Nephrology and Urology for the Pediatrician

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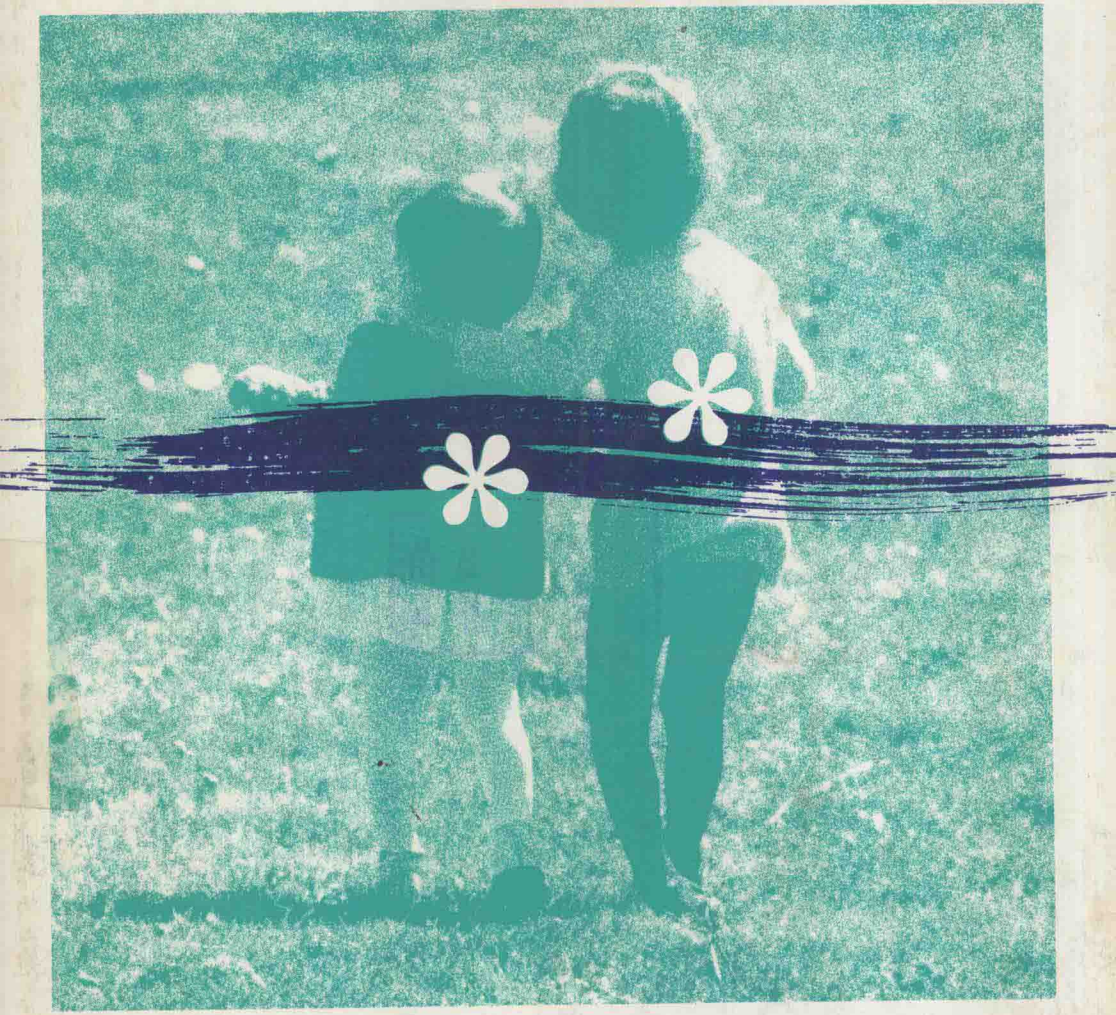

Nephrology and Urology for the Pediatrician

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Nephrology and Urology for the Pediatrician

To Jellie, Norma, and Shirley

Preface

Renal and urologic diseases are among the common chronic illnesses that are replacing acute diseases as major health problems in children. In this process the role of the pediatrician (this term is used throughout to refer to all physicians and other health professionals who provide general or other types of special care for infants, children, and adolescents) in all aspects of good general care of children with such chronic illnesses and handicaps has become increasingly important. Concomitantly, however, the greater biomedical and technological complexity in the diagnosis and treatment of some patients has required the development of highly trained organ system specialists such as pediatric nephrologists and urologists. The expanding complexity in the care of these patients has tended to persuade many pediatricians to refer increasing numbers of patients with renal or urologic diseases earlier and earlier to specialists.

This new text on renal and urologic diseases in children has features that distinguish it from others. As pediatric nephrologists, we have undertaken the task of defining how much of the differential diagnosis and treatment of renal and urologic disease in children can be done confidently by the pediatrician and when consultation with a pediatric nephrologist or urologist is indicated. We believe that the investigation and treatment of most of these children can and should be done initially (and in many instances throughout) by the pediatrician. Reluctance to assume these responsibilities, heightened often by warnings from pediatric nephrologists and urologists, stems from the concern that the diagnosis of a condition requiring early specialized treatment may be delayed, or that important new aspects of treatment may have been missed. This reluctance and these concerns may be justified unless the pediatrician is able to maintain a knowledge of advances in the field. All physicians must have accurate, current information on diagnosis and treatment of the patients they manage themselves. Equally important, pediatricians must understand the problems of the children for whom they seek consultation and for whose care they will continue to share the responsibility. Major goals of this book are to lessen these concerns, to encourage pediatricians to care for children with renal and urologic diseases with greater confidence, and to assist them in seeking consultation more selectively.

The text is problem oriented. It presents the differential diagnosis of the signs and symptoms of renal and urologic disease that constitute the initial complaints presented to the pediatrician. Discussion of the differential diagnosis and treatment of the various clini-

cal entities representing the major renal and urologic diseases in children follows.

It is anticipated that *Nephrology and Urology for the Pediatrician* will serve to fulfill an unmet need of medical students, residents, pediatricians, family physicians, and other health professionals. If it contributes to increasing the knowledge of all professionals concerning advances in the diagnosis and treatment of children with renal and urologic diseases, and especially if it encourages pediatricians to care for more of these patients on their own, and to refer patients more selectively, it will have fulfilled its goals.

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B. G.
C. M. E., Jr.
H. L. B.

Introduction

The content and structure of this book are designed to fulfill the purposes described in the Preface. An important feature is that it is closely related and cross-referenced extensively to a comprehensive textbook (edited by one of the present authors) of renal and urologic disease in children (C. M. Edelmann, Jr. [Ed.], *Pediatric Kidney Disease*. Boston: Little, Brown, 1978.). Accordingly, the text does not attempt to be encyclopedic in the coverage of its subjects, nor does it include detailed discussions of the etiology, pathogenesis, and pathophysiology of renal and urologic diseases of infants and children. Rather, major emphasis is placed on diagnosis and management; the symptoms and diseases included have been selected because they represent the great majority of those for which children are brought to the pediatrician.

Recommendations are made about the point in the management of a child with a renal or urologic disorder when the pediatrician should seek consultation. However, it should be noted that the decision to seek consultation will vary under different circumstances, such as the amount of experience the pediatrician has in caring for children with renal and urologic disorders and the facilities that are available. The recommendations also will change as knowledge of renal disorders increases.

The evaluation and initial differential diagnosis of the major signs and symptoms associated with renal or urologic disease are presented in Section I. Section II discusses clinical features and differential diagnosis of the major clinical entities, each of which may be associated with one or several underlying diseases. Treatment and prognosis of major renal and urologic syndromes are covered in Section III, and management of specific disorders is discussed in Section IV. Some normal laboratory values and methods for determinations that are readily done in the office of the pediatrician are given in the Appendix.

The most effective use of this text can be demonstrated by two examples. A newborn infant is discovered to have an abdominal mass. In approaching the differential diagnosis of this problem, the physician can turn to Chap. 1, Manifestations of Renal and Urologic Disease in the Newborn, and to Chap. 5, Abdominal Mass. On the basis of the initial examination, it is apparent that the infant has a urologic disorder, and consultations with the pediatric urologist and pediatric radiologist are requested. If, for example, the diagnosis of hydronephrosis were made, further information about this condition could be obtained from Chaps. 18 and 38, which deal with the clinical features and management of this disorder. Had this infant

proved to have infantile polycystic disease rather than hydronephrosis, consultation and joint management with the pediatric nephrologist would have been indicated. The discussion of polycystic kidney disease in Chap. 18 is intended to provide the physician with a sufficient understanding of the disorder to be able to deal effectively with the patient and his family and to work cooperatively with the pediatric nephrologist. If a more detailed exposition of the pathogenesis and histologic features of polycystic kidney disease were desired, the reader could refer to Chap. 40 of *Pediatric Kidney Disease*. If the infant were found to be uremic, principles of management of renal insufficiency could be reviewed in this text in Chap. 31 and 32, with a more extensive discussion being available in *Pediatric Kidney Disease*.

As another example, a 7-year-old boy is brought to his practicing physician because of the abrupt onset of painless gross hematuria noted on arising that morning. It seems apparent that the child has a renal or urologic disorder. The physician could consult Chap. 13, Gross Hematuria, for information concerning differential diagnosis. In that chapter, it is pointed out that red urine is not invariably indicative of hematuria, and that other conditions must also be considered. Red blood cells are found in the urine, however, and the diagnosis of hematuria is confirmed.

From the history, physical examination, and urinalysis with the finding of red blood cell casts, a differential diagnosis is made of some form of acute glomerulonephritis. Further evaluation may require the aid of laboratory examinations not available in the pediatrician's office, but unless a renal biopsy or urologic procedure is required or there are indications of renal failure, no consultation would be necessary at this point. The pediatrician might then turn to Chap. 15 for a more comprehensive discussion. In approaching the clinical problem of acute glomerulonephritis, Chap. 28 provides guidelines concerning management and prognosis.

From the initial evaluation, response to treatment, and early course, the diagnosis of acute poststreptococcal glomerulonephritis is made. Most children with this disease do not require consultation; if, however, there are early complications or if the usual benign course is aggravated by symptoms or findings such as persistence of hypertension, elevated serum creatinine after 4 to 6 weeks, or persistent proteinuria or hematuria for more than 3 to 6 months, consultation with a pediatric nephrologist may be warranted. If a renal biopsy is done at that time and the tissue diagnosis proves to be a form of chronic disease (such as membranoproliferative glomerulonephritis), the continuing care will usually be undertaken jointly by the pediatrician and the pediatric nephrologist. Additional information about membranoproliferative glomerulonephritis can be found

in Chaps. 16 and 33. The pediatrician sharing in the care of the patient may want to refer also to Chap. 51 in *Pediatric Kidney Disease*, or to other references, for more detailed discussions of the pathogenesis, histopathology, course, and treatment of this disease.

These examples demonstrate how the structure and content of this book permit a different manner of use from that of others on renal and urologic disease in children. The intent is to provide pediatricians with the knowledge and confidence necessary to deal with the majority of children with renal and urologic disorders who come under their care, to seek consultation selectively, and to work effectively with consultants in the management of the occasional child with a complex disorder. Used as suggested, we believe the book can lead to better care of children with renal and urologic disease through better identification of the roles of the pediatrician and of the pediatric nephrologist and urologist, and more effective cooperation between them.

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I. Differential Diagnosis of Signs and Symptoms That May Be Associated with Renal or Urologic Disease

NOTICE

The indications and dosages of all drugs in this book have been recommended in the medical literature and conform to the practices of the general medical community. The medications described do not necessarily have specific approval by the Food and Drug Administration for use in the diseases and dosages for which they are recommended. The package insert for each drug should be consulted for use and dosage as approved by the FDA. Because standards for usage change, it is advisable to keep abreast of revised recommendations, particularly those concerning new drugs.

1. Manifestations of Renal and Urologic Disease in the Newborn

Symptoms due to abnormalities of the urinary tract in the newborn are quite variable and they may include those suggestive of renal insufficiency, with or without oliguria, an abdominal mass, edema, dehydration or acidosis, or unexplained fever. They may or may not be accompanied by urinary abnormalities such as proteinuria, hematuria, or pyuria. On very rare occasions, careful examination may reveal hypertension as the presenting manifestation.

SYMPTOMS DUE TO RENAL INSUFFICIENCY

Renal insufficiency must be suspected in any infant with oliguria or anuria, with or without accompanying symptoms. Failure to thrive, poor feeding, vomiting, lethargy, and seizures in the newborn also must suggest the possibility of renal insufficiency. These symptoms may be due to many nonrenal causes, including heart failure, a variety of metabolic disorders, adrenal insufficiency (particularly congenital adrenal hyperplasia), sepsis, and neurologic disease.

Heart failure is evidenced by the presence of organic murmurs, cardiac enlargement (however, this is not always present and is often difficult to assess in the neonate), reduced blood pressure and pulse pressure, pale and cool extremities with weak pulses, and marked enlargement of the liver. Generalized sweating may be particularly prominent in infants.

Infants with metabolic disorders that are manifested in the newborn period usually are seriously ill. They often have severe metabolic acidosis, with an increased anion gap. Appropriate metabolic studies of blood and urine are necessary to establish the diagnosis (Chap. 8).

Infants with adrenal insufficiency present with dehydration and circulatory inadequacy and develop cardiovascular collapse. Female infants may show evidence of virilization of the external genitalia. Their serum sodium may be normal or low, but the serum potassium is almost invariably elevated to 6 mEq per liter or greater. Despite these abnormalities, the urinary concentration of sodium may be high and that of potassium low. The diagnosis is established by performing appropriate studies of adrenocortical metabolism.

The presence of sepsis can only be suspected clinically; the diagnosis is established by obtaining cultures of stool, urine, spinal fluid, and blood.

The general differential diagnosis of renal insufficiency in the

Table 1-1. Differentiation of Functional from Organic Renal Failure in Infants

Measurement	Functional	Organic
U_{Na}	<20 mEq/L	>70 mEq/L
U/P osmolality	>2	<1.1
U/P urea	>15	<5
U/P creatinine	>15	<5
Fractional excretion of sodium	<1.8%	>10%

Note: U/P = urinary to plasma ratio; fractional excretion of sodium = proportion of filtered sodium excreted in urine, calculated as $(U_{Na} \times P_{Cr}) \div (U_{Cr} \times P_{Na}) \times 100$.

Source: A. Spitzer. Diseases of the Kidney in the Newborn. In C. M. Edelmann, Jr. (Ed.), *Pediatric Kidney Disease*. Boston: Little, Brown, 1978.

newborn, as in older patients, is initially between functional, organic, and obstructive renal failure. Functional renal failure occurs secondary to inadequate perfusion of the kidneys, due to dehydration, shock, sepsis, heart failure, or blood loss. The underlying disturbance usually is apparent clinically, but at times discrimination between functional and organic renal failure may be difficult. Laboratory assessment, as shown in Table 1-1, may be helpful.

Obstructive renal failure may occur in newborns with obstructive uropathy or in those with a neurogenic bladder, due usually to myelomeningocele.

Organic renal failure in the newborn is associated most often with hypoplastic or dysplastic kidneys, urologic abnormalities, cystic disorders, or with sepsis. It may develop in infants with bilateral renal venous thrombosis or as a nephrotoxic reaction to drugs, such as the aminoglycoside antibiotics. Renal insufficiency in the newborn is rarely secondary to some form of glomerulonephritis.

Oligohydramnios should raise the suspicion of severe renal impairment, since urine excreted by the fetus is a major source of amniotic fluid. Renal agenesis, severe hypoplasia or cystic disease, or urinary tract obstruction may be associated with the so-called Potter facies, consisting of hypertelorism, large low-set ears, pronounced epicanthic folds, micrognathia, and a broad flattened nose (Fig. 1-1). These abnormalities often coexist with pulmonary hypoplasia. The entire constellation has been described in infants born of mothers with oligohydramnios secondary to chronic leakage of amniotic fluid. Since these infants have normal kidneys, it would appear that fetal compression secondary to oligohydramnios, rather than renal insufficiency per se, is the cause of the Potter syndrome.