

**DISEASES OF
THE
NEWBORN**

SCHAFFER

DISEASES OF THE NEWBORN

SECOND EDITION • 1965

Diseases of the Newborn

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Harlequin color change. Note the sharp line of demarcation in the midline of the face and trunk separating the reddened right half of the body and the blanched left half. (From M. Birdsong and J. E. Edmunds: *Obst. & Gynec.*, Vol. 7. Reproduced with permission of the senior author.)

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FOREWORD

ALTHOUGH there is controversy about "new" and "old" pediatrics, everyone recognizes the responsibility of physicians—be they obstetricians, pediatricians or general practitioners—to newborn infants. Dr. Ethel C. Dunham, a pioneer in this field, posed the problem as follows: One must learn new facts about the newborn; one must spread more widely what is already known; one must make it possible to apply these facts. Dr. Schaffer's book is an important direct step toward these goals. Out of his extensive experience as a critical clinician and teacher he has written a book to help physicians judge the significance of symptoms in newborn infants. Appraisal of the neonate is most difficult, but careful history and careful physical examination are, as in all medicine, the basic modalities with which the physician must deal. The art is to know how to interpret findings, how to know when laboratory assistance is required. Direct experience with large numbers of newborn infants and understanding of their physiology are the bases for expert clin-

ical judgment. Neither is a substitute for the other. During his more than thirty-five years as a leading practitioner of pediatrics in Baltimore Dr. Schaffer has made careful clinical observations of thousands of newborn infants not only in the nurseries of Johns Hopkins, Sinai and the Women's Hospitals, but also as the infants have grown. He has maintained a continuing interest in the anatomic, biochemical, immunologic, pathologic, physiologic and psychologic peculiarities of the newborn infant, as befits a man who was Dr. John Howland's chief resident, and who became a leading teacher of pediatrics at Johns Hopkins University under Dr. E. A. Park and his successors.

Dr. Schaffer, and his associate Dr. Milton Markowitz, have for years demonstrated their consummate skill in diagnosis and treatment of newborn infants to those colleagues fortunate enough to work with them on a day-to-day basis. This book extends their influence for the benefit of newborn infants and their parents, wherever they are.

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PREFACE TO THE SECOND EDITION

The widespread acceptance of *Diseases of the Newborn* as a primary reference source for diagnostic and therapeutic problems in the neonatal period has been gratifying. At the same time it has made it necessary that the book be kept up to date. This is no easy task, since the explosive outburst of new information which stimulated the original compilation has proceeded with even greater momentum over the past five years. Some of our original discussions read as though they were written in medieval times rather than a few short years ago.

This second edition is an attempt to accomplish three ends. The first is to correct those few inevitable errors which escaped our most critical editing. We hope we have added no new ones. The second is to fill in omissions. We were appalled to discover that we had inadvertently neglected to include so important and so characteristically neonatologic a problem as tetany of the newborn. This hiatus has been filled, as well as those concerning idiopathic hypercalcemia, *Pneumocystis carinii* infection, the Wilson-Mikity syndrome, amyotonia congenita and a number of others. And, finally, we have tried to update many of the sections treating dis-

orders about which much new knowledge has accumulated in the intervening period. New material has been added on Gross Chromosomal Aberrations and on recognizable Constellations of Congenital Malformations for which no gross chromosomal defect has been established as the cause. The chapter on Inborn Errors of Metabolism has been enlarged in keeping with the augmented knowledge acquired recently, much of it pertinent to proper diagnosis and care of the newborn infant. The immunologic status of the newborn has been re-examined, with its fascinating corollaries, the gamma globulins, the role of the thymus and the problem of immunologic tolerance. Many other disorders, ignored in the original edition, have been described, if only briefly, for the sake of that unattainable ideal, completeness.

We are indebted to Dr. Lawrence Finberg for having written a section on Fluid and Electrolyte Balance in the Neonatal Period. Dr. Milton Markowitz has thoroughly revised his section on Cardiovascular Disorders in order to make it more useful to pediatric practitioners, residents and advanced students. We think that he has succeeded admirably.

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Dr. Barbara Migeon was good enough to read over the new chapter on Gross Chromosomal Aberrations and to make valuable suggestions which we have tried to incorporate. She also let us use several of her own illustrations.

We have again resisted the urge to

have subspecialists, other than the two mentioned above, write chapters on their particular subspecialties, because we feel that the orientation originally decided upon, from one practitioner to another, has proved its usefulness.

ALEXANDER J. SCHAFFER

PREFACE TO THE FIRST EDITION

THIS BOOK was intended to be an Atlas of Diseases of the Newborn. It was to consist of a large number of illustrations and a minimum number of words. Justifiably or not, we soon found ourselves changing this plan. The chief reason was that many important topics were simply not amenable to photographic treatment. How does one go about handling galactosemia or phenylketonuria pictorially? Of what use to a student or a practitioner would a book on the newborn be which omitted any discussion of an inborn error of metabolism whose early recognition spells the difference between vision and blindness, intelligence and stupidity, longevity and early death?

The second consideration which changed the structure of this book was the gigantic growth within the past decade of the corpus of knowledge concerning the newborn. Pathologists, physiologists of many varieties, radiologists and clinicians have begun to swarm over the newborn in ever-increasing numbers. Much has been learned, even though much still remains to be discovered. We felt that the time was ripe for this new knowledge to be collected and to be in-

tegrated with the old into an omnium gatherum of Diseases of the Newborn. This could not be accomplished with pictures alone.

We were then confronted with the apparently insurmountable obstacle of our own limitation of knowledge. Clearly no one man can hope to know as much about disturbances in bodily chemistry *and* endocrinology *and* congenital heart disease *and* the premature infant as do the various subspecialists in these limited fields. Would not a collection of essays written by eight or ten of these specialists constitute a superior kind of volume to this necessarily less erudite and less detailed one? We ventured to guess that, for the purpose we had in mind, it would not be as useful. For we practitioners of pediatrics are the newborns' first and primary physicians. We are the ones who should be aware of genetic predispositions which may dictate laboratory studies even before our newborn becomes ill. In this connection we call to your attention the proper management of babies born in families which contain known sufferers from congenital galactosemia or phenylpyruvic oligophrenia. We are the ones who should not, indeed

must not, allow even the most trifling deviations from normal to escape our attention in our original examination. Overlooking a cornea which is larger than it should be may spell the difference between ultimate good vision and blindness. Not noticing, or attaching no significance to, a tiny red spot over the spine may mean that that baby will suffer one or two bouts of staphylococcal meningitis before his dermal sinus is diagnosed and excised. In these situations the ophthalmologist and the neurologic surgeon are of absolutely no help to us until we have made the all-important original observation. Finally we are the ones called when cyanosis, dyspnea, fever or convulsions appear, and we must make rapid decisions as to immediate treatment and further study. We must categorize the illness accurately and ask help, when needed, of the proper subspecialist. We have quoted in the text the unfortunate story of a newborn with dextrocardia for whom the aid of a cardiologist was sought. Both practitioner and cardiologist stood by while the infant died of untreated pneumothorax.

In actuality the situation of the pediatrician practicing neonatology differs but little qualitatively from his everyday posture with respect to older infants and children. Much of his function consists in screening, expediting and directing his patients to other subspecialists. But quantitative differences exist in the neonatal period. When newborns are sick, they are often so terribly sick that one is loath to endanger their lives by performing diagnostic procedures that would be sheerest routine in older infants. At the same time physical examination is less rewarding in them. Finally the clinical entities peculiar to their age group are just beginning to be defined and are far from being neatly classified in any fixed nosologic schema. Thus even the simplest decisions, for instance whether cyanosis is caused by a congenital heart defect or by pulmonary or intracranial disorder, can be far from simple in the neonate. We believe the practitioner needs a reference book which brings

these matters up to date and which may permit him to make these important decisions more promptly and more accurately. Detailed information about pathologic physiology, pathology and embryology may be obtained from other sources.

This, then, is a book on clinical neonatology, written by a practitioner who is neither pathologist nor physiologist, neither biochemist nor virologist. It will therefore have suffered from lack of detailed knowledge in these fields. It is our hope that it will have gained something by virtue of the author's preoccupation for many busy years with the diagnosis, natural history and treatment of disorders of the sick newborn.

Many acknowledgments are in order, too many to detail in this place. The first must go to my wife, who accepted with extraordinary good grace almost complete withdrawal from social life plus the inconvenience of having been awakened at or before dawn every morning for about five years. The second is directed to Dr. Harry H. Gordon, who stimulated me to begin this work and whose constant interest and affectionate concern were mine throughout its long-drawn-out course. He must not be held responsible for any of its imperfections. Neither must my associate, Dr. Milton Markowitz, who not only wrote the section devoted to cardiac disorders, but also struggled with me over most of the sick infants who formed the basis of such knowledge of neonatology as we may possess. Dr. Anthony Perlman, my former associate, was equally conscientious in the matters of diagnosis and treatment of many of these infants and in the mechanical job of keeping detailed day-by-day records of their progress. Pathologists at three hospitals have cooperated freely at the expense of much of their valuable time. Dr. William J. Lovitt, Dr. Ella Oppenheimer and Dr. Tobias Weinberg of The Hospital for the Women of Maryland, The Johns Hopkins and the Sinai Hospitals of Baltimore, respectively, deserve my thanks. The interpretations placed upon their

observations are my own. So do the librarians of the Medical and Chirurgical Faculty of Maryland, chiefly Miss Louise D. C. King and Miss M. Florence Woods, and my own secretary, Miss Patricia Lilly. I am obligated deeply to all those physicians and surgeons who have given permission to utilize cases and have supplied me with prints of illustrations which my own files did not contain. In this connection the photographers at the various hospitals must be thanked, especially Mr. Harold A. Thomas at The Sinai Hospital of Baltimore.

I am singularly indebted to three good friends whose financial aid made it possible for me to amass an extensive and expensive collection of prints, lantern slides and color transparencies. They are the Messrs. Alan Wurtzburger, James H. Levi and the late Stuart M. Weiler and their wives. Dr. Markowitz is equally

grateful to the Benjamin and Minnie Landsberg Memorial Foundation for their support of his studies in the field of heart disease in infancy and childhood.

I must mention my special feeling of gratitude to my publishers, W. B. Saunders Company. My contacts and correspondence with them were effected largely through the medium of John Dusseau, less often through Robert Rowan. Their help, their encouragement, their sound advice and, not least, their exhilarating senses of humor have carried me over many rough spots.

Finally I must thank the administrators of the aforementioned hospitals, plus those of the Union Memorial, University and Lutheran Hospitals of Baltimore, and the heads of their respective Pediatric and Obstetrical Departments for their permission to utilize their cases to illustrate many of my points.

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