SPLENECTOMY IN HEMORRHAGIC PURPURA*

IDIOPATHIC PURPURA, ESSENTIAL THROMBOPENIE (FRANK). PURPURA HEMORRHAGIC PROTOPATHIQUE (HAYEM)

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The hemorrhagic diatheses have always been of considerable importance to the surgeon, because of the bleeding which occurs in these conditions following simple surgical procedures. This interest has recently been stimulated by Kaznelson, who recommended splenectomy upon a patient with essentielle thrombopenie (Frank) in October, 1916, with rather striking results.

Hayem has given a fairly definite set of clinical phenomena as characteristic of essential or idiopathic purpura (purpura hemorrhagique protopathique). The patient has an anæmia of the secondary type, with multiple spontaneous hemorrhages into the skin, from the mucous membranes, etc., recurring at irregular intervals with the following essential features: 1. Absence of any changes in the red blood-cells. 2. Marked diminution in the number of the blood platelets. 3. No constant variation in the white blood-cells. 4. Normal coagulation time of the blood. 5. A marked increase in the bleeding time. 6. Loss of contractility of the blood clot.

Frank, under the term "Die essentielle Thrombopenie," separates this type from the other varieties of purpura and considers it a clinical entity.

Fonio separates the hemorrhagic diatheses (other than hæmophilia) into three groups:

1. The Secondary Purpuras.—These have a known etiology and include the purpuras which occur in the febrile diseases (small-pox, typhus, ulcerative endocarditis, etc.), in peritonitis, in blood diseases (the leukæmias, pernicious anæmia); in diseases of the liver; due to the action of such poisons as phosphorus, benzol, snake venom, etc.; in scurvy, melena neonatorum, etc.

2. The Anaphylactoid Purpuras.—The etiology is unknown and the hemorrhagic diathesis is merely a part of the reactive symptom complex of some anaphylactic agent.

In this group the attacks recur at varying intervals with a free interval in between the attacks. The symptoms are various, fever, joint pains and swelling, urticaria, erythema, œdema, polyneuritis, albuminuria, hemorrhagic nephritis, colic, melena, etc. The occurrence of the various symptoms, the petechial eruptions, ecchymosis, etc., overlap and blend in the different forms and may exhibit their most marked symptoms in varying areas.

The blood platelets are slightly increased or very slightly below normal in

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number, the clotting time and the bleeding time are normal, and the blood clot contracts normally.

3. Idiopathic Purpura.—In this group he places those various forms of the hemorrhagic diatheses which are primary. The etiology is unknown. Without warning bleeding into the skin, from the mucous membranes, melena,

	Anaphylactic purpura.	Idiopathic purpura.
Etiology.	Known or suspected causative agent. Primary fever. Premonitory symptoms.	Unknown. No fever. No premonitory symptoms.
Premonitory symptoms.	Primary fever, urticaria, œdema, joint pains, hemorrhagic nephritis, colic, melena, etc.	None.
Bleeding time.	Normal.	Lengthened.
Clotting time.	Normal.	Normal.
Retraction of clot.	Normal.	Absent or markedly diminished.
Blood platelets.	Increased or slightly decreased.	Markedly decreased. May be absent during attack.

	Hæmophilia.	Idiopathic purpura.
Etiology.	Unknown.	Unknown.
History.	Classical history of bleeders in family involving males transmitted through female line.	No typical history. May have history of similar condition in the family but no definiteness in the transmission.
Cause of bleeding.	Traumatic.	Usually spontaneous, frequently multiple. Periodic with free interval. Trauma may cause bleeding but is not so definite as in hæmophilia.
Bleeding time.	Markedly lengthened.	Lengthened. Increased (especially during attacks).
Clotting time.	Markedly lengthened.	Normal.
Blood platelets.	Normal or increased in number.	Markedly decreased or absent during attacks.

hæmatemesis, nose bleed and hæmaturia occur. Fever is absent unless the case is otherwise complicated.

The characteristic features are, increased (lengthened) bleeding time, the absence of retraction of the blood clot or great diminution of this con-

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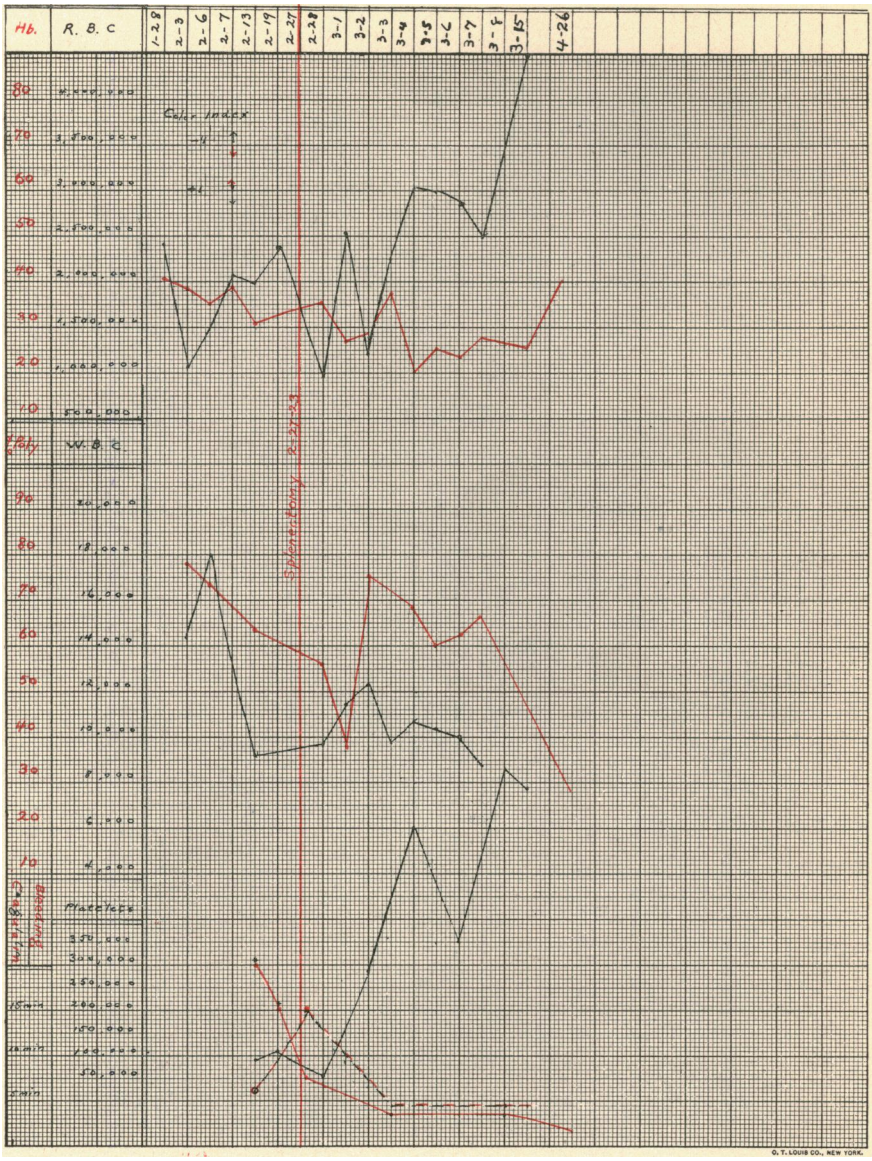

FIG. 1.—Platelets. — — — Bleeding time.

traction, marked diminution in the number of the blood platelets while the clotting time of the blood is normal.

Fonio gives the schematic forms for the differential diagnosis between anaphylactic purpura and idiopathic purpura, and hæmophilia and idiopathic purpura in the above table.

Kaznelson was struck by the observation of Frank, that in the conditions to which Frank had given the name "essentielle thrombopenie" there was a constant enlargement of the spleen, and formulated the hypothesis that the disease was due to some destructive agent which destroyed the blood platelets and that the spleen, because of its enlargement had some definite relationship to the process of platelet destruction. Acting on this hypothesis in October, 1916, he submitted a female patient of thirty-six who had had the characteristic features of the essential purpura, described by Hayem, Frank, and others, to splenectomy with remarkable results. The blood platelets which were 300 to 600 before operation rose rapidly to 600,000 on the second day after operation. The clot contracted early and the bleeding time diminished, nose bleed stopped and the menses became normal in character. He has had the patient under observation for three years and the improvement has been constant.

Since then Kaznelson has reported two other cases submitted to splenectomy with a satisfactory outcome. Schmidt, Minkowski, Ehrenberg, Keisman, Beneke, Cori have reported other similar cases in the German literature. In America, Bowen reports a satisfactory case from the Buffalo General Hospital and Brill showed two cases at the meeting of the Medical Section of the New York Academy of Medicine on February 15, 1923, submitted to splenectomy with a satisfactory outcome.

Case Report.—Esther C., eight years old, was admitted to the First Medical Division, Doctor Connor's service, on January 27, 1923 and discharged March 16, 1923.

Her present illness began two days before admission with almost constant bleeding from the nose and gums, and the appearance of red spots on the body. She has not vomited blood or passed blood in urine or stools.

Past History.—Patient was well up to three years ago, then had scarlet fever; she recovered and was well up to one year ago. At that time she began to have repeated nose bleeds which were difficult to stop, at intervals of every two or three weeks. One week ago she had a bronchopneumonia from which she is now convalescing.

Family History negative for family bleeding, two brothers and sister well.

Examination showed well nourished, pale child, with hemorrhagic spots on face. Bleeding from left nostril and gums. Heart, lungs, abdomen were negative. Examination of skin showed hemorrhagic areas over the entire body most marked over the lower extremities. In one area on the hip there was a large ecchymotic area resembling a bruise.

Clinical pathological report:

Red cells, 2,400,000—H. B. 40%, Color index 0.8.

White cells, 38,000—Polymorphonuclears 85%, Lymphocytes 11%, Large mononuclears 4%.
Platelets, 40,500.
Coagulation time 6 minutes.
Bleeding time more than 20 minutes.
Clot did not contract.
Fragility test—Hemolysis began at 0.45, complete at 0.36.
Temperature 99, Pulse 116, Respiration 26.
Chronological Course:
Day of admission 15 c.c. whole blood intramuscularly.
15 c.c. horse serum intramuscularly.

First day after admission.	Vomited blood.
Second day after admission.	Nose bleed for three hours, packing.
Third day after admission.	Nose bleed, slight, packing.
Fourth day after admission.	Transfusion 150 c.c. of blood intravenous (Unger method) followed by chill.
Fifth day after admission.	No bleeding since transfusion.
Sixth day after admission.	Bleeding from right nostril, packed.
Seventh day after admission.	Nose bleed, practically throughout day, partially stopped by packing. 15 c.c. horse serum intramuscularly.
Eighth day after admission.	Enema, large stool containing altered blood.
Ninth day after admission.	Nose bleed, hæmatemesis, transfusion 340 c.c. blood, followed by chill.
Tenth day after admission.	No bleeding.
Eleventh day after admission.	Melena, nose bleeds.
Twelfth day after admission.	Nose bleed not stopped by packing, cocaine, adrenalin, monochloracetic acid.
	Stopped with thromboplastin.
Fifteenth day after admission.	No bleeding, palpable spleen noted.
Sixteenth day after admission. Seventeenth day after admission.	Nose bleed slight.
Eighteenth day after admission.	Transfusion 250 c.c. followed by chill.
Nineteenth day after admission.	Severe hemorrhage from left nostril for five hours, vomited two pus basins of blood.
	Nose bleed all afternoon. Not controlled by packing.
Twentieth day after admission.	Melena, vomitted blood during day. Constant oozing from nostrils not stopped by thromboplastin.
Twenty-first day after admission. to Twenty-ninth day after admission.	No bleeding.
Thirtieth day after admission.	Profuse nose bleed, chills, much worse. Consultation with Dr. R. G. Stillman.

On February 27th, thirty-one days after her admission, at the suggestion of Dr. R. G. Stillman, of the First Medical Division, splenectomy was done through a left rectus incision and a somewhat enlarged spleen easily removed. No

hæmolymph-nodes were noted along the splenic vein. The liver and gall-bladder seemed normal. The blood was quite watery. Tier closure of the abdomen without drainage. The wound healed by primary union and the patient was sent to the country on the seventeenth post-operative day.

The essential features of the case following the splenectomy were the immediate cessation of the bleeding, the marked increase in the number of blood platelets from 50,000 to over 600,000, the change in the bleeding time from 15 to less than 5 minutes and the marked improvement in the condition of the child. This improvement has continued up to the last observation made on May 25, 1923.

The last observations on the blood made by Dr. Ralph G. Stillman on April 26, 1923 are as follows:

Bleeding time 2½ minutes. Hæmoglobin, 40%
Polymorphonuclears, 24.0%
Lymphocytes, 68.8%
L.M. and Tr., 4.0%
Eosinophiles, 2.4%
Normoblasts one to each 250 white cells.

Film shows leukocytosis, well marked pallor of red blood-cells. Some granular basophilia and slight polychromatophilia. There are abundant platelets, looks like a secondary anemia.

Pathological Report.—Esther C., May 16, 1923. Hyperplasia of the pulp. Myeloidization. Specimen consists of a spleen of normal contour, rather dark bluish-red color. Capsule elastic, not very tense. Weight with contained blood 120 grams. (Patient's weight 47.5 lbs. 21-590 kg.) Size 4 mm. x 5.5 x 11 mm. On section of capsule much blood escaped estimated at about 15 c.c. Piece removed sterile for culture. Sections placed in formalin, bichloride, Zenker and absolute alcohol. On section the cut surface is smooth, bright red and very thickly studded with follicles that appear of normal size. The consistence is firm, possibly slightly firmer than normal. There was no obvious increase in connective tissue. No hemorrhages were seen. Films made from surface.

Stained film shows the usual number of lymphocytes and large mononuclear cells. Polymorphonuclear leukocytes, neutrophiles are moderate. Eosinophile leukocytes are fairly numerous. There are many normoblasts and a few megaloblasts. There are also a moderate number of myelocytes. Most of them neutrophilic but a few eosinophilic and basophilic. From the cells found in these films one would expect to find in the spleen areas of myeloidization and possibly also of erythropoiesis.

On microscopic examination there appears to be no increase in the amount of fibrous tissue present. Lymphoid follicles all have large germinal centres, but are no larger than one would expect in the spleen in a child of this age. The arteries in the follicles show almost uniformly a thickening and hyalin degeneration of the wall and a narrowing of the lumen. The venous sinuses appear to be approximately normal in size. The pulp in places shows an increased blood content and a hyperplasia of the pulp cords. There are seen in the pulp occasional nucleated red cells and a moderate number of myelocytes. There are also a number of undifferentiated mononuclear cells whose exact nature is unknown. There appears to be a slight increase in the number of eosinophiles present. Collections of blood platelets were not recognized.

Bacteriological Report.—Doctor Wheeler. Cultures from the spleen were sterile.

CONCLUSION

The removal of the spleen has a definite effect in the idiopathic type of hemorrhagic purpura and this effect seems to be related in some way to the change in the bleeding time and in the number of the blood platelets.

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