EDITORIAL

New Forms of "Idiopathic" Thrombocytopenic Purpura

THROMBOCYTOPENIA is in general due to one of several mechanisms:

1. A deficiency in the materials necessary for normal platelet production by the marrow megakaryocytes, as in pernicious anemia.

2. A reduction in the actual number of megakaryocytes, and thus in platelets, as in leukemia and aplastic anemia.

3. A reduction in platelet formation by the cytoplasm of the megakaryocytes, as in idiopathic thrombocytopenic purpura and in numerous conditions with splenomegaly, and in certain allergic states.

4. A "sweeping-up" effect, in which although the megakaryocytes are presumably normal both in number and in platelet production the latter are "lost" in the peripheral blood vessels. It is with this type of condition that the two papers of Fitzpatrick et al., and Singer, Bornstein and Wile in this issue are concerned.

In the typical form of idiopathic thrombocytopenic purpura, the megakaryocytes are numerous but greatly deficient in platelet production. We conceive of this disorder as a form of hypersplenism,[1] splenectomy being followed by an extreme increase in platelet production. It seems likely that the spleen is functionally abnormal, producing either an abnormal substance or an excessive amount of a normal inhibitor of platelet production. Other forms of idiopathic thrombocytopenia are beginning to be discriminated. Thus, the type associated with splenomegaly may also be classed as a form of hypersplenism. This may occur in such diverse conditions as rheumatoid arthritis, Boeck's sarcoid, Gaucher's disease and cirrhosis of the liver. The platelet diminution may also be due to an excess of platelet inhibitor released by the enlarged spleen.

S. O. Schwartz[2] has pointed out that an allergic reaction on the part of the marrow may result in megakaryocytic changes indistinguishable from those found in the completely idiopathic type. The allergic reaction is however associated with a marrow eosinophilia, which serves to distinguish it from the nonallergic type. Schwartz contends that those cases showing eosinophilia as a rule recover without splenectomy. Before this concept can be fully accepted, further statistical data are required.

"Thrombotic thrombocytopenic purpura," as described in this issue under different designations, poses a new problem to the clinician. Is the case of thrombocytopenic purpura at hand one of the truly idiopathic type in which the marrow megakaryocytes are functionally inadequate, or is it perhaps the disorder first described by Moschowitz, in which megakaryocytes are normal both in number and in function, but the platelets are being swept into small blood vessels where they cause multiple thromboses? Singer discusses the differential diagnostic features which may be diagnostically helpful in a given case. The condition appears to be more common than was previously considered, although the diagnosis has yet to

be made clinically. The prognosis in the thrombotic type is apparently uniformly bad, whereas in the ordinary idiopathic type it is usually good when splenectomy is performed.

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REFERENCES

[1] DAMESHEK, WILLIAM, AND MILLER, E. B.: The megakaryocytes in idiopathic thrombocytopenic purpura, a form of hypersplenism. Blood *1:* 27, 1946.

[2] SCHWARTZ, S. O.: The prognostic value of marrow eosinophils in thrombocytopenic purpura. Am. J. M. Sci. *209:* 579, 1945.