Anomalies

OF INFANTS AND CHILDREN

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FOREWORD

Anomalies of Infants and Children is written primarily for the student and general practitioner. I believe it will also be of aid to the obstetrician and pediatrician.

The book is designed as a quick reference. It helps those who are interested in this subject to discuss intelligently with the parents and others concerned the anatomical anomaly from which the patient is suffering, the resulting physiologic disturbance, the rationale of treatment, and the accepted time for repair.

The subject of anomalies of infants and children, in the opinion of many writers, has never been properly studied or correlated. Until a few years ago these anatomical defects fell into the hands of the general surgeon, who perhaps had little, if any, experience in their repair. The general surgeon was fortunate who, when called to treat congenital or acquired anatomical defects of the palate, superior maxilla, or inferior maxilla, could consult with or call to his aid a maxillofacial or plastic surgeon.

Plastic surgery may be defined as that specialty which embraces reconstruction of anomalies and anatomical derangements following injury or disease—in fact all operative measures undertaken to improve the appearance and to restore function of the human body. The authors' training has qualified them in the field of maxillofacial surgery after basic training in general surgery. Their experience in plastic surgery has been extensive, not only in civilian practice but also in wartime surgery.

Dr. William J. Mayo once wrote, "Every human being has the divine right to look human. One of the compensations of the Great War was the development of plastic surgery of the face, a new special field in surgery which has given astonishing results."

To learn technique is easy, but to acquire judgment is a different matter. Both these attributes are possessed by the authors. Plastic surgery is not for the impatient surgeon or patient. Patients and their relatives should be made to understand the complication of the congenital or acquired deformity involved, the numerous operations that may be necessary, and the length of time required.

Some might be tempted to say that this book describes nothing new or nothing that has not been described by previous writers; this is true, but the present authors are the first to correlate the available information in one volume. It was the novelist Charles Reade who, when accused of drawing to an undue extent on previous writers, said, "I have milked three hundred cows but the cheese that I made was my own."

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PREFACE

Throughout their years of experience in the observation and treatment of anomalies in infants and children, the authors have been impressed by an apparent lack of general familiarity with the causes and dangers of many anomalies, the prognoses for their correction, and the optimum time for the initiation of therapy.

This is the more surprising in view of the fact that so much well-documented scientific knowledge and experience in this field has been recorded. Unfortunately, such information is widely scattered and appears in isolated papers or in brief incidental chapters in books devoted to a single specialty, such as orthopedics, pediatrics, or urology.

Until now it has been necessary, therefore, to refer to many sources in order to locate references to a specific anomaly. This book is an attempt to place in a single volume our present knowledge of the more common types of congenital and acquired abnormalities.

The chapters have been organized for simple and rapid reference. Their subjects include the role of genetics in the

production of congenital malformations; the increasing incidence of the acquired deformities; and cancer in infants and children.

Because not only the patient but also the family—especially the mother—may often suffer psychological trauma in connection with anomalies, considerable attention is given to facts which can be used to explain congenital anomalies as a natural hazard of conception and birth rather than a consequence of prenatal influences. In each case the authors indicate what can and should be done about the anomaly, when such steps ought to be initiated, and what final outcome can reasonably be expected.

The authors call attention to the danger of radiation for the removal of benign skin lesions and have devoted an entire chapter to the management of burns. They also stress the critical importance of initial treatment of a burn patient and the avoidance of dressings and medication which may complicate later professional care. With these exceptions, little reference has been made to the details of treatment of anomalies, in recognition of the fact that many medical specialties are involved in such treatment.

It is the authors' hope that this volume will prove to be a simple and ready reference and will make more general the knowledge of a subject of very pressing importance to thousands of young patients, their parents, their families, and their friends.

Grateful acknowledgment is made to the many who helped and advised in the compilation of this book: in particular to Majorie Foley, our photographer; to Stanley Q. Schwartz and Masayuki M. Nakamura of the Medical Illustrations Department, Veterans Administration Hospital, East Orange, New Jersey; and to David Q. Hammond, Vice President of Fairleigh Dickinson University.

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CHAPTER 1

Introduction

Although a certain amount of surgery has been performed in the young for many years, it is only in the last couple of decades that extensive surgery has been carried out in the young as well as in the very old. Surgical horizons have been extended in every direction. Much of this advance can be attributed to the marked improvements in anesthesiology. The development of many new anesthetic agents, as well as the perfection of additional techniques in the use of the older agents, offers a wide enough choice for the anesthetic to be fitted to the patient rather than the patient to it. The present growth of anesthesiology as a medical specialty and the dependence on it by surgeons stimulate better training of anesthetists.

The progress of surgery has kept pace with anesthesiology. The major strides have occurred in the physiologic aspect of surgery. The electrolyte and fluid-balance features of preoperative and postoperative treatment have been recognized and embodied in the general knowledge of surgery. There is continual improvement in various techniques. As new information is uncovered by research, better procedures are de-

veloped. Procedural controversies are resolved by common interest and combined effort of surgeon and anesthetist.

The impact of an anomaly, especially a visible one, is fully realized only by the one afflicted or by those close to him. Others of us feel compassion toward such an unfortunate person but we can never, of course, measure his profound psychologic depression, which is the same whether the deformity is congenital or acquired.

A conspicuous anomaly initiates a marked personality change in most adult victims. In the infant and the child, the effect is first of all on the immediate family, particularly the mother, until the child enters the preschool age, when he meets new friends and playmates, some of whom may be unkindly critical. At this time the child realizes how different he is from other children. To protect himself from unkind nicknames and remarks, his tendency is to avoid others altogether, and thus the psychologic harm begins.

This emphasizes the importance of early surgery. For the child's benefit, surgery should be performed well before he has reached the immediate preschool age. For the family's benefit (or peace of mind), surgical correction should be made as soon as possible after birth.

In many anomalies of the congenital type, the correction can be made shortly after birth. The only deterring factors are possible interference with the growth centers and the patient's ability to withstand surgery, including anesthesia. If the growth centers are traumatized by the procedure, a cessation of growth may result in a new abnormality superimposed on the original one. In fact, the postoperative defect may be worse than the original anomaly, which may have involved only the soft tissues. The concern for the growth centers is especially important in skeletal anomalies, more so than in those affecting soft tissue. This possibility usually precludes the use of radiation of any type in infants and children.

Among the facial structures, the nose is the least amenable

to early surgery. The nose continues to grow until about the sixteenth year. If surgical trauma interrupts the normal growth, some degree of saddlenose or other deformity may occur. Small nasal corrections, particularly those of the nasal tip or other soft tissue, are permissible if the deformity warrants it. However, correction of the nose may not be lasting, as the eventual shape is unpredictable even when growth centers are not involved. Nasal corrections in youth are usually limited to the secondary nasal-tip deformity associated with cleft lip and palate, and traumatic defects that demand immediate repair, such as dislocation of the parts resulting in obstruction.

Very early surgery, especially in the treatment of palatal defects, is imperiled by so many anesthetic or physiologic problems that it is usually not attempted. Many surgeons feel that the period from the twelfth to the eighteenth month is the optimal time for corrective surgery of the palate. Others prefer to defer surgery until the maxillary arch has had more time to develop unhindered. This ensures more normal dentition. In treatment of cleft palate, one of the chief aims is normal speech. This is rarely achieved by early cleft-palate surgery without an effective speech-training program for the child as soon as possible after closure. A dental prosthesis or an obturator helps in swallowing and permits the patient to form good speech habits. The normal growth of the maxillary arch is not restricted, and good dentition is encouraged. At a later date, about the fifth year or later, surgical correction can be done more safely and with a chance of anatomic perfection.

The reconstruction of the cleft lip need not await the cleft-palate closure. As no skeletal interference is present in the lip proper, the closure of the upper lip should be performed soon after the child has regained its birth weight. If other pertinent factors are favorable, a protruding premaxilla of a double cleft can be repositioned at this time, in order to effect a closure. Complete or nearly complete cleft of the upper lip should be repaired within the first three months.

In general, other deformities, congenital or acquired, should be eradicated by surgery as early as possible. Some birthmarks, it must be remembered, regress or disappear. Therefore surgery may be delayed indefinitely, as these defects are of a cosmetic nature and seldom involve function. It is well to inform parents of this.

Many pediatricians measure an infant's health by its gain in weight. When the infant has oriented itself to its new environment well enough to regain the ounces which are normally lost shortly after birth, it is able to withstand the shock of minimal surgery. With acquired defects, the time to operate depends on the degree of the trauma rather than on the general good health of the patient. It is best to combine the reconstructive surgery with first-aid treatment, as frequently a secondary procedure can be eliminated in this way. The patient's contours as they will be in adulthood—particularly the face—must be carefully considered when reconstructive work is performed in youth. If skeletal corrections are necessary, it is advisable to defer any attempt at final reconstruction until after puberty.

An axiom to follow concerning all deformities is that when restoration of function is imperative, cosmesis is secondary. The surgical plans should, however, consider both the functional and the cosmetic results.

It has been the authors' aim to compile material that will be of service to the general practitioner, the dentist, and the pediatrician in acquainting their patients or patients' relatives with the best time for and the possibilities of reconstructive surgery. Therefore, little space has been spent on techniques and operative procedures. Just enough of the method is presented to equip the reader with the essential means for visualizing the problem. Readers who wish to investigate more thoroughly all the steps of the procedures can find them in any of a number of textbooks on general plastic surgery.

CHAPTER 2

General Aspects of Pediatric Surgery

Surgery in the infant or child is a specialized field, not merely a scaled-down form of adult surgery. Besides the variations of immature anatomy and physiology, there are specific indications and special skills with which the pediatric surgeon must be familiar. Patients in this age group show a tolerance for surgery in direct proportion to the surgeon's ability and planning. They particularly lack tolerance for loss of blood, and their sensitivity to exposure and trauma requires short operative procedure, gentle tissue manipulation, and expert administration of anesthesia.

HISTORY

Much can be learned of the patient and his condition by careful history taking, including the past history as well as the illness or anomaly currently under consideration. In recording data, it is best to express age in hours for the newborn infant

and in months for the child under two years. Sex should be recorded, not only because of its bearing on such anomalies as malformation of the genitourinary organs, but also because it may affect the cosmetic importance or the possibility of concealment of certain abnormalities. For instance, the significance of an abnormality in the beard area of a male will differ from scarring on the chin of a female; a female is better able to conceal an ear abnormality because her hair will cover it; the use of cosmetics by females may hide a birthmark. It is well to bear in mind also that only male children exhibit hemophilia. Race is of interest in the study of anomalies and the type of skin healing to be expected after surgery. For instance, Negroes are more prone to keloid formation following burns, lacerations, or skin incisions than members of other races.

The history should always include evidence of recognized congenital malformation in both parental families, as this may give a clue to the cause of the child's present condition. If the findings of a careful and complete examination done shortly after delivery are charted, they will form a basis for future studies of the patient.

It must be remembered when obtaining the infant's history from the mother or other members of the family that they probably are not trained medical observers and that the intelligence of the individual giving the history determines its value to a great extent. An emotional parent may not be a reliable source for the history. For diagnostic purposes, the history may be at least as important as the physical examination.

As infants and young children cannot evaluate their subjective symptoms, the history is necessarily based on objective findings such as rashes, scars, and external anomalies, plus whatever symptoms the parents or nurses report from their observation.

PHYSICAL EXAMINATION

Although a complete physical examination is desirable, it is often advisable to proceed directly to the focal condition and evaluate the findings in the light of the proposed surgery. It is well to remember that a trusting child will frequently exhibit unusual fortitude. An explanation of what is to be done, if the child is old enough to understand and cooperate, will often be rewarded by complete cooperation. The well-advised physician will even tell the child that the examination may be painful; but if severe pain must be inflicted, sedation or, if necessary, general anesthesia should be administered.

Such objective findings as the temperature, pulse, respiration, and blood pressure must be evaluated according to the child's age. The temperature in infants and young children averages slightly higher than in adults. Extremes of temperature over a broad range are produced by slight causes in the child. However, a rise in temperature does not always mean that infection is present. Infants and young children show temperature elevations unaccountably or for trivial causes, such as slight gastric upsets, colds, teething, or mild neurogenic disturbances such as might be induced by environmental changes.

The pulse is normally irregular in the infant. It ranges from 130 to 140 beats per minute at birth to 110 to 120 in the first and second years, and 90 to 100 from the second to the fourth year, finally slowing to the level of 75 to 80 at puberty. Crying or any trivial excitement may increase the rate 20 or 30 beats per minute even in adults.

The respirations of the infant are irregular, varying from an average of 25 to 35 per minute up to the first year and gradually diminishing to about 18 to 20 per minute during the fifth year. The breathing is usually of the abdominal type until the tenth year, after which the costal type predominates.

The blood pressure is impossible to determine by the usual

methods prior to the third year. From the third to the eighth year, the pressure averages 85 to 95 mm Hg in systole to 70 in diastole. The pressure rises slowly to 105 or 110/80 at puberty.

LABORATORY EXAMINATION

Urine

Urinalysis for children and infants does not vary from the adult standard. It may be difficult to collect a urine specimen from the infant. For the male child a test tube can be strapped to the penis with adhesive; for the female a special apparatus can be used which is similar to a rubber-bag urinal, or catheterization may be resorted to. However, catheterization of small children should not be attempted unless it is clearly indicated, as when a sterile specimen is necessary.

Blood

Blood studies may include volumetric determination, cellular contents, and bleeding and coagulation time. There may be indication for such tests as CO_2 combining power and level of blood chlorides, sugar, or proteins, especially in severe burn cases.

The blood volume averages one-nineteenth of the body weight at birth and increases to one-thirteenth to one-tenth by the end of the first year. The loss of 10 per cent of the blood volume is a serious threat to the life of the infant or young child, while a loss of 20 per cent is generally fatal. The surgeon's efforts to preserve the blood volume at the highest possible level will go far to ensure rapid postoperative recovery.

Caution is needed in obtaining a blood specimen from an infant. The vascular tree of the infant is not readily accessible except in the larger vessels. There are certain dangers associated with collecting blood from a vessel of the extremities. Although the femoral vessels may be readily accessible, puncture can cause spasm throughout this terminal vascular tree