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Preface: A Gamut of Kidney Diseases in Children xv
Tej K. Mattoo

Assessment of Kidney Function in Children 1017
Emil den Bakker, Arend Bökenkamp, and Dieter Haffner

A good understanding of kidney function tests is essential for patient care. Urinalysis is the commonest used test for screening purposes in ambulatory settings. Glomerular function is assessed further by urine protein excretion and estimated glomerular filtration rate and tubular function by various tests such as urine anion gap and excretion of sodium, calcium, and phosphate. In addition, kidney biopsy and/or genetic analyses may be required to further characterize the underlying kidney disease. In this article, we discuss maturation and the assessment of kidney function in children.

Evaluation of Proteinuria and Hematuria in Ambulatory Setting 1037
Abubakr A. Imam and Sermin A. Saadeh

Proteinuria and/or hematuria are common findings in ambulatory settings. Proteinuria can be glomerular and/or tubular in origin and it may be transient, orthostatic, or persistent. Persistent proteinuria may be indicative of a serious kidney pathology. Hematuria, which denotes the presence of an increased number of red blood cells in the urine, can be gross or microscopic. Hematuria can originate from the glomeruli or other sites of the urinary tract. Asymptomatic microscopic hematuria or mild proteinuria in an otherwise healthy child is less likely to be of clinical significance. However, the presence of both requires further workup and careful monitoring.

Acute Postinfectious Glomerulonephritis 1051
Minh Dien Duong and Kimberly J. Reidy

Postinfectious glomerulonephritis (PIGN) is a leading cause of acute glomerulonephritis in children. The presentation of PIGN can vary from asymptomatic microscopic hematuria incidentally detected on routine urinalysis to nephritic syndrome and a rapidly progressive glomerulonephritis. Treatment involves supportive care with salt and water restriction, and the use of diuretic and/or antihypertensive medication, depending on the severity of fluid retention and the presence of hypertension. PIGN resolves completely and spontaneously in most children, and the long-term outcomes are typically good with preserved renal function and no recurrence.
Current Understanding of Nephrotic Syndrome in Children

Tej K. Mattoo and Sami Sanjad

Nephrotic syndrome in children is mostly idiopathic in origin. About 90% of patients respond to corticosteroids; 80-90% have at least one relapse and 3-10% become corticosteroid resistant after the initial response. A kidney biopsy is seldom indicated for diagnosis except in patients with atypical presentation or corticosteroid resistance. For those in remission, the risk of relapse is reduced by the administration of daily low dose corticosteroids for 5-7 days at the onset of an upper respiratory infection. Some patients may continue having relapses through adult life. Many country-specific practice guidelines have been published, which are very similar with clinically insignificant differences.

Urinary Tract Infection in Children

Per Brandström and Sverker Hansson

Symptoms of urinary tract infection (UTI) in young children are nonspecific and urine sampling is challenging. A safe and rapid diagnosis of UTI can be achieved with new biomarkers and culture of clean-catch urine, reserving catheterization or suprapubic aspiration for severely ill infants. Most guidelines recommend ultrasound assessment and use of risk factors to direct further management of children at risk of kidney deterioration. The increasing knowledge of the innate immune system will add new predictors and treatment strategies to the management of UTI in children. Long-term outcome is good for the majority, but individuals with severe scarring can develop hypertension and decline in kidney function.

Primary Vesicoureteral Reflux and Renal Scarring

Tej K. Mattoo and Dunya Mohammad

Vesicoureteral reflux (VUR) is the commonest congenital anomaly of urinary tract in children. It is mostly diagnosed after a urinary tract infection or during evaluation for congenital anomalies of the kidney and urinary tract. High-grade VUR, recurrent pyelonephritis, and delayed initiation of antibiotic treatment are important risk factors for renal scarring. The management of VUR depends on multiple factors and may include surveillance only or antimicrobial prophylaxis; very few patients with VUR need surgical correction. Patients with renal scarring should be monitored for hypertension and those with significant scarring should also be monitored for proteinuria and chronic kidney disease.

Antenatally Diagnosed Kidney Anomalies

Caoimhe S. Costigan and Norman D. Rosenblum

Congenital anomalies of the kidney and urinary tract encompass a broad spectrum of developmental conditions that together account for the majority of childhood chronic kidney diseases. Kidney abnormalities are the most commonly diagnosed congenital anomaly in children, and detection of this anomaly is increasing as a result of improved antenatal care and widespread access to more sensitive screening ultrasonography. Most paediatricians will encounter children with congenital kidney anomalies across a wide spectrum of disorders, and a broad understanding of the
classification, investigation, and basis of management is important to appropriately direct their care.

Diagnosis and Management of Nephrolithiasis in Children

Larisa Kovacevic

The incidence of kidney stones in children is increasing. Approximately two-thirds of pediatric cases have a predisposing cause. Children with recurrent kidney stones have an increased higher risk of developing chronic kidney. A complete metabolic workup should be performed. Ultrasound examination is the initial imaging modality recommended for all children with suspected nephrolithiasis. A general dietary recommendation includes high fluid consumption, dietary salt restriction, and increased intake of vegetables and fruits. Depending on size and location of the stone, surgical intervention may be necessary. Multidisciplinary management is key to successful treatment and prevention.

Hypertension in Children and Young Adults

Emily Haseler and Manish D. Sinha

Primary hypertension (PH) is most common during adolescence with increasing prevalence globally, alongside the epidemic of obesity. Unlike in adults, there are no data on children with uncontrolled hypertension and their future risk of hard cardiovascular and cerebrovascular outcomes. However, hypertension in childhood is linked to hypertensive-mediated organ damage (HMOD) which is often reversible if treated appropriately. Despite differing guidelines regarding the threshold for defining hypertension, there is consensus that early recognition and prompt management with lifestyle modification escalating to antihypertensive medication is required to ameliorate adverse outcomes. Unfortunately, many unknowns remain regarding pathophysiology and optimum treatment of childhood hypertension.

Hemolytic-Uremic Syndrome in Children

Olivia Boyer and Patrick Niaudet

Hemolytic uremic syndrome is characterized by a triad of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney failure. Most cases are caused by Shiga-toxin-producing bacteria, especially Escherichia coli. Transmission occurs through ground beef and unpasteurized milk. STEC-HUS is the main cause of acute renal failure in children. Management remains supportive. Immediate outcome is most often. Atypical HUS represents about 5% of cases, has a relapsing course with more than half of the patients progressing to end-stage kidney failure. Most cases are due to variants in complement regulators of the alternative pathway. Complement inhibitors, such as eculizumab, have considerably improved the prognosis.
Pediatric vasculitis is a complex group of disorders that commonly presents with multisystem involvement. Renal vasculitis can be isolated to the kidneys or can occur as part of a broader multiorgan vasculitis. Depending on severity, renal vasculitis may present as acute glomerulonephritis (AGN) often associated with hypertension and sometimes with a rapidly deteriorating clinical course. Prompt diagnosis and initiation of therapy are key to preserving kidney function and preventing long-term morbidity and mortality. This review focuses on the clinical presentation, diagnosis, and treatment objectives for common forms of renal vasculitis seen in pediatric patients.

Acute kidney injury (AKI) is common in children and is associated with significant morbidity and mortality. In the last decade our understanding of AKI has improved significantly, and it is now considered a systemic disorder that affects other organs including heart, lung, and brain. In spite of its limitations, serum creatinine remains the mainstay in the diagnosis of AKI. However, newer approaches such as urinary biomarkers, furosemide stress test, and clinical decision support are being increasingly used and have the potential to improve the accuracy and timeliness of AKI diagnosis.

Chronic kidney disease (CKD) in children occurs mostly due to congenital anomalies of kidney and urinary tract and hereditary diseases. For advanced cases, a multidisciplinary team is needed to manage nutritional requirements and complications such as hypertension, hyperphosphatemia, proteinuria, and anemia. Neurocognitive assessment and psychosocial support are essential. Maintenance dialysis in children with end-stage renal failure has become the standard of care in many parts of the world. Children younger than 12 years have 95% survival after 3 years of dialysis initiation, whereas the survival rate for children aged 4 years or younger is about 82% at one year.