**Nephrology**

**Randomised trial comparing short and long intravenous antibiotics in children with acute pyelonephritis: DMSA scan evaluation at 6-month follow-up**

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**Background:** Urinary tract infection (UTI) is a common infection in infants and children. It may lead to irreversible changes in renal parenchyma and long term complications. In pediatric patients with febrile UTI the administration of intravenous antibiotics is often recommended. However, there is no general agreement regarding the duration of intravenous treatment, ranging from a few days to more than two weeks. A potential advantage of prolonged parenteral treatment might be that it would decrease the frequency of renal scar formation. The aim of this prospective study was to compare the prevalence of renal scarring following initial treatment with antibiotics administered intravenously for 14 or 3 days.

**Methods:** In a prospective study (November 2005 to March 2007) 73 children (67 girls (91.8%) and 6 boys (8.2%)) aged 3 months to 12 years with positive urine culture and acute renal lesions on initial DMSA scan were randomly assigned to receive intravenous ceftriaxone (75 mg/kg in two divided doses) for 14 days or 3 days, followed by oral cefixime (4 mg/kg/bid) to complete a 14 days course. After six months, scintigraphy was repeated in order to diagnose renal scars. All children studied with DMSA scan and ultrasonography within 3 days of admission. Investigations were completed by voiding cystourethrography to detect vesicoureteric reflux when urine culture became negative.

**Results:** Renal scarring developed in 19.2% of the 37 children in the 14 days intravenous group and 24% of the 36 children in the 3 days group. Of 29 kidneys with reflux, 28 (96%) were found to have abnormal renal scan. Among 115 kidneys with non-refluxing ureters 99 (86%) revealed parenchymal changes on renal cortical scintigraphy (P >0.05). Children between 1 to 5 years had more renal scarring than infants and group older than 5 years [38 (52%), 18 (24.6%) and 17 (23.3%), respectively]. After adjustment for age, sex, delay of treatment (>7 days) degree of inflammation, presence of vesicoureteral reflux there was no significant difference between the two treatment groups on renal scarring (P=0.3). Also there was no significant difference in creatinine clearance (CCr) at the first and six months later between two treatment groups. In final analysis rate of remission on DMSA scan was significantly high in both groups (P=0.001).

**Conclusion:** In children with acute pyelonephritis initial intravenous treatment for 14 days, compared with 3 days, dose not significantly reduce the development of renal scar formation process. Additionally, we found that an antibiotic course of 3 days is safe when powerful antibiotic such as ceftriaxone is used

**Keywords:** Acute pyelonephritis, Children, DMSA scan, Vesicoureteral reflux, Scar

**A case report of bladder extrophy in 4 years old girl complicated with bilateral vesicoureteral reflux in a 4 years follow up**

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A case report of bladder extrophy in 4 years old girl complicated with bilateral vesicoureteral reflux in a 4 years follow up

**Background:** The bladder extrophy is a rare cloacal membrane defect with failure of fusion of the lower abdominal wall, the symphysis pubis, urinary tract and the external genitalia to different degrees such as exteriorization of the pelvic viscera on the abdominal surface, inferiorly displaced umbilicus and abnormal exterior genitalia. Complete bladder extrophy and epispadias with concomitant separation of the pubic bones is the most common in clinical cases. Bladder extrophy is rare, occurring once in 25 000–50 000 births, with a 2 : 1 male to female ratio. There is some evidence that genetic factors may play a role to this pathology and other cloacal malformations.

**Case presentation:** The Case was a 4 years old girl, born after a normal term pregnancy with a normal birth weight. She was the child of a healthy mother. On physical examination all the characteristic features of bladder extrophy were present. No other pathological findings was found. Neck of bladder was open in cystoscopy. The patient underwent the excision of exstrophied bladder through a successful operative process. 4 years after the surgery the patient referred to childeren’s medical center with the chief complaint of incontinency. A kidney, ureter, and bladder (KUB) x-ray showed evidences of exstrophied bladder and grade 2 of bilateral vesicourethral reflux. ureteric orifices was abnormal (stadium or the golf hole). There was no evidence of trabeculation, stenosis and/or diverticulum and the bladder was in normal size. The length of the duct was about 10 cm. After the assessments through cystoscopic procedure the diagnosis changed in to the presence of short urethra and bilateral Vesicoureteral reflux (VUR). The patient was discharged with good general condition and further surgery was
recommended in order to treat Vesicoureteral reflux.

**Conclusion:** Nowadays, death from extrophy alone is uncommon and improvement of surgical techniques have made it possible to cure children with this abnormality.

**Keywords:** Bladder extrophy, Vesicoureteral reflux, VUR

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**Prevalence and symptoms of Idiopathic Hypercalciuria in primary school children of Rasht**

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**Background:** Hypercalciuria is defined by urinary calcium excretion more than normal rate for age. Hypercalciuria causes many of urinary symptoms like hematuria, frequency, dysuria, UTI and et. It will be silent in children. For determine the prevalence and clinical symptoms of Hypercalciuria in primary, school children in Rasht, we did a descriptive and cross – sectional study. The aim of this study was to determine the prevalence of idiopathic hypercalciuria (IH) in school children in rasht.

**Methods:** This study was descriptive and cross – sectional we had 30 primary school. We evaluated 340 primary school children (age 6-11, mean 9.1 years) in two steps: first (Screening test), we measured urine calcium to urine creatinine ratio (UCa/UCr) and in the second step (Definitive test), for those children who had UCa/UCr ratio more than 0.21 mg/mg we measured 24 hours urine calcium excretion. Children with secondary forms of hypercalciuria were excluded from the study. In the end of study we take a good history and did physical examination, kidney sonography and necessary laboratory exam to determine the cause of hypercalciuria

**Results:** Of 340 children, 180 were males and 160 females. The mean age of patients were 9.3 years old. In the first screening, 47 (13.8%) children (26 males, 21 females) had an abnormal UCa/UCr ratio. But in the end only 19 had the criteria of IH, i.e. the prevalence of IH was Prevalence of Idiopathic Hypercalciuria in primary school children of Rasht 5.6%. The prevalence in females and males was 3.3% and 2.3%, respectively. Of these children 3 had hematuria (including 2 cases of gross hematuria), 5 children gave a history of recurrent abdominal pain, 3 children suffered from dysuria and 12 persons had a history of personal or familial (first or second degree)urolithiasis.

**Conclusion:** Prevalence of IH in our children was 5.6% and its manifestations were: hematuria, dysuria, recurrent abdominal pain, incontinence, urgency, urinary tract infections and urolithiasis.

**Keywords:** Renal stone; Hypercalciuria; Hematuria; Abdominal pain; Calcium

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**Non-Calculus Signs and Symptoms of Hyperoxaluria and Hyperuricosuria in Children: A Single Experience**

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**Background:** Non-calculus presentations of hyperoxaluria (HX) and Hyperuricosuria (HU) are not common. The aim of this study was to investigate the relationship of symptomatic non-calculus idiopathic HX, HU and both of them with dysuria, failure to thrive (FTT), recurrent urinary tract infection (UTI), dysmorphic red blood cells (RBCs) and abdominal pain in children.

**Methods:** A cross sectional study was done on 58 children who were aged less than 14 years with history of persistent microscopic or macroscopic hematuria with HX and/or HU, regardless of having renal calculi, between October 2007 and October 2008. The patients were divided into three groups according to the type of crystalluria (I, 10 HX; II, 20 HU; and III, 28 HX+HU).

**Results:** The common presenting symptoms were: abdominal pain (63%) and dysuria (45%). FTT frequently occurred in female (68%). No significant relation was seen between the groups in terms of gender, macroscopic hematuria and recurrent UTI. We found that dysuria, positive family history, FTT, abdominal pain and dysmorphic RBCs in patients with HX were higher compared to HU group. Moreover, logistic regression analysis showed the higher odds ratio of FTT, abdominal pain and dysmorphic RBCs in patients with HX+HU group when compared to patients with HU.

**Conclusion:** Although our study showed that non-calculus symptoms and signs of crystalluria such as dysmorphic RBCs, FTT, abdominal pain and dysuria are frequently seen in children with HX, however, further studies are needed.

**Keywords:** Hyperoxaluria, Hyperuricosuria, Non-Calculus Signs and Symptoms

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**Relationship between Pathologic and Laboratory Data of Children Suffering from Hemolytic Uremic Syndrome (HUS): A Center study**

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**Background:** Hemolytic uremic syndrome (HUS) is the most prevalent cause of children renal insufficiency which in many cases (90%) occurs following diarrhea.
Hemolytic microangiopathic anemia, thrombocytopenia, and renal insufficiency are main symptoms of hemolytic uremic syndrome. This study aims to consider the relationship between pathologic data of nephro-biopsy and laboratory data of children suffering from the disease. 

**Methods:** This study has been carried out in retrospective, cross-sectional and descriptive procedures. For this purpose, 28 patients with an average age of 6 years suffering from uremic hemolytic syndrome referred to Ali Asghar Hospital over the last 10 years. Light microscopic data of glomeruli, arterioles, arteries, interstitial tissue, medullary vessels and tubules were evaluated. Laboratory data including hematology, biochemistry, and urinary tests were extracted from patients’ files. Data were analyzed using SPSS software.

**Results:** The most prevalent damages in glomeruli were decreased capillary lumen and thickening of its wall and in arterioles were mild decrease of lumen and in artery thickening of intima and mild infiltration of inflammatory cells and mild edema in interstitial and hyperemia in vaso recta and the most prevalent pathology in tubules was the existence of cast. Significant relationship was found out between time of recovery of hematological disorders and medullary vessels congestion and reduplication of arterial inner elastic lamina and also improvement of biochemistry changes with glomerulus necrosis and leucocytes assembly in vaso recta. Arteriolar rate with creatinine serum level at discharge time was related and tubular rate with platelet count at discharging time was also related.

**Conclusion:** Biopsy is an important tool for prognosis and determination of disease intensity. There was valuable statistical relationship between some laboratory data at the time of referral and pathological data which even could influence intensity or prognosis of disease.

**Keywords:** Hemolytic Uremic Syndrome, Histopathology, Laboratory

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**Subcutaneous terbutaline use in CKD to reduce potassium concentrations**

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Background: Acute hyperkalaemia is a common, potentially life-threatening problem for patients receiving maintenance haemodialysis. Patients with chronic kidney disease (CKD) can tolerate only a small potassium load, such as that from dietary ingestion, because of the inability of the kidney to excrete potassium. Maintenance of potassium homeostasis is therefore dependent on extrarenal mechanisms and intermittent haemodialysis. Extrarenal potassium balance is governed by mechanisms that include β-adrenergic-receptor (βAR) mediated stimulation, insulin, aldosterone, acid-base balance and plasma osmolality, but some or all of these mechanisms may be impaired in patients with CKD. βAR agonists reproducibly decrease plasma potassium concentrations in healthy volunteers, suggesting that they might be useful for treating symptomatic hyperkalaemia in patients with CKD. β2AR agonists can be administered by a variety of routes, including nebulization, metered-dose inhalation and intravenous infusion. Each route of administration has its pros and cons. The aim of the study was to assess the effectiveness of weight-based subcutaneous terbutaline dosing in reducing the plasma potassium levels of patients with CKD who require haemodialysis. STUDY DESIGN: Clinical trial. ENDPOINTS: Changes over time in plasma potassium concentration and heart rate in response to terbutaline; and percentage change in responses from baseline.

**Method:** Fourteen patients with CKD (aged 10-15 years) on long-term haemo- dialysis received terbutaline 7 µg/kg subcu- taneously. Heart rates and the potassium concentration in blood samples were measured serially for 7 hours. The effects of terbutaline on heart rate and potassium responses were determined for each patient.

**Results:** Terbutaline significantly reduced plasma potassium concentrations and significantly increased heart rates during the study (p<0.001, repeated-measures analysis of variance). The mean reduction in peak potassium concentration (-1.31±0.5 mEq/L) and the mean increase of peak heart rate (25.8± 10.5 beats/min) relative to baseline were statistically significant (p<0.001, base- line versus peak for both responses). No significant adverse effects were observed.

**Conclusion:** Subcutaneously administered terbutaline is an effective acute treat- ment for hyperkalaemia in patients with CKD undergoing long-term haemodialysis. In patients without active ischaemic heart disease, use of subcutaneous terbutaline renders intravenous access unnecessary and should be considered as an alternative to nebulized or inhaled β-agonists. Close cardiovascular monitoring is necessary to minimize treatment-related toxicity, as with any β- adrenergic receptor agonist.

**keywords:** hyperkalaemia-ckd-terbutaline

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**Undescended testis among six-year-old boys in I.R.Iran - 2009**

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Background: Screening can be considered as the best way for early diagnosis of many diseases to prevent subsequent complications and imposed socio-economic costs to the communities. Some diseases such as
undescended testis have a golden time to be diagnosed and any delay may lead to irreversible results including infertility, carcinogenesis and some other health complications. This study performed to assess the prevalence of undescended testis in all six-year old boys over the country (I.R.Iran) in 2009.

Method: This study was based on a national screening program for screening six-year–old children before entering primary schools, in all the country. As a rule, all children before registration in primary schools must refer to specified assessment centers to be screened for probable diseases or disabilities by trained General Practitioners. Suspicious cases referred to related specialists for more assessment. All findings were registered.

Results: Out of all 955,388 children at the said age group, 484,891 were boys among which undescended testes were detected in 2261 cases (0.47%).

Conclusion: Regarding the golden time for treatment of undescended testis and probable serious complications of the disease, reaching this large number of involved children to age six without any diagnosis or treatment, as an alarm emphasizes on more attention of health decision makers to design earlier diagnosis or treatment, as an alarm emphasizes on more attention of health decision makers to design earlier screening programs and physicians, especially pediatricians to examine testes in all referred boys, before expiration of golden time.

Keywords: Undescended Testis, Screening, Children

The evaluation roles of hypercalciuria and hypocitraturia in children with urolithiasis in the north west of Iran

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Background: now the prevalence of renal stone are increasing because of the change in diet (uses high level protein) and life style. Renal stone is not a disease but it is the side effect of the other disease. in the most of the studies hypercalciuria is the most frequent metabolic Disorders in children with renal stone In this study we evaluated the metabolic factors that they may cause renal stones in children

Method: it is a cross sectional study. in this study we studied 56 children with renal stones in one years from north west of Iran. The mean ages of them were 2.4 years old. (Boys 46%–54% girls). we wanted urine analysis about metabolic elements, we found that only 3 of them had hypercalciuria(0.05) but 47 of them had hypocitraturia(83%) and 1 child had hyperexcretion of uric acid and non of them had oxaloria.

Result: this study showed that in our patient are not statistical significancy about sex and age. the evaluation of metabolic elements in children with renal stone in this region of Iran showed that hypocitraturia had the more important role in producing stones than hypercalciuria.

Conclusion: the evaluation of the level of urine citrate in patient with renal stones in this region of country is necessary. but other studies had need.

Keywords: hypercalciuria-hypocitraturia-renal stone

Kideny involvement has been reported in different bacterial, viral, fungal and parasitic infections.

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Kideny involvement has been reported in different bacterial, viral, fungal and parasitic infections. According to better control of infectious disease, renal involvement has been reduced recently, occurs more in primary severe infections, late diagnosis of native infection with progressive manifestations and inappropriate treatment. Immunologic pathway (immune complex deposition), toxemia (endotoxemia and other toxins), ischemic damage, direct invasion, antimicrobial nephrotoxicity and combination of these are the pathophysiological mechanisms in kidney involvement. Renal vasculature, glomeruli, tubulointerstitium and collecting system may be involved. Different pathologic features has been reported from focal or diffuse proliferative Glomerulonephritis, tubulo interstitial disorder (interstitial edema, mononuclear infiltration and tubular degeneration), MPGN, MGN, glomerular sclerosis, crescentic GN and hyalinosis. Complement, immunoglobulin, antigenic deposition and organism detection in renal biopsy is possible in some of the infections. Some of them have also characteristic pathologic features. Clinical manifestations range from urinary tract infection, renal tubular acidosis, isolated hematuria or proteinuria, hematuria plus proteinuria, sterile pyuria, urinary spreading, acute nephritis, nephrotic syndrome, acute renal failure and renal transplant dysfunction. Characterisitic radiologic manifestation such as renal cyst, necrosis, calcification, abcess formation (direct invasion) and obstructive uropathy has been reported in some infections. Improvement of primary infection could improve renal manifestations. Adjunctive treatments is necessary for better control. An interesting case of hydatid cyst renal involvement with a rare clinical manifestation is discussed in this lecture.

Keywords: kidney, infection, pathogenesis, pathology, symptoms

Persian manna as a prebiotic

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Background: "Taranjabin" or Persian manna has been used for centuries as a remedy to improve infantile health in Iranian traditional medicine, it supposed by some to have been the “manna” of holy books Koran and scripture. Peribiotic are new healthy foods that improve microbial flora of GI system to have beneficial effects on host's by increasing number of bifidobacter and lactobacilli.

Methods: This before and after study conducted on 20 formula fed infant aged between 2-12 months in Yazd. Stool culture was taken before and after Taranjabin consumption (30 gram/ two weeks) and counted for lactobacilli, Ecoli and bifidobacter.

Results: Data showed significant increase in number of lactobacilli, bifidobacter & decrease in E coli count(P<0.05) mean while there was correlation between duration of usage and its bifidogenic effects of the manna (P<0.05) . According to Mother’s opinion their child sleep improved and less sweating took place during study, meanwhile stool was softer than previous. Four Infant developed diarrhea at the beginning of study which exclude from study they all improved without medical intervention no other complication was reported.

Conclusion: "Taranjabin” has perbiotic effect and its effect is depending on duration of consumption, it may improve child wellbeing.

key word : Peribiotic, Taranjabin, Persian manna, Bifidogenic

New Renal Function Tests in Neonates

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Renal function tests are common procedures used to evaluate renal function. Plasma creatinine and urea are relatively insensitive. Limitations of serum creatinine as an assessment of GFR in neonates have been attributed to effects of maternal load, tubulal secretion of creatinine, passive back diffusion of creatinine through tubules particularly in preterm infants. The serum creatinine concentration immediately after birth reflects the maternal creatinine concentration, neonatal muscle mass and GFR at the time of delivery. There is thus a practical need for an easy alternative to plasma creatinine, which would be more specific-sensitive and reliable form the analytical and clinical viewpoint. These tests include cystatin C, beta trace protein, NGAL,... Cystatin C is a nonglycosylated protein produced in all nucleated cells with a constant rate. Cystatin C is freely filtrated by glomeruli and is catabolized by proximal tubules without any tubular secretion. In neonates the cys C concentrations are not influenced by gender, weight, height gestational age and serum bilirubin level. Moreover the cystatin C doesn’t cross through the placenta and neonatal cystatin C concentrations does not correlate with maternal cystatin C level. In contrast to creatinine, the cystatin C protein is not reabsorbed by immature kidney. Some authors believe that cystatin C is a better marker of GFR than creatinine in neonates and pediatric age group because it mirrors the maturation of renal function more closely. In this study we measured serum cystatin C and creatinine in 50 neonates. We used several equations for estimation of GFR by serum cystatin C and compared the GFR calculated by these equations with creatinine based GFR.

Keywords: neonate, renal function test, cystatin C, creatinine

The renal stone in children

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Iran is considered as a country with high prevalence of nephrolithiasis among children, hence we decided to study the epidemiology and etiology of this common disease in Iran. In this cross sectional study, 177 children with nephrolithiasis from 1382 till 1388 were evaluated. The mean age was 6.91 (S.D: 6.15) years. Male to female ratio was 1.88/1. Abdominal pain (51.9%) and hematuria (37.5%) were the main clinical presentation. FTT (5.1%) and UTI (36.4%) were other clinical manifestations. Of the 177 patients 46% had a family history of stones. RTA was seen in 6.5% of cases. Hypercalciuria (6.5%) and hyperoxaluria (5.1%) were the main metabolic disorders among our patients. 63% of patients had opaque stones. Nephrocalcinosis was observed in 5% of our patients. Stage horn stone was in 12.7%. 50% of patients treated only medically. Chronic renal failure and end stage renal disease was seen 3% and 1.5% of patients respectively.

Keywords: stone, children, epidemiology

The Incidence and Outcome of Focal Segmental Glomerulosclerosis in Iranian Children

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Focal segmental glomerular sclerosis (FSGS) is a cause of nephrotic syndrome characterized by proteinuria with and without renal insufficiency in children. The incidence of FSGS among children diagnosed with nephrotic syndrome has been increased in recent years in some studies. In contrast, some other studies did not show any increase in the incidence of FSGS. Since ethnicity seems to play an important role in the incidence of FSGS, multiple studies in different races should be done to determine disease frequency in varying demographics. To date, there has been no study of the outcome in children with FSGS and also its
frequency over several decades in Iran. In this study, we determined the frequency of FSGS in children who underwent renal biopsy over several decades in a major pediatric nephrology center in Tehran, Iran. Other aims of this study were to analyze clinical characteristics, course, resistance to steroid and other immunosuppressive medications and outcome of Iranian children with primary FSGS referred to Ali Asghar Children Hospital for the first time in Iran. Between 1982 and 2009, 716 renal biopsies were performed in Aliasghar children hospital. Eighty four children diagnosed as FSGS at Ali Asghar Children Hospital. In three periods of time we evaluated the incidence of FSGS in patients who underwent renal biopsy. Between 1982 and 1990 the incidence of FSGS was 10.1, between 1991 and 2000, this incidence was 9.2%. After 2000 the incidence of FSGS increased significantly and reached 20.5% (Pv=0.001). In our study, fifty eight patients with FSGS were followed for the mean of 5.7 yr (range: 3 months-20 years). Among these 58 cases, initial steroid resistance was seen in 47 (81.3%) and late resistance in 5 patients (8.6%). In contrast, two patients completely recovered and responded to steroid without any recurrence. Other four patients responded to steroid but suffered the recurrence. The clinical status at last visit in patients with initial and/or late steroid resistance showed that 14 patients (26.9%) gained complete recovery, while 32 patients (61.5%) were resistant to immunosuppressive drugs other than steroid. In 14 patients who recovered, nine patients responded completely to CsA. Four patients had recurrence on CsA therapy and responded completely to the combination of CsA and MMF. Additionally, one patient received MMF only and recovered. Twenty patients (20/58; 34.4%) progressed to ESRD in a mean time of 4.9 yr (range: 3 mo-12 yr) and the mean survival time of 11.45 [Standard Error (SEM) = 1.34] yr. The kidney survival rates were 90.4%, 69% and 47% at 1, 5 and 10 years of follow-up, respectively.

**Keywords:** FSGS, outcome, incidence, children

**Genetic in congenital nephritic syndrome**

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Congenital nephritic syndrome (CNS) is defined as proteinuria manifesting in the first 3 months of life. NS appearing later during the first year (4-12 months) is defined infantile, and NS manifesting thereafter is called childhood NS. Primary CNS is typically caused by mutations in genes encoding for components of the glomerular filtration barrier. The classical form is the Finnish type of CNS (CNF), which is caused by mutations in the nephrin gene (NPHS1) leading to massive proteinuria, hypoproteinemia and edema in the newborn period. Other known genes causing CNS are podocin gene (NPHS2), wilms’ tumor factor 1 gene (WT1), laminin β2- gene (LAMB2), and PLCe1 gene (PLCe1). Glomerular Filtration Barrier: This barrier is located in the glomerular capillary wall and comprises three layers: fenestrated endothelium, glomerular basement membrane (GBM), and epithelial cell (podocyte) layer with distal foot processes and interposed slit diaphragms (SDs). The GBM is a protein network formed by type IV collagen, laminin, nidogen and negatively charged proteoglycans. The role of GBM in glomerular permselectivity has been debated, but it is now known that a primary defect in a GBM component results in heavy proteinuria. Podocytes and SD are, however, even more important in preventing proteinuria that the GBM. The glomerular basement membrane also depicts the five important molecules in the pathogenesis of CNS. WT1 is a transcription factor important for podocyte functions. PLC1 is cytoplasmic enzyme involved in cell signaling. Nephrin is a major component of SD and podocin is an adapter and scaffold protein for the SD components. Laminin is an important component of GBM. CNF originally denoted to a severe form congenital nephritic syndrome typically seen in the Finnish newborns. The disease (also called as NPHS1) is highly enriched in Finland, the incidence being 1:8,200 live births. However, patients are reported all over the world among various ethnic groups.

**Genetics:** CNF is inherited as an autosomal-recessive trait. The NPHS1 gene is located to chromosome 19q13.1 and exon sequencing analyses have revealed two important mutations in over 90% of the Finnish patients (Fin-major and Finminor). Both mutations result in a stop codon and a truncated nephrin protein not expressed in SD. Several reports on NPHS1 mutations in non-Finnish patients have been published. The patients come from Europe, North America, North Africa, Middle East and Asia. Most non-Finns have individual mutations including deletions, insertions, nonsense, missense, and splicing mutations spanning over the whole gene. Missense mutations are all located within the extracellular part, and clustering to. Congenital Nephrotic Syndrome 25603 exons coding for the Ig-like motif two, four and seven have been reported. The Fin-major and Fin-minor mutations are rare outside Finland, but enrichment of other mutations has been reported also in non-Finns. In Mennonites, 1481delC mutation is common and leads to a truncated protein of 547 residues. On the other hand, a homozygous nonsense mutation R1160X in exon 27 has been found in all Maltese cases. Importantly, six of the 16 cases with this mutation had an atypically mild disease. The same mutation has been reported in six French patients and two of them had a mild disease.

**Podocin Gene Mutations Genetics:** Mutations in the NPHS2 gene, encoding for a podocyte protein podocin, are a common cause of a steroid resistant NS (SRNS) in children and adults, accounting for up to 28% of sporadic and over 40% of familial cases of SRNS manifesting at various ages. The podocin gene mutations, however, are also an important cause of CNS. In a recent report, recessive NPHS2 mutations accounted for half of the CNS cases in 80 European families, while NPHS1 mutations were responsible for only one third of the cases.
Validation of Persian version of PedsQLTM end stage renal disease module version 3 in children under 18 years


Background: to evaluate the quality of life in Iranian children with ESRD, we validate the Farsi version of PedsQLTM End Stage Renal Disease Module version 3 in children with ESRD in Isfahan (2008-2010).

Method: To evaluate the reliability, validity and feasibility of the Farsi version of PedsQL TM ESRD module version 3 in children with ESRD ages 2-18 we followed the translation methodology proposed by developer (Forward-backward translation). A sample of 25 children with ESRD ages 2-18 (chosen by census method from patients come to Alzahra hospital in Isfahan 2009-2010) and their parents completed the questionnaires. The data after collection analyzed by SPSS 18.0 soft ware.

Results: In the Farsi version of PedsQL TM End Stage Renal Disease Module version 3 Cronbach Alpha's internal consistency values was 0.82 for children self reports and 0.88 for proxy reports. Constructive, criterion face and content validity were good. Missing items were less than 5%. The Farsi version of PedsQL TM End Stage Renal Disease Module version 3 was feasible.

Conclusion: Results show The Farsi version of PedsQL TM End Stage Renal Disease Module version 3 is suitable for pediatric health researches children with ESRD and their parents.

Keywords: quality of life, ESRD, children

Renal calculi due to cystinuria in children

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Background: Cystinuria is a rare autosomal recessive disorder of proximal tubular amino acid transport, characterized by excessive urinary excretion of cystine and dibasic amino acids lysine, arginine, and ornithine. Its overall prevalence is 1/7000-1/15000, with an estimated gene frequency of 1/100. Cystinuria results in considerable morbidity as recurrent stone formation. Its incidence in pediatric urolithiasis has been reported from 1% to 12.9% in different studies. First line therapy consists of high oral fluid intake and a low salt diet with urinary alkalinization to a PH of 7. Sulphhydryl agents such as tiopronin, D-penicilamin can be added when stone formation is not well controlled. The aim of this study was to investigate the role of cystinuria in pediatric urolithiasis in our institution.

Method: Medical records of 25 children with cystine calculi who referred to Children’s Hospital of Tabriz from 1999 to 2010 were studied retrospectively. Cystinuria was confirmed by measurement of cystine in urine, urine nitroprusside test or stone analysis.

Results: From 356 children with urolithiasis 25 children had Cystinuria (7%). Male to female ratio was 14/11. Eighteen patients (72%) had bilateral and 28% had unilateral kidney stones. Mean stone diameter was 11.4±5.9 mm (3-23 mm). Consanguinity was found in 16 (64%) patients. Four patients (16%) presented with acute renal failure due to bilateral urinary obstruction. Staghorn stone and pyonephrosis detected each in one patient. Twelve patients (48%) experienced at least one episode of urinary tract infection. All patients received conservative management with hydration and urine alkalinization with potassium citrate. Captopril was added to treatment in 6 (24%) patients. ESWL and surgical interventions were performed in 7 (28%) and 14 (56%) patients respectively. DMSA scan revealed unilateral scarred kidney in 5 patients (20%). All patients had normal serum creatinine level at the end of follow up period.

Conclusion: The incidence and clinical course of cystinuria in this study was similar to literature. Even with good medical management long-term outcome may be poor due to insufficient efficiency and low compliance. All children with urolithiasis need complete metabolic evaluation for cystinuria. Key words: Urolithiasis, Cystinuria, Children

Keywords: Urolithiasis, Cystinuria, Children