydatid cysts, simple cysts, angiomas, abscess, infarcts, lardaceous disease.

Those associated with Hepatic Enlargement or Cirrhosis.—Chronic venous congestion, other forms of hepatic disease.

In order to aid diagnosis, a useful method of procedure consists in examining each case in the following manner :—

HISTORY.

In congenital syphilis, von Jaksch's pseudo-leukaemia infantum, the purpuras, Hodgkin's disease, malaria, Banti's disease, Gaucher's disease, &c., the history is of

obvious value. Inherited disease, familial affections, duration of symptoms, residence in the tropics and the special clinical features of the case are all of great importance. The previous medical history may greatly help, as osteomyelitis in amyloid disease and a history of tuberculosis in suspected tuberculous spleen.

CLINICAL EXAMINATION.

This should be most carefully carried out and, in addition to examination of all the different systems, the precise size, shape and consistence of the spleen should be noted and the liver and glandular system equally accurately palpated. It will be important to observe signs of rickets, discoloration of the skin, ascites, pallor, breathlessness, faintness, weakness and any tendency to hæmorrhages.

BLOOD EXAMINATION.

Leukæmia : Myelogenous.—The red blood-corpuscles are much reduced in numbers. Normoblasts are present in considerable numbers in some cases. The leucocytes may number 100,000 (or less) up to 500,000, and 50 per cent. may be myelocytes.

Lymphatic.—Dr. Knyvett Gordon finds that the red cells are usually unaffected except in the terminal stage, but their number may be reduced to 2,000,000 or less. The white blood-corpuscles may number 200,000 or more. In some cases they are less. The large lymphocyte is especially noted among the leucocytes. Mixed forms showing great increase of myelocytes and lymphocytes are believed to occur in early childhood, but this is denied by some observers.

Aplastic Anæmia.—The idiopathic form is infrequent in children. The red blood-corpuscles may be fewer than 1,000,000, but show very slight alterations in size and shape. Although the granular leucocytes are reduced, there may be an increase of mononuclears. The platelets are distinctly reduced. Myelocytes are rare. This disease only lasts a few weeks and is always fatal.

Pernicious Anæmia.—In this rare disease in children the well-known peculiarities of the red blood-corpuscles, the hæmoglobin and the colour index, &c., are present.

Infantile Splenic Anæmia.—The red blood-corpuscles and Hb. are reduced. The colour index is low. Anisocytosis, polychromasia and poikilocytosis are seen, also nucleated red cells and megaloblasts. There may be a leucocytosis of 20,000 to 60,000, in which lymphocytes predominate to 50 and 70 per cent. Transitionals average 12 per cent. Myelocytes are usually found. The platelets are increased.

Banti's Disease.—There is leucopænia and in the latter stages the lymphocytes are chiefly affected.

Blood-culture.—For septicæmia.

Special Tests.—Wassermann reaction for syphilis. Widal for enteric fever. Increased fragility of the red blood-corpuscles to salt solution is present in congenital acholuric jaundice. (Compare with ordinary obstructive jaundice in which there is an increased resistance to salt solution.) Malaria: The presence of the plasmodium is positive of this disease. Kala-azar: The presence of *Leishmania donovani* in the blood, spleen or liver makes the diagnosis positive. Gaucher's disease: Gaucher cells may be found in lymphatic glands, &c. Lymphadenoma: Finding the typical cells in an excised gland. Lardaceous disease: Waxy casts may be found in the urine. Hydatid disease: The finding of hooklets, &c. Tuberculin tests: Sometimes of use in a doubtful tuberculous case. Capillary resistance test in essential thrombocytopenic purpura hæmorrhagica. Bleeding time in essential thrombocytopenic purpura hæmorrhagica.

THE ACTION OF THE SPLEEN IN CERTAIN DISEASES AFFECTING CHIEFLY THE RED BLOOD - CORPUSCLES AND PLATELETS.

As Knyvett Gordon states, we have to remember that the red bone-marrow forms

the red blood-corpuscles, the hæmoglobin, the polymorphonuclear leucocytes and the platelets. When free from disease all of them are well-formed and normal physiologically, but under certain conditions the red corpuscles are abnormally formed, as in pernicious anæmia and aplastic anæmia, and under these circumstances the spleen acts as a destroyer of ill-formed red cells, hence, the less matured formed cells, the marrow cells, nucleated red cells, &c., make their appearance in the blood.

We can picture to ourselves again a condition in which the red blood-corpuscles are normally formed in the bone-marrow, but a toxin acting in the blood alters, or partly destroys, them while in the circulation. Perhaps this happens in acholuric jaundice. Here the spleen would play its usual part of destroying the ill-made red blood-corpuscles.

But a third condition might exist, where the red blood-corpuscles or platelets reach the spleen as perfectly made cells, then a toxin in the spleen or some pathological action of the cells in the splenic pulp alters or destroys these cells; again, we can further appreciate how a splenic toxin could affect the endothelium of the capillaries and smallest arteries so that bleedings are enabled to take place.

Where the defect is in the spleen only, or where the toxin affects the cells in the circulation only, we can see that the removal of the spleen would leave many more blood-cells in the circulation, and if any other existing toxin can be successfully combated, obviously much good will result.

Diagnosis.—This needs careful consideration of the history, symptoms, physical signs, blood-examination and other special tests. By this means the special features in favour of particular diseases are elucidated and the diagnosis established.

Thus, let us take for example the diagnosis of the much discussed Banti's disease.

Banti's disease commences in childhood and slowly progresses, although the diagnosis is frequently not established until about the

age of 20 years or older. It is hereditary but is occasionally familial. There are three stages in this disease, and during the first leucopænia is the first blood-change to be looked for, the lymphocytes being chiefly affected. There is no increase of fragility of the red blood-corpuscles.

In the first stage, which lasts several years, increasing pallor and weakness occur and digestive symptoms. The spleen enlarges, hæmatemesis may occur and some anæmia of the chlorotic type. The urine contains an excess of urobilin. In the second stage the symptoms progress and there is digestive disturbance and slight increase in the size of the liver. There is hepatic cirrhosis and ascites in the third stage.

It is diagnosed by the Wassermann reaction from syphilis; the history and symptoms are obviously also important.

In leukæmia the blood-examination should leave little doubt as a rule as to the diagnosis, although it is possible to have leucopænia during periods of aplasia.

Hodgkin's disease has its own special history, symptoms, exacerbations, and examination of a section from a gland, if necessary, can be carried out.

Splenic anæmia of infants usually shows a leucocytosis and the other symptoms already mentioned; there is a history of malnutrition and rickets in most cases.

Gaucher's Disease.—In this disease the liver as well as the spleen may show very distinct increase in size, there is also the skin discoloration and the conjunctivæ display peculiar wedge-shaped thickening, which is observed on either side of the canthus. Again, Gaucher's disease not infrequently is seen in several members of a family. Banti's disease shows a more rapid development in the second and third stages than occurs in this disease; Banti also shows a more obvious anæmia, hæmorrhagic tendency, while the third stage is often marked with ascites and jaundice. These symptoms are quite infrequent in Gaucher's disease.

Malaria is diagnosed by the history,

generally a residence in hot countries, and clinched by finding the plasmodia.

Congenital hæmolytic jaundice is rarely difficult to differentiate and diagnose from the history, its acholuric jaundice, increased fragility of the red blood-corpuscles and presence of urobilin in the urine.

Hanot's cirrhosis is a disease in which the liver is in the first place affected; chronic jaundice, an enlarged spleen, abdominal pain, fever and leucocytosis are present, and later on ascites.

Chronic infections usually show a leucocytosis.

Intestinal parasites may give rise to enlargement of the spleen. Eosinophilia should be looked for, and it will be remembered that, on the contrary, the eosinophils are decreased in Banti's disease. The important thing is to examine the stools for evidence of the parasite. Banti's disease is much more chronic than the parasitic.

In amyloid disease there is a history of prolonged suppuration as obtains in osteomyelitis, &c., and waxy cells may perhaps be found in the urine.

Treatment.—This will obviously depend upon the cause; thus, where there is a syphilitic splenomegaly, the appropriate treatment will be that for syphilis; in tuberculous splenomegaly the treatment for tuberculosis, but where the spleen be greatly enlarged from caseous tuberculosis splenectomy may have to be considered if the rest of the body is not much affected with the disease. Splenomegaly due to any of the infectious fevers would obviously receive the treatment of the particular fever; when the cause is heart disease or an affection of the liver, the treatment would again concern the cause. In a malarial case and in one due to kala-azar the usual treatment of both these conditions would obtain.

There are certain cases of splenomegaly in which splenectomy has given good results and in many cases apparent cure, such as Banti's disease, congenital acholuric jaundice and essential thrombocytopenic pur-

pura hæmorrhagica. In conjunction with Dr. Knyvett Gordon and Mr. Rodney Maingot I have had several such cases, and also one serious case of Henoch's purpura, which would not re-act to medical treatment. A brief résumé of some of the cases may prove interesting.

A girl, aged 10, who had suffered from essential thrombocytopenic purpura hæmorrhagica for a year, and for three months preceding splenectomy there had been constant and continuous oozing of blood from the gums which defied all the usual treatment, both local and general, including transfusion of blood. Dr. Knyvett Gordon found that the platelets in this girl were either absent or present in very small numbers compared to a normal girl of her age. The capillary resistance test was positive, i.e., when an elastic band was fastened just above the elbow so as to constrict the artery by about a half, a purpuric crop appeared just below the elbow within two minutes. The bleeding test was likewise positive for this disease. Normally, when the ear is pricked with the bayonet-pointed needle bleeding ceases within two to three minutes, but in essential thrombocytopenic purpura hæmorrhagica this takes fifteen or twenty minutes, probably due to the deficient number of platelets. It is eighteen months since the splenectomy was performed and this girl is now in perfect health; she has not suffered from oozing from the gums or elsewhere, nor have purpuric spots occurred on the skin. Her platelets increased greatly and the capillary resistance test was no longer positive. The bleeding time also became normal. There has not been any similar case in her family.

A girl, aged 12, had suffered from acholuric jaundice for five years. When she was admitted to the Royal Waterloo Hospital she was nearly *in extremis*. Her temperature was 103° F; she was very pale and weak, her red blood-corpuscles were just over 2,000,000, and although four transfusions of blood were given it was impossible to in-

crease the number beyond 2,200,000. Mr. Maingot kindly operated, and thanks to a very speedy operation and the leaving of normal saline in the peritoneal cavity she ultimately made a splendid recovery. When seen a week ago (four and a half months after the operation) she looked excellent, and felt in robust health. She had gained $2\frac{1}{2}$ st. in weight; her red blood-corpuscles were 5,000,000, and no abnormality was found in the complete blood-count. An aunt of hers suffered from the same disease and had also been cured by splenectomy.

A girl, aged 5, was sent to London from the West Indies, where she had suffered for over a year from bleeding from the kidneys following a fairly severe attack of Henoch's purpura. She was taken into a nursing home in London for some months, but medical means were quite unavailing to stop the severe renal hæmorrhage, and all other likely sources of the blood being placed out of count by cystoscopic examination, I asked Mr. Rodney Maingot to perform splenectomy as the serious loss of blood would ultimately have had a fatal issue. Dr. Knyvett Gordon found a great reduction in the number of circulating platelets, and in the spleen itself phagocytic cells, enclosing dead platelets. The case is described in the *British Journal of Children's Diseases*. It is the first recorded instance of Henoch's purpura treated by splenectomy and in which the peculiar phagocytic cells were found. For two months after the operation there was still some bleeding from the kidneys but only in greatly reduced amount; it then ceased. We have just heard that now fifteen months after the operation, the urinal continues to be quite free from blood, her pallor has disappeared and she enjoys the same good health as she did previous to the attack.