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NEPHROLOGY - I

APPROACH TO HEMATURIA

***Seher Kamal**
****Arpana A. Iyengar**

Abstract: *Hematuria is defined as the presence of five or more red blood cells (RBCs) per high-power field of view (×40) on microscopic examination. There are many causes of hematuria and the differential diagnosis depends on whether it is glomerular or non-glomerular in origin. Clinical presentation and urine microscopy can differentiate glomerular from non-glomerular causes. Rarer causes of hematuria may need more detailed evaluation with a renal biopsy, phase-contrast microscopy and other modalities. The common causes of hematuria and basic approaches to its diagnosis are discussed in this review.*

Keywords: *Urine microscopy, Hematuria, Red blood cells.*

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Points to Remember

- ***It is mandatory to determine the origin of hematuria whether it is glomerular or non-glomerular.***
- ***A detailed history, careful physical examination, and focussed laboratory investigations are essential in the work up.***
- ***The presence of hypertension and proteinuria should prompt investigation for glomerular involvement.***
- ***An early and accurate diagnosis in children with familial hematuria provides important information regarding the plan of management and possible need for intervention.***
- ***Genetic counselling plays an important role in familial forms of hematuria.***

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NEPHROLOGY - I

ACUTE KIDNEY INJURY - CLASSIFICATION AND MANAGEMENT

***Uma Maheshwari M**
****Alpana Ohri**

Abstract: *Acute kidney injury is one of the common complications in critically ill children with increasing incidence. The criteria, 'pediatric reference change value optimized for AKI in children' defines acute kidney injury as an increase in serum creatinine of ≥ 20 μmol/L with an increase of ≥ 30% within 7 days. Evaluation involves the early detection of at risk patients with the help of biomarkers and renal angina index. Early acute kidney injury is managed by maintaining euvolemia, sustaining normotension, avoiding nephrotoxicity and treating concomitant sepsis. Advanced acute kidney injury needs renal replacement therapies such as continuous renal replacement therapy or sustained low-efficiency dialysis in hemodynamically unstable patients using special and conventional hemodialysis machines respectively. Peritoneal dialysis is adopted for small children with difficult vascular access. Survivors of acute kidney injury need long term monitoring of renal functions.*

Keywords: *Acute kidney injury, Classification, Renal replacement therapy, Hemodialysis.*

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Points to Remember

- ***Microcirculatory dysfunctions is an important mechanism of renal injury, where there is normal or increase in global renal blood flow rather than reduced renal perfusion.***
- ***pROCK criteria gives a newer definition of staging of AKI based on rise in serum creatinine value over a 7 day period, and it is less sensitive than KDIGO criteria.***
- ***Renal angina index and biomarkers play a crucial role in predicting severe AKI.***
- ***Conservative management of AKI includes fluid management, e.c. restricting or liberal fluids based on fluid status, avoiding nephrotoxic drugs, correcting dyselectrolytemias and hyperuricemia.***
- ***Renal replacement therapy is indicated in a fluid overloaded state with oliguria, dyselectrolytemias (when not medically controlled) and hypercatabolic state.***
- ***Modality of RRT is chosen depending on the age, hemodynamic stability, need for solute removal, cost and duration of RRT.***

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NEPHROLOGY - I

RENAL TUBULAR ACIDOSIS - APPROACH AND MANAGEMENT

***Mohamed Azarudeen J**
****Mehul A Shah**

Abstract: *Renal tubular acidosis is a group of inherited and acquired tubular disorders presenting as hyperchloremic normal anion gap metabolic acidosis in the presence of normal kidney function. The four phenotypes include: Type 1- distal renal tubular acidosis, Type 2- proximal renal tubular acidosis (isolated or renal Fanconi syndrome), Type 3- combined proximal and distal tubular dysfunction and Type 4- aldosterone deficiency / resistance. Clinical presentation includes polyuria, polydipsia, failure to thrive and occasionally, refractory rickets and evaluation for renal tubular functions and genetic studies are essential for making the diagnosis. Management includes supplementation of alkali, potassium, phosphorus and active vitamin D3 and specific therapy for identified etiology. Early diagnosis and management improve long term outcomes.*

Keywords: *RTA, Renal tubular acidosis, Renal Fanconi syndrome, Normal anion gap metabolic acidosis, Renal rickets.*

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Points to Remember

- ***RTA is characterised by hyperchloremic normal anion gap metabolic acidosis.***
- ***Presenting features include failure to thrive, polyuria and polydipsia.***
- ***Proximal RTA is characterised by impaired proximal tubule bicarbonate reabsorption and distal RTA due to impaired distal hydrogen secretion.***
- ***The complete normalization of plasma bicarbonate levels with alkali supplementation suggests a diagnosis of dRTA.***
- ***Early diagnosis, adequate alkali supplementation and regular long-term follow-up with monitoring of growth, bony deformities and renal function is required.***

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NEPHROLOGY - I

NEPHROLITHIASIS

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Abstract: *Nephrolithiasis or kidney stones are being increasingly recognised in children over the last few decades. The etiology is multifactorial, but underlying metabolic disorders are the most common cause in children, warranting thorough evaluation. The clinical presentation is highly variable and ultrasound is the imaging modality of choice owing to its safety and availability. Careful identification of risk factors and modification, of them either through dietary changes or pharmacological interventions, form the mainstay of therapy. The goal should be appropriate treatment and diligent surveillance to prevent progression to chronic kidney disease. This review discusses the key aspects of nephrolithiasis in children.*

Keywords: *Nephrolithiasis, Nephrocalcinosis, Children, Renal stones, Hypercalciuria.*

Points to Remember

- ***Underlying metabolic abnormalities are the most important causes of pediatric nephrolithiasis, with predominantly hypercalciuria contributing to 70-80% of cases.***
- ***Clinical presentation is widely variable from asymptomatic presentation to chronic kidney disease.***
- ***Extensive metabolic workup with urine and blood parameters is mandatory in childhood nephrolithiasis.***
- ***Dietary interventions and medical management with closed surveillance can help prevent stone formation and recurrences.***
- ***Symptomatic children or those with failed medical therapy will require definitive surgical management.***

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NEPHROLOGY - I

APPROACH TO RICKETS

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Abstract: *Rickets is a disease of the growing bone, primarily caused by abnormal calcium and phosphate homeostasis that leads to impaired mineralisation of osteoid at the growth plate. Growth failure and bony deformities are the common features. The diagnosis is established through a clinical evaluation, radiological findings, and biochemical abnormalities, particularly elevated levels of alkaline phosphatase The broad classification of rickets if it is calcipenic or phosphopenic helps in the clinical evaluation and diagnosis. The approach to rickets should include systematic analysis of clinical features in the scheme of diagnostic work up. An overview of etiopathogenesis, clinical features, diagnostic workup and management of rickets are discussed.*

Keywords: *Calcipenic rickets, Phosphopenic rickets, Vitamin D, Genetics.*

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Points to Remember

- ***Rickets is a disease of the growing bone due to abnormal calcium and phosphate homeostasis and recent evidence suggests that hypophosphatemia (low serum Pi levels) is the denominator of all forms of rickets.***
- ***The broad classification into calcipenic and phosphopenic rickets help in the clinical evaluation and diagnosis.***
- ***The approach to rickets should include systematic analysis of clinical features coupled with diagnostic work up including genetic evaluation in situations indicated.***
- ***Nutritional deficiency of vitamin D or calcium is still the commonest cause of calcipenic rickets while genetic mutations affecting vitamin D metabolism or action, FGF23 production or degradation, renal phosphate handling or bone mineralization may cause refractory rickets.***
- ***Burosumab has been proven to be highly successful in treating XL HPR and Tumor induced osteomalacia.***

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NEPHROLOGY - I

RECENT ADVANCES IN PEDIATRIC NEPHROLOGY

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******Dua Priyanka**

Abstract: *The world has witnessed tremendous developments in the field of pediatric nephrology over the last two decades. Genetic analysis has played a significant role in the diagnosis and planning of preventive and therapeutic strategies for several kidney disorders. Numerous structural and functional biomarkers for acute kidney injury have now been identified. The application of monoclonal antibodies has led to improved survival. Dialysis and transplantation services for children are now more accessible than before. Recent evidence-based consensus guidelines on steroid-sensitive and steroid-resistant nephrotic syndrome, congenital nephrotic syndrome, hemolytic uremic syndrome, hypertension, chronic kidney disease and urinary tract infection have harmonized the evaluation and management of these common disorders.*

Keywords: *Pediatric nephrology, Recent advances, Kidney diseases, Genetics.*

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Points to Remember

- ***Next-generation sequencing can help establish a diagnosis or identify a monogenic cause in up to 30% of cases of steroid-resistant nephrotic syndrome, 60-70% of complement-mediated thrombotic microangiopathy, 50-80% of renal tubular disorders and 15-20% of severe kidney malformations.***
- ***Identification of a genetic cause will help in counselling the family, prognostication, and deciding on further therapeutic options.***
- ***Access to dialysis and transplantation services for children has significantly improved over the past decade and understanding the basic tenets of these techniques is necessary for the pediatricians to provide basic care and emergency services, whenever required.***
- ***Standard guidelines have now been framed for evaluating and managing common kidney disorders.***

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NEPHROLOGY - I

NEPHROTIC SYNDROME - WHAT IS NEW IN THE GUIDELINES

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Abstract: *This article summarizes the key essentials of nephrotic syndrome guidelines and briefly discusses its pathophysiology and clinical presentation. Recent treatment recommendations, evaluations, indications for genetic testing and kidney biopsies are also discussed. The indications for steroid-sparing agents, monitoring and follow-up have been objectively outlined as presented in recent guidelines. Supportive care measures and transition to adult care are explicated. The guidelines underscore evidence-based management, incorporating ungraded "practice points" when applicable. These are largely concordant with international recommendations, with limited exceptions. These comprehensive guidelines provide a framework for the management of nephrotic syndrome in diverse clinical scenarios.*

Keywords: *Nephrotic syndrome, Genetic testing, Renal biopsy, Transition.*

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Points to Remember

- ***Nephrotic syndrome is characterized by edema, severe proteinuria and hypoalbuminemia.***
- ***The pathophysiology involves a combination of podocyte dysfunction, immune-mediated mechanisms, hemodynamic factors, secondary causes, and genetic predisposition.***
- ***Nearly 10-20% of patients do not respond to steroids and are at risk of adverse outcomes and calcineurin inhibitors are the therapy of choice for these patients.***
- ***Kidney biopsy is required for steroid-resistant disease, persistent hematuria, acute kidney injury, and systemic features.***
- ***The Indian guidelines largely align with the international recommendations from the IPNA, with a few exceptions.***

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NEPHROLOGY - I

URINARY TRACT INFECTION AND VESICOURETERIC REFLUX - AN UPDATE

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Abstract: *Diagnosing urinary tract infections in young children presents unique challenges due to non-specificity of clinical manifestations and difficulty in collecting urine specimens. Identification of bacteriuria and leukocyturia while providing good diagnostic accuracy, is cumbersome to perform. Current evidence recommends using urine dipstick tests for a presumptive diagnosis of UTI. Most children diagnosed with a UTI can be effectively managed with oral antibiotics. Additionally, the approach to imaging after an initial UTI is becoming less aggressive, largely because there are limited effective interventions to prevent kidney scarring associated with vesicoureteral reflux. Majority of patients with vesicoureteric reflux can be managed without surgical intervention as it tends to resolve spontaneously.*

Keywords: *Acute pyelonephritis, Antimicrobials, Cystitis, Micturating cystourethrography, Vesicoureteral reflux.*

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Points to Remember

- ***Urine dipstick based on leukocyte esterase and nitrite can be used for presumptive diagnosis of urinary tract infections in children.***
- ***Ultrasound scan should be performed in all children following an episode of urinary tract infection; micturating cystourethrography and DMSA scan should be reserved for selected cases.***
- ***Majority of the patients with urinary tract infection can be managed with oral antibiotic therapy***
- ***Antibiotic prophylaxis is effective in preventing recurrence of urinary tract infection only in children with high grade vesicoureteric reflux***
- ***As primary vesicoureteric reflux often resolves spontaneously, majority of these patients can be managed without surgical intervention***

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GENERAL ARTICLE

HIGH-RISK NEONATES - FOLLOW UP PLANNING

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Abstract: *High-risk neonates require comprehensive follow-up planning to ensure optimal development and health outcomes. Timely identification and enrolment in follow-up programs are crucial for monitoring growth and addressing neurodevelopmental challenges. A multidisciplinary approach involving healthcare professionals from various fields enhances the effectiveness of interventions. Regular assessments and family education are essential to support the infant's development and facilitate early detection of potential issues. Ultimately, a well-structured follow-up program aims to improve the quality of life for high-risk infants and their families.*

Keywords: *High - risk neonate, Neuro developmental outcome, Follow-up plan.*

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Points to Remember

- ***Identify high-risk infants before NICU discharge to enable timely enrolment in a follow-up program.***
- ***Adopt a multidisciplinary approach involving pediatricians, developmental clinicians, therapists, and social workers for comprehensive care.***
- ***Conduct regular assessments at appropriate intervals using standardized tools like DASII or Hammersmith examinations to detect developmental delays.***
- ***Educate and support families on the importance of follow-up care, developmental expectations, available resources, and necessary interventions to foster optimal growth and development.***

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DRUG PROFILE

DOSAGE ADJUSTMENT OF MEDICATIONS IN OBESE ADOLESCENTS

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Abstract: *The obesity epidemic is a reality today in India. It alters the pharmacokinetics of medications, impacting loading doses, dosage intervals, plasma half-lives and the time to achieve steady-state concentrations. Weight-based dosing in obese children raises the risk of medication errors, as there are no readily available dosing guidelines. An attempt is made in this article to discuss some of the issues involved and also possible adjustments that could be made to prevent toxicity under-dosing of medications in this group of children.*

Keywords: *Obesity, Dosage adjustment, Drug dosage calculations, Pharmacokinetics.*

Points to Remember

- ***In obese children, drug pharmacokinetics are significantly altered due to changes in body composition, including a higher proportion of body fat that affects the volume of distribution (Vd) for lipophilic medications, which may require adjustments in dosing to avoid therapeutic failure or toxicity.***
- ***Awareness of dosing adjustments required for obese adolescents is a must, to avoid toxicity or under-dosing of various medications***
- ***Traditional weight-based dosing methods (e.g., total body weight) are often inadequate for this population, however alternative methods such as adjusted body weight (ABW) or body surface area (BSA) may be more appropriate, as they account for the unique physiological changes associated with obesity.***
- ***As pediatricians may not be able decide on the size descriptors to base doses for drugs and calculate doses accordingly, it may be prudent to have Table 1 on the desk top to help with dosing of various commonly used drugs in obese children.***
- ***Clinicians must be cautious and consider both the pharmacokinetic changes and the specific properties of the drug being prescribed to ensure safe and effective treatment and reduce risk of toxicity.***
- ***More studies are needed to establish specific dosing guidelines for obese pediatric patients.***

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CASE REPORT

A CASE OF GATA2 MUTATION PRESENTING AS AUTOIMMUNE HEMOLYTIC ANEMIA

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Abstract: *Inborn errors of immunity are one of the important causes of immune cytopenias. In patients with immune cytopenias genetic mutations are being recognized more often in recent times with the advent of molecular testing laboratory facilities. Identification of these genetic mutations plays an important role in the management and prognostication in these diseases. GATA 2 mutation is one such mutation associated with lymphopenia and immunodeficiency which presented as autoimmune hemolytic anemia in one of our patients.*

Keywords: *Autoimmune hemolytic anemia, GATA2 mutation.*

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