**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 \[\text{weight (kg)} / \text{height (M)}^2\]. 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, 7.2 and 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 (-2.6 SDS) and weight, 45 kg (BMI SDS, -1.0).

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 indomethacin 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 P was 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.
**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 nephronophthisis 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 reflux. 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 determineth 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-03**

**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-04**

**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 polyarticular and/or systemic JIA (SjJIA). 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, Methotrexate 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, such 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 –05**

**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-microglobulin 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 –06**

**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
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children is accounted for by Nephropathic Cystinosis, a typical presentation of failure to thrive, acidosis, renal tubular Fanconi's syndrome, muscle hypotonia, rickets, polyuria, polydipsia, 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 above 38 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 (3 case). 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- 07**

**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 with 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°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 to 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 –08**

**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.
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.

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.

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-011**

**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 osteopenosis 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 3 and 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-012**

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

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2www.pedpd.org

**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/m²/BSA), lower total night-time fill volume (6137 vs. 9407 ml/m²/BSA), and lower dialysate turnover (4796 vs. 9711 ml/m²BSA). 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-013**

**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/m² and 132.2 ± 42.5 ml/min/m² respectively, p = 0.8.

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

**Thurs-014**

**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 -015

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 eosinophilic peritonitis during intraperitoneal prescription of vancomycin.

Case presentation: This is a 3- year – old girl, known case of hypoplastic-agenesis 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 pruritic papules appeared on whole body skin. The PD effluent turned turbid and the analysis revealed WBC 1700 /mico/L (71% Eosi). The systemic sign improved dramatically in less than a week (table-1).

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

<table>
<thead>
<tr>
<th>Day</th>
<th>0</th>
<th>4</th>
<th>8</th>
<th>11</th>
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<td>Allergic cough</td>
<td>Cough</td>
<td>Pruritus</td>
<td>Skin rash</td>
<td>Improvement</td>
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Conclusions: Drug induced peritonitis should be considered in different diagnosis of each patient on CAPD before considering administering another new antibiotic.

Thurs -016

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. Audio tapes 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 nontreating 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 -017**

**Measurement 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. Polymorphonuclears (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.

Third day Poster Presentations

Thurs P1

**Incidence of Meatal Stenosis Following Circumcision Done in Nappy Aged Children**

Esmaeeli M

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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**

Esmaeeli M, Azarfar A

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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**

Esmaeeli M

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A female newborn with history of prenatal hydronephrosis showed dilated calyces of right kidney without renal pelvic enlargement or hydrourer. 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 calyces 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 hydrenephrosis.
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 change 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.

Eshagh-Hoseini SA, Otukesh H, Hooman N.

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/m²/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.

Thurs P6

Beta Trace Protein As GFR Marker In Children

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Pediatric transplantation and dialysis research center, Iran University of Medical Science, Tehran, Iran

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): \( \text{GFR}=167.8\times\text{BTP}^{-0.758}\times\text{creatinine}^{-0.204} \), Poge formula (2): \( \text{GFR}=974.31\times\text{BTP}^{-0.2594}\times\text{creatinine}^{-0.647} \) and Benlamri formula (3): \( \text{GFR}=10^{(1.902+(0.9515\times\text{LOG(1/BTP))})} \).

**Results:** Twenty seven children were included in this study. All patients had Schwartz and DTPA GFR more than 50 cc/min/1.73m². We showed that there was not any significant correlation between DTPA GFR and Schwartz estimated GFR \((r=-0.1, P_v=0.5)\). There also was not any association between GFR estimated by Poge or Benlamri 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**

Moghiseh L, Saffar Z, Ghafari L, Hooman N

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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 Barter 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.