Blood Diseases of Infancy and Childhood

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Preface

The purpose of this book is to present the essentials of pediatric hematology in concise form for the medical student and practitioner. In lectures over a period of many years to members of these groups, I have been impressed with the need for providing a textbook in which the salient features of blood dyscrasias are presented against the background of normal development of infancy and childhood.

Although these hematologic disorders correspond closely to those occurring in adult life, the approach to diagnosis is frequently complicated by concomitant alterations which normally take place during growth within and outside of the hematopoietic system. Certain of the blood disorders occur almost exclusively in early life and are best described in a pediatric setting.

The rapid development of pediatric hematology can be strikingly illustrated by recalling the fate of von Jaksch's anemia. This syndrome which dominated the discussions of anemia in pediatric textbooks thirty years ago receives scant mention or is entirely omitted from present-day publications. The dismemberment of this heterogeneous group of blood disorders replaced by more soundly based conditions reflects the evolution of new concepts in a rapidly expanding field of medicine. Similarly, such diverse conditions as thrombocytopenic purpura, erythroblastosis, and the hemolytic anemias and disorders of the coagulation mechanism, which occur so prominently in younger persons, have been clarified by recent contributions from the growing field of immunohematology and by the identification of multiple factors responsible for blood clotting. In the hereditary hemolytic syndromes important information has been acquired by the application of genetic analysis and the characterization of abnormal types of hemoglobin by electrophoretic examination. The discovery that drug sensitivity and favism are based on a genetically transmitted enzyme deficiency suggests that other unexplained hematologic entities may also eventually be included in the growing list of inborn errors of metabolism.

The search for a fetal etiology of congenital anomalies has also had its impact on the blood dyscrasias in the pediatric age group. This orientation has been so attractive that the pathogenesis and interpretation of the blood disorders especially in the early months and years of life require an examination of the maternal-fetal relationships and of environmental and genetic influences and a consideration of normal embryologic development and its aberrations.

Although the blood diseases of younger patients are effectively presented in the comprehensive hematologic textbooks currently available, nevertheless there is much to be gained by their separate consideration. This book is not intended, however, to supplant these larger works; rather it is designed to serve as a companion volume since few diseases in this or other specialties can be arbitrarily restricted to a specific age period. It is planned throughout to emphasize the pathogenesis of hematologic disorders of the pediatric age group in the light of established concepts as a basis for rational treatment.

From the rapidly enlarging mass of hematologic and pediatric contributions, only that segment is represented which bears pertinently on the interpretation, diagnosis, and management of the blood diseases encountered in pediatric practice.

Elaborate techniques have been developed within recent years permitting visualization of physiologic forces controlling blood formation and their abnormalities. Although certain important data can be acquired with these facilities, I have always felt that the detection of the common blood disorders lies within the province of every medical practitioner by careful history and physical examination with the use of simple instruments and techniques and by coordinating information derived from a variety of interrelated sources. In striving to achieve this objective, it is hoped that an easily understandable and practical book has been prepared.

I wish to acknowledge my indebtedness to the authors of the standard hematologic, pathologic, and pediatric textbooks which were of inestimable value for orientation and subject matter. Thanks are due to my colleagues and associates who offered suggestions in various aspects of the text, many of whom are identified in papers written conjointly and are mentioned in the bibliographies. I wish particularly to express my appreciation to those who reviewed one or more chapters of the book, and whose advice and criticism were invaluable; notably Dr. Marion E. Erlandson, Dr. Sydney S. Gellis, Miss Jane Haber, Dr. S. Frederick Rabiner, and Dr. Marjorie B. Zucker. Above all, I am everlastingly indebted to my wife, Margaret, for supervising and contributing to every phase of the preparation of the manuscript, thereby relieving me of numerous tasks that disturb the equanimity needed for writing. For added participation of my family, thanks are due my daughter, Christine, for help in revising the manuscript and my son, Carl, for emphasizing the point of view and needs of the medical student. My deep appreciation is extended to Dr. Samuel Z. Levine, Professor of Pediatrics of the Cornell University Medical College, who by his constant support and warm friendship provided me with the needed encouragement and stimulation to develop the field of pediatric hematology in his department and the material on which this book is based. Thanks are due Mr. Percy Brooks and his associates of the Department of Illustration of The New York Hospital-Cornell Medical Center for cooperation in the preparation of the figures. Sincere appreciation is due Mrs. Pandora Manning for her patience in typing and retyping large sections of the manuscript.

CARL H. SMITH

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