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DISEASES OF THE NEWBORN

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FOREWORD

ALTHOUGH there is controversy about "new" and "old" pediatrics, everyone recognizes the responsibility of physiciansbe 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 of 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 clinical judgment. Neither is a substitute for the other. During his more than thirty 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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Baltimore, Maryland

FOREWORD

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PREFACE

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?

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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 integrated 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 situa-

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tions 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 newborn 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 longdrawn-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,

PREFACE

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.

ALEXANDER J. SCHAFFER Baltimore, Maryland

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INTRODUCTION

THE AIMS OF NEONATOLOGY

We trust we shall be forgiven for coining the words "neonatology" and "neonatologist." We do not recall ever having seen them in print. The one designates the art and science of diagnosis and treatment of disorders of the newborn infant, the other the physician whose primary concern lies in this specialty. The words follow logically the stem "neonate," in common usage, and seem to be at least as appropriate as the neologisms "geriatrics" and "geriatrist" or "gerontologist." We are not advocating now that a new subspecialty be lopped from pediatrics. Yet such a subdivision has, we suspect, as much merit as does pediatric hematology, pediatric epileptology, even pediatric endocrinology and pediatric cardiology. For the period of greatest mortality in life is the first day, and a high mortality rate characterizes the entire first month (Fig. 1). If we add to these the deaths which take place in the last months of pregnancy and during labor and delivery, the sum total of socalled perinatal deaths adds up to a staggering figure (Fig. 2). And it is a figure which has not fallen nearly as rapidly since the dawn of the era of modern medicine as have the mortality rates for any of the other age groups (Fig. 3). The neonatal period therefore represents the last frontier of medicine, territory which has just begun to be cleared of its forests and underbrush in

preparation for its eagerly anticipated crops of saved lives. Until such time as a new subspecialty may be established it behooves the pediatrician and the general practitioner who supervises the growth and development of infants and children to become more adept in diagnosis, therapy and prevention of the disorders of the newborn infant.

The Original Examination. The first task is the performance of a thorough physical examination. This does not differ in technique from the one the physician performs upon an older infant, but its orientation is different because of the special problems which are unique

in this age group.

The first examination of a newborn baby should be an exciting event. It is unfortunate that because there are so many babies born and because one so seldom discovers deviations from the normal, the average practitioner or house officer assigned that duty is inclined to look upon it as a chore. In actuality it is a task of the utmost importance which, performed conscientiously, pays large dividends.

It should be routine for the examiner to scan the labor sheet with care before looking at the baby. On this he should be able to find at a glance the mother's age, parity and the outcome of her previous pregnancies, if any, her blood type and that of her husband. Matters of significance are her estimated date of con-

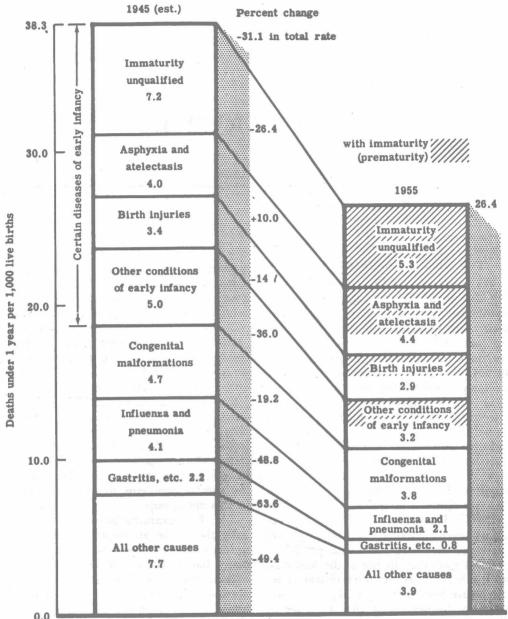


Fig. 1. Infant mortality. Main causes by Sixth Revision of the International Lists, United States, 1945 and 1955. (From Children's Bureau Statistical Series, No. 50.)

finement, the total duration of labor and the length of the second stage, the day and hour her membranes ruptured, so that he may calculate the length of time they had been ruptured prior to delivery. Any instrumental aid to delivery should be known, as well as the presentation of the fetus. Several notations of fetal heart rate should appear on this sheet. The characteristics and quantity of amniotic fluid are of no less importance. Finally there should appear on this same sheet the obstetrician's or anesthesiologist's estimate of the baby's condition one minute after delivery, à la Apgar. All this consumes but a few moments. To the neonatologist these obstetrical data are as significant as a detailed record of