

Oral / Poster Presentation

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Second day Oral Presentations

Wed- 01

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- 02

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), nephronophthisis (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, pseudogypoadosteronism 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					Total
	north	south	west	east	center	
Nephronophthisis	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
Bartter	7	34	19	27	44	131
Gitelman	2	7	3	1	59	72
hypocacemic hypercalciuria familial			20	7	121	148
hypocalcemia AR				3	123	126
Hypomagnesemia					7	7
hypomagnesemic hypercalciuria			1	1		2
hypomagnesemic+hypocalcemia		10	1			11
hypomagnesemia mitochondrial		1				1
Liddle syndrome		6	1	1	1	9
GRA				3		3
Gordon II		3		1	14	18
pseudohypoadosteronism 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
fanconi bickel			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-03

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-04

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-05

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 m² 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 $\pm$ 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 1 α -caldiol 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	1709(1180)	6580	200
Phosphatas			
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-06

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 m², 78.42 ± 21.15 ml/min/1.73 m², and 80.5 ± 22.43 ml/min/1.73 m², respectively. GFR based on Cystatin C on these days were 116.23 ± 58.9 ml/min/1.73 m², 116.49 ± 53.31 ml/min/1.73 m² and 108.37 ± 51.02 ml/min/1.73 m².

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-07

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 microglobulin, 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- 09

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- 010

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- 011

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.

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- 013

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

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Wed-012

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.

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 nephropathic 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 nephropathic cystinosis at least in Iranian patients.

Wed- 014

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- 015

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 2005 were 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 dysfunction in these children, encourage prompt gastrointestinal evaluation, and encourage treatment of more severely affected patients.

Wed- 016

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 hypothyroidism. 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 ($\pm$ 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 ($\pm$ 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- 017

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 $\geq$ 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- 018

Endocrine Disorders in Childhood Cystinosis

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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- 019

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 oligohydramniotic in fetal period; IUGR with birth weight of 2.3 kg. He was admitted to our hospital with the history of polyuria and polydipsia 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 11 β 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). Laboratory 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 HSD11 β 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- 020

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/H² . 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- 021

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- 022

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- 023

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 creatinine 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- 024

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- 025

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, Hco₃ and 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 respectively. 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- 026

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- 027

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- 028

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- 029

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 .

Second day 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), fairly good (n=32), fairly 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 Bartter 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 hydrochlorothiazide 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 $<3\mu\text{g/ml}$ in patients ≤ 10 years and levels $<6\mu\text{g/ml}$ in teenagers (>10 years).

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) $\mu\text{g/ml}$. Six CAPD (2/3) and 11 hemodialysis (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 $\pm$ 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 $\pm$ 32.68). Serum vitamin C levels were measured and values 0.6-2mg/dl in patients ≤ 20 years and 5-18mg/dl in patients >20 years defined as normal value respectively. Serum levels <0.6 and $<5\text{mg/dl}$ in age groups <20 and ≥ 20 years 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 $\pm$ 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 hemodialysis patients were 0.96 $\pm$ 0.24 and 0.87 $\pm$ 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 polycitra 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.