Oral / Poster Presentation

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Tuesday Oral Presentations

Tues- O1

Genetic Aspects of Distal Renal Tubular Acidosis (dRTA) Update

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Introduction: Distal renal tubular acidosis (dRTA) is characterized by impaired renal H+ secretion resulting in metabolic acidosis. In Autosomal recessive dRTA, a substantial fraction of patients have progressive bilateral sensorineural hearing loss. This coexistence is due to the mutations of a gene expressed both in the intercalated cells of the kidney and the apical membrane of the interdental cells and the epithelium of endolymphatic sac in cochlea. The gene ATP6V1B1, is responsible for encoding β subunit of the H^+-ATPase pump which plays an important role in the maintenance of acid- base homeostasis and endolymphatic acid secretion.
The aim of this study was to assess the mutations in ATP6V1B1 gene in our patients with dRTA.
Materials & Methods: In this study 52 children admitted with the diagnosis of RTA at nephrology clinic were evaluated. Diagnosis of dRTA was based on clinical manifestations and detection of normal anion gap metabolic acidosis, hypokalemia and urine PH which was never under 5.5 and positive urinary anion gap. Audiometry was performed in all patients with dRTA who were cooperative and evoked potential in infants for hearing evaluation. Sequencing of the ATP6V1B1 gene was performed in patients with dRTA and sensorineural hearing loss. Statistical analysis was performed using Excel software by Student t test and differences with P value lower than 0.05 were considered significant.
Results: Twenty eight patients (53.8%) had dRTA. Fifty one percent were under the age of one and 49% were between 1-12 years. Sixteen patients (57%) were male and 12 (43%) female. Twelve patients (42%) had bilateral sensorineural hearing loss consisting 6 of 15 boys (40%) and 6 of 12 girls (50%). There was no correlation between hearing loss and the gender (P= 0.38). Four patients with hearing loss had mutation in ATP6V1B1 gene (14% of patients with dRTA and 33% of patients with dRTA and hearing loss).
Conclusions: This study indicated that significant percentage of the children with dRTA had sensorineural hearing loss and mutation in ATP6V1B1 gene. It is recommended to perform audiometry in all children with dRTA.

Tues - O2

Causes of Fanconi Syndrome in Children's Hospital of Tabriz

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Introduction: Fanconi syndrome (FS) is a generalized dysfunction of renal proximal tubules leading to urinary wasting of glucose, amino acids, bicarbonate, phosphate and other solutes. The causes of FS are divided into: hereditary, acquired and exogenous substances. The aim of this study was to detect the causes of FS in children admitted in Children's Hospital of Tabriz.
Materials & Methods: In a descriptive and cross sectional study medical documents of all children who admitted in Children's Hospital of Tabriz from 2000-2010 with diagnosis of FS were studied. Demographic characteristics and clinical and laboratory findings were gathered in designed form.
Results: During ten years 39 patients with FS were diagnosed. Age of patients at the diagnosis was 2 months- 9 years (mean: 23.6± 12.7 months). Twenty three patients were male (58.9%) and 16

patients (41%) were female. Consanguinity was present in 23 (64%) cases. The causes of FS in a descending order were: cystinosis in 14 patients (36%) , tyrosinemia in 5 (12.8%), glycogen storage disease type I (Von-Gierke) in 4 (10%), galactosemia in 3 (7.6%), chemotherapy with cisplatin in 3 (7.6%), Fanconi-Bickel syndrome in 2 (5%), idiopathic FS in 2 (5%), nephritic syndrome in 2 (5%), Wilson disease in 1 (2.5%), Dent disease in 1 (2.5%), intoxication with heavy metals (mercury) in 1 (2.5%), and side effect of Na-valproate in 1 (2.5%) patient. Failure to thrive was observed in all patients with hereditary FS and in 89.7% of all patients. Nine patients (23%) progressed to end stage renal failure and 7 patients (17.9%) died.

Conclusions: In our area hereditary FS is more common than acquired forms and among them cystinosis is the most common cause with a poor prognosis.

Tues -O3

Plasma Neutrophil Gelatinase-associatde Lipocalin (NGAL) in Infants With and Without Sepsis Who Admitted in Ali-Asghar Hospital during 2011

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Introduction: Neutrophil gelatinase-associated lipocalin (NGAL) is an inflammatory maker of cell injury which increases significantly in situations such as ischemic damage, cytokine release, and sepsis. There are different studies about the relationship between serum levels and urinary levels of NGAL and acute kidney injury (AKI) and sepsis in adult and pediatric population. The aim of this study is to evaluate plasma NGAL levels in the diagnosis of neonatal sepsis.

Materials & Methods: In this study, 120 neonates who admitted in NICU of Ali-Asghar hospital included in the study. They were divided in two group; neonates with sepsis (n=52) and control group (n=68). Serum levels of NGAL in the two groups (cases/controls) were evaluated on admission, using (NGAL Rapid ELIZA Kit kit 037) and were measured by ELISA. NGAL levels in both groups were compared. Analysis for the ROC cut off level of serum NGAL in diagnosis neonatal sepsis and its diagnostic accuracy was calculated.

Results: 120 infants were examined in this study (56.7% male) and mean gestational age of them was 36.7±3.7 weeks (range 26 to 40 weeks). Mean NGAL plasma level was significantly higher in patients with sepsis than controls (102.9±65.9 vs. 29.2±13.3 ng/ml; P= 0.0001). Plasma NGAL was significantly higher in male infants than female infants (respectively, 73.5±68.7 vs. 45.1±32.3 ng/ml; P= 0.007) and also it has positive relationship with the duration of hospital stay (r=0.640, P=0.0001). In the cut-off point of 48 ng/ml, plasma NGAL had 92% sensitivity, 91% specificity, and accuracy of 80%, which it seems to be the most appropriate cut-off point of plasma NGAL for diagnosis of neonatal sepsis.

Conclusions: Regarding to high sensitivity, specificity and accuracy of NGAL plasma levels, it seems a good biomarker for early diagnosis of neonatal sepsis.

Tues -O4

Assessment of Prognostic Factors in Children RTA Type IV Overwhelmed By Bilateral Obstructive Uropathy.

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Introduction: Hyperkalemic (type IV) renal tubular acidosis occurs secondary to impaired renal responsiveness to Aldosterone. This can occur chronically in infants and children with a history of obstructive uropathy. The aim of this study is to assess contributory factors in improving conditions like hyperkalemia and metabolic acidosis as well as growth status in children suffering from bilateral obstructive uropathy and RTA type IV.

Materials & Methods: In this study, we recruited and observed 48 patients affected with both bilateral obstructive uropathy at urinary bladder outlet and RTA type IV for two years, during, children's growth, sonographic data, renal function and serum electrolytes underwent serial assessment and in case of clinical indication, the patients were treated with drugs like citrate sodium and Kayexalate. Noteworthy patient's death resulted in exclusion from the study.

Results: Frequent urinary tract infection (p=0.0011), infants and children with abnormal <20 weeks gestational sonography results like bilateral hydronephrosis (p=0.00001), birth weight below 2500 gr (LBW) (p=0.0014), preterm delivery (p=0.001), maternal age below 20 years (p=0.0018), pregnancy more than two times (p=0.004), admission due to respiratory problems during infancy period (p=0.003) were discretely accompanied with no disease convalescence while other factors were associated with no significant difference.

Conclusions: Regarding the paper results, it seems logic to consider abortion in case of renal hydronephrosis and dysplasia in gestational age below 20 weeks. Moreover, medical care during pregnancy for a term delivery with suitable weight (resulting in better maturation of lung parenchyma and precluding hypoxia related renal injuries) prevents

Tues -O5

Distal Renal Tubular Acidosis: From Clinics to Molecular Mechanisms

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Systemic acid-base homeostasis is the product of complex interactions between metabolism, regulated exhalation of CO_2 by the lungs, and acid or base excretion by the kidneys. The importance of renal acid-base transport has been highlighted by mutations identified in several proteins involved in this task in patients with inherited forms of renal tubular acidosis. In humans, distal renal tubular acidosis (dRTA) type I can be caused by mutations in the anion exchanger AE1 (SLC4A1) and the B1 and a4 subunits of the V-type H^+-ATPase. Type I dRTA is usually characterized by hypokalemic hyperchloremic metabolic acidosis. To better understand the pathophysiology of dRTA and the underlying mechanisms of defective distal urinary acidification we have generated two different mouse models that are genetically altered for the *AE1* and *B1* subunit genes. Furthermore, we developed several rat models to study mechanisms contributing to acquired forms of dRTA.

Tues -O6

Evaluation of Effect of Heparin Sodium a Diluting Factor on Blood Gas Analysis Outcomes

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Introduction: Sodium heparin solution is widely used as an anticoagulant in blood gas analysis. Although alterations in blood gas measurements may occur when small sample volumes are diluted by heparin. So we aimed to determine correlation between heparin diluted status and Blood gas analyses.

Materials & Methods: A cross sectional study was conducted on 100 pediatric ward admitted patients. We take two different samples by similar syringes, one contained 5% extra heparin sodium. Both blood samples were analyzed and compared together.

Results: Our results shown that heparin contained samples had wrong blood gas analyses reports which acidotic axis may had shown imbalance cause of H^+ imbalance.

Tues -O7

Fanconi-Bickel Syndrome Versus Osteogenesis Imperfecta: An Iranian Case With A Novel Mutation In Glucose Transporter 2 Gene, And Review Of Literature

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Case report: Fanconi-Bickel syndrome is an extremely rare hereditary metabolic disease, characterized by hepatomegaly due to glycogen storage, refractory hypophosphatemic rickets, marked growth retardation and proximal renal

tubular acidosis. Recurrent bone fractures are one of the hallmark findings. It is a single gene disorder; the responsible gene belongs to the facilitative glucose transporters 2 (GLUT2) family gene or (SLC2A2) mapped to the q26.1-26.3 locus on chromosome 3, and encodes the GLUT protein 2. This protein is expressed in pancreatic ί-cells, hepatocytes, renal tubules, and intestinal mucosa. Several mutations in the GLUT2 gene have been reported in different ethnicities. Herein we report an Iranian girl with a missed diagnosis of osteogenesis imperfecta. She was referred with the history of frequent fractures, and severe motor delay and was suspected to osteogenesis imperfecta. Following the case we detected refractory rickets instead of OI, sever growth failure, proximal renal tubulopathy and RTA, and enlarged kidneys, progressive hepatomegaly, and GSD on liver biopsy. Glucose and galactose tolerance tests confirmed abnormal carbohydrate metabolism. Molecular analysis on GLUT2 gene revealed a homozygous novel mutation in exon 5; it was 15 nucleotide deletion and 7 nucleotide insertion and caused a frame shift mutation, produced a premature truncated protein (P.A229QFsX19). This mutation has not been reported before in the relevant literature.

Tues - O9

Phenotype and Outcome of Bartter Syndrome in Iranian Children: Single Center Experience.

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Introduction: Bartter syndrome is a rare inherited tubulopathy that is characterized by renal salt wasting, hypokalemic metabolic alkalosis, normal blood pressure despite of high blood level of renin and aldosterone. Here, we describe the phenotype- based classification of children diagnosed as bartter syndrome and the outcome of these cases.

Materials & Methods: This is a retrospective study. The data of all of the patients recorded as ICD E268, admitted in pediatric nephrology department between 2004 and 2013, were reviewed. The inclusion criteria were: 1- hypochloremic metabolic alkalosis, 2- normal blood pressure, 3- high urine chloride (>20 mEq/L), 4- high renin and aldosterone. Demographic data, laboratory tests, and the outcome were recorded.

Results: From 23 children who had ICD E268 code, 19 children (10 males,9 females) fulfilled the criteria. The mean (range) age of presentation was 3.2 m (birth-12 m), the mean birth weight was 2.7 kg (1.5-4.5 kg), and the mean gestational age was 33.14 wks (28-38 wks). The most common manifestations were prematurity (83.3%), polyhydraminios (78%), growth retardation and failure to thrive (94.7%), delayed cognitive development (31.6%), polyuria/ polydipsia or dehydration episodes (94%), muscle weakness or cramps (17.6%), nephroclacinosis (22%), sensorineural deafness (16.7%), temporarily high blood pressure in one. Consanguinity was reported in 58% and affected siblings were in 39%. The classifications of patients according to their phenotype were: aBS-I-II (58%), cBS-III (21%), SND-IV (16%), CaSR-V (5%). The patients were followed for mean 7.95 years (range: 9 m – 17 yrs). One (6%) died of recurrent sepsis, one (6%) fully recovered with no need to medication while his brother with the same diagnosis led to ESRD. Two (12%) cases were transplanted, and the others have glomerular filtration rate between 60-90 ml/min/1.73 m^2. The observed co-morbidities were hyperlipidemia and obesity in two, severe varicose in leg and tight and gynecomastia in one, tonic-colonic generalized seizure and spasticity in one.

Conclusions: Bartter syndrome is more prevalent due to consanguinity marriage in Iran. However it can be classified clinically, but genetic analysis is more powerful for early diagnosis and start proper treatment.

Tues - O10

Relationship between Serum Level of Calcium and Urinary Levels of Calcium, Citrate and Urine Ph in Children with Renal Stones

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Introduction: Prevalence of urolithiasis in childhood is increasing. Many children with stone disease have a metabolic abnormality. Hypercalciuria and hypocitraturia are considered

the most important risk factors for urolithiasis. The main aims of this study were to assess serum level of Ca, calciuria, citraturia and urinary pH and to determine whether urinary Ca concentration is a helpful biomarker in metabolic evaluation of children with renal stone.

Materials & Methods: This cohort study was performed at Qom University of Medical Sciences on 100 pediatric patients with documented urolithiasis. We collected 24-h urine samples from 100 stone-forming children and adolescents with hypocitraturia and from 121 healthy controls. Urinary calcium, pH, citrate, and oxalate were assessed and compared between the two groups. Data were analyzed using SPSS-13 software.

Results: Patient's age was ranged from 20 days to 14y/o with mean age of 3.32+2.53 years; 54 patients were male (54%) and 46 patients were female (46%).In both stone-formers and controls, hypercalciura was inversely related to citraturia and urinary pH. Metabolic disorder was detected in 95% of patients; the most prevalent urine metabolic abnormalities were hypocitraturia (56.8%), hypercalciuria (29.4%), hyperuricosuria (26.3%), hyperoxaluria (14.7%), phosphaturia (8.4%) and cystinuria (6.3%).

Conclusions: This study similar to other studies in Iran have shown that the prevalence of hypercalciuria is significantly higher compare to other countries, it may be associated with excessive intake of calcium and sodium. Compared to controls, stone-formers with hypocitraturia demonstrated a higher urinary Ca concentration, but this was proportional to calciuria. However, the Ca/Citrate ratio may be a useful clinical tool in evaluating children with urolithiasis.

Tues -O11

Normal Ca/Cr, Na/Cr and K/Cr Ratio in Healthy Adolescents (North Of Iran)

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Introduction: Calcium is one of the most important ions for normal function of many organs. But with increase urinary excretion of calcium (hypercalciuria), the risk of renal stone may be increased. Due to difficulty of obtaining a 24h urine (especially in children), a random urine calcium sample is recommended to detect hypercalciuria. However, recent studies have shown that the urinary calcium/creatinine ratio varies with age and geographic areas. So, the purpose of this study was to determine normal values of urinary calcium to creatinine ratio in healthy adolescents' children.

Materials & Methods: 480 children aged 12 to 14 years were randomly selected from middle school in Babol (north of Iran) and early morning urine samples of them were studied for determine normal urine Ca / Cr, Na /Cr and K/Cr ratios. Children with family history of renal disease were excluded from this study.

Results: In our study, 50% and 95% of urinary Ca/Cr ratio were 0.078 $\pm$0.025 and 0.13 mg/mg in the group. The mean of urinary Ca/Cr ratio in boys and girls were 0.079 $\pm$0.028 and 0.077 $\pm$0/022, respectively. The mean of urinary Na/Cr ratio in boys and girls were 1.39 $\pm$0.48 and 1/21 $\pm$0/33 respectively. The mean of urinary K/Cr ratio in boys and girls were 0.30$\pm$0.11 and 0.29 $\pm$0.10, respectively.

Conclusions: This study was shown that urinary Ca/Cr ratio of these children is different from other geographic areas. Also, a direct relationship was seen between urinary Ca/Cr ratio, Na/Cr and k/Cr ratios.

Tues -O12

Hypocitraturia as Second Cause of Infantile Renal Stone in South-West of Iran

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Introduction: Renal stones in infants are relatively infrequent but its incidence has increased in recent decades. The aim of this study was to investigate the clinical presentation, metabolic risk factors, and urinary tract abnormalities in infants suffering from renal stone.

Materials & Methods: All of 152 infants that were admitted in our department between 2009 and 2012 with ultrasonography proven urolithiasis were included. Unlike previous studies we used foley catheter for accurately collecting of the urine. Ultimately 24-h urine samples were analyzed for calcium, citrate, oxalate, uric acid and magnesium. For detecting cystinuria, qualitative measurement of urinary cystine was done by

nitroprusside test. Also, urinary tract structural abnormalities were evaluated in these patients.
Results: The average age at diagnosis of stone was 5.46 months (range, 15 days-12 months). The most common clinical findings in our patients were restlessness and urinary tract infection UTI. Family history of kidney stone found in 67.1% of patients and 68.4% of them were born from consanguineous marriages. Metabolic abnormalities and urinary tract abnormalities were found in 96.1% and 15.1% of children respectively. The most common metabolic risk factors were hypercalciuria (79.6%) and hypocitraturia (40.9%). Hypocitraturia was significantly more common in hypercalciuric and normal uric acid excreted infants (69.4%, *p* value=0.002 and 77.4%, p=0.018 respectively). Hyperoxaluria and hypomagnesuria were found in about 28% of patients and both of them were significantly associated with bilateral urolithiasis (*p*=0.006 and *p*=0.005 respectively). Cystinuria was detected in 3.3% of patients of which 60% were bilateral. Urinary tract abnormalities were diagnosed in 15.1% of patients and was significantly more common in girls (*p*=0.001).
Conclusions: Our results showed that urinary metabolic abnormalities especially hypercalciuria and hypocitraturia are very common in infants with urolithiasis; so appropriate evaluation of urinary metabolic parameters can lead us to proper diagnosis and treatment.

Tues -O13

Relation between symptomatic IHC with UTI in patients attending to Children Hospital of Bandarabbas

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Introduction: Urinary tract infection (UTI) is known as one of the most frequent diseases in pediatric medicine. A new addition risk factor of predisposing patients to UTI is mentioned as idiopathic hypercalciuria (IHC). IHC is defined as a hypercalciuric status with normal level of calcium in serum and absence of any secondary cause of hypercalciuria. In this study our aim was to evaluate relation between symptomatic IHC with UTI in children.
Materials & Methods: This prospective case-control study includes 251 patients who were presented to outpatient clinic of Children hospital of Bandar Abbas. Patients divided in two groups. Case group contains 182 patients with proven UTI and control group contains 69 patients without any signs or symptoms of UTI. Mid-stream urine sample to determine Ca/Cr ratio collected from all patients. Twenty four hours urine collection was ordered for patients with Ca/Cr ration more than 0.21mg/mg. Diagnosis of IHC was based on Ca/Cr ratio more than 4 mg/mg/kg in 24 hours with no secondary cause of IHC.
Results: We evaluated association between IHC and UTI in 251 patients. In case group after collecting 24 hours urine, 66 (36.3%) patients and in control group 3 (4.3%) patients detected as IHC. We discovered that frequency of hypercalciuria is higher in female (66.6%) than male (34.4%) and IHC was found significantly more in patients with UTI (P=0.001); but we did not found any evidence to prove relation between recurrent UTI and IHC (P=0.64). Our findings show, the most common manifestations of IHC were dysuria, frequency and hematuria.
Conclusions: On the basis of these results, we confirmed the hypothesis that IHC is an important risk factor of UTI (P=0.001) but there was no significant association between recurrent UTI and IHC.

Tues -O14

Evaluation of Predisposing Factors for Nephrolithiasis and Renal Impairment in Patients with Cystic Fibrosis: A preliminary data.

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Introduction: Patients with cystic fibrosis (CF) are frequently at risks for nephrolithiasis or renal function impairment .For investigation of this complication; renal function tests and metabolic evaluation were performed in these patients.
Materials & Methods: Twenty CF patients (10 male, 10 female), mean age 12.3(5-22) years were

enrolled. All patients underwent renal ultrasonography (US) and special blood and 24 hours urinary evaluations.

Results: No patient had urolithiasis on US. The mean GFR (Schwartz) was 77.2(range 45.33-143.24 ml/min/1.73m2) and 6/20 patients had GFR<60 ml/min/1.73m2. Hypercalciuria was detected in 8 patients (range1.05-8.05 mg/kg 24h).Hyperuricosuria, hypocitraturia and hyperoxaluria were found in 13(range 3.03-29.25 mg/kg/24h), 11(range 0.23-1.28 mmol/1.73m2/24h) and 2 patients, respectively. Mild proteinuria was observed in 12/20 patients and no one showed proteinuria on urinalysis.

Conclusions: There was low level of renal impairment and low molecular weight proteinuria in CF patients. Hypercalciuria, hyperuricosuria, hypocitraturia and hyperoxaluria might lead to the higher risk of urolithiasis in patients with CF.

Tues -O15

Clinical Features and Outcome in Children with Primary Hyperoxaluria: Single Center Experience

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Introduction: Primary hyperoxaluria (PHO) consists of enzyme defect of glyoxylate metabolism including AGT and GRHPR .Type 1 is more prevalent than type II or non-type. The aim of this study was to present the clinical presentation and outcome of children with the diagnosis of PHO.

Materials & Methods: In this retrospective study, we reviewed data of all patients with diagnosis of PHO who admitted between 2001 and 2012.The criteria of diagnosis were: Recurrent / bilateral/ or multiple urolithiasis; Urine oxalate (mmol/L) to urine creatinin (mmol/L) more than percentile of 95 or 24 hour urine oxalate more than 0.5mmol/1.73/m^2; History of calcium oxalate renal stone in relatives or consanguinity of parents; with or without severe chronic kidney disease.

Results: From all of patients admitted with the diagnosis of urolithiasis or renal failure, 18patients (12 Females, 6 Males) diagnosed as PHO. The mean age of diagnosis was 4.36 years (range 3 m-13 yrs). The clinical presentations were urolithiasis (n=12) and severe chronic kidney disease (n=6). Five out of six had bilateral nephrocalcinosis and the last one diagnosed by finding oxalate crystals in renal necropsy. The ranges of creatinine at presentation were 0.3 mg/dl to 10.6 mg/dl. From 12 children with urolithiasis, 75% were multiple and 83.3% were bilateral. Failure to thrive was in 44%. Family history was positive for either urolithiasis or hyperoxaluria in 57% and the consanguinity of parents reported in 67%. The patients were followed up for seven years, seven died on CAPD or HD, one is still on HD, four have moderate CKD (creatinin 1.3 to 2.4 mg/dl), three have normal GFR, and three were lost to follow up.

Conclusions: The outcome of PHO is poor, high percentage of presentation with renal failure prompt to launch accurate genetic or enzymatic diagnosis in our country to make earlier precise decision for appropriate intensive treatment.

Tues -O16

Neurologic Disorders in Children with Cystinosis

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Introduction: Cystinosis is a metabolic disease characterized by an accumulation of cystine in different organs and tissues, leading to potentially severe organ dysfunction. Three forms of cystinosis have been described: the infantile (nephropathic) form, the intermediate (adolescent, late-onset) form; and the adult (benign) form. Cystinosis is transmitted as an autosomal recessive trait. The gene for nephropathic cystinosis has been mapped to chromosome 17p13 and identified. Now, patients with cystinosis have survived through their fifth decade, but the unremitting accumulation of cystine has created significant non-renal morbidity and mortality. In this present study we investigate neurologic disorders in children with cystinosis.

Materials & Methods: In this descriptive, cross sectional study, we evaluated 40 patients with cystinosis referred to neurology clinic in Ali-asghar children hospital between 2008 and 2013. Data was recorded on age, sex, and neurologic disorders.

Results: Forty patients with 3-23 years of age had cystinosis. Mean age was 8.65±4.74 years. Twenty cases (50%) were male. Patients had neurologic disorders including seizures (25%), tension type headache (15%), distal muscle weakness (5%), increased ICP (Intracranial pressure) (2.5%), and optic atrophy (2.5%). None of them had swallowing problem. One case died with refractory seizures and increased ICP.
Conclusions: Severe neurologic complications such as cerebral atrophy, encephalopathy, increased ICP and progressive myopathy in patients with cystinosis are rare especially in childhood period, after early treatment with Cysteamin, and renal transplantation. Other neurologic disorders should be evaluated and treated properly.

Tues -O17

The Effect of Phototherapy on Urinary Calcium Excretion Term Neonates with Hyperbilirubinaemia

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Introduction: Phototherapy plays a significant role in the treatment and prevention of hyperbilirubinaemia in neonates. However, this treatment modality may itself result in the development of some complications. A less-known complication of the phototherapy is hypocalcemia. Some studies reported hypocalcemia accompanied by increase in urinary excretion of calcium. The aim of this study was to evaluate the effect of phototherapy on urinary excretion of calcium in term neonates.
Materials & Methods: In a before-after study 80 term icteric newborns undergoing phototherapy were selected through accidental sampling. The survey was approved by the ethics committee of Guilan University and informed consent was given by all parents. Neonates who needed antibiotic therapy or blood exchange were excluded. Indication for phototherapy was a serum bilirubin more than 15mg/dl concentration. Continuous phototherapy was used for treatment. Serum samples for calcium, bilirubin and sodium and urine samples for calcium, sodium and creatinine before and after phototherapy (48 hour) were checked. UCa/UCr ratio (mg/mg) was determined. Hypercalciuria was defined by a ratio >0.85. Paired t-test and Wilcoxon test were used for statistically analysis.
Results: In this study 80 term newborns (46 male and 34 female) with mean age of 7.01±4.13 days (3-26 days), gestational age 38.4±0.54 weeks (38-40 weeks), birth weight 3198±373.2 gr 2500-4420 gr, serum bilirubin 16.54±0.92 mg/dl (15-19 mg/dl) were included. The mean level of serum calcium before and after phototherapy were 9.37mg/ml, 9.25 mg/dl respectively (p>0.05). The difference between pre- and post-phototherapy urine calcium levels were found to be statistically significant (p<0.05). The mean fractional excretion of sodium and mean of QTC before and after phototherapy had no statistical difference (p>0.05).
Conclusions: The result of this study showed that phototherapy may increase urinary calcium excretion in term neonates. Further investigation for clarifying the importance of this phenomenon is recommended. A more complex, controlled study has been designed to solve these questions. In conclusion, phototherapy might increase urinary calcium excretion in infants so a further investigation should be conducted on more newborns.

Tues -O18

The Therapy of Nephropathic Cystinosis

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Supportive treatment: The water intake must be adjusted to diuresis and weight variations. Fluid requirement increases with external temperature and with fever. It is also increased by the required mineral supplements.
Sodium and potassium bicarbonate must be given in order to obtain a plasma bicarbonate level between 21 and 24 mmol/l. This is sometimes difficult and may require large amounts of buffer, up to 10–15 mmol/kg. Hypokalemia requires 4 to 10 mmol/kg potassium supplements in order to maintain serum potassium above 3 mmol/l. Hypophosphatemia must be corrected with a supplement of sodium/potassium phosphate at a dose of 0.3–1 g/day. Excessive phosphorus prescription may lead to nephrocalcinosis. Since tubular 1α□-hydroxylation is diminished in this

disease, it is justified to give 1α- or 1α-25-OHD_3 (0.10–0.50 μg/day), especially in cases of symptomatic rickets. Carnitine is given at a dose of 100 mg/kg per day. Feeding problems may require tube or gastric button feeding. Urinary losses may be reduced by the prescription of indomethacin at a dose of 1.5–3 mg/kg. It has been shown that the angiotensin converting enzyme (ACE) inhibitor, enalapril, diminishes albuminuria and possibly slows down the degradation of renal function.

Hypothyroidism, even if asymptomatic, should be treated with L-thyroxine supplementation. Growth failure is improved by administration of recombinant growth hormone at a dose of 1 U/kg/week.

Specific Therapy: Cysteamine (Cystagon) prevents cystine accumulation in various organs. The dose is progressively increased from 10 to 50 mg/kg per day. The effect, assessed by cystine assay in leukocytes lasts no longer than 6 h. Consequently, it has to be given in 4 separate doses – one every 6 hours . It was recently shown that a twice daily administration of an enteric release formulation of cysteamine bitartrate was as effective as the current formulation of cysteamine. The aim is to keep cystine content under 2, or better, under 1 nmol of 1/2 cystine per mg of protein. The drug should be started as soon as the diagnosis is confirmed. Side effects of the drug include nausea and vomiting and can be managed with omeprazole. Less commonly, allergic rashes, seizures and neutropenia are seen. In addition, cysteamine is responsible for an unpleasant breath odour so that compliance with 4 doses per day is difficult to maintain in the long term, especially in adolescents. Cysteamine eye drops are able to prevent corneal deposits, and may decrease and even suppress the deposits already present.A new therapeutic approach was tested on the animal model using bone marrow cell transplantation with encouraging results

Tuesday Poster Presentations

Tues - P1

Fanconi Syndrome: Review of Diagnosis and Management in Patients- Single Center Report

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Introduction: Fanconi syndrome is a generalized inherited or acquired disturbance of renal tubular transport. The aim of the present study was retrospectively review of the initial clinical feature, laboratory tests disturbance, and management.

Materials & Methods: We retrospectively reviewed 14 patients who admitted in Ali-Asghar children hospital between March 2003 and May 2013. Inclusion criteria were the presence of proximal renal tubular acidosis (normal anion gap hyperchloremic metabolic acidosis, urine pH less than 5.6), glycosuria (positive urine dipstick for glucose), generalized aminoaciduria, normal blood aminoacid chromaography, phosphaturia (fractional excretion of Phosphate more than 25%), and rickets.

Results: From all children with diagnosis of renal tubular acidosis, 14 had Fanconi syndrome, 20 Von –Gierke, Fanconi Bickel in four, galactosemia in 22, and tyrosinemia in 9. Of 14 patient, 8 (57%) were females and 6 (43%) males. The mean age of children was 9.1 years (range: 3 -18 years). The mean age of patients at presentation was 7 months (range: 3-11 months). Consanguinity reported in 12 (85.7%) children. The most frequent manifestations were impaired growth (92.8%), polyuria and polydypsia (78.6%), dehydration (64.3%), and rickets (57.2%). All of the patients had a normal anion gap renal tubular acidosis, aminoaciduria, glycosuria, bicarbonaturia, phosphaturia, hypophosphatemia, and hypokalemia.

All children received citrate solution, fluid, phosphate, and vitamin D.

Conclusion: The low rate of idiopathic Fanconi syndrome urges the evaluation for secondary causes of Fanconi syndrome.

Tues - P2

Pseudohypoaldosteronism in Iranian Children-Case Series

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Introduction: Pseudohypoaldosteronism is a rare disease that characterized by low Na serum and

elevated K serum. We are going to report clinical and laboratory finding of six patients.
Materials & Methods: This is a case series study. The data of children, who were admitted between 2005-2009 in Ali-Asghar children hospital, have been reviewed. Inclusion criteria were hypoaldosteronism and hyperreninemia in the presence of hyponatremia and hyperkalemia.
Demography data, laboratory tests, treatment, and outcome have been recorded.
Results: Six patients (5 males and 1 female) with median age of 2 months (range 10 day-5.5 years) had sufficient criteria. Median body weight at presentation was 3.6 Kg (range 2.95-4.55).. All patients admitted with poor feeding and failure to gain weight. Familial history was positive in only two patients. All of the patients went under treatment with hydrocortisone and standard medical treatment for hyperkalemia. One patient died and other discharged with maintenance medical therapy. Outcome: All patients have been followed for median of 5.5 years and except one, the rest of 5 patients are still coming for follow up in our.

Tues - P3

Barrter Syndrome and Sensorineural Deafness: A Case Report of Type IV Barrter Syndrome

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Introduction: Barrter syndrome type IV is a rare autosomal recessive disease characterized by hypokalemic metabolic alkalosis, severe renal salt wasting and sensorineural hearing loss without nephrocalcinosis. Its pathophysiology is dysfunction of barttin which is an accessory subunit of chloride channels located in thick ascending loop of henle, descending convoluted tubules of kidney and inner ear. Barttin modulates the stability, cell surface localization and function of ClC-K channels. Various mutations of causal gen, BSN, lead to phenotypes of varying severity. We report a case of this disease from Children's Hospital of Tabriz.
Case report: A 50- days old girl with 2 kg weight was admitted in surgery ward with vomiting and abdominal distention with probability of Hirschsprung's disease. She had a history of premature birth at 34 weeks of gestational age. Laboratory tests revealed severe hyponatremia (113 meq/L) and hypokalemic metabolic alkalosis. Serum chloride was 91 and urine chloride was 115 meq/L. Imaging studies for Hirschsprung's disease, renal ultrasonography, and sweet test were normal. In nephrology consult diagnosis of Barrter syndrome was made and she was discharged with K-Cl tablets and Indomethacin capsule. At 1.5 years of old, parents noted hearing difficulty and she underwent insertion of bilateral Ventilation Tube and adenoidectomy with diagnosis of refractory serous otitis media. However, hearing impairment was continued after surgery. Auditory brain stem response (ABR) and Otoacoustic emission (OAE) tests showed a sensorineural deafness and now the patient is a candidate for Cochlear implantation.
Conclusion: All patients of Bartter syndrome should be evaluated for sensorineural deafness in infancy because undiagnosed deafness may have detrimental effect on development of the child.

Tues - P4

Senior-Loken Syndrome, Role of Cilia in Tubulopathies.

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Introduction: Nephronophthisis is an autosomal recessive cystic renal disease and is supposed to be the most common genetic cause of ESRD in first 3 decades of life. This disease can be present in infancy, adolescence (juvenile type) and adulthood. In rare instances it may be associated with extrarenal involvement such as retinitis pigmentosa (Senior-Loken syndrome) and cerebellum (Joubert syndrome). Recently, role of ciliary dysfunction is emphasized in many diseases and syndromes which their manifestations previously seemed to be not related to each other at all. In "Ciliopathies" kidney involvement presents as cystic or dysplastic renal disease resulting in ESRD eventually, eye involvement in form of retinitis pigmentosa can cause blindness, cerebellar involvement causes nystagmus and ataxia and so on. Here we present a case of Senior-Loken

syndrome with ESRD and her brother with only ophthalmic involvement.

Case Report: A 13- year old female with past history of blindness presented with pallor, pain on lower extremities and polydypsia. Blood pressure was normal. Initial laboratory investigation showed: Hb= 7.5 g/dl, BUN=33 mg/dl, and creatinine = 2.8 mg/dl. Urinalysis was normal. Parents were relatives (cousins) and she had younger 6- year old brother with severe visual impairment but his renal function was within normal limit. Ophthalmologic examination of our case was in favor of retinitis pigmentosa. Renal biopsy findings was suggestive of juvenile nephronophthisis with tubular atrophy, thickened tubular basement membrane , interstitial fibrosis and glomerulosclerosis. Because of these clinical and pathological findings she was labeled as a case of Senior -Loken syndrome. During follow- up, her creatinine rose up and she underwent hemodialysis after her family rejected peritoneal dialysis as a renal replacement option and now she is preparing for renal transplantation. Literature search about this case gave valuable information about "Ciliopathies", a group of previously supposed unrelated organ involvement in various syndromes which now could be described as ciliary dysfunction in kidney, eye, ear, adipose tissue, CNS and probably other organs. In kidneys ciliopathy results in abnormal tubular development, cystic and dysplastic changes that may ultimately deteriorate to end stage renal disease.

Conclusions: Multiple organ involvement in a patient with chronic kidney disease should raise suspicion of possible "ciliopathy" syndromes and because of strong probability of autosomal recessive and dominant inheritance, all first degree relatives should be screened and followed for renal involvement.

Tues - P5

Carnitine Deficiency in Chronic Kidney Disease Stage V: Comparing Hemodialysis with Peritoneal Dialysis Subjects

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Introduction: Carnitine deficiency is common in chronic hemodialysis patients. This study aims to define the frequency of Carnitine deficiency with considering the effects of age, gender, duration and modality of dialysis.

Materials & Methods: Forty seven dialysis patients including 20 girls (42.5%) and 27 boys (57.5%), aged 19-300 (166.02 ±76.09) months including 13 (31.7%) CAPD and 28(68.3%) hemodialysis cases enrolled the study. Times from onset of dialysis were 1to128 months (mean: 44.28±31.26SD). The deficiency was defined as serum levels of free carnitine less than 7 µmol/l and plasma levels of Acylcarnitine below 15 µmol/l. Patients divided into two groups: Patients with normal and those with low plasma levels. Chi square and student T tests were used. P values <0.05 were considered as statistically significant differences.

Results: Serum levels of free carnitine were normal in 45(95.7%) and high in 2(4.3%) patients. Total serum carnitine levels were normal in 44(93.6%) and high in 3(6.4%) subjects. Plasma levels of Acylcarnitine were low in 23(48.9%) and normal in 24(51.1%) of enrolled cases. There was no significant difference in frequencies of Acylcarnitine deficiency in hemodialysis versus peritoneal dialysis subjects (P=0.135), girls versus boys (P=0.76), mean ages of patients (P=0.179), and dialysis duration (P=0.126) between groups. Twenty two patients received oral carnitine 250-1000mg/day, while 13 did not receive the drugs. Low plasma levels of Acylcarnitine were reported in 11 and 6 subjects, respectively (P>0.05).

Conclusions: Acylcarnitine deficiency was common in our series. The deficiency was as common in hemodialysis as peritoneal dialysis cases. Although in normal population the deficiency is greater in females; in dialysis patients the deficiency was as common in boys as girls.

Tues - P6

Association Study of TLR4 (Asp299Gly, Thr399Ile) Gene Polymorphisms in the Severity (Amyloidosis) of Familial Mediterranean fever in North-West of Iran

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Introduction: Familial Mediterranean Fever (FMF) is an autosomal recessive auto-inflammatory disorder with more than 60 disease-associated mutations in the responsible gene, *MEFV*. Amyloidosis is the major complication of Familial Mediterranean Fever. A less frequent but most severe complication of FMF is the development of renal amyloidosis, ultimately leading to end-stage kidney failure. Toll-like receptors (TLR) are involved in the activation of an innate immune system. TLR-4(Asp299Gly and Thre399Ile) polymorphisms down-regulate inflammation. We investigated the effect of these polymorphisms on the development or resistance of amyloidosis in FMF patients.

Materials & Methods: In this study we investigated 86 FMF patients (27 with amyloidosis) and 66 matched control subjects. TLR-4(Asp299Gly and Thr399Ile) polymorphisms were analyzed with the polymerase chain reaction restriction fragment length polymorphism method (PCR-RFLP).

Results: The frequency of these polymorphisms were not different in FMF patients (with or without amyloidosis) compared to the control group. Comparison between FMF patients and control subjects revealed no significant association of TLR-4(Asp299Gly and Thre399Ile) polymorphisms, however significant associations between two groups of patients with specific symptoms were observed.

Conclusions: Previous studies suggest the association between TLR-4(Asp299Gly and Thr399Ile) polymorphisms and severity of FMF; However results of present study demonstrated no association between these polymorphisms and frequency of attacks or development of amyloidosis.

Tues – P7

Distal Renal Tubular Acidosis Associated with Hemihypertrophy (Case Report)

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Introduction: Primary distal renal tubular acidosis(dRTA) is an inherited disease characterized by a normal serum anion gap (AG) with hyperchloremic metabolic acidosis (HClMAC.), high urinary pH(UpH), growth retardation, nephrocalcinosis (NC), and renal stone.

Case report: A 6-months boy was referred to pediatric nephrology clinic with history of hemihypertrophy of right side of body and failure to thrive(FTT) and bilateral nephrolithiasis (NL) without NC He was a full term baby with a birth weight of 2.85 Kg and height of 49 cm. His parents were not first degree relatives and family history was negative for renal stone. His physical examination revealed a weight of 4.5 kg and height of 54 cm at age 6 mon. Initial laboratory investigations showed HClMAC. [Cl=119 mmol/L, PH=7.26, HCO_3= 13.7, BE = 11.4mmol/L] with hypokalemia (hypoK), normal serum AG):11meq/L, and positive urine AG: 4 meq/L. Random Uca/Ucr was 1 and 24- hour Uca 5 mg/kg/day. Other hormonal profile such as FT4, TSH, LH, FSH, prolactin, and growth hormone level were normal. Serum phosphate: 5.6 mg/dl, Ca: 8.6 mg/dl, ALKP (350 IU/L), total serum protein:60 mg/dl, Cr:0.4 mg/dl, and UpH: 6.3 . The ultrasound scan of patient revealed NL without NC and normal position of both kidneys.

Conclusions: Results presented with unusual clinical feature (hemihypertrophy) of this disease described so far. Distal RTA is a rare renal disorder characterized by normal AG - HClMAC. and hypoK, UpH consistently above 5.5, hypercalciuria, hypocitraturia, potassium wasting in urine, NL, and NC. Our patient is an atypical presentation of dRTA. Renal acidification defect with hypercalciuria and hypocitrturia are important factors in the pathogenesis of nephrolithiasis Urinary citrate are an inhibitor of crystal aggregation and precipitation. When citrate excretion is reduced, more calcium to be chelated, thus renal stone develops. RTA may be accompanied by clinically hypoK due to renal potassium wasting which leads to impaired growth and FTT. It is important to differentiate primary causes of FTT from secondary causes such as d RTA and also hemihyperthrophy associated with neurofibromatosis and vascular

disease may be an associated finding with other disorders such as d RTA as a rare and new report.

Tues – P8

Novel Mutations in PKD1 Gene in An Iranian Patients Family With Autosomal-Dominant Polycystic Kidney Disease

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Introduction: Autosomal dominant polycystic kidney disease (ADPKD) is one of the most common genetic kidney disorders with the incidence of 1 in 1000 births. ADPKD is genetically heterogeneous, with two genes identified: PKD1 (16p13.3, 46 exons) and PKD2 (4q21, 15 exons). 85% of patients with ADPKD have at least one mutation in the PKD1 gene. Genetic studies have demonstrated an important allelic variability among patients but very few data are known about the genetic variation in Iranian populations.
Materials & Methods: In this study, coding exons analysis of PKD1 by exon direct sequencing was performed in a 7-year old boy with ADPKD and his parent. The patient's father is ADPKD affected without any kidney dysfunction and the patient's mother is congenitally missing one kidney
Results: Molecular genetic testing fund a doubted pathogenic mutation in all 3 members of this family. It was a missense mutation GTG>ATG at position 3057 in exon 25 of PKD1. On the other hand two Novel missense mutations were reported, ACA>GCA found in exon 11 at codon 2241 and CAC>AAC found in exon 38 at codon 3710.
Conclusions: GTG>ATG causes the conversion of amino acids V to M. then for checking the validation of the software reports based on the pathogenicity of this mutation, exon 25 of 50 unrelated normal cases matched by sex and Ethnicity were sequenced. Our findings suggested that this mutation is a polymorphism with high frequency (60%) in the population of south west Iran. The second and third mutations were Novel polymorphisms that changed Threonine to Alanine at codon 2241 and Histidine to Asparagine at codon 3710.

Wednesday Oral Presentations

Wed- O1

Importance of Hereditary Influence of Tubulo-Interstitial Changing in the Diagnosis of Chronic Renal Diseases.

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Introduction: Until the second half of twentieth century, many tubulo-interstitial anomalies have been labeled as chronic pyelonephritis. Progresses in physiology, embryology, molecular biology and related disciplines have demonstrated that many of them depend on the anomaly of embryogenesis and fetal life; perhaps most of these anomalies have a genetic origin. During my practice at Children's Hospital (Tehran), I had the opportunity to meet children suffering from chronic renal diseases, presenting mainly tubulo-interstitial alterations. Some of them presented anomalies of other organs that I introduced those patients as "associated of anomalies over different organs or as a new title".
Materials & Methods: Clinical investigations:- Familial History– Personal History, including embryo-fetal periods. The patients were studied thoroughly for family history, personal presentation, clinical and biological anomalies. Our most relevant investigations concerned, familial history, clinical presentation and course based by fine histo-pathological investigations.
Clinical presentation, laboratory investigations as main example:
-Two brothers, suffering from chronic renal disease, associated with Congenital liver fibrosis, retinitis pigmentosa and cone shaped epiphysis (1)*
-Three infants, with early chronic renal failure, two of them with hepatic fibrosis entitling; Infantile Nephronophthisis!(2)
Results: Genetic disorders take more and more importance in the origin of human health

problems. Many Scholars are pioneers in this field taking part to the discovery of such origin instead of admitting the old terms like chronic pyelonephritis. G.Fanconi, Pierre Royer and Rene'e Habib have been the pioneers in the fields of renal and metabolic disorders and their hereditary origin.

Conclusions: Many renal disorders during childhood have their root in hereditary anomalies. Consanguineous marriages do have some responsibilities for the occurrence of these misfortunes.

Embryo-fetal observation, explorations if necessary are important to prevent the disease or intervene in most protective period.

Wed- O2

The Etiology of Tubulopathy in Iranian Children- Iranian Society of Pediatric Nephrology Collaboration.

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Introduction: To estimate the prevalence of the etiology of tubulopathy in different region of Iran.

Materials & Methods: This is a descriptive study conducted from March 2013 to October 2013.. A table consists of the list of 34 tubulopathy disorders were emailed to 70 members of paediatric nephrology in different cities of Iran and requested to report the number of patients with the specific IC code who admitted between 2006 and 2013 in their hospital. The following IC codes were used: Q61(1,3, 5,8) E72(0,4,5,6,8), E26.8, E83(4,5),E27.8, W25.8, E23.2, E10.8, E74.8, I15.2,N10, N4.9,W20(0,9),N13.7,Q61.3, E83(0,5).

Results: From 31 centres where paediatric nephrologists work, 23 filled out and returned the tables. The most frequent tubulopathies in order were renal tubular acidosis (n=437), fanconi syndrome (secondary in 245, Von-Gierke in 8 and Fanconi Bickel in 35,Lowe syndrome in 4), PCKD (229), other cystic disease (n=145), nephronopthisis (n=130),MSKD(n=102), Bartter syndrome (n=131), Gitelman (n=72), cystinosis (n=130), nephrogenic DI(n=21), Wolfram(n=21), hyperoxaluria(n=31), cystinuria(n=102), TIN (acute in 44, chronic in 20, drug induced in 49, idiopathic in 79). Hypercalciuria in 148, pseuogypoaldostronism in 39, and Gordon in 18. Table 1 shows the distribution of tubulopathy in different geographical division.

Conclusion: However this study included duplicate hospital admission but it would help to organize multicentre studies according the frequency of each tubulopathy in specific geographic region of Iran.

Table 1- Case Summaries

	Geographical division					
	north	south	west	east	center	Total
Nephronophtisis	6	48	21	12	43	130
PCKD	23	34	38	16	118	229
other cystic disease	56	25	14	3	47	145
aminoaciduria	3			12	1	16
Glycosuria	9	8	11	5	4	37
Barrter	7	34	19	27	44	131
Gitelman	2	7	3	1	59	72
hypocacemic hypercalciura familial			20	7	121	148
hypocalcmia AR				3	123	126
Hypomagnesemia					7	7
hypomagnesemic hypercalciuria			1	1		2
hypomagnesemic+hypo calcemia		10	1			11
hypomagensemia mitochondrial		1				1
Liddle syndrome		6	1	1	1	9
GRA				3		3
GordonII		3		1	14	18
pseudohypoaldosteroni sm I		4	1		34	39
Renal Tubular Acidosis	19	80	52	106	177	434
Diabetes Insipidus	15	8	9	6	20	58
Wolfram	1	3	3	7	7	21
Cystinosis	2	21	35	12	60	130
Von Gierke		1	6		1	8
fanconibickel			3		32	35
Secondary fanconi Syndrome	9	28	11	20	177	245
Lowe Syndrome	1	2		1		4
Hyperoxaluria	6	12	10	3		31
Acute tubulointerstitial nephritis	7	17	7	5	8	44
Chronic tubulointerstitial nephritis	3	6	2	8	1	20
Idiopathic Fanconi Syndrome	1	6	3	69		79
drug induced tubulopathy	7	6	30	6		49
medullary sponge kidney	11	2	10	17		40
cystinuria	6	27	30	3	36	102
AD- Hypercalciuria	54	271	42	27	122	516

Wed-O3

Different Aspects of Kidney Function in Well-Controlled Congenital Hypothyroidism

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Introduction: Congenital hypothyroidism (CH) increases the prevalence of kidney and urogenital malformations. There are limited studies considering different aspects of kidney function in well-controlled CH patients. We evaluated some features of kidney function in euthyroid children with CH who have been receiving thyroxin hormone since early life.

Materials & Methods: This cross-sectional study was conducted in Isfahan, Iran, on 74 children aged 2-15 years old (36 CH patients and 38 healthy children). Inclusion criteria for CH patients were euthyroidism at the time of the survey and initiation of replacement therapy during the early neonatal period. Kidney ultrasound evaluation was performed in all participants. Serum biochemistry included urea, creatinine, sodium (Na), potassium (K), magnesium, calcium, and cystatin C levels. Urine electrolytes, fraction excretion (FE) of electrolytes and microalbumin, and glomerular filtration rate (GFR) were also determined.

Results: The male to female ratio was 0.8 and 1.5 in the patient and control groups, respectively. Mean age and height did not differ significantly between the two groups. Ultrasound evaluation of the kidney revealed that the anteroposterior diameter of the right kidney was significantly higher in CH patients as compared to healthy subjects. No significant difference was observed between GFRs in patients with CH and healthy children. The mean values for FENa and FEK were significantly higher in the patient group.

Conclusions: Increased FENa and FEK may be a manifestation of impaired tubular maturation in CH. More longitudinal studies are needed to evaluate kidney function in CH patients.

Wed-O4

Prevalence of Nephrotic Range Proteinuria in Urinary Tract Infection

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Introduction: Urinary tract infection is one of the most common diseases in neonate, infant and child and has been considered as an important risk factor for development of renal scar, hypertension and end stage renal disease. Proteinuria is an important sign for differentiation between upper and lower urinary tract infection. The aim of this study was to determine the role of urinary tract infection, in production of proteinuria and also nephrotic range proteinuria in children.

Materials & Methods: This is a Quasi Experimental and also a before and after study conducted on pyelonephritic children admitted in Mofid children hospital. Diagnosis of pyelonephritis was carried out by standard criteria (Clinical manifestation-urinalysis- urine culture – complete blood count-C-reactive protein-erythrocyte sedimentation rate-DMSA Scintigraphy). All children treated with intravenous ceftriaxone (75 mg/kg). The first fresh urine sample was collected before any treatment and analyzed for protein and creatinine. The second urine sample collected at the 7-9th day of admission.

Results: we studied 152 children (123 were female and the remaining were male) between the ages of 1 month and 12 year. In our study only 8 children (5.2%) showed normal urine protein/creatinine ratio at the admission time and others had proteinuria (94.8%).We also found nephrotic range proteinuria in 20% of our patients that all of them had normal urinalysis and normal urine protein – creatinine ratio after antibiotic therapy for pyelonephritis. There is a significant difference between first and second U prot./Cr ($p<0.005$)

Conclusions: We conclude that proteinuria and also nephrotic range proteinuria may be an important contributing factor in pyelonephritis and almost often it ameliorates after successful treatment of infection. The severe inflammation of the kidney, interstitial nephritis, antibiotic therapy and fever may explain this complication.

Wed-O5

Fanconi Syndrome with Cataract in Sistan and Baloochestan Province

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Introduction: Fanconi renotubular syndrome 2 (FRTS2) is a consequence of decreased solute and water reabsorption in the proximal tubule of the kidney. Patients have polyuria, polydipsia and phosphaturia with glycosuria and generalized aminoaciduria as the cardinal features. Hypophosphatemic rickets, osteomalacia, and Muscle weakness are important clinical manifestations. General laboratory abnormalities consist of glucosuria with a normal serum glucose, hyperaminoaciduria, low-molecular-weight proteinuria, acidosis, progressive renal insufficiency, renal sodium and potassium wasting, hypophosphatemia and uricosuria. Proteinuria is made up of albumin, low-molecular-weight proteins and tubular enzymes, such as retinol binding protein (RBP), α-1 microglobulin, β-2microglobulin, N-acetylglucosaminidase, and alanine aminopeptidase. The urinary level of these very sensitive markers of proximal tubular dysfunction is markedly elevated in FS. Albuminuria precedes glomerular dysfunction in Fanconi syndrome and while elevated, does not reach nephrotic range proteinuria. This reflects the amount of filtered albumin requiring tubular reabsorption, which has been estimated at 0.4 to 1 g of albumin per 1.73 m^2 per day. Tubular proteinuria may be seen in some forms of nephrotic syndrome, reflecting associated tubulointerstitial damage. The aminoaciduria seen in FS is generalized and its pattern is influenced by plasma values, so that in rare situations of severe protein malnutrition, aminoaciduria, as analyzed on thin-layer chromatography, may be recorded as “normal” or “mild.”Quantitative analysis by ion-exchange chromatography should be used to determine the degree of aminoaciduria. Phosphaturia and glycosuria were key features of the original descriptions of FS, and huge losses are seen in severe cases. However, milder cases may not have clinically evident losses. Renal glucosuria in FS is characterized by a low threshold, and a low maximal glucose reabsorption at saturation

glucose concentrations in blood, but normal values of maximal reabsorptive capacity (TmG) during excessive glucose loading. Bicarbonaturia reflects a reduced threshold for reabsorption and is again variable in extent, according to the underlying
cause of FS. In severe acidosis, filtered bicarbonate is reduced to a level below the threshold for proximal reabsorption and urine pH falls below. Renal insufficiency will eventually develop. An autosomal recessive form of Fanconi renotubular syndrome (FRTS2; 613388) is caused by mutation in the SLC34A1 gene, maps to 5q35.3. So far only 2 patients have been reported and only one mutation has been described in the literature.

Results: We present 85 patients with Fanconi renotubular syndrome that referred to pediatric nephrology clinic in Zahedan .The median age of these patients were 6.63±3.7 and 47% were male.38.8% of patients had positive family history in first degree and 58.8% lived in Khash. Ophthalmological evaluation showed cataract in 34 patients that were bilateral in 24 of them. The most cataract were in female but male bilateral cataract were in male.51.8% of patients had glycosuria in urinalysis. Other laboratory investigations were summarized in table1. Patients were treated with sodium bicarbonate, phosphate supplementation and 1α-calcidol or calcitriol. In some, provision of all the above supplements failed to correct the biochemical disturbances and growth suffers as a result. Renal insufficiency were developed in 12 of them that 3 of them had peritoneal dialysis and a 2 of them were received kidney transplant .In follow up of patients in during of 12 years 4 of patients died due to sepsis and pneumonia.

Variables	Median (SD)	Maximum	Minimum
Na	138.16(5.8)	149	112
K	3.84(0.64)	5	2
Ca	9.12(0.88)	11	5
p	3.41(0.84)	5	2
Alkh Phosphatas	1709(1180)	6580	200
HCO3	15.08(3.55)	24	8
Creatinine	0.76(0.37)	2	0.2
BUN	13.26(5.49)	27	3
UpH	5.79(0.83)	9	5
Specific gravity	990(141.6)	1209	1007
WBC	5486(3412)	25000	2100
Hb	10.51(2.7)	14	6
Plt	2.85(1.06)X10^6	564000	32000

Wed-O6

Comparison of Amikacin Nephrotoxicity by Cystatin C and Creatinin in Children with Acute Pyelonephritis

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Introduction: Measurement of GFR (Glomerular Filtration Rate) in patients with renal diseases and patients receiving nephrotoxic drugs is important. Recently, several studies have shown that Cystatin C as a better marker for evaluation renal function than creatinin. So, this study was done to evaluation of Amikacin nephrotoxicity by serum Cystatin C and creatinin in children with acute pyelonephritis.

Materials & Methods: All children with acute pyelonephritis who were admitted in nephrology ward were evaluated. Serum creatinin, serum Cystatin C and the GFR values were measured in patients before admission (day zero) and then on days 3 and 7 after start of treatment by Amikacin (15mg/kg/day). SPSS software was used for statistical analysis and P-value (predictive value) less than 0.05 was considered significant.

Results: Among 70 children, 61 children were female and the others were males and mean age was 42.66 ± 41.53 months. Estimated GFR based on creatinin on days 0, 3 and 7 were 72.41± 20.89 ml/min/1.73 m^2, 78.42 ± 21.15 ml/min/1.73 m^2, and 80.5 ± 22.43 ml/min/1.73 m^2 , respectively. GFR based on Cystatin C on these days were 116.23 ± 58.9 ml/min/1.73 m^2, 116.49 ± 53.31 ml/min/1.73 m^2 and 108.37± 51.02 ml/min/1.73 m^2.

Conclusions: According to this study the GFR based on creatinin didn't decrease but the GFR based on Cystatin C showed decrease. Then we recommend Cystatin C for renal function monitoring in patients treated with nephrotoxic drugs such as Amikacin.

Wed-O7

Cisplatin Induced Nephrotoxicity in Children with Solid Tumors: The Effect of Preventive Measures

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Introduction: Cisplatin is a major antineoplastic drug used for treatment of solid tumors, but the chief dose limiting side effect is nephrotoxicity; 20% of the patients receiving high dose cisplatin would develop renal dysfunction. This study was aimed to investigate the effect of cisplatin and its preventive measures on renal tubular function in children with solid tumor.
Materials & Methods: The study was performed on 20 children <15 years who received cisplatin in oncology ward. Patients receiving other nephrotoxic drugs and any renal involvements were excluded. Urine was examined before the first , third and fifth doses of cisplatin administration for Na, Mg, uric acid, P, and Cr. Ca, beta2 microglobulin, and NAG were measured in 24 hour urine . The associated blood samples for calculation of fraction excretion of electrolytes were also taken in each session. All children were hydrated before and during chemotherapy and received Mg Sulfate in order to prevent cisplatin induced nephrotoxicity. GFR, FE Mg, FE uric acid, TPR, FE Na were calculated. 24 hour urine Ca, beta2 microglobin, NAG and the calculated variables were compared before and after chemotherapy administration.
Results: FE Na, FE Mg, and urine beta2microglobulin increased, but TPR, FE uric acid, and NAG decreased after the second and third doses of cisplatin infusion compared to the first session (before the first dose of cisplatin administration). All of the changes were not significant by statistical analysis. There was no significant difference between three periods of cisplatin infusion in terms of GFR. Urine calcium was decreased significantly (P=0.001) after the second and third chemotherapy course.
Conclusions: Regarding preventive measures including hydration and Mg sulfate supplementation, we didn't find significant tubular dysfunction in children receiving cisplatin as a chemotherapeutic agent for treatment of solid tumors. Cisplatin induced injury on more distal nephron segments [thiazide sensitive Na Cl cotransporter] might be responsible for reduced Ca excretion (Gitelman-like syndrome). Further studies are warranted in order to examine the possible protective effect of Mg-supplementation and the mechanism of cisplatin induced nephrotoxicity.

Wed- O9

Acquired Proximal Renal Tubular Dysfunction in Beta Thalassemia Patients Treated With Deferasirox

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Introduction: Deferasirox is an oral chelator which is used in beta thalassemia patients. This is very useful and effective in the decrease of iron content to prevent the body from harmful side effects. We did this study to know the prevalence of proximal tubulopathy in beta thalassemia patients treated with Deferasirox.
Materials & Methods: In this study, 71 beta thalassemia Major patients treated with Deferasirox for at least 12 months, with no history of diabetes mellitus and chronic renal disease, were evaluated. Serum and urine creatinine, calcium, phosphorus, protein, glucose, sodium, potassium, chloride, and uric acid were measured. Patients were evaluated at the beginning of the study, 6 months later, and 12 months later.
Results: Among 71 patients 44.9% were female, and 45.1% were male. Mean age was 14 (the range was 3-27 years). Dosage of Deferasirox was 30 mg/kg. 6 months after the study, 5 patients had proteinuria that continued until the end of the study. One patient had isolated glucosuria. According to fraction excretion of phosphate, 5 patients had phosphaturia; so the prevalence of proximal tubulopathy was 15.4%. GFR remained normal during the study. Comparing 2 groups (patients with and without tubulopathy) who were under treatment with Deferasirox, there were no correlation of tubulopathy with age, sex, initial level of serum ferritin, and level of creatinine. (p>0.05) But in patients with tubulopathy, the level of urine protein was significantly higher than others. (p=0.003)
Conclusions: We recommended a check of serial renal function test in beta thalassemia patients who used Deferasirox.

Wed- O10

Electrolyte and Acid-base Disturbances in Acute Pyelonephritis in Children

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Introduction: Urinary tract infection is one of the most common infections in children and has been considered as an important risk factor for development of interstitial nephritis, renal scar, hypertension and end stage renal disease. Infectious interstitial nephritis due to pyelonephritis can present as electrolyte and acid base disturbance. Quick diagnosis and treatment of acid-base and electrolyte disturbance in pyelonephritic patients can prevent of mention complications. The aim of this study was evaluation of prevalence of acid base and electrolyte imbalance due to pyelonephritis.
Materials & Methods: In this Quasi Experimental and also before and after study 101 patients with pyelonephritis have been investigated in a prospective study for changes in their electrolyte and acid base. Serum Sodium, Potassium, Bicarbonate and PH have been measured before and after treatment of pyelonephritis with antibiotics and also urine random for Na and K have been measured.
Results: Results of this study were shown increase in urine sodium and potassium and decrease of serum bicarbonate in pyelonephritic patients before treatment. This study was also revealed a significant decrease in random urine Na ($p<0.002$), K ($p<0.017$) and increase in serum bicarbonate ($p<0.000$) after treatment of pyelonephritis.
Conclusions: Hyponatremia, hypokalemia and metabolic acidosis occurs in young infants with severe acute pyelonephritis in the absence of obstructive uropathy or vesico-ureteral reflux. The severe inflammation of the kidney itself may explain the electrolyte disturbance by a transient resistance of the distal tubule to aldosterone (pseudohypoaldosteronism).

Wed- O11

Acute Tubulointerstitial Nephritis in a 14-Year-Old Girl after Taking Ibuprofen

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Introduction: Acute tubulointerstitial nephritis (TIN) is a frequent cause of acute renal failure, characterized by the presence of inflammatory cell infiltrate in the interstitium of the kidney. Immuno-allergic reaction to certain medications, mainly non-steroidal anti-inflammatory drugs and antibiotics are by far the most important etiology for TIN today. Kidney biopsy is the only method of confirming the diagnosis.
Case presentation: This case report describes a 14-year-old girl with acute TIN that induces following single dosage of ibuprofen. First presented with weight loss, decrease appetite, nausea and vomiting. On initial evaluation in this patient, routine laboratory investigation were performed. Increasing serum creatinine and BUN was detected. In this case because of renal failure symptom, hemodialysis was done. Kidney ultrasound showed normal sized kidney and with hyperechogenicity, so kidney biopsy was done. Pathologic examination of the kidney specimen showed infiltration of inflammatory cells in the renal interstitium with local oedema, compatible with acute TIN.
The patient treated with prednisolone. Kidney function is restored within several weeks. In long-term follow-up, the patient is in the complete recovery.

Wed-O12

Effect of iron deficiency anemia on renal tubular function in children

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Introduction: Little is known about renal function in children with iron deficiency anemia. The aim of this study was to investigate renal tubular function in these children.

Materials & Methods: This is a Descriptive study conducted on patients suffer from iron deficiency in Mofid children's hospital. We studied 60 iron deficient patients and evaluated their serum for hemoglobin, hematocrit, iron, total iron binding capacity, sodium, potassium, calcium, phosphor and creatinine and also their urine sample for sodium, potassium, calcium, phosphor, protein and creatinine. Then we measured fractional excretion of sodium (FE Na) and potassium (FE K), urine calcium – creatinine ratio (U Ca/Cr), urine protein – creatinine ratio (U Pr/Cr), and TMP/GFR (renal threshold phosphate concentration). In this study sampling was census, plan for data collection was observation and we used from t-test and regression analysis for statistical analysis.
Results: 65% of our patients were male and 35% were female. We detected abnormal levels of FE Na, FE K, U Ca/Cr, TMP/GFR and U Pr/Cr in 28.3%, 68/3%, 45%, 30% and 80% of our patients respectively. Total prevalence of tubulopathy in our study was 95% (confidence interval 90-99%). There was a correlation between FE NA, FE K, TMP/GFR and MCV (mean corpuscular volume). (r = 0.4, r = 0.4, r = - 0.5 respectively).
Conclusions: The results suggest that children with iron deficiency anemia have impaired renal tubular function.

Wed- O13

The first Molecular Genetics Analysis of Individuals Suffering from Nephropatic Cystinosis in the South-western Iran

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Introduction: Cystinosis is a rare metabolic disorder with an incidence rate of 1 per 100,000-200,000 live births. It has three clinical types, among which Nephropatic Cystinosis (NC) is the most common and severe. The *CTNS*- causative gene for cystinosis- is located on the 17p13 locus and encodes, a lysosomal protein called cystinosin. More than 90 mutations have been described in different regions of the *CTNS* gene, as a 57-kb deletion with relative high frequency in northern Europe.
Materials & Methods: In the present study, molecular characterization of the gene was demonstrated in 25 patients from 24 unrelated Iranian families with NC. The present data exhibits the first molecular carrier detection and prenatal diagnosis of a relative large percentage of Iranian patients suffering nephropatic cystinosis, at least in southwest Iran.
Results: None of the patients showed the 57-kb deletion in heterozygous or homozygous manner. Afterwards, the coding exons, splicing boundaries and the promoter region of the *CTNS* gene were sequenced in all patients. According to the HGMD database and to our best knowledge, we found two novel mutations in exon 5 and exon 1. It's observed some mutations happen in special population with the high frequency. This can be the result of founder effect in these populations. However, this hypothesis must be evaluated in larger size in future population studies.
Conclusions: No mutation was observed in 44% of individuals. Consequently, mRNA analysis appears to be necessary for further analysis. In the case of observing no mutation in patients' mRNA in future study and uncertified role of *CTNS* as the only gene related to the disease, this study supports the hypothesis which other unknown mechanisms might be involved in the pathogenesis of the nephropatic cystinosis at least in Iranian patients.

Wed- O14

Renal Transplantation Outcome in Children with Cystinosis, Is It Better Than Patients With Other Causes of Renal Failure?

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Introduction: Cystinosis is a rare inherited disease that leads to renal failure. Fanconi syndrome is the major renal involvement in

cystinosis patients. Renal transplantation is the treatment of choice in cystinosis children with ESRD.

Materials & Methods: This study is a follow up of 21 cystinosis children transplanted in Labafi nejad hospital. We compared the graft and patient survival between cystinosis patients and children with other causes of end stage renal disease.

Results: Three cystinosis patients involved by primary non function because of graft vein thrombosis or severe acute tubular necrosis. All these three patients had low weight. The remaining cystinosis patients had excellent graft survival rate and only one patient of them lost her graft 3 years post-transplant due to noncompliance. The graft survival rate after excluding the patients with primary non function was 100%, 94%, 94% and 94% at 1, 3, 5 and 10 years after transplant. This graft survival rate especially in long term was significantly better than patients with other causes of ESRD. The mean serum creatinine in patients with functioning graft 10 years after transplant was 1.6 mg/dl.

Conclusions: We showed that cystinosis patients had much better graft function in long term after transplant.

Wed- O15

Gastrointestinal Manifestations of Nephropathic Cystinosis In Children- Single Center Experience

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Introduction: Cystinosis is an autosomal recessive disorder which is characterized by both renal and extrarenal symptoms. Gastrointestinal dysfunction has been reported in adolescent with cystinosis, and it is rarely considered in the infants. The present case series reviewed gastrointestinal manifestations of these patients.

Materials & Methods: Gastrointestinal signs and symptoms of 23 children aged 5.99 ± 0.50 years (range, 1.0 to 12.5 years) on average with cystinosis, admitted to our department of nephrology between 1996 and 2005were retrospectively reviewed. The inclusion criteria were the presence of the crystals of cystine in bone marrow aspiration and corneal deposition detected by slit lamp examination.

Results: Gastrointestinal signs and symptoms were as follows: vomiting in 16 patients (69.6%), hepatomegaly in 8 (34.8%), diarrhea in 6 (26.1%), splenomegaly in 5 (21.7%), constipation in 4 (17.4%), anorexia in 4 (17.4%), abdominal pain in 3 (13.0%), nausea in 2 (8.7%), and ascites in 2 (8.7%). Height below the 3rd percentile in was seen in 16 patients (69.6%) and weight below the 3rd percentile, in 17 (73.9%). Fifteen patients (65.2%) had both low weight and low height. Esophagogastroduodenoscopy had been performed in 6 cases and chronic inactive gastritis with H pylori infection was detected in 2 patients (8.7%).

Conclusions: Our study revealed a wide spectrum of gastrointestinal disturbances in young patients with cystinosis. Such findings should lead to greater awareness of the presence of gastrointestinal dysfunctionin these children, encourage prompt gastrointestinal evaluation, and encourage treatment of more severely affected patients.

Wed- O16

Effect of Thyroid Function in Growth of Patients With Nephropathic Cystinosis

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Introduction: Cystinosis is an autosomal recessive disorder associated with growth retardation. Growth failure in this patient is multifactorial which is attributed to metabolic, nutrition status, renal function and endocrinopathy including hypothyroidisam.
The objective of the study is to review impact of hypothyroidism on growth in patients with nephropathic cystinosis.

Materials & Methods: Fourteen patients with nephropathic cystinosis who diagnosed at Ali-Asghar Children's Hospital were included in this study. Kidney transplantation, renal failure (Glomerular filtration rate less than 60), usage of corticosteroid considered as exclusion criteria.

Results: Male to female ratio of the cases was 1 and mean age was 5 years and 4 months. Based on TSH level, patients were divided into two groups:

hypothyroids (43%) and euthyroids (57 %). Mean serum levels of TSH in hypothyroids (43%) and euthyroids patients were 14.9 (±13) and 2.2 (± 0.8), respectively.
Mean (±SD) height z scores at diagnosis of nephropathic cystinosis in hypothyroid and euthyroid patients were -2.8(± -0.4) and -1.2 (± -1.9), respectively (P value: 0.03). Mean (±SD) weight z scores at diagnosis of nephropathic cystinosis in hypothyroids and euthyroid patients were -2.6 (± -0.5) and -1.1 (± -2.2) ,respectively (P value: 0.03).
Conclusion: This study demonstrates significant difference in terms of height and weight z scores among nephropathic cystinosis patients with normal thyroid function and those who were hypothyroid. Physicians should be evaluate thyroid function of patients with cystinosis periodically in need of treatment.

Wed- O17

Anemia Is A Common Finding At Presentation Among Patients With Infantile Nephropathic Cystinosis

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Introduction: Cystinosis is a metabolic disorder with deposition of cystine in various organs, including Kidneys and bone marrow (BM). It notified that normochrom normocytic anemia is usually present in later stages. In this study, we evaluated hematologic manifestation at presentation among these patients.
Materials and Methods: In the retrospective analytic study, we studied the files of all known cases of infantile nephropathic Cystinosis admitted to Ali-Asghar Children hospital between years 1998 and 2013. Available records including initial CBC, BUN, Cr and height at presentation were reviewed and analyzed with SPSS ver. 20 program.
Results: The file of 34 known cases (12 male, 22 female) were reviewed. Mean age at diagnosis was 8.5 ± 2.4 months. The files of 23 patients completed for initial paraclinical study. Mild normochrom - normocytic Anemia [Hb = 10.48 ± 0.31 mg/dl (8.9-12.4)] was detected at presentation among 63.60% of them. Bone marrow involvement was detected among 80% of anemic patients at presentation. 85.7% of patients with GFR <60 ml/min had anemia at presentation while this rate for the patients with GFR ≥ 60 ml/min was 14.3%. There were not significant correlation between anemia at presentation and initial GFR (P= 0.3), perhaps due to low number of the sample.
Conclusions: In spite of the other studies, mild anemia is a common finding at presentation among patients with infantile nephropathic Cystinosis. It appears that anemia is associated with Cystinosis (anemia of chronic disease) and two main causes of it may be precipitation of cystine crystal on the bone marrow and decreased renal production of erythropoietin.

Wed- O18

Endocrine Disorders in Childhood Cystinosis

Salehpour S

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In the past, the treatment of cystinosis was limited to treating metabolic acidosis and, often, replacing electrolytes lost in the urine; later during the course of the disease, chronic renal disease was treated. Today, the wide availability of an effective drug, phosphocysteamine, and kidney replacement therapy with transplantation has dramatically improved the outlook for patients and altered management strategies.
Treatment with recombinant human growth hormone improves growth velocity. Long-term recombinant human growth hormone treatment in young children with nephropathic cystinosis prior to renal replacement therapy is safe and efficient. Growth hormone treatment is less effective for peripubertal or adolescent patients on renal replacement therapy. Treatment with recombinant human growth hormone does not accelerate a decline in kidney function in children with chronic kidney disease.
Thyroid hormone replacement is indicated in patients diagnosed with hypothyroidism.
We used growth hormone therapy for 11 patients with cystinosis in whom regardless of phosphocysteamine therapy and good control of acid-base and electrolytes states, an acceptable

growth rates had not been achieved. Each case was followed by a pediatric endocrinologist and pediatric nephrologists over 4 years in 3 month intervals. The response was significantly good with no major adverse effect.

Wed- O19

Apparent Mineralocorticoid Excess Syndrome: Report of One Family with Three Affected Children

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Introduction: Apparent mineralocorticoid excess (AME) is characterized by low birth weight, failure to thrive, polyuria, hypertension in combination with hypokalemia, and metabolic alkalosis. It is caused by mutations in the *HSD11B2* gene (16q22), which leads to a loss of function of 11-hydroxysteroid dehydrogenase type 2 (11-HSD2). This enzyme is predominantly present in mineralocorticoid target tissues, such as kidney, and is responsible for the conversion of cortisol to cortisone. Similar to aldosterone, cortisol has a stimulatory effect on the mineralocorticoid receptor (MCR). In contrast, cortisone has no effect on the MCR. The loss of enzyme activity with AME leads to excessive stimulation of the MCR by cortisol, followed by an increased expression of the epithelial sodium channel (ENaC) and Na/K-ATPase in the cortical collecting tubule, which results in sodium retention and potassium loss. The diagnosis can be made by demonstrating an increased urinary ratio of the metabolites of cortisol versus cortisone (tetrahydrocortisol and tetrahydrocortisone, respectively). Treatment consists of a low-sodium diet in combination with potassium supplementation and potassium-sparing diuretics.

Case Report: Case-1**:** The 12 years old boy was born in an Iranian family with two siblings with AME. He was product of a consanguineous marriage with oligohydaminouse in fetal period; IUGR with birth weight of 2.3 kg. He was admitted to our hospital with the history of polyuria and polydypsia in 18 months. On examination, the child's weight and height were both below 3rd centile (weight=7800gr; Height=71cm). His blood pressure was normal at the time of admission. Biochemical findings indicated hypokalemia with metabolic alkalosis. With this clinical and biochemical presentation Bartter syndrome was suspected, but the patient was further investigated to rule out other possibilities. In 6 years old revealed hypertension (145/70 mm Hg [95th percentile:114/77mm Hg]), hypokalemia, and mild metabolic alkalosis. His plasma renin activity was low (<0.01 pmol/L/mL/h); serum aldosterone was low (8 pmol/L); low serum renin and aldosterone level were against the diagnosis of Bartter syndrome. High ratio of cortisol to cortisone metabolites was suggestive of defect in 11β hydroxysteroid dehydrogenase type 2 enzyme. Renal ultrasound revealed the presence of bilateral nephrocalcinosis. Treatment was started with triamterene and spironolactone and later switched to amiloride and spironolactone, which corrected the hypokalemia, but her hypertension persisted. At the age of 12 years, Marfan syndrome was suspected on the basis of dilated aorta descendens. Atenolol and Nifedipine was added to his treatment and aortic aneurism was operated in 13 years. At the time this report was written, he was 15 years old and was still being treated with Triamterene, Amiloride, Atenolol. His blood pressure and serum potassium level have been within normal limits. The addition of calcium canal blocker resulted in normalization of her blood pressure.

Case 2: His brother is 9 years old that in screening of other sibling, revealed hypertension (160/70 mm Hg [95th percentile:114/77mm Hg]), hypokalemia, and mild metabolic alkalosis. Other laboratory data were: Cortisol=12.1, ACTH=43.8, Aldosterone=6.4 Renin=0.01, Na=142, K=3.9 , PH=7.81 , HCO3=28.2 .Echocardiography was shown left ventricular hypertrophy without aortic dilation.

Case3: Another brother with 6 years old also had above abnormalities in examination and laboratory finding .His blood pressure was 150/70mmHg(95th percentile:109/72mm Hg).laburatory data also was: Cortisol=15.3 ACTH=40 Aldosterone=5.6 Renin=0.01 k=2.5 PH=7.50 HCO3=26 Left ventricular hypertrophy without aortic dilation were detected in his echocardiography. They also treated same to case 1.

Discussion: The syndrome of apparent mineralocorticoid excess of AME is a form of low-

renin hypertension that is caused by congenital deficiency in the activity of the enzyme HSD11β 2. AME is usually diagnosed within the first years of life and is characterized by polyuria and polydipsia, failure to thrive, severe hypertension with low renin and aldosterone levels, profound hypokalemia with metabolic alkalosis, and most often nephrocalcinosis . Stroke has been observed before the age of 10 years in untreated children. Transmission is autosomal recessive and AME is caused by homozygous or compound heterozygous loss-of-function mutations or deletions in the *HSD11B2*gene (16q22). In all cases, these mutations lead to abolition or a marked decrease in the activity of 11-beta-hydroxysteroid dehydrogenase type 2 (11-beta-HSD2), an enzyme involved in the conversion of cortisol to cortisone. Diagnosis should be suspected on the basis of the clinical and biochemical characteristics. Detection of a marked increase (10 to 100-fold) in the ratio of cortisol/cortisone (F/E) or of the tetrahydroxylated metabolites (THF+alloTHF/THE) in plasma and urine is a strong indication for diagnosis. Differential diagnoses include pseudohyperaldosteronism (particularly Liddle syndrome), as well as other forms of early-onset childhood hypertension (particularly renal hypertension) . For families in which the disease- causing mutation has already been identified, prenatal diagnosis may be considered in case of a life- threatening event in a previous child. Early diagnosis and treatment is important to prevent end-organ damage (central nervous system, kidney, heart and retina). Two main strategies can be used to treat AME. The first is the blockade of the mineralocorticoid receptor by spironolactone (2-10 mg/kg/day), combined with Thiazides to help to normalize blood pressure and reduce hypercalciuria and nephrocalcinosis. The second and complementary strategy is the administration of exogenous corticoids to block ACTH and suppress the endogenous secretion of cortisol. This strategy has proven efficacy on blood pressure, renin and aldosterone levels but has little effect on urinary cortisol, cortisone and corticosterone concentrations. The loss of functional epithelial sodium channel (ENaC) explains why Amiloride is only an effective means of long term blood pressure control. In the absence of treatment, the prognosis for AME is severe with malignant hypertension, stroke, cardiac and renal insufficiency. However, the prognosis for patients with appropriate treatment appears to be good.

Wed- O20

Restrictive Pulmonary Dysfunction in Cystinosis

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Introduction: Accumulation of cysteine in cells of cystinosis causes multiorgan damage. Although many organs are involved in this disease, impairment of pulmonary parenchyma is not obvious and lung dysfunction commonly is due to hypoventilation (diaphragm myopathy) and extrinsic muscle involvement.

Materials and Methods: We did spirometry for 12(7 males, 5 females) patients with cystinosis From April to October 2013.Lung parenchyma was evaluate by Chest X ray(CXR) and pulse oximetry. Forced vital capacity (FVC) and Forced expiratory volume in first second (FEV1), Peak expiratory (PEF) and Inspiratory(PIF) flow , Forced expiratory flow 25-75 (FEF25-75) were measured as indicators of pulmonary function.

Results: Mean age of the patients was 10.79 years (range 5.5-23 yrs) mean height was 122 cm, weight was 26.3 kg, and BMI 17.8 Kg/H^2 . The average of being on Cystagon treatment was 8.75 yrs(range: 4.5 -20 yrs). The mean of FVC was 88% (range: 57-116%), of FEV1 was 80% (range: 51-108%), of PIF was 69.6% (range: 35-130%), of PEF was 89.4% (range: 46-148%), of FEF25-75 was 97.58 (range: 26-180%) , of FEV1 /FVC was 0.84 (range: 0.60-1).None of the patients had abnormality in CXR or pulse oximetry. From all of the cases, one had obstructive pattern (FEV1 51%, Tiffneau 0.6, FEF25-75=29%) and two patients had restrictive pattern FEV1 and FVC (<80%) and normal Tiffneau(FEV1/FVC=>80%).

Conclusions: These findings can indicate respiratory restriction without parenchymal involvement. Therefore, we suggest to do serial Pulmonary Function in all cystinosis patients.

Wed- O21

The Evaluation of The Assessment of The Length of Consolidation Course of

The Childhood Idiopathic Nephrotic Syndrome on Relapse Risk

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Introduction: Idiopathic nephrotic syndrome (INS) is characterized by relapsing proteinuria with potential risk of the adverse effect of corticosteroid therapy. Although considerable evidence suggests an overall reduction of the relative risk of relapse with longer duration of daily steroid therapy, there is no definite clue to find out which patient needs this elongation.
Initial daily steroid therapy can be divided into two parts by determination of remission point; as induction phase, since the start to remission point, and consolidation phase, from remission till end of daily steroid therapy. The propose of this study is to compare the relapse rate among the patients with constant 3 weeks consolidation course despite different remission point at initial steroid therapy.
Materials & Methods: All new cases of childhood INS selected. Oral prednisolone at 2mg/kg/day (max.60mg), as a single morning dose was started. Parents asked to examine daily morning spot urine with dip stick to find out remission point and treatment was continued for the next 3 weeks as consolidation course. Further maintenance therapy was done based on ISKDC recommendation. Patients with induction phase less than 3 days or more than 21 days were excluded. Patients categorized into 3 groups; G1: an induction phase ≤1week (9 patients), G2: >1week≤2 weeks (8 patients), G3: >2weeks≤3 weeks (8 patients) and followed 2 years for relapse rate.
Results: Twenty-five out of 69 INS patients enrolled study. Two years relapse rate in G1, G2, and G3 were found 3.9(33%), 3.8(37%) and 5.8(62.5%), respectively. Average relapse episode/2 years were 7.9(0.78) in G1, 7/8(0.87) in G2 and 11.8(1.38) in the G3. Mean time to first relapse (interval time) in each group was 106.7days, 86days and 67days, respectively. There were no significant differences between groups regarding relapse rate, relapse episodes and interval time (p-value= 0.43, =0.71, &=0.73, respectively).
Conclusions: Consolidation course has important predictive value for relapse risk, thereby directly impacting decision-making and treatment plan in INS. More studies with greater sample size suggested.

Wed- O22

Lowering Effect of Valsartan on Fetuin-A in Type 1 Diabetes

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Introduction: Fetuin-A is a protein that plays several functions in human physiology and pathophysiology. The role of Fetuin-A in type 1 diabetes (T1D) has been less studied. Herein, we have studied the serum levels of Fetuin-A in T1D patients with microalbuminuria. Furthermore, the blocking effect of renin-angiotensin-aldosterone system (RAS) on serum levels of Fetuin-A was assessed.
Materials & Methods: From January 2010 to May 2011, 32 eligible T1D patients with confirmed microalbuminuria were included in this cross sectional study in Isfahan, Iran. Serum Fetuin-A levels before and 8-weeks after valsartan administration were measured. In addition, serum lipid profile, fasting blood sugar (FBS), creatinine, hemoglobin A1C, and urine microalbumin were determined.
Results: The mean age of participants was 21.65 ± 0.38 years, with the median value of 19 years. Before valsartan administration, mean values of Fetuin-A were not significantly different between males and females (64.2208 ± 1.77426 vs. 61.3931 ± 3.35136 ng/ml, respectively; p>0.05). After valsartan administration, serum levels of Fetuin-A and urine microalbumin/Cr decreased significantly (p< 0.05). Nonetheless, a negative correlation was observed between serum Fetuin-A level after valsartan administration and serum LDL level (p=0.007, r= -0.507).
Conclusions: Valsartan (ARBs) administration concomitantly decreases Fetuin-A levels and urine microalbumin levels.

Wed- O23

Causes of Hematuria In Children Referred To Pediatric Nephrology Clinic

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Introduction: Hematuria is one of the common complaints of children referring to pediatric nephrology and urology clinics and may be discovered in 0.5 – 2% school age children in urinalysis screening. Causes of hematuria in children encompass a wide spectrum of diseases from benign conditions to life threatening events and are in many aspects different from causes of hematuria in adults. This study was designed to investigate common causes of pediatric hematuria in an outpatient clinic in order to suggest reasonable approach and avoiding unnecessary investigations.

Materials & Methods: this observational cross – sectional study was performed on 200 infants and children from 1 month to 18 years who visited at pediatric nephrology clinic with chief complaint of gross or microscopic hematuria. If there was no indication for hospital admission, repeat of urinalysis, urine culture calcium to creatinine ratio, uric acid to creatinin ratio and sonography of urinary system were done. If there were any finding in favor of glomerular hematuria, serum level of C3 and ASO titer were investigated too. All imaging and laboratory findings for each patient gathered in pre-designed forms and analyzed with SPSS 16.

Results: Patients consisted of 131(65.5%) males and 69(34.5%) females. Most of them were between 3 – 12- years old. In 74 patients (37%) no cause of hematuria was found (idiopathic), in 36 (18%) urinary tract stones, in 31(15.5%) urinary tract infection, in 26 cases (13%) hypercalciuria, in 20(10%) hyperuricosuria, in 11(5.5%) anomalies of urinary tract and in 2 patients (1%) glomerulonephritis were detected.

Conclusions: This study shows that most the causes of childhood hematuria are benign and in 56% of patients, urinary stones, urinary tract infection and crystaluria are responsible for this condition. If initial history and physical examination does not lead to a diagnosis and urinalysis is negative for dysmorphic RBC and RBC cast then urine culture, random urine for crystalluria and sonography would be sufficient for initial investigations and there is no need to perform invasive and expensive procedures such as IVP, cystoscopy and VCUG for patients in early steps of evaluation.

Wed- O24

Assessment of therapeutic efficacy of Vitamin C on pediatrics urinary tract infection

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Introduction: Urinary tract Infection (UTI) is the most common disorders in children. Complications of disease, antibiotic resistance and the need for admission are justified using appropriate pharmaceutical supplements for faster improvement of patients.Vitamin C enhances the host response to many infections and has an important role in immune homeostasis. The aim of this work is to assess possible effect of Vitamin C in treatment of children with UTI admitted in Amir Kabir Hospital in Arak city.

Materials & Methods: This is a clinical trial, recruiting 159 patients admitted due to UTI. We randomized the patients into two groups of case and control. In case group, children were treated with Vitamin C 250 mg daily for 14 days. Both groups were given routine antibiotic regime for UTI. At the end, 152 admitted patients suffering with UTI (with no underlying disease) were assessed in terms of treatment efficacy according to their clinical and urine culture results. Finally, all data were analyzed by SPSS software version 16.

Results: This study showed that children treated with Vitamin C have shorter periods of fever, urgency, dribbling and dysuria compared to control group. Other complaints such as frequency, abdominal pain, incontinence and time to make a negative urinary culture results demonstrated no significant difference in two groups.

Conclusions: According to this paper, Vitamin C is useful in shorter periods of fever, dysuria, urgency and dribbling while regarding its safety, it is also recommended in management of other clinical complaints of UTI.

Wed- O25

Renal Tubulopathy Following Vesicoureteral Reflux In Children

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Introduction: Vesicoureteral reflux (VUR) is one of the most common congenital malformations in the urinary system. In a number of cases with VUR Abnormalities in renal tubular function such as urine concentration capability and distal acidification mechanisms are impaired. The purpose of this study was to evaluate the frequency of these abnormalities in children suffering from primary VUR.
Materials & Methods: We studied 126 children 1 to 16 years (mean 7.2 ± 2.3) old followed for primary VUR during a 3-year period. Patients had normal GFR, no history of surgical intervention in the urinary system and were urinary infection-free for the last 8 weeks. Blood tests including Cr, BUN, electrolytes, Hco3and PH, as well as urine culture, urine pH, and specific gravity were measured every 3 months.
Results: Bilateral and unilateral refluxes with varied severity were observed in 39 and 87 of patients respectivly.15 cases had renal tubular acidosis and 20 had defects in urinary concentrating ability. All children in the tubulopathy group suffered from moderate to severe VUR (78% bilateral), 40 % had renal cortical scarring and 2.1% had short stature.
Conclusions: Renal tubular dysfunction is relatively frequent in bilateral and unilateral VUR, although it is especially prevalent in the former. Grade, duration, and bilaterality of reflux were more important in the tubulopathy group. Renal scarring was similar in patients with and without renal tubulopathy. Renal tubular acidosis was the main explanation for growth failure in these patients.

Wed- O26

Urinary Neutrophil-Gelatinase Associated Lipocalin Is a More Prognostic Biomarker To Distinguish Antenatal Hydronephrosis in Neonates

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Introduction: Routine diagnostic methods of Vesicoureteral reflux (VUR) are invasive and can cause exposure to radiation and may increase risk of urinary tract infections. Therefore, introducing reliable, non invasive methods might be more interested in pediatric nephrology. The objective of this prospective case control study was to evaluate the prognostic value of urinary neutrophil-gelatinase associated lipocalin (uNGAL) on antenatal hydronephrosis (AH) with and without VUR.
Materials and Methods: A total of 50 patients diagnosed with AH; 78% males with mean age 5.71± 2.1 years, including 27 AH with VUR and 23 AH without VUR, and 19 normal healthy children; 78.9% males with mean age 5.63 ± 1.89 years, were enrolled in this study. Urinary NGAL levels were measured by enzyme linked immunosorbent assay (ELISA).
Results: There was a significant difference in uNGAL concentration between AH patients and controls (0.80 ± 0.26 and 0.29 ± 0.27 ng/ml, $p<0.0001$). However, the levels of uNGAL was not significantly deviated between AH patients with VUR compared to those without VUR (0.84 ± 0.34 vs. 0.75 ± 0.13, $p=0.419$). Standardization of NGAL based on urinary creatinine (uNGAL/uCr) showed a significantly difference between AH neonates with VUR compared to those without VUR (2.43±1.61 vs. 1.91±0.79, $p<0.0001$). Receiver operator characteristic (ROC) analysis revealed higher prognostic power of uNGAL for identifying AH with a sensitivity; 95.7%, and specificity; 84.2%. Meanwhile, the levels of uNGAL or NGAL/uCr ratio did not correlate with reflux grade or laterality.
Conclusions: The urinary level of NGAL and NGAL/Cr ratio might be a surrogate non invasive, reliable tool to distinguish hydronephrosis.

Wed- O27

The Effect of L-Carnitine Supplementation on Hyperlipidemia in Pediatric Nephrotic Syndrome

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Introduction: Hyperlipidemia is a major risk factor for atherosclerosis and cardiovascular accidents. Increased plasma lipoproteins in childhood can be a prodrome for atheroma formation. Abnormalities in lipoproteins and lipids profile are common in patients with pediatric idiopathic nephrotic syndrome and may contribute to atherosclerosis and cardiovascular accidents in near future in persistent cases. Although lipid lowering agents such as statins have been investigated in adult patients, because of myopathy and growth derangement, their prescription are not recommended in children aged groups. L-carnitine as a major catalyzer in lipid metabolism has been used as oral supplementation in patients on hemodialysis and peritoneal dialysis with beneficial effects on several parameters of lipid metabolism. Up to now, there is no investigation on carnitine effects on lipids profiles of childhood nephrotic syndrome that is our study proposes.
Materials & Methods: In this study treatment cases group included 16 patients in age 2-12 old years with steroid resistant nephrotic syndrome that they were receiving prednisolone, cyclosporine and carnitine (25mg/kg/day) in 3 months and control group included 17 matched age and sex patients receiving standard treatment (prednisolone, cyclosporine).In both groups, biochemical parameters like triglyceride, cholesterol, VLDL, LDL, HDL, BUN, creatinine and serum albumin were measured.
Results: At the end of the study period, L-carnitine treated group showed no significant improvements in the biochemical markers compared with the control group. In each group there was lowering of hyperlipidemia at the end of the study period because of expected effects of immunosuppressive agents on clinic course.
Conclusions: Our study indicates that oral administration of L-carnitine has no effect on lipoprotein profile of persistent childhood nephrotic syndrome. However, higher dosage and longer time period carnitine supplementation needs more investigations.

Wed- O28

Association of Hyponatremia with Febrile Urinary Tract Infection in Children

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Introduction: Urinary tract infection (UTI) is a common infectious disease in children. Pyelonephritis results in complications such as scar formation, hypertension and renal failure. According to some studies there is an association between UTI and electrolyte disturbances especially hyponatremia. The present study has been designed for the assessment of association between UTI and hyponatremia.
Materials and Methods: The study is a retrospective and descriptive research in which 120 children with UTI were included. They were divided into two groups of sixty children; those with and those without hyponatremia. Study parameters included serum sodium level, white blood cell count (WBC), erythrocyte sedimentation rate (ESR), C - reactive protein (CRP), and also scan findings by cortical scintigraphy with technetium-99m dimercaptosuccinic acid (99mTc DMSA). Data were collected from patients admitted between 2011 and 2012 and were used to collect the necessary information. Each patient's data were transferred onto a checklist. Data analysis was performed using descriptive statistics, chi-square, and independent t- test with Mann-Whitney test by SPSS version 19.
Results: In this study there was a significant association between hyponatremia on the one hand and WBC count ($p=0.003$), ESR ($p<0.001$), CRP ($p=0.004$), duration of fever ($p=0.002$) and abnormal DMSA scan findings ($p=0.002$) on the other hand.
Conclusions: There is a significant association between hyponatremia and severity of UTI in children.

Wed- O29

Incidence and Severity of Vesicoureteral Reflux in Children Affected By Upper Urinary Tract Infection

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Introduction: Vesicoureteral reflux incidence and indications for imaging survey is the aim of many studies for many years, in this study we try to evaluate this matter in our area.
Materials & Methods: 125 patients entered to this study aged between 2 and 96 months affected to urinary tract infection, all were febrile (central temperature above 38.3 °C) with positive urine culture, after urine culture was sterile, imaging study include voiding cystogram has been done . The severity of reflux was defined to five grades. All information include age and sex were recorded and data analyzed with independent Student T-test and Chi square test in SPSS 16 , and P value less than 0.05 was significant .
Results: 125 patients with urinary tract infection include 80 females, 45 males. Vesicoureteral reflux was detected in 55 cases (44%). Reflux occurred in male more than female (49% vs 39%). The incidence of reflux in aged under one was similar in female and male (50% in both sex roughly).
Conclusions: Urinary tract infection occurred more frequently in females compared to male (1.7 times). Although the incidence of reflux in male is more than female but, this rate is same in aged under one .

Wednesday Poster Presentations

Wed- P1

The Correlation between Sleep Quality and Ambulatory Blood Pressure in Patient with History of Urinary Tract Infection

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Introduction: Reflux nephropathy is a common problem in childhood and the leading cause of hypertension in this age group. Hypertension has been shown to be accompanied by sleep problem in adults. However, this correlation has not been studied in children. Considering the importance of sleep quality in growth and development, we performed this study to investigate sleep quality parameters in children suffering from hypertensive nephropathy.
Materials & Methods: Eighty six patients aged 5-15 years with at least one year history of reflux nephropathy were included in this study. They underwent 24-h blood pressure (BP) monitoring. Systolic BP (SBP), diastolic BP (DBP), SBP load, DBP load as well as mean arterial pressure (MAP) were measured at day and night, separately. In addition, Pittsburg Sleep Quality (PSQ) index questionnaire was filled-out and its data were compared in hypertensive and normotensive groups.
Results: After excluding duplicate cases and those ABPM with insufficient data, 78 children entered into study. Sleep quality was very good (n=38), fairy good (n=32), fairy bad (n=3), very bad (n=5). Mean diastolic blood pressure load (p=0.019), diastolic load awake-time (p=0.045), mean systolic sleep-time (p=0.022), systolic drop (p=0.009) were statistically different among groups. By dividing the children to two groups of good and poor quality sleep, the parameters of ABPM were not different between groups(P>0.05). In addition, there was no correlation between blood pressure classifications and sleep latency, duration of sleep, sleep efficiency, sleep disturbance , day dysfunction due to sleepiness, and overall sleep quality score(P>0.05).

Conclusions: Our study could not show any correlation between sleep quality and ambulatory blood pressure monitoring (ABPM) parameters in children with abnormal blood pressure but we suggested a bigger sample size and longer period of patients follow up for more precise findings.

Wed- P2

Bartter Syndrome in Neonate, a Case Report

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Introduction: Bartter syndrome is a rare autosomal recessive renal tubular disorder affecting the thick ascending limb of Henle's loop, and Characterized by hypochloraemic metabolic alkalosis, persistent hypocalcaemia, hyperreninemia with hyperaldosteronism, and normal blood pressure. The majority of patients present with failure to thrive, dehydration, vomiting and constipation during the first 2 years of life. We are reporting a neonate with Bartter syndrome diagnosed in the third week of his life, which are few reports in the literature.

Case presentation: A 23 day old-boy was born following of first degree consanguineous marriage by a preterm pregnancy complicated by polyhydramnios. There was a history of failure to thrive and death because of severe dehydration with unexplained etiology in the previous sibling. The baby presented with infrequent vomiting, and failure to thrive in spite of supervised feeding. He admitted to the hospital with severe dehydration, and laboratory investigation showed persistent hypokalemia, hypochloremic metabolic alkalosis, and excessive loss of sodium, potassium and chloride in urine. There was no evidence of sepsis and his blood pressure was normal. The diagnosis was confirmed by high plasma renin and aldosterone level. The child was put on potassium and sodium supplementation initially and after that indomethacin in divided dose. After one year follow his growth and development was normal

Conclusions: Early diagnosis and treatment of Barter syndrome is important, and treatment with indomethacin will blunt the prostaglandin overproduction and correct of this metabolic disturbance.

Wed- P3

Renal Tubular Acidosis and Muscular Paralysis

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Introduction: Renal tubular acidosis (RTA) is non-uremic defects of urinary acidification. Renal tubular acidosis is characterized by a normal anion gap hyperchloremic metabolic acidosis; plasma potassium may be normal, low or high- depending on the type of RTA. Type 1 or distal RTA is referred to as the classic RTA and is a disorder of acid excretion involving the distal tubules**.** The disorder is characterized by a hypokalemic, hyperchloremic metabolic acidosis and hypercalciuria**.** Patients with hypokalemia often have no symptoms, particularly when the disorder is mild (serum potassium, 3.0 to 3.5 mmol per liter). With more severe hypokalemia, nonspecific symptoms, such as generalized weakness, lassitude, and constipation, are more common. When serum potassium decreases to less than 2.5 mmol per liter, muscle necrosis can occur, and at serum concentrations of less than 2.0 mmol per liter, an ascending paralysis can develop, with eventual impairment of respiratory function. The likelihood of symptoms appears to correlate with the rapidity of the decrease in serum potassium. In patients without underlying heart disease, abnormalities in cardiac conduction are extremely unusual, even when the serum potassium concentration is below 3.0 mmol per liter. In distal RTA in spite of the presence of persistent hypokalemia, muscular paralysis is uncommon and rarely described in children. Here we report 2 patients with distal RTA and hypokalemia muscular paralysis.

Case 1- The first patient was a 5.5 years old boy with a history of fever and cough from the day before and walking inability and pain in lower limbs. He was the known case of distal RTA and nephrocalcinosis and was on the maintenance treatment of hydrochlorothiazide and k-citrate from 1 year old. In addition he had received inhaling salbutamol and the acetaminophen syrup because of the respiratory symptoms from the last day. On admission he had not weight bearing and

the lower limbs had tenderness on touch. He had a normal complete blood cell counts (CBC), Na=148 meq/lit, k=2.1, BUN=10mg/dl, serum Cr=0.7 mg/dl meq/lit, PH=7.32, PCo2=33.9mm Hg, HCO3=17.7 meq/lit.

Case 2- The second one was a 7.5 years old boy with a history of fever, vomiting and walking inability and pain in lower limbs from 2days before. He was the known case of distal RTA and was on the maintenance treatment of k-citrate from 1 year old. He had received no other medications. On admission he was moderately dehydrated and could not sit or stand because of the sever weakness and had lower limb tenderness. He had normal CBC, Na=154 meq/lit, k=2.5 meq/lit, BUN=16mg/dl, serum Cr=1 mg/dl, ph=7.18, HCO3=10.9 mg/dl, PCO2=28.4 mm Hg.

Both patients were rehydrated and received oral and intravenous k and potassium citrate. On day 2 of admission they were able to walk and the muscle pain was disappeared with the rising of serum k above 2.5 meq/lit. Both patients were discharged from the hospital with oral k citrate, and oral kcl and hyrochloropthiazide because of the hypercalciuria and nephrocalcinosis.

Conclusions: RTA is a known cause of hypokalemia, but in spite of the presence of persistent hypokalemia muscular paralysis is uncommon and rarely described in children, and the onset of paralysis may initially be misinterpreted particularly if the patient is attended by a physician who is not a pediatric nephrologist. Therefore parents must be informed about this possibility and be aware of the drugs which can induce hypokalemia such as salbutamol.

Wed- P4

Vitamin E Deficiency Common Findings in Dialysis Patients

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Introduction: Vitamin E is an antioxidant that prevents propagation of free radicals in membranes and plasma lipoproteins. Oxidative stresses are increased in end stage renal disease. Avoiding supplements of vitamin E in chronic renal failure is recommended. This study aimed to define serum levels of vitamin E in dialysis patients and determine whether vitamin E supplements are needed.

Materials & Methods: 26 dialysis patients; 10 girls (38.5%) and 16 boys (61.5%) aged 37-300 months enrolled the study .They included 9 (34.5%) peritoneal and 14 hemodialysis (53.9%) cases .3(11.6%) have received both modalities separately. Duration of dialysis was 7-128 months .Low serum levels of vitamin E defined as levels <3µ/ ml in patients ≤ 10 years and levels <6 µ/ml in teenagers (>10years).

Results: Serum levels of vitamin E was normal in 4(14.8%), low in 19(70.4%) and high in 3 (11.1%) patients. It ranged 0.6-20 (3.93) µ/ml.Six CAPD (2/3) and 11hemodialysis (78.5%) patients had vitamin E deficiency. Age, modality and duration of dialysis and characteristics of dialysis session (number of cycle /day, duration and volume of each dwelling time in CAPD patients; number of dialysis sessions per week and duration of each session in hemodialysis cases), mean Serum BUN and albumin levels were compared between patients with and without vitamin E deficiency. We didn't find any significant statistical differences between groups (P>0.05 for all).

Conclusions: Vitamin E deficiency is not unusual in dialysis patients, so screening for vitamin E deficiency to define those that need vitamin E supplements is recommended.

Wed- P5

Vitamin C Deficiency: A Common Finding in Hemodialysis Patients

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Introduction: Deficiencies of water soluble vitamins are common in dialysis subjects and supplements of vitamin B and C are recommended. We aimed to determine vitamin C deficiency in our dialysis centers.

Materials & Methods: 37 dialysis subjects ,15 girls(22.4%)and 22(59.5%)boys aged 19-

300(165.78±78.34)months including 9(24.3%)CAPD,23(62.2%) hemodialysis and 5(13.5%) patients who received both modalities separately enrolled the study .low flux membranes (R3-R5 and polysulfone) were used in hemodialysis subjects . The duration of dialysis was 1-128 months (44.74±32.68). Serum vitamin C levels were measured and values 0.6-2mg/dl in patients ≤20 years and 5-18mg/dl in patients >20years defined as normal value respectively. Serum levels < 0.6 and <5mg/dl in age groups <20 and ≥20years defined low respectively. For data analysis Chi square and student T tests were used and P values <0.05 considered as statistical significant differences.

Results: 18 (48.6%) subjects were received vitamin C supplement (60-125mg/daily), whereas 11(29.7%) case did not receive it .in 8 patients (21.6%) they were not sure about the drug consumption. Serum vitamin C levels were 0.45-1.45(0.92±0.27) mg/dl.The serum levels were low in 11(29.7%) and normal in 26(70.3%). Ten of 11(90.9%) subjects with vitamin C deficiency were hemodialysis patients and 1(9.1%) has been received both modalities. Mean serum vitamin C concentration in CAPD and hemodialys patients were 0.96±0.24 and 0.87± 0.28 mg/dl respectively (P=0.255).Vitamin C deficiency was significantly more prevalent in hemodialysis versus CAPD patients (P=0.017)

Conclusions: Vitamin C deficiency is common in hemodialysis patients despite using the supplement of the vitamin.

Wed- P6

Cystinuria – Clinical Presentation and Outcome- Case Series

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Introduction: Cystinuria is an autosomal-recessive defect in reabsorptive transport of cystine and the dibasic amino acids such as ornithine, arginine, lysine from the luminal fluid of the renal proximal tube and small intestine. The only phenotypic manifestation of Cystinuria is cystine urolithiasis which often recurs throughout and affected individual's life time. Surgical intervention is necessary but the cornerstones of treatment are dietary and medical prevention of recurrent stone formation. The aim of the present study was retrospectively review of the initial clinical features, laboratory tests disturbance and management.

Materials & Methods: We retrospectively review 7 patients who admitted in Ali-Asghar children hospital, between March 2003 and May 2013.Inclusion criteria were the presence of urolithiasis, positive nitropruside test, and the presence of cystin in stone analysis.

Results: Of the 7 patients, 3 (42.85%) were female and 4 (57.14%) were male. The mean age was 6.85 years (range: 1-16 yrs). The mean age of presentation of the patients was 3.85 years. (Range newborn -10 years). In 6 patients (85.71%) of the cases, the parents were close relatives. The most frequent manifestations were renal stone (57.14%), FTT (28.57%), renal stone with FTT (14.28%), and obstructive renal failure. One patient had proteinuria and one had glucosuria. Of six patients who had follow up, renal filtration rate was more than 90ml/min/1.73m2 (in 14.28%), between 60 to 90 (in 28.57%), between 30 to 59 (in 28.57), less than 15 (in 14.28%).

Six patients were treated by polycitrate potassium and one patient with captopril and Penicillamine. Outcome: Four patients are still on medication, one stopped the medication, one died, and one lost to follow up.

Thursday Oral Presentations

Thurs- O1

Renal Tubular Dysfunctions and Height Growth

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Introduction: Every chronic systemic disorder results in growth failure in children. Therefore, evaluation of growth is a valuable device for monitoring the health in children. Here three patients with undiagnosed tubulopathy are described who came because of short stature to Endocrinology Clinic. Physical examination was done first. Body mass index (BMI) was calculated as [weight (kg)/height $(M)^2$]. Their height and BMI compared with curves and tables of CDC2000. Standard deviation scores of height and BMI were calculated from (Patient measurement – 50% of

the growth tables)/SD for age and sex. Laboratory tests were done at standard level.

Case Presentation: 1st Case: A 9.33 yr girl with 105 Cm Height (H) (-4.8 SDS) and 15 kg weight (W) (BMI, -0.7 SDS) was diagnosed as distal renal tubular acidosis due to venous blood gas (VBG) as: pH, 7.276; HCO3, 12.5 mmol/L; base excess, -12.3 and urine pH, 7.2and nephrocalcinosis in kidney sonography. She had polyuria and polydipsia .Blood urea nitrogen (BUN), creatinine, blood Na, K and Ca were normal but phosphorus was 3.6 mg/dL and Alkaline phosphatase (AlkP) 1034 U/L (normal, up to 400) .She was treated by polycitra solution for 7 years. At 16.25 yr of age her height was 145.5 Cm (HSDS, -2.6) and weight, 45 kg (BMI SDS, 0.1).

2nd Case: A 13.66 yr boy with height, 112.5 Cm (-6.0 HSDS) and weight, 15 kg (BMI, -1.2 SDS) came due to short stature. On PE there was not dysmorphic feature or any anomaly. Laboratory tests showed: serum K, 2.2 mEq/L; Na, 136 mEq/L; Ca, 9 mg/dL; P, 3.8 mg/dL and AlkP, 580 U/L (normal 44 – 450). BUN, Creatinine and cell blood count were normal. Venous blood gas showed alkalosis and plasma renin activity was 86.5 ng/ml/hr (0.6 – 1.9); aldosterone, 630 pg/mL (30 – 355). There was no hypertension. Therefore, he was treated with potassium chloride and indomethacine with diagnosis of Bartter syndrome. At the age of 27.83 yr, his height was 161 Cm (-2.2 SDS) and weight, 45.5 (BMI SDS, -1.0).

3rd Case: A 9 yr old boy with height, 99.5 (-5.7 SDS); weight 13 kg (BMI SDS, -1.2) had normal anion gap metabolic acidosis (pH, 7.121; HCO3, 11.2 mmol/L; BE, -16.3 mmol/L) and urine pH, 7.5. Serum K was 3.3 mEq/L,Na, 139 mEq/L and Urea, creatinine and Ca were normal. Serum Pwas 3.5 mg/dL and AlkP, 1800 U/L (150 – 850). He also had nephrocalcinosis in sonography of the kidneys. He treated with polycitra K solution with the diagnosis of distal renal tubular acidosis. At 15.33 yr of age he had 160 Cm height (-1.4 SDS) and 43 kg weight (-0.7 SDS).

Conclusions: All of the children were the product of first cousin consanguineous marriage. All of them had mild rickets that were treated. So, in every short stature child in every age, we should think about tubulopathy and appropriate work up should be done for the diagnosis.

Thurs-O2

Clinicopathologic Study of Pediatric Tubulointerstitial Nephritis in Ali-Asghar Children Hospital During Past 30 Years

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Introduction: Tubulointerstitial disorders are characterized by diseases that affect the vascular and interstitial compartments of the kidney with relative sparing of the glomeruli. It may be either acute or chronic. Acute tubulointerstitial nephritis (TIN) is associated with acute renal failure due to either acute infection of kidneys or reaction to medication. Chronic interstitial nephritis is associated with progressive loss of glomerular filtration rate over time and characterized by many syndromes of renal tubular dysfunction that may be primary or secondary due to renal tubular damage from wide variety of causes. The aim of this study is to see the pathologic characteristics of the acute and chronic TIN and the probable causes of them.

Materials & Methods: All the patients with diagnosis of tubulointerstitial nephritis from 1983 to 2013 referred to Ali-Asghar hospital were determined. Demographic data and pathologic findings of these patients extracted from their archives in the hospital and then were analyzed.

Results: 44 patients, 18 males and 26 females with a mean age of 8.8 years (4SD) were enrolled in this study. 37 (84%) had chronic and 7 (16%) had acute TIN. 32(72%) were primary with a diagnosis of familial nephronophtisis and medullary cystic disease, in addition 12 (28%) were secondary to other diseases such as Alport disease, glomerulonephritis, hyperoxaluria, congenital nephrotic syndrome and amyloidosis. Seven of chronic cases led to secondary focal and segmental glomerulosclerosis, presenting as nephrotic syndrome and 11 of them were due to chronic pyelonephritis or vesicoureteral relux. Kidney biopsy showed similar pathologic findings, with periglomerular fibrosis (72%), interstitial fibrosis/tubular atrophy with different scores (91%), inflammatory cells infiltration, acute or

chronic (100%) and glomerular sclerosis, segmental and global (89%).

Conclusions: The clinical and pathologic findings of acute and chronic tubulointerstitial nephritis are the same and in many of these renal disorders, pathologic findings of the biopsy cannot determine th e etiology. Most cases of untreated TIN are presented as end stage kidney disease and some show nephrotic syndrome due to secondary involvement of the glomeruli. Early detection of TIN according to clinical data can prevent an irretrievable condition.

Thurs- O3

Relapsing Polychondritis with Renal Involvement: A Case Report

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Introduction: Relapsing polychondritis is a rare systemic inflammatory disease, characterized by episodic cartilage inflammation such as ear, nose and laryngotracheal Tree. Relapsing polychondritis occurs rarely in childhood but is equal in both sexes. Renal involvement is a very unusual feature of relapsing polychondritis.

Case Representation: This is a case report of an 11 years old girl presenting with microscopic hematuria arthritis and preorbital edema. During admission swelling, erythema and tenderness appeared in both auricles (auricular chondritis). Then biopsy of auricle was done and chondritis confirmed. Also renal biopsy showed focal and segmental glomerulosclerosis. Relapsing polychondritis was diagnosed by criteria of McAdam et al.

Conclusions: Relapsing polychondritis should be considered in differential diagnosis of microscopic hematuria with chondritis. Steroids and Immunosuppressive drugs can be useful for clinical improvement.

Thurs –O4

Renal Tubulopathy in Juvenile Idiopathic Arthritis

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Juvenile Idiopathic Arthritis (JIA) is the most common chronic rheumatic disorders in children. The main symptom of JIA is chronic arthritis but in this systemic disease extra articular involvement is common, especially in poly articular and/or systemic JIA (SoJIA). However, renal involvement (rheumatoid nephropathy) is uncommon as an extra articular manifestation of JIA. Renal involvement in JIA is due to side effect of treatment with non-steroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen and indometacine or disease-modifying antirheumatic drugs (DMARDs) such as anti-malaria, Meotraxate and/or gold therapy. Renal involvement can be seen as a complication of inflammatory chronic disease in JIA especially in SoJIA. Tubulointerstitial involvement is a complication of SoJIA and another serious, but rare complications is amyloidosis. Renal tubulopathy is a rare complication or side effect of treatment in JIA. Tubular enzymuria, sucha as N -acetyl glucosaminidase (NAG) and NAG/ Cr increases during the active phase of JIA especially with systemic symptoms, but it has not an association with permanent renal damage. This index has been higher in patient with decreased creatinine clearance. In a study methotrexate didn't have negative effect on tubular NAG-enzymuria in short time treatment in adult patients with RA. In the pat, gold tubulopathy was a complication of gold therapy in RA patients but it decreases after the cessation of gold therapy.

Urinary excretion of renal tubular basement membrane antigen (TBM), NAG, and beta-2-microglobulin are common tests in evaluation of renal tubular dysfunction in patient with active JIA.

Thurs –O5

Predictive Accuracy of Beta-2 Microglobulin For Kidney Injury In Children With Acute Pyelonephritis

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Introduction: Urinary Tract Infection (UTI) is one of the most common infections in childhood and infancy. Renal scar is the important complication of pyelonephritis. The aim of current study is to evaluate the diagnostic accuracy of beta-2 microglobulin test in detection of renal scar in children with acute pyelonephritis.

Materials & Methods: Eighty-nine Children between 2 months to 14 years old with the diagnosis of acute pyelonephritis who were referred as outpatient or for hospitalization to Koodakan hospital in Bandarabbas, southern Iran who hadn't past history of infection in urinary system. A standard urine sample according to patients' age was obtained for Urine Culture, Urine Analysis, and β2 Microglobulin tests. Also a blood sample was obtained for CBC, Creatinine, BUN, CRP, ESR, WBC and electrolytes tests. All patients were undergone DMSA scan. Data was analyzed using SPSS 20.0 and MedCalc software.

Results: The cut off point for beta-2-microglubuline for prediction of positive DMSA scan was > 0.8 with sensitivity of 40.91% (95% CI 26.3 – 56.8) and specificity of 84.09% (95% CI 69.9 – 93.4), Positive Predictive Value of 72% (95% CI 50.6 – 87.9) and Negative Predictive Value of 58.7% (95% CI 45.6-71). The cut off point for WBC for prediction of DMSA scan was > 12900 with sensitivity of 45.45% (95% CI; 30.4 – 61.2%) and specificity of 84.09% (95% CI 69.9 – 93.4), PPV of 74.1% (95% CI; 53.7 - 88.9%) and NPV of 60.7% (95% CI; 47.3 – 72.9). The cut off point for ESR for prediction of DMSA scan was > 56 with sensitivity of 31.82% (95% CI; 18.6 – 47.6%), specificity of 97.62% (95% CI; 87.4 – 99.9), PPV of 93.3% (95% CI; 68.1 – 99.8), NPV of 57.7% (95% CI; 45.4 – 69.4).

Conclusions: Beta-2 microglobulin is not enough sensitive and specific to be used as a diagnostic marker for prediction of renal scar. Other common markers such as ESR, WBC count, and CRP can be used in combination to predict kidney injury in UTI children.

Thurs –O6

Infantile Cystinosis (A Single Center Experience)

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Introduction: Cystinosis is a rare inherited lysosomal storage disorder with: reduced efflux of the amino acid cystine from lysosomes due to mutated cystinosin, accumulation of variable amounts of cysteine, cellular and tissue dysfunction ,5% of all chronic renal failure in children is accounted for by Nephropathic Cystinosis, a typical presentation of failure to thrive, acidosis, renal tubular Fanconi's syndrome, muscle hypotonia, rickets, polyuria, polydypsia, photophobia, and renal failure at age 10 years.

Materials & Methods: Patients with the following criteria were considered as cases of infantile cystinosis: growth retardation under third percentile, presence of glucosuria with normoglycemia and finding suggested renal tubular Fanconi's syndrome, typical corneal cystine crystals on slit lamp examination. Nineteen patients with fulfilled criteria of nephropathic cystinosis were evaluated based on personal and family history, clinical examination, blood picture and measurement of blood urea, creatinine, electrolytes, glucose, acid-base status, Free T4 and TSH. Additional investigations were performed in patients with anemia and those with renal impairment. Complete ophthalmologic examination was conducted for all patients at the Ophthalmology unit.

Results: Diagnosed patients included five boys and fifteen girls with a mean age at diagnosis of 10 ± 3 months (6-38 months). Two patients were above two years at the time of diagnosis, including one above38 month and the other above 25 month of age. The most frequent presentations were including: failure to thrive, renal tubular acidosis, glycosuria, advanced rickets, polyuria, and polydipsia, Irritability (100%). Other associated symptoms were: diarrhea (10%), constipations

and abdominal distension (40%), vomiting (20%), hypotonia (one case), Urolethiasis (3case) . Laboratory findings included: hypocalcaemia, hypophosphatemia, hypokalemia, hyponatremia. Regarding family history, pa-rents were consanguineous in 8 families and sibling deaths were reported in 3 families. These deaths occurred at a mean age of 23.4 ± 5 months (8-38 months). One infant died due to poor general condition+ metabolic disturbances and Tow patient died due to end stage renal failure. These two patients referred to our hospital without previous diagnosis of cystinosis. They had suffered from failure to thrive and had been hospitalized for many times because of dehydration in other centers (misdiagnosis).One of them had very poor general condition+ metabolic disturbances and decompensated state at hospitalization in our center. The other had got peritoneal dialysis and was stable .Parent had not follow up and child have been died because of discontinuation of dialysis and medications. Other seventeen patients were treated with oral cysteamine 50-60 mg/kg/day. Apart from cysteamine, the most frequently used medications were one alpha hydroxy vitamin D3 ,phosphorous and sodium-potassium citrate (polycitra) supplements (all cases), L-carnitine (one case with hypotonia). During follow-up, our study patients demonstrated partial improvement of height, with a median increase 6cm/year without the need for growth hormone. 3/17 patients under follow up reach ESRD by 12,14,15 years of age which got renal transplantation .

Conclusions: Most patients with infantile nephropathic cystinosis present during the first year of life with failure to thrive, polyuria, polydipsia, and/or dehydration and are found to have Fanconi's syndrome with normal anion gap metabolic acidosis. Some of these patients may develop vitamin D-resistant rickets due to phosphaturia. Renal function is generally normal at presentation, but without treatment most individuals progress to end-stage renal disease after 5 years of age, and only rarely before then. End stage renal disease in patients treated with cysteamine occurs ~ 15-28 years. IN our report patients went to end stage renal disease before 16 years old age. Successful treatment of nephropathic cystinosis requires early diagnosis and Specific therapy with cysteamine has improved the prognosis.

Thurs- O7

The Effect of Methylprednisolone on Urinary Level Of Interleukin-6 And Interleukin-8 In Children With Acute Pyelonephritis

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Introduction: Urinary tract infection is common in childhood urinary tract infection complications such as hypertension, renal scar; chronic renal failure is frequently occurs who may lead to dialysis and renal transplantation. UTI is the most frequent infection following the upper respiratory tract infection. Cytokines play a major role in renal scar formation following pyelonephritis. Subject of this study is the efficacy of the use of steroid in decrease of urinary level of cytokines. At this study we investigated the role of 5mg/kg intravenous methylprednisolone combined with antibiotics in diminishing urinary interleukin6 and interleukin8 concentration during acute phase of pyelonephritis compared with standard anti biotic therapy.

Materials and Methods: Urinary IL6 and IL8 concentration were determined by enzyme immunoassay in 25 children whit pyelonephritis who were treated with ceftriaxone plus 5mg/kg intravenous methylprednisolone (case group) and in 25 children with the same diagnosis treated with ceftriaxone alone (control group).Urine sample obtained at the time of presentation prior to drug administration and at 72 hours after initiation of medication. Urine sample stored at $20^{\circ c}$ used for cytokine measurement.

Results: Urinary level of IL6 and IL8 were high in the case and control groups prior to initiation of medication (33.58±22.46 pg/ml, 176±225.11 pg/ml and 27.26±24.28 pg/ml, 90.76±92.21 pg/ml respectively) in compare with normal population .Urinary levels of interleukin6 and interleukin 8 were decrease at two groups after treatment in comparing with before treatment and this difference was significantly valuable. In addition, combined ceftriaxone and methyl-prednisolone significantly decrease ($p<0/01$) urinary level of IL6 (three folds) and IL8 (eleven folds) in case group 72 hours after initiation of medication

compared with control group.
Measurement of cytokines: IL6 and IL8 were measured by using an enzyme linked immune sorbent assay (ELISA) method. Detection kits were interleukin6 and interleukin8 kits that were made in eBioscience co of Austria.
Data were expressed as the mean± SD. Normality of data were assessed with Kolmogrov Smirnov test and we found that distribution of data at each group were not normal, thus data assessed with non parametric Wilcoxon test for compare differences of pre and post medication urinary interleukin 6 and 8 levels between case and control group. There is no significant difference in mean age (P = 0.819), gender distribution (P=0.334) of two groups (P>0/1).All of the patients were febrile and E-coli was the commonest microbial germ who reported from urine cultures in two groups. Erythrocyte sedimentation rate from 63/8±29/3 at case group decrease to 44/5±17/1 and this decrease had significant difference (P<0/01).
Conclusions: We conclude that intravenous methyl-prednisolone combined with antibiotics significantly decrease urinary IL6 and IL8 levels in patients with acute pyelonephritis .this suggests that the clinical use of corticosteroids may prevent scar formation following pyelonephritis.

Thurs –O8

Kern- Sayer Syndrome Presenting As Fanconi Syndrome

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Introduction: Fanconi syndrome is a generalized dysfunction of renal proximal tubule resulting in bicarbonaturia, phosphaturia, aminoaciduria .This tubulopathy presents with severe metabolic acidosis, failure to thrive, and rickets. Fanconi syndrome can be inherited or acquired. Mitochondrial cytopathies are among rare causes of inherited fanconi syndrome. A case of Kern-Sayer syndrome presented here as a cause of Fanconi syndrome.
Case report: A 4- year old boy presented with severe lower limb deformity, fatigability, severe metabolic acidosis, and hypokalemia. Further laboratory data showed normal anion gap metabolic acidosis, phosphaturia, and aminoaciduria so Fanconi syndrome was diagnosed for him. No evidence of cystinosis was and Neostigmine was started for him with no benefit. After some months the patient presented with decreased level of consciousness (stupor), hypotonia, and significant worsening of ptosis. With high suspicion of mitochondrial cytopathy, serum and CSF lactate and pyruvate were measured which were in favor of this diagnosis. There were recurrent episodes of decreased level of consciousness and during one of them irregular pulse rate was detected for the patient and ECG showed complete AV block. Hence, definitive diagnosis of Kern-Sayer syndrome was made after about 8 years of initial presentation as Fanconi syndrome.
Conclusions: Fanconi syndrome may be caused by congenital inherited diseases which some of their signs and symptoms may present many years later, so close follow up of patients and notice to any new or unexplainable presentations may help reaching proper diagnosis and treatment.

Thurs -O9

Chronic Kidney Disease Stages 3-5 In Iranian Children: Need For A School-Based Screening Strategy: The CASPIAN-III Study

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Introduction: There are scarce epidemiological data on early and asymptomatic stages of chronic kidney disease (CKD) in children, especially from developing countries. In this study, we investigated the frequency of CKD stages 3-5 among the general students of Isfahan (a large province of Iran), and compared the findings with those derived from the main pediatric nephrology referral center of province.
Materials & Methods: This study was performed among 712 Isfahan school students (377 boys) aged 7-18 years, as part of the baseline survey of a national surveillance system. Blood samples were analyzed for blood urea nitrogen, creatinine, and cystatin C. Glomerular filtration rate (GFR) was calculated based on two 2009 Schwartz equations

(the "updated" and the "new" equations). CKD was defined as GFR <60 ml/min/1.73 m2. Additionally, a retrospective analysis of clinical records of children with stages 3-5 CKD referred to main referral center of province from November 2001 to December 2011 was made.

Results: The mean age of students was 12.2 ± 2.4 years. In students' screening, the frequency of CKD was 1.3% and 1.7% based on the updated Schwartz and the new Schwartz equation, respectively. The main referral center survey revealed an annual incidence of 14.5 per million age-related populations (pmarp), and a prevalence of 118.8 pmarp in our province.

Conclusions: The prevalence of asymptomatic and undetected low GFR in Iranian children is higher than what is reflected from the reports of referral centers. Simple screening programs like annual urinalysis among high-risk school students should be considered.

Thurs- O10

Are Serum and Urine Neutrophil Gelatinase Associated Lipocalin Predictive of Renal Graft Function In Short Term?

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Introduction: NGAL is a member of the lipocalin protein family that has diverse function but similar structure. The functions of NGAL are not clear, but it appears to be expressed in stress conditions and in tissues undergoing involution. Varied studies have shown increased levels of plasma or urinary NGAL in diverse renal damages.

Materials & Methods: The aim of this study was the serial measurement of serum and urinary NGAL within the first week after renal transplantation in children to predict immediate and short-term graft function.

Results: A total of 27 patients were assessed. These patients were classified into those with rapid reduction in serum creatinine (more than 50% reduction in serum creatinine in the first day after transplantation) and patients with slow reduction in serum creatinine (<50% reduction in serum creatinine). We also assessed the absolute reduction in serum creatinine before and after transplantation. Serum and urinary NGAL on the first day post-transplantation were higher in recipients with slow reduction in serum creatinine (urinary NGAL at the first day: 197 ± 153 [s.e.m.] vs. 22.54 ± 8.5 [s.e.m.],p = 0.04; serum NGAL at the first day: 199 vs. 69.8, p = 0.003). For prediction of slow creatinine reduction, the cutoff point of serum NGAL at the first day after transplantation was 174 ng/mL (sensitivity:100%, specificity: 95.5%). However, we did not find association between the absolute reduction in serum creatinine before and after transplantation with the amount of serum and urinary NGAL posttransplant.

Additionally, we did not find any effect of high serum and urine NGAL concentration on the graft function at the first year posttransplant.

Conclusions: Although it is supposed that high serum and urine NGAL may predict ischemia of graft in early phases; however, it appears that this mild ischemic injury to graft without DGF or SGF cannot affect the graft function in short-term period. Further studies are needed using larger transplant recipients in pediatric age group. It is also needed to determine the effects of mild ischemic injuries on the graft function in long-term period in future studies.

Thurs-O11

To Assay The Effect of Nandrolone Decaonate on Body Weight And Anemia In Patients Receiving Hemodialysis and Peritoneal Dialysis in Dr. Sheikh Hospital.

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Introduction: Anemia and disordering in weight, malnutrition, dysfunction of hypothalamic pituitary gonads axis, delayed puberty, sex dysfunction , delayed bone age and osteoporesis are the most common problems in chronic renal Failure child. This study was done to check the effects of Nandrolone in improvement of growth body parameters and Anemia.

Materials and Methods: In this study 20 ESRD patients at the ages of 13 to 22 were selected in 3

months without any changes in diet and drugs. (Control period)
All parameters in growth body (height, weight, fat under the skin and BMI) were measured monthly. Clinical examinations of puberty sex based on stages of Tanner and sonography of ovaries in girls and testis in boys were done. Radiography of hand and wrist to control the bone age were done. Laboratories including hemoglobin, hematocrit, Fe, TIBC, triglyceride, cholesterol, PTH, Vit D3, albumin and ferritin were done. Hormonal evaluation for LH, IGF-1, and IGFBP 3and DHEAS were done in all patients. FSH and Esteradiol measurement were done only in girls and Testosterone measurement only in boys. Then patients received treatment with ND, 2 mg/ kg intra muscular injection once a week for 3 months (Max 50 mg for girls and 100 mg for boys) (Intervention period). At the End all parameters repeated and their averages compared.
Results: There were 10 boys and 10 girls and totally 20 ESRD patients. The average of weight, BMI and fat under the skin, Albumin, Hemoglobin, Ferritin, Fe, transferrin saturation, esteradiol, IGF-1,DHEAS, Vit D in patients were studied in intervention period and increased in comparison with the control period and the average of IGFBP 3 and PTH decreased ($P < 0.05$). But the average of height, arm circumference, puberty parameters, TIBC, LH, FSH and testosterone had no differences. ($P> 0.05$)
Conclusions: According to these results, ND is recommended as an effective drug in increasing the growth body parameters and improving anemia in chronic renal failure child.

Thurs -O12

A Report on CAPD Patients From Ali-Asghar Children Hospital- The Importance of An Online Simple Registry

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Introduction: Continuous ambulatory peritoneal dialysis is growing up in our country. Haemodialysis circuit suitable for very small children and automated peritoneal dialysis is still unavailable in our country. The care and management of CAP children are not uniform and very individualized. Despite the survival of patients improved, but the rate of morbidity and technical failure are still needs to be challenged.
Materials and Methods: Clinical, laboratory, and peritoneal adequacy of CAPD patients of Ali-Asghar Children hospital entered to a IPPN registry. The information updated every six months online. Data of eighteen out of 25 children on CAPD entered into registry so far.
Results: The mean age at start of PD was 5.2(4.8SD) years in our centre but in All IPPNR mean age was 8.4 years (6.1). Renal malformation is still the top of list of the underlying disease (66.7 % vs. 45%). Height SDS at PD initiation and recent follow up was (-5.36) compared to (-2.37). Nutritional supplementation is only provided to 12.5% (by PEG) and 62.5% (by Oral supplements) .Mean duration of PD was similar in two groups. Peritonitis incidence in our centre was one peritonitis episode per 2 treatment months compared to 1 episode per 27.7 months in all IPPNR. The incidence of exit site /tunnel infection was 1 episode per 5.9 treatment months but in IPPNR was one per 9.3.treatment months. The estimated GFR at PD initiation was lower in our centre compared to registry (9.1 vs. 12.3). We used higher daytime fill volume (585 vs. 448 ml/m^2/BSA), lower total night-time fill volume (6137 vs. 9407 ml/m2/BSA), and lower dialysate turnover (4796 vs. 9711 ml/m^2BSA). There was different in the PD fluid usage and the dialysis modalities. Patients with haemoglobin less than 11 g/dl were in 87.5% (vs. 52.4%). PTH K/DOQI target range was 75% below, and 25% above (Vs. 42.6% below, 14.3% within, and 43.2% above). Small molecule clearances were significantly higher than whole registry.
Conclusions: Designing an online network program accessible to all registered help to first compare to other centres and provide instant measurement of improving or deterioration of clinical and laboratory condition of the PD patients.

Thurs-O13

Virus Excretion In Acquired Immunocompromised Children: A Comparison Between Kidney Transplant Recipients And Steroid Resistant Nephrotic Syndrome

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Introduction: BK Virus (BKV) is ubiquitous in human beings. Virus reactivation may occur in immunocompromised settings. The aim of this study was to compare BKV excretion in acquired immunocompromised children (kidney transplant recipients and steroid resistant nephrotic syndrome) with normal population. .

Materials & Methods: One hundred and thirty one participants less than 20 years were recruited in the case control study from June 2009 to December 2010. The participants consisted of 40 patients with steroid resistant nephrotic syndrome (subgroup 1), 39 kidney transplant recipients (subgroup 2) and 52 normal populations as control group. The first morning urine samples were analyzed in duplicate by conventional polymerase chain reaction (PCR) method for BKV.

Results: Nine participants out of 131 had positive results for BKV. Three patients in subgroup 1 (7.5%), two patients in subgroup 2 (5.1%) and six people (11.5%) in the control group had positive PCR results for urinary BKV. No significant difference was noted among groups, p = 0.53. The mean of glomerolar filtration rates in participants with positive and negative results for BKV were 125.5 ± 30.8 ml/min/m^2 and 132.2 ± 42.5 ml/min/m^2 respectively, p = 0.8.

Conclusions: Acquired immunocompromised conditions did not increase the chance of urine BKV.

Thurs- O14

Thyroid Disorders In Children With Chronic Renal Failure

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Introduction: There are various changes in thyroid gland and its function in chronic renal failure (CRF). This study is designed to investigate the frequency of goiter and thyroid disorders in patients with CRF.

Materials & Methods: Forty (16 females and 24 males) patients with CRF and 40 (18 females and 22 males) healthy individuals as controls were included with mean age of 11.28 ± 4.87 years and age spectrum of 0.7 -18 years.

All patients received medical services from Dr. Sheikh Hospital. Examination of thyroid gland using ultrasonography and physical examination along with simultaneous measurement of blood level of thyroid hormone, TBG, were made for every individual. Results were recorded from Day 1386 to Day 1387.

Results: By physical examination the presence of goiter was found in 23% of CRF patients and 5% of controls. The presence of goiter demonstrated by ultrasonography was found in 10% of CRF patients and 0% of controls. One patient had hypothyroidism and the remainder patients and controls did not have hypo or hyperthyroidism. The patients and controls did not have nodule. The serum level of TT3, TT4, Hb, and Alb significantly were lower and TSH was significantly higher as compared to controls. Between CRF patients and controls in serum level of TBG and FT4 there were not meaningful significant difference.

Conclusions: According to thyroid disorders in CRF patients, serial physical examination, sonography and hormonal Lab. tests is recommended. Further study in this filed is recommended.

Thurs -O15

Vancomycin Induced Peritonitis – A Case Report

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Introduction: Drug induced peritonitis is a rare but important complication in CAPD. Hereby, we report on a case that developed esosinophilic

peritonitis during intraperitoneal prescription of vancomycin.

Case presentation: This is a 3- year – old girl, known case of hypoplastic-agnesis kidneys, has been on CAPD since 2010 (neonatal period). Because of Dacron sheet extrusion, she went under surgerical replacement of swan neck Tenckhoff catheter. Four days later, she returned to clinic for cloudiness of effluent and mild abdominal symptoms. The analysis of effluent showed 145 WBC/micr/L(60% PMN) that increased to 1400 WBC /micro/L(80%PMN). The dialysate culture was staphylococcus epidermidis resistant to methicillin and aminoglycoside, and sensitive to glycopeptide. Therefore, intraperitoneal vancomycin started and continued for 11 days. During therapy she had persistently allergic cough, and the 10th day pruretic papules appeared on whole body skin. The PD effluent turned turbid and the analysis revealed WBC 1700 /mico/L (71% Eosi). The PD culture was negative for any microorganism. With suspicious to vancomycin induced eosinophilic peritonitis, vancomycin discontinued. Antihistamine medication started and the PD effluent two days after stopping medication revealed 85 WBC (10% Eosi, 71% Lymph). The systemic sign improved dramatically in less than a week (table-1).

Table 1- The progress of sign, symptoms and effluent analysis

Day	0	4	8	11	18	23
sign	Cuff Extrusion		Allergic cough	Skin rash Pruritis Cough	improvement	
D_{WBC}		145	1700	650	83	22
D_{PMN}		60	3	10	2	
D_{LYM}		40	26	10	71	
D_{EOS}		-	71	80	10Neg	
Dialysate Culture		MRSE	Neg	Neg		
Exit culture		Neg				
Treatment	Cath. Replacement	Vancomycin (V)	V+ Diphen	Stop V Ketotifen	Ketotifen	

Conclusions: Drug induced peritonitis should be considered in different diagnosis of each patient on CAPD before considering administering another new antibiotic.

Thurs -O16

Treatment Adherence in Pediatric Patients

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Introduction: To discuss the issue of treatment adherence in children and their parents.
Materials & Methods: A review on relevant data bases are done and presented.
Results: Adherence refers to the extent a person's behavior corresponds with agreed recommendations from a health care provider. It emphasizes the patient's/caregiver's role as a partner in the treatment and decision-making process. Average medical adherence is around 50% in the pediatric population. Communication is a central element in adherence. Effective communication causes perception of interest, caring, warmth, responsiveness, empathy, respect, and honesty. One-on-one relationship between one doctor and one patient, working in collaboration and cooperation with the patient and the family, and familiarity of the physician and the office staff with the patient, family, and the treatment program are important elements that can enhance doctor-patient relationship which in turn ends up in higher levels of adherence. Parent's beliefs in the seriousness of their child's conditions and the severity of the complications their child suffers if they fail to adhere, can increase adherence.
There are often large discrepancies between what the health care staff feel they have told and what patients actually recall. Thus, it is necessary to carefully check the understanding of parents and children about what is expected of them. Education of parents and children is highly helpful, although they forget much of the information, soon after the session. The most retained information is the part presented during the first third of the meeting. Researches indicate that parents forget almost 50% of the information presented during a 15-minute meeting, therefore short and repeated educational sessions are the

best recommended way to overcome this problem. Audiotapes are useful in allowing parents to listen to the information repeatedly and to disseminate them to important others (e.g., extended family members, other practitioners, other parents, religious leaders, and tribal elders). Parents of chronically ill children need more and clearer information about their child's condition, the treatment plans, and opportunity for advance care planning, be shared with them as soon as it is known. In addition, they want advice about their child's behavior, child's development, genetic implications, and the long-term care plan. They need their views and concerns be factored into the care plan, being treated like partners in their child's care, affirmation of their efforts, support for child care, and support for professional services. Parents need regular meetings with the physician to discuss the "big picture"and to feel that they have a "medical home". Availability and continuity of care (e.g., telephone availability 24 hours a day and seven days a week, off-hours availability, and consistent response to questions or problems) are another elements which are important in adherence promotion. Note that in cases of no adherence, a nonthreatening and nonjudgmental manner in determining the extent of and the reasons for it, is more helpful.

Conclusions: Adherence depends on the patient's and physician's committing to the same objectives. Good doctor-patient relationship has the most dramatic effect on adherence. When parents and children are addressed in information gathering and treatment plan, they will be active participants in their care, will be more satisfied, and their adherence improves.

Thurs -O17

Measurment of Cystine in PMN Cells by Liquid Chromatography –Tandem Mass Spectrometry

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Introduction: Cystinosis is a rare autosomal recessive disorder caused by mutations in the CTNS gene, encoding the lysosomal cystine transporter cystinosin characterized by an accumulation of intra lysosomal cystine due to a defect in cystine transport across the lysosomal membrane. This disorder can be treated specifically using high doses of cysteamine. Accurate measurement of intracellular cystine content is necessary for the diagnosis and monitoring of treatment with cysteamine. Here we describe a method to measure intracellular cystine. It relies on a liquid chromatography-tandem mass spectrometry assay.

Materials & Methods: Seven to 10 ml venous blood is collected in an ACD or heparin anticoagulant, for monitoring of treatment with Cystamine. Samples should be collected within 6 hr after treatment. Polymorphoneuclears (PMNs) are isolated within 24 hr according to 2001 guidelines from the group "cystine in WBCs" and lysed in the presence of N-ethylmaleimide to avoid interference from cysteine. After deproteinization, addition of stable isotope d6- cystine and butylation, cystine is measured using an API 3000 MSMS.

Conclusions: Liquid chromatography-tandem mass spectrometry method makes it possible to measure very low concentrations of intracellular cystine in blood. This allows to quantify Cystine levels in PMNs in levels greater than 2 nmol ½ cystine/mg protein in cystinosis patients and lower than <.025nmol ½ cystine/mg protein in normal subjects. This method also allows identification of patients with cystiuria.

Thursday Poster Presentations

Thurs P1

Incidence of Meatal Stenosis Following Circumcision Done in Nappy Aged Children

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Introduction: Meatal stenosis is a recognized complication of circumcision done during nappy aged boys as a late presentation. The objective of this study was to describe the incidence of documented meatal stenosis in boys circumcised in infantile period.

Materials & Methods: Meatal stenosis was defined as change in appearance of the delicate

lips of the urinary meatus, with loss of ellipticated shape to a circular shape because of scar and visually apparent narrowing.
The study included 356 consecutive samples of boys aged 6 months to 6 years (mean 2.5 y/o), circumcised in first 2 months of life, who visited in my private office referred for reasons of urinary or non urinary complaints.
Results: The diagnosis was made in 32 of 365. Four of 32 stenotic cases were asymptomatic, common symptoms in other 28 boys were decreased urine caliber (11), crying before or during voiding (8), dysuria (5), penile deviation (3), and bloody spotting (3). None cases had urinary tract infection; urinary tract sonography was within normal limit in all cases.
Conclusions: Meatal stenosis is a complication of circumcision done during nappy aged boys as a late presentation. A careful meatal examination is indicated in any boys with history of circumcision during nappy age. We recommend to use lubricant or anti inflammatory ointments for prevention of meatal fibrosis and stenosis.

Thurs P2

Renal Glycosuria in Febrile Urinary Tract Infection

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Introduction: Renal glycosuria is excretion of glucose in urine in a normoglycemic condition although it is a benign, occasionally familial condition, but may be result from renal tubular dysfunction or immaturity of tubular function in the neonatal period.
Materials & Methods: Admission charts of 134 children aged 3month to 9 years with acute pyelonephritis. That were admitted in January 2011- March 2012 were reviewed.
Results: We found glucose (one plus) in urinalysis of 18 cases (7.1%) in first urine sample at admission time that was absent in discharged or followed up times.
Conclusions: Renal glycosuria is a benign and reversible condition that may be observed in children with acute pyelonephritis.

Thurs - P3

Neonatal Hydronephrosis Due to Congenital Megacalycosis

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A female newborn with history of prenatal hydronephrosis showed dilated calyces of right kidney without renal pelvic enlargement or hydroureter. As she was taking antibiotic prophylaxis, result of voiding cystourethrography was normal without vesicoureteral reflux. Diuretic renogram showed non obstructive pattern. Because of persistent dilated calyses in 2 year. Intravenous pyelography showed normal function of kidneys with dilated calyces in favor of megacalycosis. Congenital megacalycosis is a rare congenital renal abnormality with a benign course and good prognosis that must be differentiated from obstructive hydronephrosis.

Thurs P4

Clinical Course of Congenital Nephrotic Syndrome

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Introduction: Congenital nephrotic syndrome is a nephrotic condition presents in first 3 months of life. Histological diagnosis and management is a challenge for pediatric nephrologists. We present clinical course and pathologic findings in patients with CNS followed by pediatric nephrology center in Mashhad.
Materials & Methods: We reviewed medical charts of 19 cases with CNS that were admitted in 2000 to 2010.
Results: Our patients were aged 20 days to 2.3 years old with mean age of 5.5 months, including 11 boys and 8 girls that presented with generalized edema. All patients presented in first two months of life. Family history with involvement of sibling was present in 15 cases that their parents were relative. Kidney Biopsy

had done in 7 cases. Finish type of CNS was in 4 cases, diffuse mesangial sclerosis in 2 cases and minimal charge in one case. Low birth weight was noted in 16 cases and prematurity in 5 cases. There was no history of placental weight or labor problem. There was history of infection as sepsis or fever of unknown origin in 11 cases. The patient with minimal change histology responded to steroid with complete remission of 3 months therapy. Four cases were expired in admission time; due to infection.

Conclusions : Parents of our patients were not cooperative for kidney biopsy procedure. Finnish type is an important cause of CNS. Infection is a major complication that may terminate to death.

Thurs P5

Prognostic Value of Serum and Urine NGAL in Response to Corticosteroid in Children with Nephrotic Syndrome.

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Introduction: The aim of this study was to determine the diagnostic value of serum and urinary NGAL in response to steroid in nephrotic children.

Materials & Methods: Twenty-four children aged one to 18 years with idiopathic nephrotic syndrome enrolled in the study. Urine sample and blood were collected to measure protein and NGAL at the time of admission and before starting therapy. All children went on standard therapy of steroid with dosage of 60mg/m^2/day for six weeks and the children were followed up for response to therapy and relapse. Mean of variables were compared in two groups.

Results: Form 24 patients, 5 relapsed in three months of changing the dosage of steroid. However, there was correlation between serum creatinine and urinary NGAL level (r = 0.65, P = 0.001), this correlation was borderline between serum creatinine and serum NGAL level (r = 0.36, P = 0.088). Two cut point was identified: the first was for uNGAL that four out of five relapsed cases and three out of 19 non-relapsing children had uNGAL above 11.9 (p=0.014). The second was for blood NGAL. Three out of five relapsing children compared to 2 of non-relapse children had bNGAL above 93(p=0.042)

Conclusions: According to this study, it was shown that urine and blood NGAL levels might help to predict earlier the dependency to steroid in nephrotic children..

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Thurs P6

Beta Trace Protein As GFR Marker In Children

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Introduction: Serum creatinine is the most used endogenous marker of GFR but also has multiple limitations. Therefore, some surrogate GFR markers have been introduced for GFR estimation such as beta trace protein. The aim of our study was to estimate GFR by serum beta trace protein..

Materials & Methods: We used three available equations and compared them with DTPA GFR as gold standard and Schwartz GFR. The three beta trace protein (BTP) related GFR formulas were White formula (1): $GFR=167.8\times BTP^{-0.758}\times creatinine^{-0.204}$, Poge formula (2): $GFR=974.31\times BTP^{-0.2594}\times creatinine^{-0.647}$ and Benlamri formula (3): GFR= 10^(1.902 + (0.9515×LOG(1/BTP).

Results: Twenty seven children were included in this study. All patients had Schwartz and DTPA GFR more than 50 cc/min/1.73m^2. We showed that there was not any significant correlation between DTPA GFR and Schwartz estimated GFR (r=-0.1, Pv=0.5), There also was not any association between GFR estimated by Poge or Benllamri formulas and DTPA scan, in contrast, there was significant association between DTPA GFR and White BTP formula estimated GFR (r=0.77 r=0.00).

Conclusions: This study has shown that GFR estimated by serum beta trace protein and White formula had accuracy over Schwartz formula in children with normal or mild reduced GFR. This result needs to more studies with more cases for confirmation.

Thurs P7

Ambulatory Blood Pressure Monitoring in CAPD Patients

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Introduction: Hypertension is a risk factor for cardiovascular disease which is the main cause of morbidity and mortality in the dialysis population. Volume overload is prevalent in PD patients. The aim of this study was to assess blood pressure statues in children on CAPD.

Materials & Methods: Between 2010 and 2013, 14(9 Males, 5 females) patients on CAPD aged 2-10 years old went on Ambulatory Blood Pressure Monitoring. Mean blood pressure more than 95%, and blood pressure load more than 25% considered hypertension. In the case of normal mean blood pressure and blood pressure load more than 25% defined as pre-hypertension. If the drop of blood pressure at night was less than 10%, non-dipper statute was defined.

Results: The majority of patients received one or more antihypertensive medications. The underlying disease of patients were PCKD (n=4), nephrotic syndrome (n=3), Agenais/ hypoplasia of kidney (n=3), Atypical HUS (n=1), barter syndrome (n=2), cystinosis (n=1). From 18 ABPM measurements in 14 patients, high blood pressure was detected in 42.8% of the patients (one of nephrotic, one PCKD, one Barrter syndrome). Non-dipper status was detected in 85.7% of the patients. Three patients transplanted, two patients passed away, and eight of them continued on CAPD

Conclusions: The ABPM is a useful instrument for early detection of hemodynamic changes in patients of CAPD and their compliance with medication.

Thurs P8

Tacrolimus administration for a diabetic patient with steroid resistant nephrotic syndrome Do we have much fear?

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Case presentation

A 5-years-old female with type 1 diabetes for four months was referred to our department after complaining of an edema that was 14 days in duration. She was being treated with insulin. Physical examination revealed blood pressure of 95/65 mmHg, pulse of 80 beats/min, normal breathing, and no fever.

The patient's eye ground was examined; no abnormalities were found. Her weight at that time was 13 kg, and her baseline weight was approximately 12 kg. She had pitting edema on the legs and face. Her heart and lungs were normal, and no liver, spleen or lymph node enlargements were detected.

Urinalysis showed 6-8 white blood cells, no red blood cells, and 20-25 granular casts per high-power field.Proteinuria was 3.14 g/24 h; blood urea, 12 mg/dL; serum creatinine, 0.5 mg/dL; serum total protein, 4.8g/ dL; albumin, 1.8 g/dL; total cholesterol, 444 mg/dL; blood glucose, 487 mg/dL; hemoglobin, 10.2 g/ dL; Hemoglobin A1c (HbA1c) 6.7 and proteinuria, 4763 mg/24 hours. Serology for hepatitis B and C, HIV, vasculitis and lupus were negative. A percutaneous renal biopsy was performed. The biopsy specimen contained 11 glomeruli. Megangial hypercellularity was seen in less than 50% glomeruli. No capillary wall thickening was identified. Tubular atrophy was seen in 30% of tissue surface. Interstitial fibrosis was seen in 30% of the specimen. Interstitial inflammation was evident in 20% of tissue surface. No vasculitis was identified. Renal histopathologic findings were compatible with FSGS.

Prednisone therapy using 2 mg/kg/day was started, and the patient's insulin dosage not changed during the course of treatment. Prednisone dose discontinued after 2 months of treatment due to massive proteinuria (2223 mg/24 hr). Steroid therapy changed to cyclophosphamide 2 mg/kg/day. Treatment with cyclophosphamide discontinued after 7 weeks because of high proteinuria. Tacrolimus 0.1 mg/kg/day was administered. Six months after the treatment, her urine protein decreased from 450 to 205 mg/day.

There was no evidence of relapse over 3.5-year follow-up.