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Comparing the Type and Antibiotic Resistance of Microorganisms Causing Pyelonephritis in Children With and Without Functional Constipation

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ABSTRACT

Background and Aim: Constipation is one of the risk factors for pyelonephritis in children. This study compared the type and resistance of microorganisms causing pyelonephritis in children with and without functional constipation.

Methods: This cross-sectional study was done on children with pyelonephritis admitted to Amirkola Children's Hospital in Babol City, Iran, from 2014 to 2021. Children were included in the study using the census method and according to the inclusion criteria. The study population comprised all children aged 2 months to 18 years old, without consumption of any oral or injection of antibiotics 48 hours before admission. The microorganisms' sensitivity and drug resistance were evaluated using disk diffusion antibiograms and Clinical and Laboratory Standard Institute (CLSI) guidelines. A gastroenterologist diagnosed and confirmed constipation based on ROME III or IV criteria.

Results: Among 270 children with pyelonephritis, about 56 patients (20.7%) had functional constipation. Nitrofurantoin (95.2%) and ampicillin (67.2%) were the most sensitive and resistant drugs. Escherichia coli was the most common microorganism in both groups ($P<0.05$), and there was no difference in drug sensitivity or resistance in the two groups ($P<0.05$).

Conclusion: In the present study, about one-fifth of children with pyelonephritis had functional constipation. The type and antibiotic resistance of microorganisms causing pyelonephritis in children with and without functional constipation was not different. So, in children with pyelonephritis in both study groups (with and without functional constipation), initial empiric treatment can be performed based on a standard protocol.

Keywords: Children, Pyelonephritis, Functional constipation, Drug resistance

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Acute Kidney Injury and Hemolytic Uremic Syndrome in Scorpion Stung Children

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ABSTRACT

Background and Aim: Hemiscorpion lepturus (HL) scorpion stung children are often accompanied by acute kidney injury (AKI). In addition, hemolytic uremic syndrome (HUS) is seen in some patients, and some evidence suggests ADAMTS13 deficiency.

Methods: In a double-blind, open-label, randomized controlled trial, 98 patients with massive hemoglobinuria following HL scorpion sting were included in the intervention and control groups (49 patients per group).

Results: We observed AKI in 27.4% of patients. They were significantly younger and lighter (P=0.006, P=0.011, respectively). Also, significant differences existed between groups with and without AKI regarding their clinical findings, including fever (P=0.003), hypertension (P<0.001), hemolytic anemia (P<0.001), thrombocytopenia (P<0.001), massive proteinuria (P<0.001), hemoglobinuria (P<0.001), pyuria (P<0.001), and hematuria (P=0.004). HES was detected in 5.5% and disseminated intravascular coagulation in 14.6% of patients with a significant association with AKI (P<0.001). A multivariate regression model found several independent predictors for AKI: thrombocytopenia (P=0.002), pyuria (P=0.01), proteinuria (P=0.01), and fever (P=0.02). We also evaluated the role of fresh frozen plasma (FFP) in preventing kidney injuries in complicated cases. In a nut shell, FFP can significantly stop the deterioration of blood urea nitrogen (BUN) and creatinine (Cr) in the intervention group (P<0.001 and P=0.003, respectively). In analogy with the control group, proteinuria was significantly lower (P=0.012), reduction of platelet count was significantly less (P=0.044), and lactate dehydrogenase (LDH) and leukocytosis rise were lower in the intervention group (P=0.037 , P=0.007, respectively).

Conclusion: The predictors for AKI in children after HL scorpion sting comprise young age, delay in medical care, pigmenturia, microangiopathic hemolytic anemia, proteinuria, and pyuria. FFP can significantly avert kidney damage after an HL scorpion sting. Along with other standard treatments, avoiding the deadly venom complications in severe cases is a safe approach.

Keywords: Acute kidney injury, Hemolytic uremic syndrome, Child

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The Post Remission of Hyperlipidemia in Children With Idiopathic Nephrotic Syndrome

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ABSTRACT

Background and Aim: Idiopathic nephrotic syndrome is a common and troublesome chronic disease in children, which requires complex management and has many complications. Hyperlipidemia is part of the laboratory profile of patients with nephrotic syndrome due to a decrease in lipoprotein lipase and response to hypoalbuminemia. However, we have many cases of hyperlipidemia after the recovery of nephrotic syndrome. This study investigated the frequency and predictivity of clinical hyperlipidemia to reach appropriate management and improve outcomes.

Methods: A total of 256 children with idiopathic steroid-sensitive nephrotic syndrome were evaluated for hyperlipidemia. We compared the clinical characteristics of 2 groups with and without hyperlipidemia. Hyperlipidemia was defined as hypercholesterolemia and hypertriglyceridemia for age and gender. We also divided the patients into two groups: steroid-dependent and non-steroid-dependent idiopathic nephrotic syndrome. Steroid-dependent nephrotic syndrome (SDNS) was defined as ≥2 relapses during steroid-reducing treatment or 28 days after discontinuation of corticosteroids.

Results: About 58.5% of the patients had hyperlipidemia during remission, with a higher incidence in patients with SDNS (P=0.04). The mean age at the last relapse was significantly higher in children with hyperlipidemia than in those without hyperlipidemia (P=0.003). The relapse rate was significantly higher in children with hyperlipidemia compared to those with low frequent relapse (P=0.004). Hyperlipidemia was not statistically correlated with gender, younger than 2 years age at presentation, allergy history, hypertension, and body mass index (BMI) at diagnosis. SDNS was found in 57% of all cases, and 65.5% were male. Compared to those with low frequent relapse, children with SDNS showed higher BMI at diagnosis (P=0.002), relapse/year (P<0.001), total relapses (P<0.001), and age of last relapse (P=0.006). However, with immunosuppressive treatment, the mean relapse rate decreased significantly in SDNS (P<0.001). Patients with SDNS had significantly longer follow-up than the other group (P<0.001).

Conclusion: Hyperlipidemia is a frequent finding after remission of idiopathic nephrotic syndrome, and it was more frequent in SDNS and older children. Follow-up of these cases for hyperlipidemia is recommended.

Keywords: Hyperlipidemia, Child, Nephrotic Syndrome

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Evaluation of the Frequency of Vesicoureteral Reflux in Infants With Urolithiasis

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ABSTRACT

Background and Aim: Urolithiasis encompasses calculi formation in the urinary tract; if the calculi size is <3 mm, it is called microlithiasis. There is controversy about the role of urolithiasis in children as a predisposing factor for vesicoureteral reflux without significant hydronephrosis and urinary tract infection (UTI). In this study, we evaluated the frequency of vesicoureteral reflux in infants with urolithiasis but without some falsifying factors such as UTI and hydronephrosis.

Methods: In this cross-sectional study, we evaluated 144 infants with urolithiasis referred to the Nephrology Clinic of Urmia University of Medical Sciences. We recorded the results of voiding cystourethrography and sonography and demographic findings in a designed checklist.

Results: Our patients were 144 infants <1 year old, comprising 81 boys (56.2%) and 63 girls (43.8%); M/F=1.28:1 with a mean age of 5.09±2.44 months. About 84% of patients were <7 months. The mean number of stones in each patient was 7.42±4.72. Also, 110 infants (76%) had microlithiasis (stones <3 mm), and others (24%) had larger stones. Vesicoureteral reflux (VUR) was found in 66 infants (45.8%), consisting of 37 girls (56%) and 29 boys (44%) (P=0.006). In addition, 27 patients (41%) had unilateral reflux (16 girls and 11 boys), and 39 (59%) had bilateral VUR (21 girls and 18 boys) (P=0.021). The frequency of VUR in children under 7 months was 50% (n=61) and 22% in children older than 7 months (P=0.018). The frequency of VUR was 51.8% in infants with microlithiasis and 26% in infants with stones larger than 3 mm (P=0.001)

Conclusion: Regarding the partially high frequency of VUR (45.8%) in our patients with urolithiasis who had no history of UTI and hydronephrosis, it seems that urolithiasis has a predisposing role for vesicoureteral reflux in infants.

Keywords: Vesicoureteral reflux, Nephrolithiasis, Child

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Comorbid Psychiatric Disorders in an Adolescent With Cloacal Malformation: A Case Report

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ABSTRACT

Cloacal anomaly is a rare and complex multisystem anomaly in girls that results from the failure of the genitourinary septum to separate the cloacal membrane, leading to the opening of the vagina and the urethra and rectum in a common channel. Despite the available surgical treatments and survival to adulthood, its long-term consequences and psychological complications are still less investigated. We report a 15-year-old teenage girl with a history of cloacal anomaly. She had frequent hospitalizations since birth for multiple therapeutic surgeries and also due to frequent urinary tract infections (UTIs). She had undergone frequent clean intermittent catheterization (CIC) since she was about 5. The patient was hospitalized due to the worsening of behavioral problems and academic performance decline. Conducting additional interviews and questionnaires confirmed the presence of concurrent disorders of attention-deficit/hyperactivity disorder, oppositional defiant disorder, autism spectrum disorder, and obsessive-compulsive disorder, in addition to medical problems. The overprotective parenting style of the parents, along with the obsessive-compulsive disorder in the mother, had led to her deprivation from acquiring many self-care and communication skills and the aggravation of her behavioral problems and oppositional behaviors. When she entered her teenage years, many of her problems escalated. During hospitalization, the patient's symptoms were controlled with medication and non-pharmacological interventions (communication and problem-solving skills training, family therapy, parenting management training). In chronic congenital diseases, especially anorectal diseases, it is essential to take into account the possibility of simultaneous occurrence of psychiatric disorders and the need to investigate and treat these disorders, along with the need for these patients to receive more support from the family, school, and treatment team (surgeon, nephrologist, and psychiatrist). These supports and appropriate psychoeducation and parenting interventions should be provided to help these patients communicate better with their peers and society. They should have been taught relevant skills to prevent behavioral and mood problems.

Keywords: Psychiatric disorders, Cloacal anomaly, Adolescent

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Blood Pressure Criteria in Term and Preterm Neonates

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ABSTRACT

Background and Aim: In preceding decades, many techniques for measuring arterial blood pressure (BP) have been developed, and numerous tables of observed BP values have been published for neonates across gestational ages at birth and postnatal ages. Although BP data are widely available, attempts to describe normal BP for neonates, particularly preterm infants, are challenging. This is partly because of the various techniques used to measure BP, rapid changes in the physiology in the immediate postnatal period as the neonate adapts to the extra uterine environment, the presence of factors that can impact BP values in the neonatal period, and difficulties defining normal values in an inherently abnormal patient population, such as preterm infants in a neonatal intensive care unit.

Methods: Methods for measuring blood pressure may be invasive intra-arterial blood pressure or noninvasive oscillometric devices. However, significant differences have been reported in measuring BP values obtained by intra-arterial versus oscillometric methods.

Results: These differences may represent true differences in the BP value obtained or may be related to patient selection, device or algorithm used, or slight differences in the postnatal age at the study time. BP values outside the range of commonly observed values, such as those greater than the 90th percentile or less than the 10th percentile for commonly cited reference, may indicate underlying pathology.

Conclusion: Assessment of BP values should be considered regarding the entire clinical picture, including the perinatal history, the infant's size and age, and the physical examination.

Keywords: Blood pressure, Neonate, Nomogram, Normal values

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Hemodiafiltration Versus Hemodialysis in the Management of End-stage Renal Failure Children

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ABSTRACT

Background and Aim: Hemodiafiltration (HDF) has a convective component (hemofiltration) more than hemodialysis (HD), thereby providing more blood purification by adding a convective mass elimination of medium and large molecular-weight uremic toxins to diffusive mass transport of small uremic toxins by HD. This convective volume is replaced 1 for 1 mL with an intravenous (IV) infusion of substitution fluid (ultrapure dialysate) produced "online" from the dialysate by refiltration. Recently, many researchers have reported that HDF can significantly improve the outcomes of the management of end-stage renal failure (ESRD) patients.

Discussion: Many studies have shown that HDF as a renal replacement therapy is better for ESRD children than HD because of its superiority in removing medium and large uremic toxins that induce ESRD complications; some advantages are mentioned below.

Anemia: Children on HDF have a lower anemia rate, and their response to erythropoiesis-stimulating agents is prominent. This condition is due to the more efficient clearing of medium-sized proinflammatory molecules, such as hepcidin. It helps iron utilization as well.

Metabolic Bone Disease: Fibroblast growth factor 23, parathyroid hormone, osteocalcin, and osteoprotegerin are implicated in abnormal bone metabolism.

Inflammation and Oxidative Stress: HDF decreases β2-microglobulin, interleukin(IL)-6, IL-10, tumor necrosis factor-α, nitrotyrosine, high-sensitive C-reactive protein, asymmetric dimethyl arginine, symmetric dimethyl arginine, advanced glycation end-products, oxidized low-density lipoprotein, free chains of immunoglobulins, hepcidin and homocysteine. It also increases total antioxidant capacity compared to HD. Thus, HDF improves the endothelial risk profile.

Hypertension: Stable blood pressure is more prevalent in children on HDF than those on HD who frequently have significant and persistent hypertension despite an equivalent dialysis dose.

Cardiovascular Disease: HDF removes indoxyl sulfate and p-cresol sulfate, which mainly bind to albumin more efficiently. In addition, carotid intima-media thickness SD score rises significantly in children on HD but remains stable in the HDF cohort.

Anorexia: Adipokine, leptin, IL-6, tumor necrosis factor-α, and IL-1b reduce appetite in patients on HD, which are associated with lower albumin and pre-albumin levels and an inverse correlation with muscle mass and lower levels of physical tolerance.

Impaired Immune System and Infections: Free light chains of immunoglobulins, retinol-binding protein-4, fibroblast growth factor-23, α-1 glycoprotein, degranulation-inhibiting protein, and granulocyte inhibitory protein inhibit in vitro polymorphonuclear neutrophils-leukocyte chemotaxis.

Growth Retardation: Intensive HDF (6 sessions/week) promotes a positive effect on growth, even more than using growth hormone alone.

Hemodynamic Instability: HDF improves hemodynamic tolerance by decreasing episodes of symptomatic hypotension.

Amyloidosis: HDF results in a significant reduction in β2-microglobulin levels, compared to high-flux HD, in conjunction with the attenuation of the inflammatory milieu, contributing to a lower incidence of dialysis-related amyloidosis.

Neuropathy: HDF may prevent or slow down the progression of peripheral neuropathy. Evidence indicates nerve excitability stays nearly normal, and uremic pruritus and restless legs reduce significantly.

Conclusion: According to the results of the DOPPS (Dialysis Outcomes and Practice Patterns Study), patients with HDF who are treated with a high convection volume (>15 L) will benefit from significantly longer survival, confirmed in several clinical trials.

Keywords: Hemodiafiltration, Hemofiltration, Hemodialysis, End-stage renal failure (ESRD), Children

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Necessity of Chronic Kidney Disease Screening Program in Developing Countries

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ABSTRACT

Background and Aim: Consanguineous marriage may increase the prevalence of congenital anomalies of kidney and urinary tract and related chronic kidney disease (CKD). Iran is reported high in this kind of marriage. Therefore, the prevalence of CKD was assessed by biochemical and kidney ultrasound measurements in the first-grade pupils.

Methods: The study population of this cross-sectional study comprised all children aged 6 to 7 years. They were evaluated through urine analysis, serum creatinine, urine microalbumin to creatinine ratio, and kidney ultrasound.

Results: A total of 653 children participated in the study. Stage 1 and stage 2 systolic hypertension were detected in 6.5% and 1% of them, respectively. Also, stage 1 and stage 2 diastolic hypertension were found in 1.3% and 0.3% of them, respectively. In addition, weight and waist Z-scores are positively associated with systolic and diastolic blood pressure. Microalbuminuria (in 2.5%) was not correlated with hypertension, body mass index, microscopic hematuria, glomerular filtration rate, kidney sonographic abnormalities, kidney parenchymal thickness, or family history of kidney transplantation. About 1.8% of the students had a glomerular filtration rate of <90 mL/min/1.73 m^2, and only 1.7% had urine red blood cells of >5 in each high-power field. Also, 1.5% had an anatomical abnormality of the kidney and urinary tract (hydronephrosis or hydroureter).

Conclusion: Considering the higher rates of blood pressure and microalbuminuria in Iranian children, a CKD screening program based on evaluating microalbuminuria and blood pressure is mandatory. However, regardless of the high prevalence of consanguineous marriage in Iran, using kidney ultrasound as a screening tool is not recommended.

Keywords: Children, Chronic Kidney Disease, Screening, Microalbuminuria

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Efficacy of Vitamin E on Renal Function and Preventing Proximal Tubulopathy Caused by Iron Chelation Therapy in Thalassemia Major Patients

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ABSTRACT

Background and Aim: Proximal tubulopathy, due to deferasirox administration, contributes to acquired kidney failure. However, the preventive role of antioxidants in acute kidney injury has remained inconclusive. This study explored the efficacy of vitamin E in stopping acute kidney injury and proximal tubulopathy in thalassemia major patients under deferasirox (Nanojade) treatment.

Methods: This randomized controlled trial was conducted at the Thalassemia Center of Amirkabir Hospital in Arak City, Iran. A total of 60 patients with thalassemia major receiving Nanojade (20 mg/kg) were recruited for the study. The experimental group (n=30) received 400 IU of vitamin E per day for one month, while the control group (n=30) received no vitamin E. Acute kidney injury (AKI) was the primary outcome measure, defined by >50% increase in serum creatinine after 7 days of deferasirox administration. Also, the indicators of proximal tubulopathy were serum phosphate, venous blood gas, and urinalysis after one month. Finally, the glomerular filtration rate (GFR) and blood urea nitrogen to creatinine ratio (BUN/Cr) were compared between the two groups.

Results: Because AKI or proximal tubulopathy did not happen in either group, our investigation was limited to the preventive effect of vitamin E in these conditions. However, several notable findings emerged from our analysis. Regarding GFR and blood urea nitrogen to creatinine ratio, there was no significant difference between the experimental and control groups after one month (P=0.985 and P=0.063, respectively). The serum creatinine increase during the first week was significantly lower in the experimental group than in the control group (P=0.019). However, no difference was seen after one month (P=0.984). Mainly, the experimental group exhibited a significantly lower decrease in serum bicarbonate (HCO_3) and pH after one month (P=0.013 and P=0.003, respectively). Both groups had no significant differences regarding serum phosphate reduction (P=0.391).

Conclusion: One week's administration of vitamin E effectively prevented serum creatinine levels increase and deferasirox-induced pH and HCO_3 decrease in thalassemia patients. However, it does not affect the GFR.

Keywords: Thalassemia, Deferasirox, Proximal tubulopathy, Vitamin E

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A Novel Gene Variant in a 9-year-old Girl With Proteinuria and Loss of Consciousness, Later Diagnosed With Schimke Immunoosseous Dysplasia

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ABSTRACT

Schimke immunoosseous dysplasia (SIOD) is an ultra-rare autosomal recessive inherited disease. Clinical manifestations include growth retardation, skeletal dysplasia, progressive proteinuric glomerulopathy, episodic lymphopenia, and defective cellular immunity with different ranges of deterioration. We present a 9-year-old girl who was referred to our hospital with complaints of acute loss of consciousness and edema. Laboratory findings indicated urgent hemodialysis. She had a history of proteinuria and was later diagnosed with SIOD following other symptoms and immunological tests. We also found different gene mutations with variants of significant effect confirmed by next-generation sequencing.

Keywords: Focal segmental glomerulosclerosis, Schimke immunoosseous dysplasia, SMARCAL1, Mutational analysis

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Fundamental Attitudes and Paying Attention to the Differences and Clinical Manifestations in Children With Pheochromocytoma Compared to Adults With Pheochromocytoma, a Unique and Easy Solution for Early Diagnosis

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ABSTRACT

Pheochromocytoma is related to the involvement of the adrenal medulla. Due to the high production of catecholamines and excessive sympathetic stimulation, it manifests with hypertension in most cases. Although pheochromocytoma disease is known with the classic triad of headache, palpitations, and sweating, almost less than half of affected patients present with these characteristics. Meanwhile, one of the characteristics of this disease in children is the presence of continuous hypertension. In other words, unlike adult patients, where hypertension caused by this disease can be found sometimes and sometimes not, in children, we expect to have constant hypertension at any time. In this section, two patients with a very rare disorder are introduced. The first patient was an 8-year-old boy who was initially examined by different doctors with a complaint of headache, and even after being diagnosed with migraine for 6 months under the supervision of a neurologist, he was treated for migraine and stopped the treatment due to lack of proper response. Finally, with the care of a general practitioner, the family noticed high blood pressure in their son, and with the investigations done, he underwent surgery with the diagnosis of pheochromocytoma. The second patient was a 19-year-old boy who was referred due to high blood pressure. At the age of 11, this patient was diagnosed with hypertension and underwent surgery and adrenalectomy. For this patient, pheochromocytoma was diagnosed, and due to the recurrence of the disease, he was operated even once more. Finally, he was operated on for the third time and recovered with a bilateral adrenalectomy. Of course, he had to use prednisolone and fludrocortisone, as it became lifelong.

Keywords: Pheochromocytoma, Adrenal medulla, Hypertension, Adrenalectomy, Catecholamines

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The Effect of Magnesium Supplementation on Vascular Calcification and Cardiac Function in Pediatric Patients With Chronic Kidney Disease on Hemodialysis

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ABSTRACT

Background and Aim: The pathogenesis of cardiovascular disease (CVD) in patients on hemodialysis (HD) includes atherosclerosis, arteriosclerosis, and myocardial damage. The harmful effect of hypomagnesemia on cardiovascular mortality has been documented in several adult studies. The present study aimed to determine the impact of magnesium (Mg) supplementation on carotid intima medial thickness (CIMT) and cardiac function in pediatric HD patients.

Methods: A total of 22 pediatric HD patents were included in the study. Serum biochemistry markers, Doppler and tissue Doppler echocardiography, and CIMT were measured at baseline and after 6 months of every other day supplementation of Mg-citrate (6 mg/kg, maximum 300 mg).

Results: Our patients had both systolic and diastolic dysfunction compared to their healthy counterparts. About 40% of our pediatric HD patients had increased CIMT. Baseline and 6-month serum Mg levels were not correlated with CIMT and echocardiographic measures. Following 6 months of Mg supplementation, we observed a non-significant improvement in systolic and diastolic cardiac function measures ($P>0.05$) and a significant improvement in CIMT on both sides ($P=0.014$ on the right side and $P=0.001$ on the left side). Moreover, serum phosphate, calcium×phosphate product, parathyroid hormone, alkaline phosphatase, and uric acid decreased significantly after 6 months.

Conclusion: Mg supplementation may be protective in reversing vascular calcification in pediatric hemodialysis patients. Future investigations with a larger sample size and longer therapy duration may reveal more accurate results

Keywords: Magnesium, Children, Hemodialysis, Cardiac dysfunction, Carotid intima media thickness

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Comparing Urine Solute Composition in Nephrolithiasis According to Sex Differences

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ABSTRACT

Background and Aim: The prevalence of kidney stones is rising due to environmental factors. Men have always been at higher risk of developing stones; however, recent data suggest a changing pattern, with women being relatively more affected than before. This research aims to explore the prevalence of abnormal concentrations of urinary solutes as potential risk factors for kidney stones in Iranian children under the age of 14 according to sex.

Methods: Through renal sonography, a cross-sectional investigation was conducted on children diagnosed with nephrolithiasis or urolithiasis from May 2022 to May 2023. They were subsequently referred to the nephrology outpatient department of our hospital or private pediatric nephrology office. Laboratory assessments encompassed measuring urine calcium, oxalate, citrate, uric acid, creatinine, and urine cysteine. Demographic attributes, stone composition (when available), and urinary metabolic disorders were scrutinized. Data analysis involved descriptive statistics such as mean, standard deviation, and frequency. The chi-square tests were employed for inferential statistics. All statistical analysis was done using SPSS software, version 24, at a significance level of $P<0.05$.

Results: The study encompassed 486 children, with a mean age of 55.58 months and a male majority of 50.8%. Predominant urinary metabolic disorders included hypercalciuria (30.2%), hyperuricosuria (22.6%), and hyperoxaluria (20.6%). Metabolic abnormalities were more prevalent in males (63.3%) compared to females (53.1%), representing a significant difference ($P=0.020$). Gender-based analysis revealed a significant difference in hypercalcemia, with rates of 34.8% in boys and 25.5% in girls ($P=0.016$). No statistically significant differences were observed with regard to other underlying disorders.

Conclusion: Metabolic abnormalities were more prevalent in males with nephrocalcinosis difference and a significant difference in hypercalcemia, with rates of 34.8% in boys and 25.5% in girls ($P=0.016$).

Keywords: Nephrolithiasis, Urolithiasis, Nephrocalcinosis, Pediatrics, Metabolic disorders, Urinary stone

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Gitelman Syndrome Concurrent AD Congenital Myopathy 1A

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ABSTRACT

Gitelman syndrome is a rather rare renal tubular disorder. Although it is a recessively inherited disorder, sporadic cases are also seen. Customarily, Gitelman syndrome creates a benign or mild tubulopathy. We present a 4-year-old male patient with hypokalemia and congenital myopathy 1A. He reported 3 episodes of generalized weakness along with sudden drops and falling. The patient was admitted to our hospital for the first time in January 2019. His electrocardiogram on admission showed evidence of prolonged QTc interval (0.46 s), resolved after treating with potassium. Subsequent laboratory investigation revealed renal wasting hypokalemia, mild hypomagnesemia, and hypercalciuria, indicating that the patient might have had a renal tubular disorder. A genetic study that revealed SLC12A3 confirmed Gitelman syndrome. With the administration of spironolactone, potassium chloride, and magnesium supplement, the serum potassium and magnesium levels were maintained within normal ranges. We are presenting our case, seeing that the patient was shown a triangular face with a hypokalemic episode at an early age (2 years). The face of the patient made it confused with Bartter syndrome. We identified compound mutations of SLC12A3 (chr16) associated with AD congenital myopathy 1A (chr19). Further efforts are needed to investigate the diversity in clinical manifestations of Gitelman syndrome and its correlation with this specific mutation. However, the patient was susceptible to malignant hyperthermia characterized by muscle weakness primarily affecting the proximal muscles of the limb. Additional features include mild facial weakness. The plan for the patient is a muscle biopsy that will reveal areas of lack of mitochondrial activity in type 1 muscle fiber.

Keywords: Gitelman syndrome, Congenital myopathy, Hypokalemia

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Cumulative Doses of Corticosteroid and Bone Mineral Density in Children With Nephrotic Syndrome

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ABSTRACT

Background and Aim: The present study aimed to determine the association of the different cumulative doses of corticosteroids with bone mineral density in children with nephrotic syndrome.

Methods: This cross-sectional study was performed on children who suffered from a nephrotic syndrome referred to Children's Medical Center in Tehran City, Iran, in 2016. The subjects were categorized into three groups based on the cumulative corticosteroid dose (received <200 mg/kg, 200-400 mg/kg, and >400 mg/kg of corticosteroid). In all groups, bone densitometry was employed to determine bone mineral density (BMD), bone mineral concentration, and Z score of the lumbar spine and neck of the femur.

Results: Fifty-seven patients (19 in each arm) were included in the study. A cumulative dose of corticosteroids and lumbar spine and neck of femur BMD and lumbar spine BMC were found to be moderately and negatively correlated (τ_{57}=-0.31, P=0.003; τ_{57}=-0.293, P=0.005; and τ_{57}=-0.234, P=0.025, respectively). Increases in cumulative dose of corticosteroids were strongly correlated with a decrease in BMD of the lumbar spine and femoral neck (τ_{56}=-0.61, P<0.001; and τ_{57}=-0.490, P<0.001, respectively).

Conclusion: High cumulative doses of corticosteroids can significantly lower bone mineral density in children who suffer from nephrotic syndrome and need to be prevented.

Keywords: Bone density, Nephrotic syndrome, Child

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Prevalence of Abnormality in Number, Position, and Location of Kidneys in Pediatric Patients Referred to Nephrology Clinic of Dr Sheikh Children Hospital: A 17-year Evaluation

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ABSTRACT

Background and Aim: Congenital anomalies of the kidneys and urinary tract account for approximately 20% to 30% of all abnormalities detected in the prenatal period. Diagnosing this group of abnormalities is crucial as they are associated with other urological abnormalities, especially vesicoureteral reflux. Perinatal ultrasounds, performed routinely in recent decades, have made possible early detection of abnormalities. This 17-year study aimed to determine the prevalence of this abnormality regarding the number, position, and location of kidneys in symptomatic pediatric patients referred to the Nephrology Clinic of Dr Sheikh Children's Hospital in Mashhad City, Iran.

Methods: In this cross-sectional study, patients under 18 referred to the Nephrology Clinic of this Hospital for various reasons over 17 years (2003-2009) underwent ultrasonography of the kidneys and urinary system, and a clinical record was prepared for them. Based on the designed questionnaire, information about paraclinical examinations and tests was extracted from the records of these patients, and statistical analyses were performed using SPSS software.

Results: A total of 92 eligible patients with a mean age of 53.44±45.12 months entered the study. Of them, 49 (53.26%) were boys. The most common urinary tract abnormalities found in ultrasound of these patients were renal agenesis (n=41, 44.56%), ectopic kidney (n=26, 28.26%), and horseshoe kidney (n=25, 27.17%). The most common findings in the clinical examination of these patients included urinary tract infection (n=17, 18.5%), abdominal pain (n=15, 16.3%), neonatal colic (n=8, 8.7%), and anorexia (n=6, 6.5%). In 60 available voiding cystourethrogram cases, 20 (33.3%) were reported as abnormal, of which 15 patients (75%) had vesicoureteral reflux (VUR). It was also found that there is no statistically significant relationship between gender, presence or absence of kidney stones, vesicoureteral reflux, and urinary tract infection (UTI) with the occurrence of the mentioned anomalies of the genitourinary system (P=0.58, P=0.246, P=0.728, and P=0.648, respectively).

Conclusion: The present study showed that renal agenesis (RA), ectopic kidney, and horseshoe kidney were the most common renal and urinary system abnormalities in symptomatic children who participated in this study. Also, in this study, gender, presence of kidney stones, UTI, and VUR were not significantly associated with the occurrence of three main anomalies in this design (RA, ectopic kidney, and horseshoe). The most common disorders of the genitourinary system associated with renal agenesis include VUR, undescended testis, and UTI. In combination with horseshoe kidneys, the common abnormalities include VUR, UTI, and kidney stones. In association with the ectopic kidney, they included hydronephrosis, VUR, and kidney stones.

Keywords: CAKUT, Child, Vesicoureteral reflux

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The Effect of L-carnitine on Improving Cardiac Function in Pediatric Patients Under Hemodialysis

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ABSTRACT

Background and Aim: Chronic renal failure is a critical urinary system dysfunction among children, and its diagnosis and treatment are essential to improve the conditions and its prognosis. Cardiac function is a critical disordered item in these children. Therefore, this study assessed the effect of L-carnitine on improving cardiac function in hemodialysis patients.

Methods: In this before-and-after clinical trial, 13 consecutive hemodialysis children in Bahrami Pediatric Hospital in 2020 were enrolled. The plasma carnitine level and cardiac function were initially assessed; L-carnitine 40 mg/g per day was administered for three months, the plasma level was evaluated after one month, and echocardiography was taken after three months.

Results: The mean left ventricular internal diameter end diastole (P=0.054), left ventricular internal diameter end-systole (P=0.796), posterior wall thickness at end-diastole (P=0.690), septal wall thickness during diastole (P=0.715), fractional shortening (P=0.162), ejection fraction (P=0.596), peak early filling velocity (E)/peak atrial filling velocity (A) (P=0.358), left atrium/ peak atrial filling velocity (P=0.245), isovolumic relaxation time (P=0.643), left ventricular mass (P=0.060) were not differed. Hemoglobin (P=0.008), hematocrit (P=0.027), serum iron (P=0.039), and carnitine (P=0.039) were significantly increased after treatment.

Conclusion: According to the results, L-carnitine has no significant effect on hemodialysis children's cardiac function, and its use is not recommended.

Keywords: L-carnitine, Cardiac function, Hemodialysis, Children

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Vaccination of Pediatric Candidates for Kidney Transplant in Iran

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ABSTRACT

Pediatric organ transplantation, especially kidney, has progressed dramatically in Iran in recent decades. In this regard, pre-transplant vaccination is an effective preventive tool because infections are the most common cause of morbidity and mortality among transplanted children. In addition, administering some vaccines is contraindicated in the post-transplant period, and authorized vaccines' efficacy and immunogenicity may become suboptimal compared to the average population. Therefore, the pre-transplant period offers an outstanding chance to increase the immunization of this population. In this population, customized vaccination guidelines must be prepared for nephrologists and other transplant team clinicians in Iran. This study aims to provide a comprehensive overview of the vaccines recommended for these cases regarding the expanded program on immunization and available vaccines in Iran. In addition, general principles of vaccination, the use of specific vaccines, and accelerated vaccination in this population are discussed in this article.

Keywords: Vaccination, Child, Kidney transplantation

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Evaluation of Hearing Impairments in Pediatric Patients With Chronic Kidney Disease

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ABSTRACT

Background and Aim: Hearing impairments are prevalent in patients with chronic kidney disease (CKD). Although the pathophysiology of hearing loss in patients with CKD is still unclear, some theories include the side effects of ototoxic drugs, elevated urea, electrolyte disturbances, edema, anemia, and changes in blood pressure. Our objective was to determine the prevalence of hearing impairments in children and adolescents with CKD.

Methods: In this prospective descriptive study, we assessed the hearing of 20 children and adolescents with CKD. Eighteen underwent pure tone audiometry tests, while two underwent auditory brainstem response tests. Patients' demographics, including height, weight, and laboratory results were recorded.

Results: The study included 12 male and 8 female children and adolescents with CKD, with a mean age of 8.7±4.43 years old. Of the 20 participants, 12 were on dialysis (6 peritoneal, 6 hemodialysis), and 8 had a glomerular filtration rate above 15, receiving supportive therapies. The causes of CKD were diverse: renal hypoplasia (40%), reflux nephropathy (25%), obstructive uropathy (20%), nephrotic syndrome (10%), and polycystic kidney disease (5%). In this study, 20% of patients exhibited hearing impairments, with 15% (n=3) having conductive hearing loss and 5% (n=1) experiencing sensorineural hearing loss. All participants were on CKD-related medications (calcium carbonate, sevelamer, ferrous sulfate, calcitriol). Hemodialysis patients were periodically given nephrotoxic drugs, including vancomycin, due to PermCath infections, while peritoneal dialysis patients intermittently received nephrotoxic medications due to peritonitis. Moreover, individuals with reflux nephropathy and obstructive uropathy, facing recurrent urinary infections, also received nephrotoxic drugs like aminoglycosides.

Conclusion: This study revealed a 20% prevalence of hearing impairments in pediatric CKD patients, with 15% having conductive hearing loss and 5% experiencing sensorineural hearing loss. Regular screening for hearing status is recommended for all children with CKD.

Keywords: Pediatrics, Chronic kidney disease, Hearing loss, Hemodialysis

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Haplotype Analysis and Sanger Sequencing in Patients Suspected of Type 3 Hyperoxaluria

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ABSTRACT

Background and Aim: Primary hyperoxaluria (PH) is an autosomal recessive hereditary condition causing excess oxalate production. There are three forms of PH; the most common is PH type 1, caused by a mutation in the *AGXT* gene. PH type 2 is brought on by a deficiency of the enzyme glyoxylate reductase/hydroxy pyruvate reductase (GRHPR). In contrast, type 3 of the disorder is brought on by mutations in the *HOGA1* gene that induce malfunctions or impairment. HOGA1 is sometimes known as dihydrodipicolinate synthase-like. This protein forms a tetrameric structure containing 327 amino acids, 17 helices, 8 B strands, 3 twists, and 2 dimers. Various techniques, like haplotype analysis and single-nucleotide polymorphisms (SNPs), can be used to determine the causative mutation. Haplotype analysis uses that patients were born in consanguineous marriages or from a small geographic area that probably inherited two recessive copies of a mutant allele from a common ancestor.

Methods: Fourteen individuals from 11 unrelated families suspected of having PH type 3 were referred to the Ali Asghar Children's Hospital. SNPs were used for haplotype analysis. The *HOGA1* gene is surrounded by four separate SNPs, namely rs2275090, rs1124116, rs3750614, and rs2296438. The SNPs have been genotyped using amplification refractory mutation system PCR. Affected individuals with identical haplotypes underwent Sanger sequencing of the *HOGA1* gene. One patient undergoing investigation who did not exhibit a homozygous haplotype was referred for next-generation sequencing.

Results: Only 2 patients had the homozygous haplotypes. Sanger sequencing was done in all coding exons of the *HOGA1* gene for both families, and one of them was identified to have a unique homozygous mutation of c.266G>A p.(Arg89His). It was fond in the exon 2 of the *HOGA1* gene. Her parents were heterozygous for the identified mutation. Another patient had a homozygous haplotype. After checking and analyzing all exons of the target gene, no mutation was found in those exons. Following next-generation sequencing analysis in the other patient, no pathogenic or likely pathogenic variants were discovered. Additional investigation revealed the ROH (run of homozygosity) regions. The only mutation that can cause this disease is the *GRHPR* gene, which may have occurred in the non-exonic region of the *GRHPR* gene. It is better to perform a whole genome sequence for this case.

Conclusion: A new missense mutation of c.266G>A p.(Arg89His) in the homozygote family was confirmed. Moreover, the mutant residue is smaller than the wild-type residue, and its charge is neutral, whereas the wild-type residue was charged positively. The size difference between the wild-type and mutant residue means that the new residue is not in the correct position to form the same hydrogen bond as the original wild-type residue, according to

HOPE (The HOPE server is an internet-based resource used for protein prediction). Each amino acid in this change has its specific size, charge, and hydrophobicity-value. This variant is a VUS (variant of uncertain significance) one based on the ACMG (the American College of Genetics and Genomics) guideline. One novel mutation in the *HOGA1* gene was identified, including c266G>A. More sample size is necessary to extend the mutation spectrum in Iran

Keywords: Haplotype analysis, Sanger sequencing, Type 3 hyperoxaluria

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A 14-year-old Boy With Post-transplant Lymphoproliferative Disorder: A Case Report

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ABSTRACT

One of the most severe and potentially deadly complications of solid organ and hematopoietic stem cell transplantation is post-transplant lymphoproliferative disorder (PTLD). Most cases of PTLD are linked with Epstein-Barr virus (EBV). After transplantation, the T cell number is decreased, resulting in uncontrolled proliferation of EBV-positive B cells. EBV turns positive in 60%-80% of PTLD cases. It is usually associated with early-onset PTLD. On the other hand, EBV-negative PTLD is mainly late onset. A 14-year-old male patient who underwent renal transplantation because of polycystic kidney disease 6 years ago was admitted to the hospital with abdominal pain and generalized lymphadenopathy. Physical examination revealed splenomegaly and a non-significant lymph node in the right axilla. He had been on mycophenolate mofetil, tacrolimus, and prednisolone since then. Ultrasound confirmed porta hepatis lymph node and portal hypertension. Moreover, EBV and CMV virus polymerase chain reaction were negative. He had undergone a biopsy of the axillary lymph node, which appeared to be reactive lymphadenopathy. Finally, the biopsies of abdominal lymph nodes revealed Burkitt lymphoma. The patient was treated with a reduction of the immunosuppressive drugs and received 2 cycles of chemotherapy. He died in the second course of chemotherapy due to complications of renal failure.

Keywords: Kidney transplantation, Children, Post-transplant lymphoproliferative disorder (PTLD)

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A Challenging Case of Nephrotic Range Proteinuria

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ABSTRACT

A 2-year-old girl was presented with periorbital edema and nephrotic-range proteinuria for the past 3 months. She was born to consanguineous parents and had a brother who passed away on peritoneal dialysis. The brother had cherry red spots and splenomegaly, and the necropsy showed foam cells. Investigation of metabolic disorders yielded inconclusive results. The patient had a mix of tubular and glomerular proteinuria. Her bone marrow had many small vacuoles in peripheral lymphocytes and foamy macrophages. A kidney biopsy was performed, and foamy cells aggregated in glomeruli, tubules, and interstitium. The genetic study was normal, but the second analysis confirmed sialidosis type 2. Our goal is to raise awareness about lysosomal storage disorders among clinicians and to diagnose this disease earlier, through the patient's clinical findings and attention to peripheral blood smear, to achieve the diagnosis.

Keywords: Sialidosis, Nephrotic syndrome, Vacuolated lymphocyte

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The Effect of Phototherapy in Infants With Non-physiological Jaundice in the Occurrence of Urinary Stones

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ABSTRACT

Background and Aim: Jaundice is one of the common diseases of infancy, and in severe cases, the first treatment is to use phototherapy. This study investigates the effect of phototherapy in increasing urinary excretion of calcium and the occurrence of kidney stones.

Methods: This research was a before-and-after study. A total of 100 infants aged <2 weeks with normal birth weight who were hospitalized due to non-physiological jaundice were included in the study. Urine samples were collected from infants at the beginning of hospitalization, and 48 hours after phototherapy, random urine Ca and Cr were measured. Then, the Ca/Cr ratio was calculated. The obtained data were entered into SPSS software, version 19, and subjected to statistical analysis.

Results: The Mean±SD ratio of Ca/Cr before and after phototherapy was 0.48±0.02 and 0.75±0.3, respectively. There was a significant relationship between the mean Ca/Cr ratio of urine in infants with jaundice before and after phototherapy (P<0.001). Hypercalciuria (Ca/Cr >0.8) was observed in 45.4% of infants.

Conclusion: The above findings indicate an increased urinary calcium excretion during phototherapy. Regarding the role of urinary calcium in urinary stones, phototherapy can be a risk factor for hypercalciuria and urinary stones.

Keywords: Phototherapy, Infants, Hypercalciuria, Urinary stone

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The Amount of KIM-1 Biomarker in the Urine of Children With Diabetes

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ABSTRACT

Background and Aim: KIM-1 is a membrane protein at the tip of proximal tubule cells. This biomarker is present in small amounts in the urine of healthy people, while its amount in urine increases in proximal tubule damage.This study aimed to investigate the amount of KIM-1 in the urine of children with diabetes.

Methods: This descriptive-analytical study employed a cross-sectional design. The study population comprised all children under 18 years old with diabetes living in Sanandaj City, Iran in 2019. In this study, children with diabetes who had been at least 3 years since the onset of their disease were included, and their morning urine samples were taken. After measuring KIM-1 in urine using the ELISA method, the results were subjected to statistical analysis using the chi-square, t-test, and Pearson correlation in SPSS software, version 22. A significance level of <0.05 was considered for all tests.

Results: A total of 40 children were included in this study. About 58% were girls, and 42% were boys; their mean age was 12.2 years. The mean HbA1c was around 7.95%. Also, 32.5% of patients had positive proteinuria.

KIM-1 was not related to the gender of patients (P>0.05), but it was significantly related to proteinuria. In people with positive proteinuria, the mean of KIM-1 was higher (P<0.05).

Conclusion: KIM-1 has a significant positive relationship with proteinuria in diabetes and can be used as a new predictor for early detection of kidney damage in children with diabetes.

Keywords: KIM-1 protein, Proteinuria, Diabetes

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Asthma as a Common Pulmonary Manifestation in Antineutrophil Cytoplasmic Antibody Associated Vasculitis

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ABSTRACT

Background and Aim: The antineutrophil cytoplasmic antibody (ANCA) associated vasculitis (AAV) has significant morbidity and mortality. Lung involvement is one of the most common clinical features. This study reviews the lung imaging and clinical history.

Methods: This is a case series study consisting of 10 patients by ICD-10 admitted as having AAV between 2013 to 2023. The participants' ages ranged from 2 to 16 years (mean 9.4 y). The patients' follow-up dates, first creatinine and the last proteinuria, chest X-ray (CXR) reports, pathology, and treatment were reviewed. Eight patients were male, and 2 were female. Hyperinflation (as a sign of hyperreactive airway disease) was defined by >14 anterior and posterior visible ribs. CXRs were reported by the radiologist.

Results: Follow-up dates ranged from 6 to 99 months. The antineutrophil cytoplasmic antibody (c-ANCA) was positive in 9 cases. All patients received corticosteroids. Five patients (50%) received CellCept, and 3 (33.3%) received plasma phrases. Of the 10 patients included, all had a history of cough, and 9 (90%) had hyperinflation in CXR. Also, 3 patients (33.3%) had pulmonary hemorrhage. One patient was incubated due to a respiratory problem.

Conclusion: AAV can present with a wide range of pulmonary manifestations, including nodules, interstitial lung disease, pleural effusion, asthma, and pulmonary embolism. Asthma was the most common pulmonary involvement in our series.

Keywords: Vasculitis, Asthma, Hyperinflation

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Rituximab: What Do We Expect From Difficult-to-treat Nephrotic Syndrome?

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ABSTRACT

Background and Aim: Rituximab (RTX), as a chimeric CD20 blocker, is widely prescribed to treat idiopathic nephrotic syndrome (INS). However, it is not so effective in treating steroid-resistant nephrotic syndrome (SRNS). In this study, we evaluated the response to RTX in INS and explored the predicting factors in relapse-free time and outcome.

Methods: The study sample consisted of 38 patients under 22 with INS. The study was carried out in Isfahan City, Iran, in 2014. The primary endpoint was a relapse-free period for 1 year or achieving a glomerular filtration rate (GFR) of <70 mL/min. RTX protocol was 375 mg/m^2 /dose/4 times. Biochemical and urinary parameters and immunoglobulin G (IgG), CD19, and CD20 levels were assessed before the first and after the last dose of RTX.

Results: The mean age of the participants was 12.86±4.5 years. Histopathology of focal and segmental glomerular sclerosis was predominant. A 1-year follow-up demonstrated that 42% (n=16) of patients were still in remission, and 34% (n=13) were in partial remission. However, 24% (n=9) ended up with chronic renal failure even after 4 doses of RTX. The absolute CD20 levels at the time of the first course and after stopping RTX predicted the final 24-h urine protein and the GFR (P>0.05). However, GFR (P<0.05, R^2=0.56) and age of diagnosis (P<0.05, R^2=0.591) were the predictors of final 24-h urine protein (P<0.05). In addition, response to steroids (SDNS, FRNS) had a positive effect on final GFR (P<0.05).

Conclusion: RTX proves to be an effective monoclonal antibody in keeping INS patients in remission. Although its efficacy in CNI-unresponsive SRNS is not equal to SDNS, RTX may keep most INS cases in complete or partial remission.

Keywords: Nephrotic syndrome, Rituximab, Children, Recurrence

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Febrile Proteinuria in Children: An Evaluation of Causes and Findings

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ABSTRACT

Background and Aim: High fever can cause transient proteinuria, which does not involve underlying kidney disease. The present study explored the prevalence of transient proteinuria in children with febrile diseases.

Methods: A total of 168 febrile children were chosen according to the enumeration method and studied for one year between August 2017 and 2018. The patients with renal diseases were excluded. The urine analysis was performed at admission and after the fever improvement. C-reactive protein, erythrocyte sedimentation rate, and white blood cells were evaluated using the patients' blood samples.

Results: Proteinuria was detected in 21 patients (11.4%). Among them, 8 patients (38%) were male, and 13 (62%) were female, with an mean age of 1.87±1.94 years. No significant differences were found in patients with or without proteinuria (P>0.05) regarding their sex and age. The concentration of proteinuria at the time of admission was a trace in 4 patients (19%), one plus (+) in 12 patients (57%), two plus (++) in 4 patients (19%), and three plus (+++) in 1 patient (5%). However, proteinuria was cleared in all patients after fever improvement. Febrile convulsion and gastroenteritis were the common diagnosis. The mean values of fever, urine specific gravity, erythrocyte sedimentation rate, and white blood cells in children with proteinuria were 38.78±0.560 cell/mm^3, 1017.58±8.58, 45.05±10.92 mm/h, and 13247.05±6501.06 cell/mm^3, respectively. CRP was positive in 16 (76%) and 73 patients (44.24%) with and without proteinuria, respectively; the difference was significant (P=0.01).

Conclusion: The prevalence of proteinuria in febrile children was 11.4%. Febrile children with proteinuria most commonly suffer from gastroenteritis and febrile convulsion.

Keywords: Child, Fever, Proteinuria

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Kidney and Urinary Tract Problems in Pediatric Down Syndrome

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ABSTRACT

Background and Aim: Kidney and urinary tract diseases are not common complications in children with Down syndrome (DS). However, various urological abnormalities and glomerulopathies have been described in these children, and some develop chronic kidney disease (CKD).

Objective: This study intended to enrich our understanding of renal diseases in children with DS by estimating their incidence and range of kidney and urological abnormalities.

Methods: A cross-sectional study was conducted on diagnosed DS patients referred for kidney evaluation. Medical records were reviewed, and physical examinations, including blood pressure measurements, were done for all of them. Kidney and urinary tract ultrasound were performed on all children after urinalysis and urine culture. Laboratory data, including plasma urea, creatinine, uric acid, and nuclear medicine scans, were performed for any possible renal disorder.

Results: A total of 110 patients, aged 8 months to 20 years, were recruited (59 females). Pathological findings included only one case of hypertension in a 10-year-old girl. Ten patients (11%) had urinary tract infections without any significant symptoms. Urinalysis in 15 patients (13.6%) had hyperuricosuria, and 10 had hyperuricemia without clinical gout. Furthermore, 3 cases had mild proteinuria, and 2 patients showed microscopic hematuria. Twenty-seven patients (24.5%) had abnormal sonography (small kidney in 5, hydronephrosis in 10, and thick bladder wall in 12). DMSA (dimercaptosuccinic acid) scans were done in 27 children, and the renal scar was reported in 3 and kidney dysplastic in 5. Two patients (1.8%) had CKD based on creatinine measurement (estimated glomerular filtration rate <75 mL/min/1.73 m^2).

Conclusion: Renal diseases in patients with Down syndrome are more than we used to think. According to this study and pathologies found, it is necessary to regularly monitor renal function in these patients to detect irreversible renal injury early.

Keywords: Down syndrome, Trisomy 21, Kidney anomalies, Kidney size, Congenital anomalies of the kidneys and urinary tracts (CAKUT)

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Research and Review on Kidney Transplant Laws in Iran

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ABSTRACT

Background and Aim: With the progress of science, organ transplantation has entered society as one of the new achievements of medical sciences. From the medicine perspective, organ transplantation refers to transferring cells, tissues, or organs so that their function is preserved. Organ transplantation, like all new areas of science, besides its advantages, accompanies legal and ethical challenges for the people of society. These challenges affect patients, medical staff, and donors in areas such as the individual's authority to donate, transplant from a corpse, brain death, and obtaining consent from the organ donor. It also causes financial relations, foreign nationals, etc. In this regard, the legislator has established laws and regulations for mutual protection of the rights of the members of the society and unity of action and citizenship from the single model.

Methods: This article reviews existing laws and regulations in Iran's organ transplantation field. To clarify the religious and legal aspects of organ transplantation in Iran, the Supreme Council was formed, the Organ Transplantation and Brain Death Act was approved in the Islamic Parliament in 1999, and the Ministry of Health prepared the executive order. We will mention the legal sources of organ transplantation in Iran, especially the constitution, the Islamic Penal Code, health and medical laws and regulations, etc

Conclusion: Important legal points in kidney transplantation include the informed consent of the transplant donor during his lifetime, having an organ donation card or obtaining permission from the recipient's relatives within the framework of the laws, death confirmation, and legal challenges related to it, respecting the organ donors' health and his sufficient knowledge about complications and lack of financial abuse and respect for the deceased, donor conditions, foreign nationals, violations in this field, how to deal with these violations, organ harvesting from children, free people, vulnerable groups, risks of surgery and other cases in this research are checked.

Keywords: Kidney transplantation, Legal standards, Legal resources, Iran

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Order Disruption and Adherence Failure in Children and Adolescents With Nephrotic Syndrome

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ABSTRACT

Background and Aim: A large number of children and adolescents with Nephrotic syndrome experience difficulties after catching this syndrome and its consequences. They feel psychological distress and show difficulties in adherence.

Methods: This research conducted a grounded theory study on how these people face the syndrome and its consequences. The research sample comprised children and adolescents with nephritic syndrome who were hospitalized at Ali-Asghar Hospital in Iran. Of them, 15 patients (and also their mothers) were selected using purposive sampling and reaching data saturation. Unstructured interviews were conducted with patients and their mothers. After transcribed interviews, they were analyzed in three stages: open, axial, and selective coding.

Results: The present qualitative study generated a model of adherence difficulties, with a main concept called "order disruption", which causes problems in adherence in children or adolescents with nephrotic syndrome. Order disruption is understandable based on problems in the individual, family, and health team levels of functional order. These disruptions occur following illness symptoms (such as distress from diagnosis, physical symptoms, functional limitations, and treatment outcomes).

Conclusion: To understand and improve the non-adherence of children or adolescents with nephrotic syndrome, it is necessary to pay attention to the disruption of usual order at different levels from micro to macro.

Keywords: Nephrotic syndrome, Adherence, Order disruption, Children, Adolescents

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Comparing Early Surgical and Conservative Therapy in Children With Ureteropelvic Junction Obstruction

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ABSTRACT

Background and Aim: Ureteropelvic junction obstruction (UPJO), a urologic problem in children, usually appears with pelvicalyceal dilatation in antenatal. Most UPJO cases are handled using surgical methods. However, many patients have recently been treated by nonsurgical observational plans. This study compared the outcomes of children with UPJO who were treated with surgical and observational methods.

Methods: In this retrospective study, the medical history of UPJO patients was investigated from March 2011 to March 2021. The cases were confirmed as having grade 3-4 hydronephrosis and obstructive pattern in dynamic renal isotopes scan. Patients were assigned to two groups: group 1, treated with a surgical procedure, and group 2, without any surgical procedure, for at least six months after diagnosis. We evaluated long-term outcomes and improvement of obstruction.

Results: A total of 78 children (mean age 7.32 mo., 80% male) were recruited in the study (55 patients in group 1 and 23 in group 2). Severe hydronephrosis was the problem of 96% of all patients, which decreased significantly to 20% in group 1 and 9% in group 2 (P<0.001). Severe kidney involvement was detected in 91% of patients in group 1 and 83% in group 2. These numbers decreased to 15% and 6%, respectively (P<0.001). No significant differences were observed between the two intervention groups regarding sonographic and functional improvement. Long-term prognostic issues, growth, functional impairment, and hypertension were comparable between the two groups. However, group 1 experienced more recurrence of UTI than group 2.

Conclusion: Conservative management and early surgical treatment are both effective in the management of infants with severe UPJO.

Keywords: Hydronephrosis, Ureteropelvic junction obstruction (UPJO), Child

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A Rare Cause for Atypical Hemolytic Uremic Syndrome

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ABSTRACT

A 15-year-old boy with symptoms of uremia, oliguria, severe anemia, and thrombocytopenia was referred to Abouzar Children's Medical Center in Ahvaz City, Iran. He was admitted to the nephrology intensive care unit. The initial diagnosis was hemolytic uremic syndrome (HUS), and diagnostic and therapeutic investigations were started. Hemodialysis was started on the third day of hospitalization due to continued oliguria and then anuria. He underwent daily hemodialysis for several days and simultaneously received frozen fresh plasma (FFP) and several blood transfusions due to repeated hemoglobin drops. After about three weeks, anuria and uremia resolved, and hemodialysis was stopped, but the pancytopenia continued, especially the severe leukopenia that had started gradually. Several hematology consultations and two bone marrow aspiration were performed during hospitalization, which were negative for malignancy.Many other examinations, such as ultrasound, CT, and bone scans, showed no abnormalities. Fortunately, we finally managed to identify the possible cause of the patient's multiple renal and non-renal manifestations.

Keywords: Child, Case Report, Hemolytic uremic syndrome

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Vitamin D Deficiency and Kidney Scarring in Children With Febrile Urinary Tract Infections: A Correlational Study

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ABSTRACT

Background and Aim: Urinary tract infection (UTI) is a significant issue in childhood because of its high prevalence and potential long-term complications. Several studies have suggested that vitamin D deficiency is an influential factor in the progression of infection. Our study aimed to investigate the correlation between serum vitamin D levels and the risk of renal scarring in children with febrile UTIs.

Methods: This cohort study was performed on 50 children with febrile urinary tract infections referred to 22 Bahman Hospital in Mashhad City, Iran, from 2020 to 2021. All patients were diagnosed with urinary tract infection (UTI) and evaluated for the levels of 25-hydroxy vitamin D, creatinine, blood urea nitrogen (BUN), calcium, phosphorus, and parathyroid hormones in serum samples. After six months of treatment, all patients were assessed for renal scarring using radioisotope scans.

Results: A total of 50 children with a mean age of 3.56±2.13 years (28% male) participated in this study. About 56% of the children had vitamin D deficiency. Levels of 25-hydroxy vitamin D were significantly associated with kidney scarring (P=0.0001). Moreover, the incidence of renal scarring did not correlate with age or sex (P>0.05). The receiver operating characteristic curve for the serum level of vitamin D revealed an area under the ROC curve of 0.856, suggesting that it is helpful in predicting renal abnormality on the DMSA (dimercaptosuccinic acid) scan. A serum vitamin D level of <30 ng/mL was the best predictor for post-UTI renal Scar (Positive LR 4.246, Youden's j index 0.676).

Conclusion: Vitamin D deficiency is prevalent in children, and there is a significant association between vitamin D levels and kidney scarring. Based on this study, there was a reverse correlation between vitamin D deficiency and the incidence of kidney scars caused by urinary tract infections in children. Therefore, we suggest screening to identify high-risk groups and to prescribe vitamin D supplements for susceptible children.

Keywords: Vitamin D, Urinary tract infection, Pyelonephritis, Scar, Immune system

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Nutritional Management in Pediatric Kidney Diseases

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ABSTRACT

Since children with kidney diseases have unique nutritional needs related to decreased kidney function, nutrition treatment of pediatrics is a big challenge, certainly for infants and young children. Previous studies show that protein-energy wasting and malnutrition are strongly associated with chronic kidney diseases. Abnormalities related to metabolism, fluids, and electrolyte balance increase the risk of morbidity and mortality by impairing growth, neurocognitive development, heart malformations, and bone damage. So sufficient growth and individual nutrient needs based on primary renal disease are all challenges for pediatric patients with kidney diseases. It seems that alternative formulas, such as complete formulas that meet this population's dietary requirements, are needed. Limited commercially available options exist for children and adolescents who require formula supplementation. Renastart is a powdered formula containing whey protein, carbohydrates, fat, vitamins, and minerals. It is restricted in protein, potassium, chloride, and phosphorus to be used in the dietary management of pediatric kidney disease from 1 year of age. Restricted to essential nutrients and electrolytes, it can help adjust the feeding regimen to match biochemical results. Some studies have illustrated that nutrition management with Renastart improves weight and linear growth and normalizes electrolyte serum levels.

Keywords: Pediatrics, Nutrition, Renastart, Kidney diseases

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Renal Replacement Therapies for Children With Acute Kidney Injury and Heart Failure

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ABSTRACT

Acute kidney injury (AKI) in children coupled with higher morbidity and mortality in critically ill patients. Continuous renal replacement therapy (CRRT) may be suggested in small children who cannot tolerate hemodialysis because of life-threatening hyperkalemia, acidosis, intoxications/ingestions, hyperammonemia/inborn errors of metabolism, severe symptomatic uremia, oliguria/anuria, and volume overload associated with AKI in children with low blood pressure or cardiac heart failure. After cardiac surgery, patients may require CRRT compared to other patients (4.9% vs 1.6%). Of children requiring CRRT after cardiac surgery, 38% were <6 months old and weighed <10 kg. Also, more than 3 organ failure has been detected in 80.2% of patients requiring CRRT after cardiac surgery. The mean arterial pressure in children with heart disease was significantly lower than in other patients, so 95% needed vasoactive drugs with a higher inotropic index. In addition, many of these patients were on mechanical ventilation at the time of starting CRRT. Early diagnosis and start of CRRT will reduce morbidity and complications

Keywords: Acute kidney injury (AKI), Continuous renal replacement therapy (CRRT)

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Two Almost Missed Unusual Cases of Hypercalcemia Presenting With Urinary Tract Infection

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ABSTRACT

Case 1: A 3-month-old girl with poor feeding and weight gain, developmental delay, polyuria, and constipation for 1.5 months was admitted with a probable urinary tract infection with a positive urine culture for *E. coli*. On physical examination, she was emaciated, hypotonic, and had difficulty holding her neck. The parents were first cousins. Bilateral nephrocalcinosis accompanied by hypercalciuria (urine calcium/creatinine: 1.1) was noted on subsequent evaluation for urinary tract infection. The patient was almost released from the hospital without being tested for her blood calcium and phosphorus levels despite having hypercalciuria. However, when the levels were finally checked, her calcium was 18 mg/dL, and her phosphorus was 3.7 mg/dL, which was relatively high. This level of hypercalcemia in the presence of low phosphate level and phosphaturia was indicative of neonatal severe hyperparathyroidism (NSHPT), the disorder of calcium-sensing receptors. However, the suppressed parathormone level (2.1 pg/mL) and hypercalciuria were inconsistent with NSHPT. The hypercalcemia was treated symptomatically until genetic testing revealed a CYP24A1 loss-of-function mutation compatible with idiopathic infantile hypercalcemia type 1. The CYP24A1 gene encodes the enzyme 24-hydroxylase, which converts active vitamin D metabolites such as 1,25-(OH)2-vitamin D to its inactive form. Although phosphate levels are typically normal in these patients, this patient had hypophosphatemia and phosphaturia, possibly due to excessive stimulation of FGF23 by the high levels of 1,25 OHD. The patient is now on vitamin D and ketoconazole to reduce CYP27B1 expression and activity, and on these therapies, her calcium and phosphorus levels are within normal ranges.

Case 2: A 15-month-old boy was admitted to the hospital because of his high creatinine (1.01 mg/dL) level. He had a history of recurrent episodes of nausea, vomiting, constipation, and polyuria over the past three months, resulting in multiple admissions. On his last admission, he was diagnosed with pyelonephritis and was treated with ibuprofen, cephalexin, ketotifen, and nystatin before being discharged. Upon admission to our center, given his drug history and renal insufficiency, the first impression was tubulointerstitial nephritis. He was developmentally delayed and could not walk, unlike his twin sister. Renal ultrasound showed bilateral increased cortical echogenicity in favor of nephrocalcinosis. The patient's creatinine levels returned to near normal with hydration alone. However, during the patient's discharge process, elevated levels of serum calcium (12.5 mg/dL) and phosphate (4.4 mg/dL), which had not been noticed until then, were discovered incidentally. Subsequent laboratory studies showed serum calcium of 13.5 mg/dL, urine calcium/creatinine of 0.8, serum phosphate of 7.2 mg/dL,

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PTH of 1 pg/mL, and 25-hydroxy vitamin D of 33.3 ng/mL. High levels of calcium and phosphate, along with suppressed PTH levels, suggest vitamin D toxicity or disorders of its metabolism. Upon further investigation, it was discovered that both twins had received a stoss dose of vitamin D3 and regular doses. However, the 25-hydroxy vitamin D levels were in the normal range. The hypercalcemia was treated symptomatically with bisphosphonate, and whole exome sequencing was performed. During the next three months, the patient was readmitted twice with the same clinical picture and was again treated with bisphosphonate. The patient's calcium phosphate and creatine levels subsequently remained in the normal range, his general condition improved, and he began to walk. However, genetic testing and re-analysis of the genetic data returned negative for any related disorder, and the diagnosis remained unexplained.

Keywords: Hypercalcemia, Child, Urinary tract infection (UTI)

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Effect of Exercise Training on Functional Capacity, Muscle Strength, Exercise Capacity, Dialysis Efficacy, and Quality of Life in Children and Adolescents With Chronic Kidney Disease: A Systematic Review and Meta-analysis

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ABSTRACT

Background and Aim: We aimed to investigate the effect of exercise training on functional capacity, muscle strength, exercise capacity, dialysis efficacy, and quality of life (QOL) in children and adolescents with chronic kidney disease (CKD).

Methods: We searched PubMed/Medline, Scopus, PEDro, Web of Science, CINAHL, Cochrane, and Embase databases from inception to September 30, 2023. In this review, we included randomized control trials (RCTs) and clinical trials that explored the effect of exercise training programs on functional capacity, muscle strength, exercise capacity, dialysis efficacy, and QOL in children and adolescents with CKD. We employed a random effect model and meta-regression to conduct the meta-analysis.

Results: Four clinical trials and three RCTs were eligible for this review. The results indicate that exercise training improves strength, but meta-analysis did not reveal a significant effect of exercise on functional capacity (WMD: 1.02; 95% CI, −0.14 to 2.18; P=0.083) and QOL (WMD: 8.00; 95% CI, −3.90 to 19.91; P=0.187). Subgroup analysis showed that more than 25 sessions and 45 min per intervention session, a PEDro score of more than 5, and being younger than 12 years significantly affect functional capacity and QOL results. Due to the limited number of studies on the effect of exercise on dialysis efficacy and exercise capacity, the findings were inconclusive in this area.

Conclusion: Exercise training profits children and adolescents with CKD by increasing their strength. Longer exercise interventions may help improve functional capacity and QOL. Future well-designed RCTs could overcome the existing limitations using adequate sample sizes with longer exercise durations.

Keywords: Chronic kidney disease, Dialysis, Child, Exercise

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Recurrence of Urinary Stones in Children

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ABSTRACT

Background and Aim: A recurrence rate of 50% has been reported for urinary stones in a 7-year follow-up. We aimed to estimate the recurrence rate of nephrolithiasis in a follow-up ≥1 year and determined factors associated with the recurrence.

Methods: A cross-sectional study was performed on children ≤18 years old diagnosed with nephrolithiasis at the Nephrology Clinic of Dr Sheikh Hospital from March 1989 to September 2022. These cases were followed up for ≥1 year in the study. Cases with anatomical urinary obstruction, neurogenic bladder, and kidney dysfunction were excluded. Increased stone size for ≥1 mm or the appearance of new stone was considered a stone recurrence. The correlation between recurrence to gender and the following factors were assessed at presentation: age, the size of the biggest stone, number of stones, unilateral versus bilateral stones, positive family history of stone, presence of hypercalciuria, and hyperuricosuria.

Results: In total, 147 patients were enrolled, including 53.1% boys. The median age at presentation was 21 months. There was a positive family history of stones in 65.3%, hypercalciuria in 20.4%, and hyperuricosuria in 19% of patients. Bilateral urinary stones were found in 28.6% of patients. The median size of the largest stone and the number of stones were 2.5 mm and 2, respectively. The patients were followed up for a median of 22 months. Recurrence of stone was reported in 43 cases (29.2%). Stone recurrence was significantly higher in the group with a positive family history of stone than those without (P=0.033). Also, patients with stone recurrence compared to those without had a significantly higher duration of follow-up (P=0.022). Other factors did not significantly correlate with stone recurrence (P>0.05). The recurrence rates in follow-ups of 1 year, 1 to 3 years, 1 to 5 years, and >5 years were 18.2%, 23.8%, 26.4%, and 45.5%, respectively.

Conclusion: The recurrence of urinary stones in children is not uncommon and increases with the duration of follow-up. It is more common in children with a family history of stones and a longer duration of follow-up.

Keywords: Urinary stone, Children, Recurrence, Hypercalciuria, Hyperuricosuria

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Acute Axonal Motor Neuropathy in a Child With Continuous Ambulatory Peritoneal Dialysis After Remdesivir Therapy

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ABSTRACT

Food and Drug Administration approved remdesivir for managing hospitalized pediatric patients aged 12–17 years (weighing ≥40 kg) with SARS-COVID-19. It is authorized to be given in patients with estimated glomerular filtration (eGFR) <30 mL/min. However, it was recommended that the risk be outweighed in patients with end-stage kidney disease. The reason for avoidance of remdesivir in the setting of chronic kidney disease stage V is the concern for accumulation of the excipient Betadex sulfobutyl ether sodium. It is frequently known as sulfobutyl ether beta-cyclodextrin-sodium (SBECD). At this moment, we represent a child on continuous ambulatory peritoneal dialysis (CAPD) who received remdesivir for SARS-COVID-19. An 18-month-old girl known for focal segmental glomerulosclerosis (FSGS), who recently underwent CAPD, was admitted to the Pediatric ICU because of respiratory distress due to SARS-COVID-19 infection. She received dexamethasone, enoxaparin, remdesivir, antibiotics, and IVIG during 2-week-admission. Two days after discharge, she re-admitted because of continued excessive irritability, vomiting, and diarrhea. Neurological examination revealed axial hypotonia, lower limb weakness in the distal part, and hyporeflexia. Electromyography and nerve conduction velocity revealed acute generalized motor axonal neuropathy more prominent in the distal part of the extremities. The patient was treated with IVIG and physiotherapy. Gradually, the muscle weakness began to improve, and after 45 days, he completely recovered. Guillain-Barré syndrome and toxic neuropathy due to remdesivir were the most important causes of neuropathy in this patient. Of course, it is not possible to find the main culprit of neuropathy out of these two etiologies.

Keywords: Remdesivir, Continuous ambulatory peritoneal dialysis (CAPD), Neuropathy, Chronic kidney disease (CKD), Guillain-Barré syndrome

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Renal Manifestations in Pediatric Patients With Familial Mediterranean Fever: A Case Series

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ABSTRACT

Background and Aim: Familial Mediterranean fever (FMF) is an autoinflammatory disorder typically characterized by recurrent episodes of fever and serositis. The most severe complication of FMF is the development of AA amyloidosis from prolonged inflammation, which can cause end-organ damage, including nephropathy, glomerulonephritis, hematuria, hypertension, urinary tract infection (UTI) and amyloidosis-related nephrotic syndrome or end-stage renal disease (ESRD). We present a case series of 5 young girls with FMF and varying presentations of kidney disease to highlight the importance of screening and early management.

Methods: This retrospective case series reviewed the medical records of 5 female pediatric patients diagnosed with FMF who presented to Ali-Asghar Children's Hospital in Tehran City, Iran, between 2013 and 2023. All patients had genetic testing confirming mutations in the MEFV gene. We collected data on clinical features, laboratory investigations, medical imaging, histopathology reports, treatments, and outcomes related to renal manifestations.

Results: All patients had recurrent fever episodes. Case 1 had seizures, Crohn disease, arthritis, hematuria, hypertension, and proteinuria. Case 2 had a UTI, and case 3 had proteinuria. Case 4 developed nephrotic syndrome and end-stage renal disease from AA amyloidosis, requiring peritoneal dialysis. Case 5 had asthma and focal segmental glomerulosclerosis (FSGS), leading to ESRD, necessitating hemodialysis. Among patients with detailed lab records, elevated acute phase reactants were noted during attacks across cases. Hypertension requiring an ACE inhibitor was seen in case 1. Proteinuria ranged from subnephrotic (<350 mg/d) in case 3 to heavy nephrotic range (>3.5 g/d) in case 4. When performed, renal biopsies showed AA amyloidosis deposits (case 4) and FSGS (case 5).

With colchicine treatment, fever attack severity, infection rate, and proteinuria decreased in cases 2 and 3. Case 1 also received steroids, immunomodulators for Crohn disease, and antihypertensives with partial control of hematuria and proteinuria. Cases 4 and 5 underwent regular dialysis without renal transplantation owing to high costs.

Conclusion: Our case series showcases the broad spectrum of renal involvement in pediatric FMF patients, from mild disease with microscopic hematuria to ESRD requiring dialysis. Amyloidosis and FSGS were confirmed histologically as the underlying etiologies. Early screening and treatment are vital to prevent amyloid deposition and irreversible kidney damage. Future multicenter prospective studies should delineate the rates of various renal complications in larger pediatric FMF cohorts and test new therapies.

Keywords: Familial Mediterranean fever, Kidney diseases, Proteinuria, Renal insufficiency, Dialysis

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Rare Post-streptococcal Glomerulonephritis Presentations Including Back Pain and Constipation

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ABSTRACT

Post-streptococcal glomerulonephritis (PSGN) is commonly seen after pharyngitis or dermatitis due to the streptococcus genus. PSGN complications vary from rare neurological disorders like to posterior reversible encephalopathy syndrome (PRES). However, PSGN with PRES is completely curable after prompt diagnosis and treatment. The disease has some common symptoms like hypertension, oliguria or anuria, and neurological defects; however, we present a child with PSGN who complained of severe back pain or constipation. Our case is a child with severe back pain and constipation who was diagnosed as PRES after PSGN. During his hospitalization, symptomatic treatment started due to renal and neurological involvement. Also, corticosteroid therapy resulted in a dramatic response to his constipation and back pain. Noticing unusual and rare associated symptoms could help practitioners accurately diagnose PSGN. In addition, these findings help us investigate the disease in more detail.

Keywords: Post-streptococcal glomerulonephritis, Back pain, Constipation, Posterior reversible encephalopathy syndrome

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Tunnel Infection in Child With Continuous Ambulatory Peritoneal Dialysis: A Case Report

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ABSTRACT

A tunnel infection is defined as erythema, tenderness, or edema over the subcutaneous pathway of the catheter. Sonographic studies of the tunnel can help confirm the tunnel infections. Approximately 50% of exit-site infections have an associated tunnel infection. The infection may be limited to the outer cuff of the catheter and inter-cuff pathway or extend to involve the inner cuff. The involvement of the inner cuff is associated with a high risk of peritonitis in the same organism. A 3-year-old boy who had continuous ambulatory peritoneal dialysis (CAPD) since he was 6 months old was admitted with a fever, restlessness, and loss of appetite. He was born with IVF (in vitro fertilization), and end-stage renal disease (ESRD) had been due to renal dysplasia. He had a past history of peritonitis 3 times. This time, the dialysate was clear, and there was no discharge from the existing site, but the tunnel was seen as edematous and tender. In laboratory data, leukocytosis and a rise in erythrocyte sedimentation rate and C-reactive protein were reported. He was admitted and has been started on vancomycin and ceftazidime for him. At first, dialysate was clear, and exist site was without any discharge. After two days, peritonitis and site infection appeared, so both cultures were staph epidermidis. Despite appropriate antibiotic therapy, we had to exit CAPD and insert a new one after one week. Peritoneal catheter exit site and tunnel infections may cause peritonitis and catheter loss. Tunnel infection is presented with subcutaneous tunnel erythema or purulent drainage from the site. Staphylococcus aureus is the most common pathogen at the exit site and in tunnel infections. S. aureus nasal carriage is a significant risk factor for S. aureus catheter infections. Catheter infections are treated by antibiotics, often prolonged, and escalation of exit-site care. Refractory cases can be treated by adjusting the tunnel and exit site and removing the superficial cuff. However, some patients will develop peritonitis after this procedure.

Keywords: Tunnel infection, CAPD, Peritonitis, Case report

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An Infant Girl With Bilateral Multicystic Dysplastic Kidney: A Case Report

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ABSTRACT

Multicystic dysplastic kidney (MCDK) is a congenital abnormality seen in 1 per 4300 live births, more common in males. In addition, It is the most prevalent prenatal diagnosed cystic lesion of the kidney. It may occur unilaterally or bilaterally and is usually seen with other structural ureteral anomalies. As the reports of bilateral alive MCDK cases are limited, we present a 6-day-old girl infant born with bilateral MCDK. Our case had bilateral MCDK complicated by hypertension, severe vesicoureteral reflux (VUR), and urinary tract infection (UTI). She was stabilized during hospitalization with prospective kidney transplantation due to a poor prognosis. Bilateral MCDK is a rare disease with many complications and a poor prognosis, so management and treatment of a patient's symptoms are more challenging. Bilateral MCDK frequently results in death in infancy. Thus, our case report could help manage complicated bilateral MCDK manifestations, especially to improve the patient's survival.

Keywords: Bilateral multicystic dysplastic kidney, Vesicoureteral reflux, Hypertension

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Bartter Syndrome Type 4a in an Adolescent With Lower Extremities Pain: A Case Report

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ABSTRACT

Bartter syndrome is a rare genetic disease with 5 subtypes. In Bartter syndrome type 4, patients are deaf with renal dysfunction since infancy. In this report, we present a 16-year-old girl with congenital deafness with no previous renal complaints. She was referred to our center due to ankle pain. The patient's metabolic conditions were treated. Based on the presence of deafness in the patient and her family, mental retardation, and the age of the patient, Bartter syndrome was suggested. Whole exome sequencing (WES) confirmed the type 4a Bartter syndrome diagnosis.

Keywords: Bartter syndrome, Case report, Child

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Refractory Pericardial Effusion Due to Acid-fast Bacillus Positive Smear of Pericardial Fluid in Down Syndrome Girl Presented With End-stage Renal Disease: A Case Report

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ABSTRACT

Non-tuberculosis mycobacterial pericarditis is identified by acid-fast bacillus positive smear in pericardial space. Pericarditis due to non-tuberculosis mycobacterial infections is very rare and is associated with severe sepsis, cardiac tamponade, and high mortality. Pericardial involvement with non-tuberculosis mycobacterial infections in Down syndrome with end-stage renal disease is unusual. We present a case of Down syndrome with recurrent severe pericardial effusion and profound anemia, along with end-stage renal disease and near tamponade presentation. She was not responsive to dialysis, and after an acid-fast bacillus-positive smear in pericardial space, her treatment began. She got better, and after 45 days of hospitalization, she was discharged.

Keywords: Pericarditis, Non-tuberculosis mycobacterial infections, Down syndrome, End-Stage renal disease, Case report

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The Frequency of Kidney And Urinary System Abnormalities In Children With Imperforate Anus Anomaly

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ABSTRACT

Background and Aim: Imperforate anus is a congenital anorectal anomaly. We investigated the frequency of kidney and urinary system abnormalities in children with an imperforate anus.

Methods: This cross-sectional study was conducted on 70 children with imperforate anus who underwent surgery. Renal ultrasound, urinalysis, culture test, serum urea, and creatinine were assessed. Cystourethrography or kidney core scans such as DMSA (dimercaptosuccinic acid) or DTPA (diethylene-triamine-pentaacetate) were performed in abnormal ultrasound and tests.

Results: The mean age of the study participants was 6.56±4.16 years, and 52.9% were females. Fever (17.1%) and lack of weight gain (15.7%) were the most common clinical complaints. Abdominal examination was abnormal in two subjects (2.9%), and lumbosacral examination was abnormal in five subjects (7.1%). Five subjects (7.1%) had minor labial adhesions and five (7.1%) had hypospadias. Urine culture was positive in 15 cases (23.4%). Abnormal ultrasound findings were reported in 36 subjects (51.4%). Voiding cystourethrogram was performed for 43 patients, and 20 (28.6%) had urinary reflux. Twenty-five patients (78.1%) had abnormal findings in DMSA. DTPA was performed in 11 patients (15.7%), two of which had evidence of ureteropelvic junction obstruction on the right and three on the left.

Conclusion: Abnormal renal findings were detected in 51.4% of the patients. Considering the high frequency of congenital anomalies of the kidney and urinary system in children with imperforate anus, it is recommended to examine the kidney and urinary system of all these patients.

Keywords: Imperforate anus, Kidney abnormalities, Child

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Infant With Polydipsia, the Value of Re-history Taking

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ABSTRACT

History taking in clinical practice assesses each symptom in detail, highlighting what essential questions are important to ask and which critical differential diagnosis is important to rule out. In some cases, after taking the initial history, the exact diagnosis of the disease is not precise, and it is necessary to ask more detailed questions. Our patient presented with polyuria, polydipsia, and Fanconi syndrome. However, after carefully reviewing his medical history, we found out that he has a cousin with cystinosis, which leads us to more investigation for the diagnosis of cystinosis. The point of this case report was to encourage fellow physicians to pay more attention to each sign and symptom and clinical and family history. A 12-month-old boy born into a consanguinity marriage was presented with excessive thirst and heavy diapers due to excessive urination volume. Previous deprivation and DDAVP (desmopressin) tests were inconclusive. He had diluted urine while dehydrated and a mild low estimated glomerular filtration rate (Cr= 0.8 mg/dL). The other findings were mild to normal anion gap metabolic acidosis (HCO_3=17), hypokalemia (K=2.3 mEq/L), and hypophosphatemia (Phosphorus=3 mg/dL). Urine and blood amino acid chromatography results were in a normal range. Ophthalmic exam and bone marrow aspiration were normal. After genetic consulting and requesting a genetic test, indomethacin (2 mg/kg/d) and hydrochlorothiazide (1 mg/kg/d) were given. One month later, the lab test revealed glucosuria, aggravated hypophosphatemia, and hypokalemia. The cystine level of leukocytes was 0.14 (normal range). Re-history taking and emphasis on affected family members revealed that one of the cousins had cystinosis. Cystagon was started for the patient. This case report highlights the significance of re-history taking and not confining on the early information.

Keywords: History taking, Cystinosis, Polydipsia

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A Rare Case of Concomitant Parvovirus B19 Infection and Cytomegalovirus Infection in a Kidney Transplanted Child

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ABSTRACT

Parvovirus B19 is a common human infection worldwide. It is a member of the Parvoviridae family and Erythroparvovirus genus. Parvovirus B19 is a small, non-enveloped, single-stranded DNA (ssDNA) virus infecting only immunocompromised persons and is uncommon in normal children. However, immunosuppressive patients are prone to it. We present a kidney-transplanted child infected by parvovirus B19 presented with intractable anemia. The patient was diagnosed after evaluation for dizziness and lethargy. A complete anemia work-up revealed a low reticulocyte count and hemoglobin <6 g/L. Parvovirus PCR (polymerase chain reaction) was positive, so we decreased immunosuppressive drugs and removed prophylactic Valcyte and cotrimoxazole. Anemia improved after receiving intravenous immunoglobulin 2 g/kg given four doses per week. Unfortunately, he came back with cytomegalovirus infection and a rise in creatinine. Parvovirus B19 infection is typically self-limiting, but immunocompromised patients demand specific treatment. pretransplant screening seems to be unimportant.

Keywords: Kidney transplantation, Child, Parvovirus B19, Cytomegalovirus

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Platelet Indices in Children With Nephrotic Syndrome

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ABSTRACT

Background and Aim: Nephrotic syndrome is a prevalent kidney disease characterized by a significantly elevated risk of thrombotic events. The present study aimed to assess mean platelet volume in children with nephrotic syndrome and compare between the patient groups.

Methods: This cross-sectional study was conducted in the Pediatric Nephrology ward at Zahedan University, Zahedan City, Iran. The study sample consisted of 491 children (333 children with idiopathic nephrotic syndrome and 156 healthy children). Changes in platelet count and volume were evaluated in the groups of nephrotic syndrome and healthy children in steroid-resistant and steroid-responsive patients. Data analysis was performed in SPSS software, version 20 (SPSS, Inc., Chicago, IL). The significance value and confidence coefficient levels were considered 0.05 and 95%, respectively.

Results: The mean platelet count changed significantly in the steroid-responsive, steroid-resistant, and control groups. The mean platelet volume was significantly different in the steroid-responsive, steroid-resistant, and control groups. In the patients, the means of platelet count were 413.26±131.81 and 331.26±102.28 ($P<0.001$) in those with active phase and remission, respectively. The difference between means of mean platelet volume were 8.28±1.22 and 7.96±1.04 ($P<0.001$) in those with active phase and remission, respectively. Platelet and mean platelet volume had no significant correlation with the patients' cholesterol, albumin, and protein/creatinine ratio ($P>0.05$).

Conclusion: The study concluded that platelet count was the highest value in resistance compared with responders and controls but a converse result for mean platelet volume. We suggest to evaluate these indicators to predict the prognosis of nephrotic syndrome in children.

Keywords: Platelet count, Mean platelet volume, Nephrotic syndrome, Children

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Comparing Mean D-dimer Levels Between Children With Urinary Tract Infection and Healthy Children

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ABSTRACT

Background and Aim: Urinary tract infection (UTI) is a common disease in children. This infection is the most common serious bacterial infection in children with fever. It may be associated with complications such as involvement of the renal parenchymal infection (acute pyelonephritis) followed by late renal scarring. Due to the increase in D-dimer in infection and the possibility of differentiating it from non-infectious cases, we decided to conduct a study to determine and compare the mean serum levels of D-dimer in healthy children and children with urinary tract infections.

Methods: This case-control study was performed on 130 children in two groups (65 children with culture-confirmed urinary tract infections and 65 healthy children) in 2020. In these children, demographic data, complete blood count (CBC) findings, and D-dimer level (measured by ELISA method) were assessed and analyzed using SPSS software.

Results: This study showed that the mean age of children with UTI was 4.42 years, and that of healthy children was 5.35 years. Also, the mean age of all subjects was 10.10. It was 4.88 years. Also, most subjects were children with urinary tract infections and healthy girls (with a frequency of 78.46% and 60%, respectively). White blood cell count in children with urinary tract infections was significantly higher than in healthy children ($P<0.001$). The mean D-dimer level in children with UTI was significantly higher than in healthy children ($P<0.001$). However, there was no significant difference in hemoglobin and platelet levels.

Conclusion: According to the results of this study, in children with UTI aged 1 month to 15 years, the level of D-dimer is significantly higher than that of healthy children. Therefore, D-dimer can be considered a potential diagnostic marker of UTI in children.

Keywords: D-dimer, Pediatrics, Urinary tract infection (UTI)

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Intradialytic Hypotension in End-stage Renal Disease Children

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ABSTRACT

Hypotension usually happens during hemodialysis in pediatric patients. Intradialytic hypotension (IDH) may decrease the effectiveness of the dialysis, resulting in increasing morbidity and mortality related to hemodialysis. Although IDH has many definitions over the years, absolute systolic blood pressure (SBP) correlates significantly with patient outcomes. However, Kidney Disease Outcomes Quality Initiative guidelines have defined IDH as either a decrease in SBP ≥20 mmHg or mean arterial blood pressure ≥10 mmHg in combination with hypotension symptoms. Kidney Foundation Kidney Disease Outcomes Quality Initiative or European Best Practice Guidelines definitions, with some variations, included a fall in SBP of ≥30 mmHg. The cardiovascular response to hypovolemia encompasses heart rate, contractility, and peripheral vascular resistance alterations. Sodium is crucial to preserving plasma tonicity and vascular refilling. Single or dual-center studies have established that high dialysate sodium can increase interdialytic weight gain and hypertension but less IDH. However, lower dialysate sodium reduces interdialytic weight gains.

Keywords: Hypotension, Hemodialysis, Child

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Risk Factors of Hyperkalemia in Dialysis Patients: A Single-center Study

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ABSTRACT

Background and Aim: We investigated the association between hyperkalemia with demographic characteristics, types of dialysis, acid-base conditions, serum sodium, bicarbonate, and pH levels in venous blood and drugs.

Methods: This was a six-month cross-sectional study (July to December 2020). Patients managed by the dialysis centers of Dr Sheikh Hospital entered the study. Potassium levels > 5.5 meq/L were considered hyperkalemia. Patients were divided into groups with hyperkalemia and non-hyperkalemia. The following items were compared between groups: median age and duration from placing on dialysis, gender, modality of dialysis, venous pH <7.31 and > 7.41, hyponatremia (sodium <135 meq/L), mean serum sodium levels, bicarbonate, and venous pH levels as well as using furosemide, angiotensin-converting enzyme inhibitor, angiotensin receptor blocker, and Shohl's solution.

Results: A total of 71 dialysis children (57.75% boys) entered the study (median age: 12 years), including 54.9% peritoneal dialysis and 45.1% hemodialysis cases. In total, 38 patients (53.5%) had at least one episode of hyperkalemia during the study. Hyperkalemia was found in 53.3% of girls and 53.6% of boys (P=0978), and 81.2% of hemodialysis and 30.8% of peritoneal dialysis patients (P<0.001). In total, 12.8% and 25.8% of episodes of hyperkalemia and non-hyperkalemia were accompanied by a venous pH >7.41 (P=0.027). A venous pH < 7.31 was noted in 11.5% and 8.1% of episodes of hyperkalemia and non-hyperkalemia, respectively (P=0.41). Hyponatremia was present in 15.8% of episodes of hyperkalemia and 10.4% of episodes with non-hyperkalemia (P=0.206). There was no significant difference in using furosemide, captopril, losartan, enalapril, and Shohl's solution between the two groups (P>0.05 for all). The median age and duration from being placed on dialysis were not significantly different between groups (P>0.05 for both). The mean serum sodium levels were significantly lower in hyperkalemic patients (P=0.008). Logistic regression analysis showed that neither hyponatremia nor serum sodium, pH<7.31 and pH > 7.41, and hemodialysis modality are risk factors for the development of hyperkalemia.

Conclusion: We found that hemodialysis was significantly associated with hyperkalemia. Also, a venous pH >7.41 was significantly more common in non-hyperkalemic episodes. However, we could not find any factor that increases the risk of hyperkalemia in dialysis patients.

Keywords: Hemodialysis, Peritoneal dialysis, Acid-base disorders, Hyperkalemia, Medications

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Evaluation of Urine Citrate, Magnesium Levels, and Prolonged Hematuria in Children

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ABSTRACT

Background and Aim: Hematuria is one of the most common complaints of children in the nephrology clinic. Hematuria is defined as the excretion of >5 red blood cells (RBCs)/high power field (HPF) in uncentrifuged urine. It can be divided into macroscopic, microscopic, transient, or persistent types. One important etiology of hematuria is the increased excretion of promotors of crystallization and stone formation like calcium, oxalate, and uric acid, while the decreased urinary excretion of inhibitors of crystallization like magnesium and citrate may also be very important. This study aimed to evaluate and determine the excretion of urinary promoters and inhibitors of crystallization in children with continuous microscopic hematuria in Taleghani Hospital in Gorgan City, Iran, in 2021.

Methods: Following ethical principles and confidentiality, the urine samples were examined after obtaining informed consent from 246 children with microscopic hematuria. The enzymatic method was used to evaluate oxalate and citrate content. Also, the automatic analysis method was used to measure magnesium content. For patients, after proving microscopically isolated hematuria, a random urine test for urinary minerals citrate, magnesium, calcium, oxalate, and uric acid was performed.

Results: The Mean±SD age of the subjects was 4.79±3.67 years; 129 (52.4%) were boys, and 117 (47.6%) were girls. The Mean±SD urinary citrate to Cr was 5.86±3.37 mg/mg. In our study, 19 individuals (7.7%) had hypocitraturia, and 227 (92.3%) had normal urinary citrate. The Mean±SD urinary magnesium to Cr was 0.263±0.225 mg/mg. In our study, 183 patients (74.4%) had hypomagnesuria, and 63(25.6%) had normal urinary magnesium levels. This study's Mean±SD urinary calcium to Cr was 0.7±0.4 mg/mg. In our study, 68 (27.6%) hematuria patients had hypercalciuria, and 178 (72.4%) had normal urinary calcium levels. The Mean±SD uric acid to Cr was 1.42±1.06 mg/mg. In our study, 34 hematuria patients (13.8%) had hyperuricosuria, and 212 patients (86.2%) had normal uric acid levels. The Mean±SD urinary oxalate to Cr was 0.372±0.149 mg /mg. In our study, 99 patients with hematuria (40.2%) had hyperoxaluria, and 147 patients (59.8%) had normal urinary oxalate levels.

Conclusion: In this study, we observed that a significant percentage of children aged 1 to 14 years with hematuria had urinary mineral abnormalities; the most frequent abnormality was hypomagnesuria, followed by hypercalciuria, hyperuricosuria, hyperoxaluria, and hypocitraturia. This finding shows the important role of urinary crystallization inhibitors in producing hematuria other than the urinary promoters. More precise studies, including interventional studies with supplementation of magnesium and citrate, are required to investigate the role of urinary crystallization inhibitors in hematuria.

Keywords: Microscopic hematuria, Urinary citrate, Urinary magnesium, Minerals

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Research on Legal Challenges and Their Consequences in the Treatment of Chronic Kidney Diseases in Children

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ABSTRACT

Dealing with various problems and issues in the diagnosis and treatment of patients with chronic kidney diseases in pediatric departments is one of the serious, time-consuming, and tiring challenges for doctors and medical staff. Years of experience working in children's hospitals, examining relevant cases, being a member of medical malpractice investigation commissions, and consulting with parents and doctors made the author think about investigating and researching the causes of these challenges and ways to prevent them. In examining the cases, there were various reasons; the most common and important ones include non-acceptance and denial of the diagnosis at first by the parents, request to leave the hospital with personal consent and refer to a doctor or other centers, multiple referrals to different doctors, referral to alternative medicine such as traditional medicine, etc., refusal of treatment and parental intervention in treatment, request to remove part of the treatment from the parents, parents' or child's disobedience in accepting the need for hospitalization and requesting outpatient treatment, lack of trust in the treating physician and ask to change the doctor or center and discharge with personal consent, non-acceptance of complications, failure to refer on time for follow-up, and so on. Regarding the refusal to follow the doctor's orders for sick children, it is a different matter from adult patients because the decision maker here is not the sick child but the legal guardian of the patient. According to the law, the guardians include the father, paternal grandfather, or a substitute person according to the order of the judicial authority, etc. In this article, two concrete examples of legal challenges in these patients are referred to for a better understanding: refusal of treatment and parental involvement. The first case was a child with nephrotic syndrome whose mother prevented him from receiving regular treatment, medication, and related follow-ups. Finally, the child died, and the parents complained to the treating doctors. The second case was a 3-year-old child with a rare metabolic disease and kidney failure who was a candidate for peritoneal dialysis, and the parents prevented him from performing peritoneal dialysis according to the procedure in the government hospital. This article will discuss issues on how to properly deal with these cases in medical centers that can be associated with the risk of death or irreversible injuries, the role of the hospital forensic doctor and psychiatrist, the responsibility of the attending physician, the role of parents and their decisions, the complaint, and the handling process, auxiliary measures dealing with parents and the need to record documents in the clinical file and how to respond to judicial authorities, etc.

Keywords: Chronic kidney diseases, Sick child, Parents, Doctor, Law, Nephrotic syndrome

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Comparing the Efficacy of Tolterodine Versus Oxybutynin in Treating Children With Desmopressin-resistant Enuresis

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ABSTRACT

Background and Aim: Enuresis is defined as involuntary nocturnal urination without an underlying physical debility in a child old enough to control urination. We evaluated the efficacy of tolterodine and oxybutynin in patients with primary desmopressin-resistant enuresis.

Methods: This randomized clinical trial was performed on 68 patients aged 5 to 16 years who suffered from primary enuresis. They were randomly assigned to two treatment groups for three months: group 1, oxybutynin + desmopressin, and group 2, tolterodine+ desmopressin. Patients' demographic characteristics, clinical and laboratory findings, and subjective response to treatment were documented based on the number of wets per night and per week and compared with pre-treatment.

Results: Patients were put in two groups (30 patients in group 1 and 30 in group 2). The mean age of patients was 88.97±27.09 months. Six out of 30 patients (20%) in the first group and 5 out of 38 (13.2%) in the second group recovered completely without a statistically significant difference between the two groups. Seven patients (23%) in the oxybutynin group and 13 (34%) in the tolterodine group showed no response to treatment, with no significant difference between the two groups. Also, the minimum response to anti-muscarinic agents in patients with enuresis was significantly higher in males (P=0.026). Based on gender, other response parameters were not significantly different.

Conclusion: In desmopressin-resistant patients, adding anticholinergic drugs leads to a significant response in more than half of the patients. However, oxybutynin and tolterodine are both equally effective in treating desmopressin-resistant enuresis.

Keywords: Enuresis, Oxybutynin, Tolterodine, Desmopressin

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Bartter Syndrome and Congenital Chloride Diarrhea: A Case Report

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ABSTRACT

Bartter syndrome is an autosomal recessive disorder manifested by persistent polyuria, dehydration, salt wasting, hypokalemic hypochloremia alkalosis with different genetic origins and molecular pathophysiology. Although prompt diagnosis and appropriate treatment of Bartter's syndrome may improve the outcome and prevent complications, other symptoms, such as chronic diarrhea, are sometimes added, the cause of which is important. We report a 6-year-old girl who was admitted to the Nephrology Department of Hazrat Masoumeh Hospital of Qom University of Medical Sciences, Qom City, Iran, on September 5, 2023, due to congenital chloride diarrhea and electrolyte imbalance. The patient had electrolyte disorders since birth and was under follow-up and treatment due to the diagnosis of Bartter syndrome. Now, the patient has chronic chloride diarrhea and is not growing well. In this syndrome, if there is diarrhea at the same time, a correct diagnosis of the cause of diarrhea and its treatment will help prevent complications.

Keywords: Dehydration, Diarrhea, Metabolic alkalosis, Hypokalemia

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A Novel Mutation in the Alanine–glyoxylate Aminotransferase Gene in an Iranian Patient: A Case Report

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ABSTRACT

Primary hyperoxaluria 1 is a rare genetic disorder that typically results in nephrolithiasis and or nephrocalcinosis in patients. It is caused by a deficiency of alanine–glyoxylate aminotransferase. This study aims to report a novel mutation in the alanine–glyoxylate aminotransferase (*AGXT*) gene in an Iranian patient. The patient's clinical data and family history were collected, and the salting out technique was used to extract the genomic DNA from the whole blood. Sanger sequencing was performed for all coding exons of the *AGXT* gene. The variants were interpreted according to The American College of Medical Genetics and Genomics (ACMG) guideline. The patient was a 24-year-old female who suffered from recurrent nephrolithiasis since birth. She also revealed bilateral nephrocalcinosis. The affected individual was born into a consanguineous marriage. Genetic analysis revealed a previously unreported homozygous missense mutation in the *AGXT* gene (c.846G>T/p.Gln282His). Based on the ACMG classification, this mutation is classified as likely pathogenic. In this study, we identified a novel mutation in an Iranian patient with hyperoxaluria that can extend our knowledge about the genotype of the *AGXT* gene and understanding the etiology of hyperoxaluria type 1.

Keywords: Case report, *AGXT* gene, Missense mutation